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Peter C. Laussen *Editors*

Pediatric and Congenital Cardiac Care

Volume 1: Outcomes Analysis

Springer

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We would like to dedicate this book to all patients receiving pediatric and congenital cardiac care and their families.
Each of the Editors would like to make the following additional dedications:

To Elijah, to Tore, to Harrison, and to the love of my life, Julie, who makes it all possible.

Paul R. Barach

To my parents David and Marilyn Jacobs for giving me the opportunity, to my wife Stacy for supporting and loving me, to my children Jessica and Joshua for making me proud and motivated, and to my patients, who represent the rationale for this initiative.

Jeffery P. Jacobs

To my wife Tracie and our children Hannah, Zach, Emma, and Sarah, who have always inspired me to do everything possible to improve the health of children. Their motivation has guided me, in the words of others, to "treat every child and family I have the privilege to care for as if they were my first and every day that I am entrusted with their care as if it was my last." These volumes are part of the covenant I have made with many patients and their families to do our very best for them and those who follow. This has been made possible by the wisdom and collaboration of my cherished colleagues and mentors.

Steven E. Lipshultz

To Julia and Joan for their wisdom, guidance, and love.
To my children and precious Emery, an inspiration for the future. To the patients I care for and who teach me every day.

Peter C. Laussen

Foreword

A parent taking a child into hospital for diagnosis and treatment hopes for a cure and the restoration of a normal quality of life in the future. They fear many things, ranging from the worst—the death of the child—through their pain and suffering to uncertainty about how to manage the complexity of their own lives, which have so suddenly been disturbed. What they have a right to expect is that the people treating their child know what they are doing, are well trained, and particularly will put the needs of the their child at the center of their decision making.

They are handing over their precious bundle of joy to strangers to care for, aware that that very care might actually threaten the life of that child as well as offer treatment. They are *loaning* their child to these professionals. It demands an enormous amount of trust to do that. As professionals, we need to be able to recognise that level of trust and repay it. Trust is a two-way thing.

The remarkable fall in the mortality for repair of congenital heart defects over the last 60 years could lead to complacency. But we must not forget that mortality is only one outcome measure and cannot reflect all the issues which concern parents. Medicine is dangerous. Many readers will have seen the famous diagram which charts the relative risk of accidents affecting users of organisations, which shows that there are high-reliability organisations like European railroads, western airlines and the nuclear industry but that medicine is about as safe as bungee jumping. This is due to errors that we make, problems we fail to address, complications we fail to tackle. There is no room for complacency if we want to deserve the trust of the parents who have loaned us their child.

If it were my child being treated, this is what I expect:

- I *expect* that my child will be cared for safely in a modern hospital.
- I *expect* my child to be looked after by a well-functioning multi-disciplinary team.
- I *expect* the staff will know the results of the treatment they propose not just in the literature but in their own hands.
- I *expect* the staff to know the complication rates in their hospital and put in place ways to reduce them.
- I *expect* that they will be collecting complete and validated data on all they do and that they will share those data openly with other professionals and the public.

- I *expect* the staff will do all they can to mitigate the certain human error that will occur, by putting in place systems which limit both risk and harm.
- I *expect* that the staff will be honest, open and transparent in all their dealings with me and that if they don't know something, they will say so and let me get a second opinion.
- I *expect* to be involved in decisions about the care of my child and to have my views respected.
- I *expect* that any harmful incident will be fully, openly and honestly investigated as quickly as possible and that learning from the incident will spread widely so that no one else can suffer.
- I *expect* that the team will be interested in the long-term outcome of treatment, not just in hospital, and that they will have mechanisms in place to gather the relevant information.
- I *expect* the truth and to be treated as if I were a friend, with warmth and empathy.

The Editors of this timely book have gathered an array of experts to give guidance as to how these expectations should be met. They give valuable insight into methods and use their own experience to highlight what we can do to be better. Being better, continuous improvement is what it is all about. Our speciality has done well with a relentless pursuit of excellence and is further advanced than many in being open about its results. Yet, it has much to learn from other disciplines, particularly oncology, about the benefits of collaboration over competition. Our discipline was built on the drive and energy of highly competitive alpha males and the disruptive technology of cardiopulmonary bypass. A second wave of disruption has followed the introduction of trans-catheter interventions. But this too has resulted in the same kind of rush to glory that we saw in the 1970s with surgical heart valve implementation and design. We need good studies, strong data and multi-center collaboration if we want to give the best care as quickly as possible.

This book exemplifies the move to collaboration and the drive towards openness and transparency. All our patients and their families are now 'digital natives'. They access the collective memory of Google just as we do. They expect to see our results and can quickly find their way around PubMed. We have a duty to give them insight into the facts they can read. The information provided in this text will help units realise both the importance of good data but also the methods by which it can be used, evaluated, interpreted and reported.

Don't forget, your duty is to keep the child safe and make it as well as you can. This book will help.

London, UK Martin Elliott, MD, FRCS

Preface

The idea that clinical data could be analyzed by multiple congenital heart centers was shared by many enlightened individuals who foresaw the utility of such an organizational structure in the early 1980s. Discussions led to ideas that resulted in primitive data collection systems that catalogued diagnoses, procedures, complications, and survival statistics. The difficulty with these systems was that the nomenclature was not uniform and the challenge of comparing diagnoses and procedures prevented accurate analysis. In short, nomenclature categories were diverse owing to substantial and justified differences of opinion by many leading anatomists. Parallel publications by surgeons and cardiologists resulted in more uniform parochial nomenclature systems, but still there were significant differences between the two that challenged future collaborative efforts. The call to arms was answered by concerned clinicians and anatomists and resulted in a computer mapping strategy that was successful in categorizing diagnoses and procedures by what is actually described and performed and not by what it is called. As a result, the types of ventricular septal defects, atrial septal defects, truncus arteriosus, and the like now had a computer number and not a name. It was revolutionary in concept and comprehensive in scope. It was as if the world had one language even if the cultures varied. Before long, North and South America, Europe, Asia, and Africa were using the standard nomenclature.

This was just the beginning. Data were collected, analyzed, and interpreted to reveal or contradict theretofore clinical assumptions, biases, and largely undocumented hearsay conclusions. Data verification strategies by professional volunteers were planned, and audit visits were instituted. Concurrently, participating center data were to be assessed and compared with the combined experience of the participating centers. This allowed the possibility of program assessment and quality improvement. Complexity scores were developed based on Delphian principles until the time that enough data were collected to allow data-driven risk stratification.

The subsequent analysis of the databases and the developed nomenclature became exponential. Government agencies accepted the documents and instituted registries based on the developed principles. Long-term outcome analyses became a reality with database linking to both the Department of Health and Human Services Centers for Medicare and Medicaid Services Database and the Social Security Death Master File. Ethical issues were being discussed and used to clarify rules and regulations. In addition to these innovations, database documentation of complications has been used to guide

the clinician to perform more extensive data-driven informed consent. In an interesting twist of phrases, the database was used to inform the informed consent process.

The benefits of the database systems and the supporting nomenclature were simply too much to document in an expanded treatise. It could only have been accomplished by a book, the like of which is offered in this informative and excellent text. The reader will enjoy this book not only for the rich references that accompany the prose but also for the enjoyable historical account of what some people refer to as simply unbelievable.

Orlando, FL, USA Constantine Mavroudis, MD

Acknowledgments

The Editors of *Pediatric and Congenital Cardiac Care: Volume 1 – Outcomes Analysis and Volume 2 – Quality Improvement and Patient Safety,* Paul R. Barach, Jeffrey P. Jacobs, Peter C. Laussen, and Steven E. Lipshultz, would like to thank all the authors of chapters in this two-volume set of textbooks, the families of these authors, our administrative staff, and our Editorial and Publishing team.

- Our authors represent an international community of scholarship, with chapters written by luminaries and cutting-edge thinkers.
- All the family members of these authors are indeed owed a debt of gratitude because writing chapters markedly decreases the time available with them.
- Finally, this set of textbooks is possible only because of the tremendous efforts of our administrative staff and the Editorial and Publishing team, and we especially acknowledge the coordination throughout this project by Mitzi Wilkinson and the hundreds of hours devoted to this project by Flora Kim and Grant Weston.

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Part I

Introduction

Introduction

1

Paul R. Barach, Jeffrey P. Jacobs, Peter C. Laussen, and Steven E. Lipshultz

Keywords
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This book, entitled "***Pediatric and Congenital Cardiac Care***: ***Outcomes Analysis***, ***Quality Improvement***, ***and Patient Safety***," is Volume 1 of one of a two volume textbook. The focus of Volume 1 is outcomes analysis. The focus of Volume 2 is quality improvement and patient safety. The first volume of this textbook concentrates on measurement and analysis of health outcomes. Leading work has been undertaken in pediatric cardiac care to understand and measure improved patient outcomes and how to establish collaborative definitions and tools of measurement. The book highlights best practices for measuring outcomes of pediatric cardiac care. Meaningful analyses of outcomes requires a database that can incorporates the following seven essential elements: (1) Use of a common language and nomenclature; (2) Use of a database with an established uniform core dataset for collection of information; (3) Developing a mechanism for evaluating case complexity; (4) Using a mechanism to assure and verify the completeness and accuracy of the data collected; (5) Collaboration between medical and surgical subspecialties with assistance by health service researchers; (6) Standardization of data collection protocols; and (7) Incorporation of strategies

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for quality assessment and quality improvement. Volume 1 of this textbook will focus on these seven essential areas while, volume 2 will cover both implementation science for continuous quality improvement, safety science and systems improvement.

The fields of pediatric cardiology and cardiac surgery have grown and developed faster than most other fields in medicine. The fundamental biological embryological causes contributing to congenital heart disease are far from understood. There are great variations in the complexity of congenital cardiac defects, but nevertheless there are well established treatment options for correction and palliation of most defects. It seems, however, that despite unprecedented levels of spending on pediatric cardiac care, preventable medical errors have not been reduced, uncoordinated care continues to frustrate patients, parents and providers, and healthcare costs continue to rise [1]. The US Institute of Medicine estimates that 100 patients die each day in the United States from iatrogenic causes. There are many possible factors related to this unexpected circumstance, including the introduction of new technology that alters rather than improves systems for care, the lack of engagement of front line staff in decision making the complexity of patient disease and the increasing toxicity of medical treatments.

Delivering safe pediatric cardiac care is complex and complicated. The way, we organize as teams, the systems of care we develop, and the means by which we collaborate and share information are crucial for delivering safe and cost effective care [2]. Indeed, the delivery of safe and reliable patient care is an international health system priority. In the early days of pediatric cardiac surgery, mortality rates were very high. During the past three decades, survival among children born with even the most complex cardiac defects has increased substantially so that from 2005 to 2009, the discharge mortality of index cardiac operations was 4.0 % (3,418/86,297) in the Congenital Heart Surgery Database of the Society of Thoracic Surgeons (85 centers from the United States and Canada) [3, 4]. Across the world, mortality figures have declined, suggesting that perhaps this outcome variable is perhaps no longer the best metric by which cardiac surgery programs can be evaluated. However, the mortality rates between institutions continues to vary up to sixfold, suggesting there is still many modifiable factors related to case volume, experience, and practice variability [5]. Morbidity and preventable adverse events are better metrics for the evaluation of performance and competence, but are difficult to measure, vary between and by systems of care, and are dependent on the socio-technical interactions of the care we provide and decisions we make [6]. Complications and adverse events result in higher morbidity, and the potential for longer-term disability and decreased quality of life. The quality of life achieved by our patients following the care we deliver is arguably the most important outcome metric for children with heart defects.

Rapid advancements that followed from improved diagnostic modalities (i.e., 2D echocardiography among others), improved technology in cardiopulmonary bypass, and new management paradigms and prostaglandin E1 infusions to maintain patency of the arterial duct, have all contributed to the remarkable successes in treating these children. Despite remarkable advances, there still remains a relatively high rate of early and late adverse events (mortality and morbidity), particularly in newborns and infants. The frequency of events and the focused patient population means that providers caring for children with congenital and pediatric cardiac disease are compelling model for investigating resilient systems, human errors, and their impact on patient safety [2].

This first of a kind cross-disciplinary collaboration by four lead clinician editors from disparate medical disciplines (i.e., cardiac surgery, cardiology, anesthesia, and critical care), has pulled together an international community of scholarship with articles by luminaries and cutting edge thinkers on the current and future status of pediatric and congenital cardiac care.

Intense scrutiny and measurement of clinical outcomes is increasing at a rapid rate, beyond institutions, regions, and borders. Simultaneously, the requirement and demand for more transpar-

ency and more public reporting, new regulations, and penalties when reported outcomes do not meet expectations is increasing. We believe the current multi-disciplinary approaches in pediatric cardiac care can provide a collaborative road map for other disciplines and fields in healthcare such as medicine, surgery and general practice. Proscriptive rules, guidelines, and checklists are helping to raise awareness and prevent harm. However, to provide an ultra-safe system for patients and their families, we need to engage users in more creative ways that rely on systems thinking, involved redesign of work practices [2].

Although the field of pediatric and congenital cardiac care has received worldwide recognition as a leader in outcomes analysis, quality improvement, and patient safety and has advocated for system-wide changes in organizational culture, opportunities remain to lower costs, reduce risks, and improve performance. The field has many complex procedures that depend on a sophisticated organizational structure, the coordinated efforts of a team of individuals, and high levels of cognitive and technical performance. In this regard, the field shares many properties with high-technology systems such as aviation and chemical manufacturing in which performance and outcomes depend on complex individual, technical, and organizational factors and the interactions among them [6].

Several factors have been linked to poor outcomes in pediatric cardiac care, including institutional and surgeon- or operator-specific volumes, case complexity, team coordination and collaboration, and systems failures [7]. Safety and resilience in these organizations are ultimately understood as a characteristic of the system—the sum of all its parts plus their interactions. Further, many regulatory and government agencies are examining more closely the utility, management of risk, relationships of programmatic volume, and outcomes in the field.

Interventions to improve quality and strategies to implement change should be directed to improve and reduce variations in outcomes. It is imperative that there be an appreciation of the impact of human factors in the field, including an understanding of the complexity of interactions between:

- The technical task,
- The stresses of the treatment settings,
- The consequences of rigid hierarchies within the staff,
- The equipment and physical architecture,
- The lack of time to brief and debrief, and
- Cultural norms that resist change.

Technical skills are fundamental to good outcomes, but non-technical skills—coordination, followership, cooperation, listening, negotiating, and so on—also can markedly influence the performance of individuals and teams and the outcomes of treatment [8].

Pediatric cardiac surgical care has been the subject of well publicized inquiries. A consistent theme from these inquiries is that many staff, patients, and managers had raised concerns about the standard of care provided to patients before the sentinel event. The events surrounding the Bristol Royal Infirmary [9], the Manitoba Healthcare [10], and the Mid Staffordshire [11] inquiries highlight the importance of engaged leaders and clinicians who appreciate the impact of human factors and systems improvement in improving outcomes in pediatric cardiac surgery.

The accidents and adverse events that still occur within systems that possess a wide variety of technical and procedural safeguards (such as operating rooms and intensive care units) have been termed organizational accidents [11, 12]. These are mishaps that arise not from single errors or isolated component breakdowns, but from the accumulation of delayed action failures lying mainly within system flaws that set up good people to fail [13]. People often find ways of getting around processes which seem to be unnecessary or which impede the workflow. This concept is known as normalization of deviance [14]. This accumulated and excepted acceptance of cutting corners or making work-arounds over time poses a great danger to healthcare systems. Similar findings have been described in other investigations into major episodes of clinical failure, and healthcare systems need to heed similar lessons from other industries [15, 16]. This concept is shown schematically in Fig. 1.1.

Fig. 1.1 High reliability organizations and their organizational culture (Reprinted from Berg et al. [30])

The study of human factors is fundamentally about appreciating the nature of socio-technical systems and optimizing the relationship between people, tasks, and dynamic environments [17]. Although a particular human action or omission may be the immediate or suspected cause of an incident, closer analysis in pediatric care usually reveals a preceding series of events and departures from safe practice, potentially influenced by the working environment and the wider organizational context [18]. An organizational accident model proposes that adverse incidents be examined both [19]:

- From an organizational perspective that incorporates the concept of active and latent conditions, and
- From an individual perspective that considers the cascading nature of human error.

To improve outcomes of children with heart defects, we need to create and support an organizational conditions, resources, and culture in which clinicians can produce safe outcomes. Leaders in our field must create the climate that allows people to acknowledge mistakes and encourages clinicians to innovate. There is tight coupling and complexity across pediatric cardiac care, and the ability of the team to recognize and respond quickly and appropriately to errors and threats is essential to minimize the consequences and ensure recovery [20, 21].

High reliability—or consistent performance at high levels of safety over prolonged periods—is a hallmark for non-health-related, high-risk industries, such as aviation and nuclear power generation [22]. High reliability is centered on supporting and building a culture of trust, transparency, and psychological safety [23]. In the face of health reform and increased competition in the market, moving to high reliability requires adopting and supporting a culture that appreciates the relationships among a variety of organizational risk factors and their effect on patient harm and procedural inefficiency. Improving safety and quality, and providing true value in pediatric cardiac care, will require clinicians to acknowledge their primary responsibility to the care of their patients and their families, as well as managing processes for optimization, standardization, and continuous measuring and monitoring of outcomes [24].

Finally, trust and collaboration within teams, between institutions, and across institutional and jurisdictional borders are essential elements in pediatric cardiac care to ensure clinicians feel safe and empowered to speak up and talk about processes and outcomes that could be improved [25–27].

This book came about from a long standing friendship and camaraderie of the editors who collectively believe that we should continuously strive to do much better for our patients, and their families, in delivering safer, higher value, and patient centered pediatric cardiac care. The book evolved from two successful special issues of Pediatric Cardiology [28, 29]. The editor's feel strongly that no one repository exists for the three inter-related domains of outcomes analysis, quality improvement, and patient safety.

We believe that innovation in patient care is best designed in concert with those on the front lines of healthcare delivery—patients and clinicians—and incorporating relevant knowledge from other scientific disciplines such as operations research, organizational behavior, industrial engineering, and human factors psychology. In order to best engage with medical staff, the focus of improvement efforts should be on bringing even more scientific discipline and measurement to the design of healthcare delivery. The need exists to develop innovative models of care that lower the complexity and cost of delivering health care, while simultaneously

improving clinical outcomes and the patient experience.

The editors are indebted to the wonderful contributions from leaders across the world from a wealth of disciplines with expertise in pediatric cardiac care. The authors are all "thought leaders," have lead important change, and are visionaries. We hope this book provides readers with a roadmap and a common reference source of current initiatives in outcomes analysis, quality improvement, and patient safety in our field of pediatric and congenital cardiac care. Moreover, we hope the content and the authors of this text will inspire readers, foster engagement, and change, and that through collaboration and sharing, pediatric cardiac care with be enriched and improved.

References

1. Committee on Quality of Health Care in America, Institute of Medicine. To err is human: building a safer health system. Washington, DC: National Academy Press; 1999.
2. Amalberti R, Auroy Y, Berwick DM, Barach P. Five system barriers to achieving ultra-safe health care. Ann Intern Med. 2005;142(9):756–64.
3. Jacobs JP, O'Brien SM, Pasquali SK, Jacobs ML, Lacour-Gayet FG, Tchervenkov CI, Austin 3rd EH, Pizarro C, Pourmoghadam KK, Scholl FG, Welke KF, Mavroudis C, Richard E. Clark paper: variation in outcomes for benchmark operations: an analysis of the Society of Thoracic Surgeons Congenital Heart Surgery Database. Ann Thorac Surg. 2011;92(6):2184–92.
4. Jacobs JP, O'Brien SM, Pasquali SK, Jacobs ML, Lacour-Gayet FG, Tchervenkov CI, Austin 3rd EH, Pizarro C, Pourmoghadam KK, Scholl FG, Welke KF, Gaynor JW, Clarke DR, Mayer Jr JE, Mavroudis C. Variation in outcomes for risk-stratified pediatric cardiac surgical operations: an analysis of the STS Congenital Heart Surgery Database. Ann Thorac Surg. 2012;94(2):564–72.
5. Jacobs JP, Jacobs ML, Austin EH, Mavroudis M, Pasquali SK, Lacour–Gayet FG, Tchervenkov CI, Walters III HW, Bacha EA, del Nido PJ, Fraser CD, Gaynor JW, Hirsch JC, Morales DLS, Pourmoghadam KK, Tweddell JT, Prager RL, Mayer JE. Quality measures for congenital and pediatric cardiac surgery. World J Pediatr Congenit Heart Surg. 2012;3(1):32–47.
6. deLeval MR, Carthey J, Wright DJ, Farewell VT, Reason JT. Human factors and cardiac surgery: a multi-center study. J Thorac Cardiovasc Surg. 2000;119:551–672.
7. Schraagen JM, Schouten T, Smit M, et al. A prospective study of paediatric cardiac surgical microsystems: assessing the relationships between non-routine events, teamwork and patient outcomes. BMJ Qual Saf. 2011;20:599–603. doi:10.1136/bmjqs.2010.048983.
8. Catchpole KR, Mishra A, Handa A, et al. Teamwork and error in the operating room: analysis of skills and roles. Ann Surg. 2008;247:699–706.
9. Kennedy I. Learning from Bristol: the report of the public inquiry into children's heart surgery at the Bristol Royal Infirmary 1984–1995. London: Crown Copyright; 2001. Department of Health.
10. Manitoba pediatric cardiac surgery inquest report. http://www.pediatriccardiacinquest.mb.ca/pdf/pcir_intro.pdf. Accessed 10 Aug 2011.
11. Francis R, QC (6 February 2013). Report of the Mid Staffordshire NHS Foundation Trust Public Inquiry. (Report). House of Commons. ISBN 9780102981476. Retrieved 17 Mar 2014.
12. Cover-up over hospital scandal. Daily Telegraph. 20 June 2013.
13. Rasmussen J. The role of error in organizing behavior. Ergonomics. 1990;33:1185–99.
14. Vaughan D. The dark side of organizations: mistake, misconduct and disaster. Annu Rev Sociol. 1999;25:271–305.
15. Norman D. The psychology of everyday things. New York: Basic Books; 1988.
16. Reason J. Managing the risks of organizational accidents. Aldershot: Ashgate; 1997.
17. Sagan SD. The limits of safety: organizations, accidents, and nuclear weapons. Princeton: Princeton University Press; 1994.
18. Catchpole KR, Giddings AE, de Leval MR, Peek GJ, Godden PJ, Utley M, Gallivan S, Hirst G, Dale T. Identification of systems failures in successful paediatric cardiac surgery. Ergonomics. 2006;49:567–88.
19. Cassin B, Barach P. Making sense of root cause analysis investigations of surgery-related adverse events. Surg Clin NA. 2012;92:1–15. doi:10.1016/j.suc.2011.12.008.
20. Westrum R. Organizational and inter-organizational thought: World Bank Workshop on Safety Control and Risk Management. Washington, DC; 1988.
21. Perrow C. Normal accidents: living with high-risk technologies. New York: Basic Books; 1984.
22. Weick K, Sutcliffe K, Obstfeld D. In: Boin A, editor. Organizing for high reliability: processes of collective mindfulness in crisis management. Thousand Oaks: Sage Press; 2008. p. 31–67.
23. Edmondson A. Psychological safety and learning behaviours in work teams. Adm Sci Q. 1999;44(2):350–83.
24. The more I know, the less I sleep. Global perspectives on clinical governance. KPMG Global Health Practice. US. Dec 2013.
25. Langer EG. Mindfulness. Boston: Da Capo Press; 1990. ISBN 9780201523416.

26. Bognar A, Barach P, Johnson J, Duncan R, Woods D, Holl J, Birnbach D, Bacha E. Errors and the burden of errors: attitudes, perceptions and the culture of safety in pediatric cardiac surgical teams. Ann Thorac Surg. 2008;85(4):1374–81.
27. Barach P, Small DS. Reporting and preventing medical mishaps: lessons from non-medical near miss reporting systems. Br Med J. 2000;320: 753–63.
28. Lipshultz S, Barach P, Jacobs J, Laussen P, editors. Quality and safety in pediatric cardiovascular care. Prog Pediatr Cardiol. 2011;1.
29. Lipshultz S, Barach P, Jacobs J, Laussen P, editors. Quality and safety in pediatric cardiovascular care. Prog Pediatr Cardiol. 2011;2.
30. Berg M, et al. The more I know, the less I sleep: global perspectives on clinical governance. Switzerland: KPMG International Cooperative; 2013.

Introduction: The History of Statistics in Medicine and Surgery

2

Eugene H. Blackstone

Abstract

Cardiac surgery has been quantitative from its onset. As the field progressed, surgeons encountered questions that required going beyond existing and traditional methods, fostering both adoption of analytic methods from non-medical fields (communication, industrial sciences, and physics, for example) and development of new ones. These were underpinned by specific philosophies of science about uncertainty, causes of surgical failure as a result of human error on the one hand and lack of scientific progress on the other, and how to express effectiveness and appropriateness to inform the timing of surgery and its indications. Included were traditional methods such as confidence limits and P-values, but also appreciation of why human error takes limited forms, as studied by human factors and cognitive researchers. The "incremental risk factor concept" reinterpreted variables associated with outcomes, initially in the context of congenital heart disease. New methods were either developed within the discipline or introduced, including those for survival analysis and competing risks that accounted for non-proportional hazards by temporal decomposition and separate risk factors for different time frames of follow-up. More recently, longitudinal methods to examine binary, ordinal, and continuous outcomes were developed. Propensity-score–based methods for comparative effectiveness studies, particularly in light of the limited ability to randomize treatments, enabled identifying complementary rather than competing techniques. However, just as the evolution of surgery has not stopped, neither has the quest for better methods to answer surgeons' questions. Increasingly, these require advanced algorithmic data analytic methods, such as those developing in the field of genomic informatics.

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Cardiac surgery—and particularly surgery for congenital heart disease—was quantitative from the outset [1], more so than most other medical specialties. This was largely stimulated by John Kirklin, who said "our true progress must continue to be based on research done painstakingly and accurately, and on experience honestly and wisely interpreted." As time went on, he, his colleagues, and others in the field embraced and advocated for statistical methods that answered increasingly important and complex questions. They fostered development of new methods "born of necessity" when they encountered questions existing methods could not answer [2].

With time, however, the underlying philosophical underpinnings and assumptions, limitations, and rationale for use and development of these methods can be forgotten, leading to less understanding and even misunderstanding. Readily available do-it-yourself statistical packages consisting of a limited repertoire of "standard" procedures encourage use of methods, applied with little understanding, that may be inappropriate. Economics may also drive a wedge between collaborating statisticians and clinicians, as meanwhile there is explosive development of statistical techniques, some of which may be perfectly suited to answering the question clinicians are asking.

Therefore, this introductory chapter traces historical roots of the most common qualitative and quantitative statistical techniques that play an important role in assessing early and late results of therapy for pediatric and congenital heart disease. I will introduce them in roughly the order they came into use in this field, which roughly recapitulates the simple to the more complex.

Uncertainty

Confidence Limits

In 1968 at the University of Alabama Hospital, outcomes of portacaval shunting for esophageal varices were presented at Saturday morning Surgical Grand Rounds: 10 hospital deaths among 30 patients. An outside surgeon receiving the live televised feed called in and began, "My experience has been exactly the same.…" Dr. Kirklin asked the caller how many portacaval shunts he had performed. "Three, with one death, the same mortality as you have experienced."

Dr. Kirklin had no doubt that the caller was being truthful, but intuitively believed that one must know more from an experience on 30 than 3. I indicated that there was a formal way to quantify his intuitive belief: confidence limits. Confidence limits are expressions of uncertainty. It is not that the data are uncertain—indeed, if one just wants to report facts and draw no inferences from them, such as risk for future patients, expressions of uncertainty are not needed. Confidence limits transform historic records of achievement into honest assessments of therapy for future patients, accounting for limited experience.

Underlying the concept of uncertainty, and confidence limits as their reflection, are at least two essential philosophical premises. First, unlike the nihilist, we embrace the philosophy that the future can be predicted, at least to some extent. Second, when we say we are predicting, we are referring to a prediction concerning the probability of an event for a future patient; we generally cannot predict exactly who will experience an event or when that event will occur.

Historically, the roots of confidence limits can be traced to Galileo, seventeenth century gamblers, and alchemists [3, 4]. If three dice are thrown at the same time, the gamblers wanted to know, what is the total score that will occur most frequently, 10 or 11? From 1613 to 1623, increasingly meticulous experiments were done to guarantee fair throws. To everyone's astonishment, these yielded equal occurrences of every possibility. From these 10 years of experimentation, Galileo developed what became known as the Laws of Chance, now known as the *theory of*

probability [5]. The laws were derived from the ordering logic of combination and permutations that had been developed by the alchemists.

We postulated that events and phenomena of cardiac surgery can also be considered to behave in accordance with this theory. These laws indicate that the larger the sample, the narrower the confidence limits around the probabilities estimated for the next patient. For treatment of patients with congenital heart disease, *n*—the number of patients—tends to be small. Confidence limits around point estimates of adverse events, therefore, are essential for interpreting the results and drawing inferences about risk for the next patient.

But what confidence limits should we use? We cannot use 100 % confidence limits because they always extend from 0 to 100 %. In the late 1960s, we decided on 70 % confidence limits. This was not an arbitrary decision, but was carefully considered. Seventy percent confidence limits (actually 68.3 %) are equivalent to ±1 standard error. This is consistent with summarizing the distribution of continuous variables with mean and standard deviation, and of model parameter estimates as point estimates and 1 standard error. Further, overlapping vs. nonoverlapping of confidence limits around two point estimates can be used as a simple and intuitive scanning method for determining whether the difference in point estimates is likely or unlikely to be due to chance alone [6]. When 70 % confidence limits just touch, the *P* value for the difference is likely between 0.05 and 0.1. When confidence limits overlap, the difference in point estimates is likely due to chance; when they are not overlapping, the difference is unlikely to be due to chance alone.

P Values

In the context of hypothesis (or significance) testing, the *P* value is the probability of observing the data we have, or something even more extreme, if a so-called *null hypothesis* is true [7]. The phrase "statistically significant," generally referring to *P* values that are small, such as less than 0.05, has done disservice to the understanding of truth, proof, and uncertainty. This is in part because of fundamental misunderstandings, in part because of failure to appreciate that all test statistics are specific in their use, in part because *P* values are frequently used for their effect on the reader rather than as one of many tools useful for promoting understanding and framing inferences from data [8–10], and in part because they are exquisitely dependent on *n*.

Historically, hypothesis testing is a formal expression of English common law. The null hypothesis represents "innocent until proven guilty beyond a reasonable doubt." Clearly, two injustices can occur: a guilty person can go free or an innocent person can be convicted. These possibilities are termed type I error and type II error, respectively. Evidence marshalled against the null hypothesis is called a *test statistic,* which is based on the data themselves (the exhibits) and *n.* The probability of guilt (reasonable doubt) is quantified by the *P* value or its inverse, the odds $[(1/P) - 1]$.

Some statisticians believe that hypothesis or significance testing and interpretation of the *P* value by this system of justice is too artificial and misses important information [11–13]. For example, it is sobering to demonstrate the distribution of *P* values by bootstrap sampling—yes, *P* values have their own confidence limits, too! These individuals would prefer that *P* values be interpreted simply as "degree of evidence," "degree of surprise," or "degree of belief" [14]. We agree with these ideas and suggest that rather than using *P* values for judging guilt or innocence (accepting or rejecting the null hypothesis), the *P* value itself should be reported as degree of evidence. In addition, machine learning ideas, which view data as consisting of signals buried in noise, have introduced multiple alternatives to *P* values that are less sensitive to *n*.

In using *P* values, some threshold is often set to declare a test statistic "significant." Sir Ronald Fisher, who introduced the idea of *P* values, wrote, "Attempts to explain the cogency of tests of significance in scientific research by hypothetical frequencies of possible statements being right or wrong seem to miss their essential nature. One who 'rejects' a hypothesis as a matter of habit, when $P \geq 1$ %, will be mistaken in not more than 1 % of such decisions. However, the

calculation is absurdly academic. No scientific worker has a fixed level of significance at which from year to year, and in all circumstances, he rejects hypotheses; he rather gives his mind to each particular case in the light of his evidence and his ideas" [15].

Human Error

Although it may seem that human error is distanced far from statistics, in fact, qualitative analysis of human error played a prominent role in how we approached statistics in the early 1970s. As early as 1912, Richardson recognized the need to eliminate "preventable disaster from surgery" [16]. Human errors as a cause of surgical failure are not difficult to find [17–19], particularly if one is careful to include errors of diagnosis, delay in therapy, inappropriate operations, omissions of therapy, and breaches of protocol. When we initially delved into what was known about human error in the era before Canary Island (1977), Three-Mile Island (1979), Bhopal (1984), Challenger (1986), and Chernobyl (1986), events that contributed enormously to developing formal knowledge of the cognitive nature of human error, we learned two lessons from investigating occupational and mining injuries [20, 21]. First, successful investigation of the role of the human element in injury depends on establishing an environment of *non-culpable error* [21]. The natural human reaction to investigation of error is to become defensive and to provide no information that might prove incriminating. An atmosphere of blame impedes investigating, understanding, and preventing error. How foreign this is from the culture of medicine! We take responsibility for whatever happens to our patients as a philosophic commitment [22, 23]. Yet cardiac operations are performed in a complex and imperfect environment in which every individual performs imperfectly at times [24]. It is too easy, when things go wrong, to look for someone to blame [25]. Blame by 20/20 hindsight allows many root causes to be overlooked [26]. Second, we learned that errors of *omission* exceed errors of *commission.* This is exactly what we found in ventricular septal defect repair, our first published study of human error [19], suggesting that the cardiac surgical environment is not so different from that of a gold mine and that we can learn from that literature.

Those who study human error suggest constructive steps for reducing it and, thus, surgical failure [27–30]. They affirm the necessity for intense apprentice-type training that leads to automatization of surgical skill and problem-solving rules [31], the value of simulators for acquiring such skills [32], and creating an environment that minimizes or masks potential distractions while supporting a system that discovers errors and allows recovery from them before injury occurs. In this regard, the pivotal study of human error during the arterial switch operation for transposition of the great arteries by de Leval and colleagues found that major errors were often realized and corrected by the surgical team, but minor ones were not, and the number of minor errors was strongly associated with adverse outcomes [33, 34].

This led Dr. Kirklin to suggest that there were two causes of surgical failure: lack of scientific progress and human error. The former meant that there were still gaps in knowledge that must be filled (research) in order to prevent these failures. The latter meant that we possess the knowledge to prevent human errors, but yet a failure occurred. A practical consequence of categorizing surgical failures into these two causes is that they fit the programmatic paradigm of "research and development": discovery on the one hand and application of knowledge to prevent failures on the other. The quest to reduce surgical failure by these two mechanisms is what drove us to use or develop methods to investigate these failures in a quantitative way.

Understanding Surgical Failure

Surgeons have intuitively understood that surgical failures, such as hospital mortality, may be related to a number of explanatory variables, such as renal and hepatic function [35]. However, we rarely know the causal sequence and final

mechanism of these failures, particularly when they occur after a complex heart operation. There is simply no way of knowing absolutely everything that may have influenced outcome. Although it is not at all satisfying, an alternative to a mechanistic explanation is to identify variables that appear to increase the risk of a patient experiencing an adverse event. This actually is a seminal idea in the history of biostatistics, and it was born and developed in the field of heart disease by investigators in the Framingham epidemiologic study of coronary artery disease [36]. Two papers are landmarks in this regard. In 1967, Walker and Duncan published their paper on multivariable analysis in the domain of logistic regression analysis, stating that "the purpose of this paper is to develop a method for estimating from dichotomous (quantal) or polytomous data the probability of occurrence of an event as a function of a relatively large number of independent variables" [37]. Then in 1976, Kannel and colleagues coined the term "risk factors" (actually "factors of risk"), noting that (1) "a single risk factor is neither a logical nor an effective means of detecting persons at high risk" and (2) "the risk function … is an effective instrument … for assisting in the search for and care of persons at high risk for cardiovascular disease" [38]. In 1979 the phrase "incremental risk factors" was coined at UAB to emphasize that risk factors add in a stepwise, or incremental, fashion to the risk present in the most favorable situation, as we will describe subsequently [39].

A Mathematical Framework for Risk

Multivariable analysis as described by the Framingham investigators required a model as the framework for binary outcomes such as death. The model they chose was the logistic equation, which had been introduced by Verhulst between 1835 and 1845 to describe population growth in France and Belgium [40, 41]. It describes a simple, symmetric S-shaped curve much like an oxygen dissociation curve, in which the horizontal axis is risk (where the lowest possible risk is at – infinity and the highest possible risk is at + infinity, and 50 % risk is at 0) and the vertical axis is the probability of experiencing an event. The model reappeared in the work of Pearl and Reed at Johns Hopkins University in 1920 [42], and then prominently at Mayo Clinic in the 1940, where Berkson described its use in bioassay (introducing terms such as the LD50 dose). The logistic equation was made a multivariable model of risk in the 1960s by Cornfield and colleagues [43] and Walker and Duncan [37].

Unlike most investigators, however, Dr. Kirklin and I approached the risk factor analysis differently from most. We wanted to know how best to use logistic regression to understand surgical failure. This led us to develop a framework to facilitate this process. It was steeped in a concept of incremental risk factors, philosophy, and nomograms.

Incremental Risk Factor Concept

As I described in 1980 at the congenital heart disease meeting in London, an incremental risk factor is a variable identified by multivariable analysis that is associated with an increased risk of an adverse outcome (surgical failure) [6, 39]. In the context of other simultaneously identified factors, the *magnitude* (strength) and *certainty* (*P* value) of an incremental risk factor represent its contribution over and above those of all other factors. Thus, it is incremental in two ways: (1) with respect to being associated with increased risk and (2) with respect to other factors simultaneously incorporated into a risk factor equation.

In understanding surgical failures, we believed the incremental risk factor concept was useful in several ways.

- Incremental risk factors are variables that reflect increased difficulty in achieving surgical success.
- Incremental risk factors are *common denominators* of surgical failure.
- Some incremental risk factors reflect *disease acuity.*
- Some incremental risk factors reflect *immutable conditions* that increase risk. These include extremes of age, genetic disorders, gender, and ethnicity.

- Some incremental risk factors reflect influential coexisting *noncardiac diseases* that shorten life expectancy in the absence of cardiac disease.
- Incremental risk factors are usually *surrogates* for true, but unmeasured or unrecognized, sources of surgical failure.
- An incremental risk factor may be a cause or mechanism of *surgical failure.* It is difficult to establish a causal mechanism outside the scope of a randomized, well-powered, and well-conducted generalizable clinical trial. This is because of confounding between selection factors influencing treatment recommendations and decisions and outcome. Balancing score methods (such as propensity score) attempt to remove such confounding and approach more closely causal inferences [44].
- Some incremental risk factors reflect *temporal experience*. The "learning curve" idea is intended to capture variables relating to experience of the surgical team, but also those representing temporal changes in approach or practice.
- Some incremental risk factors reflect *quality of care* and, as such, "blunt end" ramifications of institutional policies and practices, health care systems, and national and political decisions.
- Incremental risk factors reflect individual patient *prognosis.* They cannot be used to identify *which* patient will suffer a surgical failure, but they can be used to predict the *probability* of failure.
- However, incremental risk factors may be *spurious associations* with risk.

Philosophy

The inferential activity of understanding surgical failure, aimed at improving clinical results, is in contrast to pure description of experiences. Its motivation also contrasts with those aspects of "outcomes assessment" motivated by regulation or punishment, institutional promotion or protection, quality assessment by outlier identification, and negative aspects of cost justification or containment. These coexisting motivations stimulated us to identify, articulate, and contrast philosophies that informed our approach to analysis of clinical experiences. I have described these in detail in the Kirklin/Barratt-Boyes text *Cardiac Surgery,* but a few that were central to how we interpreted risk factors bear repeating [45].

Continuity Versus Discontinuity in Nature

Many risk factors related to outcome are measured either on an ordered clinical scale (ordinal variables), such as New York Heart Association (NYHA) functional class, or on a more or less unlimited scale (continuous variables), such as age. Three hundred years after Graunt, the Framingham Heart Disease Epidemiology Study investigators were faced with this frustrating problem [36, 46]. Many of the variables associated with development of heart disease were continuously distributed ones, such as age, blood pressure, and cholesterol level. To examine the relationship of such variables to development of heart disease, it was then accepted practice to categorize continuous variables coarsely and arbitrarily for cross-tabulation tables. Valuable information was lost this way. Investigators recognized that a 59-year-old's risk of developing heart disease was more closely related to that of a 60-year-old's than to that of the group of patients in the sixth versus seventh decade of life. They therefore insisted on examining the entire spectrum of continuous variables rather than subclassifying the information. What they embraced is a key concept in the history of ideas, namely, *continuity in nature.* The idea has emerged in mathematics, science, philosophy, history, and theology [47]. In our view, the common practice of stratifying age and other more or less continuous variables into a few discrete categories is lamentable, because it loses the power of continuity (some statisticians call this "borrowing power"). Focus on small, presumed homogeneous, groups of patients also loses the power inherent in a wide spectrum of heterogeneous, but related, cases. After all, any trend observed over an ever-narrower framework looks more and more like no trend at all! Like the Framingham

investigators, we therefore embraced continuity in nature unless it can be demonstrated that doing so is not valid, useful, or beneficial.

Linearity Versus Nonlinearity

Risk factor methodology introduced a complexity. The logistic equation is a symmetric S-shaped curve that expresses the relationship between a scale of risk and a corresponding scale of absolute probability of experiencing an event [39, 48]. The nonlinear relationship between risk factors and probability of outcome made medical sense to us. We could imagine that if all else positions a patient far to the left on the logit scale, a 1-logit-unit increase in risk would result in a trivial increase in the probability of experiencing an event. But as other factors move a patient closer to the center of the scale (0 logit units, corresponding to a 50 % probability of an event), a 1-logit-unit increase in risk makes a huge difference. This is consistent with the medical perception that some patients experiencing the same disease, trauma, or complication respond quite differently. Some are medically robust, because they are far to the left (low-risk region) on the logit curve before the event occurred. Others are medically fragile, because their age or comorbid conditions place them close to the center of the logit curve. This type of sensible, nonlinear medical relation made us want to deal with absolute risk rather than relative risk or risk ratios [49]. Relative risk is simply a translation of the scale of risk, without regard to location on that scale. Absolute risk integrates this with the totality of other risk factors.

Nihilism Versus Predictability

One of the important advantages of using equations such as the logistic equation is that they can be used to predict results for either groups of patients or individual patients. We recognize that when speaking of individual patients, we are referring to a prediction concerning the probability of events for that patient; we generally cannot predict exactly who will experience an event or when an event will occur. Of course, the nihilist will say, "You can't predict." A doctor cannot treat patients if he or she is a nihilist and believes that there is no way to predict if therapy will have an effect. Thus, although we do not want to overinterpret predictions from logistic models, we nevertheless believe the predictions made are better than "seat of the pants" guesses.

Parsimony Versus Complexity

Although clinical data analysis methods and results may seem complex at times, as in the large number of risk factors that must be assessed, an important philosophy behind such analysis is parsimony (simplicity). There are good reasons to embrace parsimony to an extent. One is that clinical data contain inherent redundancy, and one purpose of multivariable analysis is to identify that redundancy and thus simplify the dimensionality of the problem. A corollary is that there are likely variables that just introduce noise, and what we want to find is real signal. A second reason is that assimilation of new knowledge is incomplete unless one can extract the essence of the information. Thus, clinical inferences are often even more digested and simpler than the multivariable analyses. We must admit that simplicity is a virtue based on philosophic, not scientific, grounds. The concept was introduced by William of Ocken in the early fourteenth century as a concept of beauty—beauty of ideas and theories [50]. Nevertheless, it is pervasive in science. There are dangers associated with parsimony and beauty, however. The human brain appears to assimilate information in the form of models, not actual data [51]. Thus, new ideas, innovations, breakthroughs, and novel interpretations of the same data often hinge on discarding past paradigms (thinking "outside the box") [52]. There are other dangers in striving for simplicity. We may miss important relations because our threshold for detecting them is too high. We may reduce complex clinical questions to simple but inadequate questions that we know how to answer.

Nomograms

One of the most powerful tools to understand the relationship of incremental risk factors to surgical failure is graphics [53]. An important reason

for our using and even developing completely parametric models such as the logistic model was that they so easily allow graphics to be generated in the form of *nomograms,* as advocated by the Framingham investigators [49]. For example, if an analysis indicates an association of survival with young age, we want to know what the shape of that relationship is. Is it relatively flat for a broad range of age and then rapidly increasing at neonatal age? Or does risk increase or decrease rather linearly with age? Although the answers to these questions are contained within the numbers on computer printouts, these numbers are not easily assimilated by the mind. However, they can be used to demonstrate graphically the shape of the age relation with all other factors held constant.

Because graphics are so powerful in the process of generating new knowledge, an important responsibility is placed on the statistician to be sure that relations among variables are correct. Often, variables are examined and statistical inferences made simply to determine whether a continuous variable is a risk factor, without paying particular attention to what the data convey regarding the shape of the relationship to outcome. Instead, the statistician needs to focus during analysis on linearizing transformations of scale that may be needed to faithfully depict the relationship. Our experience indicates that most relations of continuous variables with outcome are smooth. They do not show sharp cut-offs, something we think investigators should be discouraged to look for.

Effectiveness, Appropriateness, Timing, and Indications for Intervention

Our initial focus was on surgical success and surgical failure (safety), but we soon began to investigate the effectiveness, appropriateness, and timing of intervention. The concept evolved that, only after we knew about the safety, effectiveness, long-term appropriateness, and optimal timing of possibly alternative interventions versus the natural history of disease, would we be able to define indications for intervention. This was subsequently embodied in the organization of each chapter of the Kirklin/Barratt-Boyes text *Cardiac Surgery* [45]. This was backward to the usual surgical thinking of the time, which began at indications rather than ended there.

What we immediately realized was that for most congenital heart lesions, knowledge of natural history was scant. In seeking sources of that information, we were faced with time-related mortality data in multiple different formats. Some data were typical right-censored (meaning that we knew the time of death—uncensored observations—and the time of follow-up of persons still surviving—censored observations). Others were presented as temporal blocks of data and counts of deaths (eg, died within first year, first year to age 5, 5–10, and so forth). Statisticians call this interval-censored count data. Yet others came from census data for which we knew nothing about deaths, only about patients in various time frames who were still alive (cross-sectional censored data). Dr. Kirklin was aware of, and had himself performed the calculations, for Berkson's life table method [44, 54], and I had worked for Dr. Paul Meier of Kaplan-Meier fame using the product-limit method [55]. But this heterogeneous type of data necessitated forging new collaboration with an expert in such matters, Dr. Malcolm Turner. He indicated to us that our problem went deeper than just the data: We needed to figure out how we would manipulate those data once we had answers to the natural history question. It was his belief that we once again needed to formulate equations that could be mathematically manipulated to identify, for example, optimal timing of intervention. Thus began a decade-long quest for a parametric model of survival, culminating in May 1983.

By that time two important things had happened. First, D. R. Cox in the United Kingdom had proposed a proportional hazards, semiparametric approach to multivariable analysis of survival data [56]. Dr. Naftel visited him, showed him many of the survival curves we had generated, and asked for his advice. To our dismay, he responded that it was highly unlikely that the idea of proportional hazards was appropriate for the

intervention data we showed him. Immediately after surgery mortality was elevated, and he opined that risk factors for this likely were very different from those pertaining to long-term mortality. Second, he thought the curves could be characterized as being of "low order" (that is, they could be described by a model with few parameters to estimate). Third, he believed that a fully parametric model is what we should be looking for so it could be easily manipulated not only for display of results, but for use in determining optimal timing of operation. Finally, he thought our group had enough mathematical firepower to figure this all out.

The second event was failure of the Braunwald-Cutter valve [57]. Advice was sought from all quarters, including industry, on how to analyze the data and possibly make the tough and potentially dangerous decision to remove these prostheses. This brought us into contact with Wayne Nelson, a General Electric statistician who was consulting for Alabama Power Company. He introduced us to the cumulative hazard method for calculating the life table, which brought several advantages [58]. First, it could be used to analyze repeatable events, such as repeat hospitalizations and adverse events. Second, each event could be coupled with what he called a cost value, such as severity of a non-fatal event, e.g., a stroke [59, 60]. Third, we needed to consider the competing risk of death as we thought about potentially non-fatal events.

Thus, in developing a comprehensive model for time-related events, of necessity we knew we had to take into account simultaneously the multiple formats the data might come in, repeatable events, weighting applied to these events, competing risks, and mathematical manipulation of all these.

Time-Related Events

Time-related events are often analyzed by a group of methods commonly called "actuarial." The word *actuarial* comes from the Latin *actuarius,* meaning secretary of accounts. The most notable actuarius was the Praetorian Prefect Domitius Ulpianus, who produced a table of annuity values in the early third century AD [4]. With emergence of both definitive population data and the science of probability, modern actuarial tables arose, produced first by Edmund Halley (of comet fame) in 1693 [61]. He was motivated, as was Ulpianus, by economics related to human survival, because governments sold annuities to finance public works [4]. Workers in this combined area of demography and economics came to be known as *actuaries* in the late eighteenth century. In the nineteenth century the actuary of the Alliance Assurance Company of London, Benjamin Gompertz, developed mathematical models of the dynamics of population growth to characterize survival [62]. In 1950, Berkson and Gage published their landmark medical paper on the life-table (actuarial) method for censored data, which they stated was no different from that used by others as early as at least 1922 [44, 54]. In 1952, Paul Meier at Johns Hopkins University and, in 1953, Edward Kaplan at Bell Telephone Laboratories submitted to the *Journal of the American Statistical Association* a new method for survival analysis, the product-limit method, that used more of the data. Estimates were generated at the time of each occurrence of an event. Further, the basis for the estimates was grounded in sound statistical theory. The journal editor, John Tukey, believed the two had discovered the same method, although presented differently, and insisted they join forces and produce a single publication. For the next 5 years, before its publication in 1958 [55], the two hammered out their differences in terminology and thinking, fearing all the while they would be scooped. The product-limit method (usually known as the Kaplan-Meier method), after considerable delay awaiting the advent of high-speed computers to ease the computation load, became the gold standard of nonparametric survival analysis. Until 1972, only crude methods were available to *compare survival curves* according to different patient characteristics [63–70]. The introduction by Cox of a proposal for multivariable survival analysis based on a semi-parametric proportional hazard method revolutionized the field [56].

Unlike nonparametric and even semi-parametric survival estimation based on counting (martingale) theory, model-based or parametric survival estimation arose out of biomathematical consideration of the force of mortality, the hazard function [71]. The hazard function was a unidirectional rate value or function that transported, as it were, survivors to death with the same mathematical relations as a chemical reaction (compartmental theory). This idea arose during the Great Plague of the sixteenth century. John Graunt, a haberdasher, assumed a constant risk of mortality (the mortality rate or force of mortality), which generates an exponentially decreasing survival function (as does radioactive decay). He called this constant unidirectional rate the *hazard function* after a technical term for a form of dicing that had by then come into common usage to mean "calamity" [71]. Because a constant hazard rate presumes a mathematical model of survival, his was a *parametric method.* Today, this expression of hazard is called the *linearized rate*.

Although linearized rates have been used to characterize time-related events after cardiac surgery, particularly by regulatory agencies, it is uncommon for hazard to be constant [72]. A challenge in devising, however, a time-varying parametric hazard model was that we often had only a small portion of the complete survival curve, such as 5- or 10-year survival after repair of a ventricular septal defect. The approach we finally figured out in the spring of 1983 was a temporal decomposition, much like putting light through a prism and depicting its colors [73]. Each component of the decomposition dominated a different time frame and could be modulated by its own set of risk factors, all estimated simultaneously.

Repeatable Events

Unlike death, morbid events such as thromboembolism or infection after transplantation may recur. The most common method of analysis is to focus only on its first occurrence, ignoring any further information beyond that point for the patients experiencing the event. However, true repeated-events analysis can be performed using the Nelson estimator. Basically, patients are not removed from the patients at risk after they experience the event. Thus, they are at risk of it again after experiencing it. A special case of repeated events is the industrial method known as "modulated renewal" [74]. The idea behind a modulated renewal process is that the industrial machine (or patient) is restarted at a new time zero each time the event occurs. This permits (1) ordinary Kaplan-Meier methods to be used, (2) the number of occurrences and intervals between each recurrence to be used in multivariable analyses, and (3) change in patient characteristics at each new time zero to be used in analyses. Thus, if the modulated renewal assumption can be shown to be valid, it increases the power and utility of the analysis tremendously.

Competing Risks

Competing risks analysis is a method of time-related data analysis in which multiple, mutually exclusive events are considered simultaneously [75, 76]. It is the simplest form of continuous-time Markov process models of transition among states [77]. In this simplest case, patients make a transition from an initial state (called event-free survival) to at most one other state that is considered to be terminating. Rates of transition from the initial state to one of the events (called an *end state*) are individual, independent functions.

Analysis of a single time-related event is performed in isolation of any other event. This is ideal for understanding that specific phenomenon. In contrast, competing risks analysis considers multiple outcomes in the context of one another. It is thus an integrative analysis.

In the early eighteenth century, some progress was made in the war against smallpox by inoculating people with small doses of the virus to establish immunity to the full-blown disease. Because governments at that time were supported in part by annuities, it was of considerable economic importance to know the consequences a cure of smallpox might bring upon the government's purse. Daniel Bernoulli tackled this

question by classifying deaths into mutually exclusive categories, one of which was death from smallpox [78]. For simplicity, he assumed that modes of death were independent of one another. He then developed kinetic equations for the rate of migration from the state of being alive to any one of several categories of being dead, including from smallpox. He could then compute how stopping one mode of death, smallpox, would influence both the number of people still alive and the redistribution of deaths into the other categories. (The triumph of the "war on smallpox" came in 1796, just 36 years after his publication).

Weighted Events

As noted in previous text, once one thinks "out of the box" beyond probability theory, one can begin to imagine that any non-fatal event could be characterized not only as having occurred, but with a "cost" associated with it. This might be actual cost of a medical readmission, for example [79], or length of stay, or a functional health assessment metric.

Longitudinal Data Analysis

Today, we look beyond occurrences of clinical events. How does the heart morphology and function change across time? How does a patient's functional health status change across time? How often does supraventricular tachycardia occur? What are the variables that modulate these longitudinal values? Importantly, do they influence clinical events? This is today's frontier of statistical methods.

Severe technologic barriers to comprehensive analysis of longitudinal data existed before the late 1980s [80]. Repeated-measures analysis of variance for *continuous* variables had restrictive requirements, including fixed time intervals of assessment and no censored data. *Ordinal* logistic regression for assessment of functional status was useful for assessments made at cross-sectional follow-up [81, 82], but not for repeated assessment at irregular time intervals with censoring. In the late 1980s, Zeger and his students and colleagues at Johns Hopkins University incrementally, but rapidly, evolved the scope, generality, and availability of what they termed "longitudinal data analysis" [83]. Their methodology accounts for correlation among repeated measurements in individual patients and variables that relate to both the ensemble and the nature of the variability. Because average response and variability are analyzed simultaneously, the technology has been called "mixed modeling." The technique has been extended to continuous, dichotomous, ordinal, and polytomous outcomes using both linear and nonlinear modeling.

Because of its importance in many fields of investigation, the methodology acquired different names. In 1982, Laird and Ware published a *random effects model* for longitudinal data from a frequentist school of thought [84]. In 1983, Morris presented his idea on *empirical Bayes* from a Bayesian school of thought [85]. In the late 1980s, members of Zeger's department at Johns Hopkins University developed the *generalized estimating equation* (GEE) approach [83]. Goldstein's addition to the Kendall series in 1995 emphasized the hierarchical structure of these models [86]. His is a particularly apt description. The general idea is that such analyses need to account for covariables that are measured or recorded at different hierarchical levels of aggregation. In the simplest cases, time is one level of aggregation, and individual patients with multiple measurements is another. These levels have their corresponding parameters that are estimated, and each may require different assumptions about variability (random versus fixed-effects distributions). Except under exceptional circumstances, these techniques have replaced former restrictive varieties of repeated-measures analysis, which we now consider of historical interest except for controlled experiments designed to exactly meet their assumptions.

Using the same strategy and mathematical formulation that Naftel, Blackstone, and Turner did for time-related events [73], we have introduced a longitudinal data analysis method by which the

temporal occurrence of a binary event, such as presence or absence of atrial fibrillation, is conceived as the addition of a number of temporal components, or phases. Each phase is modulated simultaneously by a log-linear additive function of risk factors. However, like all current methods, there is only primitive built-in capability for selecting variables for modulating the temporal components. Therefore, with a number of our colleagues and funding from the National Institutes of Health, we are actively developing new comprehensive methods for longitudinal data analysis.

Comparison of Treatments

Clinical Trials with Randomly Assigned Treatment

Controlled trials date back at least to biblical times, when casting of lots was used as a fair mechanism for decision-making under uncertainty (Numbers 33:54). An early clinical trial of a high protein vs. high calorie diet took place in the Court of Nebuchadnezzar, king of Babylon (modern Iraq). The first modern placebo-controlled, double-blinded, randomized clinical trial was carried out in England by Sir Austin Bradford Hill on the effectiveness of streptomycin versus bed rest alone for treatment of tuberculosis [87], although seventeenth and eighteenth century unblinded trials have been cited as historical predecessors [88–90]. Multi-institutional randomized clinical trials in pediatric and congenital heart disease have been championed by the Pediatric Heart Network over the last decade.

Randomization of treatment assignment has three valuable and unique characteristics:

- It eliminates selection factors (bias) in treatment assignment (although this can be defeated at least partially by enrollment bias).
- It distributes patient characteristics equally between groups, whether they are measured or not, known or unknown (balance), a well-accepted method of risk adjustment [91–94].
- It meets assumptions of statistical tests used to compare end points [93].

Randomized clinical trials are also characterized by concurrent treatment, excellent and complete compilation of data gathered according to explicit definitions, and proper follow-up evaluation of patients. These operational by-products may have contributed nearly as much new knowledge as the random assignment of treatment.

Unfortunately, it has become ritualistic for some to dismiss out of hand all information, inferences, and comparisons relating to outcome events derived from experiences in which treatment was not randomly assigned [95]. If this attitude is valid, then much of the information now used to manage patients with congenital heart disease would need to be dismissed and ignored!

Clinical Studies with Nonrandomly Assigned Treatment

Clinical studies with nonrandomly assigned treatment produce little knowledge when improperly performed and interpreted. Because this is often the case, many physicians have a strong bias against studies of this type. However, when properly performed and interpreted, and particularly when they are multi-institutional or externally validated, clinical studies of real-world experience can produce secure knowledge. During the 1980s, federal support for complex clinical trials in adult heart disease was abundant. Perhaps as a result, few of us noticed the important advances being made in statistical methods for valid, nonrandomized comparisons, now called “comparative effectiveness studies.” One example was the seminal 1983 *Biometrika* paper “The Central Role of the Propensity Score in Observational Studies for Causal Effects,” by Rosenbaum and Rubin [96]. In the 1990s, as the funding climate changed, interest in methods for making nonrandomized comparisons accelerated [97]. This interest has accelerated further in the twenty-first century.

Apples-to-apples nonrandomized comparisons of outcome can be achieved, within certain limitations, by use of so-called *balancing scores*, of which the *propensity score* is the simplest [96]. Balancing scores are a class of multivariable

statistical methods that identify patients with similar chances of receiving one or the other treatment. Perhaps surprisingly, even astonishingly, patients with similar balancing scores are well balanced with respect to at least all patient, disease, and comorbidity characteristics taken into account in forming the balancing score. This balancing of characteristics permits the most reliable nonrandomized comparisons of treatment outcomes available today [98].

The essential approach to a comparison of treatment outcomes in a nonrandomized setting is to design the comparison as if it were a randomized clinical trial and to interpret the resulting analyses as if they emanated from such a trial. This essential approach is emphasized in Rubin's 2007 article, "The Design Versus the Analysis of Observational Studies for Causal Effects: Parallels with the Design of Randomized Trials [99]. As noted by Rubin, "I mean all contemplating, collecting, organizing, and analyzing data that takes place prior to seeing any outcome data." He emphasizes by this statement his thesis that a nonrandomized set of observations should be conceptualized as "a broken randomized experiment…with a lost rule for patient allocation, and specifically for the propensity score, which the analysis will attempt to construct." For example, the investigator should ask, "Could each patient in all comparison groups be treated by all therapies considered? If not, this constitutes specific inclusion and exclusion criteria. If this were a randomized trial, when would randomization take place? One must only use variables to construct a propensity score that would be known at the time randomization would have occurred, not after that; this means that variables chosen in the propensity score analysis are not those that could possibly be affected by the treatment."

The most common use of the propensity approach is to match pairs of patients on the basis of their propensity score alone. Outcomes can then be compared between groups of matched pairs. However, just as in a randomized trial, the results will be applicable to patients who match the characteristics of the propensity groups.

Where Have We Been and Where Are We Headed?

Analysis, as expressed by Sir Isaac Newton, is that part of an inductive scientific process whereby a small part of nature (a phenomenon) is examined in the light of observations (data) so that inferences can be drawn that help explain some aspect of the workings of nature [100].

Philosophies underpinning methods of data analysis have evolved rapidly since the latter part of the nineteenth century and may be at an important crossroad. Stimulated in large part by the findings of his cousin Charles Darwin, Sir Francis Galton, along with Karl Pearson and Francis Edgeworth, established at that time what has come to be known as *biostatistics* [101]. Because of the Darwinian link, much of their thinking was directed toward an empirical study of genetics versus environmental influence on biological development. It stimulated development of the field of eugenics (human breeding) [102] and the study of mental and even criminal characteristics of humans as they relate to physical characteristics (profiling). The outbreak of World War I led to development of statistics related to quality control. Sir Ronald Fisher formalized a methodologic approach to experimentation, including randomized designs [103]. The varying milieus of development led to several competing schools of thought within statistics, such as frequentist and Bayesian, with different languages and different methods [104]. Formalization of the discipline occurred, and whatever the flavor of statistics, it came to dominate the analytic phase of inferential data analysis, perhaps because of its empirical approach and lack of underlying mechanistic assumptions.

Simultaneously, the discipline of *biomathematics* arose, stimulated in particular by the need to understand the growth of organisms (allometric growth) and populations in a quantitative fashion. Biomathematicians specifically attempt to develop mathematical models of natural phenomena such as clearance of pharmaceuticals, enzyme kinetics, and blood flow dynamics. These continue to be important today in understanding such altered physiology as cavopulmonary shunt

flow [105]. Many of the biomathematical models came to compete with statistical models for distribution of values for variables, such as the distribution of times to an event.

Advent of the fast Fourier transform in the mid-1960s [106] led to important medical advances in filtering signal from noise and image processing. The impetus for this development came largely from the communications industry, so only a few noticed that concepts in communication theory coincided with those in statistics and mathematics.

As business use of computers expanded, and more recently as genomic data became voluminous, computer scientists developed methods for examining large stores of data [107]. These included data mining in business and computational biology and bioinformatics in the life sciences. Problems of classification (such as of addresses for automating postal services) led to such tools as neural networks [16], which have been superseded in recent years by an entire discipline of machine learning [107, 108].

In the past quarter century, all these disciplines of mathematics, computer science, information modeling, and digital signal processing have been vying for a place in the analytic phase of clinical research that in the past has largely been dominated by biostatistics. Specifically, advanced statistics and algorithmic data analysis have conquered the huge inductive inference problem of disparity between number of parameters to be estimated and number of subjects (e.g., in genetics, hundreds of thousands of variables for n = 1) [109]. Advanced high-order computer reasoning and logic have taken the Aristotelian deterministic approach to a level that allows intelligent agents to connect genotype with phenotype [110]. It may be rational to believe that the power of these two divergent approaches to science can be combined in such a way that very "black-box" but highly predictive methods can be explored by intelligent agents that report the logical reasons for a black-box prediction [52].

Fortunately for those of us in cardiac surgery, we need not be threatened by these alternative voices, but rather can seize the opportunity to discover how each can help us understand the phenomena in which we are interested.

References

1. Kirklin JW, Dushane JW, Patrick RT, Donald DE, Hetzel PS, Harshbarger HG, et al. Intracardiac surgery with the aid of a mechanical pump-oxygenator system (Gibbon type): report of eight cases. Proc Staff Meet Mayo Clin. 1955;30:201–6.
2. Blackstone EH. Born of necessity: the dynamic synergism between advancement of analytic methods and generation of new knowledge. J Heart Valve Dis. 1995;4:326–36.
3. David FN. Games, gods & gambling: a history of probability and statistical ideas. London: Charles Griffin; 1962.
4. Hacking J. The emergence of probability. Cambridge: Cambridge University Press; 1975. p. 102.
5. Galilei G. Sopra le scoperte dei dadi, as summarized in R. Langley: practical statistics simply expanded. New York: Dover; 1970.
6. Blackstone EH. Thinking beyond the risk factors. Eur J Cardiothorac Surg. 2006;29:645–52.
7. Ware JH, Mosteller F, Delgado F, Donnelly C, Ingelfinger AJ. P values. In: Bailar JC, Mosteller F, editors. Medical uses of statistics. 2nd ed. Boston: NEJM Books; 1992.
8. Grunkemeier GL, Wu Y, Furnary AP. What is the value of a p value? Ann Thorac Surg. 2009;87:1337–43.
9. Hubbard R, Lindsay RM. Why P values are not a useful measure of evidence in statistical significance testing. Theor Psychol. 2008;18:69–88.
10. Senn S. Two cheers for P-values? J Epidemiol Biostat. 2001;6:194–204.
11. Barnard GA. Must clinical trials be large? The interpretation of P-values and the combination of test results. Stat Med. 1990;9:601–14.
12. Kempthorne O. Of what use are tests of significance and tests of hypotheses? Commun Statist Theor Method A. 1976;5:763.
13. Salsburg D. Hypothesis versus significance testing for controlled clinical trials: a dialogue. Stat Med. 1990;9:201–11.
14. Burack JH, Impellizzeri P, Homel P, Cunningham Jr JN. Public reporting of surgical mortality: a survey of New York State cardiothoracic surgeons. Ann Thorac Surg. 1999;68:1195–200.
15. Fisher RA. Statistical methods and scientific inference. 3rd ed. New York: Hafner; 1973.
16. Richardson MH. The gradual elimination of the preventable disaster from surgery. Thorac Med Assoc. 1912:181.
17. Gawande A. Complications: a surgeon's notes on an imperfect science. New York: Metropolitan Books; 2002.
18. Kirklin JW, Karp RB. The tetralogy of Fallot, from a surgical point of view. Philadelphia: WB Saunders; 1970.

19. Rizzoli G, Blackstone EH, Kirklin JW, Pacifico AD, Bargeron Jr LM. Incremental risk factors in hospital mortality rate after repair of ventricular septal defect. J Thorac Cardiovasc Surg. 1980;80:494–505.
20. Lawrence AC. Human error as a cause of accidents in gold mining. J Safety Res. 1974;6:78–88.
21. Wigglesworth EC. A teaching model of injury causation and a guide for selecting countermeasures. Occup Psychol. 1972;46:69–78.
22. Berry DA, Stangl DK. Meta-analysis in medicine and health policy. New York: Marcel Dekker; 2000.
23. McIntyre N, Popper K. The critical attitude in medicine: the need for a new ethics. Br Med J (Clin Res Ed). 1983;287:1919.
24. Christensen JF, Levinson W, Dunn PM. The heart of darkness: the impact of perceived mistakes on physicians. J Gen Intern Med. 1992;7:424–31.
25. Wu AW, Folkman S, McPhee SJ, Lo B. Do house officers learn from their mistakes? JAMA. 1991;265:2089–94.
26. Reason JT, Carthey J, de Leval MR. Diagnosing "vulnerable system syndrome": an essential prerequisite to effective risk management. Qual Health Care. 2001;10 Suppl 2:ii21–5.
27. Cook RI, Wreathall J, Smith A, Cronin DC, Rivero O, Harland RC, et al. Probabilistic risk assessment of accidental ABO-incompatible thoracic organ transplantation before and after 2003. Transplantation. 2007;84: 1602–9.
28. Cooper JB, Newbower RS, Kitz RJ. An analysis of major errors and equipment failures in anesthesia management: considerations for prevention and detection. Anesthesiology. 1984;60:34–42.
29. Leape LL. Error in medicine. JAMA. 1994;272: 1851–7.
30. Leape LL, Lawthers AG, Brennan TA, Johnson WG. Preventing medical injury. QRB Qual Rev Bull. 1993;19:144–9.
31. Reason J. Combating omission errors through task analysis and good reminders. Qual Saf Health Care. 2002;11:40–4.
32. Hinske LC, Sandmeyer B, Urban B, Hinske PM, Lackner CK, Lazarovici M. The human factor in medical emergency simulation. AMIA Annu Symp Proc. 2009;2009:249–53.
33. Carthey J, de Leval MR, Reason JT. The human factor in cardiac surgery: errors and near misses in a high technology medical domain. Ann Thorac Surg. 2001;72: 300–5.
34. de Leval MR, Carthey J, Wright DJ, Farewell VT, Reason JT. Human factors and cardiac surgery: a multicenter study. J Thorac Cardiovasc Surg. 2000;119: 661–72.
35. Lew RA, Day Jr CL, Harrist TJ, Wood WC, Mihm Jr MC. Multivariate analysis. Some guidelines for physicians. JAMA. 1983;249:641–3.
36. Gordon T. Statistics in a prospective study: the Framingham Study. In: Gail MH, Johnson NL, editors. Proceedings of the American Statistical Association: sesquicentennial invited paper sessions. Alexandria: ASA; 1989. p. 719–26.
37. Walker SH, Duncan DB. Estimation of the probability of an event as a function of several independent variables. Biometrika. 1967;54:167–79.
38. Kannel WB, McGee D, Gordon T. A general cardiovascular risk profile: the Framingham Study. Am J Cardiol. 1976;38:46–51.
39. Kirklin JW. A letter to Helen (presidential address). J Thorac Cardiovasc Surg. 1979;78:643–54.
40. Verhulst PF. Notice sur la loi que la population suit dans son accroissement. Math Phys. 1838;10:113.
41. Verhulst PF. Recherches mathematiques sur la loi d'accroissement de la population. Nouv Mem Acad R Sci Belleslett Brux. 1845;18:1.
42. Pearl R, Reed LT. On the rate of growth of the population of the United States since 1790 and its mathematical representation. Proc Natl Acad Sci. 1920;6:275–88.
43. Cornfield J, Gordon T, Smith WS. Quantal response curves for experimentally uncontrolled variables. Bull Int Stat Inst. 1961;3:97–115.
44. Berkson J, Flexner LB. On the rate of reaction between enzyme and substrate. J Gen Phys. 1928;11: 433–57.
45. Kouchoukos NT, Blackstone EH, Hanley FL, Kirklin J. Cardiac surgery. 4th ed. Philadelphia: Elsevier; 2012. p. 251–352.
46. Gordon T, Kannel WB. Predisposition to atherosclerosis in the head, heart, and legs. The Framingham study. JAMA. 1972;221:661–6.
47. Bland JM. Sample size in guidelines trials. Fam Pract. 2000;17:S17–20.
48. Berkson J. Application of the logistic function to bioassay. J Am Stat Assoc. 1944;39:357–65.
49. Gordon T, Kannel WB. Multiple risk functions for predicting coronary heart disease: the concept, accuracy, and application. Am Heart J. 1982;103:1031–9.
50. Formigari L. Chain of being. In: Wiener PP, editor. Dictionary of the history of ideas: studies of selected pivotal ideas. New York: Charles Scribner's Sons; 1968. p. 325–35.
51. Reason JT. Human error. Cambridge: Cambridge University Press; 1990.
52. Hrushesky WJ. Triumph of the trivial. Perspect Biol Med. 1998;41:341–8.
53. Kirklin JK, Blackstone EH. Notes from the editors: figures. J Thorac Cardiovasc Surg. 1994;107:1175–7.
54. Murphy RD, Papps PC. Construction of mortality tables from the records of insured lives. New York: The Actuarial Society of America; 1922.
55. Kaplan EL, Meier P. Nonparametric estimation from incomplete observations. J Am Stat Assoc. 1958;53: 457–81.
56. Cox DR. Regression models and life tables. J Roy Statist Soc Ser B. 1972;34:187–220.
57. Blackstone EH, Kirklin JW, Pluth JR, Turner ME, Parr GV. The performance of the Braunwald-Cutter aortic prosthetic valve. Ann Thorac Surg. 1977;23: 302–18.
58. Nelson W. Theory and applications of hazard plotting for censored failure data. Technometrics. 1972;14: 945–66.

59. Nelson W. Graphical analysis of system repair data. J Qual Technol. 1988;20:24–35.
60. Nelson W. Confidence limits for recurrence data: applied to cost or number of product repairs. Technometrics. 1995;37:147–57.
61. Halley E. An estimate of the degrees of the mortality of mankind, drawn from curious tables of the births and funerals of the city of Breslau. Philos Trans R Soc Lond. 1693;17:596.
62. Gompertz B. On the nature of the function expressive of the law of human mortality. Philos Trans R Soc Lond. 1825;115:513–83.
63. Gehan EA. A generalized Wilcoxon test for comparing arbitrarily singly-censored samples. Biometrika. 1965;52:203–25.
64. Lilienfeld DE, Pyne DA. On indices of mortality: deficiencies, validity, and alternatives. J Chronic Dis. 1979;32:463–8.
65. Mantel N. Evaluation of survival data and two new rank order statistics arising in its consideration. Cancer Chemother Rep. 1966;50:163–70.
66. Mantel N, Haenszel W. Statistical aspects of the analysis of data from retrospective studies of disease. J Natl Cancer Inst. 1959;22:719–48.
67. O'Neill TJ. Distribution-free estimation of cure time. Biometrika. 1979;66:184–7.
68. Peto R, Peto J. Asymptotically efficient rank invariant test procedures. J R Statist Soc A. 1972;135:185–207.
69. Prentice RL, Marek P. A qualitative discrepancy between censored data rank tests. Biometrics. 1979;35:861–7.
70. Wilcoxon F. Individual comparisons by ranking methods. Biomet Bull. 1947;1:80–3.
71. Graunt J. Natural and political observations made upon the bills of mortality. Baltimore: Johns Hopkins University Press; 1939. 1662. Reprint.
72. Grunkemeier GL, Thomas DR, Starr A. Statistical considerations in the analysis and reporting of time-related events. Application to analysis of prosthetic valve-related thromboembolism and pacemaker failure. Am J Cardiol. 1977;39:257–8.
73. Blackstone EH, Naftel DC, Turner Jr ME. The decomposition of time-varying hazard into phases, each incorporating a separate stream of concomitant information. J Am Stat Assoc. 1986;81:615–24.
74. Kalbfleisch JD, Prentice RL. The statistical analysis of failure time data. 2nd ed. New York: Wiley; 2002.
75. David HA, Moeschberger ML. The theory of competing risks. New York: Macmillan; 1978.
76. Prentice RL, Kalbfleisch JD, Peterson Jr AV, Flournoy N, Farewell VT, Breslow NE. The analysis of failure times in the presence of competing risks. Biometrics. 1978;34:541–54.
77. Andersen PK, Hansen LS, Keiding N. Assessing the influence of reversible disease indicators on survival. Stat Med. 1991;10:1061–7.
78. Berkson J, Hollander F. Chemistry–on the equation for the reaction between invertase and sucrose. J Wash Acad Sci. 1930;20:157.
79. Smedira NG, Hoercher KJ, Lima B, Mountis MM, Starling RC, Thuita L, et al. Unplanned hospital readmissions after HeartMate II implantation. J Am Coll Cardiol Heart Fail. 2013;1:31–9.
80. Blackstone EH. Actuarial and Kaplan-Meier survival analysis: there is a difference. J Thorac Cardiovasc Surg. 1999;118:973–5.
81. Fontan F, Kirklin JW, Fernandez G, Costa F, Naftel DC, Tritto F, et al. Outcome after a "perfect" Fontan operation. Circulation. 1990;81:1520–36.
82. Hickey MS, Blackstone EH, Kirklin JW, Dean LS. Outcome probabilities and life history after surgical mitral commissurotomy: implications for balloon commissurotomy. J Am Coll Cardiol. 1991;17:29–42.
83. Diggle PJ, Heagerty PJ, Liang KY, Zeger SL. Analysis of longitudinal data. 2nd ed. New York: Oxford University Press; 2002.
84. Laird NM, Ware JH. Random-effects models for longitudinal data. Biometrics. 1982;38:963–74.
85. Morris CN. Parametric empirical Bayes inference: theory and applications. J Am Stat Assoc. 1983;78:47–55.
86. Goldstein H. Multilevel statistical models. 2nd ed. London: Arnold; 1995.
87. Steyerberg EW, Eijkemans MJ, Harrell Jr FE, Habbema JD. Prognostic modelling with logistic regression analysis: a comparison of selection and estimation methods in small data sets. Stat Med. 2000;19:1059–79.
88. Chalmers I. Comparing like with like: some historical milestones in the evolution of methods to create unbiased comparison groups in therapeutic experiments. Int J Epidemiol. 2001;30:1156–64.
89. Dodgson SJ. The evolution of clinical trials. J Eur Med Writers Assoc. 2006;15:20–1.
90. Milne I, Chalmers I. A controlled clinical trial in 1809? J Epidemiol Community Health. 2002;56:1.
91. Bhudia SK, McCarthy PM, Kumpati GS, Helou J, Hoercher KJ, Rajeswaran J, et al. Improved outcomes after aortic valve surgery for chronic aortic regurgitation with severe left ventricular dysfunction. J Am Coll Cardiol. 2007;49:1465–71.
92. Bunker JP, Barnes BA, Mosteller F, editors. Costs, risks, and benefits of surgery. New York: Oxford University Press; 1977.
93. Burton PR, Gurrin LC, Campbell MJ. Clinical significance not statistical significance: a simple Bayesian alternative to p values. J Epidemiol Community Health. 1998;52:318–23.
94. Weinstein MC. Allocation of subjects in medical experiments. N Engl J Med. 1974;291:1278–85.
95. Burdette WI, Gehan EA. Planning and analysis of clinical studies. Springfield: Charles C Thomas; 1970.
96. Rosenbaum PR, Rubin DB. The central role of the propensity score in observational studies for causal effects. Biometrika. 1983;70:41–55.
97. Rubin DB. Estimating causal effects from large data sets using propensity scores. Ann Intern Med. 1997;127:757–63.

98. Blackstone EH. Comparing apples and oranges. J Thorac Cardiovasc Surg. 2002;123:8–15.
99. Rubin DB. The design versus the analysis of observational studies for causal effects: parallels with the design of randomized trials. Stat Med. 2007;26:20–36.
100. Newton I. Philosophiae Naturalis Principia Mathematica; 1687.
101. Stigler SM. Statistics on the table: the history of statistical concepts and methods. Cambridge: Harvard University Press; 2002.
102. Gillham NW. Sir Francis Galton and the birth of eugenics. Annu Rev Genet. 2001;35:82–101.
103. Fisher RA. Statistical methods for research workers. 1925. Reprint, 14th edition. New York: Hafner; 1970.
104. Howie D. Interpreting probability: controversies and developments in the early twentieth century. Cambridge: Cambridge University Press; 2002.
105. de Leval MR, Dubini G, Migliavacca F, Jalali H, Camporini G, Redington A, et al. Use of computational fluid dynamics in the design of surgical procedures: application to the study of competitive flows in cavopulmonary connections. J Thorac Cardiovasc Surg. 1996;111:502–13.
106. Cooley JW, Tukey JW. An algorithm for machine calculation of complex Fourier series. Math Comput. 1965;19:297–301.
107. Breiman L. Bagging predictors. Mach Learn. 1996; 24:123–40.
108. Hastie T, Tibshirani R, Friedman JH. The elements of statistical learning: data mining, inference, and prediction. New York: Springer; 2001.
109. Zhang M, Zhang D, Wells MT. Variable selection for large p small n regression models with incomplete data: mapping QTL with epistases. BMC Bioinforma. 2008;9:251–9.
110. Giesl J. Current trends in automated deduction. Künstl Intell. 2010;24:11–3.

3 Introduction: Using Data to Drive Change and Improvement: The Legacy of Florence Nightingale

Kathleen Mussatto and Maryanne Kessel

Abstract

This introduction summarizes the legacy of Florence Nightingale's important work as a leader of nursing on modern day nursing, evidence-based practice, and health care administration. The relevance of Florence Nightingale's teachings to the care of children and adults with pediatric heart disease is highlighted.

Keywords

Florence Nightingale • Evidence-based practice • Nursing • Health care administration

> Were there none who were discontented with what they have, the world would never reach anything better.
>
> —Florence Nightingale

Florence Nightingale, a celebrated British reformer, statistician, and the founder of modern nursing, is credited as being the first clinician (nurse) and epidemiologist to use data to describe patient experiences and outcomes. She was a master in creating visualizations of statistics to document the experiences of soldiers during the Crimean War (Fig. 3.1). She was a pioneer of evidence-based practice in nursing. Her legend continues to guide nursing practice, including the care of children with congenital heart disease, at all levels, from direct care, to research, and administration.

Nightingale identified that more soldiers were dying in the Crimean War in 1854 from infection due to poor health care conditions, than from wounds inflicted on the battlefield. She instituted nurse-led infection control strategies that reduced mortality from 42 % to 2 %. Poor nutrition, inadequate supplies, and stress on the soldiers were cited as major contributors to poor outcomes. These same risk factors continue to contribute to patient morbidity.

Health care providers are called to practice from a sound evidence base in order to provide

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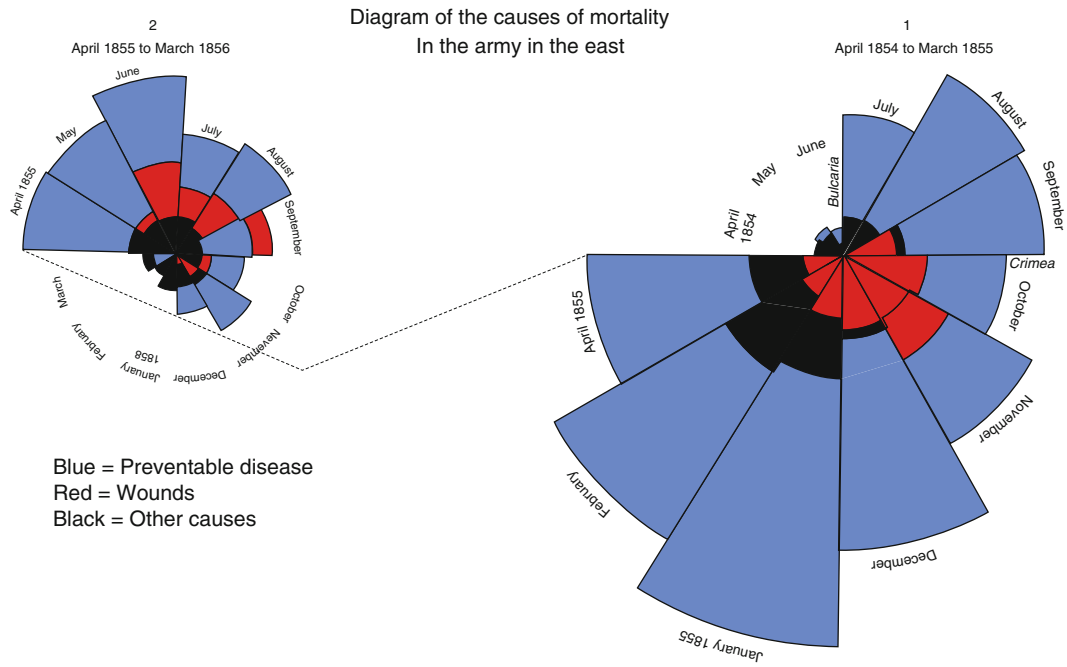

Fig. 3.1 An example of Florence Nightingale's wedge diagram

care of the highest value. This means that we must move beyond dogma, expert opinion, and practice driven by tradition. Instead, we must learn how to find, interpret, and use evidence and data to ground our practice in a sound, scientifically-validated base. Our policies and procedures should reflect the state of the science and set a standard of practice that promotes provision of the best and safest care available to our patients.

It is imperative that our nursing, medical, and allied health professional schools teach the skills necessary to critically evaluate evidence presented in the literature. Study design, the source of the data, inclusion and exclusion criteria, potential for bias, and the analytical approach all need to be examined with a critical lens. We must also be critical in evaluating whether the available evidence is sufficient to guide practice or whether further research is needed. Nightingale's work was key in revolutionizing hospital data collection. She knew that rigorous research and the best and safest care depended on accurate data that could be easily transformed into information to influence care decisions.

We do not practice in isolation. We practice within systems, large and small, academic and non-academic. High quality data is equally important at the administrative level. The role of nursing leaders in administration has evolved to emphasize collaboration with physician leaders. We recognize that administration and clinicians have a singular, common purpose, delivering the best and safest care to our patients. Working together will improve our progress toward this goal.

In our current healthcare environment of decreased reimbursement and with the data that disputes that "more care is better care" [1] there has never been a more compelling need for administrative and clinical data to ground the new direction that we as healthcare leaders must navigate. Total expenditure on health continues to rise exponentially. Whenever possible, medicine based on anecdotal evidence must be replaced by fully transparent data-driven models of care with appropriate governance. Providers

and administrators have the opportunity to collaborate in a focused and disciplined way to provide appropriate resources to maintain high quality care in a cost effective manner, all of which will contribute to healthcare sustainability. Florence Nightingale would have been proud to see nursing administrators present at this innovative table where tough decisions will need to be made when we have to make do with less.

In summary, the legacy of Florence Nightingale has cast its shadow on all aspects of health care delivery, not just nursing. She recognized that knowledge and skill were needed to improve public health. With great courage and determination she single-handedly set about changing conditions for the injured soldiers of the Crimean War. She used data and statistics to support her efforts and gained the respect of health care providers and public policy makers worldwide. Nightingale's greatest contribution to nursing may have been her emphasis on nursing integrity and the role of the nurse in helping patients to live their fullest regardless of the conditions they faced. In the 1880s, Nightingale wrote that it would be 100–150 years before educated, experienced nurses reached large enough numbers to change the healthcare system. We are those nurses today, we need to be leaders in both practice and administration. We must carry the legacy of Florence Nightingale forward by creating and using high-quality data, striving for evidence-based practice, and taking an active role in health care administration and public policy.

The chapters in this book will guide you in developing skills in seeking, finding, interpreting, and implementing meaningful data into evidence-based practice and administrative decision making. Optimizing the health and healing of children, adults, and families facing congenital heart disease requires the courage and determination that Nightingale modeled. We will do well to walk in her shadow.

Reference

1. World Health Organization. World health statistics 2013: a wealth of information on global public health. Geneva: WHO Press; 2013. Available at: http://apps.who.int/iris/bitstream/10665/82058/1/WHO_HIS_HSI_13.1_eng.pdf?ua=1&ua=1. Accessed 14 June 2014.

4 Introduction: Quality Improvement and Databases in the Context of Professionalism

John E. Mayer Jr.

Abstract

Throughout much of modern history, the physician has had two roles, "healer of the sick" and "member of a profession". Although the distinction between these two roles has not been commonly appreciated, these roles have different historical origins and involve different activities. Viewed from this context, quality improvement is a fundamental responsibility of a profession, and databases and registries, created and maintained by professional groups that are focused on patient outcomes, are a critically important tool by which quality improvement can occur. Database/registry participation, careful review of the resulting outcomes data, and active attempts to improve the quality of our patients' outcomes are thus fundamental to both being better "healers of the sick" and responsible "members of a profession." .

Keywords

Profession • Quality Improvement • Outcomes Databases

Throughout much of modern history, the physician has had two roles, "healer of the sick" and "member of a profession" [1]. Although the distinction between these two roles has not been commonly appreciated, these roles have different historical origins and involve different activities [1]. Professions were created by and exist for the benefit of the general society as a means of organizing the delivery of complex services which society requires, including that of the healer. Characteristics of a profession include (1) an occupation whose core element is work based upon mastery of a complex body of knowledge and skills (2) knowledge or practice of a knowledge-based art that is used in the service of others, (3) governance by codes of ethics, (4) commitments to competence, integrity and morality, and (5) altruism and promotion of the public good [2]. The relationship between society and a profession has been described as a "social contract" with implied prerogatives and responsibilities for each profession [1]. Among the most

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important of these professional responsibilities is the expectation that the profession will act in the societal interest and not its own [3]. Other responsibilities articulated by a number of different individuals and organizations include (1) maintaining, advancing, and disseminating a body of knowledge, (2) setting and enforcing its own standards and values, i.e. self-regulation, (3) cherishing performance above personal reward, (4) placing patients' interests above their own, and (5) fairly distributing finite medical resources [2].

Viewed from this context, quality improvement is a fundamental responsibility of a profession, and databases and registries, created and maintained by professional groups that are focused on patient outcomes, are a critically important tool by which quality improvement can occur. These databases foster the dissemination of knowledge and mastery of a complex body of knowledge and can be an important vehicle for setting and enforcing standards and evaluating performance. Ultimately these databases and registries can serve as a mechanism for the wise allocation of societal resources for healthcare, provided that both government and private payors will provide the resource utilization data. Database/registry participation, careful review of the resulting outcomes data, and active attempts to improve the quality of our patients' outcomes are thus fundamental to both being better "healers of the sick" and responsible "members of a profession." The accompanying body of work represents a clear manifestation of these professional efforts. This herculean effort led by 4 internationally respected leaders and drawing the best wisdom around the world represents a major milestone in the development and progress of safe and high valued pediatric cardiac care.

References

1. Creuss RL, Creuss SR. The Osler Fellows program: physicianship, professionalism, and medicine's social contract with society [Powerpoint file]. Available at: http://www.medicine.mcgill.ca/physicianship/Presentations/Intro%20to%20Preofessional%20for%20Osler%20Fellows.ppt. Accessed 13 Apr 2014.
2. Mayer Jr JE. Is there a role for the medical profession in solving the problems of the American health care system? Ann Thorac Surg. 2009;87:1655–61.
3. Krause EA. Death of the guilds: professions, states, and the advance of capitalism, 1930 to the present. New Haven: Yale University Press; 1996. p. 29–49.

Part II

Nomenclature and Taxonomies

5 Nomenclature for Congenital and Pediatric Cardiac Disease: Historical Perspectives and the International Pediatric and Congenital Cardiac Code

Rodney C.G. Franklin, Jeffrey P. Jacobs, Otto N. Krogmann, and Marie J. Béland

Abstract

The International Pediatric and Congenital Cardiac Code (IPCCC) was created by the International Society of Nomenclature for Paediatric and Congenital Heart Disease (ISNPCHD) to name and classify pediatric and congenital cardiac disease and its treatment. It is a comprehensive code that can be freely downloaded from the internet (http://www.IPCCC.net) and is already in use worldwide, particularly for international comparisons of outcomes. The goal of this effort is to create strategies for stratification of risk and to improve healthcare for the individual patient. The collaboration with the World Health Organization, the International Health Terminology Standards Development Organization, and the healthcare industry, will lead to further enhancement of the IPCCC, and to its more universal use.

Keywords

Nomenclature • Database • Classification • ICD • SNOMED-CT • IPCCC

on behalf of the International Society for Nomenclature of Paediatric and Congenital Heart Disease

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Introduction

Clinicians working in the field of congenital and pediatric cardiology have long felt the need for a common diagnostic and therapeutic nomenclature and coding system with which to classify patients of all ages with congenital and acquired cardiac disease. Over the last 15 years, this desire has been heightened by the need to provide national and international comparisons of surgical results between centers caring for these patients, following high profile enquiries such as that examining the outcome of pediatric cardiac surgery at the unit in Bristol, in the United Kingdom [1, 2], as well as similar events in Sydney Australia [3], Winnipeg Canada [4], Denver Colorado [5–11], and Lexington Kentucky [12].

The incorporation of effective clinical governance and best practice into our specialty requires the harvesting of accurate and validated data on the diagnosis, treatment, and outcome of these patients from prenatal life through to adulthood. Such a system facilitates the comparison of outcomes following interventions between individual centers, whilst taking into account the mix of cases involved, accompanying risk factors and comorbidities, as well as postprocedural complications. Benchmarking against those units who perform best allows analysis of relevant and genuine factors underlying differing outcomes, and instigation of improvements, in terms of both mortality and morbidity. For this objective to be achieved, it is essential to have a comprehensive and standardized system of coding and classification, using mutually exclusive and unambivalent terms. The system must be easy to use, preferably in multiple languages, be digitally compatible with different software database systems, and also fulfil the needs and expectations of widely different cultures of practice.

Although historically many centers developed their own systems of classification for internal audit, and some co-operative work between centers nationally and across international boundaries has occurred, these systems were dissimilar enough to preclude the large scale studies needed to understand outcomes from the heterogeneous population of patients with congenitally malformed hearts. A cohesive and comprehensive system of nomenclature, suitable for setting a global standard for multicentric analysis of outcomes and stratification of risk, has only relatively recently emerged, namely, the freely available International Pediatric and Congenital Cardiac Code (IPCCC), as developed and copyrighted by the International Society for Nomenclature of Paediatric and Congenital Heart Disease (ISNPCHD). This review gives an historical perspective on the development of systems of nomenclature in general, and specifically with respect to the diagnosis and treatment of patients with pediatric and congenital cardiac disease, with particular reference to the International Classification of Diseases (ICD) and its various iterations since 1900, emphasizing the current work on the 11th Revision, with collaboration between the World Health Organization (WHO) and the ISNPCHD. Finally, current and future efforts to merge such systems into the paperless environment of the electronic health or patient record on a global scale are briefly explored.

History of the Classification of Disease [13]

Although the gathering of population based information, or censuses, originated in Babylonian times (3800 BC), it was not until the early seventeenth century that the focus shifted away from taxation and military conscription, to causes of death, with the weekly publication from 1603 through the 1830s of the Bills of Mortality in London [14]. In 1662, these statistics were first systematically analyzed by John Graunt (1620–1674), when he estimated, for instance, the mortality of children before the age of 6 years to be 36 %, an estimate later proven to be quite accurate. In the eighteenth century François Bossier de Lacroix (1706–1767), better known as Sauvages, is credited with the first formal classification of diseases based on the methodology of the Swede Carl Linnaeus (1707–1778). This 1,763 system of classification developed by Sauvages contained 2,400 maladies divided into class, order and species. However, by the early nineteenth century, the classification most in use was that published by the Scottish

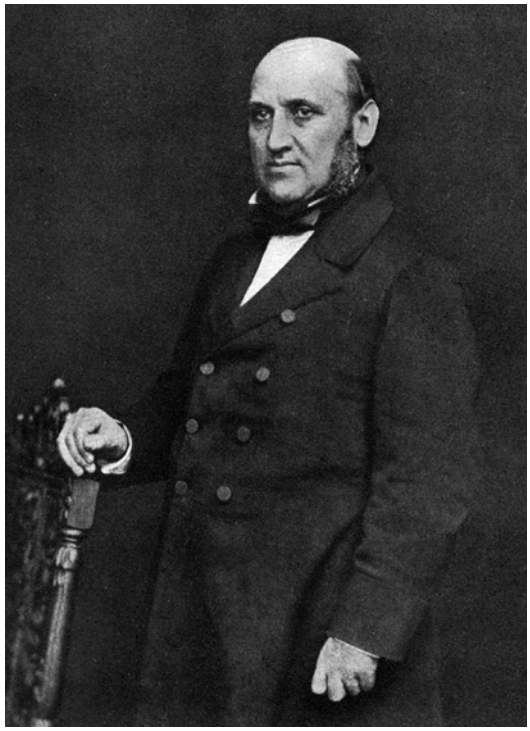

Fig. 5.1 Dr William Farr (1807–1883) in 1850: physician and first medical statistician for the General Register Office of England and Wales from 1839 to 1879. He pioneered the statistical analysis and development of a system of classification for causes of death and disease-related morbidity, emphasizing the need for a common international lexicon to allow the epidemiological study of diseases and thereby their causes, whilst incorporating medical advances (This figure was made available at http://en.wikipedia.org/wiki/File:William_Farr_2.jpg by Materialscientist/Public Domain)

physician William Cullen (1710–1790) in 1769, Synopsis Nosologiae Methodicae, with four main categories:

- "Pyrexiae", that is pyrexial, or febrile diseases, such as typhus fever
- "Neuroses", or nervous diseases, such as epilepsy
- "Cachexiae", that is cachexial illnesses, or diseases resulting from a bad habit of body, such as scurvy
- "Locales", or local diseases, such as cancer.

In 1837, the General Register Office of England and Wales was established, with William Farr (1807–1883), as its first medical statistician (Fig. 5.1). Farr revolutionized the principles of statistical classification and nomenclature, emphasizing the need for a common international lexicon to allow the epidemiological study of diseases and thereby their causes, whilst incorporating medical advances. His initial report, published in 1839, resonates through to the late twentieth century:

> The advantages of a uniform statistical nomenclature, however imperfect, are so obvious, that it is surprising that no attention has been paid to its enforcement in Bills of Mortality. Each disease has, in many instances, been denoted by three or four terms, and each term has been applied to as many different diseases: vague inconvenient names have been employed, or complications registered instead of primary diseases. The nomenclature is of as much importance in this department of inquiry, as weights and measures in the physical sciences, and should be settled without delay. [15]

William Farr later used these methods to help prove the water-born origin of cholera, the so-called "germ theory", providing evidence that countered the previously supported miasmatic, or "bad air theory" of disease, leading to the treatment of sewage. The eminence of Farr in the field was recognized at the first International Statistical Congress, held in Brussels in 1853, when he and Marc d'Espine, of Geneva, were asked to prepare an internationally applicable and uniform classification of causes of death. The approach of Marc d'Espine was based on the nature of disease, such as gouty, herpetic, or hematic, whilst the classification proposed by Farr divided into five groups:

- Epidemic diseases
- Constitutional, or general, diseases
- Local diseases arranged according to anatomical site
- Developmental diseases
- Diseases that are the direct result of violence.

It was the arrangement suggested by Farr which was dominant in the classification of 139 categories accepted by the Congress in 1855, and over the subsequent four revisions through to 1886, particularly the principle of classifying causes of death by anatomical site in distinction to generalized processes of disease. The failure of this classification to achieve wide international recognition, however, led the International Statistical Institute, which had developed from the former Congress, to commission in 1891 a

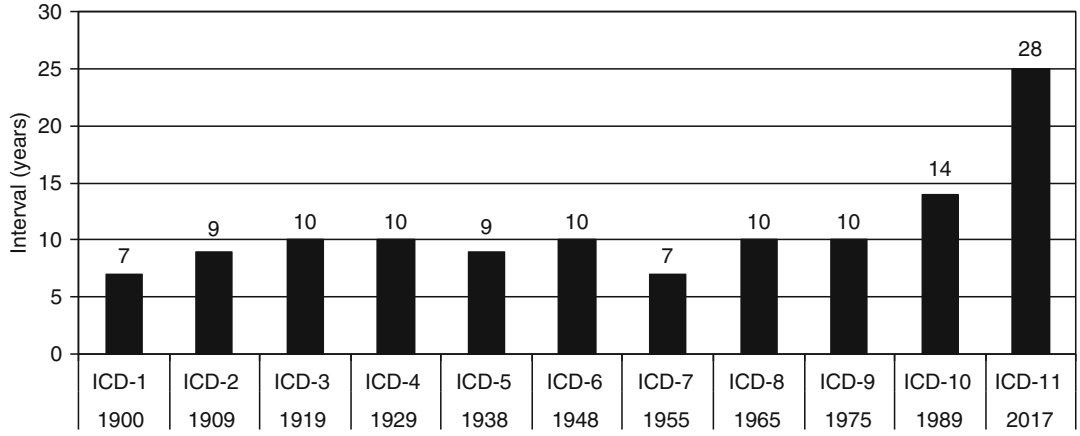

Fig. 5.2 This bar chart documents the time interval between each Revision of the International Classification of Diseases. Bertillon presented his [International] Classification of Causes of Death at the meeting of the International Statistical Institute in Chicago in 1893, where it was adopted and taken up by several cities and countries. In 1898, the American Public Health Association recommended its adoption in North America and that the classification be revised every 10 years. The First International Conference to revise the Bertillon Classification of Causes of Death was held in Paris in 1900. In 1909 non-fatal diseases were added (morbidity) and in 1948 the WHO took over its promotion and management as the 6th revision of the International Classification of Diseases, Injuries and Causes of Death (Adapted from WHO presentation made by Dr. Kentaro Sugano at the Ninth NWG meeting, Keio Plaza Hotel, Tokyo, JAPAN, July 2007)

committee chaired by Jacques Bertillon (1851–1922), Chief of Statistical Services of the City of Paris, to create what became the Bertillon [International] Classification of Causes of Death. This classification was based on the principles propounded by Farr and consisted of three levels of classification, with 44, 99, and 161 titles, respectively. Over the following decades, it was adopted by many countries in the Americas and Europe, with conferences for revision occurring roughly decennially to take note of medical advances (Fig. 5.2).

During the latter half of the nineteenth century, there was increasing recognition of the need for a parallel list of non-fatal diseases. Farr was again instrumental here, stating the need to:

> extend the same system of nomenclature to diseases which, though not fatal, cause disability in the population, and now figure in the tables of the diseases of armies, navies, hospitals, prisons, lunatic asylums, public institutions of every kind, and sickness societies, as well as in the census of countries like Ireland, where the diseases of all the people are enumerated

He submitted a list of these entities to the second meeting of the International Statistical Congress, held in 1856. At the fourth meeting, held in 1860, Florence Nightingale urged its adoption for the tabulation of hospital morbidity in her paper *Proposals for a uniform plan of hospital statistics* [16]. In 1900 and 1909, at the first two International Conferences to revise the Bertillon International Classification of Causes of Death, a parallel classification of diseases for use in statistics of sickness was adopted, but with limited scope. The Health Organization of the League of Nations, and the International Statistical Institute, recommended at the 4th and 5th revision conferences of the International List of Causes of Death (as it was then known), held in 1928 and 1938, respectively, that individual countries develop and promote their own systems of classification for morbidity statistics, using the Causes of Death structure as a template. It was not until the 6th revision conference, held in Paris in 1948, that a single comprehensive list was approved as the International Classification of Diseases, Injuries and Causes of Death; later

shortened to the International Classification of Diseases (ICD) by the time of the 7th revision in 1955. This was endorsed by the First World Health Assembly in the same year, along with rules for selecting a cause of death, and an agreement for international cooperation under the jurisdiction of the recently constituted WHO.

The 9th ICD Revision (ICD-9), in 1975, saw further expansion and structural change, with the addition of a fourth digit, as well as an optional fifth digit to facilitate greater detail where needed by subspecialties. The dagger and asterisk system was introduced to allow the linkage of anatomically specific areas of disease to more generalized diseases. The first International Classification of Procedures in Medicine was also published, in response to international pressure for a lead in this important area. By the time of the 10th revision (ICD-10), in 1993, the decennial revision schedule was abandoned (Fig. 5.2) due to the enormous amount of work involved with each revision and an alphanumeric coding structure was introduced. The promised revision of the listing of Procedures in Medicine never materialized, leaving countries to develop further their own systems of classification for interventions. Currently, the 11th ICD revision is well under way with planned dissemination following World Health assembly approval in 2017 (see below).

Up until the eleventh ICD iteration, congenital cardiac diseases have remained very poorly represented. Outdated terminology, inconsistent logic, and little appreciation of the inherent complexity of lesions, has meant that the ICD system of classification has never been robust enough for the purposes of relevant clinical governance. In the 9th and 10th revisions, there are a total of only 29 and 73 individual codes, respectively, for congenital cardiovascular lesions (Fig. 5.2). Despite these limitations, both versions remain in use, mostly for the purpose of 'billing', returns to central governments, and crude epidemiological surveillance. A comprehensive, clinically acceptable system for the naming and coding of congenital cardiac disease, relevant acquired cardiac disease, and, most importantly, procedures to treat congenital and relevant acquired cardiac lesions, was needed.

Pediatric and Congenital Cardiac Nomenclature and the International Pediatric and Congenital Cardiac Code

Although there were many descriptive publications of individuals with various congenital cardiac malformations in the nineteenth and early twentieth centuries, it was the publication in 1936 of the *Atlas of Congenital Cardiac Disease*, by the distinguished Canadian physician Maude Abbott, that saw the first systematic classification of congenital cardiac lesions [17]. Over the following decades, several centers developed more inclusive systems of classification for both the diagnosis and treatment of congenital cardiac disease. It was not until the 1980s that advances in the hardware and software underpinning information technology made it feasible to have a uniform and internationally acceptable system of nomenclature, with facilitation of entry of data and exchange of information.

In Europe, this era was marked by the publication in 1985 of the Brompton Hospital Diagnostic Code, with 507 items classified using an associated six digit code [18]. This diagnostic system was expanded to 1,717 terms in the Netherlands in the late 1980s, including sections on acquired cardiac disease, arrhythmias, relevant non-cardiac anomalies, and, most importantly, surgical and transcatheter procedures. From 1989 to 1994, further enhancements occurred in all areas, with the introduction of terms for postprocedural complications and qualifiers, leading to a single hierarchical tree with over 4,300 terms, each with its own six digit code [19]. These terms were incorporated into the United Kingdom panmedical nomenclature and coding initiative within the National Health Service, becoming Clinical Terms version 3.1, or the Read Codes, which later formed a fundamental part of the Systematized Nomenclature of Medicine Clinical Terms (SNOMED-CT), described below. This 1994 version was used by The European Congenital Heart Surgeons Foundation to audit surgical outcomes across much of Europe. Finally, in 1998, this system of nomenclature was

adopted by the Association for European Paediatric Cardiology (AEPC) as the standard system for databases across Europe, and was titled the European Paediatric Cardiac Code [20]. The publication of this system included rule-based crossmapping to the 9th and 10th revisions of the ICD [19, 20].

In the mid to late 1990s, a second major and parallel initiative emerged to list and classify pediatric and congenital cardiac diagnoses and procedures. Two large multi-institutional surgical database projects were in process:

- The Congenital Heart Surgery Database of the Society of Thoracic Surgeons (STS) in North America, and
- The European Congenital Heart Defects Database of the European Congenital Heart Surgeons Foundation, which in 2003 was renamed the European Congenital Heart Surgeons Association (ECHSA). In the mid 1990s, this database received data from 18 countries.

Both project teams identified a need for an international structure that would standardize nomenclature and strategies for reporting. There was, for example, disharmony amongst the many centers and countries on the terms used to describe identical congenital cardiac lesions, such as subtypes of ventricular septal defects. This led the STS, the ECHSA, and the European Association for Cardio-Thoracic Surgery (EACTS) to set up the International Congenital Heart Surgery Nomenclature and Database Project in 1998 [21]. Over the next 2 years, a series of meetings between a core group of experts in congenital cardiac surgery and pediatric cardiology met to create a standardized inclusive hierarchical nomenclature, with a generous use of synonyms, based on consensus, scientific principles and popular usage [22].

In early 2000, both of these two systems of nomenclature were published [20, 22]

- The nomenclature of the International Congenital Heart Surgery Nomenclature and Database Project of the STS, the ECHSA, and the EACTS
- The European Paediatric Cardiac Code of the AEPC

Both systems of nomenclature included a comprehensive Long List, with thousands of terms, and a Short List designed to be used as part of a Minimum Dataset for audit and research purposes, with up to 650 terms [20, 22]. The comprehensive datasets include all the imagined variables, in a hierarchical structure, and are detailed enough to enable analyses that incorporate stratification of risk. The minimum dataset includes sufficient data points to enable easy and mandatory sharing of interinstitutional data for basic analysis of mortality, governance, and interpretation of trends. Both Long Lists map fully to their respective Short Lists. The nearly simultaneous publication of these two complementary systems of nomenclature led to the problematic situation of having two lexicons that were to be widely adopted, with the potential risks of invalidating multicentric projects due to confusion between the two systems and duplicate or inaccurate entries within institutions.

A meeting was convened, therefore, between representatives from the AEPC, the STS, and the EACTS on October 6, 2000, in Frankfurt, Germany. It was agreed to establish The International Nomenclature Committee for Pediatric and Congenital Heart Disease (which later evolved into the ISNPCHD), including representatives of the three societies, as well as representatives from the remaining continents of the world (Africa, Australia, Asia, and South America), to work in partnership and produce a reconciliatory bidirectional map between the two systems. Fortuitously, the International Congenital Heart Surgery Nomenclature and Database Project did not feature a numerical code, and it was therefore resolved to use the six digit numerical code derived from the European Paediatric Cardiac Code as the backbone for mapping the two systems. The feasibility of this project was established by the creation of a rule-based bidirectional crossmap between the two Short Lists [23, 24]. This work was then presented and endorsed at the Third World Congress of Pediatric Cardiology and Cardiac Surgery in Toronto, Canada on May 27, 2001, during the First International Summit on Nomenclature for Congenital Heart Disease, which was attended by

representatives from at least ten Societies and five continents [25]. This bidirectional crossmap between the two Short Lists therefore established precedent to achieve the main goal of mapping the two comprehensive lists to each other to create the International Pediatric and Congenital Cardiac Code (IPCCC), for subsequent presentation at the Fourth World Congress in 2005.

The working component of this International Nomenclature Committee has been the International Working Group for Mapping and Coding of Nomenclatures for Pediatric and Congenital Heart Disease, with 12 members, better known as the Nomenclature Working Group (NWG). On September 19, 2005, the Nomenclature Working Group was able to report to the Second International Summit on Nomenclature for Congenital Heart Disease at the Fourth World Congress in Buenos Aires, Argentina, that they had met seven times, over a total period of 33 days, and had succeeded in crossmapping the majority of congenital cardiac lesions [26]. The IPCCC at that point consisted of 7,623 items, each with a six digit code, in two dominant versions:

- The IPCCC derived from the European Paediatric Cardiac Code of the AEPC;
- The IPCCC derived from the International Congenital Heart Surgery Nomenclature and Database Project of the STS, the ECHSA, and the EACTS.

Each unique six-digit code in the IPCCC corresponds to a single entity, whether it be a morphological phenotype, procedure, symptom, or genetic syndrome, with the mapped terms in the two versions being synonymous to each other. Additional systems of nomenclature, for pediatric cardiology and cardiac surgery, which are mapped to the common spine, include the Boston-based Fyler codes, and the Canadian nomenclature system. There is also mapping to ICD-9 and ICD-10, usually in a many to one fashion, given the limitations of these ICD revisions. Subsequent meetings of the NWG, in 2006 and 2007, expanded the IPCCC further to cover fetal cardiology, arrhythmias, congenital coronary arterial anomalies, echocardiography, and interventional cardiology procedures, with the help of several invited experts. A separate parallel process also involved members of the NWG, in developing a nomenclature and system of classification for complications during and following interventions for patients with pediatric and congenital cardiac disease, as supported by The MultiSocietal Database Committee for Pediatric and Congenital Heart Disease. This was published in 2008 as a Supplement in Cardiology in the Young [27]. During this process, the IPCCC expanded its list from 1,422 codes related to complications to a listing of over 2,500 items, each with its own numerical six digit code and consensus derived definition.

In early 2013, there were 12,168 items in the IPCCC version derived from the European Paediatric Cardiac Code, and 17,176 in the IPCCC version derived from the International Congenital Heart Surgery and Nomenclature Database Project, with an additional hundreds of qualifiers, some specific such as anatomical sites and some generic such as gradings of severity. The IPCCC is available for download without cost from the internet at http://www.IPCCC.net.

The Nomenclature Working Group has also published review articles which provide a unified and comprehensive classification, with definitions, for several complex congenital cardiac malformations, along with a complete listing of the relevant codes and terms in both versions of the IPCCC: the functionally univentricular heart [28], hypoplastic left heart syndrome [29], discordant atrioventricular connections [30] and cardiac structures in the setting of heterotaxy [31].

The IPCCC, in its parallel systems of nomenclature, is made up of eight fundamental components:

- Diagnoses related to congenital cardiac malformations
- Diagnoses related to acquired cardiac disease, primarily but not exclusively focusing, on those occurring at in patients under 18 years of age. These diagnoses related to acquired cardiac disease include entities such as cardiomyopathies, endocarditis, rheumatic heart disease, and acquired abnormalities related to congenital cardiac malformations, such as acquired right pulmonary arterial stenosis.

- Relevant diagnoses related to genetic syndromes, chromosomal abnormalities, non-cardiac congenital malformations, and acquired diseases, such as Marfan syndrome, trisomy 21, intestinal malrotation, and kyphoscoliosis.
- Signs, symptoms, and cardiac related diagnostic investigations
- Preprocedural factors
- Procedures: surgical, transcatheter interventions, and hybrid procedures
- Arrhythmias and inherited cardiac conditions, related procedures and their complications
- Intraoperative and postoperative complications

Further Developments Related to the IPCCC

The crossmapping of the Short Lists of the International Congenital Heart Surgery Nomenclature and Database Project and the European Paediatric Cardiac Code [24], which are used primarily for analyses of multi-institutional and international outcomes following operations and procedures for patients with congenitally malformed hearts, and which are derived directly from their respective Long Lists, enables the huge number of over 500,000 patients registered in their respective databases to be used together. Work has shifted in recent years to focus on risk adjusted outcomes in order to compare the outcomes of similar groups of patients that have been stratified into categories of increasing complexity and hence higher operative risk. Initial efforts in this regard were based on an expert panel's subjective assessment of the risks associated with individual operations, namely **R**isk **A**djustment in **C**ongenital **H**eart **S**urgery-1 methodology (RACHS-1 method) and the Aristotle Complexity Score (ABC Score) [32, 33]. More recently two empirical systems have emerged using actual patient outcomes as the basis for adjusting relative risks: the **STS**-E**A**C**T**S Congenital Heart Surgery Mortality Categories (STS-EACTS Mortality Categories) (STAT Mortality Categories), derived from the STS and EACTS databases [34], and the Partial Risk Adjustment in Surgery (PRAiS) system derived from the National Institute of Cardiovascular Outcomes Research (NICOR) Congenital Audit in the United Kingdom (previously known as the Central Cardiac Audit Database), which stratifies risk based on operation type, comorbidities, and diagnoses [35]. Both systems depend upon the IPCCC to ensure a common lexicon of terms between institutions submitting data and both perform better than the systems based on the subjective assessment of risk.

The structure and content of the Short Lists remain the purview of the Societies and organizations that created them. It is, of course, possible to shorten further, or create Short Lists specific to a subspecialty, or minimum datasets to suit individual projects and research aims, provided that those using the Long List as the data entry mechanism, focus on similar areas, and ensure no orphan terms are produced during the process of electronic conversion of the terms in the Long List to specific terms in the Short List of interest, based on the resultant crossmap.

In January, 2005, the International Nomenclature Committee was incorporated in Canada as the International Society for Nomenclature of Paediatric and Congenital Heart Disease (ISNPCHD) [26]. On July 9, 2007, during its third annual meeting, the ISNPCHD created two new Working Groups, so that the Society now has the following three committees:

- The International Working Group for Mapping and Coding of Nomenclatures for Pediatric and Congenital Heart Disease, also known as the Nomenclature Working Group (NWG). This Group continues to maintain, develop, expand, update, and preserve the IPCCC. It also provides ready access to the IPCCC for the global pediatric and congenital cardiology and cardiac surgery communities, related disciplines, the healthcare industry, and governmental agencies, both electronically and in published form. Table 5.1 lists the current membership of the NWG.
- The International Working Group for Defining the Nomenclatures for Pediatric and Congenital Heart Disease, also known as the Definitions Working Group (DWG). This Group has been engaged in writing definitions

Table 5.1 Membership of The International Working Group for Mapping and Coding of Nomenclatures for Pediatric and Congenital Heart Disease, also known as the Nomenclature Working Group (NWG)

1. Marie J. Beland, M.D. (Co-Chair)
2. Rodney C.G. Franklin, M.D. (Co-Chair)
3. Jeffrey P. Jacobs, M.D. (Co-Chair)
4. Vera D. Aiello, M.D.
5. Steven D. Colan, M.D.
6. J. William Gaynor, M.D.
7. Otto N. Krogmann, M.D.
8. Hiromi Kurosawa, M.D.
9. Bohdan Maruszewski, M.D.
10. Giovanni Stellin, M.D.
11. Christo I. Tchervenkov, M.D.
12. Paul M. Weinberg, M.D.

for the terms in the IPCCC, building on the previously published definitions from the Nomenclature Working Group [28–31] and more recently focusing on the 11th revision of the ICD (see below).

- The International Working Group for Archiving and Cataloguing the Images and Videos of the Nomenclatures for Pediatric and Congenital Heart Disease, also known as the Archiving Working Group (AWG). This Group has been engaged in linking images and videos to the IPCCC, with monthly teleconferences to discuss, label, and code individual images [36] (http://www.IPCCC-awg.NET). The images and videos are acquired from cardiac morphologic specimens and imaging modalities such as echocardiography, angiography, computerized axial tomography and magnetic resonance imaging, as well as intraoperative images and videos.

Crossmapping Issues and the Development of Crossmapping Rules

The process of crossmapping of systems of nomenclature has allowed the ISNPCHD to clarify several issues concerning nomenclature and databases that had been difficult to resolve [37]:

- Generic terms in the lists, that is terms ending in *NOS* (*not otherwise specified*) or (*unspecified*)
- Nonspecific terminology meant to allow further description in the nomenclature lists, that is terms ending in *Other* or (*DESCRIBE*)
- The meaning of the words *right* and *left* in the nomenclature lists, or lateralization
- Structural differences between nomenclature systems

Optimal performance from systems of nomenclature can be expected in an environment where the database, or system for entry of data, has certain standard regulations and requirements. The person entering the data, the "nomenclature coder", must be forced to choose from the choices in the list of nomenclatures, and not be allowed to type free text directly into the fields for "Diagnoses" and "Procedures". A separate "Comments" field will then allow further free text to add additional description to any individual diagnosis or procedure that has been chosen. The crossmapping, and the systems themselves, will work effectively in environments that follow this basic rule or principle. This fundamental principle also leads to logical solutions for the first two issues above.

All terms in the nomenclature lists theoretically end in *NOS* or (*unspecified*), in that one can always create further subdivisions for virtually any diagnosis or procedure. Therefore, the generic term on its own is self-explanatory, without the need for other clarifying nomenclature, such as *NOS* or (*unspecified*) being affixed. These suffixes are consequently not necessary.

Terms ending in *Other* are problematic for several reasons. The appendage *Other* could confer different meanings to a term depending on the list in which it is included, and any entry containing the appended term *Other* may change meaning over time as additional terms are added to the parent list from which the term is derived. In some systems of nomenclature the intent of the terms with the appendage *Other* may be to allow for the further description of related terms or choices not appearing in the list, similar to the use of the suffix (*DESCRIBE*). The use of the suffix (*DESCRIBE*) is preferable to the suffix *Other* because the suffix

(*DESCRIBE*) circumvents the above shortcomings and implications inherent in the word *Other*. It is apparent, however, it is actually completely unnecessary to specify that a family of terms can have further items added, when the database environment follows the rule discussed above; namely, that no free text is permitted in the fields for "Diagnoses" or "Procedures", whilst a separate "Comments" field exists to allow further description of any chosen item. Thus, theoretically, all terms in the lists are suffixed with (*DESCRIBE*), and the coder has the option to add further detail to any selected term. As a consequence, generic family terms ending in (*DESCRIBE*) or *Other* become redundant.

When discussing cardiac chambers, such as atriums and ventricles, and spatial relationships, the words *left* and *right* can be confusing. Rules were therefore created to provide consistency and accuracy of descriptive terms of anatomical phenotypes. For cardiac chambers, unless otherwise stated, *left* refers to morphologically left, and *right* refers to morphologically right. Thus, left ventricle means the morphologically left ventricle, left atrium refers to the morphologically left atrium, and right atrial appendage refers to the morphologically right atrial appendage, and so on. When discussing cardiac chambers, the words *left* and *right* do not imply sidedness or position. If one wishes to describe the position or sidedness of a cardiac chamber, it is necessary to use terms such as *left-sided ventricle*. The term left ventricle, therefore, merely means the morphologically left ventricle, and does not mean or imply left-sidedness or right-sidedness. Similarly, it does not imply connections to the right or left atrium, or the pulmonary or systemic circulations. In contrast, when describing the superior caval vein, and using the prefix *left* or *right*, it is the spatial position that is being alluded to, rather than any other connection or phenotypic variation that may exist.

A separate issue is the fundamental structure of systems for nomenclature. Some systems of nomenclature use a *molecular* structure, with an incrementally more complex diagnostic or procedural combination of terms. Each combination is considered a single diagnostic unit, which theoretically could have its own numerical code. In contrast, other systems of nomenclature use an *atomic* structure, so that a complex diagnosis would have separate numerical codes for each element. This means that a map between an atomic system and a molecular system would have a series of atomic codes being equivalent to one molecular code. Thus, the combination term from the molecular nomenclature "*TGA*, *VSD – LVOTO*" is equivalent to the three entries in an atomic system: *Discordant VA connections* (01.05.01), *VSD* (07.10.00), and *LV outflow tract obstruction* (07.09.01). Exceptions to this configuration are a few common combinations of lesions that are so routinely associated with each other that they have been grouped as one discrete diagnosis or procedure in both systems. Examples are: *Pulmonary atresia + VSD* (*including Fallot type*) (01.01.06), or *Arterial and atrial switch procedures* (*double switch*) (12.29.25).

The Electronic Health Record and the Systematized Nomenclature of Medicine Clinical Terms (SNOMED-CT)

Currently, the use of the IPCCC has largely been limited to individuals working in the field of pediatric cardiology and pediatric cardiac surgery. There is a need to expand this usage to other areas of global healthcare, where the individual with a congenitally malformed heart may be referred for care, or have an impact, such as other medical subspecialties, hospital administrative and insurance systems, and the WHO. A key element in achieving this has been the development of the electronic health record, facilitated by the continued advancements in computers and information technology [38]. Healthcare providers are now demanding digitization of the healthcare record and clinical support systems, so as to move towards a paperless environment. This advance involves creating a standardized system for obtaining accurate and detailed clinical information on the history of the patient, as well as diagnosis, and treatment, using a reliable and easily validated methodology. The hope is that the savings made from this

patient-centric system, free from duplication of data entry, would free up resources for comparative studies across units and nations of clinical and cost-related outcomes. This achievement would allow risk-adjusted benchmarking, and identification of best practice, as well as the generation of effective guidelines and tools to support decision-making. The prerequisite for this accomplishment is an underlying, all encompassing, common nomenclature and system of coding for healthcare, with clinician-led and validated entry of data, the qualities of which should include the following specifications:

- An 'atom' based, clinically sensitive structure, so that each numerical code corresponds to the lowest denominator concept, based on a multidisciplinary clinical knowledgebase. This atomically oriented system would not preclude higher level more complex concepts which are in common clinical use, such as 'hypoplastic left heart syndrome'
- The ability to code relevant qualities of severity and complexity, for stratification of risk
- Standardization of underlying terminologies and hierarchical classifications across multiple specialties
- An aim for one preferred representation of a concept or phenotype, but with explicit integration of synonyms and commonly used abbreviations. The user would then have a choice of synonymous terms, enabling entry of data that easily encompasses different cultures of practice and different medical specialties. This use of synonyms should abrogate multiple, same-meaning, redundant codes
- The ability to access terms through multiple intuitive hierarchies, such as concepts for 'ventricular septal defect' via pathways based on either septal defect, tetralogy of Fallot, or functionally univentricular heart
- Routine clinically sensitive updates for new procedures and scientific advances. This specification is the "responsiveness" of the system of nomenclature
- The ability to move a concept to a new hierarchy, whilst maintaining its unique code, in response to scientific or clinical advances, such as genetics or transcatheter procedures
- The ability to account for diagnostic uncertainty or negative findings, using an additional attribute, such as 'suspected', 'uncertain' or 'ruled out'
- A definition for each term provided by an overseeing expert authority. We suggest that the DWG should oversee definitions for codes related to congenital cardiology and acquired pediatric cardiac disease
- The ability to deal with obsolete or scientifically inaccurate concepts by reassignment to the correct term and using an 'obsolete' label for the term itself, whilst retaining historical data
- Multilingual translations

These specifications are not available in current international systems, such as ICD-9 and ICD-10. As a consequence, many non-integrated self-made, or industry-created, solutions exist, which are often expensive and of variable quality and integrity.

In 1974, the College of the American Pathologists created the Systematized Nomenclature of Medicine from their more restrictive listing of 1965, the Systematized Nomenclature of Pathology, generating an electronic format in 1977. After considerable expansion over the next 25 years, with the endorsement of the National Library of Health and American National Standards Institute, it combined with the Clinical Terms version 3 project based in the United Kingdom to become, in 2002, the Systematized Nomenclature of Medicine Clinical Terms, or 'SNOMED-CT' [39]. During this time, and subsequently, it has achieved wide acceptance as an effective tool to classify diseases, and is being promoted as the optimal product for the electronic health record, having most of the above specifications, although notably without definitions (and multilingual translations remain an ambition). In a précis of their own words: it is a dynamic, scientifically validated clinical reference terminology that makes health care knowledge more usable and accessible by providing a common language that enables a consistent way of capturing, sharing, and aggregating health data across all specialties and sites of care. There are over 365,000 concepts, 730,000 attributes or

descriptors, and one million relational connections, inclusive of diagnoses, procedures, complications and relevant qualifiers. In 2007, nine countries grouped together to purchase the product as the not-for-profit International Health Terminology Standards Development Organization (IHTSDO),[1] whilst having the College of American Pathologists continuing in a managerial role. It is in use in over 50 countries worldwide, with 19 co-owner countries. The aim is to have a global, validated, and stable system of nomenclature with shared ownership, transparent processes of management, and a secure governance structure, as well as financial sustainability. Arrangement for licensing with vendors from industry is to be simple, clear, and inclusive, whilst the user license is free to member states and 40 nations classified by the World Bank as "low income countries".

With respect to congenital and pediatric cardiology and surgery, over 4,000 terms were integrated into SNOMED-CT in 2002 when amalgamated with the UK Clinical Terms system, which itself had taken these terms into its structure in 1994, as described above. These 4,000 terms are also a core part of the IPCCC, with often exact or near-exact matching to the version derived from the European Paediatric Cardiac Code. Unfortunately, many categories also exist with redundant and obsolete terms, as well as areas of non-intuitive hierarchy, as this section of the lexicon has not had input from relevant experts since 1994. One of the remits of the IHTSDO is to promote the development of subsets for individual medical disciplines that already have a system of nomenclature in active use. This process is well underway after an agreement between the IHTSDO and ISNPCHD to create a congenital cardiology subset by fully incorporating the IPCCC into the SNOMED-CT, whilst 'cleaning up' the latter to a clinically sound product, and mapping to the six digit numerical code. Over 2,500 terms have to date been incorporated and mapped.

Discussions with the cardiovascular specialist healthcare industry have been ongoing and led to the successful incorporation of the IPCCC into standalone database software, as well as echocardiographic and Catheterisation based software, either independently or as a specialist subset of SNOMED-CT.

[1]Australia, Canada, Denmark, Lithuania, New Zealand, Netherlands, Sweden, United Kingdom, United States.

The 11th Revision of the International Classification of Diseases (ICD-11)

Discussions began in 2007, in Tokyo Japan, between ISNPCHD and representatives of the steering group responsible for the creation of ICD-11, administered by the WHO, and now scheduled for launch in 2017. The ICD-11 mission is 'To produce an international disease classification that is ready for electronic health records that will serve as a standard for scientific comparability and communication [40].' It is planned to incorporate most of the above prerequisites, including textual definitions, and will have logical linkages to other standard terminologies such as SNOMED-CT. In addition, the system will be explicitly stratified to cater for different users such as primary care and public health, so called the linearizations. For the first time the revision process has moved away from reliance on large meetings of national delegations of health statisticians with manual archiving (curation) and wherein those who voiced their opinion strongest would dominate ('decibel' diplomacy), to being dependent upon international expert clinicians with digital curation and incorporating wide peer review. The work is divided into content specific Topic Advisory Groups (TAGs) and related Working Groups, as well as cross sectional TAGs to ensure structural uniformity (Fig. 5.3). The authoring process utilizes a web based platform for its entire content, the ICD Collaborative Authoring Tool (iCAT), which allows online global peer review and comment, as well as editing by designated Managing Editors [40]. ICD-11 utilizes an ontological content model that was derived from computer science. Ontology in this context can be defined as the explicit, operational description of the concepts within a domain; in other words, its qualities, properties and attributes. In ICD-11 the evidence based attributes of each individual disease are in the process of being delineated, including a textual definition, synonyms, inclu-

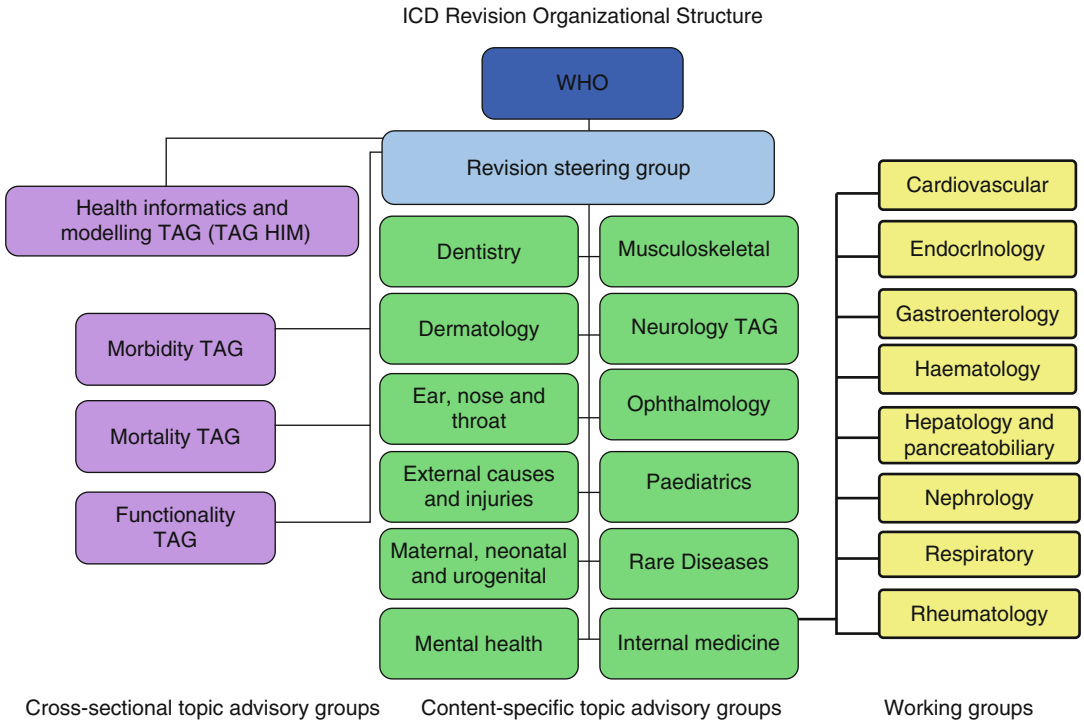

Fig. 5.3 The workflow for the collaborative authoring of ICD is built around the ICD organizational structure. Branches of ICD classification are assigned to at least one Topic Advisory Group (*TAG*), led by a Managing Editor. Congenital cardiology falls under the remit of the Cardiovascular Working Group of the Internal Medicine TAG with collaborative input and ratification by the Pediatric and Rare Disease TAGs. The members of each Working Group and TAG have the task of entering the content of the diseases in their domain of expertise, such as textual definition, synonyms, clinical description, diagnostic criteria, causal mechanisms, and so on

sions, exclusions, diagnostic criteria, causal mechanisms, functional impact, and so on. This ontological content model will result in a common vocabulary with robust data standards, enabling a shared understanding and exchange of information (data) between individuals and institutions, permitting interoperability and like-for-like comparisons across all fields of medicine. Congenital and pediatric cardiac disease are under the auspices of the Cardiovascular Working Group of the Internal Medicine Topic Advisory Group (TAG), in liaison with the Rare Diseases and Pediatric TAGs. Recent NWG and DWG annual meetings have been devoted to creating the ICD-11 content for the Cardiovascular Working Group; and consequently, ISNPCHD has submitted a short list of over 300 congenital cardiology terms, mostly with definitions and synonyms and based upon the IPCCC short lists described above. This draft version is currently available for comment and endorsement within the ICD-11 Beta Draft (http://apps.who.int/classifications/icd11/browse/f/en). Each item has IPCCC code(s) attached, and there will be a corresponding map from the respective diagnostic sections of the IPCCC Long List to ICD-11 to allow a further expansion of international and national comparisons of outcomes and quality assurance for those with congenital cardiac malformations.

Differences Between Clinical and Administrative Nomenclature

Several studies have examined the relative utility of clinical and administrative nomenclature for the evaluation of quality of care for patients undergoing treatment for pediatric and congenital cardiac disease. Evidence from four recent investigations suggests that the validity of coding of lesions seen in the congenitally malformed

heart via ICD-9 as used currently in administrative databases in the United States of America is poor [41–44]. First, in a series of 373 infants with congenital cardiac defects at Children's Hospital of Wisconsin, investigators reported that only 52 % of the cardiac diagnoses in the medical records had a corresponding code from the ICD-9 in the hospital discharge database [41]. Second, the Hennepin County Medical Center discharge database in Minnesota identified all infants born during 2001 with a code for congenital cardiac disease using ICD-9. A review of these 66 medical records by physicians was able to confirm only 41 % of the codes contained in the administrative database from ICD-9 [42]. Third, the Metropolitan Atlanta Congenital Defect Program of the Birth Defect Branch of the Centers for Disease Control and Prevention of the United States government carried out surveillance of infants and fetuses with cardiac defects delivered to mothers residing in Atlanta during the years 1988 through 2003 [43]. These records were reviewed and classified using both administrative coding and the clinical nomenclature used in the Society of Thoracic Surgeons Congenital Heart Surgery Database. This study concluded that analyses based on the codes available in ICD-9 are likely to "have substantial misclassification" of congenital cardiac disease. Fourth, a study was performed using linked patient data (2004–2010) from the Society of Thoracic Surgeons Congenital Heart Surgery (STS-CHS) Database (clinical registry) and the Pediatric Health Information Systems (PHIS) database (administrative database) from hospitals participating in both in order to evaluate differential coding/classification of operations between datasets and subsequent impact on outcomes assessment [44]. The cohort included 59,820 patients from 33 centers. There was a greater than 10 % difference in the number of cases identified between data sources for half of the benchmark operations. The negative predictive value (NPV) of the administrative (versus clinical) data was high (98.8–99.9 %); the positive predictive value (PPV) was lower (56.7–88.0 %). Overall agreement between data sources in RACHS-1 category assignment was 68.4 %. These differences translated into significant differences in outcomes assessment, ranging from an underestimation of mortality associated with truncus arteriosus repair by 25.7 % in the administrative versus clinical data (7.01 % versus 9.43 %; $p = 0.001$) to an overestimation of mortality associated with ventricular septal defect (VSD) repair by 31.0 % (0.78 % versus 0.60 %; $p = 0.1$). For the RACHS-1 categories, these ranged from an underestimation of category 5 mortality by 40.5 % to an overestimation of category 2 mortality by 12.1 %; these differences were not statistically significant. This study demonstrates differences in case ascertainment between administrative and clinical registry data for children undergoing cardiac operations, which translated into important differences in outcomes assessment.

Several potential reasons can explain the poor diagnostic accuracy of administrative databases and codes from ICD-9:

- Accidental miscoding
- Coding performed by medical records clerks who have never seen the actual patient
- Contradictory or poorly described information in the medical record
- Lack of diagnostic specificity for congenital cardiac disease in the codes of ICD-9
- Inadequately trained medical coders.

Although one might anticipate some improvement in diagnostic specificity with the planned adoption of ICD-10 by the United States, it is likely to still be far short from that currently achieved with clinical registries using IPCCC derived Short Lists. (ICD-9 has only 29 congenital cardiac codes and ICD-10 has 73 possible congenital cardiac terms.) It will not be until there is implementation of the pediatric and congenital cardiac components of ICD-11, as developed by the ISNPCHD, that harmonization of clinical and administrative nomenclature will be achieved with the resolution, therefore, of many of these challenging issues.

Conclusions

The IPCCC was created by specialists in the field to name and classify pediatric and congenital cardiac disease and its treatment. It is a comprehensive code that can be freely down-

loaded from the internet (http://www.IPCCC.net) and is already in use worldwide, particularly in its Short List formats for international comparisons of risk adjusted outcomes. This latter work is being used to compare performance between units, and even individual clinicians, to create strategies for stratification of risk, and to improve healthcare for the individual patient. Such comparisons have already been shown to be culturally reassuring when no statistically different outcomes can be demonstrated across a nation [45]. We anticipate that future cooperative multi-institutional studies will enable the optimization of the quality and effectiveness of healthcare for our patients with congenital cardiac malformations, whilst influencing the allocation of increasingly limited resources.

The collaboration with the ISNPCHD, the WHO, the IHTSDO and the healthcare industry, will lead to further enhancement of the IPCCC, and to its more universal use. Future work of the ISNPCHD, and its three working groups, should produce in the next few years a unique, multifaceted lexicon of terms related to congenital cardiac disease and acquired pediatric cardiac disease, for clinical, governance, educational, research, and administrative purposes. This system will be replete with comprehensive definitions, and the ability to visualize the respective lesions, along with their modes of therapy, across a full array of imaging platforms. All will be available at the click of a mouse, and free throughout the world.

References

1. Smith R. All changed, changed utterly. Brit Med J. 1998;316:1917–8.
2. Learning from Bristol: the report of the public inquiry into children's heart surgery at the Bristol Royal Infirmary 1984–1995. http://webarchive.nationalarchives.gov.uk/+/www.dh.gov.uk/en/Publicationsandstatistics/Publications/PublicationsPolicyAndGuidance/DH_4005620. Accessed 14 Oct 2013.
3. Wright J. Putting a surgeon under: a personal story of hospital politics. http://www.uow.edu.au/arts/sts/bmartin/dissent/documents/Wright. Accessed 14 Oct 2013.
4. The report of the Manitoba pediatric cardiac surgery inquest: an inquest into twelve deaths at the Winnipeg Health Sciences Centre in 1994. http://www.pediatriccardiacinquest.mb.ca/. Accessed 14 Oct 2013.
5. Sherry A. Children's hospital cardiology chief told to resign. Denver Post. Article Published: March 1, 2001. http://www.denverpost.com/news/news0301b.htm. Accessed 21 Mar 2001.
6. Sherry A. Hospitals shield mortality rates. Denver Post. Article Published: March 2, 2001. http://www.denverpost.com/news/news0302d.htm. Accessed 21 Mar 2001.
7. The Denver Post editorial board. At the heart of the problem. Denver Post. Article Published: March 2, 2001. http://www.denverpost.com/opinion/edits0302c.htm. Accessed 21 Mar 2001.
8. Hernandez J. Other options. Denver Post. Article Published: March 3, 2001. http://www.denverpost.com/opinion/lett0311.htm. Accessed 21 Mar 2001.
9. Johnson L. Baby's death at children's turns parents to their faith. Denver Post. Article Published: March 3, 2001. http://www.denverpost.com/opinion/lett0311.htm. Accessed 21 Mar 2001.
10. White S. Kids' best interests: Re: "Children's hospital cardiology chief told to resign," March 1. Denver Post. Article Published: March 3, 2001. ttp://www.denverpost.com/opinion/lett0311.htm. Accessed 21 Mar 2001.
11. Weinberg S. Rare look inside a surgeon's sanctum. Denver Post. Article Published: Sunday, April 20, 2003. http://www.denverpost.com/Stories/0%2C1413%2C36~28~1333663%2C00.html. Accessed 22 Oct 2004.
12. Hudson W, Cohen E. After CNN investigates babies' deaths, hospital releases mortality data. Available at: http://edition.cnn.com/2013/08/12/health/kentucky-children-update/index.html, Accessed 12 Aug 2013.
13. World Health Organization. History of the development of the ICD. http://www.who.int/classifications/icd/en/HistoryOfICD.pdf. Accessed 14 Oct 2013.
14. Greenwood M. Medical statistics from Graunt to Farr. Cambridge: Cambridge University Press; 1948.
15. Farr W. First annual report. London: Registrar General of England and Wales; 1839. p. 99.
16. Nightingale F. Proposals for a uniform plan of hospital statistics. London: Fourth International Statistical Congress; 1860.
17. Abbott ME. Atlas of congenital cardiac disease. New York: The American Heart Association; 1936.
18. Miller GAH, Anderson RH, Rigby ML. The diagnosis of congenital heart disease; incorporating the Brompton hospital diagnostic code. Tunbridge Wells: Castle House; 1985. p. 110–20.
19. Franklin RCG, Anderson RH, Daniëls O, et al. The European paediatric cardiac code. Cardiol Young. 1999;9:633–57.
20. Association for European Paediatric Cardiology. The European paediatric cardiac code. Cardiol Young. 2000;10 Suppl 1:1–146.
21. Mavroudis C, Jacobs JP. Congenital heart surgery nomenclature and database project: overview and minimum dataset. Ann Thorac Surg. 2000;69:S2–17.

22. Mavroudis C, Jacobs JP, editors. Congenital Heart Surgery Nomenclature and Database Project. Ann Thorac Surg. 2000;69 Suppl:S1–S372.
23. Béland M, Jacobs JP, Tchervenkov CI, Franklin RCG. The international nomenclature project for paediatric and congenital heart disease: report from the executive of the International Working Group for Mapping and Coding of Nomenclatures for Paediatric and Congenital Heart Disease. Cardiol Young. 2002;12: 425–30.
24. Franklin RCG, Jacobs JP, Tchervenkov CI, Béland MJ. The international nomenclature project for congenital heart disease: bidirectional crossmap of the short lists of the European paediatric cardiac code and the international congenital heart surgery nomenclature database project. Card Young. 2002;12:431–5.
25. Béland MJ, Franklin RCG, Jacobs JP, et al. Update from the international working group for mapping and coding of nomenclatures for paediatric and congenital heart disease. Cardiol Young. 2004;14:225–9.
26. Franklin RC, Beland MJ, Krogmann ON. Mapping and coding of nomenclatures for paediatric and congenital heart disease. Cardiol Young. 2006;16:105–6.
27. Multi-Societal Database Committee for Pediatric and Congenital Heart Disease. Databases and the assessment of complications associated with the treatment of congenital of patients with congenital cardiac disease. Cardiol Young. 2008;18 Suppl 2:1–530.
28. Jacobs JP, Franklin RCG, Jacobs ML, et al. Classification of the functionally univentricular heart: unity from mapped codes. Cardiol Young. 2006;16 Suppl 1:9–21.
29. Tchervenkov CI, Jacobs JP, Weinberg PM, et al. The nomenclature, definition and classification of hypoplastic left heart syndrome. Cardiol Young. 2006;16:339–68.
30. Jacobs JP, Franklin RC, Wilkinson JL, et al. The nomenclature, definition and classification of discordant atrioventricular connections. Cardiol Young. 2006;16 Suppl 3:72–84.
31. Jacobs JP, Anderson RH, Weinberg P, et al. The nomenclature, definition and classification of cardiac structures in the setting of heterotaxy. Cardiol Young. 2007;17 Suppl 2:1–28.
32. Jenkins KJ, Gauvreau K, Newburger JW, Spray TL, Moller JH, Iezzoni LI. Consensus-based method for risk adjustment for surgery for congenital heart disease. J Thorac Cardiovasc Surg. 2002;123:110–8.
33. Lacour-Gayet F, Clarke D, Jacobs J, Gaynor W, Hamilton L, Jacobs M, et al. The Aristotle score for congenital heart surgery. Semin Thorac Cardiovasc Surg Pediatr Card Surg Annu. 2004;7:185–91.
34. O'Brien SM, Clarke DR, Jacobs JP, Jacobs ML, Lacour-Gayet FG, Pizarro C, et al. An empirically based tool for analyzing mortality associated with congenital heart surgery. J Thorac Cardiovasc Surg. 2009;138:1139–53.
35. Crowe S, Brown KL, Pagel C, et al. Development of a diagnosis- and procedure-based risk model for 30-day outcome after pediatric cardiac surgery. J Thorac Cardiovasc Surg. 2013;145(5):1270–8.
36. Giroud JM, Jacobs JP, Spicer D, et al. Report from the international society for nomenclature of paediatric and congenital heart disease: creation of a visual encyclopedia illustrating the terms and definitions of the international pediatric and congenital cardiac code. World J Pediatr Congenit Heart Surg. 2010;1(3): 300–13.
37. Franklin RCG, Jacobs JP, Tchervenkov CI, Béland M. Report from the executive of the International Working Group for Mapping and Coding of Nomenclatures for Paediatric and Congenital Heart Disease: bidirectional crossmap of the short lists of the European paediatric cardiac code and the international congenital heart surgery nomenclature and database project. Cardiol Young. 2002;12(Suppl II):18–22.
38. Jacobs JP, Maruszewski B, European Association for Cardio-thoracic Surgery-Society of Thoracic Surgeons Joint Congenital Heart Surgery Nomenclature and Database Committee. Computerized outcomes analysis for congenital heart disease. Curr Opin Pediatr. 2005;17:586–91. Review.
39. College of American Pathologists. SNOMED historical perspectives. Available at: http://www.cap.org/apps/cap.portaL?_nfpb=true&cntvwrPtlt_actionOverride=%2Fportlets%2FcontentViewer%2Fshow&_windowLabel=cntvwrPtlt&cntvwrPtlt%7BactionForm.contentReference%7D=snomed%2FhistPersp.html&_state=maximized&_pageLabel=cntvwr. Accessed Oct 2013.
40. Tudorache T, Falconer S, Nyulas C, et al. Supporting the collaborative authoring of ICD-11 with WebProtégé. AMIA Annu Symp Proc. 2010;2010:802–6.
41. Cronk CE, Malloy ME, Pelech AN, et al. Completeness of state administrative databases for surveillance of congenital heart disease. Birth Defects Res A Clin Mol Teratol. 2003;67:597–603.
42. Frohnert BK1, Lussky RC, Alms MA, et al. Validity of hospital discharge data for identifying infants with cardiac defects. J Perinatol. 2005 Nov;25(11):737–42.
43. Strickland MJ, Riehle-Colarusso TJ, Jacobs JP, et al. The importance of nomenclature for congenital cardiac disease: implications for research and evaluation. In: Jacobs JP, editor. 2008 Cardiology in the Young Supplement: Databases and The Assessment of Complications associated with The Treatment of Patients with Congenital Cardiac Disease. Prepared by: The Multi-Societal Database Committee for Pediatric and Congenital Heart Disease. Cardiology in the Young, vol. 18, issue S2 (Suppl 2), p. 92–100, December 9, 2008.
44. Pasquali SK, Peterson ED, Jacobs JP, He X, Li JS, Jacobs ML, Gaynor JW, Hirsch JC, Shah SS, Mayer JE. Differential case ascertainment in clinical registry versus administrative data and impact on outcomes assessment for pediatric cardiac operations. Ann Thorac Surg. 2013;95(1):197–203. doi:10.1016/j.athoracsur.2012.08.074. Epub 2012 Nov 7. PMID: 23141907.
45. Gibbs JL, Monro JL, Cunningham D, Rickards A. Survival after surgery or therapeutic catheterisation for congenital heart disease in children in the United Kingdom: analysis of the central cardiac audit database for 2000-1. Brit Med J. 2004;328:611–5.

Defining Terms in Lists of Nomenclature

6

Henry L. Walters III and Steven D. Colan

Abstract

A hierarchical organization of terms with definitions for pediatric and congenital heart disease (PCHD) comprises a language or system of communication that can be used to accurately describe the diagnoses and procedures associated with developmental cardiac malformations as well as acquired cardiac diseases that affect children and may persist into adulthood. However, some of the existing terminology is regional, disorganized, redundant, ambiguous and imprecise. As such, an internationally accepted, cohesive and comprehensive set of terms with definitions for PCHD is required to unify the subspecialty. To achieve this goal, the mission of the Definitions Working Group (DWG) of the International Society for the Nomenclature of Pediatric and Congenital Heart Disease (ISNPCHD) is to create scientifically accurate, precise and concise definitions for all of the diagnostic and procedural terms encompassed by the International Paediatric and Congenital Cardiac Code (IPCCC). A hierarchy and definitions for many of the parent terms of the IPCCC will also be used to populate the PCHD terms for the upcoming International Classification of Diseases (ICD-11) published by the World Health Organization (WHO). The ongoing work of the DWG ultimately has the potential to create a universally accepted, cohesive and comprehensive set of terms for PCHD with scientifically accurate and clear definitions. The ultimate realization of this goal would greatly facilitate and improve international PCHD outcomes analyses and quality improvement strategies.

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Cardiac • Congenital • Database • Defect • Definition • Disease • Heart • Hierarchy • Pediatric • Term

Introduction

A hierarchical organization of terms with definitions for pediatric and congenital heart disease (PCHD) comprises a language or system of communication that can be used to accurately describe the diagnoses and procedures associated with developmental cardiac malformations as well as acquired cardiac diseases that affect children and may persist through adulthood. Numerous PCHD terms are derived from Latin and Greek roots. Although intimate familiarity with these classical languages is not common, the PCHD terms derived therefrom are readily recognized and understood simply because they permeate the medical curricula and literature and are frequently used. Some Latin/Greek terms, such as *truncus arteriosus*, are one step further removed from intuitive understanding because they are additionally based upon embryology and, as such, these terms are intrinsically less descriptive, even to those with a basic understanding of Latin and Greek. Nonetheless, the sheer prevalence and common usage of Latin and Greek PCHD terms makes them universally familiar and, therefore, useful. English translations of PCHD terms can also be used as substitutes for the Latin/Greek terms (Anderson RH, June 2013, personal communication). For example, the *ductus arteriosus* can be called the *arterial duct* and *truncus arteriosus* can be called *common arterial trunk*. The journal, *Cardiology in the Young*, implements this process of anglicisation of PCHD terms in its editorial process [1] to improve grammatical precision, literary style and clarity (Anderson RH, June 2013, personal communication). Some very common Latin/Greek terms that seem intrinsically obvious, such as *atrial septal defect*, are not so straightforward, however, when one considers that not all atrial septal defects are, in fact, *defects* in the atrial septum. An interatrial communication of the sinus venosus type is just such an example. The problems caused by the diverse and sometimes unclear or scientifically incorrect PCHD terms that exist worldwide underscore the need for building crossmaps between existing terms and for creating accurate and internationally accepted definitions for these terms so that clinicians, researchers, epidemiologists and administrators can communicate precisely and can begin comparing apples to apples. During the process of agreeing upon these definitions it will sometimes become clear that certain terms should be retired to the status of synonyms and be replaced with terms that are more clear, intuitive and/or scientifically correct.

Currently, major international classifications such as the International Classification of Diseases (ICD) [2, 3] and the Systematized Nomenclature of Medicine Clinical Terms (SNOMED CT) [3, 4] do not include term definitions. The World Health Organization (WHO) and the International Health Terminology Standards Development Organization (IHTSDO), the respective parent organizations of ICD and SNOMED CT, both understand the importance of definitions and are committed to including definitions in their respective updated versions.

A universally accepted, cohesive and comprehensive set of terms for PCHD, it seems apparent, would be desirable to facilitate international outcomes analyses and quality improvement. Such a language, though, for medicine in general has historically been elusive, as articulated by William Farr (b1807-d1883). He emphasized the need for a common international lexicon to allow for the epidemiological study of diseases and their causes. As the first medical statistician of the General Register Office of England and Wales, Farr noted, in his first report, published in 1839:

> The advantages of a uniform statistical nomenclature, however imperfect, are so obvious, that it is surprising that no attention been paid to its enforcement in Bills of Mortality. Each disease has,

> in many instances, been denoted by three or four terms, and each term has been applied to as many different diseases: vague inconvenient names have been employed, or complications registered instead of primary diseases. The nomenclature is of as much importance in this department of inquiry, as weights and measures in the physical sciences, and should be settled without delay [3, 5].

The Problem

The terminology of PCHD shares many deficiencies in common with other disciplines of medicine. These include:

1. Using multiple terms for a solitary disease
2. Using a solitary term for multiple diseases
3. Classifying or defining according to the clinical presentation
4. Using terms that are unfamiliar or uninformative
5. Using shorthand or abbreviated terms
6. Using eponyms
7. Classifying or defining based upon the approach to the treatment or surgical repair and, finally,
8. Classifying or defining according to embryology or genetics.

Multiple Terms – Solitary Disease: It is notable that the same types of nomenclature problems that confronted medicine in Farr's nineteenth century exist today in the terminology of PCHD. For example, as it pertains to the problem of "multiple terms for a solitary disease", the same type of *ventricular septal defect* (*VSD*) is alternately termed *subarterial*, *juxtaarterial*, *doubly committed juxtaarterial*, *conal septal*, *conoseptal hypoplasia*, *absent outlet septum*, *intraconal*, *supracristal*, *infundibular or subpulmonary*, all depending upon local custom [6].

Solitary Term – Multiple Diseases: As regards a solitary term that is inappropriately applied to many congenital cardiac diseases, the term *single ventricle* is used to encompass a variety of diverse congenital heart diseases both with and without an anatomically *single ventricle* [7–9].

Clinical Presentation: In addition to pointing out these two pitfalls, Farr also suggested that various causes of death, identified in his day, be classified, not according to the type of disease presentation, such as its symptoms and findings, (i.e. pyrexial, cachexial, neurotic diseases) but rather according to the anatomical location of the disease [5]. An example of such a "clinical phenotypic" classification might include dividing PCHD lesions into three physiological groups, those associated with cyanosis, pulmonary overcirculation or low cardiac output. While such a classification may sometimes be useful for understanding and categorizing the physiology of PCHD, it is a poor choice for the classification thereof in a database because of the extensive overlap that can occur between these categories. For example, both cyanotic lesions and those associated with pulmonary overcirculation can also be associated with low cardiac output.

Shorthand/Abbreviations: The hazards of using shorthand or abbreviated terms are inherently obvious. A few examples of such terms are *tet* for tetralogy of Fallot, *transpo* for transposition of the great arteries and *total veins* for totally anomalous pulmonary venous connection. For example, the term "plast" is often used as shorthand for any lesion treated with a Norwood-type procedure, obscuring the marked differences between lesions such as hypoplastic left heart syndrome and unbalanced atrioventricular septal defect. These casual terms constitute a subtle type of "insider's language" and tend to be readily adopted due to their brevity, peer pressure and the ease and frequency with which they are used. Yet they are also poor and imprecise substitutes for the name of the actual lesion. These terms can, therefore, result in lost information and important miscommunication and, perhaps most importantly, can create bad habits and misconceptions among students and trainees.

Eponyms: The use of eponyms, while common, is not optimal for terms describing congenital heart disease. Some examples of such diagnostic, anatomical or procedural terms include Marfan syndrome, Kawasaki's disease, sinus of Valsalva or Waterston shunt. These terms, while often yielding important contextual information, do not, intrinsically, convey precise information about the meaning or nature of the term and, hence, should be reserved as historical footnotes or listed as important synonyms.

Treatment/Surgical Repair: Classifying or defining congenital heart disease according to the type of treatment used should also be avoided. For example, while it is true that patients with double outlet right ventricle (DORV) may require tunnel closure of the VSD to the malposed aortic valve whereas patients with a simple VSD can undergo a flat-patch closure, using the mode of repair as the primary basis for defining DORV or VSD or distinguishing these entities from each other is not optimal for several reasons. Firstly, the diagnosis should remain the same regardless of whether an intervention is undertaken or not, and secondly, the type of intervention may well evolve over time (for example, aortic translocation in DORV or device closure of VSD) even though the morphological entity does not [10, 11].

The term *single ventricle* is a controversial example in which it is common to define a PCHD term according to its mode of repair. First of all, the term, itself, is immediately inadequate because patients with a so-called *single ventricle* often have somewhat more than one complete ventricle, albeit usually less than two complete ventricles. One may argue that the term *single ventricle* is just a name that refers to an entity and that the term is, therefore, no more important than a name like *John* or *Mary*. The logical extension to this argument is that it is the definition that really matters and not the term itself. The term, one may say, is simply a name. However logical as this may seem, the term *single ventricle* is, nonetheless, an example of a PCHD term that compromises on scientific accuracy. While *single ventricle* should be retained as an important synonym because of its prevalence in the medical literature and its undeniable place in the history of PCHD, the term *functionally univentricular heart* is an imperfect, but more scientifically accurate, replacement term [7–9, 12] in part because the introduction of the modifier, *functionally*, makes it clear that this is a category based on more than just the anatomic findings since, as Jacobs and Anderson have said, "The entire ventricular mass is *functionally univentricular* whenever one or the other ventricle is incapable, for whatever reason, of supporting either the systemic or the pulmonary circulation" [9]. What may be considered an adequate definition for *functionally univentricular heart* is usually more detailed and more complex because this is a broad term that encompasses a wide spectrum of diverse congenital cardiac lesions. Because of this morphological diversity, a *functionally univentricular heart* cannot be defined solely according to its anatomy, as one would ideally like to define a congenital cardiac lesion. Rather, one may define *functionally univentricular heart* as "a spectrum of congenital cardiovascular malformations in which the ventricular mass may not readily lend itself to partitioning that commits one ventricular pump to the systemic circulation, and another to the pulmonary circulation. A heart may be functionally univentricular because of its anatomy or because of the lack of feasibility or lack of advisability of surgically partitioning the ventricular mass. Common lesions in this category typically include double inlet right ventricle (DIRV), double inlet left ventricle (DILV), tricuspid atresia, mitral atresia, and hypoplastic left heart syndrome. Other lesions which sometimes may be considered to be a functionally univentricular heart include complex forms of atrioventricular septal defect, double outlet right ventricle, congenitally corrected transposition, pulmonary atresia with intact ventricular septum, and other cardiovascular malformations. Specific diagnostic codes should be used whenever possible, and not the term 'functionally univentricular heart'" [13]. This is certainly an example of an imperfect term with an equally imperfect definition in that the term encompasses a multitude of diverse congenital cardiac lesions and its definition, by necessity, invokes the surgical procedure(s) used for the surgical repair. This underscores the difficulty in both naming and defining this complex group of congenital heart lesions.

Embryology/Genetics: While there may be some value in classifying or naming anatomical congenital heart disease terms according to their embryology or genetics, this approach should be reserved for special situations where development is the focus of the database. There are many examples of congenital cardiac terms that are based upon embryology, two of which, for example, are *truncus arteriosus* and *sinus venosus*

atrial septal defect. Many of these embryologically oriented terms are deeply imbedded in the terminology of congenital heart disease and will likely persist over time due to their overwhelming prevalence and frequency of use. Nonetheless, currently the classification, management and measurement of outcomes in congenital heart disease emphasize the morphology of the congenital heart defects as opposed to the proposed embryological origins or to various identified genetic defects. While PCHD may ultimately be distilled to and classified by its genotype, the current state of knowledge of this important field of investigation is not yet specific enough to allow us to propose such an organization. For example it is known that certain myosin binding protein mutations are associated with cardiomyopathy, either dilated, restrictive or hypertrophic. The same mutations, however, can also be associated with non-compaction or even no detectable disease at all [14]. Hence, while it remains important to capture and categorize genetic information related to congenital heart disease, the emphasis is currently on phenotype rather than genotype. This emphasis may very well change over time as more is learned about the genetics of congenital heart disease.

In summary, the language of PCHD should continue to be rooted primarily in its structural aspects (morphology) and not in its physiology, mode of repair, embryology or genetics. Our understanding of etiology (including the genetic basis) and of treatment are the most evanescent and dynamic aspects related to PCHD. Definitions should, therefore, be designed so that they remain relevant and as accurate as possible, even while therapies evolve and our knowledge of molecular biology increases exponentially.

The Solution

Though a universally accepted, cohesive and comprehensive set of terms and definitions for PCHD, avoiding the pitfalls described above, is desirable, nonetheless an eclectic list of congenital heart disease terms already exists stratified within a number of different classifications. For example, the Society of Thoracic Surgeons' Congenital Heart Surgery Database (STS-CHSD) and the European Association for Cardiothoracic Surgery European Congenital Heart Defects Database (EACTS-ECHDD) both participated in the International Congenital Heart Surgery Nomenclature and Database Project (ICHSNDP) to standardize the nomenclature and reporting strategies that would establish the foundations for an international congenital heart disease database. The work product of the ICHSNDP was reported as the EACTS-STS Database short and long lists in a special supplement of the Annals of Thoracic Surgery published in April of 2000 [15]. This so-called "molecular" approach to the stratification of the nomenclature of PCHD can be compared to the more "atomic" structure of the Association for European Paediatric Cardiology's European Paediatric Cardiac Code (AEPC-EPCC) that was published independently in Cardiology in the Young in January of the same year [16]. Examples of other types of databases related to congenital heart disease, in addition to those mentioned above, include but are not limited to: (1) institutional congenital heart disease databases such as the Fyler Codes of Boston Children's Hospital [17]; (2) research-focused databases such as the Congenital Heart Surgeons Society Database (CHSS Database) [18]; (3) specialty databases such as the Pediatric Heart Transplant Study Group database (PHTSG) [19]; (4) pediatric cardiac catheterization databases such as the IMPACT Registry (IMproving Pediatric and Adult Congenital Treatment) [20, 21]; (5) databases related to supporting subspecialties such as pediatric cardiac anesthesiology (STS Congenital Database Anesthesia Module) [22] and pediatric critical care (Virtual Pediatric Intensive Care Unit Performance System (VPS)) [23]; and (6) international administrative databases such as the ICD [2, 3].

The diverse hierarchies and terms populating the multitude of databases that contain PCHD terms would seem to mitigate against the successful creation of a unified international congenital heart disease nomenclature, much less one with agreed upon definitions. This task was initiated by the leadership of two of the most

Table 6.1 Names, medical specialties and countries of origin of the members of the Definitions Working Group (DWG) of the International Society for the Nomenclature of Pediatric and Congenital Heart Disease (ISNPCHD)

Definitions Working Group (DWG)		
Member name	Member specialty	Member institution/country
Vera D. Aiello	Cardiac Morphologist	Heart Institute of San Paulo University, Brazil
Robert H. Anderson	Cardiac Morphologist	Inst. Medical Genetics, Newcastle University, UK
Marie J. Beland	Pediatric Cardiologist	The Montreal Children's Hospital, Canada
Steven D. Colan (Co-Chair)	Pediatric Cardiologist	Boston Children's Hospital, USA
Rodney C. Franklin	Pediatric Cardiologist	Royal Brompton Hospital, UK
J. William Gaynor	Pediatric Cardiac Surgeon	Children's Hospital of Philadelphia, USA
Jorge Giroud	Pediatric Cardiologist	All Children's Hospital, USA
Lucile Houyel	Pediatric Cardiologist	Hôpital Marie – Lannelongue, France
Christopher Hugo-Hamman	Pediatric Cardiologist	University of Stellenbosch, South Africa
Jeffrey P. Jacobs	Pediatric Cardiac Surgeon	All Children's Hospital, USA
Marshall L. Jacobs	Pediatric Cardiac Surgeon	Johns Hopkins University SOM, USA
Howard Jeffries	Pediatric Cardiac Critical Care	Seattle Children's Hospital, USA
Amy Juraszek	Pediatric Cardiologist	UT Southwestern Medical Center, USA
Otto N. Krogmann	Pediatric Cardiologist	CHD Heart Center Duisburg, Germany
Hiromi Kurosawa	Pediatric Cardiac Surgeon	Former, Tokyo Women's Medical Univ., Japan
Bohdan Maruszewski	Pediatric Cardiac Surgeon	The Children's Memorial Health Institute, Poland
Stephen Seslar	Pediatric Cardiologist	Seattle Children's Hospital, USA
Giovanni Stellin	Pediatric Cardiac Surgeon	University of Padova, Italy
Christo I. Tchervenkov	Pediatric Cardiac Surgeon	The Montreal Children's Hospital, Canada
Henry L. Walters (Co-Chair)	Pediatric Cardiac Surgeon	Children's Hospital of Michigan, USA
Paul M. Weinberg	Pediatric Cardiologist/Morphologist	Children's Hospital of Philadelphia, USA
Jim Wilkinson	Pediatric Cardiologist	Royal Children's Hospital, Australia

widely used databases dedicated solely to congenital heart disease, the EACTS-STS [15] and the AEPC-EPCC [16] databases, along with other international experts. In 2000 they formed the International Society for the Nomenclature of Pediatric and Congenital Heart Disease (ISNPCHD) [3]. Over the course of the next decade members of the ISNPCHD fulfilled their mission of creating an international database for congenital heart disease by crossmapping the EACTS-STS and the AEPC-EPCC terms into what is now called the International Paediatric and Congenital Cardiac Code (IPCCC) [3, 24]. This work, performed by the Nomenclature Working Group (NWG) of the ISNPCHD, preserved the integrity of the hierarchy and terms of the individual databases by using an inclusive crossmap technique that matched terms between the two databases thereby creating the codes of the IPCCC [24]. The Nomenclature Working Group has also previously published review articles which provide a unified and comprehensive classification, with definitions, for several complex congenital cardiac malformations, along with a complete listing of the relevant codes and terms in both versions of the IPCCC: the functionally univentricular heart [8], hypoplastic left heart syndrome [25], discordant atrioventricular connections [26] and cardiac structures in the setting of heterotaxy [27].

In 2007 at the ISNPCHD meeting in Tokyo, Japan, the Definitions Working Group (DWG) was established (Table 6.1) with the mandate to build upon the initial efforts of the NWG by

creating definitions for all of the diagnostic and procedural terms encompassed by the IPCCC. These definitions were to be scientifically accurate, precise and as concise as possible. Inclusivity was assured by choosing the IPCCC as the list of terms to define since it cross-mapped the EACTS-STS and the AEPC-EPCC database terms and since the IPCCC was freely available online for download to be used by other institutions or for crossmapping to their databases [24].

According to its Latin root, *-finire*, to *define* a term is to fix or to mark its limits, thereby determining not only what it is but also what it is not. In so doing one identifies the essential qualities or the meaning of the entity to which the term applies as opposed to establishing quantitative diagnostic criteria or listing an expansive description of all possible associations and variations. As stated earlier, anatomic elements should be defined anatomically and physiologic ones should be defined physiologically. Definitions should, most importantly, be scientifically accurate. For example, although the term *sinus venosus atrial septal defect* is commonly used, it is more scientifically accurate to call this a sinus venosus *interatrial communication* because, while it *functions* as an interatrial communication in the mouth of the superior vena cava, this lesion is not an actual *defect* of the atrial septum but rather results from the biatrial connection of the superior vena cava and right upper pulmonary veins [28–30]. For this same reason of scientific accuracy, the parent term *atrial septal defect* is better called an *interatrial communication* since not all interatrial communications are actual *defects* of the interatrial septum. In addition to being scientifically accurate, PCHD definitions should be clear, consistent, incisive, and, whenever possible, concise. An example definition that aptly illustrates all of these attributes is that of ventricular septal defect (VSD): "A congenital cardiovascular malformation in which there is a hole between the ventricular chambers or ventricular remnants" [6, 31]. Whenever possible, for the sake of consistency, the definitions begin with the same phrase, "A congenital cardiac malformation in which …". Since some terms, like VSD, can be considered *parent terms*, the definitions of any derived terms, like perimembranous VSD, should use the parent term, itself, rather than repeat the definition thereof. According to this rule the definition of inlet VSD would then be: "A congenital cardiovascular malformation in which there is a ventricular septal defect that permits direct flow between the inlet components of the ventricles" [32, 33]. PCHD definitions should, however, not sacrifice scientific accuracy and clarity for the sake of being incisive and concise. Hence the somewhat longer and convoluted definition of perimembranous VSD is: "A congenital cardiovascular malformation in which there is a ventricular septal defect contiguous with the site of the membranous septum, defined as the area of the septum contiguous with the fibrous continuity between the leaflets of an atrioventricular valve and an arterial valve" [6, 34–36].

While most PCHD definitions may stand on their own merits, there are some situations in which supplemental explanation is required to promote clarity, to explain variable interpretations and/or to allow for an expression of controversy. Hence, a *commentary* is required and is added to supplement some definitions. An example supplement for the definition of VSD is : "The VSD is defined on the basis of its margins as seen from the aspect of the morphologically right ventricle. In the setting of double outlet right ventricle, the defect provides the outflow from the morphologically left ventricle. In univentricular atrioventricular connections with functionally single left ventricle with an outflow chamber, the communication is referred to by some as a bulboventricular foramen" [37].

While the most clear and scientifically accurate PCHD term is listed as the one to be defined, it is important to remain inclusive by retaining as many synonyms, historical, local/regional or institutional terms as possible. The synonym does not constitute a part of the definition, but is used to prevent the loss of important data and facilitate accurate searches for identical terms.

This goal is accomplished by creating a list of acceptable *synonyms*. *Synonyms* are defined as terms that have the identical meaning in all senses, as the term being defined. When the synonym is more frequently used than the primary term, it is listed immediately beside the primary term to be defined. For example, while the most scientifically accurate term may be *interatrial communication*, the term *atrial septal* defect is more widely used and, as such, is listed immediately beside *interatrial communication* in the hierarchy [38]. On the other hand *perimembranous VSD* can also be called *paramembranous VSD or Type 2 VSD*. Since these two synonyms are no longer widely used, they are placed in a separate list of synonyms linked to the primary term [39]. Similarly, a separate list of acceptable abbreviations linked to the primary term is also created. Finally, in the interest of total inclusivity, a list of poor synonyms and abbreviations is also maintained and linked to each principle term to be defined. As described earlier, *sinus venosus ASD*, is not a scientifically accurate term and, as such, it is placed in this list of poor synonyms [40].

After the DWG established the principles for creating definitions for the terms of PCHD, the actual process for crafting the definitions was developed. Individual members of the DWG were assigned terms to define. These definitions were then debated, modified and ratified at the subsequent six annual meetings of the DWG held from 2008 to 2013 (Table 6.2). While an exhaustive listing of all of the definitions completed to date is outside the scope of this chapter, some examples of these definitions are listed in Table 6.3. With more than 8,000 diagnostic and procedural terms contained within the IPCCC the decision of where to actually start the process of defining was established when the DWG accepted the challenge of establishing the diagnostic PCHD terms, hierarchy and definitions for the upcoming International Classification of Diseases (ICD-11) published by the World Health Organization (WHO). In previous versions (ICD-9 and ICD-10) there were relatively few PCHD terms included, with 35 and 73 terms respectively. The hierarchy of these terms was not optimal and they were placed within the "Rare Diseases" section. For ICD-11 the decision was made to place PCHD within the Internal Medicine Topic Advisory Group, assigned to the Cardiovascular Working Group. Through a process of consensus a final list of approximately 311 terms were selected and organized into a six-level hierarchy by coalescing the best of the diagnostic short lists of both the EPCC of the AEPC and the EACTS-STS databases [41]. In creating this hierarchy, with its list of terms, emphasis was placed upon scientific accuracy, comprehensiveness and the creation of a logical categorization. The starting points for definitions have been assigned to each of these terms using source material that includes the papers of the ICHSNDP published in the special supplement of the Annals of Thoracic Surgery in April of 2000 [15] and previous publications of the ISNPCHD [3, 8, 27, 42]. These starting definitions are further refined by discussion/debate during full session of the DWG. Thus far 187 definitions have been fully ratified by the DWG and working definitions for future discussion/debate by the DWG for the remaining 107 terms have been assigned but have yet to be ratified [39]. Since these ICD-11 definitions comprise most of the parent diagnostic terms for the IPCCC, the definitions for the subsidiary diagnostic terms in the IPCCC will eventually build upon the definitions of these parent terms.

Table 6.2 Locations and dates of the working meetings of the Definitions Working Group (DWG) of the International Society for the Nomenclature of Pediatric and Congenital Heart Disease (ISNPCHD)

Meetings of the Definitions Working Group (DWG)	
Location	Year
Cape Cod, MA, USA	July 2008
Boston, MA, USA	May 2009
County Donegal, Republic of Ireland	July 2010
Wild Dunes, SC, USA	July 2011
St Goar, Germany	July 2012
Holetown, Barbados	December 2013
New York, NY	September 2014

Table 6.3 Examples of some of the definitions of pediatric and congenital heart disease terms created by the DWG during annual meetings from 2008 to 2013

Term	Definition
Interatrial communication (Atrial septal defect)	A congenital cardiac malformation in which there is a hole or pathway between the atrial chambers
Ventricular septal defect	A congenital cardiac malformation in which there is a hole or pathway between the ventricular chambers or ventricular remnants
Tetralogy of Fallot	A group of congenital cardiac malformations with biventricular atrioventricular alignments or connections characterized by anterosuperior deviation of the conal or outlet septum or its fibrous remnant, narrowing or atresia of the pulmonary outflow, a ventricular septal defect of the malalignment type, and biventricular origin of the aorta. Tetralogy of Fallot will always have a ventricular septal defect, narrowing or atresia of the pulmonary outflow, aortic override, and most often right ventricular hypertrophy
Atrioventricular septal defect (Atrioventricular canal defect)	A congenital cardiac malformation with a common atrioventricular junction and an atrioventricular septal defect
Functionally univentricular heart	The term "functionally univentricular heart" describes a spectrum of congenital cardiac malformations in which the ventricular mass may not readily lend itself to partitioning that commits one ventricular pump to the systemic circulation, and another to the pulmonary circulation
Hypoplastic left heart syndrome	A congenital cardiovascular malformation where there is a spectrum of cardiovascular malformations with normally aligned great arteries without a common atrioventricular junction and significant hypoplasia of the left ventricle associated with atresia, stenosis, or hypoplasia of the aortic or mitral valve, or both valves, and hypoplasia of the ascending aorta and aortic arch. A spectrum of congenital cardiovascular malformations with normally aligned great arteries without a common atrioventricular junction with significant hypoplasia of the left ventricle and including atresia, stenosis, or hypoplasia of the aortic or mitral valve, or both valves, and hypoplasia of the ascending aorta and aortic arch.
Visceral heterotaxy (Abnormal arrangement of thoraco-abdominal organs)	A congenital malformation in which the internal thoraco-abdominal organs demonstrate abnormal arrangement across the left-right axis of the body. By convention, heterotaxy syndrome does not include patients with complete mirror-imaged arrangement of the internal organs along the left-right axis also known as "situs inversus totalis"
Transposition of the great arteries (Discordant ventriculo-arterial connections)	A congenital cardiovascular malformation in which the morphologically right ventricle connects to the aorta and the morphologically left ventricle connects to the pulmonary trunk
Congenitally corrected transposition (Discordant atrioventricular and ventriculo-arterial connections)	A congenital cardiovascular malformation in which the morphologically right atrium connects to the morphologically left ventricle, the morphologically left atrium connects to the morphologically right ventricle, the morphologically right ventricle connects to the aorta, and the morphologically left ventricle connects to the pulmonary trunk

Conclusion

The treatment of PCHD is improved more by cooperation than by competition. Cooperation is enhanced by improving the precision of communication amongst all those who are involved in the field. Communication is enhanced by using the same diagnostic and procedural terms and definitions. The ongoing work of the DWG ultimately has the potential to create a universally accepted, cohesive and comprehensive set of terms for PCHD with scientifically accurate and clear definitions. The ultimate realization of this goal would greatly facilitate and improve international PCHD outcomes analyses and quality improvement strategies.

References

1. Cardiology in the young: instructions for contributors [Internet]. 2013 [cited 2013 Jul 1]. Available from: http://assets.cambridge.org/CTY/CTY_ifc.pdf.
2. World Heath Organization. History of the development of the ICD [Internet]. 2013 [cited 2013 Jul 1]. Available from: http://www.who.int/classifications/icd/en/HistoryOfICD.pdf.
3. Franklin RC, Jacobs JP, Krogmann ON, Béland MJ, Aiello VD, Colan SD, Elliott MJ, Gaynor JW, Kurosawa H, Maruszewski B, Stellin G, Tchervenkov CI, Walters III HL, Weinberg P, Anderson RH. Nomenclature for congenital and paediatric cardiac disease: historical perspectives and the international pediatric and congenital cardiac code. Cardiol Young. 2008;18 Suppl 2:70–80.
4. International Health Terminology Standards Development Organization [Internet]. 2013 [cited 2013 Jul 1]. Available from: http://www.ihtsdo.org/snomed-ct/.
5. Farr W. First annual report of the registrar-general of births, deaths, and marriages in England. London: His Majesty's Stationery Office; 1839. p. 99–102.
6. Jacobs JP, Burke RP, Quintessenza JA, Mavroudis C. Congenital heart surgery nomenclature and database project: ventricular septal defect. Ann Thorac Surg. 2000;69:S25–35.
7. Jacobs ML, Mayer JE. Congenital heart surgery nomenclature and database project: single ventricle. Ann Thorac Surg. 2000;69:S197–204.
8. Jacobs JP, Franklin RC, Jacobs ML, Colan SD, Tchervenkov CI, Maruszewski B, Gaynor JW, Spray TL, Stellin G, Aiello VD, Béland MJ, Krogmann ON, Kurosawa H, Weinberg PM, Elliott MJ, Mavroudis C, Anderson RH. Classification of the functionally univentricular heart: unity from mapped codes. Cardiol Young. 2006;16 Suppl 1:9–21.
9. Jacobs ML, Anderson RA. Nomenclature of the functionally unventricular heart. Cardiol Young. 2006;16 Suppl 1:3–8.
10. Walters HL, Mavroudis C, Tchervenkov CI, Jacobs JP, Lacour-Gayet F, Jacobs ML. Congenital heart surgery nomenclature and database project: double outlet right ventricle. Ann Thorac Surg. 2000;69:S249–63.
11. Walters HL, Mavroudis C. Double-outlet ventricles. In: Mavroudis C, Backer CL, editors. Pediatric cardiac surgery. Oxford: Wiley-Blackwell; 2013. p. 457–91.
12. Van Praagh R, Ongley PA, Swan HJC. Anatomic types of single or common ventricle in Man: morphologic and geometric aspects of sixty necropsied cases. Am J Cardiol. 1964;13:367–86.
13. ICD11 Beta Draft, World Health Organization. Functionally univentricular heart [Internet]. 2013 [cited 2013 Jul 1]. Available from: http://apps.who.int/classifications/icd11/browse/f/en#/http%3a%2f%2fid.who.int%2ficd%2fentity%2f5417233.
14. McNally EM, Golbus JR, Puckelwartz MJ. Genetic mutations and mechanisms in dilated cardiomyopathy. J Clin Res. 2013;123:19–26.
15. Mavroudis C, Jacobs JP. Congenital heart surgery nomenclature and database project. Ann Thorac Surg. 2000;69:S1–357.
16. Franklin RCG, Anderson RH, Daniels O, Elliot M, Gewillig MHML, Renzo G, Krogmann ON, Ulmer HE, Stocker FP. Report of the coding committee of the association for european paediatric cardiology. Cardiol Young. 2000;10:S1–146.
17. Triedman JK. Methodologic issues for database development: trends. In: Keane JF, Lock JE, Fyler DC, editors. Nadas' pediatric cardiology. 2nd ed. Philadelphia: Saunders/Elsevier; 2006. p. 323–36.
18. Patel A, Hickey E, Mavroudis C, et al. Impact of noncardiac congenital and genetic abnormalities on outcomes in hypoplastic left heart syndrome. Ann Thorac Surg. 2010;89(6):1805–13.
19. Hsu DT, Naftel DC, Webber SA, Morrow WR, Canter CE, Chinnock RE, Clark ML, Kirklin JK. Lessons learned from the pediatric heart transplant study. Congenit Heart Dis. 2006;1(3):54–62.
20. Martin GR, Beekman RH, Ing FF, Jenkins KJ, McKay CR, Moore JW, Ringel RE, Rome JJ, Ruiz CE, Vincent RN. The IMPACT Registry™: IMproving Pediatric and Adult Congenital Treatments. Semin Thorac Cardiovasc Surg Pediatr Card Surg Annu. 2010;13(1):20–5.
21. Jenkins KJ, Beekman III RH, Bergersen LJ, Everett AD, Forbes TJ, Franklin RCG, Klitzner TS, Krogman ON, Martin GR, Webb CL. Databases for assessing the outcomes of the treatment of patients with congenital and paediatric cardiac disease – the perspective of cardiology. Cardiol Young. 2008;18 Suppl 2:116–23.
22. Vener DF, Guzzetta N, Jacobs JP, Williams GD. Development and implementation of a new data registry in congenital cardiac anesthesia. Ann Thorac Surg. 2012;94:2159–65.
23. LaRovere JM, Jeffries HE, Sachdeva RC, Rice TB, Wetzel RC, Cooper DS, Bird GL, Ghanayem NS, Checchia PA, Chang AC, Wessel DL. Cardiol Young. 2008;18 Suppl 2:130–6.
24. International Paediatric and Congenital Cardiac Code: IPCCC [Internet]. 2013 [cited 2013 Jul 1]. Available from: http://www.ipccc.net/.
25. Tchervenkov CI, Jacobs JP, Weinberg PM, Aiello VD, Béland MJ, Colan SD, Elliott MJ, Franklin RC, Gaynor JW, Krogmann ON, Kurosawa H, Maruszewski B, Stellin G. The nomenclature, definition and classification of hypoplastic left heart syndrome. Cardiol Young. 2006;16(4):339–68.
26. Jacobs JP, Franklin RCG, Wilkinson JL, Cochrane AD, Karl TR, Aiello VD, Béland MJ, Colan SD, Elliott MJ, Gaynor JW, Krogmann ON, Kurosawa H, Maruszewski B, Stellin G, Tchervenkov CI, Weinberg PM. The nomenclature, definition and classification of discordant atrioventricular connections. In: 2006 Supplement to cardiology in the young: controversies and challenges of the atrioventricular junctions and other challenges facing paediatric cardiovascular practitioners and their patients, Jacobs JP, Wernovsky

G, Gaynor JW, and Anderson RH (Editors). Cardiol Young. 2006;16(Suppl 3):72–84.
27. Jacobs JP, Anderson RH, Weinberg PM, Walters HL, Tchervenkov CI, Del Duca D, Franklin RC, Aiello VD, Béland MJ, Colan SD, Gaynor JW, Krogmann ON, Kurosawa H, Maruszewski B, Stellin G, Elliott MJ. The nomenclature, definition and classification of cardiac structures in the setting of heterotaxy. Cardiol Young. 2007;17 Suppl 2:1–28.
28. Lewis FJ, Taufic M, Varco RL, Niazi S. The surgical anatomy of atrial septal defects: experiences with repair under direct vision. Ann Surg. 1955;142(3): 401–16.
29. Geddes AC. Abnormal superior vena cava. Anat Anz. 1912;41:449–53.
30. Ingalls NW. Report communication between the right pulmonary veins and the superior vena cava. Anat Rec. 1907;1:14–22.
31. ICD-11 Beta Draft, World Health Organization. Ventricular septal defect [Internet]. 2013 [cited 2013 Jul 1]. Available from: http://apps.who.int/classifications/icd11/browse/f/en-/http%3a%2f%2fid.who.int%2ficd%2fentity%2f1908503567.
32. Spicer DE, Anderson RH, Backer CL. Clarifying the surgical morphology of inlet ventricular septal defects. Ann Thorac Surg. 2013;95:236–41.
33. ICD-11 Beta Draft, World Health Organization. Inlet ventricular septal defect [Internet]. 2013 [cited 2013 Jul 1]. Available from: http://apps.who.int/classifications/icd11/browse/f/en-/http%3a%2f%2fid.who.int%2ficd%2fentity%2f1491280288.
34. Soto B, Becker AE, Moulaert AJ, Lie JT, Anderson RH. Classification of ventricular septal defects. Br Heart J. 1980;43:332–43.
35. Soto B, Ceballos R, Kirklin JW. Ventricular septal defects: a surgical viewpoint. JACC. 1989;14(5):1291–7.
36. ICD-11 Beta Draft, World Health Organization. Perimembranous ventricular septal defect [Internet]. 2013 [cited 2013 Jul 1]. Available from: http://apps.who.int/classifications/icd11/browse/f/en-/http%3a%2f%2fid.who.int%2ficd%2fentity%2f2023258628.
37. ICD-11 Beta Draft, World Health Organization. Commentary: ventricular septal defect [Internet]. 2013 [cited 2013 Jul 1]. Available from: http://apps.who.int/classifications/icd11/browse/f/en-/http%3a%2f%2fid.who.int%2ficd%2fentity%2f1908503567.
38. ICD-11 Beta Draft, World Health Organization. Synonym: interatrial communication [Internet]. 2013 [cited 2013 Jul 1]. Available from: http://apps.who.int/classifications/icd11/browse/f/en-/http%3a%2f%2fid.who.int%2ficd%2fentity%2f1285985084.
39. ICD-11 Beta Draft, World Health Organization. Synonym: perimembranous ventricular septal defect [Internet]. 2013 [cited 2013 Jul 1]. Available from: http://apps.who.int/classifications/icd11/browse/f/en-/http%3a%2f%2fid.who.int%2ficd%2fentity%2f1930019148.
40. ICD-11 Beta Draft, World Health Organization. Synonym: sinus venosus defect [Internet]. 2013 [cited 2013 Jul 1]. Available from: http://apps.who.int/classifications/icd11/browse/f/en-/http%3a%2f%2fid.who.int%2ficd%2fentity%2f2004408087.
41. ICD-11 Beta Draft, World Health Organization. Congenital anomalies of heart and/or great vessels and related acquired abnormalities [Internet]. 2013 [cited 2013 Jul 1]. Available from: http://apps.who.int/classifications/icd11/browse/f/en-/http%3a%2f%2fid.who.int%2ficd%2fentity%2f2004408087.
42. Jacobs JP, Pasquali SK, Morales DLS, Jacobs ML, Mavroudis C, Chai PJ, Tchervenkov CI, Lacour-Gayet FG, Walters HL, Quintessenza JA. Heterotaxy: lessons learned about patterns of practice and outcomes from the congenital heart surgery database of the society of thoracic surgeons. WJPCHS. 2011;2: 278–86.

Illustrating Terms in Lists of Nomenclature

7

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Abstract

Survival for children with cardiac disease has dramatically increased in the past four decades with the advent of improvements in diagnosis and treatment. In order to further decrease morbidity and mortality, optimization of outcomes must be vigorously pursued, and this optimization requires a common language when discussing and comparing results of the available diagnostic and therapeutic options. This common language exists and is named The International Pediatric and Congenital Cardiac Code (IPCCC). In order to make the IPCCC more universally understood, a 'virtual visual encyclopedia' has been created that links and illustrates the terms and definitions of the IPCCC with images of all types. The Archiving Working Group (AWG) of the International Society for Nomenclature of Paediatric and Congenital Heart Disease (ISNPCHD) is an organization composed of members of the international pediatric cardiac medical and surgical community that collaborate to illustrate, with representative images of all types and formats, the pertinent aspects of cardiac diseases that affect all pediatric patients, using the codes and definitions of the IPCCC as the organizational structure. This chapter describes

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the process of linking illustrations and nomenclature in the effort to better understand congenital and acquired cardiac disease and improve outcomes.

Keywords

Databases • Cardiac nomenclature • Illustrations • Congenital heart disease • Internet • Cardiac encyclopedia • Cardiac images • International Pediatric and Congenital Cardiac Code

Abbreviations

AEPC	Association for European Paediatric Cardiology
AWG	Archiving Working Group
CPT	Current procedural terminology
DWG	Definitions Working Group
EACTS	European Association for Cardio-Thoracic Surgery
ICD	The International Classification of Diseases
IPCCC	International Pediatric and Congenital Cardiac Code
ISNPCHD	International Society for Nomenclature of Paediatric and Congenital Heart Disease
JPEG	Joint Photographic Expert Group
MRI	Magnetic resonance imaging
NWG	Nomenclature Working Group
STS	Society of Thoracic Surgeons

Introduction

Attempts to understand, classify, and illustrate the various medical afflictions of mankind have been part of the human legacy from the earliest days of recorded antiquity. The best well documented of these histories come from the Greco-Roman world and were reintroduced into Western Europe at the time of the 'Golden Age' of Arabic-Islamic science [1]. The intellectual ferment of the Renaissance changed the prevalent static nature of medical thinking and encouraged the approach of direct observation. This process slowly changed the philosophical underpinnings of learning and practicing medicine, from the study of the writings of Galen to that of empirical observations. As the study of normal and pathologic anatomy grew from isolated instances to the systematic review of the available pathologies, catalogues or atlases linking illustrations with the prevailing terms grew in importance and availability. In the modern era, this phenomenon has become even more important; and now, terms of nomenclature, describing the diagnosis and treatments for the diverse forms of cardiac disease, and based on logic and the best available science, have become the standard way to classify and catalogue the diverse manifestations of cardiac disease of neonates, infants, children, and young adults. As discussed in previous chapters, the International Pediatric and Congenital Cardiac Code (IPCCC) is one of the most commonly used international systems of nomenclature for cardiac disease. The IPCC was copyrighted in 2005 by the International Society for Nomenclature of Paediatric and Congenital Heart Disease (ISNPCHD) and is freely available for download at http://www.ipccc.net/. The IPCCC consists of a cross-map linking the following systems of classification:

1. The International Congenital Heart Surgery Nomenclature and Database Project of the European Association for Cardio-Thoracic Surgery (EACTS) and the Society of Thoracic Surgeons (STS).
2. The European Paediatric Cardiac Coding (EPCC) of the Association for European Paediatric and Congenital Cardiology (AEPC).
3. The Fyler Codes of Boston Children's Hospital and Harvard University.
4. The International Classification of Diseases (ICD-9 and ICD-10) of the World Health Organization.

5. The Current Procedural Terminology (CPT) of the American Medical Association.

As one of the three working groups of the ISNPCHD, the Archiving Working Group (AWG) was formed in 2007 with the mandate of using images and illustrations to complement the written codes and definitions available within the IPCCC. The AWG became operational in 2010 with the development of a process and web presence (ipccc-awg.net) that strives to identify, certify, and display images in all formats, of the best available phenotypes that illustrate the list of terms of the IPCCC. Therefore, the purpose of this chapter is to discuss the process followed by the AWG and the lessons learned on how to develop a comprehensive system of illustrations of nomenclatures. This effort is in keeping with the ultimate goal of the authors, in promoting the use of a universal system of nomenclature that facilitates the large scale adoption of pediatric cardiac databases. The long term effect will be to improve communications between all pediatric cardiac specialties and promote continued advancements in the diagnosis and treatment of the neonate, infant, child, and young adult with the various forms of pediatric and congenital cardiac disease.

Historical Background

Hippocrates of Kos (460–370 BCE) is credited with being the first reject the widely held belief of the divine origin of disease and to argue for the practice of medicine based on observation and rational thought. "On Fractures" as well as in other treatises and part of the Hippocratic Corpus (a collection of writings ascribed to Hippocrates but with likely contributions by others), reflected a significant body of knowledge of anatomy that could only be gained by direct observations on the human body [2, 3]. In Hellenistic Alexandria, Herophilos (280 BCE) began the systematic use of dissections on human cadavers to study anatomy; and as this knowledge advanced, illustrations were used to help in the teaching of this new found knowledge [4–6]. Galen (129–200 CE), the most famous physician of Roman times, and personal doctor to the emperors Marcus Aurelius, Commodus and Septimus Severus, was born in Pergamun in Asia Minor. Galen was the intellectual heir to the Greek traditions of medicine, wrote profusely on many subjects, including philosophy, and understood the relationship between anatomy and physiology. Unfortunately, due to the Roman prohibition of human dissections, Galen performed his anatomical studies almost exclusively on animals, principally the pig and Barbary ape. He wrote many of his works on the various aspects of medicine, and by some accounts his literary output was in the order of as many as 400–600 manuscripts. Some of his works were illustrated to complement his concepts and descriptions of health and disease, based on the Hippocratic humoral theory of disease. Sadly most of his manuscripts were destroyed in a fire a few years before his death [7, 8]. The assumptions made by Galen in conjunction with the social and religious changes that occurred after the fall of the Western Roman Empire went unchallenged until the Renaissance and the advent of the rebirth in the study of human diseases and their classification.

It is suggested that the great Renaissance artists such Raphael and Michelangelo performed their own dissections. However, it was Leonardo da Vinci (1452–1519 CE) who, in order to understand and improve his renditions of the human body, performed as many as thirty (30) dissections in his study of human anatomy. Nevertheless it is understood that da Vinci's interest was primarily artistic, a means to 'perfect' the anatomical detail of his paintings and sculptures [6]. These innovations by Leonardo da Vinci, paved the way for Andreas Vesalius (1514–1564 CE) epic study of the human body, 'De Humani Corporis Fabrica Libri Septem' (1543). This document is likely one of the most important and influential anatomical medical works ever published, because it not only reflected Vesalius' careful, direct observations based on his own dissections, but also on the artistry of the 186 illustrations that accompanied his descriptions. Vesalius' observations rejected and in other cases confirmed many of Galen's

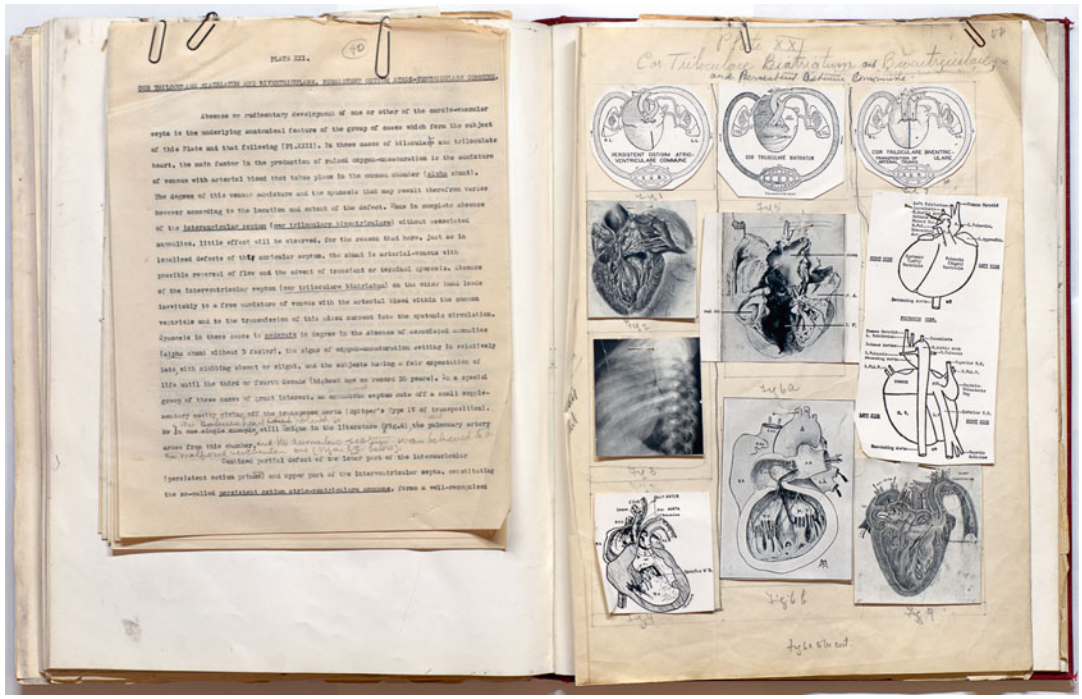

Fig. 7.1 This is a photograph of a draft copy of Maude Abbott's Atlas that was given by Lois Hawkins of the Division of Paediatric Cardiology in Edmonton, Canada to the Osler Library at McGill University in 2008 (Reproduced by permission of the Osler Library of the History of Medicine, McGill University)

observation and theories. For his investigations and publications, Vesalius received both condemnation and praise. He became court physician to Charles V, Holy Roman Emperor, and traveled throughout many parts of Europe. He died in 1564 following a shipwreck on the Greek island of Zante, after returning from a pilgrimage to Jerusalem, and on his way to Padua where he had been appointed to a prestigious chair in anatomy [5, 7–10].

The seventeenth and eighteenth centuries saw an increase in the interest of defining and illustrating the diverse forms of cardiac disease and saw anecdotal contributions from well known figures in the history of medicine such as LeCat, Morgagni and Hunter. The nineteenth century saw contributions by Farre, Gintrac, Meckel and Paget, as well as Fallot [11]. In 1858, Thomas Peacock published his book 'On Malformations of the Human Heart', where he outlined a system of classification based on cardiac anatomy and embryology. This book was the outgrowth of a series of lectures given to students at St. Thomas Hospital [12]. Then, in 1875, Carl von Rokitansky published his 'Defects of the Cardiac Septa' (Die Defekte der Scheidewände des Herzens) [13]. As curator of the McGill Medical Museum and inspired by Sir William Osler, Maude Abbott, in 1901, published an article in the Montreal Medical Journal of a congenitally malformed heart given to McGill University by Andrew Holmes, one of the founders of the McGill Medical School, in 1823. Abbott's work with congenitally malformed hearts proved to be the most important aspect of her academic career [14]. In 1936, she published the Atlas of Congenital Cardiac Diseases (Fig. 7.1), which consisted of 75 pages with 25 illustrated plates, grouped under her system of classification [15]. Dr. Abbott's work can be considered the "first systematic classification of congenital cardiac lesions" [16]. In 1947, Helen Taussig published her two volume book "Congenital Malformations of the Heart" [11]. The last half of the twentieth

century saw the contributions to the understanding of congenital cardiac morphology by Maurice Lev, Jesse Edwards, Richard and Stella Van Praagh, Robert Anderson and Anton Becker, among others. At this time, systems of classification were neither uniform nor universal, but Van Praagh and Van Praagh and coworkers, as well as Anderson and colleagues, proposed different systems of classification and organization based on a segmental approach. These two systems were in some ways similar, but they were different enough that two competing systems developed and were used separately or in combination by different practitioners and institutions [17].

In a parallel effort, the pediatric cardiac surgeons were also developing additional systems of nomenclature for use in the coding of cardiac surgical diagnoses and procedures. Fortunately, by the first decade of the twenty-first century, developments within the pediatric cardiology and pediatric cardiac surgical community led to the recognition that a universal and comprehensive system of nomenclature was within reach. In 2000, representatives from The Association for European Paediatric Cardiology (AEPC), The Society of Thoracic Surgeons (STS), and The European Association for Cardio-Thoracic Surgery (EACTS), as well as other societies and entities, agreed to establish The International Nomenclature Committee for Pediatric and Congenital Heart Disease, which later became The International Society for Nomenclature of Paediatric and Congenital Heart Disease (ISNPCHD). In 2001, at The First International Summit on Nomenclature for Pediatric and Congenital Heart Disease held at The Third World Congress of Pediatric Cardiology and Cardiac Surgery in Toronto, Canada, the Nomenclature Working Group (NWG) of The International Nomenclature Committee for Pediatric and Congenital Heart Disease was established. The initial goal of the NWG was to work in partnership and produce a reconciliatory bidirectional map between the predominant nomenclature systems for pediatric and congenital cardiac disease; this bidirectional map and system of nomenclature was ultimately named The International Pediatric and Congenital Cardiac Code (IPCCC) [16]. By 2005, the NWG had nearly completed the cross-map and presented the results at The Second International Summit on Nomenclature for Pediatric and Congenital Heart Disease held at The Fourth World Congress of Pediatric Cardiology and Cardiac Surgery in Buenos Aires, Argentina. In 2009, an updated version of the IPCCC was presented at The Third International Summit on Nomenclature for Pediatric and Congenital Heart Disease held at The Fifth World Congress of Pediatric Cardiology and Cardiac Surgery in Cairns, Australia. In 2013, an additional updated version of the IPCCC was presented at The Fourth International Summit on Nomenclature for Pediatric and Congenital Heart Disease held at The Sixth World Congress of Pediatric Cardiology and Cardiac Surgery in Cape Town, South Africa.

