Defining Terms in Lists of Nomenclature

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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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P.R. Barach, J.P. Jacobs, S.E. Lipshultz, P.C. Laussen (eds.),
Pediatric and Congenital Cardiac Care: Volume 1: Outcomes Analysis,
DOI 10.1007/978-1-4471-6587-3_6, © Springer-Verlag London 2015

Keywords
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.

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