In 2007, in a parallel development, the ISNPCHD created two additional Working Groups, so that the ISNPCHD now has the following three committees or working groups [18–25]:

1. The Nomenclature Working Group (NWG) was created in 2001 and is the oldest and original working group of the ISNPCHD. The purpose of the NWG is to maintain, develop, expand, update, and preserve the IPCCC. It also has the ancillary responsibility to provide ready access to the IPCCC for the global pediatric and congenital cardiology and cardiac surgery communities as well as related disciplines and interested parties and individuals. The IPCCC is available free of charge from the Internet at http://www.ipccc.net/.
2. The Definitions Working Group (DWG) was created in 2007. The purpose of the DWG is to write definitions for the terms in the IPCCC, building on the previously published definitions from the Nomenclature Working Group.
3. The Archiving Working Group (AWG) was also created in 2007. The purpose of the AWG is to link images and videos of all types to illustrate the terms and definitions of the IPCCC. The images and videos may be from cardiac morphologic specimens as well as a variety of other sources including

echocardiograms, angio-cardiograms, computerized axial tomographic images, magnetic resonance images, and intra-operative images and videos.

Illustration of the Terms of the IPCCC

In the effort to illustrate, with representative images, the terms and definitions of the IPCCC, the AWG has developed and maintains an active web presence known as the Archiving Working Group Web Portal, which may be accessed at http://www.ipccc-awg.net/. The remainder of this chapter will review the organization of the AWG Web Portal and the lessons that have been learned in the process of promoting and illustrating the lists of terms and definitions that compose the IPCCC.

1. **AWG Organization:**
   - 1.1. AWG Workflow: The workflow structure follows a peer-reviewed process. The identification of images, with accompanying textual explanations, may be solicited or unsolicited. The members of the AWG project (Table 7.1), and in particular the Senior Archivist, share in the responsibility of identifying the images that illustrate the codes and definitions of the IPCCC. The Senior Archivist and the three Co-Chairpersons of the AWG work closely during the initial review and assignment of the codes and definitions to the images identified and submitted. After the process is completed, a web page is created or modified, and the images, codes, definitions, and explanatory text are posted to the internet presence of the AWG called the AWG Web Portal. The initial publication of the images to the AWG Web Portal is posted with the label: "Pending' certification".
   - 1.2. Review Process: On a periodic basis, typically every other month, the members of the AWG review the posted images, codes, and text for accuracy, quality, and suitability. The typical review process is carried out trans-telephonically using an international call center and a specially created; internet based, closed 'wiki' [26]. The participants of the conference are able to view the images and posted comments on their computers while discussing the images and terms by telephone. The suggestions are incorporated into the 'wiki' concurrently in real time

Table 7.1 Members of the Archiving Working Group

Co-chairpersons
Vera D. Aiello (Cardiac Pathologist, Brazil)
Robert H. Anderson (Cardiac Morphologist, UK and USA)
Jorge M. Giroud (Pediatric Cardiologist, USA)
ISNPCHD executive committee
Rodney C. G. Franklin (Pediatric Cardiologist, UK) (President, NWG Co-Chair)
Jeffrey P. Jacobs (CV Surgeon, USA) (Vice President, NWG Co-Chair)
Christo I. Tchervenkov (CV Surgeon, Canada) (Past President)
Marie J. Béland (Pediatric Cardiologist, Canada) (NWG Co-Chair)
Steven D. Colan (Pediatric Cardiologist, USA) (DWG Co-Chair)
Henry Walters III (CV Surgeon, USA) (DWG Co-Chair)
Editorial members
Carl Backer (CV Surgeon, USA)
Frederique Bailliard (Pediatric Cardiologist, USA)
Meryl Cohen (Pediatric Cardiologist, USA)
Andrew Cook (Cardiac Morphologist, UK)
Allen D. Everett (Pediatric Cardiologist, USA)
J. William Gaynor (CV Surgeon, USA)
Lucile Houyel (Pediatric Cardiologist, France)
Marina Hughes (Pediatric Cardiologist/MRI, UK)
Marshall L. Jacobs (CV Surgeon, USA)
Amy Juraszek (Pediatric Cardiologist, USA)
Otto N. Krogmann (Pediatric Cardiologist, Germany)
Hiromi Kurosawa (CV Surgeon, Japan)
Leo Lopez (Pediatric Cardiologist, USA)
James St. Louis (CV Surgeon, USA)
Bohdan Maruszewski (CV Surgeon, Poland)
Charles Shepard (Pediatric Cardiologist, USA)
Giovanni Stellin (CV Surgeon, Italy)
Paul M. Weinberg (Pediatric Cardiologist/ Morphologist, USA)
Senior Archivist
Diane Spicer (Cardiac Morphologist, USA)

Fig. 7.2 Home (Landing) Page of the AWG Web Portal (http://ipccc-awg.net). This figure illustrates the homepage and gives a brief overview of the AWG Web Portal. Please note the Navigation Bar that gives the user the capacity to review and image or video, submit an image or video, or view other features of the AWG Web Portal such as our sponsors and membership of the AWG. This home page is linked to the website of The International Society for Nomenclature of Paediatric and Congenital Heart Disease (ISNPCHD): http://www.ipccc.net/

and immediately updated. This process ensures a timely and simultaneous review of the images linked to the corresponding terms and definitions of the IPCCC. After the evaluation of the posted image is completed, the images with the accompanying web page are officially certified and rated on a scale of 1–4 hearts, with four hearts being a superb example of the phenotype encoded by the IPCCC term. The web page is updated with the modifications, date of certification, and rating, in order to reflect the date of final approval.

1.3. AWG Web Portal Navigation: The IPCCC is organized in a hierarchal structure and is composed of Long Lists of nomenclature containing thousands of terms that are mapped to Short Lists of nomenclature that contain hundreds of terms. The navigation of the web site is based on the use of the IPCCC Short Lists. To navigate the web site, the user selects from drop-down menus to reach the areas of interest (Fig. 7.2). The user clicks on the image and code of interest to access the web page, where the codes, images, and explanatory texts are displayed (Figs. 7.3 and 7.4). The images and videos reflect a variety of modalities, including still images and videos of from cardiac morphologic specimens, echocardiograms, angio-cardiograms, computerized axial tomographic images, magnetic resonance

Fig. 7.3 This is an example of the first portion of a finished page with images, codes, and, when available, definitions and comments. The images for this page are shown in Fig. 7.4

images, and intra-operative images and videos. These still images and videos are stored and displayed using Web enabled standard formats such as JPEG (Joint Photographic Expert Group) for still images and Flash animation (Adobe Systems, San Jose, California, USA) for video clips.

1.4. Copyright Protection: An important feature of the AWG Web Portal is that the contributing author retains all copyrights to the images and has given permission for the portal visitor to view the images and to use them for not-for-profit, instructional, or educational purposes only. If the AWG Web Portal visitor wishes to use the images displayed for publications or for-profit-use, the visitor is free to contact the contributor of the image, who is identified and credited in each of the web pages posted, to obtain the necessary permissions.

2. **Lessons Learned**:

2.1. Senior Archivist: In spite of best intentions, unsolicited images with descriptions have been a minor portion of the images published in this 'virtual web-based encyclopedia' to date. This represents the most important lesson learned so far: the professional identification, photography and labeling of cardiac images is an 'absolute' necessity for a project of illustration of terms of classifications to be successful. Although not glamorous by current standards, cardiac morphologic specimens remain the

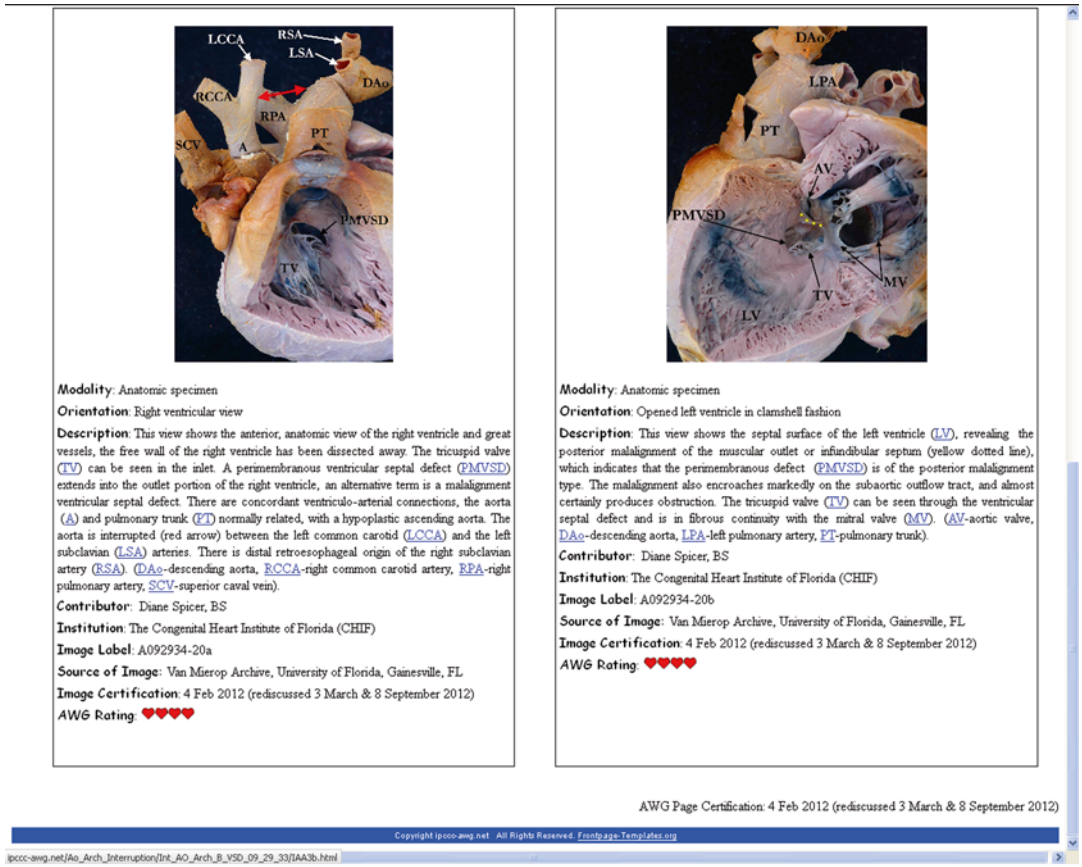

Fig. 7.4 This figure illustrates the images associated with the terms and definitions of the IPCCC shown in Fig. 7.2. Any of the images displayed, if selected, link to the full-resolution version. This allows for review of the image in greater detail or downloads for not-for-profit use. Please note the 'Certification' and 'AWG Rating' status listed for each image. The date of certification or review is added to the bottom of the page as well. This is done after the review process is completed by the AWG Editorial Board

cornerstone for the illustration of the definitions and terms of the IPCCC. In spite of increasing technological wonders, such as three dimensional reconstructions by magnetic resonance imaging (MRI) or echocardiography, a well photographed and labeled cardiac morphologic specimen continues to be an essential component of any 'encyclopedia' that wishes to illustrate terms and definitions of cardiac malformations. In a project such as the AWG Web Portal, it is of paramount importance to have a well educated and experienced Senior Archivist that is versed in the current embryologic and morphologic debates as well as the essential techniques of dissection and photography, and this experience is reflected in the quality of morphologic illustrations used in the AWG Web Portal.

2.2. Webmaster: It has been our experience that a technologically experienced member or 'Webmaster' must be identified and given the support necessary to create and maintain the process. This is a crucial component of any project that seeks to popularize the linkage of illustrations with terms using the Internet as the publishing medium. Technological familiarity is important but most be complemented with knowledge of congenital cardiac disease. This collaboration of this

Webmaster and the Senior Archivist is the key that helps in the creation of a web based, technological platform that reflects the current understanding of the meanings inherent in the terms and the illustration of the accompanying images.

2.3. Membership: Another crucial aspect in the organizational structure of a project that seeks to link images with list of terms is that the best available images that represent the phenotypes must not only be identified but also reviewed by a group of knowledgeable practitioners that are recognized within the field. The responsibility of the members is to insure that the images collected and linked are indeed the best examples of the terms and definitions listed in the posted pages. The international nature of such a project demands that the certifying members represent the global perspective of the modern standard of pediatric cardiac classification. Additionally, as in daily clinical practice, areas of particular expertise must be recognized and utilized in the process of identification, review, and certification. For example, with over 9,000 diagnostic terms in the EACTS-STS version of the IPCCC, and a similar number in the AEPC version of the IPCCC, it is imperative that an 'expert' for each of the coding systems be available for each review and certification session. In a similar vein, it is required that a cardiac morphologist participate in each review and certification session. We find that this participation significantly improves the accuracy and utility of each posted image and page. Additionally, at least one cardiac surgeon must participate in each session to complement the expertise of the morphologists and coding 'experts'. Finally, if the image that is being reviewed is that of a complex three dimensional reconstruction either by echocardiography or MRI, the participation of a member of the AWG that is familiar with these techniques is also required.

2.4. Funding: A project that seeks to illustrate terms with images using technology must be adequately funded in order to maintain its web presence. The domain name must be registered and maintained. The images and web pages must be hosted with enough capacity to both support traffic as well storage of images and text. The Senior Archivist must be funded in order to identify and photograph available morphologic specimens and help with the linkage of the diverse images identified and described. The telephony costs for the review and certification sessions must also be incorporated into the budget. The approach followed by the AWG for the AWG Web Portal is to allow full access with no cost incurred by the user or viewer. To date the AWG has been successful in identifying and obtaining funding for the project by grants and donations, in particular from The Children's Heart Foundation: http://www.childrensheartfoundation.org/. A research grant from The Children's Heart Foundation (Table 7.2) was critical to the development of the AWG Web Portal.

Summary

The effort to develop a web based platform to illustrate, with representative images and videos, the terms and definitions of The International Pediatric and Congenital Cardiac Code has been successfully implemented by the members of the Archiving Working Group (AWG) of The International Society for Nomenclature of Paediatric and Congenital Heart Disease (ISNPCHD). The AWG maintains an active web presence known as the Archiving Working Group Web Portal. This 'virtual encyclopedia' combines the tools of classification incorporated into the lists of terms of the IPCCC, with the ancient tradition of using illustrations to help in the understanding of diseases of the heart. The goal of this chapter, 'Illustrating Terms in Lists of Nomenclature', guided by the modern

Table 7.2 The Children's Heart Foundation grant

Period of award: January 1, 2011 to December 31, 2012	
Title of project:	
Creation of a visual encyclopedia illustrating the terms and definitions of the International Paediatric and Congenital Cardiac Code, a system of nomenclature developed by The International Society for Nomenclature of Paediatric and Congenital Heart Disease	
Granting agency: the Children's Heart Foundation	
Amount of funding = $100,000 over 2 years	
Name of principal investigator: Jeffrey Phillip Jacobs, MD, FACS, FACC, FCCP	
Investigative team:	
Jeffrey Phillip Jacobs, MD, FACS, FACC, FCCP	Principal investigator
Jorge Manuel Giroud, MD, FACC	Co- principal investigator
Robert Anderson	Co-investigator
Marshall Lewis Jacobs, MD	Co-investigator
Hal Walters, MD	Co-investigator
Diane Spicer, BS	Senior Archivist
Tracey Griffith, LPN	Research nurse coordinator

understanding of the various forms of cardiac disease that affect neonates, infants, children and increasingly the young adult, has been effectively implemented by the Archiving Working Group of the International Society for Nomenclature of Paediatric and Congenital Heart Disease. Please visit us at http://www.ipccc-awg.net/.

References

1. Falagas ME, Zarkadoulia EA, Samonis G. Arab science in the golden age (750–1258 C.E.) and today. FASEB J. 2006;20(10):1581–6.
2. McRae C. Fathers of biology. London; 1890.
3. Orfanos CE. Hippocrates to modern medicine. J Eur Acad Dermatol Venereol. 2007;21(6):852–8.
4. von Staden H, editor trans. Herophilos: the art of medicine in early Alexandria. Cambridge University Press; 1989.
5. Loechel WE. The history of medical illustration. Bull Med Libr Assoc. 1960;48(2):168–71.
6. McFall KJ. A critical account of the history of medical photography in the United Kingdom. IMI fellowship submission, June 2000. Available from http://www.migroup.co.uk/. Accessed 11 Jan 2013 at 17:48.
7. Aufderheide AC. The scientific study of mummies. Cambridge University Press; 2003. p 5.
8. Dunn PM. Galen (AD 129–200) of Pergamun: anatomist and experimental physiologist. Arch Dis Child Fetal Neonatal Ed. 2003;88:F441–3.
9. U.S. National Library of Medicine. Vesalius: De Humani Corporis Fabrica Libri Septem. Available from http://www.nlm.nih.gov/exhibition/historicalanatomies/vesalius_bio.html. Accessed 26 Jan 2013 at 13:22.
10. National Library of Medicine. De Humani Corporis Fabrica Libri Septem. Available from http://archive.nlm.nih.gov/proj/ttp/flash/vesalius/vesalius.html. Accessed 26 Jan 2013 at 13:22.
11. Rashkind WJ. Pediatric cardiology: a brief historical perspective. Pediatr Cardiol. 1979;1(1):63–71.
12. Porter IH. The nineteenth-century physician and cardiologist Thomas Bevill Peacock (1812–82). Med Hist. 1962;6(3):240–54.
13. Rokitansky C. Die Defekte der Scheidewände des Herzens. Wien: Braumüller; 1875.
14. Fraser R. Maude Abbott and the "atlas of congenital cardiac disease". Cardiovasc Pathol. 2006;15:233–5.
15. [No author listed]. Dr. Maude Abbott's atlas of congenital cardiac disease. Can Med Assoc J. 1936;34(2):194–5.
16. Franklin RCG, Jacobs JP, Krogmann ON, et al. Nomenclature for congenital and paediatric cardiac disease historical perspectives and the International Pediatric and Congenital Cardiac Code. Cardiol Young. 2008;18 Suppl 2:70–80.
17. Freedom RM, Lock J, Bricker JT. Pediatric cardiology and cardiovascular surgery: 1950–2000. Circulation. 2000;102:IV-58–68.
18. Giroud JM, Jacobs JP, Spicer D, Backer C, Martin GR, Franklin RCH, Béland MJ, Krogmann ON, Aiello VD, Colan SD, Everett AD, Gaynor JW, Kurosawa H, Maruszewski B, Stellin G, Tchervenkov CI, Walters III HL, Weinberg P, Anderson RH, Elliott MJ. Report from the International Society for Nomenclature of Paediatric and Congenital Heart Disease: creation of a visual encyclopedia illustrating the terms and definitions of the International Pediatric and Congenital Cardiac Code. World J Pediatr Congenit Heart Surg. 2010;1:300–13. doi:10.1177/2150135110379622.
19. Giroud JM, Jacobs JP, Fricker FJ, Spicer D, Backer C, Franklin RCH, Béland MJ, Krogmann ON, Aiello VD, Colan SD, Everett AD, Gaynor JW, Kurosawa H, Maruszewski B, Stellin G, Tchervenkov CI, Walters III HL, Weinberg P, Fogel MA, Jacobs ML, Elliott MJ, Anderson RH. Web based global virtual museum of congenital cardiac pathology. Prog Pediatr Cardiol. 2012;33(1):91–7. doi:10.1016/j.ppedcard.2011.12.015. In: Lipshultz SE, Barach P, Jacobs JP, Laussen P, editors. Progress in pediatric cardiology: the future of pediatric and congenital cardiac care special part 2. 2012;33(1):1–101.
20. Giroud JM, Aiello VD, Spicer DE, Anderson RH. The Archiving Working Group of the International Society

for Nomenclature of Paediatric and Congenital Heart Disease: a visual encyclopedia illustrating the terms and definitions of the International Paediatric and Congenital Cardiac Code. Congenit Cardiol Today. 2012;10(8):8–10.
21. Aiello VD, Anderson RH, Giroud JM, Spicer DE. Image of the month (aortic valve pathology, bicuspid and pulmonary valve pathology, bicuspid) – August 2012 – Presented by The Archiving Working Group. Congenit Cardiol Today. 2012; 10(8):14–5.
22. Anderson RH, Aiello VD, Spicer DE, Jacobs JP, Giroud JM. Image of the month #2 (interrupted aortic arch (IAA), Type B2 (interruption between the carotid and subclavian arteries with both subclavian arteries arising from the aorta distal to the interruption)) – October 2012 – Presented by The Archiving Working Group. Congenit Cardiol Today. 2012;10(10):20–1.
23. Aiello VD, Spicer DE, Jacobs JP, Giroud JM, Anderson RH. Image of the month #3 (total anomalous pulmonary venous connection (TAPVC), Type 4 (mixed)) – December 2012 – Presented by The Archiving Working Group. Congenit Cardiol Today. 2012;10(12):8–9.
24. Spicer DE, Jacobs JP, Giroud JM, Anderson RH, Aiello VD. Image of the month #4 – February 2013 (single ventricle, DILV) – Presented by The Archiving Working Group. Congenit Cardiol Today. 2013;11(2):13–4.
25. Jacobs JP, Giroud JM, Anderson RH, Aiello VD, Spicer DE. Image of the month #5 (VSD, Type 2 (perimembranous) (paramembranous)) – May 2013 – Presented by The Archiving Working Group. Congenit Cardiol Today. 2013;11(5):8–9.
26. WIKI: a web site that allows visitors to make changes, contributions, or corrections. Available from http://www.merriam-webster.com/dictionary/wiki. Accessed 31 Jan 2013 at 16:44.

Part III

Databases

8 Databases for Assessing the Outcomes of the Treatment of Patients with Congenital and Pediatric Cardiac Disease: The Perspective of Cardiac Surgery

Jeffrey P. Jacobs

Abstract

This chapter discusses the historical aspects, current state of the art, and potential future advances in the areas of nomenclature and databases for the analysis of surgical outcomes of treatments for patients with congenitally malformed hearts. We will consider the current state of analysis of outcomes, lay out some principles which might make it possible to achieve life-long monitoring and follow-up using our databases, and describe the next steps those involved in the care of these patients need to take in order to achieve these objectives.

In order to perform meaningful multi-institutional analyses of outcomes, any database must incorporate the following seven essential elements: (1) Use of a common language and nomenclature, (2) Use of a database with an established uniform core dataset for collection of information, (3) Incorporation of a mechanism of evaluating case complexity, (4) Availability of a mechanism to assure and verify the completeness and accuracy of the data collected, (5) Collaboration between medical and surgical subspecialties, (6) Standardization of protocols for life-long follow-up (7) Incorporation of strategies for quality assessment and quality improvement.

During the 1990s, both The European Association for Cardio-Thoracic Surgery and The Society of Thoracic Surgeons created databases to assess the outcomes of congenital cardiac surgery. Beginning in 1998, these two organizations collaborated to create the International Congenital Heart Surgery Nomenclature and Database Project. By 2000, a common

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nomenclature, along with a common core minimal dataset, were adopted by The European Association for Cardio-Thoracic Surgery and The Society of Thoracic Surgeons, and published in the *Annals of Thoracic Surgery*. In 2000, The International Nomenclature Committee for Pediatric and Congenital Heart Disease was established. This committee eventually evolved into the International Society for Nomenclature of Paediatric and Congenital Heart Disease. The original working component of this international nomenclature society was The International Working Group for Mapping and Coding of Nomenclatures for Pediatric and Congenital Heart Disease, also known as the Nomenclature Working Group. By 2005, the Nomenclature Working Group crossmapped the nomenclature of the International Congenital Heart Surgery Nomenclature and Database Project of The European Association for Cardio-Thoracic Surgery and The Society of Thoracic Surgeons with the European Paediatric Cardiac Code of the Association for European Paediatric Cardiology, and therefore created the International Pediatric and Congenital Cardiac Code (IPCCC), which is available for free download from the internet at [http://www.IPCCC.NET]. This common nomenclature, the International Pediatric and Congenital Cardiac Code, and the common minimum database data set created by the International Congenital Heart Surgery Nomenclature and Database Project, are now utilized by both The European Association for Cardio-Thoracic Surgery (EACTS), The Society of Thoracic Surgeons (STS), and The Japan Congenital Cardiovascular Surgery Database (JCCVSD). As of January 1, 2014, the STS Congenital Heart Surgery Database contains data from 292,828 operations, the EACTS Congenital Heart Surgery Database contains data from over 157,772 operations, and the JCCVSD contains data from over 29,000 operations. Therefore, the combined dataset of the STS Congenital Heart Surgery Database, the EACTS Congenital Heart Surgery Database, and the JCCVSD contains data from over 479,000 operations performed between 1998 and January 1, 2014 inclusive, all coded with the EACTS-STS derived version of the IPCCC, and all coded with identical data specifications.

Three major multi-institutional efforts have attempted to measure the complexity of congenital cardiac surgical operations: **R**isk **A**djustment in **C**ongenital **H**eart **S**urgery-1 methodology (RACHS-1 method), **A**ristotle **B**asic **C**omplexity Score (ABC Score), and **STS**-E**A**C**T**S Congenital Heart Surgery Mortality Categories (STS-EACTS Mortality Categories) (STAT Mortality Categories). RACHS-1 and the ABC Score were developed at a time when limited multi-institutional clinical data were available and were therefore based in a large part on subjective probability (expert opinion). The STAT Mortality Categories are a tool for complexity stratification that was developed from an analysis of 77,294 operations entered into the EACTS Congenital Heart Surgery Database (33,360 operations) and the STS Congenital Heart Surgery Database (43,934 patients) between 2002 and 2007. Procedure-specific mortality rate estimates were calculated using a Bayesian model that adjusted for small denominators. Operations were sorted by increasing risk and grouped into five categories (the STS–EACTS Congenital Heart Surgery Mortality Categories) that

were designed to be optimal with respect to minimizing within-category variation and maximizing between-category variation. STS and EACTS have transitioned from the primary use of Aristotle and RACHS-1 to the primary use of the STAT Mortality Categories.

Collaborative efforts involving The European Association for Cardio-Thoracic Surgery and The Society of Thoracic Surgeons are under way to develop mechanisms to verify the completeness and accuracy of the data in the databases. Under the leadership of The MultiSocietal Database Committee for Pediatric and Congenital Heart Disease, further collaborative efforts are ongoing between congenital and pediatric cardiac surgeons and other subspecialties, including pediatric cardiac anesthesiologists, via The Congenital Cardiac Anesthesia Society, pediatric cardiac intensivists, via The Pediatric Cardiac Intensive Care Society, and pediatric cardiologists, via the Joint Council on Congenital Heart Disease and The Association for European Paediatric Cardiology. Analysis of outcomes must move beyond mortality, and encompass longer term follow-up, including cardiac and noncardiac morbidities, and importantly, those morbidities impacting health related quality of life. Methodologies must be implemented in these databases to allow uniform, protocol driven, and meaningful, long term follow-up and quality improvement.

Keywords

Database • Outcomes • Quality • Pediatric cardiac surgery • Congenital cardiac surgery

Introduction

Although significant progress has been made in the care of patients with pediatric and congenital cardiac disease, complications and death still occur. As a result, optimization of outcomes remains a constant goal. Substantial efforts has been devoted to advancing the science of assessing the outcomes and improving the quality of care associated with the treatment of patients with pediatric and congenital cardiac disease [1–227]. The importance of these efforts is supported by the fact that congenital heart defects are the most common birth anomalies, with moderate to severe variants occurring in approximately 6 per 1,000 live births [228].

In order to perform meaningful multi-institutional outcomes analyses and quality improvement, any database must incorporate the following seven essential elements:

1. **Use of a common language and nomenclature** [1–52, 54, 55, 62–64, 66–71, 75, 77, 79, 81, 82, 87, 88, 93, 94, 96, 100, 103, 104, 110–112, 114–116, 128–140, 148, 152, 155, 162, 167–169, 171, 172, 178, 179, 188, 191, 200–202, 209, 210, 213, 216, 218, 221]
2. **Use of a database with an established uniform core dataset for collection of information** [1–23, 55, 58–60, 63, 64, 71, 77, 79–82, 87, 88, 90, 93, 95, 98, 100, 104–106, 110–113, 115, 117–123, 145, 146, 148, 152–155, 161, 163, 164, 171, 172, 174, 178, 179, 185, 188, 189, 204, 207, 210, 212, 214, 216, 220–227]
3. **Incorporation of a mechanism of evaluating case complexity** [56, 57, 61, 65, 72–74, 76–79, 81–84, 88–91, 97–102, 104, 106, 107, 110–112, 124, 125, 141, 142, 147–150, 152, 178, 179, 188, 204, 215–217, 221]
4. **Availability of a mechanism to assure and verify the completeness and accuracy of the data collected** [77, 81, 85, 86, 88, 100, 104, 110–112, 126, 148, 152, 178, 179, 188, 216, 221]

5. **Collaboration between medical and surgical subspecialties** [81, 100, 104, 110–140, 148, 152, 178, 179, 188, 216, 221]
6. **Standardization of protocols for life-long follow-up** [104, 109–112, 127, 145, 146, 152, 164, 173, 178, 179, 184, 188, 189, 214, 216, 221]
7. **Incorporation of strategies for quality assessment and quality improvement** [108, 110, 115, 143–148, 151, 152, 154, 156–160, 165–167, 170, 175–183, 186–188, 190, 192–199, 203, 205, 206, 208, 210, 211, 216, 219, 221, 222]

The foundation of these seven elements is the use of a common language and nomenclature. The remaining six elements are all dependent on this nomenclature; and therefore, quality improvement in the domain of congenital cardiac disease depends on a solid understanding of cardiac morphology and nomenclature.

Events at Bristol, England [229], Denver, Colorado, United States of America [230–236], Winnipeg, Canada [237], Mid Staffordshire, England [238] and Lexington, Kentucky, United States of America [239] have clearly demonstrated the importance of clinically driven analysis of outcomes. For example, the Bristol Report presents the results of the inquiry into the management of the care of children receiving complex cardiac surgical services at the Bristol Royal Infirmary between 1984 and 1995 and relevant related issues. Approximately 200 recommendations are made, many of which relate to the need for accurate multi-institutional outcomes databases to quantitate outcomes of care rendered to patients with congenital cardiac disease. Perhaps less well-known than the Bristol Report, the Report of the Manitoba Pediatric Cardiac Surgery Inquest presents data from an inquest involving 12 children who died while undergoing, or soon after having undergone, cardiac surgery at the Winnipeg Health Sciences Centre in 1994. Clearly, these events demonstrate the importance of a meaningful and fair method of multi-institutional analysis of outcomes for congenital cardiac surgery.

Nomenclature

Substantial effort has been devoted to the standardization of nomenclature and definitions related to surgery for pediatric and congenital cardiac disease. During the 1990s, both The European Association for Cardio-Thoracic Surgery (EACTS) and The Society of Thoracic Surgeons (STS) created databases to assess the outcomes of congenital cardiac surgery. Beginning in 1998, these two organizations collaborated to create the International Congenital Heart Surgery Nomenclature and Database Project. By 2000, a common nomenclature and a common core minimal dataset were adopted by EACTS and STS and published in the *Annals of Thoracic Surgery* [21]. In 2000, The International Nomenclature Committee for Pediatric and Congenital Heart Disease was established. This committee eventually evolved into the International Society for Nomenclature of Paediatric and Congenital Heart Disease (ISNPCHD). By 2005, members of the ISNPCHD crossmapped the nomenclature of the International Congenital Heart Surgery Nomenclature and Database Project of the EACTS and STS with the European Paediatric Cardiac Code (EPCC) of the Association for European Paediatric Cardiology (AEPC), and therefore created the International Pediatric and Congenital Cardiac Code (IPCCC) [114], which is available for free download from the internet at [http://www.IPCCC.NET].

Most international databases of patients with pediatric and congenital cardiac disease use the IPCCC as their foundation. Two versions of the IPCCC are used in the overwhelming majority of multi-institutional databases throughout the world:

1. The version of the IPCCC derived from the nomenclature of the International Congenital Heart Surgery Nomenclature and Database Project of the EACTS and the STS
2. The version of the IPCCC derived from the nomenclature of the EPCC of the AEPC

These two versions of the IPCCC are also often referred to with the following abbreviated short names:

1. EACTS-STS derived version of the IPCCC
2. AEPC derived version of the IPCCC

The STS Congenital Heart Surgery Database, the EACTS Congenital Heart Surgery Database, and The Japan Congenital Cardiovascular Surgery Database (JCCVSD) all use the EACTS-STS derived version of the IPCCC.

The ISNPCHD has published review articles which provide a unified and comprehensive classification, with definitions, for several complex congenital cardiac malformations: the functionally univentricular heart [92], hypoplastic left heart syndrome [94], discordant atrioventricular connections [96] and cardiac structures in the setting of heterotaxy [103]. These review articles include definitions and a complete listing of the relevant codes and terms in both versions of the IPCCC.

In collaboration with the World Health Organization (WHO), the ISNPCHD is developing the pediatric and congenital cardiac nomenclature that will be used in the eleventh version of the International Classification of Diseases (ICD-11). With a grant funded by The Children's Heart Foundation [http://www.childrensheart-foundation.org/], the ISNPCHD has also linked images and videos to the IPCCC. These images and videos are acquired from cardiac morphologic specimens and imaging modalities such as echocardiography, angiography, computerized axial tomography, and magnetic resonance imaging, as well as intraoperative images and videos [162, 191, 200–202, 209, 213, 218]. These images and videos are available for free download from the internet at [http://www.IPCCC-awg.NET]. The IPCCC itself is available for free download from the internet at [http://www.IPCCC.NET].

The EACTS-STS derived version of the IPCCC [110, 112, 114], and the common minimum database data set created by the International Congenital Heart Surgery Nomenclature and Database Project [208], are now utilized by the STS Congenital Heart Surgery Database, the EACTS Congenital Heart Surgery Database, and the JCCVSD. Between 1998 and January 1, 2014 inclusive, this nomenclature and database was used by STS, EACTS, and JCCVSD to analyze outcomes of 479,000 operations.

Several studies have examined the relative utility of clinical and administrative nomenclature for the evaluation of quality of care for patients undergoing treatment for pediatric and congenital cardiac disease. Evidence from four recent investigations suggests that the validity of coding of lesions seen in the congenitally malformed heart via 9th ICD Revision of the International Classification of Diseases (ICD-9) as used currently in administrative databases in the United States of America is poor [116, 210, 240, 241]. First, in a series of 373 infants with congenital cardiac defects at Children's Hospital of Wisconsin, investigators reported that only 52 % of the cardiac diagnoses in the medical records had a corresponding code from the ICD-9 in the hospital discharge database [240]. Second, the Hennepin County Medical Center discharge database in Minnesota identified all infants born during 2001 with a code for congenital cardiac disease using ICD-9. A review of these 66 medical records by physicians was able to confirm only 41 % of the codes contained in the administrative database from ICD-9 [241]. Third, the Metropolitan Atlanta Congenital Defect Program of the Birth Defect Branch of the Centers for Disease Control and Prevention of the United States government carried out surveillance of infants and fetuses with cardiac defects delivered to mothers residing in Atlanta during the years 1988 through 2003 [116]. These records were reviewed and classified using both administrative coding and the clinical nomenclature used in the Society of Thoracic Surgeons Congenital Heart Surgery Database. This study concluded that analyses based on the codes available in ICD-9 are likely to "have substantial misclassification" of congenital cardiac disease. Fourth, a study was performed using linked patient data (2004–2010) from the Society of Thoracic Surgeons Congenital Heart Surgery (STS-CHS) Database (clinical registry) and the Pediatric Health Information Systems (PHIS) database (administrative database) from hospitals participating in both in order to evaluate differential coding/classification of operations between datasets

and subsequent impact on outcomes assessment [210]. The cohort included 59,820 patients from 33 centers. There was a greater than 10 % difference in the number of cases identified between data sources for half of the benchmark operations. The negative predictive value (NPV) of the administrative (versus clinical) data was high (98.8–99.9 %); the positive predictive value (PPV) was lower (56.7–88.0 %). These differences translated into significant differences in outcomes assessment, ranging from an underestimation of mortality associated with truncus arteriosus repair by 25.7 % in the administrative versus clinical data (7.01 % versus 9.43 %; $p=0.001$) to an overestimation of mortality associated with ventricular septal defect (VSD) repair by 31.0 % (0.78 % versus 0.60 %; $p=0.1$). This study demonstrates differences in case ascertainment between administrative and clinical registry data for children undergoing cardiac operations, which translated into important differences in outcomes assessment.

Several potential reasons can explain the poor diagnostic accuracy of administrative databases and codes from ICD-9:

- Accidental miscoding
- Coding performed by medical records clerks who have never seen the actual patient
- Contradictory or poorly described information in the medical record
- Lack of diagnostic specificity for congenital cardiac disease in the codes of ICD-9
- Inadequately trained medical coders.

Although one might anticipate some improvement in diagnostic specificity with the planned adoption of ICD-10 by the United States, it is likely to still be far short from that currently achieved with clinical registries. (ICD-9 has only 29 congenital cardiac codes and ICD-10 has 73 possible congenital cardiac terms.) It will not be until there is implementation of the pediatric and congenital cardiac components of ICD-11 that harmonization of clinical and administrative nomenclature will be achieved with the resolution, therefore, of many of these challenging issues.

Database

The STS Congenital Heart Surgery Database is the largest database in North America dealing with congenital cardiac malformations [117, 152]. It has grown annually since its inception, both in terms of the number of participating centers submitting data, and the number of operations analyzed (Figs. 8.1, 8.2 and 8.3). As of January 1, 2014, the STS Congenital Heart Surgery Database currently has 111 Participating Centers representing 120 hospitals performing pediatric and congenital cardiac surgery in North America: 117 out of an estimated 125 centers from the United States of America that perform pediatric and congenital heart surgery and 3 out of centers 8 from Canada that perform pediatric and congenital heart surgery [95, 174]. (The Report of the 2005 STS Congenital Heart Surgery Practice and Manpower Survey, undertaken by the STS Workforce on Congenital Heart Surgery, documented that 122 centers in the United States of America perform pediatric and congenital heart surgery and 8 centers in Canada perform pediatric and congenital heart surgery [95]. The Report of the 2010 STS Congenital Heart Surgery Practice and Manpower Survey, undertaken by the STS Workforce on Congenital Heart Surgery, documented that 125 centers in the United States of America perform pediatric and congenital heart surgery and 8 centers in Canada perform pediatric and congenital heart surgery [174].)

The STS Congenital Heart Surgery Database therefore contains data from an estimated 93.6 % of hospitals (117 out of 125) performing pediatric cardiac surgery in the United States. With penetrance of over 90 %, the data in the STS Congenital Heart Surgery Database is representative of pediatric and congenital heart surgery in the United States of America. As of January 1, 2014, the number of cumulative total operations in the STS Congenital Heart Surgery Database is 292,828 [19]. The aggregate Participant Feedback Report from the Fall 2013 Harvest of the STS Congenital Heart Surgery Database includes 136,617 operations performed in the 4 year analytic window of July 1, 2009 to June 30, 2013,

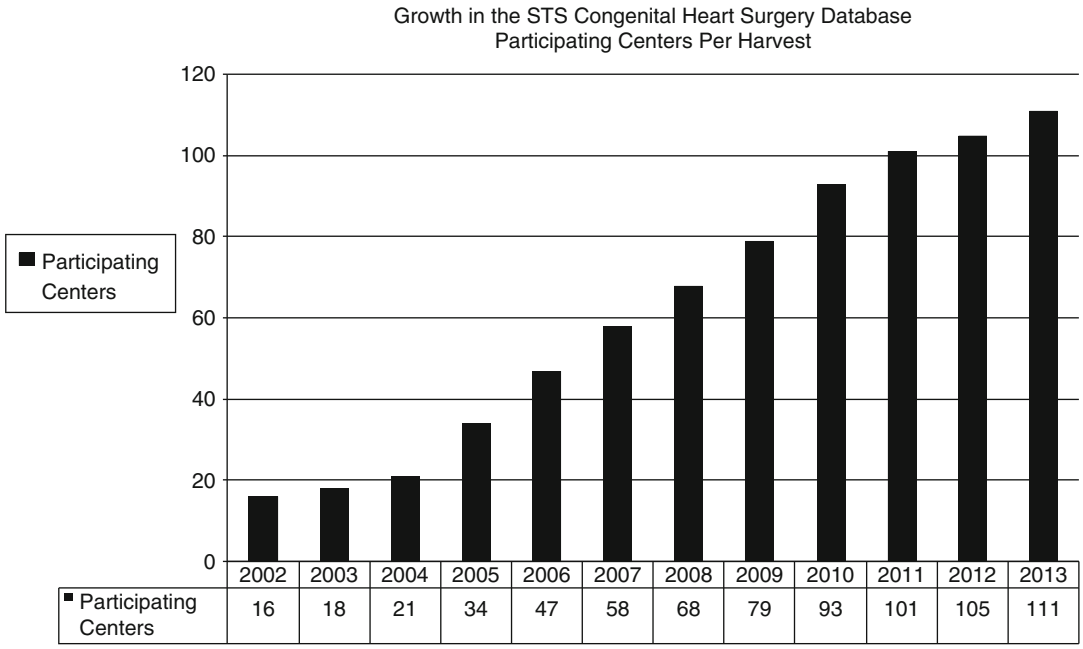


Fig. 8.1 The graph documents the annual growth of the STS Congenital Heart Surgery Database by number of participating centers submitting data. The aggregate report from the Fall 2013 Harvest of the STS Congenital Heart Surgery Database [19] includes data from 111 North American Congenital Database Participants representing 120 Congenital Heart Surgery hospitals in the North America, 117 in the United States of America and 3 in Canada

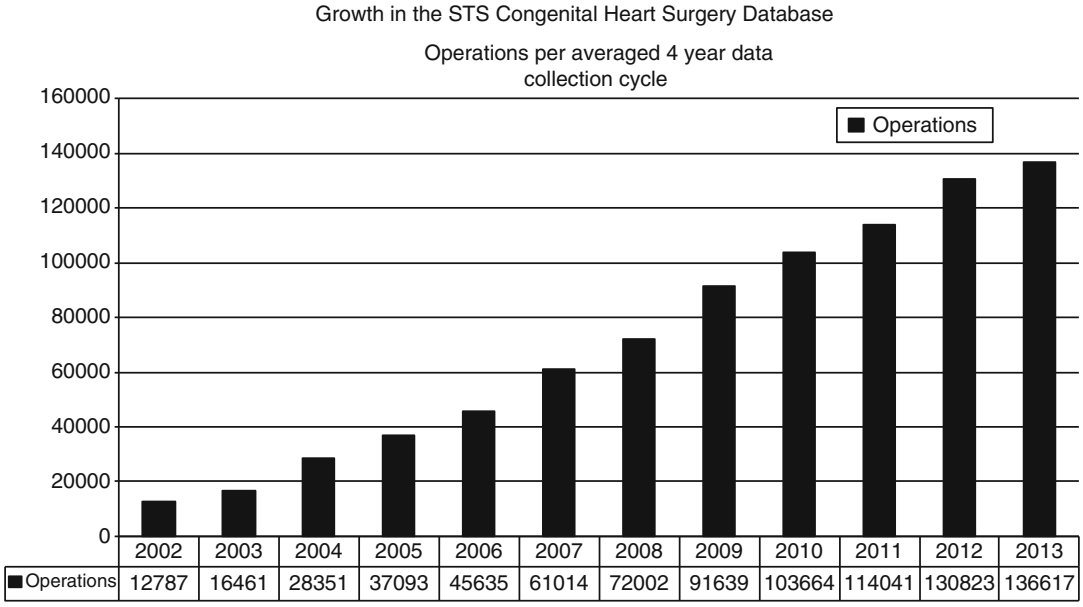
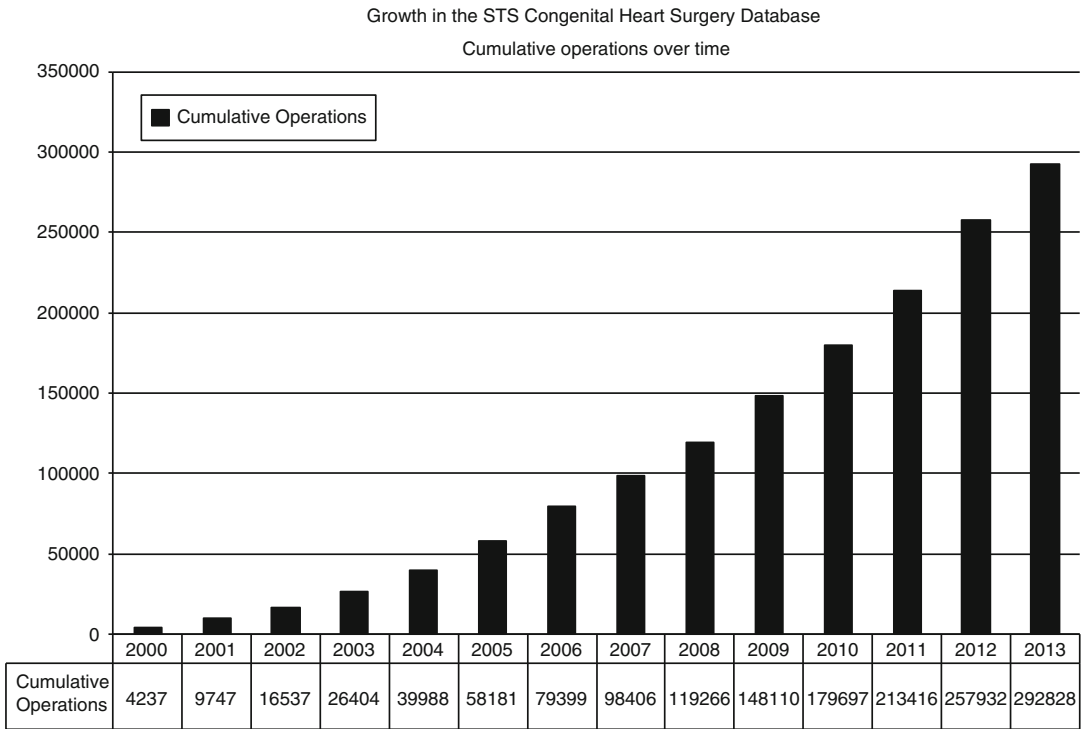


Fig. 8.2 The graph documents the annual growth of the STS Congenital Heart Surgery Database by the number of operations per averaged 4 year data collection cycle. The aggregate report from the Fall 2013 Harvest of the STS Congenital Heart Surgery Database [19] includes 136,617 operations performed in the 4 year period of July 1, 2009–June 30, 2013, inclusive, submitted from 120 hospitals in North America, 117 in the United States of America and 3 in Canada



	2000	2001	2002	2003	2004	2005	2006	2007	2008	2009	2010	2011	2012	2013
Cumulative Operations	4237	9747	16537	26404	39988	58181	79399	98406	119266	148110	179697	213416	257932	292828

Fig. 8.3 The graph documents the annual growth of the STS Congenital Heart Surgery Database by the cumulative number of operations over time. As of January 1, 2014, the number of cumulative total operations in the STS Congenital Heart Surgery Database is 292,828. The aggregate report from the Fall 2013 Harvest of the STS Congenital Heart Surgery Database [19] includes 136,617 operations performed in the 4 year period of July 1, 2009–June 30, 2013, inclusive, submitted from 120 hospitals in North America, 117 in the United States of America and 3 in Canada

inclusive, submitted from 120 hospitals in North America, 117 in the United States of America and 3 in Canada. In collaboration with EACTS, the STS has developed standardized methodology for tracking mortality and morbidity associated with the treatment of patients with congenital and pediatric cardiac disease [93, 105].

The EACTS Congenital Heart Surgery Database is the largest database in Europe dealing with congenital cardiac malformations (Fig. 8.4) [112, 117]. As of May 2013, the EACTS Congenital Heart Surgery Database contained 157,772 operations performed in 130,534 patients. As of May, 2013, the EACTS Congenital Heart Surgery Database had 348 Centers from 76 countries registered, with 173 active Centers from 46 countries submitting data.

The JCCVSD has recently been operationalized based on identical nomenclature and database standards as that used by EACTS and STS [117]. The JCCVSD began enrolling patients in 2008. By December 2011, over 100 hospitals were submitting data, and by April 2013, over 29,000 operations were entered into the JCCVSD, in just under 5 years of data collection (Fig. 8.5). In Japan, it is mandatory for specialists to enroll in

Fig. 8.5 The graph documents the initial growth of The Japan Congenital Cardiovascular Surgery Database (JCCVSD). The JCCVSD has recently been operationalized based on identical nomenclature and database standards as that used by EACTS and STS. The JCCVSD began enrolling patients in 2008. By December 2011, over 100 hospitals were submitting data, and by April 2013, over 29,000 operations were entered into the JCCVSD, in just under five years of data collection. The developers of the JCCVSD hope to collaborate with their colleagues across Asia to create an Asian Congenital Heart Surgery Database (This graph is provided courtesy of Arata Murakami, MD of The University of Tokyo in Tokyo, Japan)

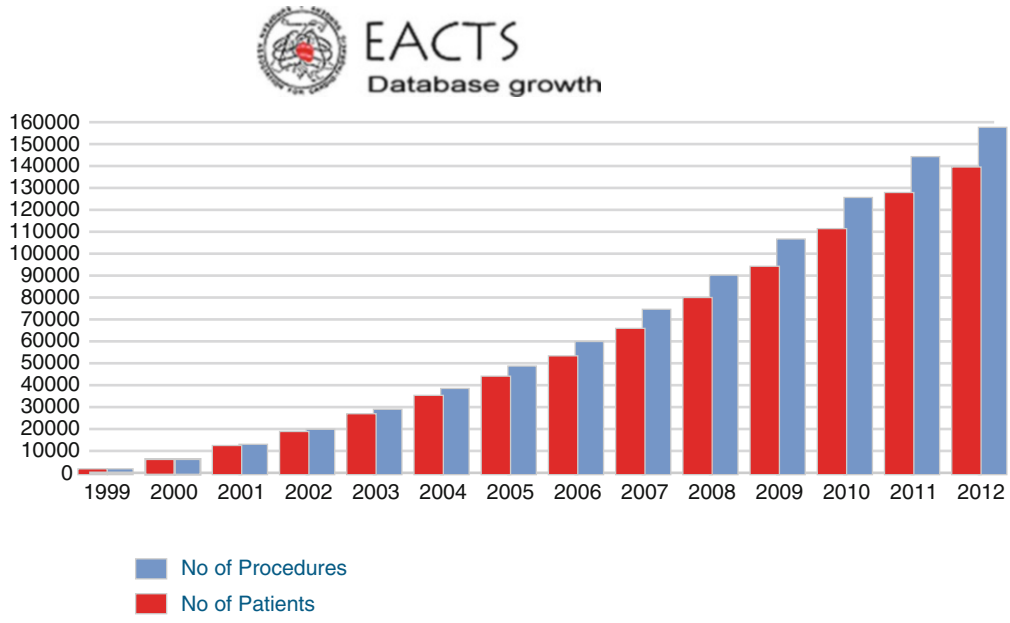
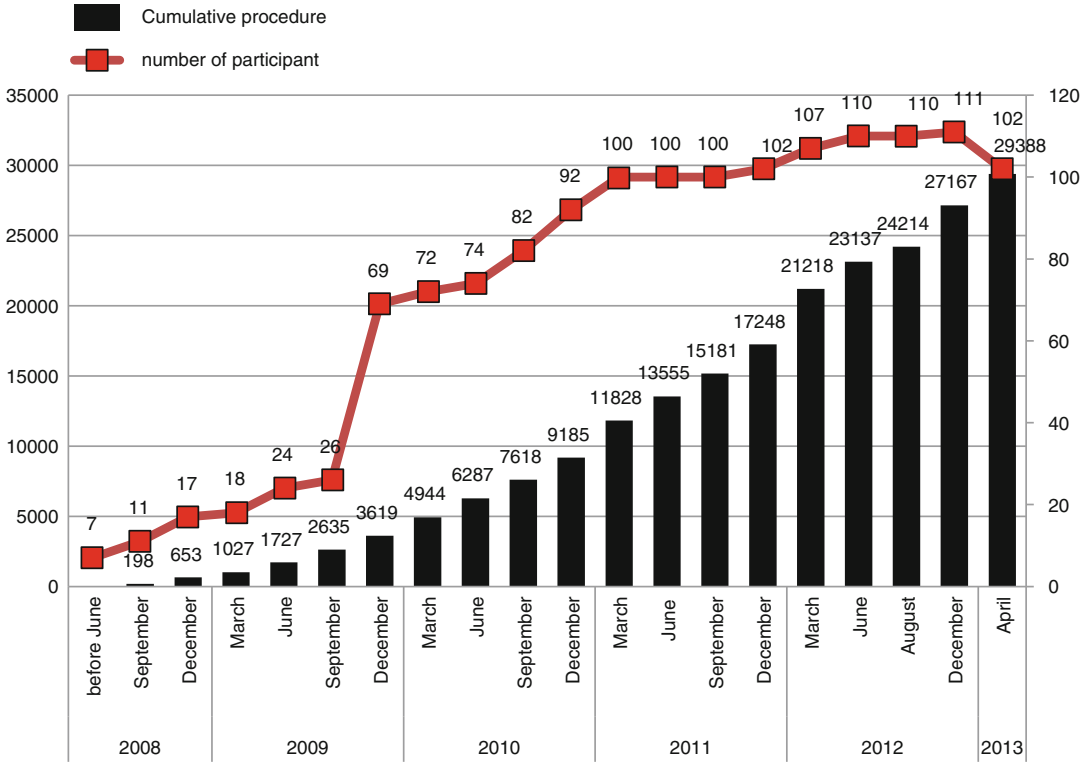


Fig. 8.4 The graph documents the annual growth in The European Association for Cardio-Thoracic Surgery Congenital Database by both number of patients and number of operations. As of May 2013, the EACTS Congenital Heart Surgery Database contained 157,772 operations performed in 130,534 patients. As of May, 2013, the EACTS Congenital Heart Surgery Database had 348 Centers from 76 countries registered, with 173 active Centers from 46 countries submitting data (This graph is provided courtesy of Bohdan Maruszewski of the Children's Memorial Health Institute in Warsaw, Poland, Director of The European Association for Cardio-Thoracic Surgery Congenital Database, and President of The European Congenital Heart Surgeons Association (ECHSA))



this benchmarking project in order to objectively examine their own performance and make efforts for continuous improvement. In the future, certification in Japan is to be performed solely on the basis of empirical data registered by the project. The developers of the JCCVSD hope to collaborate with their colleagues across Asia to create an Asian Congenital Heart Surgery Database.

In the United Kingdom, the United Kingdom Central Cardiac Audit Database (UKCCAD) uses the AEPC derived version of the IPCCC as the basis for its national, comprehensive, validated, and benchmark-driven audit of all pediatric surgical and transcatheter procedures undertaken since 2000 [152]. All 13 tertiary centers in the United Kingdom performing cardiac surgery or therapeutic cardiac catheterization in children with congenital cardiac disease submit data to the UKCCAD. Data about mortality is obtained from both results volunteered from the hospital databases, and by independently validated records of deaths obtained by the Office for National Statistics, using the patient's unique National Health Service number, or the general register offices of Scotland and Northern Ireland. Efforts are underway to link the UKCCAD to The EACTS Congenital Heart Surgery Database. Linkage of the UKCCAD to The EACTS Congenital Heart Surgery Database will require use of the crossmap of the AEPC derived version of the IPCCC (used by the UKCCAD) to the EACTS-STS derived version of the IPCCC (used by the EACTS, STS, and JCCVSD).

As of January 1, 2014, the STS Congenital Heart Surgery Database contains data from 292,828 operations, the EACTS Congenital Heart Surgery Database contains data from over 157,772 operations, and the JCCVSD contains data from over 29,000 operations. Therefore, the combined dataset of the STS Congenital Heart Surgery Database, the EACTS Congenital Heart Surgery Database, and the JCCVSD, contains data from over 479,000 operations, all coded with the EACTS-STS derived version of the IPCCC [100, 110, 112, 114], and all coded with identical data specifications [208].

Complexity Stratification

The importance of measurement of complexity derives from the fact that analysis of outcomes using raw measurements of mortality, without adjustment for complexity, is inadequate. The mix of cases can vary greatly from program to program. Without stratification of complexity, the analysis of outcomes will be flawed [56, 61, 73, 74, 76, 82, 106, 149, 150].

The analysis of outcomes after surgery requires a reliable method of estimating the risk of adverse events. However, formal risk modeling is challenging for rare operations. Complexity stratification provides an alternative methodology that can facilitate the analysis of outcomes of rare operations. Complexity stratification is a method of analysis in which the data are divided into relatively homogeneous groups (called strata). The data are analyzed within each stratum.

Three major multi-institutional efforts have attempted to measure the complexity of congenital cardiac surgical operations:

1. **R**isk **A**djustment in **C**ongenital **H**eart **S**urgery-1 methodology (RACHS-1 method) [56, 73, 149]
2. **A**ristotle **B**asic **C**omplexity Score (ABC Score) [61, 74, 76, 82, 106, 149]
3. **STS**-E**A**C**T**S Congenital Heart Surgery Mortality Categories (STS-EACTS Mortality Categories) (STAT Mortality Categories) [150].

RACHS-1 and the ABC Score were developed at a time when limited multi-institutional clinical data were available and were therefore based in a large part on subjective probability (expert opinion). The STAT Mortality Categories are a tool for complexity stratification that was developed from an analysis of 77,294 operations entered into the EACTS Congenital Heart Surgery Database (33,360 operations) and the STS Congenital Heart Surgery Database (43,934 patients) between 2002 and 2007. Procedure-specific mortality rate estimates were calculated using a Bayesian model that adjusted for small denominators. Operations were sorted by increasing risk and grouped into five categories (the STS–EACTS Congenital Heart Surgery Mortality Categories) that were designed to be

Table 8.1 Method of modeling procedures. Shows the results of comparing the STS–EACTS Categories (2009) to the RACHS-1 Categories and the Aristotle Basic Complexity Score using an independent validation sample of 27,700 operations performed in 2007 and 2008. In the subset of procedures for which STS–EACTS Category, RACHS-1 Category, and Aristotle Basic Complexity Score are defined, discrimination was highest for the STS–EACTS categories (C-index = 0.778), followed by RACHS-1 categories (C-index = 0.745), and Aristotle Basic Complexity scores (C-index = 0.687)

	Model without patient covariates	Model with patient covariates	Percent of operations that can be classified (%)
STS-EACTS Congenital Heart Surgery Mortality Categories (2009)	C = 0.778	C = 0.812	99
RACHS-1 categories	C = 0.745	C = 0.802	86
Aristotle basic complexity score	C = 0.687	C = 0.795	94

optimal with respect to minimizing within-category variation and maximizing between-category variation.

Table 8.1 compares RACHS-1, the ABC Score, and the STS-EACTS Mortality Categories. Table 8.2 shows the application in the STS Congenital Heart Surgery Database of the STAT Congenital Heart Surgery Mortality Categories [198]. STS and EACTS have transitioned from the primary use of Aristotle and RACHS-1 to the primary use of the STAT Mortality Categories because of three reasons:

1. STAT Score was developed primarily based on objective data while RACHS-1 and Aristotle were developed primarily on expert opinion (Subjective probability)
2. STAT Score allows for classification of more operations than RACHS-1 or Aristotle
3. STAT Score has a higher c-statistic than RACHS-1 or Aristotle.

Meaningful evaluation and comparison of outcomes require consideration of both mortality and morbidity, but the latter is much harder to measure. The STAT Mortality Categories provide an empirically based tool for analyzing mortality associated with operations for congenital heart disease [150]. STS has developed the STAT Morbidity Categories [215] based on major postoperative complications and postoperative length of stay. Both major postoperative complications and postoperative length of stay were used because models that assume a perfect one to one relationship between postoperative complications and postoperative length of stay are not likely to fit the data well. Incorporation of both major postoperative complications and postoperative length of stay allows creation of a much more informative model. The STAT Morbidity Categories provide an empirically based tool for analyzing morbidity associated with operations for congenital heart disease [215].

Table 8.2 Shows the discharge mortality in an analysis of patients in the STS Congenital Heart Surgery Database who underwent surgery between January 1, 2005 and December 31, 2009, inclusive [198], stratified by STAT Mortality Categories (STS–EACTS Congenital Heart Surgery Mortality Categories)

	Total number of operations	Discharge mortality (%)
STAT mortality category 1	15,441	0.55
STAT mortality category 2	17,994	1.7
STAT mortality category 3	8,989	2.6
STAT mortality category 4	13,375	8.0
STAT mortality category 5	2,707	18.4

Data Verification

Collaborative efforts involving EACTS and STS aim to enhance mechanisms to verify the completeness and accuracy of the data in the databases [21, 126]. A combination of three strategies may ultimately be required to allow for optimal verification of data:

1. Intrinsic data verification (designed to rectify inconsistencies of data and missing elements of data)
2. Site visits with "Source Data Verification" (in other words, verification of the data at the primary source of the data)
3. External verification of the data from independent databases or registries (such as governmental death registries)

Data quality in the STS Congenital Heart Surgery Database is evaluated through intrinsic data verification by Duke Clinical Research Institute (DCRI) (including identification and correction of missing/out of range values and inconsistencies across fields). DCRI is the data warehouse and analytic center of the STS Congenital Heart Surgery Database.

In addition to intrinsic data verification by DCRI, each year, approximately 10 % of participants are randomly selected for audits of their center, in accordance with their STS Congenital Heart Surgery Database Participation Agreement. The audit is designed to complement the internal quality controls, with an overall objective of maximizing the integrity of the data in the STS Congenital Heart Surgery Database by examining the accuracy, consistency, and completeness of the data. STS has selected Telligen to perform an independent, external audit of the STS Congenital Heart Surgery Database. As the state of Iowa's Medicare Quality Improvement Organization (QIO), Telligen partners with health care professionals to assure high quality, cost effective health care. As a Quality Improvement Organization, Telligen is compliant with the Health Insurance Portability and Accountability Act of 1996 of the United States of America (HIPAA) and performs audits adhering to strict security policies. Additionally, an STS congenital heart surgeon volunteer leader participates in the audit.

In the STS Congenital Heart Surgery Database, the audit process includes:

- Completion of the STS Data Collection Questionnaire and review of responses with the primary data contact, data manager, and/or other relevant personnel
- Review of the data collection process and documentation to determine case eligibility for submittal to the STS Congenital Heart Surgery Database
- Comparison of facility operative case logs with cases submitted to the STS Congenital Heart Surgery Database
- Data abstraction (from original source documents) of congenital heart surgery records randomly selected by DCRI and all operative mortality cases for the preceding calendar year.
- A summary conference with the surgeon representative, primary data contact, data manager, and/or other relevant personnel to discuss general trends in data collection and submission processes.

In 2013, the audit of the STS Congenital Heart Surgery Database included the following documentation of rates of completeness and accuracy for the specified fields of data:

- Primary Diagnosis (Completeness = 100 %, Accuracy = 96.2 %),
- Primary Procedure (Completeness = 100 %, Accuracy = 98.7 %),
- Mortality Status at Hospital Discharge (Completeness = 100 %, Accuracy = 98.8 %)

In 2014, 11 Participants in the STS Congenital Heart Surgery Database will be audited.

Subspecialty Collaboration

Under the leadership of The MultiSocietal Database Committee for Pediatric and Congenital Heart Disease [110–112], further collaborative efforts are ongoing between congenital and pediatric cardiac surgeons and other subspecialties, including

1. Pediatric cardiac anesthesiologists, via The Congenital Cardiac Anesthesia Society [105, 119, 139, 207],
2. Pediatric cardiac intensivists, via The Pediatric Cardiac Intensive Care Society [190], and
3. Pediatric cardiologists, via the Joint Council on Congenital Heart Disease, the American College of Cardiology, and The Association for European Paediatric Cardiology [118].

Strategies have been developed to link together databases [109, 164, 189, 193, 194, 210, 219]. By linking together different databases, one can capitalize on the strengths and mitigate some of the

weaknesses of these databases and therefore allow analyses not possible with either dataset alone. Linked databases have facilitated both comparative effectiveness research [193, 194, 219] and longitudinal follow-up [145, 146, 173, 214]. Under the leadership of The MultiSocietal Database Committee for Pediatric and Congenital Heart Disease [110–112], further collaborative efforts are ongoing between congenital and pediatric cardiac surgeons and other subspecialties.

The Multi-Societal Database Committee for Pediatric and Congenital Heart Disease has held ten annual meetings, each lasting 1 or 2 days, in 2005, 2006, 2007, 2008, 2009, 2010, 2011, 2012, 2013, and 2014. The 11th Multi-Societal Meeting has already been scheduled for 2015 in Prague, the Czech Republic:

1. The First Annual Meeting of The Multi-Societal Database Committee for Pediatric and Congenital Heart Disease. Chicago, Illinois, Chicago Hilton, Thursday August 25, 2005 and Friday August 26, 2005. (At the inception of this first meeting, the meeting was named the "VPS/STS/PCICS Combined Database Meeting". VPS=The Virtual Pediatric Intensive Care Unit Systems, STS=The Society of Thoracic Surgeons, PCICS=The Pediatric Cardiac Intensive Care Society.)
2. The Second Annual Meeting of The Multi-Societal Database Committee for Pediatric and Congenital Heart Disease. Chicago, Illinois, Thursday August 17, 2006 and Friday, August 18, 2006.
3. The Third Annual Meeting of The Multi-Societal Database Committee for Pediatric and Congenital Heart Disease. Hotel George in Washington, DC, Thursday September 27, 2007 and Friday, September 28, 2007.
4. The Fourth Annual Meeting of The Multi-Societal Database Committee for Pediatric and Congenital Heart Disease. Omni Mount-Royal Hotel, Montreal, Canada, Saturday October 4, 2008 and Sunday October 5, 2008.
5. The Fifth Annual Meeting of The Multi-Societal Database Committee for Pediatric and Congenital Heart Disease of The Global Organization for Pediatric and Congenital Heart Disease: "The Transition from Outcomes Analysis to Quality Improvement". The Emory Conference Center, Atlanta, Georgia, Wednesday September 16, 2009 and Thursday, September 17, 2009.
6. The Sixth Annual Meeting of The Multi-Societal Database Committee for Pediatric and Congenital Heart Disease of The Global Organization for Pediatric and Congenital Heart Disease: "Creating a Multidisciplinary Strategy for Improving the Quality of HealthCare Delivered to Patients with Pediatric and Congenital Heart Disease". The Emory Conference Center, Atlanta, Georgia, Thursday, August 26, 2010 and Friday, August 27, 2010.
7. The Seventh Annual Meeting of The Multi-Societal Database Committee for Pediatric and Congenital Heart Disease: "The relationship between (1) Outcomes Analysis, (2) Quality Improvement, and (3) Patient Safety". University of Cambridge, Cambridge, United Kingdom, Tuesday, September 20, 2011 and Wednesday, September 21, 2011.
8. The Eighth Annual Meeting of The Multi-Societal Database Committee for Pediatric and Congenital Heart Disease: "New Initiatives in Outcomes and Quality". Chair: Jeffrey P. Jacobs, MD, Local Hosts: Robert Campbell, MD and Robert Vincent, MD. The Emory Conference Center, Atlanta, Georgia (404) 712–6000. Thursday, August 23, 2012 and Friday, August 24, 2012.
9. The Ninth Annual Meeting of The Multi-Societal Database Committee for Pediatric and Congenital Heart Disease: "Bridging the Gap from Outcomes to Quality". Chair: Jeffrey P. Jacobs, Local Host: Shakeel Qureshi, President, The Association for European Paediatric and Congenital Cardiology, Meeting held at the 47th Annual Meeting of The Association for European Paediatric and Congenital Cardiology (AEPC), London, England, United Kingdom, Thursday, May 23, 2013.
10. The Tenth Annual Meeting of The Multi-Societal Database Committee for Pediatric and Congenital Heart Disease: "Dashboards for Pediatric and Congenital Cardiac Care". Chair: Jeffrey P. Jacobs, MD, Local Hosts:

Robert Campbell, MD and Robert Vincent, MD. The Emory Conference Center, Atlanta, Georgia (404) 712–6000. Thursday, September 4, 2014 and Friday, September 5, 2014.

11. The Eleventh Annual Meeting of The Multi-Societal Database Committee for Pediatric and Congenital Heart Disease: "Improving the quality of congenital cardiology healthcare by harmonizing international databases". Chair: Jeffrey P. Jacobs, MD, Robert Vincent, MD, and Rodney Franklin, MD. Meeting held at the 49th Annual Meeting of The Association for European Paediatric and Congenital Cardiology (AEPC), Prague, the Czech Republic, Wednesday, May 20, 2015.

The various organizations and Societies whose members have participated in the meetings and activities of The Multi-Societal Database Committee for Pediatric and Congenital Heart Disease as well as the various participants themselves have previously been published [111], although the group continues to grow and involve multiple professional medical and nursing societies as well as multiple governmental and nongovernmental agencies. Some notable accomplishments of this multidisciplinary group are worth brief mention. At the first meeting of the Multi-Societal Database Committee, initial discussions took place about the possibility of linking together the various databases of the subspecialties of pediatric cardiac surgery, pediatric cardiology, pediatric cardiac anesthesia, and pediatric critical care. The Multi-Societal Database Committee rapidly realized that it would be essential to collaborate in multiple areas:

1. Use of a common language and nomenclature
2. Use of a database with an established uniform core dataset for collection of information
3. Incorporation of a mechanism of evaluating case complexity
4. Availability of a mechanism to assure verification of the completeness and accuracy of the data collected
5. Collaboration between medical and surgical subspecialties,
6. Standardization of protocols for life-long longitudinal follow-up.

Each of these six areas is discussed in detail in the following 530 page Supplement published in Cardiology in the Young by the Multi-Societal Database Committee for Pediatric and Congenital Heart Disease [110]. Initial discussions of the Multi-Societal Database Committee identified that it was essential for the various subspecialty databases to use identical nomenclature in order to allow them to communicate with each other with meaning. Various lists of terminology would need to be harmonized:

1. Diagnoses
2. Procedures
3. Complications
4. Preoperative Factors

The Multi-Societal Database Committee agreed to use The International Pediatric and Congenital Cardiac Code (IPCCC) (http://www.ipccc.net/) as the basis of communication. Mature and well developed Short Lists and Long Lists of Diagnoses and Procedures are available via The International Pediatric and Congenital Cardiac Code, and these diagnostic and procedural lists have been incorporated into the various subspecialty databases and harmonized.

At the second meeting of The Multi-Societal Database Committee, the diagnostic and procedural lists of nomenclature were harmonized across the multiple databases of pediatric cardiac surgery, pediatric cardiology, pediatric cardiac anesthesia, and pediatric critical care. These harmonized lists were based on the IPCCC. Because the diagnostic and procedural lists in The International Pediatric and Congenital Cardiac Code are matured and functional, the Multi-Societal Database Committee adopted these lists and harmonized them across their databases. The Multi-Societal Database Committee then elected to focus on developing a mature list of Complications and defining these complications [110–140].

At the third and fourth meeting of The Multi-Societal Database Committee, the topic of complications associated with the treatment of patients with pediatric and congenital cardiac disease was discussed in detail. The Multi-Societal Database Committee ultimately developed and published a Long List of Complications [110, 140] and a Short List of Complications [110–112], with consensus-based definitions provided in each List:

1. The Long List of Complications contains and defines 2,836 terms and is named: "The Long List of Complications of The Multi-Societal Database Committee for Pediatric and Congenital Heart Disease", with the abbreviated short name: "Multi-Societal Long List of Complications". Although the act of navigating a list with 2,836 terms can initially seem quite daunting, it can become quite simple and enjoyable with the aid of computerized navigation tools designed to support the hierarchal structure of the list.
2. The Short List of Complications contains and defines 56 terms.

At the fifth meeting of The Multi-Societal Database Committee, the Committee transitioned from collaborative efforts related to databases to collaborative initiatives related to quality improvement. The sixth meeting of The Multi-Societal Database Committee focused on "Creating a Multidisciplinary Strategy for Improving the Quality of HealthCare Delivered to Patients with Pediatric and Congenital Heart Disease". The first and second meetings were organized and hosted by the VPS Database, and the National Association of Children's Hospitals and Related Institutions (NACHRI). The third and fourth meetings were organized and hosted by the Society of Thoracic Surgeons (STS), and the fifth, sixth, eighth, and tenth meetings were organized and hosted by Emory University. The seventh meeting was hosted the Pediatric Cardiac Intensive Care Society. The ninth meeting was hosted by The Association for European Paediatric and Congenital Cardiology (AEPC), and the AEPC will host the eleventh meeting. The Multi-Societal Database Committee for Pediatric and Congenital Heart Disease is a platform that facilitates the ability for databases in the domain of pediatric cardiac care to span conventional subspecialty and temporal boundaries.

Longitudinal Follow-Up

The transformation of the STS Database to a platform for longitudinal follow-up will ultimately result in higher quality of care for all cardiothoracic surgical patients by facilitating longitudinal comparative effectiveness research on a national level [127, 173, 184, 214]. Several potential strategies will allow longitudinal follow-up with the STS Database, including the development of clinical longitudinal follow-up modules within the STS Database itself, and linking the STS Database to other clinical registries, administrative databases, and national death registries:

1. Using probabilistic matching with shared indirect identifiers, the STS Database can be linked to administrative claims databases (such as the CMS Medicare Database [145, 146] and the Pediatric Health Information System (PHIS) database [109, 164, 189, 193, 194, 210, 219]) and become a valuable source of information about long-term mortality, rates of re-hospitalization, long-term morbidity, and cost [208].
2. Using deterministic matching with shared unique direct identifiers, the STS Database can be linked to national death registries like the Social Security Death Master File (SSDMF) and the National Death Index (NDI) in order to verify life-status over time [127, 173, 184, 214].
3. Through either probabilistic matching or deterministic matching [184], the STS Database can link to multiple other clinical registries, such as the National Cardiovascular Data Registry (NCDR) of the American College of Cardiology (ACC), in order to provide enhanced clinical follow-up.
4. The STS Database can develop clinical longitudinal follow-up modules of its own to provide detailed clinical follow-up [109, 127, 173, 184, 214].

Quality Assessment and Quality Improvement

The STS Database is increasingly used to document variation in outcomes [182, 198] and measure quality [179, 186]. Funnel plots may be used to demonstrate this variation in outcome and to facilitate the identification of centers that are outliers in performance (Fig. 8.6). Quality improvement initiatives can be initiated in "low

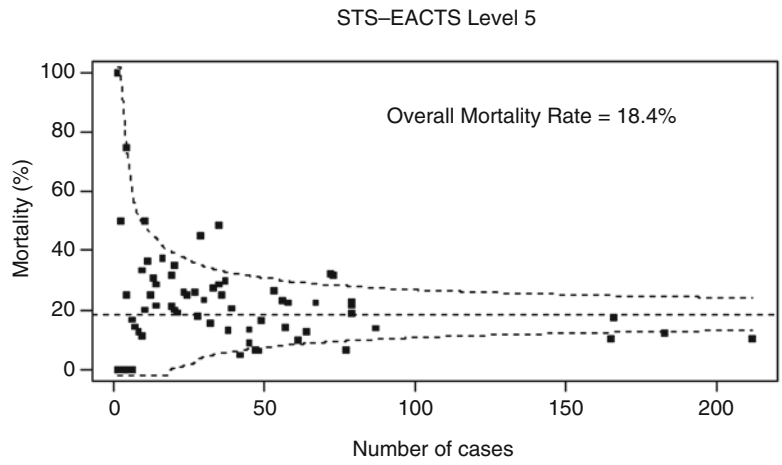


Fig. 8.6 In this graph, data about mortality is displayed as a funnel plot for STAT Category five operations [198]. The horizontal dashed line depicts aggregate STS mortality before discharge. Dashed lines depicting exact 95 % binomial prediction limits were overlaid to make a funnel plot. Squares represent the number of cases and mortality before discharge for individual STS Congenital Heart Surgery Database participants (centers). This analysis includes patients undergoing surgery during the 5 year analytic window of 2005 –2009, inclusive, and includes 70 STS centers in the STS Congenital Heart Surgery Database and 2,707 operations. Centers that were identified as outliers represented 18.6 % of participating centers (13 out of 60): 10 % (7 out of 70) were "high-performing outliers" and 8.6 % (6 out of 70) were "low-performing outliers". Quality improvement initiatives can be initiated in "low performing centers" and best practices can be obtained from "high performing centers". (**STS**-E**A**C**T**S Congenital Heart Surgery Mortality Categories = STS-EACTS Mortality Categories = STAT Mortality Categories [150], *STS* The Society of Thoracic Surgeons, *EACTS* European Association for Cardio-Thoracic Surgery)

performing centers" and best practices can be obtained from "high performing centers".

STS has collaborated with the Congenital Heart Surgeons' Society (CHSS) to develop and endorse metrics to assess the quality of care delivered to patients with pediatric and congenital cardiac disease [186]. Tables 8.3, 8.4, and 8.5 presents 21 "Quality Measures for Congenital and Pediatric Cardiac Surgery" that were developed and approved by the Society of Thoracic Surgeons (STS) and endorsed by the Congenital Heart Surgeons' Society (CHSS). These Quality Measures are organized according to Donabedian's Triad of Structure, Process, and Outcome [242]. It is hoped that these quality measures can aid in congenital and pediatric cardiac surgical quality assessment and quality improvement initiatives. These initiatives will take on added importance as the public reporting of cardiac surgery performance becomes more common [143, 176, 177].

Summary: Bridging the Gap Form Analysis of Outcomes to Improvement of Quality

Clinical registries represent a foundational tool in the following inter-related process:

1. Measuring the outcomes of medical and surgical practices,
2. Developing evidence for best medical and surgical practices,
3. Providing actionable feedback to clinicians, and
4. Improving the quality of care and outcomes.

Clinical registries are the best tool for measuring the outcomes of the processes of care [220, 221]. As described in this chapter, the ability to measure clinical outcomes properly requires using standardized clinical nomenclature, uniform standards for defining elements of data and collecting these data, strategies to adjust for the complexity of patients, techniques

Table 8.3 Quality measures for congenital and pediatric cardiac surgery

1. Participation in a National Database for Pediatric and Congenital Heart Surgery
2. Multidisciplinary rounds involving multiple members of the healthcare team
3. Availability of Institutional **Pediatric ECLS** (**Extracorporeal Life Support**) **Program**
4. **Surgical volume** for Pediatric and Congenital Heart Surgery: Total Programmatic Volume and Programmatic Volume Stratified by the **Five STS-EACTS Mortality Categories**
5. **Surgical Volume** for **Eight Pediatric and Congenital Heart Benchmark Operations**
6. Multidisciplinary **preoperative planning conference** to plan pediatric and congenital heart surgery operations
7. Regularly Scheduled **Quality Assurance and Quality Improvement Cardiac Care Conference**, to occur no less frequently than once every two months
8. Availability of **intraoperative transesophageal echocardiography** (**TEE**) and epicardial echocardiography
9. **Timing of Antibiotic Administration** for Pediatric and Congenital Cardiac Surgery Patients
10. Selection of **Appropriate Prophylactic Antibiotics and Weight-Appropriate Dosage** for Pediatric and Congenital Cardiac Surgery Patients
11. Use of an **expanded pre-procedural and post-procedural** "**time-out**"
12. Occurrence of new post-operative **renal failure** requiring dialysis
13. Occurrence of new post-operative **neurological deficit** persisting at discharge
14. Occurrence of arrhythmia necessitating **permanent pacemaker** insertion
15. Occurrence of **paralyzed diaphragm** (possible phrenic nerve injury)
16. Occurrence of need for **postoperative mechanical circulatory support** (IABP, VAD, ECMO, or CPS)
17. Occurrence of **unplanned reoperation** and/or interventional cardiovascular catheterization procedure
18. **Operative Mortality** Stratified by the Five STS-EACTS Mortality Levels
19. **Operative Mortality** for Eight Benchmark Operations
20. Index Cardiac **Operations Free of Mortality and Major Complication**
21. Operative **Survivors Free of Major Complication**

to verify the completeness and accuracy of data, and collaboration across medical and surgical subspecialties. All of these elements exist in the ideal clinical registry.

Clinical registries can be used as a platform for developing evidence for best medical practices and performing comparative effectiveness research. The NIH-funded linkage of the STS Congenital Heart Surgery Database to the Pediatric Health Information System (PHIS) Database exemplifies this approach [164, 189, 193, 194, 210, 219]. This linkage of clinical and administrative data facilitated comparative effectiveness research in the domains of perioperative methylprednisolone and outcome in neonates undergoing heart surgery [193] and antifibrinolytic medications in pediatric heart surgery [194]. Similarly, The NIH-funded ASCERT trial (American College of Cardiology Foundation—Society of Thoracic Surgeons Collaboration on the Comparative Effectiveness of Revascularization sTrategies trial) also used linked clinical and administrative data to compare surgical and transcatheter strategies of coronary revascularization [243, 244]. Although randomized trials have been considered by many to be the gold standard of comparative effectiveness research, recent efforts have examined the possibility of using a clinical registry as a platform for randomized trials [245, 246], potentially accomplishing the dual objectives of decreasing the cost of the trial and increasing the generalizability of the patients enrolled.

Clinical registries can provide actionable feedback to clinicians and therefore aid in initiatives to improve quality. Clinical registries can provide practitioners with accurate and timely feedback of their own outcomes and can benchmark these outcomes to regional, national, or even international aggregate data [182, 198, 247–249].

The ultimate goal of clinical registries is to improve quality of care and outcomes. Clinical

Table 8.4 Definitions of quality measures for congenital and pediatric cardiac surgery

	Number	Type	Title of indicator	Description
1	S-1	Structure	Participation in a National Database for Pediatric and Congenital Heart Surgery	Participation in at least one multi-center, standardized data collection and feedback program that provides regularly scheduled reports of the individual center's data relative to national multicenter aggregates and uses process and outcome measures
2	S-2	Structure	Multidisciplinary rounds involving multiple members of the healthcare team	Occurrence of daily multidisciplinary rounds on pediatric and congenital cardiac surgery patients involving multiple members of the healthcare team, with recommended participation including but not limited to: cardiac surgery, cardiology, critical care, primary caregiver, family, nurses, pharmacist, and respiratory therapist. Involvement of the family is encouraged
3	S-3	Structure	Availability of Institutional Pediatric ECLS (Extracorporeal Life Support) Program	Availability of an institutional pediatric Extracorporeal Life Support (ECLS) Program for pediatric and congenital cardiac surgery patients. Measure is satisfied by availability of ECMO equipment and support staff, but applies as well to Ventricular Assist Devices (including extracorporeal, paracorporeal, and implantable)
4	S-4	Structure	Surgical volume for Pediatric and Congenital Heart Surgery: Total Programmatic Volume and Programmatic Volume Stratified by the Five STS-EACTS Mortality Categories	Surgical volume for Pediatric and Congenital Heart Surgery
				STS version 2.5: All Index Cardiac Operations (A Cardiac Operation is defined as an operation of Operation Type "CPB" or "No CPB Cardiovascular".)
				STS version 3.0: Same
				Surgical volume for pediatric and congenital heart surgery stratified by the five STS-EACTS Mortality Levels, a multi-institutional validated complexity stratification tool
				See *J Thorac Cardiovasc Surg* 2009;138:1139–1153. **O'Brien et al. An empirically based tool for analyzing mortality associated with congenital heart surgery. Table 1, pp 1140–1146**
5	S-5	Structure	Surgical volume for Eight Pediatric and Congenital Heart Benchmark Operations	Surgical volume for Eight Benchmark Pediatric and Congenital Heart Operations:
				These 8 Eight Benchmark Pediatric and Congenital Heart Operations are tracked when they are the primary procedure of an Index Cardiac Operation. (A Cardiac Operation is defined as an operation of Operation Type "CPB" or "No CPB Cardiovascular")
				Procedure type Abbreviation STS–CHSDB diagnostic and procedural inclusionary and exclusionary criteria

	1. VSD repair	VSD	**Procedural inclusionary criteria**:
			100 = VSD repair, Primary closure
			110 = VSD repair, Patch
			120 = VSD repair, Device*
			*(Please note that this measure is applicable when one or more septal occluder devices are implanted in the course of a surgical operation for which the Primary Procedure of an Index Cardiac Operation is VSD repair. [A Cardiac Operation is defined as an operation of Operation Type "CPB" or "No CPB Cardiovascular".] A VSD device that is placed as a purely transcatheter technique and not as a component of a cardiac operation is classified as an Interventional Cardiology Procedure and is not tracked as part of this measure.)
			Diagnostic inclusionary criteria:
			71 = VSD, Type 1 (Subarterial) (Supracristal) (Conal septal defect) (Infundibular)
			73 = VSD, Type 2 (Perimembranous) (Paramembranous) (Conoventricular)
			75 = VSD, Type 3 (Inlet) (AV canal type)
			77 = VSD, Type 4 (Muscular)
			79 = VSD, Type: Gerbode type (LV-RA communication)
			Diagnostic exclusionary criteria:
			80 = VSD, Multiple
	2. TOF repair	TOF	**Procedural inclusionary criteria:**
			350 = TOF repair, No ventriculotomy
			360 = TOF repair, Ventriculotomy, Nontransanular patch
			370 = TOF repair, Ventriculotomy, Transanular patch
			380 = TOF repair, RV-PA conduit
			Diagnostic inclusionary criteria:
			290 = TOF
			2140 = TOF, Pulmonary stenosis
			Diagnostic exclusionary criteria
			300 = TOF, AVC (AVSD)
			310 = TOF, Absent pulmonary valve
			320 = Pulmonary atresia
			330 = Pulmonary atresia, IVS
			340 = Pulmonary atresia, VSD (Including TOF, PA)
			350 = Pulmonary atresia, VSD-MAPCA (pseudotruncus)
			360 = MAPCA(s) (major aortopulmonary collateral[s]) (without PA-VSD)

(continued)

Table 8.4 (continued)

Number	Type	Title of indicator	Description	
		3. Complete AV canal repair	AVC	**Procedural inclusionary criteria**
				170 = AVC (AVSD) repair, Complete (CAVSD)
				Diagnostic inclusionary criteria:
				100 = AVC (AVSD), Complete (CAVSD)
				Diagnostic exclusionary criteria:
				110 = AVC (AVSD), Intermediate (transitional)
				120 = AVC (AVSD), Partial (incomplete) (PAVSD) (ASD, primum)
				300 = TOF, AVC (AVSD)
		4. Arterial switch	ASO	**Procedural inclusionary criteria**:
				1110 = Arterial switch operation (ASO)
				Procedural exclusionary criteria:
				1120 = Arterial switch operation (ASO) and VSD repair
				1123 = Arterial switch procedure + Aortic arch repair
				1125 = Arterial switch procedure and VSD repair + Aortic arch repair
				1050 = Congenitally corrected TGA repair, Atrial switch and ASO (double switch)
		5. Arterial switch + VSD repair	ASO + VSD	**Procedural inclusionary criteria**:
				1120 = Arterial switch operation (ASO) and VSD repair
				Procedural exclusionary criteria:
				1110 = Arterial switch operation (ASO)
				1123 = Arterial switch procedure + Aortic arch repair
				1125 = Arterial switch procedure and VSD repair + Aortic arch repair
				1050 = Congenitally corrected TGA repair, Atrial switch and ASO (double switch)
		6. Fontan	Fontan	**Procedural inclusionary criteria**:
				950 = Fontan, Atrio-pulmonary connection
				960 = Fontan, Atrio-ventricular connection
				970 = Fontan, TCPC, Lateral tunnel, Fenestrated
				980 = Fontan, TCPC, Lateral tunnel, Nonfenestrated
				1000 = Fontan, TCPC, External conduit, Fenestrated
				1010 = Fontan, TCPC, External conduit, Nonfenestrated
				1030 = Fontan, Other
				2340 = Fontan + Atrioventricular valvuloplasty
				Procedural exclusionary criteria:
				Exclude patients age≥7 years
				1025 = Fontan revision or conversion (Re-do Fontan)

				7. Truncus repair	Truncus	**Procedural inclusionary criteria**:
						Primary procedure must be:
						230= Truncus arteriosus repair
						Procedural exclusionary criteria:
						Exclude any operation if any of the component procedures is:
						240 = Valvuloplasty, Truncal valve
						2290 = Valvuloplasty converted to valve replacement in the same operation, Truncal valve
						250 = Valve replacement, Truncal valve
						2220 = Truncus + Interrupted aortic arch repair (IAA) repair
				8. Norwood	Norwood	**Procedural inclusionary criteria**:
						870 = Norwood procedure
6	P-1	Process	Multidisciplinary preoperative planning conference to plan pediatric and congenital heart surgery operations	Occurrence of a pre-operative multidisciplinary planning conference to plan pediatric and congenital heart surgery cases. This conference will involve multiple members of the healthcare team, with recommended participation including but not limited to: cardiology, cardiac surgery, anesthesia, and critical care		
				This measure will be coded on a per operation basis. Reporting of compliance will be as a fraction of all Cardiac Operations. A Cardiac Operation is defined as an operation of Operation Type “CPB” or “No CPB Cardiovascular”		
7	P-2	Process	Regularly scheduled Quality Assurance and Quality Improvement Cardiac Care Conference, to occur no less frequently than once every 2 months	Occurrence of a regularly scheduled Quality Assurance and Quality Improvement Cardiac Care Conference to discuss care provided to patients who have undergone pediatric and congenital cardiac surgery operations, including reporting and discussion of all major complications and mortalities, and discussion of opportunities for improvement		
				Reporting of compliance will be by reporting the date of occurrence. Annual compliance of 100 % equals no fewer than six conferences per year		
8	P-3	Process	Availability of intraoperative transesophageal echocardiography (TEE) and epicardial echocardiography	Availability of intraoperative transesophageal echocardiography (TEE) and appropriate physician and sonographer support for pediatric and congenital cardiac operations. Epicardial echocardiography and appropriate physician and sonographer support should be readily available for those patients in whom TEE is contraindicated or less informative. Availability means presence and availability of equipment and staff		
				This measure will be coded on a per operation basis. Reporting of compliance will be as the fraction of all Cardiac Operations with availability (as opposed to use) of TEE and/or epicardial echocardiography. (A Cardiac Operation is defined as an operation of Operation Type “CPB” or “No CPB Cardiovascular”)		
9	P-4	Process	Timing of Antibiotic Administration for Pediatric and Congenital Cardiac Surgery patients	Measure is satisfied for each Cardiac Operation, when there is documentation that the patient has received prophylactic antibiotics within the hour immediately preceding surgical incision (2 h if receiving vancomycin). (A Cardiac Operation is defined as an operation of operation type “CPB” or “No CPB Cardiovascular”)		

(continued)

Table 8.4 (continued)

	Number		Type	Title of indicator Description
10	P-5	Process	Selection of appropriate prophylactic antibiotics and weight-appropriate dosage for pediatric and congenital cardiac surgery patients	Measure is satisfied for each Cardiac Operation, when there is documentation that the patient received body weight appropriate prophylactic antibiotics as recommended for the operation. (A Cardiac Operation is defined as an operation of operation type "CPB" or "No CPB Cardiovascular")
11	P-6	Process	Use of an expanded pre-procedural and post-procedural "time-out"	Measure is satisfied for each Cardiac Operation when there is documentation of performance and completion of an expanded pre-procedural and post-procedural "time-out" that includes the following four elements (A Cardiac Operation is defined as an operation of operation type "CPB" or "No CPB Cardiovascular"):
				1. The conventional pre-procedural "time-out", which includes identification of patient, operative site, procedure, and history of any allergies
				2. A pre-procedural briefing wherein the surgeon shares with all members of the operating room team the essential elements of the operative plan; including diagnosis, planned procedure, outline of essentials of anesthesia and bypass strategies, antibiotic prophylaxis, availability of blood products, anticipated or planned implants or device applications, and anticipated challenges
				3. A post-procedural debriefing wherein the surgeon succinctly reviews with all members of the operating room team the essential elements of the operative plan, identifying both the successful components and the opportunities for improvement. This debriefing should take *place prior to the patient leaving the operating room or its equivalent*, and may be followed by a more in-depth dialogue involving team members at a later time. (The actual debriefing in the operating room is intentionally and importantly brief, in recognition of the fact that periods of transition may be times of instability or vulnerability for the patient)
				4. A briefing and execution of a hand-off protocol at the time of transfer (arrival) to the Intensive Care Unit at the end of the operation, involving the anesthesiologist, surgeon, physician staff of the Intensive Care Unit (including critical care and cardiology) and nursing

12	O-1	Outcome	Occurrence of new post-operative renal failure requiring dialysis	For each surgical admission (Index Cardiac Operation) code whether the complication occurred during the time interval beginning at admission to operating room and ending 30 days post-operatively or at the time of hospital discharge, whichever is longer (A Cardiac Operation is defined as an operation of operation type “CPB” or “No CPB Cardiovascular”): STS version 2.5: 220 = Acute renal failure requiring temporary dialysis 230 = Acute renal failure requiring permanent dialysis STS version 3.0: 230 = Renal failure – acute renal failure, acute renal failure requiring dialysis at the time of hospital discharge 223 = Renal failure–acute renal failure, acute renal failure requiring temporary dialysis with the need for dialysis not present at hospital discharge 224 = Renal failure–acute renal failure, acute renal failure requiring temporary hemofiltration with the need for dialysis not present at hospital discharge Please note: Unless a patient requires dialysis prior to surgery, renal failure that requires dialysis after surgery constitutes an operative complication, despite the fact that pre-operative diminished renal perfusion may have contributed to the development of this complication This measure will be reported as percentage of all Index Cardiac Operations. This measure will also be reported stratified by the 5 STS-EACTS Congenital Heart Surgery Mortality Categories. (STS is developing Congenital Heart Surgery Morbidity Categories. When these STS Congenital Heart Surgery Morbidity Categories are published and available, this metric will be stratified by the STS Congenital Heart Surgery Morbidity Categories instead of the STS Congenital Heart Surgery mortality categories)

(continued)

Table 8.4 (continued)

	Number	Type	Title of indicator	Description
13	O-2	Outcome	Occurrence of new post-operative neurological deficit persisting at discharge	For each surgical admission (Index Cardiac Operation) code whether the complication occurred during the time interval beginning at admission to operating room and ending 30 days post-operatively or at the time of hospital discharge, whichever is longer (A Cardiac Operation is defined as an operation of operation type "CPB" or "No CPB Cardiovascular"):
				320 = Neurological deficit, neurological deficit persisting at discharge
				This measure tracks "new post-operative neurological deficits" that (1) occur during the time interval beginning at admission to operating room and ending at the time of hospital discharge and (2) persist at discharge
				Such new post-operative neurological deficits may or may not be related to a stroke. If the new post-operative neurological deficit is the result of a stroke (that occurs during the time interval beginning at admission to operating room and ending at the time of hospital discharge) and the neurological deficit persists at discharge, then the following two complications should both be selected:
				320 = Neurological deficit, neurological deficit persisting at discharge
				420 = Stroke
				Thus, this complication (320 = Neurological deficit, neurological deficit persisting at discharge) should be coded in situations where a patient has a stroke (during the time interval beginning at admission to operating room and ending at the time of hospital discharge) and the neurological deficit persists at discharge
				This measure does not include a neurologic deficit (which may or may not be related to a stroke) that does not persist at discharge
				Please note that this complication (320 = Neurological deficit, neurological deficit persisting at discharge) should be coded even in the situation where the patient has a neurological deficit that is present prior to admission to operating room and this neurological deficit worsens (or a new neurological deficit develops) during the time interval beginning at admission to operating room and ending at the time of hospital discharge
				This measure will be reported as percentage of all Index Cardiac Operations. This measure will also be reported stratified by the 5 STS-EACTS Congenital Heart Surgery Mortality Categories. (STS is developing Congenital Heart Surgery Morbidity Categories. When these STS Congenital Heart Surgery Morbidity Categories are published and available, this metric will be stratified by the STS Congenital Heart Surgery Morbidity Categories instead of the STS Congenital Heart Surgery Mortality Categories)
14	O-3	Outcome	Occurrence of arrhythmia necessitating permanent pacemaker insertion	For each surgical admission (Index Cardiac Operation) code whether the complication occurred during the time interval beginning at admission to operating room and ending 30 days post-operatively or at the time of hospital discharge, whichever is longer (A Cardiac Operation is defined as an operation of operation type "CPB" or "No CPB Cardiovascular".):
				STS version 2.5:
				60 = Postoperative AV block requiring permanent pacemaker
				STS version 3.0:
				74 = Arrhythmia necessitating pacemaker, permanent pacemaker
				This measure will be reported as percentage of all Index Cardiac Operations. This measure will also be reported stratified by the 5 STS-EACTS Congenital Heart Surgery Mortality Categories. (STS is developing Congenital Heart Surgery Morbidity Categories. When these STS Congenital Heart Surgery Morbidity Categories are published and available, this metric will be stratified by the STS Congenital Heart Surgery Morbidity Categories instead of the STS Congenital Heart Surgery Mortality Categories)

15	O-4	Outcome	Occurrence of paralyzed diaphragm (possible phrenic nerve injury)	For each surgical admission (Index Cardiac Operation) code whether the complication occurred during the time interval beginning at admission to operating room and ending 30 days post-operatively or at the time of hospital discharge, whichever is longer (A Cardiac Operation is defined as an operation of operation type "CPB" or "No CPB Cardiovascular"):
				STS version 2.5:
				300=Phrenic nerve injury/paralyzed diaphragm
				STS version 3.0:
				300=Paralyzed diaphragm (possible phrenic nerve injury)
				This measure will be reported as percentage of all Index Cardiac Operations. This measure will also be reported stratified by the 5 STS-EACTS Congenital Heart Surgery Mortality Categories. (STS is developing Congenital Heart Surgery Morbidity Categories. When these STS Congenital Heart Surgery Morbidity Categories are published and available, this metric will be stratified by the STS Congenital Heart Surgery Morbidity Categories instead of the STS Congenital Heart Surgery Mortality Categories)
16	O-5	Outcome	Occurrence of need for Postoperative mechanical circulatory support (IABP, VAD, ECMO, or CPS)	For each surgical admission (Index Cardiac Operation) code whether the complication occurred during the time interval beginning at admission to operating room and ending 30 days post-operatively or at the time of hospital discharge, whichever is longer (A Cardiac Operation is defined as an operation of operation type "CPB" or "No CPB Cardiovascular".):
				STS version 2.5:
				40=Postoperative mechanical circulatory support (IABP, VAD, ECMO, or CPS)
				STS version 3.0:
				40=Postoperative/Postprocedural mechanical circulatory support (IABP, VAD, ECMO, or CPS)
				Please note that this complication should be coded even in the situation where the patient had preoperative mechanical circulatory support if the patient has mechanical circulatory support postoperatively at any time until 30 days post-operatively or the time of hospital discharge, whichever is longer
				This measure will be reported as percentage of all Index Cardiac Operations. This measure will also be reported stratified by the 5 STS-EACTS Congenital Heart Surgery Mortality Categories. (STS is developing Congenital Heart Surgery Morbidity Categories. When these STS Congenital Heart Surgery Morbidity Categories are published and available, this metric will be stratified by the STS Congenital Heart Surgery Morbidity Categories instead of the STS Congenital Heart Surgery Mortality Categories)

(continued)

Table 8.4 (continued)

	Number		Type	Title of indicator Description
17	O-6	Outcome	Occurrence of unplanned reoperation and/or interventional cardiovascular catheterization procedure	For each surgical admission (Index Cardiac Operation) code whether the complication occurred during the time interval beginning at admission to operating room and ending 30 days post-operatively or at the time of hospital discharge, whichever is longer (A Cardiac Operation is defined as an operation of operation type "CPB" or "No CPB Cardiovascular"):
				STS version 2.5:
				20 = Reoperation during this admission (unplanned reoperation)
				240 = Bleeding requiring reoperation
				STS version 3.0:
				22 = Unplanned cardiac reoperation during the postoperative or postprocedural time period
				24 = Unplanned interventional cardiovascular catheterization procedure during the postoperative or postprocedural time period
				26 = Unplanned non-cardiac reoperation during the postoperative or postprocedural time period
				240 = Bleeding, Requiring reoperation
				n.b. does not include delayed sternal closure
				This measure will be reported as percentage of all Index Cardiac Operations. This measure will also be reported stratified by the 5 STS-EACTS Congenital Heart Surgery Mortality Categories. (STS is developing Congenital Heart Surgery Morbidity Categories. When these STS Congenital Heart Surgery Morbidity Categories are published and available, this metric will be stratified by the STS Congenital Heart Surgery Morbidity Categories instead of the STS Congenital Heart Surgery Mortality Categories)
				This measure counts all patients who require any additional unplanned cardiac or non-cardiac operation and/or interventional cardiovascular catheterization procedure occurring (1) within 30 days after surgery or intervention in or out of the hospital, or (2) after 30 days during the same hospitalization subsequent to the operation or intervention
				A cardiac operation is defined as any operation that is of the Operation Type of "CPB" or "No CPB Cardiovascular"
				The following operations will always be coded as "Planned Reoperation": (1) Delayed Sternal Closure, (2) ECMO Decannulation, (3) VAD Decannulation, (4) Removal of Broviac catheter
				The following operations will always be coded as "Unplanned Reoperation": (1) Reoperation for bleeding, (2) Reoperation for infection, (3) Reoperation for hemodynamic instability, (4) Reoperation for initiation of ECMO or VAD, (5) Reoperation for residual or recurrent lesion
18	O-7	Outcome	Operative Mortality Stratified by the Five STS-EACTS Mortality Levels	Operative mortality stratified by the five STS-EACTS Mortality Levels, a multi-institutional validated complexity stratification tool
				See *J Thorac Cardiovasc Surg* 2009;138:1139–1153. O'Brien et al. **An empirically based tool for analyzing mortality associated with congenital heart surgery. Table 1, pp 1140–1146**
19	0–8	Outcome	Operative Mortality for Eight Benchmark Operations	Operative Mortality for Eight Benchmark Pediatric and Congenital Heart Operations:
				These 8 Eight Benchmark Pediatric and Congenital Heart Operations are tracked when they are the Primary Procedure of an Index Cardiac Operation. (A Cardiac Operation is defined as an operation of Operation Type "CPB" or "No CPB Cardiovascular")
				(These 8 Eight Benchmark Pediatric and Congenital Heart Operations are listed and described in this table in Measure Number S-5)

20	O-9	Outcome	Index Cardiac Operations Free of Mortality and Major Complication	"Index Cardiac Operations free of mortality and major complication" is defined as the percent of pediatric and congenital heart surgery Index Cardiac Operations free all of the following: (1) Operative mortality, (2) any one or more of the following major complications occurring or diagnosed during the time interval beginning at admission to operating room and ending 30 days post-operatively or at the time of hospital discharge, whichever is longer:
				(a) Renal failure. Acute renal failure requiring temporary or permanent dialysis (220, 230, 223, 224)
				STS version 2.5:
				220=Acute renal failure requiring temporary dialysis
				230=Acute renal failure requiring permanent dialysis
				STS version 3.0:
				230=Renal failure – acute renal failure, Acute renal failure requiring dialysis at the time of hospital discharge
				223=Renal failure – acute renal failure, Acute renal failure requiring temporary dialysis with the need for dialysis not present at hospital discharge
				224=Renal failure – acute renal failure, Acute renal failure requiring temporary hemofiltration with the need for dialysis not present at hospital discharge
				(b) Neurological deficit, neurological deficit persisting at discharge
				STS version 2.5:
				320=Postoperative neurological deficit persisting at discharge
				STS version 3.0:
				320=Neurological deficit, Neurological deficit persisting at discharge
				(c) Arrhythmia necessitating pacemaker, Permanent pacemaker (60, 74)
				STS version 2.5:
				60=Postoperative AV block requiring permanent pacemaker
				STS version 3.0:
				74=Arrhythmia necessitating pacemaker, Permanent pacemaker
				(d) ECMO/VAD. Postop mechanical circulatory support (IABP, VAD, ECMO or CPS) (40)
				STS version 2.5:
				40=Postoperative mechanical circulatory support (IABP, VAD, ECMO, or CPS)
				STS version 3.0:
				40=Postoperative/Postprocedural mechanical circulatory support (IABP, VAD, ECMO, or CPS)
				(e) Paralyzed diaphragm (possible phrenic nerve injury)
				STS version 2.5:
				300=Phrenic nerve injury/paralyzed diaphragm
				STS version 3.0:
				300=Paralyzed diaphragm (possible phrenic nerve injury)
				(f) Unplanned reoperation. (20, 22, 26 or 240)
				STS version 2.5:

(continued)

Table 8.4 (continued)

Number		Type	Title of indicator	Description
				20 = Reoperation during this admission (unplanned reoperation)
				240 = Bleeding requiring reoperation
				STS version 3.0:
				22 = Unplanned cardiac reoperation during the postoperative or postprocedural time period, exclusive of reoperation for bleeding
				24 = Unplanned interventional cardiovascular catheterization procedure during the postoperative or postprocedural time period
				26 = Unplanned non-cardiac reoperation during the postoperative or postprocedural time period
				240 = Bleeding, Requiring reoperation
				This measure will be reported as percentage of all Index Cardiac Operations. This measure will also be reported stratified by the 5 STS-EACTS Congenital Heart Surgery Mortality Categories. (STS is developing Congenital Heart Surgery Morbidity Categories. When these STS Congenital Heart Surgery Morbidity Categories are published and available, this metric will be stratified by the STS Congenital Heart Surgery Morbidity Categories instead of the STS Congenital Heart Surgery Mortality Categories)
21	0–10	Outcome	Operative survivors free of major complication	"Operative survivors free of major complication" is defined as the percent of all surviving (live at discharge and 30 days postoperatively) pediatric and congenital heart surgery index operations free all of the following itemized major complications:
				(a) Renal failure. Acute renal failure requiring temporary or permanent dialysis (220, 230, 223, 224)
				STS version 2.5:
				220 = Acute renal failure requiring temporary dialysis
				230 = Acute renal failure requiring permanent dialysis
				STS version 3.0:
				230 = Renal failure – acute renal failure, Acute renal failure requiring dialysis at the time of hospital discharge
				223 = Renal failure – acute renal failure, Acute renal failure requiring temporary dialysis with the need for dialysis not present at hospital discharge
				224 = Renal failure – acute renal failure, Acute renal failure requiring temporary hemofiltration with the need for dialysis not present at hospital discharge
				(b) Neurological deficit, Neurological deficit persisting at discharge
				STS version 2.5:
				320 = Postoperative neurological deficit persisting at discharge
				STS version 3.0:
				320 = Neurological deficit, Neurological deficit persisting at discharge
				(c) Arrhythmia necessitating pacemaker, Permanent pacemaker (60, 74)
				STS version 2.5:
				60 = Postoperative AV block requiring permanent pacemaker
				STS version 3.0:
				74 = Arrhythmia necessitating pacemaker, permanent pacemaker
				(d) ECMO/VAD. Postop mechanical circulatory support (IABP, VAD, ECMO or CPS) (40)
				STS version 2.5:
				40 = Postoperative mechanical circulatory support (IABP, VAD, ECMO, or CPS)

	STS version 3.0:
	40 = Postoperative/Postprocedural mechanical circulatory support (IABP, VAD, ECMO, or CPS)
	(e) Paralyzed diaphragm (possible phrenic nerve injury)
	STS version 2.5:
	300 = Phrenic nerve injury/paralyzed diaphragm
	STS version 3.0:
	300 = Paralyzed diaphragm (possible phrenic nerve injury)
	(f) Unplanned reoperation. (20, 22, 26 or 240)
	STS version 2.5:
	20 = Reoperation during this admission (unplanned reoperation)
	240 = Bleeding requiring reoperation
	STS version 3.0:
	22 = Unplanned cardiac reoperation during the postoperative or postprocedural time period, exclusive of reoperation for bleeding
	24 = Unplanned interventional cardiovascular catheterization procedure during the postoperative or postprocedural time period
	26 = Unplanned non-cardiac reoperation during the postoperative or postprocedural time period
	240 = Bleeding, requiring reoperation
	This measure will be reported as percentage of all Index Cardiac Operations. This measure will also be reported stratified by the 5 STS-EACTS Congenital Heart Surgery Mortality Categories. (STS is developing Congenital Heart Surgery Morbidity Categories. When these STS Congenital Heart Surgery Morbidity Categories are published and available, this metric will be stratified by the STS Congenital Heart Surgery Morbidity Categories instead of the STS Congenital Heart Surgery Mortality Categories)

Table 8.5 Consensus definitions of the morbidities

Measure	Organ system	Complication	Definitions
12	Renal	Renal failure – acute renal failure, Acute renal failure requiring dialysis at the time of hospital discharge	Renal failure – acute renal failure (ROOT Definition) + With new postoperative/postprocedural requirement for dialysis, including peritoneal dialysis and/or hemodialysis. Code this complication if the patient requires dialysis at the time of hospital discharge or death in the hospital. (This complication should be chosen only if the dialysis was associated with acute renal failure.) {"Renal failure – acute renal failure" ROOT Definition = Acute renal failure is defined as new onset oliguria with sustained urine output < 0.5 cc/kg/h for 24 h and/or a rise in creatinine >1.5 times upper limits of normal for age (or twice the most recent preoperative/preprocedural values if these are available), with eventual need for dialysis (including peritoneal dialysis and/or hemodialysis) or hemofiltration. Acute renal failure that will be counted as an operative or procedural complication must occur prior to hospital discharge or after hospital discharge but within 30 days of the procedure. (An operative or procedural complication is any complication, regardless of cause, occurring (1) within 30 days after surgery or intervention in or out of the hospital, or (2) after 30 days during the same hospitalization subsequent to the operation or intervention. Operative and procedural complications include both intraoperative/intraprocedural complications and postoperative/postprocedural complications in this time interval.) The complication is to be coded even if the patient required dialysis, but the treatment was not instituted due to patient or family refusal}
12	Renal	Renal failure – acute renal failure, Acute renal failure requiring temporary dialysis with the need for dialysis not present at hospital discharge	Renal failure – acute renal failure (ROOT Definition) + With new postoperative/postprocedural requirement for temporary dialysis, including peritoneal dialysis and/or hemodialysis. Code this complication if the patient does not require dialysis at the time of hospital discharge or death in the hospital. (This complication should be chosen only if the dialysis was associated with acute renal failure.) {"Renal failure – acute renal failure" ROOT Definition = Acute renal failure is defined as new onset oliguria with sustained urine output <0.5 cc/kg/h for 24 h and/or a rise in creatinine >1.5 times upper limits of normal for age (or twice the most recent preoperative/preprocedural values if these are available), with eventual need for dialysis (including peritoneal dialysis and/or hemodialysis) or hemofiltration. Acute renal failure that will be counted as an operative or procedural complication must occur prior to hospital discharge or after hospital discharge but within 30 days of the procedure. (An operative or procedural complication is any complication, regardless of cause, occurring (1) within 30 days after surgery or intervention in or out of the hospital, or (2) after 30 days during the same hospitalization subsequent to the operation or intervention. Operative and procedural complications include both intraoperative/intraprocedural complications and postoperative/postprocedural complications in this time interval.) The complication is to be coded even if the patient required dialysis, but the treatment was not instituted due to patient or family refusal}

Table 8.5 (continued)

12	Renal	Renal failure – acute renal failure, Acute renal failure requiring temporary hemofiltration with the need for dialysis not present at hospital discharge	Renal failure – acute renal failure (ROOT Definition) + With new postoperative/postprocedural requirement for temporary hemofiltration. Code this complication if the patient does not require dialysis at the time of hospital discharge or death in the hospital. (This complication should be chosen only if the hemofiltration was associated with acute renal failure.) {"Renal failure – acute renal failure" ROOT Definition = Acute renal failure is defined as new onset oliguria with sustained urine output <0.5 cc/kg/h for 24 h and/or a rise in creatinine >1.5 times upper limits of normal for age (or twice the most recent preoperative/preprocedural values if these are available), with eventual need for dialysis (including peritoneal dialysis and/or hemodialysis) or hemofiltration. Acute renal failure that will be counted as an operative or procedural complication must occur prior to hospital discharge or after hospital discharge but within 30 days of the procedure. (An operative or procedural complication is any complication, regardless of cause, occurring (1) within 30 days after surgery or intervention in or out of the hospital, or (2) after 30 days during the same hospitalization subsequent to the operation or intervention. Operative and procedural complications include both intraoperative/intraprocedural complications and postoperative/postprocedural complications in this time interval.) The complication is to be coded even if the patient required dialysis, but the treatment was not instituted due to patient or family refusal}
13	Neurologic	Neurological deficit, Neurological deficit persisting at discharge	Newly recognized and/or newly acquired deficit of neurologic function leading to inpatient referral, therapy, or intervention not otherwise practiced for a similar unaffected inpatient, With a persisting neurologic deficit present at hospital discharge. In other words, new (onset intraoperatively or postoperatively – or intraprocedurally or postprocedurally) neurological deficit persisting and present at discharge from hospital
13	Neurologic	Stroke	"Stroke" ROOT Definition = A stroke is any confirmed neurological deficit of abrupt onset caused by a disturbance in blood flow to the brain, when the neurologic deficit does not resolve within 24 h
13	Neurologic	Spinal cord injury, Neurological deficit persisting at discharge	Spinal cord injury (ROOT Definition) + With a persisting neurologic deficit present at hospital discharge. {"Spinal cord injury" ROOT Definition = Newly acquired or newly recognized deficit of spinal cord function indicated by physical exam findings, imaging studies, or both}
13	Neurologic	Peripheral nerve injury, Neurological deficit persisting at discharge	Peripheral nerve injury (ROOT Definition) + With a persisting neurologic deficit present at hospital discharge. {"Peripheral nerve injury" ROOT Definition = Newly acquired or newly recognized deficit of unilateral or bilateral peripheral nerve function indicated by physical exam findings, imaging studies, or both}
14	Arrhythmia – Arrhythmia necessitating pacemaker	Arrhythmia necessitating pacemaker, Permanent pacemaker	Implantation and utilization of a permanent pacemaker for treatment of any arrhythmia including heart block (atrioventricular [AV] heart block)

(continued)

Table 8.5 (continued)

Measure	Organ system	Complication	Definitions
15	Neurologic	Paralyzed diaphragm (possible phrenic nerve injury)	Presence of elevated hemi-diaphragm(s) on chest radiograph in conjunction with evidence of weak, immobile, or paradoxical movement assessed by ultrasound or fluoroscopy
16	Mechanical support utilization	Postoperative/Postprocedural mechanical circulatory support (IABP, VAD, ECMO, or CPS)	Utilization of postoperative/postprocedural mechanical support, of any type (IABP, VAD, ECMO, or CPS), for resuscitation/CPR or support, during the postoperative/postprocedural time period. Code this complication if it occurs (1) within 30 days after surgery or intervention regardless of the date of hospital discharge, or (2) after 30 days during the same hospitalization subsequent to the operation or intervention
17	Operative/procedural	Unplanned cardiac reoperation during the postoperative or postprocedural time period, exclusive of reoperation for bleeding	Any additional unplanned cardiac operation occurring (1) within 30 days after surgery or intervention in or out of the hospital, or (2) after 30 days during the same hospitalization subsequent to the operation or intervention. A cardiac operation is defined as any operation that is of the operation type of "CPB" or "No CPB Cardiovascular". The following operations will always be coded as "Planned Reoperation": (1) Delayed Sternal Closure, (2) ECMO Decannulation, (3) VAD Decannulation, (4) Removal of Broviac catheter. The following operations will always be coded as "Unplanned Reoperation": (1) Reoperation for bleeding, (2) Reoperation for infection, (3) Reoperation for hemodynamic instability, (4) Reoperation for initiation of ECMO or VAD, (5) Reoperation for residual or recurrent lesion
17	Operative/procedural	Unplanned interventional cardiovascular catheterization procedure during the postoperative or postprocedural time period	Any unplanned interventional cardiovascular catheterization procedure occurring (1) within 30 days after surgery or intervention in or out of the hospital, or (2) after 30 days during the same hospitalization subsequent to the operation or intervention
17	Operative/procedural	Unplanned non-cardiac reoperation during the postoperative or postprocedural time period	Any additional unplanned non-cardiac operation occurring (1) within 30 days after surgery or intervention in or out of the hospital, or (2) after 30 days during the same hospitalization subsequent to the operation or intervention

registries have been used to create standardized measures of quality that have been endorsed by multiple professional medical societies and the National Quality Forum [186, 250]. Compliance with these measures and the public reporting of these measures should lead to improvements in the overall quality of care delivered to our patients [143, 176, 177].

Figure 8.7 is a Venn Diagram that demonstrates the close and overlapping relationships between the three domains of this textbook: Outcomes Analysis, Quality Improvement, and Patient Safety. These relationships compose the underlying theme of this textbook and are fundamental to improving the state of the art of pediatric and congenital cardiac care.

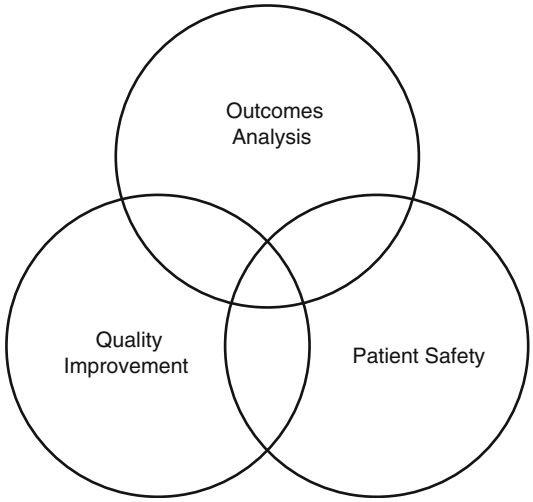


Fig. 8.7 This Venn Diagram demonstrates the close and overlapping relationships between the three domains of this textbook: outcomes analysis, quality improvement, and patient care. These relationships compose the underlying theme of this textbook and are fundamental to improving the state of the art of pediatric and congenital cardiac care

References

1. Mavroudis C (Chairman) and Congenital Database Subcommittee: Backer CL, Bove E, Burke RP, Cameron D, Drinkwater DC, Edwards FH, Grover FL, Hammon JW Jr, Jacobs JP, Kron IL, Mayer JE, Myers JL, Ring WS, Siewers RD, Szarnicki RJ, Watson DC Jr. Data analyses of the society of thoracic surgeons national congenital cardiac surgery database. 1994–1997, Summit Medical, Minnetonka; Sept 1998.
2. Jacobs JP, Jacobs ML, Mavroudis C, Lacour-Gayet FG. Executive summary: the society of thoracic surgeons congenital heart surgery database – second harvest – (1998–2001) beta site test. The Society of Thoracic Surgeons (STS) and Duke Clinical Research Institute (DCRI), Duke University Medical Center, Durham; Fall 2002 Harvest.
3. Jacobs JP, Jacobs ML, Mavroudis C, Lacour-Gayet FG. Executive summary: the society of thoracic surgeons congenital heart surgery database – third harvest – (1998–2002). The Society of Thoracic Surgeons (STS) and Duke Clinical Research Institute (DCRI), Duke University Medical Center, Durham; Spring 2003 Harvest.
4. Jacobs JP, Jacobs ML, Mavroudis C, Lacour-Gayet FG. Executive summary: the society of thoracic surgeons congenital heart surgery database – fourth harvest – (2002–2003). The Society of Thoracic Surgeons (STS) and Duke Clinical Research Institute (DCRI), Duke University Medical Center, Durham; Spring 2004 Harvest.
5. Jacobs JP, Jacobs ML, Mavroudis C, Lacour-Gayet FG. Executive summary: The Society of Thoracic Surgeons Congenital Heart Surgery Database – fifth harvest – (2002–2004). The Society of Thoracic Surgeons (STS) and Duke Clinical Research Institute (DCRI), Duke University Medical Center, Durham; Spring 2005 Harvest.
6. Jacobs JP, Jacobs ML, Mavroudis C, Lacour-Gayet FG, Tchervenkov CI. Executive summary: The Society of Thoracic Surgeons Congenital Heart Surgery Database – sixth harvest – (2002–2005). The Society of Thoracic Surgeons (STS) and Duke Clinical Research Institute (DCRI), Duke University Medical Center, Durham; Spring 2006 Harvest.
7. Jacobs JP, Jacobs ML, Mavroudis C, Lacour-Gayet FG, Tchervenkov CI. Executive summary: The Society of Thoracic Surgeons Congenital Heart Surgery Database – seventh harvest – (2003–2006). The Society of Thoracic Surgeons (STS) and Duke Clinical Research Institute (DCRI), Duke University Medical Center, Durham; Spring 2007 Harvest.
8. Jacobs JP, Jacobs ML, Mavroudis C, Lacour-Gayet FG, Tchervenkov CI. Executive summary: The Society of Thoracic Surgeons Congenital Heart Surgery Database – eighth harvest – (January 1, 2004–December 31, 2007). The Society of Thoracic Surgeons (STS) and Duke Clinical Research Institute (DCRI), Duke University Medical Center, Durham; Spring 2008 Harvest.
9. Jacobs JP, Jacobs ML, Mavroudis C, Lacour-Gayet FG, Tchervenkov CI. Executive summary: The Society of Thoracic Surgeons Congenital Heart Surgery Database – ninth harvest – (July 1, 2004–June 30, 2008). The Society of Thoracic Surgeons (STS) and Duke Clinical Research Institute (DCRI), Duke University Medical Center, Durham; Fall 2008 Harvest.
10. Jacobs JP, Jacobs ML, Mavroudis C, Lacour-Gayet FG, Tchervenkov CI. Executive summary: The Society of Thoracic Surgeons Congenital Heart Surgery Database – tenth harvest – (January 1, 2005–December 31, 2008). The Society of Thoracic Surgeons (STS) and Duke Clinical Research Institute (DCRI), Duke University Medical Center, Durham; Spring 2009 Harvest.
11. Jacobs JP, Jacobs ML, Mavroudis C, Lacour-Gayet FG, Tchervenkov CI. Executive summary: The Society of Thoracic Surgeons Congenital Heart Surgery Database – eleventh harvest – (July 1, 2005–June 30, 2009). The Society of Thoracic Surgeons (STS) and Duke Clinical Research Institute (DCRI), Duke University Medical Center; Fall 2009 Harvest.
12. Jacobs JP, Jacobs ML, Mavroudis C, Lacour-Gayet FG, Tchervenkov CI. Executive summary: The Society of Thoracic Surgeons Congenital Heart Surgery Database – twelfth harvest – (January 1, 2006–December 31, 2009). The Society of Thoracic Surgeons (STS) and Duke Clinical Research Institute (DCRI), Duke University Medical Center; Spring 2010 Harvest.

13. Jacobs JP, Jacobs ML, Mavroudis C, Lacour-Gayet FG, Tchervenkov CI. Executive summary: The Society of Thoracic Surgeons Congenital Heart Surgery Database – thirteenth harvest – (July 1, 2006–June 30, 2010). The Society of Thoracic Surgeons (STS) and Duke Clinical Research Institute (DCRI), Duke University Medical Center, Durham; Fall 2010 Harvest.
14. Jacobs JP, Jacobs ML, Mavroudis C, Lacour-Gayet FG, Tchervenkov CI. Executive summary: The Society of Thoracic Surgeons Congenital Heart Surgery Database – fourteenth harvest – (January 1, 2007–December 31, 2010). The Society of Thoracic Surgeons (STS) and Duke Clinical Research Institute (DCRI), Duke University Medical Center, Durham; Spring 2011 Harvest.
15. Jacobs JP, Jacobs ML, Mavroudis C, Lacour-Gayet FG, Tchervenkov CI. Executive summary: The Society of Thoracic Surgeons Congenital Heart Surgery Database – fifteenth harvest – (July 1, 2007–June 30, 2011). The Society of Thoracic Surgeons (STS) and Duke Clinical Research Institute (DCRI), Duke University Medical Center, Durham; Fall 2011 Harvest.
16. Jacobs JP, Jacobs ML, Mavroudis C, Lacour-Gayet FG, Tchervenkov CI. Executive summary: The Society of Thoracic Surgeons Congenital Heart Surgery Database – sixteenth harvest – (January 1, 2008–December 31, 2011). The Society of Thoracic Surgeons (STS) and Duke Clinical Research Institute (DCRI), Duke University Medical Center, Durham; Spring 2012 Harvest.
17. Jacobs JP, Jacobs ML, Mavroudis C, Lacour-Gayet FG, Tchervenkov CI. Executive summary: The Society of Thoracic Surgeons Congenital Heart Surgery Database – seventeenth harvest – (July 1, 2008–June 30, 2012). The Society of Thoracic Surgeons (STS) and Duke Clinical Research Institute (DCRI), Duke University Medical Center, Durham; Fall 2012 Harvest.
18. Jacobs JP, Jacobs ML, Mavroudis C, Lacour-Gayet FG, Tchervenkov CI, Pasquali SK. Executive summary: The Society of Thoracic Surgeons Congenital Heart Surgery Database – eighteenth harvest – (January 1, 2009–December 31, 2012). The Society of Thoracic Surgeons (STS) and Duke Clinical Research Institute (DCRI), Duke University Medical Center, Durham; Spring 2013 Harvest.
19. Jacobs JP, Jacobs ML, Mavroudis C, Lacour-Gayet FG, Tchervenkov CI, Pasquali SK. Executive summary: The Society of Thoracic Surgeons Congenital Heart Surgery Database – nineteenth harvest – (July 1, 2008–June 30, 2013). The Society of Thoracic Surgeons (STS) and Duke Clinical Research Institute (DCRI), Duke University Medical Center, Durham; Fall 2013 Harvest.
20. Mavroudis C, Gevitz M, Ring WS, McIntosh C, Schwartz M. The Society of Thoracic Surgeons National Congenital Cardiac Surgery database. Ann Thorac Surg. 1999;68:601–24.
21. Mavroudis C, Jacobs JP. The International Congenital Heart Surgery Nomenclature and Database Project. Ann Thorac Surg. 2000;(Suppl):S1–372.
22. Mavroudis C, Jacobs JP. Congenital Heart Surgery Nomenclature and Database Project: Introduction. In: Mavroudis C, Jacobs JP, editors. The Annals of Thoracic Surgery April 2000 Supplement: The International Congenital Heart Surgery Nomenclature and Database Project. The Annals of Thoracic Surgery, April 2000, Supplement, S1.
23. Mavroudis C, Jacobs JP. Congenital Heart Surgery Nomenclature and Database Project: Overview and Minimum Dataset. In: Mavroudis C, Jacobs JP, editors. The Annals of Thoracic Surgery April 2000 Supplement: The International Congenital Heart Surgery Nomenclature and Database Project. The Annals of Thoracic Surgery, April 2000, Supplement, S2–17.
24. Jacobs JP, Quintessenza JA, Burke RP, Mavroudis C. Congenital Heart Surgery Nomenclature and Database Project: atrial septal defect. In: Mavroudis C, Jacobs JP, editors. The Annals of Thoracic Surgery April 2000 Supplement: The International Congenital Heart Surgery Nomenclature and Database Project. The Annals of Thoracic Surgery, April 2000, Supplement, S18–24.
25. Jacobs JP, Burke RP, Quintessenza JA, Mavroudis C. Congenital Heart Surgery Nomenclature and Database Project: ventricular septal defect. In: Mavroudis C, Jacobs JP, editors. The Annals of Thoracic Surgery April 2000 Supplement: The International Congenital Heart Surgery Nomenclature and Database Project. The Annals of Thoracic Surgery, April 2000, Supplement, S25–35.
26. Jacobs JP, Burke RP, Quintessenza JA, Mavroudis C. Congenital Heart Surgery Nomenclature and Database Project: Atrioventricular Canal Defect. In: Mavroudis C, Jacobs JP, editors. The Annals of Thoracic Surgery April 2000 Supplement: The International Congenital Heart Surgery Nomenclature and Database Project. The Annals of Thoracic Surgery, April 2000, Supplement, S36–43.
27. Jacobs JP, Quintessenza JA, Gaynor JW, Burke RP, Mavroudis C. Congenital Heart Surgery Nomenclature and Database Project: aortopulmonary window. In: Mavroudis C, Jacobs JP, editors. The Annals of Thoracic Surgery April 2000 Supplement: The International Congenital Heart Surgery Nomenclature and Database Project. The Annals of Thoracic Surgery, April 2000, Supplement, S44–9.
28. Jacobs M. Congenital Heart Surgery Nomenclature and Database Project: truncus arteriosus. In: Mavroudis C, Jacobs JP, editors. The Annals of Thoracic Surgery April 2000 Supplement: The International Congenital Heart Surgery Nomenclature and Database Project. The Annals of Thoracic Surgery, April 2000, Supplement, S50–55.
29. Herlong JR, Jaggers JJ, Ungerleider RM. Congenital Heart Surgery Nomenclature and Database Project:

pulmonary venous anomalies. In: Mavroudis C, Jacobs JP, editors. The Annals of Thoracic Surgery April 2000 Supplement: The International Congenital Heart Surgery Nomenclature and Database Project. The Annals of Thoracic Surgery, April 2000, Supplement, S56–69.
30. Gaynor JW, Weinberg P, Spray T. Congenital Heart Surgery Nomenclature and Database Project: Systemic Venous Anomalies. In: Mavroudis C, Jacobs JP, editors. The Annals of Thoracic Surgery April 2000 Supplement: The International Congenital Heart Surgery Nomenclature and Database Project. The Annals of Thoracic Surgery, April 2000, Supplement, S70–6.
31. Jacobs M. Congenital Heart Surgery Nomenclature and Database Project: tetralogy of Fallot. In: Mavroudis C, Jacobs JP, editors. The Annals of Thoracic Surgery April 2000 Supplement: The International Congenital Heart Surgery Nomenclature and Database Project. The Annals of Thoracic Surgery, April 2000, Supplement, S77–82.
32. Lacour-Gayet F. Congenital Heart Surgery Nomenclature and Database Project: right ventricular outflow tract obstruction – intact ventricular septum. In: Mavroudis C, Jacobs JP, editors. The Annals of Thoracic Surgery April 2000 Supplement: The International Congenital Heart Surgery Nomenclature and Database Project. The Annals of Thoracic Surgery, April 2000, Supplement, S83–96.
33. Tchervenkov CI, Roy N. Congenital Heart Surgery Nomenclature and Database Project: pulmonary atresia – ventricular septal defect. In: Mavroudis C, Jacobs JP, editors. The Annals of Thoracic Surgery April 2000 Supplement: The International Congenital Heart Surgery Nomenclature and Database Project. The Annals of Thoracic Surgery, April 2000, Supplement, S97–105.
34. Dearani JA, Danielson GK. Congenital Heart Surgery Nomenclature and Database Project: Ebstein's anomaly and tricuspid valve disease. In: Mavroudis C, Jacobs JP, editors. The Annals of Thoracic Surgery April 2000 Supplement: The International Congenital Heart Surgery Nomenclature and Database Project. The Annals of Thoracic Surgery, April 2000, Supplement, S106–17.
35. Nguyen KH. Congenital Heart Surgery Nomenclature and Database Project: aortic valve disease. In: Mavroudis C, Jacobs JP, editors. The Annals of Thoracic Surgery April 2000 Supplement: The International Congenital Heart Surgery Nomenclature and Database Project. The Annals of Thoracic Surgery, April 2000, Supplement, S118–31.
36. Mitruka SN, Lamberti JJ. Congenital Heart Surgery Nomenclature and Database Project: mitral valve disease. In: Mavroudis C, Jacobs JP, editors. The Annals of Thoracic Surgery April 2000 Supplement: The International Congenital Heart Surgery Nomenclature and Database Project. The Annals of Thoracic Surgery, April 2000, Supplement, S132–46.
37. Ring WS. Congenital Heart Surgery Nomenclature and Database Project: aortic aneurysm, sinus of valsalva aneurysm, and aortic dissection. In: Mavroudis C, Jacobs JP, editors. The Annals of Thoracic Surgery April 2000 Supplement: The International Congenital Heart Surgery Nomenclature and Database Project. The Annals of Thoracic Surgery, April 2000, Supplement, S147–63.
38. Myers JL, Mehta SM. Congenital Heart Surgery Nomenclature and Database Project: aortico-left ventricular tunnel. In: Mavroudis C, Jacobs JP, editors. The Annals of Thoracic Surgery April 2000 Supplement: The International Congenital Heart Surgery Nomenclature and Database Project. The Annals of Thoracic Surgery, April 2000, Supplement, S164–9.
39. Tchervenkov CI, Jacobs M, Tahta SA. Congenital Heart Surgery Nomenclature and Database Project: hypoplastic left heart syndrome. In: Mavroudis C, Jacobs JP, editors. The Annals of Thoracic Surgery April 2000 Supplement: The International Congenital Heart Surgery Nomenclature and Database Project. The Annals of Thoracic Surgery, April 2000, Supplement, S170–9.
40. Delius RE. Congenital Heart Surgery Nomenclature and Database Project: pediatric cardiomyopathies and end-stage congenital heart disease. In: Mavroudis C, Jacobs JP, editors. The Annals of Thoracic Surgery April 2000 Supplement: The International Congenital Heart Surgery Nomenclature and Database Project. The Annals of Thoracic Surgery, April 2000, Supplement, S180–90.
41. Myers JL, Mehta SM. Congenital Heart Surgery Nomenclature and Database Project: diseases of the pericardium. In: Mavroudis C, Jacobs JP, editors. The Annals of Thoracic Surgery April 2000 Supplement: The International Congenital Heart Surgery Nomenclature and Database Project. The Annals of Thoracic Surgery, April 2000, Supplement, S191–6.
42. Jacobs M, Mayer JE. Congenital Heart Surgery Nomenclature and Database Project: single ventricle. In: Mavroudis C, Jacobs JP, editors. The Annals of Thoracic Surgery April 2000 Supplement: The International Congenital Heart Surgery Nomenclature and Database Project. The Annals of Thoracic Surgery, April 2000, Supplement, S197–204.
43. Jaggers JJ, Cameron DE, Herlong JR, Ungerleider RM. Congenital Heart Surgery Nomenclature and Database Project: transposition of the great arteries. In: Mavroudis C, Jacobs JP, editors. The Annals of Thoracic Surgery April 2000 Supplement: The International Congenital Heart Surgery Nomenclature and Database Project. The Annals of Thoracic Surgery, April 2000, Supplement, S205–35.
44. Wilkinson JL, Cochrane AD, Karl TR. Congenital Heart Surgery Nomenclature and Database Project: corrected (discordant) transposition of the great

arteries (and related malformations). In: Mavroudis C, Jacobs JP, editors. The Annals of Thoracic Surgery April 2000 Supplement: The International Congenital Heart Surgery Nomenclature and Database Project. The Annals of Thoracic Surgery, April 2000, Supplement, S236–48.
45. Walters III HW, Mavroudis C, Tchervenkov CI, Jacobs JP, Lacour-Gayet F, Jacobs ML. Congenital Heart Surgery Nomenclature and Database Project: double outlet right ventricle. In: Mavroudis C, Jacobs JP, editors. The Annals of Thoracic Surgery April 2000 Supplement: The International Congenital Heart Surgery Nomenclature and Database Project. The Annals of Thoracic Surgery, April 2000, Supplement, S249–63.
46. Tchervenkov CI, Walters III HW, Chu VF. Congenital Heart Surgery Nomenclature and Database Project: double outlet left ventricle. In: Mavroudis C, Jacobs JP, editors. The Annals of Thoracic Surgery April 2000 Supplement: The International Congenital Heart Surgery Nomenclature and Database Project. The Annals of Thoracic Surgery, April 2000, Supplement, S264–9.
47. Dodge-Khatami A, Mavroudis C, Backer CL. Congenital Heart Surgery Nomenclature and Database Project: anomalies of the coronary arteries. In: Mavroudis C, Jacobs JP, editors. The Annals of Thoracic Surgery April 2000 Supplement: The International Congenital Heart Surgery Nomenclature and Database Project. The Annals of Thoracic Surgery, April 2000, Supplement, S270–97.
48. Backer CL, Mavroudis C. Congenital Heart Surgery Nomenclature and Database Project: patent ductus arteriosus, coarctation of the aorta, and interrupted aortic arch. In: Mavroudis C, Jacobs JP, editors. The Annals of Thoracic Surgery April 2000 Supplement: The International Congenital Heart Surgery Nomenclature and Database Project. The Annals of Thoracic Surgery, April 2000, Supplement, S298–307.
49. Backer CL, Mavroudis C. Congenital Heart Surgery Nomenclature and Database Project: vascular rings, tracheal stenosis, and pectus excavatum. In: Mavroudis C, Jacobs JP, editors. The Annals of Thoracic Surgery April 2000 Supplement: The International Congenital Heart Surgery Nomenclature and Database Project. The Annals of Thoracic Surgery, April 2000, Supplement, S308–18.
50. Deal BJ, Jacobs JP, Mavroudis C. Congenital Heart Surgery Nomenclature and Database Project: arrhythmias. In: Mavroudis C, Jacobs JP, editors. The Annals of Thoracic Surgery April 2000 Supplement: The International Congenital Heart Surgery Nomenclature and Database Project. The Annals of Thoracic Surgery, April 2000, Supplement, S319–31.
51. Rocchini AP. Congenital Heart Surgery Nomenclature and Database Project: therapeutic cardiac catheter interventions. In: Mavroudis C, Jacobs JP, editors. The Annals of Thoracic Surgery April 2000 Supplement: The International Congenital Heart Surgery Nomenclature and Database Project. The Annals of Thoracic Surgery, April 2000, Supplement, S332–42.
52. Gaynor JW, Bridges ND, Spray T. Congenital Heart Surgery Nomenclature and Database Project: end-stage lung disease. In: Mavroudis C, Jacobs JP, editors. The Annals of Thoracic Surgery April 2000 Supplement: The International Congenital Heart Surgery Nomenclature and Database Project. The Annals of Thoracic Surgery, April 2000, Supplement, S343–57.
53. Mehta SM, Myers JL. Congenital Heart Surgery Nomenclature and Database Project: cardiac tumors. In: Mavroudis C, Jacobs JP, editors. The Annals of Thoracic Surgery April 2000 Supplement: The International Congenital Heart Surgery Nomenclature and Database Project. The Annals of Thoracic Surgery, April 2000, Supplement, S358–68.
54. Joffs C, Sade RM. Congenital Heart Surgery Nomenclature and Database Project: palliation, correction, or repair. In: Mavroudis C, Jacobs JP, editors. The Annals of Thoracic Surgery April 2000 Supplement: The International Congenital Heart Surgery Nomenclature and Database Project. The Annals of Thoracic Surgery, April 2000, Supplement, S369–72.
55. Lacour-Gayet F, Maruszewski B, Mavroudis C, Jacobs JP, Elliott MJ. Presentation of the International Nomenclature for Congenital Heart Surgery – the long way from nomenclature to collection of validated data at the EACTS. Eur J Cardiothorac Surg. 2000;18(2):128–35.
56. Jenkins KJ, Gauvreau K, Newburger JW, Spray TL, Moller JH, Iezzoni LI. Consensus-based method for risk adjustment for surgery for congenital heart disease. J Thorac Cardiovasc Surg. 2002;123:110–8.
57. Mavroudis C, Jacobs JP. Congenital heart disease outcome analysis: methodology and rationale. J Thorac Cardiovasc Surg. 2002;123(1):6–7.
58. Mavroudis C, Gevitz M, Elliott MJ, Jacobs JP, Gold JP. Virtues of a worldwide congenital heart surgery database. Semin Thorac Cardiovasc Surg Pediatr Card Surg Annu. 2002;5(1):126–31.
59. Williams WG, McCrindle BW. Practical experience with databases for congenital heart disease: a registry versus an academic database. Semin Thorac Cardiovasc Surg Pediatr Card Surg Annu. 2002;5(1):132–42.
60. Maruszewski B, Tobota Z. The European congenital heart defects surgery database experience: pediatric European cardiothoracic surgical registry of the European Association for Cardio-Thoracic Surgery. Semin Thorac Cardiovasc Surg Pediatr Card Surg Annu. 2002;5(1):143–7.
61. Lacour-Gayet F. Risk stratification theme for congenital heart surgery. Semin Thorac Cardiovasc Surg Pediatr Card Surg Annu. 2002;5(1):148–52.

62. Jacobs JP. Software development, nomenclature schemes, and mapping strategies for an International Pediatric Cardiac Surgery Database System. Semin Thorac Cardiovasc Surg Pediatr Card Surg Annu. 2002;5(1):153–62.
63. Maruszewski B, Lacour-Gayet F, Elliott MJ, Gaynor JW, Jacobs JP, Jacobs ML, Tchervenkov CI, Kurosawa H, Mavroudis C. Congenital Heart Surgery Nomenclature and Database Project: update and proposed data harvest. For simultaneous publication: The Annals of Thoracic Surgery 2002, The European Journal of Cardio-thoracic Surgery 2002, and The Jpn J Thorac Cardiovasc Surg 2002. The European Journal of Cardio-thoracic Surgery, 21(1):47–9, January 2002.
64. Gaynor JW, Jacobs JP, Jacobs ML, Elliott MJ, Lacour-Gayet F, Tchervenkov CI, Maruszewski B, Mavroudis C. Congenital Heart Surgery Nomenclature and Database Project: update and proposed data harvest. For simultaneous publication: The Annals of Thoracic Surgery 2002, The European Journal of Cardio-thoracic Surgery 2002, and The Jpn J Thorac Cardiovasc Surg 2002. The Annals of Thoracic Surgery, 73(3):1016–8, March 2002.
65. Jenkins KJ, Gauvreau K. Center-specific differences in mortality: preliminary analyses using the Risk Adjustment in Congenital Heart Surgery (RACHS-1) method. J Thorac Cardiovasc Surg. 2002;124:97–104.
66. Franklin RCG, Jacobs JP, Tchervenkov CI, Béland M. Report from the Executive of The International Working Group for Mapping and Coding of Nomenclatures for Paediatric and Congenital Heart Disease: Bidirectional Crossmap of the Short Lists of the European Paediatric Cardiac Code and the International Congenital Heart Surgery Nomenclature and Database Project. Cardiol Young. 2002;(Suppl II):8–22.
67. Franklin RCG, Jacobs JP, Tchervenkov CI, Béland M. European Paediatric Cardiac Code Short List crossmapped to STS/EACTS Short List with ICD-9 & ICD-10 crossmapping. Cardiol Young. 2002;(Suppl II):23–49.
68. Franklin RCG, Jacobs JP, Tchervenkov CI, Béland M. STS/EACTS Short List mapping to European Paediatric Cardiac Code Short List with ICD-9 & ICD-10 crossmapping. Cardiol Young. 2002;(Suppl II):50–62.
69. Béland M, Jacobs JP, Tchervenkov CI, Franklin RCG. The International Nomenclature Project for Paediatric and Congenital Heart Disease: report from the Executive of The International Working Group for Mapping and Coding of Nomenclatures for Paediatric and Congenital Heart Disease. Cardiol Young. 2002;12:425–30.
70. Franklin RCG, Jacobs JP, Tchervenkov CI, Béland M. The International Nomenclature Project for Pediatric and Congenital Heart Disease: bidirectional crossmap of the short lists of the European Paediatric Cardiac Code and the International Congenital Heart Surgery Nomenclature and Database Project. Cardiol Young. 2002;12:431–5.
71. Kurosawa H, Gaynor JW, Jacobs JP, Jacobs ML, Elliott MJ, Lacour-Gayet F, Tchervenkov CI, Maruszewski B, Mavroudis C. Congenital Heart Surgery Nomenclature and Database Project: update and proposed data harvest. For simultaneous publication: The Annals of Thoracic Surgery 2002, The European Journal of Cardio-thoracic Surgery 2002, and The Jpn J Thorac Cardiovasc Surg 2002. The Jpn J Thorac Cardiovasc Surg, 50(11):498–501, November 2002.
72. Allen SW, Gauvreau K, Bloom BT, Jenkins KJ. Evidence-based referral results in significantly reduced mortality after congenital heart surgery. Pediatrics. 2003;112(1 Pt 1):24–8.
73. Jenkins KJ. Risk adjustment for congenital heart surgery: the RACHS-1 method. Semin Thorac Cardiovasc Surg Pediatr Card Surg Annu. 2004;7:180–4.
74. Lacour-Gayet FG, Clarke D, Jacobs JP, Gaynor JW, Hamilton L, Jacobs ML, Maruszewski B, Pozzi M, Spray T, Tchervenkov CI, Mavroudis C, Aristotle Committee. The Aristotle score for congenital heart surgery. Semin Thorac Cardiovasc Surg Pediatr Card Surg Annu. 2004;7:185–91.
75. Béland MJ, Franklin RCG, Jacobs JP, Tchervenkov CI, Aiello VD, Colan SD, Gaynor JW, Krogmann ON, Kurosawa H, Maruszewski B, Stellin G, Weinberg PM. Update from The International Working Group for Mapping and Coding of Nomenclatures for Paediatric and Congenital Heart Disease. Cardiol Young. 2004;14(2):225–9.
76. Lacour-Gayet FG, Clarke D, Jacobs JP, Comas J, Daebritz S, Daenen W, Gaynor JW, Hamilton L, Jacobs ML, Maruszewski B, Pozzi M, Spray T, Stellin G, Tchervenkov CI, Mavroudis C, Aristotle Committee. The Aristotle score: a complexity-adjusted method to evaluate surgical results. Eur J Cardiothorac Surg. 2004;25(6):911–24.
77. Jacobs JP, Mavroudis C, Jacobs ML, Lacour-Gayet FG, Tchervenkov CI, Gaynor JW, Clarke DR, Spray TL, Maruszewski B, Stellin G, Elliott MJ, Dokholyan RS, Peterson ED. Lessons learned from the data analysis of the second harvest (1998–2001) of The Society of Thoracic Surgeons (STS) congenital heart surgery database. Eur J Cardiothorac Surg. 2004;26(1):18–37.
78. Boethig D, Jenkins KJ, Hecker H, Thies WR, Breymann T. The RACHS-1 risk categories reflect mortality and length of hospital stay in a large German pediatric cardiac surgery population. Eur J Cardiothorac Surg. 2004;26:12–7.
79. Welke KF, Jacobs JP, Jenkins KJ. Evaluation of quality of care for congenital heart disease. Semin Thorac Cardiovasc Surg Pediatr Card Surg Annu. 2005;8:157–67.
80. Jacobs JP, Elliott MJ, Anderson RH, Quintessenza JA, Chai PJ, Morell VO, Botero LM, van Gelder HM, Badhwar V, Kanani M, Cohen GA, Burke

RP. Creating a Database with Cardioscopy and Intra-Operative Imaging. In: Jacobs JP, Wernovsky G, Gaynor JW, Anderson RH, editors. 2005 Supplement to Cardiology in the Young: Controversies of the Ventriculo-Arterial Junctions and Other Topics. Cardiology in the Young, vol. 15, Supplement 1:184–9, February 2005.

81. Jacobs JP, Maruszewski B, Tchervenkov CI, Lacour-Gayet FG, Jacobs ML, Clarke DR, Gaynor JW, Spray TL, Stellin G, Elliott MJ, Ebels T, Franklin RCG, Béland MJ, Kurosawa H, Aiello VD, Colan SD, Krogmann ON, Weinberg P, Tobota Z, Dokholyan RS, Peterson ED, Mavroudis C. The Current Status and Future Directions of Efforts to create a Global Database for the Outcomes of Therapy for Congenital Heart Disease. In: Jacobs JP, Wernovsky G, Gaynor JW, Anderson RH, editors. 2005 Supplement to Cardiology in the Young: Controversies of the Ventriculo-Arterial Junctions and Other Topics. Cardiology in the Young, vol. 15, Supplement 1:190–8, February 2005.
82. Jacobs JP, Lacour-Gayet FG, Jacobs ML, Clarke DR, Tchervenkov CI, Gaynor JW, Spray TL, Maruszewski B, Stellin G, Gould J, Dokholyan RS, Peterson ED, Elliott MJ, Mavroudis C. Initial application in the STS congenital database of complexity adjustment to evaluate surgical case mix and results. Ann Thorac Surg. 2005;79(5):1635–49; discussion 1635–49.
83. Lacour-Gayet F, Clarke DR, Aristotle Committee. The Aristotle method: a new concept to evaluate quality of care based on complexity. Curr Opin Pediatr. 2005;17(3):412–7.
84. Lacour-Gayet F, Jacobs JP, Clarke DR, Gaynor JW, Jacobs ML, Anderson RH, Elliott MJ, Maruszewski B, Vouhe P, Mavroudis C. Performance of surgery for congenital heart disease: shall we wait a generation or look for different statistics? J Thorac Cardiovasc Surg. 2005;130(1):234–5.
85. Maruszewski B, Lacour-Gayet F, Monro JL, Keogh BE, Tobota Z, Kansy A. An attempt at data verification in the EACTS congenital database. Eur J Cardiothorac Surg. 2005;28(3):400–4; discussion 405–6.
86. Jacobs ML. Editorial comment. Eur J Cardiothorac Surg. 2005;28(3):405–6.
87. Jacobs JP, Maruszewski B, The European Association for Cardio-Thoracic Surgery (EACTS) and The Society of Thoracic Surgeons (STS) Joint Congenital Heart Surgery Nomenclature and Database Committee. Computerized outcomes analysis for congenital heart disease. Curr Opin Pediatr. 2005;17(5):586–91.
88. Jacobs JP, Jacobs ML, Maruszewski B, Lacour-Gayet FG, Clarke DR, Tchervenkov CI, Gaynor JW, Spray TL, Stellin G, Elliott MJ, Ebels T, Mavroudis C. Current status of the European Association for Cardio-Thoracic Surgery and the Society of Thoracic Surgeons Congenital Heart Surgery database. Ann Thorac Surg. 2005;80(6):2278–83; discussion 2283–4.
89. Larsen SH, Pedersen J, Jacobsen J, Johnson SP, Hansen OK, Hjortdal V. The RACHS-1 risk categories reflect mortality and length of stay in a Danish population of children operated for congenital heart disease. Eur J Cardiothorac Surg. 2005;28:877–81.
90. Miyamoto T, Sinzobahamvya N, Kumpikaite D, et al. Repair of truncus arteriosus and aortic arch interruption: outcome analysis. Ann Thorac Surg. 2005;79:2077–82.
91. Maruszewski B, Tobota Z, Kansy A, Lacour-Gayet F, Jacobs JP, Clark D, Elliott MJ. The Aristotle score methodology for evaluation of outcomes in congenital heart surgery. Standardy Med Pediatr. 2005;7 Suppl 22:29–33.
92. Jacobs JP, Franklin RCG, Jacobs ML, Colan SD, Tchervenkov CI, Maruszewski B, Gaynor JW, Spray TL, Stellin G, Aiello VD, Béland MJ, Krogmann ON, Kurosawa H, Weinberg PM, Elliott MJ, Mavroudis C, Anderson R. Classification of the Functionally Univentricular Heart: Unity from mapped codes. In: Jacobs JP, Wernovsky G, Gaynor JW, Anderson RH, editors. 2006 Supplement to Cardiology in the Young: Controversies and Challenges in the Management of the Functionally Univentricular Heart. Cardiology in the Young, vol. 16, Supplement 1:9–21, February 2006.
93. Jacobs JP, Mavroudis C, Jacobs ML, Maruszewski B, Tchervenkov CI, Lacour-Gayet FG, Clarke DR, Yeh T, Walters 3rd HL, Kurosawa H, Stellin G, Ebels T, Elliott MJ. What is operative mortality? Defining death in a surgical registry database: a report from the STS congenital database task force and the joint EACTS-STS congenital database committee. Ann Thorac Surg. 2006;81(5):1937–41.
94. Tchervenkov CI, Jacobs JP, Weinberg PM, Aiello VD, Béland MJ, Colan SD, Elliott MJ, Franklin RC, Gaynor JW, Krogmann ON, Kurosawa H, Maruszewski B, Stellin G. The nomenclature, definition and classification of hypoplastic left heart syndrome. Cardiol Young. 2006;16(4):339–68.
95. Jacobs ML, Mavroudis C, Jacobs JP, Tchervenkov CI, Pelletier GJ. Report of the 2005 STS Congenital Heart Surgery Practice and Manpower survey: a report from the STS work force on congenital heart surgery. Ann Thorac Surg. 2006;82(3):1152–8, 1159.e1–5; discussion 1158–9.
96. Jacobs JP, Franklin RCG, Wilkinson JL, Cochrane AD, Karl TR, Aiello VD, Béland MJ, Colan SD, Elliott, MJ, Gaynor JW, Krogmann ON, Kurosawa H, Maruszewski B, Stellin G, Tchervenkov CI, Weinberg PM. The nomenclature, definition and classification of discordant atrioventricular connections. In: Jacobs JP, Wernovsky G, Gaynor JW, Anderson RH, editors. 2006 Supplement to Cardiology in the Young: Controversies and Challenges of the Atrioventricular Junctions and Other Challenges Facing Paediatric Cardiovascular Practitioners and their Patients. Cardiology in the Young, vol. 16 (Supplement 3):72–84, September 2006.

97. Sinzobahamvya N, Photiadis J, Kumpikaite D, et al. Comprehensive Aristotle score: implications for the Norwood procedure. Ann Thorac Surg. 2006;81:1794–800.
98. Atrip JH, Campbell DN, Ivy DD, et al. Birth weight and complexity are significant factors for the management of hypoplastic left heart syndrome. Ann Thorac Surg. 2006;82:1252–7; discussion 1258–9.
99. Al-Radi OO, Harrell Jr FE, Caldarone CA, McCrindle BW, Jacobs JP, Williams MG, Van Arsdell GS, Williams WG. Case complexity scores in congenital heart surgery: a comparative study of the Aristotle basic complexity score and the Risk Adjustment in Congenital Heart Surgery (RACHS-1) system. J Thorac Cardiovasc Surg. 2007;133(4): 865–75. Epub 2007.
100. Jacobs JP, Mavroudis C, Jacobs ML, Maruszewski B, Tchervenkov CI, Lacour-Gayet FG, Clarke DR, Gaynor JW, Spray TL, Kurosawa H, Stellin G, Ebels T, Bacha EA, Walters HL, Elliott MJ. Nomenclature and databases – the past, the present, and the future: a primer for the congenital heart surgeon. Pediatr Cardiol. 2007;28(2):105–15. Epub 2007.
101. Lacour-Gayet F, Jacobs ML, Jacobs JP, Mavroudis C. The need for an objective evaluation of morbidity in congenital heart surgery. Ann Thorac Surg. 2007;84(1):1–2.
102. Lacour-Gayet FG, Jacobs JP, Clarke DR, Maruszewski B, Jacobs ML, O'Brien SM, Mavroudis C. Evaluation of quality of care in congenital heart surgery: contribution of the Aristotle complexity score. Adv Pediatr. 2007;54:67–83. PMID: 17918467, 2007.
103. Jacobs JP, Anderson RH, Weinberg P, Walters III HL, Tchervenkov CI, Del Duca D, Franklin RCG, Aiello VD, Béland MJ, Colan SD, Gaynor JW, Krogmann ON, Kurosawa H, Maruszewski B, Stellin G, Elliott MJ. The nomenclature, definition and classification of cardiac structures in the setting of heterotaxy. In: Anderson RH, Jacobs JP, Wernovsky G, editors. 2007 Supplement to Cardiology in the Young: Controversies and Challenges Facing Paediatric Cardiovascular Practitioners and their Patients. Cardiology in the Young, vol. 17, Supplement 2, p. 1–28, doi:10.1017/S1047951107001138, September 2007.
104. Jacobs JP, Wernovsky G, Elliott MJ. Analysis of outcomes for congenital cardiac disease: can we do better? Cardiol Young. 2007;17 Suppl 2:145–58. doi:10.1017/S1047951107001278.
105. Jacobs JP, Jacobs ML, Mavroudis C, Maruszewski B, Tchervenkov CI, Lacour-Gayet FG, Clarke DR, Yeh T, Walters 3rd HL, Kurosawa H, Stellin G, Ebels T, Elliott MJ, Vener DF, Barach P, Benavidez OJ, Bacha EA. What is operative morbidity? Defining complications in a surgical registry database: a report from the STS congenital database task force and the joint EACTS-STS congenital database committee. Ann Thorac Surg. 2007;84:1416–21.
106. O'Brien SM, Jacobs JP, Clarke DR, Maruszewski B, Jacobs ML, Walters III HL, Tchervenkov CI, Welke KF, Tobota Z, Stellin G, Mavroudis C, Lacour-Gayet FG. Accuracy of the Aristotle basic complexity score for classifying the mortality and morbidity potential of congenital heart surgery procedures. Ann Thorac Surg. 2007;84(6):2027–37. PMID: 18036930.
107. Derby CD, Kolcz J, Kerins PJ, Duncan DR, Quezada E, Pizarro C. Aristotle score predicts outcome in patients requiring extracorporeal circulatory support following repair of congenital heart disease. ASAIO J. 2007;53:82–6.
108. Curzon CL, Milford-Beland S, Li JS, O'Brien SM, Jacobs JP, Jacobs ML, Welke KF, Lodge AJ, Peterson ED, Jaggers J. Cardiac surgery in infants with low birth weight is associated with increased mortality: analysis of the Society of Thoracic Surgeons Congenital Heart Database. J Thorac Cardiovasc Surg. 2008;135(3):546–51. Epub 2008.
109. Jacobs JP, Haan CK, Edwards FH, Anderson RP, Grover FL, Mayer JE Jr, Chitwood WR Jr. Editorial for Ann Thorac Surg: the rationale for incorporation of HIPAA compliant unique patient, surgeon, and hospital identifier fields in the STS database. Ann Thorac Surg. 2008;86(3):695–8. doi:10.1016/j.athoracsur.2008.04.075.
110. Jacobs JP, editor. 2008 Cardiology in the Young Supplement: Databases and The Assessment of Complications associated with The Treatment of Patients with Congenital Cardiac Disease. Prepared by: The Multi-Societal Database Committee for Pediatric and Congenital Heart Disease, Cardiology in the Young, vol. 18, Supplement S2, p. 1–530, December 9, 2008.
111. Jacobs JP. Introduction – Databases and the assessment of complications associated with the treatment of patients with congenital cardiac disease. In: JP Jacobs, editor. 2008 Cardiology in the Young Supplement: Databases and The Assessment of Complications associated with The Treatment of Patients with Congenital Cardiac Disease. Prepared by: The Multi-Societal Database Committee for Pediatric and Congenital Heart Disease. Cardiology in the Young, vol. 18, issue S2 (Suppl 2), p. 1–37, December 9, 2008.
112. Jacobs JP, Jacobs ML, Mavroudis C, Backer CL, Lacour-Gayet FG, Tchervenkov CI, Franklin RCG, Béland MJ, Jenkins KJ, Walters III H, Bacha EA, Maruszewski B, Kurosawa H, Clarke DR, Gaynor JW, Spray TL, Stellin G, Ebels T, Krogmann ON, Aiello VD, Colan SD, Weinberg P, Giroud JM, Everett A, Wernovsky G, Martin J, Elliott MJ, Edwards FH. Nomenclature and databases for the surgical treatment of congenital cardiac disease – an updated primer and an analysis of opportunities for improvement. In: Jacobs JP, editor. 2008 Cardiology in the Young Supplement: Databases and The Assessment of Complications associated with The Treatment of Patients with Congenital Cardiac Disease. Prepared by: The Multi-Societal Database Committee for Pediatric and Congenital Heart

Disease. Cardiology in the Young, vol. 18, issue S2 (Suppl 2), p. 38–62, December 9, 2008.
113. Tchervenkov CI, Jacobs JP, Bernier P-L, Stellin G, Kurosawa H, Mavroudis C, Jonas RA, Cicek SM, Al-Halees Z, Elliott MJ, Jatene MB, Kinsley RH, Kreutzer C, Leon-Wyss J, Liu J, Maruszewski B, Nunn GR, Ramirez-Marroquin S, Sandoval N, Sano S, Sarris GE, Sharma R, Shoeb A, Spray TL, Ungerleider RM, Yangni-Angate H, Ziemer G. The improvement of care for paediatric and congenital cardiac disease across the World: a challenge for the World Society for Pediatric and Congenital Heart Surgery. In: JP Jacobs, editor. 2008 Cardiology in the Young Supplement: Databases and The Assessment of Complications associated with The Treatment of Patients with Congenital Cardiac Disease. Prepared by: The Multi-Societal Database Committee for Pediatric and Congenital Heart Disease. Cardiology in the Young, vol. 18, issue S2 (Suppl 2), p. 63–9, December 9, 2008.
114. Franklin RCG, Jacobs JP, Krogmann ON, Béland MJ, Aiello VD, Colan SD, Elliott MJ, Gaynor JW, Kurosawa H, Maruszewski B, Stellin G, Tchervenkov CI, Walters HL 3rd, Weinberg P, Anderson RH. Nomenclature for congenital and paediatric cardiac disease: historical perspectives and The International Pediatric and Congenital Cardiac Code. In: Jacobs JP, editor. 2008 Cardiology in the Young Supplement: Databases and The Assessment of Complications associated with The Treatment of Patients with Congenital Cardiac Disease. Prepared by: The Multi-Societal Database Committee for Pediatric and Congenital Heart Disease. Cardiology in the Young, vol. 18, issue S2 (Suppl 2), p. 70–80, December 9, 2008.
115. Jacobs JP, Benavidez OJ, Bacha EA, Walters HL 3rd, Jacobs ML. The nomenclature of safety and quality of care for patients with congenital cardiac disease: a report of the Society of Thoracic Surgeons Congenital Database Taskforce Subcommittee on Patient Safety. In: Jacobs JP, editor. 2008 Cardiology in the Young Supplement: Databases and The Assessment of Complications associated with The Treatment of Patients with Congenital Cardiac Disease. Prepared by: The Multi-Societal Database Committee for Pediatric and Congenital Heart Disease. Cardiology in the Young, vol. 18, issue S2 (Suppl 2), p. 81–91, December 9, 2008.
116. Strickland MJ, Riehle-Colarusso TJ, Jacobs JP, Reller MD, Mahle WT, Botto LD, Tolbert PE, Jacobs ML, Lacour-Gayet FG, Tchervenkov CI, Mavroudis C, Correa A. The importance of nomenclature for congenital cardiac disease: implications for research and evaluation. In: Jacobs JP, editor. 2008 Cardiology in the Young Supplement: Databases and The Assessment of Complications associated with The Treatment of Patients with Congenital Cardiac Disease. Prepared by: The Multi-Societal Database Committee for Pediatric and Congenital Heart Disease. Cardiology in the Young, vol. 18, issue S2 (Suppl 2), p. 92–100, December 9, 2008.
117. Jacobs ML, Jacobs JP, Franklin RCG, Mavroudis C, Lacour-Gayet F, Tchervenkov CI, Walters III H, Bacha EA, Clarke DR, Gaynor JW, Spray TL, Stellin G, Ebels T, Maruszewski B, Tobota Z, Kurosawa H, Elliott M. Databases for assessing the outcomes of the treatment of patients with congenital and paediatric cardiac disease – the perspective of cardiac surgery. In: Jacobs JP, editor. 2008 Cardiology in the Young Supplement: Databases and The Assessment of Complications associated with The Treatment of Patients with Congenital Cardiac Disease. Prepared by: The Multi-Societal Database Committee for Pediatric and Congenital Heart Disease. Cardiology in the Young, vol. 18, issue S2 (Suppl 2), p. 101–15, December 9, 2008.
118. Jenkins KJ, Beekman III RH, Bergersen LJ, Everett AD, Forbes TJ, Franklin RCG, Klitzner TS, Krogmann ON, Martin GR, Webb CL. Databases for assessing the outcomes of the treatment of patients with congenital and paediatric cardiac disease – the perspective of cardiology. In: Jacobs JP, editor. 2008 Cardiology in the Young Supplement: Databases and The Assessment of Complications associated with The Treatment of Patients with Congenital Cardiac Disease. Prepared by: The Multi-Societal Database Committee for Pediatric and Congenital Heart Disease. Cardiology in the Young, vol. 18, issue S2 (Suppl 2), p. 116–23, December 9, 2008.
119. Vener DF, Jacobs JP, Schindler E, Maruszewski B, Andropoulos D. Databases for assessing the outcomes of the treatment of patients with congenital and paediatric cardiac disease – the perspective of anaesthesia. In: Jacobs JP, editor. 2008 Cardiology in the Young Supplement: Databases and The Assessment of Complications associated with The Treatment of Patients with Congenital Cardiac Disease. Prepared by: The Multi-Societal Database Committee for Pediatric and Congenital Heart Disease. Cardiology in the Young, vol. 18, issue S2 (Suppl 2), p. 124–9, December 9, 2008.
120. LaRovere JM, Jeffries HE, Sachdeva RC, Rice TB, Wetzel RC, Cooper DS, Bird GL, Ghanayem NS, Checchia PA, Chang AC, Wessel DL. Databases for assessing the outcomes of the treatment of patients with congenital and paediatric cardiac disease – the perspective of critical care. In: Jacobs JP, editor. 2008 Cardiology in the Young Supplement: Databases and The Assessment of Complications associated with The Treatment of Patients with Congenital Cardiac Disease. Prepared by: The Multi-Societal Database Committee for Pediatric and Congenital Heart Disease. Cardiology in the Young, vol. 18, issue S2 (Suppl 2), p. 130–6, December 9, 2008.
121. Welke KF, Karamlou T, Diggs BS. Databases for assessing the outcomes of the treatment of patients with congenital and paediatric cardiac disease – a

comparison of administrative and clinical data. In: Jacobs JP, editor. 2008 Cardiology in the Young Supplement: Databases and The Assessment of Complications associated with The Treatment of Patients with Congenital Cardiac Disease. Prepared by: The Multi-Societal Database Committee for Pediatric and Congenital Heart Disease. Cardiology in the Young, vol. 18, issue S2 (Suppl 2), p. 137–44, December 9, 2008.

122. O'Brien SM, Gauvreau K. Statistical issues in the analysis and interpretation of outcomes for congenital cardiac surgery. In: Jacobs JP, editor. 2008 Cardiology in the Young Supplement: Databases and The Assessment of Complications associated with The Treatment of Patients with Congenital Cardiac Disease. Prepared by: The Multi-Societal Database Committee for Pediatric and Congenital Heart Disease. Cardiology in the Young, vol. 18, issue S2 (Suppl 2), p. 145–51, December 9, 2008.
123. Hickey EJ, McCrindle BW, Caldarone CA, Williams WG, Blackstone EH. Making sense of congenital cardiac disease with a research database: the Congenital Heart Surgeons' Society Data Center. In: Jacobs JP, editor. 2008 Cardiology in the Young Supplement: Databases and The Assessment of Complications associated with The Treatment of Patients with Congenital Cardiac Disease. Prepared by: The Multi-Societal Database Committee for Pediatric and Congenital Heart Disease. Cardiology in the Young, vol. 18, issue S2 (Suppl 2), p. 152–62, December 9, 2008.
124. Jacobs ML, Jacobs JP, Jenkins KJ, Gauvreau K, Clarke DR, Lacour-Gayet FL. Stratification of complexity: the Risk Adjustment for Congenital Heart Surgery-1 Method and The Aristotle Complexity Score – past, present, and future. In: Jacobs JP, editor. 2008 Cardiology in the Young Supplement: Databases and The Assessment of Complications associated with The Treatment of Patients with Congenital Cardiac Disease. Prepared by: The Multi-Societal Database Committee for Pediatric and Congenital Heart Disease. Cardiology in the Young, vol. 18, issue S2 (Suppl 2), p. 163–8, December 9, 2008.
125. Clarke DR, Lacour-Gayet F, Jacobs JP, Jacobs ML, Maruszewski B, Pizarro C, Edwards FH, Mavroudis C. The assessment of complexity in congenital cardiac surgery based on objective data. In: Jacobs JP, editor. 2008 Cardiology in the Young Supplement: Databases and The Assessment of Complications associated with The Treatment of Patients with Congenital Cardiac Disease. Prepared by: The Multi-Societal Database Committee for Pediatric and Congenital Heart Disease. Cardiology in the Young, vol. 18, issue S2 (Suppl 2), p. 169–76, December 9, 2008.
126. Clarke DR, Breen LS, Jacobs ML, Franklin RCG, Tobota Z, Maruszewski B, Jacobs JP. Verification of data in congenital cardiac surgery. In: Jacobs JP, editor. 2008 Cardiology in the Young Supplement: Databases and The Assessment of Complications associated with The Treatment of Patients with Congenital Cardiac Disease. Prepared by: The Multi-Societal Database Committee for Pediatric and Congenital Heart Disease. Cardiology in the Young, vol. 18, issue S2 (Suppl 2), p. 177–87, December 9, 2008.
127. Morales DLS, McClellan AJ, Jacobs JP. Empowering a database with national long-term data about mortality: the use of national death registries. In: Jacobs JP, editor. 2008 Cardiology in the Young Supplement: Databases and The Assessment of Complications associated with The Treatment of Patients with Congenital Cardiac Disease. Prepared by: The Multi-Societal Database Committee for Pediatric and Congenital Heart Disease. Cardiology in the Young, vol. 18, issue S2 (Suppl 2), p. 188–95, December 9, 2008.
128. Bacha EA, Cooper D, Thiagarajan R, Franklin RCG, Krogmann O, Deal B, Mavroudis C, Shukla A, Yeh Jr T, Barach P, Wessel D, Stellin G, Colan SD. Cardiac complications associated with the treatment of patients with congenital cardiac disease: consensus definitions from the Multi-Societal Database Committee for Pediatric and Congenital Heart Disease. In: Jacobs JP, editor. 2008 Cardiology in the Young Supplement: Databases and The Assessment of Complications associated with The Treatment of Patients with Congenital Cardiac Disease. Prepared by: The Multi-Societal Database Committee for Pediatric and Congenital Heart Disease. Cardiology in the Young, vol. 18, issue S2 (Suppl 2), p. 196–201, December 9, 2008.
129. Deal BJ, Mavroudis C, Jacobs JP, Gevitz M, Backer CL. Arrhythmic complications associated with the treatment of patients with congenital cardiac disease: consensus definitions from the Multi-Societal Database Committee for Pediatric and Congenital Heart Disease. In: Jacobs JP, editor. 2008 Cardiology in the Young Supplement: Databases and The Assessment of Complications associated with The Treatment of Patients with Congenital Cardiac Disease. Prepared by: The Multi-Societal Database Committee for Pediatric and Congenital Heart Disease. Cardiology in the Young, vol. 18, issue S2 (Suppl 2), p. 202–5, December 9, 2008.
130. Shann KG, Giacomuzzi CR, Harness L, Myers GJ, Paugh TA, Mellas N, Groom RC, Gomez D, Thuys CA, Charette K, Ojito JW, Tinius-Juliani J, Calaritis C, McRobb CM, Parpard M, Chancy T, Bacha E, Cooper DS, Jacobs JP, Likosky DS, on behalf of the International Consortium for Evidence-Based Perfusion. Complications relating to perfusion and extracorporeal circulation associated with the treatment of patients with congenital cardiac disease: Consensus Definitions from the Multi-Societal Database Committee for Pediatric and Congenital Heart Disease. In: Jacobs JP, editor. 2008 Cardiology

in the Young Supplement: Databases and The Assessment of Complications associated with The Treatment of Patients with Congenital Cardiac Disease. Prepared by: The Multi-Societal Database Committee for Pediatric and Congenital Heart Disease. Cardiology in the Young, vol. 18, issue S2 (Suppl 2), p. 206–14, December 9, 2008.

131. Cooper DS, Jacobs JP, Chai PJ, Jaggers JJ, Barach P, Beekman III RH, Krogmann O, Manning P. Pulmonary complications associated with the treatment of patients with congenital cardiac disease: consensus definitions from the Multi-Societal Database Committee for Pediatric and Congenital Heart Disease. In: Jacobs JP, editor. 2008 Cardiology in the Young Supplement: Databases and The Assessment of Complications associated with The Treatment of Patients with Congenital Cardiac Disease. Prepared by: The Multi-Societal Database Committee for Pediatric and Congenital Heart Disease. Cardiology in the Young, vol. 18, issue S2 (Suppl 2), p. 215–21, December 9, 2008.
132. Welke KW, Dearani JA, Ghanayem NS, Béland MJ, Shen I, Ebels T. Renal complications associated with the treatment of patients with congenital cardiac disease: consensus definitions from the Multi-Societal Database Committee for Pediatric and Congenital Heart Disease. In: Jacobs JP, editor. 2008 Cardiology in the Young Supplement: Databases and The Assessment of Complications associated with The Treatment of Patients with Congenital Cardiac Disease. Prepared by: The Multi-Societal Database Committee for Pediatric and Congenital Heart Disease. Cardiology in the Young, vol. 18, issue S2 (Suppl 2), p. 222–5, December 9, 2008.
133. Checchia PA, Karamlou T, Maruszewski B, Ohye RG, Bronicki R, Dodge-Khatami A. Haematological and infectious complications associated with the treatment of patients with congenital cardiac disease: consensus definitions from the Multi-Societal Database Committee for Pediatric and Congenital Heart Disease. In: Jacobs JP, editor. 2008 Cardiology in the Young Supplement: Databases and The Assessment of Complications associated with The Treatment of Patients with Congenital Cardiac Disease. Prepared by: The Multi-Societal Database Committee for Pediatric and Congenital Heart Disease. Cardiology in the Young, vol. 18, issue S2 (Suppl 2), p. 226–33, December 9, 2008.
134. Bird GL, Jeffries HE, Licht DJ, Wernovsky G, Weinberg PM, Pizarro C, Stellin G. Neurological complications associated with the treatment of patients with congenital cardiac disease: consensus definitions from the Multi-Societal Database Committee for Pediatric and Congenital Heart Disease. In: Jacobs JP, editor. 2008 Cardiology in the Young Supplement: Databases and The Assessment of Complications associated with The Treatment of Patients with Congenital Cardiac Disease. Prepared by: The Multi-Societal Database Committee for Pediatric and Congenital Heart Disease. Cardiology in the Young, vol. 18, issue S2 (Suppl 2), p. 234–9, December 9, 2008.
135. Ghanayem NS, Dearani JA, Welke KF, Béland MJ, Shen I, Ebels T. Gastrointestinal complications associated with the treatment of patients with congenital cardiac disease: consensus definitions from the Multi-Societal Database Committee for Pediatric and Congenital Heart Disease. In: Jacobs JP, editor. 2008 Cardiology in the Young Supplement: Databases and The Assessment of Complications associated with The Treatment of Patients with Congenital Cardiac Disease. Prepared by: The Multi-Societal Database Committee for Pediatric and Congenital Heart Disease. Cardiology in the Young, vol. 18, issue S2 (Suppl 2), p. 240–4, December 9, 2008.
136. Walters HL 3rd, Jeffries HE, Cohen GA, Klitzner T. Congenital cardiac surgical complications of the integument, vascular system, vascular-line(s), and wounds: consensus definitions from the Multi-Societal Database Committee for Pediatric and Congenital Heart Disease. In: Jacobs JP, editor. 2008 Cardiology in the Young Supplement: Databases and The Assessment of Complications associated with The Treatment of Patients with Congenital Cardiac Disease. Prepared by: The Multi-Societal Database Committee for Pediatric and Congenital Heart Disease. Cardiology in the Young, vol. 18, issue S2 (Suppl 2), p. 245–55, December 9, 2008.
137. Dickerson H, Cooper DS, Checchia PA, Nelson DP. Endocrinal complications associated with the treatment of patients with congenital cardiac disease: consensus definitions from the Multi-Societal Database Committee for Pediatric and Congenital Heart Disease. In: Jacobs JP, editor. 2008 Cardiology in the Young Supplement: Databases and The Assessment of Complications associated with The Treatment of Patients with Congenital Cardiac Disease. Prepared by: The Multi-Societal Database Committee for Pediatric and Congenital Heart Disease. Cardiology in the Young, vol. 18, issue S2 (Suppl 2), p. 256–64, December 9, 2008.
138. Jeffries H, Bird G, Law Y, Wernovsky G, Weinberg P, Pizarro C, Stellin G. Complications related to the transplantation of thoracic organs: consensus definitions from the Multi-Societal Database Committee for Pediatric and Congenital Heart Disease. In: Jacobs JP, editor. 2008 Cardiology in the Young Supplement: Databases and The Assessment of Complications associated with The Treatment of Patients with Congenital Cardiac Disease. Prepared by: The Multi-Societal Database Committee for Pediatric and Congenital Heart Disease. Cardiology in the Young, vol. 18, issue S2 (Suppl 2), p. 265–70, December 9, 2008.
139. Vener DV, Tirotta CF, Andropoulos D, Barach P. Anaesthetic complications associated with the treatment of patients with congenital cardiac disease: consensus definitions from the Multi-Societal Database Committee for Pediatric and Congenital

Heart Disease. In: Jacobs JP, editor. 2008 Cardiology in the Young Supplement: Databases and The Assessment of Complications associated with The Treatment of Patients with Congenital Cardiac Disease. Prepared by: The Multi-Societal Database Committee for Pediatric and Congenital Heart Disease. Cardiology in the Young, vol. 18, issue S2 (Suppl 2), p. 265–270, December 9, 2008.

140. The Multi-Societal Database Committee for Pediatric and Congenital Heart Disease. Part IV – the dictionary of definitions of complications associated with the treatment of patients with congenital cardiac disease. In: Jacobs JP, editor. 2008 Cardiology in the Young Supplement: Databases and the Assessment of Complications associated with the Treatment of Patients with Congenital Cardiac Disease. Prepared by: The Multi-Societal Database Committee for Pediatric and Congenital Heart Disease. Cardiology in the Young, vol. 18, issue S2 (Suppl 2), p. 282–530, December 9, 2008.
141. Tsang VT, Brown KL, Synnergren MJ, Kang N, de Leval MR, Gallivan S, Utley M. Monitoring risk-adjusted outcomes in congenital heart surgery: does the appropriateness of a risk model change with time? Ann Thorac Surg. 2009;87(2):584–7.
142. Jacobs JP, Shahian DM, Jacobs ML, Mavroudis C. Invited commentary of "monitoring risk-adjusted outcomes in congenital heart surgery: does the appropriateness of a risk model change with time?". Ann Thorac Surg. 2009;87(2):587–8. PMID: 19161784.
143. Jacobs JP, Cerfolio RJ, Sade RM. The Ethics of Transparency: Publication of Cardiothoracic Surgical Outcomes in the Lay Press. Accepted for publication Friday December 12, 2008. The Annals of Thoracic Surgery. 2009;87(3):679–86. PMID: 19231369, March 2009.
144. Welke KF, O'Brien SM, Peterson ED, Ungerleider RM, Jacobs ML, Jacobs JP. The complex relationship between pediatric cardiac surgical case volumes and mortality rates in a national clinical database. J Thorac Cardiovasc Surg. 2009;137(5):1133–40. Epub 2009 Mar 17, PMID: 19379979.
145. Dokholyan RS, Muhlbaier LH, Falletta J, Jacobs JP, Shahian D, Haan CK, Peterson ED. Regulatory and Ethical Considerations for Linking Clinical and Administrative Databases. The American Heart Journal, accepted for publication Tuesday, April 14, 2009. Am Heart J. 2009;157(6):971–82. PMID: 19464406, June 2009.
146. Hammill BG, Hernandez AF, Peterson ED, Fonarow GC, Schulman KA, Curtis LH. Linking inpatient clinical registry data to Medicare claims data using indirect identifiers. The American Heart Journal, accepted for publication Tuesday, April 14, 2009. Am Heart J. 2009;157(6):995–1000, June 2009.
147. DeCampli WM, Burke RP. Interinstitutional comparison of risk-adjusted mortality and length of stay in congenital heart surgery. Ann Thorac Surg. 2009;88(1):151–6.
148. Jacobs JP, Quintessenza JA, Burke RP, Bleiweis MS, Byrne BJ, Ceithaml EL, DeCampli WM, Giroud JM, Perryman RA, Rosenkranz ER, Wolff G, Posner V, Steverson S, Blanchard WB, Schiebler GL. Regional congenital cardiac surgery of outcomes in Florida using The Society of Thoracic Surgeons Congenital Heart Surgery Database. Cardiol Young. 2009;19:360–9. doi:10.1017/S1047951109990151. First published online 6 July 2009.
149. Jacobs JP, Jacobs ML, Lacour-Gayet FG, Jenkins KJ, Gauvreau K, Bacha E, Maruszewski B, Clarke DR, Tchervenkov CI, Gaynor JW, Spray TL, Stellin G, O'Brien SM, Elliott MJ, Mavroudis C. Stratification of complexity improves the utility and accuracy of outcomes analysis in a Multi-Institutional Congenital Heart Surgery Database: application of the Risk Adjustment in Congenital Heart Surgery (RACHS-1) and Aristotle systems in the Society of Thoracic Surgeons (STS) Congenital Heart Surgery Database. Pediatr Cardiol. 2009. doi:10.1007/s00246-009-9496-0.
150. O'Brien SM, Clarke DR, Jacobs JP, Jacobs ML, Lacour-Gayet FG, Pizarro C, Welke KF, Maruszewski B, Tobota Z, Miller WJ, Hamilton L, Peterson ED, Mavroudis C, Edwards FH. An empirically based tool for analyzing mortality associated with congenital heart surgery. J Thorac Cardiovasc Surg. 2009;138(5):1139–53. PMID: 19837218.
151. Barker GM, O'Brien SM, Welke KF, Jacobs ML, Jacobs JP, Benjamin Jr DK, Peterson ED, Jaggers J, Li JS. Major infection after pediatric cardiac surgery: a risk estimation model. Ann Thorac Surg. 2010;89(3):843–50. PMID: 20172141 [PubMed – indexed for MEDLINE].
152. Jacobs JP, Maruszewski B, Kurosawa H, Jacobs ML, Mavroudis C, Lacour-Gayet FG, Tchervenkov CI, Walters 3rd H, Stellin G, Ebels T, Tsang VT, Elliott MJ, Murakami A, Sano S, Mayer Jr JE, Edwards FH, Quintessenza JA. Congenital heart surgery databases around the world: do we need a global database? Semin Thorac Cardiovasc Surg Pediatr Card Surg Annu. 2010;13(1):3–19. PMID: 20307856.
153. Jacobs JP, Jacobs ML, Mavroudis C, Maruszewski B, Lacour-Gayet FG, Tchervenkov CI. A correction to an analysis from the EACTS and STS congenital heart surgery databases. Ann Thorac Surg. 2010;89(4):1339.
154. Burstein DS, Rossi AF, Jacobs JP, Checchia PA, Wernovsky G, Li JS, Pasquali SK. Variation in models of care delivery for children undergoing congenital heart surgery in the United States. World J Pediatr Congenit Heart Surg. 2010;1:8–14.
155. Shann KG, Giacomuzzi CR, Jacobs JP, Myers GJ, Paugh TA, Mellas N, Puis L, Ojito JW, Gomez D, Olshove V, Fitzgerald DC, Itoh H, Brabant C, Thuys CA, Charette K, Calaritis C, Parpard M, Chancy T, Baker RA, Pourmoghadam KK, Likosky DS. Rationale and use of perfusion variables in the 2010 update of the Society of Thoracic Surgeons

Congenital Heart Surgery Database. World J Pediatr Congenit Heart Surg. 2010;1:34–43.
156. Jacobs JP, Jacobs ML, Mavroudis C, Chai PJ, Tchervenkov CI, Lacour-Gayet FG, Walters III H, Quintessenza JA. Atrioventricular septal defects: lessons learned about patterns of practice and outcomes from the congenital heart surgery database of the Society of Thoracic Surgeons. World J Pediatr Congenit Heart Surg. 2010;1:68–77.
157. Johnson JN, Jaggers J, Li S, O'Brien SM, Li JS, Jacobs JP, Jacobs ML, Welke KF, Peterson ED, Pasquali SK. Center variation and outcomes associated with delayed sternal closure after stage 1 palliation for hypoplastic left heart syndrome. J Thorac Cardiovasc Surg. 2010;139(5):1205–10. Epub 2010 Feb 18.
158. Patel A, Hickey E, Mavroudis C, Jacobs JP, Jacobs ML, Backer CL, Gevitz M, Mavroudis CD. Impact of noncardiac congenital and genetic abnormalities on outcomes in hypoplastic left heart syndrome. Ann Thorac Surg. 2010;89(6):1805–13; discussion 1813–4. PMID: 20494032.
159. Fudge Jr JC, Li S, Jaggers J, O'Brien SM, Peterson ED, Jacobs JP, Welke KF, Jacobs ML, Li JS, Pasquali SK. Congenital heart surgery outcomes in down syndrome: analysis of a national clinical database. Pediatrics. 2010;126(2):315–22. Epub 2010 Jul 12.
160. Al Habib HF, Jacobs JP, Mavroudis C, Tchervenkov CI, O'Brien SM, Mohammadi S, Jacobs ML. Contemporary patterns of management of tetralogy of Fallot: data from the Society of Thoracic Surgeons Database. Ann Thorac Surg. 2010;90(3):813–9; discussion 819–20.
161. Jonas RA, Jacobs JP, Jacobs ML, Mavroudis C. Letter to the editor: reporting of mortality associated with pediatric and congenital cardiac surgery. J Thorac Cardiovasc Surg. 2010;140(3):726; author reply 726–7. PMID: 20723742.
162. Giroud JM, Jacobs JP, Spicer D, Backer C, Martin GR, Franklin RCG, Béland MJ, Krogmann ON, Aiello VD, Elliott MJ, Colan SD, Everett AD, Gaynor JW, Kurosawa H, Maruszewski B, Giovanni S, Tchervenkov CI, Walters III HL, Weinberg P, Anderson RH. Report from The International Society for Nomenclature of Paediatric and Congenital Heart Disease: creation of a visual encyclopedia illustrating the terms and definitions of the International Pediatric and Congenital Cardiac Code. World J Pediatr Congenit Heart Surg. 2010;1:300–13.
163. Shahian DM, Edwards F, Grover FL, Jacobs JP, Wright CD, Prager RL, Rich JB, Mack MJ, Mathisen DJ. The Society of Thoracic Surgeons National Adult Cardiac Database: a continuing commitment to excellence. J Thorac Cardiovasc Surg. 2010;140(5):955–9. PMID: 20951246.
164. Pasquali SK, Jacobs JP, Shook GJ, O'Brien SM, Hall M, Jacobs ML, Welke KF, Gaynor JW, Peterson ED, Shah SS, Li JS. Linking clinical registry data with administrative data using indirect identifiers: implementation and validation in the congenital heart surgery population. Am Heart J. 2010;160:1099–104.
165. Jacobs JP, Jacobs ML, Mavroudis C, Chai PJ, Tchervenkov CI, Lacour-Gayet FG, Walters III H, Quintessenza JA. Transposition of the great arteries: lessons learned about patterns of practice and outcomes from the congenital heart surgery database of the Society of Thoracic Surgeons. World J Pediatr Congenit Heart Surg. 2011;2(1):19–31. doi:10.1177/2150135110381392.
166. Jacobs JP, Pasquali SK, Morales DLS, Jacobs ML, Mavroudis C, Chai PJ, Tchervenkov CI, Lacour-Gayet FG, Walters III H, Quintessenza JA. Heterotaxy: lessons learned about patterns of practice and outcomes from the congenital heart surgery database of the Society of Thoracic Surgeons. World J Pediatr Congenit Heart Surg (WJPCHS). 2011;2(2):278–86. doi:10.1177/2150135110397670.
167. Wallace MC, Jaggers J, Li JS, Jacobs ML, Jacobs JP, Benjamin DK, O'Brien SM, Peterson ED, Smith PB, Pasquali SK. Center variation in patient age and weight at Fontan operation and impact on postoperative outcomes. Ann Thorac Surg. 2011;91(5):1445–52. PMID: 21524453.
168. Bergersen L, Everett AD, Giroud JM, Martin GR, Franklin RC, Béland MJ, Krogmann ON, Aiello VD, Colan SD, Elliott MJ, Gaynor JW, Kurosawa H, Maruszewski B, Stellin G, Tchervenkov CI, Walters HL, Weinberg P, Jacobs JP. Report from The International Society for Nomenclature of Paediatric and Congenital Heart Disease: cardiovascular catheterisation for congenital and paediatric cardiac disease (Part 1 – Procedural nomenclature). Cardiol Young. 2011;21(3):252–9. PMID:21310103.
169. Bergersen L, Giroud JM, Jacobs JP, Franklin RC, Béland MJ, Krogmann ON, Aiello VD, Colan SD, Elliott MJ, Gaynor JW, Kurosawa H, Maruszewski B, Stellin G, Tchervenkov CI, Walters HL, Weinberg P, Everett AD. Report from The International Society for Nomenclature of Paediatric and Congenital Heart Disease: cardiovascular catheterisation for congenital and paediatric cardiac disease (part 2 –nomenclature of complications associated with interventional cardiology). Cardiol Young. 2011;21(3):260–5. PMID: 21310094.
170. Burstein DS, Jacobs JP, Li JS, Sheng S, O'Brien SM, Rossi AF, Checchia PA, Wernovsky G, Welke KF, Peterson ED, Jacobs ML, Pasquali SK. Care models and associated outcomes in congenital heart surgery. Pediatrics. 2011;127(6):e1482–9. Epub 2011 May 16. PMID: 21576309.
171. Weintraub WS, Karlsberg RP, Tcheng JE, Boris JR, Buxton AE, Dove JT, Fonarow GC, Goldberg LR, Heidenreich P, Hendel RC, Jacobs AK, Lewis W, Mirro MJ, Shahian DM. ACCF/AHA 2011 Key data elements and definitions of a base cardiovascular vocabulary for electronic health records: a report of the American College of Cardiology Foundation/American Heart Association Task Force on Clinical Data Standards. Writing Committee Members:

Weintraub WS, Chair; Karlsberg RP, Vice-Chair; Tcheng JE, Boris JR, Buxton AE, Dove JT, Fonarow GC, Goldberg LR, Heidenreich P, Hendel RC, Jacobs AK, Lewis W, Mirro MJ, Shahian DM. ACCF/AHA Task Force on Clinical Data Standards: Hendel RC, Chair; Bozkurt B, Fonarow GC, Jacobs JP, Peterson PN, Roger VL, Smith EE, Tcheng JE, Wang T, Weintraub WS. Circulation. 2011. [Epub ahead of print]. PMID: 21646493. Circulation July 5, 2011.

172. Weintraub WS, Karlsberg RP, Tcheng JE, Boris JR, Buxton AE, Dove JT, Fonarow GC, Goldberg LR, Heidenreich P, Hendel RC, Jacobs AK, Lewis W, Mirro MJ, Shahian DM. ACCF/AHA 2011 Key data elements and definitions of a base cardiovascular vocabulary for electronic health records: a report of the American College of Cardiology Foundation/American Heart Association Task Force on Clinical Data Standards. Writing Committee Members: Weintraub WS, Chair; Karlsberg RP, Vice-Chair; Tcheng JE, Vice-Chair; Boris JR, Buxton AE, Dove JT, Fonarow GC, Goldberg LR, Heidenreich P, Hendel RC, Jacobs AK, Lewis W, Mirro MJ, Shahian DM. ACCF/AHA Task Force on Clinical Data Standards: Hendel RC, Chair; Bozkurt B, Fonarow GC, Jacobs JP, Peterson PN, Roger VL, Smith EE, Tcheng JE, Wang T, Weintraub WS. J Am Coll Cardiol. Published online 6 June 2011. doi:10.1016/j.jacc.2011.05.001. J Am Coll Cardiol. 2011;58(2). © 2011 by the American College of Cardiology Foundation and the American Heart Association, Inc. ISSN 0735-1097. Published by Elsevier Inc.
173. Jacobs JP, Edwards FH, Shahian DM, Prager RL, Wright CD, Puskas JD, Morales DL, Gammie JS, Sanchez JA, Haan CK, Badhwar V, George KM, O'Brien SM, Dokholyan RS, Sheng S, Peterson ED, Shewan CM, Feehan KM, Han JM, Jacobs ML, Williams WG, Mayer Jr JE, Chitwood Jr WR, Murray GF, Grover FL. Successful linking of the Society of Thoracic Surgeons Database to Social Security data to examine survival after cardiac operations. Ann Thorac Surg. 2011;92(1):32–9. PMID: 21718828.
174. Jacobs ML, Daniel M, Mavroudis C, Morales DLS, Jacobs JP, Fraser CD, Turek JW, Mayer JE, Tchervenkov C, Conte JV. Report of the 2010 Society of Thoracic Surgeons Congenital Heart Surgery Practice and Manpower survey. Ann Thorac Surg. 2011;92:762–9.
175. Petrucci O, O'Brien SM, Jacobs ML, Jacobs JP, Manning PB, Eghtesady P. Risk factors for mortality and morbidity after the neonatal Blalock-Taussig shunt procedure. Ann Thorac Surg. 2011;92(2):642–51; discussion 651–2. Epub 2011 May 8. PMID: 21550583.
176. Shahian DM, Edwards FH, Jacobs JP, Prager RL, Normand SL, Shewan CM, O'Brien SM, Peterson ED, Grover FL. Public reporting of cardiac surgery performance: part 1-history, rationale, consequences. Ann Thorac Surg. 2011;92(3 Suppl):S2–11. PMID: 21867789.
177. Shahian DM, Edwards FH, Jacobs JP, Prager RL, Normand SL, Shewan CM, O'Brien SM, Peterson ED, Grover FL. Public reporting of cardiac surgery performance: part 2-implementation. Ann Thorac Surg. 2011;92(3 Suppl):S12–23. PMID: 21867788.
178. Jacobs JP, Pasquali SK, Gaynor JW. Invited commentary: the assessment of outcomes and the improvement of quality of the treatment of patients with congenital and pediatric cardiac disease. World J Pediatr Congenit Heart Surg (WJPCHS). 2011;2(4):597–602. doi:10.1177/2150135111418258.
179. Jacobs JP, Pasquali SK, Jeffries H, Barnett-Jones S, Cooper DC, Vincent R. Outcomes analysis and quality improvement for the treatment of patients with pediatric and congenital cardiac disease. World J Pediatr Congenit Heart Surg (WJPCHS). 2011;2(4):620–33. doi:10.1177/2150135111406293.
180. Hornik CP, He X, Jacobs JP, Li JS, Jaquiss RD, Jacobs ML, O'Brien SM, Peterson ED, Pasquali SK. Complications after the Norwood operation: an analysis of The Society of Thoracic Surgeons Congenital Heart Surgery Database. Ann Thorac Surg. 2011;92(5):1734–40. Epub 2011 Sep 19. PMID: 2193702.
181. Mascio CE, Pasquali SK, Jacobs JP, Jacobs ML, Austin 3rd EH. Outcomes in adult congenital heart surgery: analysis of the Society of Thoracic Surgeons Database. J Thorac Cardiovasc Surg. 2011;142(5):1090–7. Epub 2011 Sep 10. PMID: 21911232.
182. Jacobs JP, O'Brien SM, Pasquali SK, Jacobs ML, Lacour-Gayet FG, Tchervenkov CI, Austin 3rd EH, Pizarro C, Pourmoghadam KK, Scholl FG, Welke KF, Mavroudis C. Richard E. Clark paper: Variation in outcomes for benchmark operations: An analysis of the Society of Thoracic Surgeons Congenital Heart Surgery Database. Ann Thorac Surg. 2011;92(6):2184–92. PMID: 22115229.
183. Barach PR, Jacobs JP, Laussen PC, Lipshultz SE. Outcomes analysis, quality improvement, and patient safety for pediatric and congenital cardiac care: theory, implementation, and applications. Progress Pediatr Cardiol. 2011;32:65–7. In: Lipshultz SE, Barach P, Jacobs JP, Laussen P, editors. Progress in pediatric cardiology: the future of pediatric and congenital cardiac care special issue 1, vol. 32, issue 2, p. 65–153.
184. Jacobs JP, Morales DLS. Strategies for longitudinal follow-up of patients with pediatric and congenital cardiac disease. Progress Pediatr Cardiol. 2011;32:97–102. doi:10.1016/j.ppedcard.2011.10.007. In: Lipshultz SE, Barach P, Jacobs JP, Laussen P, editors. Progress in pediatric cardiology: the future of pediatric and congenital cardiac care special issue 1, vol. 32, issue 2, p. 65–153.
185. Winlaw DS, Large MM, Jacobs JP, Barach PR. Leadership, surgeon well-being and non-technical competencies of pediatric cardiac surgery. Progress Pediatr Cardiol. 2011;32:129–33. doi:10.1016/j.ppedcard.2011.10.011. In: Lipshultz

SE, Barach P, Jacobs JP, Laussen P, editors. Progress in pediatric cardiology: the future of pediatric and congenital cardiac care special issue 1, vol. 32, issue 2, p. 65–153.
186. Jacobs JP, Jacobs ML, Austin EH, Mavroudis M, Pasquali SK, Lacour–Gayet FG, Tchervenkov CI, Walters III HW, Bacha EA, del Nido PJ, Fraser CD, Gaynor JW, Hirsch JC, Morales DLS, Pourmoghadam KK, Tweddell JT, Prager RL, Mayer JE. Quality measures for congenital and pediatric cardiac surgery. World J Pediatr Congenit Heart Surg. 2012;3(1):32–47. doi:10.1177/2150135111426732.
187. Russell HM, Pasquali SK, Jacobs JP, Jacobs ML, O'Brien SM, Mavroudis C, Backer CL. Outcomes of repair of common arterial trunk with truncal valve surgery: a review of The Society of Thoracic Surgeons Congenital Heart Surgery Database. Ann Thorac Surg. 2012;93(1):164–9. Epub 2011 Nov 16. PMID:22088417.
188. Barach PR, Jacobs JP, Laussen PC, Lipshultz SE. Outcomes analysis, quality improvement, and patient safety for pediatric and congenital cardiac care: theory, implementation, and applications part 2. Progress Pediatr Cardiol. 2012;33(1):1–3. doi:10.1016/S1058-9813(12)00003-3. In: Lipshultz SE, Barach P, Jacobs JP, Laussen P, editors. Progress in pediatric cardiology: the future of pediatric and congenital cardiac care special part 2. 2012;33(1):1–101.
189. Pasquali SK, Li JS, Jacobs ML, Shah SS, Jacobs JP. Opportunities and challenges in linking information across databases in pediatric cardiovascular medicine. Progress Pediatr Cardiol. 2012;33(1):21–4. doi:10.1016/j.ppedcard.2011.12.004. In: Lipshultz SE, Barach P, Jacobs JP, Laussen P, editors. Progress in pediatric cardiology: the future of pediatric and congenital cardiac care special part 2. 2012;33(1):1–101.
190. Gaies MG, Jeffries HE, Jacobs JP, Laussen PC. Measuring quality and outcomes in pediatric cardiac critical care. Progress Pediatr Cardiol. 2012;33(1):33–6. doi:10.1016/j.ppedcard.2011.12.006. In: Lipshultz SE, Barach P, Jacobs JP, Laussen P, editors. Progress in pediatric cardiology: the future of pediatric and congenital cardiac care special part 2. 2012;33(1):1–101.
191. Giroud JM, Jacobs JP, Fricker FJ, Spicer D, Backer C, Franklin RCG, Béland MJ, Krogmann ON, Aiello VD, Colan SD, Everett AD, Gaynor JW, Kurosawa H, Maruszewski B, Stellin G, Tchervenkov CI, Walters III HL, Weinberg P, Fogel MA, Jacobs ML, Elliott MJ, Anderson RH. Proposal for a web based "global virtual museum of congenital cardiac pathology". Progress Pediatr Cardiol. 2012;33(1):91–7. doi:10.1016/j.ppedcard.2011.12.015. In: Lipshultz SE, Barach P, Jacobs JP, Laussen P, editors. Progress in pediatric cardiology: the future of pediatric and congenital cardiac care special part 2. 2012;33(1):1–101.
192. Pasquali SK, Li JS, Burstein DS, Sheng S, O'Brien SM, Jacobs ML, Jaquiss RD, Peterson ED, Gaynor JW, Jacobs JP. Association of center volume with mortality and complications in pediatric heart surgery. Pediatrics. 2012;129(2):e370–6. Epub 2012 Jan 9. PMID: 22232310.
193. Pasquali SK, Li JS, He X, Jacobs ML, O'Brien SM, Hall M, Jaquiss RDB, Welke KF, Peterson ED, Shah SS, Gaynor JW, Jacobs JP. Perioperative methylprednisolone and outcome in neonates undergoing heart surgery. Pediatrics. 2012;129(2):e385–91. Epub 2012 Jan 23. PMID: 22271697.
194. Pasquali SK, Li JS, He X, Jacobs ML, O'Brien SM, Hall M, Jaquiss RDB, Welke KF, Peterson ED, Shah SS, Jacobs JP. Comparative analysis of antifibrinolytic medications in pediatric heart surgery. J Thorac Cardiovasc Surg. 2012;143(3):550–7. Epub 2012 Jan 20. PMID: 22264414.
195. Stewart RD, Pasquali SK, Jacobs JP, Benjamin DK, Jaggers J, Cheng J, Mavroudis C, Jacobs ML. Contemporary Fontan operation: association between early outcome and type of cavopulmonary connection. Winner of the 2011 Clifford Van Meter President's Award for the best scientific paper presented at the 2011 STSA 58th annual meeting in San Antonio. Ann Thorac Surg. 2012;93(4):1254–61. PMID: 22450074.
196. Pasquali SK, Jacobs JP, He X, Hornik CP, Jaquiss RDB, Jacobs ML, O'Brien SM, Peterson ED, Li JS. The complex relationship between center volume and outcome in patients undergoing the Norwood operation. Ann Thorac Surg. 2012;93(5):1556–62. Epub 2011 Oct 19. PMID: 22014746.
197. Hornik CP, He X, Jacobs JP, Li JS, Jaquiss RD, Jacobs ML, O'Brien SM, Welke K, Peterson ED, Pasquali SK. Relative impact of surgeon and center volume on early mortality after the Norwood operation. Ann Thorac Surg. 2012;93(6):1992–7. Epub 2012 Apr 18. PMID: 22516833.
198. Jacobs JP, O'Brien SM, Pasquali SK, Jacobs ML, Lacour-Gayet FG, Tchervenkov CI, Austin 3rd EH, Pizarro C, Pourmoghadam KK, Scholl FG, Welke KF, Gaynor JW, Clarke DR, Mayer Jr JE, Mavroudis C. Variation in outcomes for risk-stratified pediatric cardiac surgical operations: An analysis of the STS congenital heart surgery database. Ann Thorac Surg. 2012;94(2):564–72. Epub 2012 Jun 15. PMID: 22704799.
199. Pasquali SK, He X, Jacobs JP, Jacobs ML, O'Brien SM, Gaynor JW. Evaluation of failure to rescue as a quality metric in pediatric heart surgery: an analysis of the STS congenital heart surgery database. Ann Thorac Surg. 2012;94(2):573–80. Epub 2012 May 24. PMID: 22633496.
200. Giroud JM, Aiello VD, Spicer DE, Anderson RH. The archiving working group of the international society for nomenclature of paediatric and congenital heart disease: a visual encyclopedia illustrating the terms and definitions of the international paediatric and congenital cardiac code. Congenit Cardiol Today. 2012;10(8):8–10.
201. Aiello VD, Anderson RH, Giroud JM, Spicer DE. Image of the month (Aortic valve pathology, bicuspid and pulmonary valve pathology, bicuspid) –

August 2012 – presented by The Archiving Working Group. Congenit Cardiol Today. 2012;10(8):14–5.
202. Anderson RH, Aiello VD, Spicer DE, Jacobs JP, Giroud JM. Image of the month #2 (Interrupted aortic arch [IAA], Type B2 [Interruption between the carotid and subclavian arteries with both subclavian arteries arising from the aorta distal to the interruption]) – October 2012 – presented by The Archiving Working Group. Congenit Cardiol Today. 2012;10(10):20–1.
203. Woods RK, Pasquali SK, Jacobs ML, Austin EH, Jacobs JP, Krolikowski M, Mitchell ME, Pizarro C, Tweddell JS. Aortic valve replacement in neonates and infants: an analysis of the Society of Thoracic Surgeons Congenital Heart Surgery Database. J Thorac Cardiovasc Surg. 2012;144(5):1084–90. doi:10.1016/j.jtcvs.2012.07.060. PMID: 22921819. Epub 2012 Aug 24.
204. Jacobs JP, Jacobs ML, Maruszewski B, Lacour-Gayet FG, Tchervenkov CI, Tobota Z, Stellin G, Kurosawa H, Murakami A, Gaynor JW, Pasquali SK, Clarke DR, Austin 3rd EH, Mavroudis C. Initial application in the EACTS and STS Congenital Heart Surgery Databases of an empirically derived methodology of complexity adjustment to evaluate surgical case mix and results. Eur J Cardiothorac Surg. 2012;42(5):775–80. doi:10.1093/ejcts/ezs026. PMID: 22700597. Epub 2012 Jun 14.
205. Dibardino DJ, Pasquali SK, Hirsch JC, Benjamin DK, Kleeman KC, Salazar JD, Jacobs ML, Mayer JE, Jacobs JP. Richard E. Clark paper: effect of sex and race on outcome in patients undergoing congenital heart surgery: an analysis of the Society of Thoracic Surgeons Congenital Heart Surgery Database. Richard Clark Award recipient for best use of the STS Congenital Heart Surgery Database. Ann Thorac Surg. 2012;94(6):2054–60. doi:10.1016/j.athoracsur.2012.05.124. Epub 2012 Aug 9. PMID: 22884593.
206. Kansy A, Jacobs JP, Pastuszko A, Mirkowicz-Małek M, Manowska M, Jezierska E, Maruszewski P, Burczyński P, Maruszewski B. Major infection after pediatric cardiac surgery: external validation of risk estimation model. Ann Thorac Surg. 2012;94(6):2091–5. doi:10.1016/j.athoracsur.2012.07.079. Epub 2012 Oct 4. PMID: 23040826.
207. Vener DF, Guzzetta N, Jacobs JP, Williams GD. Development and implementation of a new data registry in congenital cardiac anesthesia. Ann Thorac Surg. 2012;94(6):2159–65. doi:10.1016/j.athoracsur.2012.06.070. PMID: 23176940.
208. Pasquali SK, Gaies MG, Jacobs JP, William Gaynor J, Jacobs ML. Centre variation in cost and outcomes for congenital heart surgery. Cardiol Young. 2012;22(6):796–9. PMID: 23331604.
209. Aiello VD, Spicer DE, Jacobs JP, Giroud JM, Anderson RH. Image of the month #3 (Total anomalous pulmonary venous connection [TAPVC], Type 4 [mixed]) – December 2012 – presented by The Archiving Working Group. Congenit Cardiol Today. 2012;10(12):8–9.
210. Pasquali SK, Peterson ED, Jacobs JP, He X, Li JS, Jacobs ML, Gaynor JW, Hirsch JC, Shah SS, Mayer JE. Differential case ascertainment in clinical registry versus administrative data and impact on outcomes assessment for pediatric cardiac operations. Ann Thorac Surg. 2013;95(1):197–203. doi:10.1016/j.athoracsur.2012.08.074. Epub 2012 Nov 7. PMID: 23141907.
211. Ungerleider RM, Pasquali SK, Welke KF, Wallace AS, Ootaki Y, Quartermain MD, Williams DA, Jacobs JP. Contemporary patterns of surgery and outcomes for aortic coarctation: an analysis of the Society of Thoracic Surgeons Congenital Heart Surgery Database. J Thorac Cardiovasc Surg. 2013;145(1):150–8. doi:10.1016/j.jtcvs.2012.09.053. PMID: 23098750. Epub 2012 Oct 23.
212. Overman D, Jacobs JP, Prager RL, Wright CD, Clarke DR, Pasquali S, O'Brien SM, Dokholyan RS, Meehan P, McDonald DE, Jacobs ML, Mavroudis C, Shahian DM. Report from The Society of Thoracic Surgeons National Database Work Force: clarifying the definition of operative mortality. World J Pediatr Congenit Heart Surg (WJPCHS). 2013;4(1):10–2. doi:10.1177/2150135112461924.
213. Spicer DE, Jacobs JP, Giroud JM, Anderson RH, Aiello VD. Image of the month #4 (Single ventricle, DILV) – February 2013 – presented by The Archiving Working Group. Congenit Cardiol Today. 2013;11(2):13–4.
214. Jacobs JP, O'Brien SM, Shahian DM, Edwards FH, Badhwar V, Dokholyan RS, Sanchez JA, Morales DL, Prager RL, Wright CD, Puskas JD, Gammie JS, Haan CK, George KM, Sheng S, Peterson ED, Shewan CM, Han JM, Bongiorno PA, Yohe C, Williams WG, Mayer JE, Grover FL. Successful linking of the Society of Thoracic Surgeons Database to Social Security data to examine the accuracy of Society of Thoracic Surgeons mortality data. J Thorac Cardiovasc Surg. 2013;145(4):976–83. doi:10.1016/j.jtcvs.2012.11.094. PMID: 23497944.
215. Jacobs ML, O'Brien SM, Jacobs JP, Mavroudis C, Lacour-Gayet F, Pasquali SK, Welke K, Pizarro C, Tsai F, Clarke DR. An empirically based tool for analyzing morbidity associated with operations for congenital heart disease. J Thorac Cardiovasc Surg. 2013;145(4):1046–1057.e1. doi:10.1016/j.jtcvs.2012.06.029. Epub 2012 Jul 24. PMID: 22835225.
216. Pasquali SK, Jacobs JP. The role of databases in improving the quality of care for congenital heart disease. World J Pediatr Congenit Heart Surg. 2013;4(2):139–41. doi:10.1177/2150135113480221. PMID: 23799726.
217. Kansy A, Maruszewski B, Jacobs JP, Maruszewski P. Application of four complexity stratification tools (Aristotle Basic Score, RACHS-1, STAT Mortality Score, and STAT Mortality Categories) to evaluate early congenital heart surgery outcomes over 16 years at a single institution. Kardiochirurgia i Torakochirurgia Polska. 2013;10(2):115–9. doi:10.5114/kitp.2013.36129.

218. Jacobs JP, Giroud JM, Anderson RH, Aiello VD, Spicer DE. Image of the month #5 (VSD, Type 2 [Perimembranous] [Paramembranous]) – May 2013 – presented by The Archiving Working Group. Congenit Cardiol Today. 2013;11(5):8–9.
219. Pasquali SK, He X, Jacobs ML, Hall M, Gaynor JW, Shah SS, Peterson ED, Hill KD, Li JS, Jacobs JP. Hospital variation in postoperative infection and outcome after congenital heart surgery. Ann Thorac Surg. 2013;96(2):657–63. doi:10.1016/j.athoracsur.2013.04.024. Epub 2013 Jun 28. PMID: 23816416.
220. Shahian DM, He X, Jacobs JP, Rankin JS, Peterson ED, Welke KF, Filardo G, Shewan CM, O'Brien SM. Issues in quality measurement: target population, risk adjustment, and ratings. Ann Thorac Surg. 2013;96(2):718–26. doi:10.1016/j.athoracsur.2013.03.029. Epub 2013 Jun 29. PMID: 23816415.
221. Shahian DM, Jacobs JP, Edwards FH, Brennan JM, Dokholyan RS, Prager RL, Wright CD, Peterson ED, McDonald DE, Grover FL. The Society of Thoracic Surgeons National Database. Heart. 2013;99(20):1494–501. doi:10.1136/heartjnl-2012-303456. PMID: 23335498. Epub 2013 Jan 18.
222. Jacobs JP, Maruszewski M. Functionally univentricular heart and the Fontan operation: lessons learned about patterns of practice and outcomes from the congenital heart surgery databases of the European Association for Cardio-Thoracic Surgery and the Society of Thoracic Surgeons. World J Pediatr Congenit Heart Surg. 2013;4:349–55. doi:10.1177/2150135113494228.
223. Mascio CE, Austin 3rd EH, Jacobs JP, Jacobs ML, Wallace AS, He X, Pasquali SK. Perioperative mechanical circulatory support in children: an analysis of the Society of Thoracic Surgeons Congenital Heart Surgery Database. J Thorac Cardiovasc Surg. 2013. doi:10.1016/j.jtcvs.2013.09.075. PMID: 24246548.
224. Hendel RC, Bozkurt B, Smith EE, Fonarow GC, Tcheng JE, Jacobs JP, Wang TY, Lichtman JH, Weintraub WS, ACC/AHA Task Force on Clinical Data Standards. ACC/AHA 2013 methodology for developing clinical data standards: a report of the American College of Cardiology/American Heart Association Task Force on clinical data standards. J Am Coll Cardiol. 2013. doi:10.1016/j.jacc.2013.11.006. PMID: 24246166.
225. Hendel RC, Bozkurt B, Fonarow GC, Jacobs JP, Lichtman JH, Smith EE, Tcheng JE, Wang TY, Weintraub WS. ACC/AHA 2013 methodology for developing clinical data standards: a report of the American College of Cardiology/American Heart Association Task Force on Clinical Data Standards. Circulation. 2013. [Epub ahead of print]. PMID: 24243855.
226. Jacobs JP, Pasquali SK, Austin E, Gaynor JW, Backer C, Romano JCH, Williams WG, Caldarone C, McCrindle BW, Graham K, Dokholyan RS, Shook G, Poteat J, Baxi M, Karamlou T, Morris JA, Blackstone EH, Mavroudis C, Mayer Jr JE, Jonas RA, Jacobs ML. Linking the congenital heart surgery databases of the Society of Thoracic Surgeons (STS) and the Congenital Heart Surgeons' Society (CHSS): part 1 – rationale and methodology. World J Pediatr Congenit Heart Surg (WJPCHS). 2014;5(2):256–71. doi:10.1177/2150135113519454.
227. Jacobs JP, Pasquali SK, Austin E, Gaynor JW, Backer C, Romano JCH, Williams WG, Caldarone C, McCrindle BW, Graham K, Dokholyan RS, Shook G, Poteat J, Baxi M, Karamlou T, Morris JA, Blackstone EH, Mavroudis C, Mayer Jr JE, Jonas RA, Jacobs ML. Linking the congenital heart surgery databases of the Society of Thoracic Surgeons (STS) and the Congenital Heart Surgeons' Society (CHSS): part 2 – lessons learned and implications. World J Pediatr Congenit Heart Surg (WJPCHS). 2014;5(2):272–82. doi:10.1177/2150135113519455.
228. Hoffman JI, Kaplan S. The incidence of congenital heart disease. J Am Coll Cardiol. 2002;39:1890–900.
229. Kennedy I. Learning from Bristol: the report of the public inquiry into children's heart surgery at the Bristol Royal Infirmary 1984–1995. Department of Health, London: Crown Copyright; July 2001. http://www.bristol-inquiry.org.uk/. Accessed 21 May 2005.
230. Sherry A. Children's hospital cardiology chief told to resign. Denver Post. Article published: March 1, 2001. http://www.denverpost.com/news/news0301b.htm. Accessed 21 Mar 2001.
231. Sherry A. Hospitals shield mortality rates. Denver Post. Article published: March 2, 2001. http://www.denverpost.com/news/news0302d.htm. Accessed 21 Mar 2001.
232. The Denver Post Editorial Board. At the heart of the problem. Denver Post. Article published: March 2, 2001. http://www.denverpost.com/opinion/edits0302c.htm. Accessed 21 Mar 2001.
233. Hernandez J. Other options. Denver Post. Article published: March 3, 2001. http://www.denverpost.com/opinion/lett0311.htm. Accessed 21 Mar 2001.
234. Johnson L. Baby's death at children's turns parents to their faith. Denver Post. Article published: March 3, 2001. http://www.denverpost.com/opinion/lett0311.htm. Accessed 21 Mar 2001.
235. White S. Kids' best interests: Re: "Children's Hospital cardiology chief told to resign," March 1. Denver Post. Article published: March 3, 2001. http://www.denverpost.com/opinion/lett0311.htm. Accessed 21 Mar 2001.
236. Weinberg S. Rare look inside a surgeon's sanctum. Denver Post. Article published: Sunday, April 20, 2003. http://www.denverpost.com/Stories/0%2C1413%2C36,28,1333663%2C00.html. Accessed 22 Oct 2004.
237. The Report of the Manitoba pediatric cardiac surgery inquest: an inquest into twelve deaths at the Winnipeg Health Sciences Centre in 1994. http://www.pediatriccardiacinquest.mb.ca/pdf/pcir_intro.pdf. Accessed 14 June 2014.

238. Francis R, QC. 2013. Report of the Mid Staffordshire NHS Foundation Trust Public Inquiry (Report). House of Commons. ISBN 9780102981476. http://www.midstaffspublicinquiry.com/report]. Accessed 14 June 2014.
239. Hudson W, Cohen E. After CNN investigates babies' deaths, hospital releases mortality data. http://edition.cnn.com/2013/08/12/health/kentucky-children-update/index.html. Accessed 14 June 2014.
240. Cronk CE, Malloy ME, Pelech AN, et al. Completeness of state administrative databases for surveillance of congenital heart disease. Birth Defects Res A Clin Mol Teratol. 2003;67:597–603.
241. Frohnert BK, Lussky RC, Alms MA, Mendelsohn NJ, Symonik DM, Falken MC. Validity of hospital discharge data for identifying infants with cardiac defects. J Perinatol. 2005;25(11):737–42.
242. Donabedian A. Evaluating the quality of medical care. Milbank Mem Fund Q. 1966;44(Suppl):166–206.
243. Shahian DM, O'Brien SM, Sheng S, Grover FL, Mayer JE, Jacobs JP, Weiss JM, Delong ER, Peterson ED, Weintraub WS, Grau-Sepulveda MV, Klein LW, Shaw RE, Garratt K, Moussa I, Shewan CM, Dangas GD, Edwards FH. Predictors of long-term survival following coronary artery bypass grafting surgery: results from The Society of Thoracic Surgeons Adult Cardiac Surgery Database (The ASCERT Study). Circulation. 2012;125(12):1491–500. Epub 2012 Feb 23. PMID: 22361330.
244. Weintraub WS, Grau-Sepulveda MV, Weiss JM, O'Brien SM, Peterson ED, Kolm P, Zhang Z, Klein LW, Shaw RE, McKay C, Ritzenthaler LL, Popma JJ, Messenger JC, Shahian DM, Grover FL, Mayer JE, Shewan CM, Garratt KN, Moussa ID, Dangas GD, Edwards FH. Comparative effectiveness of revascularization strategies. N Engl J Med. 2012;366(16):1467–76. doi:10.1056/NEJMoa1110717. Epub 2012 Mar 27. PMID: 22452338.
245. Fröbert O, Lagerqvist B, Olivecrona GK, Omerovic E, Gudnason T, Maeng M, Aasa M, Angerås O, Calais F, Danielewicz M, Erlinge D, Hellsten L, Jensen U, Johansson AC, Kåregren A, Nilsson J, Robertson L, Sandhall L, Sjögren I, Ostlund O, Harnek J, James SK. Thrombus aspiration during ST-segment elevation myocardial infarction. N Engl J Med. 2013;369(17):1587–97. doi:10.1056/NEJMoa1308789. Epub 2013 Aug 31. PMID: 23991656 [PubMed – indexed for MEDLINE].
246. Lauer MS, D'Agostino Sr RB. The randomized registry trial – the next disruptive technology in clinical research? N Engl J Med. 2013;369(17):1579–81. doi:10.1056/NEJMp1310102. Epub 2013 Aug 31. PMID: 23991657.
247. Shahian DM, O'Brien SM, Filardo G, Society of Thoracic Surgeons Quality Measurement Task Force, et al. The Society of Thoracic Surgeons 2008 cardiac surgery risk models: part 1—coronary artery bypass grafting surgery. Ann Thorac Surg. 2009;88(Suppl):2–22.
248. O'Brien SM, Shahian DM, Filardo G, Society of Thoracic Surgeons Quality Measurement Task Force, et al. The Society of Thoracic Surgeons 2008 cardiac surgery risk models: part 2—isolated valve surgery. Ann Thorac Surg. 2009;88(Suppl):23–42.
249. Shahian DM, O'Brien SM, Filardo G, Society of Thoracic Surgeons Quality Measurement Task Force, et al. The Society of Thoracic Surgeons 2008 cardiac surgery risk models: part 3—valve plus coronary artery bypass grafting surgery. Ann Thorac Surg. 2009;88(Suppl):43–62.
250. The National Quality Forum website: http://www.qualityforum.org/. Accessed 23 Dec 2013

9 Databases for Assessing the Outcomes of the Treatment of Patients with Congenital and Pediatric Cardiac Disease: The Perspective of Cardiology

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Abstract

The development of the conceptual and technological infrastructure of a health care system is primarily driven by a society's obligation to provide for its people. Few people today would deny that organized data collection, processing and presentation is an increasingly key component of good patient care. Given the "wild west" nature of the medical informatics free market today, there is certainly no standardized database structure or architecture. A viable, forward looking database design must be able to interface in a variety of ways, including ways unforeseen at this time.

The fields of congenital cardiology and cardiac surgery are poised to benefit from, as well as promote, new developments in data structuring, storage, retrieval, processing and analysis. Databases are comprised of a variety of platforms, software and architectures depending on the size and purpose of the database. Databases at and above a hospital department level should be organized around purpose, utility, flexibility and growth.

A state-of-the-art database should have the following core architectural components: an On-Line Transactional Processing system, an On-Line Analytical Processing system, and we propose an On-Line Semantics Processing system. Such databases could place a modern congenital cardiac center in an ideal position to take full advantage of the technological advancements of today as well as tomorrow, including artificial intelligence, cloud based systems, and evolving human device-interfaces,

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ultimately providing unmatched facility in data management, which leads to similar advances in patient management.

Keywords

Pediatric • Congenital • Cardiac • Center • Database • Architecture • Ontology

Background, History and Current Status

The need for accurate, timely and complete collection of data has never been as paramount in the medical field as it is today. While it is vitally important that clinicians strive for excellence in the technical aspects of their specialties, clinical excellence alone will add up to little if we cannot assess the outcomes of our practice or fully understand the complexities of the diseases we treat.

Our world is an increasingly technical, complex world and this is nowhere more apparent than in the field of congenital heart disease (CHD). There is great variation in the manifestation of congenital heart defects, as well as significant variation in the treatment of these defects. The process of caring for individuals born with congenital heart disease is an ever evolving, continuous learning process requiring the commitment of lifetimes. Today's pediatric cardiologists (and cardiovascular surgeons) are inundated with data. The great challenge is to make sense of all of this information. Databases are needed to catalog, organize, store, retrieve and analyze this over-whelming amount of raw data.

Two things are needed to provide a high fidelity database. The first component is the *database* itself. The second component is comprised of the *interfaces* into and out of the database. The word "interface" can be defined as a "*surface forming a common boundary between adjacent regions*, *bodies*, *substances*, *or phases* [1]." The key word is "boundary." Like any boundary, or border between two regions or countries, an interface will need to control and define what commerce (in this case information) is allowed to cross, in what manner it crosses and in what direction.

It is difficult to consider databases without discussing the interfaces regulating information flow into and out of those databases. Interfaces may be of many types. Broad categories of interfaces would include human-database, database-database, machine-database, and database-web. In the following discussion databases and interfaces will be discussed concurrently. Because database information is presented via an interface, interfaces define the way we perceive a database. Good interfaces will promote data collection, speed workflow, perform precise translations, provide error checking, and present, organize and sort information.

The technology exists today to create interfaces and databases which can meet every conceivable information need. As Health Care Providers we need to understand the technical aspects of these important tools to be able to use them to best advantage. We need to know how a database is organized to understand what it can and cannot do. A database designed and built for a specific task will not perform well when forced to do something different. Optimal configuration and planning is important when setting up a database and its interfaces.

At first, it might seem like the requirements for a clinical database and a research database are incongruent. Technological improvements have allowed interface developments to expand exponentially, ultimately facilitating human interaction with data. The impact of databases on our practices and lives continues to evolve. Ultimately, transactions through interfaces will come to define both our perception of data and the utility of data. We are in a time of transformation and rapid evolution, with a future that will allow us to evolve what we can do for our patients and practice of medicine.

A well thought out, well designed, and well utilized departmental level cardiac center database (CCDB) will be able to facilitate information exchange between a hospital information system (HIS), cardiovascular modality systems (electrocardiogram [ECG], echocardiography, magnetic resonance imaging [MRI], angiography), an exercise physiology laboratory, as well as physicians, nurses, technicians, and other cardiac division health care providers. The CCDB will promote and enable registry submissions, intra- and extra-mural research, and will greatly facilitate quality assurance/quality improvement (QA/QI) initiatives.

We have come to the point in this day and age when it is feasible and desirable to have as much data as possible reside in a departmental CCDB. While we cannot predict today precisely what the practice of medicine will look like tomorrow, we can build departmental level databases which will support every activity involved in caring for patients with congenital heart disease as well as advancing our knowledge and practice both from within institutional walls as well as outside.

Database History

Databases are already widely used in our field. They are highly scalable, ranging from small information repositories for short-term research and running up in size to the large multi-center complex registry-type databases (examples include European Association for Cardio-Thoracic Surgery [EACTS], Society of Thoracic Surgeons [STS], Congenital Cardiovascular Interventional Study Consortium [CCISC], Improving Pediatric and Adult Congenital Treatment [IMPACT], Central Cardiac Audit Database [CCAD], Pediatric Cardiac Critical Care Consortium [PC4], and various National Database Initiatives). The simplest database is nothing more complex than an electronic spreadsheet, while the most complex CCDB will be comprised of sub-components including complex interfaces, relational databases, semantic layers and data warehouses.

One form of database, important in assessing outcomes, is the medical registry. A medical registry [2] is a collection of secondary data related to patients with a specific disease, diagnosis, condition, treatment or procedure. Compared to an electronic medical record (EMR), a registry only keeps track of a small sub-population of patients with a specific condition. A registry needs to assure that data it receives is valid and has been verified. It is common for registries to regularly audit contributing entities to verify that data submitted to the registry is valid.

The American College of Cardiology Foundations (R) has developed the IMPACT Registry™ (IMproving Pediatric and Adult Congenital Treatment) to capture diagnostic cardiac catheterization and catheterization-based outcomes data [3]. This registry is administered by the National Cardiovascular Data Registry (R) (NCDR) of the American College of Cardiology®, a primary resource for measuring and quantifying outcomes and identifying gaps in the delivery of quality cardiovascular patient care in the United States. The IMPACT Registry™ has grown rapidly since its inauguration in 2009, and receives data from more than 80 institutions encompassing over 12,000 procedures. Goals of the IMPACT Registry™ are to improve the quality of cardiovascular patient care and support research that improves patient care and outcomes.

Similarly, the Congenital Cardiovascular Interventional Study Consortium (CCISC) is an industry sponsored, not-for-profit organization that assists and coordinates the design, conduct and reporting of scientific studies in interventional cardiovascular care for individuals with congenital heart disease [4].

Cardiologists are also interested in other less procedure based outcome measures. While there is a wealth of pharmacological data from large adult studies, no such similar data is available in children. Such studies would require large numbers of patients, larger databases and longer periods of time to aggregate meaningful outcomes. Yet these types of multi-center trials could be greatly facilitated by participating centers sharing pre-certified retrospective data.

As clinicians, we are honestly often more interested in the data that goes into a database than the actual database structure and function. This is analogous to us being more interested in the congenital heart defect we are imaging with echocardiography and angiography rather than the equipment we are using to do the imaging. Never the less, we do spend a fair amount of time making sure that our imaging equipment is technically adequate, and we prefer state-of-the-art equipment as it allows us to gather information more accurately and make more precise diagnoses. This analogy holds true with databases. The interfaces, structure, granularity and function (the technology) behind a database will have an impact on the quality of information stored in that database. This has to be finely balanced with the burden of data entry so that data capture is complete.

We need to understand that the type of database used to gather, organize, store and display our data is every bit as important as the type of catheterization laboratory equipment or echocardiography carts we use. It's not just what data are being stored, but how data are stored, organized, summarized and displayed back to the clinician that can make a world of difference in our clinical practice and the progression of our ability to care for those born with congenital heart disease. Three problems must be overcome in order to transform data into actionable knowledge are (1) duplication of data in separate systems, (2) non-standard and proprietary coding systems, and (3) the patient-centric nature of the electronic medical record.

It is common and usual for various silos of clinical data to be housed within separate functional systems. There are electronic medical records from hospital information systems, echocardiogram imaging and reporting systems, cardiac catheterization hemodynamic, imaging, and reporting systems, ECG monitoring and recording systems, cardiac MRI systems, and electrophysiology laboratory systems. Each of these functional systems will keep some of the same, or similar data housed within separate local repositories. For instance, when considering patient demographics each system keeps some form of patient identifier information. It can be a medical record number, name, address, gender, date of birth, or various combinations of the above. Some data are entered manually, and some are automated. Some are correct, and some are incorrect or incomplete. In some systems, a medical record number may be stored as an integer value, but in others that same designator may be kept as a text field. This non-uniformity across systems creates problems with data validation and standardization.

The second problem has to do with coding systems and identifiers. Any structured reporting system, including a code set, is based on an internally standardized selectable set of items. These could include categorized lists of diagnoses, procedures, and risk factors to name a few. Although most categories contain common meanings, they do not often contain common terms and identifiers. Some systems will utilize broadly recognized coding representations, such as the 10th International Classification of Diseases Revision (ICD-10) or Current Procedural Terminology (CPT), some will use registry-based coding, such as STS or IMPACT, while still others will use their own specific designators recognized by no other systems. To further complicate the situation, in one system an attending physician may be identified as "Smith, John M.D." while in another system that same physician will be identified as employee number "1234567". This lack of standardization creates problems with data translation and consolidation.

The third problem is that the data contained within these disparate functional systems are, by their very nature, extremely patient-centric. While this is the most logical data structure for individual patient care and cardiology department modeling, this is not an efficient organization for reporting, evaluating key performance indicators, analytics, decision support, quality improvement, and research-based endeavors. The patient centric nature of these functional systems creates challenges with data aggregation, manipulation, and representation.

Database Architecture

There are three major required sub-systems within a complete Cardiac Center Database solution. These consist of (1) an on-line transaction processing (OLTP) system with an application (user interface) front end, (2) an on-line analytical processing (OLAP) or data warehouse system with an analytics and data visualization layer, and (3) an interface to national registries and external databases. Other components, including an on-line semantics processing system, artificial intelligence, and a cloud component are optional, but add significant value and utility to the CCDB. Each one of these systems will be discussed in detail.

On-Line Transactional Processing System

OLTP systems are typically relational databases. Relational Databases are databases comprised of a collection of formally organized or related tables. Each table entry is defined by a 'key' which is the first value of an entry in that table. An entry in a second table will have another 'key,' and can be related to an entry in the first table by including the 'foreign key' from the first table. This can lead to a complex series of links between data tables, particularly when a Relational Database can contain hundreds of different tables.

An online transaction processing system has three architectural requirements. First, it must be designed around the data flow and dependency of a business or, in this case, a Cardiac Center. As an example, the most important single unit within a Cardiac Center is the patient. This then becomes the highest level in the hierarchical database schema. Each patient can have multiple visits, or encounters, with the department, creating a one-to-many relationship between the patient and each visit. A particular visit may be an observation visit in which one cardiac catheterization is performed, or an inpatient visit (admission) in which several cardiac characterizations and surgeries are performed. It follows that there is at the potential of a one-to-many relationship between each visit and multiple procedures or tests. This particular data dependency-based model is shown in Fig. 9.1.

The strength of relational databases is that they are well suited towards organizing workflow and business processes. Data obtained at a single encounter or session is easily sorted to go into organized tables. It immediately comes to mind, however, that extracting information out of a relational database is not necessarily straightforward. Because related information can be placed in multiple tables, extracting data from relational databases using a data query often requires specialized skill and technical knowledge beyond the interest- if not beyond the ability- of most clinical practitioners, even if one could get past an institution's Database Systems Administrator to directly access a database [5]!

The second requirement is that the OLTP system provides transactional, or real-time, data to the system. The OLTP system needs to pick up data as soon as data are available. Many interfaces, including interfaces to the echocardiography reporting system, the hospital hematology and chemistry laboratories, and radiology picture archiving and communications systems (PACS) and others previously mentioned are required. Additionally, the right kind of interface can display a very complex set of numbers much more easily that simply listing tables. Figure 9.2 shows a well designed CCDB display interface with a graphical display of "ins and outs" for a given patient [6].

Graphical patient information can also be depicted in other unique and intuitive ways. For instance, diagnosis or findings codes, if granular enough, might allow for the generation of Mullins-types diagrams using the CCDB internal code set [7]. A prototype system was been implemented using Protégé 1 and the Java Advanced Imaging package in 2005 at Children's Mercy Hospital. This type of system would allow creation of Mullins type diagrams to display serial chronologic changes in anatomy and physiology independent of imaging modality, since the diagrams would be generated by database codes.

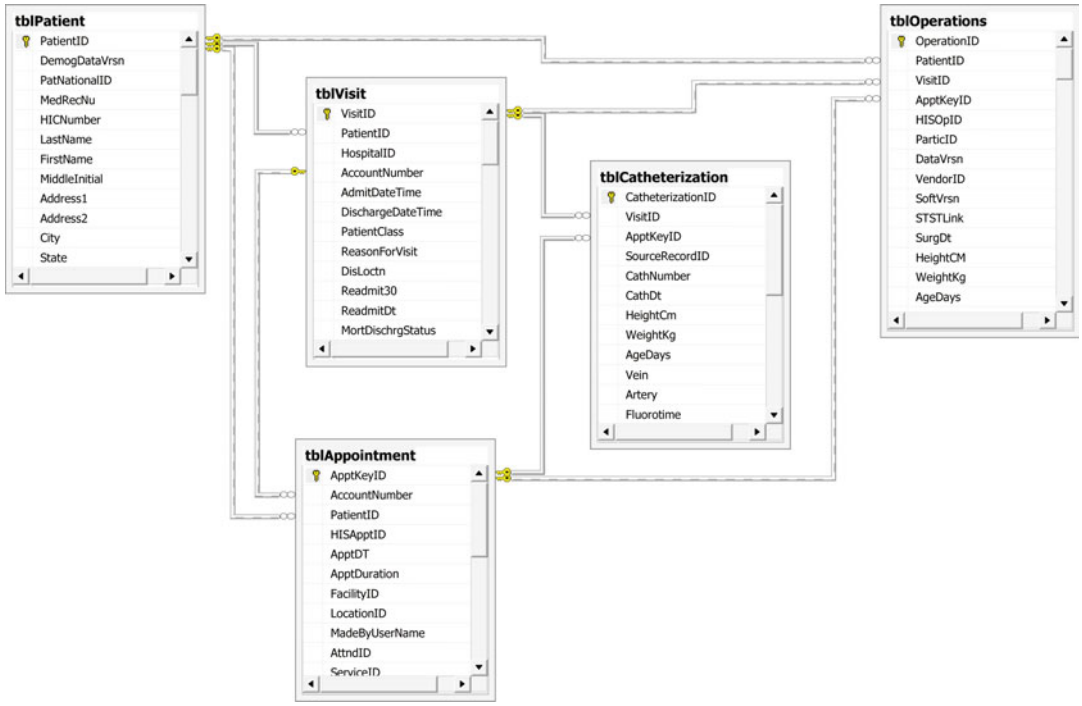

Fig. 9.1 A relational data model containing demographic data (tblPatient), visit data (tblVisit), appointment data (tblAppointment, and procedure data (tblCatheterization, tblOperations). The tblPatient is the highest-level table. It contains a 'key' called PatientID which is a 'foreign key' for the tblVisit, tblOperations and tblAppointment tables. Since one patient can have potentially many admissions, there may be multiple entries in tblVisit for the patient with the same PatientID. Likewise, it is possible for several entries in tblCatherization to have the same VisitID if a patient has multiple catheterization procedures during the same admission

Figure 9.3 shows the prototype Ontodiagram query interface which generated a diagram of a heart with dextro-transposition of the great arteries (D-TGA) and ventricular septal defect (VSD). This type of diagram generation would optimally allow a cardiologist or surgeon to overlay cath, echo and surgical data on the diagram framework. The diagram would automatically "change" as codes and findings in the CCDB update, providing an evolving diagrammatic modality to assist in management, reflecting the latest knowledge available in the CCDB.

Lastly, and perhaps most importantly for outcomes and QA/QI, the OLTP system becomes the vehicle for receiving, gathering and extracting data from all of the systems (human, machine, and database) through appropriate interfaces to enable normalization and storage within one local data store, the On-Line Analytical Processing (OLAP) System.

On-Line Analytical Processing System (Data Warehouse)

The on-line analytical processing system, commonly called a Data Warehouse, is profoundly different than the on-line transaction processing system described above. OLTP (transactional) systems are designed mainly for large numbers of transactions in real time. The high performance and constant load requirements are the driving force behind the entity relationship data model, where discrete tables can be updated on an immediate and independent basis. As noted above, the large number of tables and complex relationships within an entity relationship data model are nearly impossible to comprehend without extensive training, rendering routine data query, data extraction, and decision making processes virtually impossible. A data warehouse, however, is a "subject-oriented, integrated, time-varying,

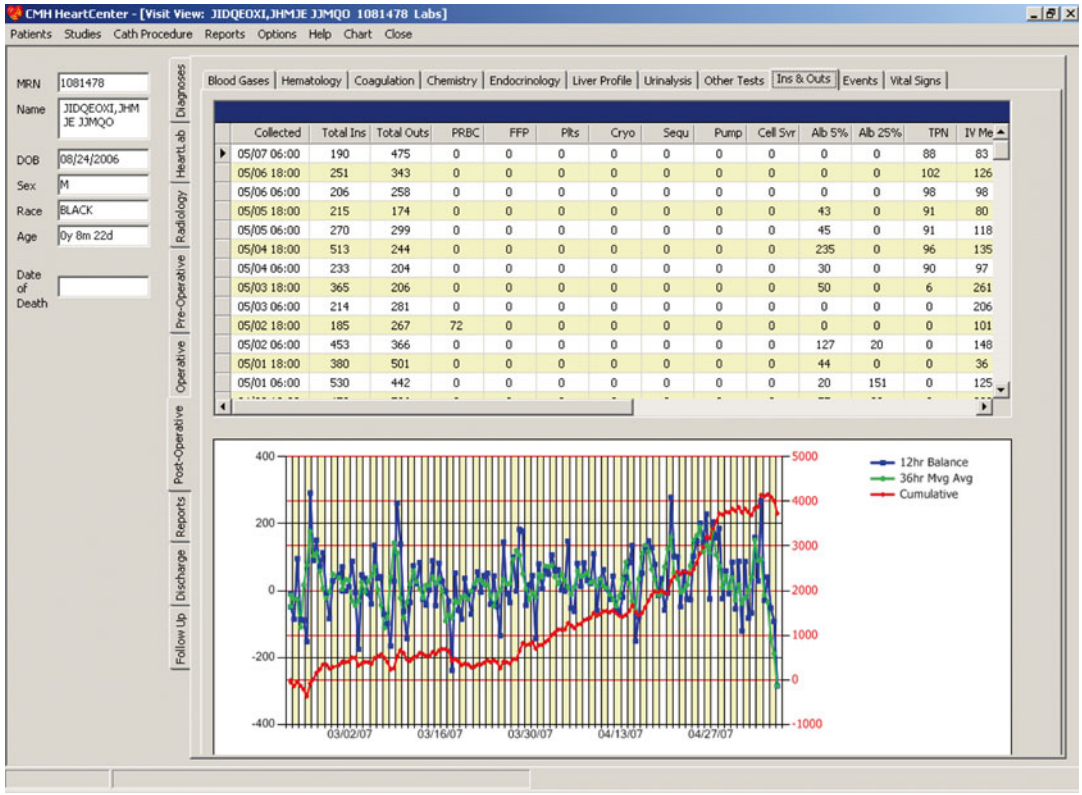

Fig. 9.2 Daily "ins and outs" for a patient over a 2 month period graphically displayed with 12 h balance, 36 h moving average, and cumulative plots over time. The advantage of being able to discern daily variance and longer term cumulative changes is easily appreciated

non-volatile collection of data that is used primarily in organizational decision making" [5]. As such, an OLAP (data warehouse) system is designed to assess outcomes, measure quality, and provide critical input for strategic decision making.

At the heart of a data warehouse is a common organization known as a multidimensional data model. As shown in Fig. 9.4, the central table within a multidimensional data model is the fact table (fct_Patients). This database organization is sometimes referred to as a "star schema" due to its symmetrical look and well defined hierarchical structure [8].

The purpose of the fact table is to contain all of the measurements of interest when analyzing a particular process or unit type. A cardiac center data warehouse would very likely have separate fact tables for cardiac catheterizations and echocardiograms. Examples of measures that would be stored in a cardiac catheterization fact table would be fluoroscopy time, contrast, age, and weight, amongst many others. For an interventional catheterization procedure, measures would be saved on a pre-interventional and post-interventional basis. The degree of granularity in the fact table for interventional catheterization cases should be defined by the set-based intra-procedural measurements. Other fact tables require temporal analysis, in which case subsequent stored measures would depend upon the minimum time interval requirements for the analysis (for instance measures being stored at ten minute intervals during a cardiac catheterization).

Aggregate measures, including sub-totaling, multiplying, averaging, and others also are best placed in the data warehouse. Aggregations can occur on an as-needed basis when measures are being displayed, during the analysis cube calculation process, or when data is imported from the

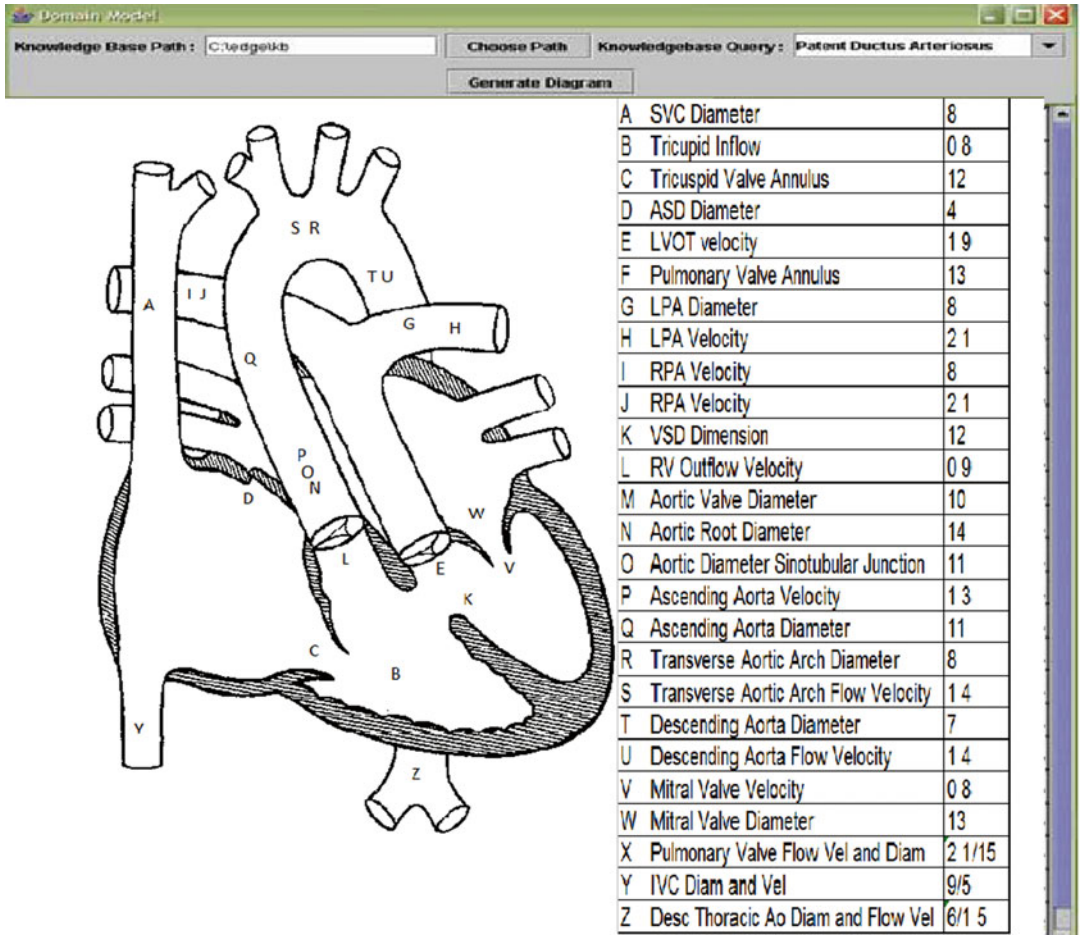

Fig. 9.3 The Ontodiagram query interface. Measurement mappings are shown. Diagrams are generated by breaking each diagram into very granular, atomic components. For instance, the concept "normal pulmonary veins" is a parent concept that actually consists of a number of child concepts, including "normal right upper pulmonary vein," "normal left upper pulmonary vein," "normal right lower pulmonary vein," "normal left lower pulmonary vein," and "all pulmonary veins go to leftward atrium." These concepts can be mapped to a CCDB internal code set, which optimally should be even more granular than the concepts described here

OLTP system into the data warehouse. The most important factor in determining where to process aggregations is the impact on overall database efficiency and performance.

Temporal aggregate measures often require special attention, with differentiation between a set date and time (start time of a cardiac catheterization) compared to a time interval (minutes a series of arterial blood gases [ABGs] are obtained post initiation of nitrous oxide [NO]).

The dimension table is the final important concept in the cardiac center data warehouse. Each unique row of measures in the fact table depends on a set of dimensions which provide the context for that particular set of measures. The dimensions together uniquely determine the measure. Therefore, the multidimensional data views a measure as a value in the multidimensional space of dimensions [9]. Each dimension is a closed set of attributes that more distinctly defines or stratifies a particular measure or set of measures. Figure 9.5 shows a simple example of a multidimensional cube space in a cardiology data warehouse.

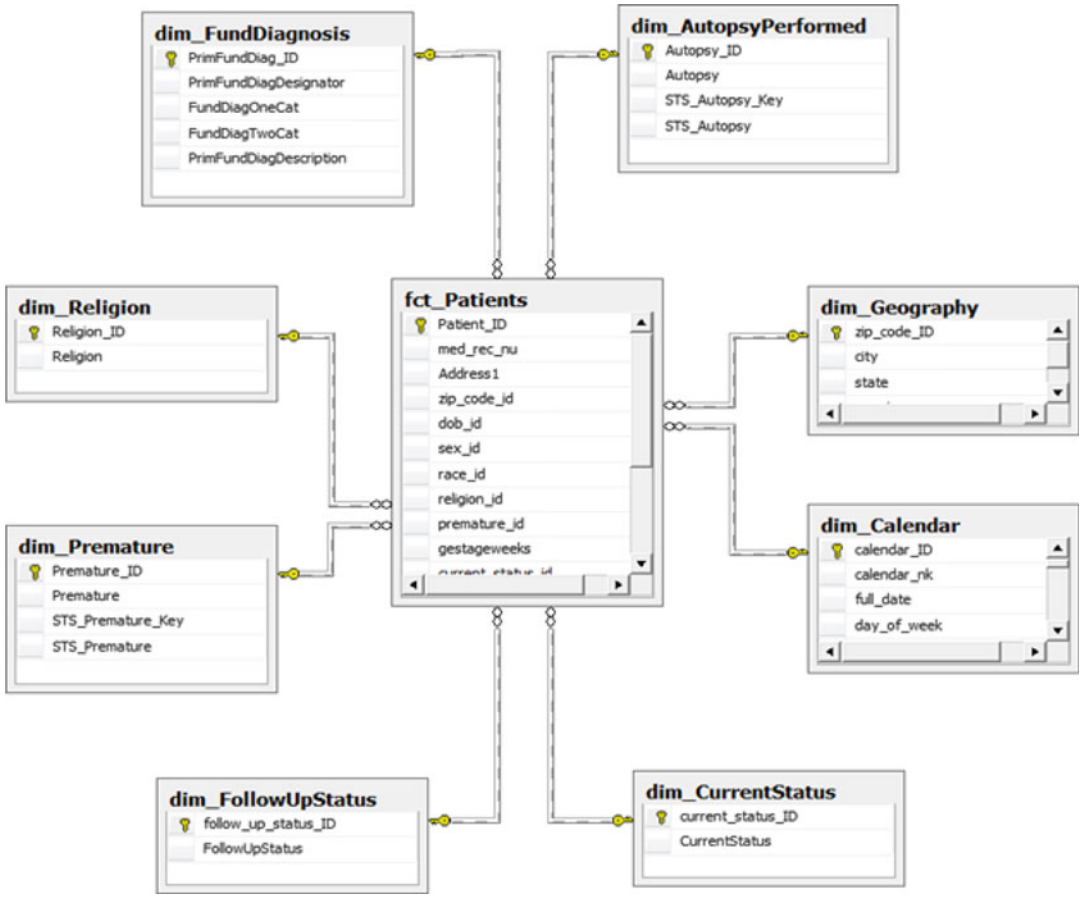

Fig. 9.4 The Online Analytical Processing (OLAP) system (Data Warehouse) star schema. Surrounding the fact table, and connected to it though primary to foreign key relationships, are any number of dimension tables (i.e. dim_CurrentStatus or dim_Geography). A fact table contains all of the measurements of interest in analyzing a particular process or unit type. A cardiac center data warehouse would very likely have separate fact tables for cardiac catheterizations and echocardiograms. Examples of measures that would be stored for a cardiac catheterization fact table would be fluoroscopy time, contrast volume, age, and weight, and any other data that might be submitted to a registry, for instance

Registry Interface

As noted above, data submitted to a registry is a sub-set of data in an EMR. Indeed, a registry submission should be a sub-set of the CCDB. If a CCDB exists only for the purposes of submitting to a single registry, then it is limited in scope factually, temporally and in utility.

With regards to quality, a registry must have assurance that incoming data meets a minimum standard. This is most commonly done by auditing submitting entities to verify that the submitted data sets are accurate and complete.

From the registry perspective, audits are required simply because there is no other way to certify incoming data accuracy. Typically there is no alternate independent entity which can audit and certify the veracity of data submitted from an entity. A CCDB with a data warehouse can simplify the audit process since the 'data trail' in the database is inherently explicit. In the example shown in Fig. 9.4, data to be submitted to a given registry would occupy one or several dimension tables in addition to the central fact tables.

With this type of systems approach to CCDB organization, it would seem reasonable that an

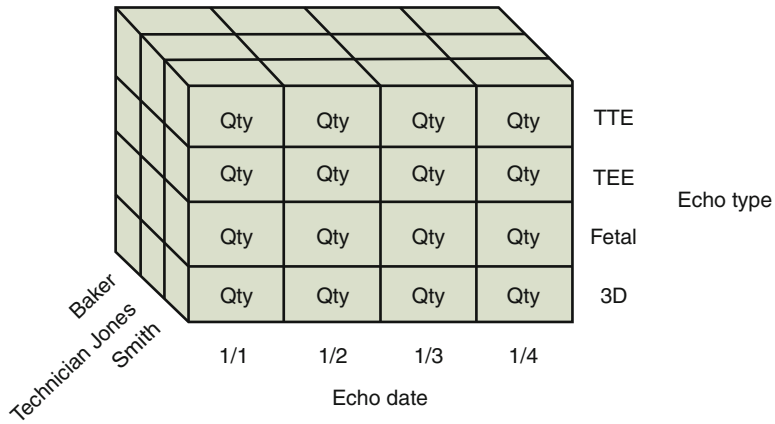

Fig. 9.5 Fact table. The fact table would be representative of echocardiogram measures (Qty). In addition, each row in the echocardiogram fact table would also include the key value for each of the associated dimension tables. In this case, there would be one key for each echo row that pointed to the primary date key of the date dimension, one key that pointed to the primary technician key of the person who performed the echocardiogram in the technician dimension, and one key that pointed to the primary echocardiogram type key in the echo type dimension table

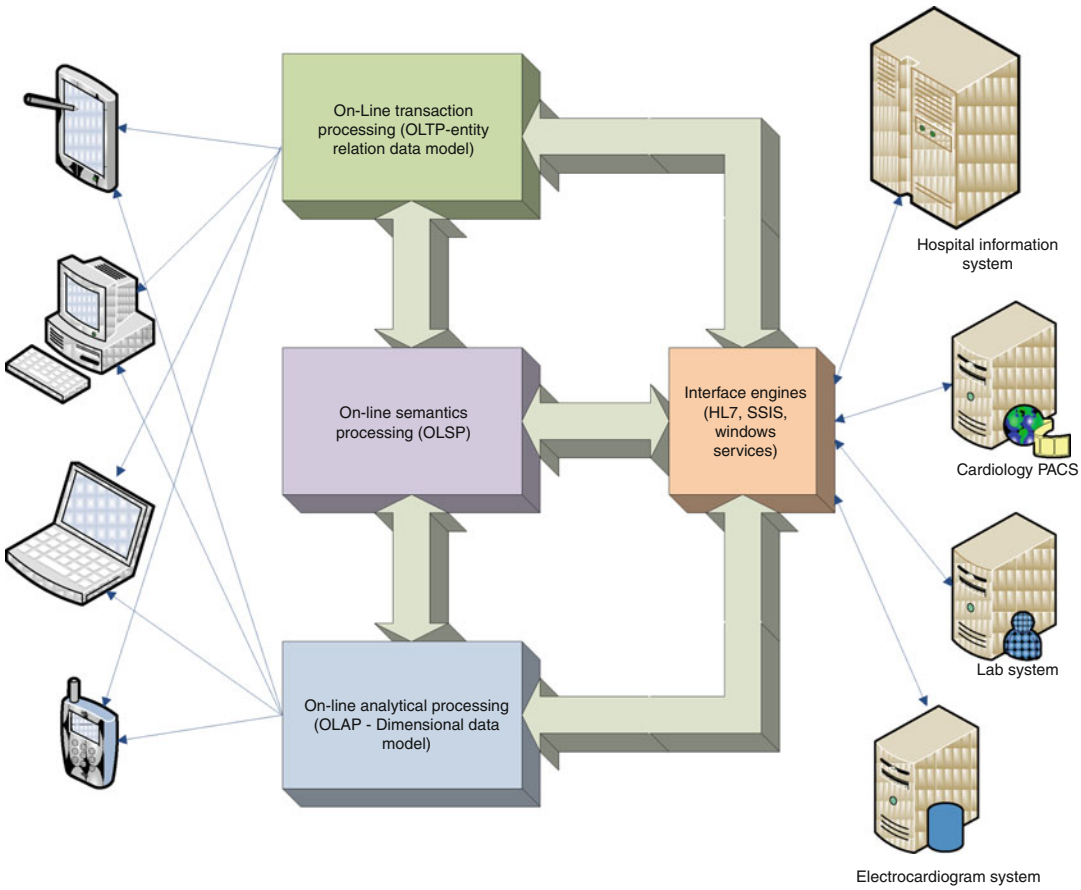

Fig. 9.6 The cardiac center database. Data can arrive from any number of sources. Human interfaces can include smart phones, tablets, laptops, and PCs. Various machine systems connect through appropriate interfaces (hospital information system via Health Level 7 [HIS via HL-7], Cardiology PACS via Open Database Connectivity, etc.). Outbound interfaces allow CCDB components to connect to the web, desktop clients, registries, and cloud aggregate databases

audit of the entire CCDB at regular time intervals, with a resulting 'clean bill of health', allows a CCDB permission to submit to accepting registries until the next audit cycle. This type of audit could assure uniformity between institutional CCDB participants with regards to coding, data handling, practices and work flow.

Prior to submitting data to a registry the database should run an internal error checking script. This would be performed to assure that there are no null values, date ranges are valid (make sure that admission dates are not prior to birth dates, etc.), and values generally fall within realistic ranges (i.e., hematocrit is not 139 %). The script produces a list of anomalies that can be double checked prior to submission.

A good registry interface from a CCDB will then do what any good interface does: promote and facilitate data submission, provide error checking, and organize and sort information.

On-Line Semantics Processing (OLSP) System

While not traditionally considered a core component of advanced database systems, the OLSP system is an important component in the context of congenital heart disease databases. It creates and supports internal hierarchical structure building, translation and mapping.

From the psychological standpoint, we all do semantic processing when we understand the meaning of a word and can relate it to similar words with similar meaning. The computational meaning is similar. The OLSP system can look at code, understand the meaning behind the code, and relate it to similar codes with similar meanings despite the fact that they may be from different code sets.

The OLSP system resides between the transactional layer and the data warehouse, as can be seen when registry tables are being stored in the data warehouse. All databases have an internal code set. For instance, the code set for a spreadsheet might consist of the address "A4," referring to the field in column "A" and row "4." More sophisticated databases have more sophisticated internal code sets, which may be defined by almost any variable, including an address (as in the spreadsheet example), a number, or a term such as "peak_sys_RV_pressure_echo_TR_jet_estimate_apical4chamber." The internal code set provides a ground, a basis and framework with which to begin relating different sets of codes.

This allows the OLSP system to relate any other outside codeset, database, or machine interface to the transactional (OLTP) system. In fact, an effective OLSP system resides in the middle of just about every transaction in the CCDB (Fig. 9.6).

The OLSP is essentially an intelligent mapping system, which understands hierarchical code set organization and facilitates rapid 'translation' between different systems. While interfaces can be built for each different data source, an effective OLSP system can otherwise ameliorate the need for new and unique interface construction requisite with the acquisition of new systems and hardware. The OLSP system adds flexibility and robustness, and benefits from the integration of artificial intelligence.

Artificial Intelligence

Once data are extracted, transformed, and loaded into the data warehouse, knowledge discovery is the next crucial step. In addition to standard statistical analysis, significant advances in automated understanding and interpretation of large amounts of data have been made in recent years. This field of research, known as Artificial intelligence (AI), includes the use of intelligent agents, artificial neural networks, Bayesian belief networks, and other machine learning algorithms. These tools are used to transform discrete data into actionable knowledge, and can be implemented with data sets of almost any size [10].

Intelligent agents [11] are autonomous software programs which accommodate new problem solving rules adaptively in real time. They can sense and interact with their environment, and are able to develop goal-directed behavior based upon the combination of their current environment and prior positively reinforced goal-seeking actions.

Artificial neural networks [12] are similar to intelligent agents, except that they are designed

to solve only one specific problem. Neural networks can be thought of as "black boxes," since neural networks are initially provided with training data representing groups of inputs along with corresponding decisions classifications (correct outputs). Neural networks then build an internal model utilizing any approach that maximizes correct decision classifications. Once trained, a neural network always uses the learned model to make decisions. When faced with new or additional data, an artificial neural network will persist in its trained pattern until such time as it is retrained to be able to handle the new or additional data.

Bayesian Networks are conditional probabilistic models based on Bayes Theorem [13]. If provided with a *directed acyclic graph* (decision state diagram) and prior probabilities, a Bayesian network will make likelihood predictions based upon current (posterior) information. Bayesian networks can work with relatively smaller data sets. Such models can be trained with either real data or a probability estimate if no actual prior data exists. As a Bayesian network is trained it will become more accurate with its probability estimates. The power of a Bayesian network is in its ability to provide more accurate probabilities of remaining variables as known classifications of other variables become facts. For instance, if a patient presents with pulmonary atresia and an intact interventricular septum (PAIVS), the physician will know if that patient is male or female. That variable is then changed from a probability (56 %/44 %) to a fact (100 % probability of female). All other remaining conditional probabilities (in this case mortality) are then recalculated more accurately based upon the uncertainty of gender being removed.

For analyzing large amounts of data, other standard machine learning techniques are useful, including classification, association, clustering, and numeric prediction. Classification is a structured machine learning method in which software learns how to predict new categorical dependent variables based upon previously learned examples. Association is an unstructured machine learning technique in which any interesting relationships, between all attributes, is sought. Clustering is a machine learning technique that combines individual variables into coherent groups that seem to fall naturally together. Numeric Prediction is a machine learning technique that builds models that accurately predict numeric values based upon previously learned examples.

The Cloud

With the above tools available to institutions, each one having a sophisticated CCDB, it can be seen that the potential for sharing large amounts of certified, coded, and similarly organized data is greatly facilitated. In fact, the potential amount of data shared could dwarf the data contained in any single registry. Properly de-identified, certified inter-institutional data could be pooled and analyzed, in essence creating 'virtual registries.' Data parameters could be identified to study a particular quality or outcome study, and large amounts of certified data from large numbers of patients could be available to immediately study the question. While these types of studies are not truly prospective, if care is taken when collecting and storing the data then data collection is in a way already blinded and prospective.

Summary

What data are pediatric cardiologists interested in putting into a departmental database? Given the diversity of pediatric cardiology practice, the easy, glib and likely true answer is "all of it."

The desire to understand outcomes and the need for continued quality improvement are prime drivers for the development of effective registries and databases. Pediatric Cardiology interests include understanding both surgical and catheter based procedure outcomes, as well as non-procedure based indices such as tracking carotid artery intimal thickness in preventative cardiology clinics and understanding the cumulative risk of necrotizing enterocolitis factoring for prematurity, mean arterial blood pressure, and feeding rates in babies with congenital heart disease.

The Cardiac Center Departmental Database will ideally collect and organize data from inpatient and outpatient patient encounters, the operating room, cardiac catheterization laboratory, electrophysiology laboratory, exercise laboratory, imaging encounters (echo, MRI, computed tomography [CT], etc.), hospital chemistry and hematology laboratories, pathology and genetics laboratories, and any other connected system of interest.

Good data collection will facilitate not only understanding outcomes and quality measures, but improve our knowledge of aspects of pathophysiology and wellness. While it is of course important to track changes in the physical exam, ECG, echocardiogram, and MRI/CT it is also important to track feedings, collect parent reports and follow serial neurologic tests. This means that data collection should occur not only from within a hospital, but from outside locations as well. Cloud and web-based interfaces to and from the CCDB will facilitate data collection directly from patients' homes and primary care physician offices. The opportunity to build educational and data collection protocols into outside home and physician office interfaces means that quality of care for cardiac patients can be incrementally improved while at the same time the amount of coded observational information available for analysis can be increased.

Human-database interfaces often rely on structured reporting, which has at least a partially deserved reputation for being insufficiently flexible and incomplete. While it is likely that structured reporting is here to stay in one form or another, semantic interfaces which can search text files for information are becoming increasingly common. These semantic interfaces could use natural language processing [14] to coded database entries from raw text files. Data gathered in this fashion needs to be fastidiously verified, but this technique holds promise for increasing the information gathered into a CCDB.

Ideally, every datum should be coded from the moment it enters a database. The underlying code set of a departmental CCDB need not be identical to any particular registries or published code set, but it does need to translate into various registry (STS, CCISC, IMPACT, EACTS) and common external code sets (International Pediatric and Congenital Cardiac Code [IPCCC]). The internal CCDB code set should therefore be as granular as possible to allow for the highest internal data resolution.

We are living in an age of change- the Silicon Revolution- at least as significant if not more so that the Industrial Revolution. It is truly impossible to anticipate all of the possibilities and opportunities in front of us. We have the ongoing challenge of building the best "systems of care" to take the best care of our patients. These systems will integrate machines, databases, physicians, health care professionals, schools, families and patients to provide cutting edge, top tier therapies for the most complex diseases. This is a good time to be practicing medicine. And we will make it better.

References

1. The free online dictionary. http://www.thefreedictionary.com/interface. Accessed 3 June 2013.
2. Wikipedia – disease registries. http://en.wikipedia.org/wiki/Disease_registry. Accessed 3 June 2013.
3. IMPACT Registry™ website. https://www.ncdr.com/webncdr/impact/. Accessed 3 June 2013.
4. Jenkins KJ, Beekman III RH, Bergersen LJ, Everett AD, Forbes TJ, Franklin RC, Klitzner TS, Krogman ON, Martin GR, Webb CL. Databases for assessing the outcomes of the treatment of patients with congenital and paediatric cardiac disease – the perspective of cardiology. Cardiol Young. 2008;18:116.
5. Kimball R, Reeves L, Ross M, Thornthwaite W. The data warehouse lifecycle toolkit: expert methods for designing, developing, and deploying data warehouses. New York: Wiley; 1998.
6. Dennis PA, Grist GE, Lofland GK Jr., O'Brien JE, Stroup R, Tarrants ML. System and method for collecting, organizing and presenting research-oriented medical information. U.S. Patent Number 7,512,541, issued 2009.
7. Vishwanath K, Viswanath V, Drake W, Lee Y. OntoDiagram: automatic diagram generation for congenital heart defects in pediatric cardiology. AMIA Annu Symp Proc. 2005;754–758.
8. William H. Inmon. 1992. Building the Data Warehouse. John Wiley & Sons, Inc., New York, NY, USA.
9. Chaudhuri S, Dayal U. An overview of data warehousing and Olap technology. ACM Sigmod Rec. 1997;26(1):65–74.
10. Stroup R, O'Brien J, et al. A data model for congenital heart disease research. Orlando: American Medical Informatics Association (AMIA) Spring Congress; 2007.

11. Russell S, Norvig P. Artificial intelligence: a modern approach. 2nd ed. Upper Saddle River: Prentice Hall; 2003. Chapter 2.
12. Mehrota K, Mohan CK, Ranka S. 1996. Elements of Artificial Neural Networks. MIT Press, Cambridge, MA, USA.
13. Dunson D. Commentary: practical advantages of Bayesian analysis of epidemiologic data. Am J Epidemiol. 2001;153(12):1222–6.
14. Lee Y, Supekar K, Geller J. Ontology integration: experience with medical terminologies. Comput Biol Med. 2006;36(7–8):893–919.

10 Databases for Assessing the Outcomes of the Treatment of Patients with Congenital and Pediatric Cardiac Disease: The Perspective of Anesthesia

David F. Vener

Abstract

The Joint Congenital Cardiac Anesthesia Society-Society of Thoracic Surgeons Congenital Cardiac Anesthesia Database is a multi-institutional registry that tracks variables related to the anesthetic management of patients with pediatric and congenital cardiac disease. This registry is an optional module of The Society of Thoracic Surgeons Congenital Heart Surgery Database and is part of the Congenital Cardiac Anesthesia Society's commitment to patient care and research on outcomes improvement. Patients in the registry include not only cardiac surgical patients but also those with pediatric and congenital cardiac disease undergoing procedures in locations other than the operating room, including in the cardiac catheterization laboratory, intensive care unit, general operating room, and radiology suite. Because of the relative infrequency of anesthesia-related events in this low-volume procedure, a multi-institutional database is the most reasonable approach to capture a sufficient number of patient encounters in a timely manner to support outcomes analysis, quality assessment, and quality improvement.

Children undergoing repair of congenital heart defects are among the sickest population treated by anesthesiologists. The incidence of complications such as cardiac arrest related to anesthesia is proportionally much higher, the difficulties associated with airway and vascular access are well known, and the time and money spent on their care is substantial. To date there have been no systematic reviews of their anesthetic care and the associated complications, particularly those that do not result in cardiac arrest. The Joint Congenital Cardiac Anesthesia Society-Society of

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Thoracic Surgeons Congenital Cardiac Anesthesia Database marks the first real-time picture of the "state-of-the-art" of anesthetic care for patient with pediatric and congenital cardiac disease. This information will help guide future care as well as provide better information for the patients and their families.

Keywords

Anesthesia • Pediatric cardiac disease • Congenital cardiac disease • Joint Congenital Cardiac Anesthesia Society-Society of Thoracic Surgeons Congenital Cardiac Anesthesia Database

Background

Anesthesia for patients with congenital heart disease (CHD) is a frequent occurrence in children's hospitals as well as outpatient surgical centers and clinics worldwide. While many of the procedures are specifically related to the patient's heart defect, it is also common for these patients to undergo diagnostic and therapeutic interventions unrelated to their heart defect. Multiple investigations have now shown that this patient population is particularly vulnerable to anesthesia-related complications both in the cardiac operating rooms and in other locations as compared to the non-CHD patients [1–4]. There are ongoing discussions within the pediatric cardiac anesthesia community about both where and who should be caring for these patients and about stratifying patients into higher- and lower-risk populations. For example, a patient with an unrepaired ASD, restrictive VSD or other stable two-ventricle lesions probably represent a different risk group than those with pulmonary hypertension, single ventricle anatomy (functionally univentricular hearts), heart failure, severe aortic stenosis or shunt-dependent lesions. Until recently, data about improvements in outcomes in these patients has been limited due to the relative infrequency of cardiac arrest, the primary endpoint measurement of most outcome studies; meanwhile, almost nothing has been published about either associated morbidities or need-to-rescue interventions that were successful. Improved outcomes, or at least fewer cardiac arrests, are potentially possible by identifying the higher-risk patients and involving physicians familiar with congenital cardiac physiology in their care early on to prevent problems before they arise [5]. Unfortunately, without continuous surveillance monitoring by external observers, it is exceedingly difficult to document "prevented" adverse events or "near-misses."

Children's Hospital Boston and the Mayo Clinic have previously published their single-center results of anesthesia-related cardiac arrest in congenital heart disease patients [1, 3, 6]. Odegard et al. have reported on cardiac arrests, defined as cessation of circulation requiring chest compressions, at their institution in both the cardiac operating rooms and cardiac catheterization labs at Children's Hospital Boston. Their first publication reviewed 5,213 cardiac surgical patients cared for over a 6 year period between January 2000 and December 2005, during which they found 41 episodes of cardiac arrest probably related to anesthesia in 40 patients for an overall frequency of 0.79 % [3]. All of these children were cared for by a dedicated congenital cardiac anesthesia group. Their second publication examined the anesthesia records of 7,289 cardiac catheterizations from 2004 to 2009 and found 70 episodes of cardiac arrest (0.96 arrests per 100 procedures), of which only seven were felt to be likely related to anesthesia or nurse-managed sedation with no mortality [6]. Faculty at the Mayo Clinic in Rochester, Minnesota reviewed the incidence of perioperative cardiac arrest in 92,881 children undergoing all types of surgery

at their facility from November 1988 to June 2005. Four thousand two hundred forty-two of those patients were undergoing cardiac surgery during that 17 year period. They found that the incidence of cardiac arrest was 2.9 per 10,000 patients in the non-cardiac procedures compared to 127 per 10,000 in the cardiac surgical group. Anesthesia was found to be the primary cause of the arrest in only 7.5 % of all the 80 recorded cardiac arrests – the remainder due to factors other than anesthesia. Within the 80 patients who suffered cardiac arrests under anesthesia, 88 % occurred in patients with a history of congenital heart disease, regardless of the type of surgery being performed – a testament to the critical nature of these patients regardless of the procedure being performed. Anesthesia-related adverse events other than those leading to cardiac arrest are not discussed in any of these publications.

For pediatric anesthesia practitioners, the Peri-Operative Cardiac Arrest (POCA) Registry was one of the first multi-site studies examining the etiology and incidence of cardiac arrests in children. The registry, which was active from 1994 to 2005, was a voluntary reporting survey which compiled extensive data concerning cardiac arrests in patients less than 18 years of age. Participating institutions agreed to provide the POCA investigators with detailed information any time a cardiac arrest, defined for their purposes as the initiation of chest compressions or death, occurred. Independent examiners then determined whether the cardiac arrest was due to anesthesia-related factors versus non-anesthesia elements such as surgical manipulation. At various times the POCA registry had between 58 and 79 participants, ranging from free-standing pediatric hospitals to pediatric units located within larger adult institutions. Comparing the two Peri-Operative Cardiac Arrest (POCA) Registry results from the initial publication in 2000 to the update published in 2007 clearly illustrates the effect of the changes in anesthesia practice over long time periods – a major flaw in long-term longitudinal studies [4, 7, 8]. From the 1960s through the 1990s, the general anesthetic halothane was commonly used despite its widely known negative inotropic and chronotropic effects. This agent was the only one then available that was well-tolerated for inhalation induction of anesthesia. Beginning in the late 1990s, a newer agent, sevoflurane, was introduced into practice in the United States. Like halothane, it is readily tolerated for inhalation induction of anesthesia but is not associated with the negative cardiac effects at therapeutic levels that halothane is. In the 2000 report, containing data collected from 1994 to 1997, medication-related cardiac arrests accounted for 37 % of the reported arrests, while the results from 1998 to 2004 showed a medication-related incidence of 18 %, which the authors largely ascribed to the decline in halothane usage during this time period.

At its conclusion, the POCA registry had collected information on 373 anesthesia-related cardiac arrests. 127 of the 373 (34 %) patients determined to have an anesthesia-related event had congenital or acquired CHD. Ramamoorthy et al. examined the POCA data specifically to determine the effects of CHD on arrest etiology and outcomes [2]. They found that the 127 children with underlying CHD were both sicker than their non-CHD counterparts and more likely to arrest from cardiovascular-related events. Fifty-four percent of the arrests reported in the POCA registry in children with CHD occurred outside of the cardiac ORs, while 26 % were from cardiac ORs and 17 % in the cardiac catheterization labs. The lesion most associated with cardiac arrest was "single ventricle," while those most likely to have the highest mortality were aortic stenosis and cardiomyopathy – the former can be very difficult patients to resuscitate once the arrest has occurred, while the latter is associated with a significant incidence of sudden cardiac death [9, 10]. Because the POCA data did not have sufficient information about the total numbers of procedures performed on children less than 18 years of age at the reporting institutions, the investigators could not determine an accurate incidence of arrest. Analysis by Ramamoorthy et al. of the POCA data led to their recommendation that those involved in the care of these children understand the physiology of CHD, particularly those patients with unrepaired or

partially-palliated single ventricle as well as the pharmacodynamics of anesthetic agents in patients with impaired ventricular function [2].

Outside of pediatric anesthesia there are at least two major multi-site collection efforts ongoing utilizing Automated Anesthesia Information Systems (AIMS) data. The American Society of Anesthesiology's Anesthesia Quality Institute has developed a National Anesthesia Clinical Outcomes Registry (NACOR) while the Department of Anesthesia at the University of Michigan has developed the Multicenter Perioperative Outcomes Group. Both systems utilize a complete download of de-identified physiologic and anesthetic data from their participating centers. This data harvest can then be "mined" extensively to determine relationships between anesthetic management strategies and physiologic readings and subsequent outcomes [11–13]. The knowledge gained from these processes may eventually prove helpful in developing similar mechanisms of data-pooling for pediatric anesthesia.

Multi-Societal Collaboration

A considerable effort has been made over the last 15 years to develop a common language for international usage for patients with pediatric and congenital cardiac disease. As congenital heart programs evolved over the years, cardiac lesions and their treatments took on a wide variety of names, occasionally reflecting the embryological origin of the affected anatomy, the final appearance of the anatomy, and frequently an eponym for the name of the person most associated with first describing either the lesion itself or its surgical repair. Thus, a Central Shunt, a "Melbourne" Shunt and a "Mee" Shunt all describe variations on a surgical procedure providing systemic blood flow to the pulmonary arterial system directly off of the aorta. As described in the chapter in this book by Franklin and colleagues titled: "***Nomenclature for Congenital and Pediatric Cardiac Disease***: ***Historical Perspectives and the International Pediatric and Congenital Cardiac Code***", in 2000, an international group of physicians from multiple professional medical societies began to meet with the goals of developing a standardized global nomenclature for patients with pediatric and congenital cardiac disease, resulting in the creation of ***The International Pediatric and Congenital Cardiac Code*** (***IPCCC***) [www.ipccc.net], which has standardized the nomenclature for congenital cardiac malformations and the procedures associated with their repair [14]. The IPCCC is maintained by ***The International Society for Nomenclature of Paediatric and Congenital Heart Disease***. Without this dictionary of terminology, it would be impossible to develop and implement databases both nationally and internationally. The IPCCC is being incorporated into upcoming revision of the International Classification of Diseases (ICD), ICD-11, and is freely available to interested users at www.ipccc.net.

These efforts have been expanded now to an international group of specialists including cardiac surgeons, cardiologists, anesthesiologists, cardiac intensive care specialists, anatomists, nurses, and government representatives who meet annually as: ***The Multi-Societal Database Committee for Pediatric and Congenital Heart Disease***. The Multi-Societal Database Committee for Pediatric and Congenital Heart Disease meets annually to agree on definitions of database standards, strategies for linking databases across geographical, temporal, and subspecialty boundaries, and even the definition of complications associated with pediatric and congenital cardiac care [15–29].

The Joint Congenital Cardiac Anesthesia Society-Society of Thoracic Surgeons Congenital Cardiac Anesthesia Database

The Congenital Cardiac Anesthesia Society (CCAS), an affiliate of the Society of Pediatric Anesthesia, was formed in 2005. Its membership is open to anesthesiologists worldwide who either care for or have an interest in patients with congenital cardiac defects. Among its primary goals, the CCAS has committed to developing a

multi-site database covering anesthesia-related information in patients undergoing surgery or procedures in and out of the cardiac surgical suites, including non-cardiac surgery on patients with congenital heart disease (CHD). The CCAS chose to partner with the Society of Thoracic Surgeons' Congenital Heart Surgery Database (STS-CHSD) because of the enormous amount of shared data elements as well as the STS's long history of successful implementation of their databases. Because of our "shared" patient population, the collaboration between the two societies was a natural fit and strongly supported by the membership of both organizations. As part of the agreement, STS, for the first time, allowed the collection of data from non-cardiac surgical and cardiology procedures occurring in patients with a history of congenital heart defects – which, as discussed above, is one of the major areas where anesthesia-related morbidity occurs [30, 31].

The goal of the Joint CCAS-STS Congenital Cardiac Anesthesia Database is to provide a more "real-time" picture of the state of anesthetic care for these patients with pediatric and congenital cardiac disease, as well as information about outcomes related to the incidence of anesthetic complications beyond cardiac arrest. Single-site analysis of this patient population requires years of data collection because of the relatively small number of cases at any one institution. Multi-site data collection will allow investigators to collect and analyze data from a much larger patient population and report the results back to the CCAS membership and participating institutions in a timely manner [17, 32].

Joint CCAS-STS Congenital Cardiac Anesthesia Database: Mechanisms

The STS data is harvested semi-annually in the Spring and Fall. The Spring harvest, typically occurring in mid-March, captures and reports data on a calendar year basis, while the Fall harvest does so on an academic July–June calendar. Programs may choose to report their data at one or both of the harvests, and "back-filling" of data from previous years is allowable and encouraged. The requirements and forms for becoming a participant in the database are available online via the STS [33]. Because transmission of potential Private Health Information (PHI) is involved in the process, it is necessary for a business agreement to be in place prior to submission of data in order to be compliant with the Health Insurance Portability and Accountability Act of 1996 of the United States of America (HIPAA) Privacy and Security Rules [33].

Groups submitting data for the anesthesia portion of the STS-CHSD are responsible for paying a flat $3,300 per annum, regardless of the number of anesthesiologists participating or the quantity and type of cases submitted. This fee is in addition to the surgical participation fee. The CCAS negotiated a flat-fee with the STS because the traditional fee mechanism the STS had utilized, a charge for each surgeon and an additional charge per case submitted, would potentially discourage groups from participating because of the high annual costs. There are typically significantly more anesthesiologists providing care in a given program to patients with pediatric and congenital cardiac disease than there are surgeons, as many institutions utilize their cardiac anesthesiologists to cover "remote" locations such as cardiac catheterization labs, intensive care units, and diagnostic and interventional radiology suites, as well as care for patients with pediatric and congenital cardiac disease undergoing non-cardiac procedures. At other locations, the cardiac anesthesia care team is an integral part of the overall anesthesia staffing and many physicians may rotate only intermittently into the cardiac operating rooms. At Texas Children's Hospital in Houston, Texas, for example, there are currently 12 cardiac anesthesiologists and 5 congenital heart surgeons. In addition to the three cardiac ORs, the cardiac anesthesia group is responsible for three cardiac catheterization labs, Cardiac ICU coverage, and one radiology site, as well as acting as consultants for the non-cardiac operating rooms when patients with pediatric and congenital cardiac disease receive care for their for non-cardiac procedures. Additionally, there are several more anesthesiologists in the general anesthesia division with extensive cardiac experience who routinely provide care for cardiac patients having

non-cardiac procedures in the inpatient and outpatient operating rooms. Each of these physicians must sign the business agreement before their data can be submitted. It was felt that a fixed fee approach would encourage greater participation and enrollment of patients, especially among those being cared for outside of the cardiac operating rooms, by not financially penalizing institutions for reporting on this critical information.

It is necessary to have the appropriate software to collect and transmit the data to the Duke Clinical Research Institute (DCRI), the data warehouse and analytic center for the STS-CHSD. All STS-approved vendors for the STS-CHSD are required to include the anesthesia data elements as part of their software package, so there should be no additional fees associated with that element of the data management. Some programs have chosen to utilize locally developed software not commercially available in order to retain access to historical data collected prior to their participation in the STS-CHSD. These programs must follow the same guidelines as commercial products and undergo the same data validation and testing. Meanwhile, other programs who utilize commercially available software have transferred their historical data into their new commercially available vendor supplied software.

The most expensive component of any database is the manpower involved in accurately collecting and entering the data. Data entry may be completed by surgeons, anesthesiologists, cardiac perfusionists, nurses, research assistants or any combination of the above. In some institutions the data is entered directly into the software, some collect the data on paper records for later entry, while still other sites abstract the data from the records post-operatively [34].

Unlike the surgical data elements, the CCAS at this time has not committed to a mechanism of data audit to ensure both data completeness and accuracy. This is a conscious decision on the part of the CCAS Database Committee as various programs are working out the mechanics of their anesthesia data entry and the extent to which they will submit data. Ideally, all patients with pediatric and congenital cardiac disease are being entered at a given site, but for a combination of factors including manpower and training, some participating sites are currently only entering the cases performed in the cardiac operating room, while participating sites are including the procedures performed in the cardiac catheterization lab but not other hospital locations [20]. Regardless, in order to submit the anesthetic data, the cardiac surgical program at the submitting hospital must be a participant in the STS-CHSD. In other words, for a program to participate in the Joint CCAS-STS Congenital Cardiac Anesthesia Database, the program must also participate in the STS-CHSDB itself.

Joint CCAS-STS Congenital Cardiac Anesthesia Database: Data Reporting and Analysis

Anesthesia departments participating in the Joint CCAS-STS Congenital Cardiac Anesthesia Database receive their individual Feedback Reports approximately 2 months after the close of data submission in the Spring and Fall. The report consists of two sets of data:

- The site specific data and
- The aggregate national values.

Uses for the data include tracking personnel activity, the occurrence of complications, statistics about usage of medications, time to extubation, and other important variables. As is the case with the surgical data, participants do not receive information about other locations except in the context of the pooled aggregate national values, and occasionally deidentified hospital specific data. This precludes a site from directly comparing their outcomes against another specific individual site. The business agreement between the sites and the STS explicitly forbids this sort of site-to-site comparison. The anesthesia report is developed by members of the CCAS Database Committee and represents an abstract of the submitted data elements felt important to report, such as types of medications, monitoring modalities, airway management, and complications. Data is broken down into three sections:

- An overall anesthesia report,
- A section specific to cardiac operating room cases (both on-pump and off-pump), and

- A section on non-cardiac OR cases specifically from the cardiac catheterization laboratory.

All sites "own" their own data at all times and are free to conduct whatever research or publications on it that they desire (with appropriate IRB approval). Sites are also welcome to request from DCRI specific information from the national data. Any requests for data from the overall database are routed through the STS Access and Publications Task Force Congenital Subcommittee, which includes a representative from CCAS.

Joint CCAS-STS Congenital Cardiac Anesthesia Database: Dataset Management

The Joint CCAS-STS Congenital Cardiac Anesthesia Database went "live" on January 1, 2010 after several years of development and programming. The data set in use through December 31, 2013 is available through the STS website, as is the newer dataset which went into operation on January 1, 2014 [35]. The data specified for collection is reviewed on a triennial basis, with the latest version, operationalized on January 1, 2014, also available through the STS website. As users have gained experience with the database, changes have been made in this next iteration to "bundle" some drug categories and simplify data entry, while expanding other drug categories such as pulmonary vasodilators, antifibrinolytic agents, and procoagulant medications. The CCAS Database Committee has been in communication regularly to facilitate these changes and communicate them to the STS-CHSD Task Force. Efforts have also been made to eliminate the redundancy of data entry between the surgical side of the data set and the anesthesia portion. For example, blood component usage and near infrared spectroscopy data was previously collected in both areas, but is now concentrated in the anesthesia section in version 3.2, the most recent iteration which became effective in January 2014. Table 10.1 includes the listing and definition of the adverse events being collected in

Table 10.1 Adverse anesthesia event categories in joint CCAS-STS database (v3.2)

Event	Definition
None	No anesthesia-related adverse events noted in the perioperative period
Oral/nasal injury-bleeding	Bleeding noted in oropharynx or epistaxis, dental, lip or nasal injury
Respiratory arrest	Need to intervene in airway management in unplanned way (i.e. converting from cannula to ETT or LMA to ETT)
Laryngospasm requiring medication	Laryngospasm requiring medical intervention other than positive pressure
Difficult intubation/reintubation	Unplanned difficult intubation or reintubation
Bronchospasm	Wheezing requiring medical intervention other than suctioning
Hemoptysis/pulmonary hemorrhage	Bleeding either from endotracheal tube or post-op hemoptysis
Stridor/subglottic stenosis	New onset stridor noted after extubation requiring intervention
Extubation	Unplanned extubation (except if TEE-Related (see below)
Endotracheal tube migration	Endotracheal tube needing to be repositioned in ICU on arrival CXR
Airway injury	Barotrauma/pneumothorax secondary to positive pressure ventilation
Pulmonary hypertensive crisis	Probable or definite PH crisis requiring intervention
Unplanned need to remain intubated due to anesthesia	Need to remain intubated at conclusion of procedure due to anesthesia factors (oversedation, muscle relaxation)
Hypercyanotic episode (Tet spell)	Hypercyanotic episode (decrease in SpO2 >20 % from baseline) requiring intervention other than establishing airway ("Tet" Spell)
Arrhythmia – CVL placement	Arrhythmia therapy needed other than withdrawing wire or catheter
Myocardial injury – CVL placement	Myocardial perforation
Vascular compromise – CVL placement	Extremity ischemia or compromise with CVL placement
Pneumothorax – CVL placement	Pneumothorax during placement of CVL

(continued)

Table 10.1 (continued)

Event	Definition
Vascular access	Inability to obtain desired all desired vascular access within 1 h of induction anesthesia (PIV/Aline/CVL)
Hematoma requiring relocation of catheter	Significant Hematoma that requires changing site of desired access
Arterial puncture	Inadvertent arterial puncture during CVL placement
Intravenous/intraarterial air embolism	Air embolism causing hemodynamic change or ischemia
Arterial line placement – extremity ischemia	Extremity ischemia or compromise with arterial line placement
Intravenous infiltration	Peripheral or central IV infiltration
Bleeding – regional anesthestic site	Bleeding at site of regional anesthetia
Intrathecal puncture – regional	Inadvertent intrathecal puncture during caudal or epidural placement
Local anesthetic toxicity – regional	Systemic evidence of local anesthesia toxicity (ECG changes, CNS changes)
Neurologic injury – regional	Injury to peripheral nerve during regional nerve block
Anaphylaxis/anaphylactoid reaction	Suspected anaphylactic/anaphylactoid reaction requiring intervention for either hemodynamic support or respiratory intervention
Non-allergic drug reaction	Non-anaphylactic reaction such as “Red Man” syndrome or hypotension
Medication administration	Wrong medication administered
Medication dosage	Wrong dosage of correct medication
Intraoperative recall	Recall of intraoperative events
Malignant hyperthermia	Suspected or confirmed MH reaction requiring dantrolene
Protamine reaction	Significant reaction to protamine requiring intervention other than slowing administration
Cardiac arrest – anesthesia related	Cardiac arrest requiring CPR during anesthesia care NOT related to surgical or catheter manipulation
Cardiac arrest – not anesthesia related	Cardiac arrest requiring CPR during surgical or catheter manipulation
TEE-related esophageal bleeding/injury	TEE-related esophageal bleeding noted during or after TEE removal
Esophageal chemical burn	TEE-related injury to esophageal mucosa due to TEE cleaning chemicals
TEE-related airway compromise	TEE-related compromise of ventilation or oxygenation requiring removal of TEE
TEE-related extubation	TEE-related inadvertent extubation of patient
Complications during patient transfer	Any event occurring during movement of patient into/out of procedure, such as loss of IV or arterial line, airway compromise, disconnection of lines
Peripheral nerve injury due to positioning	Temporary or permanent nerve injury noted post-operatively due to positioning during procedure
Integument injury under anesthesia	Skin breakdown or dehiscence or alopecia noted post-operatively due to positioning during procedure
Ocular injury (corneal abrasion or injury)	Ocular injury noted post-operatively such as corneal abrasion
Post-operative nausea/vomiting	PONV requiring unplanned admission
Emergence delirium requiring medication	Emergence agitation or delirium requiring medication
Anesthesia equipment malfunction/failure	Any anesthesia equipment malfunction or failure during procedure
Other	Any event related to anesthesia care not otherwise listed

version 3.2. Updates have been made in this version to better clarify specific adverse events as well as include events not previously listed.

Joint CCAS-STS Congenital Cardiac Anesthesia Database: Results

The Fall 2013 Harvest of the Joint CCAS-STS Congenital Cardiac Anesthesia Database includes operations from January 1, 2010 to June 30, 2013, inclusive, and includes information from 37 programs. These programs were diverse in both geographic location and in case volume. A total of 41,008 discrete records had been submitted for the 42 months since the inception of the Joint CCAS-STS Congenital Cardiac Anesthesia Database on January 1, 2010, covering a wide spectrum of surgical types and ages. These data include:

- 26,953 cardiac surgery procedures,
- 7,532 interventional cardiology procedures, and
- 6,523 anesthetics for "Non-Cardiac, Non-Thoracic Procedure on a Cardiac Patient with Cardiac Anesthesia."

As mentioned, the latter cases will include everything from radiologic procedures to bronchoscopies to general surgeries such as a Ladd's Procedure on a patient with heterotaxy or a tracheostomy on a ventilator-dependent cardiac patient.

The overall anesthesia-related adverse event rate was 1.9 %, with unexpected difficulty with intubation or reintubation being the highest reported complication (0.4 %) followed by vascular access taking more than 1 h (0.3 %). Cardiac arrest not due to surgical manipulation occurred in 76 cases (0.2 %). The full range and incidence of complications reported to date is shown in Table 10.2. Because the reporting is voluntary and there is no audit process in place for the Joint CCAS-STS Congenital Cardiac Anesthesia Database at this time, there is no way to verify the completeness or accuracy of the data and there is a likely bias towards underreporting. With sufficient time and funding however, it is hoped that the Joint CCAS-STS Congenital Cardiac Anesthesia Database will become part of the STS audit process. Another mechanism for ensuring data completeness and accuracy would be the integration of AIMS information similar to the Multicenter Perioperative Outcomes Group or

Table 10.2 CCAS reported adverse events (January 2010–June 2013)

Event	N=	% of total
None/missing	40,218	98.1
Any event	790	1.9
Dental injury	8	0.0
Respiratory arrest	22	0.1
Difficult intubation/reintubation	145	0.4
Stridor/sub-glottic stenosis	56	0.1
Inadvertent extubation	26	0.1
Endotracheal tube malposition	23	0.1
Airway injury/barotrauma	10	0.0
CVP-arrhythmia	15	0.0
CVP-myocardial injury	0	0.0
CVP – vascular compromise	25	0.1
CVP – pneumothorax	1	0.0
Vascular access >1 h	141	0.3
Access related hematoma	21	0.1
Inadvertent arterial puncture	61	0.1
IV/IA air embolus	1	0.0
Regional anesthesia – bleeding	1	0.0
Regional anesthesia – intrathecal puncture	0	0.0
Regional anesthesia – local anesthesia toxicity	0	0.0
Regional anesthesia – neurologic injury	1	0.0
Anaphylaxis/anaphylactoid reaction	35	0.1
Non-allergic drug reaction	22	0.1
Medication error – wrong drug	13	0.0
Medication error – wrong dose	19	0.0
Intraoperative recall	2	0.0
Suspected malignant hyperthermia	1	0.0
Protamine reaction	32	0.1
Cardiac arrest – unrelated to surgery	76	0.2
TEE-related – esophageal bleeding/injury	13	0.0
TEE-related – chemical burn	0	0.0
TEE-related – airway compromise	48	0.1
TEE-related – extubation	16	0.0
Patient transfer event	8	0.0
Peripheral neurologic injury	17	0.0

the NACOR data [8, 9]. These data could be filtered appropriately and fed directly into STS-compliant software, eliminating much of the legwork required currently to enter data manually, and largely eliminating the inherent bias involved in selective reporting of cases or underreporting of adverse events.

Difficult Intubation

A recent large review of intubation in 11,219 pediatric patients from Germany showed that patients undergoing cardiac surgery were associated with a significantly higher rate of difficulty with laryngoscopy visualization (Grade III/IV laryngoscopy view with an age-appropriate Macintosh blade) [36]. The Spring 2012 Feedback Report of the Joint CCAS-STS Congenital Cardiac Anesthesia Database showed a self-reported incidence of "difficult intubation or re-intubation" of 62 cases out of 17,047 cardiac surgical records (0.4 %), which appears to be lower than that reported in previous case series [37]. For the purposes of our dataset, we attempted to focus our definition on "unexpected" difficult intubation rather than capture those patients who were known or suspected to have difficult to manage airways. This may account for the observed discrepancy.

Arterial and Venous Line Placement and Complications

Patients undergoing congenital heart surgery can be particularly difficult to obtain both peripheral and central venous access and arterial access due to their need for repeated access both in the hospital and the cardiac catheterization laboratory. Many of these patients have had thromboses in major vessels over the course of their treatments or repeated access causing scarring or collateral formation. As a result, line placement intraoperatively can take a significant amount of time after induction of anesthesia and may require surgical placement of lines such as radial or ulnar arterial catheters. Additionally, both central venous and arterial line attempts are associated with a significant number of complications which may impact both patient morbidity and mortality as well as adding to hospital length of stays, including vessel injury and thrombosis, myocardial injury, catheter-related blood stream infections, arrhythmia, chylothorax and others [38–42]. The Joint CCAS-STS Congenital Cardiac Anesthesia Database collects information on many of these events, particularly those occurring in the perioperative period such as venous or arterial occlusion, hematoma formation, arrhythmias, or other complications. Additionally, a "complication" category of difficulty with line access requiring more than 1 h after induction of anesthesia is included to try and document the incidence of this event occurring, even though it does not necessarily represent an adverse event. The new Version 3.2 of the Joint CCAS-STS Congenital Cardiac Anesthesia Database includes information about whether ultrasound guidance was utilized in the placement of arterial, venous, or both catheters. Based on practice patterns observed at Texas Children's and discussions with other providers, it appears that the need for surgical arterial cutdowns is decreasing as ultrasound utilization for this procedure is increasing. Additionally, many sites are now taking advantage of this technology to aid in peripheral intravenous line placement both preoperatively by specially trained IV teams and in the operating room and other sites by anesthesiologists [43].

Future Developments and the Unique Patient Identifier

Management and interpretation of data will be a critical component in providing members of CCAS with accurate information about the current state of anesthetic care and will provide direction for future research. It is hoped that the CCAS-STS collaboration will serve as a model for incorporating other specialties into the dataset with the goal of creating a "cradle-to-grave" model [44]. Because cardiac patients may see multiple providers at multiple locations over their lifetime,

members of the STS-CHSD Task Force are striving to collaborate with other subspecialties and professional organizations including The National Institutes of Health and The American College of Cardiology in order to develop a Unique Patient Identifier (Global Unique Identifier or GUID) based upon other variables in the medical record. This confidential alphanumeric identifier, which could potentially be calculated at any treating site, could then be used to track an individual patient across multiple care locations and time. Efforts are also ongoing to link records in the STS-CHSD to the Social Security Master Death File and National Death Index. Duke University's IRB, which covers the research activity at DCRI, has reviewed the ongoing submission of unique identifiers to the STS-CHSD and found this activity to be within the guidelines of the Health Insurance Portability and Accountability Act of 1996 of the United States of America (HIPAA), as has outside counsel retained by the STS to evaluate the process [45]. Nonetheless, not all programs are able yet to provide access to these unique identifiers because of local HIPAA-related concerns. Many Institutional Review Boards (IRBs), whose permission is required, have balked at this level of submission of data, despite the presence of a signed business contract. Commercial vendors supporting the STS-CHSD have included functionality in their software to "strip" the unique identifiers from patient records prior to transmission of data to DCRI for those programs unable to share these data with DCRI.

As described in the chapter in this book by Morales and colleagues titled: "***Use of National Death Registries to Empower Databases in Reporting Longitudinal Follow-up***", efforts are ongoing to transform the STS-CHSD into a platform for longitudinal follow-up. It is only through this process that the STS-CHSD will be able to determine accurately the long-term mortality and morbidity associated with the treatments of patients with pediatric and congenital cardiac disease. For example, a patient with hypoplastic left heart disease will typically undergo at least three major cardiovascular surgical procedures and numerous non-surgical procedures prior to age 4, sometimes at multiple institutions. If this patient happens to suffer a major morbidity or mortality after discharge from the hospital at any time, the institution(s) where the procedures were performed may have no way to record these data in their current STS-CHSD. Alternatively, if a patient undergoes a procedure at Hospital A and then is subsequently admitted to Hospital B due to complications or the need for revision, then the patient may be included in two separate data sets. The Unique Patient Identifier (Global Unique Identifier or GUID) is an attempt to control for these not uncommon scenarios and present a more accurate picture of the time course of various cardiac defects and their repairs. Including anesthetic data in this process will help augment the time-course of these patients and the various interventions involved in their care by adding more data elements beyond cardiac surgery.

Conclusion

Children undergoing repair of congenital heart defects are among the sickest population treated by anesthesiologists. The incidence of complications such as cardiac arrest related to anesthesia is proportionally much higher, the difficulties associated with airway and vascular access are well known, and the time and money spent on their care far outweighs their numbers. To date there have been no systematic reviews of their anesthetic care and the associated complications, particularly those that do not result in cardiac arrest. The Joint CCAS-STS Congenital Cardiac Anesthesia Database marks the first real-time picture of the "state-of-the-art" of anesthetic care for patient with pediatric and congenital cardiac disease [46]. This information will help guide future care as well as provide better information for the patients and their families.

References

1. Flick RP, Sprung J, Harrison TE. Perioperative cardiac arrests in children between 1988 and 2005 at a tertiary referral center: a study of 92,881 patients. Anesthesiology. 2007;106:226–37.
2. Ramamoorthy C, et al. Anesthesia-related cardiac arrest in children with congenital heart disease: data from the

Pediatric Perioperative Cardiac Arrest (POCA) registry. Anesth Analg. 2010;110(5):1376–82.
3. Odegard KC, et al. The frequency of anesthesia-related cardiac arrests in patients with congenital heart disease undergoing cardiac surgery. Anesth Analg. 2007;105:335–43.
4. Bhananker SM, et al. Anesthesia-related cardiac arrest in children: update from the pediatric perioperative cardiac arrest registry. Anesth Analg. 2007;105(2):344–50.
5. Gupta P, et al. Sudden cardiac death under anesthesia in pediatric patient with Williams syndrome: a case report and review of the literature. Ann Card Anaesth. 2010;13(1):44–8.
6. Odegard KC, et al. The frequency of cardiac arrests in patients with congenital heart disease undergoing cardiac catheterization. Anes Analg. 2014;118(1):175–82.
7. Morray JP, et al. Anesthesia-related cardiac arrest in children: initial findings of the Pediatric Perioperative Cardiac Arrest (POCA) registry. Anesthesiology. 2000;93(1):6–14.
8. Caplan L, Vener DF. Databases and outcomes in congenital cardiac anesthesia. World J Pediatr Congenit Heart Surg. 2011;2(4):586–92.
9. Burch TM, et al. Congenital supravalvar aortic stenosis and sudden death associated with anesthesia: what's the mystery? Anesth Analg. 2008;107(6):1848–54.
10. Pahl E, et al. Incidence and risk factors for sudden cardiac death in children with dilated cardiomyopathy: a report from the pediatric cardiomyopathy registry. J Am Coll Cardiol. 2012;59(6):607–15.
11. Dutton RP, DuKatz A. Quality improvement using automated data sources: the anesthesia quality institute. Anesthesiol Clin. 2011;29:439–54.
12. Freundlich RE, Kheterpal S. Perioperative effectiveness research using large databases. Best Pract Res Clin Anesthesiol. 2011;25:489–98.
13. Ramachandran SK, Kheterpal S. Outcomes research using quality improvement databases: evolving opportunities and challenges. Anesthesiol Clin. 2011; 29:71–81.
14. International Society for Nomenclature of Paediatric and Congenital Heart Disease. International Pediatric and Congenital Cardiac Code. IPCCC. [Online] [Cited 9 Apr 2012]. www.ipccc.net.
15. Jacobs JP, et al. The nomenclature of safety and quality of care for patients with congenital cardiac disease: a report of the Society of Thoracic Surgeons Congenital Database Taskforce Subcommittee on patient safety. Cardiol Young. 2008;13 Suppl 2:81–91.
16. Jacobs JP, et al. What is operative mortality? Defining complications in a surgical registry database: a report of the STS congenital database taskforce and the joint EACTS-STS congenital database committee. Ann Thorac Surg. 2006;81:1937–41.
17. Vener DF, et al. Databases for assessing the outcomes of the treatment of patients with congenital and paediatric cardiac disease – the perspective of anaesthesia. Cardiol Young. 2008;18 Suppl 2:124–9.
18. Jacobs JP, et al. The current status and future direction of efforts to create a global database for the outcomes of therapy for congenital heart surgery. Cardiol Young. 2005;1(Suppl):190–7.
19. Bergersen L, et al. Report from the International Society for Nomenclature of Paediatric and Congenital Heart Disease: cardiovascular catheterisation for congenital and paediatric cardiac disease (part 1 – procedureal nomenclature). Cardiol Young. 2011;21(3):252–9.
20. Bergersen L, et al. Report from the International Society for Nomenclature of Paediatric and Congenital Heart Disease: cardiovascular catheterisation for congenital and paediatric cardiac disease (part 2 – nomenclature of complications associated with interventional cardiolo. Cardiol Young. 2011;13(1):260–5.
21. Society of thoracic surgeons. Database HIPAA FAQ. [Online] [Cited 9 Apr 2012.] http://sts.org/sites/default/files/documents/pdf/DCRI_FAQ-Technical_Document_re_Identifiers_in_Database_Final.pdf.
22. Zgleszewski SE, et al. Anesthesia-related cardiac arrest: five year analysis at an academic pediatric medical center. Anesthesiology. 2006;105:A134.
23. Strickland MJ, et al. The importance of nomenclature for congenital cardiac disease: implications for research and evaluation. In: Jacobs JP, editor. 2008 Cardiology in the Young Supplement: Databases and The Assessment of Complications associated with The Treatment of Patients with Congenital Cardiac Disease. Prepared by: The Multi-Societal Database Committee for Pediatric and Congenital Heart Disease. Cardiology in the Young, vol. 18, issue S2 (Suppl 2), p. 92–100, December 9, 2008.
24. Pasquali SK, et al. Linking clinical registry data with administrative data using indirect identifiers: implementaion and validation in the congenital heart surgery population. Am Heart J. 2010;160(6):1099–104.
25. O'Brien SM, et al. An empirically based tool for analyzing mortality associated with congenital heart surgery. J Thorac Cardiovasc Surg. 2009;138(5):1139–53.
26. Jacobs ML, et al. Report of the 2010 Society of Thoracic Surgeons congenital heart surgery practice and manpower survey. Ann Thorac Surg. 2011;92(2):762–8.
27. Jacobs JP, et al. Successful linking of the Society of Thoracic Surgeons database to social security data to examine survival after cardiac operations. Ann Thorac Surg. 2011;92(1):32–7.
28. Patel A, et al. Impact of noncardiac congenital and genetic abnormalities on outcomes in hypoplastic left heart syndrome. Ann Thorac Surg. 2010;89:1805–14.
29. Johnson JN, et al. Center variation and outcomes associated with delayed sternal closure following stage 1 palliation for hypoplastic left heart syndrome. J Thorac Cardiovasc Surg. 2010;139:1205–10.
30. Fudge J, et al. Outcomes in patients with down syndrome undergoing congenital heart surgery: analysis of a national clinical database. Pediatrics. 2010;126: 315–22.
31. Vener DF, Guzzetta N, Jacobs JP, Williams GD. Development and implementation of a new data registry in congenital cardiac anesthesia. Ann Thorac Surg. 2012;94(6):2159–65. doi:10.1016/j.athoracsur.2012.06.070. PMID: 23176940.

32. Society of Thoracic Surgeons Congenital Heart Surgery Database Executive Summary 2010. [Online] [Cited 28 Oct 2011] http://www.sts.org/sites/default/files/documents/STSCONG-%20Executive%20Summary-All%20patients-Spring2011.pdf.
33. STS database collection of protected health information FAQ for STS database participants. Society of Thoracic Surgery. [Online] [Cited: 19 Oct 2011] http://www.sts.org/sites/default/files/documents/pdf/DCRI_FAQ-Technical_Document_re_Identifiers_in_Database_Final.pdf.
34. Vener DF. Quality, outcomes and databases in congenital cardiac anesthesia. In: Andropoulos DB et al., editors. Anesthesia for congenital heart disease. 2nd ed. Chichester: Wiley; 2010. p. 29–36.
35. Society of Thoracic Surgeons. How to become a participant. [Online] [Cited 19 Mar 2012]. http://www.sts.org/sts-national-database/database-participants/how-become-participant.
36. STS congenital heart surgery database v3.0. [Online] 2010. [Cited 19, Mar 2012]. http://www.sts.org/sites/default/files/documents/pdf/ndb/CongenitalDataCollectionForm3_0_Annotated_20090916.pdf.
37. Heinrich S, et al. Incidence and predictors of difficult laryngoscopy in 11,219 pediatric anesthesia procedures. Paediatr Anaesth. 2012;22:729–36.
38. Akpek EA, Mutlu H, Kayhan Z. Difficult intubation in pediatric cardiac anesthesia. J Cardiothorac Vasc Anesth. 2004;18(5):610–2.
39. Elella RA, et al. Impact of bloodstream infection on the outcome of children undergoing cardiac surgery. Pediatr Cardiol. 2010;31:483–9.
40. King MA, et al. Complications associated with arterial catheterization in children. Pediatr Crit Care Med. 2008;9(4):367–71.
41. da Silva PSL, Waisberg J. Induction of life-threatening supraventricular tachycardia during central venous catheter placement: an unusual complication. J Pediatr Surg. 2010;45:E13–6.
42. Manlhiot C, et al. Risk, clinical features, and outcomes of thrombosis associated with pediatric cardiac surgery. Circulation. 2011;124:1511–9.
43. Vener DF. Mechanisms of reporting anesthesia information into the CCAS – STS database. [Abstract]. Las Vegas: Society of Pediatric Anesthesia Annual Meeting; 2013.
44. Rice-Townsend S, et al. Analysis of adverse events in pediatric surgery using criteria validated from the adult population: justifying the need for pediatric-focused outcome measures. J Pediatr Surg. 2010;45:1126–36.
45. Heinrichs J, et al. Ultrasonographically guided peripheral intravenous cannulation of children and adults: a systematic review and meta-analysis. Ann Emerg Med. 2013;61(4):444–54.
46. Vener DF, et al. Anaesthetic complications associated with the treatment of patients with congenital cardiac disease: consensus definitions from the multi-societal database committee for pediatric and congenital heart disease. Cardiol Young. 2008;18 Suppl 2:271–81.

11 Databases for Assessing the Outcomes of the Treatment of Patients with Congenital and Pediatric Cardiac Disease: The Perspective of Critical Care

Michael G. Gaies, Howard E. Jeffries, Randall Wetzel, and Steven M. Schwartz

Abstract

Several barriers exist that make measuring critical care outcomes and quality a challenge. Particularly in the field of pediatric cardiac critical care it can be difficult to disentangle an intensive care unit's contribution to patient outcome from those of other services (e.g. surgery), and appropriate risk-adjustment remains an elusive goal. Databases provide a key source of information that can be used to overcome some of these barriers. We explain the key database components necessary to provide clinicians and researchers with the foundation to measure and improve quality in this clinical arena. Databases that are currently used in the critical care community are described, including the Virtual PICU System (VPS) database. The chapter concludes with a discussion of how to move from simply assessing patient outcomes using databases to achieving quality improvement through collaborative. We review new collaborative in pediatric cardiac critical care and cardiac surgery, the Pediatric Cardiac Critical Care Consortium (PC4), that is implementing the lessons learned from successful quality improvement pioneers.

Keywords

Pediatric • Cardiac • Critical care • Database • Registries • Outcomes • Quality

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Introduction

Pediatric cardiac critical care's maturation as a specialty almost certainly has contributed to improvements in morbidity and mortality after pediatric and congenital heart surgery observed in the modern era [1]. Yet, critical care specialists still seek a deeper understanding of the impact intensive care interventions and morbidities have on short- and long-term patient outcomes in children and adults with critical cardiovascular disease. In order to build upon the progress made to-date, pediatric cardiac critical care clinicians and scientists will need robust data infrastructures to measure cardiac critical care outcomes, assess quality of care, and test new interventions. The pediatric cardiac critical care community faces inherent challenges to achieving these goals. However, multi-disciplinary databases that offer solutions are being created.

This chapter begins with a framing of the barriers to measuring critical care outcomes and quality, focusing on the challenge of disentangling critical care's contribution to patient outcome from other services (e.g. surgery) and on the issue of risk-adjustment. Key components for a successful critical care database are explained. Databases that are currently used in the critical care community are described, including the Virtual PICU System (VPS) database that has been used to track general pediatric critical care, and more recently cardiac critical care, outcomes in North America. The chapter concludes with a discussion of how to move from simply assessing patient outcomes using databases to achieving quality improvement through collaborative learning. A new collaborative in pediatric cardiac critical care and cardiac surgery, the Pediatric Cardiac Critical Care Consortium (PC^4), is implementing the lessons learned from other surgical and cardiology quality collaborations.

Challenges to Measuring Outcomes and Quality of Care

Heterogeneous Patient Populations

Patients from birth to adulthood, with a wide variety of clinical conditions and pathophysiology, are admitted to pediatric cardiac intensive care units (CICU). An important distinction is between patients admitted to the CICU directly from the operating room after palliative or corrective surgery for structural heart disease and those admitted for medical conditions including, but not limited to, pre-operative care, congestive heart failure exacerbation, arrhythmia management, or after cardiac arrest. Defining metrics specific to cardiovascular disease that apply to all patients in this setting remains a difficult task; for example, while surgical site infection may be an important measure of CICU performance in a surgical population, it is not informative for patients with non-surgical conditions. Furthermore, complication rates of cardiac arrest, development of acute kidney injury, and new neurologic injury may be important performance metrics in both populations, but sub-analysis of outcomes within each group, perhaps using separate risk adjustment methods, is necessary.

Defining Critical Care Patient Quality Outcomes and Quality Metrics

The ideal independent outcome measure for pediatric cardiac critical care databases would be one that reflects the competence and quality of care provided by the CICU team, and is unaffected by care prior or subsequent to the CICU admission. While an argument can be made that outcomes measured at the level of the hospitalization episode (e.g. discharge mortality) are most important, these metrics do not allow for a granular understanding of why hospital performance varies, nor do they provide the detailed data necessary to implement improvement strategies. Critical care specialists intent on improving their individual and unit performance need quality metrics that effectively disentangle critical care's contribution to outcomes from other domains.

Post-operative care in the CICU is the best example of this conundrum: it is extremely difficult to determine how both the CICU and surgical care contribute to individual patient and population outcomes, let alone the baseline anatomy, physiology and functional state of the patient.

For example, average length of mechanical ventilation after a particular operative procedure will depend on the ventilation strategy and weaning practices of a CICU team, but will also be influenced by the preoperative physiology, frequency of residual anatomic defects, anesthetic practices and complicating surgical morbidities (e.g. phrenic nerve injury). Propensity to develop acute kidney injury will reflect not just the fluid and hemodynamic management in the intensive care unit (ICU), but may also depend on the preoperative condition of the patient, surgical results, postoperative ventricular function and cardiac output, and perhaps on strategies used during cardiopulmonary bypass for organ protection and mitigating the inflammatory response. While the search to define an "ideal" pediatric cardiac critical care outcome measure in post-operative patients continues, it is probably necessary in the short-term to view outcome measurement as an assessment of an entire pediatric cardiac surgical program.

Outcomes of medical patients with cardiovascular disease treated in the CICU may be more likely to be influenced by the interventions and care of the CICU team. Use of risk-adjustment strategies employed in the general pediatric critical care setting (see below) may be more applicable in these patients than in a post-surgical population. However, it is not clear that benchmarks for certain outcomes (e.g. catheter-associated bloodstream infections, resource use, frequency of cardiac arrest) are applicable to a group of patients with unique pathophysiology compared to a post-cardiac surgical or general pediatric intensive care patient population. Determining how to appropriately risk-adjust surgical and medical patients in the CICU, separately or together, will be critical, especially when considering comparative analyses across centers where the relative percentage of surgical to medical patients may be markedly different.

Risk Adjustment

Adequate risk adjustment is necessary to correctly interpret outcomes and performance and quality metrics collected in multi-institutional registries. Risk stratification systems like the Risk Adjustment for Congenital Heart Surgery-1 (RACHS-1) method [2, 3], the Aristotle methodology [4–6], and the STS-EACTS (STAT) Mortality Score and Categories [7] predict mortality after congenital and pediatric heart surgery and are used for adjusted analyses in surgical databases. However, they do not account for physiologic differences on or after admission to the CICU, and therefore paint an incomplete picture when trying to measure critical care performance. Measuring performance in the CICU must take this physiologic variability and severity of illness into account to best analyze the impact of pediatric cardiac critical care on eventual patient outcome. The procedure-based adjustment methods also do not apply to non-surgical patients with critical cardiovascular disease. Thus, other tools must be incorporated in critical care databases to provide adequate risk-adjustment for outcome interpretation in the CICU.

The Pediatric Index of Mortality-2 (PIM-2) is a physiology-based risk prediction scale developed from and subsequently validated in a general mix of pediatric ICU patients, including cardiac surgery patients, from Australia, New Zealand, and the United Kingdom [8]. Though PIM-2 discriminated well in the original validation study with reasonable calibration in patients with critical cardiac disease, other literature demonstrates that heavy shifts in the patient mix of a cohort, as may be seen in specialized CICUs, may decrease the accuracy of a risk model's predictive ability [9–12]. In a recent study by Czaja and colleagues [9], the PIM-2 demonstrated relatively poor predictiveness in a cohort composed entirely of cardiac surgical patients, particularly with regards to expected mortality in the highest-complexity patients. The newly developed Pediatric Index of Mortality-3 (PIM-3) [13] has yet to be similarly studied. Variables that make up the PIM-3 are shown in Table 11.1. The Pediatric Risk of Mortality (PRISM-III) [14], shown in Table 11.2, also falls short in adequately risk-adjusting outcomes for mixed populations of pediatric cardiac surgical patients [15]. While these scales may perform better in cardiac medical patients, or when comparing patients within the same procedural category, the imperfections of the currently used risk-adjustment systems highlight

Table 11.1 Pediatric index of mortality-3 variables [13]

Pupils fixed to light? (Yes/No)
Elective admission (Yes/No)
Mechanical ventilation in the first hour (Yes/No)
Absolute value of base excess (mmol/L)
SBP at admission (mmHg)
SBP2/1,000
$100 \times Fio_2/Pao_2$ (mmHg)
Recovery post procedure?
Yes, recovery from a bypass cardiac procedure
Yes, recovery from a non-bypass cardiac procedure
Yes, recovery from a noncardiac procedure
Very high-risk diagnosis
High-risk diagnosis
Low-risk diagnosis

Table 11.2 PRISM-III variables [14]

Systolic blood pressure (mmHg)
Heart rate (beats per minute)
Temperature (C°)
Pupillary reflexes
Mental status (Glasgow coma score)
Total CO_2 (mmol)/pH
PaO_2 (mmHg)
PCO_2 (mmHg)
Glucose (mg/dL or mmol/L)
Potassium (mmol/L)
Creatinine (mg/dL or μmol/L)
Blood urea nitrogen (mg/dL or μmol/L)
White blood cell count (cells/mm^2)
Prothrombin time or Partial thromboplastin time (seconds)
Platelet count (cells/mm^2)
Other factors (diagnosis, source of PICU admission)

the need to develop new methods validated primarily in cardiac critical care patients. Without these tools pediatric cardiac critical care databases will fall short in accurately measuring outcomes and quality.

The "Ideal" Cardiac Critical Care Database

The ideal cardiac critical care database for improving quality of care would successfully deal with the challenges described previously around heterogeneity of patients, separating CICU care from other domains, and risk-adjustment. In addition, these databases should include two key components: a standard nomenclature and mechanisms that facilitate linkage between registries.

Common Nomenclature

Accurate measurement and comparison of clinical outcomes is dependent on a common nomenclature and standardized data collection. Thanks to the efforts of The International Society of Nomenclature for Paediatric and Congenital Heart Disease (ISNPCHD) [http://www.ipccc.net/] and the Multi-Societal Database Committee for Pediatric and Congenital Heart Disease (MSDC), a consensus-based, comprehensive nomenclature now exists for the diagnosis, procedures, and complications associated with the treatment of patients with pediatric and congenital cardiac disease [16, 17]. This nomenclature has been adopted by several clinical databases, including most notably:

- The Society of Thoracic Surgeons (STS) Congenital Heart Surgery Database,
- The European Association for Cardio-Thoracic Surgery (EACTS) Congenital Heart Surgery Database,
- The IMPACT Interventional Cardiology Registry™ (**IM**proving **P**ediatric and **A**dult **C**ongenital **T**reatment) of the National Cardiovascular Data Registry® of The American College of Cardiology Foundation® and The Society for Cardiovascular Angiography and Interventions (SCAI),
- The Joint Congenital Cardiac Anesthesia Society- Society of Thoracic Surgeons Congenital Cardiac Anesthesia Database
- The Virtual PICU System (VPS).
- The Pediatric Cardiac Critical Care Consortium (PC^4)

A common nomenclature is crucial for data sharing which allows robust analyses of large integrated datasets.

Linking Databases

Reflecting the nature of clinical care of patients with pediatric and congenital cardiac disease, database creation for measuring outcomes and

quality requires true multi-disciplinary collaboration. Linkage of existing and future multi-institutional subspecialty databases to promote seamless sharing of longitudinal data across temporal, geographical, and subspecialty boundaries will be necessary to achieve the desired objective of quality improvement [18–20]. Clinical and administrative databases have been successfully linked using indirect identifiers [21], and similar techniques could be used to link clinical databases. The key for new database projects is one of design; careful thought must be given when developing the list of data variables and data harvesting procedures to ensure the possibility of future linkage. In addition, innovative new software platforms can also facilitate sharing of data variables between registries more effectively and efficiently than the techniques involving indirect linkage.

Linkages between CICU databases with other large datasets holds promise to answer questions not currently accessible to CICU clinicians. For example, determining the clinical factors associated with variation in resource utilization in the CICU could be approached by linking detailed patient registries with administrative databases. Further, CICU databases, as currently designed, focus on short term, CICU-based outcome measures. Linkages with other registries, like the STS, are likely necessary to determine the impact of CICU practice on intermediate and longer-term outcomes. As with any collaboration, it is crucial to form productive partnerships with the professional organizations and other entities that administer these clinical registries, as there are several issues around data management, ownership, and analysis that that require concerted efforts to manage.

Existing Critical Care Databases

Virtual PICU Systems (VPS) Database

In 1997, the Laura P. and Leland K. Whittier Virtual Pediatric Intensive Care Unit (VPICU) was founded after a generous grant to Children's Hospital Los Angeles from the Whittier Foundation. At the same time the PICU Focus Groups, sponsored by the National Association of Children's Hospitals and Related Institutions (NACHRI), needed a software tool to facilitate multisite quality improvement and research. Both of these organizations realized that a poor understanding of pediatric critical care practice, coupled with the lack of even simple descriptive statistics, left large knowledge gaps around critical care for children. ICUs were neither connected to the Internet nor sharing data. The VPICU and NACHRI collaborated in a development process that included 15 pediatric intensivists. The result was a demographic, diagnostic and severity of illness adjusted software tool that collected information from member units, on all patients, with actual outcomes collected. The VPICU focuses on bringing information technologies to serve pediatric critical care medicine by supporting patient care, quality improvement, distance learning and research. This platform provided a necessary foundation to investigate how pediatric critical care was practiced across the United States.

The original development group refined the data collection and reporting, and in partnership with The National Outcomes Center at Children's Hospital and Health Systems, Milwaukee, improved the quality of comparative reports, the scientific rigor applied to data acquisition, and data integrity. The data collected thus far over 10 years from hundreds of thousands of cases allows for reliable, reproducible, and actionable information for critical care clinicians and hospital adminstrators. VPS currently has 120 member PICUs and data on more than 600,000 children to serve member data needs and quality reports, and is the largest pediatric critical care data repository available. Further, VPS has enabled important research in pediatric critical care, facilitated by a research oversight committee.

A separate cardiac module within the VPS database is available, and in concert with the Society of Thoracic Surgeons (STS), a multi-societal database created to facilitate sharing of information and cultivate data platforms that utilize shared definitions. The VPS adopted the International Pediatric and Congenital Cardiac Code [http://www.ipccc.net/] nomenclature for cardiac diagnoses, cardiac surgical procedures

and cardiac surgical complications. In addition, cardiac complexity scores were introduced into the cardiac module, including RACHS-1 [2, 3], Aristotle Basic Complexity [4–6], and STAT scores [7]. As the number of participating Cardiac Intensive Care Units and mixed PICUs with cardiac surgical populations grow, population-specific mortality and morbidity predictive tools will be developed.

The Future of Cardiac Critical Care Databases

One of the central purposes of any clinical registry or database project should be to collect outcomes data that can be used to drive quality improvement. The core tenets of collaborative quality improvement are:

1. Purposeful collection of granular clinical data,
2. Providing timely performance feedback to clinicians, and
3. Continuous quality improvement based on empirical analysis and collaborative learning.

The Northern New England Cardiovascular Disease Study Group (NNE) is an illustrative example of a successful regional quality collaborative [22]. Formed in 1987 to study outcomes of adult coronary bypass grafting, valve, and percutaneous cardiac procedures, the NNE developed a detailed clinical registry on practice and outcome variables and collected data from voluntary participants in the region. The collaborative described the clinical epidemiology of variation in outcomes between participants, and through subsequent empirical analysis identified key processes of care underlying the observed variation. These findings subsequently informed targeted quality improvement efforts; as a result, mortality rates became among the lowest in the nation, declining from 5 % to under 2 % by the end of the 1990s [22].

A database, by itself, does not facilitate quality improvement; the data must be actionable, rigorous scientific methods should be applied to analyses of the data, and the users of those data must facilitate learning through transparent sharing of practice and outcomes with one another. Effective quality improvement collaboratives first ask the question "Who are the best performers?" but they take the next step by using their data to answer the question of "*Why* do the best performers achieve better outcomes?" The answers to this second question are what inform successful quality improvement initiatives.

Share and colleagues have described the difference between a simple database project and one that effectively drives quality improvement collaboratives [23]. They analyzed rates of complications and mortality for adult patients undergoing general, vascular, and bariatric surgical procedures in statewide quality collaborative programs for these specialties in Michigan. Hospitals participating in a national database project (National Surgical Quality Improvement Program, NSQIP) were compared with the Michigan hospitals. Hospitals participating in the collaboratives had lower adjusted rates of morbidity and mortality compared to hospitals that were contributing data to a simple clinical registry. This analysis highlights the gap between simply measuring outcomes and promoting quality improvement.

The Pediatric Cardiac Critical Care Consortium (PC^4) was created to mimic the structure, function, and success of the NNE and the Michigan surgical collaboratives. Formed in 2009 with funding from the National Institutes of Health, PC^4 is an international, multi-institutional collaborative for the cardiac critical care community. The Consortium has created a clinical registry with outcome and critical care practice variables designed specifically to identify the key drivers of variation between hospitals. PC^4 employs a data platform that has analytic functions capable of empirically deriving risk-adjustment models from the entered data, offering the promise that outcomes will be comparable across centers. This data platform is also capable of providing real-time risk- and reliability-adjusted data to participants to facilitate local and collaborative-wide quality improvement efforts. PC^4 has also partnered with professional organizations and database projects, including the Society of Thoracic Surgeons, Pediatric Cardiac Intensive Care Society and American College of Cardiology, to

standardize data definitions, harmonize data collection efforts, and share data variables across registries through direct and indirect linkage. Data collection with this new platform commenced in 2013.

Conclusion

Though there are unique challenges to measuring and improving quality in the CICU environment, the success of other database and collaborative quality improvement projects provides a road map for success in pediatric cardiac critical care. Adherence to the conceptual foundation on which these successful projects are built should propel current and future database projects in the field to provide risk-adjusted outcome and quality metrics to clinicians and scientists. These data will drive analyses that reveal the modifiable reasons for variation in performance between centers, and fueled by a collaborative spirit the pediatric cardiac critical care community can raise the quality of care for children and adults with critical pediatric and congenital cardiovascular disease throughout the world.

References

1. Boneva RS, Botto LD, Moore CA, Yang Q, Correa A, Erickson JD. Mortality associated with congenital heart defects in the United States: trends and racial disparities, 1979–1997. Circulation. 2001;103:2376–81.
2. Jenkins KJ, Gauvreau K. Center-specific differences in mortality: preliminary analyses using the Risk Adjustment in Congenital Heart Surgery (RACHS-1) method. J Thorac Cardiovasc Surg. 2002;124:97–104.
3. Jenkins KJ, Gauvreau K, Newburger JW, Spray TL, Moller JH, Iezzoni LI. Consensus-based method for risk adjustment for surgery for congenital heart disease. J Thorac Cardiovasc Surg. 2002;123:110–8.
4. Jacobs JP, Jacobs ML, Lacour-Gayet FG, et al. Stratification of complexity improves the utility and accuracy of outcomes analysis in a multi-institutional congenital heart surgery database: application of the Risk Adjustment in Congenital Heart Surgery (RACHS-1) and Aristotle Systems in the Society of Thoracic Surgeons (STS) congenital heart surgery database. Pediatr Cardiol. 2009;30:1117–30.
5. Jacobs JP, Lacour-Gayet FG, Jacobs ML, et al. Initial application in the STS congenital database of complexity adjustment to evaluate surgical case mix and results. Ann Thorac Surg. 2005;79:1635–49; discussion -49.
6. Jacobs ML, Jacobs JP, Jenkins KJ, Gauvreau K, Clarke DR, Lacour-Gayet F. Stratification of complexity: the risk adjustment for congenital heart surgery-1 method and the Aristotle complexity score–past, present, and future. Cardiol Young. 2008; 18 Suppl 2:163–8.
7. Jacobs JP, Jacobs ML, Maruszewski B, et al. Initial application in the EACTS and STS congenital heart surgery databases of an empirically derived methodology of complexity adjustment to evaluate surgical case mix and results. Eur J Cardiothorac Surg. 2012;42:775–9; discussion 779–80.
8. Slater A, Shann F, Pearson G, Paediatric Index of Mortality Study Group. PIM2: a revised version of the paediatric index of mortality. Intensive Care Med. 2003;29:278–85.
9. Czaja AS, Scanlon MC, Kuhn EM, Jeffries HE. Performance of the Pediatric Index of Mortality 2 for pediatric cardiac surgery patients. Pediatr Crit Care Med. 2011;12:184–9.
10. Metnitz PG, Lang T, Vesely H, Valentin A, Le Gall JR. Ratios of observed to expected mortality are affected by differences in case mix and quality of care. Intensive Care Med. 2000;26:1466–72.
11. Murphy-Filkins R, Teres D, Lemeshow S, Hosmer DW. Effect of changing patient mix on the performance of an intensive care unit severity-of-illness model: how to distinguish a general from a specialty intensive care unit. Crit Care Med. 1996;24:1968–73.
12. Patel PA, Grant BJ. Application of mortality prediction systems to individual intensive care units. Intensive Care Med. 1999;25:977–82.
13. Straney L, Clements A, Parslow RC, et al. Paediatric index of mortality 3: an updated model for predicting mortality in pediatric intensive care*. Pediatr Crit Care Med. 2013;14:673–81.
14. Pollack MM, Patel KM, Ruttimann UE. PRISM III: an updated pediatric risk of mortality score. Crit Care Med. 1996;24:743–52.
15. Mildh L, Pettila V, Sairanen H, Rautiainen P. Predictive value of paediatric risk of mortality score and risk adjustment for congenital heart surgery score after paediatric open-heart surgery. Interact Cardiovasc Thorac Surg. 2007;6:628–31.
16. Jacobs JP. Introduction–databases and the assessment of complications associated with the treatment of patients with congenital cardiac disease. Cardiol Young. 2008;18 Suppl 2:1–37.
17. Jacobs JP, Jacobs ML, Mavroudis C, et al. Nomenclature and databases for the surgical treatment of congenital cardiac disease–an updated primer and an analysis of opportunities for improvement. Cardiol Young. 2008;18 Suppl 2:38–62.
18. Chang RK, Klitzner TS. Can regionalization decrease the number of deaths for children who undergo cardiac surgery? A theoretical analysis. Pediatrics. 2002; 109:173–81.

19. Bazzani LG, Marcin JP. Case volume and mortality in pediatric cardiac surgery patients in California, 1998–2003. Circulation. 2007;115:2652–9.
20. Welke KF, Diggs BS, Karamlou T, Ungerleider RM. The relationship between hospital surgical case volumes and mortality rates in pediatric cardiac surgery: a national sample, 1988–2005. Ann Thorac Surg. 2008;86:889–96; discussion -96.
21. Pasquali SK, Jacobs JP, Shook GJ, et al. Linking clinical registry data with administrative data using indirect identifiers: implementation and validation in the congenital heart surgery population. Am Heart J. 2010;160:1099–104.
22. O'Connor GT, Plume SK, Olmstead EM, et al. A regional intervention to improve the hospital mortality associated with coronary artery bypass graft surgery. The Northern New England Cardiovascular Disease Study Group. JAMA. 1996;275:841–6.
23. Share DA, Campbell DA, Birkmeyer N, et al. How a regional collaborative of hospitals and physicians in Michigan cut costs and improved the quality of care. Health Aff (Millwood). 2011;30:636–45.

Early Database Initiatives: The Fyler Codes

12

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Abstract

The escalating movement in the health care industry towards electronic medical recordkeeping has been substantially impeded by the lack of clinical data standards. The classification systems that are in common use, such as the ICD-9-CM, rely on broad disease categories with insufficient granularity to permit valid analysis of outcomes. This shortcoming is particularly evident in the field of congenital heart disease, where the highly heterogeneous congenital malformations are grouped into just 39 categories in the ICD-9 nomenclature. As described in other sections of this book, for over a decade there has been a concerted effort by various consortia of congenital cardiologists and cardiac surgeons to rectify this situation. When undertaking an effort of this magnitude it is useful to examine the successes and failures of existing nomenclatures in order to improve on history rather than to simply repeat it. In this regard, the Fyler Coding System (FCS) is one of the earliest (if not ***the*** earliest) experience with introduction of a comprehensive congenital heart disease classification into the routine delivery of cardiac medical and surgical care and 45 years of continuous use are available for analysis of successes and failures. This chapter reviews the history of the FCS with particular emphasis on recognizing the lessons that should be learned by those who seek to implement something better.

Keywords

Taxonomy • Nomenclature • Classification • Database • Congenital heart disease

Introduction

In 1968, Donald C. Fyler, through the Department of Cardiology at Boston Children's Hospital, organized The New England Regional Infant

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Cardiac Program (NERICP), a clinical care program that was designed to enhance the delivery of care to infants born in the New England area with life threatening congenital heart disease [1]. In addition to establishing a cooperative referral network, an important element of this program was the creation of a centralized regional registry of incident cases of congenital heart disease. This database provided epidemiologic and descriptive data concerning the frequency of various forms of heart disease [2], and original insights into factors associated with the presence of congenital heart disease such as birth order, population density, and season [3, 4]. Key to this effort was a system of disease categorization. A frequently quoted but still germane observation by William Farr in 1839 while working as an epidemiologist for the city of London is that "nomenclature is of as much importance in this department of inquiry, as weights and measures in the physical sciences, and should be settled without delay" [5]. Dr. Fyler completely understood the need for a systematized nomenclature of congenital heart disease for the NERICP but could not identify an adequate existing resource. He therefore initiated the construction of a coding system to fill this void, a nomenclature that in recognition of his contribution is generally referred to as the Fyler Coding System (FCS). The FCS has succeeded from both the clinical and research perspective and is still the primary system for disease and intervention classification at the Boston Children's Hospital Department of Cardiology and is also used at many other pediatric congenital heart programs both nationally and internationally. However, with regard to the purposes of this book, the shortcomings of the FCS are of more importance than the successes. Dr. Fyler worked to improve the FCS for more than 30 years during the remainder of his career, but remained invariably dissatisfied with his efforts. The reasons for this dissatisfaction can be readily described and are by no means unique in the field of medical nomenclatures. In fact, many new nomenclature efforts continue to make the same mistakes in the design of their systems. This chapter recounts in brief the Fyler experience and the lessons that can be learned from it.

Medical Record Keeping in the Nascent Electronic Era

At the time of initiation of the NERICP, affordable minicomputers were just making their way into the commercial market and from the beginning the NERICP data storage was computer based. The FCS was therefore created to optimize the efficiency of digital storage and retrieval, particularly during the early experience when digital computing was billed based on microseconds of CPU use and digital storage was both slow and expensive. Although the NERICP was initiated during the punch card era and initially relied on this technology, the transition to terminal-based data entry enabled wider access to computer resources. Dr. Fyler responded to this technical advance by quickly introducing this nomenclature into the clinical care arena at Boston Children's Hospital, constituting one of the earliest forays into the realm of electronic medical records. Surgical and catheterization data that preceded the availability of this database were retrospectively entered (back to 1950!) and the diagnoses, findings, and procedures were coded using the Fyler Codes. Prospective data collection was expanded to include outpatient clinic visits, electrocardiograms, and eventually other diagnostic and therapeutic modalities including echocardiography, electrophysiology, cardiac magnetic resonance, and cardiac computed tomography were similarly captured and coded using the same systematized nomenclature system. During the subsequent 45 year period of continuous use and modification of the Fyler Codes in response to clinical use and feedback, much has been learned. To date, over 3.3 million Fyler codes have been entered into clinical echocardiographic reports issued by the Department of Cardiology at Boston Children's Hospital alone.

Numeric Versus Text Codes

The Fyler Codes were originally based on a list of numeric codes, each of which represented a specific textual description. More recently, this

system has been modified to an alphanumeric system, but conceptually the approach is the same insofar as the code represents a specific concept that may be a diagnosis, finding, risk factor, intervention, etc. In retrospect, this was a decision that proved to be an important success of the FCS. The alternative approach, as has been implemented in some coding systems, is to rely on the textual description alone. There are some important advantages to the use of an abstract alphanumeric representation. The FCS stores only the numeric code in the database, resulting in very compact data storage, which was certainly a significant issue in the original design of the FCS whereby the average reduction of data storage requirements was >10 fold. This advantage is considerably less important in the current era of abundant storage capacity and high capacity networks. The primary disadvantage to coding systems based on numeric codes is the need for translation between text and numbers during data storage and retrieval, but again this was primarily an issue when printed manuals were the primary resource for this translation whereas now digital resources can be used to make this process completely transparent, effectively insulating the user from the abstract numeric codes. In contrast, the disadvantages of the text-based codes cannot be as easily overcome. Nomenclature evolves over time and changing the text of a specific code disrupts the validity of the previously stored data if the text is changed, an issue that does not arise as long as the data are stored using an abstract representation. Perhaps the most important advantage to the use of an abstract alphanumeric representation is that it enables synonyms and multilingual translations, such that the same alphanumeric code can be presented to the user as "atrioventricular canal", "atrioventricular septal defect", "canale atrioventricolare completo" or other locally specified designations or languages, thereby greatly facilitating collaborative work. The use of abstract concept representations is widely viewed as essential within the medical informatics community and must be viewed as one of the FCS successes.

Intrinsic Hierarchy Based on Numeric Order

The FCS was predicated on a hierarchy based on severity of disease. Major group categories (for example single ventricle, transposition of the great arteries (TGA), double outlet right ventricle, and tetralogy of Fallot) were assigned a numeric value with an ordered sequence such that those with the greatest clinical and anatomic abnormality were assigned the lowest numbers. The sequence was based primarily on probability of early survival, such that hypoplastic left heart syndrome was one of the lowest number groups whereas atrial septal defect was much higher in the sequence. In patients with multiple lesions, the lowest number code was typically the "primary", or most important diagnosis. Conceptually, this approach enabled very simple assignment of risk hierarchy based on rank order of numeric code in addition to enabling very simple statistical description of population disease severity. This approach proved moderately successful for comparison of complex versus simpler forms of congenital heart disease (such as hypoplastic left heart versus coarctation of the aorta) but fails for comparison within these groups (such as atrial septal defect versus pulmonary stenosis), where the modifying influence of severity of pulmonary stenosis or size of the atrial septal defect is a more important determinant of outcome than the category of that anatomic abnormality. For patients with multiple diagnoses, the numeric sequence of the codes therefore often fails to designate which of the several codes represents the patient's "primary" or most important diagnosis. This shortcoming led Dr. Fyler to ultimately consider the FCS to have been a failure, and is one of the reasons he never chose to publish this system. This experience provided the important lesson that the numeric (or alphanumeric) representation should be abstract and code properties such as severity or relative importance of disease should be maintained as a separate property either of the code or a property determined by the code in conjunction with one or more modifiers.

Code Organization Based on Alphanumeric Representation

Most nomenclatures require an organizational structure for their efficient use. For example, in the FCS diagnoses were grouped according to the nature of the anatomic abnormality, such that all forms of d-loop TGA were positioned numerically between 0700 and 0799 and entities with l-loop TGA were numbered as 0800–0899. From a database and software design perspective this approach provided very efficient search routines and by intrinsically grouping the related codes it facilitated manual code searches by grouping related terms together in printed manuals. This is the approach taken in the International Classification of Disease (ICD) coding system, now in its 10th revision (ICD-10). The ICD-10 has 16 sections in the procedure coding system with all members of a particular section having an alphanumeric code that begins with the same character (http://www.cms.gov/Medicare/Coding/ICD10/Downloads/pcs_final_report2013.pdf). If the groupings are truly mutually exclusive, as is the case with the biological classification into domains, kingdoms, phylums, classes, orders, families, genus, and species, such an approach can be useful and can assist in code identification, hierarchical designation and relationships between categories. However, congenital and acquired heart disease nomenclatures are based on combinations of anatomic and physiologic relationships that do not stratify into mutually exclusive categories. For congenital heart disease individual diseases often consist of "complexes" consisting of multiple anatomic abnormalities, leading to ambiguity as to where in the hierarchy the code should be positioned. For example, a heart with the combination of L-loop TGA, tricuspid atresia, and ventricular septal defect could justifiably be positioned amongst the disorders of the ventriculoarterial junction, ventricles, or atrioventricular junction. Electronic representations can easily accommodate this ambiguity by displaying the same code on multiple branches of the tree, enabling users to find the correct code regardless of which of these categories they prefer to search. However, support for multiple hierarchies requires maintenance by some other method than the alphanumeric code, which must be unique and therefore cannot be used to designate multiple locations in the tree. This is another instance where the design of the FCS failed by attempting to attach meaning to the alphanumeric code beyond simply designating the text it represents. The FCS shares this failure with the ICD and other systems. In general, the network of relationships between concepts and terms must be encoded separately from the encoding of the concepts and terms themselves.

Atomic Versus Molecular Design

As is the case with many disease entities, patients with congenital heart disease manifest one or more specific anatomic or physiologic abnormalities in a variety of combinations. For example, a ventricular septal defect can be observed as an isolated finding but is often found in association with other structural anomalies such as aortic coarctation, and in the case of more complex abnormalities such as double outlet right ventricle the ventricular septal defect is generally integral to the disease. Coding systems have generally taken two different approaches to classifying these hearts. The "atomic" approach to coding identifies each of these lesions as an independent finding and the codes for each component are captured independently. The "molecular" approach involves assigning individual codes to both the individual components and to each of the valid combinations, which constitute the "molecular" codes, also referred to as composite codes. For example, the molecular approach permits the combination of ventricular septal defect and coarctation to be captured as a single code whereas the atomic approach would require each to be entered separately. The Fyler system and the European Pediatric Cardiology Codes (EPCC) [6] have taken a more atomic approach, whereas the Society of Thoracic Surgery (STS) [7] coding system has taken a molecular approach. The Fyler system is not purely "atomic". For example, the FCS has "anatomic" codes for d-loop TGA, ventricular septal defect, and intact

ventricular septum, but also has molecular codes for d-loop TGA with ventricular septal defect and for d-loop TGA with intact ventricular septum. In contrast, the STS system is highly skewed towards the molecular approach by including combination codes such as "Hypoplastic left heart syndrome (HLHS), Without intrinsic valvar stenosis, Hypoplastic aortic valve + mitral valve + left ventricle (Hypoplastic left heart complex = HLHC), VSD, Nonrestrictive VSD". Is there in fact a reason to prefer one approach over the other? There are advantages and disadvantages to each approach and these differences are worth discussing.

Insofar as the structure of the nomenclature should be optimized for the desired functionality, functional specifications can be generally grouped into three categories: (1) maintenance of the coding system, (2) code capture (code finding in conjunction with data entry), and (3) code retrieval (code finding in conjunction with database searches). The first issue (code maintenance) is the simplest to discuss. The atomic approach results in a relatively short list of codes whereas the molecular approach, in which all of the atoms are designated in addition to all valid combinations of the individual atoms, results in a set of codes that may be orders of magnitude larger. In addition, insofar as the molecular system attempts to represent the universe of possible combinations, maintaining a complete "molecular" system is far more subject to error. In general, the cost and complexity of code system maintenance increase in proportion to the number of terms in the system, and therefore an atomic system is far more economical to maintain.

Code finding in conjunction with data entry involves searching a repository of codes and selecting those appropriate to a particular patient or patient-related event. The shorter list of codes in an atomic system makes it easier to locate individual codes at the time of code entry, but searches for specific complex cases with multiple anatomic abnormalities requires repeated searches for each component. The primary advantage to the molecular code schema is that a much smaller set of codes needs to be selected, but it is often necessary to search through a very long list of codes to find the best match. Even with a feature-rich search facility this can be very time consuming and the net tradeoff between these costs and benefits is unclear. There is a theoretical advantage to the molecular approach because the universe of valid code combinations is pre-specified. Because the user of an atomic system has the freedom to choose any combination of terms, invalid combinations (such as mitral regurgitation plus mitral atresia) can be selected for the same patient. However, because the molecular system must by necessity include all the individual atoms, such an error can be made in this system as well.

Code finding in conjunction with database searches has a different trade-off. Since the same atomic entry will appear in many molecular terms, searches for a specific atom (such as ventricular septal defect) are more complex because of the need to specify a large number of molecular entries to capture all instances of that atom. Searches for composite diagnoses are also more complex in a molecular system since they may have been coded using the composite code ("TGA with ventricular septal defect") or the individual components may have been selected separately ("TGA" and "ventricular septal defect" as atomic codes). Again, the consensus in the health informatics community is that concepts should be represented as their constituent components as is done in an atomic system.

Provisions for Expansion and Modification of the Coding System

The pace of change in medicine is fairly remarkable and as new knowledge is gained the need for new terminology and codes is relentless. Many of the standard coding systems, such as the ICD system, operate on revisions that are issued in cycles measured over years, which is completely inadequate from a clinical perspective. New procedures, for example, need to be captured in near-real time because back-coding is a notorious source of data loss. From the beginning, the FCS was, and remains, a dynamic system with

expansion of the codes to accommodate missing elements, newly recognized diseases, and most importantly the continuous evolution of new therapeutic options. When the first arterial switch operation was performed at Boston Children's Hospital, the appropriate code was added the next day. This responsiveness considerably enhances the value of the coding system, but also creates a higher risk of incorporating terminology that is rapidly outdated or duplicates existing codes (for example separate codes may be created for "LEOPARD syndrome" and "Noonan syndrome with multiple lentigines" because of a failure to recognize these as synonyms). Management of the coding system therefore requires a system of governance, generally by individuals with considerable interest in maintaining the integrity of the system, who have sufficient knowledge and are willing to put in the effort to confirm accuracy and need for new entries.

Coding Data Capture Workflow

Early on in the FCS experience it became apparent that disease classification was most accurately performed by the physician at the time of care delivery rather than later translation of free text reports based on chart review, a function that is often performed by personnel not directly involved in care delivery. The cardiology information system at Boston Children's hospital was therefore constructed with this goal in mind, implementing capture of these data as a function of the clinical workflow. It was relatively straightforward to introduce this model in the diagnostic testing environment, as the physicians performing and interpreting the echocardiograms, cardiac catheterizations, cardiac magnetic resonance imaging, exercise stress testing, and electrocardiograms entered these codes as part of the electronic documentation process. The coding process was used to facilitate the reporting process because the Fyler code translations were included in the final reports, thereby supporting clinical care delivery as well as billing requirements. Positioning this process as a critical step in care delivery ensures a higher degree of accuracy than relegating code capture to an administrative role where mistakes and inaccuracies have less opportunity for critical review and error recognition by the entire care delivery team. Capturing these data in other environments such as the inpatient service and intensive care unit has been more challenging and this remains an area where development of better systems for information capture remains an important goal.

Lessons Learned

A significant effort has been devoted toward the theoretical specification of the features of a terminology system that will best accomplish the multiple purposes that such a system is intended to accomplish. A position paper from the American National Standards Institute (ANSI) regarding informatics standards for health terminology [8] documented that many of the same problems we experienced with the FCS are present in most of the other health care terminology systems (such as the ICD and CPT systems). These other systems also tend to be constructed as a list of terms with numeric designations that are intended to have meaning beyond an arbitrary alphanumeric code, such as group membership or hierarchical relationships, and are rarely atomic. The ANSI recommendations are in alignment with overcoming the shortcomings we have experienced with the original design of the Fyler codes, such as the need for context-free identifiers and support for synonyms, properties (also known as attributes), and multiple hierarchies. The ANSI report also documented the need for mapping between systems, language independence, and other features that are time consuming and expensive to implement but are ultimately vital to the utility of these classification systems for robust data analysis.

The ANSI working group identified explicit definitions as one of the primary requirements

for a health system terminology, noting that the definitions are inconsistent in medical dictionaries and are subject to even greater variance between practitioners. This is unquestionably one of the most glaring shortcomings of the various lexicons that are currently in use for congenital heart disease, including the Fyler system. The controversies concerning the proper use and understanding of the congenital heart disease lexicon has been well documented for many years. The work in progress by the International Working Group for Defining the Diagnostic and Procedural Terms for Pediatric and Congenital Heart Disease, a subcommittee of the International Society for Nomenclature of Pediatric and Congenital Heart Disease (ISNPCHD), to correct this situation was presented in February, 2013 at The Sixth World Congress of Pediatric Cardiology and Cardiac Surgery in Cape Town, South Africa. The goal of this group is to create concise definitions for the diagnostic and procedural terms in the International Pediatric and Congenital Cardiac Code (IPCCC) and the committee has been working towards this goal since its formation in 2007. The FCS will benefit from this work, through a process of cross-mapping and transparent incorporation of these definitions.

References

1. Fyler DC, Parisi L, Berman MA. The regionalization of infant cardiac care in New England. Cardiovasc Clin. 1972;4:339–56.
2. Chameides L, Galioto F, Fyler DC. The regional infant cardiac program. A six year evaluation. Conn Med. 1975;39:707–10.
3. Rothman KJ, Fyler DC. Association of congenital heart defects with season and population density. Teratology. 1976;13:29–34.
4. Rothman KJ, Fyler DC. Sex, birth order, and maternal age characteristics of infants with congenital heart defects. Am J Epidemiol. 1976;104:527–34.
5. Farr W. Annual report of the registrar-general of the births deaths, and marriages in England. London; 1839. p. 99.
6. Franklin RC. The European paediatric cardiac code long list: structure and function. Cardiol Young. 2000;10 Suppl 1:27–146.
7. Mavroudis C, Jacobs JP. Congenital Heart Surgery Nomenclature and Database Project: Overview and Minimum Dataset. In: Mavroudis C, Jacobs JP, editors. The Annals of Thoracic Surgery April 2000 Supplement: The International Congenital Heart Surgery Nomenclature and Database Project. The Annals of Thoracic Surgery, April 2000, Supplement, S2–17.
8. Chute CG, Cohn SP, Campbell JR. A framework for comprehensive health terminology systems in the United States: development guidelines, criteria for selection, and public policy implications. ANSI healthcare informatics standards board vocabulary working group and the computer-based patient records institute working group on codes and structures. J Am Med Inform Assoc. 1998;5:503–10.

The Academic Database: Lessons Learned from the Congenital Heart Surgeons' Society Data Center

13

Christopher A. Caldarone, Jeffrey A. Poynter, and William G. Williams

Abstract

During its more than 40 year history, The Congenital Heart Surgeons Society (CHSS) has evolved from an informal club to a mature organization. In 1985, Drs. John W. Kirklin and Eugene H. Blackstone founded the CHSS Data Center. Its purpose was to develop disease-specific inception cohorts of congenital heart disease (CHD) patients and extract knowledge from the combined clinical experience of centers across North America. The mission has evolved to training of research fellows, prospective testing of patients in our lifelong cohorts, organization of a tissue bank registry, and provision of quality improvement tools for members. The hub of this activity is in the CHSS Data Center, housed within the Hospital for Sick Children in Toronto. Our review will highlight lessons learned during the course of this evolution.

Keywords

Statistics • Database structure • Clinical trials • Outcomes research • Survival analysis • Registry • Quality improvement

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Background

The rationale for establishing the Congenital Heart Surgeons Society (CHSS) Data Center was the recognition among congenital heart surgeons that pooling clinical information in an organized fashion to facilitate data analysis would help improve patient outcomes. The rarity of congenital heart disease (CHD), the wide spectrum of anatomic and physiologic variations in presentation and the extensive array of available medical and surgical management strategies contribute to the difficulties faced by any one surgeon (or institution) in determining the optimal management for a given lesion. Acknowledging these fundamental difficulties, the CHSS embarked upon a collaborative venture in 1985 to share experiences and analyze aggregate data to improve the CHD management.

The first cohort assembled by the CHSS between 1985 and 1989 enrolled patients with transposition of the great arteries. During the first 4 years of enrollment, 985 neonates admitted to a CHSS institution within the first 2 weeks of life were enrolled. There were few concerns with institutional review boards and obtaining patient/family consent in this era. The robust enrollment of patients in this cohort was fueled by an urgent desire to rapidly develop a knowledge base on which to compare more traditional atrial switch strategies with the newer arterial switch strategy. Thus, the CHSS rapidly established itself as an organization that could address contemporary clinical problems in direct response to the academic needs of the membership.

The success of this cohort was followed by the conception of 11 other cohorts with over 5,400 patients enrolled for long-term follow-up. The 12 CHSS Study Cohorts are displayed in Table 13.1. Seven of these studies are no longer actively enrolling patients, and five of these studies are still actively enrolling patients. These cohorts have provided the data for numerous analyses and publications on behalf of the CHSS. A list of CHSS publications is available on our website at www.CHSSdc.org.

Table 13.1 Twelve CHSS diagnostic cohorts

Diagnostic cohort	Enrollment	Number of patients enrolled
Transposition of the Great Arteries (TGA) study	1985–1989	891
Interrupted Aortic Arch (IAA) study	1987–1997	470
Coarctation study	1990–1993	883
Pulmonary Atresia Intact Ventricular Septum (PAIVS) study	1987–1997	444
Pulmonary Stenosis with Intact Ventricular Septum (PSIVS) study	1987–1997	187
Critical aortic stenosis study	1987–1997	422
Aortic valve atresia study	1987–1997	563
Tricuspid Atresia (TA) study	1999–2013	307
Pulmonary Conduit (PC) study	2002–2013	591
Critical Left Ventricular Outflow Tract (LVOTO) study	2005–2013	674
Anomalous Aortic Origin of a Coronary Artery (AAOCA) study	1998–2013	284
Unbalanced atrioventricular septal defect (uAVSD) study	2012–2013	84

CHSS Data Center Structure

Personnel

The CHSS Data Center employs a Research Program Manager, a Database Programmer with statistical expertise, two Clinical Research Project Assistants, and two data abstraction nurses who have extensive clinical experience with CHD. In 2001, a Research Fellowship was created (the Kirklin/Ashburn Fellowship, discussed below). The Data Center is housed within the Hospital for Sick Children in Toronto with two suites including 1,200 sq. ft. of office space with all required computers and information technology resources, as well as secure storage for all electronic and hard copy data. The active interchange of information and ideas among the Data Center staff (i.e., teamwork) is

essential for continuous improvement in data management practice.

Legal/Ethical Issues

The Data Center seeks Research Ethics Board (REB) approval on an annual basis to insure that general operations of Data Center comply with the Health Insurance Portability and Accountability Act of 1996 of the United States of America (HIPAA), and other laws and regulations regarding patient confidentiality and data security. In addition, each participating CHSS institution requires their institutional REB approval for every cohort being followed. Direct patient consent for yearly follow up by Data Center staff is also obtained. Direct patient consent facilitates communication with patients who relocated to new caregivers and hospitals. Sharing of patient data also requires a Data Use Agreement between the Data Center and each CHSS institution.

Communication

The Data Center provides bi-monthly Newsletters to the CHSS members and their data managers. We also maintain a Website (www.CHSSdc.org) to provide members, our patients and the general public with current activities, publication access, lay summaries of publications, a patient blog, and links to relevant Websites. Extensive use is made of emails to members and a web-based dropbox for secure data transfer. The Data Center website also posts inclusion/exclusion criteria for each cohort and templates of REB applications for each institution to use in their institutional REB application. The availability of templates avoids duplication of effort and facilitates institutional enrollment in CHSS studies

Work Weekends

The Data Center organizes a semi-annual 3-day weekend for interested members to work in the Data Center. The members 'brain-storm' to develop new cohorts, direct statistical analyses, construct abstracts and manuscripts, and refine presentations.

Finances

Each CHSS institution is required to support the Data Center with an annual contribution. Support is mandatory. Additional funds are sought from peer reviewed grant applications, industry partners and philanthropic individuals and institutions.

Voluntary Contribution of Data

An important lesson learned in the CHSS Data Center is that our reliance upon voluntary enrollment of data creates the potential for failure to include all eligible patients in a cohort and introduction of selection bias into our cohorts. Parameters that influence enrollment are not well-studied but are likely to include the clinical 'urgency' associated with the research question that was the rationale for inception of the cohort. For example, as noted above, the cohort of patients with transposition of the great arteries acquired patients with extreme velocity. (985 neonates were enrolled from 100 % of all CHSS institutions (24 at that time) within 4 years). In contrast, a recent inception cohort of patients with critical left ventricular outflow tract obstruction (LVOTO) has enrolled relatively slowly (718 patients from 16 institutions over 4 years). The LVOTO enrollment can be compared to the contemporaneous National Institutes of Health (NIH) funded Pediatric Health Network Single Ventricle Reconstruction (SVR) Trial, which had a more narrow diagnostic range of entry criteria and fewer participating institutions but enrolled at a far greater rate than the LVOTO cohort. It is likely that the presence of paid coordinators 'on the ground' in each institution with scrutinized enrollment rates, and potential for financial penalties for failure to enroll contributed to the far more complete enrollment in the funded SVR trial when compared to the voluntary CHSS cohort.

Centralized Abstraction of Data

The CHSS relies upon centralized data abstraction. This lesson was learned after an unsuccessful

attempt to develop a web-based data entry system for patients enrolled in our pulmonary conduit cohort. Our expectation was that surgeons or their delegates would enter the data and this would improve overall efficiency of the data collection process. In fact, the absence of training in data entry led to a high proportion of records with incomplete data entry, a high frequency of errors, and a lack of means to redress these deficiencies. Furthermore, addition of new data fields to the research data set was difficult because there was no easy mechanism to recall the person entering data at the first setting and induce them to find an old record, abstract the required new data points, and enter the data in the web-based system. To redress these problems, the Data Center currently uses centralized data abstraction where paper and electronic medical records for each patient are collected and stored within the Data Center. Data are abstracted by specially trained personnel who are conversant with the data entry forms and have a vested interest in the accuracy of entered data.

Because hard copies of the patient records are available for review, a 'follow-on' unplanned analysis can be undertaken whenever needed. For example, a detailed analysis of the aortic valve stenosis/aortic valve atresia cohort enabled a complex analysis of the role of the Ross-Konno and Yasui procedures many years after inception of the cohort [1]. For this subset of patients, unique data fields were required and the analysis would have been impossible if the raw data were not available in the Data Center.

The Kirklin/Ashburn Fellowship

The John W. Kirklin/David A. Ashburn Fellowship is a central component of the CHSS research model and represents an important 'lesson learned' in the CHSS. Employing a dedicated Fellow to undertake complex statistical analyses transformed the research activity in the CHSS Data Center from an intermittent effort predicated on part time efforts of CHSS members to a continuous effort led by the Kirklin/Ashburn Fellow – with a fundamental transformation in the productivity of the Data Center.

In exchange for the high level of productive work performed, the Kirklin/Ashburn Fellow enjoys many academic benefits. The Kirklin/Ashburn Fellows typically have studied in the Data Center for 2 years and have enrolled in concurrent Masters or PhD programs at the University of Toronto. Their tenure in the Data Center has been supported by intensive tutelage from Drs. Eugene Blackstone and Brian McCrindle, and Sally Cai. Using this support network, the Fellows have forged new analyses of CHSS cohorts using state of the art statistical techniques. The Fellows have led the analysis through collaboration with participating members from the inception of addressable questions, 'cleaning' of the data, development of an analysis plan, correspondence with working groups, creation of presentations, and writing of manuscripts. All these activities have been supported by the Data Center staff in Toronto to provide the Fellows with mentorship, and help to focus their analyses and fine-tune interpretation of results. The Fellowship is highly visible amongst congenital heart surgeons and has allowed the intellectual firepower of future congenital heart surgeons to shine among the membership where prospects for future employment are bright. Building a training program into the structure of the Data Center has promoted academic output and helped to keep the CHSS Data Center as a hub of activity within the CHSS.

Research Strategies in the CHSS Data Center

The Research Question

Clinical research should be driven by a research question or questions. The question(s) define the dataset; therefore, any proposed analysis begins with the identification of one or more specific research questions. This process typically requires several hours of thoughtful discussion. One must collect the data points that will address the proposed research question(s), including the specific information on outcomes to be determined. A common error in the early years of the

Data Center was to try to collect too much information including information which did not contribute to the central research questions.

Focus on Diagnosis-Based Inception Cohorts

During the years from 1985 to 2003, the CHSS focused on diagnosis-based cohorts over procedure-based cohorts. The rationale was to capture the wide variety of potential operative and non-operative management strategies utilized across institutions. This approach allowed evaluation of important patient subsets that are typically excluded from procedure-based surgical reports. For example, inclusion of non-operated patients who die prior to operation is an important tool to compare management strategies across institutions. Using the all-comers approach, the CHSS endeavors to avoid the filtering of patients that is often a foundation of published procedure-based reports. One institution may exclude certain patient subsets from consideration for surgical therapy whereas another institution may choose to provide therapy – making comparison of published reports from different institutions problematic. An example of the importance of inclusion of all patients is demonstrated in an analysis of the CHSS pulmonary atresia cohort [2]. In Fig. 13.1, the transition from entry in the study (diagnosis) to a definitive single-, 1.5-, or two-ventricle repair is shown. Note that a large proportion of the patients never achieved one of these 'endstates'. Consequently, a procedure-based surgical report might have neglected to account for the substantial proportion of patients who died without undergoing a 'definitive' procedure. The inclusion of non-operative patients also allows comparison between institutions by using statistical methods to control for differences in patient selection.

Data Entry

The Data Center constructs a database to record all data required to address the research question.

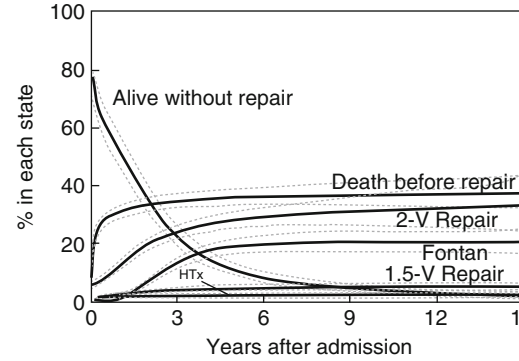


Fig. 13.1 Non–risk-adjusted competing-risks depiction of end states in 408 neonates with PAIVS illustrating the proportion of children reaching each end state over time after initial hospital admission. All patients begin alive at the time of initial admission (time = 0) and migrate to an end state at a time-dependent rate defined by the hazard functions. At 5 years, the estimated prevalences of end states are as follows: 2-ventricle repair, 28 %; Fontan operation, 19 %; 1.5-ventricle repair, 5 %; cardiac transplantation, 2 %; death before reaching a repair state, 36 %; and alive without end state, 11 % (Reprinted with permission from Ashburn et al. [2])

In the past we have collected hard copies of specified parts of the patient's hospital chart, such as admission sheet, admission history and physical, all diagnostic reports, operative reports and follow-up investigation and reports. These data are extracted by highly knowledgeable and experienced professionals. We are making a transition to collect these data via Internet e-based records using secure file transfer. Annual cross-sectional follow-up data is conducted within specific months of each year and the information, including interval procedures and/or investigations added to the dataset.

Data Integrity (The Essential Underappreciated Integral Step)

Prior to beginning any data analysis it is essential to check the dataset for errors, omissions, outliers, unknown data points, and any possible misinterpretation of data extraction. Even the most committed and compulsive professional cannot be perfect extracting data. More commonly the information from clinical records will contain

typographic errors, omissions, and outliers. We cannot overstress the essential nature of this very labor intensive requiring considerable time, effort and ingenuity to make the dataset as accurate and complete as possible.

Diagnostic Images

Institutional reports of diagnostic images vary in consistency. We have found that obtaining copies of the actual echo, computed tomography (CT) or magnetic resonance imaging (MRI) studies is relatively easy. And it has been amazing to us to see the enthusiasm of expert reviewers in undertaking detailed review of each image. The quality and detail of the diagnostic data is enormously enhanced by expert review. To do the reviews, the reviewers needed to work in the Data Center for many days at their own expense. As a further step to facilitate this expert review process we have setup a CHSS Core Lab to upload de-identified images to our central server so that the experts can review each image from the convenience of their home institution. In addition, the data from each expert review is entered into an Internet-based database (RedCap) [3] housed within the Data Center file server. The merged data is then available for analysis.

Evaluation of Uncommon Lesions

The CHSS Data Center is well suited to examine uncommon lesions because of the large number of participating institutions. For example, the CHSS assembled a large cohort of patients with interrupted aortic arch representing a relatively rare congenital heart lesion. Interestingly, among the 472 patients with interrupted aortic arch, concomitant aortopulmonary window was identified in 20 patients. Because of the large number of centers enrolling patients, the analysis spawned an important sub-analysis of patients with an extremely rare combination of interrupted aortic arch and aortopulmonary window – a feat that could not be accomplished in a single center cohort (Fig. 13.2).

The CHSS Data Center has leveraged its multi-institutional resources to develop a prospective inception cohort of patients with anomalous aortic origin of a coronary artery (AAOCA). AAOCA is diagnosed when one or both coronary arteries arise from outside their appropriate sinus of Valsalva (Fig. 13.3). Although relatively rare, the diagnosis of AAOCA provokes intense anxiety among patients (and clinicians) because there are no clearly defined management algorithms and the potential for sudden death is not well understood. Currently (as of August 2013),

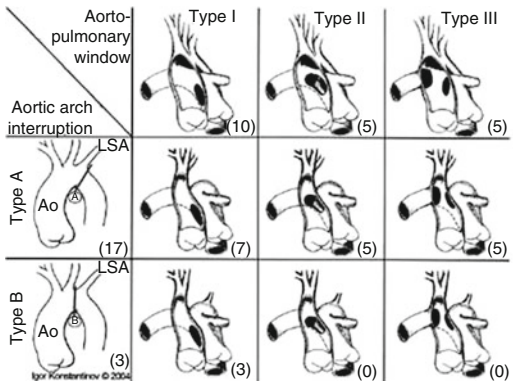
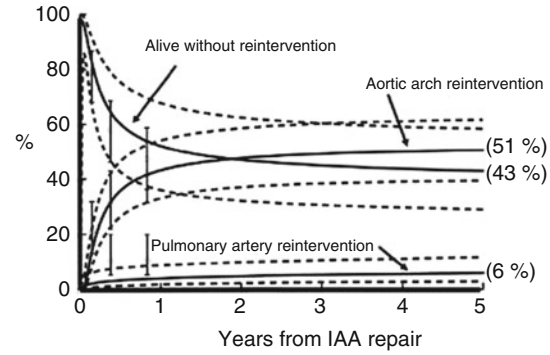


Fig. 13.2 Rare combinations of lesions can be examined within larger CHSS cohort. Konstantinov et al. examined outcomes in patients with interrupted aortic arch and concomitant aortopulmonary window and were able to make important inferences to assist in clinical decision making.



This would be difficult or impossible for any single institution to perform due to the rare prevalence of this combination of lesions (Reprinted with permission from Konstantinov et al. [9])

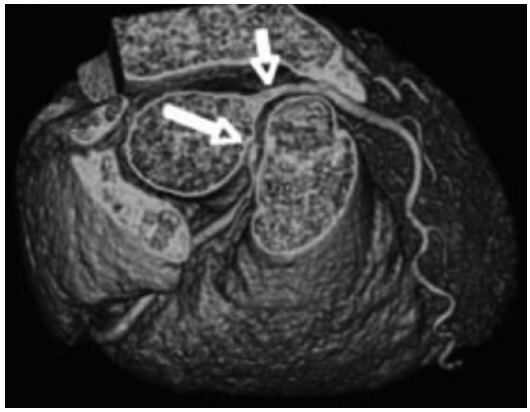


Fig. 13.3 Anomalous aortic origin of the right coronary artery (*large arrow*) from the left coronary sinus (*small arrow* identifies left main coronary artery) [10]

the Data Center has enrolled 249 patients and continues to enroll. This rapid accrual of a large cohort of patients with a relatively rare lesion will allow unprecedented analysis of the relationships between symptomatology, preoperative diagnostic data, surgical findings, operative and non-operative treatment strategies, and long term outcomes.

Data Analysis

Complex Cohorts Require Complex Statistical Techniques

An important lesson learned in the CHSS Data Center has resulted from the use of complex statistical techniques to evaluate the complex management strategies utilized in our multi-institutional patient cohort. This strategic focus distinguishes the CHSS from more traditional large-scale research ventures that tend to focus on straightforward comparisons using prospective randomized trial designs. Although randomized trials can facilitate direct evaluation of very specific hypotheses, randomized designs are less appropriate when a wide range of management options exist and/or the condition to be studied is rare. Under such circumstances, it is frequently not feasible to incorporate multiple management options into a randomized controlled trial design.

Evolution of Statistical Techniques

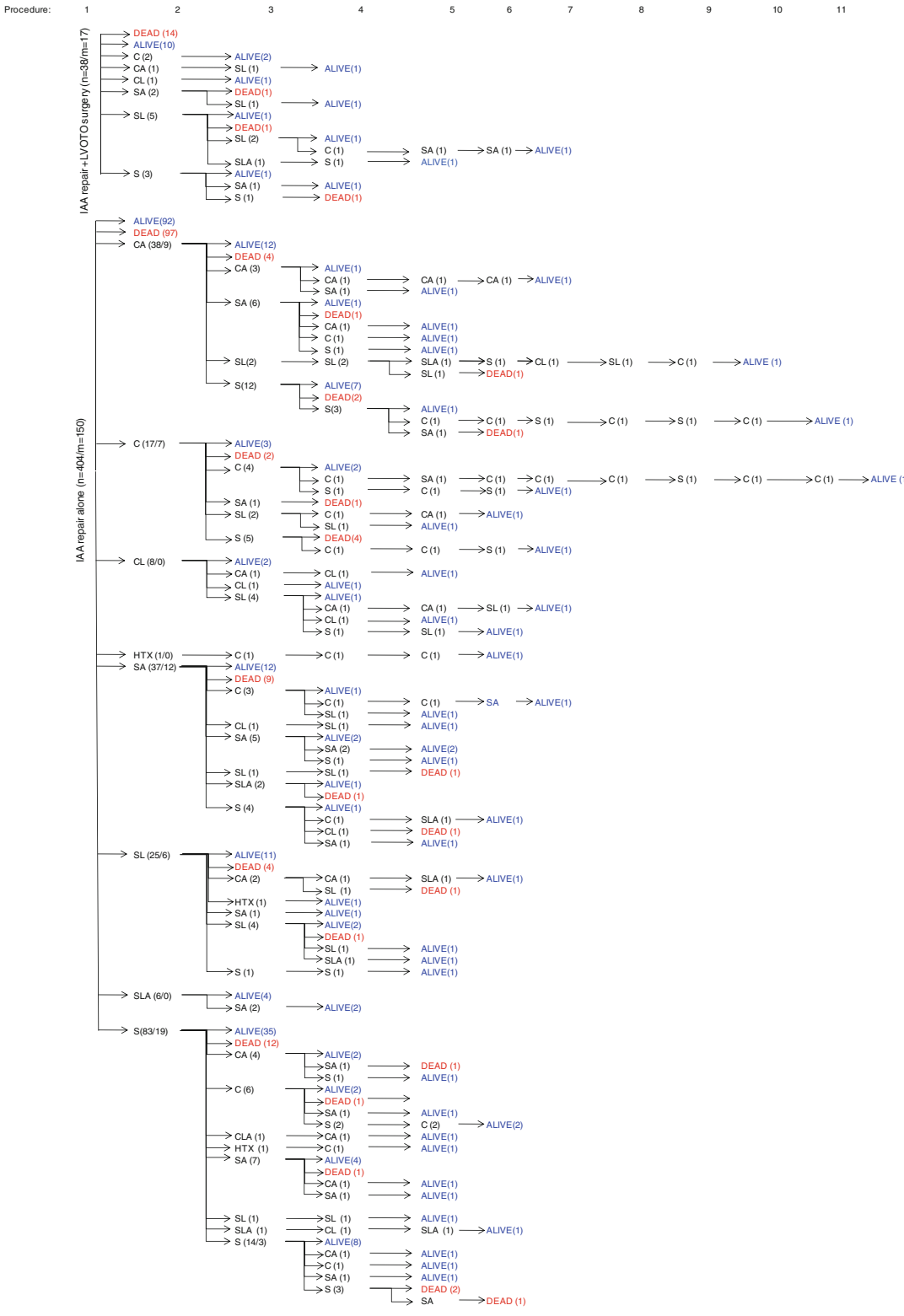
The data abstracted in the Data Center is derived from highly complex cohorts and the complexity in our cohorts is a direct result of the interaction between a wide array of management options in common clinical practice, the high degree of variability in the timing of these interventions between institutions, and the relatively high probability of multiple reinterventions and diagnostic procedures. An example of a complex cohort is demonstrated in Fig. 13.4. In order to deal with this complexity, the Data Center has evolved to utilize a wide array of advanced statistical techniques to facilitate analysis of extremely complex cohorts. The statistical evolution of the CHSS Data Center has been led by Drs. Eugene Blackstone and Brian McCrindle, Sally Cai, and the Kirklin/Ashburn Fellows.

Statistical Techniques in Use at the CHSS Data Center

Parametric Hazard Phase Decomposition

CHSS studies incorporating parametric hazard analysis nearly always incorporate a method pioneered by Eugene Blackstone and colleagues to decompose the overall time-related hazard (Fig. 13.5a) into as many as three 'phases' [4] (Fig. 13.5b). This method is well-suited to model the hazard of various outcomes surgical patients because it can account for transient but high 'early phase' of postoperative risk, a period of attrition at a constant rate (the 'constant phase'), and a 'late phase' of increasing risk. This method permits the identification of risk factors unique to each phase. Risk factors which modify one phase may not always be incorporated into the model describing another phase. For example, factors which are associated with early postoperative risk of death may quite different than factors associated with death in the late postoperative period.

The decomposition of hazard into phases is a fundamental strategy and serves as the statistical method of choice for almost all of our analyses. Adoption of this method has permitted accurate


Procedure: 1 2 3 4 5 6 7 8 9 10 11
IAA repair+LVOTO surgery (n=38/m=17)
DEAD (14)
ALIVE(10)
C (2) → ALIVE(2)
CA (1) → SL (1) → ALIVE(1)
CL (1) → ALIVE(1)
SA (2) → DEAD(1)
SL (1) → ALIVE(1)
SL (5) → ALIVE(1)
DEAD(1)
SL (2) → ALIVE(1)
C (1) → SA (1) → SA (1) → ALIVE(1)
SLA (1) → S (1) → ALIVE(1)
S (3) → ALIVE(1)
SA (1) → ALIVE(1)
S (1) → DEAD(1)
IAA repair alone (n=404/m=150)
ALIVE(92)
DEAD (97)
CA (38/9) → ALIVE(12)
DEAD (4)
CA (3) → ALIVE(1)
CA (1) → CA (1) → ALIVE(1)
SA (1) → ALIVE(1)
SA (6) → ALIVE(1)
DEAD(1)
CA (1) → ALIVE(1)
C (1) → ALIVE(1)
S (1) → ALIVE(1)
SL(2) → SL (2) → SLA (1) → S (1) → CL (1) → SL (1) → C (1) → ALIVE (1)
SL (1) → DEAD(1)
S(12) → ALIVE(7)
DEAD(2)
S(3) → ALIVE(1)
C (1) → C (1) → S (1) → C (1) → S (1) → C (1) → ALIVE (1)
SA (1) → DEAD(1)
C (17/7) → ALIVE(3)
DEAD (2)
C (4) → ALIVE(2)
C (1) → SA (1) → C (1) → C (1) → C (1) → S (1) → C (1) → C (1) → ALIVE (1)
S (1) → C (1) → S (1) → ALIVE(1)
SA (1) → DEAD(1)
SL (2) → C (1) → CA (1) → ALIVE(1)
SL (1) → ALIVE(1)
S (5) → DEAD(4)
C (1) → C (1) → S (1) → ALIVE(1)
CL (8/0) → ALIVE(2)
CA (1) → CL (1) → ALIVE(1)
CL (1) → ALIVE(1)
SL (4) → ALIVE(1)
CA (1) → CA (1) → SL (1) → ALIVE(1)
CL (1) → ALIVE(1)
S (1) → SL (1) → ALIVE(1)
HTX (1/0) → C (1) → C (1) → C (1) → ALIVE(1)
SA (37/12) → ALIVE(12)
DEAD (9)
C (3) → ALIVE(1)
C (1) → C (1) → SA → ALIVE(1)
SL (1) → ALIVE(1)
CL (1) → SL (1) → ALIVE(1)
SA (5) → ALIVE(2)
SA (2) → ALIVE(2)
S (1) → ALIVE(1)
SL (1) → SL (1) → DEAD (1)
SLA (2) → ALIVE(1)
DEAD (1)
S (4) → ALIVE(1)
C (1) → SLA (1) → ALIVE(1)
CL (1) → DEAD (1)
SA (1) → ALIVE(1)
SL (25/6) → ALIVE(11)
DEAD (4)
CA (2) → CA (1) → SLA (1) → ALIVE(1)
SL (1) → DEAD (1)
HTX (1) → ALIVE(1)
SA (1) → ALIVE(1)
SL (4) → ALIVE(2)
DEAD (1)
SL (1) → ALIVE(1)
SLA (1) → ALIVE(1)
S (1) → S (1) → ALIVE(1)
SLA (6/0) → ALIVE(4)
SA (2) → ALIVE(2)
S(83/19) → ALIVE(35)
DEAD (12)
CA (4) → ALIVE(2)
SA (1) → DEAD (1)
S (1) → ALIVE(1)
C (6) → ALIVE(2)
DEAD (1)
SA (1) → ALIVE(1)
S (2) → C (2) → ALIVE(2)
CLA (1) → CA (1) → ALIVE(1)
HTX (1) → C (1) → ALIVE(1)
SA (7) → ALIVE(4)
DEAD (1)
CA (1) → ALIVE(1)
SA (1) → ALIVE(1)
SL (1) → SL (1) → ALIVE(1)
SLA (1) → CL (1) → SLA (1) → ALIVE(1)
S (14/3) → ALIVE(8)
CA (1) → ALIVE(1)
C (1) → ALIVE(1)
SA (1) → ALIVE(1)
S (3) → DEAD (2)
SA → DEAD (1)

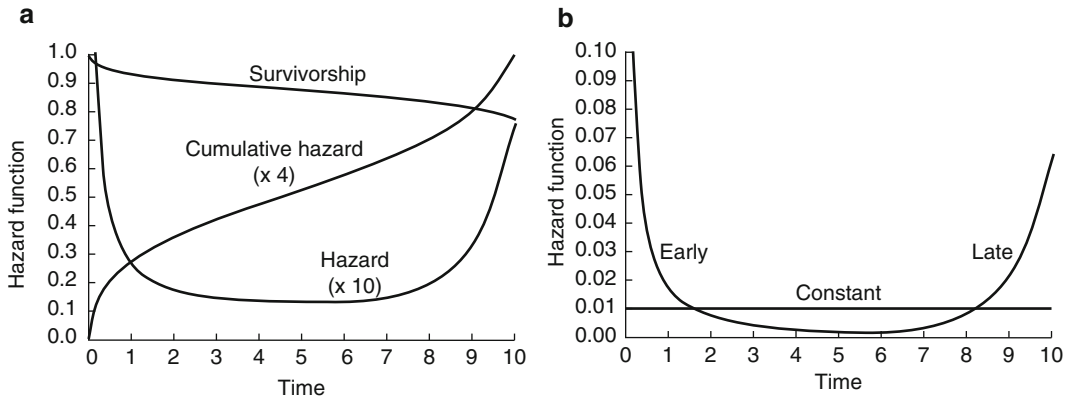


Fig. 13.5 Functions used to describe time-related events. (**a**) The cumulative hazard function accelerates rapidly before reaching a constant slope. Later, it begins to rise rapidly again. This cumulative hazard function corresponds to a survivorship function which decreases rapidly, then stabilizes before accelerating again. The hazard function is infinite at time zero, decreases to a constant rate and then increases late in follow-up. (**b**) The hazard function, decomposed into early, constant and late phases. Each phase quantifies the hazard according to time. The sum of these three phases of hazard is equivalent to the overall hazard function from the upper panel (Reprinted from Blackstone et al. [4])

modeling of every 'shape' of hazard curve yet observed, permitted the identification of risk factors unique to each phase, and facilitated the use of parametric analyses.

Time Zero

Nearly every Data Center study uses survival analysis methods which require that one define a "time zero" as the starting time for the analysis. We define time zero as the time at which the patients becomes at risk of the defined outcome. For example, a study of surgical techniques which evaluate survival after tetralogy of Fallot repair would specify time zero as the initiation of the surgical repair. In contrast, a diagnosis-based study of tetralogy of Fallot would specify time zero as the moment the diagnosis was made – irrespective of a surgical procedure. The latter example would include patients who are not expected to undergo surgery or die prior to planned surgery.

Practical aspects of data abstraction present certain challenges. For example, the date of diagnosis may actually precede birth when the diagnosis is made in utero. Similarly, if the diagnosis is made outside the CHSS member institution, the precise date may be unavailable if the Data Center does not have access to records of that initial diagnostic study (which is often the case). When formulating the precise research question it is best to specify time zero with careful consideration of these practical issues, in order to ensure that all patients in the study can be proven to have become at risk at a precise date (and not the date of their transfer to the CHSS member institution).

Advantages of Parametric Hazard Analysis

A major benefit of parametric hazard analysis over more traditional non-parametric and semi-parametric methods (e.g., Cox regression) is the generation of prediction plots by solving the

Fig. 13.4 A wide variety of management strategies were utilized in the care of patients with Interrupted Aortic Arch. This figure illustrates the complex of management strategies. There is significant crossover amongst treatment pathways, and a large number of unplanned intervening procedures between planned stages of repair. An analysis of outcomes amongst this difficult group of patients necessitates that the statistical methods account for the complexity of management (Reproduced with permission from the Congenital Heart Surgeons' Society Data Center)

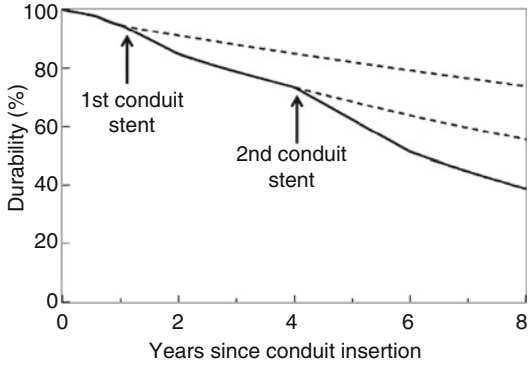


Fig. 13.6 Solution of the parametric equation illustrating the durability of a non-decellularized aortic allograft with z-score +2 inserted in a child with no other risk factors, who underwent two successive conduit stentings at 1 and 4 years after insertion. The *solid line* depicts predicted durability over years since conduit insertion on the x-axis. *Dotted lines* denote predicted durability in a child who did not undergo the first or second conduit stenting procedures. Each stenting is associated with an approximate 18 % reduction in 8-year durability (From Poynter et al. [5], copyright 2013, with permission from Elsevier)

parametric equation to reflect changing values of one or more risk factors included in the model. While Cox regression can support basic plots in which a single covariates may change, the results of Cox regression are generally presented in tables due to the poor compatibility of this method with graphical display of results. Cox regression does not support the creation of prediction plots based upon multiple changing values of covariables. Parametric hazard analysis, on the other hand, can easily support the creation of prediction plots reflecting any clinically feasible combination of values amongst the covariables (so long as they do not exceed the bounds of the original data). As a result, the reader can readily differentiate between a statistically significant result of low (and clinically unimportant) magnitude, and a statistically significant covariable that is associated with clinically important changes in outcome when modulated. The ability to directly compare time-related prediction estimates between multiple significant covariables which are simultaneously changing permits the graphical presentation of as many prediction curves as desired, with any combination of changes amongst any or all of the covariables included in the model. Feedback from our studies incorporating these prediction plots has been very positive because they facilitate visual representation of the impact of alterations in a risk factor. An example is shown in Fig. 13.6. This plot shows the predicted durability of a hypothetical aortic allograft inserted into a child with no other known risk factors for allograft failure and undergoes (or does not undergo) conduit stenting at 1 and 4 years after insertion. The dotted lines indicate the predicted time-related allograft durability if the conduit were not stented at the 1 or 4 year mark. The table within this manuscript [5] listed a parameter estimate of 0.33 and a p value <0.01. These numerical data are not nearly as informative as a plot of time-dependent durability according to whether the conduit was stented 0, 1 or 2 times.

Competing Risks

More traditional mortality studies can often be addressed by consideration of a simple binary outcome – e.g., survival versus death. Children with congenital heart disease, however, are often subject to multiple mutually exclusive 'competing risks' that often include death but may also include cardiac transplantation, re-operation or the achievement of one or more types of 'definitive' repair. Competing risks can be separated to form multiple estimates of time-related hazard of each competing risk [2]. At any point in time the survival estimate of the sum of the various competing risks always equals 100 %. Thus, a survival curve can be generated for each competing endstate and then decomposed into hazard phases to identify risk factors specific to each hazard phase for the respective end-state. Opting not to use competing risks methods requires that the researcher either ignore certain end-states, or combine the competing risks of different end-states into a binary outcome. If this is done, the patients at risk can be inappropriately censored, falsely reducing the denominator. Thus the estimate of risk for the binary outcome may be overestimated if the Kaplan-Meier method is used when there are more than two mutually exclusive possible outcomes [6]. Figure 13.7, from a seminal paper by McGiffin and colleagues, demonstrates the differences in estimates of cardiac transplantation depending on whether death is considered as a separate outcome from transplant (i.e., competing risks) or is not (i.e., the Kaplan-Meier method).

The Data Center began using the competing risks method over a decade ago to evaluate our

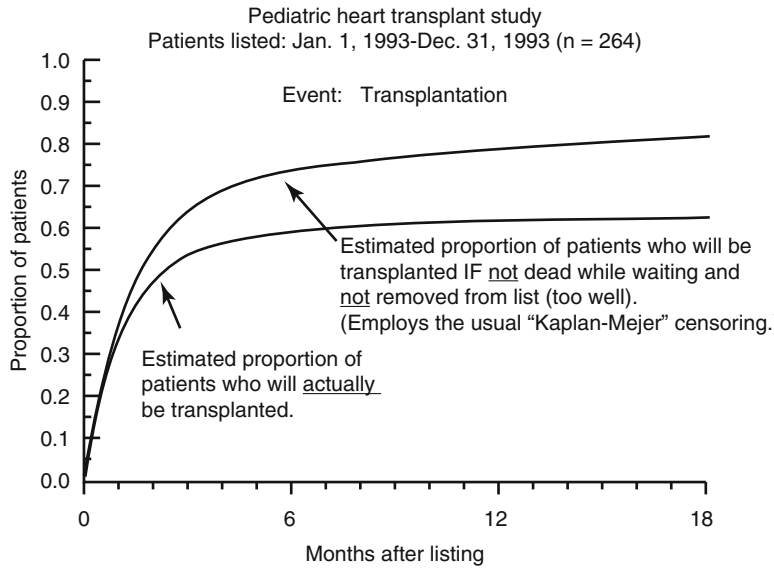


Fig. 13.7 Comparison of the Kaplan-Meier method versus competing risks methodology in analyzing outcomes of patients awaiting heart transplantation. The parametric estimate of proportion of patients who are transplanted by use of Kaplan-Meier right-censoring to remove patients from the denominator at risk and dying while waiting (*upper curve*), and parametric estimate of proportion of patients who will actually undergo transplantation (*lower curve*) are shown. The upper curve overestimates the transplantation probability by removing patients who have died while awaiting transplant. The lower curve is a more accurate estimate of the proportion of patients who will actually be transplanted, because it has considered the competing risk of death separately (From McGiffin et al. [6])

pulmonary atresia with intact ventricular septum cohort [2]. The analysis utilized the mutually exclusive end-points of: survival without repair, death, transplantation, univentricular repair, biventricular repair or 1.5-ventricle repair (Fig. 13.1). Factors associated with each of these unique end-states were identified. Had we performed a more traditional, separate sub-analysis of each of these important groups, important information about those who did not experience each outcome of interest would have been unaccounted for. The lesson here is that utilizing a competing risks analysis is essential when study subjects are at risk of multiple mutually exclusive outcomes (Fig. 13.8).

Segmentation of Longitudinal Records to Facilitate Analysis

A further extension of parametric hazard analysis can also include the incorporation of time-dependent covariables into the record. Using this method, each patient's longitudinal record may be divided into multiple intervals punctuated by interval events (e.g., multiple catheter interventions between surgical procedures) that are expressed as separate observations within the dataset. Each evaluated time interval contains time-independent variables (constant values such as gender) and time-dependent variables (such as the number of cumulative catheter interventions on the repair). Along with those variables, the dataset structure includes a mechanism to 'stitch together' each segment to replicate the longitudinal record as certain values change over time (Fig. 13.9). This technique was used most recently by the Data Center in an analysis by Poynter and colleagues to analyze the durability of right ventricle to pulmonary artery conduits [5].

Propensity Score Matching

The Data Center performs observational inception cohort studies rather than randomized controlled trials. Although observational studies are well suited to the study of congenital heart disease, on occasion we have had a need to balance groups for comparison. We have used balancing scores in order to provide more meaningful comparisons of groups. One type of balancing score called a propensity score has proven to be of

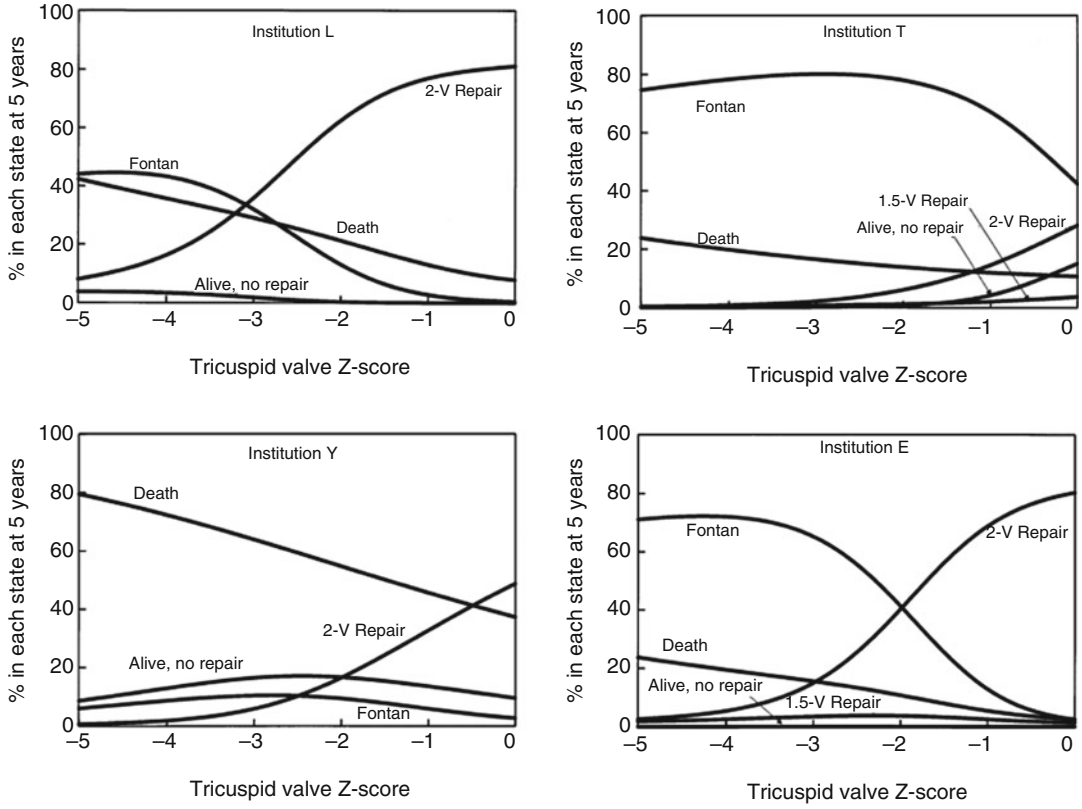
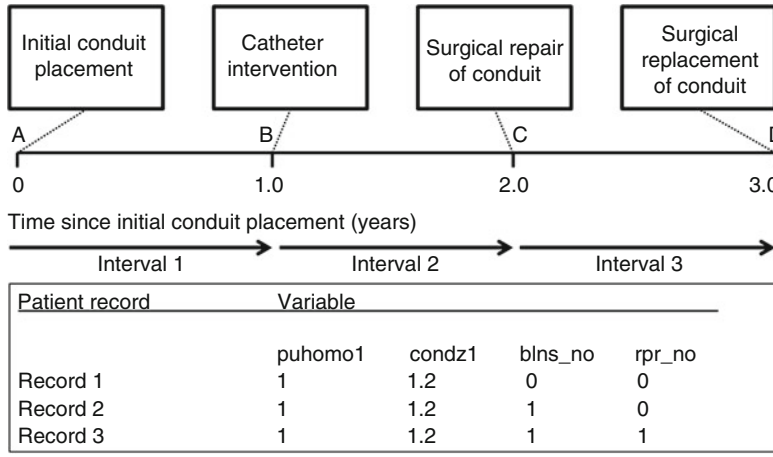


Fig. 13.8 A competing risks analysis is used to examine the relationship between tricuspid valve z-score, inter-institutional management patterns, and a group of mutually exclusive endstates. The z-score was previously found to be a useful surrogate for the size and adequacy of right heart structures in pulmonary atresia with intact ventricular septum. Using this analysis, it is apparent that there are differences in the relationship between tricuspid valve z-score and death between institutions. Importantly, it is also apparent that some institutions (e.g. Institution T) use a Fontan strategy across a wide spectrum of tricuspid valve z-scores with a low death rate, but at the expense of failing to offer two-ventricle palliation to patients with relatively large tricuspid valves (e.g. z-scores between 0 and −2). In contrast, Institution L chose more frequent two-ventricle repairs (and less frequent Fontan strategies) in patients with small tricuspid valves with a relative increase in the death rate in the patients with the smallest tricuspid valves. Finally, Institution E had a balanced strategy with two-ventricle repairs in patients with larger tricuspid valves and Fontan strategies in patients with smaller tricuspid valves – and a corresponding low death rate across the spectrum of tricuspid valve z-scores (Reprinted with permission from Ashburn et al. [2]

particular value. These scores reflect the probability of a given patient to have fallen in one group or the other, based upon various demographic and morphologic characteristics. The scores are included in the parametric hazard analysis to adjust for differences in baseline characteristics among the groups for comparison. This well-established statistical method has proven to be an essential tool to minimize (but not eliminate) an important limitation of our observational studies – i.e., that patients are not randomized into treatment groups.

Modulated Renewal

Management of patients with congenital heart disease often requires post-operative reintervention. Thus the repair is 'renewed,' just as if an old pair of shoes was re-soled. Using the re-soled shoe analogy, we hypothesize that the new sole may be 'better than new', 'as good as new', or 'worse than new'- depending on the durability of the new sole. Similarly, a surgical repair may have any of the same three potential trajectories depending on the durability of the re-intervention. The statistical technique is called



Patient record	Variable			
	puhomo1	condz1	blns_no	rpr_no
Record 1	1	1.2	0	0
Record 2	1	1.2	1	0
Record 3	1	1.2	1	1

Fig. 13.9 Example of "chopping" of a hypothetical child's longitudinal record into multiple discrete segments. Time zero is set to the date of initial pulmonary allograft insertion, which was followed by conduit balloon dilatation with stenting, surgical conduit repair, and finally conduit replacement, at 1-year intervals. Thus, the four events are used to punctuate three segments that are used to model the occurrence of these events. Records 1, 2 and 3 refer to segments AB, BC and CD along the timeline. Record 1 has a left censoring time of A and a right-censoring time of B, record 2 is left censored by B and right censored by C, and so on. The variables puhomo1 and condz1 are constant because they refer to the initial type and z-score of the conduit, respectively. However, the variables blns_no (cumulative number of conduit dilatations with stenting) and rpr_no (cumulative number of surgical conduit repairs) are time-varying; each variable turns from 0 to 1 when the corresponding interval arises. These two variables track the cumulative exposure to these interval treatment events, thus the values are ordinal and do not decrease with time. A proposed method to incorporate many longitudinal echocardiographic measurements of conduit dysfunction – a heretofore unmet challenge in parametric hazard analysis of this type – would involve the expansion of the "events" bracketing each interval by including various echocardiographic measurements from dozens of diagnostic studies performed across the entirety of the patient record (Reproduced from Poynter [11])

modulated renewal. Jegatheeswaran and colleagues used modulated renewal methodology to characterize the risk of re-interventions among children with repaired interrupted aortic arch [7] and the analysis provides insight into the degree to which interrupted aortic arch is a chronic condition that frequently requires multiple subsequent reinterventions.

Scoring Systems Based on Common Dataset Integrated Parametric Models

Another important lesson learned through experience concerns the creation of clinical calculators. Hickey and colleagues developed a parametric equation to facilitate decision making in neonates with critical aortic stenosis in whom one- and two- ventricle repairs were being considered [8]. The authors used a parametric equation to estimate 5-year survival if the same (theoretical patient) had a one-ventricle repair compared to a two-ventricle repair. Surgeons using the calculator could input specific patient variables to generate an estimate the relative advantage (or disadvantage) of a one-ventricle over a two-ventricle repair. The calculator translates a complex statistical model into a practical tool to facilitate clinical decisions at the bedside.

Conclusion

The CHSS Datacenter is a unique research organization that has evolved and learned since 1985. The focus on rare lesions and complex management strategies has played to the strengths of the Data Center: superb statistical leadership, a dedicated Data Center staff collecting data from institutions across North America, and lifetime follow up of our cohorts. The institution of the Kirklin/Ashburn Fellowship has been a critical ingredient in our ongoing academic

success. All this, however, would never have been possible without the steadfast financial support and academic contributions of the membership of the Congenital Heart Surgeons Society.

References

1. Hickey EJ, Yeh T, Jacobs JP, Caldarone CA, Tchervenkov CI, McCrindle BW, Lacour-Gayet F, Pizarro C. Ross and Yasui operations for complex biventricular repair in infants with critical left ventricular outflow tract obstruction. Eur J Cardiothorac Surg. 2010;37(2):279–88.
2. Ashburn DA, Blackstone EH, Wells WJ, et al. Determinants of mortality and type of repair in neonates with pulmonary atresia and intact ventricular septum. J Thorac Cardiovasc Surg. 2004;127(4):1000–7; discussion 7–8.
3. Harris PA, Taylor R, Thielke R, Payne J, Gonzalez N, Conde JG. Research electronic data capture (REDCap) – a metadata-driven methodology and workflow process for providing translational research informatics support. J Biomed Inform. 2009;42(2):377–81.
4. Blackstone EH, Naftel DC, Turner Jr ME. The decomposition of time-varying hazard into phases, each incorporating a separate stream of concomitant information. J Am Stat Assoc. 1986;81:615–24.
5. Poynter JA, Eghtesady P, McCrindle BW, Walters HL, Kirshbom PM, Blackstone EH, et al. Association of pulmonary conduit type and size with durability in infants and young children. Ann Thorac Surg. 2013;96(5):1695–702.
6. McGiffin DC, Naftel DC, Kirklin JK, et al. Predicting outcome after listing for heart transplantation in children: comparison of Kaplan-Meier and parametric competing risk analysis. J Heart Lung Transplant. 1997;16:713–22.
7. Jegatheeswaran A, McCrindle BW, Blackstone EH, et al. Persistent risk of subsequent procedures and mortality in patients after interrupted aortic arch repair: a congenital heart surgeons' society study. J Thorac Cardiovasc Surg. 2010;140:1059–75. e1052.
8. Hickey EJ, Caldarone CA, Blackstone EH, et al. Critical left ventricular outflow tract obstruction: the disproportionate impact of biventricular repair in borderline cases. J Thorac Cardiovasc Surg. 2007;134(6):1429–36; discussion 36–7.
9. Konstantinov IE, Karamlou T, Williams WG, Quaegebeur JM, del Nido PJ, Spray TL, et al. Surgical management of aortopulmonary window associated with interrupted aortic arch: a congenital heart surgeons society study. J Thorac Cardiovasc Surg. 2006;131:1136–41.
10. Friedman AH, Fogel MA, Stephens P, Hellinger JC, Nykanen DG, Tweddell J, Feltes TF, Rome JJ. Identification, imaging, functional assessment and management of congenital coronary arterial abnormalities in children. Cardiol Young. 2007;17 Suppl 2: 56–67.
11. Poynter JA. Application of parametric hazard analysis with time-dependent covariables to analyze durability of right ventricle-to-pulmonary artery conduits implanted in infants and young children with congenital heart disease [MSc thesis]. Toronto: University of Toronto; 2014.

Clinical Versus Administrative Data

14

Sara K. Pasquali and J. William Gaynor

Abstract

Clinical and administrative databases have become increasingly utilized in the fields of pediatric cardiology and pediatric cardiac surgery for research, health policy activities, quality improvement, and evaluation of hospital performance. This chapter reviews the attributes of both types of datasets and discusses their strengths and weaknesses with regard to case ascertainment, risk adjustment, and outcomes assessment.

Keywords

Database • Outcomes • Congenital heart disease

Background

Large multicenter databases and registries have become increasingly utilized in the fields of pediatric cardiology and pediatric cardiac surgery over the past two decades [1]. These datasets serve several functions. First, they have allowed several different types of clinical research analyses including outcomes and comparative effectiveness type studies [2–4]. Due to the relative rarity of congenital heart disease, studies from a single center often lack statistical power or may have limited generalizability due to variation in practice and outcomes across centers. The use of large multicenter datasets helps to overcome these limitations. Health policy type analyses have also been conducted, such as investigations of the impact of center surgical volume or other structure and process measures on patient outcomes [5, 6]. In addition, large multicenter databases have been used for quality improvement purposes [7, 8]. As discussed in the previous chapters, several professional societies and other organizations now collect data across many centers regarding specific groups of patients with congenital heart defects, or those undergoing certain procedures. These data are then used to provide feedback to programs regarding outcomes benchmarked to national averages and

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peer institutions. This information may also be used for collaborative learning purposes where centers with the optimal outcomes are identified, best practices elucidated, and shared with other participating centers with the aim of improving overall quality of care and outcomes. Finally, information from large multicenter pediatric cardiac datasets has also been used for the purposes of public reporting of outcomes, ranking of hospital performance, and in some cases by large payers as evidence with which to base selective contracting, etc. [9–11].

Types of Datasets

In general there are two main types of datasets used for the purposes described above (Table 14.1).

Clinical Registries/Databases

These datasets are often run by professional societies or research groups for the purposes of quality improvement and/or research investigation. They are most often specific to a particular type of congenital heart disease, or collect information on patients undergoing specific types of procedures, e.g. congenital heart surgery, or cardiac catheterization. As described in the previous chapters, many of these types of datasets now exist in the field; examples include the Society of Thoracic Surgeons (STS) Congenital Heart Surgery Database, and the American College of Cardiology Improving Pediatric and Adult Congenital Treatment (IMPACT) Registry [7, 12]. Clinical registries and databases collect detailed information using standardized definitions specific to patients with congenital heart disease. Outcomes data captured are most often focused on clinical outcomes and may not include resource utilization data. Data are usually collected and entered by trained data managers under the direction of the clinical care team, or in some cases directly by clinicians. While participation is usually "voluntary", there are generally guidelines regarding the inclusion/exclusion of cases, and many datasets perform audits to evaluate the inclusion of eligible cases, and assess the degree of accuracy of important variables [7].

Administrative Datasets

Administrative datasets contain information already being collected for the purposes of hospital billing or insurance claims. They are not specific to congenital heart disease and most often collect information regarding all hospitalized patients at a state or national level, or all patients covered by a certain payer or insurer. Alternatively, some employ sampling strategies that allow a fixed sample of patients or hospitals to represent the overall sample [13]. Examples of these types of datasets include the Children's Hospital Association Pediatric Health Information Systems (PHIS) Database, the

Table 14.1 Administrative vs. clinical data

	Type of dataset	
	Clinical	Administrative
Population	Specific to CHD patients	All hospitalized patients
Purpose	Run by professional societies or research groups for purposes of research or QI	Data collected for hospital billing purposes
Coding	CHD-specific codes	ICD-9 codes
Data collection	Trained data managers/clinicians	Billing personnel
Data		
CHD comorbidities/clinical outcomes	Detailed	Limited
Resource utilization data	Limited	Detailed

CHD congenital heart disease, *ICD* International Classification of Diseases, 9th Revision, *QI* quality improvement

Agency for Healthcare Research and Quality's Healthcare Cost and Utilization Project datasets such as the Kid's Inpatient Database (KID), and state Medicaid datasets. Administrative datasets most often capture information in the form of a Uniform Hospital Discharge Dataset, which is a uniform minimum dataset that captures information about a hospitalization including demographics, International Classification of Diseases, version 9 (ICD-9) diagnosis and procedure codes, outcomes such as mortality, and important resource utilization data such as hospital charges [1]. Some datasets such as the PHIS database capture additional resource utilization data such as utilization of medications, laboratory tests, imaging, etc. The codes and data captured in administrative data sources are not specific to congenital heart disease. The data are captured by hospital coding and billing professionals. Data capture and submission is generally mandatory, and inclusive by the nature of these types of datasets.

Strengths and Weaknesses of Clinical vs. Administrative Datasets

The relative merits of these different types of datasets have been debated for several years. In cardiac surgery, many concerns were initially raised in the 1980s when the U.S. Health Care Financing Administration (HCFA) sent reports to each US cardiac surgeon informing them of their outcomes based on analyses of administrative data [14]. While the validity of the data was questioned in many cases, there were no alternative data sources at the time that could be analyzed. This prompted the formation of the Northern New England Cardiovascular Disease Study Group, a group of cardiac surgeons and epidemiologists representing several programs in Northern New England. This group conducted pioneering work in the field to develop one of the first clinical registries designed to collect uniform information on cardiac surgery patients. In particular, given the concerns regarding the HCFA data, the group was interested in ensuring that all relevant cases were captured, and that data were captured to allow for accurate adjustment for important differences in patient characteristics and case mix when evaluating outcomes across institutions. The registry subsequently served as the foundation for collaborative learning and quality improvement, which led to an overall improvement in outcomes across the region [14]. Since that time, many other clinical registries have been developed by other groups, and the strengths and weaknesses of administrative and clinical data continue to be debated. In assessing the relative merits of these different types of datasets, there are several important issues to consider.

Case Ascertainment

The first important area to consider surrounds issues related to case ascertainment; this includes both whether the dataset captures all relevant cases (for example all patients undergoing congenital heart operations at a hospital), and also whether the cases are coded correctly. On the one hand, proponents of administrative data note that these datasets may be more generalizable since by design they are inclusive of all patients and programs either nationally or in a specific region or state [1]. In addition, it is argued that there may be less potential for "gaming the system" or omission of cases with less than optimal outcomes from inclusion in the dataset, as the individuals collecting and submitting administrative data are not being judged or evaluated based on the data, while this may not be the case with clinical registry data where practitioners whose performance is being evaluated may be involved in the collection and submission of the data. However, over the past several years as many clinical registries have expanded, issues related to generalizability have become less of a concern. For example, the STS Congenital Heart Surgery Database now represents more than 85 % of all US pediatric heart surgery programs [7]. Submission of all eligible cases is most often a stipulation of participation in a clinical registry, however mechanisms to ensure this are still under development in many cases. While some registries have audit

programs that aim in part to evaluate appropriate inclusion of all cases, this is not uniform across all registries.

The other important issue related to case ascertainment involves accurate coding of cases. It has long been known that ICD-9 codes do not cover the breadth and depth of congenital heart disease or procedures. For example, there is no ICD-9 code for the Norwood operation. Thus, a combination of various diagnosis and procedure codes are often used by investigators in order to attempt to identify patients undergoing certain types of operations, including the Norwood operation, in analyses using administrative data. In addition to problems with the codes themselves, while the administrative coding personnel are skilled at coding, they likely have limited knowledge of congenital heart disease, and do not have regular contact with the medical team to clarify conflicting data or inconsistent documentation in the medical record. Several groups have investigated the scope of this problem related to miscoding of cases. Our group recently performed an analysis of more than 55,000 children across 33 hospitals undergoing congenital heart surgery who each had information collected and coded in both a clinical registry (STS Congenital Heart Surgery Database), and an administrative dataset (PHIS Database) [15]. We compared the operation coded for each patient between datasets. Using the clinical registry data as the gold standard, we found that for four of eight benchmark operations analyzed, there was a greater than 10 % difference in the number of cases identified in the administrative vs. the clinical data. While the negative predictive value of the administrative data was high across operations (98.8–99.9 %), the positive predictive value was lower (56.7–88.0 %). This indicates that it is highly likely that a patient without a certain operation coded in the administrative data truly did not have that operation performed as assessed in the clinical registry. Conversely, the lower positive predictive value indicates that many patients coded as having undergone a certain type of operation in the administrative data are false positives and actually had a different operation performed based on evaluation of the clinical registry data. In addition to individual operations, we also evaluated categories of operations with similar mortality risk [the Risk Adjustment in Congenital Heart Surgery (RACHS) categories]. Overall, the percent agreement between the administrative and clinical registry data regarding RACHS category assignment was 68.4 %. We also found that there were relatively consistent findings across hospitals with regard to misclassification/miscoding, suggesting that these discrepancies are likely more related to the limitations of the ICD-9 system and coding methodology itself, rather than related to any hospital-specific factors such as volume or case mix [15]. Several other studies have also reported similar findings regarding differences in coding and case ascertainment between administrative and clinical datasets [16–18]. These findings may have implications for the use of administrative datasets in evaluation of the number and type of congenital heart operations or cases across institutions.

Risk Adjustment

A second important area to consider in evaluating the relative merits of clinical vs. administrative data involves risk adjustment. Accurate adjustment for potential differences in important patient characteristics and case mix is important in a variety of situations, including when comparing or reporting outcomes across hospitals, or when comparing groups of patients in an observational analysis. Because administrative datasets and the ICD-9 coding system are focused on the larger general population of hospitalized patients, they do not necessarily collect information regarding important comorbidities or patient characteristics that are more specific to patients with congenital heart disease. For example, it is well known that weight at surgery is an important factor impacting outcome following congenital heart surgery, particularly in neonates [19]. However this variable is not collected in many administrative datasets. While clinical registries often collect more specific data related to comorbidities and characteristics of patients with congenital heart disease, there are still certain limitations in that not every

potential variable of interest may be captured, particularly those that may only pertain to only a subset of diagnoses or procedures.

Another issue is related to "date-stamping." This refers to the fact that ICD-9 diagnosis codes do not differentiate between conditions present at admission vs. those that developed during the hospitalization and may be complications. Thus, there can be misidentification of post-operative complications as comorbidities, and vise versa. Some datasets have begun to address this by including a variable that indicates "present on admission."

Outcomes Assessment

A final topic of consideration relates to outcomes assessment. First, because of the general nature of administrative datasets and ICD-9 codes, they may not capture all outcomes of specific interest to the congenital heart disease population, for example certain post-operative complications. Second, studies in the adult literature suggest that there can be significant miscoding or misclassification of certain outcomes such as post-operative complications in administrative data. For example, a recent analysis compared records of patients with information collected both in a Medicare claims data set (an administrative dataset) and the American College of Surgeons National Surgical Quality Improvement Program (ACS-NSQIP) registry [20]. Across 117,752 patients from more than 200 hospitals, investigators found that the sensitivity of the administrative data for detecting post-surgical complications coded in ACS-NSQIP ranged from 0.27 to 0.78 across various major complications. Although differences in complications have not been investigated extensively in the congenital heart surgery population, coding and capture of in-hospital mortality has been evaluated. In our recent analysis of more than 55,000 children across 33 hospitals who had data collected in both the STS Congenital Heart Surgery Database (clinical registry), and the PHIS Database (administrative dataset), we found that overall there was 99.83 % agreement between databases in in-hospital mortality, suggesting that there is not significant miscoding or capture of mortality data between these datasets [15].

In addition to the capture and correct coding of outcomes themselves, a second issue relates to errors in outcomes assessment due to miscoding or misclassification of cases. In other words, are there differences in the outcomes reported for certain diagnosis or procedure groups between datasets that are not related to the coding of the outcomes themselves, but related to differences in coding of the diagnosis or procedure across datasets? Our recent work with the STS and PHIS databases has investigated this further. We found that the differences in case ascertainment between data sources described above led to significant differences in outcomes assessment, for example an underestimation of mortality associated with truncus arteriosus repair by 25.7 % to an overestimation of mortality associated with ventricular septal defect repair by 31 % [15]. Differences were also found when evaluating mortality associated with larger groups of operations (the RACHS categories) between datasets, however these did not reach statistical significance. Importantly, only patients with concordant mortality status between the datasets were included in this analysis, in order to eliminate the possibility that any difference in outcomes identified might be related to differences in the coding of the outcomes themselves, rather than difference in the coding/classification of operations [15]. These findings may have implications for the use of administrative data in outcomes assessment, particularly at the level of individual congenital cardiac operations.

A final point to consider is the type of outcomes collected by clinical vs. administrative data sources. While clinical registries often collect more detailed clinical outcomes data, they most often do not collect the valuable resource utilization information contained in administrative datasets. In this era of increasing healthcare expenditures, it has become increasingly important to incorporate measures of cost into many analyses, as hospitals are under increasing pressure to not only optimize quality of care and outcomes, but also to reduce costs [21]. The

detailed resource utilization data contained in many administrative datasets is very valuable for these types of evaluations. Finally, neither type of dataset currently contains long-term follow up information, and most are focused primarily on in-hospital or short term outcomes. Methodology to incorporate longer-term clinical outcomes, resource utilization, and neurodevelopmental outcomes and quality of life is needed.

Conclusions

In summary, there are several important points to consider when evaluating the relative merits of administrative vs. clinical registry data, including issues related to case ascertainment, risk adjustment, and outcomes assessment. These factors may impact the relative utility of each type of dataset in outcomes and quality analyses, and in the evaluation of hospital performance. It is also important to note that while each type of dataset has its advantages and disadvantages, it is possible to capitalize on the strengths and mitigate some of the weaknesses of each dataset through database linkage strategies which allow for robust investigations not possible with either type of dataset alone [22]. This will be discussed further in Chap. 30.

References

1. Welke KF, Karmlou T, Diggs BS. Databases for assessing the outcomes of the treatment of patients with congenital and paediatric cardiac disease – a comparison of administrative and clinical data. Cardiol Young. 2008;18:137–44.
2. Pasquali SK, Hall M, Li JS, Peterson ED, Jaggers J, Lodge AJ, Marino BS, Goodman DM, Shah SS. Corticosteroids and outcome in children undergoing congenital heart surgery: analysis of the pediatric health information systems database. Circulation. 2010;122:2123–30.
3. Fudge JC, Li S, Jaggers J, O'Brien SM, Peterson ED, Jacobs JP, Welke KF, Jacobs ML, Li JS, Pasquali SK. Outcomes in patients with down syndrome undergoing congenital heart surgery: analysis of a national clinical database. Pediatrics. 2010;126:315–22.
4. Karamlou T, Hirsch J, Welke K, Ohye RG, Bove EL, Devaney EJ, Gajarski RJ. A united network for organ sharing analysis of heart transplantation in adults with congenital heart disease: outcomes and factors associated with mortality and retransplantation. J Thorac Cardiovasc Surg. 2010;140:161–8.
5. Pasquali SK, Jacobs JP, He X, Hornik CP, Jaquiss RDB, Jacobs ML, O'Brien SM, Peterson ED, Li JS. The complex relationship between center volume and outcome in patients undergoing the Norwood operation. Ann Thorac Surg. 2012;93:1556–62.
6. Welke KF, Diggs BS, Karamlou T, Ungerleider RM. The relationship between hospital surgical case volumes and mortality rates in pediatric cardiac surgery: an national sample, 1988–2005. Ann Thorac Surg. 2008;86:889–96.
7. Jacobs JP, Jacobs ML, Mavroudis C, Lacour-Gayet FG, Tchervenkov CI, Pasquali SK. Executive summary: the Society of Thoracic Surgeons Congenital Heart Surgery Database – 18th Harvest – (January 1, 2009–December 31, 2013). The Society of Thoracic Surgeons (STS) and Duke Clinical Research Institute (DCRI), Duke University Medical Center, Durham, Spring 2013 Harvest.
8. Kugler JD, Beekman RH, Rosenthal GL, Jenkins KJ, Klitzner TS, Martin GR, Neish RS, Lannon C. Development of a pediatric cardiology quality improvement collaborative: From inception to implementation. From the Joint Council on Congenital Heart Disease Quality Improvement Task Force. Congenit Heart Dis. 2009;4:318–28.
9. Pediatric Congenital Cardiac Surgery in New York State 2006–2009. Available at: http://www.health.ny.gov/statistics/diseases/cardiovascular/heart_disease/docs/2006-2009_pediatric_congenital_cardiac_surgery.pdf. Accessed 10 Jul 2013.
10. U.S. News & World Report's Annual Ranking of Best Hospitals. Available at: http://www.rti.org/besthospitals. Accessed 28 May 2013.
11. OptumHealth Congenital Heart Disease Center of Excellence. Available at: https://www.myoptumhealthcomplexmedical.com/gateway/public/chd/providers.jsp. Accessed 28 May 2013.
12. Martin GR, Beekman RH, Ing FF, Jenkins KJ, McKay CR, Moore JW, Ringel RE, Rome JJ, Ruiz CE, Vincent RN. The IMPACT registry: improving pediatric and adult congenital treatments. Semin Thorac Cardiovasc Surg Pediatr Card Surg Annu. 2010;13:20–5.
13. The Nationwide Inpatient Sample Database. Available at: http://www.hcup-us.ahrq.gov/db/nation/nis/nisdbdocumentation.jsp. Accessed 10 Jul 2013.
14. Likosky DS. Lessons learned from the Northern New England Cardiovascular Disease Study Group. Prog Pediatr Cardiol. 2012;33:53–6.
15. Pasquali SK, Peterson ED, Jacobs JP, He X, Li JS, Jacobs ML, Gaynor JW, Hirsch JC, Shah SS, Mayer JE. Differential case ascertainment in clinical registry vs. administrative data and impact on outcomes assessment in pediatric heart surgery. Ann Thorac Surg. 2013;95:197–203.
16. Strickland MJ, Riehle-Colarusso TJ, Jacobs JP, et al. The importance of nomenclature for congenital cardiac disease: implications for research and evaluation. In: Jacobs JP, editor. 2008 Cardiology in the

Young Supplement: Databases and The Assessment of Complications associated with The Treatment of Patients with Congenital Cardiac Disease. Prepared by: The Multi-Societal Database Committee for Pediatric and Congenital Heart Disease. Cardiology in the Young, vol. 18, issue S2 (Suppl 2), p. 92–100, December 9, 2008.
17. Cronk CE, Malloy ME, Pelech AN, et al. Completeness of state administrative databases for surveillance of congenital heart disease. Birth Defects Res A Clin Mol Teratol. 2003;67:597–603.
18. Frohnert BK, Lussky RC, Alms MA, Mendelsohn NJ, Symonik DM, Falken MC. Validity of hospital discharge data for identifying infants with cardiac defects. J Perinatol. 2005;25(11):737–42.
19. Curzon CL, Milford-Beland S, Li JS, O'Brien SM, Jacobs JP, Jacobs ML, Welke KF, Lodge AJ, Peterson ED, Jaggers J. Cardiac surgery in infants with low birth weight is associated with increased mortality: analysis of the Society of Thoracic Surgeons Congenital Heart Surgery Database. J Thorac Cardiovasc Surg. 2008;135:546–51.
20. Lawson EH, Louie R, Zingmond DS, Brook RH, Hall BL, Han L, Rapp M, Co CY. A comparison of clinical registry versus administrative claims data for reporting of 30-day surgical complications. Ann Surg. 2012;256:973–81.
21. Pasquali SK, Gaies MG, Jacobs JP, Gaynor JW, Jacobs ML. Centre variation in cost and outcomes for congenital heart surgery. Cardiol Young. 2012;22:796–9.
22. Pasquali SK, Jacobs JP, Shook GJ, O'Brien SM, Hall M, Jacobs ML, Welke KF, Gaynor JW, Peterson ED, Shah SS, Li JS. Linking clinical registry data with administrative data using indirect identifiers: Implementation and validation in the congenital heart surgery population. Am Heart J. 2010;160:1099–104.

15 Databases for Pediatric Cardiac Transplantation: The United Network for Organ Sharing/ Scientific Registry of Transplant Recipients (UNOS/SRTR) and the Pediatric Heart Transplant Study (PHTS)

Ryan R. Davies

Abstract

Collection, analysis, and dissemination of data have been part of the transplantation since its earliest days. The two largest databases containing information on pediatric cardiac transplant patients are the United Network for Organ Sharing/Scientific Registry of Transplant Recipients (UNOS/SRTR) database and the Pediatric Heart Transplant Study (PHTS). These data have enabled examination of patients undergoing transplantation, including modeling of outcomes, analysis of allocation decisions, and the examination of criteria for listing. Extensive literature exists utilizing this data, but must be read critically, recognizing the limitations presented by missing variables (whether uncollected or collected but left blank), reproducibility, and small sample sizes among pediatric patients. However, despite these limitations, these datasets provide an important resource in the ongoing examination of cardiac transplantation in children.

Keywords

Transplantation • Cardiac failure • Pediatric cardiac transplantation • Pediatric cardiac failure • Pediatric cardiac disease • Congenital cardiac disease • United Network for Organ Sharing/Scientific Registry of Transplant Recipients (UNOS/SRTR) database • Pediatric Heart Transplant Study (PHTS)

Introduction

This chapter is based in part on a presentation at The Seventh Annual Meeting of The Multi-Societal Database Committee for Pediatric and Congenital Heart Disease: "Meeting Theme: The relationship between Outcomes Analysis,

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Quality Improvement, and Patient Safety", Tuesday, September 20, 2011 and Wednesday, September 21, 2011, Selwyn College, University of Cambridge, Cambridge, United Kingdom, and, subsequently published in the World Journal of Pediatric and Congenital Heart Surgery [1].

Even the busiest pediatric cardiac transplant centers have averaged fewer than 21 transplants per year over the past 25 years [2]. Thus, in common with other areas of pediatric heart surgery, outcomes analysis is hampered by the limited sample size available at any individual institution. In this setting, multi-institutional data collection is necessary in order to develop statistically-sound, comprehensive analyses and conclusions.

There are multiple databases, including both voluntary multi-institutional collaborations and federally mandated submissions, containing clinical outcomes data on patients undergoing cardiac transplantation. Each of these databases has advantages and disadvantages. A thorough understanding of the historical background, data collection and distribution, and techniques of analysis each dataset is important in critically evaluating published outcomes data.

The UNOS/SRTR Database

Historical Background

There is an alphabet soup of acronyms for organizations involved in administering transplantation with the United States of America. The dataset that is commonly referred to as the United Network for Organ Sharing (UNOS) Database consists of information collected by UNOS and maintained and analyzed by the Scientific Registry of Transplant Recipients (SRTR). UNOS is the public corporation which administers the Organ Procurement and Transplant Network (OPTN), under contract with the federal government. In addition to administering the OPTN, UNOS performs other functions including education and awareness of issues surrounding organ donation and transplantation. In the literature, the UNOS Database may be referred to by any of the following acronyms: SRTR database, UNOS database, OPTN database (or combinations thereof). The acronym used is often an indicator of the administrative source of data analyzed. However, in all cases the majority of the underlying data is collected by UNOS as part of the process of listing, donation, transplantation, and follow-up. SRTR combines this information with data from other sources including the Social Security Death Master File. Throughout this chapter we have referred to this database as the **UNOS/SRTR database**.

From the first successful renal transplant (in 1954) and the first organ retrieval from a cadaveric donor (in 1962) through the 1970s, transplantation was coordinated by local and regional groups of hospitals and transplant physicians [3, 4]. This resulted in variability in the provision of transplant care, including definitions of donor brain death and the allocation of donor organs. In response to this variability (as well as a concern about monetary remuneration of donors), the United States Congress passed the National Organ Transplantation Act (NOTA) in 1984. Among its effects was the creation of a single Organ Procurement and Transplantation Network (OPTN) and nationalization of the transplant lists. The United Network for Organ Sharing (UNOS) (a successor to one of the regional groups: Southeastern Organ Procurement Organization), has had the contract to administer the OPTN since its inception.

As part of developing equitable organ allocation schemes, NOTA stated explicitly that one of the roles of the OPTN was to "collect, analyze, and publish data concerning organ donations and transplants." It also established what would become the Scientific Registry of Transplant Recipients (SRTR):

> The Secretary [of Health and Human Services] shall, by grant or contract, develop and maintain a scientific registry of the recipients of organ transplants. The registry shall include such information respecting patients and transplant procedures as the Secretary deems necessary to an ongoing evaluation of the scientific and clinical status of organ transplantation [5].

The SRTR is currently administered by the Minneapolis Medical Research Foundation (MMRF) and supports ongoing evaluation of solid

organ transplantation. By providing a rigorous data and analytic component, it has an essential role in providing data to support the development of evidence-based policies of allocation through collaboration with the transplant community and OPTN. Therefore, NOTA enshrined ongoing data collection and analysis as an integral part of OPTN and organ transplantation.

In addition to the contracted analytic functions of the SRTR, the OPTN Final Rule committed to the importance of public access to scientific data:

> **Respond to reasonable requests from the public for data needed for bona fide research or analysis purposes**, to the extent that the OPTN's or Scientific Registry's resources permit, or as directed by the Secretary. The OPTN or the Scientific Registry may impose reasonable charges for the separable costs of responding to such requests. Patient-identified data may be made available to bona fide researchers upon a showing that the research design requires such data for matching or other purposes, and that appropriate confidentiality protections, including destruction of patient identifiers upon completion of matching, will be followed. All requests shall be processed expeditiously, with data normally made available within 30 days from the date of request (emphasis added) [6].

This OPTN Final Rule has resulted in a situation in which UNOS members (transplant centers, organ provider organizations (OPOs), histocompatibility laboratories) can obtain free access to the entire OPTN dataset. Fee-based access is available to interested researchers who are not UNOS members through the SRTR, and the SRTR can also provide additional programming. The UNOS/SRTR data is thereby both easily accessible to a wide-range of researchers and analyzed in a rigorous and consistent manner by the SRTR to provide comprehensive and reliable data to UNOS and the public.

Data Collection

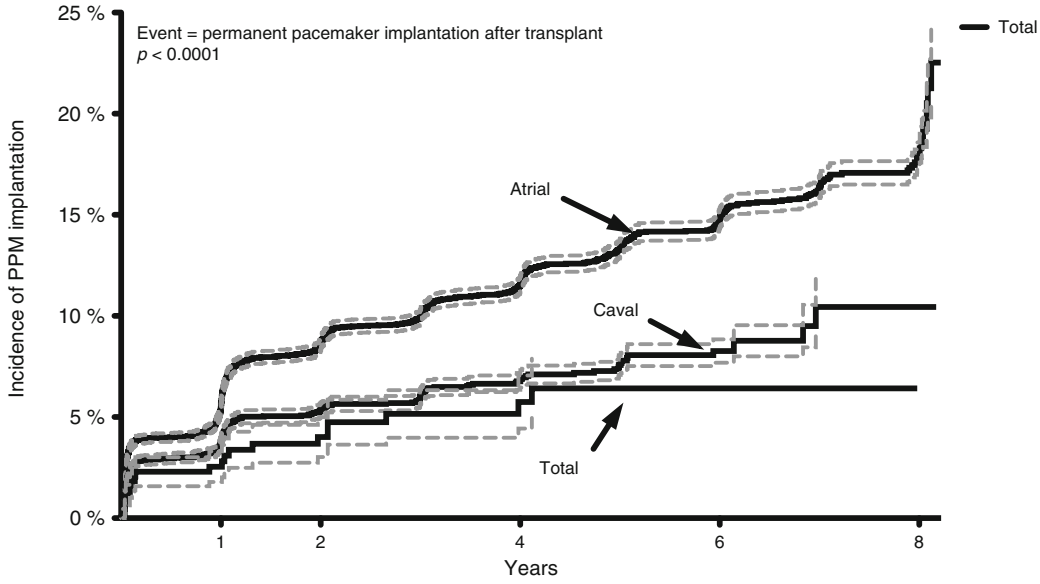
The UNOS/SRTR Database consists of all transplants performed in the United States from 1988 to the present. It consists of data collected at listing, at transplantation (both regarding the recipient and the donor), and in yearly follow-up. Data submission is a mandatory aspect of membership in UNOS and allocation of organs from the OPTN [7]. However, data submission may not necessarily be complete and missing data are a persistent problem in analyzing UNOS/SRTR data (see below) [7].

With respect to pediatric heart transplants, the primary weakness of the UNOS/SRTR dataset is the lack of data collection specifically directed at issues in pediatric and congenital patients, including exact congenital diagnosis, previous procedures, and physiologic status when listed. To date, studies evaluating congenital heart disease as a risk factor have had to use the broad and non-specific designation lumping all patients with congenital heart disease (CHD) together [8–10]. In addition, potential risk factors that may be important among pediatric patients may not be adequately captured in the adult-centric UNOS/SRTR dataset; such potential risk factors include [11–16]:

- technical aspects such as the need for pulmonary artery reconstruction,
- timing following failure of attempted surgical palliation,
- preoperative functional status, and
- the presence of non-cardiac congenital anomalies or genetic syndromes.

As the challenges with missing data make clear, another important drawback of the UNOS/SRTR dataset is the lack of an audit mechanism. While UNOS audits transplant centers and other transplant organizations on a routine basis, it is interested primarily in factors which might influence equitable allocation and quality assurance programs to prevent important errors such as misidentification of ABO type resulting in hyper-acute graft rejection. Factors which may be important to outcomes analysis, including levels of panel reactive antibodies, the numbers of previous sternotomies, are unlikely to be audited and compared to the medical record for accuracy. Therefore, the accuracy of the data (especially secondary outcomes such as lengths-of-stay, time to rejection episodes, and the incidence of non-compliance) has not been independently verified.

In addition to the potential for data inaccuracies, follow-up data within the dataset is submitted



Subjects at risk

Atrial	9,175	6,771	4,842	3,012	840
Caval	4,855	3,061	1,498	397	26
Total	438	280	159	43	2

Fig. 15.1 Kaplan-Meier estimates of the cumulative hazard of permanent pacemaker insertion after transplantation. Eight-year estimates are illustrated as a function of the type of transplant anastomosis: *CAVAL* Bicaval anastomosis, *ATRIAL* biatrial anastomosis, *TOTAL* total heterotopic transplant anastomo- sis. Numbers of subjects at risk at each time point are given across the *bottom*, and standard errors are shown by the *dashed lines*. P<0.0001 (caval vs atrial) (Reprinted from Davies et al. [72], with permission from Elsevier)

yearly by the transplant center, rather than event-driven submissions. This strategy of collection of data leads to follow-up data with steps reflecting yearly submission of follow-up data forms, rather than gradual slopes indicating the actual date of occurrence of the event (Fig. 15.1). Therefore, other than endpoints such as graft failure or death, the analysis of long-term outcomes is constrained.

Data Access and Analysis

As noted above, the legislative history of NOTA and the OPTN Final Rule has resulted in a database that is easily accessible to a wide-range of investigators. The result is that a wide-range of investigators with differing interests and differing expertise have the potential to perform complex statistical analyses of the data and provide relatively frequent updates to previous analyses. This contrasts with other multi-institutional databases within cardiac surgery, such as the Society of Thoracic Surgeons-Congenital Heart Surgery Database, the New York State Cardiac Surgery Reporting System, and the Pediatric Heart Transplant Study. In these cases, access to the raw data is limited and controlled. Also, statistical analyses are often performed by a single entity, and funding may be required to reimburse for statistical analysis (similar to the SRTR analyses of UNOS/SRTR data).

The more open nature of the UNOS/SRTR database has resulted in a wide-range of publications. Often, multiple authors may investigate the same question using different methods – as with the comparison between bicaval and biatrial anastomoses [17, 18], the impact of ventricular assist devices on post-transplant outcomes [19, 20], or outcomes following transplantation among adults with congenital heart disease [8, 9]. While this approach may result in duplicative effort, it often results in a broader understanding of the issue, and consistency of results across analytic methods reinforces the reliability of the findings.

However, the lack of standardization evident in these various analyses also illustrates an important pitfall of open access to raw data. Readers of manuscripts based on UNOS/SRTR data must be diligent in assessing the statistical methods. Different research teams may handle missing variables in importantly different ways [17, 18], or may convert raw data (especially the often informative but labor-intensive text fields) into variables using different techniques. Therefore [19, 20], the open-access nature of the UNOS/SRTR data places a higher burden on the clinician to evaluate the methods used within each individual manuscript.

Pediatric Heart Transplant Study

Historical Background

In the late 1980s, data from both the International Society for Heart and Lung Transplantation Registry (see below) and UNOS/SRTR suggested that there were significant differences in survival and risk factors for death between adult and pediatric heart transplant recipients [21]. In 1991, to address the lack of pediatric specific date, a group of pediatric heart transplant clinicians along with investigators within the Department of Surgery at the University of Alabama-Birmingham formed a voluntary, multi-institutional, collaborative effort to "advance the science and treatment of children while waiting for cardiac transplantation [21]." This collaborative effort, called the Pediatric Heart Transplant Study (PHTS), began collecting data in 1993. In 2000, the PHTS adopted a more formal structure including a governing board and standing committees to supervise various aspects of the effort. Members of the PHTS support research efforts through annual fees. Current membership consists of 44 member institutions from the United States, Canada, and the United Kingdom [22].

Data Collection

In contrast to the mandatory, publicly accessible UNOS/SRTR database, the PHTS is a voluntary association of transplant centers. Membership and data submission is voluntary. PHTS currently includes approximately 66 % of the pediatric transplants performed in the United States [4]. There are several advantages to both the data fields collected and the data collection methods as compared to the UNOS/SRTR data collection.

First, because the PHTS is a pediatric-specific database, data collection is geared toward variables of particular interest to pediatric transplantation, including congenital diagnoses and previous surgeries [23]. This advantage is somewhat mitigated by changes over time in the diagnostic and procedural categories through iterations of data collection, reinforcing the importance of standardized diagnostic and procedural coding, as described elsewhere in this book (in the chapter by Franklin and colleagues titled: ***Nomenclature for Congenital and Pediatric Cardiac Disease: Historical Perspectives and the International Pediatric and Congenital Cardiac Code***).

Second, in contrast to UNOS/SRTR, data collection is event-driven. Following entry into the database at listing, information is gathered annually but also at any of the following events [22]:

- transplantation,
- death,
- rejection,
- infection,
- malignancy,
- allograft vasculopathy, and
- retransplantation.

Time-to-event analysis is, therefore, more robust than within the UNOS/SRTR data set. In addition, the information collected at each event is much more detailed because data submission includes event-specific forms.

Data Access and Analysis

The data is not publicly accessible, nor is it directly accessible to member institutions. Instead, data is collected and centralized at the PHTS within the University of Alabama – Birmingham (UAB). Proposals for research are submitted to a Scientific Committee which meets twice a year to approve projects. Statistical analysis is performed by PHTS/UAB staff [22]. While advantages exist in

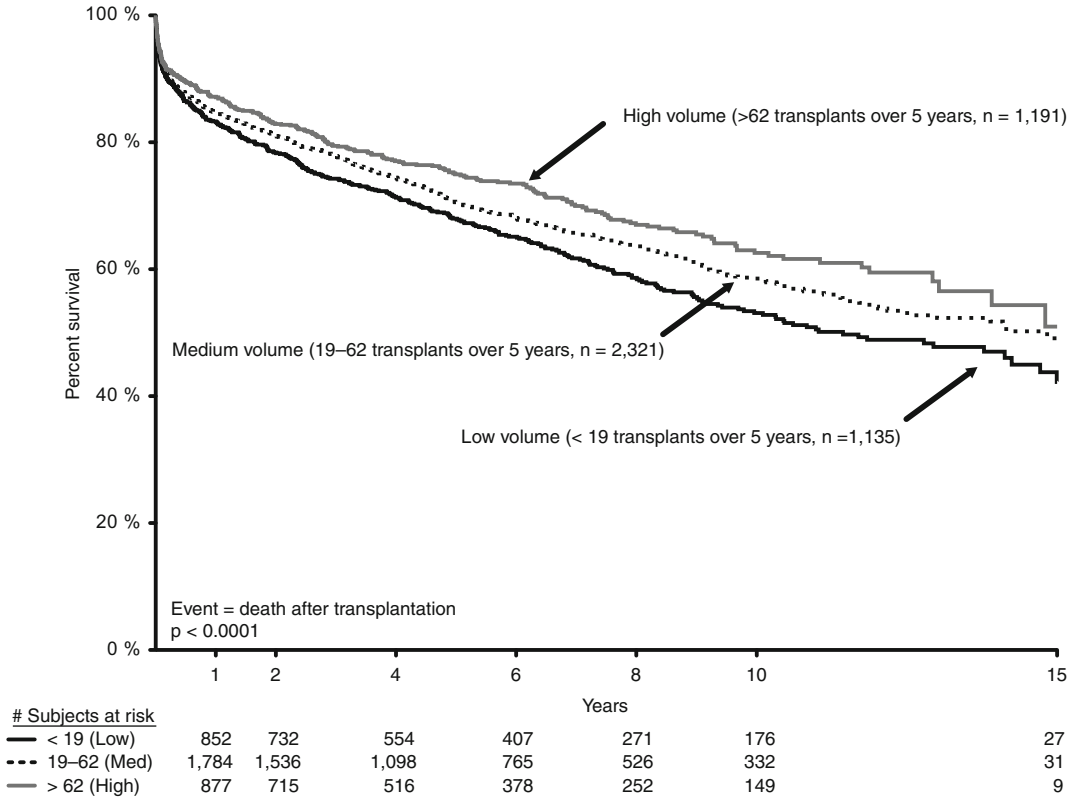


Fig. 15.2 Fifteen-year Kaplan-Meier survival estimates as a function of transplant center volume ($P<0.0001$), unadjusted for risk. The number of patients at risk is given at the *bottom* of the graph (Reprinted from Davies et al. [24])

terms of consistency and reliability, this strategy does result in a more limited number of publications derived from the data source. In addition, it is important to recognize that the PHTS transplant centers primarily higher volume, children's hospitals. The extent to which the experience within the PHTS centers is representative of the experience across the broad spectrum of transplant centers performing pediatric cardiac transplantation is not clear, especially when lower volume centers have higher mortality – especially in high risk patients (Figs. 15.2 and 15.3) [24].

Other Databases Including Pediatric Transplant Data

While the majority of multi-institutional publications regarding pediatric cardiac transplantation are based on either UNOS/SRTR or PHTS data, other databases have important applications in analyzing outcomes of children undergoing cardiac transplantation. These additional databases that capture information related to patients undergoing transplantation for pediatric and congenital cardiac disease include:

- The International Society for Heart and Lung Transplantation (ISHLT) Registry,
- INTERMACS/PEDIMACS, and
- The Society of Thoracic Surgeons-Congenital Heart Surgery Database (STS-CHSD).

International Society for Heart and Lung Transplantation (ISHLT) Registry

The ISHLT created its transplant registry to provide an ongoing assessment of the status of thoracic organ transplantation worldwide.

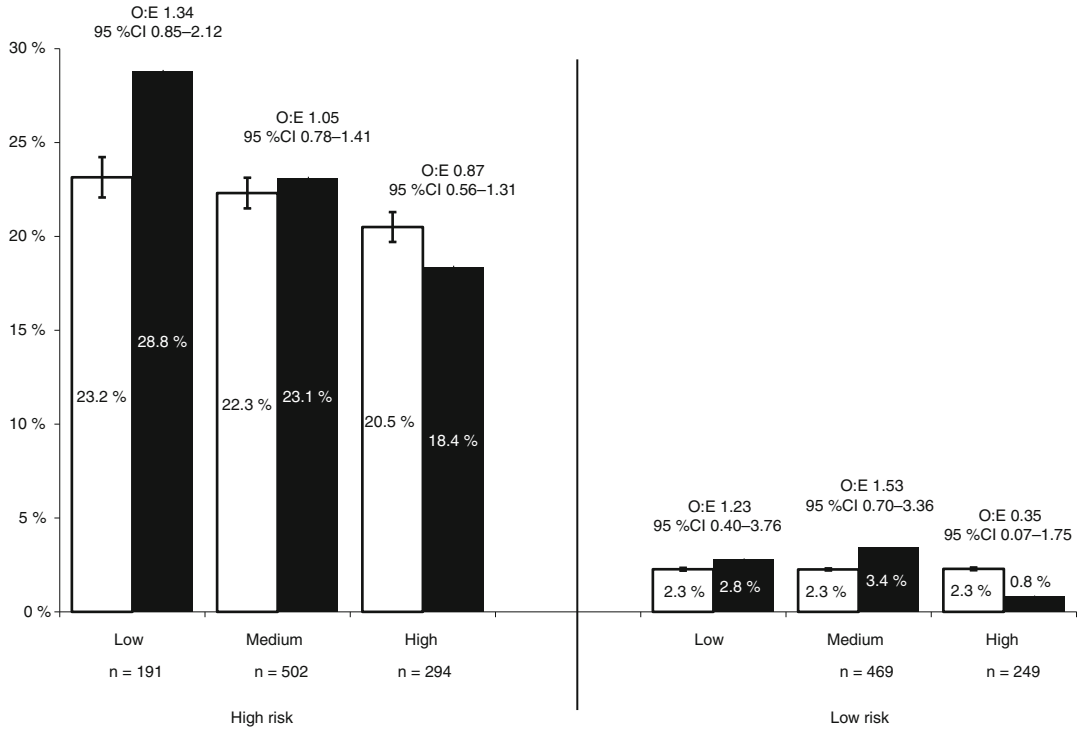


Fig. 15.3 Predicted (*white* column) versus observed (*black* column) postoperative mortality rates by volume of transplants performed in the previous 5 years by the transplant center. Results are stratified by patient risk: high-risk patients (>75th percentile for postoperative mortality) are shown at the *left*, low-risk patients (<25th percentile for postoperative mortality) are shown on the *right* (Reprinted from Davies et al. [24])

It currently aggregates data from nearly 400 transplant centers, including both individual institutions and through data interfaces with government agencies including UNOS/SRTR [25]. As such, the data has the same weaknesses as UNOS/SRTR data, magnified across the diversity of submitting institutions. The primary advantage of the ISHLT registry is that it allows for international comparisons of transplant outcomes. Otherwise, the data is essentially a subset of the data collected by UNOS/SRTR.

INTERMACS/PEDIMACS

The INTERMACS database is a voluntary database funded by a contract from the National Heart Lung and Blood Institute and currently run by staff also at the University of Alabama. Although voluntary, reimbursement for destination ventricular assist device therapy from the Centers for Medicare and Medicaid Services is contingent on submission of data to a national, audited database, and most high-volume ventricular assist device centers participate [26]. To date, submission of pediatric data is limited, but as mechanical circulatory support becomes more common in children, it will provide an important adjunct to the two previous databases in exploring the outcomes of patients with end-stage heart failure [27].

Society of Thoracic Surgeons Congenital Heart Surgery Database (STS-CHSD)

Other chapters in this text provide a more extensive description of the STS-CHSD. Currently, the STS-CHSD has not been used in analyzing pediatric transplant data, largely because it does not contain variables of critical importance to

transplant outcomes, including donor and match variables, as well as long-term graft follow-up. However, it contains substantially more data regarding congenital diagnoses, historical procedures, and technical details of the transplant operation than any of the previously described data sources. Linkage of the STS-CHSD to UNOS/SRTR or PHTS data has the potential to leverage the strengths of the different databases and mitigate the weaknesses.

Outcomes Analysis in Pediatric Cardiac Transplantation Using Large Datasets

Analysis of outcomes in transplantation is needed for several distinct but related areas: (1) prediction of individual risk of transplantation and identification of appropriate transplant candidates, (2) evaluation of individual transplant programs, including quality assessment and improvement as well as public reporting of results, and (3) evaluation of the impact of global policy decisions – especially regarding organ allocation – in order to optimize the utility of each available donor allograft.

Each of these areas of analysis has unique challenges, but they have in common the limited outcomes measures available. Whether using the UNOS/SRTR dataset or data from the PHTS, patient and graft survival are the primary outcomes available within the datasets. These data provide only a limited picture. Other outcomes that may be even more pertinent include indicators of quality of life and of functional status. With the increasing use of implantable ventricular assist devices (especially in older, near-adult, pediatric patients), quality of life rather than absolute survival will become more important as the survival following transplantation and VAD implantation becomes equivalent [28].

Currently, however, the ease of collection and analysis of a binary and easily assessed outcome, such as patient survival, graft survival, or the occurrence of drug-treated infection, make these the de facto standard for assessments of outcomes in pediatric cardiac transplantation. We will focus on these binary outcomes here.

Prediction of Individual Risk

Estimation of the risk of mortality or graft failure for an individual patient following cardiac transplantation is crucial to all aspects of outcomes analysis. Obviously counseling of individual patients and their families regarding the patient's likelihood of post-transplant survival is necessary for informed decisions regarding candidacy. In addition, without being able to risk-adjust outcomes at individual center, it becomes impossible to compare centers with each other or to assess changes outcomes at an individual center over time. Furthermore, estimates of the survival with and without an allograft are critical to optimizing allocation schema.

Models of Risk on the Waitlist

Models of waitlist outcomes have been published in both adults and children, but – in addition to the predominance of adult research – they have several drawbacks [8, 29–33]:

- First, within the pediatric population, in order to identify as many risk factors as possible, longer periods of time are used to increase the sample size [24, 34]. As the field advances and clinical care evolves, use of these longer periods of time may result in a heterogeneous population being analyzed together. Especially where mortality has decreased over time [35], much of the mortality will have occurred in the least contemporary population and the risk factors important in this earlier era will be overweighted in any combined analysis.
- Second, to date these models have been constructed to examine specific risk factors as opposed to attempting to identify the most accurate model [31, 32, 36].
- Third, reporting of waitlist mortality models often fails to include enough information (including intercepts and model parameter estimates) to reconstruct the model for use in individual risk prediction [33, 35, 37].
- Fourth, the heterogeneity of the pediatric heart failure population (including dilated cardiomyopathy [DCM] patients, patients with

CHD, and those with restrictive cardiomyopathy, among others) suggests that risk factors may vary between diagnoses. Factors important to patients with CHD may differ significantly from those important in patients with DCM [11, 31, 38].

- Finally – and most importantly – to date no waitlist model has been constructed and validated in a separate population in order to identify its independent accuracy for use outside of the derivation population.

Despite these limitations, data from both the PHTS and UNOS/SRTR has been consistent in identifying certain risk factors as predictive of waitlist mortality, including: the need for ECMO, mechanical ventilation, and a diagnosis of congenital cardiac disease [31–37, 39]. Other factors, including race, socioeconomic status, and age have varied across models. If we are to move to a model for allocation of hearts similar to the system for allocation of lungs currently in use (see below), more accurate prediction of mortality on the waitlist is required in order to allocate available allografts to those patients least likely to survive without transplantation. Not all of the limitations can be addressed, but further analysis of the UNOS/SRTR database and the PHTS database, as well as potential linkages to other datasets, should enable improvements in the accuracy of predicting mortality on the waitlist.

Models of Risk of Mortality After Transplantation

Several researchers have published risk prediction models using both UNOS/SRTR and PHTS data [10, 24, 40, 41]. These models may be helpful in estimating risk for an individual patient. However, a critical assessment of model accuracy is essential prior to using the model in a particular population of patients.

The c-statistic is valuable as a rough (though not perfect) [42] estimate of model accuracy. For example, Weiss et al. have recently published a score to be used in predicting outcomes among adults following cardiac transplantation [43]. The c-statistic for the final model was only 0.65, suggesting that it lacks significant discriminatory ability. In contrast, models used in identifying the importance of volume in outcomes following pediatric heart transplant have a c-statistic of 0.75 [24], suggesting that better discrimination is possible. However, a c-statistic of 0.75 suggests that much of the variation in outcomes results from variables not in the model. It is likely that other variables, poorly contained within the UNOS/SRTR dataset, may be particularly important including functional status, nutritional status, and the presence of cachexia. Validation of the model in a population distinct from that in which it was derived is an important step in assessing the accuracy of any model.

Table 15.1 Risk factors to derive simplified score predictive of mortality after cardiac transplantation in children

Risk factor	Weighted score
ECMO	4
Number of high risk criteria >3*	3
Reoperation	2
Stroke	2
Renal insufficiency (creatinine clearance <40 cc/min)	2
Congenital heart disease	1
Mechanical ventilation	1
Admitted to ICU	1

*High risk criteria include: PVRI >6 woods units, creatinine clearance >40 cc/min, hepatitis C positivity, donor: recipient weight ratio <0.7, panel reactive antibody >40 %, retransplantation, age ≤1 year

Given these limitations, are there models which may be useful? Prediction of risk is often a balance between accuracy and simplicity. For example, the group from Columbia has looked at estimates of mortality among high-risk transplant recipients using a simplified risk score (Table 15.1) (Fig. 15.4) [10]. While the model provides a rough estimate of outcomes and may be useful in broadly assessing the risk of any individual patient, this comes at the expense of accuracy (importantly, measures of accuracy of the model are not reported in the manuscript). The data from that study also illustrates one problem with many of these risk prediction models, which is the small number of patients at the highest risk levels. This problem often results in

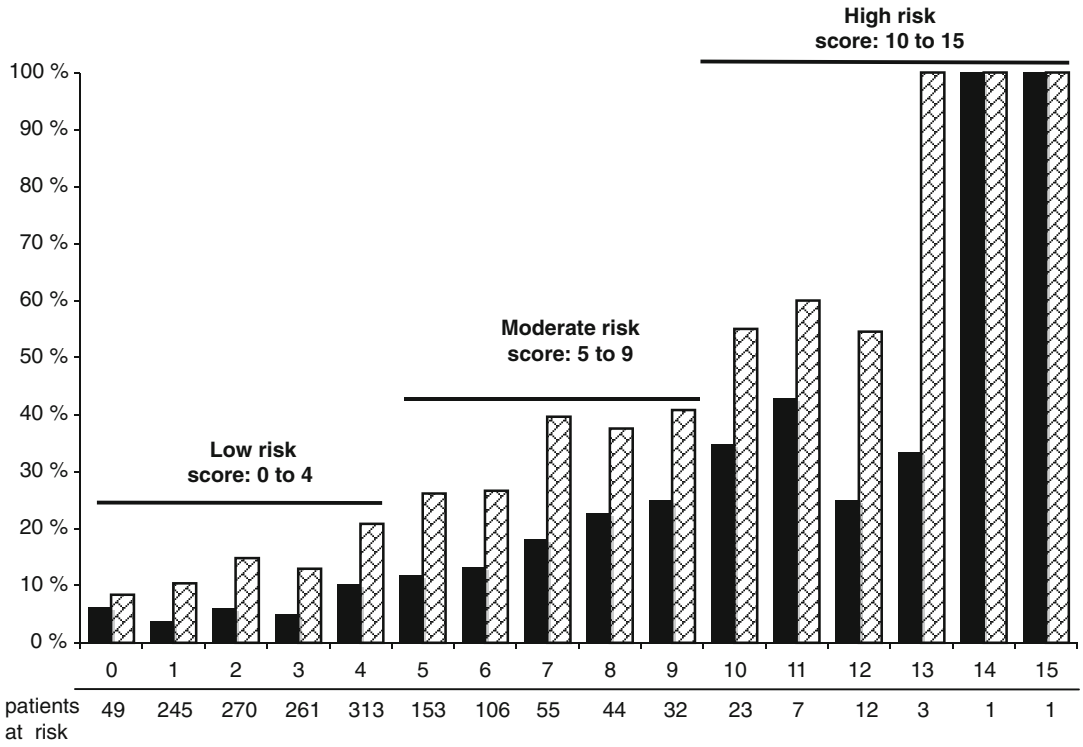


Fig. 15.4 Observed 30-day (*black*) and 1-year (*slanted brick pattern*) mortality for each risk score in patients with at least one high risk criteria ($P < 0.0001$) (Reprinted from Davies et al. [24])

models overestimating risk among the highest risk patients [44].

The PHTS dataset has the potential to provide more accurate estimates of post-transplant outcomes because it is more comprehensive with regard to variables of specific interest in pediatric cardiac transplantation [31]. A predictive model based on that data would be particularly interesting. While a predictive model based on PHTS data was presented in 2008, it has not yet been published in a form which would enable prediction of individual risk of post-transplant mortality [40]. Similarly, an attempt to investigate high-risk criteria for pediatric transplant has not moved from the abstract to completed manuscript phase [39].

As with waitlist models, there is broad agreement on several risk factors for poor outcomes after pediatric cardiac transplantation, including congenital heart disease, the need for extracorporeal membrane oxygenation or mechanical ventilation, and renal failure [10, 24, 45]. Other factors, including measurements of cardiac dimensions and the etiology of cardiomyopathies, may be important in specific subsets of pediatric patients [31]. Current data have identified risk factors for poor outcomes, and currently available modeling enables broad stratification of patients into risk categories [10]; however, accurate prediction of individual risk may require comprehensive modeling within patients subsets, likely categorized by etiology. UNOS/SRTR data may not be sufficient and linkage to other datasets may be required.

Summary

In summary, while there exist several published models of both pre-transplant mortality and post-transplant mortality among children requiring cardiac transplantation, none have been externally validated. Even the most accurate models suggest that additional variables not currently captured

within the datasets explain important amounts of variation in outcome. Ongoing research, including the potential linkage of UNOS/SRTR or PHTS data to other datasets such as the STS-CHSD may be required to provide modeling with the accuracy to predict individual outcomes in these children.

Assessment of Individual Transplant Centers

Estimates of outcomes at individual transplant centers are publicly available based on SRTR analysis of the UNOS/SRTR dataset (see SRTR website: [http://www.srtr.org/csr/current/Centers/Default.aspx]) on a semi-annual basis. The SRTR uses risk-adjustment models that are available on the website to evaluate and compare programs. Unfortunately, the SRTR models have several limitations in assessing inter-institutional variation. First, the models do not account for several factors known to influence outcomes, including

- preoperative kidney function [24, 46],
- etiology of heart failure [24, 46–50], and
- reoperative sternotomy [24, 47, 49].

Furthermore, as noted in the introduction to this chapter, pediatric cardiac transplantation at even the busiest programs is a relatively low-volume procedure.

Throughout pediatric and congenital heart surgery, low center volume for individual procedures makes statistically-valid comparison of outcomes across centers challenging [51]. Of the 51 programs performing pediatric heart transplants, the SRTR was only able to identify a single program (with a 25 % 1-year survival rate among four patients) as having a statistically significant lower than predicted survival [52]. Thus, there are important limitations to the use of the results published in the SRTR data or program-specific reports as a measure of outcomes across centers.

Improvements in predictive mortality models will not address this fundamental challenge: low volumes of patients undergoing pediatric cardiac transplantation at individual centers makes it difficult to identify significant deviations from predicted outcomes. However, accurate estimates of risk adjusted mortality may enable identification of centers with excellent outcomes and opportunities for improvement at lower performing centers even in the absence of statistically significant variation.

Evaluation of National Policies and Rules of Allocation

The UNOS/SRTR database is particularly useful in evaluating national policies and rules of allocation because it includes all of the transplants performed in the United States of America. In contrast, analysis of the more limited (by number of centers) and broader (by country) PHTS database, may not lead to conclusions valid across the entire spectrum of centers of transplantation in the United States of America. Single center studies have even more potential for findings which cannot be generalized across the entire spectrum of transplant centers.

Criteria for listing and transplant candidacy are based on a combination of anecdotal experience, individual or consensus expert opinion, published single center results, and previous research utilizing large multi-institutional datasets. In all cases, the open nature of the UNOS dataset enables clinicians and researchers with a countervailing opinion to analyze or re – analyze the data and identify areas where current criteria are not consistent with current national experience. This availability of data provides the opportunity for evidence-based refinement.

Among the important challenges to conventional listing criteria are suggestions that a body mass index (BMI) greater than 30 kg per meter squared might be a contraindication to transplant [53], and a bias against allocation across ABO blood types in infants in UNOS heart allocation rules. Evaluation of BMI has, rather than confirming the bias against patients with BMI greater than 30 kg per meter squared, suggested that (as in other areas of cardiac surgery) the association between BMI and mortality is U-shaped. Patients at both the lowest and highest BMI are at high risk

for early mortality and those with BMI between 30 and 34.9 had mortality equivalent to those with "normal" BMI [54]. Similarly, in the context of ABO-incompatible cardiac transplantation, recent research using the UNOS database (as well as outcomes from individual centers) suggests that early outcomes are equivalent among infants with ABO-compatible allografts and ABO-incompatible allografts [55, 56]. These data have contributed to recent proposals to change UNOS rules to eliminate allocation preference given to ABO compatible allograft offers for infants awaiting cardiac transplantation [30, 57].

The broad-based nature of the UNOS/SRTR dataset may also provide a more "realistic" picture of true transplant outcomes than single-center experience. While high-volume centers have demonstrated that transplantation to patients with elevated pulmonary vascular resistance index (PVRI) as high as 9 woods units is not associated with increased mortality [58], PHTS data corroborates these findings [59]. In contrast, UNOS data suggests that among patients over 1 year of age, higher PVRI is associated with poor outcomes [60]. How to reconcile these findings? Perhaps, high volume transplant centers can lessen the risk of "high-risk" transplants, so that the findings from the high-volume centers and PHTS are real, but caution should be exercised as lower volume centers embark on "high-risk" transplants. Thus the UNOS/SRTR data provides an important counter-weight to the reporting of outcomes from only high-volume institutions. In the context of transplantation where volume and outcomes are linked (especially in high-risk patients) [24], evaluation of the broader experience is critical to defining broad criteria and truly estimating risk.

Accurate estimation of both post-transplant and waiting list risk of mortality in a national sample may also be critical in refining allocation of cardiac allografts. Currently allocation of pediatric donor hearts is predicated on reducing waitlist mortality. Factors indicating more severe cardiac failure are used as the primary criteria for listing status (Table 15.2). But when donor allografts are assigned to patients at high-risk for post-transplant mortality, this strategy may result in an overall loss of efficiency and wasting of donor organs. As stated in the OPTN Final Rule: "allocation policies ….[s]hall be designed to avoid wasting organs, to avoid futile transplants, to promote patient access to transplantation, and to promote the efficient management of organ placement [7]" (Table 15.1). In contrast to the situation with cardiac transplantation, current strategies of allocation of lungs demonstrate the potential for UNOS/SRTR data to provide support for optimizing the allocation of organs using a combination of predicted survival on the waitlist and predicted survival after transplantation in each patient.

Table 15.2 Listing status criteria (Source: http://optn.transplant.hrsa.gov/PoliciesandBylaws2/policies/pdfs/policy_9.pdf)

Status 1A
Requires assistance with a ventilator
Requires assistance with a mechanical assist device (e.g. ECMO)
Requires assistance with a balloon pump
A candidate less than 6 months old with congenital or acquired heart disease and reactive pulmonary hypertension at greater than 50 % of systemic level
Requires infusion of high dose (e.g. dobutamine>/=7.5 mcg/kg/min or milrinone>/=0.50 mcg/kg/min) or multiple inotropes
A candidate who does not meet the above criteria but has a life expectancy of less than 14 days without cardiac transplantation
Status 1B
Requires infusion of low dose single inotrope
Less than 6 months old but does not meet status 1A criteria
Growth failure (i.e. 5th percentile for weight and/or height or loss of 1.5 standard deviations of expected growth (height or weight) based on the National Center for Health Statistics for pediatric growth curves)
Status 2
All other candidates

A complete description and analysis of the development of the lung allocation score (LAS) and its advantages and disadvantages is beyond the scope of this review. In brief, the LAS combines a model predicting mortality on the waitlist with one predicting survival after transplantation in order to accomplish three goals consistent with the OPTN Final Rule [29]:

1. reduction of mortality on the lung waiting list;
2. prioritization of candidates based on urgency while avoiding futile transplants; and
3. de- emphasizing the role of waiting time and geography in lung allocation within the limits of ischemic time.

Using a similar scheme for allocation of cardiac allografts would require publication and validation of accurate models of both waitlist and post-transplant survival. Early results suggest that the current allocation scheme does result in a less than optimal allocation of organs at the highest stratum of risk [61]. More accurate models, validated in an external group, are required prior to use of modeling in allocation of hearts, but eventual use of a cardiac allocation score has the potential to increase the overall survival among children with cardiac failure.

Interpreting Data Analysis in UNOS/SRTR and PHTS Studies

Handling of Missing Data

Large datasets inevitably involve missing data. Decisions regarding the handling of missing data while analyzing these datasets have important effects on the results. While a full discussion of missing data is well beyond the scope of this chapter, it is an important topic in the context of critically evaluating literature based on the UNOS/SRTR and PHTS datasets in particular (although these issues also apply more broadly to other large dataset analyses).

Several methods of handling missing data are available. The most commonly used methods in the context of medical literature are complete case analysis and overall mean imputation. Both of these methods are easy to implement and easy to understand. They are often used without explicit description. Unfortunately, they both result in a loss in sample size, and a loss in power. More importantly, they both have the potential to result in severely biased estimates of statistical significance [62, 63].

When handling of missing values is not reported in the context of multivariable logistic regression, it implies that complete case analysis has been performed. This eliminates any records where complete data for all variables in the analysis is not available. In doing so, the sample size is severely curtailed.

Imputation as a general method is simply the replacement of a missing value with an estimate of the likely value. Mean imputation involves the replacement of missing variables with the mean result across the entire population; while simple, it relies on the assumption that the variables are missing completely at random, that is the probability of a variable being missing is unrelated to the outcome of interest or to any other variable in the dataset. This assumption is rarely true, and in the unlikely event that the assumption is met, mean imputation may still result in biased estimates [62].

Rather than estimating missing values using the mean value across the population, the missing value can be estimated using regression analysis. The main drawback of this method is that in subsequent analyses the estimated value is treated as a measured value, resulting in overestimation of the precision of subsequent analyses. Multiple imputation creates multiple datasets with estimations of the missing values and then performs the analysis in each dataset, resulting in estimates of both the statistical association and the precision of that association that more accurately reflect the error introduced by substituting estimated values for missing values [18, 62, 64].

The topic of handling of missing data is not merely a topic of esoteric statistical interest, because differences in the handling of missing data may result in significant differences in the results of any analysis. For example, two separate analyses of the UNOS database examining the impact of bicaval versus biatrial anastomosis have been performed [17, 18]. Despite using nearly identical datasets, the results were different, with the study using multiple imputations [18] able to include nearly twice as many patients in the analysis as the study using complete case analysis [17]. Similarly, research in adults looking at the impact of ventricular assist devices on post-transplant outcomes resulted in significant differences based on the effort with

which missing data was augmented by examination of free-text fields (including the potential for misspellings and typographical errors) [19]. Thus, it is important in reviewing the results of studies using large datasets to critically examine the methods used to handle missing data and the completeness with which issues of missing data are reported in the manuscript.

Accuracy of Logistic Regression Models

As noted above, evaluation of published models derived from analyses of large datasets should include a critical appraisal of the accuracy. Although a complete discussion of the evaluation of statistical models of outcomes is beyond this review, some general guidelines should be enumerated. Among the criteria that should be used to evaluate a model are:

1. estimates of global model fit such as the Bayes Information Criteria,
2. indices of discrimination (how well a model discriminates between outcomes, most commonly c-statistic), and
3. indices of calibration (how well it functions across different subgroups, most commonly the Hosmer-Lemeshow test) should be evaluated [42].

Ideally, models should be derived in one population and validated in another before becoming part of clinical practice or programmatic evaluation.

Future Directions

As data continues to be collected, multiple opportunities exist for improving the usefulness of the UNOS/SRTR and PHTS datasets, including

- improving the feedback to individual transplant centers,
- developing links between the UNOS dataset and other large datasets with complementary information, and
- tailoring certain fields of data collection to pediatric and congenital cardiac surgery.

Currently, the time delay between submission of data and analysis of data by the SRTR limits the utility of the UNOS/SRTR dataset as an ongoing tool for the assessment of quality. Improvements in modeling and in the turn-around of analytics might enable application of techniques such as cumulative sum failure analysis (CUSUM) in order to provide real-time assessment of quality of transplantation. CUSUM, which is borrowed from monitoring quality on a production line, has been used both in congenital cardiac surgery [65] and in other transplantation procedures [66], as well as in a broad swath of other medical domains [67, 68]. These techniques enable ongoing monitoring of outcomes without running into problems caused by repeatedly analyzing the same data [69]. These techniques could be implemented using the ongoing submission of data to large datasets such as UNOS and provide early feedback and warning of potential problems to transplant centers. By collecting data of specific relevance to pediatric and congenital transplantation, and developing models predicting mortality [40], the PHTS might be particularly well-suited to develop an ongoing role in the assessment of quality using these types of techniques. The possibility of such techniques being used increases with contemporary rapid increases in the power of computers and the ability to collect and analyze data.

The UNOS/SRTR database – like many large datasets – is limited by the fields of data collected. No single database can be all-inclusive, and adding fields of data eventually makes the collection of data too onerous and causes the rate of missing data to increase. Linking databases with complementary information enhances opportunities for investigation without necessitating redesigning data collection or duplicating information between multiple data submissions to different entities. For example, linkage of the STS-CHSD to other databases has already been performed [70, 71]. With regard to transplant databases, linkages between the STS-CHSD and the UNOS dataset may address weaknesses in each. Specific congenital diagnosis are missing from the UNOS/SRTR database [8], while the STS-CHSD includes diagnostic information

consistent with current standards of nomenclature. In contrast, the STS-CHSD does not contain long-term follow-up data and has no provision for follow-up of specific outcomes of transplantation (graft outcomes such as rejection and allograft vasculopathy). Linking these datasets would facilitate analyses of data not available in each dataset individually.

In addition to linking currently available datasets, some alterations to the data collected by UNOS/SRTR would improve the ability to model outcomes in pediatric and congenital patients. While PHTS already has some of these data (including previous operations and congenital diagnoses), it has been collected using differing categorization schemes over time and is inconsistent with current standardized international nomenclature in pediatric and congenital cardiac disease. Improved collection of these data would enable more powerful research into issues of specific interest to pediatric and congenital patients.

Conclusion

The historical context of the UNOS/SRTR and PHTS datasets play important roles in the analysis of outcomes following pediatric cardiac transplantation. The UNOS/SRTR Database and the PHTS Database are complementary data sources, with differing strengths and weaknesses. An understanding of the limitations of each database, as well as the limitations of various analytic techniques, is essential to a critical reading of the literature based on these sources of data. Outcomes models developed using these datasets may

- enable risk-adjusted evaluation of individual transplant centers (both for internal quality improvement and external review by the public), as well as
- facilitate optimization of rules regarding allocation of organs, guidelines for transplant candidacy, and benchmarks for high quality programs.

Future directions should include

- ongoing improvements in the outcome models,
- inclusion of currently unavailable data (either through linking of databases or enhanced collection of data), as well as
- more timely feedback to enable ongoing assessment of quality in real-time.

References

1. Davies RR, Pizarro C. Using the UNOS/SRTR and PHTS databases to improve quality in pediatric cardiac transplantation. World J Pediatr Congenit Heart Surg. 2012;3(4):421–32. doi:10.1177/2150135112443971.
2. Organ Procurement and Transplantation Network (OPTN) Data, based on OPTN data as of May 17 2013. optntransplanthrsagov. Available at: http://optn.transplant.hrsa.gov/latestData/advancedData.asp. Accessed 23 May 2013.
3. Murray JE, Merrill JP, Harrison JH, Wilson RE, Dammin GJ. Prolonged survival of human-kidney homografts by immunosuppressive drug therapy. N Engl J Med. 1963;268:1315–23. doi:10.1056/NEJM196306132682401.
4. Merrill JP, Murray JE, Harrison JH, Guild WR. Successful homotransplantation of the human kidney between identical twins. J Am Med Assoc. 1956;160(4):277–82.
5. National Organ Transplant Act. 42 U.S.C. 273 et seq; 1984.
6. OPTN Final Rule. 42 C.F.R. Part 121.
7. OPTN Final Rule. 42 C.F.R. Part 121.11 (b) (2).
8. Davies RR, Russo MJ, Yang J, Quaegebeur JM, Mosca RS, Chen JM. Listing and transplanting adults with congenital heart disease. Circulation. 2011;123(7):759–67. doi:10.1161/CIRCULATIONAHA.110.960260.
9. Karamlou T, Hirsch J, Welke K, et al. A United Network for Organ Sharing analysis of heart transplantation in adults with congenital heart disease: outcomes and factors associated with mortality and retransplantation. J Thorac Cardiovasc Surg. 2010;140(1):161–8. doi:10.1016/j.jtcvs.2010.03.036.
10. Davies RR, Russo MJ, Mital S, et al. Predicting survival among high-risk pediatric cardiac transplant recipients: an analysis of the United Network for Organ Sharing database. J Thorac Cardiovasc Surg. 2008;135(1):147–55, 155.e1–2. doi:10.1016/j.jtcvs.2007.09.019.
11. Chen JM, Davies RR, Mital SR, et al. Trends and outcomes in transplantation for complex congenital heart disease: 1984 to 2004. Ann Thorac Surg. 2004;78(4):1352–61. doi:10.1016/j.athoracsur.2004.04.012; discussion 1352–61.
12. Lytrivi ID, Blume ED, Rhodes J, Dillis S, Gauvreau K, Singh TP. Prognostic value of exercise testing during heart transplant evaluation in children. Circ Heart Fail. 2013;6(4):792–9. doi:10.1161/CIRCHEARTFAILURE.112.000103.

13. Kanter KR, Mahle WT, Vincent RN, Berg AM, Kogon BE, Kirshbom PM. Heart transplantation in children with a Fontan procedure. Ann Thorac Surg. 2011;91(3):823–9. doi:10.1016/j.athoracsur.2010.11.031; discussion 829–30.
14. Bernstein D, Naftel DC, Chin C, et al. Outcome of listing for cardiac transplantation for failed Fontan: a multi-institutional study. Circulation. 2006;114(4):273–80.
15. Khairy P, Fernandes SM, Mayer JE, et al. Long-term survival, modes of death, and predictors of mortality in patients with Fontan surgery. Circulation. 2008;117(1):85–92. doi:10.1161/CIRCULATIONAHA.107.738559.
16. Davies RR, Sorabella RA, Yang J, Mosca RS, Chen JM, Quaegebeur JM. Outcomes after transplantation for "failed" Fontan: a single-institution experience. J Thorac Cardiovasc Surg. 2012;143(5):1183–92.e4. doi:10.1016/j.jtcvs.2011.12.039.
17. Weiss ES, Nwakanma LU, Russell SB, Conte JV, Shah AS. Outcomes in bicaval versus biatrial techniques in heart transplantation: an analysis of the UNOS database. J Heart Lung Transplant. 2008;27(2):178–83. doi:10.1016/j.healun.2007.11.003.
18. Davies RR, Russo MJ, Morgan JA, Sorabella RA, Naka Y, Chen JM. Standard versus bicaval techniques for orthotopic heart transplantation: an analysis of the United Network for Organ Sharing database. J Thorac Cardiovasc Surg. 2010;140(3):700–8, 708.e1–2. doi:10.1016/j.jtcvs.2010.04.029.
19. Russo MJ, Hong KN, Davies RR, et al. Posttransplant survival is not diminished in heart transplant recipients bridged with implantable left ventricular assist devices. J Thorac Cardiovasc Surg. 2009;138(6):1425–32.e1–3. doi:10.1016/j.jtcvs.2009.07.034.
20. Bull DA, Reid BB, Selzman CH, et al. The impact of bridge-to-transplant ventricular assist device support on survival after cardiac transplantation. J Thorac Cardiovasc Surg. 2010;140(1):169–73. doi:10.1016/j.jtcvs.2010.03.026.
21. Hsu DT, Naftel DC, Webber SA, et al. Lessons learned from the pediatric heart transplant study. Congenit Heart Dis. 2006;1(3):54–62. doi:10.1111/j.1747-0803.2006.00011.x.
22. Pediatric Heart Transplant Study. University of Alabama – Birmingham. Available at: http://www.uab.edu/ctsresearch/phts/archivedforms.html. Accessed Dec 2011.
23. Pediatric Heart Transplant Study (PHTS) – forms. University of Alabama (UAB). Available at: http://www.uab.edu/ctsreaserch/phts/forms.html. Accessed 1 Sept 2011.
24. Davies RR, Russo MJ, Hong KN, et al. Increased short- and long-term mortality at low-volume pediatric heart transplant centers: should minimum standards be set? Retrospective data analysis. Ann Surg. 2011;253(2):393–401. doi:10.1097/SLA.0b013e31820700cc.
25. International Society for Heart and Lung Transplantation. ISHLT registries – heart/lung registries. ishltorg. Available at: http://www.ishlt.org/registries/heartLungRegistry.asp. Accessed 24 May 2013.
26. CMS Centers for Medicare and Medicaid Services. National coverage determination. 5th ed. 2010. Available at: http://www.cmms.hhs.gov/medicare-coverage-database.
27. Blume ED, Naftel DC, Bastardi HJ, et al. Outcomes of children bridged to heart transplantation with ventricular assist devices: a multi-institutional study. Circulation. 2006;113(19):2313–9.
28. Williams ML, Trivedi JR, McCants KC, et al. Heart transplant vs left ventricular assist device in heart transplant-eligible patients. Ann Thorac Surg. 2011;91(5):1330–3. doi:10.1016/j.athoracsur.2011.01.062; discussion 1333–4.
29. Egan TM, Murray S, Bustami RT, et al. Development of the new lung allocation system in the United States. Am J Transplant. 2006;6(5 Pt 2):1212–27. doi:10.1111/j.1600-6143.2006.01276.x.
30. Everitt MD, Donaldson AE, Casper TC, et al. Effect of ABO-incompatible listing on infant heart transplant waitlist outcomes: analysis of the United Network for Organ Sharing (UNOS) database. J Heart Lung Transplant. 2009;28(12):1254–60. doi:10.1016/j.healun.2009.06.024.
31. Pietra BA, Kantor PF, Bartlett HL, et al. Early predictors of survival to and after heart transplantation in children with dilated cardiomyopathy. Circulation. 2012. doi:10.1161/CIRCULATIONAHA.110.011999.
32. Singh TP, Almond CS, Piercey G, Gauvreau K. Trends in wait-list mortality in children listed for heart transplantation in the United States: era effect across racial/ethnic groups. Am J Transplant. 2011;11(12):2692–9. doi:10.1111/j.1600-6143.2011.03723.x.
33. Almond CSD, Thiagarajan RR, Piercey GE, et al. Waiting list mortality among children listed for heart transplantation in the United States. Circulation. 2009;119(5):717–27. doi:10.1161/CIRCULATIONAHA.108.815712.
34. Davies RR, Russo MJ, Reinhartz O, et al. 483 Lower socioeconomic status predicts poor waitlist and post-heart transplant survival in children. J Heart Lung Transplant. 2011;30(4S):S164.
35. Singh TP, Almond CS, Taylor DO, Graham DA. Decline in heart transplant wait list mortality in the United States following broader regional sharing of donor hearts. Circ Heart Fail. 2012;5(2):249–58. doi:10.1161/CIRCHEARTFAILURE.111.964247.
36. Davies RR, Russo MJ, Hong KN, et al. The use of mechanical circulatory support as a bridge to transplantation in pediatric patients: an analysis of the United Network for Organ Sharing database. J Thorac Cardiovasc Surg. 2008;135(2):421–7, 427.e1. doi:10.1016/j.jtcvs.2007.09.048.
37. Singh TP, Gauvreau K, Thiagarajan R, Blume ED, Piercey G, Almond CS. Racial and ethnic differences in mortality in children awaiting heart transplant in the United States. Am J Transplant. 2009;9(12):2808–15. doi:10.1111/j.1600-6143.2009.02852.x.

38. Kirk R, Naftel D, Hoffman TM, et al. Outcome of pediatric patients with dilated cardiomyopathy listed for transplant: a multi-institutional study. J Heart Lung Transplant. 2009;28(12):1322–8. doi:10.1016/j.healun.2009.05.027.
39. Davies RR. Multiple high risk criteria predict poor survival among pediatric heart transplant recipients. J Heart Lung Transplant. 2008;27(2):255–6.
40. Naftel DC. Fourteen years of improving results illustrated by patient specific predictions. J Heart Lung Transplant. 2008;27(2):253–4.
41. Auerbach SR, Richmond ME, Chen JM, et al. Multiple risk factors before pediatric cardiac transplantation are associated with increased graft loss. Pediatr Cardiol. 2011;32(5):615–20. doi:10.1007/s00246-011-0077-7.
42. Cook NR. Use and misuse of the receiver operating characteristic curve in risk prediction. Circulation. 2007;115(7):928–35. doi:10.1161/CIRCULATIONAHA.106.672402.
43. Weiss ES, Allen JG, Arnaoutakis GJ, et al. Creation of a quantitative recipient risk index for mortality prediction after cardiac transplantation (IMPACT). Ann Thorac Surg. 2011;92(3):914–21. doi:10.1016/j.athoracsur.2011.04.030; discussion 921–2.
44. Wendt D, Osswald BR, Kayser K, et al. Society of Thoracic Surgeons score is superior to the EuroSCORE determining mortality in high risk patients undergoing isolated aortic valve replacement. Ann Thorac Surg. 2009;88(2):468–74. doi:10.1016/j.athoracsur.2009.04.059; discussion 474–5.
45. Murtuza B, Fenton M, Burch M, et al. Pediatric heart transplantation for congenital and restrictive cardiomyopathy. Ann Thorac Surg. 2013. doi:10.1016/j.athoracsur.2013.01.014.
46. Rajagopal SK, Yarlagadda VV, Thiagarajan RR, Singh TP, Givertz MM, Almond CSD. Pediatric heart failure and worsening renal function: association with outcomes after heart transplantation. J Heart Lung Transplant. 2011. doi:10.1016/j.healun.2011.08.018.
47. Joffe AR, Quiñonez LG, Robertson CMT, et al. Outcomes after heart transplantation in children under six years of age. Ann Thorac Surg. 2011;92(1):174–82. doi:10.1016/j.athoracsur.2011.02.038.
48. Dipchand A, Cecere R, Delgado DH, et al. Canadian Consensus on cardiac transplantation in pediatric and adult congenital heart disease patients 2004: executive summary. Can J Cardiol. 2005;21(13):1145–7.
49. Kirk RC, Edwards LB, Aurora P, et al. Registry of the International Society for Heart and Lung Transplantation: eleventh official pediatric heart transplantation report–2008. J Heart Lung Transplant. 2008;27(9):970–7. doi:10.1016/j.healun.2008.06.016.
50. Singh TP, Edwards LB, Kirk R, Boucek MM. Era effect on post-transplant survival adjusted for baseline risk factors in pediatric heart transplant recipients. J Heart Lung Transplant. 2009;28(12):1285–91. doi:10.1016/j.healun.2009.05.003.
51. Jacobs JP, O'Brien SM, Pasquali SK, et al. Variation in outcomes for benchmark operations: an analysis of the Society of Thoracic Surgeons Congenital Heart Surgery Database. Ann Thorac Surg. 2011;92(6):2184–91. doi:10.1016/j.athoracsur.2011.06.008; discussion 2191–2.
52. Scientific Registry of Transplant Recipients. Available at: http://www.srtr.org/csr/current/Centers/TransplantCenters_New201106x.aspx?organcode=HR. Accessed 7 Sept 2011.
53. Mehra MR, Kobashigawa J, Starling R, et al. Listing criteria for heart transplantation: International Society for Heart and Lung Transplantation guidelines for the care of cardiac transplant candidates–2006. J Heart Lung Transplant. 2006;25(9):1024–42. doi:10.1016/j.healun.2006.06.008.
54. Russo MJ, Hong KN, Davies RR, et al. The effect of body mass index on survival following heart transplantation: do outcomes support consensus guidelines? Ann Surg. 2010;251(1):144–52. doi:10.1097/SLA.0b013e3181b5db3c.
55. Dipchand AI, Pollock BarZiv SM, Manlhiot C, West LJ, VanderVliet M, McCrindle BW. Equivalent outcomes for pediatric heart transplantation recipients: ABO-blood group incompatible versus ABO-compatible. Am J Transplant. 2010;10(2):389–97. doi:10.1111/j.1600-6143.2009.02934.x.
56. Patel ND, Weiss ES, Scheel J, Cameron DE, Vricella LA. ABO-incompatible heart transplantation in infants: analysis of the united network for organ sharing database. J Heart Lung Transplant. 2008;27(10):1085–9. doi:10.1016/j.healun.2008.07.001.
57. Almond CS, Almond CS, Gauvreau K, et al. Impact of ABO-incompatible listing on wait-list outcomes among infants listed for heart transplantation in the United States: a propensity analysis. Circulation. 2010;121(17):1926–33. doi:10.1161/CIRCULATIONAHA.109.885756.
58. Chiu P, Russo MJ, Davies RR, Addonizio LJ, Richmond ME, Chen JM. What is high risk? Redefining elevated pulmonary vascular resistance index in pediatric heart transplantation. J Heart Lung Transplant. 2012;31(1):61–6. doi:10.1016/j.healun.2011.08.021.
59. Richmond ME, Law YM, Das B, Everitt MD. Elevated pre-transplant pulmonary vascular resistance is not associated with mortality in children without congenital heart disease: a multi-center study. J Heart Lung Transplant. 2013;31(4 Suppl):S35.
60. Buddhe S, Du W, L'ecuyer T. Impact of pulmonary hypertension on transplant outcomes in pediatric cardiomyopathy patients. Pediatr Transplant. 2012;16(4):367–72. doi:10.1111/j.1399-3046.2012.01678.x.
61. Singh TP, Almond CS, Piercey G, Gauvreau K. Risk stratification and transplant benefit in children listed for heart transplant in the United States. Circ Heart Fail. 2013;6(4):800–8. doi:10.1161/CIRCHEARTFAILURE.112.000280.
62. Donders ART, van der Heijden GJMG, Stijnen T, Moons KGM. Review: a gentle introduction to imputation of missing values. J Clin

Epidemiol. 2006;59(10):1087–91. doi:10.1016/j.jclinepi.2006.01.014.
63. Greenland S, Finkle WD. A critical look at methods for handling missing covariates in epidemiologic regression analyses. Am J Epidemiol. 1995;142(12):1255–64.
64. Rubin D. Multiple imputation for non-response in surveys. New York: Wiley; 1987.
65. de Leval MR, Francois K, Bull C, Brawn W, Spiegelhalter D. Analysis of a cluster of surgical failures. Application to a series of neonatal arterial switch operations. J Thorac Cardiovasc Surg. 1994;107(3):914–23; discussion 923–4.
66. Axelrod DA, Guidinger MK, Metzger RA, Wiesner RH, Webb RL, Merion RM. Transplant center quality assessment using a continuously updatable, risk-adjusted technique (CUSUM). Am J Transplant. 2006;6(2):313–23. doi:10.1111/j.1600-6143.2005.01191.x.
67. Holzhey DM, Jacobs S, Walther T, Mochalski M, Mohr FW, Falk V. Cumulative sum failure analysis for eight surgeons performing minimally invasive direct coronary artery bypass. J Thorac Cardiovasc Surg. 2007;134(3):663–9. doi:10.1016/j.jtcvs.2007.03.029.
68. Bolsin S, Colson M. The use of the Cusum technique in the assessment of trainee competence in new procedures. Int J Qual Health Care. 2000;12(5):433–8.
69. McPherson K. Statistics: the problem of examining accumulating data more than once. N Engl J Med. 1974;290(9):501–2. doi:10.1056/NEJM197402282900907.
70. Jacobs JP, Haan CK, Edwards FH, et al. The rationale for incorporation of HIPAA compliant unique patient, surgeon, and hospital identifier fields in the STS database. Ann Thorac Surg. 2008;86(3):695–8. doi:10.1016/j.athoracsur.2008.04.075.
71. Pasquali SK, Jacobs JP, Shook GJ, et al. Linking clinical registry data with administrative data using indirect identifiers: implementation and validation in the congenital heart surgery population. Am Heart J. 2010;160(6):1099–104. doi:10.1016/j.ahj.2010.08.010.

Databases for Extracorporeal Membrane Oxygenation and Ventricular Assist Devices

16

David S. Cooper, David L.S. Morales, Megan del Corral, Matthew L. Paden, and Ravi R. Thiagarajan

Abstract

The most critically ill patients can now be treated with life saving mechanical circulatory support. The development of databases to track the outcomes of patients with critical illness has been ongoing for much of the last two decades. Clinicians who engage in the use of mechanical circulatory support are interested in the analysis of outcomes of patients treated with Extracorporeal Membrane Oxygenation (ECMO) and Ventricular Assist Devices (VAD), with the ultimate aim of improvement in the quality of care.

Keywords

Ventricular assist device • Extracorporeal Membrane Oxygenation • ECMO • VAD • Database • Registry • Quality improvement

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Introduction

The most critically ill patients can now be treated with mechanical circulatory support. Extracorporeal life support using Extracorporeal Membrane Oxygenation (ECMO) is used to provide cardiopulmonary support in patients with life threatening cardiorespiratory failure unresponsive to conventional medical therapies. Mechanical circulatory support using Ventricular Assist Devices (VAD) is used to provide cardiovascular support in patients with end stage heart failure refractory to conventional medical therapies. The development of databases to track the outcomes of patients with critical illness has been ongoing for much of the last two decades, paralleled by the increasing awareness of the impact on outcomes of patients obtained from programs designed to improve quality and enhance the safety of patients. Clinicians who engage in the use of mechanical circulatory support are interested in the analysis of outcomes of patients treated with ECMO and VAD with the ultimate aim of improvement in the quality of care provided to these patients.

The Extracorporeal Life Support Organization (ELSO) is an international consortium of health care professionals and scientists who are dedicated to the development and evaluation of novel therapies for support of failing organ systems. Since 1989, ELSO has maintained a registry of the use of ECMO in active ELSO centers. The Interagency Registry for Mechanically Assisted Circulatory Support (INTERMACS) is a national registry in the United States of America for patients who are receiving mechanical circulatory support device therapy to treat advanced cardiac failure. This registry was devised as a joint effort of the National Heart, Lung and Blood Institute (NHLBI), the Centers for Medicare and Medicaid Services (CMS), the Food and Drug Administration (FDA), clinicians, scientists, and industry representatives in conjunction with the University of Alabama at Birmingham (UAB) and United Network for Organ Sharing (UNOS).

This Chapter will focus on databases used to track outcomes of critically ill patients treated with ECMO and VAD. This review discusses the historical aspects, current state of analysis of outcomes, and the potential for transitioning from acquisition of data to improvement of quality and enhanced safety in the care of these patients.

ELSO Registry

Extracorporeal Membrane Oxygenation

Extracorporeal life support using Extracorporeal Membrane Oxygenation (ECMO) is used to provide cardiopulmonary support in patients with life threatening cardiorespiratory failure unresponsive to conventional medical therapies [1]. ECMO can be lifesaving in these patients as they face imminent mortality without mechanical circulatory and/or respiratory support. Although lifesaving for many patients, ECMO does not treat primary illness that caused cardiopulmonary failure and is merely a modality of cardiopulmonary support for providing perfusion to end-organs while awaiting recovery of the primary condition or disease causing cardiopulmonary failure. Thus, outcomes following ECMO use are largely influenced by the prognosis for the primary disease (Table 16.1) [2]. Use of ECMO, indications for ECMO, and number of centers providing ECMO support have increased

Table 16.1 Indications and survival following ECMO use based on the international summary of the Extracorporeal Life Support Organization's ECMO Registry, July 2013

ECMO indication	Total patients	Survived to discharge (%)
Neonatal ECMO		
Respiratory	26,583	75
Cardiac	5,159	40
ECPR	914	39
Pediatric		
Respiratory	5,923	57
Cardiac	6,459	49
ECPR	1,878	41
Adult		
Respiratory	4,382	56
Cardiac	3,401	40
ECPR	969	28

ECPR = ECMO to support cardiopulmonary resuscitation

over the last decade. Even though ECMO offers potentially lifesaving support for many patients, many issues exist regarding the use of ECMO that require careful consideration:

- First, despite increasing use, greater experience, and newer and improved equipment and technology, mortality following ECMO support is high and has largely remained unchanged over decades.
- Second, considerable variability exists in many areas in the clinical practice of ECMO [3]. These areas include selection of patients for ECMO, timing of ECMO deployment, equipment used to provide ECMO, practice of anticoagulation while on ECMO, management of patients on ECMO, and weaning from ECMO.
- Third, safe and efficient use of ECMO requires easy availability of equipment and personnel to deploy and manage ECMO 24 hours a day, resulting in the need for a large amount of resources and expenditure [4].
- Finally, growing evidence shows that survival after ECMO is related to the characteristics of the center providing ECMO, such as volume of patients managed in the center [5, 6].

These issues have raised the need for tools to assess the quality of care provided to patients supported with ECMO and to evaluate of "best" ECMO practices. One resource available for improving care provided to ECMO patients is the ECMO registry of the Extracorporeal Life Support Organization (ELSO). The following paragraphs describe the history, details of the registry, and potential for its use for purposes of assessment of quality.

ECMO Registry of ELSO

The ECMO registry of ELSO is a data registry that collects information about ECMO across all age groups and for all ECMO indications. ELSO was formed in 1989 to improve knowledge and clinical use of Extracorporeal Life Support therapies [7, 8]. The organization's data registry contains data on patients dating as far back as 1976 and has played a pivotal role in our current understanding and practice of ECMO support. Currently over 200 International centers submit data to the registry. Membership in ELSO is required for submitting data to the registry. Data collected include:

- demographics of patients supported with ECMO,
- condition of the patient prior to ECMO,
- support of the patient prior to ECMO,
- diagnosis (using International classification of Disease; ICD-9 codes),
- procedures performed prior to and during ECMO (Common Procedural Terminology; CPT and Unique ELSO registry codes),
- details about ECMO equipment,
- duration of ECMO support,
- complications acquired during ECMO (using unique ELSO Registry codes), and
- Information about survival including survival to weaning off ECMO and survival to discharge from the hospital.

Data are submitted using standardized data collection forms. Recent changes to the process of submission of data include using a secure, web-based system of entry of data. As with other databases and registries, submission of data to ELSO requires approval by the local institutional review board at member centers. A data user agreement allows release of de-identified data to member centers for purposes of benchmarking and scientific research.

Uses for ECMO Registry Data

An important function of ELSO is maintaining a data registry for purposes of

- improving outcomes of patients supported with ECMO,
- reducing morbidity associated with ECMO, and
- fostering innovations both in the care of patients supported with ECMO and the technology associated with ECMO.

The following paragraphs describe the uses of data from the ELSO registry of these purposes.

(a) *Registry Report*: The Registry provides a yearly summary report of data contained in

the registry and has periodically published a registry report containing detailed information on trends in use of ECMO and outcomes associated with ECMO (Table 16.1) [2]. These reports serve as valuable resources for providers of ECMO and ECMO centers for

- assessment of need,
- programmatic planning, and
- identifying areas for improvement related to ECMO.

(b) *Scientific Research*: The registry provides investigators limited de-identified datasets for purposes of scientific inquiry and research. Areas of scientific research [9–15] have included

- indications for ECMO,
- analysis of outcomes stratified by specific disease or procedure,
- complications associated with ECMO, and
- assessment of outcomes associated with various technologies (e.g. type of pump and outcomes).

Many examples of these publications can be searched using commonly available search engines such as PubMed.

(c) *Data for regulatory use*: The registry collects detailed information on the various types of equipment associated with ECMO. These data can be used to serve as preliminary data for planning clinical trials, or used as comparative data for studying safety and efficacy of devices related to ECMO. One recent example is the use of data from the ELSO registry as a comparative cohort for the recently concluded trial of the Berlin Heart EXCOR Pediatric Ventricular Assist Device conducted by the Food and Drug Administration (FDA) of the United States of America [16].

(d) *Benchmarking and Assessment of Quality*: In addition to the detailed international summary containing a summarized data from all centers, member centers contributing data to ELSO are provided with a center specific data describing specific information on use of ECMO, complications associated with ECMO, and outcomes. These center specific reports categorize outcomes and complications based on indications for ECMO. Given that rates of survival after ECMO vary widely by indication for ECMO, the availability of outcomes and complications categorized by indications in the international summary and center specific reports allows center specific benchmarking of outcomes and complications without detailed risk adjustment. Center specific reports provide opportunities for a specific ECMO program to set goals for future improvement in performance.

(e) *Limitations of the ELSO registry data*: Several limitations of the data reported to the registry should be carefully considered when using these data for purposes of research and benchmarking.

- The registry does not have a robust process of verification of data to ensure the completeness and accuracy of the submitted data. The move towards web-based submission of data has allowed implementation customization of fields of data that can help assure the integrity of the submitted data and improve the validity of the submitted data.
- A robust and detailed methodology of risk adjustment is not available to facilitate comparison of center specific outcomes. Even though comparability is improved based on categorization by indication for ECMO, outcomes associated with ECMO vary widely based on diagnosis within a category, and a more advanced and detailed methodology of risk adjustment may provide improved comparability of programmatic performance (Table 16.2).
- Functional outcome after ECMO and data about quality of life are not available to improve our understanding of these important issues in survivors of ECMO.

In summary, the ECMO registry of ELSO has played an important role in our understanding of use of ECMO and outcomes associated with ECMO. It remains an important resource for benchmarking and improvement of quality for ECMO programs worldwide.

Table 16.2 The impact of diagnosis category on survival for neonates using ECMO for respiratory indications from the International Summary of the Extracorporeal Life Support Organization's ECMO Registry, July 2013

Diagnosis category	No: of patients	Survival to discharge (%)
Meconium aspiration syndrome	8,423	94
Persistent pulmonary hypertension (PPHN)	4,526	77
Respiratory distress syndrome	1,541	84
Sepsis	2,809	73
Pneumonia	364	58
Congenital diaphragmatic hernia	6,845	51
Air leak syndrome	126	73
Not categorized by diagnosis	2,318	61

Ventricular Assist Device Database

The Food and Drug Administration (FDA) entered into the arena of mechanical circulatory support devices (MCSDs) in the late 1970s with the intention to create methods of regulation of devices. The development use of MCSDs flourished, and the Institute of Medicine issued a report in 1991 stating that patients implanted with MCSDs should be followed through a registry for the remainder of their lives, recommending that this initiative be spearheaded by the National Heart, Lung & Blood Institute (NHLBI). In June of 2006, the Interagency Registry for Mechanically Assisted Circulatory Support (INTERMACS) was established as a joint effort between the NHLBI, Centers for Medicare and Medicaid Services (CMS), FDA, representatives from industry, scientists, and clinicians. The goal of this partnership was to provide contemporary data for patients who are receiving mechanical circulatory support device therapy as a treatment of advanced cardiac failure. This collaboration established a concerted effort by physicians and partners from industry

- to define adverse events precisely,
- to provide information that would allow optimal MCSD-patient matching,
- to move devices from investigational device exemption (IDE) trials into post-market approval for clinical use, and
- to create a meaningful system of classification relevant to selection of patients for support with MCSDs.

INTERMACS is the North American prospective registry utilized to collect clinical data to demonstrate outcomes over time to [17]:

1. Facilitate the refinement of selection of patients to maximize outcomes with current and new devices
2. Identify predictors of good outcomes as well as risk factors for adverse events after implantation of devices
3. Develop consensus "best practice" guideline to improve clinical management by reducing short-term and long-term complications of therapy with mechanical circulatory support devices
4. Utilize information from the INTERMACS Registry to guide improvements in technology, particularly as next generation devices evolve
5. Guide clinical testing and approval of new devices.

Information in the database includes

- data about the profile of patients,
- data about the implantation of devices,
- defined follow-up information at scheduled intervals, and
- event-driven data.

Key details that are captured by the INTERMACS registry include

- the quality of life of the patient,
- the level of function of the patient,
- death,
- occurrence of explantation of the device,
- occurrence of transplantation, and
- adverse events as defined by the registry.

These indices have become essential to the evaluation of therapy with MCSDs and the improvement of survival and functional status of the patients.

The treatment and management of advanced cardiac failure has matured over the past several years with the increased use of second and third generation mechanical circulatory support devices

and subsequent crossover into the realm of pediatrics. Accordingly, a multidisciplinary team of clinicians with expertise in pediatric cardiac failure, including cardiac surgeons led the development of the pediatric arm of the INTERMACS registry. This pediatric arm of INTERMACS, entitled pediMACS, was launched in September of 2012 with the aim to mirror the goals and expected analyses of the main INTERMACS registry, but in patients less than 19 years old at the time of implant. This new registry would contain broader data about implants, to include both durable and temporary support devices. Other subtle variances include modifications to the existing definitions of adverse events and expansion of the instruments utilized to assess quality of life. PediMACS will additionally evaluate therapy with MCSDs in the pediatric population by focusing on

- the differences in devices available for implantation, and
- the complexity surrounding the selection of both the types of devices and the appropriate children for therapy with these devices.

These data are essential for measuring outcomes of patients and evaluating new cutting edge pediatric-specific devices.

Adverse events are clearly defined by INTERMACS and pediMACS and are captured by two mechanisms:

1. The occurrence of infection, device failure, neurological injury, death (INTERMACS only) and bleeding (pediMACS only) trigger additional elements of data to be obtained; and
2. Other adverse events that have been identified are routinely collected at defined follow-up intervals. Other adverse events captured include.

- cardiac arrhythmias,
- failure of the right heart,
- arterial thromboembolic events outside of the central nervous system,
- hypertension,
- pericardial fluid collection,
- myocardial infarction,
- venous thromboembolism,
- wound dehiscence,
- renal dysfunction,
- hemolysis,
- hepatic dysfunction, and
- respiratory failure.

Quality of life assessments are also obtained from patients, parents, and caregivers at predetermined intervals (pre-implant, 3 months, 6 months, and every 6 months thereafter), with both registries utilizing uniform tools (i.e. EQ-5d and the Kansas City Cardiomyopathy Questionnaire [KCCQ]).

The acquisition of timely data is the driving force behind the pediMACS and INTERMACS registries. This information has been utilized in

- obtaining approval from the FDA,
- providing comparison arms in clinical trials,
- comparing medical therapy and therapy with MCSDs as treatment for cardiac failure, and
- creating a true venue for debate.

As the registry moves forward, these pertinent data will continue to identify risk stratifying factors and predictors for morbidity and mortality and then aid in the formulation of novel methods and strategies of care to minimize these risks. INTERMACS completed an analysis of the first full year of data on FDA-approved durable VADs in 2008 that represented 511 patients from 75 institutions [17]. Comparably, the fifth annual report was published in 2013, which summarizes and analyzes the first 6 years of collection of data, with more than 6,000 patients enrolled.

Since the inception of the INTERMACS database, it has generated useful information for a multitude of centers and has provided data for

- publications about MCSDs,
- review of outcomes, and
- programs to improve quality.

The data learned from the registry has been essential in addressing several key questions in the field and providing risk stratification (i.e. device related infection, pump thrombosis, failure of the right heart, etc.) as well as publishing rates of survival of patients having been implanted with durable VADs. The futures for the INTERMACS and pediMACS databases are to not be geographically limited. The leadership of the International Society for Heart and Lung Transplantation (ISHLT) is aiming to collaborate and consolidate European and Asian databases

(i.e.: EuroMACS, J-MACS) into a single database named the ISHLT Mechanical Assisted Circulatory Support Registry (IMACS) [18].

Conclusions

A great deal has already been accomplished to standardize and improve methodologies for the analysis of outcomes following the treatment of patients with ECMO and VAD. While these achievements have laid the groundwork, much remains to be accomplished. We can, and should, rise to the challenge, by more effectively:

- defining and measuring outcomes,
- setting standards to benchmark results, and
- using these data to change and improve upon our current practice and these results.

International efforts to standardize definitions of complications relating to perfusion and extracorporeal circulation associated with the treatment of patients with congenital cardiac disease has led to the publication of "*Consensus Definitions from the Multi-Societal Database Committee for Pediatric and Congenital Heart Disease*" [19, 20]. These publications define multiple terms related to ECMO and VAD:

- "Cardiopulmonary bypass is defined as the process of diverting venous blood from a patient's heart and lungs to a gas exchange system for the addition of oxygen, removal of carbon dioxide, and subsequent re-infusion to the patient's arterial system."
- "Extracorporeal membrane oxygenation is defined as the process of diverting venous blood from a patient to a gas exchange system for the addition of oxygen, removal of carbon dioxide, and subsequent re-infusion to the patient's arterial or venous system."
- "A mechanical circulatory support device is defined as a pump or apparatus that augments or replaces the function of the failing heart. Two types of mechanical circulatory support devices are ventricular assist devices and intra-aortic balloon pumps."

A non-comprehensive listing of areas in need of future development includes:

1. Standardizing and unifying the tools for stratification of complexity
2. Improving the tools for stratification of complexity in order to account for patient-specific variables
3. Creating methodologies for analysis beyond mortality as an endpoint
4. Improving methodologies for verification of data
5. Establishing links between databases
6. Standardizing long term follow-up

Only through a commitment to transitioning from data collection and analysis to continuous quality improvement can we leverage these data to achieve the optimal outcomes for our patients and their families.

References

1. Gaffney AM, Wildhirt SM, Griffin MJ, et al. Extracorporeal life support. BMJ. 2010;341:c5317.
2. International Summary of The Extracorporeal Life Support Organization. [www.elsonet.org].
3. Bembea MM, Annich G, Rycus P, et al. Variability in anticoagulation management of patients on extracorporeal membrane oxygenation: an international survey. Pediatr Crit Care Med. 2013;14(2):e77–84.
4. Vats A, Pettignano R, Culler S, Wright J. Cost of extracorporeal life support in pediatric patients with acute respiratory failure. Crit Care Med. 1998;26(9):1587–92.
5. Freeman CL, Bennett TD, Casper TC, et al. Pediatric and neonatal extracorporeal membrane oxygenation: does center volume impact mortality? Crit Care Med. 2013;42(3):1–8.
6. Karamlou T, Vafaeezadeh M, Parrish AM, et al. Increased extracorporeal membrane oxygenation center case volume is associated with improved extracorporeal membrane oxygenation survival among pediatric patients. J Thorac Cardiovasc Surg. 2013;145(2):470–5.
7. Conrad SA, Rycus PT. The Registry of the Extracorporeal Life Support Organization. In: Annich GM, Lynch WR, Maclaren G, Wilson JM, Bartlett RH, editors. ECMO: extracorporeal cardiopulmonary support in critical care. 4th ed. Ann Arbor: The Extracorporeal Life Support Organization; 2012. p. 87–104.
8. Paden ML, Conrad SA, Rycus PT, Thiagarajan RR. Extracorporeal Life Support Organization Registry Report 2012. ASAIO J. 2012;59(3):202–10.
9. Barrett CS, Jaggers JJ, Cook EF, et al. Outcomes of neonates undergoing extracorporeal membrane oxygenation support using centrifugal versus roller blood pumps. Ann Thorac Surg. 2012;94(5):1635–41.

10. Barrett CS, Jaggers JJ, Cook EF, et al. Pediatric ECMO outcomes: comparison of centrifugal versus roller blood pumps using propensity score matching. ASAIO J. 2013;59(2):145–51.
11. Fleming GM, Gurney JG, Donohue JE, et al. Mechanical component failures in 28,171 neonatal and pediatric extracorporeal membrane oxygenation courses from 1987 to 2006. Pediatr Crit Care Med. 2009;10(4):439–44.
12. Polito A, Barrett CS, Wypij D, et al. Neurologic complications in neonates supported with extracorporeal membrane oxygenation. An analysis of ELSO registry data. Intensive Care Med. 2013;39(9):1594–601.
13. Rollins MD, Hubbard A, Zabrocki L, et al. Extracorporeal membrane oxygenation cannulation trends for pediatric respiratory failure and central nervous system injury. J Pediatr Surg. 2012;47(1):68–75.
14. Zabrocki LA, Brogan TV, Statler KD, et al. Extracorporeal membrane oxygenation for pediatric respiratory failure: survival and predictors of mortality. Crit Care Med. 2011;39(2):364–70.
15. Thiagarajan RR, Laussen PC, Rycus PT, et al. Extracorporeal membrane oxygenation to aid cardiopulmonary resuscitation in infants and children. Circulation. 2007;116(15):1693–700.
16. Fraser Jr CD, Jaquiss RD, Rosenthal DN, et al. Prospective trial of a pediatric ventricular assist device. N Engl J Med. 2012;367(6):532–41.
17. Kirklin JK, Naftel DC, Stevenson LW, et al. INTERMACS database for durable devices for circulatory support: first annual report. J Heart Lung Transplant. 2008;27(10):1065–73.
18. Holman WL. Interagency Registry for Mechanically Assisted Circulatory Support (INTERMACS): what have we learned and what will we learn? Circulation. 2012;126(11):1401–6.
19. Shann KG, Giacomuzzi CR, Harness L, Myers GJ, Paugh TA, Mellas N, Groom RC, Gomez D, Thuys CA, Charette K, Ojito JW, Tinius-Juliani J, Calaritis C, McRobb CM, Parpard M, Chancy T, Bacha E, Cooper DS, Jacobs JP, Likosky DS, on behalf of the International Consortium for Evidence-Based Perfusion. Complications relating to perfusion and extracorporeal circulation associated with the treatment of patients with congenital cardiac disease: consensus definitions from the Multi-Societal Database Committee for Pediatric and Congenital Heart Disease. In: 2008 supplement to Cardiology in the Young: databases and the assessment of complications associated with the treatment of patients with congenital cardiac disease, prepared by: The Multi-Societal Database Committee for Pediatric and Congenital Heart Disease, Jeffrey P. Jacobs, MD, editor. Cardiol Young. 2008;18(Suppl 2):206–14.
20. Jacobs JP, editor. 2008 Supplement to Cardiology in the Young: databases and the assessment of complications associated with the treatment of patients with congenital cardiac disease, prepared by: The Multi-Societal Database Committee for Pediatric and Congenital Heart Disease. Cardiol Young. 2008;18(Suppl S2):1–530.

The United Kingdom National Congenital Heart Disease Audit

17

Rodney C.G. Franklin, David Cunningham, and John L. Gibbs

Abstract

The National Congenital Heart Disease Audit was set up as a validated electronic database in 2000 for quality assurance purposes to assess and monitor outcomes after therapeutic procedures on patients with paediatric and congenital heart disease of all ages, with over 100,000 patients currently in the database (60 % post-surgery). Mortality is tracked independently by using unique patient identifiers (NHS number). NICOR has published centre specific comparisons of activity, data quality and survival on its public portal following individual procedures for over a decade, including when units have appeared to be underperforming (web.nicor.org.uk). It now also operates at a local level with onsite near real time monitoring of 30 day mortality and re-interventions. NICOR has begun to publish risk adjusted whole centre comparative outcomes, both using novel risk adjustment methodology developed using the Audit's database. The Audit has been instrumental in improving the prenatal detection of major congenital heart malformations by publishing the regional success rates of antenatal screening.

Keywords

UK National Audit • Congenital heart disease • Database

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Introduction

The National Congenital Heart Disease Audit (NCHDA) is used to assess outcomes after therapeutic procedures in the United Kingdom (UK) and represents an excellent example of what can be achieved at a national level to monitor surgical and transcatheter cardiovascular interventions undertaken for quality assurance purposes on patients with congenitally malformed hearts of all ages. National monitoring of survival rates after congenital cardiac interventions in the UK began over 35 years ago with the voluntary submission of cardiac surgery data to the Society of Cardiothoracic Surgeons of Great Britain and Ireland in 1977. The Central Cardiac Audit Database (CCAD) was subsequently established in 1999 by the British Cardiac Society (now the British Cardiovascular Society), the Society of Cardiothoracic Surgeons, and the British Paediatric Cardiac Association (now the British Congenital Cardiac Association). This involved the establishment of a team of experts to set up computerized registries with access to sophisticated analyses of anonymised data, inclusive of robust protocols for the protection and validation of data, covering acquired and congenital heart disease for both adults and children. A major underlying principle was that the audits should compare levels of performance, so as to set standards of optimal care as a benchmark for individual hospitals. Units found to be relatively underperforming would receive constructive feedback, which might focus on, for example, surgical techniques, intensive care support, or shortcomings in the 'system' or infrastructure. The Kennedy report published in 2001 on the results of infant congenital cardiac surgery in Bristol, UK a decade earlier, underscored the need for a national system to analyze and compare outcomes after cardiovascular surgery and therapeutic cardiovascular catheterization [1–3].

In 2000 the Department of Health specifically funded the CCAD Audit to collate data from all centers undertaking procedures for congenital heart disease in the UK. This audit differed in several major aspects from previous national audit projects at that time and these features continue currently:

- data is collected electronically in a secure format, with informed patient consent (compulsory from 2006);
- data is coded with clinician guidance and then transferred into a single system to ensure relevance and avoid double counting;
- data collection is mandatory for all relevant procedures at all institutions undertaking paediatric cardiovascular procedures (under 16 years of age), aiming for 100 % case ascertainment;
- mortality and re-intervention are tracked centrally by using the unique patient identifier given to all citizens in the UK at birth, or if they are eligible for state funded health care, namely the National Health Service (NHS) number;
- independent data validation is used.

After a series of internal NHS reorganizations, in 2011 the CCAD congenital audit as a body moved away from being part of the National Clinical Audit Support Program, a subsidiary at that time of the NHS Information Centre, to being one of six audits under the auspices of the National Institute for Cardiovascular Outcomes Research (NICOR), an independent entity within University College London [4]. NICOR's mission is to provide accurate data on cardiovascular outcomes for the public, healthcare providers and the medical profession. Funding of NICOR is currently from NHS England via the Healthcare Quality Improvement Partnership (HQIP), which sets and monitors many of the deliverables of all the audits [5].

Data Collection

The initial system in 2000 consisted of the tracking of postprocedural mortality using only a standardized minimum dataset of 20 fields. This system remained unchanged for the first 2 years in order to imbed the system into stakeholder hospitals' infrastructure and culture. A gradual expansion of fields then occurred, to include whether there was an antenatal diagnosis (yes or no) and

the inclusion of adults with congenital heart disease in 2003. Fields to document additional comorbidities and the monitoring of outcomes related to specified morbidities were added a year later. Outcomes metrics now available include 30 day, in hospital and 1 year 'alive or dead' status, the length of stay in hospital, and the time to final extubation. Currently data are submitted electronically in an encrypted format with prospective tracking of mortality and re-intervention using up to a 40 field minimum dataset. To ensure patient confidentiality, advanced data encryption technology is used to control access to data through a secure key system. Patient consent for central submission is a mandatory requirement and to date there have been negligible refusals by patients or their families for data submission.

All 13 paediatric congenital heart disease centers in England, Scotland, and Northern Ireland have participated since that time, whilst the Republic of Ireland joined the NCHDA in 2012. The acquisition of local data at the point of delivery has been found to be essential in order to ensure timely and comprehensive collection of data on all cases, as has been the presence of a local database manager to encourage clinician participation and to internally validate the quality of data before submission to the NCHDA.

As detailed in Chap. 1, a common clinical language is fundamental for success when comparing institutional outcomes. The Short List of the European Paediatric Cardiac Code (EPCC) has been employed by the NCHDA since 2003 [6]. The European Paediatric Cardiac Code is a subset of the International Pediatric and Congenital Cardiac Code (IPCCC), such that all IPCCC non-qualifier codes are mapped to the EPCC, enabling those units using the full IPCCC to seamlessly map to the EPCC for NCHDA central returns of data. The EPCC is also mapped to the 10th revision of the International Classification of Diseases (ICD10), as provided by the World Health Organisation, for diagnoses, and the 4th revision of the United Kingdom specific procedure codes, as provided by the NHS Health and Social Care Information Centre, for central government returns and 'billing' (currently OPCSv4.7). By using these latter maps from the EPCC, clinicians and submitting unit managers are able to ensure that intervention linked billing is accurately ascribed for the individual patient, which should in turn ensure correct reimbursement from NHS commissioners.

Verification of Life Status and Validation of Data

The verification process begins with the database manager at the congenital cardiac center checking data accuracy with medical staff before the data is submitted. Independent validation of the patient's life status (alive or dead) is achieved by central tracking using the linkage of each patient's NHS number to the Office for National Statistics, where the life status of every resident in England and Wales is independently registered. The NCHDA is therefore periodically updated on patients' survival status, on a quarterly basis each year. Separate arrangements exist in Scotland and Northern Ireland. It is important to note that if the center undertaking the procedure records a death, then this is taken as fact, even if central mortality tracking is still recording an alive status due to legal process delays, such as delay to registration whilst awaiting the results of an inquest. This system, therefore, allows for patients to be tracked across the UK, irrespective of changes of address or hospital, minimizing the chances of being lost to follow-up, at least in terms of life status.

In addition, each unit is visited for 1 or 2 days each year by an independent specialist database nurse administrator employed by NICOR, along with a volunteer surgeon or cardiologist from another unit. A detailed pre-visit proforma is completed by each center covering such areas as security and confidentiality, in-house verification and quality assurance, training for data collection and accuracy, communication issues, accountability, health records management, and timeliness of central submission. The visits are scheduled in the year following the data submission year (1st April through to 31st March). At the visit, all operating room and catheter laboratory logbooks or electronic records are scrutinized to ensure procedural data accuracy and that

all procedures have been captured. In addition, a random selection of 20 patient hospital records is requested in advance and all data points are compared to the dataset submitted centrally to the NCHDA for missing or incorrect data. A percentage Data Quality Indicator score is then calculated. To date, the validation visit results have been very encouraging with scores improving over time from an average of 79–91 % currently (range 81–98 %). At the end of the visit, the local audit lead clinician(s), including a congenital cardiac surgeon, meet with the auditors to discuss areas of excellence and deficiencies. Within weeks, a formal report is submitted back to the hospital team and to higher management. The visiting audit team have often been able to successfully bring pressure on hospital managers to invest in the provision of manpower and/or higher quality data entry software to achieve improved standards. The visits are therefore viewed by the congenital cardiac clinicians as very positive and productive encounters.

Ideally of course, every medical record of the approximately 10,000 patients undergoing procedures each year should be examined in detail. Lack of funding and skilled manpower for such an enterprise precludes implementation of this costly, time and resource consuming strategy and this is unlikely to change in the current economic climate. In the UK it has been deemed preferable to have this quality assurance visit to all hospitals undertaking congenital cardiovascular procedures on an annual basis using this methodology of full case ascertainment along with selective in depth data quality analysis, rather than to visit a limited number of centers and comprehensively examine all of the patient records, as undertaken by some other national and international audits [7]. If a unit is under-performing in a particular area, or cases have been found which have not been submitted, the hospital team is asked to re-examine the data element and resubmit this data, along with any additional cases which may have been found when examining theatre and catheter laboratory case records. Duplicate records are also eliminated. Investigations beyond this protocol have not yet been required. As detailed below, centre specific results including the Data Quality Index are now published on the NICOR NCHDA website (web.nicor.org.uk) allowing free access to families and the media. An additional incentive, therefore, exists to provide accurate and complete data, knowing that central tracking of mortality provides external monitoring of performance.

During a recent review of the validation process, further enhancements were agreed. Data submissions will be electronically interrogated such that missing or nonsensical data field entries, will be automatically returned to the submitting center for correction, whilst still registering the individual patient concerned within the NCHDA database. An example would be the submission of a weight that is outwith three standard deviations from the mean expected for that age. This protocol will include a short timeline for resubmission of amended data for that patient or an explanation to explain the alleged variance to that expected. On site validation visits will also be completed in a more rapid timeframe so as to be completed within 6 months of the end of the submission year, with reports for that year published a month thereafter.

Procedural Activity and Mortality Tracking

Table 17.1 documents the overall numbers of surgical and transcatheter cardiovascular procedures undertaken in the United Kingdom from 2000 to 2013, with 30 day and 1 year survival [8]. Over 100,000 patients have been submitted with a current rate of over 10,000 each year. Nearly 60 % of these are surgical procedures, with just over 4,700 operations being performed currently in those under 16 years of age. The overall increase in surgical and transcatheter intervention volume over time at least in part reflects the capturing of more data from adults with congenital heart disease over recent years. When first setup there was some doubt as to the need for a separate organization to collect data for congenital cardiac procedures. A comparison was therefore made between the data collected by CCAD in 2000–2001 and the volunteered data submitted to the Society

Table 17.1 Cardiovascular surgical and transcatheter interventional procedures undertaken in the United Kingdom 2000–2013 with 30 day and 1 year survival for children (under 16 years) and adults with congenital cardiac disease, as well as children with acquired heart disease

Financial year (April to April) count	Total procedure	Surgical procedures	Surgical procedures <16 years	Transcatheter interventional procedures	Transcatheter interventional procedures <16 years	30 day survival (%)	1 year survival (%)
2012–2013	10,195	5,836	4,716	4,359	2,699	98.2	No data
2011–2012	10,207	5,716	4,649	4,491	2,626	98.1	95.4
2010–2011	10,120	5,839	4,771	4,281	2,468	97.8	94.8
2009–2010	9,209	5,264	4,238	3,945	2,153	98.1	95.3
2008–2009	8,840	4,948	4,005	3,892	2,220	97.8	94.9
2007–2008	8,354	4,770	3,959	3,584	2,022	97.9	95.2
2006–2007	8,540	4,792	4,028	3,748	2,251	97.7	94.5
2005–2006	8,089	4,632	3,919	3,457	2,212	97.7	94.8
2004–2005	7,344	4,345	3,786	2,999	2,091	97.4	93.8
2003–2004	7,376	4,489	3,985	2,887	2,211	97.4	93.9
2002–2003	6,422	4,125	3,716	2,297	1,837	97.1	93.8
2001–2002	5,943	3,787	3,311	2,156	1,720	96.6	93.4
2000–2001	5,960	4,067	3,707	1,893	1,658	96.5	91.8
Total	**105,599**	**62,610**	**52,790**	**43,989**	**28,168**		

of Cardiothoracic Surgery of Great Britain and Ireland, as well as administrative data collated by NHS coders mostly for billing purposes, known as Hospital Episode Statistics. Perhaps not surprisingly, there was marked underreporting of mortality in the voluntary data submission when compared to that submitted to CCAD with central mortality tracking. In 2000–2001 7 of 11 centers in England under-reported 30 day mortality for the 5,494 procedures undertaken (3,666 surgical and 1,828 therapeutic catheterization). Central tracking identified 194 deaths which had occurred within 30 days of the procedure, 42 of which but were not present in the volunteered data. Nineteen of the forty-two patients had been discharged alive but subsequently died within 30 days of their operation, whilst the remaining 23 patients had been incorrectly coded as alive at discharge. Voluntary reporting of discharge status, therefore, underestimated 30 day mortality by 22 % [3]. A similar picture was found with respect to administrative data acquisition, which was available for 2,716 patients. Hospital Episode Statistics data had underreported 30 day mortality by 9 %, whilst classifying 1 % of surviving patients as deceased. Administrative data had also under-reported the total number of procedures by 10 % [3]. As a consequence, volunteered and administrative data has now been completely superseded by NCHDA submissions for quality assurance purposes. Although administrative data continues to be collected in the UK, its quality remains suspect and unreliable, with little clinician input.

Validation visits have proved to be invaluable when ensuring complete case ascertainment. During the 2000–2001 validation visits 143 procedures were found to be missing from the data submissions to CCAD, predominantly related to systematic errors in data collection. The visits resulted in submission of missing or revised data from all of the 13 centers and a smaller but important number of procedures continue to be added after validation visits, as well as duplicate entries deleted.

Risk Adjusted Outcomes

Initially, in view of the heterogeneity of procedures used to treat congenital cardiac malformations and the relatively small number of cases undertaken in each center, outcomes were only reported for a limited number of ‘benchmark’

procedures: six for surgery (repairs of atrial septal defect, ventricular septal defect, atrioventricular septal defect, tetralogy of Fallot, simple transposition of the great arteries, and aortic coarctation) and three for therapeutic catheterization (atrial septal defect closure, arterial duct closure and balloon dilation of the pulmonary valve), focusing on the first year of collection [3]. Quality assurance was provided by the finding that there was no significant inter-unit or inter-operator variation detected for these procedures. Currently outcomes are stratified into 56 different procedure categories. These are tabulated and published annually, along with 3 yearly cohort analyses displayed as funnel plots (see below) which compare the 30 day mortality outcomes of the centers undertaking these procedures. Pressure to publish whole program outcomes as a single figure comparator between specialist units for postprocedural 30 day mortality have been resisted until very recently, owing to the lack of sound and validated risk adjustment methodology. Indeed outcomes were not included in the Safe and Sustainable Review of paediatric cardiac services in the UK, as fair adjustment for case mix was considered too difficult to achieve [9]. Paediatric cardiac surgery is very heterogeneous, with hundreds of IPCCC codes used in combinations to describe diagnoses and specific procedures, and it is established that the risk for individual operations varies widely [10].

Elsewhere, early efforts to adjust for case mix were based on the subjective and consensus based assessment of risks by panels of experts, creating the Risk Adjustment for Congenital Heart Surgery (RACHS-1) system [11] and Aristotle Score [12]. While certainly valuable and of some use, these methods have recently been challenged or even superseded by more empirical approaches, based on the recent availability of databases incorporating the outcomes of tens of thousands of patients. The Society of Thoracic Surgeons-European Association of Cardiothoracic Surgery (STS-EACTS) score (STAT-score), introduced in 2009, was based on data from over 75,000 paediatric cardiac surgery procedures performed between 2002 and 2007 in Europe and North America [13]. A total of 148 procedures were assigned a numeric score ranging from 0.1 to 5.0 based on the estimated mortality rate, thereby enabling benchmarking with risk adjusted outcomes based on how well the clinical teams have been performing in the recent past [14].

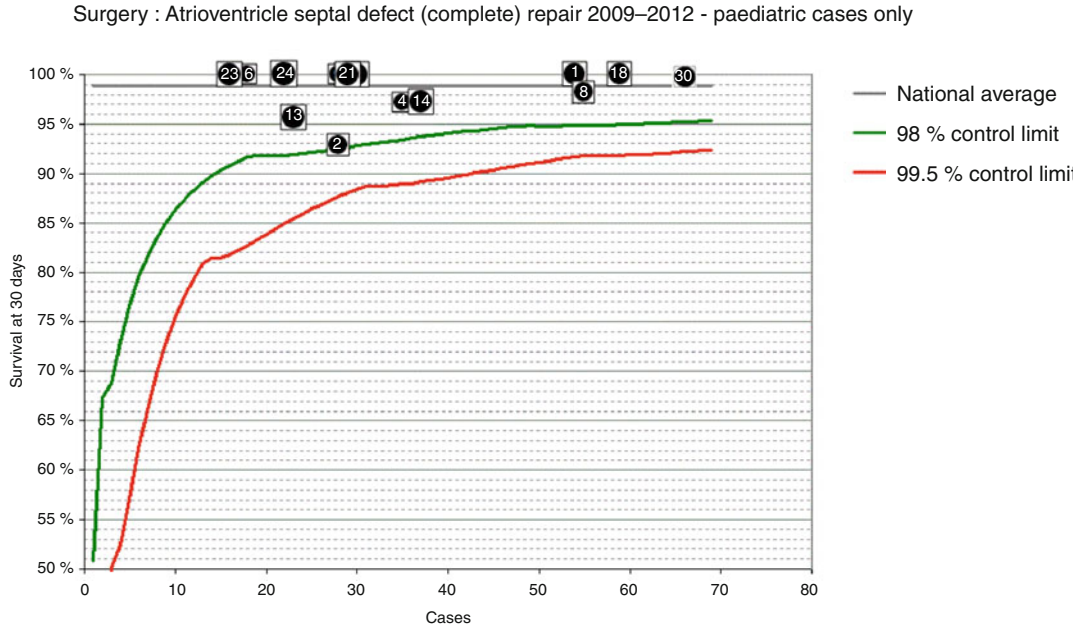
A similar empiric risk adjustment system has recently been developed in the UK based on data in the NCHDA database with respect to 30 day mortality after paediatric cardiac surgery, referred to as PRAiS (Partial Risk Adjustment in Surgery). It was created specifically to enable in house, near real time monitoring of whole program activity, so as to answer the major criticism of historical reporting of outcomes on operations, which may date from a year, if not several years earlier. This delay in analyzing and then reporting the results of surgery for quality assurance, along with the identification of potential outlier institutions, is due to the validation process and the fact that procedure specific outcomes are reported in rolling three yearly cohorts due to the relatively low numbers involved. There has been a perception by some parent groups that deaths may have occurred which were avoidable in the interim period. The PRAiS model uses relatively complex risk adjustment methodology that incorporates not only the procedure category but also includes cardiac diagnosis, univentricular status, age category (neonate, infant, child), continuous age, continuous weight and the presence of non-Down syndrome comorbidity, as well as era so as to take into account recent lower overall mortality [15]. It was developed and then validated prospectively using data from all pediatric cardiac surgery procedures performed in the UK between 2000 and 2010 as a comparator baseline [16]. It has recently been re-calibrated with data from operations between 2009 and 2012 to reflect the noted improvement of most recent 30 day mortality outcomes. All specialist centers in the UK are now mandated by NHS England to use this method for near real time in house monitoring of paediatric cardiovascular procedures, as described in detail in Chap. 25. The method enables early identification of adverse events (within a month), whether death or unplanned reoperation. Such events may be

explainable on the grounds of case mix or the presence of comorbid conditions, or alternatively generate within team discussions leading to changes in local healthcare strategies. More recently PRAiS methodology has been applied to whole program outcomes as a national comparator of performance with respect to 30 day mortality covering 2009–12 [17]. Funnel plot methodology is being used, plotting the relative risk, that is, observed survival divided by PRAiS-predicted survival, against volume of cases, such that the case complexity of each center is used as the basis for predicting performance. PRAiS has now become an integral part of UK quality assurance for congenital cardiac surgery in these complimentary capacities, for local near real time assessment of performance by individual specialist centers and for national comparisons of performance in 3 yearly cohorts.

The UK is one of only three countries with universal participation in national audit of paediatric cardiac surgery, the other two being Sweden and Poland, meaning that the UK NCHDA is a valuable resource for the exploration of trends in outcome. A recent trend analysis of over 36,600 surgical episodes undertaken in 10 consecutive years from April 2000 to March 2010, demonstrated a fall in 30 day raw mortality from 4.3 % in 2000–2001 to 2.6 % in 2009–2010 (April to March) [18]. This coincided with a significant increase in the number of pediatric congenital heart procedures undertaken in the UK; rising from 2,283 in 2000–2001 to 3,393 in 2009–2010. These UK results compare favorably with international similar era results from North America and Europe with a reported mortality of just over 4 % [14, 19]. Importantly, case mix, as assessed using four risk bands of estimated mortality calculated using PRAiS methodology, progressively increased in this decade, notably in terms of the numbers of patients weighing less than 2.5 kg and those with a high risker diagnosis such as hypoplastic left heart syndrome [18]. Concerns that the publication of comparative unit specific 30 day mortality outcomes might lead to risk averse behavior by individual surgeons or centers appear to be unfounded in the UK.

Publication of NCHDA Analyses and Quality Assurance

Since 2007, the annual results for congenital cardiac procedures have been published on the NICOR NCHDA website, with free access to families, the media and medical colleagues (web.nicor.org.uk). This Public Portal's main focus is to provide details of the 30 day and 1 year life status outcomes after 39 surgical procedures and 17 transcatheter procedures (including radiofrequency ablation and placement of implantable cardioverter defibrillators) [20]. These are presented as procedure and center specific outcomes both in tabular displays for each year and in graphical format, using funnel plots for three yearly cohorts, here plotting percentage survival against the number of cases. The funnel shape arises because Units to the left of the plot have fewer cases and so would be expected to have more variability due to chance. There are superimposed 'control limits' of two and three standard deviations below the mean, as exemplified in Fig. 17.1, which illustrates the survival plot of the 505 patients who underwent definitive repair of a complete atrioventricular septal defect (complete atrioventricular canal) in the UK and Republic of Ireland between 1st April 2009 and 31st March 2012. The overall average survival is depicted as a horizontal grey line. Two control limits are shown: a warning limit as a green line (98 % control line) and a red line for the 99.8 % control line, representing an 'alert limit'. Unit performances are shown as numbered symbols referable to the table below the plot. The Portal also has extensive sections which explain the analyses for the lay reader, details of the centers performing congenital heart procedures (provided by each center), along with the center's Data Quality Index for the most recent validated year, as well as links to explanatory pages for many congenital heart malformations. There is a technical section for contributors and a tab for antenatal diagnoses (see below). The NCHDA has not to date published individual operator results as there is an embedded belief that the responsibility for operations involving congenital cardiac



		Cases	Alive 30days	Dead 30days	Survival 30days
30	Our Lady's Dublin	66	66	0	100.0 %
18	Royal Brompton Hospital	59	59	0	100.0 %
8	Great Ormond Street Hospital for Children	55	54	1	98.2 %
1	Alder Hey Hospital	54	54	0	100.0 %
14	Leeds General Infirmary	37	36	1	97.3 %
4	Bristol Children's Hospital	35	34	1	97.1 %
21	Royal Hospital for Sick Children	29	29	0	100.0 %
2	Birmingham Childrens Hospital	28	26	2	92.9 %
10	Evelina Childrens Hospital	30	30	0	100.0 %
9	Glenfield Hospital	28	28	0	100.0 %
24	Southampton General Hospital	22	22	0	100.0 %
13	Harley Street Clinic	23	22	1	95.7 %
6	Freeman Hospital	18	18	0	100.0 %
23	Royal Victoria Belfast	16	16	0	100.0 %
20	John Radcliffe Hospital	5	5	0	100.0 %

Fig. 17.1 Funnel survival plot of the 505 complete atrioventricular septal defect (atrioventricular canal) repair operations performed in children (under age 16 years) undertaken in the United Kingdom between 1st April 2009 and 31st March 2012 (Accessed 31st March 2014)

malformations rests on the team involved (operator, intensivist, cardiologist, anesthetist, etc.) and rather than solely with the surgeon. Private, password controlled access is provided on the NCHDA Portal, however, for the individual operator to see personal details of procedures performed in comparison to the national average. This personalized data is a crucial part of the revalidation and annual appraisal processes undertaken by clinicians in the UK.

Quality assurance with respect to 30 day mortality is assessed on a rolling 3-year basis for each of the 56 procedures and displayed on the procedure specific funnel plot (Fig. 17.1). If a Unit is above the green line, then the performance is no different from the national average. If a Unit is

found to be below the warning limit, a protocol is set in motion before declaring that performance is below expected. The initial focus is on the data quality itself to ensure that a process error has not occurred when the data was submitted by the institution to the NCHDA. The Audit Lead at the Unit is asked to check with his database manager that the deaths in question have been correctly coded, particularly with respect to the exact procedure undertaken and other diagnoses present. If a potential outlier status is established, once data quality is confirmed not to be in question, then the NCHDA Clinical Lead, along with the Presidents of the Society of Cardiothoracic Surgeons of Great Britain and Ireland and British Congenital Cardiac Association formally write to the Audit Lead and Lead Surgeon at the Center requesting a report be delivered within 3 weeks to explain the outcome and any remedial healthcare changes which have been actioned as a result. The Center is expected to include an evaluation of case-mix, including comorbidities and risk factors which may have adversely effected outcome, related healthcare processes and available resources, as well as contributions from the professionals directly involved in patients' care. A similar pyramidal process has been summarized by an Australian team [21], emphasizing the complexity involved when drawing conclusions based on a single outcome variable, namely 30 day mortality. If an institution is below the alert limit (red line), a similar process follows except that the formal letter is additionally copied to the Unit's Medical Director, who then informs the appropriate NHS overarching regulatory body. Using this methodology, the spurious ranking of the centers is avoided, whilst procedural complexity and the volume of cases is taken into account and hospitals falling outside these limits incur further investigation.

Antenatal Diagnosis of Congenital Heart Malformations

Although quality assurance has been the dominant goal of the NCHDA, a parallel aim has been quality improvement in clinical practice related to congenital heart malformations. In the UK almost all women have an ultrasound scan to detect fetal anomalies in the mid second trimester including screening for major congenital heart malformations. The benefits of an antenatal detection are well established and include planed safer perinatal management, the appropriate use of medical services and parental choice. The affected child may benefit from better neurodevelopmental outcomes and a shorter hospital stay in intensive care [22]. In 2004 the Audit introduced the mandatory additional field of 'antenatal diagnosis' with the simple choice of 'yes' or 'no'. In 2008 the initial results were published on the NICOR congenital heart disease website, dividing the country into Health Authorities and Primary Care Trusts. The success rate of detected heart anomalies that require a procedure during infancy (excluding patent arterial and atrial septal defect closure) varies greatly across the UK, ranging from under 10 % to over 50 % [23]. The data do not include pregnancies that were discontinued following a prenatal diagnosis of a major cardiac anomaly, often complicated by karyotype anomalies, or in utero deaths. Subsequent analyses have shown a consistent positive trend in antenatal diagnoses across the UK (Fig. 17.2), although regional and inter-hospital variation remain, with most marked improvement seen in Wales and Scotland and some areas remaining relatively static. This improvement has largely been due to commissioned investment in local and regional training programs for sonographers as required by the NHS Fetal Anomaly Screening Program, the acquisition of high quality ultrasound equipment, and changes in national guidelines [24, 25]. It is also an excellent demonstration of how quality of care can be improved for any clinical outcome measure using a validated national database which publishes its results on a public, web-based portal, and permits a degree of 'naming and shaming' poor performance to drive improvements. This increase in performance following the incorporation of the validated 'yes/no' question into the NCHDA database has implicitly raised the standards of care for babies with major congenital heart malformations, and

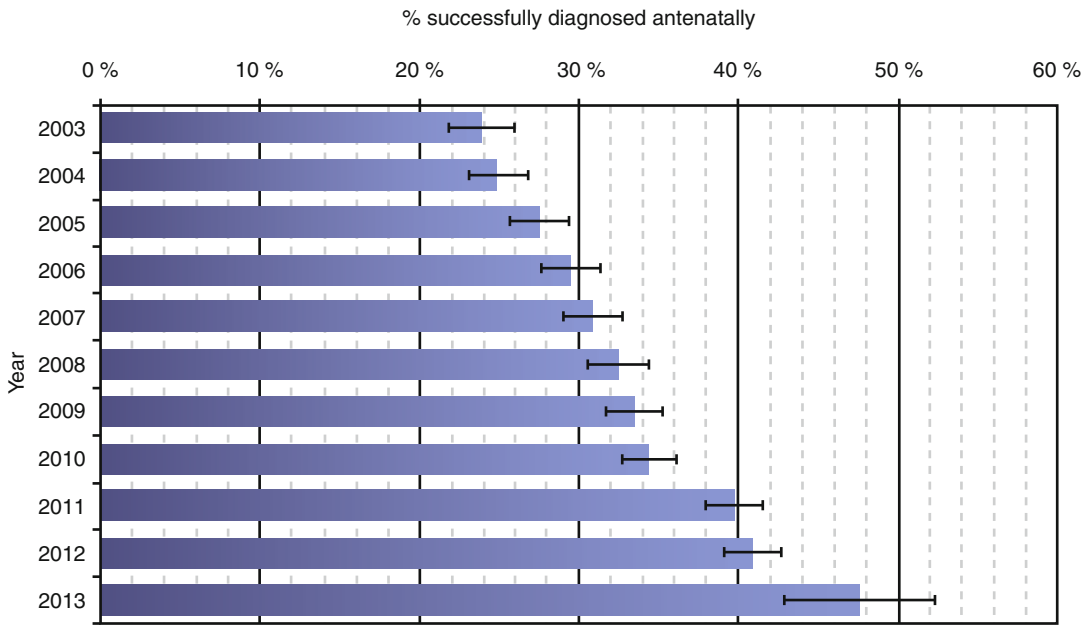


Fig. 17.2 Bar chart showing the trend in the whole of the UK towards improved antenatal diagnosis over the past 10 years for infants requiring surgery or a transcatheter intervention for congenital heart disease, excluding closure of a patent arterial duct or atrial septal defect. Less than a quarter of cases were diagnosed antenatally in 2004. This has risen to nearly half in 2013. Years are financial, running from 1st April to 31st March

supports the need for national comprehensive validated registries.

Future Developments and Conclusions

The history of pediatric cardiac surgery in the UK is such that the speciality is highly scrutinized and remains connected in the public mind to distressing past events [1, 2]. The placing of patient safety with effective clinical governance and best practice at the centre of every congenital cardiac program has been a common goal in the NHS, supported by clinicians, NICOR and related parent groups. There has been a progressive decrease in paediatric cardiac surgical mortality and world class results are now the expected norm in the UK [18]. The NCHDA has been collecting national data on all congenital cardiac surgical and transcatheter interventions in the UK since 2000, providing quality assurance on 30 day mortality rates, whilst also tracking longer term survival. It has published centre specific comparisons of activity, data quality and survival on its public portal following individual procedures for over a decade, including when units have appeared to be underperforming. It now also operates at a local level with onsite near real time monitoring of 30 day mortality and re-interventions and has begun to publish risk adjusted whole centre comparative outcomes, both using PRAiS methodology.

The NCHDA process requires the harvesting of accurate and validated data on the diagnosis, treatment, and outcome of patients with paediatric and congenital heart disease from prenatal life through to adulthood. The reporting to the public of data and outcomes, combined with the knowledge that central tracking of mortality externally monitors performance, provides added incentive to institutions to provide accurate and complete data. Planned improvements to the validation process, such as automated sense checking for accuracy and alignment of diagnosis and the procedure undertaken, should strengthen this process further.

The UK national system enables the comparison of outcomes following interventions between

individual centers, whilst taking into account the mix of cases involved, accompanying risk factors and comorbidities, as well as postprocedural complications.

Benchmarking against those units who perform best allows the analysis of relevant and genuine factors underlying differing outcomes, and should lead to the instigation of improvements, which should positively affect both mortality and morbidity. The NCHDA has a number of ongoing projects to understand better the influence of other factors on 30-day mortality. These include an assessment of the importance of ethnicity and socio-economic deprivation, as well as the importance of individual risk factors and comorbidities on short term outcome.

Although the improvements in prenatal diagnosis are gratifying and can be used to compare temporal patterns of antenatal detection, the data cannot provide a complete record of prenatal detection of major congenital heart anomalies and subsequent outcomes other than interventions. There is a requirement for mother-to-infant linkage to provide such granular and diagnosis based audit and is the aim of future modifications to the NCHDA, as well as expanding the data fields to include all possible outcomes following a prenatal diagnosis (termination of pregnancy, in utero demise, compassionate non-interventional care and interventions).

For individual patients, outcome is much more than survival alone, and their recovery usually continues long beyond the first 30 postoperative days. Modern day intensive care and life support have evolved along with surgical advances such that very long specialist hospital stays are not uncommon, some ending in death well beyond 30 days; and survival may well be accompanied by a variety of complications. These facts and the very low mortality rates currently reported in the developed world have shifted the focus of audit away from just survival, to survival with as little morbidity as possible, measures of functional status and patient reported experiences and quality of life. Databases are already being used to report both outcomes, with an emphasis on long term, complication free survival as a superior marker for quality of healthcare [26]. Such measures may also provide evidence on the comparative longer term benefits of different interventional strategies which may have commenced in the neonatal period. As part of this move, the members of NCHDA research team are undertaking analyses which examine unplanned re-interventions for specific lesions, such as for tetralogy of Fallot and transposition of the great arteries following the arterial switch procedure. A large scale project has also started to understand postoperative morbidity in the UK population [27]. An expansion of the database is planned for April 2015 to focus on complications following transcatheter interventions, including embolization of devices, the need for unplanned re-interventions (surgical or transcatheter) and local vascular compromise. Specific manufacturer details of implanted devices, including septal closure devices, implantable valves and pacemakers, will also be included.

Finally, work has begun to compare international outcomes using the IPCCC in its different versions as a common coding language to compare data and outcomes between the UK NCHDA and its equivalent in Europe and North America, the databases of the European Association of Cardiothoracic Surgery and the Society of Thoracic Surgeons. It is anticipated that such future cooperative multi-institutional and multinational studies will enable further improvements in the quality and effectiveness of healthcare for patients with congenital cardiac malformations of all ages, whilst influencing the allocation of increasingly limited resources.

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References

1. Smith R. All changed, changed utterly. Br Med J. 1998;316:1917–8.
2. Learning from Bristol: the report of the public inquiry into children's heart surgery at the Bristol Royal Infirmary 1984–1995. http://webarchive.nationalarchives.gov.uk/+/www.dh.gov.

uk/en/Publicationsandstatistics/Publications/PublicationsPolicyAndGuidance/DH_4005620. Accessed 14 Oct 2013.
3. Gibbs JL, Monro JL, Cunningham D, Rickards A. Survival after surgery or therapeutic catheterisation for congenital heart disease in children in the United Kingdom: analysis of the central cardiac audit database for 2000–1. Br Med J. 2004;328:611–5.
4. National Institute for Cardiovascular Outcomes Research home web page. http://www.ucl.ac.uk/nicor/about. Accessed 2 March 2014.
5. Barach P, Pahl R, Butcher A. Actions and Not Words, The Future of HQIP, Randwick, NSW: JBara Innovations for HQIP, National Health Service, London, 2013.
6. Franklin RC, Anderson HR, Daniels O, et al. The European Paediatric Cardiac Code. Report of the Coding Committee of the Association for European Paediatric Cardiology. Cardiol Young. 1999;9:633–58.
7. Clarke DR, Breen LS, Jacobs ML, Franklin RC, Tobota Z, Maruszewski B, Jacobs JP. Verification of data in congenital cardiac surgery. Cardiol Young. 2008;18 Suppl 2:177–87.
8. National Institute for Cardiovascular Outcomes Research, National Congenital Heart Disease Audit website. https://nicor4.nicor.org.uk/CHD/an_paeds.nsf/WMortality?Openview. Accessed 3 March 2014.
9. NHS. Safe and sustainable: children's congenital cardiac services. In: NHS, editor. NHS Specialist Services, London, UK, 2011.
10. Jacobs JP, Jacobs ML, Lacour-Gayet FG, Jenkins KJ, Gauvreau K, Bacha E, et al. Stratification of complexity improves the utility and accuracy of outcomes analysis in a Multi-Institutional Congenital Heart Surgery Database: Application of the Risk Adjustment in Congenital Heart Surgery (RACHS-1) and Aristotle Systems in the Society of Thoracic Surgeons (STS) Congenital Heart Surgery Database. Pediatr Cardiol. 2009;30(8):1117–30.
11. Jenkins KJ, Gauvreau K, Newburger JW, Spray TL, Moller JH, Iezzoni LI. Consensus based method for risk adjustment for surgery for congenital heart disease. J Thorac Cardiovasc Surg. 2002;123(1):110–8.
12. Lacour-Gayet F, Clarke D, Jacobs J, Gaynor W, Hamilton L, Jacobs M, et al. The Aristotle score for congenital heart surgery. Semin Thorac Cardiovasc Surg Pediatr Card Surg Annu. 2004;7:185–91.
13. O'Brien SM, Clarke DR, Jacobs JP, Jacobs ML, Lacour-Gayet FG, Pizarro C, et al. An empirically based tool for analyzing mortality associated with congenital heart surgery. J Thorac Cardiovasc Surg. 2009;138(5):1139–53. doi:10.1016/j.jtcvs.2009.03.071.
14. Jacobs JP, O'Brien SM, Pasquali SK, Jacobs ML, Lacour-Gayet FG, Tchervenkov CI, et al. Variation in outcomes for risk-stratified pediatric cardiac surgical operations: an analysis of the STS Congenital Heart Surgery Database. Ann Thorac Surg. 2012;94(2):564–71.
15. Crowe S, Brown KL, Pagel C, Muthialu N, Cunningham D, Gibbs J, Bull C, Franklin R, Utley M, Tsang VT. Development of a diagnosis- and procedure-based risk model for 30-day outcome after pediatric cardiac surgery. J Thorac Cardiovasc Surg. 2013;145(5):1270–8. doi:10.1016/j.jtcvs.2012.06.023.
16. Pagel C, Utley M, Crowe S, Witter T, Anderson D, Samson R, et al. Real time monitoring of risk-adjusted paediatric cardiac surgery outcomes using variable life-adjusted display: implementation in three UK centres. Heart. 2013;99(19):1445–50.
17. Cunningham D, Franklin R, Bridgewater B, Deanfield J. Investigation of mortality from paediatric cardiac surgery in England 2009–12; 8 April 2013. https://nicor4.nicor.org.uk/CHD/an_paeds.nsf/vwContent/Minutes%20and%20Newsletters.
18. Brown KL, Crowe S, Franklin RC, McClean A, Cunningham D, Barron D, Tsang V, Pagel C, Utley M. Trends in 30-day mortality rate and case mix for paediatric cardiac surgery in the UK between 2000 and 2010. Submitted for publication May 2014.
19. European Association of Cardiothoracic Surgery (EACTS). Congenital database gold standards. Warsaw: Children's Memorial Health Institute; 2013.
20. National Institute for Cardiovascular Outcomes Research congenital heart disease website. https://nicor4.nicor.org.uk/CHD/an_paeds.nsf/WBenchmarksYears?openview&RestrictToCategory=2012&start=1&count=500. Accessed 31 March 2014.
21. Duckett SJ, Coory M, Sketcher-Baker K. Identifying variations in quality of care in Queensland Hospitals. Med J Aust. 2007;187(10):571–5.
22. Brown KL, Ridout DA, Hoskote A, et al. Delayed diagnosis of congenital heart disease worsens preoperative condition and outcome of surgery in neonates. Heart. 2006;92:1298–302.
23. Antenatal diagnosis. National Institute for Cardiovascular Outcomes Research congenital heart disease website. https://nicor4.nicor.org.uk/CHD/an_paeds.nsf/vwContent/Antenatal%20Diagnosis?Opendocument. Accessed 1st June 2014
24. Antenatal Care Guideline. Published June 2008. National Institute for Health and Care Excellence, UK. http://www.nice.org.uk/CG062fullguideline. Accessed 1st June 2014.
25. Gardiner HM, Kovacevic A, vdHeijden LB, Pfeiffer PW, Franklin RCG, Gibbs JL, Averiss IE, LaRovere JM. Prenatal screening for major congenital heart disease: assessing performance by combining national cardiac audit with maternity data. Heart. 2013. doi:10.1136/heartjnl-2013-304640.
26. Jacobs ML, O'Brien SM, Jacobs JP, et al. An empirically based tool for analyzing morbidity associated with operations for congenital heart disease. J Thorac Cardiovasc Surg. 2013;145(4):1046–57.
27. Tsang V, Brown K. Selection, definition and evaluation of important early morbidities associated with paediatric cardiac surgery. On-going research project. http://www.nets.nihr.ac.uk/projects/hsdr/12500506.

The Pediatric Cardiac Care Consortium: The End of an Era and Beginning of a New Mission

18

James D. St. Louis and Lazaros K. Kochilas

Abstract

The Pediatric Cardiac Care Consortium (PCCC) was developed to provide individual institutions feedback about outcomes of common surgical and catheter based procedures and allow comparison of these outcomes on an international scale. The PCCC successfully accomplished this mission for over 30 years. The data was accurate, and provided to the individual centers on an annual basis. Recently, the database has elected to stop collecting information from the member institutions. As before, leaders within this organization have had to evolve the mission and find applications for the rich data that currently exists within the database. This chapter describes the history, current status, and future of the PCCC.

Keywords

Congenital Heart Disease • Database • Outcomes Research • Pediatric Cardiac Care Consortium

Introduction

The ***Regional Cardiac Program*** was formed in 1953 and was given the mission of providing financial support for children undergoing corrective surgery to treat congenital heart defects who were born in North and South Dakota, Montana, Wisconsin, and Minnesota. In 1979, representatives from the University of Minnesota and the Mayo Clinic organized a meeting with state and federal legislators to discuss the future of the Regional Cardiac Program. Because of the expanding availability of private and state funding, it was elected that the mission of the organization should change. A new mission was

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proposed that would provide accurate and real-time feedback to institutions providing care to children with heart defects. Thus, the ***Northern Great Plains Regional Cardiac Program*** was formed and tasked with collecting, analyzing, and providing operative and interventional outcomes on patients treated at member institutions. Subsequently, as centers from geographic areas beyond the Midwest joined, the name of the organization was changed to the ***Pediatric Cardiac Care Consortium*** (PCCC).

Although the mission of this group was clearly articulated, obstacles that confronted the group quickly became evident. Some of the obstacles included

- the very low incidence of the congenital cardiac disease,
- the diversity of anatomy,
- the lack of a detailed and uniformly accepted system of nomenclature,
- the variability of strategies of treatment between institutions, and
- the inability to risk adjust individual procedures.

Members of the PCCC were instrumental in addressing these early obstacles. Some of the early achievements of the PCCC registry were:

- the development of a system of centralized acquisition, analysis, and dissemination of data,
- the creation of a uniform system of coding and classification for congenital cardiac defects,
- the development of a methodology of risk adjustment to accurately and fairly facilitate the analysis of outcomes, and
- the implementation of a system to consistently audit the individual data that were submitted to the registry.

These accomplishment where achieved decades prior to the ongoing developments of current national and international databases for congenital cardiac disease.

The Database

The Pediatric Cardiac Care Consortium [1] database currently contains information on all cardiac procedures – including surgeries, cardiac catheterizations, and electrophysiology studies – performed at over 56 institutions over a 30 year period. Details of the creation, activities, and function of the PCCC have been described in several publications [2–4]. The database collected data about:

- demographics,
- diagnoses,
- procedures,
- history of previous cardiac operations, and
- associated non-cardiac conditions.

After these data were submitted, information was extracted and all diagnoses and procedures were coded utilizing the standardized system of nomenclature of the PCCC.

Data collected (Table 18.1) included

- patient identifier,
- patient country and state or country of origin,
- hospital name,
- encounter identifier number,
- date of birth,
- birth weight,
- date of admission to the hospital,
- weight at time of admission,
- previous cardiac operations (types and dates),
- presence and type of noncardiac malformations or conditions,
- data about cardiac catheterization, (including date, weight, hemoglobin, cardiac diagnosis, and type of procedure),

Table 18.1 Data collected in PCCC

Patient identifier
Patient country and state of origin, or country of origin
Hospital name
Encounter identification number
Birth date
Birth weight
Hospital admission data
Weight at time of admission
Previous cardiac operations (types and dates)
Presence and type of non-cardiac malformations or conditions
Cardiac catheterization data, including date, weight, hemoglobin and diagnoses
Cardiac operation data, including date, weight, hemoglobin, cardiac diagnosis, and type of procedure
Discharge, transfer, or death date, including diagnoses at death.

- date of discharge, transfer, or death (including diagnoses at death).

Diagnostic data included

- detailed cardiac diagnosis,
- cardiac procedure,
- chromosomal anomalies, and
- information on other systems including central nervous system (CNS), gastrointestinal (GI), genitourinary (GU), respiratory, and musculoskeletal.

Data about outcomes included

- 30-day survival post procedure,
- need for extracorporeal membrane oxygenation (ECMO),
- need for insertion of pacemaker or Automatic Implantable Cardio-Defibrillator (AICD),
- need for unplanned reoperation, and
- length of hospitalization.

Participating institutions submitted procedures to the PCCC in paper form that was either faxed or mailed to the University of Minnesota (Fig. 18.1). To assure quality of the data, the coders at the PCCC manually reviewed all cardiac diagnostic and procedural codes to assure consistency among institutions. A process of double data entry was employed and independent verification of the number of procedures and deaths in relationship to these procedures at each institution in a given year were obtained. If discrepancies existed between the number of cases reported to the PCCC and the number in medical records, then these discrepancies were resolved before the data from the center were analyzed. Best practices, in compliance with the Health Insurance Portability and Accountability Act of 1996 of the United States of America (HIPAA), were followed with participating institutions agreeing

- to submit data on all patients treated at their institutions, and
- to permit independent confirmation on the number of procedures performed annually to ensure compliance.

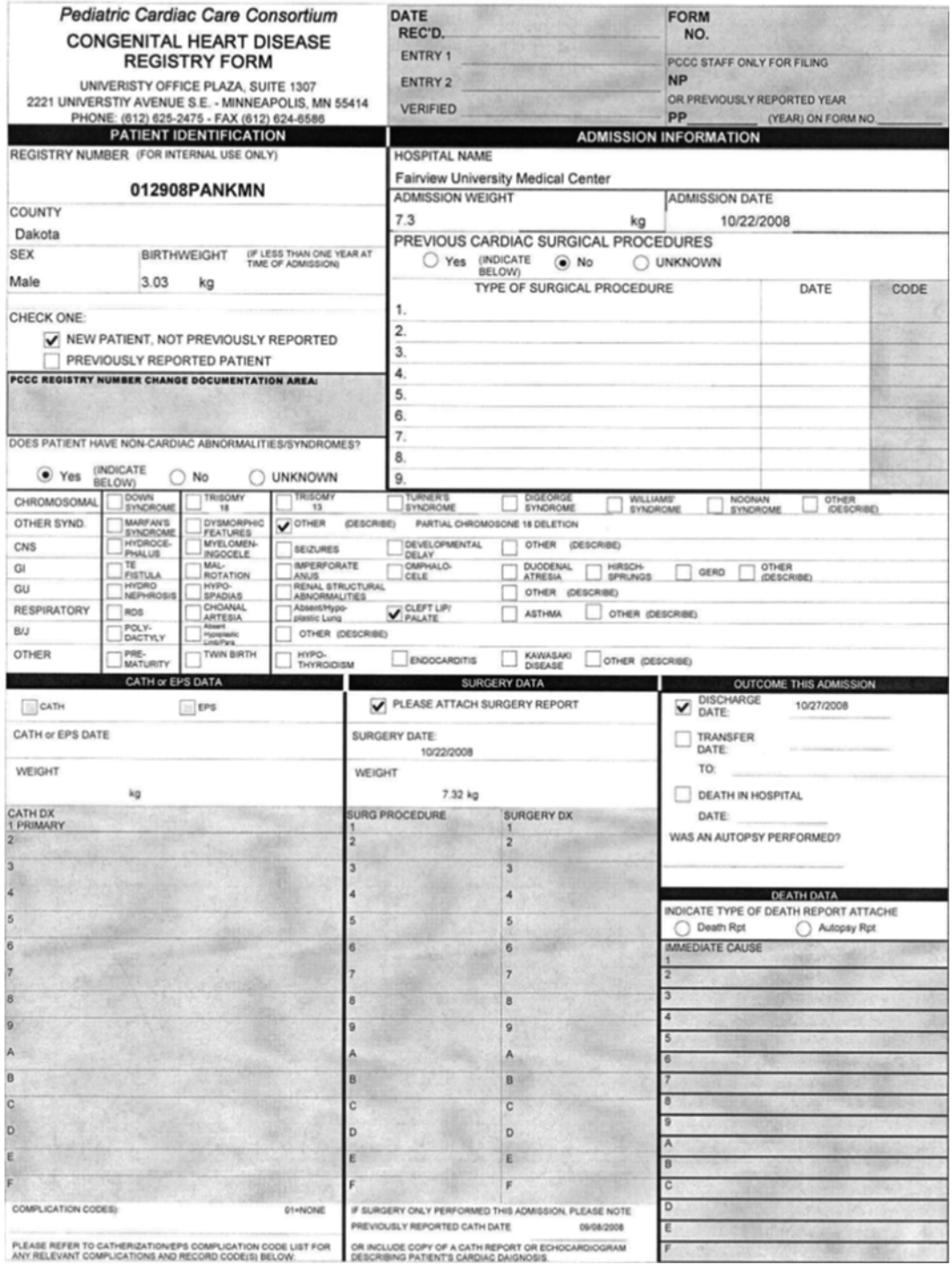
Centers also agreed to pay a nominal fee per form to support the costs of collecting and evaluating the data. Upon joining the registry, each center identified a cardiologist who was the contact person for their institution. In addition, each center identified a data collector who was responsible for the timely submission of data. This person was responsible for completing the data form and attaching the appropriate procedural reports. These forms were then forwarded to the PCCC's central office for coding and entry into the PCCC database. All diagnoses and procedures were assigned a PCCC study code number. Trained coders reviewed the reports, extracted information, and coded the cardiac diagnoses and types of operations. However, additional data were extracted from the original paper PCCC forms, such as

- procedural information (cardiopulmonary bypass time, circulatory arrest time, thymectomy, duration of exposure to radiation during catheterization procedures),
- hemodynamic findings at cardiac catheterization,
- description of operation and of devices used (pacemaker, prosthetic valve and conduit types and sizes).

At the time of the development of the PCCC, a system of coding that accurately defined congenital cardiac anomalies and allowed for clear communication between caregivers had not been adequately developed. After reviewing various classification schemes for congenital heart disease, including the International Classification of Diseases [18] of the World Health Organization, it was concluded that these systems were either too broad or imprecise. Therefore, a new coding system was developed that included separate codes for diagnoses and procedures. The coding system was developed with the goals of

- providing a detailed description of the cardiac lesion,
- evaluating the complexity of the operation,
- ensuring the completeness of the data, and
- creating a platform for collaboration between disciplines.

Codes are 5 digits in length, with each additional digit defining more precise information about anatomy and surgical intervention (Appendices 18.1 and 18.2 provide the diagnostic and procedural codes of the PCCC for atrial septal defects). For example, a diagnostic code of 13100 indicated an atrial septal defect; 13120 indicated a secundum-type atrial septal defect; while 13121 indicated a



Pediatric Cardiac Care Consortium

CONGENITAL HEART DISEASE REGISTRY FORM

UNIVERISTY OFFICE PLAZA, SUITE 1307
2221 UNIVERSTIY AVENUE S.E. - MINNEAPOLIS, MN 55414
PHONE: (612) 625-2475 - FAX (612) 624-6586

DATE REC'D.
ENTRY 1
ENTRY 2
VERIFIED

FORM NO.
PCCC STAFF ONLY FOR FILING
NP
OR PREVIOUSLY REPORTED YEAR
PP (YEAR) ON FORM NO.

PATIENT IDENTIFICATION

REGISTRY NUMBER (FOR INTERNAL USE ONLY)
012908PANKMN

COUNTY
Dakota

SEX
Male

BIRTHWEIGHT (IF LESS THAN ONE YEAR AT TIME OF ADMISSION)
3.03 kg

CHECK ONE:
- [x] NEW PATIENT, NOT PREVIOUSLY REPORTED
- [ ] PREVIOUSLY REPORTED PATIENT

PCCC REGISTRY NUMBER CHANGE DOCUMENTATION AREA:

DOES PATIENT HAVE NON-CARDIAC ABNORMALITIES/SYNDROMES?
(•) Yes (INDICATE BELOW) ( ) No ( ) UNKNOWN

CHROMOSOMAL	[ ] DOWN SYNDROME	[ ] TRISOMY 18	[ ] TRISOMY 13	[ ] TURNER'S SYNDROME	[ ] DIGEORGE SYNDROME	[ ] WILLIAMS' SYNDROME	[ ] NOONAN SYNDROME	[ ] OTHER (DESCRIBE)	
OTHER SYND.	[ ] MARFAN'S SYNDROME	[ ] DYSMORPHIC FEATURES	[x] OTHER (DESCRIBE)	PARTIAL CHROMOSONE 18 DELETION					
CNS	[ ] HYDROCEPHALUS	[ ] MYELOMENINGOCELE	[ ] SEIZURES	[ ] DEVELOPMENTAL DELAY	[ ] OTHER (DESCRIBE)				
GI	[ ] TE FISTULA	[ ] MALROTATION	[ ] IMPERFORATE ANUS	[ ] OMPHALOCELE	[ ] DUODENAL ATRESIA	[ ] HIRSCHSPRUNGS	[ ] GERD	[ ] OTHER (DESCRIBE)	
GU	[ ] HYDRONEPHROSIS	[ ] HYPOSPADIAS	[ ] RENAL STRUCTURAL ABNORMALITIES	[ ] OTHER (DESCRIBE)					
RESPIRATORY	[ ] RDS	[ ] CHOANAL ARTESIA	[ ] Absent/Hypoplastic Lung	[x] CLEFT LIP/PALATE	[ ] ASTHMA	[ ] OTHER (DESCRIBE)			
B/J	[ ] POLYDACTYLY	[ ] Absent Hypoplastic Limb/Para	[ ] OTHER (DESCRIBE)						
OTHER	[ ] PREMATURITY	[ ] TWIN BIRTH	[ ] HYPOTHYROIDISM	[ ] ENDOCARDITIS	[ ] KAWASAKI DISEASE	[ ] OTHER (DESCRIBE)			

ADMISSION INFORMATION

HOSPITAL NAME
Fairview University Medical Center

ADMISSION WEIGHT
7.3 kg

ADMISSION DATE
10/22/2008

PREVIOUS CARDIAC SURGICAL PROCEDURES
( ) Yes (INDICATE BELOW) (•) No ( ) UNKNOWN

	TYPE OF SURGICAL PROCEDURE	DATE	CODE
1.			
2.			
3.			
4.			
5.			
6.			
7.			
8.			
9.			

CATH or EPS DATA

[ ] CATH [ ] EPS

CATH or EPS DATE

WEIGHT
kg

	CATH DX 1 PRIMARY
2	
3	
4	
5	
6	
7	
8	
9	
A	
B	
C	
D	
E	
F	

COMPLICATION CODES) 01=NONE

PLEASE REFER TO CATHERIZATION/EPS COMPLICATION CODE LIST FOR ANY RELEVANT COMPLICATIONS AND RECORD CODE(S) BELOW:

SURGERY DATA

[x] PLEASE ATTACH SURGERY REPORT

SURGERY DATE:
10/22/2008

WEIGHT
7.32 kg

	SURG PROCEDURE	SURGERY DX
1		
2		
3		
4		
5		
6		
7		
8		
9		
A		
B		
C		
D		
E		
F		

IF SURGERY ONLY PERFORMED THIS ADMISSION, PLEASE NOTE
PREVIOUSLY REPORTED CATH DATE 09/08/2008
OR INCLUDE COPY OF A CATH REPORT OR ECHOCARDIOGRAM DESCRIBING PATIENT'S CARDIAC DAIGNOSIS.

OUTCOME THIS ADMISSION

[x] DISCHARGE DATE: 10/27/2008

[ ] TRANSFER DATE:
TO:

[ ] DEATH IN HOSPITAL
DATE:

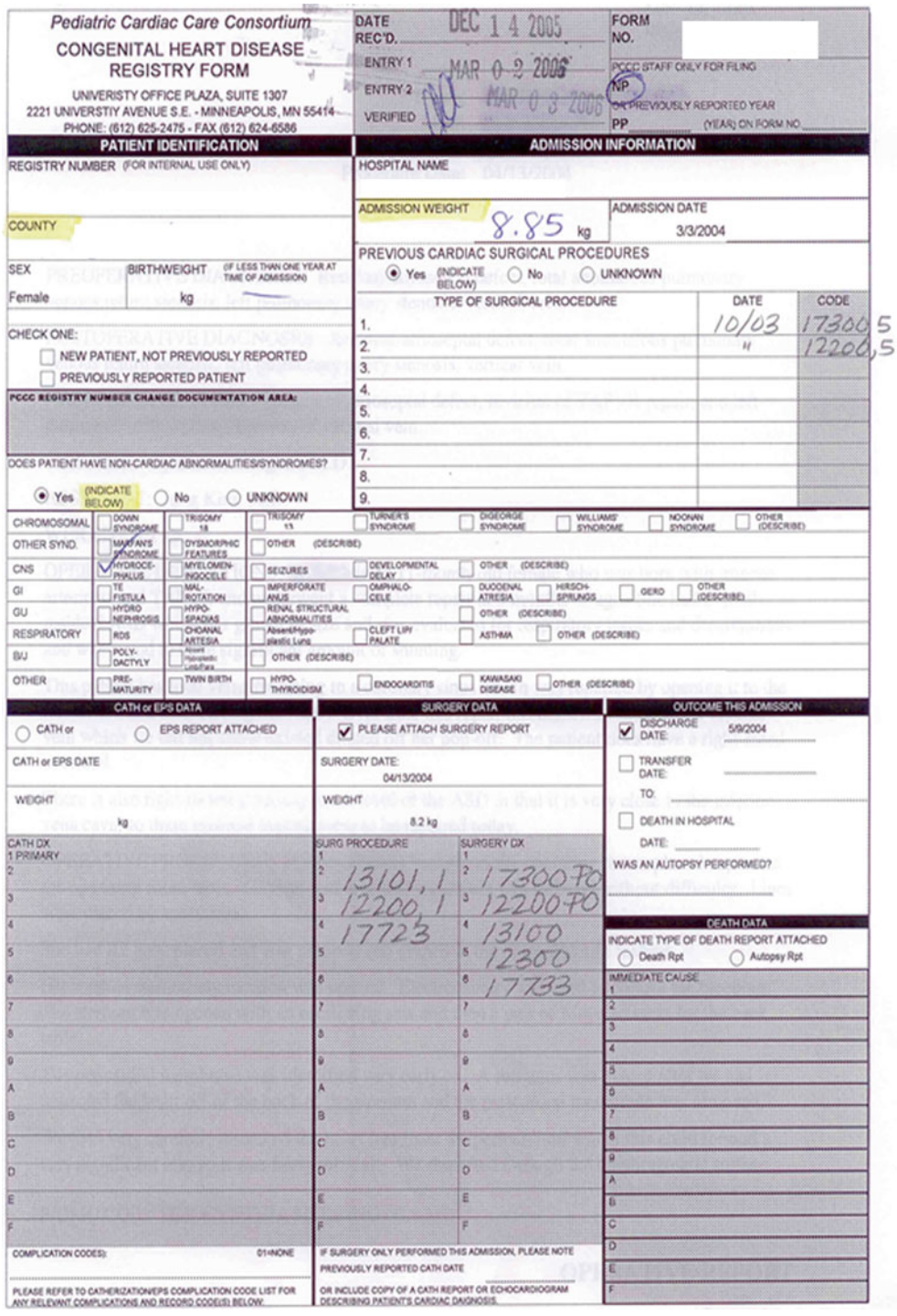
WAS AN AUTOPSY PERFORMED?

DEATH DATA

INDICATE TYPE OF DEATH REPORT ATTACHE
( ) Death Rpt ( ) Autopsy Rpt

	IMMEDIATE CAUSE
1	
2	
3	
4	
5	
6	
7	
8	
9	
A	
B	
C	
D	
E	
F	

Fig. 18.1 PCCC cover sheet



Pediatric Cardiac Care Consortium
CONGENITAL HEART DISEASE REGISTRY FORM
UNIVERISTY OFFICE PLAZA, SUITE 1307
2221 UNIVERSTIY AVENUE S.E. - MINNEAPOLIS, MN 55414
PHONE: (612) 625-2475 - FAX (612) 624-6586

DATE REC'D. DEC 14 2005
ENTRY 1 MAR 02 2006
ENTRY 2 MAR 03 2006
VERIFIED
FORM NO.
PCCC STAFF ONLY FOR FILING
NP
OR PREVIOUSLY REPORTED YEAR
PP (YEAR) ON FORM NO.

PATIENT IDENTIFICATION
REGISTRY NUMBER (FOR INTERNAL USE ONLY)
COUNTY
SEX: Female
BIRTHWEIGHT (IF LESS THAN ONE YEAR AT TIME OF ADMISSION): kg
CHECK ONE:
☐ NEW PATIENT, NOT PREVIOUSLY REPORTED
☐ PREVIOUSLY REPORTED PATIENT
PCCC REGISTRY NUMBER CHANGE DOCUMENTATION AREA:

ADMISSION INFORMATION
HOSPITAL NAME
ADMISSION WEIGHT: 8.85 kg
ADMISSION DATE: 3/3/2004
PREVIOUS CARDIAC SURGICAL PROCEDURES
◉ Yes (INDICATE BELOW) ○ No ○ UNKNOWN

TYPE OF SURGICAL PROCEDURE	DATE	CODE
1.	10/03	17300,5
2.	"	12200,5
3.		
4.		
5.		
6.		
7.		
8.		
9.		

DOES PATIENT HAVE NON-CARDIAC ABNORMALITIES/SYNDROMES?
◉ Yes (INDICATE BELOW) ○ No ○ UNKNOWN

CHROMOSOMAL	☐ DOWN SYNDROME	☐ TRISOMY 18	☐ TRISOMY 13	☐ TURNER'S SYNDROME	☐ DIGEORGE SYNDROME	☐ WILLIAMS' SYNDROME	☐ NOONAN SYNDROME	☐ OTHER (DESCRIBE)
OTHER SYND.	☐ MARFAN'S SYNDROME	☐ DYSMORPHIC FEATURES	☐ OTHER (DESCRIBE)					
CNS	☑ HYDROCEPHALUS	☐ MYELOMENINGOCELE	☐ SEIZURES	☐ DEVELOPMENTAL DELAY	☐ OTHER (DESCRIBE)			
GI	☐ TE FISTULA	☐ MAL-ROTATION	☐ IMPERFORATE ANUS	☐ OMPHALOCELE	☐ DUODENAL ATRESIA	☐ HIRSCH-SPRUNGS	☐ GERD	☐ OTHER (DESCRIBE)
GU	☐ HYDRONEPHROSIS	☐ HYPOSPADIAS	☐ RENAL STRUCTURAL ABNORMALITIES		☐ OTHER (DESCRIBE)			
RESPIRATORY	☐ RDS	☐ CHOANAL ATRESIA	☐ Absent/Hypoplastic Lung	☐ CLEFT LIP/PALATE	☐ ASTHMA	☐ OTHER (DESCRIBE)		
BJ	☐ POLYDACTYLY	☐ Absent/Hypoplastic Limb/Para	☐ OTHER (DESCRIBE)					
OTHER	☐ PREMATURITY	☐ TWIN BIRTH	☐ HYPOTHYROIDISM	☐ ENDOCARDITIS	☐ KAWASAKI DISEASE	☐ OTHER (DESCRIBE)		

CATH or EPS DATA
○ CATH or ○ EPS REPORT ATTACHED
CATH or EPS DATE
WEIGHT: kg
CATH DX 1 PRIMARY
2
3
4
5
6
7
8
9
A
B
C
D
E
F
COMPLICATION CODE(S): 01=NONE
PLEASE REFER TO CATHERIZATION/EPS COMPLICATION CODE LIST FOR ANY RELEVANT COMPLICATIONS AND RECORD CODE(S) BELOW:

SURGERY DATA
☑ PLEASE ATTACH SURGERY REPORT
SURGERY DATE: 04/13/2004
WEIGHT: 8.2 kg

SURG PROCEDURE	SURGERY DX
1	1
2 13101,1	2 17300 PO
3 12200,1	3 12200 PO
4 17723	4 13100
5	5 12300
6	6 17733
7	7
8	8
9	9
A	A
B	B
C	C
D	D
E	E
F	F

IF SURGERY ONLY PERFORMED THIS ADMISSION, PLEASE NOTE PREVIOUSLY REPORTED CATH DATE
OR INCLUDE COPY OF A CATH REPORT OR ECHOCARDIOGRAM DESCRIBING PATIENT'S CARDIAC DIAGNOSIS

OUTCOME THIS ADMISSION
☑ DISCHARGE DATE: 5/9/2004
☐ TRANSFER DATE:
TO:
☐ DEATH IN HOSPITAL
DATE:
WAS AN AUTOPSY PERFORMED?

DEATH DATA
INDICATE TYPE OF DEATH REPORT ATTACHED
○ Death Rpt ○ Autopsy Rpt
IMMEDIATE CAUSE
1
2
3
4
5
6
7
8
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A
B
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D
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F

Fig. 18.1 (continued)

secundum atrial septal defect with a left-to-right shunt. This methodology allows one to search for all diagnoses or procedures included in general categories (i.e. all 131xx codes to capture all atrial septal defects) or to search specifically for very narrow diagnoses (i.e. only secundum atrial septal defects). There are additional procedural qualifying codes which indicated if the operation was a redo, takedown, staged, etc. These qualifying codes make querying an even more specific procedure, for example takedown of a central shunt or redo of total anomalous pulmonary venous return, much easier (Appendix 18.3).

Descriptive statistics were used to describe the annual occurrence, type of patient, case mix, cardiac catheterization experience, operative experience, mortality, and length-of-stay. A statistical test of significance [17] was used to note differences between an individual center and the group. Risk adjustment was performed by utilizing ***Risk Adjusted Classification for Congenital Heart Surgery***, ***version 1*** (RACHS-1). As described elsewhere in this book (in the Chap. 26 by Thiagarajan and Laussen", RACHS-1 is a validated and widely used risk-adjustment system that classifies the congenital cardiac operations into six categories based on expected early mortality rates [19]. The PCCC presented this analyzed information in a yearly report that was distributed to the participating institutions for review. These reports summarized the center's adjusted mortality for each of the procedures for which they have had ten cases during the past 5 years. For each of these operations, a separate report was included, depicting the mean and range of values for the individual center, including nine variables predictive of mortality. This Annual Report allowed a center to compare their experience with the group, particularly for operations for which they have an elevated adjusted mortality.

Contribution to the Field of Congenital Heart Disease

Since the inception of the PCCC, the value of the database as a tool to facilitate research has been well recognized [5–10, 12–14]. Few other multicenter collaborative studies exist in the field, and none have existed for as long a time period as the PCCC [22–31]. To date, the PCCC has enrolled over 100,000 patients and has data over 130,000 procedures (surgeries, catheter-based interventions, and electrophysiological studies) since 1982 (Figs. 18.2, 18.3, and 18.4). A 25 year experience from the PCCC has recently been compiled, in addition to several articles describing composite results for specific operations, cardiac anomalies, or genetic syndromes [[1, 3, 6, 8, 9, 12, 13, 32–34]. Using multi-institutional data collected prospectively over 25 years of pediatric cardiac surgery, the PCCC analyzed trends in post-operative mortality

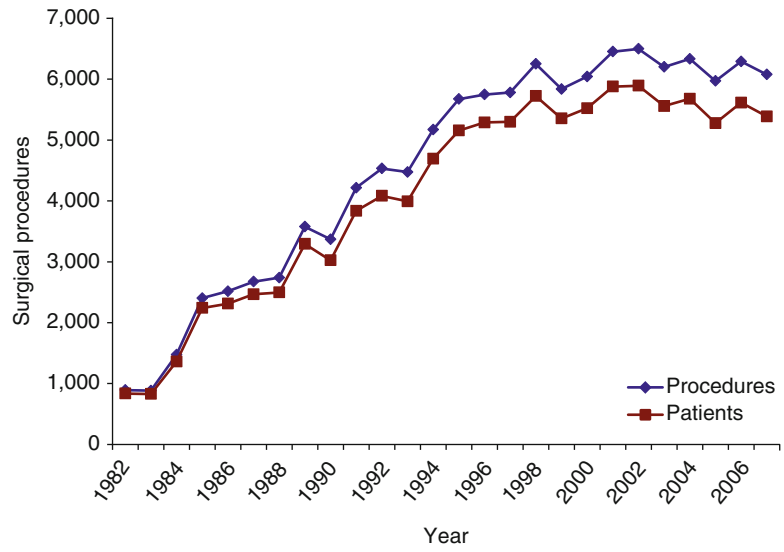

Fig. 18.2 Surgical procedures submitted to PCCC per year

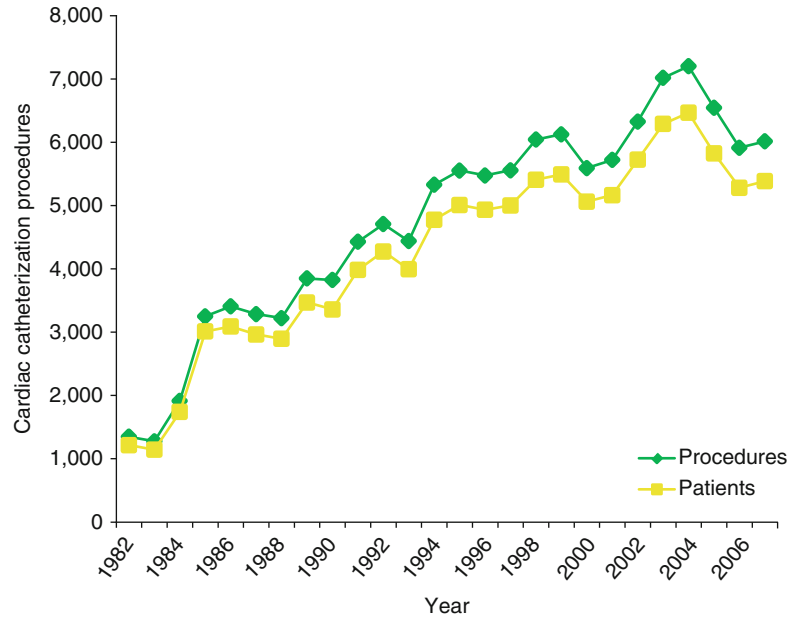

Fig. 18.3 Cardiac catheterization procedures submitted to PCCC per year

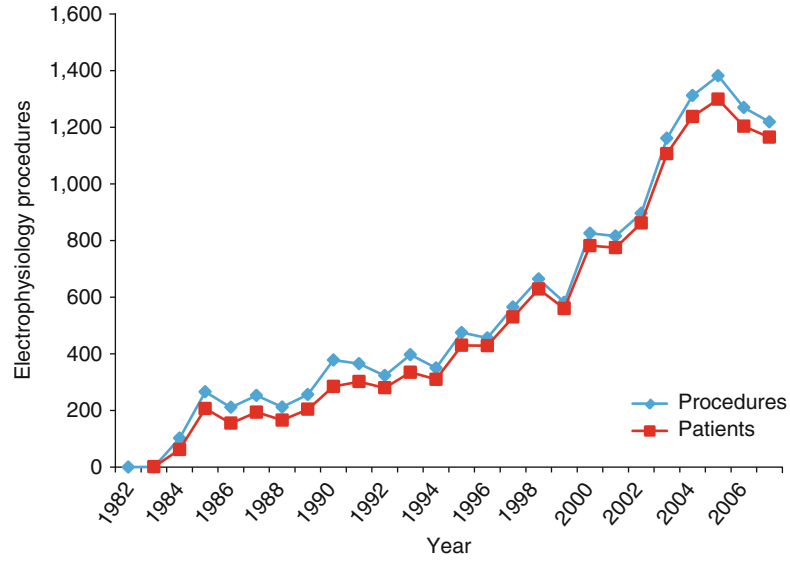

Fig. 18.4 Electrophysiological procedures submitted to PCCC per year

and quantified the influence of risk factors including institutional volume. Overall, survival after pediatric cardiac surgery improved substantially across all age and risk groups except the minimal risk category 1, which has reached a plateau. Over time, the gaps between the different risk categories narrowed, but risk severity remains by far the best predictor of post-operative mortality [35]. Residual age and sex specific risk exist that are not captured by RACHS-1, with younger age and female sex associated with increased risk of death.

The patient specific longitudinal follow-up within the registry data was invaluable when assessing long-term outcomes after operative or catheter intervention for rare forms of congenital cardiac disease. Data from the PCCC have been presented extensively in peer-reviewed manuscripts or scientific meetings, and have contributed significantly to the understanding of the long term outcomes of patients with congenital cardiac disease [6–14].

Future of the PCCC

Because of the increasing number of other nationally recognized congenital cardiac databases, in conjunction with external financial pressures, the executive board of the PCCC elected to stop accepting patient information from member institutions as of January 1, 2012. Letters were posted to each institutional representative and final quality reports were issued. The decision to stop accepting new patients was made in conjunction with a discussion concerning what the future mission of the PCCC should be. The current database has data from over 250,000 procedures stored in both digital and hard copy form. The decision was made to dedicate resources in order to transition the database from an organization that reports measures of comparative procedural outcomes to one that reports research about longitudinal clinical outcomes research. The initial step was to be able to manage the more than 100,000 aging paper forms and notes in a searchable, readily available system of information. An expensive endeavor was initiated which consisted of the organization and digitation of all forms. This process took over 4 months and entailed the scanning of hundreds of thousands of paper documents. The documents included

- operative notes,
- discharge summaries,
- autopsy reports, and
- any additional information that was attached to the record of the patient.

This initiative has recently been completed and the data is currently housed at the University of Minnesota Health System. The database is encrypted and password protected. Data are readily searchable and controlled by both the Division of Pediatric Cardiac Surgery and the Division of Pediatric Cardiology. The database is currently set up with patient-specific data and center-specific data; these data may be utilized by investigators and non-profit organizations independent of their association with the PCCC. Individuals may request, through the Division of Pediatric Cardiac Surgery, specific procedural information on large numbers of patients. With achievement of the current format of the database, multiple studies may be accomplished at a minimal fee. Utilizing these data in a retrospective fashion, many varying research initiatives may be accomplished. These include, but are not limited to:

- comparisons between surgical and catheter-based intervention;
- evaluation of the evolution of surgical techniques addressing complex cardiac defects;
- documentation of improved surgical outcomes for specific diseases.

With the massive amount of patient-specific data and procedure-specific data currently available, another critical important function of the database could be to evaluate long-term survival and outcomes. Ideally, rates of life expectancy and causes of death for patients with the various types of congenital cardiac disease should come from prospective studies of large groups of patients with these conditions and their respective interventions [15]. Currently, these types of studies are not easily feasible; and therefore, the main other source of information regarding the "unnatural" history of congenital cardiac disease remains the clinical registries. Unfortunately, a common drawback of these registries is the limited availability of long-term follow-up. An alternative approach is to enrich existing clinical registries with long-term data from other sources by linking them to these additional databases, such as national death registries, for example. Linking the PCCC with such a registry would overcome this weakness by providing long-term data about mortality and causes of death [16]. The combined post-linking database will provide a unique resource that can be used to evaluate long-term survival as well as major causes or contributing factors of death for patients surviving interventions for congenital cardiac disease.

Conclusion

Over 60 years ago, an organization was created to assist in the treatment of children diagnosed with congenital cardiac disease. Over the decades, this organization has effectively evolved to fit the particular needs of the time. Recently, the mission has changed again to provide a rich source of retrospective data that can provide an accurate picture of the evolution of the care of children with congenital cardiac disease.

Appendix 18.1: Diagnostic Codes of Atrial Septal Defect

1,3,1	Atrial septal defect
1,3,1,0	Atrial septal defect (unspecified type)
1,3,1,0,1	Atrial septal defect + LT-RT shunt
1,3,1,0,2	Atrial septal defect + RT-LT shunt
1,3,1,0,3	Atrial septal defect + bidirectional shunt
1,3,1,1	Patent foramen ovale
1,3,1,1,1	Patent foramen ovale + LT-RT shunt
1,3,1,1,2	Patent foramen ovale + RT-LT shunt
1,3,1,1,3	Patent foramen ovale + bidirectional shunt
1,3,1,2	Secundum atrial septal defect
1,3,1,2,1	Secundum atrial septal defect + LT-RT shunt
1,3,1,2,2	Secundum atrial septal defect + RT-LT shunt
1,3,1,2,3	Secundum atrial septal defect + bidirectional shunt
1,3,1,3	Sinus venosus atrial septal defect
1,3,1,3,1	Sinus venosus atrial septal defect + LT-RT shunt
1,3,1,3,2	Sinus venosus atrial septal Defect + RT-LT shunt
1,3,1,3,3	Sinus venosus atrial septal defect + bidirectional shunt
1,3,1,4	Raghib atrial septal defect (coronary sinus ASD)
1,3,1,4,1	Raghib atrial septal defect + LT-RT shunt
1,3,1,4,2	Raghib atrial septal defect + RT-LT shunt
1,3,1,4,3	Raghib atrial septal defect + bidirectional shunt
1,3,1,5	Inferior vena caval type atrial septal defect
1,3,1,5,1	Inferior vena caval type atrial septal defect + LT-RT shunt
1,3,1,5,2	Inferior vena caval type atrial septal defect + RT-LT shunt
1,3,1,5,3	Inferior vena caval type atrial septal defect + bidirectional shunt
1,3,1,6	Common atrium
1,3,1,7	Created atrial septal defects
1,3,1,7,1	Rashkind balloon septostomy
1,3,1,7,2	Blalock-Hanlon closed atrial septectomy
1,3,1,7,3	Open atrial septectomy
1,3,1,7,5	Transcatheter closure of ASD
1,3,1,7,6	Stent open ASD/atrial baffle
1,3,1,7,9	Dislodged ASD device
1,3,1,8	Fossa ovalis
1,3,1,8,1	Excision of aneurysm of fossa ovalis
1,3,1,9	Closure of adjustable ASD

Appendix 18.2: Surgical Codes of Atrial Septal Defect

1,3,1	Atrial septum surgery
1,3,1,0	Atrial septal defect closure
1,3,1,0,1	Atrial septal defect suture closure
1,3,1,0,2	Atrial septal defect patch closure
1,3,1,1	Patent forman ovale closure
1,3,1,1,1	Patent forman ovale suture closure
1,3,1,1,2	Patent forman ovale patch closure
1,3,1,2	Secundum atrial septal defect closure
1,3,1,2,1	Secundum atrial septal defect suture closure
1,3,1,2,2	Secundum atrial septal defect patch closure
1,3,1,3	Sinus venosus atrial septal defect closure
1,3,1,3,1	Sinus venosus atrial septal defect suture closure
1,3,1,3,2	Sinus venosus atrial septal defect patch closure
1,3,1,4	Raghib atrial septal defect closure
1,3,1,4,1	Raghib atrial septal defect suture closure
1,3,1,4,2	Raghib atrial septal defect patch closure
1,3,1,5	IVC type atrial septal defect closure
1,3,1,5,1	IVC type atrial septal defect suture closure
1,3,1,5,2	IVC type atrial septal defect patch closure
1,3,1,6	Common atrium closure
1,3,1,7	Creation of atrial septal defect
1,3,1,7,1	Rashkind balloon atrial septostomy
1,3,1,7,2	Blalock-Hanlon closed atrial septectomy
1,3,1,7,3	Open atrial septectomy
1,3,1,8	Create atrial septum (partition atrium)
1,3,1,8,1	Excision of aneurysm of fossa ovalis
1,3,1,9	Closure of adjustable ASD

Appendix 18.3: Qualifying Codes

1. Redo
2. Secondary procedure
3. Staged
4. Palliative
5. Corrective
6. Takedown
I Interventional cath procedure
PO postop diagnosis

References

1. Moller JH, Borbas C. The Pediatric Cardiac Care Consortium: a physician-managed clinical review program. QRB Qual Rev Bull. 1990;16(9):310–6.
2. Moller JH et al. Pediatric cardiac care consortium. Demonstrated value of a physician-directed quality assessment system. Minn Med. 1990;73(6):26–32.
3. Moller JH et al. The pediatric cardiac care consortium – revisited. Jt Comm J Qual Improv. 1994;20(12):661–8.
4. Alboliras ET et al. Left ventricular growth in selected hypoplastic left ventricles: outcome after repair of coarctation of aorta. Ann Thorac Surg. 1999;68(2):549–55.
5. Mulder TJ et al. A multicenter analysis of the choice of initial surgical procedure in tetralogy of Fallot. Pediatr Cardiol. 2002;23(6):580–6.
6. Nicolas RT et al. Early outcome after Glenn shunt and Fontan palliation and the impact of operation during viral respiratory season: analysis of a 19-year multi-institutional experience. Ann Thorac Surg. 2005;79(2):613–7; discussion 617.
7. Toro-Salazar OH et al. Long-term follow-up of patients after coarctation of the aorta repair. Am J Cardiol. 2002;89(5):541–7.
8. Tucker EM et al. Permanent pacemaker for atrioventricular conduction block after operative repair of perimembranous ventricular septal defect. J Am Coll Cardiol. 2007;50(12):1196–200.
9. Caldarone CA et al. Long-term survival after mitral valve replacement in children aged <5 years: a multi-institutional study. Circulation. 2001;104(12 Suppl 1):I143–7.
10. Hokanson JS, Moller JH. Significance of early transient complete heart block as a predictor of sudden death late after operative correction of tetralogy of Fallot. Am J Cardiol. 2001;87(11):1271–7.
11. Gatzoulis MA et al. Risk factors for arrhythmia and sudden cardiac death late after repair of tetralogy of Fallot: a multicentre study. Lancet. 2000;356(9234):975–81.
12. Pham PP et al. Cardiac catheterization and operative outcomes from a multicenter consortium for children with Williams syndrome. Pediatr Cardiol. 2009;30(1):9–14.
13. Bartlett HL et al. Early outcomes of tricuspid valve replacement in young children. Circulation. 2007;115(3):319–25.
14. Hills C et al. Cri du chat syndrome and congenital heart disease: a review of previously reported cases and presentation of an additional 21 cases from the Pediatric Cardiac Care Consortium. Pediatrics. 2006;117(5):e924–7.
15. Hoffman JI. Natural history of congenital heart disease. Problems in its assessment with special reference to ventricular septal defects. Circulation. 1968;37(1):97–125.
16. Morales DL, McClellan AJ, Jacobs JP. Empowering a database with national long-term data about mortality: the use of national death registries. Cardiol Young. 2008;18 Suppl 2:188–95.
17. Goldmuntz E et al. Evaluation of potential modifiers of the cardiac phenotype in the 22q11.2 deletion syndrome. Birth Defects Res A Clin Mol Teratol. 2009;85(2):125–9.
18. Welke KF et al. The relationship between hospital surgical case volumes and mortality rates in pediatric cardiac surgery: a national sample, 1988–2005. Ann Thorac Surg. 2008;86(3):889–96; discussion 889–96.
19. Jenkins KJ. Risk adjustment for congenital heart surgery: the RACHS-1 method. Semin Thorac Cardiovasc Surg Pediatr Card Surg Annu. 2004;7:180–4.
20. Salvin JW et al. Blood transfusion after pediatric cardiac surgery is associated with prolonged hospital stay. AnnThorac Surg. 2011;91(1):204–10.
21. Moller JH, Anderson RC. 1,000 consecutive children with a cardiac malformation with 26- to 37-year follow-up. Am J Cardiol. 1992;70(6):661–7.
22. Jacobs JP. Software development, nomenclature schemes, and mapping strategies for an international pediatric cardiac surgery database system. Semin Thorac Cardiovasc Surg Pediatr Card Surg Annu. 2002;5:153–62.
23. Jacobs JP et al. Creating a database with cardioscopy and intra-operative imaging. Cardiol Young. 2005;15 Suppl 1:184–9.
24. Jacobs JP et al. The nomenclature, definition and classification of discordant atrioventricular connections. Cardiol Young. 2006;16 Suppl 3:72–84.
25. Jacobs JP et al. Current status of the European Association for Cardio-Thoracic Surgery and the Society of Thoracic Surgeons Congenital Heart Surgery Database. Ann Thorac Surg. 2005;80(6):2278–83; discussion 2283–4.
26. Jacobs JP et al. Nomenclature and databases for the surgical treatment of congenital cardiac disease – an updated primer and an analysis of opportunities for improvement. Cardiol Young. 2008;18 Suppl 2:38–62.
27. Jacobs JP, Maruszewski B. Computerized outcomes analysis for congenital heart disease. Curr Opin Pediatr. 2005;17(5):586–91.
28. Jacobs JP et al. The current status and future directions of efforts to create a global database for the outcomes of therapy for congenital heart disease. Cardiol Young. 2005;15 Suppl 1:190–7.
29. Jacobs JP et al. Nomenclature and databases – the past, the present, and the future: a primer for the congenital heart surgeon. Pediatr Cardiol. 2007;28(2):105–15.
30. Jacobs JP, Wernovsky G, Elliott MJ. Analysis of outcomes for congenital cardiac disease: can we do better? Cardiol Young. 2007;17 Suppl 2:145–58.
31. Jacobs ML et al. Databases for assessing the outcomes of the treatment of patients with congenital and paediatric cardiac disease – the perspective of cardiac surgery. Cardiol Young. 2008;18 Suppl 2:101–15.
32. Moller JH. Congenital heart diseases and coronary artery anomalies: a comparison of experiences. Cleve Clin J Med. 1990;57(2):178–80.

33. Powell CB et al. Operative mortality and frequency of coexistent anomalies in interruption of the aortic arch. Am J Cardiol. 1997;79(8):1147–8.
34. Raghuveer G et al. Predictors of prosthesis survival, growth, and functional status following mechanical mitral valve replacement in children aged <5 years, a multi-institutional study. Circulation. 2003;108 Suppl 1:II174–9.
35. Al-Radi OO et al. Case complexity scores in congenital heart surgery: a comparative study of the Aristotle Basic Complexity score and the Risk Adjustment in Congenital Heart Surgery (RACHS-1) system. J Thorac Cardiovasc Surg. 2007;133(4):865–75.

Pediatric Cardiac Catheterization Databases

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Joshua P. Kanter, Lisa Bergersen, Sandra Coombs, Thomas J. Forbes, Allen D. Everett, and Gerard R. Martin

Abstract

This review details the major database efforts in the field of congenital cardiac catheterization. Four database projects have been developed that relate to cardiac catheterization procedures in adult and pediatric patients with congenital heart disease. The IMPACT Registry™ (IMproving Pediatric and Adult Congenital Treatment) of the National Cardiovascular Data Registry of The American College of Cardiology Foundation, the Congenital Cardiac Catheterization Project on Outcomes (C3PO), the Congenital Cardiovascular Interventional Study Consortium (CCISC), and the Mid-Atlantic Group of Interventional Cardiology (MAGIC) Catheterization Outcomes Project represent the future of evidence based practice in congenital cardiac catheterization, and demonstrate the progress in collecting accurate information to define clinical outcomes and promote quality improvement.

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The field of pediatric cardiology has seen tremendous therapeutic advancement in the last 40 years both in the scope of surgical intervention as well as catheter based intervention. Some of the greatest developments in the care of patients with congenital heart disease have occurred in the field of diagnostic and interventional cardiac catheterization. Cardiac catheterization was predominantly a diagnostic procedure in previous eras but now approximately 2/3 of all catheterization procedures performed on patients with congenital heart disease is interventional in nature. With this evolution, the evaluation of clinical efficacy, outcomes, and adverse events has been made primarily through prospective clinical trials, retrospective reviews, case reports and expert consensus opinion. These experiences have been limited by the heterogeneity of the patient population as well as the small patient numbers involved. In order to develop better evidence-based approaches to diagnostic and interventional catheterization procedures, the last 10 years have seen significant development of multi-center database registry projects to specifically define adverse event rates, develop risk stratification methodologies and to assess procedural efficacy of specific interventions.

Since 2003, four database projects have been developed that relate to cardiac catheterization procedures in adult and pediatric patients with congenital heart disease. The IMPACT Registry™ (IMproving Pediatric and Adult Congenital Treatment) of the National Cardiovascular Data Registry of The American College of Cardiology Foundation, the Congenital Cardiac Catheterization Project on Outcomes (C3PO), the Congenital Cardiovascular Interventional Study Consortium (CCISC), and the Mid-Atlantic Group of Interventional Cardiology (MAGIC) Catheterization Outcomes Project represent the future of evidence-based practice in the field of interventional pediatric cardiology. Each project has taken a different approach to database design, site inclusion, data collection methods, and data analysis, yet together they begin to describe the current practice in congenital cardiac catheterization laboratories across the clinical community.

IMPACT Registry™ (IMproving Pediatric and Adult Congenital Treatment)

Overview

The IMPACT Registry (IMproving Pediatric and Adult Congenital Treatment) is a project of the National Cardiovascular Data Registry (NCDR) of the American College of Cardiology Foundation. In 2007, two NCDR committees were formed with the task of developing a national data registry to evaluate and improve quality for diagnostic and interventional cardiac catheterization procedures on adult and pediatric patients with congenital heart disease, a Steering Committee and a Data Workgroup. The Steering Committee established the goals and strategic direction of the registry, while the Data Workgroup set out to develop the specific data elements to be collected and the quality metrics to be analyzed. Committee members represented multiple professional organizations including the American College of Cardiology, the Society for Cardiovascular Angiography and Interventions, the Society of Thoracic Surgeons (STS), the American Academy of Pediatrics, and the US Food and Drug Administration. The dataset has been harmonized with the STS surgical database, to facilitate the future collection of longitudinal data to better understand outcomes of both surgical and catheter based procedures [1].

As designed, the IMPACT Registry collects a limited dataset on all catheterization procedures

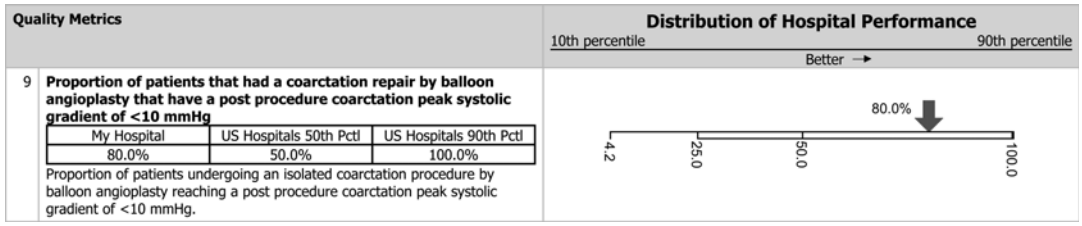

Fig. 19.1 Box and whisker plot – This plot is an example of the outcome metric reporting for a single hospital site for coarctation balloon angioplasty. The *arrow* denotes the site specific quality metric result, while the national percentiles are arrayed on the horizontal axis. This site obtained a post angioplasty peak systolic gradient <10 mmHg in 80 % of their procedures (Reprinted with permission from the National Cardiovascular Data Registry)

performed that include pre-procedural patient details, procedural details and major intra and post-procedural adverse events. A more detailed dataset is then collected on six specific interventional procedures that include ASD device closure, PDA closure, pulmonary and aortic valvuloplasty, proximal pulmonary artery stenting, and balloon/stent angioplasty for coarctation of the aorta. These data elements inform the primary outcome metrics of the registry [2]. The database went live with seven sites enrolled in 2010, and by the second quarter of 2013 had 83 sites with more than 25,000 patient records entered into the database.

Data Collection and Reporting

NCDR developed a web-based tool that sites utilize for data entry, or as an alternative, sites can purchase NCDR approved software from third party vendors to facilitate automated data entry. The IMPACT web-tool utilizes password protected, encrypted technology to insure the safety of patient data. Sites have the option of excluding all direct patient identifiers from the quarterly data upload. The procedural data is entered by the primary operator or a designee for all catheterization procedures at that institution. As each case is entered, a quality check is performed that insures that data is valid and within acceptable ranges. Data from each site is uploaded to the full registry database on a quarterly basis, at which time a more thorough data quality check (data quality review or DQR) is performed to assess data completeness. Data elements have been designated as core or supporting. Core elements feed into the quality metrics, and require a higher data completeness threshold than supporting elements. Quarterly data is then given a red, yellow or green designation whereby red data has failed the DQR and will not be included in the database. Yellow data has met certain completeness criteria to be included in the database, but is still missing key data elements and is not included in the national quality metric benchmarks. Green data is deemed complete, and is included in the national database and all data reports. Sites receive a quarterly Outcomes Report that summarizes the site's individual performance and compares them to the entire IMPACT Registry population. Comparison analysis and reporting of major adverse events is performed for all cases, and specific outcome metrics are reported for the six specific interventional procedures. On-site audits are also conducted to verify the accuracy and completeness of data entered into IMPACT as compared to the given patient's medical record.

Quarterly outcomes reports are distributed, with reporting of site specific quality metrics. Metrics are reported with a "box and whisker" plot (Fig. 19.1), a visual tool that allows easy comparison of site specific outcomes to percentile scores based on the national aggregate data. The data presented includes rates of major adverse event or death for diagnostic procedures, sub-grouped by patient age, as well as the quality metrics for the six interventional procedures. These include residual gradients, post intervention valve insufficiency, residual shunts and rates of device embolization.

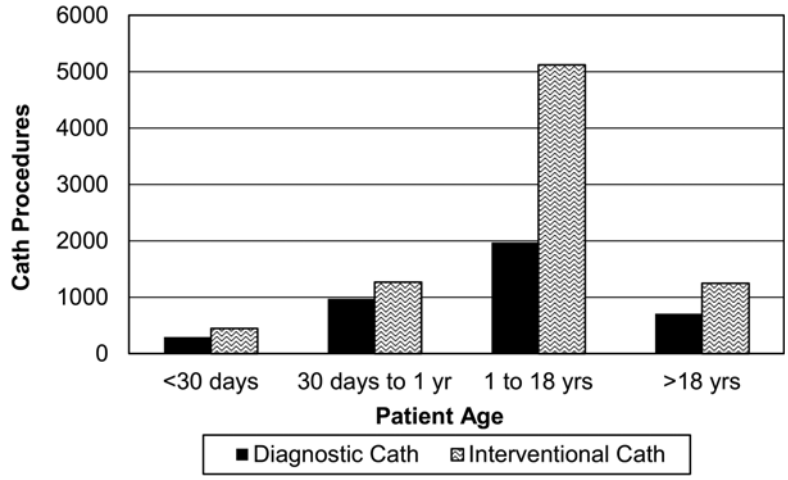

Fig. 19.2 Cath lab procedure type and patient age (Reprinted with permission from et al. [3])

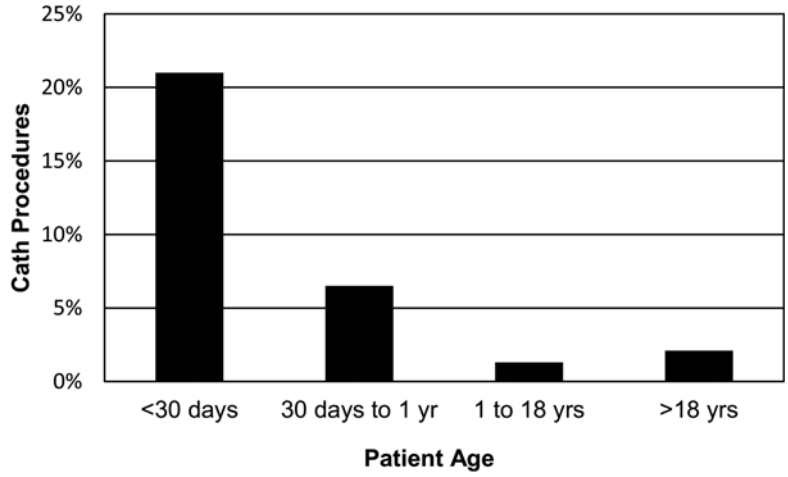

Fig. 19.3 Rates of major adverse event or death by patient age (Reprinted with permission from Vincent et al. [3])

Data

Analysis of the first seven quarters of the IMPACT database (January 2011 – September 2012) saw a significant increase in site enrollment, from 12 sites in Q1-2011 to 70 sites in Q4-2012. There were over 12,000 cath lab visits that achieved either green or yellow status in the DQR by September 2012. Of these, 32.8 % were purely diagnostic and 67.2 % included an intervention. Figure 19.2 shows the proportion of diagnostic and interventional cath procedures by patient age at the time of the procedure.

The overall rate of major adverse event (MAE) or death during a diagnostic catheterization was 4.4 %. MAE includes: cardiac arrest, tamponade requiring drainage, arrhythmia requiring intervention, air embolus, embolic stroke, pacemaker implantation, unplanned cardiac surgery (due to a cath complication) and death. Rates of MAE were highest in the youngest patients, and fell with increased patient age [3]. The IMPACT Registry does not attribute cause with the adverse event or death, except in the case of an unplanned cardiac surgery due to a cath complication. Therefore, the reported events are not necessarily related to the catheterization procedure. Adverse events or death that occur within the requisite time frame after the catheterization procedure are included and reported, as seen in Fig. 19.3.

Future Directions

Database development requires constant analysis and assessment of the data elements themselves, in order to tailor the dataset to the desired outcome measures. During the first eight quarters, there has been an ongoing effort by the Steering Committee and NCDR staff to identify data elements in need of revision in anticipation of IMPACT version 2.0. Furthermore, version 2.0 will see the inclusion of two new interventional modules; MAP-IT (**M**ulticenter Pediatric and **A**dult Congenital E**P** Qual**it**y), a module to assess electrophysiologic procedures in the pediatric and adult congenital patient population and a module to assess Transcatheter Pulmonary Valve implantation in the same patient population. Work is also ongoing to include a risk stratification methodology such as the CHARM (Congenital Heart Disease Adjustment for Risk Method) model derived from the C3PO database project. This will facilitate a more comprehensive and accurate comparison of adverse event rates across institutions and individual physician operators. In the longer term, there is an effort to harmonize the IMPACT database with other national databases such as the Congenital Heart Surgery Database of the Society of Thoracic Surgeons and the Pediatric Cardiac Care Consortium Database. Efforts have been made to harmonize the database standards used in IMPACT with the database standards used by the Society of Thoracic Surgeons (STS) Congenital Heart Surgery Database and the European Association for Cardio-Thoracic Surgery (EACTS) Congenital Heart Surgery Database. Part of this harmonization is the use of a common nomenclature in all of these databases: the International Pediatric and Congenital Cardiac Code (IPCCC). IMPACT uses the interventional cardiology component of IPCCC, and the several members of the leadership of IMPACT were instrumental in developing the interventional cardiology component of IPCCC [4, 5]. The use of common nomenclature (the IPCCC) and common database standards in IMPACT and the STS Congenital Heart Surgery Database will facilitate the linking of these 2 datasets to answer questions that neither dataset would be able to answer independently. To fully realize the potential of the IMPACT database, the inclusion of longitudinal follow-up patient data will also be critically important, and is a long-term goal of the project.

Congenital Cardiac Catheterization Project on Outcomes (C3PO)

Overview

The Congenital Cardiac Catheterization Project on Outcomes (C3PO) is an ongoing, multi-center registry that began prospectively collecting data from 8 participating centers in 2007. The goals of the C3PO collaborative group included the development of methods to assess and compare outcomes, while exploring measures of procedural efficacy. Utilizing an investigative dataset, collected in the registry between 2007 and 2010, the C3PO project yielded baseline rates for adverse events, defined procedure type risk categories, and identified indicators of hemodynamic vulnerability [6, 7]. This data was then used to develop a risk adjustment model, now known as CHARM (Congenital Heart Disease Adjustment for Risk Method), which made the equitable comparison of adverse event rates among institutions possible [6]. This metric received full endorsement from the National Quality Forum (NQF) in 2012 as a pediatric quality measure. In early 2013 the C3PO collaboration began a new phase of progress Congenital Cardiac Catheterization Project on Outcomes – Quality Improvement (C3PO-QI), and expanded their participation to 15 institutions. The C3PO-QI group is working to move beyond benchmarking and towards improving patient care practices through various quality improvement initiatives.

C3PO Data Collection and Reporting

A web-based application for data entry was created specifically for the C3PO project using

Microsoft Visual Studio NET tools (Microsoft, Redmond, Washington, USA). All data entry was securely protected by secure sockets layer (SSL) encryption and role-based security parameters were built-in to prevent unauthorized access to data. Data entry occurred at the time of the catheterization procedure and was completed by the physician performing the procedure or a designee. Patient and procedural variables as well as the occurrence of adverse events were recorded on all cases performed at the institution.

To validate the data entered, the sponsor provided monthly exception reports to highlight missing data fields or data that was out of range requiring validation. Additionally, the sponsor provided all participating sites a list of all cases that were entered in to the database to verify against their institutional records. To prevent coding variations, all adverse events (AEs) were reviewed by the principal investigator and a designee for proper definition of the seriousness and preventability of the episode. After 15 months of data collection, an independent audit was conducted at each site by the sponsor. A random sample of 10 % of the sites cases were reviewed for accuracy and completeness by comparing information entered in to the database to information in the medical records.

C3PO Development of Outcome Assessment Tools

Procedure-Type Risk Categories

A prior single center retrospective study categorized procedure types by anticipated similar risk of AE occurrence, according to expert opinion, utilizing consensus methodology. Although this worked well, the C3PO group worked to further define the procedure-type risk categories by employing both consensus and empirical methodologies to the investigative dataset (2007–2010) to improve the generalizability of the data. The cases were parsed into the appropriate risk categories (1–6) as defined by Bergersen et al. [8], and in July 2009 the C3PO group collaborated to further empirically define the categories. The outcome of the analysis done by the group was the creation of four procedure risk categories.

Hemodynamic Vulnerability

The previous single-center analysis provided a correlation between supposed hemodynamic indicators and the occurrence of adverse events through the use of subjective definitions and not data models. The C3PO group assessed eight separate hemodynamic variables for inclusion in the final risk method: cardiac index, right ventricular (RV) systolic pressure, RV to systemic pressure ratio, systemic ventricle end-diastolic-pressure, mixed venous saturation, systemic arterial saturation, main pulmonary artery systemic pressure, and main pulmonary artery mean pressure. Multivariable modeling produced four indicators of hemodynamic vulnerability independently related to the occurrence of high severity AEs: systemic ventricular end-diastolic pressure $\geq$18 mmHg, systemic arterial saturation <95 % (or <78 % if single ventricle (SV)), mixed venous saturation <60 % (or <50 % if SV), and pulmonary artery systolic pressure $\geq$45 mmHg (or mean $\geq$17 if SV).

CHARM

The final CHARM model developed by the C3PO group combined the patient and procedural characteristics that correlated to the occurrence of AEs: procedure-type risk category, number of hemodynamic indicators, and age <1 year. Using the CHARM methodology, institutions can compute the predicted probability of an AE for all cases at their institution. The sum of all predicted probabilities gives the expected number of AE occurrences, taking in to consideration the case mix within the data set.

To make a comparison between institutions or physicians at one institution, the standardized adverse event ratio (SAER) must be calculated. To compute the SAER the observed AE rate (number of level 3/4/5 AEs in the dataset divided by total number of cases) is divided by the expected AE rate (expected number of AE occurrences divided by total number of cases) (Fig. 19.4).

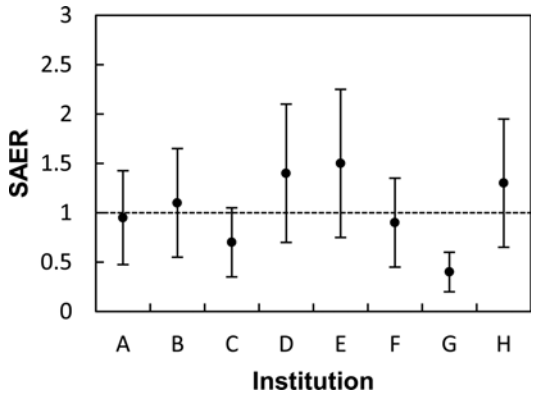

Fig. 19.4 Standardized adverse event ratio (SAER) by institution is shown with bars representing the 95 % confidence interval for each of the eight participating institutions. SAER was calculated by dividing the observed AE rate by the expected for all cases performed at the institution. Institutions C and G had lower AE rates than would be expected given their case mix, whereas institution E had a higher rate than would be expected (Reprinted from Bergersen et al. [6], copyright 2011, with permission from Elsevier.)

Efficacy and Outcome Assessment

In addition to the creation of the CHARM method, the C3PO investigative dataset also yielded measures of procedural efficacy in population subsets. PA rehabilitation was found to be associated with a 10 % incidence of high-level severity AE [9]. Hemodynamic vulnerability, young age, use of cutting balloons, and lower operator experience were significant independent risk factors for procedure-related AE [4]. Hybrid procedures were among the interventions investigated and were found to have a low incidence of major AE occurrence [10]. Procedural success is common and AEs, especially higher severity AEs, are rare for balloon pulmonary valvuloplasty in patients with isolated pulmonary valve stenosis [11]. Cardiac catheterizations involving endomyocardial biopsies can be performed in pediatric heart transplant recipients with a low AE rate and high diagnostic yield [12].

Future Directions

The next phase for C3PO is to move beyond definition toward quality improvement (QI) initiatives aimed at improving the outcomes for all patients undergoing catheterization with congenital heart disease. The group will be expanding participation to include 15 sites and will begin prospectively collecting data in May of 2013. For this new venture, the previously established web based tool has been redesigned utilizing Microsoft© Silverlight (Microsoft, Redmond, Washington, USA) technology and features improved speed, functionality, and updated nomenclature. Institutional data will be available in comparison to other participating C3PO-QI sites in aggregate including procedure counts, limited patient and procedural characteristics, and outcomes such as highest severity adverse events and transfusion rates. Quality reports will include outcome metrics such as risk adjusted adverse event ratios using CHARM. A time series analysis will allow sites to track performance over time by physician and institution in comparison to other sites in aggregate. To portray data in a fashion that is quickly responsive to system improvements, control charts will be created, and for uncommon adverse events, time and/or number of cases between specific adverse events will be reported.

The initial goal of the new C3PO-QI collaboration is to reduce radiation exposure in pediatric cardiac catheterization interventions. To work towards this reduction, established QI scientific methodology will be utilized. Initially, a key driver diagram, a common QI design tool which summarizes the key factors thought to have a potential impact on our goal, and strategies or changes for improvement will be created. Subsequently, PDSA methodology will be employed, which involves (1) proposing changes based on hunches and theories (Plan), (2) implementing the change (Do), (3) measuring or describing the effect (Study), and (4) reviewing and upgrading the process based on what is learned (Act). Participating physicians will receive Maintenance of Certification credit from the American Board of Pediatrics for these activities.

In addition to radiation reduction, the C3PO-QI group will also look at efficacy outcomes for six specific lesion types: pulmonary

stenosis, aortic stenosis, coarctation of the aorta, atrial septal defect, patent ductus arteriosus, and transcatheter pulmonary valve placement.

Congenital Cardiovascular Interventional Study Consortium

Overview

The Congenital Cardiovascular Interventional Study Consortium (CCISC) was started in 2004 by a multi-center group of pediatric cardiologists seeking answers to specific questions encountered in the practice of pediatric interventional cardiology.

The first project related to treatment of coarctation of the aorta in children greater than 4 years of age, comparing surgery vs. balloon angioplasty vs. stent treatment to compare outcomes and rates of complications in order to determine the optimal treatment choice [13, 14]. The second project compared the outcomes of two methods used to augment pulmonary blood flow; stenting the patent ductus arteriosus versus surgical creation of a modified Blalock-Taussig shunt. The third project, as discussed here, set out to develop a method of risk stratification for pediatric cardiac interventions. The focus of this project is twofold: first, to develop a pre-catheterization risk score in an attempt to quantify a particular patient's risk of encountering a significant adverse event (SAE) in the catheterization lab, while the second purpose is to use the risk score to decrease the overall SAE rates in the catheterization lab, particularly in high risk patients, through development of best practice models. This registry was started in January 2008 and currently has over 16,000 patients enrolled from 23 participating institutions. This is a worldwide registry with the majority of sites coming from the United States (n = 15) followed by South America (n = 5) and Western Europe (n = 3).

Data Collection and Reporting

Each patient who presents to the pediatric cardiac catheterization lab for a diagnostic or interventional procedure is entered into the study. Data for each procedure is manually entered at each participating site into a web-based data collection tool by the primary operator or a designee. SAEs that occur within 30 days of the catheterization procedure are entered into the database (Table 19.1). Though every patient undergoing a cardiac catheterization is entered into the CRISP (Catheterization RISk in Pediatrics) registry, only SAE's are considered for analysis. Therefore, hematomas, transient, non-life threatening arrhythmias which didn't require an intervention, or unexpected intubation during the catheterization procedure, are not considered an SAE and therefore are not entered into the registry. By definition, an event is considered significant if it: causes a potentially life-threatening event, results in a prolonged length of stay, or requires further intervention.

Data verification and validation is performed monthly. Quarterly audits are performed by the coordinating center, and include reviews of 20 % of patient charts to assure completeness and data accuracy. Normal value ranges are preprogrammed within the database, and alert the data entry designee of potentially unexpected or incongruent data entry points.

Methodology

Using the first 15,000 patients entered into the registry, a risk stratification score was developed in January 2013. Multiple patient demographic, physiologic and procedural parameters were evaluated, and a risk score developed by expert consensus along with a review of the current literature. This was then further analyzed with logistic regression analysis to tailor and develop a more predictive scoring system, and to eliminate variables that were not strongly predictive. Through multi-variate analysis, parameters which were significant in predicting SAEs were: patient weight, pre-catheterization diagnosis, physiologic parameters, the requirement of inotropic support prior to the catheterization procedure, and procedure type. Using these parameters, a 9-point risk score was generated, categorizing

Table 19.1 Significant adverse events (SAE) as recorded in the CCISC RISK project

General	Vascular	Hemodynamic	Other	Embolic
Death	AV fistula/pseudoaneurysm requiring intervention	Cardiac perforation,	Post-discharge transfusion or device migration	Embolism requiring intervention
Procedurally related cardiac arrest	Arterial compromise requiring intervention	Hemodynamically unstable arrhythmia	Renal compromise	Stroke, new seizure within 48 h of procedure
Cardiac arrest within 24 h of procedure	Venous compromise requiring intervention	Complete heart block that is unresolved at the end of procedure	Brachial plexus injury	
Unplanned CPS/ECMO	Re-bleeding requiring transfusion, retroperitoneal hematoma	Unintentionally retained device, device migration requiring retrieval	Unanticipated transfusion	
Infection (systemic/endocarditis)	Vascular injury requiring intervention	Hemopericardium, Hemothorax,	Extended length of stay	

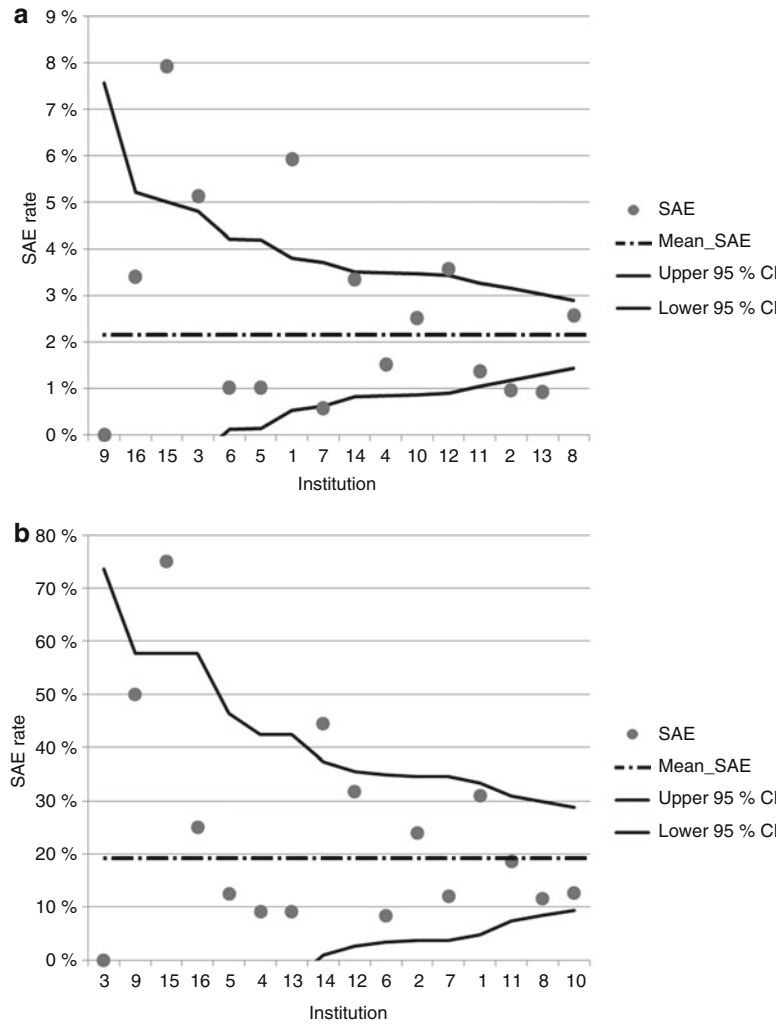

Fig. 19.5 Serious adverse events (SAE) modified funnel plot – Institutions are on the x-axis, with lower volume centers to the left and higher volume centers to the right. The mean event rate is represented by the dashed line and the 95 % confidence intervals by the solid lines. (**a**) Plots the SAE rate for the low risk group of patients. (**b**) Plots the SAE rate for the severe risk group of patients. Note the significantly higher SAE rates in the high risk group. Confidence intervals are wider towards the left as the institutional volume decreases (Reprinted with permission from the Congenital Cardiovascular Interventional Study Consortium)

the patients into low, moderate, or severe risks in encountering a SAE during the catheterization procedure. Modified funnel plots using volume adjusted standard errors of the overall mean are then created to give a brief overview to the institutions as to how they compare to other participating institutions relative to their risk score (Fig. 19.5).

The data is further broken down into specific parameters, for example, patient weight, patient pre-catheterization status, procedure type, both between institutions and within an institution (specifically evaluating the results of each interventionalist within that institution) to see if "clustering" of SAE's is present. Of note, though radiation dose is not considered a SAE, it is also being collected and currently subject to analysis.

Future Directions

Comprehensive process and outcomes failures that lead to harm must be analyzed using a variety of methods that may include Root Cause Analysis, Failure Mode and effect Analysis and Work Domain analysis [15]. The data and lessons should be regularly analyzed every 6 months to verify if a reduction in patient harm and adverse events has been achieved. The two primary areas of focus are decreasing radiation dose and decreasing overall SAE rates in children

<18 years of age. Collection of baseline data was completed in April 2013 with implantation of an RCA scheduled for May 2013.

The American Board of Pediatrics has granted maintenance certification (Maintenance of Certification or MOC) Part IV credit for participation in the RISK registry. The goal is to look specifically at decreasing the overall SAE rate and radiation dosage currently used in the catheterization laboratory.

The Mid-Atlantic Group of Interventional Cardiology (MAGIC) Catheterization Outcomes Project

Overview

The Mid-Atlantic Group of Interventional Cardiology (MAGIC) Catheterization Outcomes Project began in 2003, with the goal of developing a multi-center data registry that utilized automated data entry to facilitate the collection of data associated with cardiac catheterization in congenital heart disease. The registry grew to include 16 sites (15 in the US and one in Belgium) and collected data on eight catheter based procedures (device closure of atrial septal defect (ASD), ventrical septal defect (VSD), patent foramen ovale (PFO) and patent ductus arteriosus (PDA), pulmonary valvuloplasty, balloon angioplasty of aortic coarctation, stent angioplasty of aortic coarctation and pulmonary hypertension) to investigate short-term outcomes, procedural complications and to track long-term follow-up [16].

Data Collection and Reporting

As data collection is a labor intensive process, MAGIC utilized commercially available pediatric catheterization reporting software, PedCath™ (Scientific Software Solution, Charlottesville, VA), to automatically upload data from the individual sites to the central data warehouse. The project developers worked with representatives from the company to tailor the software to the needs of the database. This included the addition of new supplemental data fields and unique study identifiers. Once the data has been completely entered into PedCath™, a de-identified dataset is then uploaded via FTP (file transfer protocol) to the central, secure database [16]. Aggregate data is then shared with the participating sites on a weekly basis.

Results

Atrial Septal Device Closure

The MAGIC registry allowed for the first time the sampling and evaluation of the use of percutaneous therapeutic devices within the community setting after device Food and Drug Administration (FDA) approval. In 2009, MAGIC reported the first multicenter results of the ASD closure arm of the database after FDA approval of the Amplazter atrial septal occluder [17]. Data on 478 patients were submitted to the database and analyzed for the report. Of these patients, 20 did not undergo device implantation, most commonly due to insufficient rims. In those patients who underwent successful device closure, only two had a moderate sized residual shunt at 24 h. There was a 6 % rate of MAE, which included device embolization requiring surgical removal, intracardiac thrombus, and vascular injury. Minor adverse events occurred 4.8 % of the time and included device embolization requiring percutaneous removal, arrhythmia, pericardial effusion, fever, drug reaction, and access site hematoma. There were no deaths in the study population. This paper illustrated the change in practice with introduction of a new non-surgical percutaneous device option. For example a benchmark of the assessment of suitability for surgical ASD closure was determination of the left to right shunt fraction. This assessment was largely abandoned for ASD closure, replaced instead by echo determination of ASD size and right heart dilation. Twenty-nine percent of cases had a Qp/Qs ratio of <1.49, which in the surgical era would generally have not been recommended for closure. Device closure was also extended to smaller children than the 10 kg cut off in the FDA pivotal trial with 6 % of cases <8 kg (Table 19.2).

Table 19.2 Adverse events in the Multicenter MAGIC atrial septal defect (ASD) study, 2004–2007 (n = 454)[a]

	All cases (N = 454)	Simple ASD (n = 405)[b]	Complex ASD (n = 51)	<8 kg (n = 27)	Pulmonary hypertension (n = 26)[c]
Total	27 (6.0 %)	23 (5.7 %)	4 (7.8 %)	2 (7.4 %)	1 (3.9 %)
Major adverse events (total)	5 (1.1 %)	4 (1.0 %)	1 (2.0 %)	–	1 (3.9 %)
Device embolization with surgical removal	3 (0.7 %)	3 (0.7 %)	–	–	–
Intracardiac thrombus	1 (0.2 %)	1 (0.2 %)	–	–	–
Access vessel pseudo-aneurysm, requiring Treatment	1 (0.2 %)	–	1 (2.0 %)	–	1 (3.9 %)
Minor adverse events (total)	22 (4.8 %)	19 (4.7 %)	3 (5.9 %)	2 (7.4 %)	–
Cardiac arrhythmia resolving spontaneously	8 (1.8 %)	7 (1.7 %)	1 (2.0 %)	2 (7.4 %)	–
Cardiac arrhythmia requiring treatment	3 (0.7 %)	3 (0.7 %)	–	–	–
Device embolization with percutaneous removal	5 (1.1 %)	3 (0.7 %)	2 (3.9 %)	–	–
Drug reaction requiring treatment	2 (0.4 %)	2 (0.5 %)	–	–	–
Acidosis, treated	1 (0.2 %)	1 (0.2 %)	–	–	–
Fever	1 (0.2 %)	1 (0.2 %)	–	–	–
Small pericardial effusion	1 (0.2 %)	1 (0.2 %)	–	–	–
Hematoma at access site	1 (0.2 %)	1 (0.2 %)	–	–	–

Reprinted with permission from Everett et al. [17]
[a]Excludes missing complication information (n = 24)
[b]Includes multiple ASDs and patients with a weight ≤9 kg
[c]Pulmonary hypertension: Wood units >7 and/or Rp/Rs ratio ≥0.27. All had a PA mean pressure >25 mmHg

Patent Ductus Arteriosus Device Closure

As there are no guidelines for patent ductus arteriosus (PDA) device closure and multiple devices became available after FDA approval of the Amplatzer ductal occluder (ADO), in 2010 the MAGIC group published the results of 357 PDA device closures from 12 US centers comparing 3 devices, ADO, Gianturco coils (GTC) and Flipper Coils [18]. Of the cases, 82 % were less than 3 mm in diameter. In this sub-group only the ADO (n = 117) and GTC (n = 160) were utilized allowing a direct comparison of efficacy. Although both devices demonstrated equivalent efficacy at PDA closure in the catheterization laboratory, the ADO had a significantly longer fluoroscopy time, contrast volume and significantly greater cost (Table 19.3).

Pulmonary Hypertension

The only non-intervention cohort studied by MAGIC investigators was the catheterization evaluation of children with pulmonary hypertension. As this is such a rare group, a collaborative registry like MAGIC with facilitated data entry was ideal. In 2010 the MAGIC group published the aggregate results of 177 patients evaluated for pulmonary hypertension [19]. The most common pulmonary hypertension groups were pulmonary arterial hypertension associated with congenital heart disease (APAH-CHD) (34 %), idiopathic pulmonary arterial hypertension

Table 19.3 Procedural outcomes with device closure of PDA ≤3 mm

	Gianturco	Amplatzer duct occluder	
No. of cases	160	117	
Fluoroscopy time (minutes, mean + SD)	8.6 ± 7.3	12.4 ± 6.4	$P = 0.00001$
Contrast volume ($cm^3\ kg^{-1}$, mean + SD)	2.6 ± 1.3	3.5 ± 1.7	$P = 0.00001$
None-to-trivial postresidual shunt by angiography	157/159 (98.7 %)	112/117 (95.7 %)	$P = 0.14$
Complications	5/160 (3.1 %)	3/117 (2.6 %)	$P = 0.54$

Reprinted with permission from Brunetti et al. [18]
Comparison of Gianturco coils and the Amplatzer duct occluder from the MAGIC Outcomes Project

Table 19.4 A summary of baseline hemodynamics in patients with pulmonary hypertension

Diagnosis	N (%)	Age (year)	Mean PAP (mmHg)	Systolic PAP (% systemic)	PVRI (WU/m^2)	Rp: Rs
CHD related	61 (34)	0.8 (0.3–2.5)	34.0 (26–46)	67.4 (57–92)	4.7 (3.5–7.6)	0.4 (0.3–0.5)
Idiopathic	36 (20)	4.0 (0.9–11.8)	50.0 (35–67)	87.0 (66–105)	12.0 (6.3–18.9)	0.7 (0.5–1.0)
BPD related	24 (14)	0.8 (0.5–1.3)	33.0 (28–39)	61.9 (50–74)	6.3 (5.0–8.8)	0.4 (0.3–0.6)
Persistent PH of newborn	5 (3)	0.1 (0.1–0.1)	25.0 (18–52)	82.5 (54–108)	4.2 (1.0–4.9)	0.4 (0.2–0.5)
CDH related	8 (4)	1.8 (1.0–4.8)	28.0 (27–36)	56.0 (41–68)	5.9 (5.1–6.4)	0.4 (0.3–0.5)
Assoc. with left heart Dz.	9 (5)	1.9 (0.4–9.0)	36.0 (27–50)	58.5 (55–89)	6.0 (3.3–8.5)	0.3 (0.2–0.5)
Assoc. with pulm. hypoplasia	5 (3)	1.5 (0.8–10.0)	30.0 (23–47)	76.0 (46–92)	7.6 (4.1–8.7)	0.6 (0.3–0.9)
Porto-pulmonary Htn	5 (3)	2.0 (0.5–14)	56.0 (42–66)	79.2 (59–90)	10.1 (3.9–16.4)	0.6 (0.5–0.8)
Newborn GI related	4 (2)	0.3 (0.2–0.5)	23.0 (21–33)	46.7 (37–61)	3.5 (2.3–4.2)	0.3 (0.2–0.4)
Other	20 (11)	2.7 (0.9–12.5)	28.0 (21–35)	52.9 (40–67)	4.5 (3.6–6.9)	0.3 (0.2–0.4)

From the MAGIC Outcomes Project. (Reprinted with permission from Hill et al. [19]
Data represent median (25–75 inter-quartile range)
PAP pulmonary arterial pressure, *PVRI* pulmonary vascular resistance index, *WU* Wood Units, *Rp* Rs, the ratio of pulmonary to systemic vascular resistance, *CHD* congenital heart disease, *BPD* bronchopulmonary dyspnea, *PH* pulmonary hypertension, *CDH* congenital diaphragmatic hernia, *GI* gastrointestinal and refers to PH associated with omphalocele or gastroschisis

(IPAH) (20 %) and bronchopulmonary dysplasia (BPD) related (14 %). There were no deaths reported and adverse events were low (3.9 %). IPAH patients appeared to be more reactive to vasodilators compared to reported adult data and majority had a PAP > 50 % systemic illustrating that PAH in children is different than adults and thus applying adult treatment, classification and outcome strategies to children would be flawed (Table 19.4).

Future Directions

With the development of other databases, MAGIC investigators are only actively enrolling pulmonary hypertension patients into MAGIC. The focus has shifted to collecting follow up data on the over 3,000 cases currently in the database. Reports in progress are on the long the term follow up of children <8 kg with ASD device closure and the predictability of catheterization

laboratory vasodilator testing for long term outcomes in pediatric pulmonary hypertension.

Summary

In the past, the heterogeneity of congenital heart disease coupled with the relatively small number of patients has limited our ability to study outcomes and complications in the cardiac catheterization laboratory. The past 10 years have seen significant progress in the development of multiple, multi-center databases to meet this end. The projects above demonstrate that our community is able to assemble together and set up the infrastructure to contribute accurate and meaningful data to national databases. Risk stratification will be an important aspect of data analysis in this patient cohort, as identification of high risk conditions will influence clinical decisions. Interventional cardiology will require multiple registries to fully understand our patients and our practice. Registries like IMPACT with its high center enrollment and high patient volume will answer broad questions, while registries like C3PO, CCISC and MAGIC will continue to draw from a select group of centers to collect data and answer more specific questions. In addition, we will need to harmonize data across multiple databases related to cardiac surgery, cardiac intensive care and cardiac catheterization in a longitudinal fashion, to better understand the entire treatment of congenital heart disease, and hopefully lead to better outcomes for our patients.

References

1. Jenkins KJ, Beekman Iii RH, Bergersen LJ, Everett AD, Forbes TJ, Franklin RCG, Klitzner TS, Krogman ON, Martin GR, Webb CL. Databases for assessing the outcomes of the treatment of patients with congenital and paediatric cardiac disease – the perspective of cardiology. Cardiol Young. 2008;18 Suppl 2:116–23.
2. Martin GR, Beekman RH, Ing FF, Jenkins KJ, McKay CR, Moore JW, Ringel RE, Rome JJ, Ruiz CE, Vincent RN. The IMPACT registry: IMproving Pediatric and Adult Congenital Treatments. Semin Thorac Cardiovasc Surg Pediatr Card Surg Annu. 2010;13(1):20–5.
3. Vincent RN, Moore J, Benson L, Bergersen L, Holzer R, Jenkins K, Ringel R, Rome J, Michaels J, Martin G. The IMPACT Registry (IMproving Pediatric and Adult Congenital Treatment): update and trends: a report from the NCDR® IMPACT Registry TM. San Francisco, CA; 2013.
4. Bergersen L, Everett AD, Giroud JM, Martin GR, Franklin RCG, Béland MJ, Krogmann ON, Aiello VD, Colan SD, Elliott MJ, Gaynor JW, Kurosawa H, Maruszewski B, Stellin G, Tchervenkov CI, Walters 3rd HL, Weinberg P, Jacobs JP. Report from The International Society for Nomenclature of Paediatric and Congenital Heart Disease: cardiovascular catheterisation for congenital and paediatric cardiac disease (part 1 – procedural nomenclature). Cardiol Young. 2011;21(3):252–9.
5. Bergersen L, Giroud JM, Jacobs JP, Franklin RCG, Béland MJ, Krogmann ON, Aiello VD, Colan SD, Elliott MJ, Gaynor JW, Kurosawa H, Maruszewski B, Stellin G, Tchervenkov CI, Walters HL, Weinberg P, Everett AD. Report from The International Society for Nomenclature of Paediatric and Congenital Heart Disease: cardiovascular catheterisation for congenital and paediatric cardiac disease (part 2 – nomenclature of complications associated with interventional cardiology). Cardiol Young. 2011;21(3):260–5.
6. Bergersen L, Gauvreau K, Foerster SR, Marshall AC, McElhinney DB, Beekman 3rd RH, Hirsch R, Kreutzer J, Balzer D, Vincent J, Hellenbrand WE, Holzer R, Cheatham JP, Moore JW, Burch G, Armsby L, Lock JE, Jenkins KJ. Catheterization for Congenital Heart Disease Adjustment for Risk Method (CHARM). JACC Cardiovasc Interv. 2011;4(9):1037–46.
7. Bergersen L, Gauvreau K, Marshall A, Kreutzer J, Beekman R, Hirsch R, Foerster S, Balzer D, Vincent J, Hellenbrand W, Holzer R, Cheatham J, Moore J, Lock J, Jenkins K. Procedure-type risk categories for pediatric and congenital cardiac catheterization. Circ Cardiovasc Interv. 2011;4(2):188–94.
8. Bergersen L, Gauvreau K, Jenkins KJ, Lock JE. Adverse event rates in congenital cardiac catheterization: a new understanding of risks. Congenit Heart Dis. 2008;3(2):90–105.
9. Holzer RJ, Gauvreau K, Kreutzer J, Leahy R, Murphy J, Lock JE, Cheatham JP, Bergersen L. Balloon angioplasty and stenting of branch pulmonary arteries: adverse events and procedural characteristics: results of a multi-institutional registry. Circ Cardiovasc Interv. 2011;4(3):287–96.
10. Holzer R, Marshall A, Kreutzer J, Hirsch R, Chisolm J, Hill S, Galantowicz M, Phillips A, Cheatham J, Bergerson L. Hybrid procedures: adverse events and procedural characteristics – results of a multi-institutional registry. Congenit Heart Dis. 2010;5(3):233–42.
11. Holzer RJ, Gauvreau K, Kreutzer J, Trucco SM, Torres A, Shahanavaz S, Bergersen L. Safety and efficacy of balloon pulmonary valvuloplasty: a multicenter experience. Catheter Cardiovasc Interv. 2012;80(4):663–72.
12. Daly KP, Marshall AC, Vincent JA, Zuckerman WA, Hoffman TM, Canter CE, Blume ED, Bergersen L.

Endomyocardial biopsy and selective coronary angiography are low-risk procedures in pediatric heart transplant recipients: results of a multicenter experience. J Heart Lung Transplant. 2012;31(4):398–409.
13. Forbes TJ, Kim DW, Du W, Turner DR, Holzer R, Amin Z, Hijazi Z, Ghasemi A, Rome JJ, Nykanen D, Zahn E, Cowley C, Hoyer M, Waight D, Gruenstein D, Javois A, Foerster S, Kreutzer J, Sullivan N, Khan A, Owada C, Hagler D, Lim S, Canter J, Zellers T. Comparison of surgical, stent, and balloon angioplasty treatment of native coarctation of the aorta: an observational study by the CCISC (Congenital Cardiovascular Interventional Study Consortium). J Am Coll Cardiol. 2011;58(25):2664–74.
14. Holzer R, Qureshi S, Ghasemi A, Vincent J, Sievert H, Gruenstein D, Weber H, Alday L, Peirone A, Zellers T, Cheatham J, Slack M, Rome J. Stenting of aortic coarctation: acute, intermediate, and long-term results of a prospective multi-institutional registry – Congenital Cardiovascular Interventional Study Consortium (CCISC). Catheter Cardiovasc Interv. 2010;76(4):553–63.
15. Cassin B, Barach P. Balancing Clinical Team Perceptions of the Workplace: Applying 'work domain analysis' to Pediatric Cardiac Care. Progress In Pediatric Cardiology,10.1016/j.ppedcard.2011.12.005.
16. Everett AD, Ringel R, Rhodes JF, Doyle TP, Owada CY, Holzer RJ, Cheatham JP, Ringewald J, Bandisode V, Chen Y-L, Lim DS. Development of the MAGIC congenital heart disease catheterization database for interventional outcome studies. J Interv Cardiol. 2006;19(2):173–7.
17. Everett AD, Jennings J, Sibinga E, Owada C, Lim DS, Cheatham J, Holzer R, Ringewald J, Bandisode R, Ringel R. Community use of the amplatzer atrial septal defect occluder: results of the multicenter MAGIC atrial septal defect study. Pediatr Cardiol. 2009;30(3):240–7.
18. Brunetti MA, Ringel R, Owada C, Coulson J, Jennings JM, Hoyer MH, Everett AD. Percutaneous closure of patent ductus arteriosus: a multiinstitutional registry comparing multiple devices. Catheter Cardiovasc Interv. 2010;76(5):696–702.
19. Hill KD, Lim DS, Everett AD, Ivy DD, Moore JD. Assessment of pulmonary hypertension in the pediatric catheterization laboratory: current insights from the Magic Registry. Catheter Cardiovasc Interv. 2010;76(6):865–73.

Pediatric Electrophysiology Databases

20

Stephen P. Seslar and John D. Kugler

Abstract

The purpose of this chapter is to provide an overview of catheter ablation databases for outcomes analysis and quality improvement in the field of pediatric and congenital electrophysiology. We begin by discussing unique aspects of electrophysiology that impact data collection and analysis. This will be followed by a review of historic electrophysiology databases and lessons learned. The purpose of this chapter is to review past and present Pediatric and Congenital Electrophysiology Society (PACES) led registry initiatives for assessing the outcomes of catheter ablation procedures for the treatment of arrhythmias in patients with congenital and pediatric cardiac disease. Finally, we set the stage for current and future quality improvement efforts by highlighting the recently created Multicenter Pediatric and Adult Congenital Electrophysiology Quality (MAP-IT) registry and its collaboration with an existing national registry.

Keywords

Electrophysiology • Catheter ablation • Database • Pediatric • Congenital heart disease • Quality improvement

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Introduction

Catheter ablation is a minimally invasive procedure in which specialized catheters are placed into the heart from percutaneous vascular access sites (usually in the groin). Once in position, catheters are used to identify and interrupt critical elements of an arrhythmia's circuitry. Catheter ablation has become the default treatment of choice for arrhythmias in the pediatric and congenital heart disease population. These procedures are generally curative, particularly for

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patients with structurally normal hearts. Early experience culled from registry data [1–3], detailed in a subsequent section of this chapter, have validated these procedures as safe and effective for pediatric and congenital heart disease patients suffering from arrhythmias. When successful, these procedures have been shown in small case series to improve quality of life [4, 5]. It is generally accepted that catheter ablation is cost effective, particularly for highly symptomatic patients when measured against medical therapy or surgical ablation [6]. As healthcare costs rise and the application of this procedure has been extended to less symptomatic individuals, the cost analysis has become more complicated [7]. It is important to recognize, however, that neither cost nor quality of life issues have been addressed in any of the large scale registries to date. In the current environment of healthcare reform and patient-centered care, it will be critical for future databases to address both patient-centered outcomes and overall value to society.

Multicenter clinical registries are increasingly becoming part of the outcomes research and quality improvement landscape. These data repositories have particular value in medical fields where disease entities are inherently rare and the ability to acquire clinical data for meaningful research or quality improvement measures, at any one institution, is limited. Registries play a vital role in procedural subspecialties to define performance benchmarks and improve quality. The Pediatric and Congenital EP Society (PACES) is a nonprofit organization dedicated to the treatment of heart rhythm disorders in children and all individuals with congenital heart disease. The purpose of this chapter is to review past and present PACES led initiatives directed at assessing the outcomes for the treatment of arrhythmias in patients with congenital and pediatric cardiac disease.

Unique Aspects of Outcomes Analysis and Quality Improvement in Electrophysiology

The field of electrophysiology as it pertains to catheter ablation for the treatment of arrhythmias presents some unique challenges in tracking outcomes. First, the targets of catheter ablation (e.g., accessory pathways, atrioventricular nodal pathways, myocardial scar, etc.) are not visible to the unaided eye. During an electrophysiology study, the existence of such entities is inferred from the distortion of the typical electrical patterns of cardiac activation and their participation in spontaneous or inducible arrhythmias. In contrast to the surgeon who places the excised inflamed appendix in the specimen container during the procedure, the electrophysiologist must, in the acute procedural setting, infer from subtler and substantially less reliable clues whether or not the target has been permanently eliminated or only transiently impacted by catheter ablation efforts. In the later instance, the ablation target may reconstitute hours, days or even months later.

The diagnosis of arrhythmias and the tracking of recurrences also have their challenges. For example, arrhythmias are often transient and for the most part, not present during routine electrocardiography performed during office visits. Though ambulatory heart rhythm recordings can be quite effective in detecting arrhythmias, it is often difficult to know the precise mechanism from surface recordings. This can present challenges when documenting and cataloging recurrence after catheter ablation. For example, in some cases patients may have multiple loci addressed during a catheter ablation procedure. In these situations, it can be difficult based on ambulatory recordings to determine which ablation target has recurred. Alternatively, some patients (e.g., those with complex congenital heart disease) have the potential to form new arrhythmia substrates over relatively short periods of time and under unique external conditions. For this reason, it can be difficult to know precisely through non-invasive means whether arrhythmia following catheter ablation represents a recurrence of a previous target or a de novo substrate.

Finally, the field of electrophysiology is dependent on the subjective assessment of the patient as a measure of success. Of the four most frequently cited indications for arrhythmia treatment, by far the most common is “alleviation of symptoms”. It therefore stands to reason that any well designed quality assurance or quality improvement effort in

the field of electrophysiology should feature patient reported outcome measures (PROM). Yet this too is problematic, as symptoms are inherently subjective, non-specific and difficult to quantify. For example, it is often challenging after a procedure to determine whether or not the patient who reports a symptom (e.g. palpitations) actually is having an arrhythmia representing recurrence, sinus tachycardia, or inconsequential atrial or ventricular ectopy.

A patient's report can be impacted by a host of factors that are independent of whether or not the technical objectives of the procedure were successfully met. Patients may not be able to articulate symptoms, depending on the age and verbal ability, and/or the parent's/caretaker's interpretation of the symptoms may not be expressed in a manner that allows the clinician to determine if recurrence has occurred. Furthermore, the parental and child reports may be at odds [8]. Finally, even when patients are able to objectively and accurately report their symptoms, their symptoms may not correlate with a true arrhythmia. The net effect of this and the other issues outlined above is that objective outcomes assessment in the field of electrophysiology is challenging and complex, and contains a degree of inescapable uncertainty.

The History of Electrophysiology Databases

The Pediatric Radiofrequency Catheter Ablation Registry (1991–1999)

The history of catheter ablation outcomes databases for assessing the treatment of arrhythmias in pediatric and congenital cardiac patients dates back to the origins of pediatric electrophysiology as an interventional field, beginning with the Ablation Registry. Under the direction of PACES (known at the time as the Pediatric Electrophysiology Society), a voluntary procedural registry was created to assess the safety and efficacy of catheter ablation in the pediatric population. The only inclusion criterion was patient age less than 21 years. The registry cataloged patient demographic data, indications for ablation, procedural data including fluoroscopy time, acute response to treatment (success or failure), and complications. The follow-up data included recurrence which was defined as a documented return of pre-excitation, tachycardia, or both after acutely successful ablation. The data entry was paper based and forms were submitted from participating institutions to the coordinating center at the University of Nebraska.

The first data in the registry from 24 participating centers were reported in 1994 [1]. All but one center was located in the United States. The age range of this cohort was 20 days to 20.9 years, though the majority of patients in the registry were between 12 and 18 years of age (median 13.5 years). Seventy patients (11 %) had congenital heart disease. The majority of patients underwent catheter ablation due to medically refractory tachycardia, with 30 % of the procedures done at a direct request of the patient. All of the major outcome variables (fluoroscopy time, acute success, and late recurrence) were significantly impacted by the target substrate and medical center experience. The mean fluoroscopy times ranged from 45.9 to 79.6 min with atrioventricular-node reentry tachycardia requiring the shortest exposure and right free-wall accessory pathways the longest. More procedures at a center correlated with shorter fluoroscopy times. Acute success rates ranged from 38 % for ventricular tachycardia, to 89 % for left free wall accessory pathways. Overall, freedom from recurrence in patients without structural heart disease was 71 % after 2 years of follow-up. Patients with left free wall pathways experienced a 78 % freedom from recurrence, whereas in those with right free wall pathways or weight greater than or equal to 80 kg, the number decreased to approximately 50 %. Though acute success was high with ectopic atrial tachycardia, this substrate had the highest recurrence rate. The overall complication rate was low (4.8 % overall, 3.7 % acute) and correlated with body weight of less than 15 kg and institutional volume.

A subsequent study evaluated the expertise "learning curve" of proceduralists associated with catheter ablation in the pediatric population

[9]. The authors demonstrated a remarkable ability to predict performance based on experience and a negative exponential function model using acute outcome and complication data from the registry. For example, a rapid learning curve was demonstrated for left free-wall accessory pathways with a beginning proficiency of 73 % acute success and final proficiency of 94 % with 90 % of learning milestones achieved after *26 cases*. In contrast, right free-wall accessory pathways required 282 cases to achieve the maximal proficiency. The procedure and fluoroscopy times demonstrate a learning curve of intermediate slope. The authors were careful not to suggest imposing performance standards, and acknowledged the large standard deviations and confounders, measured and unmeasured, that might impact the generalizability of the model.

Inadvertent atrioventricular block during radiofrequency catheter ablation was explicitly investigated by Schaffer et al. in 1996 using data from the Ablation Registry [2]. The risk of catheter ablation related heart block was found to be 1.2 % overall and was a function of target location (mid 10 % >anterior 2 % >posteroseptal 1 %) and operator experience.

A follow-up study in 1997 from 46 medical centers participating in the ablation registry looked at outcomes of paroxysmal supraventricular tachycardia (SVT) in those without structural heart disease [3]. The short interval between this and the initial Catheter Ablation registry report in 1994 demonstrated an interesting trend in indications for cardiac ablation. Whereas approximately a third of patients underwent catheter ablation due to patient choice in the original registry report, in this cohort, the number had grown to 58 %. The association between fluoroscopy and procedure times for specific substrates were consistent with the initial report from the registry (e.g., shorter for atrioventricular node reentry tachycardia [AVNRT], longer for right-sided accessory pathways [APs]), but overall, both had decreased in this subsequent report. The acute success rates ranged from 95 % for left lateral pathways, 87 % for septal pathways, and 86 % for right free wall pathways, which was in alignment with the first report from the Ablation registry. Long-term, freedom from recurrence, defined as 3 years, remained consistent at 71 % overall for APs and 77 % for AVNRT. The authors speculated that the relatively high observed recurrence rates may reflect a bias toward under reporting for successful outcomes due to a loss to patient follow-up.

Prospective Assessment After Pediatric Cardiac Ablation (PAPCA) (1999–2003)

Prospective Assessment after Pediatric Cardiac Ablation (PAPCA) was both a prospective study and a clinical registry [10]. The PAPCA Registry, using a prospective and focused study design, set out to determine the incidence in the pediatric population of recurrence following an initially successfully ablated arrhythmia, early and late-appearing cardiac damage due to the ablation procedure, and new arrhythmias attributable to the ablation procedure. For the prospective study, inclusion and exclusion criteria included: age less than 16, no significant congenital heart disease, and only arrhythmia substrates of atrioventricular reentrant tachycardia (AVRT) or AVNRT. The registry portion of PAPCA was the natural extension of the Ablation Registry. The inclusion criteria for the registry portion was consistent with its mission to capture information about the whole of the pediatric population undergoing catheter ablation, and was similar to that of the Ablation Registry discussed above, namely for patients age less than 21 years. The quality control procedures for the prospective study patients were robust with double data entry to eliminate possible keypunch errors. Analysis of the electrophysiologic tracings to determine the mechanism of arrhythmia in 50 % of all cohort patients was done by an independent reviewer. The complications were reviewed by an independent Morbidity and Mortality Review Committee.

Several interesting trends emerged from the PAPCA study and registry data when compared with the earlier ablation registry. First, “patient choice”, as an indication for ablation in this cohort, continued to increase in frequency relative

to ablation registry era, accounting for 90 % of procedures. This, in part, likely reflects an increased awareness of and comfort level with catheter ablation in the pediatric population. There was a modest decrease in the average fluoroscopy time which was down to 28–38 min compared with that reported in the Ablation Registry. Again, this likely reflects the increased experience in the pediatric electrophysiology (EP) community rather than a major change in technology. The overall complication rate (2.9–4.2 %) was comparable to the rate seen in the Ablation Registry cohort. The rate of inadvertent atrioventricular (AV) block as a complication of radiofrequency ablation was unchanged at 1.2 %. In contrast to early reports from the Ablation Registry, no deaths were reported from the PAPCA registry. Interestingly, the acute success rate (for all substrates) was 93 % and essentially unchanged from later reports from the Ablation Registry. This suggested that the learning curve had plateaued. The patterns established for acute success rates as a function of pathway location in the Ablation Registry were reiterated in the results of the PAPCA trial (e.g. 98 % for left free-wall accessory pathways versus 88–90 % for other pathway locations) [11]. With 21 % of the patients lost to follow-up, the 12 month recurrence rate was 10.7 %. Most recurrences (70 %) occurred within 2 months. The rate of recurrence varied by substrate but it should be noted that only accessory pathways and AVNRT substrates were substantially represented in the dataset [12]. A detailed blinded analysis of valve dysfunction, regional wall dyskinesis, and left ventricular function following catheter ablation in PAPCA cohort patients identified little or no evidence for significant injury to cardiac valves or coronary arteries as a result of the radiofrequency catheter ablation [13].

The Multicenter Pediatric and Adult Congenital EP Quality (MAP-IT) Registry

The pioneering efforts discussed above provided invaluable information regarding the outcomes of radiofrequency ablation in the pediatric population. Indeed, even today these registries provide benchmarks for acute procedural success and recurrence rates. However, in the 11 years that have elapsed since the PAPCA registry closed, there have been significant and ongoing improvements in technology and changes in the patient population involved in these procedures, rendering the information progressively more outdated with each passing year. For instance, since 2003 there has been the development and broad dissemination of a new ablation energy source (i.e., cryoablation energy), enhanced three dimensional mapping platforms, and an increasing prevalence of adults with congenital heart disease and related cardiac rhythm problems. The PACES leadership, in response to these issues, sponsored the creation of the Multicenter Pediatric and Adult Congenital EP Quality (MAP-IT) Initiative in 2010. The MAP-IT registry is intended to provide the infrastructure for meaningful quality assurance, ongoing quality improvement and ultimately, a means for conducting multicenter research in the field of pediatric and congenital EP.

The MAP-IT registry was constructed using many of the terms and definitions from the earlier registries discussed above. Other terms and definitions were extracted from existing and credible sources such as the American College of Cardiology/American Heart Association/Heart Rhythm Society (ACC/AHA/HRS) 2006 Key Data Elements and Definitions for Electrophysiological Studies and Procedures [14]. Though the MAP-IT registry has many similarities to its ancestors, it differs in a couple of important aspects. First, for reasons discussed earlier, the MAP-IT registry incorporates patient-centered outcome measures, including a symptoms severity survey (Fig. 20.1) as one of its basic outcome measures. In addition, the MAP-IT registry allows meaningful, anonymized center-to-center comparisons for basic bench marking measures. Following the lessons learned from other procedural registries [15], the creators of the MAP-IT registry recognized the need for a clinical complexity score for electrophysiology procedures. Details of the first iteration of this score, known as the Complexity in Pediatric and

Adult congenital EP Study and ablation Score (COMPASS), were recently published [16]. This score should allow a level playing field from which to compare outcomes between medical centers. The MAP-IT registry will establish quality standards for acute and late technical and patient centered outcomes and adverse events in the field of pediatric and congenital EP.

Question #1

In the past 6 months, what symptom (or feeling) that comes from your (your child's) heart rhythm problem bothers you (your child) most often? (Choose only one.)

- □ None (No symptoms)
- □ Palpitations
- □ Chest Pain
- □ Shortness of breath
- □ Dizziness
- □ Fainting
- □ Fatigue

Question #2

In the last 6 months, about how often have you (or your child) had this feeling? (Choose only one.)

- □ Not applicable (no symptoms)
- □ Every day
- □ At least once per week
- □ At least once per month
- □ At least once in the last 6 months

Question #3

In the past 6 months, what symptom (or feeling) that comes from your (your child's) heart rhythm problem was the worst? (Choose only one.)

- □ None (No symptoms)
- □ Palpitations
- □ Chest Pain
- □ Shortness of breath
- □ Dizziness
- □ Fainting
- □ Fatigue

Question #4

For any heart rhythm episodes you (your child) have had in the last 6 months, what is the *most intense treatment* that you (your child) have endured to try to stop the rhythm problem? (Choose only one.)

- □ I have had no rhythm problem during this time
- □ My rhythm problem is always present and I make no effort to try to stop it.
- □ My rhythm problem episodes stop by themselves without me doing anything.
- □ I have tried bearing-down, standing on my head, blowing into straw, placing ice on my face or other vagal maneuvers.
- □ I went to the emergency department but the rhythm problem stopped by itself or with vagal maneuvers.
- □ I went to the emergency department and I received medicine to treat my rhythm problem.
- □ I stayed in the hospital for one or more days to treat my rhythm problem with medicine.
- □ I went to the hospital / emergency department and I received a shock (DC Cardioversion) to treat my rhythm problem.

Fig. 20.1 Patient symptom severity survey

Question #5

In the past 6 months, which medicine(s) have you (your child) taken for a heart rhythm problem? (Choose all that apply.)

- ◦ I have taken none of these medicines in the past 6 months.
- ◦ Amiodarone
- ◦ Beta‐blocker (atenolol, bisoprolol, metoprolol, nadolol, propranolol, timolol)
- ◦ Digoxin
- ◦ Diltiazem
- ◦ Dofetilide
- ◦ Dronedarone
- ◦ Flecainide
- ◦ Mexiletine
- ◦ Propafenone
- ◦ Sotalol
- ◦ Verapamil

Question #6

In the past 6 months, do you feel your rhythm problem has had interfered with how well you are able to work, go to school, or play?

- □ Yes
- □ No

Question #7

Only answer if catheter ablation procedure has already been performed
Compared to the 6 months before my catheter ablation procedure, in the 6 months following this procedure, I (my child) feel:

- □ Better than before
- □ Worse than before
- □ The Same as before

Fig. 20.1 (continued)

Looking Ahead: MAP-IT Pilot Testing and the IMPACT Registry

The Management Board of the ACC/NCDR approved in February, 2013 the addition of MAP-IT to the next version of the Improving Pediatric and Adult Congenital Treatment (IMPACT) Registry. In preparation for broad scale implementation, 14 centers in the US have been enrolling patients into a pilot version of the MAP-IT Registry. The MAP-IT Pilot Project is an effort to test, validate and refine the empiric constructs assembled in the first phase of the MAP-IT initiative, prior to incorporation of MAP-IT into the National Cardiovascular Data Registry (NCDR) IMPACT registry. The goals of pilot phase of the MAP-IT initiative are: to ensure that the selected data elements capture the critical information necessary to fulfill the mission of the MAP-IT registry (*validate input*), ensure consensus on definitions of terms and choices (*validate input*), establish useful strategies for dealing with data entry into the registry before, during, and after EP studies (*establish best workflow practices*), and develop recommendations on the format and content of the quarterly quality assurance (QA) report for participating centers (*validate output*). The multicenter data collection in the pilot registry has been facilitated through REDCap electronic data capture tools hosted at Seattle Children's Hospital [17]. To date over 500 procedures have been entered and a number of important improvements made to the initial construct. It is anticipated that this pilot dataset will, in turn, be used to further test and refine the COMPASS score.

A European pediatric catheter ablation registry called "Europa" involving six centers (Leiden, Prague, Bergamo, Göttingen, Milano and Leipzig) has been enrolling patients since 2013. These efforts are starting to learn from and share with the US based registries.

Conclusions

Electrophysiology presents several challenges to outcomes assessment and quality improvement. Early databases (Pediatric Radiofrequency Ablation Registry [PRAR] & PAPCA) in the field of pediatric and congenital cardiology established the safety and efficacy of catheter ablation procedures in this patient population. More recent efforts involved in the creation of MAP-IT and its incorporation into the NCDR IMPACT registry will build on earlier database experience. This should provide a robust platform upon which to perform meaningful quality assurance and improvement in the field of pediatric and congenital electrophysiology.

References

1. Kugler JD, Danford DA, Deal BJ, Gillette PC, Perry JC, Silka MJ, Van Hare GF, Walsh EP. Radiofrequency catheter ablation for tachyarrhythmias in children and adolescents. N Engl J Med. 1994;330:1481–7.
2. Schaffer MS, Silka MJ, Ross BA, Kugler JD. Inadvertent atrioventricular block during radiofrequency catheter ablation. Results of the pediatric radiofrequency ablation registry Pediatric electrophysiology society. Circulation. 1996;94:3214–20.
3. Kugler JD, Danford DA, Houston K, Felix G. Radiofrequency catheter ablation for paroxysmal supraventricular tachycardia in children and adolescents without structural heart disease. Pediatric ep society, radiofrequency catheter ablation registry. Am J Cardiol. 1997;80:1438–43.
4. DeMaso DR, Spratt EG, Vaughan BL, D'Angelo EJ, Van der Feen JR, Walsh E. Psychological functioning in children and adolescents undergoing radiofrequency catheter ablation. Psychosomatics. 2000;41: 134–9.
5. Strieper M, Leong T, Bajaj T, Huckaby J, Frias P, Campbell R. Does ablation of supraventricular tachycardia in children with a structurally normal heart improve quality of life? Congenit Heart Dis. 2010; 5:587–93.
6. Case CL, Gillette PC, Crawford FA, Knick BJ. Comparison of medical care costs between successful radiofrequency catheter ablation and surgical ablation of accessory pathways in the pediatric age group. Am J Cardiol. 1994;73:600–1.
7. Gamboa DG, Meguid CR, Kanter RJ. Disproportionate costs and charges for pediatric catheter ablation: supply and demand…or just supply? J Cardiovasc Electrophysiol. 2013;24:170–2.
8. Eiser C, Morse R. A review of measures of quality of life for children with chronic illness. Arch Dis Child. 2001;84:205–11.
9. Danford DA, Kugler JD, Deal B, Case C, Friedman RA, Saul JP, Silka MJ, Van Hare GF, Participating Members of the Pediatric Electrophysiology S. The learning curve for radiofrequency ablation of tachyarrhythmias in pediatric patients. Am J Cardiol. 1995;75:587–90.
10. Van Hare GF, Carmelli D, Smith WM, Kugler J, Silka M, Friedman R, Atkins D, Saul P, Schaffer M, Byrum C, Dunnigan A, Colan S, Serwer G. Prospective assessment after pediatric cardiac ablation: design and implementation of the multicenter study. Pacing Clin Electrophysiol. 2002;25:332–41.
11. Van Hare GF, Javitz H, Carmelli D, Saul JP, Tanel RE, Fischbach PS, Kanter RJ, Schaffer M, Dunnigan ANN, Colan S, Serwer G, Participating Members of the Pediatric Electrophysiology S. Prospective assessment after pediatric cardiac ablation. J Cardiovasc Electrophysiol. 2004;15:759–70.
12. Van Hare GF, Javitz H, Carmelli D, Saul JP, Tanel RE, Fischbach PS, Kanter RJ, Schaffer M, Dunnigan A, Colan S, Serwer G. Prospective assessment after pediatric cardiac ablation: recurrence at 1 year after initially successful ablation of supraventricular tachycardia. Heart Rhythm. 2004;1:188–96.
13. Van Hare GF, Colan SD, Javitz H, Carmelli D, Knilans T, Schaffer M, Kugler J, Byrum CJ, Saul JP. Prospective assessment after pediatric cardiac ablation: fate of intracardiac structure and function, as assessed by serial echocardiography. Am Heart J. 2007;153:815–20, 820.e811–6.
14. Buxton AE, Calkins H, Callans DJ, DiMarco JP, Fisher JD, Greene HL, Haines DE, Hayes DL, Heidenreich PA, Miller JM, Poppas A, Prystowsky EN, Schoenfeld MH, Zimetbaum PJ, Goff DC, Grover FL, Malenka DJ, Peterson ED, Radford MJ, Redberg RF. ACC/AHA/HRS 2006 key data elements and definitions for electrophysiological studies and procedures: a report of the American College of Cardiology/American Heart Association Task Force on Clinical Data Standards (ACC/AHA/HRS Writing Committee to Develop Data Standards on Electrophysiology). J Am Coll Cardiol. 2006;48:2360–96.
15. Jenkins KJ, Gauvreau K, Newburger JW, Spray TL, Moller JH, Iezzoni LI. Consensus-based method for risk adjustment for surgery for congenital heart disease. J Thorac Cardiovasc Surg. 2002;123:110–8.
16. Triedman JK, Pfeiffer P, Berman A, Blaufox AD, Cannon BC, Fish FA, Perry J, Pflaumer A, Seslar SP. Compass: a novel risk-adjustment model for catheter ablation in pediatric and congenital heart disease patients. Congenit Heart Dis. 2013;8:393–405.
17. Harris PA, Taylor R, Thielke R, Payne J, Gonzalez N, Conde JG. Research electronic data capture (redcap) – a metadata-driven methodology and workflow process for providing translational research informatics support. J Biomed Inform. 2009;42:377–81.

Using Data to Drive Improvement and Build the Science of Nursing

21

Ashley Collins, Jean Anne Connor, Sandra Mott, and Patricia Hickey

Abstract

The science and practice of pediatric cardiovascular nursing has made substantial progress since the 1970s. The use of evidence to drive clinical care has demonstrated improvement in patient outcomes including morbidity and mortality. In addition multiple, concurrent events, including dramatic innovations in technologies, medical treatment discoveries, novel surgical procedures, nursing clinical inquiry and organizational support have contributed to increasing health care quality.

Keywords

Nightingale metrics • Nurse-sensitive outcomes • Quality measurement • Process improvement • Healthy work environment • Red Zone • CHEWS • Nursing CAMEO • Consortium of Congenital Cardiac Care • Measurement of Nursing Practice (C4-MNP)

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Introduction

The science and practice of pediatric cardiovascular nursing has made substantial progress since the 1970s. Use of evidence to drive clinical care has demonstrated improvement in patient outcomes including morbidity and mortality. In addition multiple, concurrent events, including dramatic innovations in technologies, discoveries of new medical treatments, novel surgical procedures, nursing clinical inquiry and organizational support have contributed to increasing health care quality.

Use of advanced technologies and innovative medical therapies have been cited by the Institute of Medicine (IOM) as one of the four main

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attributes to increasing health care quality [1]. Several organizations, including the IOM, American Nurses Credentialing Center (ANCC) Magnet® Recognition Program, and The Joint Commission (TJC) have challenged providers to develop a practice culture that exemplifies safe, timely, effective, efficient, equitable and patient centered care systems and environments [1–4]. The application of these advances and innovations have contributed to a decrease in mortality for children with complex heart disease [5].

The professional practice environment, within which patients are cared, has been a focus of much research. In 2001, the American Association of Critical Care Nurses (AACN) and the American College of Chest Physicians (ACCP) collaborated to create standards to define and maintain a healthy work environment in the acute care setting [6]. The healthy work environment standards include:

- skilled communication,
- true collaboration,
- effective decision-making,
- appropriate staffing,
- meaningful recognition and
- authentic leadership [7].

The AACN and AACP acknowledged that a strong working relationship between cardiovascular physicians, nurses, and the multidisciplinary team is a key factor in successful patient outcomes [6]. The AACN further concluded that the links among a healthy work environment, excellent nursing practice, and outcomes of patient care were inextricable [6, 7]. Since the standards represent evidence-based and relationship-centered principles of professional performance, they provide a meaningful framework for pediatric cardiovascular programs [8].

Nurse-physician collaboration, a positive organizational climate, and nurse job satisfaction has been linked to lower mortality rates, lower complication rates, and higher levels of patient satisfaction [6–9]. Successful pediatric cardiovascular programs understand the unique contribution of each discipline and how the collective intelligence and talent of the entire multidisciplinary team is greater than any single individual or discipline [9]. Nurse and physician leaders draw on their respective scientific foundations and actively assume the responsibility for creating and supporting a professional practice milieu that fosters interdisciplinary collaboration and effective decision-making [7, 9].

The objectives for this chapter are to provide definitions of quality care, describe how quality is measured, discuss available data sources and describe how collaborations using data can drive quality cardiovascular care. Selected examples of contemporary cardiovascular nursing research and improvement science initiatives that generate high quality care for pediatric cardiovascular patients and their families are discussed as well as future directions for consideration.

Defining Quality Care

Multiple forums have suggested various elements to reflect quality care. These include but are not limited to standards and indicators of healthcare and conceptual components of care. Initially in 1998, the IOM defined quality as “the degree to which health services for individuals and populations increase the likelihood of desired health outcomes and are consistent with current professional knowledge [10].” Because “desired health outcomes” is somewhat vague and difficult to quantify, initial measures focused on negative, but more easily measured, components of quality such as death, disease, disability, discomfort, and dissatisfaction [10, 11].

Published in 2001, the most recent IOM’s guiding framework identifies the following six conceptual components of quality care for the 21st century - quality care is:

- safe,
- effective,
- patient-centered,
- timely,
- efficient, and
- equitable.

In this model, patient safety is the foundation upon which all other aspects of quality care are built [1].

Alternatively the American Academy of Nursing Expert Panel on Quality Health focused

on the following positive indicators of high-quality care they believed were sensitive to nursing input:

- achievement of appropriate self-care,
- demonstration of health-promoting behaviors,
- health-related quality of life,
- perception of being well cared for, and
- symptom management to criterion.

They concluded a quality measure should be a positive measure and mortality, morbidity, and adverse events were not only negative but represented the integration of multidisciplinary provider inputs [10–12].

Nursing and the Linkage to Patient Outcomes

Nursing's history is replete with individuals who have advanced the concept of quality in all aspects of care delivery. In 1855, Florence Nightingale analyzed mortality data among British troops and achieved a significant reduction in mortality by changing organizational and hygienic practices [13, 14]. She is also credited with creating the world's first performance measures of hospitals in 1859 [15]. In the 1970s, Wandelt reiterated the fundamental definition of quality is the characteristics and degrees of excellence, whereas standards refer to a general agreement of factors specifying what is ideal [11, 16]. At about the same time, Lang proposed a quality assurance model that has endured given its foundation of societal and professional values as well as the most current scientific knowledge [11, 17].

In the past, nursing's responsibility in patient safety has been limited to direct patient care, for example, eliminating falls and preventing skin breakdown. While these basic dimensions of safety are still important within nursing's scope of practice, they pale in comparison to the magnitude of nursing's contribution to overall patient safety and quality improvement. As nurses provide holistic patient care, they are constantly using critical thinking skills to gather, coordinate, synthesize, and interpret information and then provide data for an informed clinical decision that addresses multiple aspects of safety and quality in patient care. Nurses also provide wisdom, that deep understanding and sound judgment gained from experience, to both the decision making process and its implementation.

In addition to developing clinical expertise, it is equally important to mentor and engage bedside nurses in clinical inquiry and improvement science. Boston's Children's Hospital formed the Academy for Clinical Scholarship and Innovation in Pediatric Nursing to provide a comprehensive infrastructure for the mentorship of staff by nurse scientists. This infrastructure includes a focus on evidence-based practice, improvement science and research. The Academy for Clinical Scholarship and Innovation in Pediatric Nursing along with the Nurse Executive Committee for Research and Inquiry (NECRI) support the Nursing Science Fellowship. The Nursing Science Fellowship is a 2-year program designed to provide nurses in all roles across the hospital with foundational tools for clinical inquiry with a goal of advancing the science of nursing. Nursing science fellows participate in quarterly forums that include didactic and interactive experiences with expert nurse scientists. The forums provide basic content and in depth information on each of the clinical inquiry processes. Most significant is the one-on-one mentoring that occurs twice per month. This is independent time spent with a nurse scientist to clarify, develop, plan and implement one's own inquiry. Upon completion of the clinical inquiry, the fellow participates in its dissemination. The fellowship program provides a forum for staff to learn the value and importance of using data to advance practice.

Developing Quality Nursing Measures

Adult Versus Pediatric Measures

While nurses continue to represent the single largest provider of inpatient care, there remains a need to quantify their contributions to the provision of safe, effective, efficient, equitable, timely, and patient/family centered care [18–22]. This statement must be accentuated for pediatric

nursing care as most available standardized nursing-sensitive outcome indicators are focused on adult care [23–27]. These indicators such as prevalence of falls or failure to rescue have been shown to lack validity when applied globally to pediatrics or sub-populations of pediatrics [28, 29]. It can further be argued appropriate and high quality nursing care is especially important in children's health care as often the etiology and epidemiology of illness is different in children than adults, suggesting the need for specialized expertise. Children born with congenital heart disease explicitly exemplify this fact.

Currently the measurement of nursing care is highlighted as a major initiative across several areas of nursing and health care. The 2010 Robert Wood Johnson (RWJ)/IOM report acknowledged the USA health care system is in need of transformation to ensure higher quality, patient-centered care is provided to all citizens [30]. A major recommendation from the report is nurses function to the full extent of their education and assume a leadership role in achieving this mission. The need for nurse-sensitive quality indicators is undisputed. The National Quality Forum produced an initial consensus set of indicators in 2004 [31]. These adult based indicators, however, have yet to be adopted by national organizations, as concerns exist about the reliability of the indicators, definitions of terms, and methods of collection of data [24].

Ongoing National Database of Nursing Quality Indicators (NDNQI)

In 1994, the American Nurses Association (ANA) launched a safety & quality initiative to explore and identify the empirical linkages between nursing care and patient outcomes [32]. Information from this effort was used to develop a national database to collect data on nursing quality indicators. The NDNQI provides hospitals with unit-level performance comparison reports for state, regional, and national percentile distributions. All indicator data are reported at the nursing unit-level [32]. The NDNQI's nursing-sensitive indicators reflect the structure, process, and outcomes of nursing care (Table 21.1) [32]. While this is not specific to pediatric nursing, it does allow for pediatric participation and opportunities for benchmarking.

Table 21.1 National Database of Nursing Quality Indicators (NDNQI) measures

The current NDNQI indicators include:
Nursing hours per patient day[a]
Nursing skill mix[a]
Nurse turnover rate[a]
RN education/certification
RN survey with
Practice environment scale[a]
Job satisfaction scales
Assault/injury assault rates
Catheter-associated urinary tract infection rate[a]
Central line-associated blood stream infection rate[a]
Fall/injury fall rates[a]
Hospital/unit acquired pressure ulcer rates
Pain assessment/intervention/reassessment cycles completed
Peripheral IV infiltration rate
Physical restraint prevalence[a]
Ventilator-associated pneumonia rate[a]

[a]National Quality Forum (NQF) endorsed nursing-sensitive care measure

Ohio Children's Hospitals' Solutions for Patient Safety (OCHSPS)

Another recent national collaborative effort that is pediatric focused is the OCHSPS [33]. Launched in 2009, Solutions for Patient Safety was founded as a partnership between providers and the business community, under the leadership of the Ohio Business Roundtable, to improve quality and reduce costs among children's hospitals in the state of Ohio [33]. Experiencing initial success, they reformatted their goal to eliminating all serious harm in Ohio's children's hospitals. Initial efforts will focus on eliminating Serious Safety Events (SSEs) in Ohio children's hospitals and developing a patient harm index to capture elements of harm occurring at children's hospitals. OCHSPS has focused on tactics to reduce harm nationally in 11 healthcare acquired conditions (Table 21.2) [33].

Pediatric Cardiovascular Nursing Initiatives

The cardiovascular program at Boston Children's Hospital has a strong history in development of nurse-sensitive quality indicators of inpatient care through the creation of the Nightingale Metrics in 2003 [24]. As Nightingale said," what nursing has to do…. is to put the patient in the best condition for nature to act upon him [34]."

Table 21.2 Ohio Children's Hospitals' Solutions for Patient Safety (OCHSPS)

Adverse drug events (ADE)
Catheter-associated urinary tract infections (CAUTI)
Central line-associated blood stream infections (CLABSI)
Injuries from falls and immobility
Pressure ulcers
Surgical site infections
Ventilator-associated pneumonia (VAP)
Preventable readmissions
Obstetrical adverse events
Venous thromboembolism
Serious safety events (SSE)

The goal of this metric initiative, led by Curley and Hickey, was for nurses

- to identify nursing care activities important to their patients and families,
- to measure how often nurses performed these interventions, and
- to use the data to improve the care they provided [24].

Examples of specific cardiac nurse metrics include (1) documentation of PR interval and (2) checking central line blood return at the start of each shift. Although some of the Nightingale metrics are process measures, whenever possible, direct links to patient outcomes have been examined. Patient outcomes were found to improve with documentation of the process measure and/or compliance with the care bundles. Examples of these linkages include decreased rate of ventilator associated pneumonia (patient ventilated, oral hygiene twice in 24 h, daily holiday/twitch plan for chemically paralyzed patients) (Fig. 21.1) and decreased rate of pressure ulcer occurrence and severity (patient immobile, position changed every 2 h, heels off bed, out of bed/held in last 24 h) (Fig. 21.2).

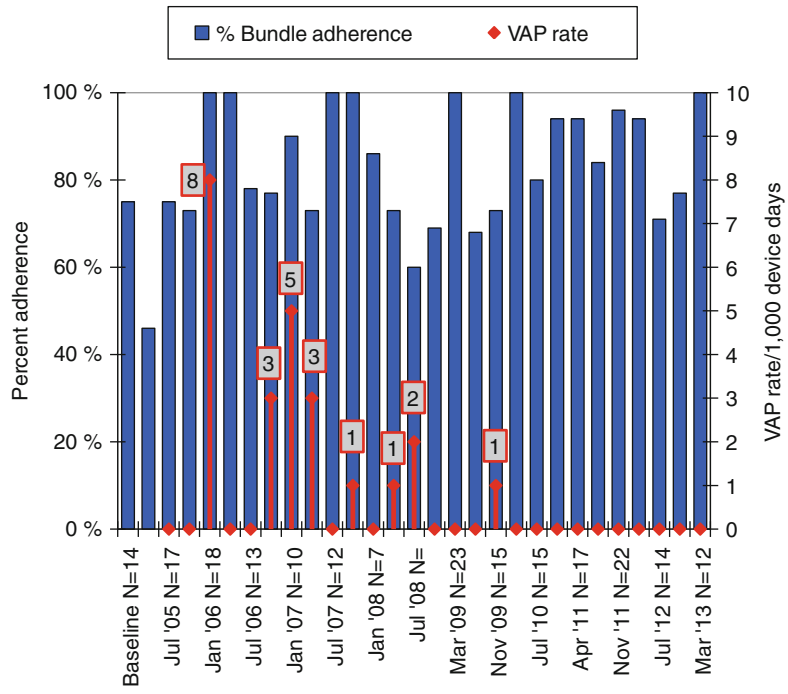

Fig. 21.1 Nightingale metric ventilator-associated pneumonia bundle

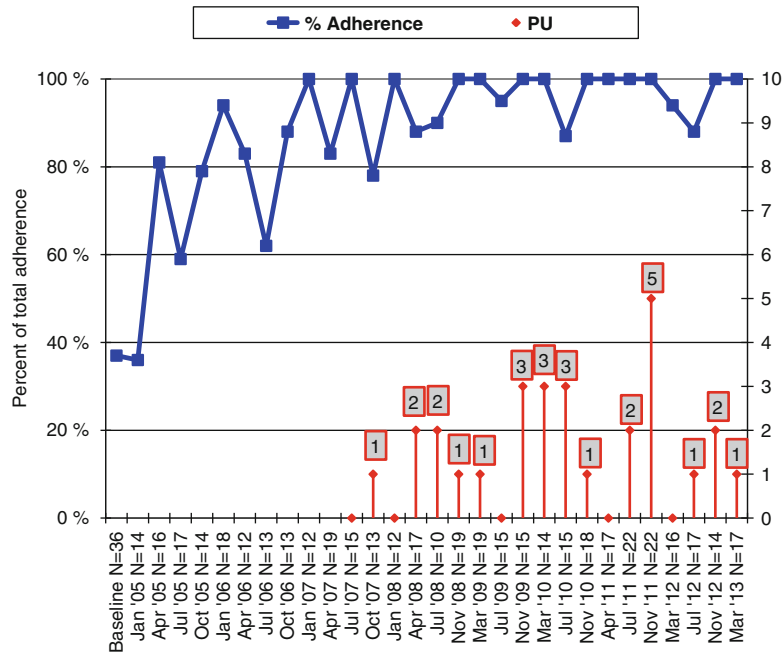

Fig. 21.2 Nightingale immobility-related pressure ulcer bundle (immobile patients only)

Table 21.3 Boston Children's Hospital Nightingale Metrics

Boston Children's Hospital Inpatient Cardiovascular Program	
Nightingale Metrics	
Cardiac Intensive Care Unit	Acute Care Cardiac Unit
Braden Q	Braden Q
Calorie counts (<1 year of age)	Chest tube bundle
Continuity of care	Continuity of care
Devices on patient	Devices on patient
Family-centered care	Family-centered care
FLACC pain scale	FLACC pain scale
Glasgow coma scale	Heparin lock PIVs
Mouth care	ID bands
Neonate care bundle	Newborn discharge procedures (48 h)
Panic labs	Newborn discharge procedures (72 h)
Parent presence	Parent presence
Pressure ulcer bundle	Patient goals
State Behavioral Scale (SBS)	PICC/central catheter
	Running IVs
	State Behavioral Scale (SBS)

Table 21.3 shows examples of Nightingale Metrics currently monitored in the cardiovascular ICU and inpatient cardiovascular unit at Boston Children's Hospital. All data are collected quarterly on a random day through chart audits, direct observation, or both methods. Chart audits are limited to the previous 7 days. When audits reveal benchmark results for 3 consecutive audit periods, the measure is retired and spot checked on a yearly basis. This methodology enables staff to evaluate their practice in real time and make data driven decisions about patient care.

National Pediatric Cardiovascular Nursing Collaboration

At the national level, the American College of Cardiology (ACC) Pediatric Quality Metric Working Group (QMWG) held an open session for participation in development of a dashboard of pediatric cardiology-sensitive metrics in 2008. Important to the ACC QMWG was the inclusion of a pediatric nursing-sensitive metric in the pilot group of proposed metrics. To this end, expert nurse clinicians, administrators, and scientists from across the country participated in the project. Fourteen nurses from 10 different institutions collaborated and conducted a literature review as the first step. Topics included in this search were:

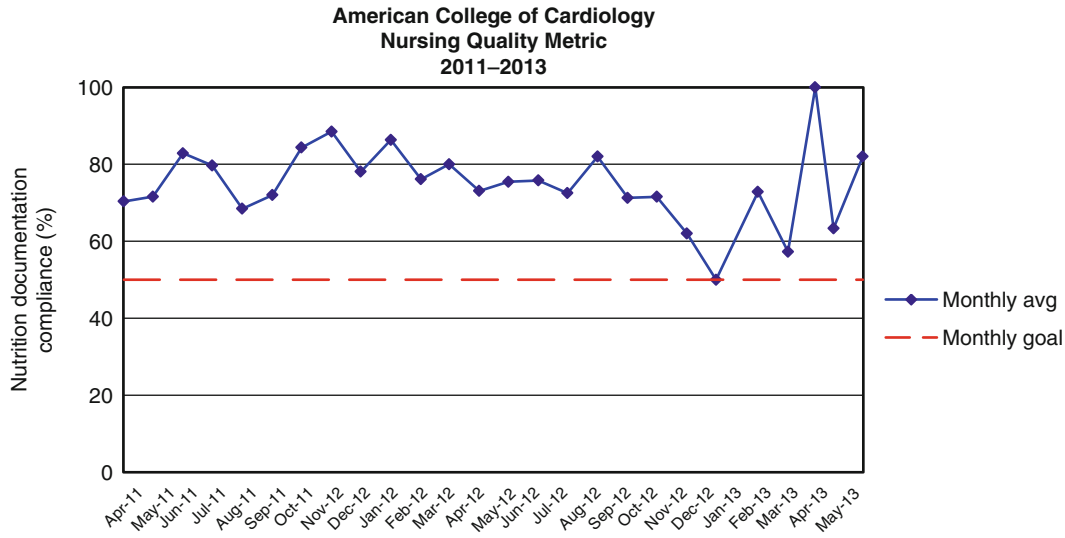

Fig. 21.3 American College of Cardiology pediatric nursing quality metric

- standardized nursing measurement,
- pediatric nurse sensitive metrics, and
- metrics related to general inpatient cardiac care.

Results revealed published measures were either not appropriate for pediatrics or not generalizable to the practice of pediatric cardiac nursing care. The charge then changed from identification of a metric to development of a metric. Through a series of conference calls, email communications, and consultation with additional nursing experts, the group reached consensus around the importance of nutrition for children with cardiac disease. Furthermore, they identified it as a critical component of pediatric cardiac nursing care that contributed to overall patient outcomes. Although documentation of daily fluid intake was a standardized activity performed by nurses, there was no consistent documentation of assessment or measurement of nutritional intake of infants during hospitalization.

In its final form "Documentation of Nutrition" is a metric of daily documentation of feeding status and Kcals/kg/day for all infants ≤30 days admitted for surgical intervention or medical intervention/management for more than one 24-h period. As of January 2011, the network of participating institutions has increased to 15 sites dedicated to the care for children with heart disease who currently are participating in the process of submitting data on the nutrition measure (Fig. 21.3).

The ACC QMWG nursing experience has demonstrated a number of successes:

1. development of a collaborative, consensus based approach among pediatric cardiac nurse scientists, administrators, and clinical experts to identify and develop a measurement of nursing quality;
2. feasibility of implementing a measure and strategy of collection of data in 15 institutions across the country; and
3. a commitment in identification and testing of other measures in a consensus based manner.

To date, this effort has provided strong foundation and a natural evolution for the development of the Consortium for Congenital Cardiac Care Measurement of Nursing Practice (C4-MNP).

Consortium for Congenital Cardiac Care Measurement of Nursing Practice (C4-MNP)

In 2011, the C4-MNP was launched to establish a national identification of pediatric cardiovascular nursing measurement with a goal of benchmarking activities of nursing care that contribute to improved outcomes in a highly complex environment and specialized population of patients. Through the structure and processes of the

consortium, a national community of researchers, administrators, and expert clinicians have come together to form a broad network committed to rigorous measurement of the quality of care required by nurses who participate in achieving optimal outcomes for children with cardiac disease. The identification of key performance measures and the articulation and measurement of the value of care delivered by nurses in the health care environment are central to improving and sustaining quality care and reducing cost. To date the effort has 20 pediatric cardiovascular programs partnered in identifying measures in seven target areas. Areas of measurement include: (1) Clinical deterioration, (2) Care of the adult congenital heart disease patient, (3) Family/patient-centered care, (4) Nutrition, (5) Pain management, (6) Prevention of pressure ulcers and (7) Work environment for the healthcare team. The developed measures will be piloted and then serve as benchmarks across pediatric cardiovascular programs.

Using Evidence to Guide Quality Nursing Care

Kid's Inpatient Database (KID) and the Pediatric Health Information Systems (PHIS)

A number of data sources are currently available for examination of outcomes for children with congenital heart disease. One such database specific to pediatrics is the KID [35]. The KID database was developed as part of the Healthcare Cost and Utilization Project (HCUP) through a Federal-State-Industry partnership sponsored by the Agency for Healthcare Research and Quality [35]. The goal of this partnership is to use these data to inform decision making at the community, state, and national levels. The KID dataset is currently available in the following years: 1997, 2000, 2003, 2006, and 2009. The latest version of the KID database consists of a stratified random sample of 3,407,146 discharges from 4,121 institutions in 44 states. The KID uses the American Hospital Association's definition of hospitals to identify all non-federal, short-term, general and other specialty hospitals. Pediatric hospitals, academic medical centers, and specialty hospitals are included. The database does not include all admissions from participating institutions, but instead includes a 10 % sample of uncomplicated, in-hospital births from these institutions, and an 80 % sample of other pediatric discharges (age <21 years). To obtain information that is nationally representative, the sample is weighted to represent the population of pediatric discharges from all community, non-rehabilitation hospitals in the United States that were open for any part of the calendar year examined. To protect patient confidentiality, the KID database does not contain specific patient or hospital identifiers.

The PHIS is another large database source for understanding pediatric outcomes [36]. The PHIS provides detailed clinical and financial information managed by the Performance Improvement Division of the Child Health Corporation of America (CHA). Data is collected from 43 participating free-standing children's hospitals in the United States [36]. Over 125 data elements are collected for each patient admission, including demographic information, admission and discharge dates, patient outcomes, and International Classification of Diseases, Ninth Revision, Clinical Modification (ICD-9-CM) codes for all diagnoses and procedures. Institutions are labeled within the database but cannot be identified in public reporting; individual patient medical record numbers and identification codes are encrypted.

Although the KID and the PHIS datasets are considered to be powerful data sources, there are universal limitations when using administrative datasets. Unique patient identifiers or record linkage numbers are not available in the datasets. Missing data, coding errors, and lack of detailed clinical information are also universal limitations in using large administrative datasets in outcomes research [37]. As described elsewhere in this book (in the chapter by Franklin and colleagues titled: ***Nomenclature for Congenital and Pediatric Cardiac Disease***: ***Historical Perspectives and the International Pediatric and Congenital Cardiac Code***), the validity of coding of lesions seen in the congenitally malformed heart via the International Classification of Diseases as used in administrative databases is likely to be poor.

Using Evidence to Impact Patient Outcomes

Registered nurses (RN) are the largest group of healthcare providers and typically account for the greatest expense in hospital operating budgets. The relationship of nursing care to patient outcomes has been well established in adult hospitals. Specifically, the nursing and organizational skills of the baccalaureate educated nurse (BSN), increased RN staffing, healthy work environments, and Magnet® recognition, are all associated with improved patient outcomes in these hospitals [38–44].

Magnet® recognition is a designation program recognizing organizational nursing excellence and has been linked to improved quality of care and patient outcomes [45]. The Magnet® Recognition Program aims to promote professional practice, excellent patient care and evidence driven practice [46]. Healthcare organizations must apply for designation and redesignation and the application and designation process requires several rigorous steps in the demonstration of excellence. The journey to Magnet® requires organizations demonstrate nursing excellence around forces of magnetism. The forces are organized into 5 overarching components:

- transformational leadership,
- structural empowerment,
- exemplary professional practice,
- new knowledge, innovations and improvements and
- empirical outcomes.

Magnet® is not a pediatric-specific designation but a significantly higher percentage of freestanding children's hospitals have Magnet® designations than do adult hospitals. Less than 6 % of hospitals across the United States have been designated in The Magnet® Recognition Program but over 80 % of pediatric hospitals are either designated as Magnet® or are on the journey [47]. The high proportion of children's hospitals with Magnet® designation demonstrates the level of nursing excellence, as well as, the commitment to quality care and excellent patient outcomes.

Little is known about the dynamic between the pediatric workforce and patient outcomes [48]. In response to this gap in the literature, Hickey and colleagues conducted a series of studies to describe and evaluate nursing's contribution to patient outcomes in the congenital heart surgery population. Although congenital heart disease is the most commonly occurring birth defect requiring surgical intervention for survival, children's hospitals in the United States have demonstrated marked variation in mortality for congenital heart surgery patients [43, 49, 50]. The group's first study revealed a negative association between BSN nurse characteristics, RN skill mix, RN staffing, or Magnet® recognition to pediatric outcomes [43]. These findings led to an examination of nursing and organizational factors not publically available that may have a protective effect on pediatric mortality. Using a survey methodology, nursing leaders from 43 free-standing children's hospitals with active cardiovascular programs provided detailed demographic data about their nursing staff members [47]. Highlights of the results are provided in Table 21.4. Data included highest level of

Table 21.4 Characteristics of pediatric intensive care nurses from 43 centers in free-standing Children's Hospitals in the United States

	Number	Percent
Highest level of education		
BSN	2,417	71
AS	598	18
Diploma	139	4
MS	153	4
PhD	3	<1
Other	103	3
Years of clinical experience		
0–2	729	24
3–5	855	28
6–10	639	21
11–15	321	11
≥16	496	16
Years of unit experience		
0–2	1,149	35
3–5	888	27
6–10	593	18
11–15	273	8
≥16	337	10
Professional nursing certification		
[a]CCRN	557	13
[b]CPN	102	2

[a]American Association of Critical Care Nurses Certified Critical Care Registered Nurse
[b]Society of Pediatric Nurses Certified Pediatric Nurse

Table 21.5 Significant relationships between nursing and organizational variables and in-hospital mortality congenital heart surgery patients in the United States

	RACHS-1 risk adjusted	
	Odds ratio	P value
Education		
% RNs with BSN or higher	0.91	0.02
Specialty certification		
% RNs with CCRN	0.93	0.06
Clinical experience		
[a]% RNs with ≤ 2 years.	1.12	<0.001
% RNs with ≥ 11 years.	0.89	0.04
% RNs with ≥ 16 years.	0.82	0.006
Quality metrics (*national*)	0.61	<0.001

[a]≥20 % of RNs have ≤ 2 years experience and the odds ratio of in-hospital mortality is 1.30 (P = 0.05)
≥25 % of RNs have ≤ 2 years experience and the odds ratio of in-hospital morality is 1.52 (P = <0.001)

education, years of clinical experience, years of critical/intensive care unit (ICU) experience, and professional certification. Seventy-one percent of the nurses were BSN prepared. The distribution of nursing experience included 52 % with less than 5 years and 48 % with more than 5 years of nursing experience; however, 62 % had less than 5 years of ICU nursing experience. Fifteen percent of the pediatric nurses were professionally certified in critical care (13 %) or pediatric nursing (2 %).

In their third study, data from the nursing and unit characteristics survey were linked to patient outcomes using the PHIS [51]. The Risk Adjustment for Congenital Heart Surgery (RACHS-1) method was used to adjust for baseline patient differences in patients <18 years of age [49]. After controlling for baseline patient risk of mortality for 20,407 patients, the odds of death increased as the institutional percentage of pediatric ICU nurses with ≤2 years clinical experience increased (OR = 1.12 for each 10 % increase, p < 0.001). The odds of mortality were highest when the percent of RNs with ≤2 years clinical experience was ≥20 % (OR 1.30, p = 0.05) and ≥25 % (OR 1.52, p = 0.001). The odds of death decreased as the institutional percentage of critical care nurses with ≥11 years clinical experience increased (OR = 0.89, p = 0.04), ≥16 years of clinical experience increased (OR = 0.82, p = 0.006), and for hospitals participating in national quality metric benchmarking (OR = 0.61, p < 0.001). Years of clinical experience was independently associated with in-hospital mortality (Table 21.5).

The third study also revealed for the first time higher levels of experience and education in the pediatric critical care nursing workforce are associated with fewer patient deaths. The identification of a cut point of ≥20 % inexperience nursing staff is associated with increased mortality may now be considered in discussions of nurse staffing and finances among nurse leaders, hospital executives, and policy makers. An additional finding was critical care units that contribute data to national quality metric registries were associated with reduced mortality. Future research is required to determine other pediatric nursing and organizational factors that may have a protective effect on mortality for critically ill patients. Although this study involves secondary analysis of a large database with broad representation of children's hospitals in the United States, it should be replicated in other populations such as non-cardiac pediatric and adult populations to determine generalizability in general hospitals.

Improving Patient Safety with the Red Zone Medication Safety Initiative

The incidence of medication errors remains a concern across the spectrum of healthcare. In hospitals, errors are common during every step of the medication process from prescription to preparation to administration; however, they occur most frequently during the prescription and administration stages. When all types of errors are taken into account, a hospitalized patient can expect on average to be subjected to more than one medication error each day. However, substantial variations in error rates are found across facilities [52]. The causes of medication errors have been well documented with many studies highlighting relationships involving structure and process of care [52–59]. Nursing studies have identified antecedents of medication errors including unit environment, communication, distractions, RN hours, and level of expertise [52–54, 56, 57, 60–62].

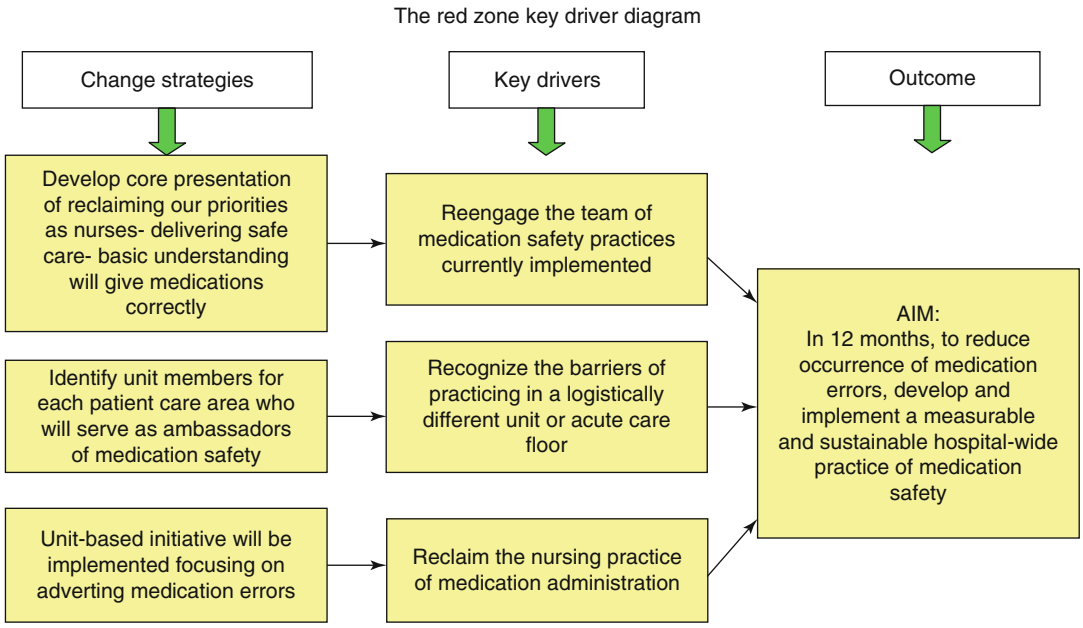

Fig. 21.4 Red Zone Medication Safety Initiative

Although preventing the cause of these errors is challenging, approaches to averting medication errors and implementing a culture of safety are key foci for most institutions. Many healthcare organizations have developed and initiated Red Zone Principles in an effort to reduce institution-wide medication error and emphasize organizational commitment to safe medication administration. The overriding belief of the Red Zone strategy is safety needs to be a priority and valued at an organizational level; overstimulation and distraction affects the precision with which the nurse is able to perform his or her job thus leading to medication errors [63]. Focus on teamwork, communication, and empowerment of staff support the guiding principles [64]. These principles have been operationalized in a number of ways depending on clinical structure and processes of care. For some units, eliminating all communication, interruptions, or distraction from staff, patients, and other health care team members at the time of medication preparation or administration has been appropriate [63, 64]. Others have developed a safety checklist for medication preparation and administration [63]. Some institutions have designated "Red Zone" areas in the unit or practice area with painting red boxes, hanging alert signs, or wearing alert vests and/or hats [63].

At Boston Children's Hospital, in the spring 2010, a multidisciplinary steering committee launched the Red Zone Medication Safety Initiative in the inpatient cardiovascular and critical care areas. Using Six Sigma and Change Acceleration Process frameworks, a baseline assessment of the number and type of medication events was reviewed. A key driver diagram was developed to guide the initiative (Fig. 21.4). For each area, a unit level Red Zone Ambassador team was convened to champion the Red Zone Principles. The Cardiac Intensive Care Unit (CICU) served as the demonstration site.

Since implementation of the Red Zone, all inpatient cardiovascular and critical care areas have experienced a sustained decrease in number of reported events from 2009 to 2013 (Figs. 21.5, 21.6, and 21.7). Rolling averages of reported medication events per month in the CICU provide an overall picture of the decrease in medication events over time (Fig. 21.6). Figure 21.7 depicts individual medication events per month in CICU as raw data points and indicates a decline in reported events. Overall, the areas experienced an average decrease in reported events of 61 %, with the cardiac intensive care unit experiencing a 68 % decrease. Unit level ambassador teams continue to report success

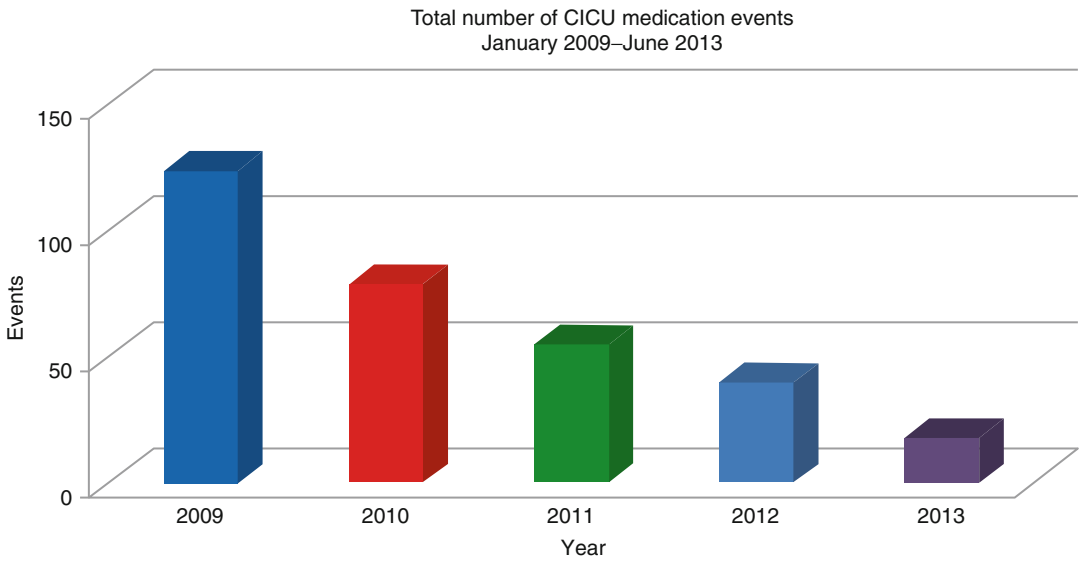

Fig. 21.5 Red Zone Medication Safety Initiative: total medication events

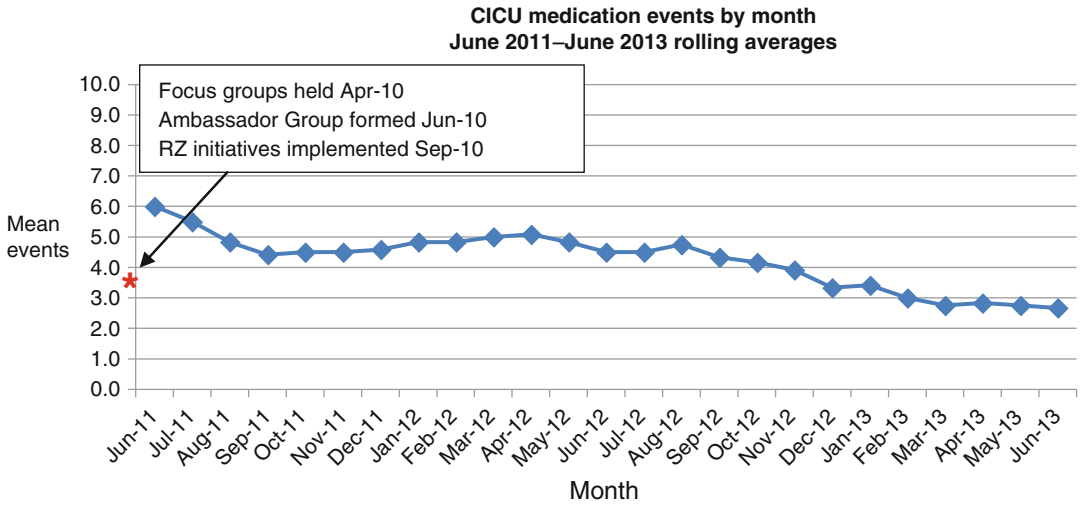

Fig. 21.6 Red Zone Medication Safety Initiative: rolling averages

with the implementation of the Red Zone initiative in terms of feasibility and sustainability.

Rescuing Patients Using the Children's Hospital Early Warning Score (CHEWS)

Pediatric cardiovascular patients have a higher incidence of cardiopulmonary arrests than other pediatric patients, with arrhythmias accounting for 41 % of these arrests [65–67]. Of the children that do experience an arrest, cardiac patients are younger than non-cardiac patients, with more than three-quarters <1 year of age compared to only one-third of non-cardiac patients [68]. In addition, cardiac patients have higher rates of preexisting arrhythmias and congestive heart failure but fewer co-morbidities than non-cardiac patients [68]. Children with congenital heart defects also have

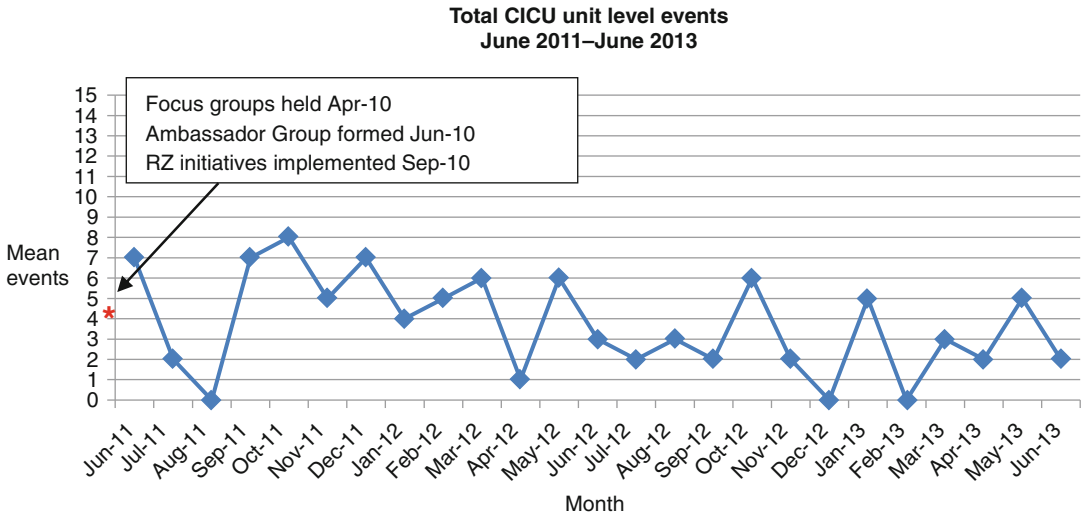

Fig. 21.7 Red Zone Medication Safety Initiative: total unit level events

baseline signs and symptoms, such as cyanosis, which are atypical of other pediatric populations.

The Pediatric Early Warning Score (PEWS) was developed in 2005, and subsequently validated in a large cohort of pediatric medical patients. The PEWS score is calculated from three domains of assessment:

- neuro/behavior,
- cardiovascular, and
- respiratory.

Each domain is scored on a scale of 0–3 with "3" representing the most concerning symptoms. Boston Children's Hospital adopted a modified PEWS with two additional domains: "staff concern" and "family concern." Nurses calculate scores during assessment of the vital signs of patients. Given the success of this tool on general medical and surgical inpatient units, the tool was piloted on the inpatient cardiovascular unit for 24 h. During the pilot, scores were compared to the assessment of clinicians and the clinical courses of patients. Surprisingly, the tool scored too low for patients who were acutely deteriorating and did not appropriately identify cardiovascular patients at risk. Expert cardiovascular nurses examined the variables in each domain and revised the tool to be more aligned with baseline and worsening signs and symptoms of children with cardiovascular disease. The new tool was named, the Cardiac Children's Hospital Early Warning Score (C-CHEWS). In September 2009, C-CHEWS was implemented on the inpatient cardiovascular unit (Table 21.6) [69]. Nurses complete the C-CHEWS in less than 10 s and document the score in the patient's record. The score is then displayed on the unit's patient census. The next step was to obtain approval from the institution's internal review board to conduct formal validity testing of the C-CHEWS in the pediatric cardiac patient population. The C-CHEWS was found to have excellent discrimination and performed substantially better than the PEWS for identifying clinical deterioration in children with cardiac disease (Fig. 21.8). When compared to the PEWS on the Escalation of Care Algorithm cut points, the C-CHEWS was found to have a higher sensitivity [70]. The C-CHEWS should help identify cardiac patients at risk for critical events and support clinicians in initiating escalation of care to prevent cardiopulmonary arrests in cardiac patients. Development of the C-CHEWS has improved safety for cardiovascular patients and increased support for cardiovascular clinicians to take action when a patient begins to deteriorate.

Table 21.6 Boston Children's Hospital early warning score

	0	1	2	3	Score
Behavior/neuro	Playing/sleeping appropriately Alert, at patient's baseline	Sleepy, somnolent when not disturbed	Irritable, difficult to console Increase in patient's baseline seizure activity	Lethargic, confused, floppy Reduced response to pain Prolonged or frequent seizures Pupils asymmetric or sluggish	
Cardiovascular	Skin tone appropriate for patient Capillary refill ≤ 2 s	Pale Capillary refill 3–4 s Mild* tachycardia Intermittent ectopy or irregular HR (not new)	Grey Capillary refill 4–5 s Moderate* tachycardia	Grey and mottled Capillary refill >5 s Severe* tachycardia New onset bradycardia New onset/increase in ectopy, irregular HR or heart block	
Respiratory	Within normal parameters No retractions	Mild* tachypnea/increased WOB (flaring, retracting) Up to 40 % supplemental oxygen Up to 1 L NC > patient's baseline need Mild desaturations < patient's baseline Intermittent apnea self-resolving	Moderate* tachypnea/ increased WOB (flaring, retracting, grunting, use of accessory muscles) 40–60 % oxygen via mask 1–2 L NC > patient's baseline need Nebs q 1–2 h Moderate desaturations < patient's baseline Apnea requiring repositioning or stimulation	Severe* tachypnea RR < normal for age Severe increased WOB (i.e. head bobbing, paradoxical breathing) >60 % oxygen via mask >2 L NC > patient's baseline need Nebs q 30 min–1 h Severe desaturations < patient's baseline Apnea requiring interventions other than repositioning or stimulation	
Staff concern		Concerned			
Family concern		Concerned or absent			
					Total

	Mild*	**Moderate***	**Severe***
Infant Toddler and older	≥10 % ↑ for age	≥15 % ↑ for age	≥25 % ↑ for age
	≥10 % ↑ for age	≥25 % ↑ for age	≥50 % ↑ for age

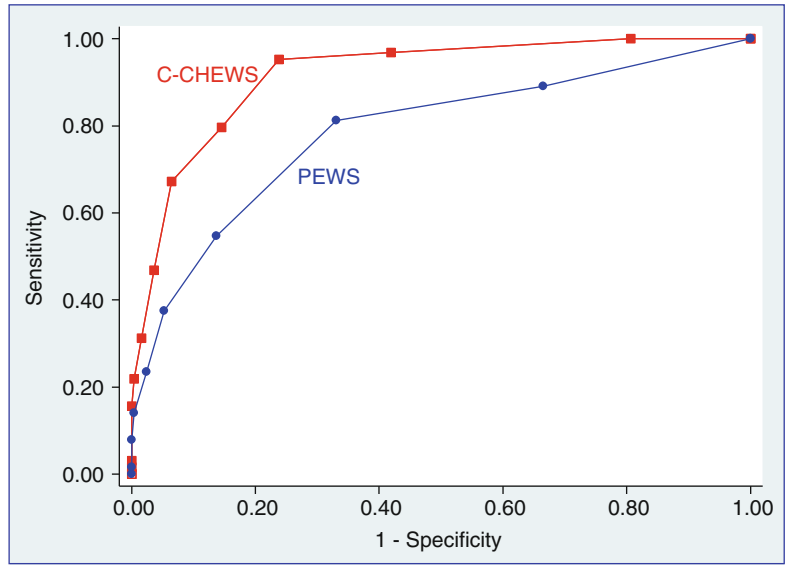

Fig. 21.8 Area under the receiver operating characteristic curve for Cardiac Children's Hospital Early Warning Score (C-CHEWS) and Pediatric Early Warning Score (PEWS) tools

Table 21.7 Complexity Assessment & Monitoring to Ensure Optimal Outcomes (CAMEO) domains of care

1. Monitoring	12. Resuscitation
2. Intermittent medications	13. Self Care
3. Vasoactive IV medications	14. Tube care
4. Continuous IV medications	15. Transfer/admissions/transport
5. Ventilatory support	16. Teaching/anticipatory guidance to patient/family
6. Dialysis	17. Assessment of anxiety/coping/mood/family adjustment
7. Interventions within the CICU	18. Infection control
8. Management within the CICU	19. Indirect/miscellaneous
9. Monitoring within the CICU	
10. Phlebotomy	
11. Procedures within the CICU	

Measuring the Complexity and Autonomy of Nursing Care in the Pediatric Cardiac Intensive Care Unit Using the Complexity Assessment & Monitoring to Ensure Optimal Outcomes (CAMEO)

Despite advances in practice, pediatric critical care nurses have been challenged to design a model for the future of healthcare that truly captures the skills required to deliver comprehensive patient care. Prior efforts to create a model have produced tools that quantify nursing workload, intensity, and resource allocation in adult intensive care units. Unfortunately, none of these tools capture the current scope and complexity of nursing practice required of pediatric critical care nurses. Information technology provides a means to track and quantify nursing skills and workload, advance the nursing profession, and improve the outcomes of patients.

In 2009, efforts began in the Boston Children's Hospital pediatric cardiac critical care unit to develop a tool that could qualify and quantify the complex assessment, monitoring, and therapeutic interventions performed by nurses caring for these patients. An expert panel of cardiac nurse clinicians used Delphi methodology to develop a detailed description of the autonomous nature and comprehensive nursing management required for a complex pediatric cardiac population. The result is a tool called CAMEO: Complexity Assessment & Monitoring to Ensure Optimal Outcomes.

CAMEO monitors nursing activity in 19 domains of care (Table 21.7). Each domain

Table 21.8 Complexity Assessment & Monitoring to Ensure Optimal Outcomes (CAMEO) classifications

I	Intensive care nursing assessment and management are focused on de-intensifying patient
	Patient is hemodynamically stable
	Patient does not require invasive hemodynamic monitoring
	The patient is near transfer to a lower level of care
II	Intensive care nursing assessment and management are required to maintain goal hemodynamics
	Patient is hemodynamically stable and is expected to remain stable or improve
	Patient may require invasive hemodynamic monitoring with noninvasive ventilatory support
III	Intensive care nursing assessment and management are required to maintain goal hemodynamics
	Patient is hemodynamically stable and is expected to remain stable or improve
	Patient requires invasive hemodynamic monitoring with invasive ventilatory support
	Respiratory support is stable or de-escalating
IV	Intensive care nursing assessment, management and frequent interventions are required to maintain goal hemodynamics
	Patient is hemodynamically unstable
	Patient requires vasoactive infusions, invasive ventilatory support and advanced hemodynamic monitoring
V	Intensive care nursing management, assessment and frequent interventions are required to maintain goal hemodynamics
	Patient is hemodynamically unstable and outcome is uncertain
	Patient requires vasoactive infusions, invasive ventilatory support, advanced hemodynamic monitoring and mechanical support therapy

contains specific items of care, scored from basic to complex. To obtain a score, each activity is given a cognitive complexity value based on a scale from 1–5. The total score is calculated across the 19 domains of care and then ranked according to the five classifications for complexity of care (Table 21.8).

To test the nursing CAMEO tool, information was gathered on 75 patients, ages 1 day to 47 years, who were admitted to the CICU for surgical recovery (86 %) and/or medical intervention (14 %). The data showed the majority of patient care activities involved complicated elements. Standard intensive care monitoring of <1 h was reported in 42 % of patients. Vasoactive intravenous medication requiring titration was noted in 78 % patients. Ventilated patients (72 %) required a number of interventions to maintain airway patency and acid-base balance as well as to achieve weaning goals.

Additional nursing activities were identified and tracked, including teaching and anticipatory guidance to patients and families, as well as coordination of services such as social work, case management, interpreter services, and clergy. Precepting new staff, quality monitoring, collection of data for research, clinical management plans, and other regulatory documentation, were identified as well. Among the 75 patients, 80 % were ranked as Class III or IV, with 7 % ranked as Class V (Fig. 21.9).

Now, the CAMEO tool is moving from "proof of concept" to daily practice in the pediatric CICU. Twice each day, the CAMEO is completed electronically to provide front line nursing staff with the ability to quantify nursing care based on the cognitive complexity required to maintain safe care practices and promote optimal healing. This real-time quantification of nursing resource use and benchmarking validates the workload of bedside clinicians and supports administrative leaders seeking to justify appropriate allocation of clinical resources.

Pressure Ulcer Prevention using the Standardized Clinical Assessment and Management Plan (SCAMPs) Methodology

Limited evidence exists about the development of pressure ulcers in pediatric patients as compared to adults. Evidence suggests pediatric patients are vulnerable to a variety of skin problems including skin tears, incontinence-associated dermatitis, epidermal stripping related to tape, and extravasation of intravenous fluid or medications infused peripherally [71]. The potential to develop serious iatrogenic injury related to hospital-acquired pressure ulcers may

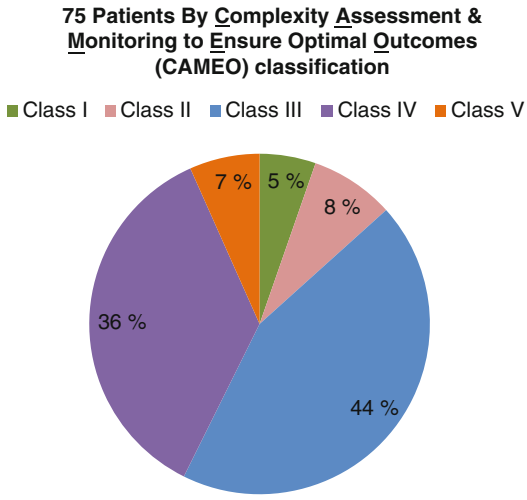

Fig. 21.9 75 patients by Complexity Assessment & Monitoring to Ensure Optimal Outcomes (CAMEO) classifications

go unrecognized or underrated, especially in pediatric populations that have been minimally studied. The risk factors related to the development of pressure ulcers in the children with congenital cardiac disease are largely unknown [72].

In October 2010, an expert panel of 13 nurses including leaders, bedside staff, and nurse scientists from across the cardiovascular program was convened to develop a Pressure Ulcer Prevention (PUP) Standardized Clinical Assessment and Management Plan (SCAMP). The key objective of the PUP SCAMP was to standardize practices to maintain skin integrity and improve the overall quality of care. Using a consensus model, key components of the SCAMP were developed. These included a background paper establishing the state of the evidence about the patient population, identification of 12 plausible findings to support the evaluation of the SCAMP, development of an enrollment form identifying inclusion and exclusion criteria, a decision tree, and the data collection form. In May of 2011, the PUP SCAMP, with data collection, was initiated. Data were collected on the 674 patients that met inclusion criteria during the 1 year time period. Thirty patients developed a pressure ulcer. Among the 30 cases, 21 (70 %) of the ulcers were deemed device related. Also of note, all cases had undergone a complex surgical intervention and 80 % of the ulcers were identified within the first 7 post-operative days.

Further work to prevent harm will include an examination to identify independent factors associated with the development of pressure ulcers in pediatric cardiac surgical patients.

Future Directions for Building the Science

The future of nursing is clear in the need to function to the full extent of education and scope of practice. Nurses are critical to the surveillance, coordination, and communication that ensure patient safety and optimal outcomes. Achieving high quality care occurs through an evidence driven practice. Use of technology, collaboration, and benchmarking (both internally and externally) are important to developing and sustaining an evidence driven practice. Opportunities in all three areas are becoming increasingly available to the field, providing a means to track and quantify nursing skills and workload, advance the nursing profession, and improve the outcomes of patients. Now more than ever, nurses must lead with evidence to advance the practice and science of pediatric cardiovascular nursing.

References

1. Institute of Medicine. Crossing the quality chasm: a new health system for the 21st century. Washington, D.C.: National Academies Press; 2001.
2. American Nurses Credentialing Center. The magnet model components and sources of evidence: Magnet Recognition Program. Silver Springs: American Nurses Credentialing Center; 2008.
3. The Joint Commission. Facts about the Joint Commission. Oakbrook Terrace: Joint Commission; 2013.
4. Institute of Medicine. The future of nursing: leading change, advancing health. Washington D.C.: National Academies Press; 2010.
5. Boneva RS, Botto LD, Moore CA, Yang Q, Correa A, Erickson JD. Mortality associated with congenital heart defects in the United States: trends and racial disparities, 1979–1997. Circulation. 2001;103(19): 2376–81.
6. Mitchell P, Armstrong S, Simpson TF, et al. American Association of Critical Care Nurses Demonstration

Project: profile of excellence in critical care nursing. Heart Lung. 1989;18(3):219–37.
7. American Association of Critical Care Nurses. AACN standards for establishing and sustaining healthy work environments: a journey to excellence. Aliso Viejo: American Association of Critical-Care Nurses; 2011.
8. McCauley K, Irwin RS. Changing the work environment in intensive care units to achieve patient-focused care: the time has come. Am J Crit Care. 2006;15(6):541–8.
9. Hickey PA. Building a culture of excellence in Boston and beyond. World J Pediatr Congenit Heart Surg. 2010;1(3):314–20.
10. Lohr KN. Outcome measurements: concepts and questions. Inquiry. 1998;25(1):37–50.
11. Mitchell PH. Defining patient safety and quality care. In: Huges RG, editor. Patient safety and quality: an evidence-based handbook for nurses. Rockville: Agency for Healthcare Research and Quality (US); 2008.
12. Mitchell PH, Heinrich J, Moritz P, et al. Outcome measures and care delivery systems: introduction and purposes of the conference. Med Care. 1997;35(11):NS1–5.
13. Nightingale, F. I have done my duty. In: Goldie SM, editor, Florence Nightingale in the Crimean War. Manchester: Manchester University Press; 1987.
14. Cohen IB. Florence Nightingale. Sci Am. 1984;250(3):128–37.
15. Monteiro LA. Florence Nightingale on public health nursing. Am J Public Health. 1985;75(2):181–6.
16. Wandelt MA. Definitions of words germane to evaluation of health care. NLN Publ. 1976;15(1611):57–8.
17. Lang N. Issues in quality assurance in nursing. Kansas City: American Nurses Association; 1976.
18. Bostick JE, Riggs CJ, Rantz MJ. Quality measurement in nursing: an update of where we are now. J Nurs Care Qual. 2003;18(2):94–104.
19. Carr-Hill RA, Jenkins-Clarke S. Measurement systems in principle and in practice: the example of nursing workload. J Adv Nurs. 1995;22(2):221–5.
20. Cho SH, Ketefian S, Barkauskas VH, Smith DG. The effects of nurse staffing on adverse events, morbidity, mortality, and medical costs. Nurs Res. 2003;52(2):71–9.
21. Smith AP. Nursing-sensitive care measures: a platform for value and vision. Nurs Econ. 2007;25(1):43–6.
22. Smith DP, Jordan HS. Piloting nursing-sensitive hospital care measures in Massachusetts. J Nurs Care Qual. 2008;23(1):23–33.
23. Lacey SR, Klaus SF, Smith JB, Cox KS, Dunton NE. Developing measures of pediatric nursing quality. J Nurs Care Qual. 2006;21(3):210–20; quiz 221–2.
24. Curley MAQ, Hickey PA. The Nightingale metrics. Am J Nurs. 2006;106(10):66–70.
25. Alexander GR. Nursing sensitive databases: their existence, challenges, and importance. Med Care Res Rev. 2007;64(2 Suppl):44S–63.
26. Naylor MD. Advancing the science in the measurement of health care quality influenced by nurses. Med Care Res Rev. 2007;64(2 Suppl):144S–69.
27. Needleman J, Kurtzman ET, Kizer KW. Performance measurement of nursing care: state of the science and the current consensus. Med Care Res Rev. 2007;64(2 Suppl):10S–43.
28. Clarke SP, Aiken LH. Failure to rescue. Am J Nurs. 2003;103(1):42–7.
29. Schwalenstocker E, Bisarya H, Lau S, Adebimpe O. Nursing sensitive indicators for Children's Hospital care quality. Childrens Health Corporation of America; 2007
30. Institute of Medicine. The future of nursing: leading change, advancing health. Washington, DC: National Academies Press; 2010.
31. National Quality Forum. National consensus standards for nursing-sensitive care: an initial performance measure set. Washington, DC: National Quality Forum; 2004. p. 40.
32. American Nurses Association. National database of nursing quality indicators. 2013 [cited 6 Aug 2013]; Available from: http://www.nursingquality.org/About-NDNQI2.
33. Ohio Children's Hospitals Solutions for Patient Safety. Solutions for patient safety. 2013 [cited 6 Aug 2013]; Available from: http://solutionsforpatientsafety.org/.
34. Skretkowicz, V. Nightingale's notes on nursing: the first version and edition. vol. 15. London; 1993.
35. Agency for Healthcare Research and Quality. Kids Inpatient Database (KID). 2013 [cited 6 Aug 2013]; Available from: http://www.hcup-us.ahrq.gov/kidoverview.jsp.
36. Children's Hospital Corporation of America. Pediatric health information systems. Shawnee Mission: Child Health Corporation of America; 2013 [cited 6 Aug 2013]; Available from: http://www.chca.com/company_profile/pi/.
37. Iezzoni LI. Using administrative data to study persons with disabilities. Milbank Q. 2002;80(2):347–79.
38. Needleman J, Buerhaus P, Mattke S, Stewart M, Zelevinsky K. Nurse-staffing levels and the quality of care in hospitals. N Engl J Med. 2002;346(22):1715–22.
39. Aiken LH, Smith HL, Lake ET. Lower medicare mortality among a set of hospitals known for good nursing care. Med Care. 1994;32:771–87.
40. Aiken LH, Clarke SP, Sloane DM, Sochalski J, Silber JH. Hospital nurse staffing and patient mortality, nurse burnout, and job dissatisfaction. JAMA. 2002;288(16):1987–93.
41. Aiken LH, Sloane DM, Cimiotti JP, et al. Implications of the California nurse staffing mandate for other states. Health Serv Res. 2010;45(4):904–21.
42. Aiken LH, Sochalski J, Lake ET. Studying outcomes of organizational change in health services. Med Care. 1997;35(11 Suppl):NS6–18.
43. Hickey PA, Gauvreau K, Connor J, Sporing E, Jenkins K. The relationship of nurse staffing, skill mix, and Magnet recognition to institutional volume and mor-

tality for congenital heart surgery. J Nurs Adm. 2010; 40(5):226–32.
44. Blegen MA, Goode CJ, Reed L. Nurse staffing and patient outcomes. Nurs Res. 1998;47(1):43–50.
45. Drenkard K. Magnet perspectives: the value of Magnet. JONA. 2013;43(10):S2–3.
46. American Nurses Credentialing Center. ANCC Magnet Recognition Program. 2014 [cited 27 March 2014]; Available from: http://www.nursecredentialing.org/magnet.aspx.
47. Hickey PA, Gauvreau K, Tong E, Schiffer N, Connor JA. Pediatric cardiovascular critical care in the United States: nursing and organizational characteristics. Am J Crit Care. 2012;21(4):242–50.
48. Mark BA, Harless DW, Berman WF. Nurse staffing and adverse events in hospitalized children. Policy Polit Nurs Pract. 2007;8(2):83–92.
49. Jenkins K, Gauvreau K. Center-specific differences in mortality: preliminary analyses using the Risk Adjustment in Congenital Heart Surgery (RACHS-1) method. J Thorac Cardiovasc Surg. 2002;124(1): 97–104.
50. Hickey PA, Gauvreau K, Jenkins K, Fawcett J, Hayman L. Statewide and national impact of California's Staffing Law on pediatric cardiac surgery outcomes. J Nurs Adm. 2011;41(5):218–25.
51. Hickey PA, Gauvreau K, Curley MAQ, Connor JA. The effect of critical care nursing and organizational characteristics on pediatric cardiac surgery mortality in the United States. J Nurs Adm. 2013;43(12): 637–44.
52. Institute of Medicine. Preventing medication errors: report brief. 2006.
53. Chang YK, Mark BA. Antecedents of severe and non-severe medication errors. J Nurs Scholarsh. 2009; 41(1):70–8.
54. Eisenhauer LA, Hurley AC, Dolan N. Nurses' reported thinking during medication administration. J Nurs Scholarsh. 2007;39(1):82–7.
55. Elganzouri ES, Standish CA, Androwich I. Medication Administration Time Study (MATS): nursing staff performance of medication administration. J Nurs Adm. 2009;39(5):204–10.
56. Kaushal R, Jaggi T, Walsh K, Fortescue EB, Bates DW. Pediatric medication errors: what do we know? What gaps remain? Ambul Pediatr. 2004;4(1):73–81.
57. Mark BA, Belyea M. Nurse staffing and medication errors: cross-sectional or longitudinal relationships? Res Nurs Health. 2009;32(1):18–30.
58. Potter P, Wolf I, Boxerman S, et al. An analysis of nurses' cognitive work: a new perspective for understanding medical errors. Adv Patient Safety. 2005;1: 39–51.
59. Nebeker J, Samore M, Barach P. Clarifying adverse drug events: a clinicians guide to terminology, documentation, and reporting. Annals of Internal Medicine. 2005;142:77–78.
60. Hussain E, Kao E. Medication safety and transfusion errors in the ICU and beyond. Crit Care Clin. 2005; 21(1):91–110. ix.
61. Kane-Gill S, Rea RS, Verrico MM, Weber RJ. Adverse-drug-event rates for high-cost and high-use drugs in the intensive care unit. Am J Health Syst Pharm. 2006;63(19):1876–81.
62. Nelson NC, Evans RS, Samore MH, Gardner RM. Detection and prevention of medication errors using real-time bedside nurse charting. J Am Med Inform Assoc. 2005;12(4):390–7.
63. Pape TM, Guerra DM, Muzquiz M, et al. Innovative approaches to reducing nurses' distractions during medication administration. J Contin Educ Nurs. 2005; 36(3):108–16; quiz 141–2.
64. Nguyen E. Medication pass time out. Stanford Nurse. 2007;2, Fall:5–8.
65. Berg MD, Nadkarni VM, Zuercher M, Berg RA. In-hospital pediatric cardiac arrest. Pediatr Clin North Am. 2008;55(3):589–604. x.
66. Hunt EA, Zimmer KP, Rinke ML, et al. Transition from a traditional code team to a medical emergency team and categorization of cardiopulmonary arrests in a children's center. Arch Pediatr Adolesc Med. 2008;162(2):117–22.
67. Samson RA, Nadkarni VM, Meaney PA, et al. Outcomes of in-hospital ventricular fibrillation in children. N Engl J Med. 2006;354(22):2328–39.
68. Ortmann L, Prodhan P, Gossett J, et al. Outcomes after in-hospital cardiac arrest in children with cardiac disease: a report from get with the guidelines–resuscitation. Circulation. 2011;124(21):2329–37.
69. McLellan MC, Connor JA. The cardiac children's hospital early warning score (C-CHEWS). J Pediatr Nurs. 2012;28(2):171–8.
70. McLellan MC, Gauvreau K, Connor JA. Validation of the Cardiac Children's Hospital Early Warning Score: An early warning scoring tool to prevent cardiopulmonary arrests in children with heart disease. Congenit Heart Dis. 2014;9(3):194–202.
71. Curley MA, Razmus IS, Roberts KE, Wypij D. Predicting pressure ulcer risk in pediatric patients: the Braden Q Scale. Nurs Res. 2003;52(1):22–33.
72. Neidig JR, Kleiber C, Oppliger RA. Risk factors associated with pressure ulcers in the pediatric patient following open-heart surgery. Prog Cardiovasc Nurs. 1989;4(3):99–106.

22 Data Standards of the American College of Cardiology Foundation (ACCF) and the American Heart Association (AHA) and the Universal Pediatric Cardiac Dataset

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Abstract

This chapter will discuss the background and the attempt to create a universal pediatric cardiac dataset for use across electronic health record platforms. This dataset, consisting of a list of data elements and their definitions, is anticipated to be used as a basis for not only standardizing nomenclature across the various clinical domains of pediatric and congenital cardiac care, but also for allowing the electronic health record to capture discrete data that can be pooled together across institutions to better strengthen the quality and depth of research for this area of patient care.

Keywords

Domain • Data elements • Data definitions • Data dictionary • Congenital heart disease • Acquired pediatric heart disease

Background

The critical importance of having a nomenclature in any field in which data is being collected is foundational to being able to progress within that field of study and practice. The lack of a cohesive and comprehensive dictionary of terms prevents adequate communication, including communication among:

- those capturing the data,
- those analyzing the data,
- those interpreting and acting on the results, and
- those trying to improve quality after data analysis.

The field of pediatric and congenital cardiology and cardiac surgery not only has adhered to that understanding, but also has actually been quite active in creating working nomenclatures for decades. Pediatric and congenital cardiology as a field has needed to work harder to aggregate its data because unlike adult cardiology, in which there is a multitude of patients with very few

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overall diseases—essentially narrow but deep—this unique patient population of patients with pediatric and congenital cardiac disease is the exact opposite; it is wide but shallow, such that there are few patients with a wide variety of diseases. The limited numbers of patients with like diseases and like interventions make it difficult to be able to advance the field with case studies, case series, and small collaborative studies, which has comprised the first iteration of the medical literature in this discipline. Thus, pediatric cardiologists and cardiac surgeons have aggressively attempted to solve this initial dilemma—so much so, that in 2000, two different major international systems of nomenclature emerged. Despite the problems inherent to the creation of two independent major international systems of nomenclature, specifically from the van Praagh-based [1] and the Anderson-based [2] groups, there have been great strides made in the harmonization of these two sets into one cross-linked dataset, the International Paediatric and Congenital Cardiac Code (IPCCC) [3, 4]. It is interesting to note that the IPCCC started out as a list of data elements without specific definitions, and now continues to undergo refinement to construct the actual data dictionary that consists of elements, definitions, images, and videos. Although clearly leading to a major advance in the standardization of nomenclature for pediatric and congenital cardiac care, this approach may have caused some difficulties, in that the terms were approved prior to linking them with definitions, which could lead to backtracking in how some of the terms are used. As described elsewhere in this book (in the chapter by Franklin and colleagues titled: ***Nomenclature for Congenital and Pediatric Cardiac Disease: Historical Perspectives and the International Pediatric and Congenital Cardiac Code***), the IPCCC is a living and breathing international system of nomenclature that continues to evolve and improve.

However, the existence of a data dictionary does not guarantee its appropriate, consistent, and widespread utilization; its presence is only the first step. Despite the explosion in information sharing and transfer, individual, institutional, and regional habits have often persisted in preventing uniform use of terms and definitions. Meanwhile, as research in congenital cardiac care becomes increasingly collaborative and based across multiple centers, maintenance of consistency of a "lingua franca" not only for use within individual studies, but also for use across multiple studies to be able to further combine data, becomes even more imperative. This scenario has led to many local or study-specific datasets being created for either individual studies or a specific series of studies. However, these datasets may or may not harmonize with any other terms and definitions within the National Library of Medicine or the Systematized Nomenclature of Medicine (SNOMED). It is interesting to note that despite these multiple smaller datasets and research initiatives, the larger effort of The Society of Thoracic Surgeons (STS) in utilizing the IPCCC has been able to mobilize and to leverage much larger amounts of data on a wider scale in order to achieve the greater order of magnitude research required to be able to

- assess overall surgical outcomes [5],
- validate models of the assessment of risk [6], and
- demonstrate variation amongst institutions [7] in congenital cardiac surgery, cardiac anesthesia, and cardiac intensive care, including performance of "deep dives" into the database to parse specific demographic aspects of patients [8].

The American College of Cardiology Foundation (ACCF) and the American Heart Association (AHA) Task Force on Data Standards

The American College of Cardiology Foundation (ACCF) and the American Heart Association (AHA) took some initial steps in this domain by creating their Task Force on Data Standards in 2000, with their first publication in 2001 addressing the key data elements and definitions for measuring the clinical management and outcomes of patients with acute coronary syndromes [9]. Their approach was to create core data dictionaries for specific cardiac processes, with

subsequent data elements and definitions for topics such as

- atrial fibrillation [10],
- chronic congestive heart failure [11],
- electrophysiology [12],
- cardiac imaging [13], and
- peripheral atherosclerotic vascular disease [14].

Of note, they have recently created an updated version of their first set of data elements and definitions for measuring the clinical management and outcomes of patients with acute coronary syndromes and coronary artery disease [15] .

One of the later projects of the ACCF/AHA Task Force on Data Standards was to create data standards for the electronic health record. This approach was taken with the hope of standardizing how data was collected for and by means of the electronic health record and, eventually, how the electronic health record could be used for larger scale aggregation of data once health information exchanges were available. The first document addressing the electronic health record was aimed toward "cardiology", and was expected to be appropriate for both adult and pediatric patients and their practitioners [16]. It became evident, though, that the needs and interests of adult practitioners and those of pediatric and congenital practitioners were quite disparate and had little overlap. Thus, although the authors of this first document addressing the electronic health record included a pediatric cardiologist and a pediatric cardiac surgeon, the leadership of the Task Force agreed that a separate document for pediatric and congenital cardiac care was necessary in order to fully encompass the needs of this group.

The approach to this new document has been relatively straightforward. Pediatric and congenital cardiology was arbitrarily divided into multiple sections, or domains, that represented the various disciplines within cardiology and cardiac surgery. These included:

1. nomenclature,
2. ambulatory cardiology,
3. echocardiography,
4. cardiac catheterization,
5. physiology,
6. electrophysiology,
7. fetal cardiology,
8. adult congenital heart disease,
9. cardiac surgery,
10. cardiac anesthesia,
11. cardiac intensive care,
12. cardiac magnetic resonance imaging,
13. research,
14. cardiac pathology, and
15. extracorporeal life support and perfusion.

Many, though not all, of these disciplines had already established their own data dictionaries that had been harmonized with the IPCCC. It was also decided that the terminology associated with cardiac pathology was already represented within the nomenclature section, was redundant, and was thus eliminated. Each of these domains would be the subjects of their own individual, but harmonized, data dictionaries. The project would be introduced with a "white paper" describing the aims of the Task Force and the Writing Committee, including

- setting forth the domains and
- having a brief description, within each domain, of the known datasets that could be subsequently used as a starting point for creation of the various datasets in subsequent papers to be associated with each domain.

Known datasets created for outside collaborative studies would be reviewed for consideration of inclusion, either in part or in whole, in the larger dataset; many of these datasets were incomplete for these domains, as they were built for the specifics of their individual projects. Finally, the project was opened to invite collaboration with various other surgical and medical groups and societies as stakeholders that would have an interest in working with this group. These collaborating organizations included:

- The American Academy of Pediatrics (AAP),
- The Association for European Paediatric and Congenital Cardiology (AEPC),
- The Society of Thoracic Surgeons (STS),
- The Congenital Heart Surgeons' Society (CHSS),
- The International Society for Nomenclature of Paediatric and Congenital Heart Disease (ISNPCHD), and

- The Child Health Corporation of America (CHCA) and The National Association of Children's Hospitals and Related Institutions (NACHRI), which have now combined to become The Children's Hospital Association.

Through the American College of Cardiology Foundation and American Heart Association Task Force for Data Standards procedures, the review process for the dataset is multi-tiered. The first level of assessment is performed by the members of the writing committee. Once approved at that level, it moves on for evaluation by the ACCF/AHA Task Force on Data Standards. After their endorsement, it is then evaluated by the Board of Trustees. Afterwards, it is then released for evaluation by the partnering organizations. Finally, it is opened up for a period of public comment, allowing for multiple experts in various domains to be able to help to further sculpt and to improve the elements and definitions in a systematic and repetitive basis. All comments from all levels at each stage are addressed by the writing committee chairman, either by incorporation into the dataset, or by a rebuttal as to why the recommendation would not be appropriate.

For completeness in transparency, all members of the writing committee as well as the task force are required to declare any relationships with industry that may be perceived to or may actually be interpreted as influencing the content of the dataset. Between the multiple reviewers and the transparency, these features offer a stringency that many other organizations attempting to create similar works for widespread use have elected not to employ.

Thus, using these procedures and guidelines, the working group pressed forward with their work as defined. However, late during the formulation of the rough draft of the manuscript related to pediatric and congenital cardiac care, the leadership of the ACCF/AHA Task Force on Data Standards decided that, since prior publications by these groups had not included a white paper, this paper should also follow previous form and not stand alone as a white paper only. After some further discussion and negotiation, the Task Force approved the decision to both continue the original plan with the white paper in combination with at least one dataset, such that at least one dataset could be put forward along with the document related to pediatric and congenital cardiac care. The dataset for ambulatory cardiology was submitted, as a group of data elements that had been assembled independently prior to the inception of this project by Jeffrey Boris, MD. These elements of data were mapped to SNOMED and reviewed by the writing committee as well as the Task Force, plus representatives from each affiliate and partner organization. One problem that immediately surfaced was that the mapped terms arose from the National Cancer Institute Enterprise Vocabulary Services data dictionary, which was created by non-cardiologists and non-cardiac surgeons. This challenge led to two specific concerns:

- lack of a complete set of definitions to which to map the terms, and
- extant definitions that were not actually accurately defined for the needs of the community of pediatric and congenital cardiac care.

Therefore, at the time of this writing, extensive revision of the definitions of the dataset continues. However, as part of the revision, harmonization with the National Cancer Institute data dictionary will also improve this larger, non-cardiac dictionary (that is maintained by the National Cancer Institute) with more accurate and complete definitions for the terminology.

Vision for the Future

These documents are created so that they will be utilized widely across the spectrum of pediatric and congenital cardiac care. The eventual plan after creating these documents is to submit these documents to the Health Information Technology Standards Panel (HITSP) to be reviewed and accepted. Once accepted, it would be then submitted to the Certification Commission for Health Information Technology (CCHIT). The CCHIT would ensure that the various vendors of the electronic health record would have access to these data dictionaries, and would be required to employ them in upgraded versions of their

products in order to maintain CCHIT certification. This way, the same dataset would be both widely available and widely utilized by those vendors in disparate electronic health records.

As mentioned, the overall strategy in pushing these datasets forward for general use is to allow multiple specialists and sub-specialists to be able to use same structured data in their documentation as well as for capture of data, such that these data can be combined across institutions, both academic and private. At first, the expectation is that these data will be used in conjunction with envisioned Health Information Exchanges, entities that will allow for transfer of information about patients across various institutions, such that patients will have seamless communication among their various providers of healthcare as they enter healthcare from any point of access. The downstream hope, though, will be the ability to utilize these data for greater clinical research to improve outcomes for these patients. The combination of these data would then be able to then be used to better understand the multiple aspects of cardiac care across the various domains. This accomplishment will then allow for improved quality of care, as both cross-sectional and longitudinal acquisition of data can be parsed for findings that will provide not only better care in the short term but also allow for better prediction and prognosis for patients and their families. The hope is to build upon the extensive work performed by the ISNPCHD, harmonize with it, and extend it beyond the initial limitations of cardiac care in the operating room, the intensive care unit, the catheterization laboratory, and the anesthesiologist's place at the head of the table in the operating room. Most recently, the Data Standards Workgroup of the National Cardiovascular Research Infrastructure (NCRI) project has been taking a larger lead on this approach, and has published their latest list of standardized cardiovascular data [17]. Per their document, it is to be used for patient care, as well as clinical research and registries. They state that the NCRI was created to make a standardized data exchange in the area above, as well as for the electronic health record. They collaborated with the Clinical Data Interchange Standards Consortium and Health Level Seven International, plus the National Cancer Institute Enterprise Vocabulary Services. They started with 353 elements, the majority of which are not utilized by or are not important to practitioners or researchers in pediatric or congenital cardiac care. However, several terms exist that are specific congenital cardiac terms, though some may or may not map to the IPCCC; it is interesting to note that, again, no pediatric cardiologists and no pediatric cardiac surgeons were involved in the creation of this document. Thus, as the Universal Pediatric Cardiac Dataset continues through its development, it will also be harmonized with the NCRI project, and further extend the ability of cardiac caregivers and researchers in both the pediatric and adult realms to be able to capture data and to be able to work together across various platforms within the electronic health record or across domains of research.

One final caveat must be kept in mind as this pathway is embarked upon. Specific research databases are curated by personnel specifically trained in and dedicated to the entry and management of data, with routine auditing to ensure the veracity of those pieces of data entered into the data warehouse; however, the electronic health record has none of these features. It is available for use by personnel often with limited training in its utilization, and likely no training in ensuring data integrity. These personnel include housestaff, nurses, allied health staff, administrative personnel, and various others. Many of these people have previously had only a small amount of exposure to electronic health records. This situation means that although there may be a robust data dictionary available for use and programmed into the electronic health record, even its very highly reviewed contents and its very presence does not guarantee the quality of data. This fact is a small but important weakness in the ability to rely on the electronic health record as a standalone platform for both entry of data as well as compilation of data for later use. Certain data that are reviewed frequently, or those that have been assigned to have associated trained personnel for their entry, including such fields as demographics, are likely to be more

accurate sources of information that can be utilized in larger scale research. As well, those data that are associated with devices that precisely record measurements, such as monitors and infusion pumps, are also likely to be quite accurate. However, those data that are manually entered by humans with little or no training on single occasions are likely to be not routinely trustworthy for use for purposes of research. There are five potential methods around this potential weakness, however:

- One is the most obvious, but also the most costly – ensure adequate and complete training of anyone entering information into the system.
- The correlate to the first potential method is to limit those personnel who have access to entering information into the electronic health record.
- A third method is to limit which data is harvested such that only those data most likely to be accurate can be used.
- Utilization of templates that requires the answers to specific questions with limited choices is also an option, although in medicine, one size certainly does not fit all and it would likely be difficult to template everything required to obtain a history, physical examination, or other piece of documentation for care of the patient. As well, templates of that size become quite unwieldy, and lead to fatigue of use of the system.
- Finally, a newer method of verifying information is to link and to combine databases with each other, something already done by the Society of Thoracic Surgeons [18–25]. This last method assumes that the data captured by the electronic health record is also available in some other database or warehouse that can then be used to confirm the information in some way.

Once further advanced methods are devised that allow larger amounts of data in the electronic health record to be verified and collated these data standards for the electronic health record, as defined by the universal pediatric cardiac dataset, will be even more valuable.

References

1. Mavroudis C, Jacobs JP (editors). Congenital heart surgery nomenclature and database project. Ann Thorac Surg 2000;69(Suppl):S1–372.
2. Association for European Paediatric Cardiology. The European paediatric cardiac code. Cardiol Young. 2000;10 Suppl 1:1–146.
3. Béland M, Jacobs JP, Tchervenkov CI, Franklin RCG. The International Nomenclature Project for Paediatric and Congenital Heart Disease: report from the Executive of The International Working Group for Mapping and Coding of Nomenclatures for Paediatric and Congenital Heart Disease. Cardiol Young. 2002;12:425–30.
4. Franklin RCG, Jacobs JP, Tchervenkov CI, Béland MJ. The International Nomenclature Project for Congenital Heart Diseaes: bidirectional crossmap of the short lists of the European Paediatric Cardiac Code and the International Congenital Heart Surgery Nomenclature database Project. Cardiol Young. 2002;12:431–5.
5. Jacobs JP, O'Brien SM, Pasquali SK, Jacobs ML, Lacour-Gayet FG, Tchervenkov CI, Austin 3rd EH, Pizarro C, Pourmoghadam KK, Scholl FG, Welke KF, Mavroudis C. Variation in outcomes for benchmark operations: an analysis of the Society of Thoracic Surgeons Congenital Heart Surgery Database. Ann Thorac Surg. 2011;92(6):2184–91.
6. Rankin JS, He X, O'Brien SM, Jacobs JP, Welke KF, Filardo G, Shahian DM. The Society of Thoracic Surgeons risk model for operative mortality after multiple valve surgery. Ann Thorac Surg. 2013;95(4):1484–90.
7. Jacobs JP, O'Brien SM, Pasquali SK, Jacobs ML, Lacour-Gayet FG, Tchervenkov CI, Austin 3rd EH, Pizarro C, Pourmoghadam KK, Scholl FG, Welke KF, Gaynor JW, Clarke DR, Mayer Jr JE, Mavroudis C. Variation in outcomes for risk-stratified pediatric cardiac surgical operations: an analysis of the STS Congenital Heart Surgery Database. Ann Thorac Surg. 2012;94(2):564–71.
8. DiBardino DJ, Pasquali SK, Hirsch JC, Benjamin DK, Kleeman KC, Salazar JD, Jacobs ML, Mayer JE, Jacobs JP. Effect of sex and race on outcome in patients undergoing congenital heart surgery: an analysis of the Society of Thoracic Surgeons Congenital Heart Surgery Database. Ann Thorac Surg. 2012;94(6):2054–9.
9. Cannon CP, Battler A, Brindis RG, Cox JL, Ellis SG, Every NR, Flaherty JT, Harrington RA, Krumholz HM, Simoons ML, Van De Werf FJ, Weintraub WS, Mitchell KR, Morrisson SL, Brindis RG, Anderson HV, Cannom DS, Chitwood WR, Cigarroa JE, Collins-Nakai RL, Ellis SG, Gibbons RJ, Grover FL, Heidenreich PA, Khandheria BK, Knoebel SB, Krumholz HL, Malenka DJ, Mark DB, Mckay CR, Passamani ER, Radford MJ, Riner RN, Schwartz JB, Shaw RE, Shemin RJ, Van Fossen DB, Verrier ED, Watkins MW, Phoubandith DR, Furnelli T. American College of Cardiology key data elements and definitions for measuring the clinical management and outcomes of patients with acute coronary syndromes.

A report of the American College of Cardiology Task Force on Clinical Data Standards (Acute Coronary Syndromes Writing Committee). J Am Coll Cardiol. 2001;38(7):2114–30.

10. McNamara RL, Brass LM, Drozda Jr JP, Go AS, Halperin JL, Kerr CR, Lévy S, Malenka DJ, Mittal S, Pelosi Jr F, Rosenberg Y, Stryer D, Wyse DG, Radford MJ, Goff Jr DC, Grover FL, Heidenreich PA, Malenka DJ, Peterson ED, Redberg RF. American College of Cardiology/American Heart Association Task Force on Clinical Data Standards (Writing Committee to Develop Data Standards on Atrial Fibrillation). ACC/AHA key data elements and definitions for measuring the clinical management and outcomes of patients with atrial fibrillation: a report of the American College of Cardiology/American Heart Association Task Force on Clinical Data Standards (Writing Committee to Develop Data Standards on Atrial Fibrillation); American College of Cardiology; American Heart Association. Circulation. 2004;109(25):3223–43.
11. Radford MJ, Arnold JM, Bennett SJ, Cinquegrani MP, Cleland JG, Havranek EP, Heidenreich PA, Rutherford JD, Spertus JA, Stevenson LW, Goff DC, Grover FL, Malenka DJ, Peterson ED, Redberg RF. American College of Cardiology; American Heart Association Task Force on Clinical Data Standards; American College of Chest Physicians; International Society for Heart and Lung Transplantation; Heart Failure Society of America. ACC/AHA key data elements and definitions for measuring the clinical management and outcomes of patients with chronic heart failure: a report of the American College of Cardiology/American Heart Association Task Force on Clinical Data Standards (Writing Committee to Develop Heart Failure Clinical Data Standards): developed in collaboration with the American College of Chest Physicians and the International Society for Heart and Lung Transplantation: endorsed by the Heart Failure Society of America. Circulation. 2005;112(12):1888–916.
12. American College of Cardiology/American Heart Association Task Force on Clinical Data Standards (ACC/AHA/HRS Writing Committee to Develop Data Standards on Electrophysiology), Buxton AE, Calkins H, Callans DJ, DiMarco JP, Fisher JD, Greene HL, Haines DE, Hayes DL, Heidenreich PA, Miller JM, Poppas A, Prystowsky EN, Schoenfeld MH, Zimetbaum PJ, Goff DC, Grover FL, Malenka DJ, Peterson ED, Radford MJ, Redberg RF. ACC/AHA/HRS 2006 key data elements and definitions for electrophysiological studies and procedures: a report of the American College of Cardiology/American Heart Association Task Force on Clinical Data Standards (ACC/AHA/HRS Writing Committee to Develop Data Standards on Electrophysiology). Circulation. 2006;114(23):2534–70.
13. Hendel RC, Budoff MJ, Cardella JF, Chambers CE, Dent JM, Fitzgerald DM, Hodgson JM, Klodas E, Kramer CM, Stillman AE, Tilkemeier PL, Ward RP, Weigold WG, White RD, Woodard PK. ACC/AHA/ACR/ASE/ASNC/HRS/NASCI/RSNA/SAIP/SCAI/SCCT/SCMR/SIR 2008 key data elements and definitions for cardiac imaging: a report of the American College of Cardiology/American Heart Association Task Force on Clinical Data Standards (Writing Committee to Develop Clinical Data Standards for Cardiac Imaging). Circulation. 2009;119(1):154–86.
14. Writing Committee to Develop Clinical Data Standards for Peripheral Atherosclerotic Vascular Disease, Creager MA, Belkin M, Bluth EI, Casey Jr DE, Chaturvedi S, Dake MD, Fleg JL, Hirsch AT, Jaff MR, Kern JA, Malenka DJ, Martin ET, Mohler III ER, Murphy T, Olin JW, Regensteiner JG, Rosenwasser RH, Sheehan P, Stewart KJ, Treat-Jacobson D, Upchurch Jr GR, White CJ, Ziffer JA, Hendel RC, Bozkurt B, Fonarow GC, Jacobs JP, Peterson PN, Roger VL, Smith EE, Tcheng JE, Wang T, Weintraub WS. 2012 ACCF/AHA/ACR/SCAI/SIR/STS/SVM/SVN/SVS key data elements and definitions for peripheral atherosclerotic vascular disease: a report of the American College of Cardiology Foundation/American Heart Association Task Force on Clinical Data Standards (Writing Committee to Develop Clinical Data Standards for Peripheral Atherosclerotic Vascular Disease). Circulation. 2012;125(2):395–467.
15. Cannon CP, Brindis RG, Chaitman BR, Cohen DJ, Cross Jr JT, Drozda Jr JP, Fesmire FM, Fintel DJ, Fonarow GC, Fox KA, Gray DT, Harrington RA, Hicks KA, Hollander JE, Krumholz H, Labarthe DR, Long JB, Mascette AM, Meyer C, Peterson ED, Radford MJ, Roe MT, Richmann JB, Selker HP, Shahian DM, Shaw RE, Sprenger S, Swor R, Underberg JA, Van de Werf F, Weiner BH, Weintraub WS. American College of Cardiology Foundation/American Heart Association Task Force on Clinical Data Standards; American College of Emergency Physicians; Emergency Nurses Association; National Association of Emergency Medical Technicians; National Association of EMS Physicians; Preventive Cardiovascular Nurses Association; Society for Cardiovascular Angiography and Interventions; Society of Cardiovascular Patient Care; Society of Thoracic Surgeons. 2013 ACCF/AHA key data elements and definitions for measuring the clinical management and outcomes of patients with acute coronary syndromes and coronary artery disease: a report of the American College of Cardiology Foundation/American Heart Association Task Force on Clinical Data Standards (Writing Committee to Develop Acute Coronary Syndromes and Coronary Artery Disease Clinical Data Standards). Circulation. 2013;127(9):1052–89.
16. Weintraub WS, Karlsberg RP, Tcheng JE, Boris JR, Buxton AE, Dove JT, Fonarow GC, Goldberg LR, Heidenreich P, Hendel RC, Jacobs AK, Lewis W, Mirro MJ, Shahian DM, Hendel RC, Bozkurt B, Jacobs JP, Peterson PN, Roger VL, Smith EE, Tcheng JE, Wang T. American College of Cardiology Foundation; American Heart Association Task Force on Clinical Data Standards. ACCF/AHA 2011 key

data elements and definitions of a base cardiovascular vocabulary for electronic health records: a report of the American College of Cardiology Foundation/American Heart Association Task Force on Clinical Data Standards. Circulation. 2011;124(1):103–23.
17. Anderson HV, Weintraub WS, Radford MJ, Kremers MS, Roe MT, Shaw RE, Pinchotti DM, Tcheng JE. Standardized cardiovascular data for clinical research, registries, and patient care: a report from the data standards workgroup of the national cardiovascular research infrastructure project. J Am Coll Cardiol. 2013;61(18):1835–46.
18. Jacobs JP, O'Brien SM, Shahian DM, Edwards FH, Badhwar V, Dokholyan RS, Sanchez JA, Morales DL, Prager RL, Wright CD, Puskas JD, Gammie JS, Haan CK, George KM, Sheng S, Peterson ED, Shewan CM, Han JM, Bongiorno PA, Yohe C, Williams WG, Mayer JE, Grover FL. Successful linking of the Society of Thoracic Surgeons Database to Social Security data to examine the accuracy of Society of Thoracic Surgeons mortality data. J Thorac Cardiovasc Surg. 2013;145(4):976–83.
19. Dokholyan RS, Muhlbaier LH, Falletta J, Jacobs JP, Shahian D, Haan CK, Peterson ED. Regulatory and ethical considerations for linking clinical and administrative databases. Am Heart J. 2009;157(6):971–82. PMID: 19464406.
20. Jacobs JP, Edwards FH, Shahian DM, Haan CK, Puskas JD, Morales DLS, Gammie JS, Sanchez JA, Brennan JM, O'Brien SM, Dokholyan RS, Hammill BG, Curtis LH, Peterson ED, Badhwar V, George KM, Mayer Jr JE, Chitwood WR, Murray GF, Grover FL. Successful linking of The Society of Thoracic Surgeons Adult Cardiac Surgery Database to Centers for Medicare and Medicaid Services Medicare Data. Ann Thorac Surg. 2010;90:1150–7.
21. Pasquali SK, Jacobs JP, Shook GJ, O'Brien SM, Hall M, Jacobs ML, Welke KF, Gaynor JW, Peterson ED, Shah SS, Li JS. Linking clinical registry data with administrative data using indirect identifiers: implementation and validation in the congenital heart surgery population. Am Heart J. 2010;160:1099–104.
22. Jacobs JP, Edwards FH, Shahian DM, Prager RL, Wright CD, Puskas JD, Morales DL, Gammie JS, Sanchez JA, Haan CK, Badhwar V, George KM, O'Brien SM, Dokholyan RS, Sheng S, Peterson ED, Shewan CM, Feehan KM, Han JM, Jacobs ML, Williams WG, Mayer Jr JE, Chitwood Jr WR, Murray GF, Grover FL. Successful linking of the Society of Thoracic Surgeons Database to Social Security Data to Examine Survival after Cardiac Operations. Ann Thorac Surg. 2011;92(1):32–9. PMID: 21718828.
23. Pasquali SK, Li JS, Jacobs ML, Shah SS, Jacobs JP. Opportunities and challenges in linking information across databases in pediatric cardiovascular medicine. Progr Pediatr Cardiol. 2012;33(1):21–4. doi:10.1016/j.ppedcard.2011.12.004. In: Steven E. Lipshultz, MD, Paul Barach, MD, MPH, Jacobs JP, and Laussen P, (Editors). Progress in pediatric cardiology: the future of pediatric and congenital cardiac care special part 2. 2012;33(1):1–101.
24. Jacobs JP, Pasquali SK, Austin E, Gaynor JW, Backer C, Romano JCH, Williams WG, Caldarone C, McCrindle BW, Graham K, Dokholyan RS, Shook G, Poteat J, Baxi M, Karamlou T, Morris JA, Blackstone EH, Mavroudis C, Mayer Jr JE, Jonas RA, Jacobs ML. Linking the congenital heart surgery databases of the Society of Thoracic Surgeons (STS) and the Congenital Heart Surgeons' Society (CHSS): part 1 – Rationale and Methodology. World J Pediatr Congen Heart Surg. 2014;5(2):256–71. doi:10.1177/2150135113519454.
25. Jacobs JP, Pasquali SK, Austin E, Gaynor JW, Backer C, Romano JCH, Williams WG, Caldarone C, McCrindle BW, Graham K, Dokholyan RS, Shook G, Poteat J, Baxi M, Karamlou T, Morris JA, Blackstone EH, Mavroudis C, Mayer Jr JE, Jonas RA, Jacobs ML. Linking the congenital heart surgery databases of the Society of Thoracic Surgeons (STS) and the Congenital Heart Surgeons' Society (CHSS): part 2 – lessons learned and implications. World J Pediatr Congen Heart Surg. 2014;5(2):272–82. doi:10.1177/2150135113519455.

Ethical Issues Confronting Outcomes Analysis and Quality Assurance

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Abstract

The Society of Thoracic Surgeons Congenital Heart Surgery Database (STS-CHSD) has served as the primary, externally validated source for outcomes from its many participating centers. Because participation is voluntary and data are not publically reported, there exists a fundamental conflict with the overwhelming trend toward public reporting in both medicine and cardiac surgery. The ethical issues surrounding this conflict, as well as those involved in public reporting of complex congenital heart disease in general are described herein. The process by which adult cardiac surgery has come to initiate public reporting, while an important model for future directions in outcome reporting, does not adequately address the unique challenges that are faced by the reporting of congenital heart surgical outcomes. If these ethical issues are addressed and public reporting can be done in a responsible manner, then it should be undertaken by the leadership of physician-based groups such as the Society of Thoracic Surgeons.

Keywords

Congenital heart surgery • Database • Public reporting

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Historical Perspective

Outcomes analysis for mortality and morbidity for congenital heart surgery on a large scale was made possible by the development of the Society of Thoracic Surgeons Congenital Heart Surgery Database (STS-CHSD) [1–4]. The intent of the STS-CHSD was to provide a voluntary data bank for individual programs to share their own de-identified data with other programs in the United States and Canada for the purpose of self-assessment, quality assurance, and quality improvement. The original and current agreement between the Society of Thoracic Surgeons (STS) and participating programs was designed so that the data from an individual institution or surgeon is confidential and is not to be divulged as a singular entity by STS. In this agreement, each program would input their own data and receive a Feedback Report comparing their individual local programmatic data to national aggregate data. These Feedback Reports were initially distributed every year and are now distributed every 6 months. In other words, the data about demographics, mortality, and morbidity from an individual program is compared with the aggregate data of all participating centers from the United States and Canada for each of the various procedures, whether they are common or rare. With the advent of risk stratification scoring systems in congenital heart surgery [1–4], an individual program can assess its own complexity, mortality, and morbidity as they relate to the aggregate data from the United States and Canada. Of paramount importance to the previous and current success of this process was the strict anonymity it provided to its participants. No program could know the identity of any other program and, if there were a particular surgeon in a group who was underperforming, only that particular group or program would know.

There has been increasing pressure to report these data to patients and families of patients, other programs, the news media, governmental bureaus, third party payers, and the like. The tenet of "raising all the boats in the harbor" is operationalized when underperforming programs are alerted and voluntarily make plans to improve their outcomes. At the time of inception of the STS-CHSD, there was no detailed long-term plan to participate in public reporting. For some, concern existed that the data could be skewed and misinterpreted by the public.

Like most established tenets, rules, and constitutions, change occurs; and in some cases, it occurs rapidly. The movement towards public reporting, participation in governmental databases, and long-term outcomes research has driven the initial agreements concerning the database to evolve. Already, the STS is exploring the methods that will allow public reporting to move forward in an organized and appropriate manner. The STS Adult Cardiac Surgery Database has recently operationalized Public Reporting. These new trends raise ethical issues that require discussion and analysis.

The Ethical Issues

Any exegesis of ethical considerations must address the four cardinal ethical principles, which include respect for patient autonomy, beneficence, non-malfeasance, and justice. These terms have been extensively described and defined and, while it is beyond the scope of this chapter to define these terms extensively, a basic review will serve to illustrate how these core principles relate to the issue at hand. Respect for patient autonomy is self-explanatory, but such respect is often challenging in the physician patient relationship. Beneficence and non-malfeasance seem to be two sides to the same coin. But while doing good and not doing harm are apparently logically equivalent, they are very much separate in the physician patient relationship—a relationship that often calls for doing harm (i.e., surgery) for a patient's health. As such, consideration of each is necessary and challenging in different ways. Justice has been a term fraught with controversy since Plato's *Republic*, and likely before. A comprehensive definition remains illusory. For ethical considerations, issues such as health policy, economics, health spending discrepancies, availability to healthcare, and ensuring consistency of care are but a few of the myriad considerations

for this multifaceted principle. Indeed, perhaps the only thing more difficult than defining the cardinal ethical principles is properly delineating the jurisdiction and limits of one versus another. Such a distinction will not be attempted herein. Suffice it to say, most of these principles are instantiated in the issues that are raised, and it is far less important to understand where an individual issue is pertinent than it is to use these principles to engage properly the proceeding ethical problems with outcomes analysis.

The root of most of the ethical issues involved with outcomes analysis and quality assurance is simple: what does one do with the information obtained? As mentioned previously, one of the founding principles of the STS-CHSD is to serve as a de-identified and voluntary exchange of information to benefit all centers; its purpose, at least from its outset, is not to identify individual programs that are underperforming in a public setting. Rather, the programs are expected to make voluntary changes to improve their outcomes relative to other sites, with the database serving as a benchmark by which it is judged. Three main dilemmas and questions emerge from such a system: (1) how are programs incentivized to improve their quality metrics relative to other programs? (2) what, if any, role does the governing body and those with access to all data have in forcing programs to change? (3) who is responsible for revealing subpar performance to patients or potential patients in a given hospital's jurisdiction?

These issues highlight the divide between the legal contract that exists among programs entering the STS-CHSD (that data are to be used to improve outcomes) and the lack of punitive measures in place for those programs that do not or cannot improve their subpar outcomes. This disconnect begs the question of why there are no punitive measures in place, but what would such punitive measures resemble? The obvious choice would be to identify those programs to an audience of their peers (i.e., other programs in the STS-CHSD), or even to the public at large.

Because there is no requirement that programs participate, the threat of public reporting or humiliation among peers might be enough for programs to cease participation. The statistical power and legitimacy of the database would suffer as a result, and the purpose of the database (i.e., to compare, standardize, and improve outcomes) would be threatened. The moral hazard thus presented is one of preserving the purpose of the database and allowing complacent or delinquent programs to put patients at risk relative to other programs to preserve the overall mission that, in time, might benefit more patients in the long term because of maintenance of broad participation of multiple institutions. If it can be assumed that programs will always reform themselves when presented with findings from the database that demonstrate relatively poor performance, then perhaps the STS-CHSD does not need any such measures to enforce quality progression and improvement.

If it cannot be assumed that programs will reform themselves, and if universal participation requires that programs not be censured or revealed for their substandard performance, then who has the authority to enforce quality improvement or to standardize outcomes where disparities exist? Such authority can lie with the physicians/surgeons who oversee the STS-CHSD, those who analyze the results, and the STS as a professional medical society. The STS and its leadership can identify which programs are underperforming, to what degree they are underperforming and, specifically if there are major discrepancies in surgical outcomes among participating programs that serve the same geographic areas. For example, what if program A and program B from the same geographic area register Norwood mortalities of 5 and 50 %, respectively? Do the physicians/surgeons in charge of the STS-CHSD have moral responsibility to the patients in the area, even though they are governed by secrecy according to the working relationship between the STS and the participating programs? In other words, does moral authority trump the black letter law/positive law/agreements that have been established by contracts and written accords? If asked by a patient/parent about any underperforming program, is the STS or its physician leadership required to divulge privileged information or is the

organization required to remain silent and to choose the supposed greater good based on the agreements that they should remain silent on the issues?

Even aligning the two sides of this moral dilemma with major debates in the history of ethics proves challenging. While the main issue is one of deontological (duty-based) versus utilitarian (doing the most good for the greatest number) ethics, the group to whom the STS leadership is duty-bound and the uncertainty of how to achieve the greater good for the greatest number is problematic. Is the STS leadership bound by the contract made among its participating members to maintain the integrity and statistical power of the database by protecting its membership or is it bound to individual patients who may be at risk at centers with worse outcomes? And if these patients are at risk, is the number of patients potentially saved by censuring or revealing lesser performing centers greater than those potentially saved through meticulous and complete data collection to accomplish the stated goal of improving national outcomes? If one perfunctory program is singled out for its lesser outcomes, patients will likely travel to a more reputable center, where they may or may not receive better care. Under such circumstances, programs would be less willing to participate in the data collection process, and the goals of the STS-CHSD would be threatened.

In many ways, the moral hazards present in outcomes analysis and quality assurance are a macrocosm of the ethical issues raised in any prospective medical study. In either case, the safety and autonomy of patients affected by any study needs to be paramount, and the benefits of continuing the study cannot be outweighed by the risks of continuing study participation. The difference in outcomes analysis and quality assurance studies is that programs, not individual patients, are the direct study participants. It is the programs, not individual patients, who consent to be studied; and these programs expect to benefit from the study's findings much like patients expect to benefit from a prospective study's findings. Viewed in this way, any benefits that may come to patients through outcomes analysis is secondary to those that may come to the participating programs. How are we to best navigate these moral hazards?

Analogues in Adult Cardiac Surgery

The identification of under and over achieving was attempted in adult cardiac surgery in the early years of public reporting, and many comparisons can be drawn from this experience. Early attempts were made on the state level, with New York State (1990) and, later, Pennsylvania (1992) releasing hospital-specific raw and risk-adjusted mortality rates for coronary artery bypass grafting (CABG) [5]. These data served as the basis for the Cardiac Surgery Reporting System (CSRS) in New York, and were used to compare outcomes for hospitals and for individual surgeons through yearly reports [6]. Initially these reports received considerable publicity by demonstrating significant variation among surgeons' mortalities and owing to the reporting of a decline in risk-adjusted mortality rates for CABG in New York. Such a decline was attributed to the advent of the reports, suggesting that public reporting improves outcomes [7–9].

The CSRS is not without its critics or its flaws, and it has been the source of controversy since its inception. Much has been written regarding the various flaws in the CSRS [10, 11], and the details of these arguments are well beyond the scope of this chapter. Briefly, the main grievances that scholars and surgeons have with the CSRS is that it does not adequately risk-stratify patients, it does not adequately stratify for case-mix among surgeons, it negatively incentivizes operating on sicker patients, its findings are difficult to validate externally, its endpoints and case classification systems have been revised significantly during the study periods, and it supposedly prompted a change in surgeons' behavior toward not operating on sicker patients [10]. Even the alleged decrease in risk-adjusted mortality is not without critics. The incidence of five major comorbidities (renal failure, chronic obstructive pulmonary disease, congestive heart failure, unstable angina, and low ejection fraction) had staggering increases

during the study period (the prevalence of congestive heart failure and renal failure increased 347 and 600 %, respectively), which were difficult to explain or interpret [12].

Were surgeons finally reporting the comorbidities of their patients accurately or were they making their patients seem sicker to increase their reported complexity or to misreport a poor outcome to a supposedly, but perhaps not genuinely, "complex" patient? Such an example is characteristic of the Hawthorne Effect, a phenomenon in epidemiology where subjects who know they are being studied alter their behavior. Such a bias is inevitable in an observational study with so many variables in place but, as the critics argue, it nonetheless harms the credibility findings of the CSRS. It is thus argued that the supposed decrease in risk adjusted mortality was more affected by the increase in predicted mortality (presumably by spurious comorbidity assignment) than it was by a decrease in observed mortality [12]. There is at least anecdotal evidence of surgeons refusing to operate on high risk patients or referring them out of state for fear that it would negatively impact their ranking [10, 13–15]. The ranking process, too, was criticized for failing to account for variations in case mix and for its considerable fluctuation in ranking which, in 1 year, saw 46 % of surgeons change from one-half of the rank list to another [12].

The STS Database was made available to STS participants in 1989 and has sought to ameliorate the problems that plagued the CSRS by standardizing surgical and procedural nomenclature throughout the field, providing a minimal data set with precise definitions of data to be reported, developing and implementing accurate and verified mechanisms of adjustment for complexity of the patients, and operationalizing methodologies of data verification [16]. Perhaps the most important of its attempts to standardize practice has been the requirement of case-mix adjustment to be reported across programs. Such a measurement serves as a direct indicator of programmatic complexity, which helps to eliminate the supposed incentives for hospitals or groups to avoid caring for more complex patients. The specifics of how the risk stratification schemes are performed in the STS Adult Cardiac Surgery Database, the STS General Thoracic Surgery Database, and the STS Congenital Heart Surgery Database are beyond the scope of this chapter, but the presence of such risk stratification schemes have been shown to correlate well with reality based outcomes in several studies [3, 4, 17–20].

In 2011, the STS launched the STS Public Reporting Online, which currently allows hospitals and programs to report voluntarily their outcomes in isolated CABG and aortic valve replacement [21–23]. This reporting allows the public to compare programmatic and hospital specific mortality and morbidity using a composite scoring system that has been similarly risk stratified. Programs and hospitals are graded on a star rating system that assigns a rating of one star, two stars, or three stars: one and three star programs have a 99 % Bayesian probability that they differ from the STS average. Anticipating the criticism that plagued the CSRS study, the STS devotes an entire page of its Web site to addressing potential pitfalls of public reporting, citing surgeon risk aversion, timeliness of outcome reporting, and how their methods help to eliminate these pitfalls [24].

Congenital Heart Surgery Outcomes Reporting as a Unique Dilemma

The preceding discussion of the evolution of aortic valve replacement and CABG outcomes reporting serves to highlight the overwhelming complexity that public reporting of outcomes measurements face in cardiac surgery. A project that has evolved since 1989 and is evidence-based, risk stratified, and objectively validated, is currently only reported publicly in two isolated adult cardiac surgical procedures. If there are difficulties associated with risk stratification, patient selection, and determination of patient clinical status in disease processes that are so widely studied as coronary artery disease and aortic valve disease, then the more esoteric and complicated world of congenital heart disease would be even more fraught with issues in public reporting.

Indeed, one can argue that the entire ethical dilemma of public reporting of cardiac surgical data is the inherent difference in understanding between the physician and patient. That is to say, the knowledge that physicians have (that patients do not) creates a disadvantage for patients that undermines their autonomy to make competent medical decisions. Such a knowledge gap is pervasive in medicine, and the preceding discussion serves to highlight just how challenging it can be to ameliorate this knowledge gap, even in disease processes about which the public has a great deal of exposure and knowledge: coronary artery disease and aortic valve disease.

The question that invariably arises is: can the public be educated sufficiently to understand complexity analysis in congenital heart malformations and the surgical procedures associated therewith? Short of providing a comprehensive medical education to everyone, such a knowledge gap seems almost insurmountable. But there needs to be a way for the public to be able to compare programs and surgeons effectively without being subject to unfair biases. Such has been the goal of the STS-CHSD, whose history closely parallels that for adult cardiac surgery. The need for proper public reporting is illustrated by a 2001 article in the *Denver Post* concerning the resignation of the hospital's chief of cardiology. The resignation, the article claims, was in response to a reported average mortality of 4.2 % for open heart surgery, in comparison to an alleged national mortality rate of 2.7 % [25, 26]. The author even went so far as to cite the STS as the source for the mortality figures. Such an article would seem scandalous if the data were true and would explain the resignation well. However, at the time of that article's printing, the STS had never published an overall national aggregate mortality rate for congenital heart surgery. Further, it is argued by Jacobs and colleagues, if such a mortality figure were to be published based on STS data, it would have been greater than 4 % at that time [27]. Furthermore, comparing and benchmarking the overall mortality of an individual program to an overall national aggregate mortality rate is meaningless without adjustment for case mix.

So, in an attempt to sell newspapers, the author interpreted one site's data without incorporating case-mix or complexity analysis and compared it with a national average that did not exist. Such an example highlights the importance of reporting correct data. To be sure, the story was well read throughout Denver and may have caused some citizens to cast a critical eye on the cardiac surgical program at Children's Hospital of Denver, but it was based on misinterpretation and misinformation. The newspaper never retracted or amended its story and did not publish rebuttals from cardiac surgeons.

Such sensationalism is made all the more challenging to combat when the disease processes and surgical procedures in congenital heart surgery are so difficult to understand among lay persons. Even databases are subject to confusion and misclassification, as several studies analyzing the validity of codes in the International Classification of Diseases have shown that it is inadequate for use as a platform for the analysis of outcomes of patients with pediatric and congenital cardiac disease because many diseases are misclassified [28–31]. Explanations for these findings are numerous and are beyond the scope of this chapter, but the challenges simply of classifying and comparing congenital malformations of the heart is a problem unique to congenital heart disease and makes proper outcomes reporting even more difficult.

Conclusion

Despite the difficulties, there can be no doubt that the public demand transparency from healthcare providers to make informed and better valued medical decisions. There are many challenges to outcomes reporting in congenital heart disease. Surgeons and other cardiac care providers are required to inform their patients and families to allow them to make the best decisions for themselves and for their children. A problem can arise, however, when there is miscommunication and misinformation that clouds a patient's judgment. Who knows what the overall effect of the *Denver Post* article was? Perhaps there were families who travelled elsewhere to seek

a cardiac surgical consultation for their child because of this seemingly negative article. Even in the gold standard of STS public reporting of aortic valve replacement and CABG data, there is the possibility that patients will feel the need to travel to have surgery at a center that is purported to be superior to others.

Publishing, as the STS does currently, the mortality and morbidity figures for specific procedures may be functional in disease processes about which the public has a great deal of knowledge, but may not work as well when the disease processes are more esoteric. For example, if a given surgeon has a reported mortality for repair of coarctation of the aorta that appears significantly higher than that of others, and there is no public reporting of her outcomes in Norwood Stage 1 operations, then how is the public to interpret this? Will a parent avoid this surgeon, even if her morbidity and mortality figures for Norwood Stage 1 operations are actually quite good but is not publically reported? The current reporting of isolated aortic valve replacement and CABG data works because the cases are the most common cardiac surgical procedures performed and are reported using validated tools that allow for the adjustment of case mix. If a reporting scheme is attempted in congenital heart surgery, one can expect that public reporting for specific procedures will start with the more common operations and will, perhaps, proceed to rarer operations. Will this discrepancy of which procedures are reported and which are not create situations where surgeons are judged by their success in but a small fraction of their practice? Most common procedures in congenital heart surgery have excellent success rates, but a concerned parent with a child with hypoplastic left heart syndrome would like to know more than a surgeon's and medical center's success with a ventricular septal defect repair.

The ultimate challenge associated with public reporting is the burden that it places on the healthcare consumer. The opportunity to be informed in the medical decision making process carries with it the right and responsibility to make the best choice for their child, even if it means seeking a consultation elsewhere. In adult cardiac surgery, there may be numerous options available in one's immediate vicinity. In congenital heart surgery, there may be no other options that are geographically convenient for a parent, who may have numerous other familial and community commitments. The increase in demands for transparency by the public, regulators and payers has brought this dilemma to the fore. The STS has been active in standardization of language and outcomes reporting in adult and pediatric cardiothoracic surgery, and these activities must continue. The STS Database remains a voluntary database; nevertheless, it is critical to ensure universal or near universal participation because, as the previous discussion shows, more data and more information will stem the tide of misinformation and sensationalism. In order to engage in responsible public reporting, STS must engage and seek approval of the participating programs given that the extant agreement that resulted in this heretofore successful enterprise did not include public reporting.

No risk stratification scheme and not even the most comprehensive database can adequately describe the complexities of individual patients. As such, public reporting will never substitute for a surgical consultation where all factors can be explored. The interaction between physician and patient where a patient's condition, a family's condition, and the proper course of action can be determined will always serve as the cornerstone for preoperative evaluation, and no public outcomes reporting can replace this aspect of informed consent. Addressing the ethical pitfalls inherent in public outcomes reporting may assuage some of the apprehension among surgeons that this project will lead to decreased case volume. When and if these concerns are assuaged and the proper methodologies are in place for responsible public reporting, then public reporting may serve to accomplish the

goal of identifying best practices and improving the overall state of congenital heart surgery. The impetus for such public reporting should not be simply a matter of inevitability, or of preventing other organizations from doing it; rather, the responsible reporting of clinical outcomes should be done to facilitate better communication between physician and patient and, in so doing, to do the best thing for our patients.

References

1. Jacobs JP, Mavroudis C, Jacobs ML, Maruszewski B, Tchervenkov CI, Lacour-Gayet FG, Clarke DR, Gaynor JW, Spray TL, Kurosawa H, Stellin G, Ebels T, Bacha EA, Walters 3rd HL, Elliott MJ. Nomenclature and databases – the past, the present, and the future: a primer for the congenital heart surgeon. Pediatr Cardiol. 2007;28(2):105–15.
2. Jacobs ML, Jacobs JP, Franklin RC, Mavroudis C, Lacour-Gayet F, Tchervenkov CI, Walters H, Bacha EA, Clarke DR, William Gaynor J, Spray TL, Stellin G, Ebels T, Maruszewski B, Tobota Z, Kurosawa H, Elliott M. Databases for assessing the outcomes of the treatment of patients with congenital and paediatric cardiac disease – the perspective of cardiac surgery. Cardiol Young. 2008;18 Suppl 2:101–15.
3. Jacobs JP, Lacour-Gayet FG, Jacobs ML, Clarke DR, Tchervenkov CI, Gaynor JW, Spray TL, Maruszewski B, Stellin G, Gould J, Dokholyan RS, Peterson ED, Elliott MJ, Mavroudis C. Initial application in the STS congenital database of complexity adjustment to evaluate surgical case mix and results. Ann Thorac Surg. 2005;79:1635–49.
4. Jacobs ML, O'Brien SM, Jacobs JP, Mavroudis C, Lacour-Gayet F, Pasquali SK, Welke K, Pizarro C, Tsai F, Clarke DR. An empirically based tool for analyzing morbidity associated with operations for congenital heart disease. J Thorac Cardiovasc Surg. 2013;145(4):1046–57.
5. Fung CH, Lim YW, Mattke S, Damberg C, Shekelle PG. Systematic review: the evidence that publishing patient care performance data improves quality of care. Ann Intern Med. 2008;148:111–23.
6. Zinman D. State takes docs' list to heart. New York Newsday. December 18, 1991:A7.
7. Coronary artery bypass graft surgery in New York State 1989–1991. Albany: New York State Department of Health; 1992.
8. Hannan EL, Kilburn Jr H, Racz M, Shields E, Chassin MR. Improving the outcomes of coronary artery bypass surgery in New York State. JAMA. 1994;271:761–6.
9. Hannan EL, Siu AL, Kumar D, Kilburn Jr H, Chassin MR. The decline in coronary artery bypass graft surgery mortality in New York State: the role of surgeon volume. JAMA. 1995;273:209–13.
10. Topol EJ, Califf RM. Scorecard cardiovascular medicine. Its impact and future directions. Ann Intern Med. 1994;120:65–70.
11. Green J. Problems in the use of outcome statistics to compare health care providers. Brooklyn Law Rev. 1992;58:55–73.
12. Green J, Wintfeld N. Report cards on cardiac surgeons. Assessing New York State's approach. N Engl J Med. 1995;332:1229–32.
13. Omoigui N, Annan K, Brown K, Miller D, Cosgrove D, Loop F. Potential explanation for decreased CABG related mortality in New York State: outmigration to Ohio. Circulation. 1994;90:I-93 (abstract).
14. Grossi EA, Green J, Galloway AC, et al. Surgical evaluation of high risk cases in the era of statewide cardiac surgery data reporting. Presented at the 20th meeting of the Western Society for Thoracic Surgery, Olympic Valley, 22–25 June 1994 (abstract).
15. Byer MJ. Faint hearts. New York Times. March 21, 1992:A23.
16. Society of Thoracic Surgeons. Welcome to the STS National Database. Available at: http://www.sts.org/sections/stsnationaldatabase/. Accessed 15 May 2013.
17. O'Brien SM, Jacobs JP, Clarke DR, Maruszewski B, Jacobs ML, Walters 3rd HL, Tchervenkov CI, Welke KF, Tobota Z, Stellin G, Mavroudis C, Hamilton JR, Gaynor JW, Pozzi M, Lacour-Gayet FG. Accuracy of the Aristotle Basic Complexity Score for classifying the mortality and morbidity potential of congenital heart surgery operations. Ann Thorac Surg. 2007;84:2027–37.
18. Welke KF, Shen I, Ungerleider RM. Current assessment of mortality rates in congenital cardiac surgery. Ann Thorac Surg. 2006;82:164–71.
19. Shroyer AL, Coombs LP, Peterson ED, et al. The Society of Thoracic Surgeons: 30-day operative mortality and morbidity risk models. Ann Thorac Surg. 2003;75:1856–65.
20. Jacobs ML, Jacobs JP, Jenkins KJ, Gauvreau K, Clarke DR, Lacour-Gayet F. Stratification of complexity: the Risk Adjustment for Congenital Heart Surgery-1 method and the Aristotle Complexity Score–past, present, and future. Cardiol Young. 2008;18 suppl 2:163–8.
21. Society of Thoracic Surgeons. STS reporting online. Available at: http://www.sts.org/quality-research-patient-safety/sts-public-reporting-online. Accessed 15 May 2013.
22. Shahian DM, Edwards FH, Jacobs JP, Prager RL, Normand SL, Shewan CM, O'Brien SM, Peterson ED, Grover FL. Public reporting of cardiac surgery performance: part 1-history, rationale, consequences. Ann Thorac Surg. 2011;92(3 Suppl):S2–11.
23. Shahian DM, Edwards FH, Jacobs JP, Prager RL, Normand SL, Shewan CM, O'Brien SM, Peterson ED, Grover FL. Public reporting of cardiac surgery performance: part 2-implementation. Ann Thorac Surg. 2011;92(3 Suppl):S12–23.
24. Society of Thoracic Surgeons. Potential unintended consequences. Available at: http://www.sts.org/

quality-research-patient-safety/sts-public-reporting-online/potential-unintended-consequences-public. Accessed 15 May 2013.
25. The Denver Post Editorial Board. At the heart of the problem. Denver Post. March 2, 2001.
26. Sherry A. Children's Hospital cardiology chief told to resign. Denver Post. March 1, 2001.
27. Jacobs JP, Cerfolio RJ, Sade RM. The ethics of transparency: publication of cardiothoracic surgical outcomes in the lay press. Ann Thorac Surg. 2009;87:679–86.
28. Cronk CE, Malloy ME, Pelech AN, et al. Completeness of state administrative databases for surveillance of congenital heart disease. Birth Defects Res A Clin Mol Teratol. 2003;67:597–603.
29. Frohnert BK, Lussky RC, Alms MA, Mendelsohn NJ, Symonik DM, Falken MC. Validity of hospital discharge data for identifying infants with cardiac defects. J Perinatol. 2005;25(11):737–42.
30. Strickland MJ, Riehle-Colarusso TJ, Jacobs JP, Reller MD, Mahle WT, Botto LD, Tolbert PE, Jacobs ML, Lacour-Gayet FG, Tchervenkov CI, Mavroudis C, Correa A. The importance of nomenclature for congenital cardiac disease: implications for research and evaluation. In: Jacobs JP, editor. 2008 Cardiology in the Young Supplement: Databases and The Assessment of Complications associated with The Treatment of Patients with Congenital Cardiac Disease. Prepared by: The Multi-Societal Database Committee for Pediatric and Congenital Heart Disease. Cardiology in the Young, vol. 18, issue S2 (Suppl 2), p. 92–100, December 9, 2008.
31. Pasquali SK, Peterson ED, Jacobs JP, He X, Li JS, Jacobs ML, Gaynor JW, Hirsch JC, Shah SS, Mayer JE. Differential case ascertainment in clinical registry versus administrative data and impact on outcomes assessment for pediatric cardiac operations. Ann Thorac Surg. 2013;95(1):197–203.

Part IV

Stratification of Complexity

Statistical Issues in the Analysis and Interpretation of Outcomes for Congenital Cardiac Surgery

24

Sean M. O'Brien

Abstract

The analysis and reporting of patient outcomes plays a major role in congenital cardiac surgery quality improvement and accountability initiatives. Although outcomes analysis can lead to important insights, factors such as wide patient heterogeneity and small sample sizes make analysis and interpretation challenging. This chapter explores issues, methods, and general principles for comparing cardiac surgery outcomes across providers.

Keywords

Case-mix adjustment • Stratification • Standardization • Confounding • Provider performance evaluation

Introduction

The analysis and reporting of health care outcomes is the centerpiece of congenital cardiac surgery quality improvement and accountability initiatives. Although outcomes analysis can generate useful information, it is inherently challenging to compare outcomes across providers because factors other than performance can impact these results. Widely differing case mix and small sample sizes can lead to perturbations which obscure a provider's true performance. This chapter explores these challenges and describes a variety of analytic approaches that can be used to address them.

In this chapter, the term "provider" is used generically to refer to the unit that is the focus of performance evaluation, for example, a hospital, surgeon, insurance plan, or any other type of healthcare unit. We focus on mortality as a recurring example, but the principles discussed apply generally to a variety of types of endpoints.

Risk Adjustment

In clinical comparative effectiveness studies, researchers often ask whether one treatment is better than another for reducing morbidity and improving survival. For example, in a randomized

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controlled trial comparing the success rate of two treatments, say "A" and "B", the average "effect" of treatment A versus B can be estimated by the difference in the proportion of patients receiving treatment A who have a successful outcome and the proportion of patients receiving treatment B who have a successful outcome, $\widehat{S}_A - \widehat{S}_B$. Randomization ensures that patients receiving the two treatments are comparable. When treatments are not randomly assigned – as in a non-randomized observational study – the observed difference $\widehat{S}_A - \widehat{S}_B$ may be biased. To the extent that patients in each treatment group differ in ways that affect outcomes (e.g., they are sicker, frailer, etc.), the observed differences in outcomes may reflect different patient characteristics rather than the treatment effect of interest. An identical confounding issue arises in studies comparing outcomes of health care providers in which patients are not randomized to providers.

Risk adjustment is a collection of techniques for reducing the effect of confounding factors in studies where patients are not randomly assigned to different treatments. In provider performance evaluation, the 'treatments' are different providers. Risk adjustment aims to control for patient factors outside the provider control so that residual differences in outcomes could, potentially, reflect true differences in quality [1].

The literature from statistics and related disciplines describes conditions in which valid inferences about treatment effects based on observational data are possible. In general, valid estimation requires the assumption that outcome differences are unconfounded conditional on a set of pre-treatment covariates [2]. This assumption means that, within blocks of patients having identical values of pre-treatment covariates, patients receiving each treatment are like a random sample from a common population. Although the unconfoundedness assumption is unlikely to be literally true in a non-randomized observational study, the risk of encountering large violations of the assumption can be minimized by careful planning to ensure collection of a wide range of suspected confounding variables.

Considerations for Variable Selection for Risk Adjustment

Risk adjustment involves an attempt to compare only patients who are similar with respect to pre-treatment covariates. In general, covariates appropriate for risk adjustment are those factors that are hypothesized to remain the same if the patient were to be re-assigned to a different provider [3]. Although it is generally desirable to adjust for all important confounding factors, theory dictates that we should not adjust for factors that are part of the treatment being evaluated [4]. Doing so may "adjust away" differences in outcomes that result from the adoption of more or less effective care practices by different providers. For example, one would generally not adjust for hand washing when comparing infection rates across hospitals because assiduous hand washing is one of the ways in which a hospital may seek to achieve a lower infection rate [3]. In some cases, the distinction between a patient factor and treatment factor may not be obvious. For example, adjusting for a neonate's age in days at the time of operation may be problematic if the provider being evaluated impacted the decision of how many days to wait after birth before operating.

Unadjusted Outcomes as Weighted Averages

Before considering methods of adjusting provider-specific outcomes, it is instructive to consider some properties of outcomes that are unadjusted. Calculations are illustrated using hypothetical data for hospitals performing cardiac surgery in neonates, categorized by birthweight, as shown in Table 24.1. When a rate is unadjusted, its value is implicitly a weighted average of the rates observed for different subgroups [5]. The weight of each subgroup is equal to the proportion of patients falling in that subgroup. For example, Hospital A had 70 deaths in a cohort of 1,000 patients for an overall unadjusted mortality rate of 70/1,000 = 0.07. This same unadjusted mortality rate can be calculated

Table 24.1 Mortality rates for cardiac operations in neonates at two hypothetical hospitals and a national registry

			Deaths		
Subgroup	Number of patients	Proportion of patients	Observed number	Observed proportion	Expected proportion[a]
Hospital A					
Overall	1,000	1.00	70	0.07	0.055
Birthweight <2.5 kg	100	0.10	16	0.16	
Birthweight ≥2.5 kg	900	0.90	54	0.06	
Hospital B					
Overall	1,000	1.00	90	0.09	0.070
Birthweight <2.5 kg	400	0.40	60	0.15	
Birthweight ≥2.5 kg	600	0.60	30	0.05	
Registry					
Overall	10,000	1.00	625	0.0625	
Birthweight <2.5 kg	2,500	0.25	250	0.10	
Birthweight ≥2.5 kg	7,500	0.75	375	0.05	

[a]Used for indirect standardization. See text for details

as $0.10 \times 0.16 + 0.90 \times 0.06 = 0.07$, which is a weighted average of Hospital A's mortality rate in patients with birthweight <2.5 kg (prevalence = 0.10; mortality = 0.16) and its mortality rate in patients with birthweight ≥2.5 kg (prevalence = 0.90; mortality = 0.06). Similarly, Hospital B had 90 deaths in a cohort of 1,000 patients for an overall unadjusted mortality rate of 90/1,000 = 0.09. This same unadjusted mortality rate can be calculated as $0.40 \times 0.15 + 0.60 \times 0.05 = 0.09$, which is a weighted average of Hospital B's mortality rate in patients with birthweight <2.5 kg (prevalence = 0.40; mortality = 0.15) and its mortality rate in patients with birthweight ≥2.5 kg (prevalence = 0.60; mortality = 0.05). As discussed below, many risk adjustment procedures also involve calculating weighted averages of subgroup-specific rates. However, the subgroup weights are artificially manipulated to ensure they are the same for both groups being compared.

Stratification

Stratification is a method for reducing the effect of confounding factors in studies where patients are not randomly assigned to different treatments [4]. In a stratified analysis, patients are divided into subgroups on the basis of one or more pre-treatment covariates in order to examine outcome differences by treatment group separately within each stratum. In general, if treatment comparisons are always performed on patients from the same stratum, then outcome differences cannot be confounded by differences in the number of patients falling in each stratum across the groups being compared.

For example, Table 24.1 presents a stratified analysis in which strata are defined by categories of birthweight. Hospital A had higher mortality than Hospital B both among patients with birthweight <2.5 kg (0.16 versus 0.15) as well as among patients with birthweight ≥2.5 kg (0.06 versus 0.05). Yet, Hospital A had a lower overall unadjusted mortality rate than Hospital B when the birthweight groups were combined. This paradoxical result occurred because Hospital B performed four times as many operations in patients with birthweight <2.5 kg (a group with relatively higher mortality) compared to Hospital A. Performing a stratified analysis by birthweight category removed the effect of different proportions of patients weighing <2.5 kg and revealed the superior outcomes of Hospital B within each birthweight subgroup.

In addition to controlling for confounding, stratification can be used to assess whether a

provider's performance differs across the various strata. For example, a provider may have acceptable outcomes in relatively low-risk patients but unacceptable outcomes in relatively high-risk patients, or vice versa. Such differences may not be apparent if outcomes are compared overall without performing a stratified analysis.

Stratification is simplest to explain with a single covariate but can also be used with two or more covariates. In that case, a "stratum" is defined for each unique combination of all possible values of each covariate. For example, with two binary (yes/no) covariates, the strata would be defined as (no, no), (no, yes), (yes, no), and (yes, yes). However, the number of strata increases quickly as the number of covariates increases. For example, controlling for ten variables with two levels each would result in $2^{10} = 1,024$ possible strata. As discussed below, statistical modeling may be more reliable than stratification when the number of strata is large and the number of patients in each stratum is small.

Direct Standardization

Reporting outcomes for each stratum separately can reduce confounding but may be unwieldy when the number of strata is large. In many situations, such as pay-for-performance, it is useful to have an overall summary of a provider's risk-adjusted outcomes. A stratum-adjusted standardized rate can be used for this purpose. Like an unadjusted rate, the standardized rate is a weighted average of a provider's stratum-specific rates. However, the weights are artificially manipulated to reflect the case mix of a "standard" population. The standard case mix can be defined in many ways; a common approach is to pool data across several providers and use the totals in each stratum in the pooled sample. Weighting in this manner answers the question: What would the provider's outcomes be if all of its stratum-specific rates remained the same but the proportion of patients in each stratum was altered to reflect the standard case-mix?

To illustrate direct standardization, we define a "standard" case mix using data from the row labeled "registry" in Table 24.1. In this hypothetical registry population, 25% of patients have birthweight <2.5 kg and 75% have birthweight ≥2.5 kg. Thus, Hospital A's directly standardized mortality rate is equal to $0.25 \times 0.16 + 0.75 \times 0.06 = 0.085$, which is a weighted average of Hospital A's mortality rate in patients with birthweight <2.5 kg (weight = 0.25, mortality = 0.16) and its mortality rate in patients with birthweight ≥2.5 kg (weight = 0.75, mortality = 0.06). Similarly, Hospital B's standardized mortality rate is $0.25 \times 0.15 + 0.75 \times 0.05 = 0.075$. As expected, direct standardization removes the effect of birthweight categories and reflects the lower mortality of Hospital B.

Direct standardization is simple and requires no modeling assumptions, but there are important limitations. First, it may not be possible to calculate a standardized rate for a provider if it has no cases in one or more strata. Even if a standardized rate is able to be calculated, the resulting estimate may be highly noisy if certain strata are sparsely populated. Like any summary measure, there is a potential for information loss when only a single summary measure is reported. For example, a hospital performing congenital cardiac surgery might have acceptable outcomes for relatively low-risk simple operations but unacceptable outcomes for relatively high-risk or complex operations. From the patient's perspective, a patient would likely prefer the hospital with the best outcomes for that patient's particular condition. This information is lost when only a single summary is reported. Finally, if a provider's performance does vary substantially across patient subgroups, then the standardized rate may be sensitive to the arbitrary choice of standard population.

Indirect Standardization

When stratum-specific numbers are small, as is often the case in congenital cardiac surgery, stratum-specific estimates may be highly noisy rendering the standardized outcome rate unreliable. An alternative approach, known as indirect standardization, addresses this issue. It

involves calculating an "expected rate" for each provider by applying stratum-specific rates from a standard population to each provider's own case mix. A provider's observed and expected rates are then compared to one another. The difference between direct and indirect standardization can be summarized as follows. Direct standardization involves re-weighting each provider's own stratum-specific rates to reflect the case mix of a standard population. In contrast, indirect standardization re-weights the stratum-specific rates of a standard population to reflect each provider's own case mix.

The ratio of a provider's observed and expected rates is called the "O/E ratio." If a provider's O/E ratio for mortality (also called "standardized mortality ratio") is significantly greater than 1, this implies that its overall mortality rate is higher than would be expected if its stratum-specific mortality rates were the same as the standard population (i.e. worse than expected mortality). If the O/E ratio is significantly less than 1, this implies that its overall mortality rate is lower than would be expected if its stratum-specific mortality rates were the same as the standard population (i.e. better than expected mortality).

Using hypothetical data from Table 24.1, Hospital A's expected mortality rate may be calculated as $0.10 \times 0.10 + 0.90 \times 0.05 = 0.055$ which is a weighted average of stratum-specific mortality rates from the standard population using a weight of 0.10 for patients with birthweight <2.5 kg and a weight of 0.90 for patients with birthweight ≥2.5 kg. Since Hospital A's actual observed mortality rate is 0.07, its O/E ratio is $0.07/0.055 = 1.27$. Thus, Hospital A's mortality rate is about 27% higher than expected compared to the standard population. Hospital B's expected mortality rate is $0.40 \times 0.10 + 0.60 \times 0.05 = 0.07$ and its observed mortality is 0.09, and so its O/E ratio is $0.09/0.07 = 1.29$. Thus, Hospital B's mortality rate is about 29% higher than expected compared to the standard population.

For presentation purposes, the O/E ratio is sometimes converted into an adjusted rate (or standardized rate), by using the formula, adjusted rate = (O/E ratio) × (overall rate of standard population). When calculated using this formula, an adjusted rate describes what a provider's outcome rate would be if the provider's performance in treating its own case mix (as reflected by the O/E ratio) was extrapolated to the overall case mix of the reference population. Using Table 24.1 data, the overall mortality rate in the standard population is 0.0625. So, continuing the calculations described above, Hospital A's adjusted mortality rate (AMR) is $(0.07/0.055) \times 0.0625 = 0.0795 \approx 0.08$ and Hospital B's AMR is $(0.09/0.07) \times 0.0625 = 0.0804 \approx 0.08$.

As noted above, indirect standardization permits adjusting for case mix even when a provider's stratum-specific sample sizes are small. In addition, because each provider is evaluated with respect to the provider's own case mix, they are evaluated based on the types of cases they perform most often, instead of a potentially irrelevant standard case mix. Despite these advantages, there are some subtle issues and caveats. Indirect standardization permits a fair comparison between each provider and the standard population, but does not necessarily permit a fair comparison between two individual providers. For example, Hospital A's mortality O/E ratio is slightly lower than Hospital B's (1.27 versus 1.29) suggesting that Hospital A has lower mortality. Yet, Hospital B has lower observed mortality rates than Hospital A in each stratum. This counterintuitive result may be explained as follows. Both hospitals have excess mortality relative to the standard population, and their excess mortality is greatest in patients with birthweight <2.5 kg. However, this group is weighted 40% in the calculation of Hospital B's mortality rate but only 10% in the calculation of Hospital A's mortality rate. The comparison between Hospitals A and B is not an "apples-to-apples" comparison because their stratum-specific results are weighted differently. The comparison between each hospital and the standard population does not have this issue because the standard population is always re-weighted to use the same weights as the hospital.

Model-Based Standardization

The simple approach to standardization described above requires an adequate number of cases in each stratum in order for the observed

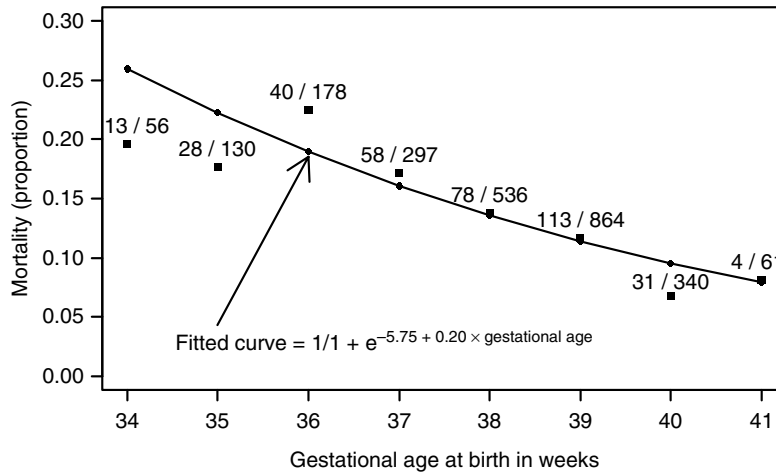

Fig. 24.1 Mortality rates stratified by gestational age at birth for patients in a hypothetical registry

stratum-specific outcome rates to be reliable. As a result, it tends to break down when the number of strata is large. For example, it cannot be used with a continuous covariate, unless it is first coarsened or categorized, because each observed value of the continuous covariate may have just a single patient. Similarly, it tends to breaks down with a large number of covariates, because the number of number of strata will be very large. Model-based standardization provides an alternative to the approach described above when strata are sparsely populated and the number of strata is large.

Model-based standardization is an extension of conventional standardization and is based on the same concepts described above. The main difference is the use of a statistical model rather than raw data to obtain stratum-specific outcome rates. Statistical modeling accommodates sparsely populated strata by making assumptions about the relationship between outcome rates in different strata. For example, if strata are defined by a continuous covariate x, such as a patient's age, we might model the stratum-specific outcome rates by assuming they increase or decrease linearly in proportion to x. Similarly, if strata are defined by the combination of k numerical covariates $x_1, x_2, \ldots, x_k$, we might assume that stratum-specific outcome rates vary as a simple additive function $f_1(x_1)+f_2(x_2)+\cdots+f_k(x_k)$ or multiplicative function $f_1(x_1)\times f_2(x_2)\times\cdots\times f_k(x_k)$. Alternatively, for binary outcomes such as mortality, stratum-specific outcome rates may be assumed to follow a multivariable logistic regression model. The logistic model equation assumes that risk $= 1/[1+\exp(-[\beta_0+\beta_1x_1+\beta_2x_2+\cdots+\beta_kx_k])]$ where $\beta_0, \beta_1, \ldots, \beta_k$ denote unknown regression coefficients to be estimated from the data.

To illustrate model-based indirect standardization, Fig. 24.1 displays mortality rates stratified by gestational age in weeks at birth for patients in a hypothetical registry of patients undergoing the Norwood Stage 1 operation. Square dots display the observed raw mortality rate (number of deaths divided by number of patients) for each gestational age. Circular dots display the estimated mortality rate for each gestational age obtained by fitting a logistic regression model to the data. The model assumes that stratum-specific mortality rates are described by a logistic regression equation of the form $1/[1+\exp(-\alpha-\beta\times\text{gestational age})]$ where α and β denote unknown regression coefficients to be estimated from the data. Since this assumption is not likely to be 100% literally correct, the estimates derived from a logistic regression model may be biased. However, if the model is approximately correct, then predictions derived from the logistic model are likely more accurate than raw stratum-specific mortality rates because the raw rates are noisy.

Let $\bar{y}_x$ denote the observed raw mortality rate among patients born at x weeks of gestational age (square dots in Fig. 24.1) and let $\hat{\pi}_x$ denote the

corresponding predicted rate according to the logistic model (circular dots in Fig. 24.1). For a particular provider, if f_x denotes the proportion of the provider's patients who were gestational age x weeks at birth, then the provider's expected mortality rate, treating the registry as the standard population, may be calculated either as $f_{34}\bar{y}_{34} + f_{35}\bar{y}_{35} + \ldots + f_{41}\bar{y}_{41}$ (conventional standardization using observed raw stratum-specific mortality rates from the registry population) or as $f_{34}\hat{\pi}_{34} + f_{35}\hat{\pi}_{35} + \ldots + f_{41}\hat{\pi}_{41}$ (model-based standardization using logistic regression to approximate the registry population outcomes). With several covariates, the form of the regression model is more complicated, but the concepts are identical.

The Problem of Small Sample Sizes

It is widely recognized that outcomes such as mortality can have limited precision when the sample size is small. Yet, even with moderate sample sizes, outcome rates can be less reliable than is commonly realized. To illustrate the potential for noisy data, we performed the following simulation. Consider comparing the mortality rate for the Norwood Stage 1 operation for two hospitals with identical sample sizes and identical case mix (thus not requiring risk adjustment). Owing to differences in quality, suppose the true (long-run) Norwood mortality rate is 15.5% at "Hospital Good Care" and 19.3% at "Hospital Average Care" These values were chosen to approximate the estimated 25th percentile and overall average mortality rate for the Norwood Operation among hospitals participating in the Society of Thoracic Surgeons (STS) Congenital Heart Surgery Database, as reported by Jacobs et al. [6]. In a small sample of patients, the observed mortality rate at each hospital will fluctuate according to the laws of probability and would not be expected to match the underlying true long-run rate. For example, if each hospital performed 10 Norwood operations, Hospital Average Results would have up to a 53% probability of having either the same or fewer deaths than Hospital Good Care—worse than a coin toss

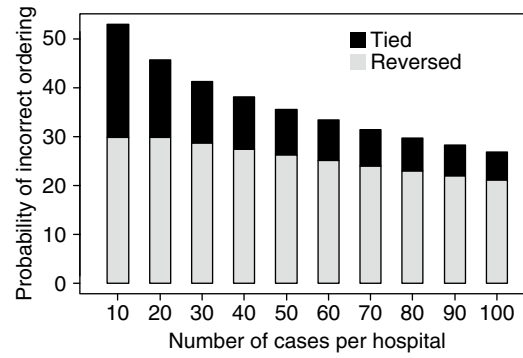

Fig. 24.2 Probability of incorrect hospital mortality rankings as a function of case volumes. Reversed indicates the probability that a hospital with a true mortality rate of 19.3% will have fewer deaths than a hospital with the same number of patients and a true mortality rate of 15.5%. "Tied" indicates the probability that the 2 hospitals will have the same number of deaths. The probability calculation is based on the binomial distribution

(Fig. 24.2). Even if these hospitals each performed 100 Norwood operations, the probability of misclassification would remain at 27%. For context, Hornik et al. reported that 64% of hospitals in the STS Congenital Heart Surgery Database performed 10 or fewer Norwood operations per year and 89% performed 20 or fewer Norwood operations per year [7].

Several approaches have been proposed for addressing the issue of small sample sizes when analyzing and comparing provider outcomes. First, one can simply exclude providers with small denominators when reporting provider-specific results. When this approach is applied to congenital cardiac surgery, a large proportion of providers may be excluded. Second, one may pool data across multiple different types of operations in order create datasets with larger provider-specific denominators. Although pooling data across operations makes the estimates less noisy, there is some risk of information loss, because a provider could perform well for some types of operations and poorly for others. Third, data can be aggregated over multiple calendar years to obtain a sufficient sample size. This makes estimates less noisy but the data are less timely. For example, older data may reflect staffing issues and care practices that are no longer relevant by the time the data are reported.

A fourth strategy is to analyze and report composite endpoints. Such composite measures are often more reliable than single endpoints because combined endpoints occur more frequently. Despite their advantages, composite endpoints may be controversial when items in the composite contribute different amounts of statistical information or when the items differ in their relative importance to patients [8, 9].

Another alternative approach to increasing reliability is to estimate outcome measures using hierarchical models [10, 11]. Such models allow "borrowing of information" across providers when estimating the outcome rate of each individual provider, in order to produce an estimate that is less noisy than the provider's raw outcome rate. The hierarchical estimator incorporates a Bayesian concept that, in the absence of data, the best estimate of a provider's outcome rate is the average outcome rate of all providers. As more data become available, the estimate shifts away from the average in the direction indicated by the provider's data. Heuristically, the hierarchical estimate is a weighted average of a participant's actual observed outcomes and the overall average outcomes of all participants. The model weights an individual participant's own data more heavily when the denominator is large enough to be reliable and weights the overall average rate more heavily when the denominator is too small to support a reliable estimate.

Extensions of hierarchical modeling allow incorporating provider-level covariates, such as surgical volume, to enhance prediction of a provider's true outcome rate. Including provider-level covariates allows "borrowing of information" from providers with similar provider-level covariates. For example, estimation of outcome rates for an individual low-volume provider will be informed by outcomes observed among other low-volume providers. Examples of other auxiliary information that can be exploited include the provider's outcome rates for procedures other than the one being evaluated and outcome rates for the same procedure in an earlier time period.

Although hierarchical modeling is a powerful tool for estimating performance, it is not a cure for small sample sizes. Hierarchical modeling can arguably produce the best statistical estimate in face of uncertainty but cannot remove the underlying uncertainty.

Prediction Intervals

In order to encourage an appropriately cautious interpretation of noisy data, provider-specific performance results are typically presented with an indication of their likely statistical precision. One popular graphical display, known as a funnel plot [12], uses "prediction intervals" for this purpose. It displays the range of outcome rates that would be expected to occur due to chance if each provider's underlying true outcome rate was the same. The prediction interval is constructed to ensure that each provider's observed outcome rate will fall within the interval with a specified probability.

Figure 24.3 illustrates the use of a funnel plot for displaying hospital-specific unadjusted mortality rates using hypothetical data for a high-risk congenital cardiac operation. In this figure, each hospital's unadjusted mortality rate is plotted against the denominator that was used for calculating the unadjusted mortality rate. Following Spiegelhalter [12], lines depicting 95% (≈2 standard deviations) and 99.8% (≈3 standard

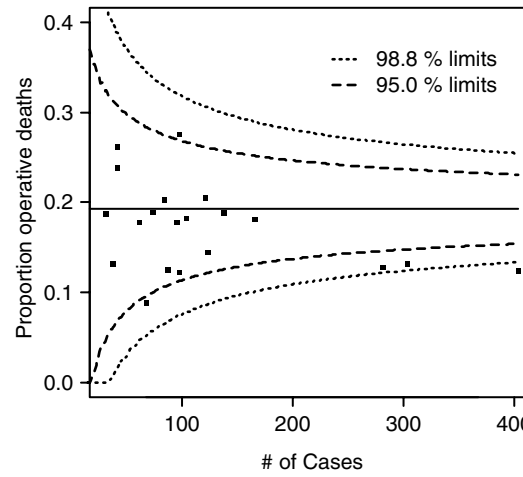

Fig. 24.3 Funnel plot of hospital-specific unadjusted mortality rates for a hypothetical congenital cardiac operation

deviations) prediction limits around the overall mortality rate are overlaid to make a funnel plot. Based on Fig. 24.3, one hospital has a mortality rate exceeding the upper limit of the 95% prediction interval, four hospitals have mortality rates falling below the lower limit of the 95% prediction interval, and one has a mortality rate following below the lower limit of the 99.8% interval. For the remaining 15 hospitals, the hospital's unadjusted mortality rate falls within the usual range of normal sampling variation. While the plot suggests some evidence of between-hospital variation in unadjusted mortality, it reveals that much of the between-hospital variation may be attributed to chance, and that precise rankings may not be possible for the majority of hospitals.

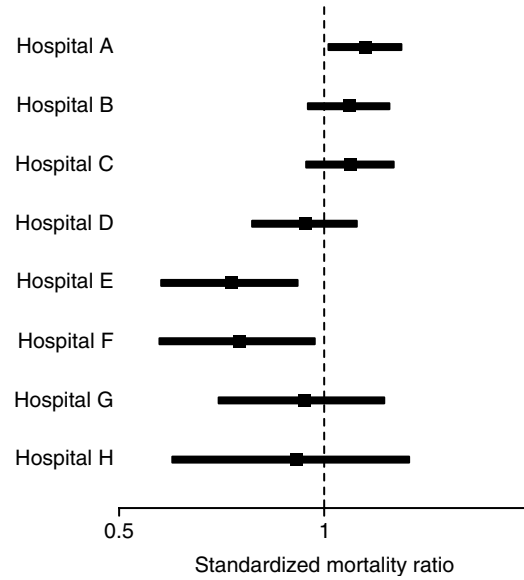

Fig. 24.4 Hospital-specific standardized mortality ratios for a hypothetical congenital cardiac operation

Interval Estimates

As noted above, a prediction interval provides the range of provider-specific outcomes that would be expected to occur by chance alone if the true underlying outcome rates did not vary. If the underlying rates do vary, then interest may focus on estimating the magnitude of true signal variation across providers and estimating true underlying outcome rates for each individual provider. Such quantities are always estimated with error, and so it is important to convey some measure of the estimate's statistical precision.

An interval estimate is a range of numbers that is expected to include the true value of the quantity being estimated. Naturally, one can be relatively confident that the true value lies in an extremely wide interval, and less confident that it lies in a narrow interval.

One type of interval estimate, known as a confidence interval, is constructed so that the interval will contain the true value with a specified probability. For example, a 95% confidence interval has the property that it will include the true value of the quantity being estimated about 95% of the time in repeated sampling. Similarly, a 99% confidence interval will include the true value about 99% of the time in repeated sampling. The width of a confidence interval depends on the desired coverage probability (e.g. 95 or 99%) as well as the sample size. If the confidence interval is very wide, this indicates that the provider's outcome rate is an imprecise estimate of the provider's true underlying outcome rate. Wide confidence intervals occur when the number of patients in the denominator is small. Naturally, a 99% confidence interval is wider than a 95% confidence interval, and the width decreases as the sample size increases.

A common graphical display in provider performance evaluation involves plotting each provider's interval estimate against a reference line representing "average" performance. For example, Fig. 24.4 illustrates this format with standardized mortality ratios (equivalent to mortality O/E ratios) compared against the null value of 1.0. Because the observed mortality rates are inherently variable, differences among hospitals' observed mortality rates should be interpreted with caution. If a hospital's 95% confidence interval for the O/E includes the reference line, then its mortality is not statistically different from expected. If a hospital's 95% confidence interval for the O/E falls entirely to the right or left of the reference line, then there is strong evidence that the hospital's mortality rate differs from the expected rate. However, it should be remembered that approximately 5% of hospitals

would be expected to have confidence intervals excluding 1.0 due to chance alone even if each hospital's true O/E was 1.0. A format such as Fig. 24.4 not only allows identifying statistically significant differences compared to the null value of 1.0, but also conveys the magnitude of the differences and a range of plausible estimates for each provider's true performance.

For simplicity, some report cards suppress the detailed information in Fig. 24.4 and report only whether each provider's performance was statistically better, worse, or same as average. Categorizing results in this manner has been criticized for being dependent on a provider's sample size and for placing too much emphasis on statistical rather than clinical significance [10].

The notion of a confidence interval, as an interval containing the true value with a specified probability, was developed within the conventional "frequentist" statistical paradigm [13]. Critics note that such intervals lack a directly useful probability interpretation. For example, if a hospital's 95% confidence interval for the O/E falls extends from 1.1 to 1.5, one cannot say there is a 95% probability that the hospital's true O/E is in the interval 1.1–1.5. One can only say that 95% of confidence intervals calculated in a similar manner would include the true O/E ratio if repeated in a series of hypothetical experiments or random samples. From a frequentist perspective, the hospital's true O/E is either in the interval 1.1–1.5 or not; there is no probability involved. An alternative statistical paradigm known as Bayesian inference can be used to construct interval estimates with a more natural interpretation, as described below.

Bayesian Inference

Although frequentist inference is the dominant paradigm, there is widespread interest in using Bayesian analysis for the evaluation of provider outcomes [10, 14–16]. In part, its appeal stems from the fact that what consumers of report cards often want to know is the likelihood (probability) that a provider's outcomes are better or worse than some benchmark, or the likelihood (probability) that the provider's outcomes differ from the benchmark by some clinically important amount. Such probability assessments are not obtainable from a conventional frequentist analysis but are available when inference is based on the Bayesian paradigm.

Unlike conventional frequentist statistics, Bayesian methodology uses the language of probability to express beliefs about unknown quantities before and after observing the data. For example, prior to observing the data, we might assign 50% probability to the hypothesis that Hospital A has a lower true underlying mortality rate than Hospital B (i.e. a coin toss). After observing the data, that probability might shift away from 50% to reflect some degree of belief about which of the two hospitals has lower mortality. Bayes' theorem provides a mathematical formula for updating prior probabilities in light of the observed data.

In order to conduct a Bayesian analysis, the analyst must specify a prior probability distribution representing the analyst's prior beliefs about the collection of all unknown quantities. In most cases, the prior distribution is chosen to reflect the absence of strong prior beliefs i.e., a "non-informative" prior. The prior distribution is then updated with the observed data using Bayes' theorem to obtain the posterior distribution. Except for special cases, the posterior distribution cannot be calculated analytically, and Markov Chain Monte Carlo (MCMC) methods are used.

Advantages of fully Bayesian estimation include the ability to perform inference about complex functions of model parameters and the ability to express the results of statistical analyses in terms of probabilities [17, 18]. Unlike frequentist confidence intervals, Bayesian probability intervals have an intuitively direct interpretation as an interval containing the true value with a specified probability [13]. In addition, Bayesian estimates are often more computationally feasible than conventional frequentist approaches for complex models or models with non-standard probability distributions.

Disadvantages of the Bayesian approach include its relative complexity and the potential for

results to be influenced by the choice of prior distribution. In addition, computation for a Bayesian analysis is technically demanding and requires the analyst to have some level of familiarity with the underlying computational methods.

Summary

The evaluation of provider performance is an integral part of efforts to improve quality, yet such assessments need to be interpreted cautiously. Case-mix and sampling variation can both have a large impact on outcomes, and should both be considered as possible explanations whenever outcomes differ between providers. An understanding of statistical tools and their limitations will help users to correctly interpret measures of performance and avoid pitfalls when making comparisons between providers.

References

1. Kuhlthau K, Ferris TG, Iezzoni LI. Risk adjustment for pediatric quality indicators. Pediatrics. 2004; 113(1 Pt 2):210–6.
2. Little RJ, Rubin DB. Causal effects in clinical and epidemiological studies via potential outcomes: concepts and analytical approaches. Annu Rev Public Health. 2000;21(1):121–45.
3. Zaslavsky AM. Statistical issues in reporting quality data: small samples and casemix variation. Int J Qual Health Care. 2001;13(6):481–8.
4. Rothman KJ, Greenland S, Lash TL. Modern epidemiology. Philadelphia: Lippincott Williams & Wilkins; 2008.
5. Schoenbach VJ, Rosamond WD. Understanding the fundamentals of epidemiology. An evolving text. Chapel Hill: University of North Carolina; 2000.
6. Jacobs JP, O'Brien SM, Pasquali SK, Jacobs ML, Lacour-Gayet FG, Tchervenkov CI, et al. Variation in outcomes for benchmark operations: an analysis of the Society of Thoracic Surgeons Congenital Heart Surgery Database. Ann Thorac Surg. 2011;92(6):2184–92.
7. Hornik CP, He X, Jacobs JP, Li JS, Jaquiss RD, Jacobs ML, et al. Relative impact of surgeon and center volume on early mortality after the Norwood operation. Ann Thorac Surg. 2012;93(6):1992–7.
8. Peterson ED, DeLong ER, Masoudi FA, O'Brien SM, Peterson PN, Rumsfeld JS, et al. ACCF/AHA 2010 Position Statement on Composite Measures for Healthcare Performance Assessment American College of Cardiology Foundation/American Heart Association Task Force on Performance Measures (Writing Committee to Develop a Position Statement on Composite Measures). J Am Coll Cardiol. 2010; 55(16):1755–66.
9. O'Brien SM, DeLong ER, Dokholyan RS, Edwards FH, Peterson ED. Exploring the behavior of hospital composite performance measures an example from coronary artery bypass surgery. Circulation. 2007; 116(25):2969–75.
10. Christiansen CL, Morris CN. Improving the statistical approach to health care provider profiling. Ann Intern Med. 1997;127(8_Part_2):764–8.
11. Krumholz HM, Brindis RG, Brush JE, Cohen DJ, Epstein AJ, Furie K, et al. Standards for statistical models used for public reporting of health outcomes: an American Heart Association Scientific Statement From the Quality of Care and Outcomes Research Interdisciplinary Writing Group: cosponsored by the Council on Epidemiology and Prevention and the Stroke Council Endorsed by the American College of Cardiology Foundation. Circulation. 2006;113(3):456–62.
12. Spiegelhalter DJ. Funnel plots for comparing institutional performance. Stat Med. 2005;24(8): 1185–202.
13. Jaynes ET. Confidence intervals vs Bayesian intervals. Foundations of probability theory, statistical inference, and statistical theories of science. W.L. Harper and C.A. Hooker (eds), D. Reidel, Dodrecht; 1976. p. 175–257.
14. Austin PC. A comparison of Bayesian methods for profiling hospital performance. Med Decis Making. 2002;22(2):163–72.
15. Normand S-LT, Glickman ME, Gatsonis CA. Statistical methods for profiling providers of medical care: issues and applications. J Am Stat Assoc. 1997; 92(439):803–14.
16. O'Brien SM, Shahian DM, DeLong ER, Normand S-LT, Edwards FH, Ferraris VA, et al. Quality measurement in adult cardiac surgery: part 2—statistical considerations in composite measure scoring and provider rating. Ann Thorac Surg. 2007;83(4):S13–26.
17. Congdon P. Bayesian statistical modelling. Chichester: John Wiley & Sons; 2006.
18. Gelman A, Carlin JB, Stern HS, Dunson DB, Vehtari A, Rubin DB. Bayesian data analysis. Boca Raton: CRC Press; 2013.

Real Time Monitoring of Risk-Adjusted Surgical Outcomes for Congenital Heart Disease

25

Kate L. Brown, Sonya Crowe, Martin Utley, and Christina Pagel

Abstract

Real time monitoring of risk-adjusted outcomes for adult cardiac surgery has been reported in the past and found to aid quality assurance efforts. Recent advances in terms of the available risk-adjustment methods for congenital heart disease and paediatric cardiac surgery, which go some way to reflecting the great complexity of this field, have opened up the opportunity to use Variable Life Adjusted Display (VLAD) charts for this context. This chapter explains what VLAD charts are, and how they differ to other retrospective methods of audit that may be familiar to readers. The chapter then provides an example of VLAD chart use for real time monitoring of risk-adjusted surgical outcomes in congenital heart disease from the United Kingdom thus illustrating how these charts may be used for quality assurance. The chapter then places this example into context covering some of the salient issues and previous experiences with VLAD for outcome monitoring.

Keywords

Outcome • Quality assurance • Variable Life Adjusted Display (VLAD) chart • Real time monitoring • Risk-adjustment

The Cumulative Sum Chart (CUSUM Chart)

The most common way to examine mortality data, risk-adjusted or not, is to convert it into a graphic or a plot that can also help to identify changes over time. Probably the most familiar chart is the Cumulative Sum Chart (CUSUM Chart). The CUSUM is constructed by plotting the cumulative number of deaths on the vertical axis against the total number of procedures on the horizontal axis. With each death the graph rises,

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while the occurrence of no deaths results in a horizontal line (Fig. 25.1) [1]. If a model exists for estimating the risk for each case (even a crude model based on average observed mortality rate), statistical limits can be calculated to give upper and lower boundaries on how many deaths would be expected (dotted line in Fig. 25.1). Often, 'alerts' are set if the observed CUSUM line is outside either boundary as a trigger for further investigation (Fig. 25.1) [2].

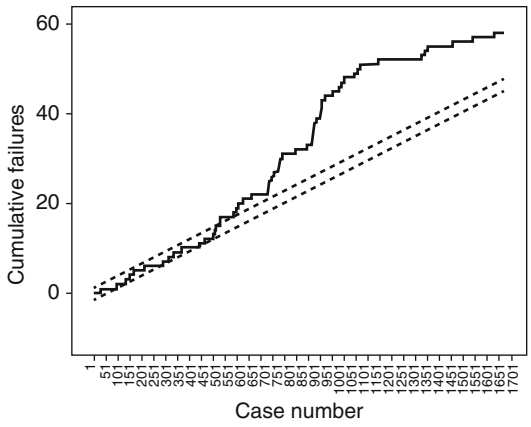

Fig. 25.1 Example of a CUSUM plot (Reprinted from Noyez [1] by permission from Oxford University Press)

Variable Life Adjusted Display (VLAD) Charts: Comparison with Other Methods of Audit

In most countries where audit of surgical outcomes occurs, underpinned by the collection of comprehensive and reliable information in a multi-institutional database, the review of outcomes is retrospective, usually presenting a single number that is the 'result' over some predefined time period (for instance 3 years) and

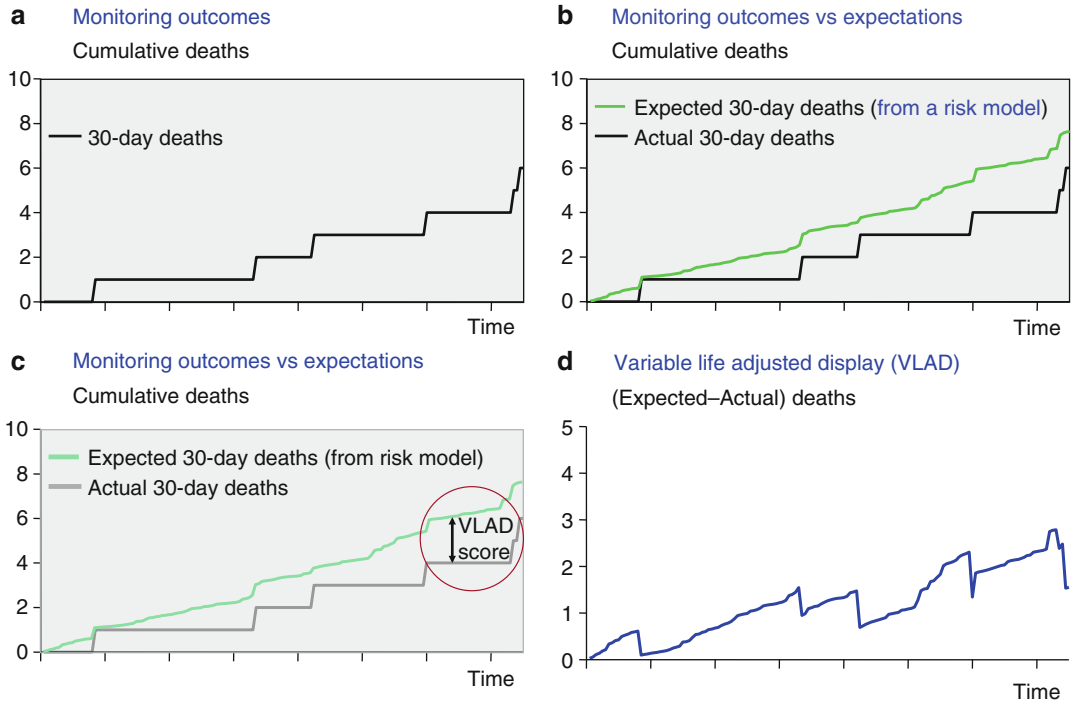

Fig. 25.2 The construction of a VLAD chart in four steps. (**a**) Shows cumulative observed deaths within 30 days displayed over time, a standard CUSUM chart. (**b**) Shows the same CUSUM plot representing observed 30 day deaths alongside a further plot depicting the expected number of deaths over the same set of operations, based on the estimated risk of death from a risk model. (**c**) Shows how the VLAD score is generated by subtracting the actual number of deaths from the expected number of deaths at each time point. (**d**) Shows the actual VLAD plot, which is then the cumulative 'Expected minus Actual Deaths' over time

often with a delay of several months after data submission.

1. Outcomes for different types of operation – currently, in the United Kingdom (UK), paediatric cardiac surgery outcomes for individual procedure types at each paediatric cardiac centre are published, along with the centre identifiers, online in the form of 30-day survival rates measured over a 3 year period displayed on funnel plots by the national audit body [3]. This represents an important contribution to benchmarking of outcomes but limitations include an interval of at least a year before data are published, the small number of patients in many procedure categories, the omission of some procedures from this audit and difficulty in obtaining an overview of a centre's outcome across their entire programme.
2. Programme based outcomes – very recently, the national audit organisation in the UK has engaged in its first attempt to audit program-based outcomes using risk adjustment, and this effort is likely to continue [4]. Internationally, several reported methods for risk-stratification or adjustment exist [5, 6] and Jacobs et al. [7] recently used the STS-EACTS score [6] to compare the risk-adjusted outcomes of paediatric cardiac centres across North America. These analyses of program based outcomes incorporating risk-adjustment may be used to review the mortality/ survival rates across programs for a given era. Despite some inherent limitations, such as the incomplete nature of all risk adjustment models, these analyses may be useful given that they provide comparison of observed outcomes to what was expected from the case mix and applied risk model. However, these retrospectively analysed data have certain disadvantages if one wishes to follow in house program based outcomes in real time for the purposes of quality assurance.

In addition to these described retrospective methods of audit, real time monitoring of risk adjusted surgical outcomes at programme level may be achieved using Variable Life Adjusted Display (VLAD) charts [8].

The VLAD chart displays more information than a conventional summary statistic of the overall mortality or survival rate for a given time era by showing the evolution of outcomes over time, and thus may be useful for qualitative review by clinical teams for the purposes of quality assurance.

An Example of VLAD Chart Use for Real Time Monitoring of Risk-Adjusted Surgical Outcomes from the UK

An example of VLAD chart implementation in the UK was recently reported by Pagel et al. [9]. based on experience of this in three UK paediatric cardiac centres (Great Ormond Street Hospital and Evelina Children's Hospital in London and The Royal Hospital for Sick Children in Glasgow). We note that sections of this chapter are adapted from this open access paper. Each centre provided audit data on all paediatric cardiac surgery procedures conducted in the period 1 January 2010 to 31 December 2011 inclusive.

For each centre, every record of a procedure was allocated to a 30-day episode of care. Each patient's first episode started with their first surgical procedure and the patient's vital status at 30 days was assigned as a primary outcome. Any further procedures within this 30-day episode constituted a secondary outcome. A surgical procedure more than 30 days after the first procedure constituted the beginning of a new episode. This episode allocation is a pre-requisite for applying the Partial Risk Adjustment in Surgery (PRAiS) risk model.

An expected risk of death was estimated for each episode of care using the PRAiS model [10], which was developed and validated using UK national audit data and contains information regarding the surgical procedure, diagnosis, age, weight, and co- morbidity. For each centre, risk-adjusted outcomes over time were displayed using the VLAD method and in addition, further cardiac surgeries and interventional catheterisations within each individual 30-day episode of care were displayed on the VLAD chart. The

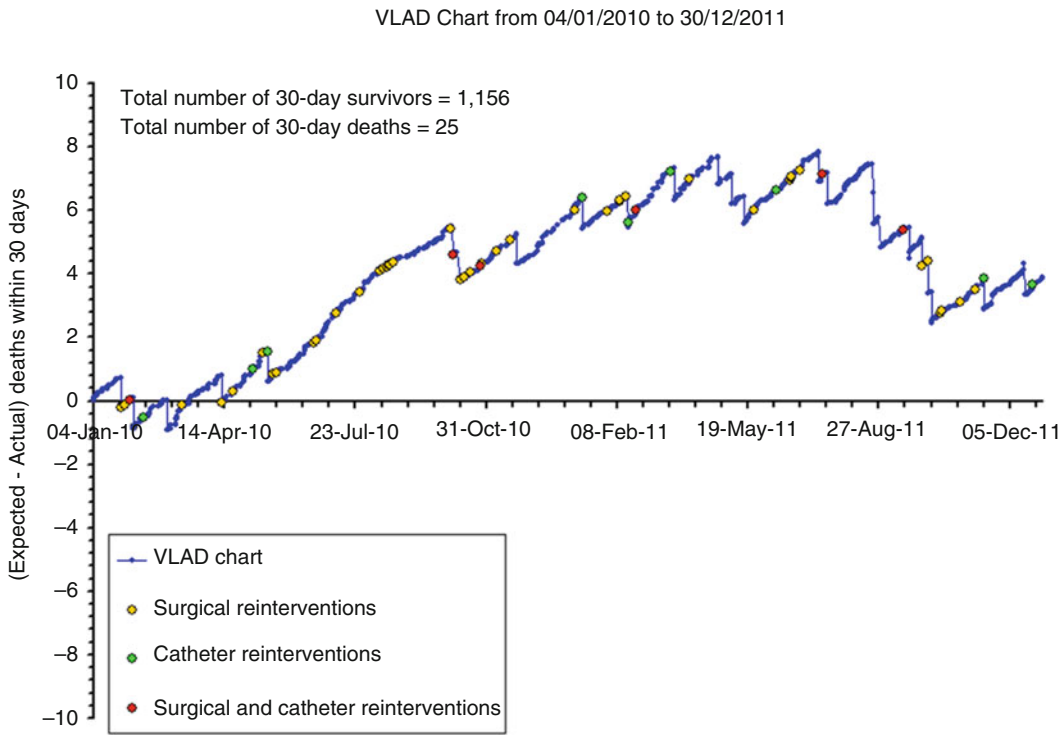

Fig. 25.3 VLAD chart for Centre A. Each *dot* represents a surgical episode (Reproduced with permission from Pagel et al. [9], copyright 2013, with permission from BMJ Publishing Group Ltd)

PRAiS model was implemented and the VLAD charts generated using a bespoke software package in excel, which is available for download under licence (http://www.e-lucid.com/i/software/clinical_data_modelling/PRAiS.html). A generic Excel spreadsheet that can be used for generating VLADs with different risk models is available free of charge with an academic licence from the UCL Clinical Operational Research Unit (http://www.ucl.ac.uk/operational-research/AnalysisTools/VLAD).

VLAD charts for the three participating centres covering the 2-year period of review are shown in Figs. 25.3, 25.4, and 25.5. If outcomes are as expected based on the risk model, the end of the VLAD plot will tend to be close to zero. Given the low 30-day mortality associated with paediatric cardiac surgery (in the UK between 2007 and 2010 UK this was 3.2 % [10]), a VLAD plot will rise much less steeply for a run of survivors than it will fall for a run of deaths. To help place the VLAD charts in context of overall program activity it may be helpful (as done here) to display the overall numbers of deaths and survivors in the top left hand side of the VLAD charts. In this example, the three participating centres all ended the 2 year period within four survivors/deaths from what would be expected using the risk model. However, a major value in the charts is not their final position but in the time evolution: the VLAD charts highlight different time periods for each centre that were of interest to the clinical teams involved as they discussed outcomes locally in conference for the purposes of quality assurance.

To help interpretation of the VLAD charts, we have shown in Fig. 25.6 an enlarged section from the VLAD chart for Centre B. Here the difference between lower- and higher-risk cases is evident: the VLAD plot for higher-risk patients who survived is steeper than for lower-risk survivors and vice versa for patients who died.

Since calendar time is plotted on the horizontal axis, VLAD plots for centres with larger case

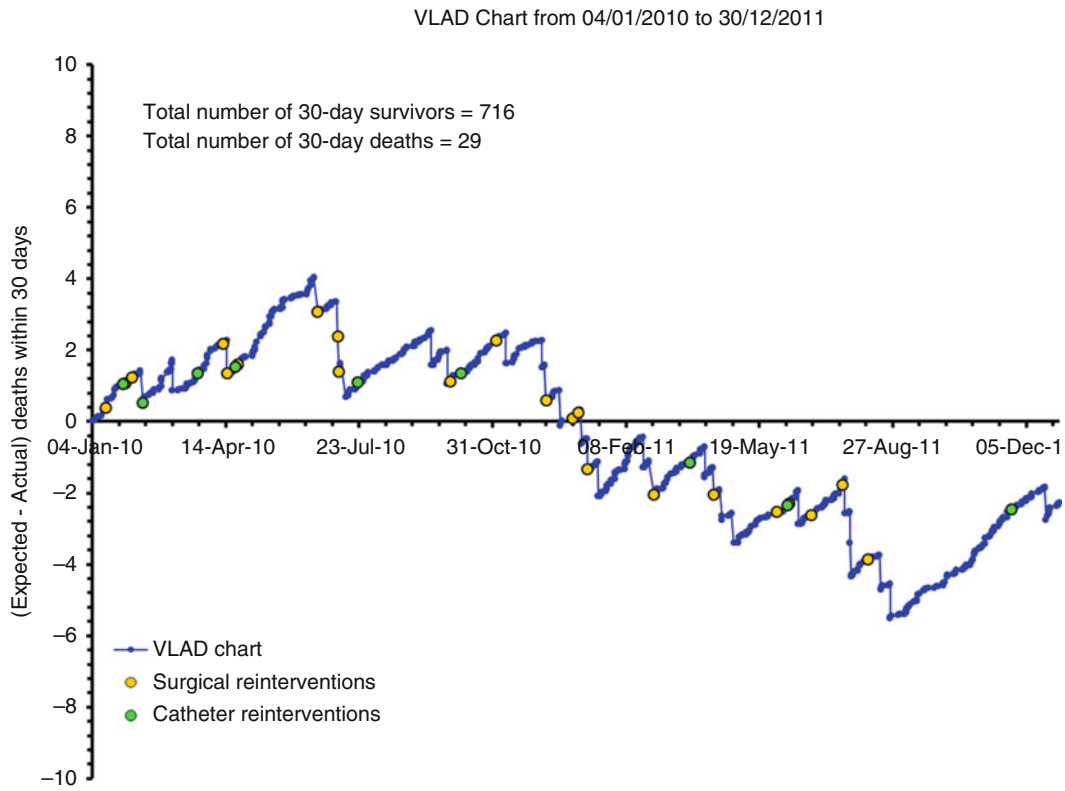

Fig. 25.4 VLAD chart for Centre B. Each *dot* represents a surgical episode (Reproduced with permission from Pagel et al. [9], copyright 2013, with permission from BMJ Publishing Group Ltd)

volumes will have steeper slopes than VLAD plots for centres with a smaller case volume over the same period of time. Since the primary aim is for units to use this software for routine in-house monitoring and not for comparison with other units, this should not represent a barrier to use.

Discussion of VLAD Charts for Real Time Monitoring of Risk-Adjusted Outcomes in 'Wider Context'

In the UK it has now been possible for all centres nationally to use the new software package to monitor their programme level, short-term outcomes with partial risk adjustment using the PRAiS risk model [10] and Variable Life Adjusted Display (VLAD) charts [8]. This necessitated a short orientation session on the methodology for practitioners at each centre delivered by the research team from University College London, with ongoing support by email as queries arise. The National Health Service in England has very recently (2014) made it a mandatory part of the local governance process for paediatric cardiac centres nationally to include presentation of VLADs to a multi-disciplinary audience regularly in the context of mortality and morbidity conferences.

This routine in house monitoring of risk adjusted outcomes using VLAD charts in UK paediatric cardiac centres complements the annual national monitoring using funnel plots already in place for a range of individual surgical procedures on the National Institute for Cardiovascular Outcomes Research (NICOR) Congenital Heart Disease web portal [3]. The VLAD charts have obvious advantages in that firstly, the VLADs incorporate an entire unit's case load and incorporate accessible display of

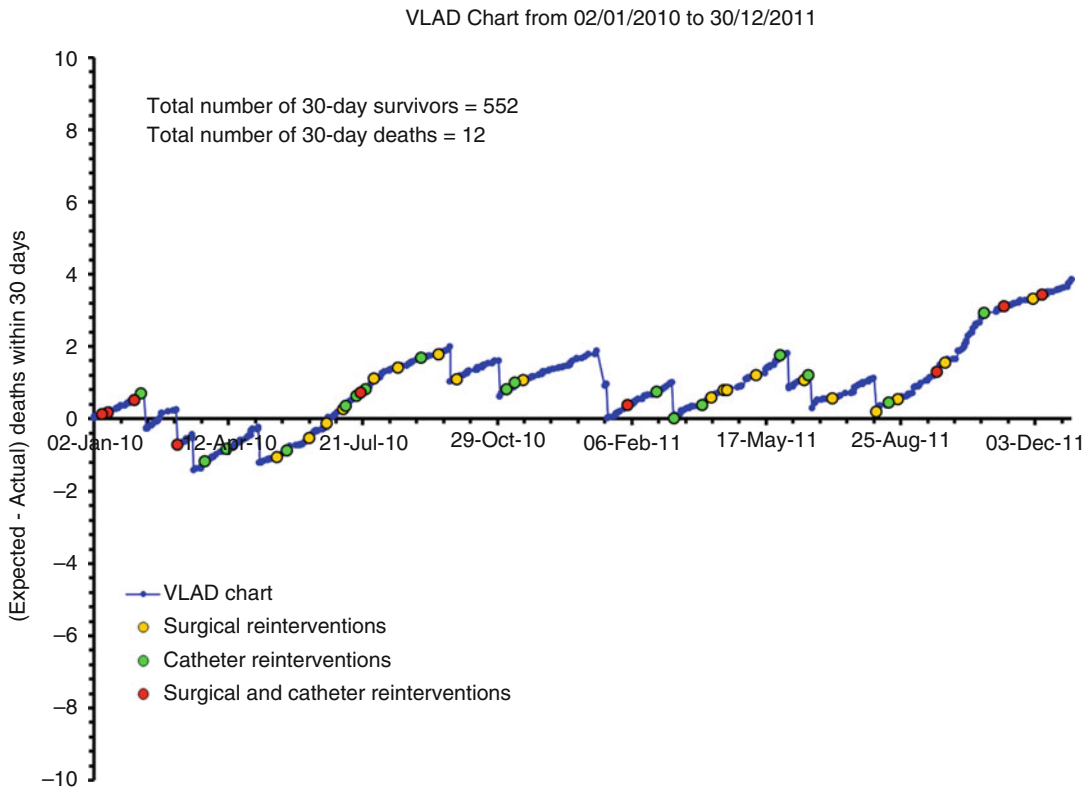

Fig. 25.5 VLAD chart for Centre C. Each *dot* represents a surgical episode (Reproduced with permission from Pagel et al. [9], copyright 2013, with permission from BMJ Publishing Group Ltd)

the individual results of cases over time, thus increasing the chance of any concerning trend being detectable, and secondly the VLADs can be delivered directly to clinical teams for very timely review of risk adjusted outcomes with obvious advantages for quality assurance.

There has been a relatively long standing experience in the use of VLAD plots to evaluate trends in the results of adult cardiac surgery, and these have used data pertaining to individual operators [11, 12] and alternatively, programme level data [8]. A choice to display programme based plots reflects the fact that practice in paediatric cardiac surgery demands high standards from cardiologists, nurses and intensive care specialists as well as surgeons as has been highlighted in recent service reviews [13]. However, anecdotal reports from the UK do indicate that surgeon specific VLAD charts are being utilised albeit for local processes since the PRAiS risk model was introduced nationally in 2013.

Statistical methods based on cumulative sum control chart (CUSUM) analysis have been used previously within VLAD plots to incorporate a 'signal' of poor outcomes [14]. Rocket tails have been applied to VLAD plots to indicate graduations of likelihood that differences between expected and observed outcomes are due to chance [15]. Given the intended use of the paediatric cardiac surgery VLAD plots in a regular continuous programme of review, the research team that developed the software in the UK chose not to include statistical signalling in order to limit scope for complacency. Of note, there is the potential for positive or negative trends to arise by chance, and it must be emphasised that VLAD plots represent a starting point for investigation, not an end.

When initiating real time monitoring of risk-adjusted outcomes within centres, the discussions

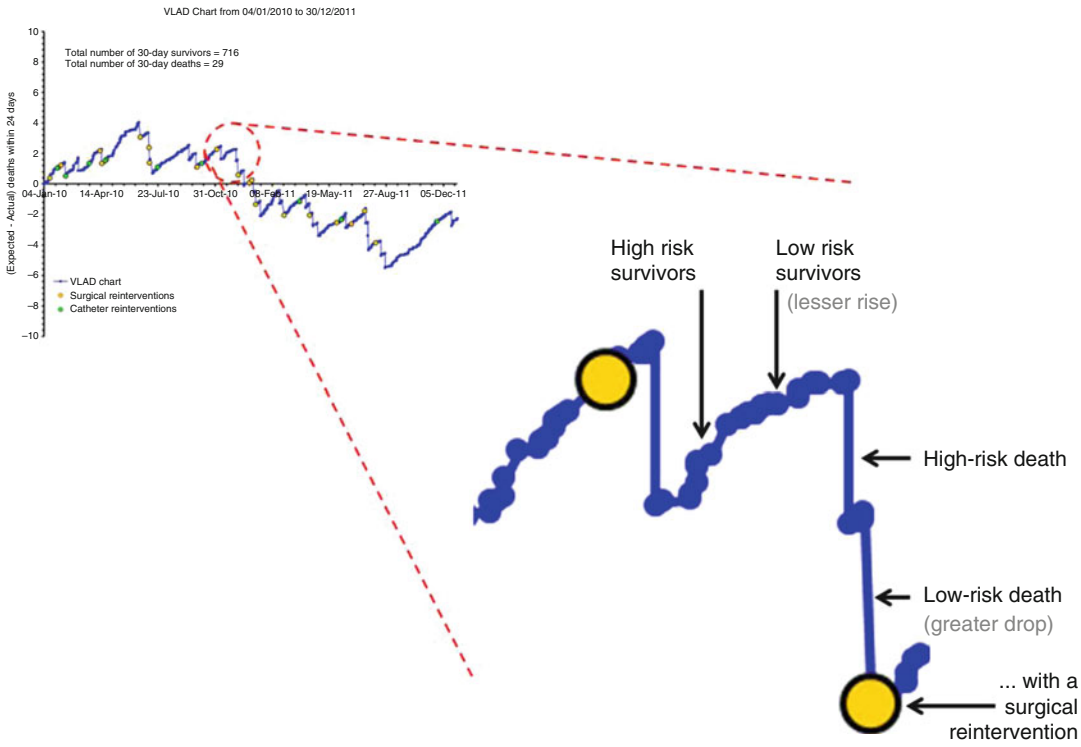

Fig. 25.6 A close-up of a section of the VLAD plot for Centre B showing some features of VLADs (Reproduced with permission from Pagel et al. [9], copyright 2013, with permission from BMJ Publishing Group Ltd)

with clinical teams around context and intended use should consider the sensitivities involved. Previous barriers to clinician participation in reviews of patient safety and quality concerns include fear of censure and reprisals, lack of trust and concern regarding confidentiality. The VLAD charts do not evaluate the programme itself – they show outcomes relative to recent national standards rather than against any absolute measure of value, and besides, quality of service is only one of many possible explanations for observed trends in a VLAD plot. The state of Queensland in Australia, which promotes the use of VLAD charts in a range of health care settings, published a 'pyramid' response to concerns that may arise from a downward trending VLAD chart. This starts with a review of data quality, then moving to patient case mix factors that may have been unaccounted for in risk adjustment, then to health services structural issues then to care processes, and finally to professional performance. Hence, the aim of real time monitoring using VLAD charts is to stimulate reflection upon practice and assist a broader programme of continuous quality improvement. An unanswered question is whether such routine monitoring will lead to service improvement in surgery for congenital heart disease.

References

1. Noyez L. Control charts, Cusum techniques and funnel plots. A review of methods for monitoring performance in healthcare. Interact Cardiovasc Thorac Surg. 2009;9(3):494–9.
2. Jensen HA, Brown KL, Pagel C, Barron DJ, Franklin RCG. Mortality as a measure of quality of care in infants with congenital cardiovascular malformations following surgery. Br Med Bull. 2014;111(1):5–15. doi: 10.1093/bmb/ldu014. Epub 2014 Jul 29.
3. NICOR. Congenital heart diseases web portal. In: London NIoCORNaUC, editor. Congenital heart disease website. London: University College London; 2013.
4. NICOR. Investigation of mortality from paediatric cardiac surgery in England 2009–2012. Report from National Institute for Cardiovascular Outcomes

Research UCL, editor. London: University College London; 2013.
5. Jenkins KJ, Gauvreau K, Newburger JW, Spray TL, Moller JH, Iezzoni LI. Consensus-based method for risk adjustment for surgery for congenital heart disease. J Thorac Cardiovasc Surg. 2002;123(1):110–8.
6. O'Brien SM, Clarke DR, Jacobs JP, Jacobs ML, Lacour-Gayet FG, Pizarro C, et al. An empirically based tool for analyzing mortality associated with congenital heart surgery. J Thorac Cardiovasc Surg. 2009;138(5):1139–53.
7. Jacobs JP, O'Brien SM, Pasquali SK, Jacobs ML, Lacour-Gayet FG, Tchervenkov CI, et al. Variation in outcomes for risk-stratified pediatric cardiac surgical operations: an analysis of the STS Congenital Heart Surgery Database. Ann Thorac Surg. 2012;94(2):564–71; discussion 71–2.
8. Lovegrove J, Valencia O, Treasure T, Sherlaw-Johnson C, Gallivan S. Monitoring the results of cardiac surgery by variable life-adjusted display. Lancet. 1997;350(9085):1128–30.
9. Pagel C, Utley M, Crowe S, Witter T, Anderson D, Samson R, et al. Real time monitoring of risk-adjusted paediatric cardiac surgery outcomes using variable life-adjusted display: implementation in three UK centres. Heart. 2013;99(19):1445–50.
10. Crowe S, Brown KL, Pagel C, Muthialu N, Cunningham D, Gibbs J, et al. Development of a diagnosis- and procedure-based risk model for 30-day outcome after pediatric cardiac surgery. J Thorac Cardiovasc Surg. 2013;145(5):1270–8.
11. Bridgewater B, Grayson AD, Jackson M, Brooks N, Grotte GJ, Keenan DJ, et al. Surgeon specific mortality in adult cardiac surgery: comparison between crude and risk stratified data. BMJ. 2003;327(7405):13–7.
12. Bridgewater B. Mortality data in adult cardiac surgery for named surgeons: retrospective examination of prospectively collected data on coronary artery surgery and aortic valve replacement. BMJ. 2005; 330(7490):506–10.
13. Moore A, Agnew T, Cole A. Bristol Royal Infirmary – the aftermath: six pages of analysis. Health Serv J. 2001;111(5764):11–7.
14. Sherlaw-Johnson C. A method for detecting runs of good and bad clinical outcomes on Variable Life-Adjusted Display (VLAD) charts. Health Care Manag Sci. 2005;8(1):61–5.
15. Sherlaw-Johnson C, Morton A, Robinson MB, Hall A. Real-time monitoring of coronary care mortality: a comparison and combination of two monitoring tools. Int J Cardiol. 2005;100(2):301–7.

26

Risk Adjustment for Congenital Heart Surgery -1 (RACHS-1) for Evaluation of Mortality in Children Undergoing Cardiac Surgery

Ravi R. Thiagarajan and Peter C. Laussen

Abstract

Assessing quality of care provided by a health care system requires a clear understanding of non-modifiable risks present prior to entry into the system. Risk adjustment is a process of understanding and accounting for these risk factors when evaluating outcomes of health care. Surgery for congenital heart disease requires the performance of a large range of procedures. These procedures vary widely in complexity, and surgical complexity strongly influences survival following congenital heart surgery. A risk adjustment method that adjusts for risk based on procedural complexity is thus required when assessing quality of pediatric cardiac surgical care provided by or between institutions. Risk Adjustment in Congenital Heart Surgery (RACHS-1) method is a simple, well-tested, widely used, and excellent risk adjustment method for assessing quality of care in children undergoing congenital heart surgery.

Keywords

Congenital heart surgery • Outcomes • Risk adjustment • RACHS-1

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Introduction

Information on quality of care provided by a system has become increasingly useful to patients and providers for making health care decisions, choosing providers or health care systems, and for purposes of reimbursement [1, 2]. Accurate assessment of health outcomes provided by a health system requires a clear understanding of the population receiving care from that system. Differences in the risk of adverse health care outcomes inherent in the

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population prior to receiving care can reduce healthcare outcomes even if the care provided by the system is exceptional. When comparing quality of care between institutions, differences in inherent risk of adverse health outcomes in patients served an institution can make comparing quality of care difficult. Thus, the types of cases receiving care in an institution or "case-mix" should be carefully considered when interpreting differences in outcomes between institutions. Adjustment for differences in case-mix or "risk adjustment" is an integral part of any analysis aimed at comparing healthcare outcomes between institutions or health care systems.

Surgical correction or palliation of congenital heart disease (CHD) is now practiced in a large number of institutions. Surgical mortality for children with CHD has been shown to be associated with patient level risk factors (e.g. prematurity), type and complexity of the procedure performed (e.g. Norwood operation), and institutional (e.g. volume of procedures performed by a center and nursing experience) and provider (surgeon volume) characteristics [3–5]. Given that a large number of surgical procedures varying widely in surgical complexity and risk of mortality are performed in children for management of CHD, accurate assessment of quality of surgical care provided adjustment of risks for children with congenital heart disease between institutions requires careful adjustment of risk due these inherent differences [6]. Several risk adjustment methods have been described for studying post-operative mortality following surgery for treatment of CHD in children. [2, 6–8]. These methods include the Risk Adjustment for Congenital Heart Surgery (version 1; RACHS-1), the Aristotle Complexity Score (basic and comprehensive), and the **S**ociety of **T**horacic Surgeons – European **A**ssociation for Cardio-**T**horacic Surgery Congenital Heart Surgery Mortality Score and Categories STAT Mortality Score and Categories. Here we describe the characteristics, uses, and limitations of the RACHS-1 score.

RACHS-1 Method

The RACHS-1 method was created to improve assessment of in-hospital mortality in children <18 years of age with congenital heart undergoing cardiac surgery [2, 6]. RACHS-1 was developed 2001 and utilized a panel of national experts to categorize pediatric cardiac surgical procedures into six categories of increasing surgical complexity based on the risk of post-operative death. Two multicenter datasets obtained from the Pediatric Cardiac Care Consortium (PCCC) and hospital discharge data from three states US were used to refine and test the RACHS-1 method. The final RACHS-1 method was published in 2002. The RACHS-1 is currently one of the most widely used risk adjustment methods for assessing mortality in children with CHD undergoing cardiac surgery. The following describe details of the RACHS-1 method.

Components of RACHS-1

RACHS-1 uses procedural and patient level information as components to provide adjustment for the influence of differences in case-mix on post-surgical mortality (Table 26.1) [2, 6]. The procedural component contains cardiac surgical procedures for CHD categorized into six complexity categories. RACHS-1 uses surgical procedures rather than CHD diagnosis for com-

Table 26.1 Components of RACHS-1 method

RACHS-1 components
Procedural information
Surgical risk category 1–6
Combination procedures
Patient level factors
Age groups
≤30 days
31 days to 1 year
>1 year
Major chromosomal and non-cardiac structural anomaly
Prematurity

plexity categorization. The categorization of CHD procedural complexity was accomplished by utilizing the consensus of a 11 member expert panel who assigned 207 pediatric cardiac surgical procedures, identified using current procedural terminology 4 (CPT 4) and International Classification of Disease, Ninth edition (ICD-9) procedure codes, into Risk Categories based on the risk death prior to hospital discharge. Patent Ductus Arteriosus ligation in neonates <30 days of age and in those weighing <2,500 g, cardiac transplantation, and interventional cardiac catheter based procedures for management of CHD are excluded from categorization. Risk category for patients undergoing multiple surgical procedures is assigned based on the procedure with highest complexity. The variable "Combination Procedure" allows further risk adjustment for those undergoing multiple procedures. Categorization of some procedures are age and diagnosis based (e.g. Coarctation of Aorta >30 days age is assigned Risk Category 1, while Coarcation of Aorta ≤30 days age is assigned to Risk Category 2). In the original article describing the RACHS-1 method, Jenkins et al. provide detailed information on categorization of surgical procedures into Risk Categories [2]. The risk of death rises across the 6 RACHS-1 categories. However the increase is not linear, and because mortality rates vary distinctly between each category, collapsing or combining risk categories may possibly diminish the utility of RACHS-1 [6]. Appendix 1 provides the algorithm for application of RACHS-1 using the International Pediatric and Congenital Cardiac Code (IPCCC) in the Society of Thoracic Surgeons Congenital Heart Surgery Database and the European Association for Cardio-Thoracic Surgery Congenital Heart Surgery Database. Because RACHS-1 category 5 is so rare, it is often combined with RACHS-1 category 6.

The second component of the RACHS-1 method contains patient level factors that may influence pediatric cardiac surgical outcomes (Table 26.1) [2]. These factors are shown in Table 26.1 and include age at surgery, prematurity (defined as <36 weeks) and major non-cardiac structural abnormalities (e.g. trachea-esophageal fistula) or major chromosomal abnormalities or syndromes (e.g. DiGeorge syndrome). This component provides risk adjustment for important patient level factors when studying mortality in this population.

RACHS-1 Performance

The ability of RACHS-1 in discriminating mortality was published in the original article describing the RACHS-1 system by Jenkins et al. [2, 6]. Overall they found excellent discriminative capabilities for the RACHS-1 method [Area under Receiver Operating Characteristics Curve (ROC): 0.811–0.814]. The discriminative capability is largely determined by the procedural complexity Risk Categories (ROC: 0.784; using the PCCC dataset), however the addition of the patient level variables improved the capacity of RACHS-1 in discriminating mortality (ROC: 0.817 with addition of the three patient level variables and using the PCC dataset). The RACHS-1 system is reported to be able to assign 80–90 % of cases in any given dataset to Risk Categories for purposes of risk adjustment [6]. Unassigned cases can cause residual confounding and these cases should be carefully examined when using the RACHS-1 method [9].

Comparison of RACHS-1 with Other Risk Adjustment Methods

A number of recent studies have compared the RACHS-1 method to other existing risk adjustment methods such as the Aristotle Basic Complexity (ABC) score and the more recent STS-EACTS score [7, 10–14]. The performance of the RACHS systems was both superior in some and inferior in others when compared to other methods in these studies. In a recent study using pediatric cardiac surgical cases from the Society of Thoracic Surgeons database by O'Brien et al., the ability to discriminate mortality across the currently available risk adjustment methods were comparable [*C-index* (*value interpreted similar to ROC*) STS-EACTS

score = 0. 816; STS-EACTS categories = 0.812; RACHS-1 = 0.802; and ABC = 0.795; all methods included patient level factors] [7]. Type and completeness of procedural data available for categorization, ability to assign the majority if not all patients to risk categories in a given dataset, and number of patient level variables available for use may all influence performance of each method.

Use of RACHS-1 Method

RACHS-1 can be used to compare mortality rates between centers and is perhaps most commonly used for this purpose [4, 15, 16]. RACHS-1 system can be used to adjust for confounding from case-mix on mortality when studying the impact of patient, institutional, and other systems level issues on post-operative mortality following pediatric cardiac surgery. Although not common, RACHS-1 has been used to evaluate some morbidity outcomes such as length of stay and duration of ventilation following pediatric cardiac surgery for CHD [17].

Limitations

All risk adjustment methods have limitations and these issues should be carefully considered prior to their use [2, 6, 13]. Some limitations for the RACHS-1 methods are described here. The RACHS-1 method was created to provide risk adjustment for comparing post-operative mortality between groups of patients undergoing congenital heart surgery. Thus RACHS-1 cannot be used to "predict" mortality for individual procedures or categories of procedures. Some cardiac surgical procedures that cannot be defined using the ICD-9 or CPT coding systems may not be assigned to a RACHS-1 Risk Category and this can diminish the utility of the method. Certain distinct congenital heart disease diagnostic details can strongly influence risk of post-operative mortality; these issues are not captured by RACHS-1 and can result in misclassification of complexity. Finally, RACHS-1 was developed in 2001 and that cardiac surgical outcomes have improved over time, its use for risk adjustment to study current post-operative mortality may require careful consideration.

Summary

The RACHS-1 method is a commonly used risk adjustment system for studying post-operative mortality in children with congenital heart disease. RACHS-1 categorizes cardiac surgical procedures based on complexity into six Risk Categories. Several patient levels factors included in RACHS-1 provide adjustment for patient level risk. The ability of RACHS-1 to discriminate post-cardiac surgical mortality in children undergoing cardiac surgery is excellent and compares well with other currently available methods. Ease of use, need for only limited number of commonly collected data fields, and the ability to use RACHS-1 method with any multicenter database, has resulted in its wide applicability.

Appendix 1: The Risk Adjustment for Congenital Heart Surgery (RACHS-1) Categories (January 1, 2010)

Appendix 1 documents how the Risk Adjustment for Congenital Heart Surgery (RACHS-1) Categories are applied in the STS Congenital Heart Surgery Database and the EACTS Congenital Heart Surgery Database [18].

Procedure	RACHS-1 Category
Aortopexy	1
ASD partial closure	1
ASD repair, Patch	1
ASD repair, Patch + PAPVC repair	1
ASD repair, Primary closure	1
Atrial fenestration closure	1
PAPVC repair	1
PAPVC Repair, Baffle redirection to left atrium with systemic vein translocation (Warden) (SVC sewn to right atrial appendage)	1
PAPVC, Scimitar, Repair	1
PFO, Primary closure	1
Aneurysm, Ventricular, Right, Repair	2
Aortic stenosis, Subvalvar, Repair	2
AP window repair	2
ASD, Common atrium (single atrium), Septation	2
AVC (AVSD) repair, Partial (Incomplete) (PAVSD)	2
Bidirectional cavopulmonary anastomosis (BDCPA) (bidirectional Glenn)	2
Bilateral bidirectional cavopulmonary anastomosis (BBDCPA) (bilateral bidirectional Glenn)	2
Coronary artery fistula ligation	2
DCRV repair	2
Glenn (unidirectional cavopulmonary anastomosis) (unidirectional Glenn)	2
Hemifontan	2
Ligation, Pulmonary artery	2
PA, reconstruction (plasty), Branch, Central (within the hilar bifurcation)	2
PA, reconstruction (plasty), Branch, Peripheral (at or beyond the hilar bifurcation)	2
PA, reconstruction (plasty), Main (trunk)	2
PA, reconstruction (plasty), NOS	2
Pulmonary artery sling repair	2
RVOT procedure	2
Sinus of Valsalva, Aneurysm repair	2
Superior cavopulmonary anastomosis(es) + PA reconstruction	2
TOF repair, No ventriculotomy	2
TOF repair, Ventriculotomy, Nontransanular patch	2
TOF repair, Ventriculotomy, Transanular patch	2
Valve replacement, Pulmonic (PVR)	2
Valve surgery, Other, Pulmonic	2
Valvuloplasty converted to valve replacement in the same operation, Pulmonic	2
Valvuloplasty, Pulmonic	2
Vascular ring repair	2
VSD repair, Patch	2
VSD repair, Primary closure	2
VSD, Multiple, Repair	2
Aortic aneurysm repair	3
Aortic arch repair + VSD repair	3
Aortic stenosis, Subvalvar, Repair, With myectomy for IHSS	3
Aortic stenosis, Supravalvar, Repair	3

Arterial switch operation (ASO)	3
Atrial baffle procedure (non-Mustard, non-Senning)	3
Atrial baffle procedure, Mustard or Senning revision	3
Atrial baffle procedure, NOS	3
Atrial baffle procedure, NOS	3
AVC (AVSD) repair, Complete (CAVSD)	3
AVC (AVSD) repair, Intermediate (Transitional)	3
Cardiac tumor resection	3
Coarctation repair + VSD repair	3
Conduit placement, LV to PA	3
Conduit placement, RV to PA	3
Cor triatriatum repair	3
DORV repair, NOS	3
DORV, Intraventricular tunnel repair	3
Fontan + Atrioventricular valvuloplasty	3
Fontan, Atrio-pulmonary connection	3
Fontan, Atrio-ventricular connection	3
Fontan, NOS	3
Fontan, Other	3
Fontan, TCPC, External conduit, Fenestrated	3
Fontan, TCPC, External conduit, Nonfenestrated	3
Fontan, TCPC, External conduit, NOS	3
Fontan, TCPC, Lateral tunnel, Fenestrated	3
Fontan, TCPC, Lateral tunnel, Nonfenestrated	3
Fontan, TCPC, Lateral tunnel, NOS	3
Hybrid Approach "Stage 1", Application of RPA and LPA bands	3
Hybrid Approach "Stage 1", Stent placement in arterial duct (PDA) + application of RPA and LPA bands	3
Mustard	3
PA banding (PAB)	3
Pulmonary artery origin from ascending aorta (hemitruncus) repair	3
Pulmonary atresia - VSD - MAPCA (pseudotruncus) repair	3
Pulmonary atresia - VSD (including TOF, PA) repair	3
Ross procedure	3
Senning	3
Shunt, Systemic to pulmonary, Central (from aorta or to main pulmonary artery)	3
Shunt, Systemic to pulmonary, Modified Blalock-Taussig Shunt (MBTS)	3
Shunt, Systemic to pulmonary, NOS	3
Shunt, Systemic to pulmonary, Other	3
Superior cavopulmonary anastomosis(es) (Glenn or HemiFontan) + Atrioventricular valvuloplasty	3
Valve closure, Semilunar	3
Valve excision, Tricuspid (without replacement)	3
Valve replacement, Aortic (AVR)	3
Valve replacement, Aortic (AVR), Bioprosthetic	3
Valve replacement, Aortic (AVR), Homograft	3
Valve replacement, Aortic (AVR), Mechanical	3
Valve replacement, Mitral (MVR)	3
Valve replacement, Tricuspid (TVR)	3
Valve surgery, Other, Aortic	3
Valve surgery, Other, Mitral	3
Valve surgery, Other, Tricuspid	3
Valvuloplasty converted to valve replacement in same operation, Tricuspid	3
Valvuloplasty converted to valve replacement in the same operation, Aortic	3
Valvuloplasty converted to valve replacement in the same operation, Aortic - with Ross procedure	3
Valvuloplasty converted to valve replacement in the same operation, Mitral	3
Valvuloplasty, Mitral	3
Valvuloplasty, Tricuspid	3
Aortic arch repair	4
Arterial switch operation (ASO) and VSD repair	4
ASD creation/enlargement	4
Congenitally corrected TGA repair, Atrial switch and ASO (double switch)	4
Interrupted aortic arch repair	4
Konno procedure	4
Rastelli	4

Truncus arteriosus repair	4
Unifocalization MAPCA(s)	4
Valvuloplasty converted to valve replacement in the same operation, Aortic - with Ross-Konno procedure	4
VSD creation/enlargement	4
Truncus + Interrupted aortic arch repair (IAA) repair	5
Damus-Kaye-Stansel procedure (DKS) (creation of AP anastomosis without arch reconstruction)	6
Hybrid Approach "Stage 2", Aortopulmonary amalgamation + Superior Cavopulmonary anastomosis(es) + PA Debanding + Aortic arch repair (Norwood [Stage 1] + Superior Cavopulmonary anastomosis(es) + PA Debanding)	6
Norwood procedure	6
Coarctation repair, End to end	1 if > 30d, 2 if <= 30d
Coarctation repair, End to end, Extended	1 if > 30d, 2 if <= 30d
Coarctation repair, Interposition graft	1 if > 30d, 2 if <= 30d
Coarctation repair, NOS	1 if > 30d, 2 if <= 30d
Coarctation repair, Other	1 if > 30d, 2 if <= 30d
Coarctation repair, Patch aortoplasty	1 if > 30d, 2 if <= 30d
Coarctation repair, Subclavian flap	1 if > 30d, 2 if <= 30d
PDA closure, Surgical	1 if > 30d, not eligible if <= 30d
TAPVC repair	2 if > 30d, 4 if <= 30d
Valvuloplasty, Aortic	2 if > 30d, 4 if <= 30d
Ebstein's repair	3 if > 30d, 5 if <= 30d
TAPVC repair + Shunt - Systemic to pulmonary	3 if age >30 days, 4 if age <=30 days

Other, Not Categorized (Eligible, but not assigned a category)
1 1/2 ventricular repair
Aneurysm, Pulmonary artery, Repair
Aneurysm, Ventricular, Left, Repair
Anomalous origin of coronary artery from pulmonary artery repair
Anomalous systemic venous connection repair
Aortic root replacement
Aortic root replacement, Bioprosthetic
Aortic root replacement, Homograft
Aortic root replacement, Mechanical
Aortic root replacement, Valve sparing
Aortic root translocation over left ventricle (Including Nikaidoh procedure)
Arterial switch procedure + Aortic arch repair
Arterial switch procedure and VSD repair + Aortic arch repair
Atrial septal fenestration
AVC (AVSD) repair, NOS
Conduit placement, NOS
Conduit placement, Other
Conduit placement, Ventricle to aorta
Conduit reoperation
Congenitally corrected TGA repair, Atrial switch and Rastelli
Congenitally corrected TGA repair, NOS
Congenitally corrected TGA repair, Other
Congenitally corrected TGA repair, VSD closure
Congenitally corrected TGA repair, VSD closure and LV to PA conduit
Coronary artery bypass
Coronary artery procedure, Other
DOLV repair
Fontan revision or conversion (Re-do Fontan)
HLHS biventricular repair
Hybrid Approach "Stage 1", Stent placement in arterial duct (PDA)
Hybrid Approach "Stage 2", Aortopulmonary amalgamation + Superior Cavopulmonary anastomosis(es) + PA Debanding + Without aortic arch repair
LV to aorta tunnel repair
Mitral stenosis, Supravalvar mitral ring repair
Occlusion MAPCA(s)
Other annular enlargement procedure
PA debanding
Partial left ventriculectomy (LV volume reduction surgery) (Batista)
Pulmonary AV fistula repair/occlusion
Pulmonary embolectomy
Pulmonary embolectomy, Acute pulmonary embolus
Pulmonary embolectomy, Chronic pulmonary embolus

Pulmonary venous stenosis repair
REV
Ross-Konno procedure
Shunt, Ligation and takedown
Systemic venous stenosis repair
TGA, Other procedures (Kawashima, LV-PA conduit, other)
TGA, Other procedures (Nikaido, Kawashima, LV-PA conduit, other)
TOF - AVC (AVSD) repair
TOF - Absent pulmonary valve repair
TOF repair, NOS
TOF repair, RV-PA conduit
Valve closure, Tricuspid (exclusion, univentricular approach)
Valve excision, Pulmonary (without replacement)
Valve replacement, Common atrioventricular valve
Valve replacement, Truncal valve
Valvuloplasty converted to valve replacement in the same operation, Common atrioventricular valve
Valvuloplasty converted to valve replacement in the same operation, Truncal valve
Valvuloplasty, Common atrioventricular valve
Valvuloplasty, Truncal valve
Ventricular septal fenestration
Ventricular septation

Other, Not Eligible (Intentionally excluded from RACHS-1)
Aortic dissection repair
Arrhythmia surgery - atrial, Surgical Ablation
Arrhythmia surgery - ventricular, Surgical Ablation
Arrhythmia surgery, NOS
ASD creation, Balloon septostomy (BAS) (Rashkind)
ASD creation, Blade septostomy
ASD repair, Device
ASD repair, NOS
Balloon dilation
Bronchoscopy
Bypass for noncardiac lesion
Cardiac procedure, Other
Cardiotomy, Other
Cardiovascular catherization procedure, Diagnostic
Cardiovascular catherization procedure, Diagnostic, Angiographic data obtained
Cardiovascular catherization procedure, Diagnostic, Electrophysiology alteration
Cardiovascular catherization procedure, Diagnostic, Hemodynamic alteration
Cardiovascular catherization procedure, Diagnostic, Hemodynamic data obtained
Cardiovascular catherization procedure, Diagnostic, Transluminal test occlusion
Cardiovascular catherization procedure, Therapeutic
Cardiovascular catherization procedure, Therapeutic Adjunctive therapy
Cardiovascular catherization procedure, Therapeutic, Balloon valvotomy
Cardiovascular catherization procedure, Therapeutic, Perforation (establishing interchamber and/or intervessel communication)
Cardiovascular catherization procedure, Therapeutic, Septostomy
Cardiovascular catherization procedure, Therapeutic, Stent re-dilation
Cardiovascular catherization procedure, Therapeutic, Transcatheter Fontan completion
Cardiovascular catherization procedure, Therapeutic, Transcatheter implantation of valve
Cardiovascular electophysiological catheterization procedure
Cardiovascular electophysiological catheterization procedure, Therapeutic ablation
Coil embolization
Decortication
Delayed sternal closure
Device closure
Diaphragm plication
Diaphragm procedure, Other
Echocardiography procedure, Sedated transesophageal echocardiogram
Echocardiography procedure, Sedated transthoracic echocardiogram
ECMO cannulation
ECMO decannulation
ECMO procedure
Esophageal procedure
Explantation of pacing system
ICD (AICD) ([automatic] implantable cardioverter defibrillator) procedure

ICD (AICD) implantation
Intraaortic balloon pump (IABP) insertion
Ligation, Thoracic duct
Lung biopsy
Lung procedure, Other
Mediastinal exploration
Mediastinal procedure
Minimally invasive procedure
Miscellaneous procedure, Other
Non-cardiovascular, non-thoracic procedure on cardiac patient with cardiac anesthesia
Organ procurement
Other procedure
Pacemaker implantation, Permanent
Pacemaker procedure
Palliation, Other
PDA closure, Device
PDA closure, NOS
Pectus repair
Pericardial drainage procedure
Pericardial procedure, Other
Pericardiectomy
Peripheral vascular procedure, Other
Pleural drainage procedure
Pleural procedure, Other
Radiology procedure on cardiac patient, Cardiac Computerized Axial Tomography (CT Scan)
Radiology procedure on cardiac patient, Cardiac Magnetic Resonance Imaging (MRI)
Radiology procedure on cardiac patient, Diagnostic radiology
Radiology procedure on cardiac patient, Non-Cardiac Computerized Tomography (CT) on cardiac patient
Radiology procedure on cardiac patient, Non-Cardiac Magnetic Resonance Imaging (MRI) on cardiac patient
Radiology procedure on cardiac patient, Therapeutic radiology
RF ablation
Right/left heart assist device procedure
Stent placement
Sternotomy wound drainage
Thoracic and/or mediastinal procedure, Other
Thoracotomy, Other
Tracheal procedure
Transplant, Heart
Transplant, Heart and lung
Transplant, lung(s)
VAD explantation
VAD implantation
VATS (video-assisted thoracoscopic surgery)
VSD repair, Device
VSD repair, NOS

References

1. Welke KF, Shen I, Ungerleider RM. Current assessment of mortality rates in congenital cardiac surgery. Ann Thorac Surg. 2006;82(1):164–70; discussion 170–1.
2. Jenkins KJ, et al. Consensus-based method for risk adjustment for surgery for congenital heart disease. J Thorac Cardiovasc Surg. 2002;123(1):110–8.
3. Hannan EL, et al. Pediatric cardiac surgery: the effect of hospital and surgeon volume on in-hospital mortality. Pediatrics. 1998;101(6):963–9.
4. Jenkins KJ, et al. In-hospital mortality for surgical repair of congenital heart defects: preliminary observations of variation by hospital caseload. Pediatrics. 1995;95(3):323–30.
5. Hickey PA, et al. The effect of critical care nursing and organizational characteristics on pediatric cardiac surgery mortality in the United States. J Nurs Adm. 2013;43(12):637–44.
6. Jenkins KJ. Risk adjustment for congenital heart surgery: the RACHS-1 method. Seminars in thoracic and cardiovascular surgery. Pediatr Cardiac Surg Annu. 2004;7:180–4.
7. O'Brien SM, et al. An empirically based tool for analyzing mortality associated with congenital heart surgery. J Thorac Cardiovasc Surg. 2009;138(5):1139–53.
8. Lacour-Gayet F, et al. The Aristotle score: a complexity-adjusted method to evaluate surgical results. Eur J Cardiothorac Surg. 2004;25(6):911–24.
9. Pasquali SK, et al. Differential case ascertainment in clinical registry versus administrative data and impact on outcomes assessment for pediatric cardiac operations. Ann Thorac Surg. 2013;95(1):197–203.
10. Al-Radi OO, et al. Case complexity scores in congenital heart surgery: a comparative study of the Aristotle Basic Complexity score and the Risk Adjustment in Congenital Heart Surgery (RACHS-1) system. J Thorac Cardiovasc Surg. 2007;133(4):865–75.
11. Bojan M, et al. Comparative study of the Aristotle Comprehensive Complexity and the Risk Adjustment in Congenital Heart Surgery scores. Ann Thorac Surg. 2011;92(3):949–56.
12. Bojan M, et al. The Aristotle Comprehensive Complexity score predicts mortality and morbidity after congenital heart surgery. Ann Thorac Surg. 2011;91(4):1214–21.
13. Kang N, et al. Risk stratification in paediatric open-heart surgery. Eur J Cardiothorac Surg. 2004;26(1):3–11.
14. Kang N, et al. Does the Aristotle score predict outcome in congenital heart surgery? Eur J Cardiothorac Surg. 2006;29(6):986–8.
15. DeMone JA, et al. Risk of death for Medicaid recipients undergoing congenital heart surgery. Pediatr Cardiol. 2003;24(2):97–102.
16. Jenkins KJ, Gauvreau K. Center-specific differences in mortality: preliminary analyses using the Risk Adjustment in Congenital Heart Surgery (RACHS-1) method. J Thorac Cardiovasc Surg. 2002;124(1):97–104.
17. Boethig D, et al. The RACHS-1 risk categories reflect mortality and length of hospital stay in a large German pediatric cardiac surgery population. Eur J Cardiothorac Surg. 2004;26(1):12–7.
18. Jacobs JP, Jacobs ML, Mavroudis C, Lacour-Gayet FG, Tchervenkov CI, Pasquali SK. Executive summary: the Society of Thoracic Surgeons Congenital Heart Surgery Database – Nineteenth Harvest – (July 1, 2009 – June 30, 2013). Durham: The Society of Thoracic Surgeons (STS) and Duke Clinical Research Institute (DCRI), Duke University Medical Center; 2013.

The Aristotle Complexity Score: A Tool to Evaluate Performance in Congenital Heart Surgery

27

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Abstract

The Aristotle Score is a complexity stratification system that was based initially on expert opinion, in absence of sufficient objective data from databases. Because the scoring system was derived from opinions, we gave the name of Aristotle to this project.

The Aristotle Score follows several rules and principles:

– The Aristotle Score measures the complexity of surgical procedures. The Aristotle Score was designed to evaluate performance and not directly to predict mortality.

– The complexity of a surgical procedure is a constant and is calculated with the following equation:

$$\textit{Complexity} = \textit{Potential for Mortality} + \textit{Potential for Morbidity} + \textit{Technical Difficulty}$$

– The calculated complexity includes both the ***Aristotle Basic Complexity Score*** (ABC Score) and the ***Aristotle Comprehensive Complexity Score*** (ACC Score).

– Performance is calculated with the following equation:

$$\textit{Performance} = \textit{Outcome} \times \textit{Complexity}$$

– Several performances can be calculated by combining complexity with various outcomes.

Two steps are defined:

1. The ***Aristotle Basic Complexity Score*** (***ABC Score***) is calculated on 15 points and is determined from the primary procedure of any operation

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2. The ***Aristotle Comprehensive Complexity Score*** (***ACC Score***) is calculated on 25 points: The Aristotle Comprehensive Complexity Score equals the Aristotle Basic Complexity Score plus 5 points for ***Procedure-dependent Factors*** and 5 points for ***Procedure-independent Factors***.

   The ***Aristotle Basic Complexity Score*** was introduced in the Congenital Heart Surgery Database of The Society of Thoracic Surgeons (STS) and The European Association for Cardio-Thoracic Surgery (EACTS) in 2002 and has been instrumental for the success of these congenital databases. The ***Aristotle Basic Complexity Score*** was validated with a C-Index for mortality and morbidity of 0.70 and 0.67 respectively. The ***Aristotle Comprehensive Complexity Score*** (www.aristotleinstitute.org) is used by many individual institutions with a C-Index of 0.860 to predict mortality.

   In the future, the next version of the Aristotle Score, named Aristotle 2, will be based on objective data provided by the new STAT Mortality Score and STAT Morbidity Score and will include an updated technical difficulty index re-calculated based on expert opinion. New basic performances for mortality, morbidity and technical difficulty will be proposed. The ***Aristotle Comprehensive Complexity Score*** will be updated and simplified and involve only around 70 procedures, including procedures performed on adults with congenital heart disease. New comprehensive performances will be proposed. The Aristotle 2 score will be ready in 2015. Once validated, it should provide a fair assessment for evaluation of performances in congenital heart surgery. Furthermore, the Aristotle Score is responsible for multiple important contributions leading to the development of newer tools to evaluate cardiac surgical performance.

Keywords

Complexity • Congenital Heart Surgery • Quality of care • Performance • Aristotle Score • Aristotle Basic Complexity Score (ABC Score) • Aristotle Comprehensive Complexity Score (ACC Score)

According to Aristotle's philosophy (Rhetoric, Book I, 350 BC):
When there is no scientific answer available, the opinion (Doxa) perceived and admitted by the majority has value of truth.

Introduction

Evaluation of quality of care in congenital cardiac surgery is challenging. Compared to adult cardiac surgery, congenital cardiac surgery covers a smaller pediatric population and deals with many times more different diagnoses and procedures. This challenge explains the delay needed in our specialty to establish professional databases in the United States of America and in Europe.

The take-off of the Congenital Heart Surgery Database of The Society of Thoracic Surgeons (STS) and The European Association for Cardio-Thoracic Surgery (EACTS) occurred following the creation of the International Congenital Heart Surgery Nomenclature and Database Project led by Constantine Mavroudis, MD and Jeffrey P. Jacobs, MD and published on The Annals of Thoracic Surgery in 2000 [1]. The other obstacle was that outcome was only based on hospital

mortality without any risk stratification or adjustment for case-mix. As a consequence, the prominent centers dealing with the most complex cases and having a greater mortality were very reluctant to send their data. The creation of the Aristotle Score [2–6], based on the International Congenital Heart Surgery Nomenclature and Database Project, contributed to the full growth of the STS and EACTS Congenital Heart Surgery Databases [7].

The Aristotle Complexity Score project started in 2002, [2] and was published in 2004 [3, 6]. The objective of the Aristotle Score is to measure performance and to allow fair and meaningful comparison between centers and surgeons. The Aristotle Score is not specifically designed to predict mortality.

This chapter will focus on the role of the Aristotle Score in evaluating performance:

- First, this chapter will describe what was accomplished in the last decade with the Aristotle Score and the STS and EACTS Congenital Heart Surgery Databases.
- Second, this chapter will present the plans for the development of the next generation of the Aristotle Score.
- Third, this chapter will present the evolution of the Aristotle Score and its contributions towards the development of newer tools to evaluate cardiac surgical performance.

Current Aristotle Score

Definition of Performance

Performance in congenital cardiac surgery is a "proteiform" concept. The outcomes of surgery depends, in most cases, not solely on the surgeon but mainly on the performance of the entire team [8]:

- the pediatric cardiologist who insure an accurate diagnosis,
- the operative team including the surgeon and also the anesthetist and the perfusionist and
- the team in the intensive care unit.

We believe that several performances should be studied and analyzed separately, depending on the outcome considered. Since the inception of analysis of outcomes of pediatric and congenital cardiac care, quality has been essentially measured based on operative mortality. Although assessment of mortality is essential, it is insufficient as it involves only around 4 % of the patients and therefore 96 % are excluded from the assessment. Instead of a assessing the unique and singular performance of mortality, the Aristotle Score is trying to evaluate all the aspect of congenital cardiac surgery, as shown in Table 27.1.

Table 27.1 Six performances can be defined according to outcomes

Outcome	Performance
Mortality	Safety
Morbidity	Efficiency
Technical difficulty	Proficiency
Long-term results	Quality
Patient satisfaction	Reputation
Cost	Economical performance

The Aristotle Score focuses only on the first three performances: safety, efficiency and proficiency. The other three are equally important

The Aristotle Score has focused on the first three performances documented in Table 27.1, based on mortality, morbidity and technical difficulty. Long term results, patient satisfaction and hospital cost are equally important but are not included in the Aristotle Score.

The Concept of Complexity

The Aristotle score propose a new and original approach to evaluate quality based on complexity. It is important to consider that complexity is different from risk [2–6]. The risk of mortality and morbidity of a Norwood operation is less in a large center with optimal experience and greater in a small center still confronted to a learning curve.

- *Risk is a variable factor and varies from center to center and even from surgeon to surgeon*
- *Complexity is designed to be a constant. Complexity is a calculated value based* on the following algorithm that is evaluated for each procedure:

$$\begin{aligned} \textit{Complexity} = {} & \textit{Potential for Mortality} \\ & + \textit{Potential for Morbidity} \\ & + \textit{Technical Difficulty} \end{aligned}$$

Initially [3], *the potentials* for mortality and morbidity were calculated by expert opinion and were subjective. Recently, the methodology for determining the first two factors, potential for mortality and potential for morbidity, has transitioned from subjective probability (expert opinion) to determination based on raw data from databases, and is therefore objective. The methodology to estimate the third factor, technical difficulty, remains subjective, but might approach more objectivity in the future in using the Technical Performance Score developed in Boston under the leadership of Emile Bacha [9–12].

The calculation of complexity using the Aristotle Score is done in *two steps*.

- *The first step gives the* ***Aristotle Basic Complexity Score*** (**ABC Score**) (Fig. 27.1). The ***Aristotle Basic Complexity Score*** is a simplified score that is calculated on 15 points and is determined from the primary procedure of any operation. The Aristotle Basic Complexity Score was ultimately also divided in four levels of complexity. The Aristotle Basic Complexity Score can be analyzed using these four categories, which are known as the ***Aristotle Basic Complexity Levels***. The ***Aristotle Basic Complexity Score*** and the ***Aristotle Basic Complexity Levels*** were introduced in the Congenital Heart Surgery Database of The Society of Thoracic Surgeons (STS) and The European Association for Cardio-Thoracic Surgery (EACTS) in 2002 and have been instrumental for the success of these congenital databases. The accuracy of the ***Aristotle Basic Complexity Score*** and the ***Aristotle Basic Complexity Levels*** are limited because of wide variations in complexity within a given procedure such as the Norwood (Stage 1) operation.
- *The second step is the* ***Aristotle Comprehensive Complexity Score*** (ACC Score) (Fig. 27.1) that increases the potential for mortality, the potential for morbidity, and technical difficulty by adding procedure dependent factors and procedure independent factors. The Aristotle Comprehensive Complexity Score is calculated on 25 points: The Aristotle Comprehensive Complexity

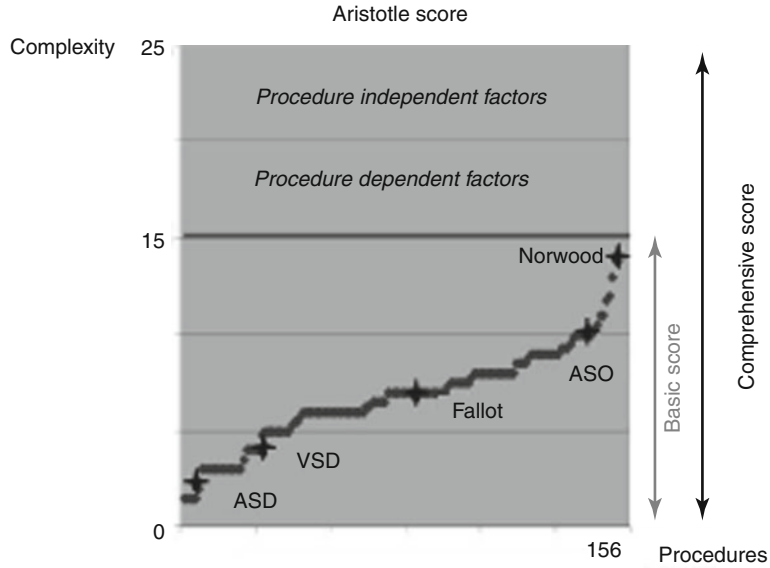

Fig. 27.1 Aristotle Score. The Aristotle Basic Complexity Score is calculated on 15 points and is determined from the primary procedure of any operation. The Aristotle Comprehensive Complexity Score equals the Aristotle Basic Complexity Score plus 5 points for Procedure-dependent Factors and 5 points for Procedure-independent Factors. Therefore, the Aristotle Comprehensive Complexity Score is calculated on 25 points (www.thearistotleinstitute.org)

Score equals the Aristotle Basic Complexity Score plus 5 points for ***Procedure-dependent Factors*** and 5 points for ***Procedure-independent Factors***. Procedure-dependent factors include anatomical factors, associated procedures, and age at procedure, and procedure independent factors include general factors, clinical factors, extracardiac factors, and surgical factors. Each factor is scored for contribution to mortality, morbidity, and technical difficulty. All complexity factors meet the following requirements: precisely quantifiable, easily available, admitted by a majority, and verifiable. These additional complexity factors (procedure dependent factors and procedure independent factors) are based on subjective opinions and will remain so for several years until the databases produce an accurate risk-stratification for each procedure. The *Aristotle Comprehensive Complexity Score* is available on the Aristotle website: [www.thearistotleinstitute.org]. The *Aristotle Comprehensive Complexity Score* has not been used so far in the STS Congenital Heart Surgery Database and the EACTS Congenital Heart Surgery Database. Multiple individual institutional studies have documented the utility of the *Aristotle Comprehensive Complexity Score* [13, 14] (see below in validation). Perhaps the most important multi-institutional contribution of the *Aristotle Comprehensive Complexity Score* to date has been that its components have been incorporated into the STS Congenital Heart Surgery Database and the EACTS Congenital Heart Surgery Database and have been used to inform the upgrade of these databases.

It is obvious that a relationship exists between complexity, outcome, and performance. We have proposed a simple equation to measure performance:

Performance = Complexity × Survival

To summarize, there are five principles behind the complexity concept:

1. ***The Aristotle Score is a tool to evaluate performance and not directly to predict mortality***
2. ***Complexity is a constant***
3. ***Complexity = Sum of Potential for Mortality + Potential for Morbidity + Technical Difficulty***
4. ***The true calculated complexity includes both the Aristotle Basic Complexity Score and the Aristotle Comprehensive Complexity Score***
5. ***Performance = Complexity × Outcome***

Why Technical Difficulty?

In sports, the concept of complexity is widely used. The complexity of ski slopes is defined by colors. In gymnastics [15], diving, and figure skating, the activities attempted by the athletes are ranked according to a complexity score established by the judges. An athlete performing a low complex activity cannot obtain the maximum score.

Two main reasons support the inclusion of technical difficulty in the evaluation of performance:

First, it has been quite a surprise to observe on raw data from the STS Congenital Heart Surgery Database and the EACTS Congenital Heart Surgery Database that several complex procedures were performed with extremely low mortality, such as the arterial switch for transposition of the great arteries with intact ventricular septum and the Ross procedure [16]. Within a system of risk stratification that is exclusively based today on mortality, these procedures are considered as average difficulty. The reality however, is that most of those complex procedures are only performed by senior experienced surgeons. The addition of the component of technical difficulty allows one to evaluate the complexity fairly, and therefore evaluate the performance of the surgeon fairly.

Furthermore, the technical difficulty of a given procedure is not constant. An arterial switch

operation in a patient with an intramural coronary artery is more challenging than with in a patient with the usual pattern of coronary arteries. The technical difficulty in the *Aristotle Comprehensive Complexity Score* integrates these anatomical variations and provides more a fair assessment.

The second reason supporting the inclusion of technical difficulty in the evaluation of performance is that the current evaluation of quality is able to say "If we do things right" but is unable to say "If we do the right things". For example, the ongoing controversy involving the management of patients with hypoplastic left heart syndrome involves uncertainty as to whether the Norwood (Stage 1) Operation is the best option for all patients or whether some patients will benefit from a less complex operation: the Hybrid Stage One. Many other examples exist where some surgeons prefer to choose a simple procedure, which may not be optimal for the long term results: Fontan versus repair of complex intracardiac repair for patients with complex double outlet right ventricle with remote ventricular septal defect, and even or mitral valve replacement versus mitral valve repair. We advocate that surgeons should perform the "right" operation, even if this operation is more demanding.

Methodology of the Aristotle Score

Methodology of the Aristotle Basic Complexity Score and Aristotle Basic Complexity Level

The Aristotle methodology to facilitate complexity adjustment is based upon the work of the Aristotle Committee. Starting in 2002, the STS Congenital Heart Surgery Database and the EACTS Congenital Heart Surgery Database incorporated the Aristotle Basic Complexity Score and Aristotle Basic Complexity Level in their analysis of discharge mortality analyses [17]. These complexity scores and levels can be reported by year, center, age group, and procedure. The complexity analysis represents a basic complexity adjustment method to evaluate surgical results (Complexity is a constant precise value for a given patient at a given point in time; performance varies between centers and surgeons. In other words, in the same exact patient with the same exact pathology, complexity is a constant precise value for that given patient at a given point in time. The risk for that patient will vary between centers and surgeons because performance varies between centers and surgeons.).

The Aristotle complexity scoring was based on the primary procedure of a given operation as defined by the short list of procedures of the EACTS-STS International Nomenclature [1] and was evaluated in two steps. The first step was the Aristotle Basic Complexity Score, defining, basically, the complexity through three factors: the potential for mortality, the potential for morbidity, and the technical difficulty of the operation, using a questionnaire filled out by 50 surgeons representing international centers. Only the Aristotle Basic Complexity Score (1.5–15) and Aristotle Basic Complexity Level (four levels: 1–4) are used in the STS Congenital Heart Surgery Database and the EACTS Congenital Heart Surgery Database (Appendix 1).

The Aristotle Basic Complexity Score is created from a survey of all 50 of the Aristotle project congenital surgeon participants. Participants were asked to rank all procedures from the EACTS-STS Minimal Database Procedure Short List [1]. Each procedure was scored with a score of 0.5–5 in three areas: potential for mortality, potential for morbidity, and technical difficulty. Guidelines were provided to the Aristotle project participants. Five levels of suggested scoring were provided for each of these three areas, with each suggested level worth 1 point: potential for mortality (less than 1, 1–5 %, 5–10 %,

10–20 %, and greater than 20 %), potential for morbidity (based on estimated intensive care unit [ICU] stay: 0–24 h, 1–3 days, 4–7 days, 1–2 weeks, and greater than 2 weeks), and technical difficulty (elementary, simple, average, important, and major). The points (0.5–5) from each of these three areas were added together to give a total of 1.5–15. For each procedure, the median value of mortality, morbidity, and technical difficulty obtained from the 50 centers was calculated. The sum of these three median values gives the final Aristotle Basic Complexity Score for each procedure (Appendix 1). The distribution of the scoring among the centers was, in general, quite uniform, although some rare or new procedures had a large dispersion.

In addition to assigning each procedure an Aristotle Basic Complexity Score ranging from 1.5 to 15, each procedure was next assigned an Aristotle Basic Complexity Level ranging from 1 through 4 based on the Aristotle Basic Complexity Score (basic score of 1.5–5.9 = basic level of 1, basic score of 6.0–7.9 = basic level of 2, basic score of 8.0–9.9 = basic level of 3, and basic score of 10.0–15.0 = basic level of 4).

In the initial application of the Aristotle Basic Complexity Score in the STS Congenital Heart Surgery Database and the EACTS Congenital Heart Surgery Database [17], 145 procedures from the EACTS-STS procedure short list were scored and 29 procedures were in level 1, 46 procedures were in level 2, 45 procedures were in level 3, and 25 procedures were in level 4. Since this initial application, additional procedures have been added to the nomenclature and have been assigned Aristotle Basic Complexity Scores and Aristotle Basic Complexity Level. The Aristotle Basic Complexity Level provides a broad generalization of complexity by dividing surgical procedures into four complexity categories. Meanwhile, the Aristotle Basic Complexity Score can provide more precise complexity stratification. Both the score and the level are useful tools; the appropriate tool can be chosen to match the required analysis.

Methodology of the Aristotle Comprehensive Complexity Score

The Aristotle Comprehensive Complexity Scores add two sorts of complexity modifiers: procedure-dependent factors (including anatomical factors, associated procedures, and age at procedure) and procedure-independent factors (including general factors, clinical factors, extracardiac factors, and surgical factors). Each factor is scored for contribution to mortality, morbidity, and technical difficulty. All complexity factors meet the following requirements: precisely quantifiable, easily available, admitted by a majority, and verifiable. The Aristotle Committee is currently involved in ongoing research to validate this complexity adjustment scoring system on a multi-institutional basis.

Results of the Aristotle Score

The Aristotle Basic Complexity Score was first introduced in the STS Congenital Heart Surgery Database and the EACTS Congenital Heart Surgery Database in 2002 [17] and allowed original risk stratification with production of quite useful graphs to evaluate performance.

Figure 27.2 is a Bubble Chart that displays the outcomes of 42 institutions and 12,576 patients [reproduced with permission from Bohdan Maruszewski, MD, Chair of the EACTS Congenital Heart Surgery Database, EACTS 2004 database annual report]. The Aristotle Basic Complexity Score is plotted against Mortality combined. Each bubble represents a different center, and the size of the bubble correlates with programmatic volume. The graph allows one to define 4 quadrants, based on averages: *the best performing centers are in the lower right quad-*

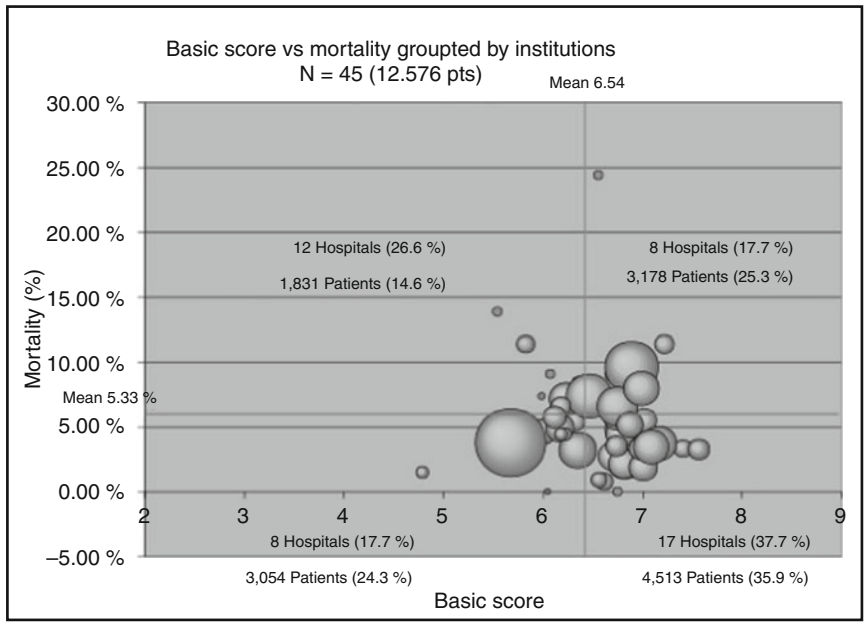

Fig. 27.2 This Bubble Chart displays the outcomes of 42 institutions and 12,576 patients. The Aristotle Basic Complexity Score is plotted against Mortality. Each bubble represents a different center, and the size of the bubble correlates with programmatic volume. Bubbles of different size represent the volume of centers. The graph allows one to define 4 quadrants, based on averages: the best performing centers are in the lower right quadrant, with lower mortality and higher complexity. Notice that the best performing centers are not always the largest ones (Reproduced with permission from Bohdan Maruszewski, MD, Chair of the EACTS Congenital Heart Surgery Database, EACTS 2004 database annual report)

rant, with higher complexity and lower mortality. Notice that the best performing centers are not always largest ones.

Figure 27.3 is a Bubble Chart that displays the outcomes of 226 surgeons [reproduced with permission from Bohdan Maruszewski, MD, Chair of the EACTS Congenital Heart Surgery Database, EACTS 2004 database annual report]. The Aristotle Basic Complexity Score is plotted against Mortality combined. Each bubble represents a different surgeon, and the size of the bubble correlates with the volume of cases performed by the individual surgeon. The graph also allows one to define 4 quadrants, based on averages: *the best performing surgeons are in the lower right quadrant*, with higher complexity and lower mortality.

The ***Aristotle Basic Complexity Score*** is calculated on 15 points and is determined from the primary procedure of any operation. The Aristotle Basic Complexity Score was ultimately divided in four levels of complexity. The Aristotle Basic Complexity Score can be analyzed using these four categories, which are known as the ***Aristotle Basic Complexity Levels*** (Fig. 27.4). Meanwhile, the ***Risk Adjustment in Congenital Heart Surgery*** (***RACHS-1***) has six categories (Fig. 27.4) [18]. Figure 27.4 displays the increment of mortality as the Aristotle Basic Complexity Levels and RACHS-1. The results are very similar, showing a good discrimination for the two systems. The ***Aristotle Basic Complexity Score*** includes 94 % of operations while the RACHS-1 includes 86 % [19].

RACHS-1 Category 5 is quite small, composed of patients who undergo combined repair of Truncus arteriosus and Interrupted aortic arch

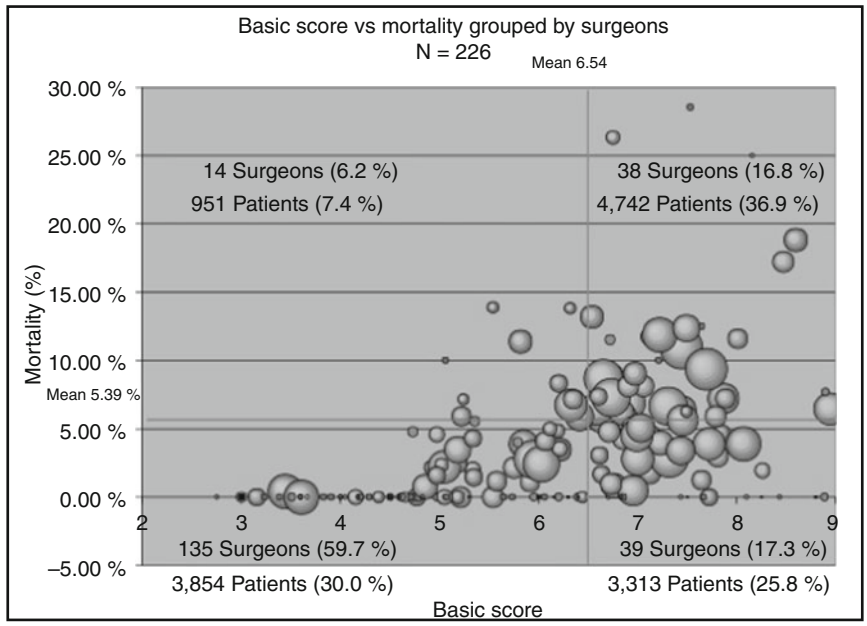

Fig. 27.3 This Bubble Chart displays the outcomes of 226 surgeons. The Aristotle Basic Complexity Score is plotted against Mortality. Each bubble represents a different surgeon, and the size of the bubble correlates with the volume of cases performed by the individual surgeon. The graph also allows one to define 4 quadrants, based on averages: the best performing surgeons are in the lower right quadrant, with lower mortality and higher complexity (Reproduced with permission from Bohdan Maruszewski, MD, Chair of the EACTS Congenital Heart Surgery Database, EACTS 2004 database annual report)

repair. Therefore, at the analytic level, RACHS-1 Category 5 is usually combined with RACHS-1 Category 6, which includes:

- Damus-Kaye-Stansel procedure (DKS) (creation of AP anastomosis without arch reconstruction)
- Hybrid Approach "Stage 2", Aortopulmonary amalgamation + Superior Cavopulmonary anastomosis(es) + PA Debanding + Aortic arch repair (Norwood [Stage 1] + SuperiorCavopulmonaryanastomosis(es) + PA Debanding)
- Norwood procedure

The exclusivity of the combined RACHS-1 Category 5 and RACHS-1 Category 6 allows for enhanced discrimination for prediction of mortality using RACHS-1. It is important to remember that the Aristotle Score was initially designed to measure performance and not to predict mortality; however, Aristotle Basic Complexity Score actually does also quite well with prediction of mortality [20].

The results of the *Aristotle Comprehensive Complexity Score* are encouraging, but have been so far only been published by individual institutions and not by the STS Congenital Heart Surgery Database and the EACTS Congenital Heart Surgery Database (Perhaps the most important multi-institutional contribution of the *Aristotle Comprehensive Complexity Score* to date has been that its components have been incorporated into the STS Congenital Heart Surgery Database and the EACTS Congenital Heart Surgery Database and have been used to inform the upgrade of these databases.). As expected, these individual institutional analyses have documented that the discrimination of the *Aristotle Comprehensive Complexity Score* is superior. The reason for

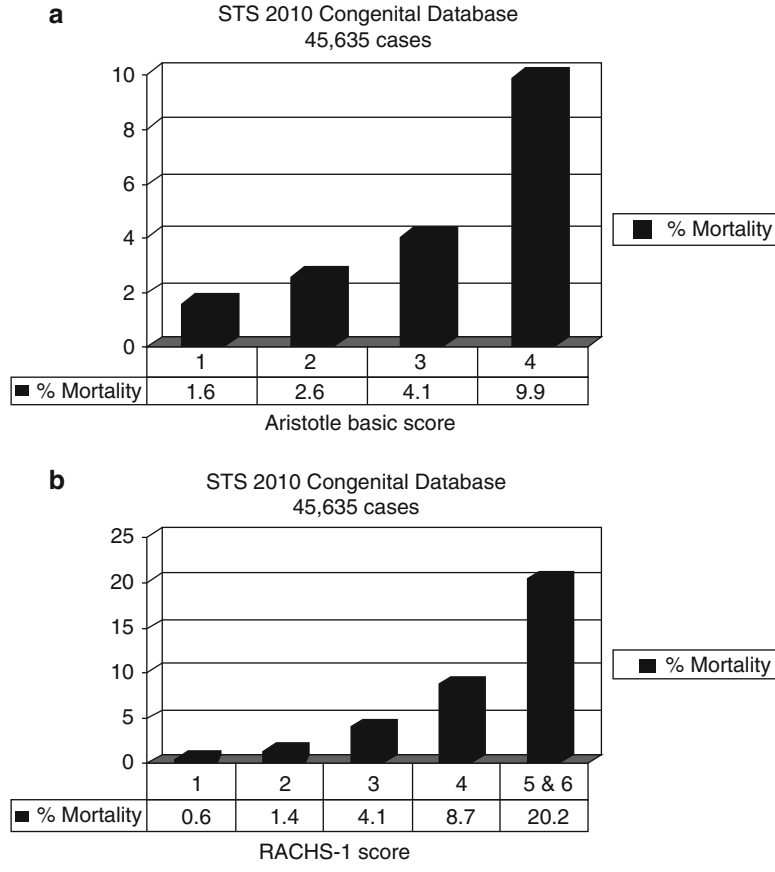

Fig. 27.4 (**a**) Aristotle Basic Complexity Score and (**b**) RACHS-1 Categories versus mortality, by categories (four categories for Aristotle Basic Complexity Score and five categories for RACHS-1)

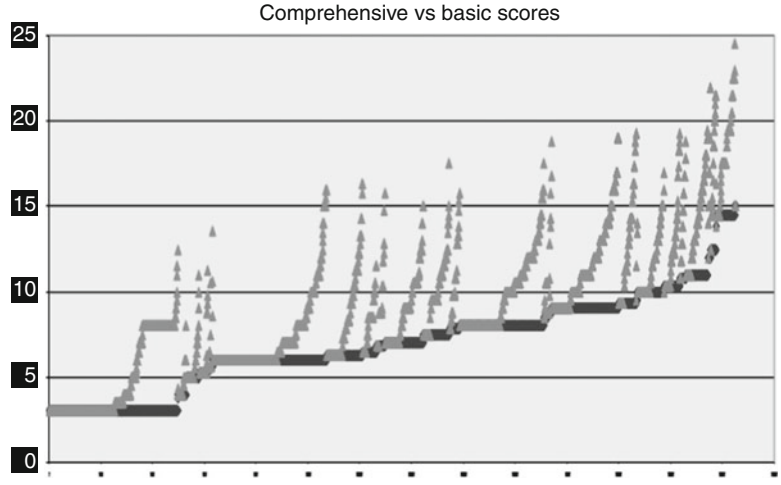

Fig. 27.5 Figure plots the Aristotle Basic Complexity Score (x-axis) versus the Aristotle Comprehensive Complexity Score (y-axis), in an analysis of 2,655 operations. Figure shows the incremental increased complexity documented in many operations due to the introduction of Procedure Dependent Factors and Procedure Independent Factors. Notice the wide increase in complexity for many operations following the addition of procedure dependent factors and procedure independent factors

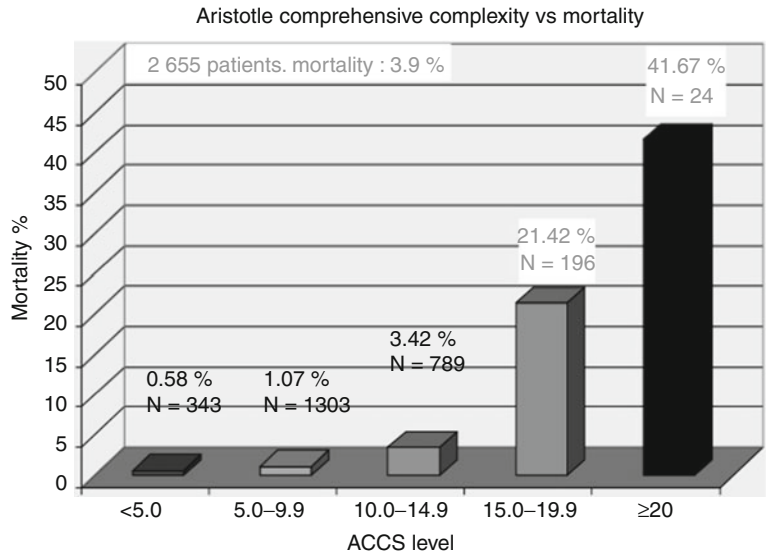

Fig. 27.6 Figure plots Aristotle Comprehensive Complexity Score (ACCS Level) versus Mortality. Notice the very severe observed mortality of 21.4 and 41.7 % when the Aristotle Comprehensive Complexity Score is beyond 15 and 20, respectively

Table 27.2 Evolution of performance within the EACTS Congenital Heart Surgery Database

Year	Performance	Mean BS	Mean MS	No. of patients	30 days mortality (%)
2012	6.71	6.93	0.68	13,870 patients	3.17
2011	6.74	6.96	0.67	14,999 patients	3.22
2010	6.7	6.92	0.65	15,312 patients	3.21
2009	6.74	7.01	0.68	12,676 patients	3.76
2008	6.77	7.05	0.7	12,012 patients	4.00
2007	6.87	7.14	0.7	11,050 patients	3.71

Reproduced with permission from Bohdan Maruszewski, MD, Chair of the EACTS Congenital Heart Surgery Database (http://www.eactscongenitaldb.org)
BS basic score, *MS* mortality score

this superior performance is that the *Aristotle Comprehensive Complexity Score* is calculated based on 25 points, with the addition of many modifiers (Procedure Dependent Factors and Procedure Independent Factors) that improve its accuracy. Figures 27.5 and 27.6 are produced from the Aristotle Comprehensive Complexity Score Study Committee, [www.aristotleinstitute.org] and include 2,655 operations from 12 centers.

The introduction of the performance equation has been a source of controversy:

$$Performance = Complexity \times Outcome$$

The EACTS [http://www.eactscongenitaldb.org] have used this equation to measure performance. Table 27.2 shows the stagnation of performance at the EACTS Congenital Heart Surgery Database, while the mortality decreases. We assume that this finding is a consequence of the decreased number of Norwood operations performed at centers participating in the EACTS Congenital Heart Surgery Database. This trend may be related to the development of the prenatal diagnosis.

Validation of the Aristotle Score

The validation of the Aristotle Basic Complexity Score was studied [20] using data from the EACTS Congenital Heart Surgery Database (17,838 operations, 56 centers) and the STS

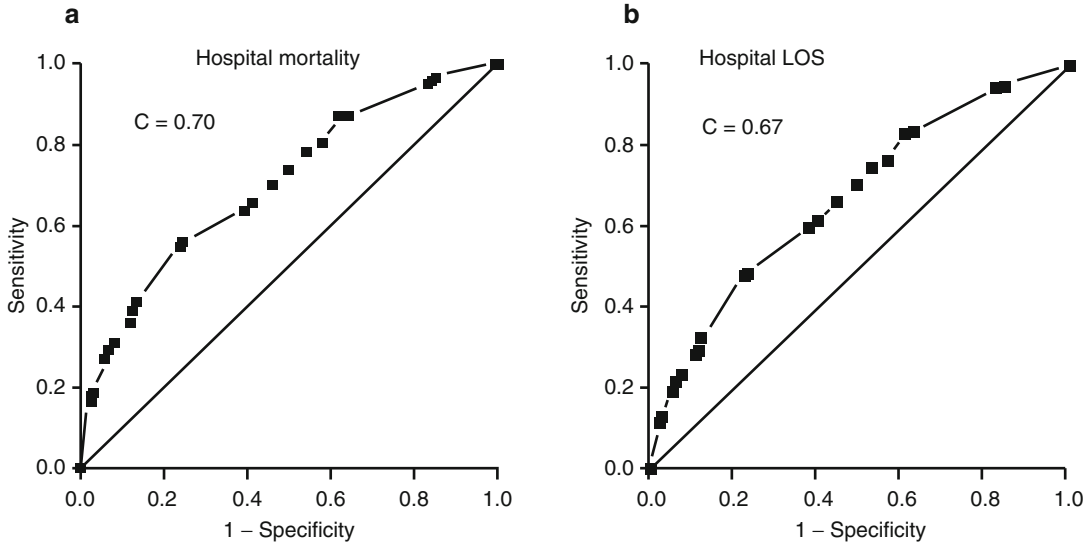

Fig. 27.7 Validation of the Aristotle Basic Complexity Score [20]. C-Index of 0.70 for prediction of mortality (**a**) and 0.67 for prediction of Hospital Length of Stay (*LOS*) (**b**)

Congenital Heart Surgery Database (18,024 operations, 32 centers). "Discrimination of the ABC score for predicting in-hospital mortality and postoperative length of stay (PLOS) of more than 21 days was quantified by the C statistic. Procedure-specific rates of mortality and prolonged PLOS were compared with predictions from a logistic regression model, and an exact binomial test was used to identify procedures that were mortality and morbidity outliers" [20]. This analysis revealed that a significant positive correlation exists between the Aristotle Basic Complexity Score of a procedure and its observed procedure-specific risk of mortality (C = 0.70) and prolonged PLOS (C = 0.67) (Fig. 27.7). It was concluded that the Aristotle Basic Complexity Score [20] "generally discriminates between low-risk and high-risk congenital procedures making it a potentially useful covariate for case-mix adjustment in congenital heart surgery outcomes analysis. Planned revisions of the ABC score will incorporate empirical data and will benefit from the large sample sizes of the STS and EACTS databases."

The validation of the Aristotle Comprehensive Complexity Score was achieved by several individual institutions and the *Aristotle Comprehensive Complexity Score* was used to study and compare outcomes of several complex procedures:

- The Aristotle Comprehensive Complexity Score was proposed in Germany as a reference for "pay for performance" and hospital reimbursement [21–28]
- For patients undergoing the Norwood (Stage 1) Operation, the Aristotle Comprehensive Complexity Score was found to be correlated with total cardiac output during the early postoperative period [29].
- In patients undergoing surgery for hypoplastic left heart syndrome, the Aristotle Comprehensive Complexity Score was correlated with survival [30, 31]
- In low weight patients placed on cardiopulmonary bypass, the Aristotle Comprehensive Complexity Score was correlated with survival [32].
- In post-operative extracorporeal membrane oxygenation (ECMO), the Aristotle Comprehensive Complexity Score was correlated with survival [33].
- In patients undergoing surgery for Truncus Arteriosus with Interrupted Aortic Arch, the Aristotle Comprehensive Complexity Score was correlated with survival [34].

- The Aristotle Comprehensive Complexity Score was used to evaluate the progress of an institution [35]. "A high correlation was found between the ACC scores and mortality, indices of morbidity and technique difficulty, Spearman's correlation coefficient r being 0.9856, 1 and 0.9429, respectively. Mortality ($p=0.037$) and morbidity ($p=0.041$) were lower in year 2007 than in 2002, surgical performance being not significantly different."
- In patients undergoing the arterial switch operation, the Aristotle Comprehensive Complexity Score was correlated with morbidity [36] and to mortality [37].
- The Aristotle Comprehensive Complexity Score was associated with the hospital length of stay in neonatal congenital cardiac surgery [38].
- In adults with congenital cardiac disease [39], the *Aristotle Comprehensive Complexity Score* was correlated with 30 day mortality with a C-Index of 0.755.
- The *Aristotle Score* was used to evaluate the impact of surgical volume on outcomes [40].
- The *Aristotle Comprehensive Complexity Score* was evaluated at the Necker Children's Hospital in Paris on a cohort of 1,454 patients. The *Aristotle Comprehensive Complexity Score* was strongly related to mortality, with a C-Index of 0.86 [41].

The *Aristotle Comprehensive Complexity Score* compares favorably with all the existing models of complexity stratification, with a C-Index of 0.860, as shown in Table 27.3.

Table 27.3 Comparison of risk stratification models, for prediction of mortality. The Aristotle Comprehensive Complexity Score has the highest score C-Statistic [20]

Risk stratification modeling	C-Index
Aristotle Comprehensive Complexity Score[a]	**0860**
STAT Mortality Score[b]	0816
STAT Mortality Categories[b]	0812
RACHS-1 Categories[b]	0802
Aristotle Basic Complexity Score[b]	0795

[a]O'Brien et al. [20]
[b]Artrip et al. [30]

Limitation of the Current Aristotle Scores

Complexity scores can incorporate only a finite number of known factors [17]. True complexity is related to both these known factors and other factors we may not know or measure. Although complexity itself is a constant precise value for a given patient at a given point in time, the Aristotle Basic Complexity Score, the Aristotle Basic Complexity Level, and the Aristotle Comprehensive Complexity Score all represent estimates to measure complexity.

Two Aristotle Scores exist: The *Aristotle Basic Complexity Score* and the *Aristotle Comprehensive Complexity Score*. Since its introduction [3], it was made clear that the *Aristotle Comprehensive Complexity Score* will be a more accurate score, not only to predict mortality but moreover to measure performance.

The expert opinion based system was initially judged inappropriate [42, 43]. In reality, the expert opinion is used in many disciplines and is objectively manageable using Bayesian statistics [16, 20, 44, 45]. When we started the new versions of the STS and EACTS Congenital Heart Surgery Databases in 2000 using the new international nomenclature based on the International Congenital Heart Surgery Nomenclature and Database Project, data about risk-stratification was not available. The only option was first to create a system of risk stratification based on expert opinion. The Aristotle Basic Complexity Score has been very instrumental to facilitate the growth of the STS Congenital Heart Surgery Database and the EACTS Congenital Heart Surgery Database: the most active centers are no longer reluctant to send their data, as a higher mortality was supported by a higher complexity. The transition and ultimate switch to a system of risk stratification based on raw, observed, objective data [16, 45] was ultimately made when enough data was accumulated in the STS Congenital Heart Surgery Database and the EACTS Congenital Heart Surgery Database (see below).

The combination of the three variables (potential for mortality, potential for morbidity, and

technical difficulty) in the Aristotle system has limited the power to predict mortality as it is also looking at two additional outcomes (potential for morbidity and technical difficulty). Nevertheless, it is noticeable that the Aristotle Comprehensive Complexity Score has today the best correlation to predict mortality (Table 27.3) [41].

To date, the Aristotle Comprehensive Complexity Score has not been introduced in the STS Congenital Heart Surgery Database and the EACTS Congenital Heart Surgery Database because it was felt that a scientific evaluation was required, which, we believe, it has since gained with many publications. However, the Aristotle Comprehensive Complexity Score is judged complicated and not user friendly. Meanwhile, the mortality in our specialty is today essentially limited to very complex patients who accumulate multiple risk factors that are not all included in any database. Using a more accurate system of system of risk stratification seems necessary to explain the cause of mortality and morbidity of the most complex patients, namely "those who die". Perhaps the most important multi-institutional contribution of the Aristotle Comprehensive Complexity Score to date has been that its components have been incorporated into the STS Congenital Heart Surgery Database and the EACTS Congenital Heart Surgery Database and have been used to inform the upgrade of these databases, with the goal of increasing the accuracy of the methodology of risk stratification.

The treatment of adults with congenital cardiac disease has become a major component of our specialty. The Aristotle Comprehensive Complexity Score has been applied quite successfully to analyze the surgical outcomes of adults with congenital cardiac disease [39]. Nevertheless, it is insufficient. The *Aristotle Comprehensive Complexity Score* for surgery in adults with congenital cardiac disease will include in the future many risk factors specific to the adult patients.

Aware of the current limitations of the Aristotle Score [38], The Aristotle Committee is today developing an Aristotle 2 model that is summarized below. The first step is to incorporate the objectively derived STAT Mortality Score and the STAT Morbidity Score.

New Aristotle 2 Scores

From Subjective to Objective Evaluation: The STAT Mortality Score and the STAT Morbidity Score

The STAT Mortality Score was introduced in 2009 [16]. *The STAT Mortality Score* is a mortality score or mortality index. Mortality risk was estimated for 148 types of operative procedures using data from 77,294 operations entered into the EACTS Congenital Heart Surgery Database (33,360 operations) and the STS Congenital Heart Surgery Database (43,934 patients) between 2002 and 2007. Procedure-specific mortality rate estimates were calculated using a Bayesian model that adjusted for small denominators. Each procedure was assigned a numeric score (the STS–EACTS Congenital Heart Surgery Mortality Score [2009] or STAT Mortality Score) ranging from 0.1 to 5.0 based on the estimated mortality rate. Procedures were also sorted by increasing risk and grouped into five categories (the STS–EACTS Congenital Heart Surgery Mortality Categories [2009] or STAT Mortality Categories) that were chosen to be optimal with respect to minimizing within-category variation and maximizing between-category variation. The STAT Mortality Score and STAT Mortality Categories could predict mortality with a C-index for the score and the categories of 0.784 and 0.773, respectively. (It was noticed that the Aristotle Basic Complexity Score, based on expert opinion, had over-evaluated the risk of mortality for several procedures.)

The STAT Morbidity Score followed in 2013 [45], after a very long debate around its definition. The STAT Morbidity Score was created because it was felt very necessary to evaluate the morbidity of the 96 % of patients surviving the operation; whose quality assessment was ignored in a system based only on mortality. The STAT Morbidity Score was developed using data from 62,851 operations in the STS Congenital Heart

Surgery Database (2002–2008). Model-based estimates with 95 %Bayesian credible intervals were calculated for each procedure's average risk of major complications and average postoperative length of stay. These 2 measures were combined into a composite morbidity score. A total of 140 procedures were assigned scores ranging from 0.1 to 5.0 and sorted into five relatively homogeneous categories. It is expected that the impact of the STAT Morbidity Score will be very important to compare hospital cost between centers in the future.

The STAT Mortality Score and the STAT Morbidity Score are objectively derived and may replace the subjectively derived "potential for mortality" and "potential for morbidity" in the Aristotle Score. *The technical difficulty component of the Aristotle Score* remains today based on expert opinion. In 2009, the Congenital Data Base Committee produced a new ranking of technical difficulty based on 148 procedures. Procedures with highest technical difficulty are listed on Table 27.4.

Table 27.4 Technical difficulty ranking of the 15 most technically demanding procedures

Technical difficulty ranking	Procedures
1	Congenitally corrected TGA repair, Atrial switch and ASO (Double switch)
2	Norwood procedure
3	Ross-Konno procedure
4	HLHS biventricular repair
5	Arterial switch procedure and VSD repair + Aortic arch repair
6	Fontan revision or conversion (Re-do Fontan)
7	Aortic root replacement, Valve sparing
8	Transplant, Heart and lung
9	Truncus + IAA Repair
10	Congenitally corrected TGA repair, Atrial switch and Rastelli
11	Arterial switch operation (ASO) and VSD repair
12	Pulmonary atresia – VSD – MAPCA (pseudotruncus) repair
13	Arterial switch procedure + Aortic arch repair
14	Congenitally corrected TGA repair, VSD closure and LV to PA conduit
15	Truncus arteriosus repair

Timing

The development of the Aristotle 2 score will start when the STS and EACTS have accumulated sufficient data on the Morbidity Score. It is expected that the new score will be *available in 2015*.

Adults with Congenital Cardiac Disease

A score for adults with congenital cardiac disease will be developed using new adult specific procedure independent factors required for the adult population [39]

Simplification of the Aristotle Scores

The Aristotle Basic Complexity Score 2 (ABS2) will be calculated, using the objective STAT Mortality Score and STAT Morbidity Score are. The technical Difficulty will remain based on expert opinion. A new Technical Difficulty may eventually be produced, incorporating the Surgical Performance data available from the Technical Performance Score developed in Boston under the leadership of Emile Bacha [9–12].

The Aristotle Basic Complexity Score 2 will be calculated with the following equation:

$$\begin{aligned} \textit{Complexity} = {} & \textit{STATM ortality Score} \\ & + \textit{STAT Morbidity Score} \\ & + \textit{Technical Difficulty} \end{aligned}$$

The Aristotle Comprehensive Complexity Score 2 (ACS2) will remain based again on expert opinion. An international committee of expert centers, surgeons, and intensivists will insure the definition of the procedure-dependent factors and procedure-independent factors [46, 47].

The Aristotle Comprehensive Complexity Score 2 will be *reduced to 70 procedures* out of

180, by selecting the 20 most frequent procedures (with the exception of "PDA closure") and the procedures for most complex pathologies. The nine pathologies studied in the Lesion Specific Section of the STS Congenital Heart Surgery Database Feedback Report will be included:

1. **Atrial Septal Defect** (**ASD**)
2. **Ventricular Septal Defect** (**VSD**)
3. **Coarctation of the Aorta** (**COA**)
4. **Tetralogy of Fallot** (**TOF**) **Palliation**
5. **Tetralogy of Fallot** (**TOF**) **Repair**
6. **AV Canal** (**AVC**) **Defect**
7. **Aortic Stenosis and Insufficiency** (**ASAI**)
8. **Transposition of the Great Arteries** (**TGA**)
9. **Hypoplastic Left Heart Syndrome** (**HLHS**) (**Norwood procedure**, **Damus–Kaye–Stansel procedure** [**DKS**] **and Hybrid procedures**)

The Aristotle Comprehensive Complexity Score 2 will also include 11 other pathologies and cardiac transplantation:

1. **Adults with Congenital Cardiac Disease**,
2. **Truncus Arteriosus**,
3. **Corrected Transposition**,
4. **Double Outlet Right Ventricle** (**DORV**),
5. **Interrupted Aortic Arch** (**IAA**),
6. **Abnormal origins of coronary arteries**,
7. **Total anomalous pulmonary venous return** (**TAPVR**),
8. **Mitral Valve Stenosis and Regurgitation**,
9. **Ebstein's repair**,
10. **Functionally univentricular heart** (**Cavopulmonary anastomoses and Fontan procedures**), **and**
11. **Cardiac Transplantation**.

Performance Measurements

A new methodology of Performance Measurement will be proposed. This new methodology of Performance Measurement will be discussed and evaluated by the Aristotle Committee.

In the Aristotle system, the Performance is defined by the axiom (axiom definition: "a basic proposition assumed to be true"):

$$\text{Performance} = \text{Outcome} \times \text{Complexity}.$$

The new system of Basic and Comprehensive Performance Measurement in Aristotle 2 will use the STAT Mortality Score and the STAT Morbidity Score based on raw data; meanwhile, technical difficulty will remain based on expert opinion.

We, along with David Clarke, coined the term Optivival [48, 49], to measure the antonym of morbidity:

$$\text{Optivival} = 100\% - \text{Morbidity}\ \%$$

(Survival is the antonym of mortality and Optivival is the antonym of morbidity).

Contributions of the Aristotle Score Towards the Development of Newer Tools to Evaluate Cardiac Surgical Performance

Perhaps the most important contribution of the Aristotle Comprehensive Complexity Score to date has been that its components have been incorporated into the STS Congenital Heart Surgery Database and the EACTS Congenital Heart Surgery Database and have been used to inform the upgrade of these databases, with the goal of increasing the accuracy of the methodology of risk stratification.

The motivation to develop the STAT Mortality Score and the STAT Morbidity Score was at least in a large part related to the Aristotle Score. The desire to transition from subjective probability to objective data within the Aristotle Score is the rational for the eventual incorporation of the STAT Mortality Score and the STAT Morbidity Score into the Aristotle Score.

Although the Aristotle Comprehensive Complexity Score has not been incorporated into the STS Congenital Heart Surgery Database and the EACTS Congenital Heart Surgery Database, many of its components have been added individually to these databases.

The listings of the procedure independent factors have been used to inform the STS Congenital Heart Surgery Database and the EACTS Congenital Heart Surgery Database. Procedure independent factors in the Aristotle Comprehensive Complexity Score include gen-

eral factors, clinical factors, extracardiac factors, and surgical factors. These procedure independent factors have been incorporated into these databases in comprehensive listings of chromosomal abnormalities, syndromes, noncardiac abnormalities, and preoperative factors (Of note, the term "preoperative factors" is used rather than "preoperative risk factors" because the data will help determine whether or not these preoperative factors are actually associated with risk.).

The listings of the procedure dependent factors have also been used to inform the STS Congenital Heart Surgery Database and the EACTS Congenital Heart Surgery Database. Procedure dependent factors in the Aristotle Comprehensive Complexity Score include anatomical factors, associated procedures, and age at procedure. Incorporation of a list of procedure dependent factors for all of the procedures in the STS Congenital Heart Surgery Database and the EACTS Congenital Heart Surgery Database would have been extremely challenging as in initial step because of the huge number of elements of data. Consequently, list of procedure dependent factors named: "Procedure Specific Factors" were incorporated into the STS Congenital Heart Surgery Database and the EACTS Congenital Heart Surgery Database for the following ten benchmark operations:

1. Ventricular Septal Defect (VSD) repair
2. Tetralogy of Fallot (TOF) repair
3. Complete atrioventricular canal repair
4. Arterial switch
5. Arterial switch + VSD repair
6. Glenn/HemiFontan
7. Fontan operation
8. Truncus arteriosus repair
9. Norwood procedure
10. Off Bypass Coarctation repair - only include cases with Operation Type = No CPB Cardiovascular

Thus, although the Aristotle Comprehensive Complexity Score has not been incorporated into the STS Congenital Heart Surgery Database and the EACTS Congenital Heart Surgery Database, many of its procedure dependent factors and procedure independent factors have been added individually to these databases.

Conclusion

In conclusion, the Aristotle Score is an original method to evaluate quality in congenital cardiac surgery. The Aristotle Basic Complexity Score has been instrumental to help the growth of the STS Congenital Heart Surgery Database and the EACTS Congenital Heart Surgery Database. The Aristotle Comprehensive Complexity Score has proven to be today the best predictor of hospital mortality with a C-Index of 0.86, and is successfully used by many institutions. The Aristotle Score system is designed to evaluate performance of centers and surgeons, not to predict individual patient mortality.

The next steps will be to incorporate the STAT Mortality Score and STAT Morbidity Score into the Aristotle Score. New measurements of Performance are proposed. The new Aristotle 2 will need more objective data on Morbidity to ultimately be constructed. The new Aristotle 2 should be available in 2015.

Finally, it is certain that the Aristotle Score is responsible for multiple important contributions leading to the development of newer tools to evaluate cardiac surgical performance. The name of the philosopher Aristotle is derived from the term "aristos", which means "the best" in Greek. Performance is therefore within the idiom "Aristotle Score".

Appendix 1: The Aristotle Basic Complexity Score (ABC Score) and the Aristotle Basic Complexity Levels (ABC Levels) (January 1, 2010)

Appendix 1 documents how the Aristotle Basic Complexity Score is applied in the STS Congenital Heart Surgery Database and the EACTS Congenital Heart Surgery Database. (Appendix 1 is reproduced with permission from Jacobs et al. [50])

Score	Mortality	Morbidity	Difficulty
1 pt	<1%	ICU 0-24H	elementary
2 pt	1-5%	ICU 1D-3D	simple
3 pt	5-10%	ICU 4D-7D	average
4 pt	10-20%	ICU 1W-2W	important
5 pt	> 20%	ICU > 2W	major

Complexity	
1.5 to 5.9	1
6.0 to 7.9	2
8.0 to 9.9	3
10.0 to 15.0	4

Procedures	Total (Basic Score)	Complexity (Basic Level)	Mortality	Morbidity	Difficulty
Pleural drainage procedure	1.5	1	0.5	0.5	0.5
Bronchoscopy	1.5	1	0.5	0.5	0.5
Delayed sternal closure	1.5	1	0.5	0.5	0.5
Mediastinal exploration	1.5	1	0.5	0.5	0.5
Sternotomy wound drainage	1.5	1	0.5	0.5	0.5
Intra-aortic balloon pump (IABP) insertion	2.0	1	0.5	1.0	0.5
Explantation of pacing system	2.5	1	1.0	1.0	0.5
PFO, Primary closure	3.0	1	1.0	1.0	1.0
ASD repair, Primary closure	3.0	1	1.0	1.0	1.0
ASD repair, Patch	3.0	1	1.0	1.0	1.0
ASD partial closure	3.0	1	1.0	1.0	1.0
Atrial fenestration closure	3.0	1	1.0	1.0	1.0
Pericardial drainage procedure	3.0	1	1.0	1.0	1.0
PDA closure, Surgical	3.0	1	1.0	1.0	1.0
Pacemaker implantation, Permanent	3.0	1	1.0	1.0	1.0
Pacemaker procedure	3.0	1	1.0	1.0	1.0
Shunt, Ligation and takedown	3.5	1	1.5	1.0	1.0
ASD, Common atrium (Single atrium), Septation	3.8	1	1.0	1.0	1.8
AVC (AVSD) repair, Partial (incomplete) (PAVSD)	4.0	1	1.0	1.0	2.0
Coronary artery fistula ligation	4.0	1	1.0	2.0	1.0
Aortopexy	4.0	1	1.5	1.5	1.0
ICD (AICD) implantation	4.0	1	1.5	1.0	1.5
ICD (AICD) (automatic implantable cardioverter defibrillator) procedure	4.0	1	1.5	1.0	1.5
Ligation, Thoracic duct	4.0	1	1.0	2.0	1.0
Diaphragm plication	4.0	1	1.0	2.0	1.0
ECMO decannulation	4.0	1	2.0	1.0	1.0
ASD creation/enlargement	5.0	1	2.0	2.0	1.0

Atrial septal fenestration	5.0	1	2.0	2.0	1.0
AVC (AVSD) repair, Intermediate (transitional)	5.0	1	1.5	1.5	2.0
PAPVC repair	5.0	1	2.0	1.0	2.0
Lung biopsy	5.0	1	1.5	2.0	1.5
Ligation, Pulmonary artery	5.0	1	1.5	2.0	1.5
Decortication	5.0	1	1.0	1.0	3.0
ASD repair, Patch + PAPVC repair	5.0	1	2.0	1.0	2.0
PAPVC Repair, Baffle redirection to left atrium with systemic vein translocation (Warden) (SVC sewn to right atrial appendage)	5.0	1	1.0	2.0	2.0
ECMO cannulation	5.0	1	2.0	1.0	2.0
Pectus repair	5.3	1	2.0	1.0	2.3
Aortic stenosis, Supravalvar, Repair	5.5	1	1.5	2.0	2.0
Valvuloplasty, Pulmonic	5.6	1	1.8	1.8	2.0
VSD repair, Primary closure	6.0	2	2.0	2.0	2.0
VSD repair, Patch	6.0	2	2.0	2.0	2.0
AP window repair	6.0	2	2.0	2.0	2.0
Valve replacement, Truncal valve	6.0	2	2.0	2.0	2.0
Cor triatriatum repair	6.0	2	2.0	2.0	2.0
Valve excision, Tricuspid (without replacement)	6.0	2	2.0	2.0	2.0
PA, reconstruction (plasty), Main (trunk)	6.0	2	2.0	2.0	2.0
Pericardiectomy	6.0	2	2.0	2.0	2.0
Coarctation repair, End to end	6.0	2	2.0	2.0	2.0
Coarctation repair, Subclavian flap	6.0	2	2.0	2.0	2.0
Coarctation repair, Patch aortoplasty	6.0	2	2.0	2.0	2.0
Vascular ring repair	6.0	2	2.0	2.0	2.0
PA banding (PAB)	6.0	2	2.0	2.0	2.0
PA debanding	6.0	2	2.0	2.0	2.0
ECMO procedure	6.0	2	2.0	3.0	1.0
Aortic stenosis, Subvalvar, Repair	6.3	2	2.0	1.8	2.5
Shunt, Systemic to pulmonary, Modified Blalock–Taussig shunt (MBTS)	6.3	2	2.0	2.0	2.3
RVOT procedure	6.5	2	2.0	2.0	2.5
Valve replacement, Pulmonic (PVR)	6.5	2	2.0	2.0	2.5
Shunt, Systemic to pulmonary, Central (From aorta or to main pulmonary artery)	6.8	2	2.0	2.0	2.8
Valvuloplasty, Truncal valve	7.0	2	2.0	2.0	3.0
Anomalous systemic venous connection repair	7.0	2	2.0	2.0	3.0
Occlusion MAPCA(s)	7.0	2	2.0	2.0	3.0
Valvuloplasty, Tricuspid	7.0	2	2.0	2.0	3.0
DCRV repair	7.0	2	2.0	2.0	3.0
Valve replacement, Aortic (AVR), Mechanical	7.0	2	2.0	2.0	3.0
Valve replacement, Aortic (AVR), Bioprosthetic	7.0	2	2.0	2.0	3.0
Atrial baffle procedure, Mustard or Senning revision	7.0	2	2.0	2.0	3.0
Aortic arch repair	7.0	2	2.0	2.0	3.0
Bidirectional cavopulmonary anastomosis (BDCPA) (bidirectional Glenn)	7.0	2	2.5	2.0	2.5
Glenn (unidirectional cavopulmonary anastomosis) (unidirectional Glenn)	7.0	2	2.5	2.0	2.5
Right/left heart assist device procedure	7.0	2	2.0	3.0	2.0
Hybrid Approach "Stage 1", Stent placement in arterial duct (PDA)	7.0	2	1.5	1.5	4.0
VAD implantation	7.0	2	2.0	3.0	2.0
VAD explantation	7.0	2	2.0	3.0	2.0
Ventricular septal fenestration	7.5	2	3.0	2.0	2.5
TOF repair, Ventriculotomy, Non-transanular patch	7.5	2	2.5	2.0	3.0
Valve replacement, Tricuspid (TVR)	7.5	2	2.5	2.0	3.0
Conduit placement, RV to PA	7.5	2	2.5	2.0	3.0
Sinus of Valsalva, Aneurysm repair	7.5	2	2.5	2.0	3.0
Valve replacement, Mitral (MVR)	7.5	2	2.5	2.0	3.0
Coronary artery bypass	7.5	2	2.5	2.0	3.0
Bilateral bidirectional cavopulmonary anastomosis (BBDCPA) (bilateral bidirectional Glenn)	7.5	2	2.5	2.0	3.0
Conduit placement, Other	7.5	2	2.5	2.0	3.0
Hybrid Approach "Stage 1", Application of RPA and LPA bands	7.5	2	2.5	2.5	2.5
Atrial baffle procedure (non-Mustard, non-Senning)	7.8	2	2.8	2.0	3.0
PA, reconstruction (plasty), Branch, Central (within the hilar bifurcation)	7.8	2	2.8	2.0	3.0
Coarctation repair, Interposition graft	7.8	2	2.8	2.0	3.0
PAPVC, Scimitar, Repair	8.0	3	3.0	2.0	3.0
Systemic venous stenosis repair	8.0	3	3.0	2.0	3.0
TOF repair, No ventriculotomy	8.0	3	3.0	2.0	3.0
TOF repair, Ventriculotomy, Transanular patch	8.0	3	3.0	2.0	3.0
TOF repair, RV-PA conduit	8.0	3	3.0	2.0	3.0
Conduit reoperation	8.0	3	3.0	2.0	3.0

Conduit placement, LV to PA	8.0	3	3.0	2.0	3.0
Valvuloplasty, Aortic	8.0	3	3.0	2.0	3.0
Aortic root replacement	8.0	3	2.5	2.0	3.5
Valvuloplasty, Mitral	8.0	3	3.0	2.0	3.0
Mitral stenosis, Supravalvar mitral ring repair	8.0	3	3.0	2.0	3.0
Coarctation repair, End to end, Extended	8.0	3	3.0	2.0	3.0
Arrhythmia surgery - atrial, Surgical ablation	8.0	3	3.0	2.0	3.0
Arrhythmia surgery - ventricular, Surgical ablation	8.0	3	3.0	2.0	3.0
Hemifontan	8.0	3	3.0	2.0	3.0
Aneurysm, Ventricular, Right, Repair	8.0	3	3.0	2.0	3.0
Aneurysm, Pulmonary artery, Repair	8.0	3	3.0	2.0	3.0
Cardiac tumor resection	8.0	3	3.0	2.0	3.0
Pulmonary embolectomy	8.0	3	3.0	3.0	2.0
Pulmonary embolectomy, Acute pulmonary embolus	8.0	3	3.0	3.0	2.0
Aortic stenosis, Subvalvar, Repair, With myectomy for IHSS	8.0	3	2.0	2.0	4.0
Valvuloplasty converted to valve replacement in the same operation, Pulmonic	8.0	3	2.5	2.5	3.0
LV to aorta tunnel repair	8.3	3	3.0	2.3	3.0
Valve replacement, Aortic (AVR), Homograft	8.5	3	3.0	2.0	3.5
Aortic root replacement, Valve sparing	8.5	3	2.0	2.0	4.5
Senning	8.5	3	3.0	2.5	3.0
PA, reconstruction (plasty), Branch, Peripheral (at or beyond the hilar bifurcation)	8.8	3	2.8	2.5	3.5
Aortic root replacement, Mechanical	8.8	3	3.3	2.0	3.5
Aortic aneurysm repair	8.8	3	3.0	2.8	3.0
VSD, Multiple, Repair	9.0	3	3.0	2.5	3.5
VSD creation/enlargement	9.0	3	3.0	3.0	3.0
AVC (AVSD) repair, Complete (CAVSD)	9.0	3	3.0	3.0	3.0
Pulmonary artery origin from ascending aorta (hemitruncus) repair	9.0	3	3.0	3.0	3.0
TAPVC repair	9.0	3	3.0	3.0	3.0
Pulmonary atresia - VSD (including TOF, PA) repair	9.0	3	3.0	3.0	3.0
Valve closure, Tricuspid (exclusion, univentricular approach)	9.0	3	4.0	3.0	2.0
1 1/2 ventricular repair	9.0	3	3.0	3.0	3.0
Fontan, Atrio-pulmonary connection	9.0	3	3.0	3.0	3.0
Fontan, Atrio-ventricular connection	9.0	3	3.0	3.0	3.0
Fontan, TCPC, Lateral tunnel, Fenestrated	9.0	3	3.0	3.0	3.0
Fontan, TCPC, Lateral tunnel, Non-fenestrated	9.0	3	3.0	3.0	3.0
Fontan, TCPC, External conduit, Fenestrated	9.0	3	3.0	3.0	3.0
Fontan, TCPC, External conduit, Non-fenestrated	9.0	3	3.0	3.0	3.0
Congenitally corrected TGA repair, VSD closure	9.0	3	3.0	3.0	3.0
Mustard	9.0	3	3.0	3.0	3.0
Pulmonary artery sling repair	9.0	3	3.0	3.0	3.0
Aneurysm, Ventricular, Left, Repair	9.0	3	3.0	2.5	3.5
Conduit placement, Ventricle to aorta	9.0	3	3.0	3.0	3.0
Pulmonary embolectomy, Chronic pulmonary embolus	9.0	3	3.0	3.0	3.0
Valvuloplasty converted to valve replacement in the same operation, Truncal valve	9.0	3	2.5	3.0	3.5
Valvuloplasty, Common atrioventricular valve	9.0	3	3.5	2.5	3.0
TOF - Absent pulmonary valve repair	9.3	3	3.0	3.0	3.3
Transplant, Heart	9.3	3	3.0	3.3	3.0
Aortic root replacement, Bioprosthetic	9.5	3	3.5	2.0	4.0
Aortic root replacement, Homograft	9.5	3	3.5	2.0	4.0
Damus–Kaye–Stansel procedure (DKS) (creation of AP anastomosis without arch reconstruction)	9.5	3	3.0	3.0	3.5
Valvuloplasty converted to valve replacement in same operation, Tricuspid	9.5	3	3.0	2.5	4.0
Superior cavopulmonary anastomosis(es) (Glenn or HemiFontan) + Atrioventricular valvuloplasty	9.5	3	3.0	3.0	3.5
Ebstein's repair	10.0	4	3.0	3.0	4.0
Arterial switch operation (ASO)	10.0	4	3.5	3.0	3.5
Rastelli	10.0	4	3.0	3.0	4.0
Coarctation repair + VSD repair	10.0	4	2.5	3.5	4.0
Aortic arch repair + VSD repair	10.0	4	3.0	3.0	4.0
Anomalous origin of coronary artery from pulmonary artery repair	10.0	4	3.0	3.0	4.0
Superior cavopulmonary anastomosis(es) + PA reconstruction	10.0	4	3.5	3.0	3.5
Hybrid Approach "Stage 2", Aortopulmonary amalgamation + Superior Cavopulmonary anastomosis(es) + PA Debanding + Without aortic arch repair	10.0	4	2.5	3.5	4.0
Hybrid Approach "Stage 1", Stent placement in arterial duct (PDA) + application of RPA and LPA bands	10.0	4	3.0	3.0	4.0
Valve replacement, Common atrioventricular valve	10.0	4	3.5	3.5	3.0
Ross procedure	10.3	4	4.0	2.3	4.0

DORV, Intraventricular tunnel repair	10.3	4	3.3	3.0	4.0
Valvuloplasty converted to valve replacement in the same operation, Aortic	10.3	4	3.5	2.5	4.3
Ventricular septation	10.5	4	3.5	3.5	3.5
Valvuloplasty converted to valve replacement in the same operation, Mitral	10.5	4	4.0	2.5	4.0
Interrupted aortic arch repair	10.8	4	3.8	3.0	4.0
Truncus arteriosus repair	11.0	4	4.0	3.0	4.0
TOF - AVC (AVSD) repair	11.0	4	4.0	3.0	4.0
Pulmonary atresia - VSD - MAPCA (pseudotruncus) repair	11.0	4	4.0	3.0	4.0
Unifocalization MAPCA(s)	11.0	4	4.0	3.0	4.0
Konno procedure	11.0	4	4.0	3.0	4.0
Congenitally corrected TGA repair, Atrial switch and Rastelli	11.0	4	4.0	3.0	4.0
Congenitally corrected TGA repair, VSD closure and LV to PA conduit	11.0	4	4.0	3.0	4.0
Arterial switch operation (ASO) and VSD repair	11.0	4	4.0	3.0	4.0
REV	11.0	4	4.0	3.0	4.0
DOLV repair	11.0	4	4.0	3.0	4.0
Aortic dissection repair	11.0	4	4.0	3.0	4.0
TAPVC repair + Shunt - Systemic to pulmonary	11.0	4	4.0	3.5	3.5
Arterial switch procedure + Aortic arch repair	11.5	4	4.0	3.5	4.0
Valvuloplasty converted to valve replacement in the same operation, Common atrioventricular valve	11.5	4	4.5	3.0	4.0
Fontan + Atrioventricular valvuloplasty	11.5	4	4.0	3.5	4.0
Pulmonary venous stenosis repair	12.0	4	4.0	4.0	4.0
Partial left ventriculectomy (LV volume reduction surgery) (Batista)	12.0	4	4.0	4.0	4.0
Transplant, Lung(s)	12.0	4	4.0	4.0	4.0
Aortic root translocation over left ventricle (Including Nikaidoh procedure)	12.0	4	3.0	4.0	5.0
Valvuloplasty converted to valve replacement in the same operation, Aortic - with Ross procedure	12.0	4	4.0	3.5	4.5
Ross–Konno procedure	12.5	4	4.5	3.0	5.0
Fontan revision or conversion (Re-do Fontan)	12.5	4	4.0	4.0	4.5
Arterial switch procedure and VSD repair + Aortic arch repair	13.0	4	4.5	4.0	4.5
Hybrid Approach "Stage 2", Aortopulmonary amalgamation + Superior Cavopulmonary anastomosis(es) + PA Debanding + Aortic arch repair (Norwood [Stage 1] + Superior Cavopulmonary anastomosis(es) + PA Debanding)	13.0	4	4.0	4.5	4.5
Transplant, Heart and lung(s)	13.3	4	4.0	5.0	4.3
Congenitally corrected TGA repair, Atrial Switch and ASO (Double switch)	13.8	4	5.0	3.8	5.0
Valvuloplasty converted to valve replacement in the same operation, Aortic - with Ross-Konno procedure	14.0	4	4.5	4.5	5.0
Norwood procedure	14.5	4	5.0	4.5	5.0
HLHS biventricular repair	15.0	4	5.0	5.0	5.0
Truncus + Interrupted aortic arch repair (IAA) repair	15.0	4	5.0	5.0	5.0

Interventional cardiology or not eligible (intentionally excluded from Aristotle) procedures:

ASD repair, Device
VSD repair, Device
PDA closure, Device
ASD creation, Balloon septostomy (BAS) (Rashkind)
ASD creation, Blade septostomy
Balloon dilation
Stent placement
Device closure
RF ablation
Coil embolization
Pulmonary AV fistula repair/occlusion
TGA, Other procedures (Kawashima, LV-PA conduit, other)
Cardiovascular catherization procedure, Therapeutic
Echocardiography procedure, Sedated transesophageal echocardiogram
Echocardiography procedure, Sedated transthoracic echocardiogram
Non-cardiovascular, non-thoracic procedure on cardiac patient with cardiac anesthesia
Radiology procedure on cardiac patient, Cardiac Computerized Axial Tomography (CT Scan)
Radiology procedure on cardiac patient, Cardiac Magnetic Resonance Imaging (MRI)
Radiology procedure on cardiac patient, Diagnostic radiology
Radiology procedure on cardiac patient, Non-Cardiac Computerized Tomography (CT) on cardiac patient
Radiology procedure on cardiac patient, Non-Cardiac Magnetic Resonance Imaging (MRI) on cardiac patient
Radiology procedure on cardiac patient, Therapeutic radiology
Cardiovascular catherization procedure, Diagnostic
Cardiovascular catherization procedure, Diagnostic, Hemodynamic data obtained
Cardiovascular catherization procedure, Diagnostic, Angiographic data obtained
Cardiovascular catherization procedure, Diagnostic, Transluminal test occlusion
Cardiovascular catherization procedure, Diagnostic, Hemodynamic alteration

Cardiovascular catherization procedure, Diagnostic, Electrophysiology alteration
Cardiovascular catherization procedure, Therapeutic, Septostomy
Cardiovascular catherization procedure, Therapeutic, Balloon valvotomy
Cardiovascular catherization procedure, Therapeutic, Stent re-dilation
Cardiovascular catherization procedure, Therapeutic, Perforation (establishing interchamber and/or intervessel communication)
Cardiovascular catherization procedure, Therapeutic, Transcatheter Fontan completion
Cardiovascular catherization procedure, Therapeutic, Transcatheter implantation of valve
Cardiovascular catherization procedure, Therapeutic Adjunctive therapy
Cardiovascular electophysiological catheterization procedure
Cardiovascular electophysiological catheterization procedure, Therapeutic ablation

Other miscellaneous, not scored:
(Either too vague or not a primary procedure)
Atrial baffle procedure, NOS
VSD repair, NOS
Valve surgery, Other, Tricuspid
Valve surgery, Other, Pulmonic
Valve surgery, Other, Mitral
Valve surgery, Other, Aortic
Tracheal procedure
TOF repair, NOS
Thoracotomy, Other
Thoracic and/or mediastinal procedure, Other
TGA, Other procedures (Nikaidoh, Kawashima, LV-PA conduit, other)
Shunt, Systemic to pulmonary, Other
Shunt, Systemic to pulmonary, NOS
Pleural procedure, Other
Peripheral vascular procedure, Other
Pericardial procedure, Other
PDA closure, NOS
Palliation, Other
PA, reconstruction (plasty), NOS
Other
Organ procurement
Miscellaneous procedure, Other
Mediastinal procedure
Fontan, TCPC, Lateral tunnel, NOS
Fontan, Other
Fontan, NOS
Esophageal procedure
DORV repair, NOS
Diaphragm procedure, Other
Coronary artery procedure, Other
Congenitally corrected TGA repair, Other
Congenitally corrected TGA repair, NOS
Conduit placement, NOS
Coarctation repair, Other
Coarctation repair, NOS
Cardiotomy, Other
Cardiac procedure, Other
AVC (AVSD) repair, NOS
ASD repair, NOS
Arrhythmia surgery, NOS
Other annular enlargement procedure
Fontan, TCPC, External conduit, NOS
VATS (video assisted thoracoscopic surgery)
Minimally invasive procedure
Bypass for non-cardiac lesion
Valve replacement, Aortic

References

1. Mavroudis C, Jacobs JP. The international congenital heart surgery nomenclature and database project. Ann Thorac Surg. 2000;(Suppl):S1–372.
2. Lacour-Gayet F. Risk stratification theme for congenital heart surgery. Semin Thorac Cardiovasc Surg Pediatr Card Surg Annu. 2002;5:148–52.
3. Lacour-Gayet F, Clarke D, Jacobs J, Comas J, Daebritz S, Daenen W, Gaynor W, Hamilton L, Jacobs M, Maruszewski B, Pozzi M, Spray T, Stellin G, Tchervenkov C, Mavroudis and the Aristotle Committee. The Aristotle score: a complexity-adjusted method to evaluate surgical results. Eur J Cardiothorac Surg. 2004;25(6):911–24.
4. Lacour-Gayet F, Jacobs JP, Clarke DR, Maruszewski B, Jacobs ML, O'Brien SM, Mavroudis C. Evaluation of the quality of care in congenital heart surgery: contribution of the Aristotle complexity score. Adv Pediatr. 2007;54:67–83.
5. Lacour-Gayet F, Clarke DR, Aristotle Committee. The Aristotle method: a new concept to evaluate quality of care based on complexity. Curr Opin Pediatr. 2005;17(3):412–7.
6. Lacour-Gayet F, Clarke D, Jacobs J, Gaynor W, Hamilton L, Jacobs M, Maruszewski B, Pozzi M, Spray T, Tchervenkov C, Mavroudis C, Aristotle Committee. The Aristotle score for congenital heart surgery. Semin Thorac Cardiovasc Surg Pediatr Card Surg Annu. 2004;7:185–91.
7. Jacobs JP, Jacobs ML, Maruszewski B, Lacour-Gayet FG, Clarke DR, Tchervenkov CI, Gaynor JW, Spray TL, Stellin G, Elliott MJ, Ebels T, Mavroudis C. Current status of the European Association for Cardio-Thoracic Surgery and the Society of Thoracic Surgeons Congenital Heart Surgery Database. Ann Thorac Surg. 2005;80(6):2278–83; discussion 2283–4.
8. Jacobs JP, Wernovsky G, Elliott MJ. Analysis of outcomes for congenital cardiac disease: can we do better? Cardiol Young. 2007;17 Suppl 2:145–58.
9. Nathan M, Karamichalis JM, Liu H, del Nido P, Pigula F, Thiagarajan R, Bacha EA. Intraoperative adverse events can be compensated by technical performance in neonates and infants after cardiac surgery: a prospective study. J Thorac Cardiovasc Surg. 2011;142(5):1098–107, 1107.
10. Nathan M, Karamichalis JM, Liu H, Emani S, Baird C, Pigula F, Colan S, Thiagarajan RR, Bacha EA, Del Nido P. Surgical technical performance scores are predictors of late mortality and unplanned reinterventions in infants after cardiac surgery. J Thorac Cardiovasc Surg. 2012;144(5):1095–101.
11. Shuhaiber J, Gauvreau K, Thiagarjan R, Bacha E, Mayer J, Del Nido P, Pigula F. Congenital heart surgeon's technical proficiency affects neonatal hospital survival. J Thorac Cardiovasc Surg. 2012;144(5):1119–24.
12. Nathan M, Pigula FA, Liu H, Gauvreau K, Colan SD, Fynn-Thompson F, Emani S, Baird CA, Mayer JE, Del Nido PJ. Inadequate technical performance scores are associated with late mortality and late reintervention. Ann Thorac Surg. 2013;96(2):664–9.
13. Bojan M, Gerelli S, Gioanni S, Pouard P, Vouhé P. Evaluation of a new tool for morbidity assessment in congenital cardiac surgery. Ann Thorac Surg. 2011;92(6):2200–4.
14. Bojan M, Gerelli S, Gioanni S, Pouard P, Vouhé P. Comparative study of the Aristotle Comprehensive Complexity and the Risk Adjustment in Congenital Heart Surgery scores. Ann Thorac Surg. 2011;92(3):949–56.
15. Code of points in artistic gymnastics. http://en.wikipedia.org/wiki/Code_of_Points_(artistic_gymnastics).
16. O'Brien SM, Clarke DR, Jacobs JP, Jacobs ML, Lacour-Gayet FG, Pizarro C, Welke KF, Maruszewski B, Tobota Z, Miller WJ, Hamilton L, Peterson ED, Mavroudis C, Edwards FH. An empirically based tool for analyzing mortality associated with congenital heart surgery. J Thorac Cardiovasc Surg. 2009;138(5):1139–53.
17. Jacobs JP, Lacour-Gayet FG, Jacobs ML, Clarke DR, Tchervenkov CI, Gaynor JW, Spray TL, Maruszewski B, Stellin G, Gould J, Dokholyan RS, Peterson ED, Elliott MJ, Mavroudis C. Initial application in the STS congenital database of complexity adjustment to evaluate surgical case mix and results. Ann Thorac Surg. 2005;79(5):1635–49; discussion 1635–49.
18. Jenkins KJ, Gauvreau K, Newburger JW, Spray TL, Moller JH, Iezzoni LI. Consensus-based method for risk adjustment for surgery for congenital heart disease. J Thorac Cardiovasc Surg. 2002;123:110–8.
19. Jacobs JP, Jacobs ML, Lacour-Gayet FG, Jenkins KJ, Gauvreau K, Bacha E, Maruszewski B, Clarke DR, Tchervenkov CI, Gaynor JW, Spray TL, Stellin G, O'Bien SM, Elliott MJ, Mavroudis C. Stratification of complexity improves the utility and accuracy of outcomes analysis in a Multi-Institutional Congenital Heart Surgery Database: Application of the Risk Adjustment in Congenital Heart Surgery (RACHS-1) and Aristotle Systems in the Society of Thoracic Surgeons (STS) Congenital Heart Surgery Database. Pediatr Cardiol. 2009;30(8):1117–30.
20. O'Brien SM, Jacobs JP, Clarke DR, Maruszewski B, Jacobs ML, Walters 3rd HL, Tchervenkov CI, Welke KF, Tobota Z, Stellin G, Mavroudis C, Hamilton JR,

Gaynor JW, Pozzi M, Lacour-Gayet FG. Accuracy of the Aristotle basic complexity score for classifying the mortality and morbidity potential of congenital heart surgery operations. Ann Thorac Surg. 2007; 84(6):2027–37; discussion 2027–37.
21. Arenz C, Asfour B, Hraska V, Photiadis J, Haun C, Schindler E, Sinzobahamvya N. Congenital heart surgery: surgical performance according to the Aristotle complexity score. Eur J Cardiothorac Surg. 2011; 39(4):e33–7.
22. Sinzobahamvya N, Photiadis J, Arenz C, Kopp T, Blaschczok HC, Hraska V, Asfour B. Congenital heart surgery: applicability of hospital reimbursement according to German diagnosis-related groups system in conformity with the Aristotle complexity score. Thorac Cardiovasc Surg. 2010;58(6):328–32.
23. Schreiber C, Hörer J. Congenital heart surgery and Aristotle complexity score. Thorac Cardiovasc Surg. 2010;58(6):333.
24. Sinzobahamvya N, Kopp T, Photiadis J, Arenz C, Schindler E, Haun C, Hraska V, Asfour B. Surgical management of congenital heart disease: correlation between hospital costs and the Aristotle complexity score. Thorac Cardiovasc Surg. 2010;58(6):322–7.
25. Williams WG. Congenital heart disease: interrelation between German diagnoses-related groups system and Aristotle complexity score. Eur J Cardiothorac Surg. 2010;37(6):1276–7.
26. Photiadis J, Sinzobahamvya N, Arenz C, Sata S, Haun C, Schindler E, Asfour B, Hraska V. Congenital heart surgery: expected versus observed surgical performance according to the Aristotle complexity score. Thorac Cardiovasc Surg. 2011;59(5):268–73.
27. Sinzobahamvya N, Photiadis J, Kopp T, Arenz C, Haun C, Schindler E, Hraska V, Asfour B. Surgical management of congenital heart disease: contribution of the Aristotle complexity score to planning and budgeting in the German diagnosis-related groups system. Pediatr Cardiol. 2012;33(1):36–41.
28. Sinzobahamvya N, Kopp T, Arenz C, Blaschczok HC, Hraska V, Asfour B. Reimbursement by current German Diagnosis-Related Groups system penalises complex congenital heart surgery. Cardiol Young. 2013;13:1–7.
29. Li J, Zhang G, Holtby H, Cai S, Walsh M, Caldarone CA, Van Arsdell GS. Significant correlation of comprehensive Aristotle score with total cardiac output during the early postoperative period after the Norwood procedure. J Thorac Cardiovasc Surg. 2008;136(1):123–8.
30. Artrip JH, Campbell DN, Ivy DD, Almodovar MC, Chan KC, Mitchell MB, Clarke DR, Lacour-Gayet F. Birth weight and complexity are significant factors for the management of hypoplastic left heart syndrome. Ann Thorac Surg. 2006;82(4):1252–7; discussion 1258–9.
31. Sinzobahamvya N, Photiadis J, Kumpikaite D, Fink C, Blaschczok HC, Brecher AM, Asfour B. Comprehensive Aristotle score: implications for the Norwood procedure. Ann Thorac Surg. 2006;81(5): 1794–800.
32. Miyamoto T, Sinzobahamvya N, Photiadis J, Brecher AM, Asfour B. Survival after surgery with cardiopulmonary bypass in low weight patients. Asian Cardiovasc Thorac Ann. 2008;16(2):115–9.
33. Derby CD, Kolcz J, Kerins PJ, Duncan DR, Quezada E, Pizarro C. Aristotle score predicts outcome in patients requiring extracorporeal circulatory support following repair of congenital heart disease. ASAIO J. 2007;53(1):82–6.
34. Miyamoto T, Sinzobahamvya N, Kumpikaite D, Asfour B, Photiadis J, Brecher AM, Urban AE. Repair of truncus arteriosus and aortic arch interruption: outcome analysis. Ann Thorac Surg. 2005;79(6):2077–82.
35. Heinrichs J, Sinzobahamvya N, Arenz C, Kallikourdis A, Photiadis J, Schindler E, Hraska V, Asfour B. Surgical management of congenital heart disease: evaluation according to the Aristotle score. Eur J Cardiothorac Surg. 2010;37(1):210–7.
36. Stoica S, Carpenter E, Campbell D, Mitchell M, da Cruz E, Ivy D, Lacour-Gayet F. Morbidity of the arterial switch operation. Ann Thorac Surg. 2012; 93(6):1977–83.
37. Lacour-Gayet F. Complexity stratification of the arterial switch operation: a second learning curve. Cardiol Young. 2012;22(6):739–44.
38. Gillespie M, Kuijpers M, Van Rossem M, Ravishankar C, Gaynor JW, Spray T, Clark 3rd B. Determinants of intensive care unit length of stay for infants undergoing cardiac surgery. Congenit Heart Dis. 2006;1(4):152–60.
39. Hörer J, Vogt M, Wottke M, Cleuziou J, Kasnar-Samprec J, Lange R, Schreiber C. Evaluation of the Aristotle complexity models in adult patients with congenital heart disease. Eur J Cardiothorac Surg. 2013;43(1):128–34; discussion 134–5.
40. Welke KF, O'Brien SM, Peterson ED, Ungerleider RM, Jacobs ML, Jacobs JP. The complex relationship between pediatric cardiac surgical case volumes and mortality rates in a national clinical database. J Thorac Cardiovasc Surg. 2009;137(5):1133–40.
41. Bojan M, Gerelli S, Gioanni S, Pouard P, Vouhé P. The Aristotle Comprehensive Complexity score predicts mortality and morbidity after congenital heart surgery. Ann Thorac Surg. 2011;91(4):1214–21.
42. Blackstone EH. Let the data speak for themselves. Semin Thorac Cardiovasc Surg Pediatr Card Surg Annu. 2004;7:192–8.
43. Blackstone EH. Statistics for the Rest of Us: monitoring surgical performance. J Thorac Cardiovasc Surg. 2004;128:807–10.
44. Lacour-Gayet F, Jacobs JP, Clarke DR, Gaynor JW, Jacobs ML, Anderson RH, Elliott MJ, Maruszewski B, Vouhé P, Mavroudis C. Performance of surgery for congenital heart disease: shall we wait a generation or look for different statistics? J Thorac Cardiovasc Surg. 2005;130(1):234–5.
45. Jacobs ML, O'Brien SM, Jacobs JP, Mavroudis C, Lacour-Gayet F, Pasquali SK, Welke K, Pizarro C, Tsai F, Clarke DR. An empirically based tool for analyzing morbidity associated with operations for congenital heart disease. J Thorac Cardiovasc Surg. 2013;145(4):1046–57.

46. Jacobs JP, Jacobs ML, Mavroudis C, Backer CL, Lacour-Gayet FG, Tchervenkov CI, Franklin RC, Béland MJ, Jenkins KJ, Walters H, Bacha EA, Maruszewski B, Kurosawa H, Clarke DR, Gaynor JW, Spray TL, Stellin G, Ebels T, Krogmann ON, Aiello VD, Colan SD, Weinberg P, Giroud JM, Everett A, Wernovsky G, Elliott MJ, Edwards FH. Nomenclature and databases for the surgical treatment of congenital cardiac disease–an updated primer and an analysis of opportunities for improvement. Cardiol Young. 2008;18 Suppl 2:38–62.
47. Jacobs ML, Jacobs JP, Jenkins KJ, Gauvreau K, Clarke DR, Lacour-Gayet F. Stratification of complexity: the risk adjustment for congenital heart surgery-1 method and the Aristotle complexity score–past, present, and future. Cardiol Young. 2008;18 Suppl 2: 163–8.
48. Kang N, Tsang VT, Elliott MJ, de Leval MR, Cole TJ. Does the Aristotle Score predict outcome in congenital heart surgery? Eur J Cardiothorac Surg. 2006;29(6):986–8.
49. Clarke DR, Lacour-Gayet F, Jacobs JP, Jacobs ML, Maruszewski B, Pizarro C, Edwards FH, Mavroudis C. The assessment of complexity in congenital cardiac surgery based on objective data. Cardiol Young. 2008;18 Suppl 2:169–76.
50. Jacobs JP, Jacobs ML, Mavroudis C, Lacour-Gayet FG, Tchervenkov CI, Pasquali SK. Executive Summary: The Society of Thoracic Surgeons Congenital Heart Surgery Database – Nineteenth Harvest – (July 1, 2009 – June 30, 2013). The Society of Thoracic Surgeons (STS) and Duke Clinical Research Institute (DCRI), Duke University Medical Center, Durham; 2013 Harvest.

28 Empirically Based Tools for Analyzing Mortality and Morbidity Associated with Congenital Heart Surgery

Marshall L. Jacobs, Sara K. Pasquali, Jeffrey P. Jacobs, and Sean M. O'Brien

Abstract

Congenital heart surgery outcomes analysis requires reliable methods of estimating the risk of adverse outcomes. In the past, methods used for risk adjusted comparisons of outcomes from congenital heart surgery relied on expert opinion about perceived complexity of treatment. The development and growth of national and international congenital heart surgery clinical registry databases has resulted in the availability of large datasets for analysis. The adoption by these registries of standardized nomenclature and definitions and their use of a uniform set of data elements has made it possible to apply robust statistical methodology to these large sets of objective data to develop empirically based tools for analysis of mortality and morbidity associated with congenital heart surgery. The Society of Thoracic Surgeons – European Association for Cardio-Thoracic Surgery Congenital Heart Surgery Mortality Score and Categories (STAT Mortality Categories) and the Society of Thoracic Surgeons Congenital Heart Surgery Morbidity Score and Categories are now widely used in reporting of outcomes, in quality assessment, and in outcomes research.

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Introduction

The rationale for the development of cardiac surgery risk models has been articulated by many groups, including by the Society of Thoracic Surgeons Workforce on Evidence Based Surgery in the following way: "Differences in medical outcomes may result from disease severity, treatment effectiveness, or chance. Because most outcome studies are observational….risk adjustment is necessary to account for case mix" [1]. Statistical models which account for these differences in case mix or severity of disease have been the most widely used approach in the evaluation and comparison of outcomes from surgery for acquired heart disease in adult patients. With respect to surgery for congenital heart disease, and all cardiac surgery for pediatric patients, the evaluation of complexity and the estimation of risk and ultimately of quality of care is a very different matter, and in some ways represents a much greater challenge. In comparison to acquired heart disease in adults, which includes ischemic heart disease, valvular heart disease and disorders affecting the aorta, congenital heart disease is characterized by a broad spectrum of anomalies encompassing hundreds of distinct diagnoses and clinical entities. The minimal datasets used in the congenital heart surgery databases of the Society of Thoracic Surgeons and the European Association for Cardio-Thoracic Surgery contain more than 150 individual diagnoses and over 200 types of procedures. Many of the diagnoses, while unique and important, are relatively rare. And many of the surgical operations are performed in small numbers at any given center. No two centers see exactly comparable patient populations. At any given center, the complexity of the patients seen in any given year is unlikely to be the same as the complexity of those managed during the preceding year, or in the following year, or at another center. Reporting of raw, unadjusted mortality data is misleading, as it fails to consider the influence of high risk patients and complex procedures on outcomes. The need to establish tools for case-mix adjustment is fundamental to any systematic attempt to measure outcomes, compare performance, and sustain a program of continual quality improvement for pediatric and congenital heart surgery.

Consensus-Based Tools for Complexity Stratification

As a response to the need for a system of outcomes assessment adjusted for case-mix, but in the absence of significant amounts of standardized multi-center data, two consensus-based tools for stratification of congenital heart surgery procedures were developed more than a decade ago. Both systems relied on the experience and opinions of panels of experts. For the Aristotle Complexity Score, three components (potential for mortality, potential for morbidity, and technical difficulty) were subjectively scored, with the sum of these scores determining the Aristotle Basic Complexity Score for a given procedure. Each procedure was then assigned to one of four levels of increasing complexity [2]. The Risk Adjustment for Congenital Heart Surgery (RACHS-1) system also relied on an expert panel to assign procedures to one of six levels of increasing risk of mortality. This allocation of procedures was subsequently refined using data from two multi-institutional registries [3]. Of the two subjectively derived tools for stratification of operative procedures, the RACHS-1 categories appeared to have better discrimination for predicting mortality, whereas the ABC score covered a larger proportion of congenital heart surgery case volume and in addition, included consideration of morbidity as an outcome measure and technical difficulty as a measure of complexity [4, 5].

In the *2004 Cardiac Surgery Annual of the Seminars in Thoracic and Cardiovascular Surgery*, Eugene Blackstone characterized contemporary methods for risk adjusted comparisons of outcomes from congenital heart surgery in the following way: "Growing in popularity

among congenital heart surgeons are methods of comparison that rely fundamentally on expert opinion about perceived complexity of treatment…" he acknowledged that, "No medical discipline has been more shaped, driven and scrutinized by outcomes data than cardiac surgery" [6]. Acknowledging the unique challenge posed by the diversity of congenital heart disease diagnoses and procedures he added, "Unlike high-volume operations for acquired heart disease, congenital heart disease is considerably more heterogeneous, many anomalies are rare, and outcomes after surgical correction are highly variable. How, then, can outcome of institutional programs be compared fairly?" His answer was "Let the data speak for themselves," upon which he elaborated as follows: "The latter is the basis for contemporary methods of risk-adjusted comparisons. The proposed international collection of a uniform set of congenital heart surgery data elements, a well-conceived and internationally accepted ontology of congenital heart disease, accurate understanding of established incremental risk factor concepts and their role in risk adjustment, advent of powerful data analysis techniques that include new types of predictive modeling, and wide understanding of risk-adjusted comparison suggest there is ample motivation and opportunity for letting data speak for themselves" [6].

While the consensus-driven stratification tools of the preceding decade filled a void and served a useful purpose, the fact that registry databases in North American and Europe had accumulated data pertaining to well over 100,000 congenital heart surgery operations using a uniform set of data elements, standardized nomenclature and definitions, and systems for data verification set the stage for the development of empirically based tools for risk adjustment and analysis of outcomes associated with congenital heart surgery.

An Empirically Based Tool for Analyzing Mortality Associated with Congenital Heart Surgery

Using a combined data set from the Congenital Heart Surgery Databases of the Society of Thoracic Surgeons (STS) and the European Association for Cardiothoracic Surgery (EACTS), an international consortium of collaborators developed an empirically based index that can be used to identify the statistically estimated risk of in-hospital mortality by procedure, and to group procedures into homogeneous risk categories [7]. Mortality risk was estimated for 148 types of operative procedures using data from 77,294 operations entered into the databases between 2002 and 2007. Procedure-specific mortality rate estimates were calculated using a Bayesian model that adjusted for small denominators. After determination of the mortality risk estimates, each procedure was assigned a numeric score (STS–EACTS score). The scores were assigned by shifting and rescaling the estimated procedure-specific mortality rates to lie in the interval from 0.1 to 5.0.

Procedures were also sorted by increasing risk and grouped into five categories (the STS–EACTS Congenital Heart Surgery Mortality Categories, or STAT Mortality Categories) that were chosen to be optimal with respect to minimizing within-category variation and maximizing between-category variation. The entire list of Mortality Scores and Mortality Categories for 148 different operative procedures, together with unadjusted and Bayesian model-based estimates of mortality risk can be viewed in table form in the original description of the development of these tools by O'Brien and associates [7]. Appendix 1 lists the STS-EACTS Mortality Scores and STAT Mortality Categories as applied in current versions of the Society of Thoracic Surgeons Congenital Heart Surgery Database and the European Association for Cardio-Thoracic Surgery Congenital Heart Surgery Database using the International Pediatric and Congenital Cardiac Code (IPCCC). Representative examples of procedures in the five STAT Mortality Categories include the following: Category 1 includes repair of partial atrioventricular canal defect and repair of ventricular septal defect, Category 2 includes aortic valvuloplasty and total cavopulmonary connection, Category 3 includes hemi-Fontan procedure and arterial switch operation, category 4 includes arterial switch with VSD repair and Ross-Konno procedure, and category 5 includes Norwood procedure and repair of truncus arteriosus with

Table 28.1 Comparison of the C-index for models using the STS-EACTS Mortality Score, STS-EACTS (STAT) Mortality Categories, RACHS-1 risk categories and Aristotle Basic Complexity Scores

Method of modeling procedures	Model without patient covariates (C-index)	Model with patient covariates (C-index)
STS-EACTS Score	0.787	0.816
STS-EACTS (STAT) categories	0.778	0.812
RACHS-1 risk categories	0.745	0.802
Aristotle Basic Complexity Score	0.687	0.795

Adapted from: O'Brien et al. [7]
Validation sample, subset of procedures for which both RACHS-1 risk categories and Aristotle Basic Complexity Scores are defined
Abbreviations: *STS-EACTS* Society of Thoracic Surgeons-European Association for Cardiothoracic Surgery, *RACHS-1* Risk Adjustment for Congenital Heart Surgery

interrupted arch. Estimated mortality rates ranged across procedure types from 0.3 % (atrial septal defect repair with patch, Category 1) to 29.8 % (truncus arteriosus plus interrupted aortic arch repair, Category 5).

The performance of the STAT model for estimated mortality risk was evaluated by applying it to an independent validation sample of data from the same databases, but entered during a 1 year period that followed the generation of the primary data set. The STS–EACTS score and STAT Mortality Categories demonstrated good discrimination for predicting mortality in the validation sample (C-index = 0.784 and 0.773, respectively). Evaluation also included comparison with two existing methods: Risk Adjustment for Congenital Heart Surgery (RACHS-1) categories and Aristotle Basis Complexity scores. In the subset of procedures for which RACHS-1 and Aristotle Basic Complexity scores are defined, the discrimination of the respective modeling procedures was compared (Table 28.1). Discrimination was highest for the STS–EACTS score, followed by the STS–EACTS Categories (STAT Categories). Adding patient-level covariates substantially improved each model's discrimination. With the addition of these patient variables, discrimination was again highest for the STS–EACTS score followed by the STS-EACTS Categories. Two important characteristics of the STS-EACTS Mortality Score and STAT Mortality Categories are that they are empirically derived, and that they compare favorably relative to the consensus based tools that preceded them. A third important characteristic is that 99 % of procedures in the STS-CHSD can be classified on the basis of STAT Mortality Categories, while this is true of only 94 % of cases using the Aristotle Basic Complexity Score and only 86 % using RACHS-1 risk categories [4, 8].

Since 2012, the outcomes reports of the STS Congenital Heart Surgery Database include stratification of all cases on the basis of STAT Mortality Categories. Risk models used for reporting observed-to-expected mortality ratios in the outcomes reports of the STS Congenital Heart Surgery Database involve adjustment for STAT Mortality Category and additional patient factors. The STAT Mortality Categories have been used in outcomes research as one means of adjustment for procedural risk in a variety of investigations [9–12]. The National Quality Forum has endorsed the following measures [13] for reporting outcomes from pediatric and congenital heart surgery:

1. **Surgical Volume for Pediatric and Congenital Heart Surgery: Total Programmatic Volume and Programmatic Volume Stratified by the Five STS-EACTS Mortality Categories**
2. **Operative Mortality Stratified by the Five STS-EACTS Mortality Categories**

Numerous quality assessment programs rely upon the use of STAT Mortality Categories as an objective measure for the description of case mix and a reliable tool for risk adjustment [8, 14].

Development of the STS-EACTS Congenital Heart Surgery Mortality Score and the STAT Mortality Categories was the result of a collaborative effort to derive a valid tool that can be used to stratify congenital heart surgery procedures based on their relative risk of in-hospital mortality. The accomplishment was made possible by combining the resources of

the STS and EACTS Congenital Heart Surgery Databases. These empirically derived tools represent an improvement over existing consensus-based methods. It will be important to periodically re-calibrate these metrics using contemporary datasets [15, 16].

An Empirically Based Tool for Analyzing Morbidity Associated with Congenital Heart Surgery

Historically, congenital heart surgery outcomes analyses have focused primarily on mortality. This, of course, is an incomplete approach to assessment of outcomes. Fortunately, we have reached a point where more than 95 % of patients who undergo surgical operations for congenital heart disease survive to hospital discharge and beyond. Therefore, it is clear that accounting not only for survival, but for other end points as well, is essential to measuring and understanding outcomes, and ultimately to measuring the effectiveness or "quality" of therapeutic approaches to congenital heart disease. Nonfatal events, such as stroke and renal failure, are major determinants of hospital cost and of patients' health status after surgery. In addition, post-procedure length of hospital stay provides useful direct information about resource use and indirect proxy information about a patient's condition [17, 18]. Although such measures are captured in clinical registries, few incorporated tools for analyzing these end points. The importance of developing a morbidity metric was articulated in 2004 by Kolh [19], who described quantitation of morbidity in cardiac surgery as follows: "Being more frequent than mortality, it could carry more information and be measured in terms of postoperative complications and length of hospital stay…. Furthermore, because of the heterogeneity of morbidity events, future scoring systems should probably generate separate predictions for mortality and major morbidity events."

After developing the previously described empirically based tool for analyzing mortality associated with congenital heart surgery, the importance of developing another empirically based tool to address morbidity was acknowledged. The objectives were:

- to develop a morbidity metric that accounts for the occurrence of complications that have a significant and durable impact on the patient's health and also accounts for utilization of health care resources
- to estimate the average amount of patient morbidity by procedure type
- to convert these procedure-specific morbidity estimates into a scale ranging from 0.1 to 5.0 (that range having been chosen specifically for consistency with the STS-EACTS Mortality Score)
- to group procedures with similar estimated morbidity risk into five relatively homogeneous categories that are designed to minimize within-category variation and to serve as a stratification variable that can be used to adjust for case mix when analyzing outcomes and comparing institutions.

Using data from the STS Congenital Heart Surgery Database, an objective, empirically based index was developed, that reflects statistically estimated risk of morbidity by procedure [20]. Using data from 62,851 operations entered in the database in 2002–2008, procedural morbidity risk was estimated using a Bayesian model that adjusted for small denominators. Morbidity was quantified for each procedure on the basis of the proportion of patients experiencing major complications and of the average postoperative length of stay (PLOS) as a measure of resource utilization. Major complication was defined as the occurrence of any one or more of six specific complications (Table 28.2). These complications represent definitive outcomes that can be ascertained reliably and that are likely to have significant and durable impact on patient health. The unadjusted rate of major complications was defined as the percent of operations that were associated with the occurrence of one or more of the major complications listed in Table 28.2.

Model-based estimates with 95 % Bayesian credible intervals were calculated for each procedure's average risk of major complications and average postoperative length of stay. Model-

Table 28.2 Major complications: STS Congenital Heart Surgery Database

Complication description (STS Congenital Heart Surgery Database code[a])	Number (%) of events[b]	Mortality N (%)
Postoperative acute renal failure requiring temporary **or** permanent dialysis (220 or 230)	705 (1.1 %)	396 (56.2 %)
Postoperative neurological deficit *persisting at discharge* (320)	500 (0.8 %)	152 (30.4 %)
Postoperative AV block requiring permanent pacemaker (60)	593 (0.9 %)	28 (4.7 %)
Postoperative mechanical circulatory support (IABP, VAD, ECMO or CPS) (40)	1,110 (1.8 %)	617 (55.6 %)
Phrenic nerve injury/ paralyzed diaphragm (300)	578 (0.9 %)	35 (6.1 %)
Unplanned reoperation (20 or 240)	2,942 (4.7 %)	636 (21.6 %)
Major complication (defined as any one or more of the above)	5,059 (8.0 %)	1,187 (23.5 %)

Adapted from: Jacobs et al. [20]
Abbreviations: *IABP* intra-aortic balloon pump, *VAD* ventricular assist device, *ECMO* extra-corporeal membrane oxygenation, *CPS* cardiopulmonary support
[a]Complication codes in the STS Congenital Heart Surgery Database Data Collection Form, Version 2.50 [ref. STS Congenital Heart Surgery Database Version 2.50 Data Collection Form Annotated "(Updated 7/10/2006)" [http://www.sts.org/sites/default/files/documents/pdf/DataCollectionForm250_07102006_Annotated.pdf]. Accessed 18 Mar 2014]
[b]Denominator is 62,851 operations

estimated risk of major complications ranged from 1.0 % for simple procedures to 38.2 % for truncus arteriosus with interrupted aortic arch repair. Procedure-specific estimates of average postoperative length of stay (PLOS) ranged from 2.9 days for simple procedures to 42.6 days for combined atrial switch and Rastelli operation. To facilitate ranking and grouping of procedures, average risk of major complications and average PLOS were combined into a single composite morbidity measure. To account for different measurement scales, the two individual measures were re-scaled to have the same standard deviation. They were then summed together. The resulting composite morbidity measure was the basis of the proposed Morbidity Scores and Categories. Each procedure was assigned a numeric score ranging from 0.1 to 5.0 (STS Congenital Heart Surgery Morbidity Score). Scores were assigned by shifting and rescaling the procedure-specific composite morbidity estimates to lie in the interval from 0.1 to 5.0 and then rounding to one decimal place. Procedures were sorted by increasing estimated morbidity and partitioned into five relatively homogeneous categories (STS Congenital Heart Surgery Morbidity Categories) using a computer program to determine cut-points that were optimal for minimizing within-category variance and maximizing between-category variance of the composite morbidity measure. In this fashion, 140 procedures were assigned scores ranging from 0.1 to 5.0 and sorted into five STS Congenital Heart Surgery Morbidity Categories (Table 28.3).

Table 28.3 Summary of STS Congenital Heart Surgery Morbidity Categories

	1	2	3	4	5
Number of procedures	36	43	36	21	4
Aggregate average postoperative length of stay	6.3	11.3	15.2	22.3	34.0
Rate of major complications (%)	3.2	6.5	11.9	15.2	30.0

Adapted from: Jacobs et al. [20]

Rate of major complications ranged from 3.2 % in category 1 to 30.0 % in category 5. Aggregate average PLOS ranged from 6.3 days in category 1 to 34.0 days in category 5. A few examples of procedures in the five STS Morbidity Categories include the following: Category 1 includes atrial septal defect repair and repair of partial atrioventricular canal defect; Category 2 includes hemi-Fontan procedure and repair of tetralogy of Fallot with transanular patch; Category 3 includes arterial switch operation and intraventricular tunnel repair of double outlet right ventricle; category 4 includes heart transplantation and repair of total anomalous pulmonary venous connection; and category 5 includes Norwood procedure, repair of truncus arteriosus with interrupted arch, and arterial switch procedure with ventricular septal

Table 28.4 Association between STS Congenital Heart Surgery Morbidity Categories and STAT Mortality Categories

	Mortality categories				
Morbidity categories	**1**	**2**	**3**	**4**	**5**
1	21	13	1	1	0
2	5	26	10	2	0
3	0	12	10	13	1
4	0	0	3	15	3
5	0	0	0	2	2

Adapted from: Jacobs et al. [20]

defect closure and aortic arch repair. The entire list of STS Congenital Heart Surgery Morbidity Scores and Morbidity Categories for 140 different operative procedures, together with unadjusted data and Bayesian model-based estimates of postoperative length of stay and rate of occurrence of any major complications can be viewed in table form in the original description of the development of these tools by Jacobs and associates [20]. Of note, Morbidity Categories were the same as the Mortality Categories for only about one half of the 140 procedures (Table 28.4) supporting the need for a separate morbidity metric to compliment the STAT Mortality metric.

Like the STAT Mortality Categories, the STS Congenital Heart Surgery Morbidity Categories have been used in outcomes research as a means of adjustment for procedural risk in investigations where the focus is on outcomes other than mortality. An example is a recent investigation of occurrence of cardiac arrest and associated outcomes following in-hospital cardiac arrest after pediatric cardiac surgery operations. In this study, multivariable models were used to evaluate association of center surgical case volume with rate of occurrence of cardiac arrest after pediatric heart surgery. Adjustment for case mix was based on classification of procedures according to the STS Morbidity Categories [21].

Conclusion

The STS Morbidity Score and Categories are empirically based tools for analyzing morbidity associated with operations for congenital heart disease and for grouping procedures with similar estimated risk of morbidity. Together with the STS-EACTS Mortality Score and STAT Categories, these tools enhance our ability to accurately characterize case mix. Together, they should add a new dimension and precision to outcome assessments and may provide important information to guide quality improvement initiatives.

Appendix 1: The Society of Thoracic Surgeons – European Association for Cardio-Thoracic Surgery Congenital Heart Surgery Mortality Scores and Mortality Categories (STAT Mortality Categories) (October 31, 2012)

Appendix 1 documents how The Society of Thoracic Surgeons – European Association for Cardio-Thoracic Surgery Congenital Heart Surgery Mortality Scores and Mortality Categories (STAT Mortality Categories) are applied in the STS Congenital Heart Surgery Database and the EACTS Congenital Heart Surgery Database. (Appendix 1 is reproduced with permission from Jacobs et al. [22])

Data version 3.0 Procedure	Procedure	STS-EACTS Mortality Score	STAT Mortality Category
30	ASD repair, Patch	0.1	1
190	AVC (AVSD) repair, Partial (Incomplete) (PAVSD)	0.1	1
10	PFO, Primary closure	0.2	1
20	ASD repair, Primary closure	0.2	1
1470	ICD (AICD) implantation	0.2	1
1480	ICD (AICD) ([automatic] implantable cardioverter defibrillator) procedure	0.2	1
1360	Vascular ring repair	0.2	1
1210	Coarctation repair, End to end	0.2	1
2110	**ASD Repair, Patch + PAPCV Repair	0.2	1
110	VSD repair, Patch	0.2	1
780	Aortic stenosis, Subvalvar, Repair	0.2	1
570	DCRV repair	0.2	1
1460	Pacemaker procedure	0.3	1
260	PAPVC repair	0.3	1
100	VSD repair, Primary closure	0.3	1
2120	*PAPVC Repair, Baffle redirection to left atrium with systemic vein translocation (Warden) (SVC sewn to right atrial appendage)	0.3	1
180	AVC (AVSD) repair, Intermediate (Transitional)	0.3	1
1680	Glenn (Unidirectional cavopulmonary anastomosis) (Unidirectional Glenn)	0.3	1
600	Valve replacement, Pulmonic (PVR)	0.3	1
1250	Coarctation repair, Interposition graft	0.3	1
680	Valve replacement, Aortic (AVR), Mechanical	0.3	1
690	Valve replacement, Aortic (AVR), Bioprosthetic	0.3	1
810	Sinus of Valsalva, Aneurysm repair	0.3	1
360	TOF repair, Ventriculotomy, Nontransanular patch	0.3	1
580	Conduit reoperation	0.3	1
350	TOF repair, No ventriculotomy	0.3	1
970	Fontan, TCPC, Lateral tunnel, Fenestrated	0.3	1
1450	Pacemaker implantation, Permanent	0.4	2
1365	Aortopexy	0.4	2

1330	PDA closure, Surgical	0.4	2
530	PA, Reconstruction (Plasty), Main (Trunk)	0.4	2
520	1 1/2 ventricular repair	0.4	2
1690	Bilateral bidirectional cavopulmonary anastomosis (BBDCPA) (Bilateral bidirectional Glenn)	0.4	2
2130	***Superior Cavopulmonary anastomosis(es) + PA reconstruction	0.4	2
660	Valvuloplasty, Aortic	0.4	2
830	Valvuloplasty, Mitral	0.4	2
1500	Arrhythmia surgery - ventricular, Surgical Ablation	0.4	2
820	LV to aorta tunnel repair	0.4	2
950	Fontan, Atrio-pulmonary connection	0.4	2
740	Ross procedure	0.4	2
930	Pericardiectomy	0.5	2
2350	*Explantation of pacing system	0.5	2
590	Valvuloplasty, Pulmonic	0.5	2
210	AP window repair	0.5	2
510	RVOT procedure	0.5	2
1670	Bidirectional cavopulmonary anastomosis (BDCPA) (Bidirectional Glenn)	0.5	2
1220	Coarctation repair, End to end, Extended	0.5	2
790	Aortic stenosis, Supravalvar, Repair	0.5	2
620	Conduit placement, LV to PA	0.5	2
1772	Conduit placement, Other	0.5	2
370	TOF repair, Ventriculotomy, Transanular patch	0.5	2
1380	Aortic aneurysm repair	0.5	2
1070	Congenitally corrected TGA repair, VSD closure	0.5	2
1730	Aneurysm, Ventricular, Left, Repair	0.5	2
720	Aortic root replacement, Mechanical	0.5	2
1291	Anomalous origin of coronary artery from pulmonary artery repair	0.5	2
715	Aortic root replacement, Bioprosthetic	0.5	2
1790	Ligation, Pulmonary artery	0.6	2
1290	Coronary artery fistula ligation	0.6	2
50	ASD, Common atrium (Single atrium), Septation	0.6	2
2270	*Valvuloplasty converted to valve replacement in the same operation, Pulmonic	0.6	2
1305	*Anomalous aortic origin of coronary artery from aorta (AAOCA) repair	0.6	2
840	Mitral stenosis, Supravalvar mitral ring repair	0.6	2
2100	*Aortic stenosis, Subvalvar, Repair, With myectomy for IHSS	0.6	2
220	Pulmonary artery origin from ascending aorta (Hemitruncus) repair	0.6	2
270	PAPVC, Scimitar, Repair	0.6	2
1000	Fontan, TCPC, External conduit, Fenestrated	0.6	2
1010	Fontan, TCPC, External conduit, Nonfenestrated	0.6	2
735	Aortic root replacement, Valve sparing	0.6	2
1230	Coarctation repair, Subclavian flap	0.7	2
85	Atrial fenestration closure	0.7	2
250	Valve replacement, Truncal valve	0.7	2
340	Systemic venous stenosis repair	0.7	2
460	Valvuloplasty, Tricuspid	0.7	2
290	Cor triatriatum repair	0.7	2

470	Valve replacement, Tricuspid (TVR)	0.7	2
310	Atrial baffle procedure (Non-Mustard, Non-Senning)	0.7	2
550	PA, Reconstruction (Plasty), Branch, Peripheral (At or beyond the hilar bifurcation)	0.7	2
2240	*Valvuloplasty converted to valve replacement in the same operation, Aortic	0.7	2
380	TOF repair, RV-PA conduit	0.7	2
1490	Arrhythmia surgery - atrial, Surgical Ablation	0.7	2
980	Fontan, TCPC, Lateral tunnel, Nonfenestrated	0.7	2
130	VSD, Multiple, Repair	0.7	2
910	Partial left ventriculectomy (LV volume reduction surgery) (Batista)	0.7	2
1630	Shunt, Ligation and takedown	0.8	3
1240	Coarctation repair, Patch aortoplasty	0.8	3
150	Ventricular septal fenestration	0.8	3
450	Occlusion MAPCA(s)	0.8	3
1740	Aneurysm, Pulmonary artery, Repair	0.8	3
330	Anomalous systemic venous connection repair	0.8	3
240	Valvuloplasty, Truncal valve	0.8	3
540	PA, reconstruction (plasty), Branch, Central (within the hilar bifurcation)	0.8	3
2280	*Valvuloplasty converted to valve replacement in same operation, Tricuspid	0.8	3
1700	Hemifontan	0.8	3
1720	Aneurysm, Ventricular, Right, Repair	0.8	3
170	AVC (AVSD) repair, Complete (CAVSD)	0.8	3
1275	**Coarctation repair + VSD repair	0.8	3
1110	Arterial switch operation (ASO)	0.8	3
1410	Transplant, Lung(s)	0.8	3
750	Konno procedure	0.8	3
70	ASD partial closure	0.9	3
1802	Pulmonary embolectomy, Acute pulmonary embolus	0.9	3
1774	Conduit placement, Ventricle to aorta	0.9	3
960	Fontan, Atrio-ventricular connection	0.9	3
1150	Rastelli	0.9	3
2290	*Valvuloplasty converted to valve replacement in the same operation, Truncal valve	1	3
700	Valve replacement, Aortic (AVR), Homograft	1	3
420	Pulmonary atresia - VSD (including TOF, PA) repair	1.1	3
1140	Mustard	1.1	3
1370	Pulmonary artery sling repair	1.1	3
1160	REV	1.1	3
1800	Pulmonary embolectomy	1.2	3
610	Conduit placement, RV to PA	1.2	3
2340	*Fontan + Atrioventricular valvuloplasty	1.2	3
2310	*Valvuloplasty converted to valve replacement in the same operation, Aortic - with Ross procedure	1.2	3
920	Pericardial drainage procedure	1.3	4
850	Valve replacement, Mitral (MVR)	1.3	4
2260	*Valvuloplasty converted to valve replacement in the same operation, Mitral	1.3	4
2300	*Valvuloplasty, Common atrioventricular valve	1.3	4
1650	PA debanding	1.4	4
1280	Aortic arch repair	1.4	4

2330	*Superior cavopulmonary anastomosis(es) (Glenn or HemiFontan) + Atrioventricular valvuloplasty	1.4	4
1760	Cardiac tumor resection	1.4	4
890	Transplant, Heart	1.4	4
1200	DOLV repair	1.4	4
1180	DORV, Intraventricular tunnel repair	1.4	4
1123	**Arterial switch procedure + Aortic arch repair	1.4	4
1120	**Arterial switch operation (ASO) and VSD repair	1.4	4
1025	Fontan revision or conversion (Re-do Fontan)	1.4	4
490	Valve excision, Tricuspid (Without replacement)	1.5	4
1590	Shunt, Systemic to pulmonary, Modified Blalock-Taussig shunt (MBTS)	1.5	4
1300	Coronary artery bypass	1.5	4
400	TOF - Absent pulmonary valve repair	1.5	4
1130	Senning	1.6	4
390	TOF - AVC (AVSD) repair	1.6	4
465	Ebstein's repair	1.6	4
760	Ross-Konno procedure	1.6	4
1640	PA banding (PAB)	1.7	4
440	Unifocalization MAPCA(s)	1.7	4
730	Aortic root replacement, Homograft	1.7	4
1285	**Aortic arch repair + VSD repair	1.7	4
1390	Aortic dissection repair	1.7	4
1080	Congenitally corrected TGA repair, VSD closure and LV to PA conduit	1.7	4
430	Pulmonary atresia - VSD - MAPCA (pseudotruncus) repair	1.7	4
140	VSD creation/enlargement	1.8	4
2230	*Valve replacement, Common atrioventricular valve	1.9	4
2250	*Valvuloplasty converted to valve replacement in the same operation, Common atrioventricular	1.9	4
280	TAPVC repair	1.9	4
880	HLHS biventricular repair	1.9	4
2320	*Valvuloplasty converted to valve replacement in the same operation, Aortic - with Ross-Konno procedure	1.9	4
300	Pulmonary venous stenosis repair	2	4
1600	Shunt, Systemic to pulmonary, Central (From aorta or to main pulmonary artery)	2.1	4
1320	Interrupted aortic arch repair	2.1	4
2210	*TGA, Other procedures (Kawashima, LV-PA conduit, other)	2.4	4
230	Truncus arteriosus repair	2.4	4
2190	*Aortic root translocation over left ventricle (Including Nikaidoh procedure)	2.4	4
1125	**Arterial switch procedure and VSD repair + Aortic arch repair	2.4	4
60	ASD creation/enlargement	2.5	4
2170	*Hybrid Approach "Stage 1", Stent placement in arterial duct (PDA)	2.5	4
80	Atrial septal fenestration	2.6	4
480	Valve closure, Tricuspid (Exclusion, Univentricular approach)	2.6	4
2160	*Hybrid Approach "Stage 1", Application of RPA and LPA bands	2.6	4
1660	Damus-Kaye-Stansel procedure (DKS) (Creation of AP anastomosis without arch reconstruction)	2.9	5
2200	*TAPVC repair + Shunt - Systemic to pulmonary	3	5
2180	*Hybrid Approach "Stage 1", Stent placement in arterial duct (PDA) + application of RPA and	3.1	5
1060	Congenitally corrected TGA repair, Atrial switch and Rastelli	3.2	5

900	Transplant, Heart and lung	3.2	5
1050	Congenitally corrected TGA repair, Atrial switch and ASO (Double switch)	3.4	5
2150	*Hybrid approach "Stage 2", Aortopulmonary amalgamation + Superior Cavopulmonary anastomosis(es) + PA Debanding + Without aortic arch repair	3.6	5
870	Norwood procedure	4	5
2140	*Hybrid approach "Stage 2", Aortopulmonary amalgamation + Superior Cavopulmonary anastomosis(es) + PA Debanding + Aortic arch repair (Norwood [Stage 1] + Superior Cavopulmonary anastomosis(es) + PA Debanding)	4.1	5
2220	**Truncus + IAA Repair	5	5

*Indicates that this Procedure, Score, and Category were not included in the original JTCVS publication [7]. The original list of procedure codes was based on Version 2.5 of the STS Congenital Heart Surgery Database. These additional procedures represent the list of new procedure codes that were added to The STS Congenital Heart Surgery Database in 2010 as part of the upgrade to version 3.0, and have also been incorporated into the EACTS Congenital Heart Surgery Database, and The Japan Congenital Cardiovascular Surgery Database (JCCVSD). To assign scores to these new procedures, a panel of highly experienced congenital heart surgeons from programs representing a variety of programmatic volume categories were surveyed and asked to provide an STS-EACTS Mortality Score for 26 procedures that were new to version 3.0, using the scores in the Table of the JTCVS article [7] as a guide. The mean of the scores from these ten surgeons was then used to assign the STS-EACTS Mortality Score and STS-EACTS Mortality Category for these 26 new procedures. (When the highest and lowest scores were discarded, the scores were essentially the same. [9/23 scores did not change, 13/23 scores change by only 0.1, and 1/23 scores change by 0.2]).
**Indicates a combined procedure (made up of two or more component procedures).
***Indicates a combined procedure and also a procedure for which the Score and Category were not part of the original JTCVS publication [7] and were assigned later as described above.

References

1. Shahian DM, Blackstone EH, Edwards FH, Grover FL, Grunkemeier GL, Naftel DC, Nashef SA, Nugent WC, Peterson ED. STS workforce on evidence-based surgery. Cardiac surgery risk models: a position article. Ann Thorac Surg. 2004;78(5):1868–77.
2. Lacour-Gayet F, Clarke D, Jacobs J, Comas J, Daebritz S, Daenen W, Gaynor W, Hamilton L, Jacobs M, Maruszsewski B, Pozzi M, Spray T, Stellin G, Tchervenkov C, Mavroudis C, The Aristotle Committee. The Aristotle score: a complexity-adjusted method to evaluate surgical results. Eur J Cardiothorac Surg. 2004;25:911–24.
3. Jenkins KJ, Gauvreau K, Newburger JW, Spray TL, Moller JH, Iezzoni LI. Consensus-based method for risk adjustment for surgery for congenital heart disease. J Thorac Cardiovasc Surg. 2002;123(1):110–8.
4. Jacobs JP, Jacobs ML, Lacour-Gayet FG, Jenkins KJ, Gauvreau K, Bacha E, Maruszewski B, Clarke DR, Tchervenkov CI, Gaynor JW, Spray TL, Stellin G, O'Bien SM, Elliott MJ, Mavroudis C. Stratification of complexity improves the utility and accuracy of outcomes analysis in a Multi-Institutional Congenital Heart Surgery Database: application of the Risk Adjustment in Congenital Heart Surgery (RACHS-1) and Aristotle Systems in the Society of Thoracic Surgeons (STS) Congenital Heart Surgery Database. Pediatr Cardiol. 2009;30(8):1117–30. doi:10.1007/s00246-009-9496-0.
5. O'Brien SM, Jacobs JP, Clarke DR, Maruszewski B, Jacobs ML, Walters HL, Tchervenkov CI, Welke KF, Tobota Z, Stellin G, Mavroudis C, Hamilton JR, Gaynor JW, Pozzi M, Lacour-Gayet FG. Accuracy of the Aristotle basic complexity score for classifying the mortality potential of congenital heart surgery operations. Ann Thorac Surg. 2007;84(6):2027–37.
6. Blackstone EH. Let the data speak for themselves? Semin Thorac Cardiovasc Surg Pediatr Card Surg Annu. 2004;7:192–8.
7. O'Brien SM, Clarke DR, Jacobs JP, Jacobs ML, Lacour-Gayet FG, Pizarro C, Welke KF, Maruszewski B, Tobota Z, Miller WJ, Hamilton L, Peterson ED, Mavroudis C, Edwards FH. An empirically based tool for analyzing mortality associated with congenital heart surgery. J Thorac Cardiovasc Surg. 2009;138(5):1139–53.
8. Jacobs JP, Jacobs ML, Maruszewski B, Lacour-Gayet FG, Tchervenkov CI, Tobota Z, Stellin G, Kurosawa H, Murakami A, Gaynor JW, Pasquali SK, Clarke DR, Austin 3rd EH, Mavroudis C. Initial application in the EACTS and STS Congenital Heart Surgery Databases of an empirically derived methodology of complexity adjustment to evaluate surgical case mix and results. Eur J Cardiothorac Surg. 2012;42(5):775–9.
9. Pasquali SK, He X, Jacobs JP, Jacobs ML, O'Brien SM, Gaynor JW. Evaluation of failure to rescue as a quality metric in pediatric heart surgery: an analysis of the STS congenital heart surgery database. Ann Thorac Surg. 2012;94(2):573–80.
10. Jacobs JP, O'Brien SM, Pasquali SK, Jacobs ML, Lacour-Gayet FG, Tchervenkov CI, Austin 3rd EH, Pizarro C, Pourmoghadam KK, Scholl FG, Welke KF, Gaynor JW, Clarke DR, Mayer Jr JE, Mavroudis C. Variation in outcomes for risk-stratified pediatric cardiac surgical operations: an analysis of the STS

congenital heart surgery database. Ann Thorac Surg. 2012;94(2):564–71.
11. Dibardino DJ, Pasquali SK, Hirsch JC, Benjamin DK, Kleeman KC, Salazar JD, Jacobs ML, Mayer JE, Jacobs JP. Effect of sex and race on outcome in patients undergoing congenital heart surgery: an analysis of the Society of Thoracic Surgeons Congenital Heart Surgery Database. Ann Thorac Surg. 2012;94(6):2054–9.
12. Mascio CE, Austin 3rd EH, Jacobs JP, Jacobs ML, Wallace AS, He X, Pasquali SK. Perioperative mechanical circulatory support in children: an analysis of the Society of Thoracic Surgeons Congenital Heart Surgery Database. J Thorac Cardiovasc Surg. 2014;147(2):658–65.
13. Jacobs JP, Jacobs ML, Austin EH, Mavroudis C, Pasquali SK, Lacour-Gayet F, Tchervenkov CI, Walters H, Bacha EA, del Nido PJ, Fraser CD, Gaynor JW, Hirsch JC, Morales DLS, Pourmoghadam KK, Tweddell JS, Prager RL, Mayer JE. Quality measures for congenital and pediatric cardiac surgery. World J Pediatr Congenit Heart Surg. 2012;3(1):32–47.
14. RTI International. Methodology, U.S. News & World Report, Best Children's Hospitals 2013–14. Available at: http://www.usnews.com/pubfiles/BCH_Methodology_2013-14.pdf. Accessed 23 Jan 2014.
15. Tsang VT, Brown KL, Synnergren MJ, Kang N, de Leval MR, Gallivan S, Utley M. Monitoring risk-adjusted outcomes in congenital heart surgery: does the appropriateness of a risk model change with time? Ann Thorac Surg. 2009;87(2):584–7.
16. Jacobs JP, Shahian DM, Jacobs ML, Mavroudis C. Invited commentary re: "monitoring risk-adjusted outcomes in congenital heart surgery: does the appropriateness of a risk model change with time?". Ann Thorac Surg. 2009;87(2):587–8.
17. Mahle WT, Wernovsky G. Long-term developmental outcome of children with complex congenital heart disease. Clin Perinatol. 2001;28:235–47.
18. Pasquali SK, Sun JL, d'Almada P, Jaquiss RD, Lodge AJ, Miller N, et al. Center variation in hospital costs for patients undergoing congenital heart surgery. Circ Cardiovasc Qual Outcomes. 2011;4:306–12.
19. Kolh P. Importance of risk stratification models in cardiac surgery. Eur Heart J. 2006;27:768–9.
20. Jacobs ML, O'Brien SM, Jacobs JP, Mavroudis C, Lacour-Gayet F, Pasquali SK, Welke K, Pizarro C, Tsai F, Clark DR. An empirically based tool for analyzing morbidity associated with operations for congenital heart disease. J Thorac Cardiovasc Surg. 2013;145(4):1046–57.
21. Gupta P, Jacobs JP, Pasquali SK, Hill KD, Gaynor JW, O'Brien SM, He M, Sheng S, Schexnayder SM, Berg RA, Nadkarni VM, Imamura M, Jacobs ML. Epidemiology and outcomes following in-hospital cardiac arrest after pediatric cardiac surgery. Proceedings of 50th annual meeting of the Society of Thoracic Surgeons; Jan 2014. Available at: http://www.sts.org/sites/default/files/documents/pdf/annmtg/2014AM/50AM_AbstractBook.pdf.
22. Jacobs JP, Jacobs ML, Mavroudis C, Lacour-Gayet FG, Tchervenkov CI, Pasquali SK. Executive Summary: The Society of Thoracic Surgeons Congenital Heart Surgery Database – Nineteenth Harvest – (July 1, 2009 – June 30, 2013). The Society of Thoracic Surgeons (STS) and Duke Clinical Research Institute (DCRI), Duke University Medical Center, Durham; 2013 Harvest.

Part V

Verification of Data Completeness and Accuracy of Data

Verification of Data Completeness and Accuracy

29

David M. Overman and David R. Clarke

Abstract

Complete and accurate data is the backbone of any database. Rigorous, objective, and transparent verification processes are vital to establishing and maintaining high quality data. In the absence of such processes, a database cannot be relied upon to properly inform database participants and users. Data about outcomes occupy an increasingly pivotal role in decisions that impact quality of care, professional reputation, reimbursement, and health care policy, to name a few. These developments underscore the importance of robust data verification processes. Data verification may be accomplished using several means, including on site visits and remote source document verification. The emergence of central statistical verification techniques using adaptive or triggered monitoring in real time is an important development. This approach to data verification has the potential of significantly improving both the volume and quality of verified data as well as limiting the resource utilization impact on databases and their participants.

Keywords

Database • Medical Audit • Data Verification • Outcomes • Congenital Heart Disease • Quality Care • Health Policy • Health Professional Affairs

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Florence Nightingale is widely credited with being the first person to advocate for the collection, analysis, and public reporting of surgical outcome data [1]. Complete and accurate data are fundamental to database quality and associated outcomes analysis. In the absence of complete and accurate data, the power of a database as an instrument of outcomes measurement and a platform for quality improvement is severely diminished.

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A process of verification must ensure the completeness and accuracy of data. To accomplish this it is essential that the process be both rigorous and independent to maximize its effectiveness and support data credibility. The importance of the latter consideration is growing as the future of health care delivery is increasingly determined by outcomes analysis as it is employed as a tool for affecting consumer choice, creation of centers of excellence, pay for performance schema, and regulations as yet undeveloped.

The following chapter examines the impact of missing or inaccurate data on outcomes analysis, features of a robust data verification processes, and various methods of data verification. It also provides a detailed description of an effective data verification process which may be used as a template for other database monitors seeking to establish their own data verification process.

The Impact of Missing or Inaccurate Data

Though both impact the ability to perform data analysis, the implications of missing and inaccurate data are notably distinct. Missing data impacts the discriminatory power of a database, in that there are fewer data elements to be analyzed. Strictly speaking, the accuracy of the data in the database is not necessarily compromised, there is simply less of it. It generally accepted that when 10 % of data is missing, analysis of the data element in question can no longer be considered reliable [2, 3]. Thus, systematic pursuit of complete data entry via a rigorous audit process is of paramount importance to the integrity of a database.

In addition to diminishing database sensitivity, missing data also raises the specter of selective omission, a fundamental threat to the integrity of the database. Indeed, it is established that patients not included in a medical audit have higher rates of morbidity and mortality than those are included in the audit [4]:

> In a multi-institutional database of vascular surgery tracking all infrainguinal bypass operations at involved institutions, independent audit revealed that sixteen per cent of eligible cases had not been reported. Mortality and the rate of amputation were twice as high among the missing cases as among the reported cases; however, no difference in patency was identified between the missing cases and the reported cases. The authors concluded that "Overall judgment of the performance of an individual department may be impaired by cases not included in the register." [4]

The above example demonstrates the importance of verifying the completeness of data within a database. In addition to verifying the completeness of data within a database, it is imperative to verify the accuracy of the data. The European Association for Cardio-Thoracic Surgery Congenital Heart Surgery Database attempted to verify the data within the databases of five European centers utilizing "source data verification" [5]. Pre-verification and post-verification mortalities in all groups showed no significant differences, although 7 deaths out of 68 (10.27 %) were missed. None of the other verified fields showed significant differences after verification. The authors stated that "source data verification" showed no statistically significant differences between verified and nonverified data on mortality at 30 days after surgery, length of stay in the hospital, age, body weight, cardiopulmonary bypass time, aortic cross-clamp time, and circulatory arrest time. The authors also state that "an international committee of experts is needed to define common data verification methodology and to apply it in future works on outcome analysis in CHS (congenital heart surgery)." Although the authors state that "source data verification" showed no statistically significant differences between verified and nonverified data in the field of mortality 30 days after surgery, it is troubling that one-tenth of these deaths were not reported. This study confirms the need for a common methodology for verification of data to be developed and implemented in all registries collecting outcomes worldwide.

The importance of the verification of the accuracy of the data is also demonstrated by a prospective, longitudinal, observational, national cohort survival study from the United Kingdom Central Cardiac Audit Database [6]. This analysis included 3,666 surgical procedures and 1,828 therapeutic catheterizations performed from 2000 to 2001, in

all 13 tertiary centers in the United Kingdom performing cardiac surgery or therapeutic cardiac catheterization in children with congenital cardiac disease. Deaths within 30 days of the procedure were established both by results volunteered from the hospital databases, and by independently validated records of deaths obtained by the Office for National Statistics, using the patient's unique National Health Service number, or the general register offices of Scotland and Northern Ireland. Central tracking of mortality identified 469 deaths, with 194 occurring within 30 days and 275 later. Of the 194 deaths occurring within 30 days, 42, or 21.6 %, were detected by central tracking but not by volunteered data. In other words, hospital-based databases underreported mortality within 30 days of the procedure by 21.6 %, even though the hospitals were aware that the data would be independently verified. The authors of the report concluded that "independent data validation is essential for accurate survival analysis" and that "1-year survival gives a more realistic view of outcome than traditional perioperative mortality".

The most spectacular example of the impact of missing data is that of the Bristol Royal Infirmary Inquiry. Some providers at the Bristol Infirmary had a sense that neonatal and infant cardiac surgery at Bristol was resulting in excess mortality, but data to corroborate this impression was lacking [7]. The United Kingdom had initiated clinical audits of pediatric cardiac surgical programs in 1989. It wasn't until several years later, however, that a secret audit [8, 9] was undertaken to objectively assess outcomes that were apparently not documented or available via the existing audit mechanism. The findings of that secret audit, which revealed excessively high mortality, eventuated in the now well known public inquiry and policy decisions regarding provision of pediatric cardiac services in the United Kingdom. The rectification of Bristol's missing data problem forever changed the face of pediatric cardiac surgery and ushered in the era of public reporting of outcomes in the United Kingdom and around the world [10]. As Dr. Stephen Bolsin, who initiated the audit which led to the Bristol Inquiry has noted, "The lesson for the future must be that all services must prospectively collect standardized outcome data for comparison with other centers and to enable performance monitoring…" [11].

Of course most missing data is not typically the result of intentional manipulation of results or other nefarious objectives. Instead, missing data most commonly results from systematic issues such as inadequate communication tools, lack of informed oversight of data entry, suboptimal documentation of procedures, staff turnover, and failure to coordinate data entry from multiple procedural locations within a given institution.

Procedures done at the bedside, usually in the cardiac or neonatal intensive care unit, are frequent sources of missing data or data inconsistencies. This occurs because the procedures are frequently done after hours and with less experienced staff and data is not recorded with the same precision as with elective cases in the operative suite. Lack of surgeon involvement in the data entry process and monitoring of data quality has been shown to result in higher rates of data inaccuracies [12]. Despite the innocuous nature of this incompleteness, however, its impact on the power of a database is substantial. Without complete data, outcomes analysis is severely hindered.

Data accuracy establishes the specificity of the information contained within the database. The issue is not the presence or absence of a value in the data field, but rather whether the values present are correct or not. The necessity of accurate data is self evident. Despite this seemingly obvious truth, examples of the use of inaccurate data and the repercussions thereof are many. Several studies have documented significant data discrepancies when comparing hospital specific databases in the U.K. with data from the United Kingdom Central Cardiac Audit Database [6, 13–15] as well the Scottish Morbidity Record of the Information and Statistics Division of the Scottish Executive (ISD). Despite this, individual centers and their physician staffs have been wrongly accused in the popular press of performing in a substandard manner [16, 17].

Administrative data are widely used to evaluate hospital and subspecialty performance in the United States and Europe. Administrative databases, created as repositories of information

regarding claims and billing of health care services, are useful for several reasons. They are very large datasets and have a broader reach than clinical, academic, or institutional databases. Coding is not performed by clinical personnel, and therefore in theory the data is more objective and less prone to manipulation. However, (though this is not a universal finding), investigators from a variety of specialties have documented the inferiority of data accuracy in administrative databases when compared to clinical databases [18–21]. Multiple factors contribute to this phenomenon [22]:

- coders who are unfamiliar with congenital cardiac disease,
- the inability of coders to clarify questions regarding data entry with clinical personnel, and
- the lack of appropriate ICD-9 codes for many congenital cardiac operations.

Clinical databases, on the other hand, benefit from direct involvement of clinical personnel in data entry. While this has been shown to improve data accuracy, it does raise the possibility of clinical personnel "gaming the system" by under reporting of adverse outcomes [23, 24].

These considerations are addressed in detail elsewhere in this textbook. The point to be understood here is that databases of any form have inherent strengths and weaknesses, and their utility is related to their particular attributes. What they share in common, however, is the need for independent verification of data to guarantee that such data is validated, properly framed, and fairly reported. Lack of verification of complete and accurate data undermines the confidence that participants, clinical investigators, and regulatory agencies have in its dataset. Without such verification, clinicians, administrators, and consumers alike have no substantiated information upon which to base decisions about therapies, program performance, resource utilization, or any one of a myriad of health care delivery concerns are based. In the absence of validated data, the ability of the database to perform multiple important functions is largely lost, including the ability to:

- thoughtfully and empirically advance quality initiatives,
- establish clinical guidelines, and
- perform maintenance of professional standards.

Data verification, then, is central to the increasing role of outcomes databases in the formation of health care structure and policy [25, 26].

Principles of Data Verification

The attributes of a rigorous and effective data verification process have been outlined by the Royal College of Surgeons [27], the Agency for Health Care Research and Quality (AHRQ) and others [28–30]. Features consistently cited as integral to this process include:

1. Confidentiality: First and foremost amongst these is confidentiality. The verification process must ensure the protection of the data being examined. Patient specific information must be protected as well as the identity of the center from which the data is submitted. The participating center must feel assured of this protection if the verification process is to be most effective. This assurance diminishes the tendency for the verification process to be perceived as an adversarial or prosecutorial exercise. Instead, it fosters a cooperative and educational atmosphere whereby deficiencies are viewed in terms of opportunities to improve data quality rather than as examples of ignorance, incompetence, or worse. Confidentiality is, of course, a covenant between the persons involved in data verification and the clinical team members. Secure, password protected files and computers are mandatory requirements.

   This concept has recently come under attack in the United States. The state of New York has required that mortality data in cardiac surgery be made public relative to hospital and even individual surgeon. The need for accurate, complete, and fairly analyzed and reported data has therefore become paramount, and it is now extremely important that clinical principles and credible data verification be applied to the entire registry data process [31–34].

2. Transparency: While confidentiality is of paramount importance, transparency of data verification results is nearly its equal. Objective scrutiny of data quality is only useful if the results of such scrutiny are readily available for review and analysis. This is true for database participants desiring to assess their performance as compared with their peer group, for database personnel who may identify problematic features of the data elements that need correcting, and for outside agencies that may use database analyses for any number of regulatory or other reasons. All of these interested parties' confidence in the ability of the database to perform the function they desire hinges on the transparent reporting of data quality in a formalized and thorough data quality report.
3. Independence: The verification team should design and perform its function without interference from database personnel or participants. The independence and objectivity of the data verification process is crucial to ensure dispassionate evaluation not only of participant performance but also issues related to database design and function. Thus, both "sides" of the database relationship (participant and database personnel or officers) must acknowledge and cooperate with the principle that the data verification team and function must "stand apart". This requirement is substantially addressed by contractual involvement of a professional third party in the data verification process. However, when surgeons are involved in the verification of data, as is necessary with the complexity of congenital heart disease, it is critical that potential conflicts of interest be scrupulously avoided.
4. Consistency: The data verification methodology should be well documented and available for review by whomever may be interested. Data dictionaries should be established and updated so adjudication of data errors is as straightforward and objective as possible. Clearly written educational materials should be maintained to provide general and specific information regarding best practices in databasing, suggested support personnel, coding, and other topics for clinical and data entry personnel. Verification processes must be uniform to facilitate data quality analysis and recognition of trends in data inaccuracies over time. Consistent verification practices assure database participants that data quality results are an "apples to apples" comparison to their peer group.
5. Efficiency: Data verification processes can be very resource intensive [35]. The methodology selected for verification should take into consideration the resource burden such processes may place on the participating center. In addition to minimizing resource utilization impact on participants, efficiencies in the verification process allows, if needed, more expansive application of data quality monitoring within the database. This, in turn, increases data quality and shortens the interval between verification episodes at each participating center.
6. Accountability: For data verification processes to be relevant, significant findings must result in action. This action may be as simple and straightforward as instituting regular coding educational sessions for data entry personnel or increased involvement of surgeons in the processes of entry of data. In the most serious of instances, it may mean corrective action to address data fraud on the part of a database participant. In any case, the performance of data verification implies an intention to act upon its findings. The governance bodies responsible for maintenance of the database should have a clearly defined reporting structure for the database and its data verification process. This most often is in the form of a data verification committee or subcommittee which reports to a larger group of stakeholders overseeing the database and its various functions. Mechanisms for addressing issues identified through data verification should be prospectively established. The Royal College of Surgeons of England, for instance, has a College Hospital Recognition Committee and a Specialist Advisory Committee which are charged with examining evidence that effective clinical audit meetings are taking place,

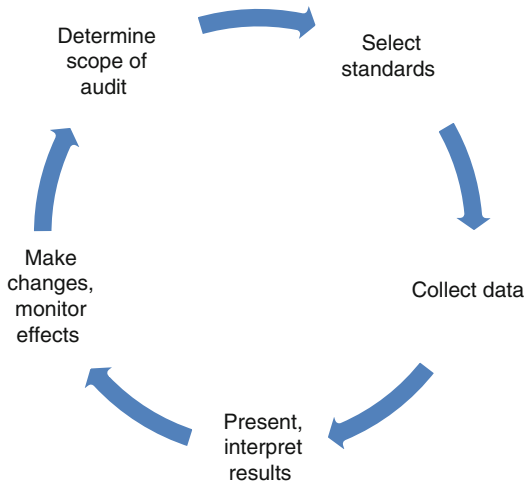

Fig. 29.1 The audit cycle (Adapted from Boult and Maddern [51])

that these are attended regularly by surgical staff, that documentation of those meetings is available, that appropriate institutional support is in place, *and that recommendations arising from the audit have been implemented* [27]. It is accountability and an active feedback mechanism that allows the audit cycle to be effective (Fig. 29.1). Without accountability, data driven improvements in the processes of data entry cannot be accomplished.

Methods of Data Verification

There are three basic approaches to data verification (also referred to as monitoring):

1. automated intrinsic verification of data
2. manual audit of data, which may be performed on-site or remotely with "virtual audit", and
3. verification of data using additional external data sources of data such as national registries of death.

Remote (or central) data verification can include both automated intrinsic monitoring (automated intrinsic verification of data) and virtual remote audit. Hybrid approaches to data verification are most common.

The methodology chosen for data verification is influenced by multiple factors:

- the objectives of the registry or database,
- the type of data being collected,
- the sources of data,
- resources available for verification, and
- the timeframe of data collection and analysis.

The core element of data verification is what is referred to as Source Document Verification (SDV). This refers to direct review of the source documents (operative logs, medical records, laboratory documents, consent forms, etc.) by the auditor. SDV usually occurs on site, but with the advent of electronic medical records, may in some instances be feasible remotely. In high complexity data situations, and where verification expectations are stringent, on site methodologies are usually employed [28].

Remote monitoring is able to identify missing values, out of range or nonsense values, and responses that are either internally (to the participant) or logically inconsistent. In addition, with increasing frequency, central statistical monitoring is employed to detect abnormal patterns of submitted data to allow for more focused analysis, either remotely or on site [36]. This approach is referred to as targeted, adaptive, or triggered monitoring. It has been argued that these statistically driven techniques may be more sensitive discriminators of data error or fraud than more traditional data verification practices [37]. The most notable example of this is the second European Stroke Prevention Study, wherein data for 438 patients were fabricated at a single center. This fraud was detected by an abnormal distribution of plasma levels of aspirin and dipyridamole as compared to other centers [38]. Remote verification procedures should be a standard aspect of data cleaning and ongoing feedback to participating centers in a registry on an ongoing basis, but they are also an integral part of the data verification process. In fact, it may be argued that, in the future, with increasingly sophisticated pattern recognition software, the highest quality data will be realized using real time remote monitoring of inputted data and the mathematical modeling referenced above.

Another significant advantage of remote verification of data is cost savings. Travel and related human resource expenditures for site monitoring can account for as much as 35 % of clinical trial costs [39] and may contribute to database participation costs in an era when institutions are facing

diminishing resources for non-clinical activities. In addition, time spent traveling is time that cannot be used for remote monitoring activities, thus diminishing the number of sites that can be monitored annually. A distinct disadvantage of remote verification is the lack of interface between the data verification team and the participating center personnel. Education and support of database managers and surgeons is a central goal of the process of data verification, and the difficulty of performing this role with remote verification is a significant issue.

Historically, on site monitoring has been the main modality for data verification. The practice of on site SDV has come under increasing scrutiny, however, due to concerns about utilization of resources [40]. SDV produced very low rates of significant data discrepancies as exemplified by the audit of a large cancer clinical trial that revealed no difference in measurements of observed serious adverse events or death compared to data without SDV [41]. These considerations can be minimized by using targeted remote monitoring techniques. The remote use of electronic medical records (EMRs) can also reduce the on site time required for data verification [42].

While 100 % SVD has been the standard in clinical trials, this is not possible in the setting of very large multi-institutional databases such as the Society of Thoracic Surgeons (STS) Congenital Heart Surgery Database (CHSD). In such instances, random sampling of 5–20 % of centers and/or documents is customary.

As mentioned above, when complexity of the dataset is pronounced, and where tolerance for inaccurate data is low (i.e., the monitoring of mortality related to intervention), on site data verification is generally required. On site monitoring gives auditors direct and complete access to source documents and the ability to immediately pursue issues involving missing or unusable (illegible or fragmentary) documentation. Auditors are able to work with on site personnel directly to resolve these issues in real time. In this way, on site verification is reliably thorough and completed in a short amount of time. Finally, on site verification has the very important attribute of face to face debriefing and education. Although verification of the completeness and accuracy of the data in the database is the most important function of the data verification process, education of the participants in the database is a secondary benefit of the data verification process which can have a substantial impact on the quality of the data in the database. Properly conducted data verification processes are educational and professionally enriching for database participants. The audit should not be viewed or experienced as punitive or prosecutorial in nature. The ultimate goal of the data verification process is to improve data completeness and accuracy, thereby amplifying the already well established power of well run databases.

A Data Verification Process: The STS Congenital Heart Surgery Database

The STS CHSD is the largest congenital heart surgery database in the world and now contains over 300,000 operations performed between 1998 and 2014. Remote, intrinsic auditing of submitted data is performed by the Duke Clinical Research Institute (DCRI), and three pilot on site audits were done prior to 2007. At that time, after a thorough investigation of legal issues was conducted by the STS, a formal data verification process was designed and initiated, which involved on site visits and more recently remote data verification. The initial experience with this data verification process has been previously reported [3].

The STS CHSD data warehouse and analytic center is at the DCRI. An independent medical audit firm, The Iowa Foundation for Medical Care (now Telligen), was retained by the STS to manage the data verification process. The data verification cycle is carried out through collaboration between STS, DCRI, and Telligen. Two congenital heart surgeons, who are members of the Audit Subcommittee of the STS Congenital Heart Surgery Database Taskforce, provide input into the logistical details of the verification process and analysis of its results. Formal data verification reports are made to the STS Database Taskforce each year at the STS Annual Meeting. The audits are designed to validate the integrity of the data contained in the database through evaluation of

the completeness and accuracy of the database, as well as the consistency and comprehensiveness of practices of collection of data among a random sample of database participants.

In 2007, the first year of the official audits, five sites were audited with site visits. In 2011, "desk audits" using remote SDV via EMR or shipped paper medical records were first conducted. In 2013, eight centers were being audited (five on site audits and three desk audits), representing nearly 10 % of the 92 participating centers that submitted data the prior year. Unless serious problem issues are discovered, centers that undergo an audit are not eligible for re-auditing until the fourth year following the previous year of audit.

The on-site data verification process is as follows:

Pre-site Visit

- At DCRI, 20 eligible sites are randomly selected for potential data verification by the end of the first quarter of the year.
- The first eight centers (or whatever is the predetermined number to be audited during the current year) receive a Site Audit Notification letter at least 6–8 weeks in advance of the proposed audit. If exigencies prevent auditing of any of the first eight centers (natural disaster or other substantial and unanticipated event) then the next center on the list from the oversample is chosen and notified.
- The population of data to be audited is operations that were performed during the calendar year prior to the audit.
- A Data Manager Questionnaire is completed by the site prior to the audit, and this is reviewed by an auditor at Telligen with extensive knowledge of the STS CHSD. It is used to obtain details regarding the data entry process at the participating center such as timing of data entry, personnel entering data, and documentation utilized. Procedures in place for quality control of data submitted are also itemized. Areas of concern regarding the process of data entry and monitoring can in this way be prospectively identified prior to performance of the audit.
- At DCRI, 30 primary procedures are randomly selected for each center from the year under review. The random list is numbered consecutively. At Telligen, the first 20 procedures are selected for the audit, starting with the first numbered case. The oversample is created in order to accommodate any possible logistical issues (unavailable record, inappropriate procedure, etc.) that might preclude a case from being reviewed.
- A data file is then created for each case selected by DCRI. These files contain data regarding the center's software and vendor, operation and patient identification numbers, and the clinical data elements that will be reabstracted by the auditors. Currently there are 37 such elements. In 2012, over 4,200 variables were audited [43].
- At DCRI, a list is created of all cases with any data element indicating mortality, and reabstraction of this list is also part of the audit.
- Finally, also at DCRI, two case log lists are created:
  - All cases submitted by the center undergoing audited for the calendar year under review.
  - All submitted cases designated CPB or Non-CPB cardiovascular. In the STS CHSD, only cases of these two types are included in the analysis of mortality.

On Site Audit Procedures

- In order to minimize bias a scripted interview is performed with the center's data manger and the surgeon responsible for the database. The Data Manager Questionnaire is reviewed and a discussion of database policies and procedures is conducted. Particularly difficult aspects of data entry, such as obtaining 30 day mortality status, are specifically targeted in this discussion.
- For data completeness, a case log comparison is performed. The data submitted to the STS

CHSD is compared with the hospital operative procedure case log. Discrepancies are identified and the reason for each one determined.

- The 37 data elements are reabstracted from the medical records of the hospital for 20 randomly selected cases and compared with the data elements submitted to the STS CHSD by the center. The following data elements are routinely abstracted by the surgeon auditor:
  - Pre-operative Factors
  - Fundamental Diagnosis
  - Primary Diagnosis
  - Primary Procedure
  - Complications
- All audited fields are adjudicated and classified by the auditors according to the following schema that assigns every audited field to one of the following four classifications:
  1. Match
  2. Acceptable Variance (minor difference in coding)
  3. Unacceptable Variance
  4. Missing Data

  (Note: "Match" is the default value; therefore, it is not necessary to make a notation for a matching value during the adjudication process.)

Rates of agreement as well as rates of completeness are calculated for each data element, using the following formulas:

Rate of agreement = (Match + Acceptable Variance)/(Match + Acceptable Variance + Unacceptable Variance)

Rate of completion = (Match + Acceptable Variance + Unacceptable Variance)/(Match + Acceptable Variance + Unacceptable Variance + Missing Data)

- All index operations associated with mortality are reabstracted by the auditors. A total of 11 data elements are reabstracted for each mortality case. The following mortality classifications are reabstracted exclusively by the surgeon auditor:
  - Mortality: status at hospital discharge (Discharge Mortality)
  - Mortality: 30-day status (30-day Mortality)

Operative Mortality includes both (1) all deaths occurring during the hospitalization in which the operation was performed, even if after 30 days; and (2) those deaths occurring after discharge from the hospital, but within 30 days of the procedure. These classifications have been described in detail previously [44, 45]. Precise definitions are critical to ensure accurate Calculation of mortality and rates of agreement.

- A summary conference is conducted with the database manager, responsible surgeon, any other interested clinical or administrative on site personnel, auditor, and surgeon-auditor. The data verification process is reviewed as are its preliminary findings. Case by case review of errors identified in the submitted data is performed. Clarifications of data definitions and data collection processes are provided. Trends in data inconsistencies are specifically identified, and structural or procedural systematic issues affecting data quality are addressed in detail. Preliminary recommendations to improve data quality are provided. The post site visit process is discussed (see below).

Post-site Visit

- A summary report is prepared within 30 days of completion of the audit by Telligen. After review by the surgeon-auditor, this report is submitted to the STS administration and after review by STS the report is forwarded to the data manager and responsible surgeon at the participating center. As discussed above, an annual compilation of the findings of data verification is presented to the STS Congenital Heart Surgery Database Taskforce.

This process of verification of data has proved to be both effective and efficient for the STS CHSD. Data quality has measurably improved since the inception of the audits. A small number of centers have (randomly) undergone repeat data verification. Repeat audits have uniformly documented improved rates of agreement, suggesting the process of data verification has a salutary and measurable impact on data quality.

Discharge mortality has been consistently captured well by participating centers with

Table 29.1 Mortality audit results from the STS congenital heart surgery database: 2008–2012

	2012	2011	2010	2009	2008
Mortality data					
Surgery date	99.44 %	91.4 %	100.0 %	100.0 %	100.0 %
Admit date	100.0 %	100.0 %	96.0 %	99.0 %	100.0 %
Discharge date	99.72 %	99.5 %	96.0 %	100.0 %	100.0 %
Date of birth	100.0 %	100.0 %	100.0 %	100.0 %	100.0 %
Age	100.0 %	93.3 %	100.0 %	97.1 %	100.0 %
Gender	100.0 %	100.0 %	98.0 %	100.0 %	100.0 %
Operation type	96.90 %	100.0 %	94.3 %	94.4 %	NA
Mortality-discharge status	99.72 %	99.5 %	98.0 %	97.3 %	100.0 %
Mortality-30-day status	97.46 %	99.0 %	93.5 %	97.1 %	72.0 %
Mortality date	99.72 %	96.6 %	96.0 %	94.3 %	88.0 %

resulting high rates of agreement. Historically, "Mortality: 30-day status (30-day Mortality)" has been more challenging to capture; however, the completeness and accuracy of the data in this field of data has improved over time because particular emphasis has been placed on improving capture of 30 day status, and therefore, operative mortality. Agreement rates have improved significantly as a result (Table 29.1).

With accurate mortality data secured, future emphasis will be placed on improving data quality and agreement rates regarding complications and morbidity. As an initial step, focus will be on previously identified major post-operative complications [46]. Educational initiatives and tracking of rates of agreement will be directed at capture of each these major morbidities individually. This evolution from focusing on the capture of mortality data, to that which addresses morbidities associated with intervention, allows the STS CHSD to keep pace with metrics other than survival that are increasingly felt to be more relevant and important to the measurement of quality in congenital heart surgery.

Verification of Data Using Additional External Sources of Data Such National Registries of Death

Both automated intrinsic verification of data and manual audit of data can be augmented by verification of data using additional external sources of data such national registries of death. In the United Kingdom Central Cardiac Audit Database, independent validation of the patient's status (alive or dead) is achieved by central tracking using the linkage of each patient's National Health Service number to the Office of National Statistics, where the death of every resident in England and Wales is registered [3]. (Separate, similar systems exist in Scotland and Northern Ireland). Early efforts are underway to operationalize similar strategies with the STS Database in the United States of America [47–50].

Conclusion

Databases continue to move toward the center of national and international initiatives directed at improvement in the quality of healthcare. Governmental agencies, private payers, professional organizations, and patients all have a vested interest in the assessment of outcomes. Increasingly, certification of professionals, reimbursement, health care policy, and structures of care delivery are driven by outcomes analyses derived from database content.

Verification of the completeness and accuracy of data is central to the legitimacy, intelligence, and credibility of these assessments. Without robust and independent data verification processes, such completeness and accuracy cannot be assumed. The consequences related to the use of incomplete or inaccurate data in the conduct of clinical care or the

formation of health care policy and structure are at best wasteful and ill advised, and at worst devastating.

Several methods of data verification exist. These have various strengths and weaknesses which potentially impact their accuracy, completeness, objectivity, and economy. Appropriate use of a particular data verification platform requires understanding the vulnerabilities of a database and where the most likely sources of data inconsistencies may be found. Judicious use of resources to support the data verification process is an important mechanism to contain costs associated with database participation, and may allow for expansion of the data verification process to include a larger percentage of submitted data. Remote data verification is one way to achieve this end. In the future, development of software specifically designed to interface with electronic medical records for the purpose of data reabstraction and auditing may facilitate the ability to perform remote audits more accurately and efficiently. This tool, coupled with pattern recognition software and the statistical approaches referenced above may provide a platform to create clean, complete, and highly accurate data in real time. Until then, however, vigorous and independent data verification processes are central to the establishment and maintenance of any clinical database.

At present, complex data elements usually require traditional on site source document verification. Though resource intensive, the value and reliability of SDV in assuring data quality justifies this expense. The Society of Thoracic Surgeons Congenital Heart Surgery Database data verification process has proven to be a reasonably effective and efficient platform for assessing and improving data quality. Other organizations wishing to perform database data verification may find this a useful template.

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References

1. Spiegelthalter D. Surgical audit: statistical lessons from Nightingale and Codman. J R Stat Soc. 1999;162(Part 1): 45–58.
2. Messenger JC, Ho KKL, Young CH, Slattery LE, Draoui JC, Curtis JP, Dehmer GJ, Grover FL, Miro MJ, Reynolds MR, Rokos IC, Spertus JA, Wang TY, Winston SA, Rumsfeld JS, Masoudi FA, On behalf of the NCDR Science and Quality Oversight Committee Data Quality Workgroup. The National Cardiovascular Data Registry (NCDR) data quality brief: the NCDR data quality program in 2012. J Am Coll Cardiol. 2012;60(16):1484–8.
3. Clarke DR, Breen LS, Jacobs ML, Franklin RCG, Tobora Z, Maruszewski B, Jacobs JP. Verification of data in congenital cardiac surgery. Cardiol Young. 2008;18 Suppl 2:177–87.
4. Elfstrom J, Stubberod A, Troeng T. Patients not included in medical audit have a worse outcome than those included. Int J Qual Health Care. 1996;8: 153–7.
5. Maruszewshi B, Lacour-Gayet F, Monro JL, Keough BE, Tobota Z, Kansy A. An attempt at data verification in the EACTS Congenital database. Eur J Cardiothorac Surg. 2005;28:400–6.
6. Westaby S, Archer N, Manning N, Adwani S, Grebenik C, Ormerod O, et al. Comparison of hospital episode statistics and the central cardiac audit database in the public reporting of congenital heart surgery mortality. BMJ. 2007;335:759.
7. Bolsin SN. Professional misconduct: the Bristol case. Med J Aust. 1998;169:369–72.
8. Stirrat GM. Audit was secret but not confidential. BMJ. 1999;318:1010.
9. Bolsin SN. Audit was not secret. BMJ. 1999;318:1010.
10. Keough BE, Bridgewater B. Toward public disclosure of surgical results: experience of cardiac surgery in the United Kingdom. Thorac Surg Clin. 2007;17:403–11.
11. Bolsin SN. Paper confirms poor quality of paediatric heart surgery at Bristol during 1991–1995. BMJ. 2002;324:1095.
12. Hunter JC, Lyon C, Galloway K, Putterill M, van Rij A. Complete clinical outcomes audit: resource requirements and validation of the instrument. Surg Endosc. 1999;13:699–704.
13. Gibbs JL, Cunningham D, de Leval M, Monro J, Keogh B. Paediatric cardiac surgical mortality after Bristol: paediatric cardiac hospital episode statistics are unreliable. BMJ. 2005;330(7481):43–4
14. Aylin P, Bottle A, Jarman B, Elliott P. Paediatric cardiac surgical mortality in England after Bristol: descriptive analysis of hospital episode statistics 1991–2002. BMJ. 2004;329:825–9.
15. Milburn JA, Driver CP, Youngson GG, King PM, MacAulay E, Krukowshi ZH. The accuracy of clinical data: a comparison between central and local data collection. Surgeon. 2007;5(5):275–8.

16. Foster K. Fears over child surgery deaths. Scotland on Sunday. 2007 June 10. http://Scotlandonsunday.com/index.cfm?id=906872007.
17. Westaby S, Archer N, Wilson N. Media attack. BMJ. 2007;335:839.
18. Shahian DM, Silverstein T, Lovett AF, Normand SLT. Comparison of clinical and administrative data sources for hospital coronary artery bypass graft surgery report cards. Circulation. 2007;115:1518–27.
19. Herbert MA, Prince SL, Williams JL, Magee MJ, Mack MJ. Are unaudited records from an outcomes registry database accurate? Ann Thorac Surg. 2004;77:1960–5.
20. Welke KF, Diggs BS, Karamlou T, Ungerleider RM. Comparison of pediatric cardiac surgical mortality rates from national administrative data to contemporary clinical standards. Ann Thorac Surg. 2009;87:216–23.
21. Woodworth GF, Baird CJ, Garces-Ambrossi G, Tonascia J, Tamargo RJ. Inaccuracy of the administrative database: comparative analysis of two databases for the diagnosis and treatment of intracranial aneurysms. Neurosurgery. 2009;65(2):251–6.
22. Vlasschaert ME, Bejaimal SA, Hackam DG, Quinn R, Cuerden MS, Oliver MJ, Iansavichus A, Sultan N, Mills A, Garg AX. Validity of administrative database coding for kidney disease: a systematic review. Am J Kidney Dis. 2011;57(1):29–43.
23. Lapar DJ, Stukenborg GJ, Lau CL, Jones DR, Kowzower BD. Differences in reported esophageal cancer resection outcomes between national clinical and administrative databases. J Thorac Cardiovasc Surg. 2012;144(5):1152–7.
24. Menyhei G, Bjorck M, Beiles B, Halbakken E, Jensen LP, Lees T, et al. Outcome following carotid endarterectomy: lessons learned from a large international registry. Eur J Vasc Endovasc Surg. 2011;41:735–40.
25. Department of Health. Good doctors, safer patients: proposals to strengthen the system to assure and improve the performance of doctors and to protect the safety of patients. A report by the Chief Medical Officer. London: Department of Health; 2006.
26. Hewitt P. Trust, assurance, and safety – the regulation of health professionals in the 21st century. White paper presented to Parliament by the Secretary of State for Health by order of Her Majesty. London: Stationary Office; 2007.
27. Royal College of Surgeons. Clinical audit in surgical practice. 1st ed. London: RCS; 1989 (revised June 1995).
28. Gliklich RE, Dreyer NA, editors. Registries for evaluating patient outcomes: a user's guide. 2nd ed. Rockville: Agency for Healthcare Research and Quality (US); 2010. Chapter 10.
29. Rostami R, Nahm M, Pieper CF. What can we learn from a decade of database audits? The Duke Clinical Research Institute experience, 1997–2006. Clin Trials. 2009;6(2):141–50.
30. Arndt S, Tyrell G, Woolson RF, Flaum M, Andreason NC. Effects of errors in a multicenter medical study: preventing misinterpreted data. J Psychiatr Res. 1994;28:447–59.
31. Jha AK, Epstein AM. The predictive accuracy of the New York State coronary artery bypass surgery report-card system. Health Aff (Millwood). 2006;25(3):844–55.
32. Smith CR. The big chill: adverse effects of public reporting on access to health care. J Am Coll Cardiol. 2006;47(8):1737; author reply 1737–8.
33. Apolito RA, Greenberg MA, Menegus MA, Lowe AM, Sleeper LA, Goldberger MH, Remick J, Radford MJ, Hochman JS. Am Heart J. 2008;155(2):267–73.
34. Resnic FS, Welt FG. The public health hazards of risk avoidance associated with public reporting of risk-adjusted outcomes in coronary intervention. J Am Coll Cardiol. 2009;53(10):825–30.
35. Eisenstein EL, Lemons 2nd PW, Tardiff BE, Schulman KA, Jolly MK, Califf RM. Reducing the cost of phase II clinical trials. Am Heart J. 2005;149:482–8.
36. Venet D, Doffagne E, Burzykowski T, Beckers F, Tellier Y, Genevois-Marlin E, Becker U, Bee V, Wilson V, Legrand C, Buyse M. A statistical approach to central monitoring of data quality in clinical trials. Clin Trials. 2012;9:705–13.
37. Buyse M, George SL, Evans S, Geller NL, Ranstam J, Scherrer B, Lesaffre E, Murray G, Edler L, Hutton J, Colton T, Lachenbruch P, Verma BL. The role of biostatistics in the prevention, detection and treatment of fraud in clinical trials. Stat Med. 1999;18(24):3435–51.
38. The ESPS2 Group. European stroke and prevention study 2. Efficacy and safety data. J Neurol Sci. 1997;151(Suppl):S1–77.
39. Duley L, Antman K, Arena J, Avezum A, Blumenthal M, Bosch J, Chrolavicius S, Li T, Ounpuu S, Perez AC, Sleight P, Svard R, Temple R, Tsouderous Y, Yunis C, Yusuf S. Specific barriers to the conduct of randomized trials. Clin Trials. 2008;5(1):40–8.
40. Grimes DA, Hubacher D, Nanda K, Schulz KF, Moher D, Altman DG. The good clinical practice guideline: a bronze standard for clinical research. Lancet. 2005;366:172–4.
41. Tudur Smith C, Stokken DD, Dunn J, Cox T, Ganeh P, Cunningham D, Neoptolemos JP. The value of source data verification in a cancer clinical trial. PLoS One. 2012;7(12):e51623.
42. Uren SC, Kirkman MB, Dalton BS, Zalcberg JR. Reducing clinical trial monitoring resource allocation and costs through remote access to electronic medical records. J Oncol Pract. 2013;9(1):e13–6.
43. Beck P, Dagget S, Naert L, Polich V. The society of thoracic surgeons congenital heart surgery database audit: telligen final report. 2012: p. 4.
44. Jacobs JP, Mavroudis C, Jacobs ML, Maruszewski B, Tchervenkov CI, Lacour-Gayet FG, Clarke DR, Yeh Jr T, Walters 3rd HL, Kurosawa H, Stellin G, Ebels T, Elliott MJ, STS Congenital Database Taskforce; Joint EACTS-STS Congenital Database Committee. What is operative mortality? Defining death in a surgical registry database: a report of the STS Congenital Database Taskforce and

the Joint EACTS-STS Congenital Database Committee. Ann Thorac Surg. 2006;81(5):1937–41.
45. Overman DM, Jacobs JP, Prager RL, Wright CD, Clarke DR, Pasquali SK, O'Brien SM, Dokholyan RS, Meehan P, McDonald DE, Jacobs ML, Mavroudis C, Shahian DM. Report from the society of thoracic surgeons national database workforce: clarifying the definition of operative mortality. World J Pediatr Congenit Heart Surg. 2013;4(1):10–2.
46. Jacobs JP, Jacobs ML, Austin 3rd EH, Mavroudis C, Pasquali SK, Lacour-Gayet FG, Tchervenkov CI, Walters 3rd H, Bacha EA, Nido PJ, Fraser CD, Gaynor JW, Hirsch JC, Morales DL, Pourmoghadam KK, Tweddell JS, Prager RL, Mayer JE. Quality measures for congenital and pediatric cardiac surgery. World J Pediatr Congenit Heart Surg. 2012;3(1):32–47.
47. Jacobs JP, Edwards FH, Shahian DM, Haan CK, Puskas JD, Morales DLS, Gammie JS, Sanchez JA, Brennan JM, O'Brien SM, Dokholyan RS, Hammill BG, Curtis LH, Peterson ED, Badhwar V, George KM, Mayer Jr JE, Chitwood WR, Murray GF, Grover FL. Successful linking of The Society of Thoracic Surgeons Adult Cardiac Surgery database to centers for medicare and medicaid services medicare data. Ann Thorac Surg. 2010;90:1150–7.
48. Jacobs JP, Edwards FH, Shahian DM, Prager RL, Wright CD, Puskas JD, Morales DL, Gammie JS, Sanchez JA, Haan CK, Badhwar V, George KM, O'Brien SM, Dokholyan RS, Sheng S, Peterson ED, Shewan CM, Feehan KM, Han JM, Jacobs ML, Williams WG, Mayer Jr JE, Chitwood Jr WR, Murray GF, Grover FL. Successful linking of the Society of Thoracic Surgeons database to social security data to examine survival after cardiac operations. Ann Thorac Surg. 2011;92(1):32–9. PMID: 21718828.
49. Jacobs JP, Shahian DM, Edwards FH, O'Brien SM, Blackstone EH, Puskas JD, Schaffer J, Grover FL, Mayer Jr JE. Reply to letter to the editor: strategies to longitudinally track mortality. Ann Thorac Surg. 2012;94(2):693–4. PMID: 22818334.
50. Jacobs JP, O'Brien SM, Shahian DM, Edwards FH, Badhwar V, Dokholyan RS, Sanchez JA, Morales DL, Prager RL, Wright CD, Puskas JD, Gammie JS, Haan CK, George KM, Sheng S, Peterson ED, Shewan CM, Han JM, Bongiorno PA, Yohe C, Williams WG, Mayer JE, Grover FL. Successful linking of the Society of Thoracic Surgeons Database to Social Security data to examine the accuracy of Society of Thoracic Surgeons mortality data. J Thorac Cardiovasc Surg. 2013;145(4):976–83. doi:10.1016/j.jtcvs.2012.11.094. PMID: 23497944.
51. Boult M, Maddern GJ. Clinical audits: why and for whom? ANZ J Surg. 2007;77:572–8.

Part VI

Subspecialty Collaboration

Linking Databases

30

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Abstract

Multicenter datasets are increasingly utilized in pediatric cardiovascular research. In this chapter, we discuss the rational for linking information across data sources, and describe different linkage mechanisms which enable analyses not possible with individual datasets alone.

Keywords

Database • Outcomes • Congenital heart disease

Introduction

As discussed in Chap. 14, multicenter databases and registries are increasingly used in pediatric cardiovascular medicine. These datasets serve a variety of functions including research, benchmarking of performance, and quality improvement [1–11]. There are now numerous different clinical registries, administrative datasets, and other large multicenter data sources used for these purposes in the field. Many of these are described in detail in Chap. 14.

However, there are also certain limitations to consider. There are relatively few existing data sources, each containing a limited set of variables. In addition, the different datasets do not readily communicate with each other. Analyses of administrative databases can be further limited as these data sources rely on International Classification of Diseases, Ninth Revision (ICD-9) diagnosis and procedure codes to identify and classify patients which is known to have limited accuracy [12].

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Finally, there are limited current mechanisms for efficient collection of additional data points or new information to answer important clinical questions that may arise.

Rationale for Linking Databases

Linkage of existing data sources can address many of the limitations associated with the use of individual datasets [13]. Linking databases expands the pool of available data for analysis and capitalizes on the strengths of different types of data sources. Linkage allows analyses otherwise not possible with single center data or individual datasets alone. Linking datasets can be more time and cost efficient than creating additional new datasets, and can involve several different methodologies.

Mechanisms for Linking Data

Linking on Unique Identifiers

Some datasets contain unique patient identifiers such as social security number and these data can facilitate linkages with other data sources. For example, the Society of Thoracic Surgeons (STS) Adult Cardiac Surgery Database recently began collecting unique identifiers, including social security number which has enabled linking of this database to the Social Security Death Master File in order to evaluate long-term mortality [14, 15]. Unique identifiers were also recently incorporated into the STS Congenital Heart Surgery Database, and this may facilitate similar linkages in the future to assess long-term mortality in this cohort. This methodology has been successfully used in the outpatient pediatric cardiology realm as well, where investigators linked outpatient records regarding pediatric cardiology visits for chest pain to the National Death Index and Social Security Death Master File to evaluate for subsequent mortality in this cohort [16]. However, new limitations on the availability of the Social Security Death Master File for research purposes may pose a greater challenge to the use of this methodology in the future. There will likely be other challenges to consider in the pediatric population as well, as many neonates undergoing cardiac surgery may not have yet had a social security number assigned, for example.

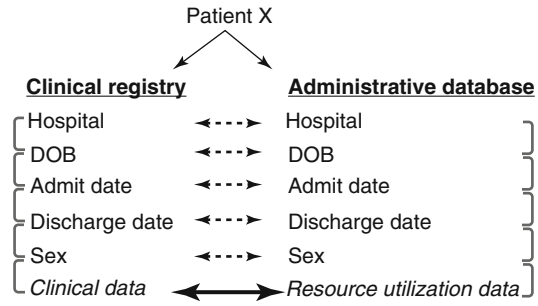

Fig. 30.1 Linkages Based on Indirect Identifiers. This figure describes how information on patient "X" entered into both a clinical registry and an administrative database may be linked through the use of indirect identifiers. Linkage on the indirect identifiers collected in both datasets (Hospital, DOB, Admit Date, Discharge Date, Sex) allows the information on patient X in one dataset to be linked to their information in the other dataset, such that clinical data and resource utilization data (for example) may be analyzed together. *DOB* date of birth

Linking on Indirect Identifiers

While linkage on direct or unique identifiers is the easiest way to accomplish linkages between datasets, these are often not collected or readily available for analysis in many databases due to a variety of regulatory requirements and concerns [17]. Thus, methodology has also been developed to link databases records through the use of "indirect" identifiers [18]. These include date of birth, date of admission, date of discharge, sex, and center where hospitalized. It has been shown that nearly all records at a given center can be uniquely identified using these indirect identifiers, and that a crosswalk can then be created between two datasets, linking patients on the values of center where hospitalized and the indirect identifiers (Fig. 30.1). This method has been used to successfully link adult cardiac databases [18].

Our group recently adapted this methodology to successfully link two large pediatric data sources [13]. The STS Congenital Heart Surgery Database is the largest pediatric heart surgery registry worldwide and contains detailed pre-operative, operative, and outcomes data on all patients undergoing pediatric heart surgery at participating centers. However, the STS Database does not collect resource utilization data. The Pediatric Health Information Systems (PHIS) Database is a large administrative database containing inpatient data from >40 US children's hospitals. It contains valuable resource utilization information such as total charges (which can be used to estimate costs), as well as data regarding medications, laboratory test, imaging, etc.

Linking these two datasets allows us to utilize the detailed operative data from the STS Congenital Heart Surgery Database, and the resource utilization data from the PHIS Database [13]. Both datasets contain useful information regarding various patient and center factors and outcomes. All of these data may be pooled together for analysis.

From 2004 to 2008, 30 US centers submitted data to both the STS Congenital Heart Surgery Database and the PHIS Database, accounting for approximately 45,000 admissions and encompassing a wide range of center surgical volume and geographic region [13]. Through using the methodology of linking on indirect identifiers, we successfully linked data on 90 % of eligible patients. We were also able to validate the linkage methodology, demonstrating in a 10 % sample of records that 100 % of the records linked using indirect identifiers were indeed true matches bases on evaluation of medical record number [13]. We have subsequently updated the link through 2010 such that linked data is now available for more than 60,000 patients from 33 US centers [19, 20]. The data have been used to successfully conduct several comparative effectiveness and health economic studies that would not have been possible using either dataset alone. For example, different regimens of peri-operative corticosteroids and anti-fibrinolytic medications have been evaluated using the linked dataset [19, 20]. We have also evaluated clinical and resource utilization outcomes associated with hospital rates of post-operative infection, and have examined the variability in hospital costs across centers performing pediatric heart surgery [21].

Center-Level Linkages

Linking registry data to other center level data through matching on center can be easily accomplished. For example, a survey regarding different center-level models of intensive care unit (ICU) care was successfully linked to the STS Congenital Heart Surgery Database. This linkage enabled evaluation of the association of the variables collected in the survey with outcomes data collected in the STS Database [22]. A similar linkage involving merging of hospital-level data regarding nursing practices with outcomes data is also ongoing.

Supplementary Data Modules

Data linkages can also be accomplished through the development and use of a modular data collection system that enables collection of supplemental data points to the main registry. The modules are generally web-based and can be quickly created and deployed to allow "real-time" collection of additional data. They are more time and cost-efficient compared with traditional data collection methods that may duplicate data already being collected in the main registry. This methodology has been recently successfully used by the Pediatric Cardiac Critical Care Consortium (PC^4) to design a module to collect supplemental data to their main registry to study the relationship between Vasoactive-Inotropic Score (VIS) and outcome after infant cardiac surgery [23]. This methodology allowed for efficient data collection, with 391 infants prospectively enrolled in the study and data collection completed during a span of 6 months. Supplementary data modules have also

been used within the STS Congenital Heart Surgery Database to collect pilot data on various quality measures in order to evaluate potential issues with definitions and data collection before these measures were considered for incorporation into the main database [24].

Collaboration/Partnering Between Databases

Data can also be shared or linked through collaboration and partnering between different organizations and datasets. For example, beginning in 2010 the STS and the Congenital Cardiac Anesthesia Society collaborated to add a new anesthesia section to the STS data collection forms [25]. Anesthesia data are now collected, harvested, reported, and analyzed along with surgical data for participating centers. This approach proved to be more time and cost efficient than creating a separate anesthesia database in which many of the fields regarding patient characteristics and the operative procedure would have been duplicated between databases. Another example of partnering between databases involves the ongoing collaboration between the Congenital Heart Surgeon's Society (CHSS) and STS. This collaboration involves leveraging the information collected in the STS Congenital Heart Surgery Database in order to help participating sites better identify patients who may be eligible for enrollment in certain CHSS longitudinal cohort studies.

Future Directions

To facilitate further linkages and expand the potential for sharing of data between multiple sources, there has been increasing interest in the creation of a Global Unique Identifier (GUID) [26]. Developed by the autism research community, the GUID allows multiple linkages and also maintains privacy. It is generated based on a set of simple identifiers unique to the patient, and undergoes encryption before being shared with a central system so that identifiers are never transmitted or stored outside the local site [27]. In autism research, the GUID is used to track patients between studies and forward in time. The National Heart, Lung, and Blood Institute Pediatric Heart Network is currently investigating mechanisms to incorporate a GUID into pediatric cardiac datasets [26].

Better integration of data in the future will also likely require investigators to take advantage of current technologies such as social media and mobile devices which may allow more efficient engagement with patients and better collection of patient reported quality of life and functional outcomes compared with older methodology relying on surveys delivered via phone or mail. Investigating better ways to harness the increasing amount of data collected in the electronic medical record will also be important.

Conclusions

In summary, linkage of information across a variety of pediatric cardiac datasets is possible and can involve several different methodologies. Linkage capitalizes on the strengths, and mitigates some of the weaknesses, of different types of data sources and can allow analyses not otherwise possible with single center data or individual datasets alone. Further development of this methodology may facilitate analyses not only of isolated short-term outcomes associated with a particular operation, device, or medication, but enable longitudinal evaluation of the child diagnosed with congenital heart disease who may receive multiple different therapies during their lifetime.

References

1. Pasquali SK, Hall M, Li JS, Peterson ED, Jaggers J, Lodge AJ, Marino BS, Goodman DM, Shah SS. Corticosteroids and outcome in children undergoing congenital heart surgery: analysis of the Pediatric Health Information Systems Database. Circulation. 2010;122:2123–30.
2. Fudge JC, Li S, Jaggers J, O'Brien SM, Peterson ED, Jacobs JP, Welke KF, Jacobs ML, Li JS, Pasquali SK. Outcomes in patients with Down syndrome undergoing congenital heart surgery: analysis of a national clinical database. Pediatrics. 2010;126:315–22.

3. Karamlou T, Hirsch J, Welke K, Ohye RG, Bove EL, Devaney EJ, Gajarski RJ. A United Network for Organ Sharing analysis of heart transplantation in adults with congenital heart disease: outcomes and factors associated with mortality and retransplantation. J Thorac Cardiovasc Surg. 2010;140:161–8.
4. Pasquali SK, Jacobs JP, He X, Hornik CP, Jaquiss RDB, Jacobs ML, O'Brien SM, Peterson ED, Li JS. The complex relationship between center volume and outcome in patients undergoing the Norwood operation. Ann Thorac Surg. 2012;93:1556–62.
5. Welke KF, Diggs BS, Karamlou T, Ungerleider RM. The relationship between hospital surgical case volumes and mortality rates in pediatric cardiac surgery: an national sample, 1988–2005. Ann Thorac Surg. 2008;86:889–96.
6. Jacobs JP, Jacobs ML, Mavroudis C, Lacour-Gayet FG, Tchervenkov CI, Pasquali SK. Executive summary: the Society of Thoracic Surgeons Congenital Heart Surgery Database – 18th Harvest – (January 1, 2009 – December 31, 2013). The Society of Thoracic Surgeons (STS) and Duke Clinical Research Institute (DCRI), Duke University Medical Center; Durham; 2013 Harvest.
7. Kugler JD, Beekman RH, Rosenthal GL, Jenkins KJ, Klitzner TS, Martin GR, Neish RS, Lannon C. Development of a pediatric cardiology quality improvement collaborative: from inception to implementation. From the Joint Council on Congenital Heart Disease Quality Improvement Task Force. Congenit Heart Dis. 2009;4:318–28.
8. Pediatric Congenital Cardiac Surgery in New York State 2006–2009. Available at: http://www.health.ny.gov/statistics/diseases/cardiovascular/heart_disease/docs/2006-2009_pediatric_congenital_cardiac_surgery.pdf. Accessed 7/10/2013.
9. U.S. News & World Report's Annual Ranking of Best Hospitals. Available at: http://www.rti.org/besthospitals. Accessed 28 May 2013.
10. OptumHealth Congenital Heart Disease Center of Excellence. Available at: https://www.myoptumhealthcomplexmedical.com/gateway/public/chd/providers.jsp. Accessed 28 May 2013.
11. Martin GR, Beekman RH, Ing FF, Jenkins KJ, McKay CR, Moore JW, Ringel RE, Rome JJ, Ruiz CE, Vincent RN. The IMPACT registry: improving pediatric and adult congenital treatments. Semin Thorac Cardiovasc Surg Pediatr Card Surg Annu. 2010;13:20–5.
12. Pasquali SK, Peterson ED, Jacobs JP, He X, Li JS, Jacobs ML, Gaynor JW, Hirsch JC, Shah SS, Mayer JE. Differential case ascertainment in clinical registry vs. administrative data and impact on outcomes assessment in pediatric heart surgery. Ann Thorac Surg. 2013;95:197–203.
13. Pasquali SK, Jacobs JP, Shook GJ, O'Brien SM, Hall M, Jacobs ML, Welke KF, Gaynor JW, Peterson ED, Shah SS, Li JS. Linking clinical registry data with administrative data using indirect identifiers: implementation and validation in the congenital heart surgery population. Am Heart J. 2010;160:1099–104.
14. Jacobs JP, Edwards FH, Shahian DM, et al. Successful linking of the Society of Thoracic Surgeons adult cardiac surgery database to Centers for Medicare and Medicaid Services Medicare data. Ann Thorac Surg. 2010;90:1150–7.
15. Jacobs JP, Edwards FH, Shahian DM, et al. Successful linking of the STS database to social security data to examine survival after cardiac surgery. Ann Thorac Surg. 2011;92:32–7.
16. Saleeb SF, Li WYV, Warren SZ, Lock JE. Effectiveness of screening for life-threatening chest pain in children. Pediatrics. 2011;128:e1062–8.
17. Dokholyan RS, Muhlbaier LH, Falletta J, et al. Regulatory and ethical considerations for linking clinical and administrative databases. Am Heart J. 2009;157(6):971–82.
18. Hammill BG, Hernandez AF, Peterson ED, Fonarow GC, Schulman KA, Curtis LH. Linking inpatient clinical registry data to Medicare claims data using indirect identifiers. Am Heart J. 2009;157:995–1000.
19. Pasquali SK, Li JS, He X, Jacobs ML, O'Brien SM, Hall M, Jaquiss RDB, Welke KF, Peterson ED, Shah SS, Gaynor JW, Jacobs JP. Perioperative methylprednisolone and outcome in neonates undergoing heart surgery. Pediatrics. 2012;129:e385–91.
20. Pasquali SK, Li JS, He X, Jacobs ML, O'Brien SM, Hall M, Jaquiss RDB, Welke KF, Peterson ED, Shah SS, Jacobs JP. Comparative analysis of antifibrinolytic medications in pediatric heart surgery. J Thorac Cardiovasc Surg. 2012;143:550–7.
21. Pasquali SK, He X, Jacobs ML, Hall M, Gaynor JW, Shah SS, Peterson ED, Hill KD, Li JS, Jacobs JP. Hospital variation in post-operative infection and outcome following congenital heart surgery. Ann Thorac Surg. 2013;96(2):657–63.
22. Burstein DS, Jacobs JP, Sheng S, et al. Care models in congenital heart surgery and associated outcomes. Pediatrics. 2011;127:6.e1482–9.
23. Gaies M, Jeffries H, Niebler R, Pasquali SK, Donohue J, Yu S, Thiagarajan R. Vasoactive inotropic score (VIS) is associated with outcome after infant cardiac surgery: a report of the Pediatric Cardiac Critical Care Consortium (PC4). J Am Coll Cardiol. 2013;61:E424.
24. Jacobs JP, Jacobs ML, Austin EH, Mavroudis C, Pasquali SK, Lacour–Gayet FG, Tchervenkov CI, Bacha EA, del Nido PJ, Fraser CD, Gaynor JW, Hirsch JC, Morales DLS, Pourmoghadam KK, Tweddell JT, Prager RL, Mayer JE. Quality measures for congenital and pediatric cardiac surgery. World J Pediatr Congenit Heart Surg. 2012;3:32–47.
25. Vener DF, Jacobs JP, Schindler E, Maruszewski B, Andropoulos D. Databases for assessing the outcomes of the treatment of patients with congenital and paediatric cardiac disease – the perspective of anaesthesia. Cardiol Young. 2008;18 Suppl 2:124–9.
26. Pearson GD, Kaltman JR, Lauer MS. Evidence-based medicine comes of age in pediatric cardiology. J Am Coll Cardiol. 2013;61:2565–7.
27. Johnson SB, Whitney G, McAuliffe M. Using global unique identifiers to link autism collections. J Am Med Inform Assoc. 2010;17:689–95.

Part VII

Longitudinal Follow-Up

Use of National Death Registries to Empower Databases in Reporting Longitudinal Follow-Up

31

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Abstract

National databases are beginning to play a key role in the care of patients by predicting prognosis of particular treatment, defining risk factors, and aiding with the selection of patients for particular treatments. These databases also help to establish standards of care, which are gaining importance as government is beginning to emphasize on "pay for performance" programs. Medical societies are therefore establishing databases that can provide relevant and accurate clinical information for these purposes. The increase in number of medical societies using their own database for research and improvement in quality of care has sparked new challenges. Societies particularly related to procedure-based specialties usually lack the ability to record longitudinal follow-up data for mortality, as providers in these specialties often follow their patients only for a short period of time after the procedure. This limitation in longitudinal data regarding "late" mortality can be overcome by linking these national databases with national registries of death. Herein, an example of a surgical society is presented to demonstrate how a national death registry is used to empower a national database. The information gathered, the comparisons outlined, and the processes used to determine the best combination of indices of death for this society should be translatable and hopefully useful for other societies and registries who wish to empower their databases with long-term national data about mortality.

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A growing number of medical societies have formed their own databases in view of their goal to provide high quality patient care through education and research. These databases have been instrumental in

- providing insight in various therapies,
- creating benchmarks,
- standardizing the reporting of outcomes,
- educating patients, and
- interacting with industry, payers, and government.

Surgical specialties have relied more on these databases to help with the care of patients by predicting prognosis, defining risk-factors, and aiding with the selection of patients who are the best candidates for particular procedures. Above all, for most surgical specialties, as governmental "pay-for-performance" initiatives evolve, these databases will be contributory in establishing the standards of care and benchmarks of performance. If these accurate clinical databases from individual societies are not available, government will use administrative databases that are available and infer the data from them. These inferences from administrative claims data will not be a correct representation of the population or their outcomes.

An inherent issue with databases of surgical societies is that after a short-term post-operative course, the surgeons are no longer the primary care provider for the patients. Therefore, their ability to track the long-term outcomes of the patients is limited, resulting in databases that usually lack the ability to report long-term outcomes. This lack of data about longitudinal outcomes represents a significant limitation of otherwise robust databases such as "The Society of Thoracic Surgeons National Database" which has more than 5.1 million surgical case records in its three components: Adult Cardiac Surgery, General Thoracic Surgery, and Congenital Heart Surgery [1].

The addition of long-term follow-up data to an otherwise comprehensive database has many advantages including:

- comparing novel surgical techniques,
- enabling longitudinal comparative effectiveness research comparing different treatments
- facilitating longitudinal surveillance of implanted medical devices, and
- educating patients about their long-term prognosis.

Herein, the Society of Thoracic Surgeons (STS) is used as an example to demonstrate how a professional medical society can use a national death registry to empower a national clinical database.

"Founded in 1964, The Society of Thoracic Surgeons is a not-for-profit organization representing over 6,800 surgeons, researchers and allied health care professionals worldwide who are dedicated to ensuring the best possible outcomes for surgeries of the heart, lung, and esophagus, as well as other surgical procedures within the chest." ([www.sts.org], accessed April 26, 2014). The mission of STS is "to enhance the ability of cardiothoracic surgeons to provide the highest quality patient care through education, research, and advocacy." By sending in their data about outcomes to the Society of Thoracic Surgery Database, surgeons are committing to improving the quality of care that their cardiothoracic surgical patients receive. In January 2007, STS established a Database Task Force to investigate ways to strengthen the database by extending its long-term follow-up. Key outputs of this initiative include:

- the inclusion of "personal health information" in the STS Database that can facilitate both longitudinal follow-up and linkages of databases
- the creation of strategies to link to other databases such as the National Cardiovascular Data Registry (NCDR) of the American

College of Cardiology, the database of the Centers for Medicare and Medicaid Services of the United States of America, the Pediatric Health Information System (PHIS) database, and various national death registries, using both probabilistic matching (matching with indirect identifiers) and deterministic matching (matching with direct identifiers).

These strategies greatly increase the ability of STS to help patients and physicians, facilitate research, support education and advocacy, and improve quality.

National Death Registries

Mortality is still considered the most important long-term outcome by patients, clinicians, and third parties, including government and industry. Therefore, the STS Task Force did due diligence regarding accessibility and accuracy of national registries of death and mainly focused its attention to the two most comprehensive registries of death in the United States of America:

- the National Death Index (NDI), and
- the Social Security Death Master File (SSDMF).

The linking of a national mortality database to a surgical database has been successfully accomplished in United Kingdom where the Central Cardiac Adult Database was linked to a national registry of death obtained by the Office of National Statistics using patients' National Health Service number [2, 3].

The National Death Index of the United States of America

Death of a person in the United States, irrespective of their citizenship, is reported to the Office of Vital Statistics of each state and a certificate is generated. This record is then used to create a registry of death at the state level. Therefore, before the establishment of the National Death Index, if someone wanted to verify the true mortality in the United States within a specific registry or database, it would have been necessary to send a standard form to each of the offices of the vital statistics in all 50 states. The office for the state would cross-reference the "personal health information" given by the investigator with the registry of the death of the state and determine which subjects in the database of the investigator had died in that state. This process would take at least 8 months to 1 year to complete, but, in the past, was the most accurate and only mechanism to determine true mortality, regardless of citizenship or age, in the United States of America.

In the 1970s, a work force committee of the National Center of Health Statistics recommended a national registry of death, or "national death index" [4], be created by working with all 50 states to centralize all state registries of death into one national registry of death. Since only the states have the authority to collect the certificates of death, the "National Death Index" could not mandate the collection of their registries. The state governments agreed to participate if their data would only be used for research. With this understanding, monetary contracts were negotiated with all 50 states to obtain their registries of death yearly.

The National Death Index began in 1979 and activated for use in 1982. Initially, the National Death Index was partially funded by the National Institute of Health, but it became self-supportive in 1992. The National Death Index is part of the Division of Vital Statistics, which falls under the authority of the National Center of Health Statistics, a division of the Centers for Disease Control and Prevention. Yearly harvests of the registries of death from the states are done 12 months after the end of each calendar year. The National Death Index then takes approximately 1 year to organize, verify, audit, and insert the data into the database. This entire process leads to a 2-year delay in the availability of the information. Therefore, in 2007, all the deaths from 1979 to 2005 were available.

To access the database, one must first submit an application stating how the information learned from the National Death Index will be utilized. There is a list of acceptable uses and criteria found on the website of the National Death Index [4]. The overriding criterion is that the

National Death Index can only be used in medical or healthcare research. It cannot be used for legal or administrative purposes. Once the application is submitted, it takes approximately 2–3 months to be processed and approved. Submission of records to the National Death Index is quite specific. One submits as many of the 9 National Death Index pre-determined items of "personal health information" into the format of an "American Standard Code for Information Interchange text file" (ASCII) as described on the National Death Index website [4]:

- social security number
- day/month/year of birth
- first name, middle initial, last name
- father's surname
- gender
- marital status
- state of birth
- state of residence
- race

The National Death Index retrieval program is the software used to determine if a particular record of death in the National Death Index qualifies as a possible match with a submitted user record. To qualify as a match, both records must share at least 1 of 7 matching criteria listed:

- Social Security Number
- Exact month and year of birth, first and last name
- Exact month and year of birth, first and middle initials, last name
- Exact month and day of birth, first and last name
- Exact month and day of birth, first and middle initials, last name
- Exact month and year of birth, first name, father's surname
- If the subject is female: exact month and year of birth, first name, last name and father's surname.

If a user record satisfies 1 of the 7 matching criteria with one or more National Death Index records, the National Death Index records are retrieved. The software then analyzes each of the matching National Death Index records with the entire "personal health information" in the user record. It determines not only how many "personal health information" data points match but the "quality" of the matching "personal health information" data. For example, matching social security numbers are of higher quality than matching first names. The retrieval program uses this analysis to determine a probabilistic score for each matching record in the National Death Index. The score represents the probability that the record of death in the National Death Index is a true match with the submitted record of the user. The report of the National Death Index for a particular submitted record lists in descending order all matching records in the National Death Index with the probabilistic score of each record.

The program also determines if the matched record in the National Death Index with the highest probabilistic score meets criteria so that the user record is marked "probably dead". This determination is based on the probabilistic score and the type and quantity of "personal health information" given by the user for this particular record. Per internal audit, the sensitivity of the National Death Index to capture death of a submitted record is 92–98 %, and if it determines that the patient is "probably dead", the specificity is 98 % [5].

The use of National Death Index is costly, as this project is self-sufficient and receives no federal funding. The National Death Index has to pay every state a fee each year to gain access to their registry. Additionally, it pays to each state a fee every time one of the records in state's registry is matched to a submitted request. To the user, the National Death Index costs US$ 350 to submit a request initially and US $100 for each subsequent submission under the same protocol. The cost of each record searched each year is US $ 0.15 and US $ 0.21 for with and without cause of death, respectively [6]. This can sum up to a considerable amount of money when considering large cohort studies over a long period of time. To give an example, if a cohort of 1,000 patients undergoing a certain procedure over a 5 year period are submitted to National Death Index to establish mortality and its cause. It will cost the investigator a sum of US $630, but if the cohort is increased to 1,000,000 patients and study time to 7 years, the cost will be US $1,176,000 [5].

National Death Index is to date the most accurate and uncontaminated registry of death records in the United States because, unlike other registries of death, it is based on actual certificates of death, which are filled out for every death in the United States of America, regardless whether the deceased is a citizen or non-citizen or a child or adult. The National Death Index does not rely on any third party or extracted data from any other database; and therefore, the National Death Index is considered the "gold standard" for determining true mortality in the United States of America.

Social Security Administration Death Master File (Index)

The "Social Security Death Master File", otherwise known as the "Social Security Death Index" was created in the year 1980 by the Social Security Administration (SSA). It contains information on anyone assigned a Social Security Number (SSN) whose death was reported to the SSA any time after 1962.

This chapter will first describe the history of the Social Security Death Master File and its status as of November 2011. ***In November 2011, major changes in the availability of data from the Social Security Death Master File dramatically decreased the utility of the Social Security Death Master File***. These changes are discussed in the final section of this chapter titled: "Future Challenges".

The Social Security Death Master File is part of the SSA's numerical identification database called "NUMIDENT" Each entry in the Death Master File contains the following variables:

- name of the person
- social security number
- date of birth
- month and year of death
- date of death if the record is from 2000 or later
- state or country of residence
- zip code of last residence
- zip code where "death benefit payment" was sent.

The Social Security Administration is not permitted to release to the public, in any form, data about death obtained from a state under the auspices of section 205(r) of the Social Security Act, unless given permission by the state. All of the states do give their data from their registries of death to the Social Security Administration to be used for internal government functions. However, half the states do not give permission to the Social Security Administration to use that data for the Death Master File or any other dissemination to the public.

Therefore, the data in the Death Master File is retrieved from other sources. Approximately 90 % of the reported deaths come from funeral homes and family members. The remaining 10 % come from various financial agencies and/or the postal service. The majority of deaths are reported to the Social Security Administration in order to receive monetary benefits such as a burial benefit, for which most are eligible. In October of 1981, Law P.L. 97–35 greatly reduced the number of people eligible for such benefits and thus the number of deaths reported to the Social Security Administration significantly decreased. This problem was remedied in 1989 by the Death Benefit Enunciation, which restored the death benefit to even more families than before. This legislation is thought to have made the Death Master File more robust than it had ever been.

A single issue of the Death Master File, which is published quarterly, costs US$1730. This option is the most economical approach for someone who needs to sample the National Vital Statistics at a given time or only yearly. This file includes data from 1937 up to 2 months prior to the request date.

When the Social Security Death Master File was compared to the National Death Index for accuracy, it was 95, 97.8, and 99.6 % for years 1990, 1995, and 1999 respectively [5]. It is impressive, however the sensitivity and specificity by external reviews usually including the known period of poor collection of data has been lower, around 73–83 % [7–9]. The sensitivity and specificity increases with increasing age, and therefore, for a more elderly Veterans Affairs population the reported sensitivity was 92.1 % [5]. Conversely, the specificity of the Social Security Death Master File drops precipitously with age.

Limitation with the Death Master File is that it is based upon availability of specific parameters:

- that the deceased was issued a Social Security Number
- that the deceased did not change their name
- that his or her death was reported to the Social Security Administration.

The Social Security Administration Vital Status Service

The Death Master File is not the only source of vital statistics that the social security offers; it also offers Vital Status Service to Epidemiological Researchers [10]. This service is available only for use in research that has been determined to contribute to a national health interest. This service searches the following three government databases for the vital status of an individual. The service informs the user not only of probable death but of probable living:

- NUMIDENT which contains information from Social Security cards and includes the Death Master File
- the Master Beneficiary Record, which contains information on all individuals receiving any governmental benefits, and
- the Master Earnings File, which contains information about earnings on all persons that both work and have a residence.

The Vital Status Service requires the following six parameters for each submitted record:

- the first name of the patient
- the middle name of the patient
- the last name of the patient
- date of birth
- social security number, and
- sex

If any of this is not provided, this service cannot be used. This requirement makes it difficult for cohorts that have predominantly pediatric population or if it is a retrospectively collected database. The cost of using Vital Status Service is 10 % less than using the National Death Index, especially for large cohorts. If the "living status" of the person is not required than Vital Status Service does not provide any advantage over Death Master File as it uses the Death Master File to determine the "death status" of an individual.

Which National Death Registry Is Better?

Comparison of two national death registries is summarized in Table 31.1. The National Death Index and the Social Security Death Index have been compared in the past [11]. When a society is deciding on which national registry of death to use to link to their database; there are many important factors to be considered such as accuracy, accessibility, and cost.

Accuracy

The National Death Index is the most accurate and reliable way to determine mortality in the United States because it is based on certificates of death. Age, citizenship, or socioeconomic class does not affect the issuing of a certificate of death, and thus does not affect the National Death Index. Therefore, the National Death Index is the "Gold Standard". The Death Master File is known to be most accurate for older patients who are beneficiaries of Social Security. The limitations in people, who are not beneficiaries of governmental funds, children, and non-citizens, are well known. There are also eras in which reporting of deaths to the Social Security Association was limited. However, since 1995, the sensitivity of the Death Master File has been greater than 97 %, reaching 99 % in the past few years. Also, over the past 5 years, internal auditing has demonstrated a specificity of 93 %. As an increasing number of governmental databases are integrated, the accuracy of the Death Master File will continue to increase.

Accessibility

In regards to accessibility, the National Death Index is limited in that there is a 2-year lag

Table 31.1 Comparison of National Death Registries

National death index		Death master file	
Pros	Cons	Pros	Cons
Gold standard (most accurate)			Less accurate depending on patient population & era (<1995) In November 2011, major changes in the availability of data from the Social Security Death Master File dramatically decreased the utility of the Social Security Death Master File
Sensitivity same for all		Good sensitivity for older adults (i.e. geriatric)	Poor sensitivity for certain populations (i.e. children)
	Very expensive	Inexpensive (one-time fee)	
	Cost dependent on number of reports, and number of patients and years in analysis	Cost independent of reports rum or number of years or patients in analysis	
Minimal data management			Complete data management
Returns probability of match; highly specific	Create program to manage data & report results to your specific needs		
Accuracy should not change			Accuracy sensitive to governmental changes in policy
	2 year delay for data availability, "Real-Time" yearly reporting impossible	Yearly reports are available with capture of 99.5 % of deaths over the past year, "Real-Time" yearly reporting possible	
	Use of data restricted to research ONLY	No restriction on the use of data	

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between collection of data and access of the user to that data. This delay makes "real-time" yearly reporting of mortality not possible with the National Death Index. However, one can update a prospective database monthly using the Death Master File.

The National Death Index is to be used for research only and cannot be used for administrative functions or interactions with payers. The Death Master File has no restriction in the use of its information. Although the primary goals of a medical society for a database are to improve the care of patients, educate patients and physicians, and improve the performance of physicians, the database will eventually be used to interact with industry, third party payers, and the government. It can be argued that the primary function of the databases of The Society of Thoracic Surgeons is "healthcare operations", which is not research. This limitation of the National Death Index must therefore be considered.

Cost

In regards to cost, the National Death Index is much more expensive when compared to the Death Master File. The cost of using the National Death Index to review retrospectively 10 years of records from The Adult Cardiac Surgery Database of The Society of Thoracic Surgeons is US$1,881,638.25 [5]. In compari-

son, the cost to perform the same analysis using the Social Security Death Master File is US$1,730.00 plus the cost of creating a program to cross reference the database of The Society of Thoracic Surgeons with the Death Master File. This program can be written to the preferences of the Society in terms of what data should be weighted stronger for matches, how much data is needed to be considered a match, and how matches are reported. The sensitivity and specificity of the results can therefore mostly be determined by the programmer. Also, once this program is created, it can be used to harvest mortality from the Death Master File in any increment of time and for any patient cohort. The cost of creating reports retrospectively becomes independent of the number of patients and years to be reviewed as well as to the number of reports one wishes to create. This cost-savings alone is a tremendous advantage of the Death Master File over the National Death Index. It is also not possible to collect data prospectively and give yearly "real-time" reports about mortality in a growing database using the National Death Index because of the 2-year delay in data collection. Therefore the report in 2014 would contain data about mortality through 2012. If one uses the Death Master File, one can give a yearly report on the mortality of the entire database, or for any particular procedure or patient population, within months of the end of the year. Once the matching program is created, the cost would be US$1,730.00 per year.

In the current environment, it is essential to link national death registries to larger multi-institutional databases for many reasons as outline elsewhere in this chapter. Linking the database to a death index like the Death Master File, that allows the Society of Thoracic Surgeons Database to use this powerful information about mortality in any manner that is beneficial to its patients and physicians, is essential. The flexibility to create multiple reports on specific procedures, populations of patient, or of particular eras, with no additional cost, is extremely beneficial. Yearly reports of mortality in a variety of different cohorts of patient and follow-up periods with no delay in time would be a tremendous service to our specialty. Also, the majority of data in The Society of Thoracic Surgeons database is after 1995 and consists of adults. Therefore, multiple reasons supported the decision to use the Social Security Death Master File as the primary index of death for the Society of Thoracic Surgeons. However, if there is a known cohort of patients that is easily identifiable and known to be grossly underrepresented in the Social Security Death Master File, such as the Society of Thoracic Surgeons Congenital Heart Surgery Database, one can consider using the National Death Index for that particular group. The importance of linking one's registry to an accurate national registry of death should not be underestimated. When choosing an index of death with which to link, it is important to examine multiple factors in one's own database including

- its patient populations
- its eras of data collection
- its available "patient health information", and
- its goals in regards to cost, reporting, and services provided to members and perhaps payers.

The Society of Thoracic Surgery has reported successfully linking their database to Social Security Death Master File on different occasions [1, 12]. One significant limitation that came across is the under-reporting of social security numbers by data participants and therefore inability to link it to the Social Security Death Master File. However, with various efforts by The Society of Thoracic Surgery over the recent years, more and more centers are reporting completed social security numbers data [12]. It is important to note that the methodology used by The Society of Thoracic Surgeons to harvest, analyze, and report these data is compliant with all federal regulations [13].

Although herein the databases of The Society of Thoracic Surgeons are used as an example, the information gathered, and the processes used to determine the best combination of indices of death for this society, should be translatable and hopefully useful for other societies and registries who wish to empower their databases with long-term national data about mortality.

Future Challenges

As mentioned earlier in this chapter, the Social Security Death Master File contains data from different federal and states sources. As of November 2011, Social Security Administration announced that death information from states cannot be disclosed except to federal benefit-paying agencies [14]. Almost 40 % of the information in Death Master File comes from states; therefore, Blackstone [14] concluded that "this renders the Social Security Death Master File useless for biomedical research". Societies now have to explore legislative means to address this issue in order to be able to resuscitate this important source of vital status.

Alternatively societies have to investigate other reliable sources of death data. The Society of Thoracic Surgeons has already demonstrated using Centers of Medicare and Medicaid Services data for Medicare beneficiaries can empower its database [15]. Similarly, the National Death Index data may be an alternative if some of the obstacles mentioned earlier in this chapter are resolved. Jacobs and colleagues proposed potential solutions to these problems with the National Death Index [1, 16]. These would include:

- the National Death Index allowing medical societies to verify life status for a society's healthcare operations;
- release the death data with a lag time of only 6 months instead of previous lag time of 2 years, and
- development of a less expensive, "bulk purchasing" platform for routine use of its data by medical societies.

Professional medical and surgical societies with databases, especially surgical ones, should not underestimate the benefit of empowering their data with long-term outcomes like re-admission, re-operation, and death. Only by ensuring there is accurate data that is weighted appropriately for factors that affect outcome, can physicians have the opportunity to be a partner with payers and governmental agencies, rather than a recipient of the changing environment of pay for performance.

References

1. Jacobs JP, O'Brien SM, Shahian DM, Edwards FH, Badhwar V, Dokholyan RS, Sanchez JA, Morales DL, Prager RL, Wright CD, Puskas JD, Gammie JS, Haan CK, George KM, Sheng S, Peterson ED, Shewan CM, Han JM, Bongiorno PA, Yohe C, Williams WG, Mayer JE, Grover FL. Successful linking of the Society of Thoracic Surgeons Database to Social Security data to examine the accuracy of Society of Thoracic Surgeons mortality data. J Thorac Cardiovasc Surg. 2013;145(4):976–83. doi:10.1016/j.jtcvs.2012.11.094. PMID: 23497944.
2. Gibbs JL, Monro JL, Cunningham D, Rickards A. Survival after surgery or therapeutic catheterisation for congenital heart disease in children in the United Kingdom: analysis of the central cardiac audit database for 2000–1. BMJ. 2004;328:611.
3. Maruszewski B, Lacour-Gayet F, Monro JL, Keogh BE, Tobota Z, Kansy A. An attempt at data verification in the European Association for Cardio-Thoracic Surgery Congenital Database. Eur J Cardiothorac Surg. 2005;28:400–4.
4. Centers for Disease Control and Prevention, National Death Index. http://www.cdc.gov/nchs/ndi.htm. Accessed June 2013.
5. Morales DLS, McClellan AJ, Jacobs JP. Empowering a database with national long-term data about mortality: the use of national death registries. In: 2008 Supplement to Cardiology in the Young: Databases and The Assessment of Complications associated with The Treatment of Patients with Congenital Cardiac Disease, Prepared by: The Multi-Societal Database Committee for Pediatric and Congenital Heart Disease, Jeffrey P. Jacobs, MD (editor). Cardiol Young. 2008;18 Suppl 2:188–95.
6. National Death Index User Fees. http://www.cdc.gov/nchs/data/ndi/Users_Fees_Worksheet.pdf. Accessed June 2013.
7. Schisterman EF, Whitcomb BW. Use of the Social Security Administration Death Master File for ascertainment of mortality status. Popul Health Metr. 2004;2:2.
8. Cowper DC, Kubal JD, Maynard C, Hynes DM. A primer and comparative review of major U.S. mortality databases. Ann Epidemiol. 2002;12:462–8.
9. Buchanich JM, Dolan DG, Marsh GM, Madrigano J. Underascertainment of deaths using Social Security Records: a recommended solution to a little-known problem. Am J Epidemiol. 2005;162:193–4.
10. Service to Epidemiological Researchers to provide Vital Status Data on Subjects of Health Research.

http://www.ssa.gov/policy/about/epidemiology.html. Accessed June 2013.
11. Lash TL, Silliman RA. A comparison of the National Death Index and Social Security Administration databases to ascertain vital status. Epidemiology. 2001;12:259–61.
12. Jacobs JP, Edwards FH, Shahian DM, Prager RL, Wright CD, Puskas JD, Morales DL, Gammie JS, Sanchez JA, Haan CK, Badhwar V, George KM, O'Brien SM, Dokholyan RS, Sheng S, Peterson ED, Shewan CM, Feehan KM, Han JM, Jacobs ML, Williams WG, Mayer Jr JE, Chitwood Jr WR, Murray GF, Grover FL. Successful linking of the Society of Thoracic Surgeons database to social security data to examine survival after cardiac operations. Ann Thorac Surg. 2011;92(1):32–9. PMID: 21718828.
13. Dokholyan RS, Muhlbaier LH, Falletta J, Jacobs JP, Shahian D, Haan CK, Peterson ED. Regulatory and ethical considerations for linking clinical and administrative databases. Am Heart J. 2009;157(6):971–82. PMID: 19464406.
14. Blackstone EH. Demise of a vital resource. J Thorac Cardiovasc Surg. 2012;143:37–8.
15. Jacobs JP, Edwards FH, Shahian DM, Haan CK, Puskas JD, Morales DLS, Gammie JS, Sanchez JA, Brennan JM, O'Brien SM, Dokholyan RS, Hammill BG, Curtis LH, Peterson ED, Badhwar V, George KM, Mayer Jr JE, Chitwood WR, Murray GF, Grover FL. Successful linking of the Society of Thoracic Surgeons adult cardiac surgery database to Centers for Medicare and Medicaid Services Medicare data. Ann Thorac Surg. 2010; 90:1150–7.
16. Jacobs JP, Shahian DM, Edwards FH, O'Brien SM, Blackstone EH, Puskas JD, Schaffer J, Grover FL, Mayer Jr JE. Reply to letter to the editor: strategies to longitudinally track mortality. Ann Thorac Surg. 2012;94(2):693–4. PMID: 22818334.

Quality of Life: The Need for a National Database

32

Bradley S. Marino and Jeffrey B. Anderson

Abstract

Due to recent advances in pediatric cardiovascular therapy, mortality rates for children with heart disease (HD) have decreased dramatically. Despite these advances, however, survivors suffer from morbidity resulting from their circulatory abnormalities and the medical and surgical therapies they have received. These morbidities significantly impact the child's neurodevelopmental, psychosocial, and physical functioning and diminish their quality of life (QOL). As a result, outcome assessment focusing on QOL has become increasingly important in this high-risk population. QOL may be described as a child's ability to function in situational contexts and derive personal satisfaction from doing so. This paper will delineate health measurement definitions including QOL and health-related QOL (HRQOL), identify inherent difficulties in HRQOL measurement in the pediatric HD population, and discuss salient aspects of HRQOL instrument evaluation. In addition, this manuscript will describe existing generic and disease-specific HRQOL measures that may be used to assess HRQOL in the pediatric HD population, what research on HRQOL in the pediatric HD population has shown, and the extent to which HRQOL evaluations are being fully utilized in clinical practice. A research and clinical agenda is proposed to harness the potential applications of HRQOL assessment. Finally, the relationship of quality of life, safety, and value will be discussed as well as the formation of a quality of life national database.

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Introduction

Ultimately, the goals of pediatric research and clinical care are to maximize health and minimize symptomatology, disability, and dysfunction that may impact the lives of children with acute and chronic disease processes. Over the last several decades, new surgical techniques and advances in cardiopulmonary bypass, intensive care, interventional cardiac catheterization, non-invasive imaging, and medical therapies have significantly lowered neonatal mortality rates for children with the most complex congenital heart disease (CHD) (e.g. hypoplastic left heart disease) to less than 10 % [1]. In addition, cardiac-related mortality in patients with congenital and acquired heart disease has diminished significantly during the first two decades of life [2, 3]. Although survival rates vary by disease complexity, long-term survival (>20 years) rates for children with HD are estimated to be 95 % for simple CHD, 90 % for moderate CHD, and 80 % for complex CHD in the current era [3].

Despite these advances, however, survivors suffer from morbidity resulting from their circulatory abnormalities and the medical and surgical therapies they have received. These morbidities significantly impact the child's neurodevelopmental [4–6], psychosocial [7–9], and physical [10–12] functioning and diminish their QOL (Fig. 32.1). Given the high incidence of functional impairment in the pediatric cardiac population, there has been a paradigm shift in clinical research from short-term mortality prevention to long-term morbidity assessment. As a result, outcome assessment focusing on QOL has become increasingly important in this high-risk population.

QOL may be described as a child's ability to function in situational contexts (family, school, and peer) and derive personal satisfaction from doing so [13–15]. The multidimensional construct of QOL is thought to include three essential domains: physical health status and physical functioning; psychological status; and social functioning (Fig. 32.2) [13–15]. QOL measurement provides a comprehensive description of an individual's health, may result in the identification of physical, functional, and psychosocial dysfunction, and is a critical component of the evaluation of long-term outcomes of chronic conditions and disease-specific therapies.

This paper will delineate health measurement definitions including QOL and health-related QOL (HRQOL), identify inherent difficulties in HRQOL measurement in the pediatric HD population, and discuss salient aspects of HRQOL instrument evaluation. In addition, this

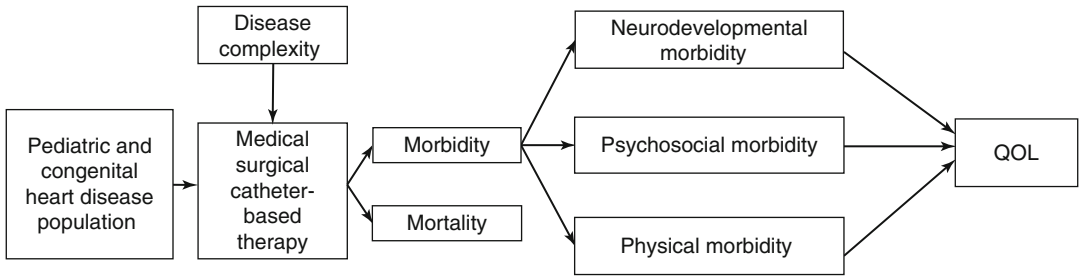

Fig. 32.1 The relationship between HD-related morbidity factors and QOL

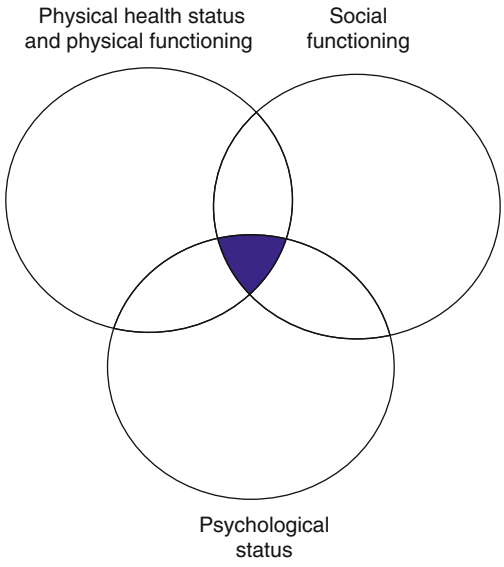

Fig. 32.2 Definition of QOL

manuscript will describe existing generic and disease-specific HRQOL measures that may be used to assess HRQOL in the pediatric HD population, what research on HRQOL in the pediatric HD population has shown, and the extent to which HRQOL evaluations are being fully utilized in clinical practice. Finally, a research and clinical agenda is proposed to harness the potential applications of HRQOL assessment.

Health Measurement Definitions: QOL and HRQOL

There are many ways to define "health". While the differences in meaning between various attributes of health may be subtle, the differences are important and have significant implications as to how clinical and research data is interpreted and findings incorporated into how we care for children with chronic disease. Health has been defined by the World Health Organization as "a state of complete physical, mental, and social well-being and not merely the absence of disease or infirmity" [16]. Indeed it was this initial definition of health that gave rise to the concept of QOL. Health status, which impacts QOL, may be thought of as a child's level of wellness versus illness, describing the impact of physiologic dysfunction, symptom burden, and/or level of illness control. Alternatively, functional status is defined as an individual's ability to perform activities of daily living, meet basic needs, fulfill roles, and maintain health and well-being within the context of various life situations [17]. Functional status is often affected by health status and has a significant impact on QOL. However, it is HRQOL, a more specific description of QOL, which is the most relevant construct relative to clinical and research data assessing outcomes and the provision of comprehensive clinical care of children and adolescents with chronic illness or injury [18]. HRQOL is defined as the influence of a specific illness, medical therapy, or health services policy on the ability of the patient to both function in and derive personal satisfaction from various physical, psychological, and social life contexts [19]. For the purposes of this manuscript all references to QOL hereafter are referring specifically to HRQOL.

Evaluating HRQOL is important because it allows for: better communication among patients, parents, and healthcare providers; prioritization of problems based partially on patient and/or parent preference; the monitoring of changes over time or in response to a specific therapy; and screening for other significant physical and psychosocial problems [20]. HRQOL measurement has emerged as a high priority not only for patients and their families, and medical caregivers, but also for the National Institutes of Health (NIH), Food and Drug Administration (FDA), and insurance providers [21]. The NIH's Patient-Reported Outcomes Measurement Information System, part of the NIH Roadmap, is a multimillion dollar effort devoted in part to improving HRQOL [22–27]. The FDA recognized the importance of such patient reported outcomes by issuing guidance to industry on the use of such outcomes in clinical trials in support of medical product claims [28]. A better understanding of the perceptions of HRQOL among patients with HD, and their parents and healthcare providers may improve treatment and patient outcome [29, 30], and the ability to perform important prospective cross-sectional, cohort, and randomized

clinical trials to improve patient outcome. Despite the known advantages, assessment of HRQOL in the pediatric cardiac population is lacking.

Measurement of HRQOL in patients with congenital and acquired HD has been limited over the last 25 years. Moons et al. noted that only 1 in 70 outcomes studies published between 1980 and 2003 that purported to assess HRQOL in pediatric cardiac patients actually measured the patients' perceived HRQOL [31]. In addition, more than half of the 70 articles did not meet any of the ten critical appraisal criteria for HRQOL research studies advocated by Gill and Feinstein in *JAMA* in 1994 [32]. The lack of rigorous research on HRQOL in the pediatric HD population is not surprising given the inherent difficulties of measuring HRQOL in this population.

Inherent Difficulties of HRQOL Evaluation in the Pediatric HD Population

HRQOL assessment in the pediatric population is challenging due to the wide age range and the changing developmental capabilities of the patients as they age. HRQOL assessment in the pediatric cardiac population is further complicated by the variety of congenital and acquired diseases, varying levels of severity, the array of therapeutic modalities that may be utilized to treat the patient (medical, surgical, and interventional), and the spectrum of outcomes. Similar to other pediatric chronic diseases, there may be variation in the perceived impact of HD on HRQOL, as many of the patients have always had HD (congenital HD) while others have been diagnosed with HD at an age when they were aware of the acute change in their health status (acquired HD).

Patient-family interactions are critical in QOL assessment, and the role of proxy-reporting (parent/guardian) and cross informant variance is often debated [33]. It has been extensively documented that HRQOL measurement in children with chronic health conditions and healthy children provided by proxy-respondents is not equivalent to that reported by the child [34, 35]. These findings indicate that proxy-reports cannot be substituted for child self-reports [36] and evaluations of pediatric cardiac patients' perspectives regarding treatment outcomes should be included in pediatric clinical care and clinical trials given the documented differences between child and proxy-reports. While pediatric patient self-report should be considered the standard for measuring patient perceived HRQOL [37], there may be situations when the child is too young, too cognitively impaired, or too ill to complete an HRQOL instrument, and proxy-reporting may be required. Further, it is typically parents' perceptions of their child's HRQOL that influences health care use [38, 39]. Ideally, parent and child QOL instruments should be chosen that measure the perspectives of both the child and proxy-reporter with the same constructs and parallel items to make comparisons between self and proxy-reports more informative and useful [40]. As noted by Moons et al. [31] most research studies assessing HRQOL in children with HD have only assessed proxy-reporters (parents/guardians) and ignored the HRQOL perceptions of the child. Studies of the cognitive development in children, psychometric studies on pediatric QOL measures that have included child self- reporting, and cognitive interviewing studies on children's abilities to respond to questionnaires indicate that self-reports from children over 7 years of age are reliable and valid [41]. Both patients and their parents provide important information, even though they may vary or even disagree significantly with one another. It may be that understanding the differences in perception of HRQOL between patients, their parent/guardians, and medical caregivers may be more important and informative than perceived agreement [30].

In addition to assessment issues related to the pediatric population in general and the pediatric HD population specifically, issues related to cultural and demographic variables (culture, race, ethnicity, income) affects HRQOL assessment in both pediatric and adult respondents. Culture, race, ethnicity, and income have critical influences on HRQOL measurement relative to the values that are attributed to various health states by children and their parents, the language in which health is described by children and their families, and the perceived functional impact of symptoms and changes in health states [42].

HRQOL Instrument Evaluation

When measuring HRQOL it is important to be clear on whether the goal of the application is to assess HRQOL or functional status or both. These measures of health are distinct constructs and are often confused with each other; note prior definitions. When assessing either HRQOL or functional status for research and clinical application, specific aspects of instrument selection (Tables 32.1 and 32.2), validation (Table 32.3), and availability (Table 32.4) must be considered.

When selecting an instrument it is important to note the instrument type, the specific construct that will be assessed, desired respondent type(s), patient and proxy-reporter age range(s), and the

Table 32.1 Instrument selection: constructs, respondent types, and domains

Instrument type	Instrument	Construct measured	Respondent type	Age range (years)	Domain(s)
Generic	CHQ [43]	Functional Status	Self-report	10–18	Physical functioning
			Proxy report[a]	5–18	Role/social emotional
					Role/social behavioral
					Role/social-physical
					Bodily pain
					General behavior
					Mental health
					Self-esteem
					General health perceptions
					Change in health
					Family activities
					Family cohesiveness
					Parental impact-time
					Parental impact-emotional[b]
	PedsQL 4.0 General Core Scales [33]	HRQOL	Self-report	5–7, 8–12, 13–18, 19–25	Physical
			Proxy report	2–4, 5–7, 8–12, 13–18	Emotional
					Social
					School[c]
Disease-Specific (CHD)	CHAT [47]	HRQOL	Self-report	11–18	Physical symptoms
					Physical limitations
					Limitations of physical education at school
					Social limitations
					External pressures
					Concerns (general, social, educational, physical, total)
	ConQol [48]	HRQOL	Self-report	8–11, 12–16	Symptoms
					Activities
					Relationships
					Coping and control[d]
	CHD-TAAQOL [45]	HRQOL	Self-report	17–32	Symptoms
					Impact cardiac surveillance
					Worries

(continued)

Table 32.1 (continued)

Instrument type	Instrument	Construct measured	Respondent type	Age range (years)	Domain(s)
Disease-Specific (HD)	PCQLI [49]	HRQOL	Self-report	8–12, 13–18	Disease impact (physical)
			Proxy report	8–12, 13–18	Psychosocial impact
	PedsQL 3.0	HRQOL	Self-report	5–7[e], 8–12, 13–18	Heart problems (symptoms)
	Cardiac Module [46]		Proxy report	2–4, 5–7, 8–12, 13–18	Treatment (barriers) Perceived physical appearance Treatment anxiety Cognitive problems Communication

Key: *CHAT*, Congenital Heart Adolescent and Teenage questionnaire, *CHD* Congenital heart disease, *CHQ* Child Health Questionnaire, *HD* heart disease, *PCQLI* Pediatric Cardiac Quality of Life Inventory, *PedsQL* Pediatric Quality of Life Inventory 4.0 Generic Core Scales

[a]Proxy reports are for parents, guardians or primary home care providers of patients

[b]The CHQ domain lists include both those for the patient and proxy reporter. Domain differences between patient and proxy report forms include: the Parental Impact-Time and Parental Impact-Emotional domains may only be found in the proxy form; the domains Role/Social-Emotional and Role/Social-Behavioral from the self report form are combined into one domain, Role/Social Emotional/Behavior in the proxy form; the listed domains except for Change in Health, Family Activities, and Family Cohesiveness create the Physical and Psychosocial summary scores for the Proxy Report Forms (CHQ-PF50 and CHQ-PF28) only

[c]The Physical domain makes up the Physical Health Summary Score, while the Emotional, Social, and School domains make up the Psychosocial Summary Score

[d]For adolescents 12–16 years old only

[e]Reliability of the PedsQL 3.0 Cardiac Module Young Child self-report Form for ages 5–7 years has not been established (ranged from 0.35 to 0.83)

domains to be assessed (Table 32.1). When selecting a specific form from within an instrument grouping, the number of items in the tool and the average completion time should be considered as it may impact the feasibility of completing the research project or clinical application (Table 32.2).

Both generic and disease-specific instruments exist that may be used to measure HRQOL or functional status in the pediatric HD population [43–49]. Whether generic or disease-specific, the "ideal" QOL measure will have a patient self-reporting mechanism with parent/guardian proxy-reporting, wide age range, will be easily self-administered in a reasonable timeframe, and have an array of relevant constructs to describe and measure HRQOL or functional status. Generic HRQOL or functional status measures assess these constructs in both healthy children and in children with chronic disease. They may be used to compare various chronic disease groups or chronic disease groups and healthy controls. Disease specific instruments assess HRQOL in a particular condition or disease, and may be more comprehensive for a specific disease, and a better discriminator of differences between sub-groups within a disease category. Disease-specific pediatric cardiac HRQOL instruments may provide new, critical information on the outcome of present and future interventional catheterization and cardiac surgical procedures in the short and long term and may be utilized for randomized clinical trials for cardiovascular drugs and new technologies and interventions. In addition, a disease-specific pediatric cardiac HRQOL instrument may define changes in HRQOL over time (evaluative tool), predict future changes in HRQOL (prognostic tool), and signal new problems or issues that might not be noted by traditional biologic markers (diagnostic tool).

Table 32.2 Instrument selection: forms

Instrument type	Instrument	Name	Respondent type/age range (years)	Items (#)	Completion time (min)
Generic	CHQ	CHQ-CF87	Child/Adolescent (10–18)	87	25
		CHQ-PF50	Parent Proxy (5–18)	50	15
		CHQ-PF28	Parent Proxy (5–18)	28	10
	PedsQL 4.0 General Core Scales	Young Child Report	Young Child (5–7)	23	10
		Child Report	Child (8–12)		
		Teen Report	Teen (13–18)		
		Young Adult Report	Young Adult (19–25)		
		Parent of Toddler Report	Parent Proxy (2–4)	23	10
		Parent of Young Child Report	Parent Proxy (5–7)		
		Parent of Child Report	Parent Proxy (8–12)		
		Parent of Teen Report	Parent Proxy (13–18)		
Disease-Specific (CHD)	CHAT	CHAT Questionnaire	Adolescent (11–18)	53	20–30
	ConQol	ConQol 8–11	Child (8–11)	29	10
		ConQol 12–16	Adolescent (12–16)	35	10
	CHD-TAAQOL	CHD-TAAQOL Questionnaire	Young Adult (17–32)	26	10
Disease-Specific (HD)	PCQLI	Child Form	Child (8–12)	24	10
		Adolescent Form	Adolescent (13–18)	30	10
		Parent of Child Form	Parent Proxy (8–12)	24	10
		Parent of Adolescent Form	Parent Proxy (13–18)	30	10
	PedsQL 3.0 Cardiac Module	Young Child Report	Young Child (5–7)	27	10
		Child Report	Child (8–12)		
		Adolescent Report	Adolescent (13–18)		
		Parent of Toddler Report	Parent Proxy (2–4)	27	10
		Parent of Young Child Report	Parent Proxy (5–7)		
		Parent of Child Report	Parent Proxy (8–12)		
		Parent of Teen Report	Parent Proxy (13–18)		

Key: *CHAT* Congenital Heart Adolescent and Teenage questionnaire, *CHQ* Child Health Questionnaire, *PCQLI* Pediatric Cardiac Quality of Life Inventory, *PedsQL* Pediatric Quality of Life Inventory 4.0, *UK* United Kingdom, *USA* United States of America

Assessing prior validation data on a given HRQOL or functional status instrument is central to the instrument evaluation process (Table 32.3). When assessing validation of a given instrument, four specific questions relative to the psychometric properties of the instrument should be asked: (1) Has the instrument been shown to be reliable in the patient population being studied?; (2) Has the instrument been shown to be internally valid in the patient population being studied?; (3) Has the instrument been shown to be externally valid in the patient population to be studied?; (4) If the study is assessing change over time or the impact of an intervention, has the instrument been shown to be responsive in the patient population to be studied?

All psychometric instruments must be shown to be reliable before validity and responsiveness may be considered [50]. An "unreliable instrument" cannot be deemed valid or responsive.

Table 32.3 Instrument validation: reliability, validity, and responsiveness

				External validity		
Instrument type	Instrument	Reliability	Internal validity	Construct validity	Generalizability	Responsiveness
Generic	CHQ	USA[a]	USA[a]	USA[a]	USA[a]	USA[a]
	PedsQL 4.0 General Core Scales	USA[b]	USA[b]	USA[b]	USA[b]	USA[b]
Disease-Specific (CHD)	CHAT	Canada	Canada	Canada	–	–
	ConQol	UK, Canada	UK, Canada	UK, Canada	–	–
	CHD-TAAQOL	Netherlands	Netherlands	Netherlands	–	
Disease-Specific (HD)	PCQLI	USA [49, 57], UK [81]	USA [49, 57], UK [81]	USA [49, 57], UK [81]	USA [57, 76]	–
	PedsQL 3.0 Cardiac Module	USA	USA	USA	–	–

Key: *CHAT* Congenital Heart Adolescent and Teenage questionnaire, *CHQ* Child Health Questionnaire, *PCQLI* Pediatric Cardiac Quality of Life Inventory, *PedsQL* Pediatric Quality of Life Inventory 4.0, *UK* United Kingdom, *USA* United States of America

[a]The CHQ has been translated and validated in multiple languages and countries. Only the original is listed here for clarity. Please see www.healthactchq.com for an exhaustive list

[b]The PedsQL has been translated and validated in multiple languages and countries. Only the original is listed here for clarity. Please see www.pedsql.org for an exhaustive list

Table 32.4 Instrument availability: authorization, cost, and languages

Instrument type	Instrument	Authorization	Cost	Language(s)
Generic	CHQ	Licensure	Licensing fee	English (USA)[a]
	PedsQL 4.0 General Core Scales	User Agreement	None/Licensing Fee	English (USA)[b]
Disease-Specific (CHD)	CHAT	None Required	None Required	English
	ConQol	None Required	None	English (UK)
	CHD-TAAQOL	NA	NA	English, Dutch
Disease-Specific (HD)	PCQLI	User Agreement	None	English (USA, UK)
	PedsQL 3.0 Cardiac Module	User Agreement	None/Licensing Fee	English (USA)

Key: *CHAT* Congenital Heart Adolescent and Teenage questionnaire, *CHQ* Child Health Questionnaire, *NA* information not available, *PCQLI* Pediatric Cardiac Quality of Life Inventory, *PedsQL* Pediatric Quality of Life Inventory 4.0, *UK* United Kingdom, *USA* United States of America

[a]The CHQ has been translated in multiple languages. Only the original is listed here for clarity. Please see www.healthact.com for an exhaustive list

[b]The PedsQL has been translated and validated in multiple languages and countries. Only the original is listed here for clarity. Please see www.pedsql.org for an exhaustive list

Demonstrating reliability involves assessing score "reproducibility" through internal consistency measurement (Cronbach α) and comparing scores on the same patient at two distinct points in time with an appropriate interval between them to minimize recall bias (test-retest reliability). Validity testing of a psychometric scale is an ongoing, evidence-based process that assesses the degree of confidence one should have in inferences made about a test-taker based on their score. Assessing validity is often divided into the domains of "internal" and "external" construct validity [51–53]. "Internal validity" [50–52, 54] may be thought of as an assessment of content validity and structural validity that includes: assessment of the theoretical conceptualization of the respective instrument; the clarity, relevance, and representativeness of the item content; and tool construction. In contrast, establishing "external validity" requires demonstrating convergent and discriminant construct validity and "generalizability" [51, 52, 55]. "Generalizability" may be defined as the ability of a tool to provide valid and reliable information when utilized in different geographic regions and patient populations [52]. Generalizable tools allow researchers to have confidence that data collected from multiple sites and regions are comparable. Substantiation of the "generalizability" part of external validity enables a HRQOL instrument to be used for clinical applications and multi-center research that may serve as a platform for future multi-site cross-sectional and prospective studies using HRQOL as an outcome. Responsiveness describes the ability of the HRQOL or functional status instrument to be sensitive to change in score after intervention or if there is meaningful change in score over time [50]. Having a responsive instrument allows the investigator to note differences in HRQOL based on a specific intervention or treatment strategy, which is critical to assessing all potential interventions in a given disease population. In addition a responsive tool will allow the clinician or researcher to see changes in HRQOL over time as a patient's health status or functional status worsens. Having a responsive instrument is critical to any interventional agenda or follow-up program for any given chronic disease population.

Existing Generic and Disease-Specific HRQOL Measures That May Be Used to Assess HRQOL in the Pediatric HD Population

Both the PedsQL 4.0 Cores scales, a generic HRQOL measure, and the Child Health Questionnaire (CHQ), a generic functional status

measure, have patient and proxy-reporting, wide age range, and may be administered in a reasonable timeframe. In addition, these commercially available tools have been shown to be reliable and internally and externally valid, and responsive in the United States and many other countries after language translation (Tables 32.1, 32.2, 32.3, and 32.4) [43, 44]. Five disease-specific pediatric cardiac HRQOL instruments have been previously described [45–49]. The Congenital Heart Disease-TNO/AZL Adult QOL (CHD-TAAQOL) questionnaire is a disease specific instrument that assesses HRQOL in young adults with CHD [45]. The PedsQL 3.0 Cardiac Module [46], Congenital Heart Adolescent and Teenager Questionnaire (CHAT) [47], ConQol [48], and Pediatric Cardiac Quality of Life Inventory (PCQLI) [49] measure HRQOL in the pediatric cardiac population. Instrument availability (form specific authorization requirements, user costs, and available language translations) are shown in Table 32.4. The PedsQL 3.0 Cardiac Module [46], created and validated by Uzark et al. at a single site, was a critical and important step forward in the objective assessment of HRQOL in children with HD. The PedsQL 3.0 Cardiac Module has been shown to be reliable, internally and externally valid. The 27-item PedsQL Cardiac Module encompasses 6 Scales: Heart Problems and Treatment; Treatment II, Perceived Physical Appearance, Treatment Anxiety, Cognitive Problems, and Communication [46, 56]. The PedsQL 3.0 Cardiac Module has not yet been shown to be generalizable to other geographic regions or demographic populations in the United States. The CHAT and ConQol questionnaires have important limitations that include: intended for use in HD patients with CHD only [47, 48]; narrow age range [47, 48]; lack of parent proxy reporting [48]; no generalizability data to support broad applicability to other geographic regions or demographics within the United States [47, 48], and/or an inadequate ability to discriminate among various types of cardiovascular disease across a wide age range [48]. The PCQLI is the most recently published disease specific instrument. Similar to the PedsQL 3.0 Cardiac Module, it has a patient self-reporting mechanism with parent/guardian proxy reporting, wide age range, is easily self-administered in a reasonable time-frame, and has an array of relevant constructs to describe and measure HRQOL in the pediatric HD population. The PCQLI is the only disease-specific measure that has been tested in a multi-center trial and shown to be reliable, valid, and generalizable in the United States [57, 58]. The ConQol and PedsQL 3.0 Cardiac Module can distinguish among disease severity subgroups but only within select subsets of a study population (age, respondent type) [46, 48]. In contrast, PCQLI Total and subscale scores (Disease Impact and Psychosocial Impact) differentiate between congenital HD severity subgroups irrespective of age category, score examined or respondent type [40]. From a research perspective, this is an important development that will facilitate cross-sectional and prospective studies of HRQOL in clinically important subgroups in the pediatric HD population. It is important to note that none of the five disease specific instruments have been shown to be responsive in the United States.

In summary, whether selecting a measurement tool for research or clinical application it is critical to define the specific hypothesis or what clinical information is desired and then match the hypothesis/clinical data requirement to potential instruments based on the constructs assessed in the specific instrument, the proposed respondents, and the feasibility of utilization. It is important to use age-appropriate measures that reflect the maturity and the cognitive development of the desired respondents. It is vital that the instrument being considered for measurement be reliable, valid, and responsive in the patient population being considered. The instrument needs to have been shown to be responsive if assessing for change in score over time or after intervention in the population being studied. Collect data from both patient and parent/guardian-proxy respondents to identify the full HRQOL impact on the patient, parent, and family. Use both a disease-specific and generic measure, to allow for discrimination between sub-groups and comparison with other chronic disease groups and/or healthy children.

Research on HRQOL in Pediatric Patients with HD: What Is Known

HRQOL Studies in the General CHD Population

Most early outcome studies in patients with CHD described mortality and morbidity or health status, including anatomic or hemodynamic outcome, electrophysiologic sequelae, and/or exercise capacity following surgical interventions or included "quality of life" parameters such as marital status, number of offspring, employment status, or educational attainment in adults with CHD [59]. More recent studies have recognized the multidimensional nature of HRQOL and have included not only physical health status and physical functioning, but also psychological status and social functioning. Unfortunately, there are only a few studies that have evaluated the patient's self-perceptions of HRQOL.

Health Status and Functional Status in the HD Population

In children with CHD as young as 1–3 years, Limperopoulos et al. reported a high incidence of functional limitations including difficulties in socialization skills [11]. Walker et al. evaluated functional status in children attending a cardiology clinic, utilizing the Child Health Questionnaire (CHQ PF-50) to describe the physical and psychosocial health status by parent-proxy report [60]. The sample of children seen in the outpatient cardiology clinic were reported to have worse scores for physical function, general health perception, assessment of family activities, and parental emotional impact, as well as more anxiety problems and learning problems. Majnemer et al. [61] used the CHQ PF-50 as well as the Child Behavior Checklist and the Parenting Stress Index in describing well-being in children 5 years of age following open-heart surgery in infancy. Mean scores on the CHQ were in the normal range, however parents more often reported problems related to anxiety, attention, developmental delays, and learning. The child's psychosocial health status was significantly correlated with parental stress. Both of the later studies acknowledge parental responses are likely influenced by their hopes and expectations for their child and how well they are coping as a family. This is consistent with one of the earliest studies of the emotional adjustment of children with CHD by DeMaso and colleagues [62] who reported that approximately 33 % of the variability in the child's adjustment was accounted for by maternal perceptions, while the medical severity accounted for less than 3 % of the variability.

HRQOL in the HD Population

Self-reported HRQOL related to physical health, psychosocial health, social functioning, and school functioning for children with CHD is reduced compared to healthy children [56, 57, 63, 64]. Mussatto and colleagues [64] found that the greatest negative impact on HRQOL was reported in the areas of social and educational functioning, despite the perception that CHD primarily has physical effects. In a large single-center study of HRQOL in children with HD, Uzark et al. evaluated both parent-proxy and self-reported perceptions utilizing the PedsQL 4.0 Core scales [56] As perceived by parents, worse physical and psychosocial HRQOL is related to the severity of HD. While most children with HD reported good overall HRQOL, 20 % of the children with HD reported significantly impaired psychosocial HRQOL, including children with mild or repaired HD. A recent systematic review of studies assessing psychological adjustment and HRQOL in children and adolescents following open-heart surgery for CHD [63] concluded that studies on self-reported psychological adjustment indicate a good outcome, however, a considerable proportion of children experienced psychological maladjustment according to their parents which was related to severity of CHD and developmental delay.

The largest multi-center study assessing HRQOL in the United States utilizing a reliable, valid, and generalizable disease-specific HRQOL measure including both child and adolescent self-report and parent/guardian proxy-report was

Table 32.5 Variation in HRQOL within CHD subgroups

PCQLI median total score (range)				
	Child	Parent of child	Adolescent	Parent of adolescent
AS (n = 75)	86.2 (51.9, 100)	86.8 (54.1, 100)	89.5 (65.1, 97.8)	85.2 (40.8, 99.3)
TOF (n = 125)	75.6 (48.7, 100)	78.6 (43.5, 100)	79.6 (39.6, 99.3)	78.7 (33.5, 100)
Fontan (n = 219)	64.4 (32.2, 99.1)	66.1 (30.7, 100)	70.5 (39.6, 100)	69.7 (26.0, 98.5)

performed by Marino et al. in the PCQLI Validation Study [40]. In this study 1,605 patient parent pairs (3,210 total respondents) participated from seven geographically diverse centers in the United States. In this study HD patients with both CHD (68 % of the cohort) and acquired HD (32 % of the cohort) were included. This study showed that lower patient and parent-reported HRQOL scores were associated with higher disease severity and increased medical care utilization, poorer patient self-perception and competency, and increased behavioral and emotional problems in the pediatric HD population. PCQLI scores (Total, Disease Impact, and Psychosocial Impact) differed significantly among disease severity subgroups (mild CHD, biventricular CHD s/p surgical repair or palliation, and single ventricle CHD s/p Fontan completion). Mild CHD was defined as CHD that had not required surgical or catheter-based intervention. Furthermore, patients in the repaired biventricular subgroup had significantly lower PCQLI Total and subscale scores than patients in the mild subgroup, and patients in the palliated single-ventricle subgroup had significantly lower PCQLI Total and subscales scores than patients in both the repaired biventricular and mild CHD subgroups. These results were reproducible across all age categories and respondent types and are consistent with widespread clinical observations that increased disease severity is associated with a lower HRQOL. Increased number of cardiac surgeries, cardiac-related hospital admissions, and doctor visits in the last 12 months were associated with lower PCQLI Total score. These results were consistent across all four forms (Child Form, Parent of Child Form, Adolescent Form, and Parent of Adolescent Form). Worse PCQLI Total score was significantly correlated with lower Global Self-Worth score on the Self Perception Profile for Children and Adolescents for both age groups. A statistically significant positive correlation was noted between the PCQLI Total score and Achenbach (Youth Self Report and Child Behavior Checklist) Total Competency score, and statistically significant inverse correlations existed between PCQLI Total score and both the Achenbach Internalizing Problems summary scale score and DSM-IV Oriented Scale scores (Affective Disorder, Anxiety Disorder, Somatic Disorder, Attention Deficit Hyperactivity Disorder) for all groups [40].

Interestingly, there was significant variation noted in the specific diagnosis and procedural groups (Table 32.5) for acyanotic two-ventricle (e.g. aortic stenosis), cyanotic two-ventricle (e.g. tetralogy of Fallot) and the palliated single ventricle Fontan populations [40]. While each specific population group segregated into a particular HRQOL score range (aortic stenosis – 80s; tetralogy of Fallot – 70s; and Fontan – 60s) based on the underlying disease severity and the medical, catheter-based, and surgical therapy required, there were complex single ventricle Fontan patients that had HRQOL scores as high as aortic stenosis patients who had not undergone intervention and aortic stenosis patients who had undergone intervention who had HRQOL scores that were worse than the typical Fontan. This data suggests that there are resilience and depressant factors that increase or decrease each individual patient's arc of HRQOL over time (Fig. 32.3). Understanding broad resilience and depressant factors across the entire HD population and/or resilience and depressant factors important for specific diagnosis or procedural groups will create opportunities to prevent the development of lower HRQOL or treat HD patients with lower HRQOL to improve it.

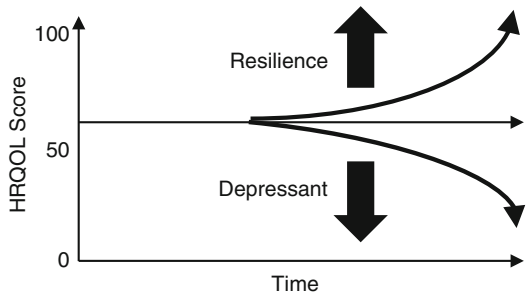

Fig. 32.3 Patient Independent ARC of HRQOL: resilience vs. depressant factors

HRQOL Studies in Specific HD Subgroups

HRQOL and functional status has also been studied in some specific patient subgroups with HD. In children with transposition of the great arteries (TGA), Culbert and colleagues from the Congenital Heart Surgeons Society [65] assessed functional status using the CHQ-CF87 completed by 306 children 11–15 years after TGA repair. Health status was perceived as excellent when compared with published normative data and was better after arterial switch operation (ASO) than after atrial switch operation. Dunbar-Masterson and colleagues [66] who used the parent version of the CHQ also found that at 8 years of age, children after the ASO had an overall physical and psychosocial health status similar to that of the general population. It was also noted that lower IQ and academic achievement was associated with worse psychosocial health status. Hovels-Gurich and associates [67] reported that children with TGA who had undergone neonatal ASO had parent-reported behavioral impairment at age 8–14 years with normal self-reported HRQOL.

Brosig and colleagues [68] compared psychosocial outcomes between preschool-aged survivors who underwent ASO for TGA and Fontan palliation for hypoplastic left heart syndrome (HLHS). By parent report, HRQOL scores in both CHD subgroups did not differ from healthy controls. Parents of children with HLHS reported more negative impact of the child's illness on the family and more parenting stress than parents of children with TGA, and children with HLHS had higher rates of inattention and externalizing behavior problems than children with TGA. In a large study of Fontan survivors [69] 6–18 years of age, parents reported CHQ functional status summary scores were significantly lower than the U.S. population for Physical Functioning and Psychosocial Functioning. Parent-reported patient conditions, including behavior, learning, anxiety, attention problems and depression explained the greatest amount of variation in the Psychosocial Functioning scores.

In a study by DeMaso et al, HRQOL has been assessed in children and adolescents with implantable cardioverter-defibrillators [70]. While the parent-reported psychosocial summary scores for children with defibrillators were not significantly different from the normative U.S. sample, the domains of social emotional behavioral roles, self-esteem, and the emotional impact of their child's health on themselves were all significantly lower than the normative sample. A recent multi-center study by Czosek and colleagues compared HRQOL scores between pediatric device patients and healthy controls, and determined the key drivers of HRQOL in pediatric device patients [71]. The study included 173 patient-parent pairs [40 implantable cardiac defibrillators (ICD)/133 pacemaker; 50 % CHD; 50 % male; median age 13 (8–18) years]. Compared to healthy controls, both patients and parents reported significantly lower PedsQL Total scores. ICD patients had significantly lower PCQLI Total scores than pacemaker patients. CHD patients had significantly lower PCQLI Total score than non-CHD patients. The key drivers of patient HRQOL were presence of ICD, CHD, and worse self-perception. For parent proxy-reporters, patient HRQOL was driven by internalizing behavioral problems (anxiety, depression, and somatization). Interestingly, activity restrictions and device complications did not impact HRQOL. Whether these factors can be mitigated through the use of psychological interventions needs to be assessed.

Finally, in the pediatric heart transplant population, using the Children's Global Assessment Scale, 27 % of children 6.1–12.9 years after transplant had emotional adjustment difficulties

[72] and there was a significant correlation between emotional adjustment and family functioning. Wray and colleagues also found that a significant number of pediatric heart transplant recipients (>33 %) had increased behavior problems and diminished social competence, especially in children with a pre-transplant diagnosis of CHD in comparison to children with cardiomyopathy pre-transplant [73].

Predictors of HRQOL in the Pediatric HD Population

Neurodevelopmental Predictors of HRQOL in the Pediatric HD Population

Few studies have investigated the impact of neurodevelopmental outcome on HRQOL in the pediatric CHD population. For children with d-TGA, Dunbar-Masterson et al. found that lower full-scale IQ (intelligence) and lower performance in reading and math (academic achievement) were associated with lower parent-reported psychosocial HRQOL scores at 8 years of age [66]. Williams et al. found that children with Fontan palliation for hypoplastic left heart syndrome displayed significant delays in communication and motor skills and lower parent-reported psychosocial HRQOL scores [74]. Of note, both of these studies used a generic QOL instrument to measure psychosocial QOL, which may not be as sensitive or accurate as a disease-specific instrument [75]. In addition, neither study measured patient-perceived HRQOL nor specifically assessed the association between neuropsychological impairments and patient-perceived HRQOL. Parent-reported and self-reported HRQOL are both important as perception of HRQOL differs between patients and parents [30, 56].

The cardiac-specific module of the PedsQL includes a cognitive problems subscale and a communication subscale [44, 46]. Using the PedsQL cardiac-specific module, Uzark and colleagues found that children with severe cardiovascular disease have lower parent-reported and self-reported HRQOL scores on the cognitive problems subscale and lower parent-reported HRQOL scores on the communications subscale than children with less severe cardiovascular disease [56]. Recently, Marino et al. demonstrated that worse executive functioning, gross motor ability, and mood (presence of anxiety and depression) significantly predicted lower PCQLI score after controlling for patient demographics and important clinical covariates [76]. Executive functioning, gross motor ability, and mood accounted for up to 50 % of the variance in patient and parent-reported HRQOL scores. These factors appear to be key drivers of HRQOL in complex CHD survivors and may be targets for future intervention [75].

Psychosocial Predictors of HRQOL in the Pediatric HD Population

While multiple studies have shown that there is psychosocial dysfunction in the pediatric heart disease population, few studies have assessed for association between psychosocial predictors in the individual or family environment and HRQOL [56, 57, 69, 77–79]. A recent multi-center multinational study in the United States and England completed by Marino and colleagues explored the relationships between HD complexity, HRQOL, and psychosocial morbidity factors. 815 patient-parent pairs participated in the study with a mean patient age of 12.5 ± 1.5 years. The study assessed the mediating impact of specific psychosocial morbidity factors (family functioning and parental stress, and patient and parent post-traumatic stress and trait anxiety) on the association between HD complexity and lower HRQOL. High complexity was associated with a lower PCQLI Psychosocial Impact subscale score. Higher parental stress, post-traumatic stress, and trait anxiety scores were associated with a lower PCQLI Psychosocial Impact subscale score. The association between High complexity and lower PCQLI PI score was mediated by worse parental stress, post-traumatic stress, and trait anxiety [Total Correlation (direct and indirect effects) $= -0.21$ to -0.46; $p < 0.001$] for both patients and parents. Interventions on these psychosocial morbidities may improve QOL [80].

Summary

Many studies of HRQOL in children with CHD have been limited by small sample size, have

often relied on parental proxy-report of the child's HRQOL, reported health status or observed functional status, or focused on a single dimension of HRQOL. Recent instrument development has allowed multidimensional, self-report of HRQOL, essential to development of interventions to improve HRQOL. Further research is needed to discover links between specific aspects of neurodevelopmental and psychosocial morbidity factors and HRQOL to identify developmental delays and psychosocial issues that may be improved through intervention. By characterizing the relationship between disease complexity, neurodevelopmental and psychosocial morbidity, and HRQOL, physicians and caregivers will be able to change the medical care delivery system to significantly improve the lives of children with CHD and ensure their future success.

Clinical Implementation (Is HRQOL Evaluation Utilized in Clinical Practice?)

The importance and utility of HRQOL assessment with reliable, valid, generalizable, and responsive instruments in the pediatric HD population is centered on the fact that they provide a means to improve patient HRQOL outcomes through: (1) The improvement of comprehensive follow-up of the pediatric HD population (surveillance and screening); (2) The identification of at-risk patients (risk stratification of HD subpopulations); (3) The identification of modifiable risk factors to prevent adverse outcomes (prevention); and (4) The design of interventions for children with poor outcomes (treatment planning). Early identification of neurodevelopmental impairments in academic achievement, language, visual construction and perception, attention, processing speed, memory, executive functioning, fine and gross motor skills, and/or ADHD in these children may allow the clinical care team to stratify populations and improve HRQOL outcomes through targeted intervention for children at-risk. Given some of the early data on the utility of physical rehabilitation in patients with CHD and diminished exercise capacity, physical functioning and HRQOL may be improved through rehabilitation or medical or device based (pacemaker) therapies. In addition, early identification of psychosocial functioning issues in the child and/or family (post-traumatic stress, trait anxiety, depression, coping, family functioning, and parental stress) may allow for risk-stratification and the incorporation of targeted interventions to prevent or treat psychological or social morbidity. Unfortunately, the clinical evaluation of neurodevelopmental impairments, physical and psychosocial morbidity on HRQOL in children with HD has not become a standard component of care and much is left to be learned. The clinical utility of current instruments is largely unknown.

Multiple barriers exist to incorporating HRQOL assessment into the clinical environment [19]. Pediatricians and cardiologists (medical home providers) have ever-increasing demands on their time and the utilization of HRQOL instruments for surveillance and screening will only occur if administration, scoring, and interpretation of measures are simple and are easily integrated into the clinical practice setting in "real-time" as part of the "paper" or electronic medical record. In addition, practitioners will only incorporate HRQOL assessment into their clinical care if there is compelling evidence that surveillance, screening, referral, evaluation, and intervention are efficient, cost-effective, and make a difference in patient specific HRQOL outcomes. One of the biggest barriers to incorporation is the lack of knowledge among practitioners of the theoretical benefits to HRQOL assessment. Only through clinical research studies focusing on harnessing the potential of HRQOL assessment will these barriers begin to fall.

Research Agenda (What Is Needed in HRQOL Research in Pediatric HD Patients?)

1. Future HRQOL research should focus on the associations of specific morbidities/phenotypes in the pediatric HD population and HRQOL to determine candidate factors for interventional studies for prevention and

treatment. Specifically, further research is needed to discover links between neurodevelopmental, psychosocial, and physical morbidity factors and HRQOL to identify specific functional deficits that may be prevented or mitigated through intervention.
2. Both patient and parent respondents need to be evaluated in HRQOL research to learn more about similarities and differences between patient and parent respondents. These patterns will inform clinical applications on how best to assess HRQOL in patients and their parent/guardians and inform how future screening and interventions programs may improve HRQOL.
3. Demonstrate that HRQOL assessment in the clinical setting will ultimately result in clinically meaningful changes in HRQOL and/or functional status. Specifically, it must be demonstrated that: (1) It is feasible to collect HRQOL data in "real-time" in the clinical setting and that patients may be stratified into low and high risk groups for neurodevelopmental, physical and psychosocial dysfunction; (2) It is feasible to refer patients with lower HRQOL scores stratified into a high risk category into interventions; and (3) Interventions in a high risk group will result in clinically meaningful changes in HRQOL.
4. A responsive instrument is required and fundamental for the field to pursue an interventional agenda that will improve current clinical practice. Efforts should be made to demonstrate responsiveness in disease-specific measures shown to be reliable, valid, and generalizable in the United States. Once responsiveness has been demonstrated, this HRQOL tool should be considered for inclusion in all randomized clinical or interventional drug, device, or surgical treatment trials, where appropriate.
5. New HRQOL instruments should be developed for aging cohorts of pediatric HD patients (young adults and adults) in the United States as there is a rapidly growing population of Adult with CHD (ACHD population) in transition with changing HRQOL assessment needs. HRQOL instruments for adults with HD are intended for patients with HD due to hypertension or coronary ischemia. A combination of an adult generic HRQOL measure and an ACHD disease-specific HRQOL measure will provide the necessary tools to provide critical information on this unique and growing population.

Clinical Agenda (What Is Needed to Harness the Potential of HRQOL Assessment for Clinical Use in Pediatric HD Patients?)

1. Begin incorporating HRQOL evaluation into the clinic visit to take full advantage of the current advances in HRQOL measurement.
2. Strive to make all HRQOL evaluations conducted in clinical settings "research quality" (or at least create standard evaluation protocols) so that the field may benefit from reliable, valid, and potentially generalizable clinical information. There is so much variability in the way evaluations are presently conducted that it makes it difficult to generalize any data collected at any particular site or clinical setting across settings and/or populations.
3. Once it has been demonstrated that HRQOL assessment may be used for neurodevelopmental, psychosocial, and physical morbidity risk stratification, HRQOL assessment should be performed in all outpatient clinic visits as part of a formal standardized surveillance and screening program to allow for referral, intervention, and follow-up.
4. Incorporate HCQOL assessment as a routine and longer term measure of quality care provided by providers and systems.

The Relationship of Quality of Life, Safety, and Value

While clinical outcomes in CHD, as with many other pediatric conditions, have improved over the last two decades, these improvements have come at an economic cost. Diagnostic and interventional technologic advances have

partially driven clinical improvements. Improvement in mortality has led to clinical focus on improving other outcomes, including improvement in HRQOL. In addition, as healthcare fiscal resources have become progressively more stretched, cost and value have increasingly worked their way into the discussion of best outcomes. In fact, economic metrics are important tools to assist decision makers in defining the value of the care we offer outpatients and families. Interventions and programs that represent good value, or best outcomes/cost, should be sought out, tested and spread. These metrics can, and will, be used to decide whether to fund or reimburse particular interventions or diagnostic tests. This movement has resulted in an increasing number of published studies that include economic appraisals of interventions and services that are aimed at children and adolescents [81, 82].

Several studies have attempted to identify metrics that define value in health care. These studies have aimed to compare specific interventions in terms of their costs and benefits. Some of the analyses that have been studied include: (1) Cost-benefit analysis: a technique in which the costs are compared with benefits of an intervention, both valued in monetary terms; (2) Cost-effectiveness analysis: a technique in which the costs of an intervention are compared with a single predefined health outcome (e.g., cost per case detected, cost per life-year gained); and (3) Cost-utility analysis: a special type of cost-effectiveness analysis that uses quality-adjusted life-years (QALYs) as an outcome measure [83]. Among all study types, cost-utility analysis, whereby health benefits are quantified in terms of QALYs, has become the standard type and is now recommended in the great majority of health economics guidelines as the analysis of choice [84, 85].

The main reason for using QALYs as an outcome measure is that improvements in HRQOL and life expectancy are captured within a single index that also incorporates individual preferences for various health outcomes. Such form of analysis therefore allows the direct comparison of the relative health benefits of interventions across different disease areas and populations. The use of QALYs, however, relies on a number of assumptions, including that the health of the patient is the only important outcome and that it is possible to trade directly between quality and quantity of life [86].

While improvement in clinical outcomes have come with increased diagnostic and interventional costs, it is important not to discount the potential gain in QALYs as we improve not only clinical outcomes but improve HRQOL in infants, children and adults with CHD. As we care for our patients we focus on doing so in the safest way possible to minimize harm, offering the best in medical care to improve clinical outcomes, and in the most efficient way possible to maximize patient value. These same areas should be focused on when addressing issues of improvement in HRQOL. The best value will be offered to our patients and families if we provide them with the best clinical and HRQOL outcomes in the most cost-effective manner and in a safe environment.

Quality of Life National Registry

Based on the unique interaction between Safety, Quality, and Value it is a national imperative to set up a national registry that will collect data on HRQOL pediatric for and congenital heart disease. By doing so we will be able to accumulate critical HRQOL data on important pediatric heart disease cohorts so we can perform cost-utility analyses to determine discrete QALY values for specific populations. These cost-utility analyses will allow clinicians and researchers to compare the impact of specific medical, surgical, and catheter-based therapies within specific high-risk pediatric and congenital heart disease populations. More importantly, clinicians and researchers will be able to compare the cost per QALY for various potential therapeutic choices for the same lesion or clinical situation. From a safest perspective the cost to provide a given QALY outcome is not linear and knowing the cost per outcome relative to various safety profiles will be fundamental to reshape our clinical environment. By pursuing this agenda we will have the opportunity to care

for our patients in the safest possible manner to minimize harm, offer the best medical care to maximize QALYs, and do so in the most efficient manner to maximize patient value (cost per outcome).

Conclusion

Over the last several decades mortality rates for children with HD have fallen. However, survivors may have neurodevelopmental, psychosocial, and physical morbidities that lower HRQOL. Although HRQOL assessment in this high-risk population has been lacking due to inherent issues in HRQOL assessment in the pediatric HD population, advances have been made in HRQOL measurement with new reliable, valid, and generalizable measures. These questionnaires may be utilized to rapidly improve HRQOL research and obtain critical information that may be translated into the clinical domain. Rigorous characterization of the relationship between neurodevelopmental, psychosocial, and physical morbidity factors and HRQOL will identify specific factors amenable to intervention and allow clinicians to modify the medical care delivery system to significantly improve the lives of children with CHD and promote their future success. Formal screening and intervention programs based on HRQOL assessment will allow clinicians to intervene in those children with significant deficits with the greatest potential to improve HRQOL. Cost-Utility analyses that will utilize HRQOL measurement to generate QALYs as an outcome measure are critical so improvements in HRQOL and life expectancy are captured within a single index that also incorporates individual preferences for various health outcomes. Cost-Utility analyses allow for the direct comparison of the relative health benefits of interventions across different disease areas and populations. It is imperative that we set up a National QOL Registry for patients with pediatric and CHD so that we can care for our patients in the safest possible manner to minimize harm, offer the best medical care to maximize QALYs, and do so in the most efficient manner to maximize patient value (cost per outcome).

References

1. Tweddell JS, Hoffman GM, Mussatto KA, et al. Improved survival of patients undergoing palliation of hypoplastic left heart syndrome: lessons learned from 115 consecutive patients. Circulation. 2002;106:I82–9.
2. Mahle WT, Spray TL, Wernovsky G, Gaynor JW, Clark 3rd BJ. Survival after reconstructive surgery for hypoplastic left heart syndrome: A 15-year experience from a single institution. Circulation. 2000;102:III136–41.
3. Warnes CA, Liberthson R, Danielson GK, et al. Task force 1: the changing profile of congenital heart disease in adult life. J Am Coll Cardiol. 2001;37:1170–5.
4. Mahle WT, Clancy RR, Moss EM, Gerdes M, Jobes DR, Wernovsky G. Neurodevelopmental outcome and lifestyle assessment in school-aged and adolescent children with hypoplastic left heart syndrome. Pediatrics. 2000;105:1082–9.
5. Wernovsky G, Stiles KM, Gauvreau K, et al. Cognitive development after the Fontan operation. Circulation. 2000;102:883–9.
6. Bellinger DC, Wypij D, duPlessis AJ, et al. Neurodevelopmental status at eight years in children with dextro-transposition of the great arteries: the Boston Circulatory Arrest Trial. J Thorac Cardiovasc Surg. 2003;126:1385–96.
7. Wray J, Sensky T. How does the intervention of cardiac surgery affect the self-perception of children with congenital heart disease? Child Care Health Dev. 1998;24:57–72.
8. Casey FA, Sykes DH, Craig BG, Power R, Mulholland HC. Behavioral adjustment of children with surgically palliated complex congenital heart disease. J Pediatr Psychol. 1996;21:335–52.
9. Davis CC, Brown RT, Bakeman R, Campbell R. Psychological adaptation and adjustment of mothers of children with congenital heart disease: stress, coping, and family functioning. J Pediatr Psychol. 1998;23:219–28.
10. Mahle WT, McBride MG, Paridon SM. Exercise performance after the arterial switch operation for D-transposition of the great arteries. Am J Cardiol. 2001;87:753–8.
11. Limperopoulos C, Majnemer A, Shevell MI, et al. Functional limitations in young children with congenital heart defects after cardiac surgery. Pediatrics. 2001;108:1325–31.
12. Paridon SM, Mitchell PD, Colan SD, et al. A cross-sectional study of exercise performance during the first 2 decades of life after the Fontan operation. J Am Coll Cardiol. 2008;52:99–107.
13. Ware Jr JE. Methodology in behavioral and psychosocial cancer research. Conceptualizing disease impact and treatment outcomes. Cancer. 1984;53:2316–26.
14. Aaronson NK. Quality of life: what is it? How should it be measured? Oncology (Williston Park). 1988;2:69–76.

15. Drotar D. Measuring child health: scientific questions, challenges, and recommendations. Ambul Pediatr. 2004;4:353–7.
16. World Health Organization. Preamble to the Constitution of the World Health Organization as adopted by the International Health Conference, New York, 19–22 June 1946; signed on 22 July 1946 by the representatives of 61 States (Official Records of the World Health Organization, no. 2, p. 100) and entered into force on 7 April 1948.
17. Stein REK, Jessop DJ. Functional status II(R): a measure of child health status. Med Care. 1990;28: 1041–55.
18. U.S. Department of Health and Human Services FDA Center for Drug Evaluation and Research, U.S. Department of Health and Human Services FDA Center for Biologics Evaluation and Research, and U.S. Department of Health and Human Services FDA Center for Devices and Radiological Health. Guidance for industry: patient-reported outcome measures: use in medical product development to support labeling claims. 2006:1–20.
19. Drotar D. Measuring health-related quality of life in children and adolescents. Mahwah: Lawrence Erlbaum Associates Publishers; 1998.
20. Higginson IJ, Carr AJ. Measuring quality of life: using quality of life measures in the clinical setting. BMJ. 2001;322:1297–300.
21. Lee CP, Chertow GM, Zenios SA. An empiric estimate of the value of life: updating the renal dialysis cost-effectiveness standard. Value Health. 2009;12:80–7.
22. Cella D, Riley W, Stone A, et al. The Patient-Reported Outcomes Measurement Information System (PROMIS) developed and tested its first wave of adult self-reported health outcome item banks: 2005–2008. J Clin Epidemiol. 2010;63:1179–94.
23. Hahn E, DeVellis R, Bode R, et al. Measuring social health in the Patient-Reported Outcomes Measurement Information System (PROMIS): item bank development and testing. Qual Life Res. 2010;19:1035–44.
24. Hays R, Bjorner J, Revicki D, Spritzer K, Cella D. Development of physical and mental health summary scores from the Patient-Reported Outcomes Measurement Information System (PROMIS) global items. Qual Life Res. 2009;18:873–80.
25. Irwin DE, Varni JM, Yeatts K, DeWalt D. Cognitive interviewing methodology in the development of a pediatric item bank: a Patient Reported Outcomes Measurement Information System (PROMIS) study. Health Qual Life Outcomes. 2009;7:3.
26. Rothrock NE, Hays RD, Spritzer K, Yount SE, Riley W, Cella D. Relative to the general US population, chronic diseases are associated with poorer health-related quality of life as measured by the Patient-Reported Outcomes Measurement Information System (PROMIS). J Clin Epidemiol. 2010;63:1195–204.
27. Ware JE. The Patient-Reported Outcomes Measurement Information System (PROMIS) seeks to improve and standardize measures of five generic health-related QOL domains. Pediatr Rep Outcomes Newsl. 2007;38:1–3.
28. U.S. Department of Health and Human Services Food and Drug Administration, Center for Drug Evaluation and Research (CDER), Center for Biologics Evaluation and Research (CBER), and Center for Devices and Radiological Health (CDRH). Guidance for industry: patient-reported outcome measures: use in medical product development to support labeling claims. 2009:1–43.
29. Janse AJ, Uiterwaal CS, Gemke RJ, Kimpen JL, Sinnema G. A difference in perception of quality of life in chronically ill children was found between parents and pediatricians. J Clin Epidemiol. 2005;58:495–502.
30. Marino BS, Tomlinson RS, Drotar D, et al. Quality-of-life concerns differ among patients, parents, and medical providers in children and adolescents with congenital and acquired heart disease. Pediatrics. 2009;123:e708–15.
31. Moons P, Van Deyk K, Budts W, De Geest S. Caliber of quality-of-life assessments in congenital heart disease: a plea for more conceptual and methodological rigor. Arch Pediatr Adolesc Med. 2004;158:1062–9.
32. Gill TM, Feinstein AR. A critical appraisal of the quality of quality-of-life measurements. JAMA. 1994;272:619–26.
33. Varni JW. The Pediatric Cancer Quality of Life Inventory (PCQL). I. Instrument development, descriptive statistics, and cross-informant variance. J Behav Med. 1998;21:179–204.
34. Eiser C, Morse R. Can parents rate their child's health-related quality of life? Results of a systematic review. Qual Life Res. 2001;10:347–57.
35. Upton P, Lawford J, Eiser C. Parent–child agreement across child health-related quality of life instruments: a review of the literature. Qual Life Res. 2008;17: 895–913.
36. Theunissen NCM, Vogels TGC, Koopman HM, et al. The proxy problem: child report versus parent report in health-related quality of life research. Qual Life Res. 1998;7:387–97.
37. Varni JW, Limbers CA, Burwinkle TM. How young can children reliably and validly self-report their health-related quality of life? An analysis of 8,591 children across age subgroups with the PedsQL 4.0 Generic Core Scales. Health Qual Life Outcomes. 2007;5:1–13.
38. Campo JV, Comer DM, Jansen-McWilliams L, Gardner W, Kelleher KJ. Recurrent pain, emotional distress, and health service use in childhood. J Pediatr. 2002;141:76–83.
39. Janicke DM, Finney JW, Riley AW. Children's health care use: a prospective investigation of factors related to care-seeking. Med Care. 2001;39:990–1001.
40. Cremeens J, Eiser C, Blades M. Characteristics of health-related self-report measures for children aged three to eight years: a review of the literature. Qual Life Res. 2006;15:739–54.
41. Riley AW. Evidence that school-age children can self-report on their health. Ambul Pediatr. 2004;4:371–6.
42. Olson LM, Lara M, Pat Frintner M. Measuring health status and quality of life for US children: relationship

to race, ethnicity, and income status. Ambul Pediatr. 2004;4:377–86.
43. Landgraf JM, Abezt L, Ware JE. The Child Health Questionnaire (CHQ): a user's manual. Boston: The Health Institute, New England Medical Center; 1996.
44. Varni JW, Seid M, Rode CA. The PedsQL: measurement model for the pediatric quality of life inventory. Med Care. 1999;37:126–39.
45. Kamphuis M, Zwinderman KH, Vogels T, et al. A cardiac-specific health-related quality of life module for young adults with congenital heart disease: development and validation. Qual Life Res. 2004;13:735–45.
46. Uzark K, Jones K, Burwinkle TM, Varni JW. The Pediatric Quality of Life Inventory in children with heart disease. Prog Pediatr Cardiol. 2003;18:141–9.
47. Kendall L, Lewin RJ, Parsons JM, Veldtman GR, Quirk J, Hardman GE. Factors associated with self-perceived state of health in adolescents with congenital cardiac disease attending paediatric cardiologic clinics. Cardiol Young. 2001;11:431–8.
48. Macran S, Birks Y, Parsons J, et al. The development of a new measure of quality of life for children with congenital cardiac disease. Cardiol Young. 2006;16:165–72.
49. Marino BS, Shera D, Wernovsky G, et al. The development of the pediatric cardiac quality of life inventory: a quality of life measure for children and adolescents with heart disease. Qual Life Res. 2008;17: 613–26.
50. Streiner DL, Norman GR. Health measurement scales. New York: Oxford University Press; 1995.
51. Koot HM, Wallander JL. Quality of life in child and adolescent illness: concepts, methods and findings. New York: Brunner-Routledge; 2001.
52. Messick S. Validity of Psychological Assessment: Validation of inferences from persons' responses and performances as scientific inquiry into score meaning. Am Psychol. 1995;50:741–9.
53. Clark LA, Watson D. Constructing validity: basic issues in objective scale development. Psychol Assess. 1995;7:309–19.
54. Lohr KN. Assessing health status and quality-of-life instruments: attributes and review criteria. Qual Life Res. 2002;11:193–205.
55. Thorndike RM, Thorndike-Christ T. Measurement and evaluation in psychology and education. Boston: Pearson; 2010.
56. Uzark K, Jones K, Slusher J, Limbers CA, Burwinkle TM, Varni JW. Quality of life in children with heart disease as perceived by children and parents. Pediatrics. 2008;121:e1060–7.
57. Marino BS, Tomlinson R, Wernovsky G, et al. Validation of the pediatric cardiac quality of life inventory. Pediatrics. 2010;126:498–508.
58. Marino BS, Drotar D, Cassedy A, et al. External validity of the pediatric cardiac quality of life inventory. Qual Life Res. 2011;20:205–14.
59. Gersony WM, Hayes CJ, Driscoll DJ, et al. Second natural history study of congenital heart defects. Quality of life of patients with aortic stenosis, pulmonary stenosis, or ventricular septal defect. Circulation. 1993;87(2 Suppl):I52–65.
60. Walker RE, Gauvreau K, Jenkins KJ. Health-related quality of life in children attending a cardiology clinic. Pediatr Cardiol. 2004;25:40–8.
61. Majnemer A, Limperopoulos C, Shevell M, Rohlicek C, Rosenblatt B, Tchervenkov C. Health and well-being of children with congenital cardiac malformations, and their families, following open-heart surgery. Cardiol Young. 2006;16:157–64.
62. DeMaso DR, Campis LK, Wypij D, Bertram S, Lipshitz M, Freed M. The impact of maternal perceptions and medical severity on the adjustment of children with congenital heart disease. J Pediatr Psychol. 1991;16:137–49.
63. Latal B. Psychological adjustment and quality of life in children and adolescents following open-heart surgery for congenital heart disease: a systematic review. BMC Pediatr. 2009;9:6.
64. Mussatto K, Tweddell J. Quality of life following surgery for congenital cardiac malformations in neonates and infants. Cardiol Young. 2005;15 Suppl 1:174–8.
65. Culbert EL, Ashburn DA, Cullen-Dean G, et al. Quality of life of children after repair of transposition of the great arteries. Circulation. 2003;108:857–62.
66. Dunbar-Masterson C, Wypij D, Bellinger DC, et al. General health status of children with D-transposition of the great arteries after the arterial switch operation. Circulation. 2001;104:I138–42.
67. Hovels-Gurich HH, Konrad K, Wiesner M, et al. Long term behavioural outcome after neonatal arterial switch operation for transposition of the great arteries. Arch Dis Child. 2002;87:506–10.
68. Brosig CL, Mussatto KA, Kuhn EM, Tweddell JS. Psychosocial outcomes for preschool children and families after surgery for complex congenital heart disease. Pediatr Cardiol. 2007;28:255–62.
69. McCrindle BW, Williams RV, Mitchell PD, et al. Relationship of patient and medical characteristics to health status in children and adolescents after the Fontan procedure. Circulation. 2006;113:1123–9.
70. DeMaso DR, Lauretti A, Spieth L, et al. Psychosocial factors and quality of life in children and adolescents with implantable cardioverter-defibrillators. Am J Cardiol. 2004;93:582–7.
71. Czosek J, Bonney WJ, Mah D, Cassedy A, Tanel RE, Imundo JR, Singh AK, Cohen M, Miyake C, Fawley K, Marino BS. Impact of cardiac devices on the quality of life in pediatric patients and parents. 2011; 124:A10016.
72. DeMaso DR, Douglas Kelley S, Bastardi H, O'Brien P, Blume ED. The longitudinal impact of psychological functioning, medical severity, and family functioning in pediatric heart transplantation. J Heart Lung Transplant. 2004;23:473–80.
73. Wray J, Radley-Smith R. Longitudinal assessment of psychological functioning in children after heart or heart-lung transplantation. J Heart Lung Transplant. 2006;25:345–52.

74. Williams DL, Gelijns AC, Moskowitz AJ, et al. Hypoplastic left heart syndrome: valuing the survival. J Thorac Cardiovasc Surg. 2000;119:720–31.
75. Drotar D, Stancin T, Dworkin PH, Sices L, Wood S. Selecting developmental surveillance and screening tools. Pediatr Rev. 2008;29:e52–8.
76. Marino BS, Beebe D, Cassedy A, et al. Executive functioning, gross motor ability and mood are key drivers of poorer quality of life in child and adolescent survivors with complex congenital heart disease. J Am Coll Cardiol. 2011;57:421.
77. American Academy of Pediatrics Committee on Children with Disabilities and Committee on Psychosocial Aspects of Child and Family Health: Psychosocial risks of chronic health conditions in childhood and adolescence. Pediatrics. 1993;92:876–8.
78. Spijkerboer A, Utens E, Bogers A, Verhulst F, Helbing W. Long-term behavioural and emotional problems in four cardiac diagnostic groups of children and adolescents after invasive treatment for congenital heart disease. Int J Cardiol. 2008;125:66–73.
79. Spurkland I, Bjørnstad PG, Lindberg H, Seem E. Mental health and psychosocial functioning in adolescents with congenital heart disease. A comparison between adolescents born with severe heart defect and atrial septal defect. Acta Paediatr. 1993;82:71–6.
80. Marino BS, Cassedy AEC, Drotar D, et al. Psychosocial morbidity factors mediate the relationship between heart disease complexity and lower quality of life. Submitted for national presentation at the 61th Scientific Sessions of the American College of Cardiology, Chicago, 2012.
81. Wray J, Brown K, Franklin R, et al. The Pediatric Cardiac Quality of Life Inventory: testing a new disease specific quality of life measure in the UK. Arch Dis Child. 2010;95:A74.
82. Ungar WJ, Santos MT. Trends in paediatric health economic evaluation: 1980 to 1999. Arch Dis Child. 2004;89(1):26–9.
83. Griebsch I, Coast J, Brown J. Quality-adjusted life-years lack quality in pediatric care: a critical review of published cost-utility studies in child health. Pediatrics. 2005;115(5):e600–14.
84. Hjelmgren J, Berggren F, Andersson F. Health economic guidelines–similarities, differences and some implications. Value Health. 2001;4(3):225–50.
85. Gold M, Russell L, Siegel J, Weinstein M. Cost-effectiveness in health and medicine. Oxford: Oxford University Press; 1996.
86. Drummond M, O'Brien B, Stoddart G, Torrance G. Methods for the economic evaluation of health care programmes. 2nd ed. Oxford: Oxford University Press; 1997.

Longitudinal Follow-Up Studies in the Pediatric Heart Network

33

Lynn Mahony, Lynn A. Sleeper, and Gail D. Pearson

Abstract

To promote collaborative research leading to evidence-based treatment options for pediatric patients with congenital and acquired heart disease, the National Heart, Lung, and Blood Institute established the Pediatric Heart Network (PHN) in 2001. The infrastructure is now well-developed and capable of implementing complex, multicenter protocols efficiently and recruiting subjects effectively. In addition, we have developed mechanisms to retain subjects and have established several cohorts that we are following long-term. The purpose of this chapter is to describe the structure of the PHN and to review studies relevant to longitudinal patient outcome. The PHN is uniquely positioned to contribute to the body of knowledge regarding evidenced-based treatment approaches for pediatric patients with cardiovascular disease.

Keywords

Randomized clinical trials • Congenital heart disease • Pediatric clinical research • Single ventricle • Congenital heart surgery • Cardiomyopathy in children • Outcomes

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Although morbidity, mortality, and quality of life have improved greatly in the past 50 years for patients with congenital and acquired heart disease, these improvements may have been limited by the fact that many contemporary treatments are based on expert opinion, single institution observational studies, or extrapolated from adult cardiovascular medicine. Barriers to developing and applying evidence-based approaches to pediatric cardiovascular conditions have often precluded the conduct of studies evaluating treatments for these patients. These barriers

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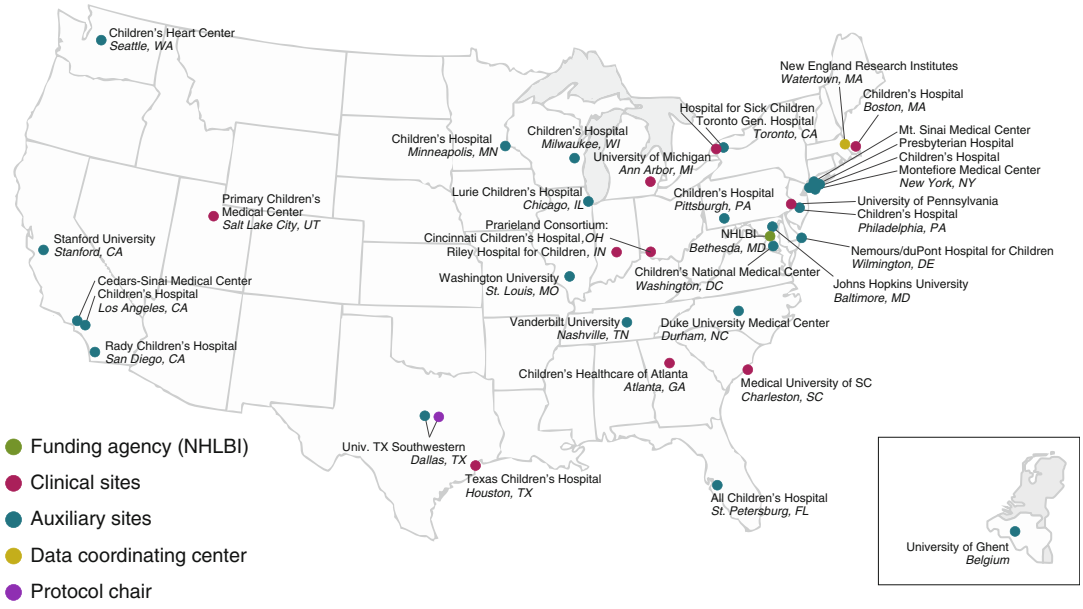

Fig. 33.1 Map showing participants in the Pediatric Heart Network (Adapted from Mahony et al. [1])

include: (1) the number of patients with a given condition being too small for research questions to be answered by a single center study; (2) ethical considerations involved in performing research studies in children; (3) potential lack of therapeutic equipoise; and (4) the high cost of pediatric research including the need for lengthy longitudinal follow-up to accurately assess outcomes.

In an effort to address this situation, the National Heart, Lung, and Blood Institute (NHLBI) established the Pediatric Heart Network (PHN) in 2001 [1]. The PHN has been steadily expanded by the NHLBI and currently has nine core Clinical Centers, one Data Coordinating Center (DCC), a biorepository, several core laboratories, and more than 25 auxiliary centers contributing to various studies (Fig. 33.1). The goals of the PHN are to accelerate research in the diagnosis and management of congenital and acquired pediatric heart disease, to standardize existing treatments, and to evaluate new therapies to bring evidence-based medicine to the care of children with heart disease. The purpose of this chapter is to describe the structure of the PHN and to review studies relevant to longitudinal patient outcome.

Structure of the PHN

The DCC is responsible for overall coordination of operations. This includes (1) maintaining systems for electronic communications, administrative management, and coordination; (2) all phases of protocol development and implementation; (3) design, testing, and maintenance of secure data collection and management systems; (4) tracking and execution of regulatory submissions and reporting; (5) systems and procedures for quality assurance; and (6) study design, data monitoring, conduct of data analyses, and collaboration on manuscripts.

The main governing body of the PHN is the Executive Committee which includes the Protocol Chair, the Principal Investigator from each participating core Clinical Center and the DCC, and the NHLBI Program Officer. The Executive Committee is responsible for developing all policies and procedures and for reviewing and prioritizing study proposals in consultation with the Steering Committee. Membership in the PHN Steering Committee is open to investigators from the core Clinical Centers and all auxiliary enrolling sites, and personnel from the DCC, core laboratories and NHLBI.

Clinical Center investigators as well as investigators from outside the PHN provide the intellectual leadership for developing proposals for new studies. For proposals approved by the Executive Committee, the investigators write the formal protocol with statistical, quality assurance, and regulatory consultation from the DCC. Every proposed study is reviewed by two independent committees established by NHLBI that consist of individuals who do not have close professional relationships to PHN investigators and who have expertise relevant to the proposed study. The Protocol Review Committee assesses the scientific merit and feasibility of all proposed protocols. The Data and Safety Monitoring Board (DSMB) reviews protocols and consent forms with special emphasis on subject safety. In addition, an independent medical monitor adjudicates serious adverse events as they are reported for all PHN studies.

The Clinical Centers and auxiliary enrolling sites are responsible for subject recruitment and execution of the study protocol. During the course of the study all data, including adverse events (by study arm, where applicable), are reviewed to ensure the safety of study subjects by the DSMB. Furthermore, the DSMB advises the NHLBI on data quality, achievement of recruitment and study aims as well as ethical and human subjects issues. If the trial design includes early stopping rules, the DSMB also provides recommendations with regard to continuing or stopping the study prematurely. Members of the Clinical Centers, auxiliary enrolling sites, the Protocol Chair, the DCC, and the NHLBI staff collaborate in reporting results of the studies through presentations, abstracts, and manuscripts.

PHN Studies

Longitudinal Fontan Studies

For patients born with single ventricle physiology, the Fontan procedure restores near-normal systemic oxygen saturation, reduces the demands on the systemic ventricle, and extends the lives of patients with even the most complex forms of congenital heart disease. Nevertheless, the resulting abnormal hemodynamic state is associated with a variety of late complications including decreased exercise performance, abnormal ventricular function, intracardiac and extracardiac thrombosis and embolic phenomena, arrhythmias and conduction system impairment, and protein-losing enteropathy [2].

Despite these well-recognized problems, few contemporary treatment strategies are evidence-based because randomized clinical trials are difficult to perform in this population. In trying to design a clinical trial, the PHN had difficulty identifying a primary endpoint that could be observed within a reasonable time period. Surrogate endpoints, such as functional health status, are often desirable to optimize the feasibility of a study, but the PHN recognized that understanding of the association between functional health status and laboratory measures was limited. We therefore designed the Fontan 1 study with the goal of identifying one or more quantifiable laboratory measures of cardiovascular performance that correlated with a validated health-related quality of life instrument (a potential surrogate outcome). We performed a cross-sectional study (Fontan 1) of 546 children who were 6–18 years of age at study entry (mean 11.9 ± 3.4 years) [3], creating a broad and unique dataset that was used to investigate the relationship of functional health status of these subjects to medical history and various laboratory measures of cardiac function [4–11]. Unfortunately, in our cohort of relatively healthy Fontan patients, laboratory measures accounted for only a small proportion of the variability in functional health status. Thus, functional health status may not be an optimal surrogate endpoint for trials of therapeutic interventions in this patient population [9].

Although this study is the largest to date of children who have undergone the Fontan operation, every study manuscript noted the limitations inherent to the cross-sectional design. Additionally, it was difficult to determine whether differences between older and younger patients were related to the length of time each had lived with Fontan physiology or to secular changes in management strategies for palliating patients with

Fontan physiology. Unfortunately, longitudinal data in this population are difficult to obtain. Of concern, the frequency and severity of many of the complications related to the Fontan procedure increases as these patients become older. Furthermore, as these patients age into young adulthood, access to specialized health care may become limited. A follow-up study (Fontan 2) at an average of 7 years after enrollment in Fontan 1 was performed that included a limited re-evaluation of the original Fontan 1 cohort using the following outcomes: vital status, functional health status, interim medical events, and access to health care. This study enrolled 427 of the 502 eligible subjects in the Fontan 1 cohort. Only 2.6 % (13 of the eligible subjects) were lost to follow-up. Of the original cohort, 5 % had died or undergone transplantation. Deterioration in physical functioning during the 7 years between studies was associated with respiratory conditions and protein-losing enteropathy [12]. In addition, the change in physical functioning score was not significantly associated with laboratory measures of exercise capacity and ventricular characteristics and function obtained 7 years earlier as part of the Fontan 1 study.

We are currently performing the Fontan 3 study. In this study, we are collecting vital and cardiac transplant status data from medical records and public records on the 546 subjects screened for the Fontan 2 study, and then approaching those alive with a Fontan circulation for enrollment in Fontan 3. Repeat administration of age-appropriate health status questionnaires, maximal exercise testing, echocardiography and B-type natriuretic peptide analysis are being performed prospectively during a visit to a participating PHN center and compared to data obtained during the previous Fontan studies. Additional data will be collected by medical record review and questionnaires to assess socioeconomic status, family functioning, and access to health care. The Fontan 3 study will also collect biological specimens for storage in a central repository for future genetic studies. Longitudinal assessment of functional health status and repeat testing of laboratory measures of ventricular function an average of 9 years after initial enrollment in the Fontan 1 study is of significant clinical importance and will provide useful comparisons to the single ventricle subjects being followed prospectively from birth in other PHN studies.

Single Ventricle Reconstruction Trial

The PHN conducted the first multi-center, randomized clinical trial that compared outcomes of the Norwood procedure with either a right-ventricular-to-pulmonary-artery shunt (RVPAS) or a modified Blalock-Taussig Shunt (MBTS), in patients with hypoplastic left heart syndrome and other single right ventricle anomalies [13]. The results showed that a Norwood procedure with RVPAS (n=274), compared to MBTS (n=275), was associated with better transplant-free survival 12 months after randomization (74 % vs. 64 %, respectively, P=0.01) [14]. Of note, the rate of unintended cardiovascular interventions (P=0.003) and complications through 12 months after randomization was higher in the RVPAS group (P=0.002).

This trial is an excellent example of the importance of longitudinal follow-up. Although the primary outcome in the trial was transplant-free survival 12 months after randomization, we noted that the difference between groups in transplant-free survival (P=0.06) did not achieve statistical significance when we used all follow-up data available at the time of the report (mean for transplant-free survivors, 32±11 months). In our ongoing follow-up of the trial cohort, we have found that by 3 years after randomization, the Norwood procedure with RVPAS, compared with MBTS, was no longer associated with superior transplant-free survival (Fig. 33.2) [15].

The PHN Single Ventricle Reconstruction trial has resulted in the largest cohort to date of children with single right ventricular anomalies. Ongoing follow-up of this cohort is underway to delineate the evolving natural history after the Norwood procedure using the RVPAS compared with the MBTS strategy [16–20]. This study cohort is very well-characterized and provides a unique opportunity to explore determinants of long-term outcome in these fragile patients. Biospecimens have been stored for genetic analysis.

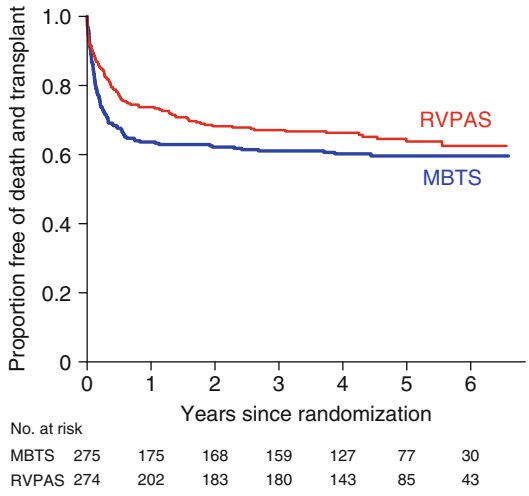

Fig. 33.2 Comparison of the shunt types by intention-to-treat analysis in their freedom from the composite endpoint of death or cardiac transplant for the cohort of subjects in the single ventricle reconstruction trial (i.e., transplant-free survival). Using all available data, transplant-free survival did not differ between groups (logrank P = .14). *RVPAS* right ventricular to pulmonary artery shunt, *MBTS* Modified Blalock-Taussig shunt, *No* number (Adapted from Newburger et al. [15])

Marfan Trial

Cardiovascular pathology, including aortic root dilation, dissection, and rupture, is the leading cause of mortality in patients with Marfan syndrome. Although advances in therapy have improved life expectancy, affected individuals continue to suffer cardiovascular morbidity and mortality. Studies in an *FBN1*-targeted mouse model of Marfan syndrome with aortic pathology similar to that seen in humans showed that treatment with losartan normalized aortic root growth and aortic wall architecture [21]. These compelling results prompted a desire to translate these results systematically to humans.

The PHN therefore designed a clinical trial to compare aortic root growth, which is the best predictor of cardiovascular outcome, in children and young adults with Marfan syndrome randomized to receive atenolol or losartan for a period of 3 years [22]. Between 2007 and 2011, with the help of the National Marfan Foundation, 21 clinical sites randomized 608 subjects, aged 6 months to 25 years who met diagnostic criteria and had a body surface area-adjusted aortic root diameter z-score > 3.0. Five echocardiograms over 3 years were submitted for each subject to assess the primary endpoint, the rate of change in maximum aortic root diameter (sinuses of Valsalva) z-score. Baseline demographic, clinical, and anthropometric characteristics of the randomized cohort are representative of patients in this population with moderate to severe aortic root dilation [23]. Mean age at study entry was 11.2 ± 6.3 years, 25 % were older teenagers and young adults, and 60 % were male. Nearly two-thirds had a family history of Marfan syndrome. The median baseline aortic root diameter z-score was 4.0. Inter- and intra-observer variability of echocardiographic measurements made at the core laboratory showed excellent reproducibility [24]. We expect that the results of the trial, which will be available in 2014, and the wealth of systematic data collected will make an important contribution to the management of individuals with Marfan syndrome.

Challenges

Despite the participation of a large number of talented investigators and the generous resources provided by NHLBI, we encounter a number of barriers in trying to plan and execute studies.

Selection of a Primary Endpoint

Selection of an appropriate primary endpoint is often quite controversial within the PHN, as it is within other groups of trialists. Ideally the primary endpoint selected for a prospective trial should be clinically meaningful; that is, it should measure how a subject survives, feels, or functions [25, 26]. These types of endpoints are sometimes called "hard endpoints". We were able to use such an endpoint in the Single Ventricle Reconstruction trial (transplant-free survival) but often these "hard" endpoints are difficult to evaluate in pediatric cardiology patients because events such as death and major complications are relatively rare [26, 27].

Because of these difficulties, we like many others have considered surrogate markers as substitute outcome measures for a "hard" endpoint [28–30]. Surrogate endpoints are usually laboratory measures (e.g., ejection fraction or peak oxygen consumption) or patient-reported measures of functional health status that are expected to predict a clinically meaningful endpoint, but it can be measured earlier in the clinical course than an outcome such as death or major complication. Aortic root diameter is a well-accepted surrogate for aortic dissection in patients with Marfan syndrome so we were able to use this echocardiographic measurement as the primary outcome in the Marfan trial. In contrast, we continue to struggle to design prospective trials in the Fontan population because of the absence of validated surrogate outcomes (see discussion above).

Estimation of Sample Size and Subject Availability

An accurate estimate of the outcome rate (or mean and variance of the outcome measure) in the control group is necessary to calculate the sample size necessary for adequate statistical power. The PHN conducts formal chart reviews during the protocol development phase to estimate current event rates in our participating sites rather than relying on historical data because event rates for many conditions of interest change relatively rapidly.

The sample size required for a study also depends on the magnitude of the treatment difference that investigators consider clinically meaningful and wish to observe. In addition to determining the outcome rate in the control group, the size of the minimum clinically significant treatment effect must be specified. For a given statistical power, more subjects are needed to evaluate smaller treatment differences. It is important to design studies that will have sufficient power to detect the minimum clinically important difference, even if the magnitude of that difference is smaller than what has been observed in a prior study. Otherwise, the trial may miss clinically-important treatment benefits. In addition, if the overall event rate is overestimated, the trial's target sample size may not yield the power necessary to detect the specified treatment difference. For this reason, interim analysis of aggregate event rates (or, for continuous measures, the standard deviation) is desirable to determine whether a modification to sample size is indicated to successfully execute the study.

Accurately defining the number of potentially available subjects can be difficult. We designed a placebo-controlled randomized trial of enalapril in children with mitral regurgitation after repair of an atrioventricular septal defect. As part of a feasibility study, individual sites reviewed their databases to determine the number of patients with mitral regurgitation based on subjective assessment from the echocardiograms. For the purposes of the actual study inclusion criteria, however, we created an algorithm requiring quantitative measurements to define mitral regurgitation. Unfortunately, application of this algorithm excluded many of the patients previously thought to be eligible. Recruitment was very poor and the trial was eventually abandoned [31]. Based on this experience, we now conduct all feasibility studies using actual trial entry criteria.

Subject Recruitment and Retention

We have found recruiting patients into our studies, especially those with severe defects, to be challenging and have therefore developed an Inventory of Best Practices which has greatly facilitated this process (Table 33.1) [32]. Clinicians and other personnel caring for patients must be involved in a collaborative manner and educated regarding the rationale and procedures for the study. The timing of a trial is also critical to recruitment success. A study cannot be started until all centers have developed sufficient experience with any new procedures or therapies, but it must be started before a new treatment becomes the standard of care despite any evidence beyond anecdotal reports that it is the best approach [33].

Table 33.1 Inventory of best recruitment practices

Partnerships with clinicians	Provide study updates and meet regularly (conferences, bedside rounds, Division and Department meetings) with staff to convey a sense of partnership, to address concerns and keep the study visible
	Determine the local practices for using the study drug routinely in this population and address issues of equipoise
Fostering relationships with families	Remain in consistent contact with the family from birth to study completion by the same members of the research team whenever possible
	Allow patients to complete one follow-up visit locally (non-Pediatric Heart Network centre) to alleviate long-distance travel for some families
	Ask the cardiac surgeon or physician who has an established relationship with the family to mention the trial and advise that study personnel will talk with them about participation
Enhancing the environment	Approach the family for study enrolment at a less stressful time – when the infant is more stable and perhaps transitioning out of the critical care unit
	Present an informational study brochure before providing the lengthier informed consent document but always as part of a verbal discussion
	Introduce the idea of research participation at a prenatal cardiology follow-up visit

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A variety of approaches facilitate recruiting and retaining subjects. For the Marfan trial, we were greatly helped by the advocacy efforts of the National Marfan Foundation to encourage enrollment in the trial. We also recognized that many families are not familiar with the processes and procedures involved in human subjects research. The PHN therefore developed the Children and Clinical Studies informational website; this is now used routinely to assist in the educational process [34].

One of the most important aspects of retaining subjects, especially in long-term follow-up studies, is the relationships that are developed with the research staff. One of the advantages of the PHN structure is that we are able to support the same clinical site staff over a number of years. Subjects and their families often feel that members of the research staff are integral to the overall care of the subject. In addition, when the families consent to our studies, they also consent to being contacted frequently, and to permit access to medical records for several years after a study ends.

Another issue that affects both recruitment and retention is that study subjects often live some distance from the study center. These patients are often followed by a pediatric cardiologist near their homes, and do not return to the study center for routine care, making studies that require long-term follow-up at the PHN Center more challenging. The trusting relationships developed between subjects, families and the research staff are particularly important to this effort. We try to allow some follow-up visits to be completed locally, but we also can pay for transportation and lodging when necessary.

Conclusions

The PHN was established evidence-based approaches to management of pediatric patients with cardiovascular disease. A large number of centers in the US that are involved in the care of these patients are now involved in one or more studies. The well-developed infrastructure allows efficient implementation of complex multicenter protocols and effective subject recruitment. In addition, we have developed mechanisms to retain subjects in our studies thereby ensuring longitudinal follow-up. The accomplishments of the PHN will permit those taking care of children with heart disease to evaluate clinical practice critically and will provide data for an evidence-based rationale for diagnostic and therapeutic options.

References

1. Mahony L, Sleeper LA, Anderson PA, Gersony WM, McCrindle BW, Minich LL, et al. The Pediatric Heart Network: a primer for the conduct of multicenter studies in children with congenital and acquired heart disease. Pediatr Cardiol. 2006;27(2):191–8. PubMed PMID: 16261271. Epub 2005/11/02. eng.
2. Khairy P, Poirier N, Mercier LA. Univentricular heart. Circulation. 2007;115(6):800–12. PubMed PMID: 17296869. Epub 2007/02/14. eng.
3. Sleeper LA, Anderson P, Hsu DT, Mahony L, McCrindle BW, Roth SJ, et al. Design of a large cross-sectional study to facilitate future clinical trials in children with the Fontan palliation. Am Heart J. 2006;152(3):427–33. PubMed PMID: 16923408. Epub 2006/08/23. eng.
4. McCrindle BW, Williams RV, Mitchell PD, Hsu DT, Paridon SM, Atz AM, et al. Relationship of patient and medical characteristics to health status in children and adolescents after the Fontan procedure. Circulation. 2006;113(8):1123–9. PubMed PMID: 16490823. Epub 2006/02/24. eng.
5. Anderson PA, Sleeper LA, Mahony L, Colan SD, Atz AM, Breitbart RE, et al. Contemporary outcomes after the Fontan procedure: a Pediatric Heart Network multicenter study. J Am Coll Cardiol. 2008;52(2):85–98. PubMed PMID: 18598886. Epub 2008/07/05. eng.
6. Paridon SM, Mitchell PD, Colan SD, Williams RV, Blaufox A, Li JS, et al. A cross-sectional study of exercise performance during the first 2 decades of life after the Fontan operation. J Am Coll Cardiol. 2008;52(2):99–107. PubMed PMID: 18598887. eng.
7. Blaufox AD, Sleeper LA, Bradley DJ, Breitbart RE, Hordof A, Kanter RJ, et al. Functional status, heart rate, and rhythm abnormalities in 521 Fontan patients 6 to 18 years of age. J Thorac Cardiovasc Surg. 2008;136(1):100–7. 7.e1. PubMed PMID: 18603061. Pubmed Central PMCID: 2525868. Epub 2008/07/08. eng.
8. Margossian R, Schwartz ML, Prakash A, Wruck L, Colan SD, Atz AM, et al. Comparison of echocardiographic and cardiac magnetic resonance imaging measurements of functional single ventricular volumes, mass, and ejection fraction (from the Pediatric Heart Network Fontan Cross-Sectional Study). Am J Cardiol. 2009;104(3):419–28. PubMed PMID: 19616678. Pubmed Central PMCID: 2741330. Epub 2009/07/21. eng.
9. McCrindle BW, Zak V, Sleeper LA, Paridon SM, Colan SD, Geva T, et al. Laboratory measures of exercise capacity and ventricular characteristics and function are weakly associated with functional health status after Fontan procedure. Circulation. 2010;121(1):34–42. PubMed PMID: 20026781. Pubmed Central PMCID: 2810546. Epub 2009/12/23. eng.
10. Anderson PA, Breitbart RE, McCrindle BW, Sleeper LA, Atz AM, Hsu DT, et al. The Fontan patient: inconsistencies in medication therapy across seven pediatric heart network centers. Pediatr Cardiol. 2010;31(8):1219–28. PubMed PMID: 20938655. eng.
11. Stephenson EA, Lu M, Berul CI, Etheridge SP, Idriss SF, Margossian R, et al. Arrhythmias in a contemporary Fontan cohort: prevalence and clinical associations in a multicenter cross-sectional study. J Am Coll Cardiol. 2010;56(11):890–6. PubMed PMID: 20813285. Pubmed Central PMCID: 3200364. Epub 2010/09/04. eng.
12. McCrindle BW, Zak V, Breibart RE, Mahony L, Lai WW, Burns KM, et al. The relationship of patient, medical and laboratory characteristics to changes in functional health status in children and adolescents after the Fontan procedure. Pediatr Cardiol. 2014;35(4):632–40. PubMed PMID:24264999. PubMed Central PMCID: 3959245 Epub 2013/11/22. eng.
13. Ohye RG, Gaynor JW, Ghanayem NS, Goldberg CS, Laussen PC, Frommelt PC, et al. Design and rationale of a randomized trial comparing the Blalock-Taussig and right ventricle-pulmonary artery shunts in the Norwood procedure. J Thorac Cardiovasc Surg. 2008;136(4):968–75. PubMed PMID: 18954638. Pubmed Central PMCID: 2745283. Epub 2008/10/29. eng.
14. Ohye RG, Sleeper LA, Mahony L, Newburger JW, Pearson GD, Lu M, et al. Comparison of shunt types in the Norwood procedure for single-ventricle lesions. N Engl J Med. 2010;362(21):1980–92. PubMed PMID: 20505177. Pubmed Central PMCID: 2891109. Epub 2010/05/28. eng.
15. Newburger J, Sleeper LA, Pearson GD, Mahle W, Chen S, Dunbar-Masterson C, et al. Transplant-free survival and interventions at 3 years in the single ventricle reconstruction trial. Circulation. 2014; 129(20):2013–20. PMID:24705119. Pubmed Central PMCID:PMC4029928. Epub 2014 Apr 4. eng.
16. Frommelt PC, Gerstenberger E, Cnota J, Cohen MS, Gorentz J, Hill KD, et al. Changes in cardiac size and function in children with single right ventricle anomalies after Norwood-RV remodeling after the first year varies by initial shunt type. J Am Coll Cardiol. 2013;61(10):E557.
17. Frommelt PC, Gerstenberger E, Baffa J, Border WL, Bradley TJ, Colan S, et al. Doppler flow patterns in the right ventricle-to-pulmonary artery shunt and neo-aorta in infants with single right ventricle anomalies: impact on outcome after initial staged palliations. J Am Soc Echocardiogr. 2013;26(5):521–9. PubMed PMID: 23540728. Epub 2013/04/02. Eng.
18. Ghanayem NS, Allen KR, Tabbutt S, Atz AM, Clabby ML, Cooper DS, et al. Interstage mortality after the Norwood procedure: results of the multicenter Single Ventricle Reconstruction trial. J Thorac Cardiovasc Surg. 2012;144(4):896–906. PubMed PMID: 22795436. Epub 2012/07/17. eng.
19. Tweddell JS, Sleeper LA, Ohye RG, Williams IA, Mahony L, Pizarro C, et al. Intermediate-term mortality and cardiac transplantation in infants with single-ventricle lesions: risk factors and their interaction with

shunt type. J Thorac Cardiovasc Surg. 2012;144(1):152–9. PubMed PMID: 22341427. Pubmed Central PMCID: 3359406. Epub 2012/02/22. eng.
20. Newburger JW, Sleeper LA, Bellinger DC, Goldberg CS, Tabbutt S, Lu M, et al. Early developmental outcome in children with hypoplastic left heart syndrome and related anomalies: the single ventricle reconstruction trial. Circulation. 2012;125(17):2081–91. PubMed PMID: 22456475. Pubmed Central PMCID: 3341507. Epub 2012/03/30. eng.
21. Habashi JP, Judge DP, Holm TM, Cohn RD, Loeys BL, Cooper TK, et al. Losartan, an AT1 antagonist, prevents aortic aneurysm in a mouse model of Marfan syndrome. Science. 2006;312(5770):117–21. PubMed PMID: 16601194. Pubmed Central PMCID: 1482474. Epub 2006/04/08. eng.
22. Lacro RV, Dietz HC, Wruck LM, Bradley TJ, Colan SD, Devereux RB, et al. Rationale and design of a randomized clinical trial of beta-blocker therapy (atenolol) versus angiotensin II receptor blocker therapy (losartan) in individuals with Marfan syndrome. Am Heart J. 2007;154(4):624–31. PubMed PMID: 17892982. Pubmed Central PMCID: 3042860. Epub 2007/09/26. eng.
23. Lacro RV, Guey LT, Dietz HC, Pearson GD, Yetman AT, Gelb BD, et al. Characteristics of children and young adults with Marfan syndrome and aortic root dilation in a randomized trial comparing atenolol and losartan therapy. Am Heart J. 2013;165(5):828–35.e3. PubMed PMID: 23622922. Epub 2013/04/30. eng.
24. Selamet Tierney ES, Levine JC, Chen S, Bradley TJ, Pearson GD, Colan SD, et al. Echocardiographic methods, quality review, and measurement accuracy in a randomized multicenter clinical trial of Marfan syndrome. J Am Soc Echocardiogr. 2013;26(6):657–66. PubMed PMID:23582510. Epub 2013/04/10. eng.
25. Zhang B, Schmidt B. Do we measure the right end points? A systematic review of primary outcomes in recent neonatal randomized clinical trials. J Pediatr. 2001;138(1):76–80. PubMed PMID: 11148516. Epub 2001/01/10. eng.
26. Silber JH. Challenges in conducting a pediatric longitudinal prevention study: lessons from the ACE-inhibitor after anthracycline trial. Prog Pediatr Cardiol. 2001;20:65–70.
27. Sinaiko AR, Lauer RM, Sanders SP. End points for cardiovascular drug trials in pediatric patients. Am Heart J. 2001;142(2):229–32. PubMed PMID: 11479459. Epub 2001/08/02. eng.
28. Bucher HC, Guyatt GH, Cook DJ, Holbrook A, McAlister FA. Users' guides to the medical literature: XIX. Applying clinical trial results. A. How to use an article measuring the effect of an intervention on surrogate end points. Evidence-Based Medicine Working Group. JAMA. 1999;282(8):771–8. PubMed PMID: 10463714. Epub 1999/08/27. eng.
29. DeMets DL, Califf RM. Lessons learned from recent cardiovascular clinical trials: part II. Circulation. 2002;106(7):880–6. PubMed PMID: 12176964. Epub 2002/08/15. eng.
30. Strand M, Jobe AH. The multiple negative randomized controlled trials in perinatology – why? Semin Perinatol. 2003;27(4):343–50. PubMed PMID: 14510325. Epub 2003/09/27. eng.
31. Li JS, Colan SD, Sleeper LA, Newburger JW, Pemberton VL, Atz AM, et al. Lessons learned from a pediatric clinical trial: the Pediatric Heart Network angiotensin-converting enzyme inhibition in mitral regurgitation study. Am Heart J. 2011;161(2):233–40. PubMed PMID: 21315203. Pubmed Central PMCID: 3053082. Epub 2011/02/15. eng.
32. Pike NA, Pemberton V, Allen K, Jacobs JP, Hsu DT, Lewis AB, et al. Challenges and successes of recruitment in the "angiotensin-converting enzyme inhibition in infants with single ventricle trial" of the Pediatric Heart Network. Cardiol Young. 2013;23(2):248–57. PubMed PMID: 22931751. Pubmed Central PMCID: 3563771. Epub 2012 Jul 5. Eng.
33. Chalmers TC. Randomization of the first patient. Med Clin North Am. 1975;59(4):1035–8. PubMed PMID: 1142862. Pubmed Central PMCID: 1142862.
34. National Heart, Lung, and Blood Institute. Children and clinical studies. Available from: http://www.ChildrenandClinicalStudies.nhlbi.nih.gov. Accessed 5 May 2013.

34 The Value of National Institutes of Health (NIH) Registry-Based Research in Identifying Childhood Cardiac Disease Outcomes: The Pediatric Cardiomyopathy Registry Experience

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Abstract

Cardiomyopathy is a serious, but rare, disease of the heart muscle. Cardiomyopathy commonly results in heart failure and is the leading cause of heart transplantation in children older than 1 year of age. The Pediatric Cardiomyopathy Registry (PCMR), which has been funded by the National Heart Lung and Blood Institute (NHBLI) since 1994, has enrolled and followed more than 3,500 children with cardiomyopathy in the US and Canada. Results from the registry have established the incidence of pediatric cardiomyopathy in North America, determined the prevalence of heart failure, have identified causes of cardiomyopathy, as well as transplant and survival patterns. In this chapter, we will describe the design and operation of the PCMR and present a summary of results. The PCMR is an example

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of how a well-designed and well-conducted patient registry can provide important insights into the etiologies, clinical course, and patient outcomes for a rare disease, in this case cardiomyopathy in children.

Keywords
Cardiomyopathy • Pediatrics • Patient registry • Heart failure • Pediatric cardiomyopathy registry

Introduction

Patient registries are important public health tools for disease surveillance as well as an important observational research design to address important research questions, such as disease outcomes, which cannot always be definitively addressed by clinical trials. The Agency for Healthcare Research and Quality (AHRQ) defines a patient registry as "an organized system that uses observational data (clinical and other) to evaluate specific outcomes for a population defined by a particular disease, condition, or exposure, and that serves one or more predetermined scientific, clinical, or policy purposes" [1]. In the AHRQ report, they suggest how to plan and design a patient registry, selection of data elements, data linking, ethical and privacy issues, data quality, registry data analysis and interpretation and other key issues important to establishing and maintaining robust patient registries [1]. These patient registries are particularly important in understanding rare diseases, which often disproportionally affect pediatric populations [2, 3]. Over 7,000 rare diseases, defined as affecting less than 200,000 US residents annually, affect almost 30 million persons in the US [3]. However, registries must collect high quality data and employ robust statistical analyses to result in the most informative results, while minimizing study bias which is intrinsic in observational studies. Dreyer and Garner state that patient registries are useful in understanding how clinical trial results can be applied in clinical practice as well as support regulatory agency decisions about product safety and indications as well as support insurance coverage for specific interventions [4]. They emphasize the importance of well-designed observational studies, including patient registries, as real-word research additions to the evidence-based understanding of a variety of health conditions. However, they caution that a clear understanding of registry methodologies and follow-up design are essential to evaluating the value of registry-based results, which echoes the AHRQ guidelines.

In this chapter, we present the results from the National Heart, Lung and Blood Institute (NHLBI)-funded Pediatric Cardiomyopathy Registry (PCMR) as an example of how a well-designed and well-conducted patient registry can provide important insights into the etiologies, clinical course, and patient outcomes for a rare disease, in this case cardiomyopathy in children. Combining robust registry data with the results of current and future clinical trials, often initiated based on patient registry observational results, can lead to important clinical advances in improving patient outcomes for a variety of diseases.

The Significance of Pediatric Cardiomyopahty as a Rare Disease

Although relatively rare, cardiomyopathy is a common cause of heart failure in children and the leading cause of heart transplantation in those over the age of 1 year [5–8]. Among the functional types, dilated and hypertrophic cardiomyopathies are the most common in children. These two functional types are characterized by abnormal cardiac structure and function and a poor prognosis. Until the mid-1990s, little was known about its incidence, prevalence, risk factors, causes, and outcomes.

Given the disease's rarity and heterogeneity, accurately estimating its incidence required a large and relatively unbiased sample of the population that in turn required an extensive and stringent recruitment strategy. Long-term follow-up

was also needed to better document the diagnosis, treatment, course, and outcome of this varied disease, as well as to identify cause-specific prevention and treatment strategies.

To study the disease to this extent, in 1994, the NHLBI funded a large, multi-center observational study of both primary and idiopathic cardiomyopathies: the Pediatric Cardiomyopathy Registry. Currently the largest pediatric cardiomyopathy registry in the world, it contains data collected annually from more than 3,500 children until heart transplant, death, or loss to follow-up. The PCMR has provided the most complete picture of pediatric cardiomyopathy and its diagnosis and treatment, and, in so doing, has provided a solid foundation on which new treatment strategies can be based.

The PCMR has evolved in response to early findings. Originally, its purpose was to document the incidence, presentation, and functional types of the disease for specific demographic groups. With this purpose largely achieved, its focus is to better characterize the functional types, the causes, and risk factors for transplant and death. In this chapter, we discuss the nature, development, and contributions of the PCMR.

The Design and Operation of the Registry

The PCMR began with two cohorts [9]. The first was a retrospective cohort of children in whom cardiomyopathy had been diagnosed between January 1, 1990, and December 31, 1995. These children were identified by a chart review from 39 tertiary care centers in the US and Canada. The purpose of this cohort was to identify potential predictors of outcomes as well as diagnostic approaches.

The second cohort was a population-based, prospective cohort of children diagnosed after January 1, 1996, by pediatric cardiologists at 98 pediatric cardiac centers in two geographically distinct regions of the US (New England, consisting of Connecticut, Maine, Massachusetts, New Hampshire, and Rhode Island; and the Central Southwest, consisting of Arkansas, Oklahoma, and Texas). These geographic areas were selected because of the local referral patterns, which allowed for the identification of nearly all cases of pediatric cardiomyopathy from tertiary centers. The purpose of studying this cohort was to accurately estimate the incidence of pediatric cardiomyopathy. Data collection in both regions was standardized by an outreach team that regularly visited each participating center to enroll new cases and abstract data from medical records.

Children were eligible for inclusion if they were up to 18 years old, had a diagnosis of cardiomyopathy based on quantitative echocardiographic criteria; a pattern of cardiomyopathy matching a defined, semi-quantitative pattern; or tissue analysis confirming the diagnosis (Table 34.1). All cases were classified as one of four main disease types:

Table 34.1 Inclusion criteria for enrollment in the Pediatric Cardiomyopathy Registry

Echocardiographic measurements
Left ventricular fractional shortening or ejection fraction >2 SD below the normal mean for age. Fractional shortening is acceptable in patients with a normal ventricular configuration and no regional wall motion abnormalities. Echocardiographic, radionuclide or contrast angiographic, or MRI evidence of abnormal ejection fraction is also an acceptable criterion, but age-appropriate norms for each laboratory must be used
Left ventricular posterior wall thickness at end-diastole >2 SD above the normal mean for body-surface area
Left ventricular posterior wall thickness at end-diastole >2 SD below the normal mean for body-surface area
Left ventricular end-diastolic dimension or volume >2 SD above the normal mean for body-surface area. Dimension data are acceptable under the conditions outlined for fractional shortening above, and volume data may be derived from the imaging methods as above
Clinical patterns
Localized ventricular hypertrophy, such as septal thickness >1.5 × left ventricular posterior wall thickness with at least normal left ventricular posterior wall thickness, with or without dynamic outflow obstruction
Restrictive cardiomyopathy: one or both atria enlarged relative to ventricles of normal or small size with evidence of impaired diastolic filling and in the absence of marked valvular heart disease
Contracted form of endocardial fibroelastosis; similar to restrictive cardiomyopathy plus echo-dense endocardium
Ventricular dysplasia/Uhl's anomaly: very thin right ventricle with dilated right atrium (usually better assessed by MRI than by echocardiography)
Concentric hypertrophy in the absence of a hemodynamic cause: a single measurement criterion of LV posterior wall thickness at end-diastole >2 SD would suffice
Left ventricular myocardial noncompaction: very trabeculated spongiform left ventricle myocardium with multiple interstices

Table 34.2 Exclusion criteria for enrollment in the Pediatric Cardiomyopathy Registry

Endocrine disease known to cause heart muscle disease (including infants of diabetic mothers)
History of rheumatic fever
Toxic exposures known to cause heart muscle disease (e.g., anthracyclines, mediastinal radiation, iron overload, or heavy metal exposure)
HIV infection or born to an HIV positive mother
Kawasaki disease
Congenital heart defects unassociated with malformation syndromes (e.g., valvular heart disease or congenital coronary artery malformations)
Immunologic disease
Invasive cardiothoracic procedures or major surgery during the preceding month, except those specifically related to cardiomyopathy, including left ventricular assist devices, extracorporeal membrane oxygenation, and automatic implanted cardioverter defibrillator placement
Uremia, active or chronic
Abnormal ventricular size or function that can be attributed to intense physical training or chronic anemia
Chronic arrhythmia, unless the inclusion criteria were documented before the onset of the arrhythmia (except that a patient with chronic arrhythmia, subsequently ablated, whose cardiomyopathy persists after 2 months is not to be excluded)
Malignancy
Pulmonary parenchymal or vascular disease (e.g., cystic fibrosis, cor pulmonale, or pulmonary hypertension)
Ischemic coronary vascular disease
Age 18 years or younger
Association with drugs known to cause hypertrophy (e.g., growth hormone, corticosteroids or cocaine)

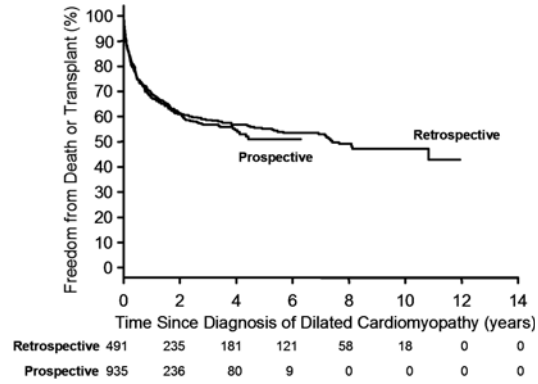

Fig. 34.1 Freedom from death or transplant for 1,426 children with dilated cardiomyopathy. Data are from 491 children in the retrospective cohort and 935 children in the prospective cohort of the Pediatric Cardiomyopathy Registry and were collected between 1990 and 2002. The groups did not differ on this endpoint (P = 0.71) (Reprinted from Towbin et al. [5]. Copyright © 2006 American Medical Association. All rights reserved)

dilated cardiomyopathy (DCM), hypertrophic cardiomyopathy (HCM), restrictive cardiomyopathy (RCM), mixed HCM and RCM, or idiopathic. Since the inception of the PCMR, more specific types have been added, such as HCM with left ventricular non-compaction (LVNC).

Exclusion criteria helped to narrow the focus of the registry (Table 34.2). For example, children were excluded if they had specific secondary causes of structural abnormalities, such as congenital heart defects, and exposure to cardiotoxic drugs, such as doxorubicin chemotherapy, which are known to cause cardiac hypertrophy. These exclusion criteria allowed for the examination of primary cardiomyopathies specifically.

Data collected from the prospective cohort included demographic information, quantitative echocardiographic measurements, a brief family history, transplant status, and clinical findings. Data from the retrospective cohort were more detailed and included a complete family history, qualitative echocardiographic measurements, electrocardiography data, and data on therapy and hospitalizations. To account for the range of body sizes and ages, echocardiographic measurements were transformed into body surface area or age-adjusted z-scores base upon a cohort of healthy children from Boston Children's Hospital. Clinical outcomes (Fig. 34.1), echocardiographic measurements, and clinical characteristics did not differ greatly between the two cohorts. Therefore, data from the cohorts were combined for most analyses.

From 2005 to 2010, 394 children were enrolled in a new prospective cohort from which blood and tissue samples were collected for analysis. The primary goal of studying this new cohort was to estimate associations between clinical outcomes and functional types of cardiomyopathy with physical and psychosocial functioning and genetic and viral status (Table 34.3). All newly enrolled children were tested for the

Table 34.3 Specific aims of the current PCMR study: 2005–2010

Specific aim 1	To integrate the PCMR and the Pediatric Heart Transplant Study Group (PHTS) databases to examine whether and how cardiac transplantation modifies the clinical course of cardiomyopathy in children
Specific aim 2	To establish the longitudinal course of functional status in children with cardiomyopathy and to determine the relationship of this course to clinical events and outcomes
Specific aim 3	To investigate how genetic and viral markers of cardiomyopathy are associated with clinical and functional outcomes

G4.5 mutation. The G4.5 gene encodes to the protein taffazin, and mutations of this gene are associated with Barth syndrome, a cause of dilated cardiomyopathy [10–12]. Also, in this cohort more detailed data on medications, echocardiography, and other cardiac studies were collected than ever before. From a subset of these children, biopsies were taken to assess the prevalence of viral cardiomyopathy using polymerase chain reaction.

The Epidemiology of Cardiomyopathy

The Incidence of Pediatric Cardiomyopathy

In the early stages of the PCMR, nearly 500 children were enrolled from the two major geographic regions. Lipshultz et al. estimated that fewer than five cases of cardiomyopathy were missed per year in these two regions [13]. The same study estimated the annual incidence of pediatric cardiomyopathy in the United States to be 1.13 cases per 100,000 children age birth to 18 years, an incidence similar to registry-based reports from Finland and Australia [14, 15].

The annual incidence was much higher in infants under the age of 1 year with 8.34 cases per 100,000 children. The incidence in boys was higher than that in girls (1.32 vs. 0.92 cases per 100,000; $P<0.001$), and the incidence in blacks was higher than that in whites (1.47 vs. 1.06 cases per 100,000; $P=0.02$). The incidence was also higher in the New England region than in the Central Southwest region (1.44 vs. 0.98 cases per 100,000; $P<0.001$). By type, DCM was the most common phenotype at 51 % of all cases, followed by: HCM (42 %), RCM and mixed HCM/RCM (3 %), and idiopathic (4 %). The same variations by sex, race, and geographic region reported above were seen in these functional types.

Unforeseen features in the original protocol may have resulted in underestimating the overall incidence of cardiomyopathy. Children presenting with sudden death may not have been properly identified because pathologist and medical examiner reports were not examined in the PCMR protocol through 2005. In addition, the PCMR definition of cardiomyopathy relies on clinically present disease. Therefore, children with asymptomatic left ventricular (LV) dysfunction may also not have been identified during this period.

Genetic and Viral Associations

The PCMR provides a unique opportunity to examine genetic causes of cardiomyopathy. Preliminary analyses found that variants of the G4.5 mutation of the taffazin (TAZ) gene were about equally prevalent in boys (25 %) and in girls (22 %) (Table 34.4) [16]. A variant was found in 7 % of males who were identified as having Barth syndrome. In children without a diagnosis of Barth syndrome, variants of uncertain importance were found in all four functional cardiomyopathy groups: HCM (20 %), DCM (24 %), RCM (30 %), and in children with mixed phenotypes (18 %).

Polymerase chain reaction analyses of myocardial tissue samples from 44 children in the PCMR were also examined. Two (4.5 %) children were positive for Epstein-Barr virus, and six (13.6 %) were positive for parvovirus. Tests for cytomegalovirus, adenovirus, and enterovirus, among other viruses, were negative.

Table 34.4 G4.5 gene variants found in 37 of 160 children enrolled in the Pediatric Cardiomyopathy Registry[a]

Sex	n/N (%)	Hemizygous SNP, n/N (%)	Intronic substitution, SNP, n/N (%)	Missense substitution, unclassified, n/N (%)	Hemizygous mutation[a], n/N (%)
Boys	27/110 (25)[b]	22/27 (81)	24/27 (89)	3/27 (11)	2/27 (7)
Girls	10/48 (22)[b]	3/10 (30)	10/10 (100)	0/10 (0)	0/10 (0)

Reprinted with permission from Towbin et al. [16]

[a]The number of children with G4.5 gene variants as a proportion of all children with the same diagnosis were: 9 of 45 (20 %) children with hypertrophic cardiomyopathy; 19 of 79 (24 %) with dilated cardiomyopathy; 3 of 10 (30 %) with restrictive cardiomyopathy; 4 of 22 (18 %) with other or mixed forms of cardiomyopathy; and 2 of 2 with unknown forms

[b]Causes of cardiomyopathy in boys included, two with Barth syndrome, two with probable myocarditis, one with Cori Disease, one with Noonan syndrome, one with familial DCM, and six with idiopathic disease; causes in girls included, two with familial HCM, two with confirmed myocarditis, and six with idiopathic disease. Children with Barth syndrome each had two variants, denoted here as hemizygous mutations (hemizygous SNPs were not counted)

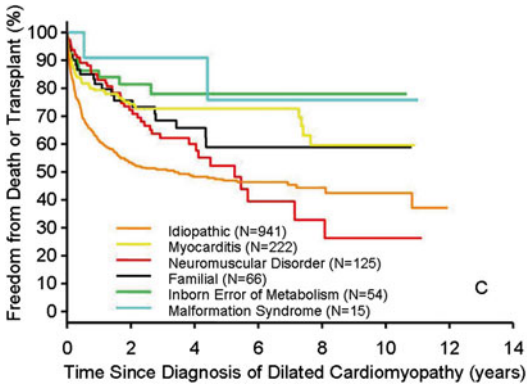

Fig. 34.2 Freedom from death or transplantation for 1,423 children with dilated cardiomyopathy, by cause. Data are from the Pediatric Cardiomyopathy Registry collected between 1990 and 2002 (Reprinted from Towbin et al. [5]. Copyright © 2006 American Medical Association. All rights reserved)

Outcomes of Cardiomyopathy by Phenotype

Analyses of the database have estimated the rates of cause-specific outcomes (death and heart transplantation) and of predictors for these outcomes.

Dilated Cardiomyopathy

In a study of more than 1,400 children with DCM, 1- and 5-year rates of death or heart transplantation were 31 and 46 %, respectively [5]. However, these rates varied widely by the cause of the disease (Fig. 34.2). Survival rates were better in children under the age of 6 years than in older children ($P<0.001$). After excluding children with neuromuscular disease or inborn errors of metabolism, congestive heart failure (CHF) at diagnosis and decreased left ventricular fractional shortening (LVFS) z-score predicted either death or transplant in children with idiopathic dilated cardiomyopathy with increased risk at older ages of diagnosis. However, an increased LV end-diastolic dimension (LVEDD) z-score was associated with transplantation but not mortality, whereas a lower height-for-age z-score was associated only with mortality for children with idiopathic dilated cardiomyopathy (IDCM) [17].

For children with DCM and neuromuscular disease, a lower LVFS z-score was associated with both death and transplant, whereas a higher LVEDD z-score was associated only with transplantation. Older age at diagnosis, CHF, and increased LVEDD z-score were associated with an increased risk of both death and transplantation for children with myocarditis, whereas the risk of both outcomes was increased equally for children with familial DCM who had CHF and lower LVFS z-scores. Thus, outcomes for children with DCM depend on an aggregate of age at diagnosis, cause, and heart failure at presentation. Although disease-specific therapies are available, most children present without an identifiable cause, which limits the therapies' usefulness.

Left ventricular dysfunction at presentation predicts death or transplantation in children with DCM, but the relationship between progressive LV dilation and wall thinning to death or transplant in this population was not well studied before the PCMR was established [18]. Analysis of data from the PCMR comparing LVFS, LVEDD, and LV

posterior wall thickness z-scores at baseline and up to 12 months later showed that children with DCM who survived 1 year had a higher baseline LVFS z-score and a lower baseline LVEDD z-score [18]. In addition, although baseline wall thickness did not differ between children who survived and children who died or received transplants, the LV posterior wall thickness-to-dimension ratio decreased among those who died or received a transplant and increased in those who survived 12 months. In Cox regression analysis, decreased thickness-to-dimension ratio predicted death or heart transplant after 12 months. In addition, a lower median LVFS z-score over time and an increase in LVEDD z-score during the first year after diagnosis predicted death or transplant in multivariate modeling. These parameters as well as increased wall thinning may be important predictors for poorer outcomes in children with DCM.

The outcomes of children with IDCM are often poor, yet some children show normalization of ventricular size and systolic function. In one study of the PCMR, echocardiographic characteristics returned to normal in 22 % of children with IDCM within 2 years of diagnosis [19]; median time to normalization was 9 months. Independent predictors of this recovery within 2 years were younger age and lower LV dilation at diagnosis. However, about 9 % of these children eventually died or underwent transplantation, indicating that follow-up of these children may be warranted.

Children with dilated cardiomyopathy are at a lower risk for sudden cardiac death (SCD) than are adults with non-ischemic, dilated cardiomyopathy. Only 13 % of these children in the PCMR experienced SCD [20]. Randomized trials have found that automatic implantable cardioverter-defibrillators improve survival in adults with non-ischemic cardiomyopathy [21–23]. However, there are no established criteria for using defibrillators in children with DCM to prevent sudden death. One analysis of the PCMR showed that the 5-year cumulative incidence of SCD in children with DCM was 2.4 % [20]. Children with CHF at diagnosis, as well as those taking anti-arrhythmic medications, were at higher risk for SCD. This analysis developed a risk stratification model to aid in identifying children with DCM at high risk for SCD. The model included an LV end-systolic dimension (LVESD) z-score greater than 2.6, age at diagnosis less than 14.3 years, and a LV posterior wall thickness to LVEDD ratio less than 0.14. The sensitivity and specificity of this model were 86 and 57 %, respectively. Children with DCM who consistently meet all these criteria should be considered for a defibrillator.

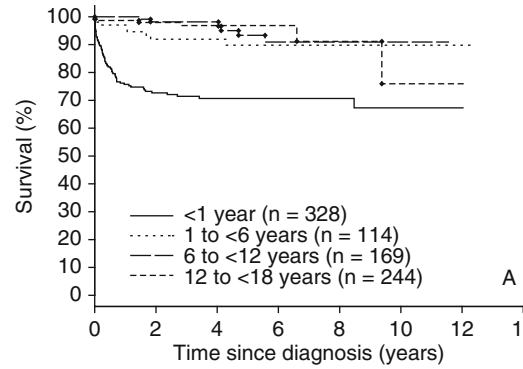

Fig. 34.3 Freedom from death or transplantation for 855 children with idiopathic hypertrophic cardiomyopathy, by age at diagnosis. Data are from the Pediatric Cardiomyopathy Registry collected between 1990 and 2002 (Reprinted with permission from Colan et al. [24])

Hypertrophic Cardiomyopathy

In children with HCM, data from the PCMR showed that outcomes varied greatly by age at diagnosis and cause (Figs. 34.3 and 34.4) [24]. The worst outcomes were in children who presented with inborn errors of metabolism, in whom the rate of death or transplant was 57 % at 2 years [25]. In addition, children with mixed types of cardiomyopathy also had poor outcomes, with rates of death or transplant at 2 years of 45 % for children with HCM and DCM and 38 % for children with HCM and RCM. The 2-year rate of death or transplant in children with HCM and a malformation syndrome was 23 %. Age at diagnosis was a risk factor among children with HCM; children diagnosed before age 1 year had a 2-year rate of death or transplant of 21 %. Children who survived to age 1 year had a 1 % mortality rate regardless of time of diagnosis.

Each of the cause-specific forms of pediatric HCM had unique risk factors for poor outcomes [25]. In general, these risk factors included

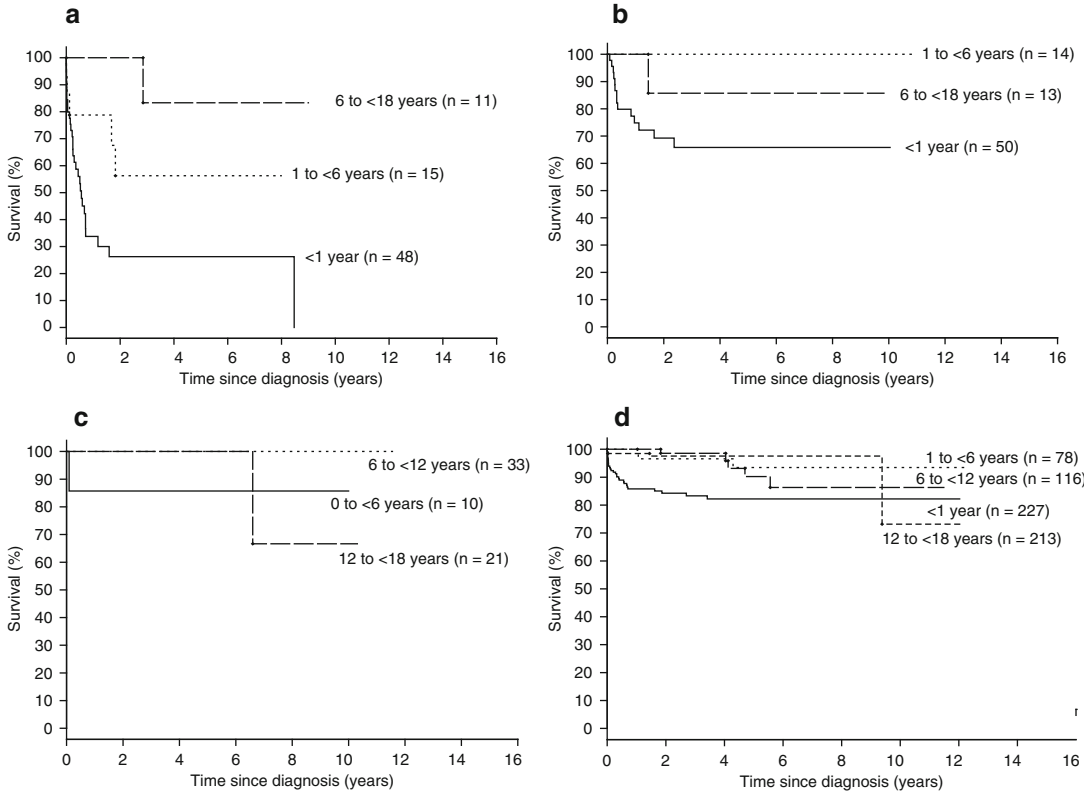

Fig. 34.4 Survival rates from the date of diagnosis of cardiomyopathy in children with (**a**) inborn errors of metabolism (N = 74, logrank P < 0.001); (**b**) malformation syndromes (N = 77, logrank P = 0.07); (**c**) neuromuscular disease (N = 64, logrank P = 0.22), and (**d**) idiopathic hypertrophic cardiomyopathy (N = 634, logrank P < 0.001), by age at diagnosis (Reprinted with permission from Colan et al. [24])

younger age at diagnosis, lower weight, presence of CHF at diagnosis, an increased LV posterior wall thickness z-score, and a decreased LVFS z-score. For all HCM subgroups by functional type and cause, the risk of death or transplantation was significantly increased by the presence of two or more of these risk factors. Further, the risk of a poor outcome also increased as the number of risk factors increased (Fig. 34.5) [25].

Restrictive Cardiomyopathy

Restrictive cardiomyopathy can occur in isolation or in combination with HCM. It is the least common form of cardiomyopathy, comprising only 4.5 % of all cardiomyopathies in the PCMR [26]. Cases of mixed RCM-HCM comprised roughly 33 % of all restrictive cardiomyopathies in the PCMR, and a family history of cardiomyopathy was present in nearly 25 % of RCM cases.

The outcomes of this phenotype are the worst of all the pediatric cardiomyopathies, especially for children who have not received a heart transplant. Overall, the 5-year survival of patients with either pure RCM or the mixed RCM-HCM phenotype was 68 %. However, transplant-free survival is much lower; 5-year transplant-free survival was 28 % for RCM and 43 % for the mixed RCM-HCM phenotype. In addition, transplant-free survival was worsened by the presence of CHF, a decreased LVFS z-score, and an increased LV posterior wall thickness z-score.

Left Ventricular Non-compaction

Left ventricular non-compaction is rare in children, accounting for only 4.9 % of the cardiomyopathies recorded in the PCMR [27]. This phenotype is a clinically heterogeneous cardiomyopathy and, like RCM, can occur in association with other

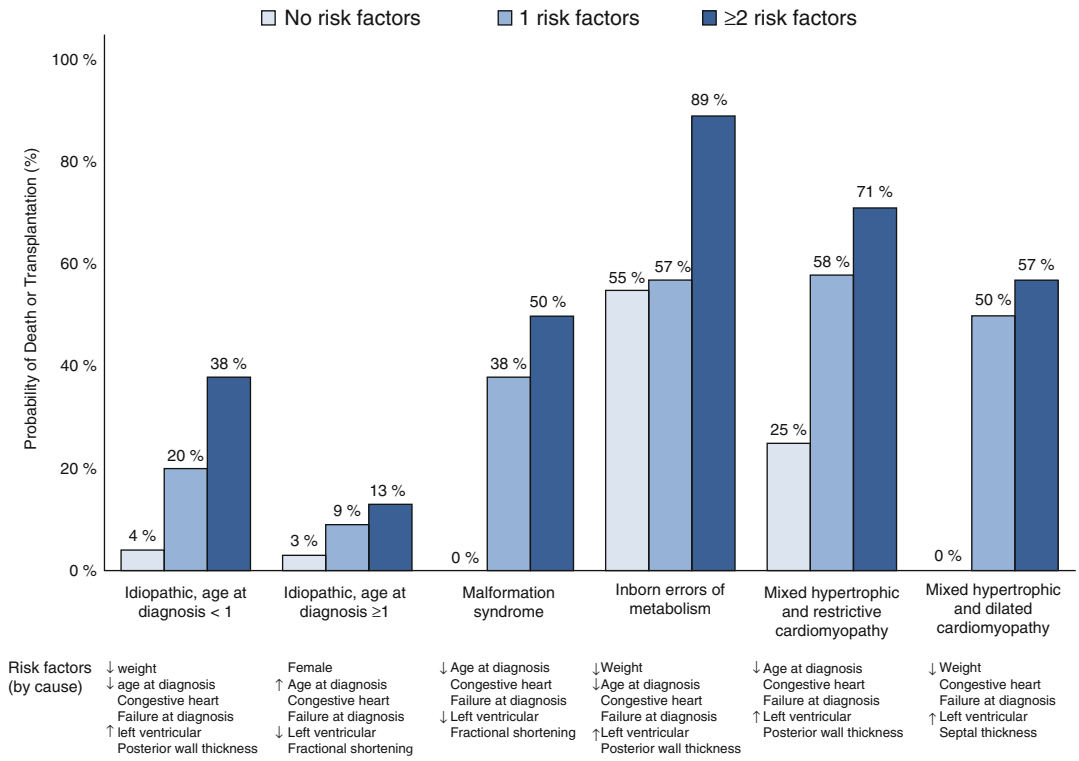

Fig. 34.5 Risk of death or transplant in 882 children with hypertrophic cardiomyopathy, by number of cause-specific risk factors. Data are from the Pediatric Cardiomyopathy Registry collected between 1990 and 2002 (Reprinted from Lipshultz et al. [25], copyright 2013, with permission from Elsevier)

phenotypes or in isolation. There is no clear consensus on the cause or diagnosis; however, echocardiographic findings are most widely accepted criteria for making the diagnosis. This disease is more often diagnosed in infancy and less likely to be idiopathic than are other cardiomyopathy phenotypes. Children in the PCMR with this phenotype and preserved systolic function have better 1- and 5-year outcomes than those of children with other cardiomyopathies, apart from HCM. However, a decreased LVFS z-score is a strong predictor for death or transplant.

Outcomes of Cardiomyopathy by Cause

The PCMR has also allowed the pediatric cardiomyopathies to be characterized by cause in addition to functional type. The cause is largely unknown for many cases of cardiomyopathy [5]. Children with a known cause of cardiomyopathy were believed to have characteristics that would distinguish them from children with idiopathic disease. Identifying the variables associated with a causal diagnosis could lead to improved diagnostic strategies and improved outcomes for these patients.

In one analysis of PCMR data, only one-third of cases had a cause identified at the time of diagnosis [28]. The proportion of cases without a known cause did not differ substantially between 1990 and 1999, indicating little improvement in the ability to establish a causal diagnosis during that time period. The rates of causal diagnoses also did not differ significantly between the four functional types of cardiomyopathy: HCM, DCM, RCM, and other/mixed cardiomyopathy. Children with a family history of cardiomyopathy were more likely to have a causal diagnosis assigned, regardless of functional type. In addition, a family history of sudden death or a genetic syndrome was also associated with having a

Table 34.5 Prevalence of congestive heart failure and median fractional shortening z-scores for 1,426 children with dilated cardiomyopathy at the time of diagnosis

Cause of DCM	Heart failure at diagnosis (%)	Fractional shortening z-score (IQR)
Idiopathic (N = 941)	74	−9.62 (−11.42 to −7.16)
Myocarditis (N = 222)	84	−9.11 (−11.05 to −6.67)
Neuromuscular disorders (N = 125)	35	−5.88 (−8.02 to −3.32)
Familial (N = 66)	53	−7.07 (−9.63 to −3.68)
Inborn errors of metabolism (N = 54)	60	−8.94 (−10.30 to −5.33)
Malformation syndromes (N = 15)	67	−5.95 (−9.49 to −5.10)

Data are from the Pediatric Cardiomyopathy Registry

Table 34.6 Prevalence of congestive heart failure and mean fractional shortening z-scores for 849 children with hypertrophic cardiomyopathy at the time of diagnosis

Cause of HCM	Heart failure at diagnosis (%)	Fractional shortening z-score (SD)
Inborn errors of metabolism (N = 74)	40.3	−1.11 (5.65)
Malformation syndromes (N = 77)	23.4	5.42 (4.31)
Neuromuscular disorders (N = 64)	6.4	3.01 (3.40)
Infantile (N = 634)	9.9	3.62 (5.15)

Data are from the Pediatric Cardiomyopathy Registry

known cause of cardiomyopathy for children with either HCM or DCM.

Certain patient characteristics were significantly associated with receiving a causal diagnosis, although these characteristics differed by type of cardiomyopathy [28]. For patients with HCM, female sex, lower height and weight for age, and increased left ventricular posterior wall thickness independently predicted receiving a causal diagnosis. For children with DCM, the rate of causal diagnosis did not differ by sex; however, children with a known cause of DCM were more likely to be older at diagnosis, to have smaller LV dimensions, and to have greater LVFS z-scores. Multivariate modeling found that children with HCM who had undergone metabolic blood and urine tests were four times more likely to have a causal diagnosis than children who did not undergo these tests. Furthermore, both endomyocardial biopsy and viral serology or culture independently predicted the establishment of a causal diagnosis for patients with DCM. This suggests that although available testing may currently be underused, increasing such testing may help identify a causal diagnosis in more patients.

Further examinations of PCMR data have confirmed that most cases of pediatric HCM and DCM lack a causal diagnosis [5, 24]. In one study, a known cause of disease was identified in only 34 % (N = 485) of children with DCM; the cause was myocarditis in 16 %, a neuromuscular disorder in 9 %, familial cardiomyopathy in 5 %, inborn errors of metabolism in 4 %, and malformation syndrome in 1 % [5]. In the same study, only 26 % (N = 215) of children with newly diagnosed HCM had a known cause of disease (9 % with a malformation syndrome, 9 % with inborn errors of metabolism, and 8 % with a neuromuscular disorder) [24]. For both groups, the prevalence of CHF varied greatly by type and cause of cardiomyopathy (Tables 34.5 and 34.6). At diagnosis, CHF was present in 71 % of children with DCM but in only 13 % of children with HCM [5, 24]. Children with DCM also presented with severely depressed left ventricular fractional shortening, regardless of cause (Table 34.5).

Neuromuscular disorders account for approximately 26 % of patients with a known cause of DCM within the PCMR [5]. Most children in the registry with neuromuscular disorders had one of two common types of muscular dystrophy, Duchenne (DMD) or Becker (BMD). Before the PCMR, the characteristics, development of cardiomyopathy, and outcomes had not been well studied in these patients [29]. Data from the PCMR were used to determine whether outcomes differed between children with either form of muscular dystrophy or from children with other

non-neuromuscular dilated cardiomyopathies (ODCM) [30]. All children with DMD and BMD included in the study had dilated cardiomyopathy, whereas the ODCM group included children with myocarditis and idiopathic dilated cardiomyopathy. Age at diagnosis of cardiomyopathy and rates of CHF at diagnosis were similar in children with DMD and BMD. Five-year survival was lower for children with DMD (57 %) than for those with BMD (100 %) or ODCM (71 %). However, children with BMD had a higher transplant rate than that of those with DMD.

Outcomes were also compared between children with ODCM and those with either type of neuromuscular disorder (BMD and DMD combined) [30]. Children in the combined DMD and BMD group had less LV dilation and higher LVFS z-scores at cardiomyopathy diagnosis than did those with ODCM [30]. Two years after cardiomyopathy diagnosis, LV dilation increased for children with DMD and BMD, but not for those with ODCM. In contrast, LVFS improved in the ODCM group during the same time period, but remained unchanged in the combined DMD and BMD group.

Although generally considered distinct diseases, myocarditis also often presents with DCM [31–33]. In one analysis of the PCMR, myocarditis was the leading cause of cardiomyopathy, accounting for 29.2 % of cases with a known cause [28]. For DCM in particular, myocarditis accounted for 47 % of cases with a known cause. Established histopathologic criteria were thought to have low sensitivity for the diagnosis of myocarditis [34]. However, an analysis of the PCMR found that time to death, transplantation, and echocardiographic normalization did not differ between children with biopsy-confirmed myocarditis ([BCM]; i.e., diagnosed clinically and with biopsy confirmation) and those with probable myocarditis ([PM]; i.e., clinical diagnosis of myocarditis alone or with inconclusive or nondiagnostic biopsy results) [35]. The proportion of death or transplant did not differ between children with BCM and PM 3 years after diagnosis, but were significantly lower than those for children with IDCM. During the same time period, nearly half of those with BCM or PM achieved echocardiographic normalization, and the proportion of echocardiographic normalization was significantly higher than for children with IDCM. These findings suggest better outcomes for children with myocarditis than those with IDCM. In children with myocarditis, lower LVFS z-scores at time of presentation predicted greater mortality, and greater left ventricular posterior wall thickness predicted transplantation.

Familial isolated cardiomyopathy, defined as having two or more family members with a history of cardiomyopathy, accounted for 24 % of cases with a known cause in the PCMR. DCM accounted for 34 % of these cases. Studies comparing outcomes in patients with familial dilated cardiomyopathy (FDCM) and outcomes in patients with IDCM have had conflicting results [5, 17]. With data from the PCMR, survival rates were compared between patients with FDCM and IDCM with no family history of cardiomyopathy. Freedom from death or transplantation was significantly higher in patients with FDCM (1-year survival, 87 %) than in patients with IDCM (1-year survival, 60 %). Patients with FDCM were less likely to present with heart failure and had higher ejection fractions, lower LV mass, LVESD, and LVEDD z-scores. Multivariate analyses found that LVEDD z-score at diagnosis independently predicted increased risk of death or transplant in patients with FDCM [36].

Mitochondrial disorders can present as either HCM or DCM in children [28]. Within the PCMR, only a few patients had undergone skeletal muscle biopsy to confirm mitochondrial disease [37]. In 40 % (N = 99) of these children, the mitochondrial disorder was diagnosed more than 30 days after presentation with cardiomyopathy. Nearly half of the patients had unspecified mitochondrial disease; the most common classification of mitochondrial disease was Barth syndrome (27 %). Mitochondrial disorders can occur with all functional types of cardiomyopathy. However, of children with both mitochondrial disease and cardiomyopathy, 49 % had DCM at diagnosis, whereas 26 % had HCM at diagnosis. Patients with HCM had the poorest survival rates (2-year survival, 51 %). Furthermore, a diagnosis of cardiomyopathy before 1 year of age was a significant risk factor for death in children with HCM and mitochondrial disease.

Hypertrophic cardiomyopathy is often present in patients with Noonan syndrome (NS) [38], the second most common syndromic cause of congestive heart disease [39]. The incidence of HCM in all NS cases is between 20 and 30 % [38, 40, 41]. Before the

PCMR, few studies compared outcomes of children with NS and HCM to those of children with other causes of HCM. Analysis of the PCMR revealed that 51 % of children with NS and HCM were diagnosed before age 6 months—a larger percentage than were children with other causes of HCM (28 %) [42]. Children with NS were also nearly three times as likely to present with CHF. Crude 3-year mortality rates for children with NS and HCM were higher than for those with other causes of HCM (26 % vs. 11 %). However, after adjusting for age at diagnosis and congestive heart failure, survival did not differ significantly between these two groups.

Within the NS cohort, the presence of CHF and age at diagnosis were important risk factors for mortality [42]. Survival was lower when CHF was present at cardiomyopathy diagnosis (1-year survival, 34 % vs. 90 %). Children with NS and HCM who presented before 6 months of age had a significantly higher rate of early mortality than that of those who presented after 6 months (1 year survival, 36 % vs. 96 %, respectively). Furthermore, for children who presented before 6 months of age and with CHF, 1-year survival was only 31 %. Other independent predictors of poor outcomes in patients with NS and HCM included a decreased LVFS z-score, and a lower height-for-age z-score. The risk profile of children presenting with HCM and NS, as well as the high mortality if diagnosed at an earlier age and when CHF is present, may warrant aggressive treatment and potential listing for transplantation.

Treating Pediatric Cardiomyopathy

Treatment for heart failure associated with cardiomyopathy includes pharmacologic interventions (such as anti-heart failure medications, angiotensin-converting enzyme inhibitors, and beta-blockers), mechanical support, and cardiac transplantation.

Medical Treatment of Pediatric Cardiomyopathy

Pharmacological treatments for children with IDCM and heart failure enrolled in the PCMR were examined over two time periods to identify treatment patterns. Treatments begun at diagnosis for children with IDCM diagnosed between 1990 and 1995 in the retrospective cohort were compared to those of similar children diagnosed between 2000 and 2006 in the prospective cohort [43]. Although clinical treatment of children with IDCM varies widely, treatments had changed little over the 16 years of this study. The use of anti-heart-failure therapy (with digoxin, a diuretic, or both) within 1 month of diagnosis did not vary greatly between the two time periods (84 % in the retrospective cohort and 87 % in the prospective cohort). Similarly, the percentage of children started on an angiotensin converting enzyme inhibitor (ACE-I) was similar between the two cohorts (66 and 70 %, respectively). Conversely, beta-blocker use increased from 4 to 18 % over the two time periods. Anti-heart-failure and ACE-I therapy were most often given to children with worsening LV dilation and LVFS. Current guidelines recommend treating nearly all heart failure patients with ACE-I therapy; anti-heart-failure therapy is recommended solely for children with symptomatic heart failure [44]. Despite these recommendations, only 47 % of patients with asymptomatic left ventricular dysfunction were started on ACE-1 therapy, although 60 % of these patients were treated with anti-heart-failure therapy.

Surgical Treatment of Pediatric Cardiomyopathy

Children with a diagnosis of cardiomyopathy have limited surgical options. For most children with end-stage cardiomyopathy, heart transplantation remains the only option for long-term survival [45]. The Pediatric Heart Transplant Study (PHTS) Group collects data on children from the time they are listed for transplant. In an analysis of their database, 10-year survival after the time of listing for patients with cardiomyopathy was 66 % [46]. However, despite the relative success of surgery, mortality remained quite high in some groups of patients between listing and transplantation [45]. Mechanical support has increasingly been used to improve outcomes during

this waiting period [47]. The problem remains, however, that the availability of ventricular assist devices (VADs) for infants and small children is still quite limited [47].

The PCMR is limited in its ability to assess children after heart transplantation or until the time of death post-transplant. In an attempt to incorporate transplant-related outcomes, the PCMR data were merged with the PHTS data. This merge allowed us to follow patients from the time of diagnosis of cardiomyopathy until after transplant. These combined data were used to assess whether different levels of heart failure severity affected survival before and after transplant [48].

For children who did not require mechanical or circulatory support (severity score 0) at the time of listing, mortality primarily occurred after transplantation. However, these children often deteriorated after listing, and therefore required increased support. A history of cardiac surgery was significantly associated with increased severity in children with a severity score of 0 in the short term, whereas a lower LV mass z-score was associated with an increased risk of deterioration 2–5 months after listing. In children who required intravenous inotropic drugs (severity score 1), death occurred both before and after heart transplant with relatively equal frequency. In contrast, for children who required mechanical or circulatory support (severity score 2), death primarily occurred while waiting for transplantation. This finding, which is similar to that in adults [49–51], suggests that in children with cardiomyopathy and higher severity scores at listing, transplantation provides a survival advantage that is not observed in children who do not require the same level of support.

In the combined PHTS and PCMR dataset, most patients who received a heart transplant had DCM [52]. In a separate analysis of these patients [53], those listed for transplant had a median age at diagnosis of 3.4 years and a median age at transplantation of 4.4 years. Pre-transplant mortality was 11 %. Among all patients listed for heart transplant, 30 % were ventilator-dependent at listing. Mechanical ventilation, as well as older age at listing for children not on mechanical ventilation, was associated with mortality while waiting for transplantation. Non-white race and lower LVEDD z-score were associated with poorer outcomes after transplant. Although most children with myocarditis do not require a heart transplant, the post-transplantation outcomes for those diagnosed with myocarditis were worse than for children without myocarditis, suggesting that infectious or immune mechanisms may affect graft survival and patient mortality [53].

Although heart transplantation was most common among patients with DCM in the combined dataset, only a proportion of these children are listed for transplant. Therefore, any factors that increase the risk of death in children with DCM after listing for heart transplant are of concern. In children diagnosed with DCM before 5 years of age, LV dilation at listing for heart transplant was significantly associated with a higher risk of death while waiting for transplant or within 6 months after receiving a transplant [54]. This association was strongest for children diagnosed with DCM before 6 months of age. Specifically, the risk of death while waiting for or within 6 months after transplant for infants less than 6 months of age was 5.8 times higher for every 1 cm increase in LVEDD at the time of listing. Additionally, the risk of death 6 months after listing for children less than 6 months of age with an LVEDD z-score above the median was 14 % higher than that for those with a z-score below median in the same age group. Thus, LV dilation (measured as LVEDD z-score) is a potential predictor of death for infants and young children who are listed for heart transplant.

Nutritional Status of Children with Cardiomyopathy

Growth retardation and malnutrition are significant clinical threats to children with cardiomyopathy. Children from the PCMR with IDCM diagnosed before 1 year of age were more likely to have lower heights, weights, and body mass indices (BMI) than were older children with the same type of cardiomyopathy [55]. In children diagnosed at an older age,

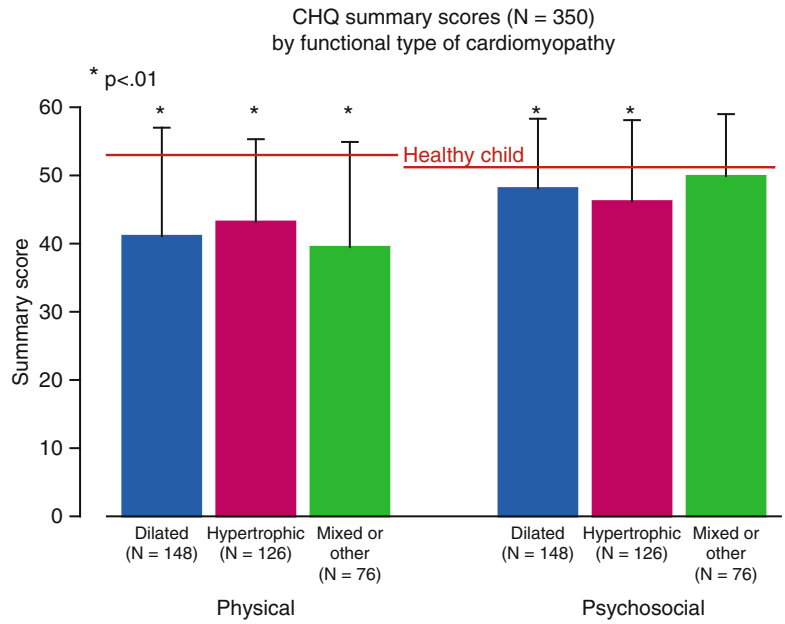

Fig. 34.6 Physical and psychosocial function of 294 children with cardiomyopathy, by functional type, compared to scores for healthy children. Data are from the Child Health Questionnaire (CHQ) administered by the Pediatric Cardiomyopathy Registry between 1990 and 2002

cardiac dysfunction was associated with lower height and lower BMI. Height was an independent predictor of death for all children with IDCM, regardless of age.

Functional Status of Children with Cardiomyopathy

Before the PCMR, there was little information on how functional status of children with cardiomyopathy compared to that of healthy children. We administered two validated surveys, the Child Health Questionnaire and the Functional Status II(R) Parent Reports Instrument, to children with cardiomyopathy at multiple PCMR centers, to obtain information on functional status. Compared to healthy children, children with cardiomyopathy were significantly impaired in physical functioning as well as in psychosocial functioning, although to a lesser extent (Figs. 34.6 and 34.7) [56]. Socioeconomic variables independently predicted impaired functional status. For example, higher total household income was independently associated with higher psychosocial functioning, whereas higher physical functioning was associated with having married parents and higher parental education level.

Impairments to functional status were also associated with cardiac size and function. Greater LV size was associated with poorer physical functioning in children with DCM, whereas the ratio of LV posterior wall thickness to end-diastolic dimension was inversely associated with physical functioning in children with HCM. Poorer functional status predicted death or transplant in children with DCM and mixed or other cardiomyopathy types, but not in children with HCM. Functional status was also positively associated with longer time since diagnosis, suggesting that many children may improve over time.

International Conferences on Pediatric Cardiomyopathy

The first International Workshop on Idiopathic and Primary Pediatric Cardiomyopathies was organized by the PCMR investigators in 2007. More than 30 participants attended the conference, which was co-sponsored by the Children's Cardiomyopathy Foundation and the NHLBI. The results of the conference were published in three issues of the journal *Progress in Pediatric Cardiology* [52, 57–92]. The Second International Conference on Cardiomyopathy in

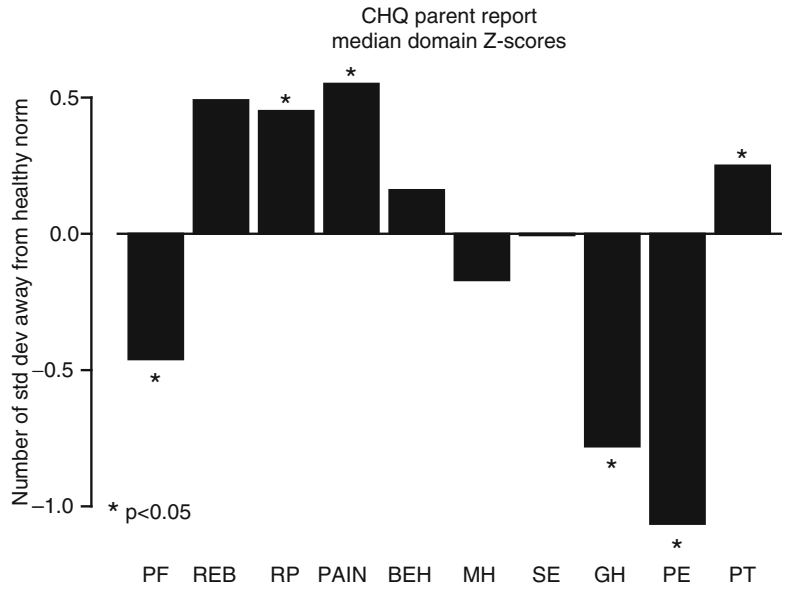

Fig. 34.7 Mean Child Health Questionnaire (CHQ) z-scores for 303 children with cardiomyopathy, by domain. Data are from the Pediatric Cardiomyopathy Registry and were collected between 1990 and 2002. *PF* physical functioning, *RE* role/social limits-emotional, *RP* role/social limits-physical, *PAIN* bodily pain, *BEH* behavior, *MH* mental health, *SE* self-esteem, *GH* general health perception, *PE* parental impact-emotional, *PT* parental impact-time

Children was held in May 2010, with more than 50 participants in attendance. Similar to the 2007 conference, the results from the second conference were published in three issues of *Progress in Pediatric Cardiology* [45, 93–123]. A third conference is currently scheduled for May 2014.

Future Directions of the Pediatric Cardiomyopathy Registry

By continuing to collect follow-up data from children enrolled in the PCMR, we can refine the description of pediatric cardiomyopathy and its clinical course in even more detail. These data will also allow us to examine risk factors more closely and to determine their long-term utility in diagnosis, prognosis, and treatment. The usefulness of registry data in guiding clinical decision making is increasingly appreciated by research methodologists and is being made more useful by advances in analytic and statistical theory [4]. In 2013, the PCMR investigators received two new awards from NHLBI which will use the PCMR research platform. The first study is enrolling 600 children with cardiomyopathy, and their parents, to have whole exome sequencing performed. One aim is gene discovery and the other is to establish genotype-phenotype associations. The phenotypes of interest are functional type of cardiomyopathy, age at presentation, and survival patterns. The second study is currently enrolling 300–400 children with cardiomyopathy and is periodically testing blood samples for a large panel of cardiac biomarkers. The association of biomarker levels with the child's clinical status, their survival patterns, and results of cardiac imaging will be analyzed.

Conclusions

The PCMR—the largest pediatric cardiomyopathy registry in the world, contains clinically important data on more than 3,500 cases of pediatric cardiomyopathy. Important contributions have included refined estimates of the incidence and outcomes of the disease, identified risk factors and predictors of outcomes for children with several cause-specific forms of cardiomyopathy, identified factors associated with making a causal diagnosis, and descriptions of the clinical care being provided to children with dilated cardiomyopathy.

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References

1. Gliklich RE, Dreyer NA, editors. Registries for evaluating patient outcomes: a user's guide. 2nd ed. Rockville: Agency for Healthcare Research and Quality; 2010. AHRQ publication No. 10-EHC049.
2. Richesson R, Vehik K. Patient registries: utility, validity, and inference. In: Rare diseases epidemiology. Dordrecht: Springer; 2010. p. 87–104.
3. Griggs RC, Batshaw M, Dunkle M, et al. Clinical research for rare diseases: opportunities, challenges, and solutions. Mol Genet Metab. 2009;96:20–6.
4. Dreyer NA, Garner S. Registries for robust evidence. JAMA. 2009;302:790–1.
5. Towbin JA, Lowe AM, Colan SD, et al. Incidence, causes, and outcomes of dilated cardiomyopathy in children. JAMA. 2006;296:1867–76.
6. Webber SA. New-onset heart failure in children in the absence of structural congenital heart disease. Circulation. 2008;117:11–2.
7. Andrews RE, Fenton MJ, Ridout DA, Burch M. New-onset heart failure due to heart muscle disease in childhood: a prospective study in the United kingdom and Ireland. Circulation. 2008;117:79–84.
8. Massin MM, Astadicko I, Dessy H. Epidemiology of heart failure in a tertiary pediatric center. Clin Cardiol. 2008;31:388–91.
9. Grenier MA, Osganian SK, Cox GF, et al. Design and implementation of the North American Pediatric Cardiomyopathy Registry. Am Heart J. 2000;139:S86–95.
10. Bione S, D'Adamo P, Maestrini E, Gedeon AK, Bolhuis PA, Toniolo D. A novel X-linked gene, G4.5. is responsible for Barth syndrome. Nat Genet. 1996;12:385–9.
11. Schlame M, Towbin JA, Heerdt PM, Jehle R, DiMauro S, Blanck TJ. Deficiency of tetralinoleoyl-cardiolipin in Barth syndrome. Ann Neurol. 2002;51:634–7.
12. Kelley RI, Cheatham JP, Clark BJ, et al. X-linked dilated cardiomyopathy with neutropenia, growth retardation, and 3-methylglutaconic aciduria. J Pediatr. 1991;119:738–47.
13. Lipshultz SE, Sleeper LA, Towbin JA, et al. The incidence of pediatric cardiomyopathy in two regions of the United States. N Engl J Med. 2003;348:1647–55.
14. Arola A, Jokinen E, Ruuskanen O, et al. Epidemiology of idiopathic cardiomyopathies in children and adolescents. A nationwide study in Finland. Am J Epidemiol. 1997;146:385–93.
15. Nugent AW, Daubeney PE, Chondros P, et al. The epidemiology of childhood cardiomyopathy in Australia. N Engl J Med. 2003;348:1639–46.
16. Towbin JA, Sleeper L, Jefferies JL, et al. Genetic and viral genome analysis of childhood cardiomyopathy: the PCMR/PCSR experience [abstract]. J Am Coll Cardiol. 2010;55:E409.
17. Alvarez JA, Orav EJ, Wilkinson JD, et al. Competing risks for death and cardiac transplantation in children with dilated cardiomyopathy: results from the pediatric cardiomyopathy registry. Circulation. 2011;124:814–23.
18. Kantor PF, Orav E, Wilkinson J, et al. Progressive left ventricular changes predict the likelihood of survival in pediatric dilated cardiomyopathy: findings from the Pediatric Cardiomyopathy Registry [abstract]. J Am Coll Cardiol. 2012;59:E740.
19. Everitt MD, Sleeper LA, Lu M, et al. Recovery of echocardiographic function in children with IDCM: results from the Pediatric Cardiomyopathy Registry. J Am Coll Cardiol. 2014;36:1405–13.
20. Pahl E, Sleeper LA, Canter CE, et al. Incidence of and risk factors for sudden cardiac death in children with dilated cardiomyopathy: a report from the Pediatric Cardiomyopathy Registry. J Am Coll Cardiol. 2012;59:607–15.
21. Kadish A, Dyer A, Daubert JP, et al. Prophylactic defibrillator implantation in patients with nonischemic dilated cardiomyopathy. N Engl J Med. 2004;350:2151–8.
22. Bansch D, Antz M, Boczor S, et al. Primary prevention of sudden cardiac death in idiopathic dilated cardiomyopathy: the Cardiomyopathy Trial (CAT). Circulation. 2002;105:1453–8.
23. Bardy GH, Lee KL, Mark DB, et al. Amiodarone or an implantable cardioverter-defibrillator for congestive heart failure. N Engl J Med. 2005;352:225–37.
24. Colan SD, Lipshultz SE, Lowe AM, et al. Epidemiology and cause-specific outcome of hypertrophic cardiomyopathy in children: findings from the Pediatric Cardiomyopathy Registry. Circulation. 2007;115:773–81.
25. Lipshultz SE, Orav EJ, Wilkinson JD, et al. Risk stratification at diagnosis for children with hypertrophic cardiomyopathy: an analysis of data from the Pediatric Cardiomyopathy Registry. Lancet. 2013;382:1889–97.

26. Webber SA, Lipshultz SE, Sleeper LA, et al. Outcomes of restrictive cardiomyopathy in childhood and the influence of phenotype: a report from the Pediatric Cardiomyopathy Registry. Circulation. 2012;126:1237–44.
27. Jefferies JL, Colan SD, Sleeper LA, et al. Outcomes and risk stratification for children with left ventricular noncompaction: findings from the pediatric cardiomyopathy registry [abstract]. Circulation. 2009;120:S794.
28. Cox GF, Sleeper LA, Lowe AM, et al. Factors associated with establishing a causal diagnosis for children with cardiomyopathy. Pediatrics. 2006;118:1519–31.
29. American Academy of Pediatrics Section on Cardiology and Cardiac Surgery. Cardiovascular health supervision for individuals affected by Duchenne or Becker muscular dystrophy. Pediatrics. 2005;116:1569–73.
30. Connuck DM, Sleeper LA, Colan SD, et al. Characteristics and outcomes of cardiomyopathy in children with Duchenne or Becker muscular dystrophy: a comparative study from the Pediatric Cardiomyopathy Registry. Am Heart J. 2008;155:998–1005.
31. Maron BJ, Towbin JA, Thiene G, et al. Contemporary definitions and classification of the cardiomyopathies: an American Heart Association Scientific Statement from the Council on Clinical Cardiology, Heart Failure and Transplantation Committee; Quality of Care and Outcomes Research and Functional Genomics and Translational Biology Interdisciplinary Working Groups; and Council on Epidemiology and Prevention. Circulation. 2006;113:1807–16.
32. Magnani JW, Dec GW. Myocarditis: current trends in diagnosis and treatment. Circulation. 2006;113:876–90.
33. Cooper Jr LT. Myocarditis. N Engl J Med. 2009;360:1526–38.
34. Baughman KL. Diagnosis of myocarditis: death of Dallas criteria. Circulation. 2006;113:593–5.
35. Foerster SR, Canter CE, Cinar A, et al. Ventricular remodeling and survival are more favorable for myocarditis than for idiopathic dilated cardiomyopathy in childhood: an outcomes study from the Pediatric Cardiomyopathy Registry. Circ Heart Fail. 2010;3:689–97.
36. Rusconi P, Wilkinson JD, Sleeper LA, et al. Outcomes in children with familial dilated cardiomyopathy compared to children with idiopathic dilated cardiomyopathy [abstract]. Circulation. 2010;122, A16092.
37. Cox GF, Colan SD, Kantor P, et al. Mitochondrial disorders: characteristics and outcomes from the pediatric cardiomyopathy registry [abstract]. In: Proceedings of the 5th World Congress of Pediatric Cardiology and Cardiac Surgery. 2009;5:134.
38. Roberts AE, Allanson JE, Tartaglia M, Gelb BD. Noonan syndrome. Lancet. 2013;381:333–42.
39. Marino B, Digilio MC, Toscano A, Giannotti A, Dallapiccola B. Congenital heart diseases in children with Noonan syndrome: an expanded cardiac spectrum with high prevalence of atrioventricular canal. J Pediatr. 1999;135:703–6.
40. Noonan J, O'Connor W. Noonan syndrome: a clinical description emphasizing the cardiac findings. Acta Paediatr Jpn. 1996;38:76–83.
41. Romano AA, Allanson JE, Dahlgren J, et al. Noonan syndrome: clinical features, diagnosis, and management guidelines. Pediatrics. 2010;126:746–59.
42. Wilkinson JD, Lowe AM, Salbert BA, et al. Outcomes in children with Noonan syndrome and hypertrophic cardiomyopathy: a study from the Pediatric Cardiomyopathy Registry. Am Heart J. 2012;164:442–8.
43. Harmon WG, Sleeper LA, Cuniberti L, et al. Treating children with idiopathic dilated cardiomyopathy (from the Pediatric Cardiomyopathy Registry). Am J Cardiol. 2009;104:281–6.
44. Rosenthal D, Chrisant MR, Edens E, et al. International society for heart and lung transplantation: practice guidelines for management of heart failure in children. J Heart Lung Transplant. 2004;23:1313–33.
45. Kirklin JK, Pearce FB, McGiffin DC, Dabal R. Surgical therapies for advanced heart failure in pediatric patients with cardiomyopathy. Prog Pediatr Cardiol. 2011;31:3–6.
46. Zangwill SD, Naftel D, L'Ecuyer T, et al. Outcomes of children with restrictive cardiomyopathy listed for heart transplant: a multi-institutional study. J Heart Lung Transplant. 2009;28:1335–40.
47. Blume ED, Naftel DC, Bastardi HJ, Duncan BW, Kirklin JK, Webber SA. Outcomes of children bridged to heart transplantation with ventricular assist devices: a multi-institutional study. Circulation. 2006;113:2313–9.
48. Larsen RL, Canter CE, Naftel DC, et al. The impact of heart failure severity at time of listing for cardiac transplantation on survival in pediatric cardiomyopathy. J Heart Lung Transplant. 2011;30:755–60.
49. Lietz K, Miller LW. Improved survival of patients with end-stage heart failure listed for heart transplantation: analysis of organ procurement and transplantation network/U.S. United Network of Organ Sharing data, 1990 to 2005. J Am Coll Cardiol. 2007;50:1282–90.
50. Jimenez J, Bennett Edwards L, Higgins R, Bauerlein J, Pham S, Mallon S. Should stable UNOS Status 2 patients be transplanted? J Heart Lung Transplant. 2005;24:178–83.
51. Krakauer H, Lin MJ, Bailey RC. Projected survival benefit as criterion for listing and organ allocation in heart transplantation. J Heart Lung Transplant. 2005;24:680–9.
52. Canter CE, Kantor PF. Heart transplant for pediatric cardiomyopathy. Prog Pediatr Cardiol. 2007;23:67–72.
53. Pietra BA, Kantor PF, Bartlett HL, et al. Early predictors of survival to and after heart transplantation in children with dilated cardiomyopathy. Circulation. 2012;126:1079–86.
54. Singh TP, Sleeper LA, Lipshultz S, et al. Association of left ventricular dilation at listing for heart transplant with postlisting and early posttransplant mortality in

children with dilated cardiomyopathy. Circ Heart Fail. 2009;2:591–8.
55. Miller TL, Orav EJ, Wilkinson JD, et al. Nutritional status is associated with cardiac outcomes and mortality in children with idiopathic dilated cardiomyopathy [abstract]. Circulation. 2009;120:S861.
56. Sleeper LA, Colan SD, Towbin JA, et al. Functional status is impaired and correlated with clinical status in pediatric cardiomyopathy [abstract]. In: Proceedings of the 5th World Congress of Pediatric Cardiology and Cardiac Surgery. 2009;5:134.
57. Lipshultz SE, Colan SD, Towbin JA, Wilkinson JD. Introduction for "idiopathic and primary cardiomyopathy in children". Prog Pediatr Cardiol. 2007;23:3.
58. Colan SD. Classification of the cardiomyopathies. Prog Pediatr Cardiol. 2007;23:5–15.
59. Weintraub RG, Nugent AW, Daubeney PEF. Pediatric cardiomyopathy: the Australian experience. Prog Pediatr Cardiol. 2007;23:17–24.
60. Alvarez JA, Wilkinson JD, Lipshultz SE. Outcome predictors for pediatric dilated cardiomyopathy: a systematic review. Prog Pediatr Cardiol. 2007;23:25–32.
61. Chung WK. Predictive genetic testing for cardiomyopathies. Prog Pediatr Cardiol. 2007;23:33–8.
62. Kishnani PS, Burns Wechsler S, Li JS. Enzyme-deficiency metabolic cardiomyopathies and the role of enzyme replacement therapy. Prog Pediatr Cardiol. 2007;23:39–48.
63. Rodrigues CO, Shehadeh LA, Webster KA, Bishopric NH. Myocyte deficiency as a target in the treatment of cardiomyopathy. Prog Pediatr Cardiol. 2007;23:49–59.
64. Jefferies JL. Novel medical therapies for pediatric heart failure. Prog Pediatr Cardiol. 2007;23:61–6.
65. Hsu DT. Age-related factors in child heart transplants. Prog Pediatr Cardiol. 2007;23:73–9.
66. Lipshultz SE, Colan SD, Towbin JA, Wilkinson JD. Idiopathic and primary cardiomyopathies in children. Prog Pediatr Cardiol. 2007;24:1.
67. Mestroni L, Miyamoto SD, Taylor MRG. Genetics of dilated cardiomyopathy conduction disease. Prog Pediatr Cardiol. 2007;24:3–13.
68. Cox GF. Diagnostic approaches to pediatric cardiomyopathy of metabolic genetic etiologies and their relation to therapy. Prog Pediatr Cardiol. 2007;24: 15–25.
69. Sheikh F, Chen J. Mouse models for cardiomyopathy research. Prog Pediatr Cardiol. 2007;24:27–34.
70. Dellefave LM, McNally EM. Cardiomyopathy in neuromuscular disorders. Prog Pediatr Cardiol. 2007;24:35–46.
71. Cooper Jr LT. Giant cell myocarditis in children. Prog Pediatr Cardiol. 2007;24:47–9.
72. Kaufman BD, Shaddy RE. Beta-adrenergic receptor blockade and pediatric dilated cardiomyopathy. Prog Pediatr Cardiol. 2007;24:51–7.
73. Miller TL, Neri D, Extein J, Somarriba G, Strickman-Stein N. Nutrition in pediatric cardiomyopathy. Prog Pediatr Cardiol. 2007;24:59–71.
74. Alcalai R, Arad M, Depreux F, Wang L, Seidman JG, Seidman CE. Hypertrophy, electrical abnormalities, autophagic vacuoles accumulation and cardiac fibrosis in LAMP2 cardiomyopathy mouse model. Prog Pediatr Cardiol. 2007;24:73–4.
75. Joshi VA, Roberts AE, Kucherlapati RS. Noonan syndrome associated congenital hypertrophic cardiomyopathy and the role of sarcomere gene mutations. Prog Pediatr Cardiol. 2007;24:75–6.
76. Rossano JW, Dreyer WJ, Kim JJ, et al. Pre-transplant serum creatinine predicts long-term outcome in pediatric heart transplant patients. Prog Pediatr Cardiol. 2007;24:77–8.
77. Taylor MRG. When echocardiogram screening "is not enough". Prog Pediatr Cardiol. 2007;24:79–80.
78. Ratnasamy C, Kinnamon DD, Lipshultz SE, Rusconi PG. Associations between neurohormonal and inflammatory activation and heart failure in children. Prog Pediatr Cardiol. 2007;24:81–2.
79. Lipshultz SE, Colan SD, Towbin JA, Wilkinson JD. Introduction for "idiopathic and primary cardiomyopathy in children". Prog Pediatr Cardiol. 2008;25:1.
80. Towbin JA. Molecular mechanisms of pediatric cardiomyopathies and new targeted therapies. Prog Pediatr Cardiol. 2008;25:3–21.
81. Lipshultz SE, Wilkinson JD. Epidemiological and outcomes research in children with pediatric cardiomyopathy. Prog Pediatr Cardiol. 2008;25:23–5.
82. Colan SD. Clinical issues in the pediatric hypertrophic cardiomyopathies. Prog Pediatr Cardiol. 2008;25:27–9.
83. Wilkinson JD, Sleeper LA, Alvarez JA, Bublik N, Lipshultz SE. The Pediatric Cardiomyopathy Registry: 1995–2007. Prog Pediatr Cardiol. 2008;25:31–6.
84. Young K, Hare JM. Stem cells in cardiopulmonary development: implications for novel approaches to therapy for pediatric cardiopulmonary disease. Prog Pediatr Cardiol. 2008;25:37–49.
85. Negro A, Dodge-Kafka K, Kapiloff MS. Signalosomes as therapeutic targets. Prog Pediatr Cardiol. 2008;25:51–6.
86. Menon SC, Olson TM, Michels V. Genetics of familial dilated cardiomyopathy. Prog Pediatr Cardiol. 2008;25:57–67.
87. Hill KD, Rizwan H, Exil VJ. Pediatric cardiomyopathies related to fatty acid metabolism. Prog Pediatr Cardiol. 2008;25:69–78.
88. Fisher SD, Pearson GD. Peripartum cardiomyopathy: an update. Prog Pediatr Cardiol. 2008;25:79–84.
89. Webber SA. Primary restrictive cardiomyopathy in childhood. Prog Pediatr Cardiol. 2008;25:85–90.
90. Somarriba G, Extein J, Miller TL. Exercise rehabilitation in pediatric cardiomyopathy. Prog Pediatr Cardiol. 2008;25:91–102.
91. Sokol KC, Armstrong FD, Rosenkranz ER, et al. Ethical issues in children with cardiomyopathy: making sense of ethical challenges in the clinical setting. Prog Pediatr Cardiol. 2007;23:81–7.
92. Bublik N, Alvarez JA, Lipshultz SE. Pediatric cardiomyopathy as a chronic disease: a look at comprehensive care programs. Prog Pediatr Cardiol. 2008;25:103–11.

93. Bernstein D, Fajardo G, Zhao M. The role of β-adrenergic receptors in heart failure: differential regulation of cardiotoxicity and cardioprotection. Prog Pediatr Cardiol. 2011;31:35–8.
94. Colan SD. Treatment of hypertrophic cardiomyopathy in childhood. Prog Pediatr Cardiol. 2011;31:13–9.
95. Dadlani GH, Harmon WG, Perez-Colon E, Sokoloski MC, Wilmot I, Lipshultz SE. Diagnosis and screening of hypertrophic cardiomyopathy in children. Prog Pediatr Cardiol. 2011;31:21–7.
96. Frazier AH, Ramirez-Correa GA, Murphy AM. Molecular mechanisms of sarcomere dysfunction in dilated and hypertrophic cardiomyopathy. Prog Pediatr Cardiol. 2011;31:29–33.
97. Hlaing WM, Messiah SE, Lipshultz SE, Ludwig DA. Obesity and length of hospital stay in children: a retrospective review of Florida Agency for Health Care Administration data. Prog Pediatr Cardiol. 2011;31:67–72.
98. Kantor PF, Rusconi P. Biomarkers in pediatric heart failure: their role in diagnosis and evaluating disease progression. Prog Pediatr Cardiol. 2011;31:53–7.
99. Messiah SE, Miller TL, Lipshultz SE, Bandstra ES. Potential latent effects of prenatal cocaine exposure on growth and the risk of cardiovascular and metabolic disease in childhood. Prog Pediatr Cardiol. 2011;31:59–65.
100. Pincott ES, Burch M. New biomarkers in heart failure. Prog Pediatr Cardiol. 2011;31:49–52.
101. Rampersaud E, Siegfried JD, Norton N, Li D, Martin E, Hershberger RE. Rare variant mutations identified in pediatric patients with dilated cardiomyopathy. Prog Pediatr Cardiol. 2011;31:39–47.
102. Shaddy RE. Randomized clinical trials and the treatment of pediatric cardiomyopathy. Prog Pediatr Cardiol. 2011;31:7–11.
103. Chung WK. Novel gene discovery in pediatric cardiomyopathy. Prog Pediatr Cardiol. 2011;31:89–91.
104. Foerster SR, Canter CE. Contemporary etiology, outcomes, and therapy in pediatric myocarditis. Prog Pediatr Cardiol. 2011;31:123–8.
105. Ho CY. New paradigms in hypertrophic cardiomyopathy: insights from genetics. Prog Pediatr Cardiol. 2011;31:93–8.
106. Hollander SA, Rosenthal DN. Cardiac resynchronization therapy in pediatric heart failure. Prog Pediatr Cardiol. 2011;31:111–7.
107. Payne RM. The heart in Friedreich's ataxia: basic findings and clinical implications. Prog Pediatr Cardiol. 2011;31:103–9.
108. Peter AK, Cheng H, Ross RS, Knowlton KU, Chen J. The costamere bridges sarcomeres to the sarcolemma in striated muscle. Prog Pediatr Cardiol. 2011;31:83–8.
109. Porter Jr GA, Hom JR, Hoffman DL, Quintanilla RA, Bentley K, Sheu S-S. Bioenergetics, mitochondria, and cardiac myocyte differentiation. Prog Pediatr Cardiol. 2011;31:75–81.
110. Wang H, Xin B. Hypertrophic cardiomyopathy in the Amish community — what we may learn from it. Prog Pediatr Cardiol. 2011;31:129–34.
111. Ware SM. Genetic diagnosis in pediatric cardiomyopathy: clinical application and research perspectives. Prog Pediatr Cardiol. 2011;31:99–102.
112. Weintraub RG, Nugent AW, Davis A, King I, Bharucha T, Daubeney PEF. Presentation, echocardiographic findings and long-term outcomes in children with familial dilated cardiomyopathy. Prog Pediatr Cardiol. 2011;31:119–22.
113. Bernstein D, Webber S. New directions in basic research in hypertrophy and heart failure: relevance for pediatric cardiology. Prog Pediatr Cardiol. 2011;32:5–9.
114. Canter CE, Cunningham MW, Cooper LT. Recent clinical and translational research on pediatric myocarditis. Prog Pediatr Cardiol. 2011;32:15–8.
115. Chung W, Towbin J. Genetic issues in pediatric cardiomyopathy: future research directions. Prog Pediatr Cardiol. 2011;32:3–4.
116. Conway J, Dipchand AI. Transplantation and pediatric cardiomyopathies: indications for listing and risk factors for death while waiting. Prog Pediatr Cardiol. 2011;32:51–4.
117. Gambetta K, Tambur A, Pahl E. Immune monitoring of pediatric heart transplant recipients through serial donor specific antibody testing — an initial experience and review of the literature. Prog Pediatr Cardiol. 2011;32:43–9.
118. Kantor PF, Rusconi P, Lipshultz S, Mital S, Wilkinson JD, Burch M. Current applications and future needs for biomarkers in pediatric cardiomyopathy and heart failure: summary from the Second International Conference on Pediatric Cardiomyopathy. Prog Pediatr Cardiol. 2011;32:11–4.
119. Kindel SJ, Pahl E. Cardiac allograft vasculopathy in children — treatment challenges. Prog Pediatr Cardiol. 2011;32:37–42.
120. Mital S. Biomarkers of cardiac fibrosis: new insights. Prog Pediatr Cardiol. 2011;32:35–6.
121. Ricci M, Lincoln J. Molecular markers of cardiomyopathy in cyanotic pediatric heart disease. Prog Pediatr Cardiol. 2011;32:19–23.
122. Wilkinson JD, Diamond M, Miller TL. The promise of cardiovascular biomarkers in assessing children with cardiac disease and in predicting cardiovascular events in adults. Prog Pediatr Cardiol. 2011;32:25–34.
123. Wilkinson JD, Zebrowski JP, Hunter JA, et al. Assessing the global and regional impact of primary cardiomyopathies: the Global Burden of Diseases, Injuries and Risk Factors (GBD 2010) Study. Prog Pediatr Cardiol. 2011;32:55–63.

Part VIII

Public Reporting of Data

Public Reporting of Cardiac Data: Pros, Cons, and Lessons for the Future

35

Edward L. Hannan

Abstract

Beginning with the Health Care Administration mortality reports in the 1980s, public reporting of health outcomes has been controversial, and its impact has been widely debated. The first public release of provider performance using clinical registry data occurred in 1989 when the New York State Department of Health released hospital risk-adjusted mortality rates for coronary artery bypass graft (CABG) surgery. Shortly afterward Pennsylvania released similar data, and currently a few other states, the Centers for Medicare and Medicaid Services, and several other organizations and professional societies release provider outcomes to the public for a variety of medical conditions and procedures. This communication summarizes and critiques some important literature that assesses the impact of these public releases with regard to quality improvement activities resulting from public reporting, improvement in health outcomes, surgeon reactions to outlier status, avoidance of high-risk patients, ability to predict future performance, and market share. Also, the communication closes with a summary of the state of public reporting and prospects for the future.

Keywords

Public reporting • CABG surgery • Percutaneous coronary intervention • New York State cardiac registries • Risk-adjusted mortality

The first large-scale initiative for public reporting of hospital outcomes was the series of annual mortality reports released by the Health Care Financing Administration (HCFA, the predecessor of the Center for Medicare and Medicaid Services [CMS]). These mortality reports were released annually between 1986 and 1992 [1]. These studies used Medicare administrative data to assign medical and surgical patients into 17 groups (16 diagnostic categories and "all patients"). Each group was analyzed separately and risk-adjusted mortality rates were developed and released to the public for each hospital/group. The HCFA mor-

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tality reports were discontinued in 1992 in the face of continuing criticism regarding the use and limitations of administrative data, the grouping of patients, and several other criticisms/concerns.

In 1989, in the wake of criticism of the HCFA studies, the New York State Department of Health (DOH) and its Commissioner, Dr. David Axelrod, were becoming increasingly concerned about the variation in hospital mortality rates for coronary artery bypass graft surgery among the hospitals with Certificate of Need approval to perform the procedure. There was roughly a five-fold variation in the annual in-hospital mortality rates for the procedure, but the only information available was the number of cases and number of deaths, and high mortality rate hospitals' claim that they were treating the sickest patients could not be examined without more detailed information. Because of the reaction to HCFA's use of administrative data for evaluating hospital performance, DOH decided to create a patient-level clinical database that could be used to assess institutional outcomes for CABG surgery while taking into account inter-hospital differences in patient acuity. Later studies conducted in New York confirmed that administrative data and clinical data arrive at different assessments of hospital quality [2, 3].

The New York State Department of Health and its Cardiac Advisory Committee (CAC) used the current literature to identify patient risk factors that were related to short-term adverse outcomes for CABG surgery, and these risk factors were included in the data system along with demographics, complications of care, admission and discharge dates, procedures performed, and patient disposition at discharge. The new registry was used for the first time to assess hospital performance in a 1990 manuscript published in the *Journal of the American Medical Association (JAMA)* [4]. This study identified independent significant risk factors for CABG/valve surgery in-hospital mortality, as well as the observed, expected and risk-adjusted mortality rates and volumes for all of the 28 hospitals (numbered from 1 to 28 but not named) in the state that were approved through Certificate of Need to perform these procedures [4]. On the same day that the *JAMA* paper was published, the Department released the names of the hospitals along with their risk-adjusted mortality rates to the *New York Times* [5]. The first formal public report was issued in 1990, and the earliest one available on the web is for 1990–1992 data [6].

Similar concerns by other regions and groups led to the establishment of public reporting for CABG surgery in Pennsylvania (using clinically enhanced administrative data) shortly after the New York initiative [7]. The Veterans Administration, Northern New England, and the Society of Thoracic Surgeons also established registries for CABG surgery in the 1990s, although those registries were not used for public reporting. Later, New Jersey, California, and Massachusetts all developed CABG surgery registries and began publicly releasing hospitals' risk-adjusted mortality rates [8–10]. New York and New Jersey also release risk-adjusted rates for surgeons. New York and Massachusetts also release risk-adjusted mortality rates for percutaneous coronary interventions (PCI), and New York releases the same data for cardiac valve surgery. New York also releases physician-level data for PCI and valve surgery.

The following few sections describe and critique peer-reviewed studies that have assessed the impact of these public releases with regard to quality improvement activities resulting from public reporting, improvement in health outcomes, surgeon reactions to outlier status, avoidance of high-risk patients, ability to predict future performance, and market share. The communication closes with a summary of the state of public reporting and prospects for the future.

Hospital Quality Improvement Activities Triggered by Public Reporting

There have been several reported hospital-specific quality improvement initiatives related to the release of risk-adjusted outcomes information. All of these reports come from New York. St. Peter's Hospital in Albany. N.Y. was identified as having significantly higher than expected

mortality in the early years of the program (1991 and 1992). The excess mortality was found to be a result of emergency cases, for which St. Peter's experienced a 26 % mortality rate compared with a 7 % rate for the state. Other cases at St. Peter's had roughly the same mortality as the statewide rate. After a multidisciplinary review of care for emergency patients, St. Peter's found that the patients were not being sufficiently stabilized prior to surgery. This led to major changes in the care management for these patients, and consequently in 1993 there were no deaths in the 54 emergency patients who underwent CABG surgery [11].

Winthrop Hospital had one of the highest risk-adjusted mortality rates in the state in the first public report, and a site visit commissioned by the DOH led to probation for Winthrop's cardiac surgery program. As a result, the hospital hired a new chief of cardiac surgery, who made several changes, including concentrating the service on a single floor, hiring clinical nurse specialists and physician assistants who were dedicated to cardiac surgery, reviewing each case pre-operatively, and installing a dedicated cardiac anesthesia service [12]. Winthrop's risk-adjusted mortality rate fell considerably from 9.2 to 4.6 to 2.3 % between 1989 and 1991 [6].

Erie County Medical Center also experienced high-risk adjusted mortality in the early years of the program, and it had the highest risk-adjusted mortality in the state for the first 6 months of 1989. A site visit by the CAC resulted in several recommended changes and the hospital voluntarily suspended operations in 1990 to implement these recommendations. Some of the changes that were implemented included the establishment of a cardiac surgery quality assurance program, credentialing and ongoing evaluation of surgeon performance, dedicated cardiac anesthesiologists and cardiac intensive care beds, and the recruitment of a permanent, full-time service chief [12]. The new chief of cardiac surgery hired new operating room (OR) nurses, cardiopulmonary bypass technicians, and intensive care staff who were all dedicated to cardiothoracic surgery. The hospital's risk-adjusted mortality dropped from 7.31 % in 1989–1991 to 2.51 % in 1993–1995, slightly lower than the statewide average of 2.57 %. The annual volume of just over 100 cases rose to 219 cases per year in the 1996–1998 time period, and the mortality dropped further to 1.77 % [6, 12, 13].

Changes in Cardiac Outcomes

A few studies that have examined the change in short-term CABG surgery mortality following the public release of CABG surgery data. In the early days of New York's system, Hannan et al. found that the in-hospital mortality for CABG patients decreased from 3.52 % in 1989 to 2.78 % in 1992. After risk-adjustment to reflect differences across the years in patient severity of illness, this decrease was assessed to be 41 %, a decrease in risk-adjusted mortality from 4.17 % in 1989 to 2.45 % in 1992 [14].

However, since CABG surgery outcomes have improved throughout the country because of new techniques and processes of care, the fairest way to assess outcome changes in New York is to compare them to other regions for the same period of time. In this regard, Ghali et al. found that between 1990 and 1994 there were similar trends in mortality and mortality reduction in Massachusetts and Northern New England as there were in New York [15]. Peterson et al. used Medicare data between 1987 and 1992 (before and after initiation of the program) to examine the CABG mortality rate and changes in the rate [16]. Conclusions were that New York had the lowest risk-adjusted mortality rate of any state in 1992, and that the decrease in mortality between 1987 and 1992 was higher than for any other state with low mortality in the earlier period [16]. Peterson et al. also found that although there were similar trends in mortality reduction in Northern New England and Massachusetts, the mortality reduction was significantly lower in Massachusetts than in New York and Northern New England. The reductions in Northern New England are very like a result of the registry they developed and the extensive quality improvement initiatives undertaken there.

Later, a study by Hannan et al. compared in-hospital/30-day risk-adjusted mortality after CABG between 1994 and 1999 in states/regions of the country with public reporting and/or formal quality improvement programs with the mortality in the remainder of the United States [17]. The study found that the risk-adjusted odds for mortality in New York for the 1994–1999 time period was only 0.66, 95 % CI (0.57, 0.77) times the odds in the remainder of the country, meaning that after adjusting for patients' pre-procedural severity of illness, patients in the remainder of the country were 50 % more likely to experience short-term mortality [17]. Another finding was that the overall risk –adjusted mortality in Northern New England 1994–1999 was lower, but not significantly lower than the remainder of the country (adjusted odds ratio = 0.92 [95 % CI {0.076, 1.11}]).

In summary, there appears to be more evidence and more convincing evidence that public reporting has been associated with larger mortality reductions than in regions without public reporting. The following section reviews the evidence that this reduction has come at a high price: the avoidance of high-risk procedures by providers who are wary of their impact on public reports.

Avoidance of High-Risk Patients by not Providing Procedures or by Out-of-State Referrals

Several studies have concluded that an unintended consequence of public dissemination of cardiac data has been for providers to avoid high-risk patients by either refusing to recommend cardiac surgery or PCI, or by referring the patient out-of-state. First, surveys of surgeons and cardiologists indicate that they feel that avoidance of high-risk patients is a problem. Burack et al. reported that 67 % of New York surgeons claimed they had refused to treat at least one patient in the previous year and 18 % refused to treat five or more patients [18]. Narins et al. found that 83 % of interventional cardiologists in New York "agreed" or "strongly agreed" that the publication of statewide PCI report cards decreased the chance that patients needing PCI actually received it [19]. Also, Schneider and Epstein conducted a survey that found that 59 % of cardiologists had more difficulty referring high-risk patients and 63 % of cardiac surgeons were less willing to operate on high-risk patients [20].

An early empirical study on avoidance of high-risk procedures by Omoigui et al., compared the number and acuity level of New York patients undergoing CABG surgery from 1980 to 1988 in the Cleveland Clinic prior to the inauguration of the New York CABG registry with similar patients in the 1989–1993 time frame [15]. Findings of the study were that there were an average of 61.4 patients New York patients per year undergoing CABG surgery at the Cleveland Clinic in the first time period, and 96.2 per year after the founding of the New York registry. Also, New York patients treated at the Cleveland Clinic were of higher risk than New York patients treated in New York [21].

Another study, by Moscucci et al., compared the pre-procedural severity of illness of New York PCI patients with PCI patients in eight Michigan hospitals in 1998–1999 [22]. New York patients were found to have significantly lower prevalences of acute myocardial infarction, cardiogenic shock, and congestive heart failure than the Michigan patients. Even though the unadjusted in-hospital mortality was significantly lower in New York than in Michigan, the risk-adjusted mortality rates in the two regions were not significantly different. Moscucci et al. concluded that New York cardiologists may not be intervening as much on high-risk patients because of their fear of public reporting [22].

A study by Dranove et al. used Medicare data to conclude that the severity of illness (measured by a proxy of costs in the year preceding hospitalization) of CABG patients vs. acute myocardial infarction (AMI) patients in New York and Pennsylvania hospitals declined between 1987 and 1994 compared to the change in states without public reporting [23].

It should be noted that there appear to be flaws in each of these empirical studies. With regard to the Omoigui study, the first public

release of New York CABG surgery data occurred in 1990 and it was not announced in advance, so the earliest that hospitals would have been tempted to refer out-of-state for fear of adverse publicity would have been in 1991, 2 years after the beginning of the "after" time period in the study. The expected mortality of the patients referred from New York, although higher than that of other Cleveland Clinic patients changed little between the 1989–1990 period and the 1991–1993 period [12, 21].

Also, in the study by Hannan et al. that compared Medicare patients undergoing CABG surgery in regions with public dissemination of outcomes and formal quality improvement programs with the remainder of the United States (US), it was found that out-of-state referrals in New York were lower in 1994 and in 1999 than the remainder of the US (9.9 % vs. 10.4 %, and 10.4 % vs. 10.5 %, respectively), so there does not appear to be any widespread outmigration in New York in comparison with states without public data releases [17].

There are also some reasons why Moscucci et al.'s conclusions may be misleading. First, although the authors mention that some risk factor definitions in the two systems are nearly identical, they do not mention definitions of all risk factors. For instance, shock is defined in the New York registry as systolic blood pressure below 80 or cardiac index below 2.0 at the time of the procedure despite pharmacologic or mechanical support. Since most definitions of shock do not require low blood pressure or low output even after treatment, it is not clear that this was the Michigan definition. Second, the prevalences of high-risk conditions are calculated by dividing the number of patients with that condition undergoing PCI by the total number of patients undergoing PCI. The number could be lower in New York because there are more low-risk patients per capita undergoing PCI instead of that there are fewer high-risk patients per capita undergoing PCI.

The measure used in the Dranove study is not straightforward and is not necessarily the best measure to use. Purportedly this unconventional measure of patient severity was used because it is not subject to surgeon manipulation, is not controlled for in the risk-adjustment methodology, and could be used by surgeons to identify lower-risk patients. This assumes the unlikely scenario that surgeons would take the time to identify patients with lower hospital utilization who appeared not to have a commensurately low severity of illness based on measures that are used in the risk-adjustment methodology [23, 24]. In a more direct and straightforward study of Medicare patients undergoing CABG surgery between 1994 and 1999 in states/regions with public reporting/quality improvement efforts, and the remainder of the United States by Hannan et al., New York CABG patients had significantly higher prevalences of acute myocardial infarction, patients 80 or more years old, emergency admissions, and females than the remainder of the country [17]. Also, Peterson et al. reported that New York Medicare CABG patients had comparable prevalences of AMI, congestive heart failure (CHF), diabetes, and peripheral vascular disease as other United States Medicare CABG patients between 1987 and 1992, and New York patients had higher rates of AMI, age >80, and females in 1992 than in 1989 [16]. The risk factors in these studies were coded by hospital personnel who do not work in the cardiac surgery departments, and are therefore not as susceptible to manipulation or subtle over-coding of risks that could bias risk assessment in registry data.

It is also important to note that in some instances the avoidance of high-risk procedures may be beneficial to patients. This includes instances in which patients are denied access by hospitals/surgeons who are not experienced or skilled enough to perform a procedure, but who refer the patient to another provider. Another example is a patient who has no chance of survival or who is terminally ill with another disease [25]. Regardless of what appears to be weak evidence of widespread avoidance of high-risk cases related to public reporting, the reports of individual clinicians that they and/or their colleagues have denied surgery to high-risk patients should be taken seriously. This doesn't mean throwing out the baby with the bathwater and scrapping all public reporting. However, there are

measures that can be considered to reduce incentives for risk aversion. One of the most effective of these measures is to be responsive to clinicians' concerns about the unfairness of including certain high-risk patients in the public reporting process. In New York, cardiologists and surgeons were particularly concerned about the inclusion of cardiogenic shock patients in the computation of risk-adjusted mortality rates. Following a recommendation by the state's Cardiac Advisory Committee, the Department of Health decided to exclude shock patients from public reporting starting with the 2006 annual report because of the concern that these patients were sometimes being refused revascularization when it would have been beneficial.

It was decided to continue to collect data on shock patients undergoing revascularization to assess the impact of the new policy. In 2005, when results for shock patients were publicly reported, 83 shock patients underwent PCI with an in-hospital/30-day mortality rate of 34 %. In 2006–2008, respective totals of 133, 146, and 138 shock patients underwent PCI with a combined mortality rate of 45 %. In conclusion, the policy of omitting shock patients from public reporting resulted in an average increase in number of shock patients undergoing PCI of 67 %/year in the next 3 years and the shock patients undergoing PCI were of higher risk on average. For CABG surgery, there were 32 shock patients in 2005, followed by 46, 41 and 43 in the next 3 years. However, the average mortality rate in the subsequent 3 years was 38 %, compared with a 22 % mortality rate in 2005.

In Massachusetts, a compassionate use variable (active cardiopulmonary resuscitation [CPR], comatose, or requiring a ventricular assist device) was added to their PCI risk-adjustment model, as opposed to exempting a group of high-risk patients from public reporting [25, 26].

In New York, a recent decision has been made to exclude from analysis and public reporting PCI patients who experience hypoxic brain injury and expire due to withdrawal of support subsequent to a documented pre-intervention AMI and cardiac arrest. This policy took effect with 2010 procedures, so the data are still being reviewed and the impact of this policy change is unknown at this time.

Other measures that have been recommended for reducing the incentives to avoid high-risk patients in a recent study on this topic are (1) the improvement of risk-adjustment methods and highlighting hospitals and physicians who undertake high-risk procures in appropriate patients, (2) providing adequate resources for assuring high-quality data collection and analysis efforts, (3) developing national standards for public reporting of risk-adjusted outcomes, and (4) reporting measures of appropriateness to complement risk-adjusted outcomes reporting [26].

Impact of Public Reporting on Surgeons

Another impact on the delivery of care that appears to have been related to the release of public cardiac reports is the decrease in prevalence of low-volume surgeons with high risk-adjusted mortality rates. Publications based on New York data highlighted the fact that lower volume surgeons were associated with worse outcomes, and this fact became clear with the release of surgeon-level data [27, 28]. As a results of these studies and reports, some New York hospitals restricted the privileges of low-volume surgeons, and between 1989 and 1992, 27 low-volume surgeons ceased practicing cardiac surgery in the state. Some of these surgeons left the state, some retired, and others restricted their practice to non-cardiac surgery [28]. The risk-adjusted CABG surgery mortality for the group who ceased practicing in the state was 11.9 % in the last year in which they practiced in the state, compared with the statewide rate of 3.1 % in that time interval [12].

A later study by Jha and Epstein used 3-year reports (the periods used to report surgeon data) from 1989 to 1991 through 1994–1996 to examine outcomes for the surgeons who discontinued performing cardiac surgery in the 2-year period following the publication of the reports [29]. The authors found that more than 20 % of surgeons with patient risk-adjusted mortality rates

in the highest (worst) quartile stopped practicing CABG surgery within 2 years after publication of the reports, in comparison to roughly 5 % of surgeons in the top three quartiles. There was a statistically significant difference when all five 3-year reports were combined [29].

The reporting of physician-level data has been one of the most controversial aspects of public reporting, and Massachusetts does not report physician-level data, although the other states that report hospital level cardiac data do report physician-level data. The findings above suggest that knowledge of physician/surgeon performance can help hospitals improve overall performance, although certainly driving poor-performing physicians out of practice or out-of-state is not the only way this can be done. Studies that examine the effectiveness of working with these physicians to improve performance while staying in practice, and studies that examine the impact of confidentially sharing physician-level information in regions without physician-level reporting would be important contributions.

Ability to Predict Performance Over Time (for Use in Choosing Providers)

The ability of public cardiac reports to predict performance over time is important because it there has to be reasonable consistence over time for the reports to be used for patients to choose hospitals and surgeons. Jha and Epstein found that if patients undergoing CABG surgery in New York in 1996 selected a hospital in the top decile with the latest available (1993) data, the mean 1996 risk-adjusted mortality rate of those hospitals was 1.82 %, compared to a rate of 2.89 % in 1996 for hospitals that were in the bottom decile in the 1993 report. Accumulating these data across the 1996–2002 period, the mean risk-adjusted mortality of hospitals that were in the top decile 3 years earlier was 1.59 %, compared to 2.78 % for hospitals in the bottom decile in the index year [29].

Glance et al. compared the ability of the New York CABG report card data to predict mortality 2 and 3 years later using a different strategy. The authors compared quality ratings in two different time periods (first 2 years apart, then 3 years apart) based on ratios of observed to expected mortality. Conclusions of the study were that hospital assessments based on 2-year old data is a strong predictor of future performance, but that 3-year old data may not be useful for identifying low-performance hospitals [30].

Impact of Public Reporting on Market Share

A few studies that have examined the impact of public reporting of CABG surgery mortality on hospitals' future market share, with contrasting results. Hannan et al. [31] found no substantial changes in hospital volumes in the early years of the New York system, between 1989 and 1992. Mukamel and Mushlin found that the New York CABG surgery reports had a small effect on hospital volume between 1990 and 1993, but a larger effect on surgeon volume [32]. Romano and Zhou found that New York CABG hospitals with significantly lower risk-adjusted mortality had higher CABG surgery volume in the first month after publication (61 patients predicted and 75 admitted, a 22 % increase) [33]. A later comprehensive study by Jha and Epstein found no evidence that performance in the reports was associated with a significance increase in market share for hospitals with the best risk-adjusted mortality or a significant decrease in market share for hospitals with the worst risk-adjusted mortality between 1989 and 2002. Also, in general, each hospital's market share remained very similar over time [29]. The fact that these studies came to different conclusions is probably more likely due to differences in the methods used than to differences in the time frames that were studied.

It is also notable that whatever changes in market share did occur, they don't seem to have been strongly impacted by changes in referrals in the early years of the system when such information was obtained. In a survey of New York cardiologists, Hannan and Stone found that only 22 %

routinely discussed the report with their patients, and only 38 % of the cardiologists used the report card information for referrals [34]. A caveat is that these findings are quite old.

Market share changes can also occur is by managed care organizations (MCOs) altering their contracting for tertiary services. Romano et al. found that about half of the hospital administrators they surveyed used the report cards in health-plan negotiations [35]. Also, Mukamel et al. [36] found that 60 % of MCOs who responded to a 1998 survey ranked surgeon quality as the most important factor in contracting, but only 20 % indicated that the report cards were a major factor in their contracting decision. However, a later study by Mukamel et al. [37] found that the probability of MCOs contracting with a surgeon in the Downstate region of New York was significantly higher if surgeon had a lower risk-adjusted mortality rate.

Regardless of the modest impact demonstrated by most of the studies just mentioned, given the explosion in the number of public reports on various aspects of the quality of health care as well as the federal healthcare reform initiative, it stands to reason that consumers will use these reports more in the future as a means of choosing healthcare providers. Shahian et al. provide an excellent discussion of the rationale for this contention [25].

Summary

In summary, the public release of cardiac outcomes has arguably been associated with larger decreases in mortality than in regions without public releases of outcomes. There has also been controversy regarding whether these decreases have been in part due to the avoidance or out-migration of high-risk patients. Given reports from individual physicians asserting that they are aware of this practice, it is certainly true to some degree. What is not known is the degree to which it happens, and whether when it does happen, patients are either referred to other (perhaps more skilled, or suited to their risk-level) providers or the patients are inappropriate for the procedure. My view, based on New York data on volumes of cases and changes in risk profile of patients, combined with data quality auditing, is that the number of high-risk patients not undergoing a procedure from some provider in the state is minimal.

Several other impacts of public reporting have been studied, including the impact on providers' market share, the impact on low-rated surgeons discontinuing practice in the region with public reports, and the ability of the reports to predict future performance. The evidence on market share is dated and ambivalent, but probably understated given the multitude of recent reports, the healthcare reform effort, and the obvious increase in patient interest in quality of care. There does seem to be evidence from the early years of New York's system that surgeons with poor ratings were more likely to discontinue cardiac surgery in relation to other surgeons, although that may no longer be true since the reports do not receive the publicity they received in the press in the early days of the system. Also, there is evidence from New York data that the reports are reasonably good predictors of future performance, although it is important that they be as timely as possible if used for this purpose.

The Future

Despite the controversy about public reporting of health outcomes, it is clear that it is here to stay for the foreseeable future. Two primary reasons for this is the public demand for this information and the embrace of public reporting by governmental agencies (e.g., Centers for Medicare and Medicaid Services) regulators and payers. Consequently, there is a need to focus on what has been learned from existing public report cards. Three of the most valuable lessons we have learned in New York are (1) it is critically important to assure the completeness and accuracy of the data being used because the reports can impact quality of patient care as well as the reputation of healthcare providers, (2) the accep-

tance and use of the reports are impacted by the manner in which they are presented to providers and the public, and by the degree to which these constituencies are part of the process of developing the reports, and (3) the fact that a provider can be identified as having significantly higher mortality than the state or region of interest is a very powerful motivator for effecting change.

Importance of Completeness and Accuracy of Data

In the New York experience over the course of releasing cardiac data for the past 20 years, there have been many instances in which inaccurate data that were threats to the validity of our reports have been discovered. Prior to public knowledge regarding efforts to assure data quality, a hospital submitted more than 300 isolated CABG cases without a reported death. When these data were matched to New York's administrative data, it was discovered that about 50 more patients had undergone surgery and 18 of them had died in the index admission. Other hospitals have maintained that their patients had extremely high prevalences (more than three times as high as the statewide average in some cases) of some risk factors, but medical record auditing uncovered overcoding of these risk factors, and these hospitals have been required to resubmit data or in extreme cases pay for an outside abstractor to submit the hospitals' data. Another hospital reported that numerous patients were discharged alive (when in-hospital mortality was used as the outcome measure) but upon review it was discovered that they were "discharged" to an in-hospital hospice that was not certified and died of complications of CABG surgery.

Also, since mortality after discharge but within 30 days of the index procedure became part of the outcome measure in addition to in-hospital mortality, hospitals were asked to report this measure to the registry. Before the hospital-reported data were used, patients were matched against reported deaths in the National Death Index, and we found that the vast majority of out-of-hospital deaths were not identified by the hospitals. As a result, New York uses the National Death Index, or New York vital statistics data limited to New York patients in conjunction with the Social Security Death Master File, to capture out-of-hospital deaths. The National Death Index (NDI) is preferable since it includes non-New York patients, but funding issues have sometimes required the use of the latter.

In conclusion, there is a strong possibility that publicly reported data will be either accidentally or deliberately inaccurate without concerted efforts to assure accuracy and completeness. With regard to assuring completeness, the best safeguard is to match the data against another database if possible. In New York, we have had access to SPARCS administrative data, and have used that (Statewide Planning and Research Cooperative System [https://www.health.ny.gov/statistics/sparcs/], accessed March 16, 2014). Completeness is probably not a problem when using CMS data because MedPAR should be complete (for Medicare patients) given that it is related to reimbursement [38].

With respect to assuring accuracy of risk factors and outcomes, the New York experience, as noted above, is that out-of-hospital death as an outcome should be captured by matching to death indices. Risk factor accuracy can probably only be assured by auditing medical records. This can be very expensive depending on the percentage of cases that is audited, and thought needs to be given to choosing cases/hospitals to be audited in a cost-effective manner. As noted earlier, New York has chosen hospitals based on past problems, time since last audit, and presence of risk factors in the statistical models with unusually high prevalences [38].

The use of administrative data in the risk-adjustment process, as is the case with the CMS report cards for AMI and CHF, is problematic because the International Classification of Diseases (ICD) codes used in MedPAR are not very detailed. For example, in the New York cardiac registries, creatinine levels are used as a measure of renal failure, and chronic obstructive pulmonary disease (COPD) is defined in terms

of expiratory volume, partial pressure of oxygen (PO_2), and partial pressure of carbon dioxide (PCO_2) levels, whereas the ICD codes have no clinically objective definition for these conditions or any other conditions. This means that more subjectivity is involved in assigning risk factors to patients. Also, for any type of patient, clinical data elements such as blood pressure, ejection fraction, and heart rate are important independent predictors of mortality that are not contained in administrative data. The discrepancies between administrative data and clinical data may result in different hospital risk-adjusted mortality and outlier status for administrative and (gold standard) clinical models.

Improving Acceptance and Use of Public Reports

To improve the acceptance and quality of public reports, it is important to seek the advice of multiple constituencies (patients, hospital administrators, clinicians from inside and outside the health care system being studied, health policy experts, ethicists, researchers, third party payers, etc.) and to keep hospitals and physicians apprised of decisions that are being made as well as to provide them with a forum for making recommendations. For example, New York's CAC has always included several of the most prominent clinicians and researchers in the world. Also, town hall meetings have been convened across the state on several occasions to present the methods used and findings as well as to entertain suggestions and questions, and this seems to be a valuable practice.

However, most people would probably agree that current cardiac and other public reports are difficult to read and comprehend by the average consumer/patient as well as clinician. Also, public reports should probably include additional outcome measures, including appropriateness. Reports also should be tailored to the disease rather than the procedure used to treat it (e.g., acute myocardial infarction reports as is done by CMS in addition to cardiac surgery reports), and possibly include process measures.

Impact of Identifying Outliers

The description above of specific initiatives taken by hospitals in New York to improve their risk-adjusted mortality rates is an indication of the motivation inspired by being identified as an outlier in public reports. In New York, we have also been privy to numerous other off-the-record reports of hospitals who confide that they would not have looked more carefully at their processes of care without having been identified as an outlier or feared being an outlier. An interesting topic for a future study would be differences in quality initiatives of hospitals that were and were not flagged as outliers in public reporting regions. Moreover, the identification of high performing outliers can create a platform for ascertaining and disseminating best practices. In conclusion, reports that contain very few outliers may not have the ability to effect change as much as reports that tend to distinguish hospitals from one another. Also, reports that do not distinguish quality among providers offer little interest or guidance to patients and referring physicians [24].

References

1. Krakauer H, Bailey C, Skellan KJ, et al. Evaluation of the HCFA model for the analysis of mortality following hospitalization. Health Serv Res. 1992;27(3):317–35.
2. Hannan EL, Kilburn Jr H, Lindsey ML, Lewis R. Clinical versus administrative databases for CABG surgery. Med Care. 1992;30(10):892–907.
3. Hannan EL, Racz MJ, Jollis JG, Peterson ED. Using Medicare claims data to assess provider quality for CABG surgery: does it work well enough? Health Serv Res. 1997;31(6):559–678.
4. Hannan EL, Kilburn Jr H, O'Donnell JF, Lukacik G, Shields EP. Adult open heart surgery in New York State: an analysis of risk factors and hospital mortality rates. JAMA. 1990;264(21):2768–74.
5. Lawrence KA. Heart surgery death rates decline in New York. New York Times, 5 Dec 1990.
6. Coronary artery bypass surgery in New York State: 1990–1992. New York State Department of Health, Dec 1993.
7. Cardiac surgery in Pennsylvania 2007–2008. Pennsylvania Health Care Cost Containment Council, Sept 2010.
8. The California report on coronary artery bypass graft surgery: 2007 hospital data. California CABG

Outcomes Reporting Program. California Office of Statewide Health Planning and Development; June 2011.
9. Adult coronary artery bypass graft surgery in the Commonwealth of Massachusetts: fiscal year 2009 report. Harvard Medical School; Jan 2011.
10. Cardiac surgery in New Jersey: 2008. Health Care Quality Assessment, Office of Policy and Strategic Planning, New Jersey Department of Health and Senior Services; June 2011.
11. Dziuban Jr SW, McIlduff JB, Miller SJ, Dal Col RH. How a New York cardiac surgery program uses outcomes data. Ann Thorac Surg. 1994;58(6):1871–6.
12. Chassin MR. Achieving and sustaining improved quality: lessons from New York State and cardiac surgery. Health Aff. 2002;21(4):40–51.
13. Coronary artery bypass surgery in New York State: 1996–1998. New York State Department of Health; Jan 2001.
14. Hannan EL, Kilburn Jr H, Racz M, Shields E, Chassin MR. Improving the outcomes of coronary artery bypass surgery in New York State. JAMA. 1994;271(10):761–6.
15. Ghali WA, Ash AS, Hail RE, Moskowitz MA. Statewide quality improvement initiatives and mortality after cardiac surgery. JAMA. 1997;277:379–82.
16. Peterson ED, DeLong ER, Jollis JG, Muhlbaier LH, Mark DB. The effects of New York's bypass surgery provider profiling on access to care and patient outcomes in the elderly. J Am Coll Cardiol. 1998;32(4):993–9.
17. Hannan EL, Sarrazin MSV, Doran DR, Rosenthal GE. Provider profiling and quality improvement efforts in coronary artery bypass graft surgery. Med Care. 2003;41:1164–72.
18. Burack JH, Impellizzeri P, Homel P, Cunningham Jr JN. Public reporting of surgical mortality: a survey of New York state cardiothoracic surgeons. Ann Thorac Surg. 1999;68(4):1195–202.
19. Narins CR, Dozier AM, Ling FS, Zareba W. The influence of public reporting of outcome data on medical decision making by physicians. Arch Intern Med. 2005;165(1):83–7.
20. Schneider EC, Epstein AM. Use of public performance reports: a survey of patients undergoing cardiac surgery. JAMA. 1998;279:1638–42.
21. Omoigui NA, Miller DP, Brown KJ, et al. Outmigration for coronary artery bypass surgery in an era of public dissemination. Circulation. 1996;93:27–33.
22. Moscucci M, Eagle KA, Share D, et al. Public reporting and case selection for percutaneous coronary interventions: an analysis from two large multicenter percutaneous coronary intervention databases. J Am Coll Cardiol. 2005;45(11):1759–65.
23. Dranove D, Kessler D, McClellan M, Satterthwaite M. Is more information better? The effects of "report cards" on health care providers. J Polit Econ. 2005;111(3):555–88.
24. Epstein AJ. Do cardiac surgery report cards reduce mortality? Assessing the evidence. Med Care Res Rev. 2006;63:403–26.
25. Shahian DJ, Edwards FH, Jacobs JP, et al. Public reporting of cardiac surgery performance: part I-history, rationale, consequences. Ann Thorac Surg. 2011;92:S2–11.
26. Resnic FS, Welt FGP. The public health hazards of risk avoidance associated with public reporting of risk-adjusted outcomes in coronary intervention. J Am Coll Cardiol. 2009;53:825–30.
27. Hannan EL, O'Donnell JF, Kilburn Jr H, Bernard H, Yazici A. Investigation of the relationship between volume and mortality for surgical procedures performed in New York State hospitals. JAMA. 1989;262:503–10.
28. Hannan EL, Siu AL, Kumar D, Kilburn Jr H, Chassin MR. The decline in coronary artery bypass graft surgery mortality in New York State: the role of surgeon volume. JAMA. 1995;272:209–13.
29. Jha A, Epstein AM. The predictive accuracy of the New York State Coronary Artery Bypass Surgery Report-Card System. Health Aff. 2006;25(3):844–55.
30. Glance LG, Dick AW, Mukamel DB, Osler TM. How well do hospital mortality rates reported in the New York State CABG report card predict subsequent hospital performance? Med Care. 2010;48(5):466–71.
31. Hannan EL, Kumar D, Racz M, Siu AL, Chassin MR. New York State's cardiac surgery reporting system: four years later. Ann Thorac Surg. 2004;58(6):1852–7.
32. Mukamel DB, Mushlin AI. Quality of care information makes a difference: an analysis of market share and price changes after publication of the New York Cardiac Surgery Mortality Reports. Med Care. 1998;36(7):945–54.
33. Romano PS, Zhou H. Do well-publicized risk-adjusted outcomes reports affect hospital volume? Med Care. 2004;42(4):367–77.
34. Hannan EL, Stone CA, Biddle TB, DeBuono BA. Public release of cardiac surgery outcomes data in New York: what do New York State cardiologists think of it? Am Heart J. 1997;134:1120–8.
35. Romano PS, Rainwater JA, Antonius D. Grading the graders: how hospitals in New York and California perceive and interpret their report cards. Med Care. 1999;35(3):295–305.
36. Mukamel DB, Mushlin AI, Weimer D, Zwanziger J, Parker T, Indridason I. Do quality report cards play a role in HMO's contracting practices? Evidence from New York State. Health Serv Res. 2000;35(1, Part II):319–32.
37. Mukamel BB, Weimer DL, Zwanziger J, Mushlin AI. Quality of cardiac surgeons and managed care contracting practices. Health Serv Res. 2002;37(5):1129–44.
38. Hannan EL. Ensuring accuracy and completeness of data used for outcomes assessment. Ann Thorac Surg. 2012;93:1172–3.

Public Reporting of Pediatric Cardiac Data

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Abstract

Public reporting of outcomes and quality in cardiac surgery is part of a national effort to improve performance and accountability in healthcare. There have been pioneering efforts to justly tabulate and report clinical risk-adjusted data with much experience gained over the last decade. We are now on the threshold of national public reporting in congenital heart surgery. Our objective is to outline the foundation for public reporting of cardiac surgical outcomes and describe the recent progress and future direction for public reporting of pediatric cardiac data.

Keywords

Outcome assessment • Database analysis • Congenital heart surgery • Public reporting

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> "The purest treasure mortal times afford…Is spotless reputation." William Shakespeare, Richard II 1:1

Improving performance and accountability in healthcare requires the development of shared goals that unite the interests of all stakeholders [1]. Public reporting of outcomes and quality in cardiac surgery is a means to strategically approach this important objective. If done properly, this effort has the potential to harmonize the goals of individual physician providers with those of hospitals, health plans, national payers, state and federal governments, and consumers. The availability of reliable and risk-adjusted outcomes data presented in a clear, concise, and audience-appropriate manner has the ability to aid patients and their families in the making of informed decisions; simultaneously, it will motivate hospitals and providers to refine their processes in order to increase the value of their healthcare delivery.

Measuring the value of pediatric cardiac surgical care provided over the last decade has been greatly facilitated by multidisciplinary international consensus regarding precise patho-anatomic classification of disease, resulting in more accurate reporting of outcome data [2–4]. Together with more precise definitions of morbidity and mortality, this collaborative process has promoted evolving international efforts to measure and improve pediatric cardiac surgical quality by procedure and disease type [4–7]. Now, in addition to measuring and reporting a single center's short-term experience with a complex multi-procedural pediatric cardiac operation, which is undeniably valuable both for the centers and for their patients, national and international benchmarks are available. Efforts are ongoing to establish strategies for the longitudinal assessment of outcomes after cardiothoracic surgery. This is of particular importance in the pediatric cardiac population in order to assess the potential impact of procedures on subsequent neurocognitive development and the durability of specific procedures over time.

As we are on the threshold of national public reporting in congenital heart surgery, the objective of this article is to outline the foundation for public reporting of cardiac surgical outcomes and describe the recent progress and future direction for public reporting of pediatric cardiac data.

The Beginning of Public Reporting

The contemporary effort to publicly report outcome data began in 1986 with a groundbreaking initiative by the United States Health Care Financing Administration (HCFA), the Medicare management predecessor to the Centers for Medicare and Medicaid Services (CMS). HCFA's goal was to address the inherent variability in reported death rates across US hospitals treating Medicare patients. Their seemingly straightforward approach involved identifying 269 hospitals that had death rates outside of the range predicted for the overall Medicare population and publicizing these findings. However, these published "death lists" were based on Medicare claims data and had numerous methodological flaws, including the lack of credible risk-adjustment [8, 9]. Because of these criticisms, the program was finally terminated in 1993. Nonetheless, through this initial organized effort to systematically assess mortality rates, HCFA brought transparency of reporting of outcomes to the national stage and ushered in our current era of public reporting.

Cardiac surgery has always been among the most resource intense specialties in healthcare, particularly in the adult Medicare population. In the late 1980s, coronary artery bypass grafting (CABG) was the most common cardiac procedure performed in the United States [10–12]. Not surprisingly, therefore, cardiac surgery became the early focus of performance assessment and improvement initiatives that were far more sophisticated than those in the initial HCFA efforts. Beginning in the late 1980s, several states began to collect, analyze and publish data on cardiac surgery outcomes. At the same time, professional organizations, most notably the Society of Thoracic Surgeons (STS), made strategic decisions to develop clinical data sources and risk-adjustment methodologies. What was set in motion by these events has forever changed the landscape of transparency and accountability, not only in cardiac surgery but across healthcare.

We will outline the key developments in each of these areas and their impact on public reporting and accountability, culminating in the pub-

lic reporting of risk-adjusted pediatric cardiac outcomes by the STS Congenital Heart Surgery Database.

Lessons from Early Experiences with Public Reporting of Cardiac Surgery Data

State-based initiatives to analyze clinical data in cardiac surgery began in New York, Massachusetts and Pennsylvania. In New York, beginning in 1988, early reports of substantial variability in cardiac surgery outcomes led to the development of a clinical data registry and hospital-level risk-adjusted outcomes analyses, which were released publicly in 1990. Because of a media suit, the state was also ultimately required to release physician-level outcomes. Short-term mortality following CABG was reduced by 41 % in the first 4 years of the program [13–15], and this program has expanded to include report cards for percutaneous coronary interventions, cardiac valve surgery, and pediatric cardiac surgery.

However, despite progressive decreases in risk-adjusted mortality, which may have been driven by the exodus of low-volume high mortality surgeons, there was little demonstrable change in the market share of hospitals with significantly higher or lower than expected mortality rates. Furthermore, numerous studies in New York and Pennsylvania raised concerns that public reporting of CABG outcomes had led to risk-averse behavior by surgeons and hospitals. In other words, because they feared the potential impact of bad outcomes on their reputations and referrals, providers were less inclined to accept higher risk patients for surgery, despite the fact that all their results would be risk-adjusted. This potential risk-aversion was, and remains, a major unintended negative consequence of cardiac surgery public reporting, and the same phenomenon has also been observed in New York and Massachusetts for percutaneous coronary interventions. The concern is that this behavior may reduce access to care for the very patients who might benefit the most, even though they are high risk [9, 14]. While New York pioneered cardiac surgical outcome reporting, it also exposed the potential negative consequences resulting from the anxiety of providers. It is hoped that by including comprehensive risk-adjustment and fostering trust in this process, the practice of risk aversion in congenital heart surgery can be avoided.

In 1986, Pennsylvania enacted legislation creating the Pennsylvania Health Care Cost Containment Council (PHC4) as an independent state agency to collect hospital and provider outcome data, with the primary purpose of making these data publicly accessible. By state law, hospitals were required to submit cardiac surgery data for hospitals and individual surgeons. However, while PHC4 results are risk-adjusted, they currently remain based primarily on claims data.

In 2000, the Massachusetts legislature mandated the development of a public report card for cardiac surgery, and this system was implemented in 2002 using STS data and locally developed risk algorithms. Also developed in the late 1980s, the Northern New England Cardiovascular Disease Study Group [16] focused on confidential feedback to its participants but did not, until recently, publish these results. Interestingly, their mortality rates and reductions in mortality over time using this confidential feedback and best practice approach were similar to those achieved in public reporting states. This reduction of mortality and morbidity by the Northern New England Cardiovascular Disease Study Group demonstrates the value of robust, risk-adjusted, confidential, peer-level feedback to achieve improvement in outcomes without the confounding influence of media scrutiny that may promote risk aversion.

In 1987, The Veterans Administration (VA) developed a clinical risk-adjusted database that served to monitor outcomes in all VA medical centers performing cardiac surgery. A short data form of 50 variables was recorded on all patients undergoing cardiac surgery, and participation in the VA Continuous Improvement in Cardiac Surgery Program was mandatory. The VA developed multivariable risk modeling to perform risk-adjusted analyses for isolated CABG, isolated valve, and great vessel cardiac surgery. The national VA Quality Oversight Committee calculated the 30-day risk-adjusted mortality rates and reported this information every 6 months to

regional and facility leadership, who then disseminated these data to front-line providers with the goal of improving quality. This program resulted in a decrease in the observed to expected mortality ratio from 1.5 to 0.9 over the first 9 years of implementation [17–19].

These early and ongoing initiatives highlight the value of publicly reporting cardiac outcomes data but also some of its potential perils, including risk aversion. To avoid such behavior, there is value in beginning such an initiative with confidential peer reporting of clinical data that are robustly risk-adjusted and communicated in a trustworthy manner. Once trust is established in the value and accuracy of the data, public reporting can be initiated with less potential for negative unintended consequences. In addition, the mode in which the data is publicly reported is clearly of importance. To foster informed, shared decision-making, the data must be credible and presented with sufficient flexibility that stakeholders with varying levels of education and sophistication can all understand and interpret them correctly. These important lessons have been incorporated into the evolution of the STS National Database.

The Society of Thoracic Surgeons (STS) National Database

In 1989, STS committed to develop a voluntary national database for cardiothoracic surgery. Similar to state-based responses to the HCFA mortality report, this national quality effort resulted from the desire of surgeons to collect their own national, risk-adjusted data to study and improve the quality of cardiac surgery [20–22]. The STS National Database has evolved to become one of the world's largest and most robust cardiac surgery databases and one of the most respected national-level participants in public reporting.

Duke Clinical Research Institute (DCRI) serves as the data warehouse and analytic center of the STS National Database. The partnership and collaboration of STS and DCRI has facilitated refinements in the statistical modeling and analytics used for risk assessment [23, 24], including the use of hierarchical approaches and longitudinal data linkages [25].

With 90–95 % of all cardiac surgery programs in the US participating, the STS National Database has become the paradigm for clinical data registries developed by professional societies in healthcare. It has been a major driving force in defining best practice in quality measurement, quality improvement, and clinical research. The STS National Database now encompasses all aspects of cardiothoracic surgery and all its major procedures, and over 40 sophisticated risk models have been developed.

STS National Database Partners with United States Healthcare for National Quality Measurement

As the US healthcare system pursued ways to improve value of care through higher quality and reduced cost, the Institute of Medicine (IOM) delivered two seminal reports in 1999 and 2001 that served as a call to arms in the pursuit of quality and transparency [26–28]. The IOM estimated that over 90,000 Americans die each year from preventable medical errors, which led to increasing demands for accountability and transparency of outcomes in healthcare. The IOM defined six aims that would broadly shape the future healthcare system — care should be safe, effective, patient-centered, timely, efficient and equitable [28]. The Agency for Healthcare Research and Quality (AHRQ) operationalized these recommendations through the identification of common priority conditions, a call for providers to use information technology and data registries to aid in this mission, and the alignment of payment policies with quality improvement.

In 1999, the National Quality Forum (NQF), a nonprofit, multi-stakeholder membership organization was created to advance efforts to improve health care quality through measurement and reporting. In 2002, NQF identified and subsequently published a report on 27 adverse events considered preventable and of concern to the public and health care providers. Since then, NQF has endorsed voluntary consensus standards for

Table 36.1 The Society of Thoracic Surgeons in collaboration with the National Quality Forum agreed on 21 outcome and process measures for cardiac operations. These measures became the standards for the first public reporting initiative for risk-adjusted clinical adult cardiac surgery data

National voluntary consensus standards for cardiac surgery
1. Participation in a systematic database for cardiac surgery
2. Surgical volume for isolated coronary artery bypass graft (CABG) surgery, valve surgery, and CABG + valve surgery
3. Timing of antibiotic administration for cardiac surgery patients
4. Selection of antibiotic administration for cardiac surgery patients
5. Pre-operative beta blockade for CABG
6. Use of internal mammary for CABG
7. Duration of prophylaxis for cardiac surgery patients
8. Prolonged intubation
9. Deep sternal wound infection rate
10. Stroke/cerebrovascular accident
11. Post-operative renal insufficiency
12. Surgical re-exploration
13. Anti-platelet medications at discharge for CABG
14. Beta blockade at discharge for CABG
15. Anti-lipid treatment at discharge for CABG
16. Risk-adjusted inpatient operative mortality for CABG
17. Risk-adjusted operative mortality for CABG
18. Risk-adjusted operative mortality for aortic valve replacement (AVR)
19. Risk-adjusted operative mortality for mitral valve replacement/repair (MVR)
20. Risk-adjusted operative mortality for MVR + CABG
21. Risk-adjusted operative mortality for AVR + CABG

Adapted from National Quality Forum Consensus Report on Cardiac Surgery, 2004. National Quality Forum, 601 Thirteenth Street, NW, Suite 500 N, Washington, DC 20005

Table 36.2 Utilizing Bayesian modeling and distribution to calculate the STS CABG Composite Scores, the majority often fall at the 2-star level with smaller proportions at the 1-star and 3-star level

Star rating	Spring 2007 (%)	Fall 2007 (%)	Spring 2008 (%)	Fall 2008 (%)	Spring 2009 (%)
1	12.9	11.4	12.1	12.1	12.2
2	72.0	76.5	76.2	74.1	72.3
3	15.1	12.1	11.6	13.9	15.5

public reporting of performance metrics across a broad range of health care settings and conditions. Through collaborations with other national healthcare entities, NQF has helped launch public reporting into the modern era, and STS has played a large part in that evolution.

In 2004, NQF endorsed 21 adult cardiac surgery measures, most of which were developed by STS (Table 36.1). These evidence-based measures have stood the test of time and have been re-endorsed in 2007 and again in 2011. STS formed its Quality Measurement Task Force in 2005 to use the NQF-endorsed cardiac surgery performance measures to develop an objective, evidence-based, statistically valid model of quality in cardiac surgery. Performance measurement in cardiac surgery can be divided into three principle categories, based on the original Donabedian taxonomy: Structural measures, Process measures, and Outcomes measures [29]. Structural measures include surgical volumes and participation in a national clinical registry. Process measures include evidence-based variables such as the use of the internal mammary artery in CABG and perioperative medications (e.g., beta blockers and statins) that have been shown to improve long term outcomes. Outcome measures include mortality and major complications such as postoperative stroke, renal failure, prolonged ventilation, surgical re-exploration, and deep sternal wound infection.

To further enhance the comprehensive measurement of cardiac surgery quality, STS developed composite measures that included multiple performance domains, not just mortality. The first of these was the STS CABG Composite Score, which was subsequently the basis for the first publicly reported outcomes by STS [28, 29]. Utilizing sophisticated statistical approaches [30], these composites were reported to the public in an easily understood star-rating format, ranging from 1 (lowest performance) to 3 (highest performance) (Table 36.2). The STS CABG Composite Score is based on 11 NQF-endorsed individual measures within four domains (Table 36.3). The first domain is avoidance of risk-adjusted operative mortality; the second is avoidance of any of

the five major risk-adjusted major complications (reoperation, stroke, renal failure, deep sternal wound infection, or prolonged ventilator support); the third domain is use of the internal mammary artery for revascularization; and the fourth is use of all evidence-based perioperative medications (aspirin, statins, beta blockers). To enhance consumer understanding, while at the same time providing drill-down capability for more sophisticated stakeholders, STS provides star ratings as well as numerical scores (Fig. 36.1).

Table 36.3 STS CABG Composite Score utilizing 4 domains and 11 cardiac surgery measures endorsed by the National Quality Forum

Perioperative medical care bundle
All four indicated medications given
Preoperative β blockade
Discharge aspirin, β blockade, and lipid-lowering agents -2° prevention
Operative care
Single process measure: IMA use
Risk-adjusted operative mortality
Postoperative risk-adjusted major morbidity bundle
Risk-adjusted occurrence of any of the following
Renal insufficiency
Deep sterna wound infection
Re-exploration
Stroke
Prolonged ventilation/intubation

IMA internal mammary artery

On September 7, 2010, voluntary public reporting of the composite scores for isolated CABG commenced with a collaborative effort between STS and Consumers Union, publisher of the widely popular Consumer Reports magazine. In January 2011, STS launched STS Public Reporting Online, a section of its website dedicated to the reporting of composite scores for isolated CABG and to providing public reporting-related resources. In January 2013, STS began the voluntary public reporting of composite scores for isolated aortic valve replacement (AVR). Moving forward, STS plans to develop and publicly report an additional composite score

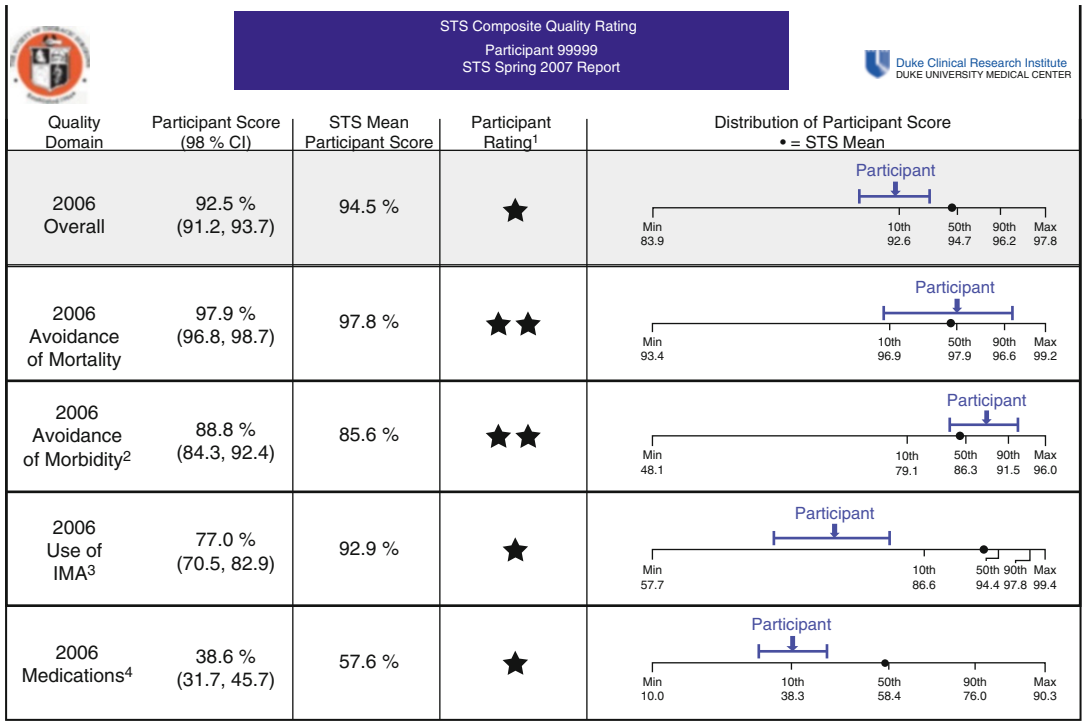

Fig. 36.1 Sample of the STS CABG Composite Score report provided to database participants semiannually (Note: sample data; not associated with a patient) (© The Society of Thoracic Surgeons 2014, used with permission from STS. All rights reserved)

every year. In 2014, a composite score for combined AVR and CABG will be added to the STS public reporting portfolio. In 2015, a composite score for mitral valve surgery will be added to the STS public reporting portfolio. Also in 2014, STS plans to begin publicly reporting pediatric cardiac surgical outcomes. Finally, in 2015, STS plans to begin publicly reporting general thoracic surgical outcomes beginning with isolated lobectomy in 2015 and likely progressing to esophagectomy in 2016.

Recently, the STS National Database has further extended its value beyond 30 day outcomes with the linkage to external data sources such as those provided by the Centers for Medicare and Medicaid Services. These data linkages have been achieved using probabilistic and deterministic matching [31], thus providing an extraordinary tool for multi-year longitudinal outcome to address important clinical questions, such as long term outcomes, readmissions, re-interventions, and costs [32]. Although its availability is currently in question, the Social Security Death Master File has also been useful in validating longitudinal mortality across all age ranges. The National Death Index is another source of longitudinal information about life status [33].

Quality Measurement and Public Reporting in Pediatric and Congenital Heart Surgery

Events at Bristol, England [34], Denver, Colorado [35–41], Winnipeg, Canada [42], and recently Lexington, Kentucky [43], have clearly demonstrated the importance of multi-institutional quality measurement in pediatric and congenital cardiac surgery. The Bristol Report presents the results of the inquiry into the management of the care of children receiving complex cardiac surgical services at the Bristol Royal Infirmary between 1984 and 1995 and relevant related issues. In the Bristol Report, approximately 200 recommendations are made, many of which relate to the need for accurate multi-institutional outcomes databases to quantitate outcomes of care rendered to patients with pediatric and congenital cardiac disease. Perhaps less well-known than the Bristol Report, the Report of the Manitoba Pediatric Cardiac Surgery Inquest presents data from an inquest involving 12 children who died while undergoing, or soon after having undergone, cardiac surgery at the Winnipeg Health Sciences Centre in 1994. These events demonstrate the importance of a meaningful and fair method of multi-institutional analysis of outcomes for congenital cardiac surgery.

Building upon the progressively data-driven approach of the STS Adult Cardiac Surgery Database, transformative developments have occurred over the past 15 years through the establishment and maturation of the STS Congenital Heart Surgery Database, including an international collaboration to define and develop global standards in outcome reporting of pediatric cardiac operations. This work has helped to reshape and clarify not only how congenital heart surgery is reported, but even how it is performed, through specific patho-anatomic definitions and tracking related outcomes. As of January 1, 2014, the STS Congenital Heart Surgery Database contains data from 117 of the 125 hospitals (93.6 % penetrance by hospital) in the United States that perform pediatric cardiac surgery and 3 of the 8 centers in Canada (Fig. 36.2).

The International Congenital Heart Surgery Nomenclature and Database Project was a joint initiative of STS and The European Association for Cardio-Thoracic Surgery (EACTS). It established data elements and key outcome variables that are the foundation for international collaboration in the recording and reporting of pediatric cardiac data [4]. The nomenclature of the International Congenital Heart Surgery Nomenclature and Database Project was cross-mapped to the nomenclature of the European Pediatric Cardiac Code of the Association of The Association of European Pediatric Cardiology (AEPC) to create the International Pediatric and Congenital Cardiac Code (IPCCC) [44]. The IPCCC and data standards of STS and EACTS are now utilized by both organizations and have provided the platform for the analysis of over 250,000 pediatric and

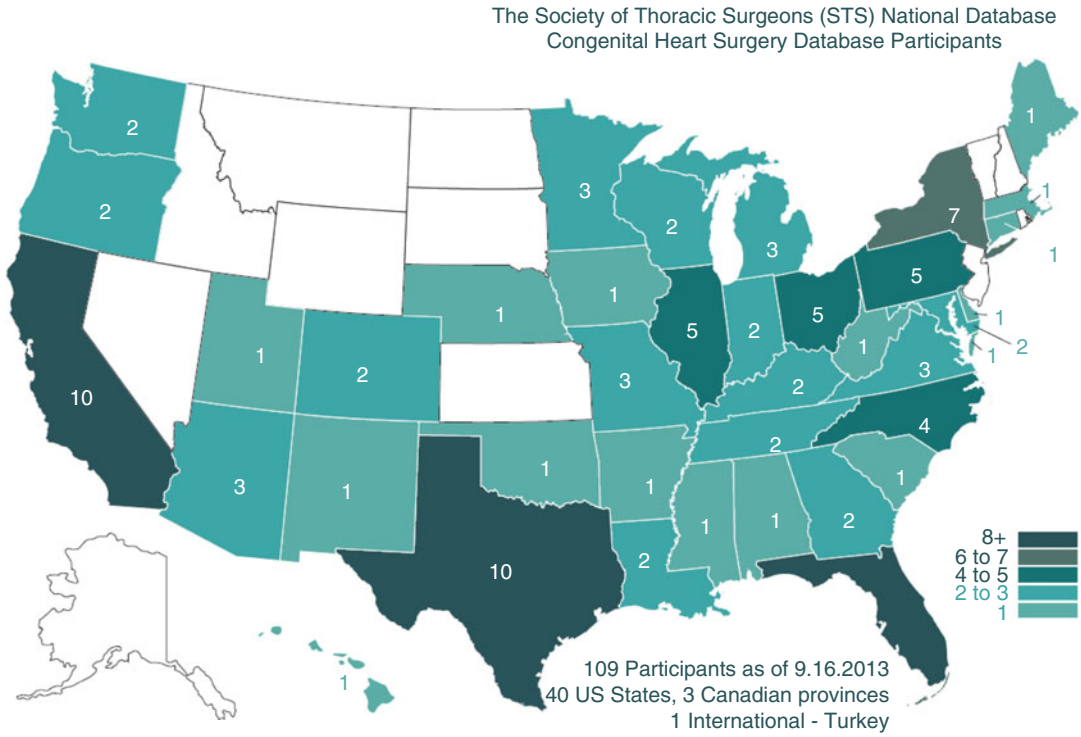

Fig. 36.2 Representation of the completeness of the STS Adult Cardiac Surgery Database in the representation of cardiac surgery in the United States (Reprinted with permission from http://www.sts.org/sites/default/files/documents/adultcardiacMap2.pdf, updated September 16, 2013, accessed January 22, 2014. © The Society of Thoracic Surgeons 2014, used with permission from STS. All rights reserved)

congenital cardiac operations. Efforts to define risk adjustment have also evolved from methodologies based primarily on subjective probability and expert opinion, (**R**isk **A**djustment for **C**ongenital **H**eart **S**urgery-1 Categories [**RACHS-1 Categories**] and **A**ristotle **B**asic Complexity **L**evels [**ABC Levels**]) to methodologies based on the use of objective data and Bayesian algorithms (The **S**ociety of **T**horacic Surgeons – European **A**ssociation for Cardio-**T**horacic Surgery Congenital Heart Surgery Mortality Categories [**STAT Mortality Categories**]) [5–7].

STS has collaborated with the Congenital Heart Surgeons' Society (CHSS) to develop and endorse metrics to assess the quality of care delivered to patients with pediatric and congenital cardiac disease [45]. The 21 "Quality Measures for Congenital and Pediatric Cardiac Surgery" that were developed and approved by the Society of Thoracic Surgeons (STS) and endorsed by the Congenital Heart Surgeons' Society (CHSS) were published in 2012 and are listed in Table 36.4. These Quality Measures are organized according to Donabedian's Triad of Structure, Process, and Outcome. Three of these measures have been endorsed by The National Quality Forum:

- Participation in a National Database for Pediatric and Congenital Heart Surgery
- Surgical volume for Pediatric and Congenital Heart Surgery: Total Programmatic Volume and Programmatic Volume Stratified by the Five STAT Mortality Categories
- Operative Mortality Stratified by the Five STAT Mortality Categories

STS has recently developed a platform for the public reporting of congenital cardiac operations based on a series of benchmark operations (Table 36.5) and the empirically derived STAT risk categories [46–48]. Previous analyses of the STS Congenital Heart Surgery Database have

Table 36.4 Quality measures for congenital and pediatric cardiac surgery

1. Participation in a **national database** for pediatric and congenital heart surgery
2. **Multidisciplinary rounds** involving multiple members of the healthcare team
3. Availability of institutional **pediatric ECLS (extracorporeal life support) program**
4. **Surgical volume** for pediatric and congenital heart surgery: total programmatic volume and programmatic volume stratified by the **five STAT mortality categories**
5. **Surgical volume** for **eight pediatric and congenital heart benchmark operations**
6. Multidisciplinary **preoperative planning conference** to plan pediatric and congenital heart surgery operations
7. Regularly scheduled **quality assurance and quality improvement cardiac care conference**, to occur no less frequently than once every 2 months
8. Availability of **intraoperative transesophageal echocardiography (TEE)** and epicardial echocardiography
9. **Timing of antibiotic administration** for pediatric and congenital cardiac surgery patients
10. Selection of **appropriate prophylactic antibiotics and weight-appropriate dosage** for pediatric and congenital cardiac surgery patients
11. Use of an **expanded pre-procedural and post-procedural "time-out"**
12. Occurrence of new post-operative **renal failure** requiring dialysis
13. Occurrence of new post-operative **neurological deficit** persisting at discharge
14. Occurrence of arrhythmia necessitating **permanent pacemaker** insertion
15. Occurrence of **paralyzed diaphragm** (possible phrenic nerve injury)
16. Occurrence of need for **postoperative mechanical circulatory support** (IABP, VAD, ECMO, or CPS)
17. Occurrence of **unplanned reoperation** and/or interventional cardiovascular catheterization procedure
18. **Operative mortality** stratified by the five STAT mortality categories
19. **Operative mortality** for eight benchmark operations
20. Index cardiac **operations free of mortality and major complication**
21. Operative **survivors free of major complication**

documented substantial variation across institutions in discharge mortality and postoperative length of stay across these benchmark operations [46] and STAT categories [47].

Table 36.5 Benchmark operations in congenital heart surgery as defined by the Society of Thoracic Surgeons Congenital Heart Surgery Database

Benchmark operations in congenital heart surgery
1. Ventricular septal defect (VSD) repair
2. Tetralogy of fallot repair
3. Complete atrio-ventricular canal repair
4. Arterial switch operation (ASO)
5. ASO + VSD repair
6. Fontan operation
7. Truncus arteriosus repair
8. Norwood procedure

Adapted from [35]
An analysis of the STS Congenital Heart Surgery Database revealed that variation in outcome was most prominent for the more complex operations

Future Directions for Public Reporting of Pediatric Cardiac Data

After 3 years of successful public reporting of adult cardiac data from the STS Adult Cardiac Surgery Database, STS is on the cusp of publicly reporting pediatric cardiac surgical data, an initiative that is expected to commence in 2014. Plans are in place to initially publicly report data from the STS Congenital Heart Surgery Database in three states: Florida, Delaware, and Pennsylvania. All participant sites and surgeons in these three states have agreed to publicly report these outcome data, and the political leadership of these states has agreed to adopt the STS public reporting methodology as the mode to disseminate this information. These statewide platforms for public reporting of pediatric and congenital cardiac data will contain identified, site-specific data compared to both national aggregate data and the state peer group cohort. The plan is to initially make this information available on state-specific websites for public dissemination within each state. Later in 2014, this initiative will expand to the national level on a voluntary basis and will be reported on the STS website.

Future plans include implementation of enhanced risk adjustment methodologies and the creation of a composite score for pediatric and congenital cardiac surgery. This composite score would likely be based on the two domains of

mortality and morbidity, each stratified by STAT categories and further informed by additional patient and procedure specific variables. This methodology will eventually allow for public reporting of observed to expected risk-adjusted mortality and morbidity, standardized mortality rates (adjusted mortality rates), and even star ratings similar to those used by the STS Adult Cardiac Database.

Pediatric cardiac data reporting is rapidly evolving and adapting to the current era of public reporting. It is through persistent evidence-based analysis and continued international collaboration that we can make clear, accurate, audience-appropriate, risk-adjusted pediatric cardiac data available to patients, families, regulators, and payers, Simultaneously, we need to motivate hospitals and providers to refine their structures and processes to augment the value of their healthcare delivery.

References

1. Porter ME. What is value in health care? N Engl J Med. 2010;363:2477–81.
2. Strickland MJ, Riehle-Colarusso TJ, Jacobs JP, et al. The importance of nomenclature for congenital cardiac disease: implications for research and evaluation. In: Jacobs JP, editor. Cardiology in the Young Supplement: Databases and The Assessment of Complications associated with The Treatment of Patients with Congenital Cardiac Disease. Prepared by: The Multi-Societal Database Committee for Pediatric and Congenital Heart Disease. Cardiology in the Young, 2008. vol. 18, issue S2 (Suppl 2), p. 92–100.
3. Lacour-Gayet F, Maruszewski B, Mavroudis C, Jacobs JP, Elliott MJ. Presentation of the International Nomenclature for Congenital Heart Surgery. The long way from nomenclature to collection of validated data at the EACTS. Eur J Cardiothorac Surg. 2000;18:128–35.
4. Mavroudis C, Jacobs JP. International congenital heart surgery nomenclature and database project. Ann Thorac Surg. 2000;69(S1):1–372.
5. Jacobs JP, Mavroudis C, Jacobs ML, et al. What is operative mortality? Defining death in a surgical registry database: a report from the STS Congenital Database Task Force and the Joint EACTS-STS Congenital Database Committee. Ann Thorac Surg. 2006;81:1937–41.
6. O'Brien SM, Clarke DR, Jacobs JP, et al. An empirically based tool for analyzing mortality associated with congenital heart surgery. J Thorac Cardiovasc Surg. 2009;138:1139–53.
7. Jacobs ML, O'Brien SM, Jacobs JP, et al. An empirically based tool for analyzing morbidity associated with operations for congenital heart disease. J Thorac Cardiovasc Surg. 2013;145:1046–57.
8. Berwick DM, Wald DL. Hospital leaders' opinions of the HCFA mortality data. JAMA. 1990;263:247–9.
9. Shahian DM, Edwards FH, Jacobs JP, et al. Public reporting of cardiac surgery performance: part 1 history, rationale, consequences. Ann Thorac Surg. 2011;92:2–11.
10. Guyatt G, Drummond M, Feeny D, et al. Guidelines for the clinical and economic evaluation of healthcare technologies. Soc Sci Med. 1986;22:393–408.
11. Cowper P, DeLong E, Peterson ED, et al. Geographic variation in resource use for coronary artery bypass surgery. Med Care. 1997;35:320–33.
12. Weinstein MC, Stason WB. Cost-effectiveness of interventions to prevent or treat coronary heart disease. Annu Rev Public Health. 1985;6:41–63.
13. Hannan EL, Kilburn Jr H, O'Donnell JF, Lukacik G, Shields EP. Adult open heart surgery in New York State. An analysis of risk factors and hospital mortality rates. JAMA. 1990;264:2768–74.
14. Chassin MR. Achieving and sustaining improved quality: lessons from New York State and cardiac surgery. Health Aff. 2002;21:40–51.
15. Hannan EL, Kumar D, Racz M, Siu AL, Chassin MR. New York State's cardiac surgery reporting system: four years later. Ann Thorac Surg. 1994;58:1852–7.
16. O'Connor GT, Plume SK, Olmstead EM, et al. Multivariate predication of in-hospital mortality associated with coronary artery bypass graft surgery. Northern New England Cardiovascular Disease Study Group. Circulation. 1992;85:2110–8.
17. Grover FL, Johnson RR, Shroyer AL, Marshall G, Hammermeister KE. The veterans affairs continuous improvement in cardiac surgery study. Ann Thorac Surg. 1994;58:1845–51.
18. Grover FL, Shroyer AL, Hammermeister K, et al. A decade's experience with quality improvement in cardiac surgery using the Veterans Affairs and Society of Thoracic Surgeons national databases. Ann Thorac Surg. 2001;234:464–72.
19. Hammermeister KE, Johnson R, Marshall G, Grover FL. Continuous assessment and improvement in quality of care. A model from the Department of Veterans Affairs Cardiac Surgery. Ann Surg. 1994;219:281–90.
20. Kouchoukos NT, Ebert PA, Grover FL, Lindesmith GG. Report of the ad hoc committee on risk factors for coronary artery bypass surgery. Ann Thorac Surg. 1988;45:348–9.
21. Clark RE. The development of The Society of Thoracic Surgeons voluntary national database system: genesis, issues, growth, and status. Best Pract Benchmarking Healthc. 1996;1:62–9.
22. Edwards FH, Clark RE, Schwartz M. Coronary artery bypass grafting: the Society of Thoracic Surgeons

National Database experience. Ann Thorac Surg. 1994;57:12–9.
23. Edwards FH, Grover FL, Shroyer ALW, et al. The Society of Thoracic Surgeons National Cardiac Surgery Database: current risk assessment. Ann Thorac Surg. 1997;63:903–8.
24. Edwards FH. Evolution of the Society of Thoracic Surgeons National Cardiac Surgery Database. J Invasive Cardiol. 1998;10:485–8.
25. Austin PC. A comparison of Bayesian methods for profiling hospital performance. Med Decis Making. 2002;22:163–72.
26. Brennan TA, Leape LL, Laird NM, et al. Incidence of adverse events and negligence in hospitalized patients: results of the Harvard Medical Practice Study I. N Engl J Med. 1991;324:370–6.
27. Kohn LT, Corrigan JM, Donaldson MS. To err is human: building a safer health system. A report from the Committee on Quality of Healthcare in America, Institute of Medicine. Washington, DC: National Academies Press; 1999.
28. Richardson WC, Berwick DM, Bisgard JC, et al. Crossing the quality chasm: a new health system for the 21st century. A report from the Committee on Quality of Healthcare in America, Institute of Medicine. Washington, DC: National Academies Press; 2001.
29. Shahian DM, Edwards FH, Jacobs JP, et al. Public reporting of cardiac surgery performance: part 2 implementation. Ann Thorac Surg. 2011;92:12–23.
30. O'Brien SM, Shahian DM, Delong ER, et al. Quality measurement in adult cardiac surgery: part 2 – statistical considerations in composite measure scoring and provider rating. Ann Thorac Surg. 2007;83(4 suppl):S13–26.
31. Jacobs JP, Edwards FH, Shahian DM, et al. Successful linking of The Society of Thoracic Surgeons adult cardiac surgery database to Centers for Medicare and Medicaid Services Medicare data. Ann Thorac Surg. 2010;90:1150–7.
32. Badhwar V, Peterson ED, Jacobs JP, et al. Longitudinal outcome of isolated mitral repair in older patients: results from 14,604 procedures performed from 1991 to 2007. Ann Thorac Surg. 2012;94:1870–7.
33. Jacobs JP, O'Brien SM, Shahian DM, et al. Successful linking of the Society of Thoracic Surgeons Database to Social Security data to examine the accuracy of Society of Thoracic Surgeons mortality data. J Thorac Cardiovasc Surg. 2013;145:976–83.
34. Kennedy I. Learning from Bristol: the report of the public inquiry into children's heart surgery at the Bristol Royal Infirmary 1984–1995. http://www.bristol-inquiry.org.uk/. Accessed 21 May 2005.
35. Sherry A. Children's Hospital cardiology chief told to resign. Denver Post. Article Published: 1 Mar 2001. http://www.denverpost.com/news/news0301b.htm. Accessed 21 Mar 2001.
36. Sherry A. Hospitals shield mortality rates. Denver Post. Article Published: 2 Mar 2001. http://www.denverpost.com/news/news0302d.htm. Accessed 21 Mar 2001.
37. The Denver Post Editorial Board. At the heart of the problem. Denver Post. Article Published: 2 Mar 2001. http://www.denverpost.com/opinion/edits0302c.htm. Accessed 21 Mar 2001.
38. Hernandez J. Other options. Denver Post. Article Published: 3 Mar 2001. http://www.denverpost.com/opinion/lett0311.htm. Accessed 21 Mar 2001.
39. Johnson L. Baby's death at Children's turns parents to their faith. Denver Post. Article Published: 3 Mar 2001. http://www.denverpost.com/opinion/lett0311.htm. Accessed 21 Mar 2001.
40. White S. Kids' best interests: re: "Children's Hospital cardiology chief told to resign," March 1. Denver Post. Article Published: 3 Mar 2001. http://www.denverpost.com/opinion/lett0311.htm. Accessed 21 Mar 2001.
41. Weinberg S. Rare look inside a surgeon's sanctum. Denver Post. Article Published: Sunday, 20 Apr 2003. http://www.denverpost.com/Stories/0%2C1413%2C36, 28, 1333663%2C00.html. Accessed 22 Oct 2004.
42. The report of the Manitoba Pediatric Cardiac Surgery Inquest: an inquest into twelve deaths at the Winnipeg Health Sciences Centre in 1994. Available at: http://www.pediatriccardiacinquest.mb.ca/pdf/pcir_intro.pdf. Accessed 12 June 2014.
43. Hudson W, Cohen E. After CNN investigates babies' deaths, hospital releases mortality data [http://edition.cnn.com/2013/08/12/health/kentucky-children-update/index.html]. Accessed 6 Nov 2013.
44. Jacobs JP, Jacobs ML, Mavroudis C, et al. Nomenclature and databases for the surgical treatment of congenital cardiac disease – an updated primer and an analysis for opportunities for improvement. Cardiol Young. 2008;18:38–62.
45. Jacobs JP, Jacobs ML, Austin III EH, et al. Quality measures for congenital and pediatric cardiac surgery. World J Pediatric Congenit Heart Surg. 2012;3:32–47.
46. Jacobs JP, O'Brien SM, Pasquali SK, et al. Variation in outcomes for benchmark operations: an analysis of the Society of Thoracic Surgeons Congenital Heart Surgery Database. Ann Thorac Surg. 2011;92:2184–92.
47. Jacobs JP, O'Brien SM, Pasquali SK, et al. Variation in outcomes for risk-stratified pediatric cardiac surgical operations: an analysis of the STS Congenital Heart Surgery Database. Ann Thorac Surg. 2012;94:564–71.
48. Jacobs JP, Jacobs ML, Maruszewski B, et al. Initial application in the EACTS and STS Congenital Heart Surgery Databases of an empirically derived methodology of complexity adjustment to evaluate surgical case mix and results. Eur J Cardiothorac Surg. 2012;42:775–80.

37 Communication Chaos: How Incomplete and Conflicting Information Prevents Improved Outcomes for Patients with Pediatric and Congenital Cardiac Disease (and What We Can Do About It)

Debra Hilton-Kamm and Helen Haskell

Abstract

Congenital heart disease (CHD) occurs in approximately 8 of 1,000 births, with approximately one-third requiring invasive treatment or resulting in death within the first year of life. Parents of children with complex CHD need timely and accurate information regarding the diagnosis, treatments, surgical outcomes, and protocols of follow-up to provide the best care for their children. Decision-making at the time of diagnosis is especially critical; hospital choice is a major predictor of survival, yet most parents do not have access to data about surgical outcomes upon which to choose the most appropriate facility for their child. The information received from physicians, the Internet, and other sources is highly variable, unverifiable, and often inaccurate, further complicating the process of decision-making. Despite the limitations and concerns of publicly reporting data about surgical outcomes, we argue that this information must be accessible to parents at diagnosis to ensure true informed consent and to achieve best outcomes. Improved communication throughout the lifetime of the patient is also needed to ensure awareness and treatment of potential co-morbidities associated with CHD. The Internet provides an unprecedented opportunity for improved education of patients and their parents as well as collection of data regarding issues related to complications and quality of life that are not yet well understood in this population. These methods

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of communication and collection of data are critical in achieving the best long-term outcomes for patients with complex congenital heart disease.

Keywords
Congenital heart disease • Publicly reported data • Patient education • Internet • Data collection • Parent survey • Facebook

Introduction

The diagnosis of a child's complex congenital heart disease (CHD) is an emotionally difficult time for parents. While they mourn the loss of their "healthy" child, they must wade through medical jargon, complex treatment options, and probabilities of outcomes, and make decisions that will affect the life of their child and the future of their family. When the diagnosis is made prenatally, parents have more time to research options for treatment and mentally prepare for the birth of their child. At this time, termination of the pregnancy may be considered, depending upon the timing of diagnosis, the beliefs of the family, and applicable laws. Post-natal diagnosis of complex CHD leaves less time for parents to research and make decisions regarding options for treatment, and may result in the mother staying in one hospital recovering from delivery while the infant is transferred to another facility for stabilization and/or surgery. Regardless of the timing of diagnosis, the facility at which the child is born is a major predictor of the child's chance of survival [1]. However, parents are often unaware of the well-documented disparities in surgical outcomes between facilities [2–5].

Limited Publicly Reported Data

The European Association for Cardio-Thoracic Surgery (EACTS) has created a voluntary database that collects information about the outcomes of patients undergoing pediatric and congenital cardiac surgery. The EACTS Congenital Heart Surgery Database consists of 265 congenital heart centers in 36 European countries and 147 centers in 43 countries outside of Europe [6]. Data are publicly available but are geared toward medical professionals with the goal of improving survival and quality of life through comparisons of mortality and morbidity between centers. Several kinds of reports can be generated online detailing surgical parameters, characteristics of patients, and the top 5 centers in a given cohort based upon criteria entered by the user. Center names are excluded; all information is anonymous and confidential [6].

In contrast, the National Institute for Cardiovascular Outcomes Research (NICOR), in the United Kingdom (U.K.), provides data targeted directly to consumers [7]. Using information collected by its Central Cardiac Audit Database (CCAD), NICOR has developed an easily accessible public website, which names each facility and its associated outcomes for patients with CHD. This CHD portal is an attempt to ensure transparency of data to avoid another incident such as the "Bristol Inquiry," in which higher than expected mortality rates were discovered at an English hospital [8, 9]. According to the CCAD website: "As a result of the Bristol Inquiry, which concluded that up to 35 babies died unnecessarily following referral to the Bristol Royal Infirmary, all paediatric cardiac centres in England and Scotland participate in the congenital heart disease audit" [10]. The Internet portal allows parents to view information by hospital name, including volume and rates of mortality stratified by procedure. An explanation of the non-risk adjusted mortality rates is provided, alerting parents that variation in outcome may be due to case mix (more high-risk or low-risk patients in comparison to the profile of risk in the aggregate population of patients), or random

variation and fluctuation [10]. Critics argue that, "Without risk adjustment, the publication of raw mortality rates of individual centres is meaningless, of no use to the public, and useless as a quality instrument for the professionals" [8]. Other surgeons, however, believe that parents will not use this data in lieu of physician consultations, but as a framework for discussion with physicians about the most appropriate location of care for their child [8].

In the United States of America (U.S.A.), public reporting of multi-institutional data about outcomes of patients with congenital heart disease data is currently limited to the legally mandated reporting by the state of New York which has provided volume and mortality data by procedure since 2004, based upon data beginning in 1997 [11]. Parents accessing this statewide data are unable to make comparisons to facilities elsewhere in the U.S.A. However, this situation will soon change. The Society of Thoracic Surgeons (STS) Congenital Heart Surgery Database is the largest clinical database in the world that tracks outcomes of patients undergoing pediatric and congenital cardiac surgery. The Spring 2014 STS Congenital Heart Surgery Database Report contains data from 119 of the 125 hospitals (95.2 % penetrance by hospital) in the U.S.A. that perform pediatric and congenital cardiac surgery and 3 of the 8 centers in Canada that perform pediatric and congenital cardiac surgery. In late 2014, the STS Congenital Heart Surgery Database is slated to begin voluntary public reporting of outcomes of patients undergoing pediatric and congenital cardiac surgery.

Current Sources of Information for Parents

In the absence of publicly reported data on congenital heart disease, parents seek information from a variety of sources, the content, scope, and accuracy of which is widely variable and unverifiable. The following are examples of sources of information parents may access at diagnosis and the limitations of each.

Physicians

Parents may receive the diagnosis from obstetricians, pediatricians, perinatologists, or pediatric cardiologists, either prenatally or after birth. The broad range of CHDs, and relatively low incidence of complex congenital cardiac conditions, make it difficult for all physicians to have in-depth knowledge of treatments and current rates of survival for each. When asked to rate educational topics in prenatal and neonatal counseling, parents rated almost all topics as more important than cardiologists did. Topics included the ability to name and describe the heart condition and understanding of medications, long-term complications, and exercise limitations, among others. Obtaining more information on CHD via websites, written information, and referrals to other families was also rated significantly higher in importance by parents than by cardiologists. These data suggest that parents view patient education differently than physicians do, and prefer more information at diagnosis than cardiologists provide [12].

Accurate information on treatments is especially important for complex heart conditions that need immediate intervention. One of the most severe forms of CHD is hypoplastic left heart syndrome (HLHS) in which the left side of the heart is underdeveloped, leading to death if surgical intervention is not performed within the first week of life [13]. Some parents report feeling pressured by pediatric cardiologists to terminate their pregnancies when receiving the diagnosis of HLHS prenatally [14]. Treatment options include: 3-stage palliation [13, 15–19], infant heart transplantation [20], hybrid procedure [21], or fetal intervention [22]. Despite vastly improved rates of survival [13], nonintervention, or "comfort care," which results in the death of the child within days or weeks of birth, is also sometimes offered for babies with HLHS [23]. Debate continues as to whether nonintervention, which is usually not offered for any other CHD, should still be offered to parents of babies with HLHS, in light of the significant improvements in outcome for patients with HLHS [24, 25].

Parents may not always receive timely and accurate information on surgical options available.

Karamlou et al. found that neonates with HLHS were less likely to undergo surgical intervention if admitted to a non-interventional or rural hospital, or to a facility that performs a lower volume of complex congenital heart surgeries. However, neonates admitted to interventional hospitals generally undergo either Norwood staged palliation or heart transplantation [1]. The study concluded, "Despite improved surgical outcome, the majority of infants continue to receive no surgical care" [1]. It is unknown how many parents at non-interventional, rural, or low-volume facilities are made aware that life-saving surgical treatments are available at other facilities.

"Why Didn't the Doctor Tell Me About the Surgeries?"

After learning of teenage and older survivors of HLHS, a parent emailed the HLHS Information Page website expressing shock and anger that surgical options were not presented to her. She had "chosen" non-intervention for her infant who passed away shortly after birth, after physicians told her that nothing could be done for her child. Learning of the options after the fact led to feelings of guilt for trusting the information given by physicians and not seeking a second opinion (Hilton-Kamm D, email correspondence Feb 2003).

Even when learning of the availability of surgical intervention, parents have little way of knowing the large disparity between hospitals in rates of survival after surgery for pediatric and congenital cardiac disease. A facility can claim a 100 % survival rate if it has completed only one surgery and that patient survived for 30 days. In the absence of comparative data, and without context, this information can be grossly misleading to parents making life-altering decisions for their children.

Significant variation in outcomes between centers is most prominent for the more complex operations [26]. Several studies have shown that lower mortality rates correlate with higher surgical volume of the Norwood procedure [2, 3, 26]; some indicate that survival rates at higher volume institutions can be more than double those at lower volume centers [4, 5]. This information is vital to parental decision-making. However, a nationwide survey of parents of children with CHD in the U.S.A. found that of 841 parents, only 16 % received information on success rates at different hospitals [27]. When asked what else would have been helpful at diagnosis, several respondents mentioned learning of outcomes at different facilities:

> It would also have been helpful to receive stats on the best hospitals and surgeons in the country for my son's particular heart defect. Parents should know they have options. It shouldn't be a given that you automatically go to the closest hospital/surgeon just b/c of proximity. Parents need to be given information, choices and some control over their child's care.

> Knowing where the best centers were located and websites. When at (*hospital name removed*) they never mentioned the amazing outcomes of (*hospital name removed*). Providers should give out the info that would best help the patient.

Institutional and physician bias has also been documented in several studies. Kon et al. found that recommendations about treatment were not associated with the belief of the physician regarding the best outcome, but instead with the procedure most commonly performed at the hospital affiliated with the physician [23]. Prsa and colleagues found that physicians did not relate all options for treatment of HLHS to parents, and that cardiologists were more likely to recommend surgical intervention if they practiced at an active cardiac surgery program, estimated higher post-operative survival, or believed they would choose surgery for their own child [28].

The decisions of parents to terminate the pregnancy or choose non-intervention rely heavily upon estimates of the survival and projected quality of life of their child. Some predict that 70 % of newborns born today with HLHS may reach adulthood [13]. However, one study of pediatric cardiologists at the largest 14 cardiac centers in the U.S.A. found that estimates of individual physicians of 1-year survival for the 3-stage surgeries and heart transplantation ranged from 30 to 88 % and 5 to 90 %, respectively [23]. Arya and

colleagues found that parents rated quality of life as the most important determinant in considering termination of the pregnancy [12]. However, physicians may have different and more negative views of quality of life for survivors of CHD than parents and the patients themselves. Marino and colleagues found that health care providers focused mainly on in-hospital issues (fear of procedures, separation anxiety) while parents focused on broader quality of life issues that were not just hospital-related. Adolescent patients and parents also attributed positive factors to the CHD, such as being a "stronger person" or "more compassionate," whereas health care providers focused more on the negative aspects of management of chronic disease [29]. Therefore, how survival and quality of life issues are presented to parents may greatly impact decision-making at diagnosis. Physicians may also inadvertently give parents the impression of a negative prognosis by the terminology they use in counseling. A nationwide survey of parents of children with CHD in the U.S.A. [27] found that of 841 parents who responded, two-thirds were told the CHD was "rare" which was interpreted to mean

- "few or no other people alive with this defect" by 27 % of respondents, and
- "occurring in less than a million births" by 25 %.

Internet Resources

In 2002, Ikemba and colleagues found that more than half of parents with access to the Internet researched information about CHD on the Internet, and that the majority of those who did found the information helpful and easy to obtain [30]. It is likely that the number of parents using the Internet has increased substantially since the publication of this study over a decade ago. Internet searches of CHD may result in

- medical websites,
- webpages of individual hospitals,
- hospital or medical advertising,
- support networks,
- personal webpages and blogs of other parents, and even
- law firms recruiting clients for malpractice suits.

While some physicians warn parents against researching the Internet due to the possibility of finding inaccurate information, some parents find more detailed information online than from the physician, according to open-ended responses from a nationwide survey [14]:

> Everything discussed around HLHS was negative and made us feel like we had no choice other than termination. Only after I googled on my cell phone while waiting did I determine that people actually can survive HLHS.

> The primary doctors who first diagnosed the condition did not give termination as an option; he told us we had no other choice. We found out that night through Internet searches that there were other options…

Parents seeking information about outcomes may have a difficult time finding data or comparing facilities due to differing ways in which data are presented. Hospital websites that offer statistics may

- present data annually or aggregated for several years,
- use risk-adjusted or non risk-adjusted rates of mortality, and/or
- include data by stratified by procedure or aggregated to include the total number of operations.

Parents accessing peer-reviewed medical journal articles may only have access to the free abstracts, which do not include the totality of the study. Without context regarding when the study was completed, where it was performed, and which populations of patients were included, the results may be misleading to parents. Parents may also not be aware that journal publications may be more heavily weighted towards major teaching institutions and may not be representative of all hospitals.

Other Parents

Parents receiving a diagnosis may seek support groups or referrals to other families by social workers or physicians to learn of first-hand

experiences. In addition to providing emotional support and empathy to parents going through similar situations, other parents are also able to share practical tips, information on resources, and issues to discuss with physicians. Those raising a child with CHD may give newly diagnosed parents different perspectives than medical professionals. One concern when connecting parents to others is ensuring that parents understand that the experience and outcome of one child is not necessarily predictive of the experience and outcome of another child. Individual webpages and posts on Facebook, CaringBridge, CarePages, or personal blogs may provide some parents with false hope if their child has a more severe condition than the child written about. Conversely, these anecdotal accounts may dash the hopes of some if they read about children with more severe CHD, those with major complications, or those who have died. Some of these personal sites may also unintentionally provide misinformation, misleading parents as to the nature of the condition, treatments, or prognosis for their child.

Many organized support groups have been created by parents, in part due to the lack of a national organization focused solely on CHD. These groups may have in-person meetings or, more commonly, online listservs. The main advantage of online groups is the ability to access information and support at any time of day or night from others who have similar diagnoses. One posting by the parent of a newly diagnosed child can result in multiple responses within hours from parents across the nation, or even from other countries, allowing for diverse perspectives and a broad range of outcomes. Parents often seek information on surgical facilities in an effort to compare hospitals and choose an appropriate facility. Support groups often include discussions of feeding issues, developmental delays, medications, and many other vital topics that benefit all who attend meetings, or read online postings. Parent-created websites such as the HLHS Information Page [31] and CHDResources [32] (created by the co-author) are dedicated to improving outcomes through facilitating communication between parents and physicians. These websites provide questions for parents to ask physicians and insurance companies to help find the most suitable surgical facility, tips on how to prepare for surgery, descriptions of medical specialists who may work with the child, and other vital information for parents (See Table 37.1). Parents who do not have Internet access or who do not actively seek information online may not benefit from these resources and differing perspectives.

Table 37.1 Selected patient educational materials for parents of children with congenital heart disease

Document	Description
Resource guide	Information for parents on finding medical information and insurance/financial information, applying for Early Intervention services, obtaining 504 plans and Individual Education Plans (IEP's) through the school system, and more
What to ask the surgeon and hospital	A list of questions to help compare hospital facilities, services, and outcomes
What to ask the insurance company and common insurance terms	Important terms to better understand coverage and costs with a list of questions to determine specific benefits, restrictions, and out-of-pocket costs
Internet resources	Selected online resources for heart diagrams and information on specific diagnoses
Preparing for your child's hospital stay	Tips and suggestions for preparing for surgery
Medical specialists	A list and description of different medical specialists and questions for each
Medication tips	Useful tips on administering medications properly
Emergency information form	Information every parent should have available in case of emergency with space to fill in important information
When to call the doctor form	Form to be filled out by physician which delineates which symptoms warrant a call to the doctor, or a trip to the ER
Follow-up care form	Timeline for follow-up visits, vaccinations, therapies, etc
Physical activity form	Form to clarify appropriate activities for the child with CHD for parents, schools, and other caretakers

All handouts and forms are downloadable for free from www.CHDResources.org

Insurance Providers

When faced with a diagnosis of CHD, parents often consult with their health insurance company to learn if they have a choice of facilities at which to deliver the baby and/or have surgical intervention. United Health Care, through OptumHealth, and Aetna have identified hospitals that achieve better outcomes through lower mortality and morbidity [33, 34]. These "Centers of Excellence" consist of facilities that meet certain criteria and are willing to contract for lower reimbursement rates. Parents are then urged by the insurance company to utilize those contracted facilities for surgical treatments. Parents, however, may not be privy to the methodology used to determine which facilities are included, and may be unaware of the financial incentive of the insurance company to refer patients to these contracted facilities. In addition, parents may not learn of facilities outside the insurance network that might be comparable or better suited to the cardiac condition of their child.

Magazine Rankings

Parents may also encounter published rankings of hospitals in their search for outcomes data. Parents Magazine publishes the "10 Best Children's Hospitals for Heart Care" with no methodology included on the website for how these facilities were chosen. The site merely states, "Survival rates for tricky heart surgeries, experience in treating heart problems, and a robust research program were among the factors that helped put these hospitals on top" [35]. In contrast, the list of "Best Children's Hospitals" by U.S. News & World Report details its complex scoring system in a 98-page online methodology report [36]. Submission of hospital data is voluntary; 98 of 178 eligible hospitals provided data to be considered for the 2012–2013 rankings. To achieve maximum points, a pediatric cardiology and cardiac surgical facility must perform the Norwood procedure and heart transplantation, and have a program for adults with congenital heart disease [37]. These criteria may not be applicable to parents whose children have congenital anomalies other than HLHS or who do not require heart transplantation. It is also unlikely that families consider a program for adults with congenital heart disease as a major factor in their decision-making when choosing a hospital for surgery for their infant. These rankings create the impression that there is one "best" hospital for all pediatric patients needing cardiac surgery. This impression may result in some parents seeking care for less complex conditions at these high volume facilities when another hospital could provide comparable services and outcomes. In 2012, US News & World Report reported that a programming error affected some scores of hospitals, leading to a mis-ranking of facilities [37]. Hospital advertising is promoted on the US News & World Report site - even within the rankings – which can be misleading to parents seeking objective information. Many hospitals with high rankings on these lists display that information on their individual institutional websites to promote their cardiology and cardiac surgical services [38–41].

Publicly Reported Data: Obstacles and Benefits

It is clear that the current information available to parents is often subjective, incomplete, or inaccurate. Depending upon where a child with CHD is born or diagnosed, the prognosis and surgical results may vary considerably. Therefore, the basic tenet of informed consent – understanding the risks and benefits of all treatment options – is currently non-existent for many parents of children with complex CHD [42]. Public reporting of surgical outcomes for patients with pediatric and congenital cardiac disease could change the course of decision-making for many families and alter the lives of their children. It could reduce the chance that parents only learn of surgical options after it is too late. However, several obstacles have prevented that information from being collected and disseminated to parents.

Availability and Verification of Data

Unlike the government-mandated collection of data about CHD in the U.K., there is no mandated, centralized source of data about CHD in the U.S.A. As of 2011, the Society of Thoracic Surgeons (STS) Congenital Heart Surgery Database collected voluntarily submitted data from 96 of the 122 hospitals in the U.S.A., providing the most complete data set of surgical outcomes for the U.S.A. [26]. By Spring 2014, as noted above, the STS Congenital Heart Surgery Database Report contained data from 119 of the 125 hospitals in the U.S.A. and 3 of the 8 Canadian centers that perform pediatric and congenital cardiac surgery. The STS data is intrinsically verified for completeness and accuracy and is randomly audited by an independent medical audit firm.

In 2011, STS reached an agreement with Consumer Reports magazine to publicly report outcomes for isolated coronary artery bypass graft (CABG) surgery in adults, for surgical groups that agreed to do so [43]. Other STS data currently remains confidential, and can only be publicly reported with specific approval from the surgeon or program in question [44]. However, STS is committed to expansion of its program of public reporting of cardiothoracic surgical outcomes. The STS has now begun to publicly report outcomes for isolated aortic valve replacement (AVR) in adults, as well as combined aortic valve replacement (AVR) and coronary artery bypass graft (CABG) surgery in adults. Plans are underway to publicly report outcomes for isolated mitral valve repair and replacement in adults, as well as for lobectomy. In late 2014, the STS Congenital Heart Surgery Database plans to begin voluntary public reporting of outcomes of patients undergoing pediatric and congenital cardiac surgery. STS is actively making efforts to expand its portfolio of publicly reported measures for all three of its databases:

- STS Adult Cardiac Surgery Database
- STS general Thoracic Surgery Database
- STS Congenital Heart Surgery Database.

Mortality Data

A major concern of publicly reporting outcomes about congenital heart disease is the possibility of misinterpretation of data about mortality. Some believe that many parents will simply choose hospitals with the lowest rates of mortality, favoring low volume centers with fewer high-risk patients and lower unadjusted rates of raw mortality [44]. However, many high volume centers with a significant portion of high-risk patients currently voluntarily publicize their rates of mortality online, apparently without fear of losing patients to lower volume centers. Many models for reporting data about mortality already exist, including the CCAD congenital heart disease database in the U.K. [7]. The funnel plots utilized on the CCAD data portal prevent patients "from making erroneous decisions in favor of surgeons and institutions with randomly low mortality rates" [44]. These funnel plots include warning limits to alert patients and providers if rates of mortality increase beyond an expected threshold [7].

The STS model of public reporting of data about isolated coronary artery bypass graft (CABG) surgery, as well as isolated aortic valve replacement (AVR) and combined aortic valve replacement (AVR) and coronary artery bypass graft (CABG) surgery, uses a star system to indicate surgical groups that are at, below, or above average on certain variables, including mortality. However, the star rating discourages direct comparisons of hospitals. The system is voluntary and in the first year of reporting included approximately one third of the groups who submit data to STS [43]. It is available for free on the STS website and to paying subscribers of Consumer Reports [45, 46]. More detailed CABG outcome data are available in some states (e.g., California [47], Massachusetts [48], New Jersey [49], New York [50], and Pennsylvania [51],) as well as through a report of the 50 Top Cardiovascular Hospitals based upon Medicare administrative claims data [52].

Another model is the data published by the Scientific Registry of Transplant Recipients (SRTR) on adult and pediatric organ transplantation, perhaps the most detailed publicly reported data in the

U.S.A. Parents and patients can view the activity of individual centers, including waitlist times, transplant volume, and post-transplant outcomes with rates of mortality [53]. As described elsewhere in this book (Chap. 15 by Ryan R. Davies, MD), the dataset that is commonly referred to as the United Network for Organ Sharing (UNOS) Database consists of information collected by UNOS and maintained and analyzed by the Scientific Registry of Transplant Recipients (SRTR).

Access to Care: Parental Considerations

Parental decision-making is a complex process with many factors. Insurance, work limitations, logistics, and caring for other children all play an important role in deciding where to have treatment. Therefore, it is unlikely that public reporting of data would result in mass migration to centers with lower published rates of mortality. Parents expecting a new baby often want to be near family and friends, especially when the child has a severe medical condition. Some parents may choose a local environment and familiar physicians for reasons of emotional stability and comfort, despite published data of lower rates of mortality elsewhere [54].

Some researchers are concerned that access to care would be reduced if hospitals refused to accept higher risk patients in order to maintain lower published rates of mortality [44]. Parents intuitively understand that volume equals experience, however, and context can be created by providing data about volume along with data about mortality. The publishing of risk-adjusted outcomes would provide comparisons of outcomes adjusted for case mix and complexity, data that parents do not currently have at their disposal. While it is possible that some lower volume centers may decide to refer high-risk cases to more experienced centers, such referrals could actually be a desirable outcome that could potentially lead to improved survival [2, 55]. Public reporting could increase access to care for children born at low volume centers or in rural areas, by making information on the larger universe of surgical facilities available to parents at the time of their decision-making.

Quality Improvement

Public reporting of CHD data could result in quality improvement, as was reported for data about CABG in New York, when mortality decreased by 41 % over a 4-year period [56]. For data about congenital heart disease, improvements in quality could be achieved "…by identifying the best providers so that we could learn from them, and by identifying low performing centers that can benefit from quality improvement initiatives" [44]. STS has already identified low performing "outliers" with statistically significant lower rates of survival than the national average [26]. To date, however, these data are not available to parents, who unknowingly "choose" these hospitals for surgical treatment of their children. However, as described earlier in this chapter, in late 2014, the STS Congenital Heart Surgery Database will begin voluntary public reporting of outcomes of outcomes of patients undergoing pediatric and congenital cardiac surgery.

The lack of mandated public reporting, then, primarily protects

- low volume centers from reporting their lack of experience, and
- poor performing outlier facilities from reporting their higher rates of mortality.

Transparency of information would send the signal that the medical community values informed parental choice over the protection of the reputations of hospitals. Some physicians understand the importance of providing these data and advocate that physicians take an active role in developing standards of public reporting "…because doing so is required by our most fundamental professional commitment: to do what is best for our patients" [44].

The Future: Improved Communication for Better Outcomes

Improved rates of survival have led to an increasing number of children and adults with congenital heart disease. Some estimate that there are currently over one million adult survivors of

CHD in the U.S.A. alone [57]. This growing population will require ongoing medical care and continuing communication with physicians to achieve optimal outcomes; however, current information on CHD is fragmented and often contradictory, causing confusion and frustration among parents and adult patients. Protocols have been developed for

- routine follow-up for those with complex CHD [58] and
- neurodevelopmental surveillance, screening, and evaluation [59].

These protocols, however, have yet to be widely adopted by physicians and hospitals and are often unknown to parents and patients. Cooperation among hospitals, parental networks of support, and other organizations is necessary to ensure that consistent and accurate information is disseminated to those with CHD. The creation of a collaborative between these entities could lead to improved flow of data to and from parents and/or patients, resulting in improved outcomes (Fig. 37.1).

Multiple studies [60–68] have shown gaps in parental knowledge, including inability to accurately name or explain the diagnosis of their child or to exhibit understanding of:

- infective endocarditis,
- respiratory syncytial virus (RSV),
- use of medications,
- physical limitations,
- symptoms of deterioration, and
- the importance of life-long cardiac follow-up.

This lack of knowledge of co-morbidities and complications may lead to inadequate medical follow-up, and may negatively impact outcomes for children with CHD. Some parents have reported that they did not receive information from healthcare providers regarding the need for life-long cardiac follow-up [64]. Other studies have found that patients with CHD and/or their parents have other unmet informational needs [12, 69]. Studying the content, scope and effectiveness of current parent/patient communication would be an important step toward developing better and more effective tools of communication.

Improved communication regarding the potential for developmental delays and neurological deficits and how these issues can affect the child's physical, academic, social, and emotional well-being is vitally needed. Parents need to understand the importance of Early Intervention services for infants or toddlers with physical delays. Through the school system, parents can advocate for their child by creating 504 Plans for classroom accommodations and Individualized Education Plans (IEPs) in which educational services are provided [70]. Children with complex CHD need their specific physical limitations and emergency protocols to be communicated effectively and accurately to the staff at their schools and other caregivers, while ensuring that the child receives appropriate levels of physical activity. For these children to thrive in the school environment and live up to their full potential, the medical community must understand these ramifications of complex CHD and effectively communicate to parents how to get the most appropriate help.

The Internet has created an unprecedented opportunity to educate parents and patients regardless of which facility they utilize, or even if they are not currently receiving medical care at all. Lesch and colleagues found that among patients aged 10–30, use of the Internet rose with the age group, with 52 % of young adult patients accessing the Internet for information specific to CHD [69]. The Internet can also be used to gather valuable data *from* survivors with CHD to help in the creation of protocols or services that would be beneficial to this population. The low incidence of complex cardiac conditions and specific complications such as Plastic Bronchitis (PB) and Protein Losing Enteropathy (PLE) necessitates multi-institutional research. Surveys using the Internet can be effective in gathering information from large numbers of respondents [14, 69], enabling the medical community to learn directly from those living with congenital heart disease.

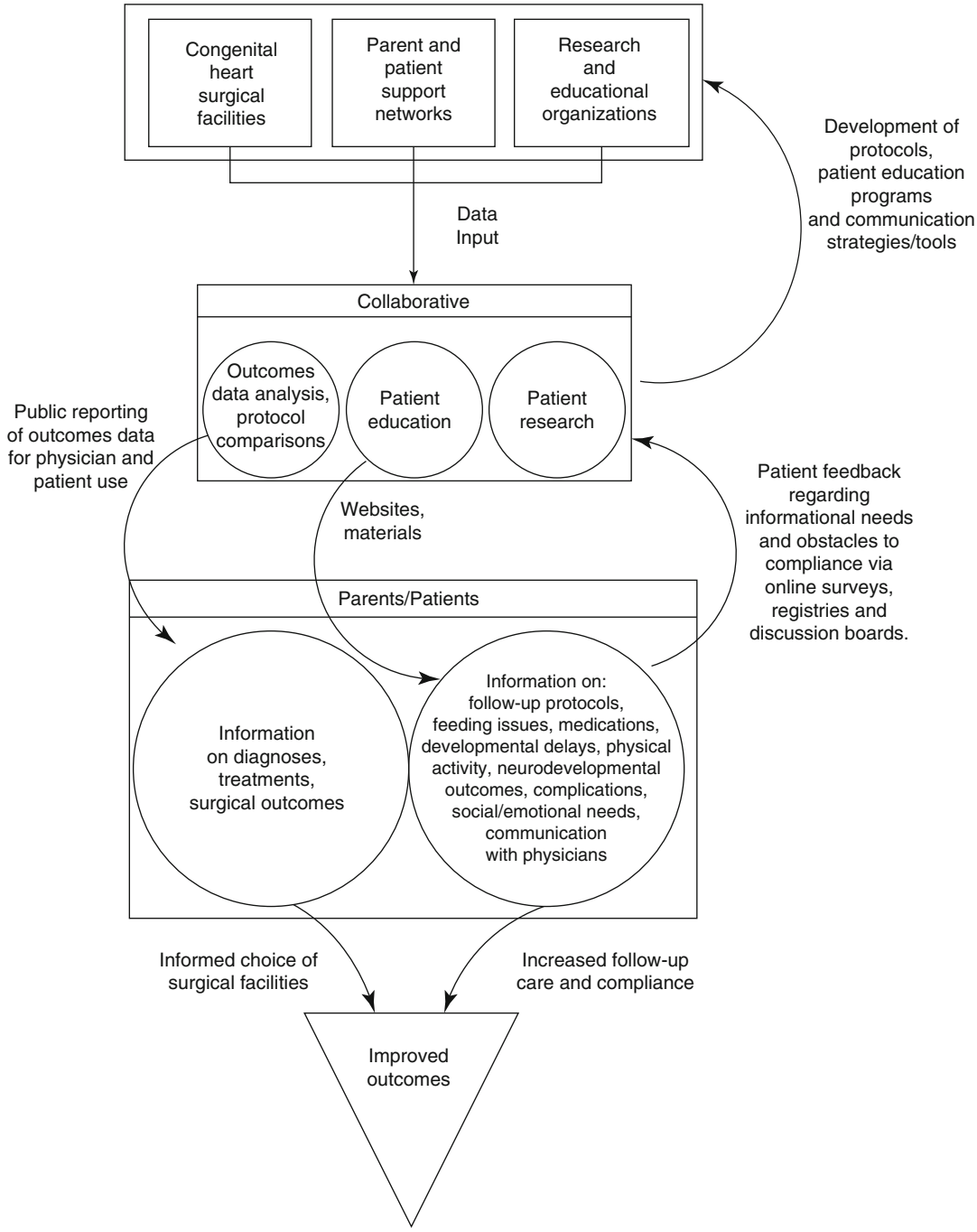

Fig. 37.1 Proposed flow of data for improved outcomes for those with congenital heart disease. Surgical facilities, parent/patient support networks and research and educational organizations provide data to a "collaborative." This centralized source analyzes data and disseminates information to parents and patients to improve communication. Feedback from patients is also gathered and analyzed to help develop protocols and create more effective strategies of communication for use by the surgical facilities, support networks and other organizations (© 2013 Debra Hilton-Kamm)

Conclusion

Advances in surgery, treatments, and availability of resources have created new opportunities for many patients and families affected by CHD. However, the disparity of information given at diagnosis results in variation in survival depending upon where a child is born and what information parents receive. While physicians are the primary source of information, recommendations for treatment options vary greatly, and can be biased towards treatments offered at the facility of the physician. Parents, therefore, are often given limited comparative information on rates of survival and treatments. With no centralized source of data about surgical outcomes, parents often seek additional information from unverified and potentially misleading sources, including the Internet, insurance providers, magazine rankings, and other parents via support networks. To create a level playing field, all parents need access to accurate and verifiable data about outcomes for all surgical facilities. Without this comparative data, parents make life and death decisions for their children without true informed consent.

As rates of survival increase, parents and older patients with CHD also have a critical and growing need for greater access to information at all stages of life. Parents need to understand

- the diagnosis,
- risks to the health of their child, and
- the necessity for life-long follow-up.

Improved communication with parents can reduce the risk of infective endocarditis and respiratory syncytial virus (RSV), and lessen the chance of errors involving medications and loss to follow-up care. Parents also need to obtain timely and accurate information on possible developmental delays, behavioral issues, and neurodevelopmental outcomes that can negatively impact the academic, social, and emotional well-being of their child. Finally, a more concerted effort to learn from parents and patients about their experiences can help further develop improved approaches to families affected by CHD. Utilizing the Internet allows for greater access to large numbers of respondents across multiple hospitals, helping to identify and ultimately treat complications that affect both large and small numbers of patients. Improved communication is not only possible, but necessary to improve the long-term outlook for the growing population of survivors with CHD.

References

1. Karamlou T, Diggs BS, Ungerleider RM, Welke KF. Evolution of treatment options and outcomes for hypoplastic left heart syndrome over an 18-year period. J Thorac Cardiovasc Surg. 2010;139(1):119–26. doi:10.1016/j.jtcvs.2009.04.061; discussion 126–7. Epub 2009 Nov 11.
2. Chang RK, Klitzner TS. Can regionalization decrease the number of deaths for children who undergo cardiac surgery? A theoretical analysis. Pediatrics. 2002;109(2):173–81.
3. Checchia PA, McCollegan J, Daher N, Kolovos N, Levy F, Markovitz B. The effect of surgical case volume on outcome after the Norwood procedure. J Thorac Cardiovasc Surg. 2005;129:754–9.
4. Hirsch JC, Gurney JG, Donohue JE, Gebremariam A, Bove EL, Ohye RG. Hospital mortality for Norwood and arterial switch operations as a function of institutional volume. Pediatr Cardiol. 2008;29(4):713–7. Epub 2007 Dec 14.
5. McHugh KE, Hillman DG, Gurka MJ, Gutgesell HP. Three-stage palliation of hypoplastic left heart syndrome in the University HealthSystem Consortium. Congenit Heart Dis. 2010;5(1):8–15.
6. European Association for Cardio-Thoracic Surgery (EACTS) Congenital Database. http://www.eactscongenitaldb.org. Accessed 25 May 2013.
7. Congenital Heart Disease Website; National Institute for Cardiovascular Outcomes Research (NICOR). https://nicor4.nicor.org.uk/CHD/an_paeds.nsf/vwContent/home?Opendocument. Accessed 25 May 2013.
8. Nicholls M. UK congenital heart centres under scrutiny. Circulation. 2007;116:F73–8. doi: 10.1161/CIRCULATION AHA.107.186287. http://circ.ahajournals.org/content/116/13/F73.full.pdf. Accessed 25 May 2013
9. The National Archives; The inquiry into the management of care of children receiving complex heart surgery at the Bristol Royal Infirmary. In: Learning from Bristol: the report of the public inquiry into children's heart surgery at the Bristol Royal Infirmary 1984–1995. Bristol Royal Infirmary Inquiry. 2001; http://webarchive.nationalarchives.gov.uk/20090811143745/http://www.bristol-inquiry.org.uk/index.htm. Accessed 25 May 2013.
10. University College London; National Institute for Cardiovascular Outcomes Research (NICOR)

Background. http://www.ucl.ac.uk/nicor/audits/congenitalheartdisease/background. Accessed 25 May 2013.
11. New York State Department of Health. State Health Department Releases reports on cardiac surgery: New York first in nation to release heart valve and pediatric cardiac surgery data. 6 May 2004. http://www.health.ny.gov/press/releases/2004/cardiac_release_05-06-2004.htm. Accessed 25 May 2013.
12. Arya B, Glickstein S, Levasseur SM, Williams IA. Parents of children with congenital heart disease prefer more information than cardiologists provide. Congenit Heart Dis. 2012. doi:10.1111/j.1747-0803.2012.00706.x [Epub ahead of print].
13. Feinstein JA, Benson DW, Dubin AM, Cohen MS, Maxey DM, Mahle WT, Pahl E, Villafañe J, Bhatt AB, Peng LF, Johnson BA, Marsden AL, Daniels CJ, Rudd NA, Caldarone CA, Mussatto KA, Morales DL, Ivy DD, Gaynor JW, Tweddell JS, Deal BJ, Furck AK, Rosenthal GL, Ohye RG, Ghanayem NS, Cheatham JP, Tworetzky W, Martin GR. Hypoplastic left heart syndrome: current considerations and expectations. J Am Coll Cardiol. 2012;59(1 Suppl):S1–42.
14. Hilton-Kamm D, Chang RK, Sklansky M. Prenatal diagnosis of hypoplastic left heart syndrome: impact of counseling patterns on parental perceptions and decisions regarding termination of pregnancy. Pediatr Cardiol. 2012;33(8):1402–10. doi:10.1007/s00246-012-0366-9.
15. Bove EL. Current status of staged reconstruction for hypoplastic left heart syndrome. Pediatr Cardiol. 1998;19:308–15.
16. Gaynor JW, Bridges ND, Cohen MI, Mahle WT, Decampli WM, Steven JM, Nicolson SC, Spray TL. Predictors of outcome after the Fontan operation: is hypoplastic left heart syndrome still a risk factor? J Thorac Cardiovasc Surg. 2002;123(2):237–45.
17. Mahle WT, Spray TL, Wernovsky G, Gaynor JW, Clark BJ. Survival after reconstructive surgery for hypoplastic left heart syndrome: a 15-year experience from a single institution. Circulation. 2000;102(19 Suppl 3):III136–41.
18. Mitchell ME, Ittenbach RF, Gaynor JW, Wernovsky G, Nicolson S, Spray TL. Intermediate outcomes after the Fontan procedure in the current era. J Thorac Cardiovasc Surg. 2006;131(1):172–80.
19. Tweddell JS, Hoffman GM, Mussatto KA, Fedderly RT, Berger S, Jaquiss RD, Ghanayem NS, Frisbee SJ, Litwin SB. Improved survival of patients undergoing palliation of hypoplastic left heart syndrome: lessons learned from 115 consecutive patients. Circulation. 2002;106:I82–9.
20. Bailey LL. Transplantation is the best treatment for hypoplastic left heart syndrome. Cardiol Young. 2004;14 Suppl 1:109–11.
21. Galantowicz M, Cheatham JP. Lessons learned from the development of a new hybrid strategy for the management of hypoplastic left heart syndrome. Pediatr Cardiol. 2005;26(3):190–9. PMID: 16179977.
22. McElhinney DB, Tworetzky W, Lock JE. Current status of fetal cardiac intervention. Circulation. 2010;121(10):1256–63.
23. Kon AA, Ackerson L, Lo B. How pediatricians counsel parents when no "best-choice" management exists. Arch Pediatr Adolesc Med. 2004;158:436–41.
24. Kon AA. Healthcare providers must offer palliative treatment to parents of neonates with hypoplastic left heart syndrome. Arch Pediatr Adolesc Med. 2008;162(9):844–8.
25. Wernovsky G. The paradigm shift toward surgical intervention for neonates with hypoplastic left heart syndrome. Arch Pediatr Adolesc Med. 2008;162(9):849–54.
26. Jacobs JP, O'Brien SM, Pasquali SK, Jacobs ML, Lacour-Gayet FG, Tchervenkov CI, Austin 3rd EH, Pizarro C, Pourmoghadam KK, Scholl FG, Welke KF, Mavroudis C. Variation in outcomes for benchmark operations: an analysis of the Society of Thoracic Surgeons Congenital Heart Surgery Database. Ann Thorac Surg. 2011;92(6):2184–91. doi:10.1016/j.athoracsur.2011.06.008; discussion 2191–2.
27. Hilton-Kamm D, Sklansky M, Chang RK. How not to tell parents about their child's new diagnosis of congenital heart disease: an Internet survey of 841 parents. Pediatr Cardiol. 2014;35(2):239–52. doi:10.1007/s00246-013-0765-6.
28. Prsa M, Holly CD, Carnevale F, Justino H, Rohlicek C. Attitudes and practices of cardiologists and surgeons who manage HLHS. Pediatrics. 2010;125(3):e625–30. Epub 2010 Feb 15.
29. Marino BS, Tomlinson RS, Drotar D, Claybon ES, Aguirre A, Ittenbach R, Welkom JS, Helfaer MA, Wernovsky G, Shea JA. Quality-of-life concerns differ among patients, parents, and medical providers in children and adolescents with congenital and acquired heart disease. Pediatrics. 2009;123(4):708–15.
30. Ikemba CM, Kozinetz CA, Feltes TF, Fraser CD, McKenzie ED, Shah N, Mott A. Internet use in families with children requiring cardiac surgery for congenital heart disease. Pediatrics. 2002;109(3):419–22. doi:10.1542/peds.109.3.419.
31. HLHS Information Page. www.HLHSinfo.org. Accessed 25 May 2013.
32. CHD Resources. www.CHDResources.org. Accessed 25 May 2013.
33. Optum Health. https://www.myoptumhealthcomplexmedical.com/gateway/public/patients/evaluationAndCostSavings.jsp. Accessed 25 May 2013.
34. Aetna Institutes of Excellence™ Pediatric Congenital Heart Surgery Facilities. http://www.aetna.com/docfind/cms/html/institutes_of_excellence_pediatric.html. Accessed 25 May 2013.
35. Parents Magazine; 10 best children's hospitals for heart care 2013. http://www.parents.com/health/doctors/best-heart-care-hospitals/. Accessed 25 May 2013.
36. U.S. News & World Report Methodology; Best children's hospitals 2012–13. http://www.usnews.com/pubfiles/7-17PedsReport2012_June25.pdf. Accessed 25 May 2013.
37. U.S. News and World Report: Top-ranked pediatric hospitals for cardiology & heart surgery. http://

health.usnews.com/best-hospitals/pediatric-rankings/cardiology-and-heart-surgery. Accessed 25 May 2013.
38. Boston Children's Hospital; #1 ranked pediatric hospital by U.S. news and world report. 2013. http://www.childrenshospital.org/microsites/Site3412/mainpageS3412P0.html. Accessed 25 May 2013.
39. Children's Hospital of Philadelphia; More no. 1 rankings than any other hospital in nation: CHOP leads U.S. News & World Report's best children's hospital list. 2013. http://www.chop.edu/about/best-in-the-nation/. Accessed 25 May 2013.
40. Mott Children's Hospital, University of Michigan; Congenital heart conditions and treatments 2013. http://mottchildren.org/medical-services/ped-heart. Accessed 25 May 2013.
41. Texas Children's Hospital; Heart Center. 2013. http://www.texaschildrens.org/heart/. Accessed 25 May 2013.
42. Hilton-Kamm, D. Hypoplastic left heart syndrome and the myths of informed consent. (Invited commentary) American Academy of Pediatrics Bioethics News Letter (Fall 2012): p. 14–17.
43. Consumer Reports Health and The Society of Thoracic Surgeons. Heart Surgery Ratings Background and Methodology, Consumers Union, August 2011. http://www.consumerreports.org/health/resources/pdf/society-of-thoracic-surgeons/Heart-Surgery-Ratings-Background-and-Methodology.pdf. Accessed 25 May 2013.
44. Jacobs JP, Cerfolio RJ, Sade RM. The ethics of transparency: publication of cardiothoracic surgical outcomes in the lay press. Ann Thorac Surg. 2009;87(3):679–86. doi:10.1016/j.athoracsur.2008.12.043.
45. The Society of Thoracic Surgeons; STS public reporting online: Heart Surgery Outcomes – public access. STS Heart Surgery Outcomes – star ratings avail by hospital or surgical group. http://www.sts.org/quality-research-patient-safety/sts-public-reporting-online. Accessed 25 May 2013.
46. Consumer Reports; Heart bypass surgery ratings. http://www.consumerreports.org/health/doctors-hospitals/surgeon-ratings/ratings-of-bypass-surgeons.htm. Accessed 25 May 2013.
47. State of California; Office of Statewide Planning and Development (OSHPD) Coronary Artery Bypass Graft (CABG) Surgery in California. http://www.oshpd.ca.gov/hid/Products/Clinical_Data/CABG/index.html. Accessed 25 May 2013.
48. Department of Health Care Policy – Harvard Medical School. Reports on risk-standardized mortality rates for hospitals performing coronary artery bypass graft surgery and percutaneous coronary interventions in the Commonwealth of Massachusetts. MASS-DAC Data Analysis Center. http://www.massdac.org/index.php/reports. Accessed 25 May 2013.
49. State of New Jersey Department of Health; Cardiac Surgery. http://www.state.nj.us/health/healthcarequality/cardiacsurgery.shtml. Accessed 25 May 2013.
50. New York State Department of Health: Cardiovascular disease data and statistics. http://www.health.ny.gov/statistics/diseases/cardiovascular/index.htm. Accessed 25 May 2013.
51. Pennsylvania Health Care Cost Containment Council (PHC4) Cardiac Surgery in Pennsylvania 2008–2009. http://www.phc4.org/reports/cabg/09/. Accessed 25 May 2013.
52. Truven Health Analytics; 50 top cardiovascular hospitals. http://www.100tophospitals.com/top-cardio-hospitals/. Accessed 25 May 2013.
53. Scientific Registry of Transplant Recipients (SRTR); US hospitals with heart transplant centers. http://www.srtr.org/csr/current/Centers/TransplantCenters.aspx?organcode=HR. Accessed 25 May 2013.
54. Finlayson SRG, Birkmeyer JD, Tosteson ANA, Nease RF. Patient preferences for local care: implications for regionalization. Med Care. 1999;37:204–9.
55. Vinocur JM, Menk JS, Connett J, Moller JH, Kochilas LK. Surgical volume and center effects on early mortality after pediatric cardiac surgery: 25-year North American Experience From a Multi-institutional Registry. Pediatr Cardiol. 2013;34(5):1226–36.
56. Hannan E, Kilburn H, Racz M, Shields E, Chassin MR. Improving the outcomes of coronary artery bypass surgery in New York State. JAMA. 1994;271(10):761–6. doi:10.1001/jama.1994.03510340051033.
57. Marelli AJ, Mackie AS, Ionescu-Ittu R, Rahme E, Pilote L. Congenital heart disease in the general population: changing prevalence and age distribution. Circulation. 2007;115:163–72.
58. Wernovsky G, Rome JJ, Tabbutt S, Rychik J, Cohen MS, Paridon SM, Webb G, Dodds KM, Gallagher MA, Fleck DA, Spray TL, Vetter VL, Gleason MM. Guidelines for the outpatient management of complex congenital heart disease. Congenit Heart Dis. 2006;1:10–26. doi:10.1111/j.1747-0803.2006.00002.x.
59. Marino BS, Lipkin PH, Newburger JW, Peacock G, Gerdes M, Gaynor JW, Mussatto KA, Uzark K, Goldberg CS, Johnson Jr WH, Li J, Smith SE, Bellinger DC, Mahle WT. American Heart Association Congenital Heart Defects Committee, Council on Cardiovascular Disease in the Young, Council on Cardiovascular Nursing, and Stroke Council. Neurodevelopmental outcomes in children with congenital heart disease: evaluation and management: a scientific statement from the American Heart Association. Circulation. 2012;126(9):1143–72. Epub 2012 Jul 30.
60. Barreira JL, Baptista MJ, Moreira J, Azevedo A, Areias JC. Understanding of endocarditis risk improves compliance with prophylaxis. Rev Port Cardiol. 2002;21(9):939–51.
61. Bulat DC, Kantoch MJ. How much do parents know about their children's heart condition and prophylaxis against endocarditis? Can J Cardiol. 2003;19(5):501–6.
62. Cheuk DK, Wong SM, Choi YP, Chau AK, Cheung YF. Parents' understanding of their child's congenital heart disease. Heart. 2004;90(4):435–9.
63. da Silva DB, Souza IP, Cunha MC. Knowledge, attitudes and status of oral health in children at risk for infective endocarditis. Int J Paediatr Dent. 2002;12(2):124–31.
64. Fernandes S, Verstappen A, Ackerman K, Adams E, Barton C, Breitinger P, Crumb S, Dummer K, Harada K, Khairy P, Landzberg M, Linstead-Goldsmith R,

Meadows AK, Nieves JA, Saidi A, Takahashi M, Zhou J, Ziniel S, Williams R, the Adult Congenital Cardiac Care Associate Research Network. Parental knowledge regarding lifelong congenital cardiac care. Pediatrics. 2011;128:e1489. doi:10.1542/peds.2010-3068.

65. Frogel MP, Stewart DL, Hoopes M, Fernandes AW, Mahadevia PJ. A systematic review of compliance with palivizumab administration for RSV immunoprophylaxis. J Manag Care Pharm. 2010;16(1):46–58.
66. Lobel A, Geyer S, Grosser U, Wessel A. Knowledge of congenital heart disease of mothers: presentation of a standardized questionnaire and first results. Congenit Heart Dis. 2012;7:31–40.
67. Williams IA, Shaw R, Kleinman C, Gersony W, Prakash A, Levasseur SM, Glickstein JS. Parental understanding of neonatal congenital heart disease. Pediatr Cardiol. 2008;29:1059–65. doi:10.1007/s00246-008-9254-8.
68. Yang HL, Chen YC, Wang JK, Gau BS, Moons P. An evaluation of disease knowledge in dyads of parents and their adolescent children with congenital heart disease. J Cardiovasc Nurs. 2013;28(6):541–9.
69. Lesch W, Specht K, Lux A, Frey M, Utens E, Bauer U. Disease-specific knowledge and information preferences of young patients with congenital heart disease. Cardiol Young. 2013. doi:10.1017/S1047951113000413. Published online: 29 April 2013.
70. Rosenfeld JS. Section 504 and IDEA: basic similarities and differences. Wrights Law. http://www.wrightslaw.com/advoc/articles/504_IDEA_Rosenfeld.html. Accessed 25 May 2013.

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