# Children's Palliative Care: An International Case-Based Manual

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*This book is dedicated to all of the children and their families that I have cared for since starting out in palliative care. Also to all my amazing colleagues and friends around the world that I have had the privilege to work with and get to know over the years, many of whom have contributed to this book. I have been inspired and motivated by you all, to continue to work to improve the quality of life for all children with life-limiting and life-threatening conditions and their families around the world.*

# Foreword

On behalf of the International Children's Palliative Care Network (ICPCN), I am pleased to commend this global resource to any individual or organisation working in the field of children's palliative care. It has been thoughtfully curated and compiled, bringing together the well-respected expertise of leading international children's palliative care (CPC) clinicians who continue to champion the delivery of CPC knowledge sharing across the world.

It is authored and edited with great energy and intent and is positioned authentically within the scaffolding of ICPCN's mission to *achieve the best quality of life for children and young people with life-limiting conditions, their families and carers worldwide*. In addition, it is a demonstrable practical and accessible resource for the CPC worldwide community that furthers the mission of ICPCN to not only raise *awareness of children's palliative care* and advance the *global development of children's palliative care services* but most significantly is underpinned by a deep commitment to *sharing expertise, skills and knowledge*.

It must be acknowledged that this unique offering to readers, as the first case-based manual for global CPC, has only been made possible by the positive international collegiality within the sector that has been strengthening over many decades. ICPCN continues to recognise the tireless work of so many who have been passionately advancing knowledge and resource collaboration globally and their dedication to ensure resources such as this are shared and distributed widely. Indeed, our hope is that a compilation of this nature is globally relevant to both readership and leadership. It is further envisaged that this publication not only serves the audience well in its very practical nature but also it can provide a foundation of inspiration to publish more books of its type for generations to come.

Finally, our belief is that this publication will not only be a crucial piece in the developed and developing countries' CPC toolkit but will also make a profound impact to global regions that are underserved by CPC services at this moment in time. Through the expert content delivered with warmth and clarity and its relevance and broad appeal, we hope it will be highly conducive to worldwide distribution and pivotal in strengthening the international collectivism for supporting children with life-limiting conditions and their families.

All involved in bringing this rich resource together, with special mention to ICPCN Chief Executive, Professor Julia Downing, as editor, should be immensely proud of this publication and the contribution it will make to a very dedicated global CPC community.

Board Chair, ICPCN, Australia Paul Quilliam

# Preface

Children's palliative care (CPC) is an important and developing field globally. The provision of children's palliative care varies around the world, with provision not meeting needs and over 65% of countries having no recognised CPC service provision. Therefore, the reality is that, whilst there are an estimated 21.1 million children who require palliative care, in many areas of the world CPC has had a poor profile, is unavailable, and children are treated like little adults without their unique needs being recognised or understood. It is therefore essential that we increase capacity for service delivery and expand service provision so that all the children, regardless of where they live, can access affordable and culturally appropriate palliative care. Too many children have been suffering needlessly due to the lack of understanding, knowledge and skills in CPC. As a growing field, evidence is emerging in all aspects of service provision, including clinical care, models of care and education, and it is important that we share this evidence and learn from each other.

Whilst the provision of CPC varies in different regions and countries, the overriding philosophy is the same, with the clear aim of improving quality of life for children and their families. However, due to the resources available, the health systems in which we work and the policies that guide us, the way that we provide care may be different. Regardless of how we provide the care, we need to be focusing on the children and their families, ensuring that we are supporting them holistically, i.e., physically, psychologically, socially and spiritually, ensuring that they are at the centre of the care that we provide.

This book will enable individuals working in CPC globally to learn through engaging in case studies. It will give them the opportunity for real-life learning, learning from each other through case histories from around the world which will draw out many of the key issues and elements of CPC. It provides a practical approach grounded in experience and the appropriate evidence base, and aims to: be practical and accessible to readers; focus on the child and the family throughout; utilise cases from around the world; demonstrate multi-disciplinarity both in terms of provision of care, but also in terms of the teams of authors contributing to the book, with each chapter being authored by a mix of individuals from low/middle and high income countries; have an international perspective, with real-life cases being presented from a wide range of countries and situations, presented by experienced international CPC practitioners; integrate physical, psychological, social and

spiritual care throughout the cases, demonstrating care across a range of ages and conditions and is one of the first books on CPC to address the concepts of Serious Health-related Suffering (SHS) from the Lancet Commission report, and Universal Health Coverage (UHC).

Every chapter starts with some key learning points, each of which will be covered through the case studies that follow. Cases are introduced and developed throughout the chapter, with new cases being introduced where appropriate. The questions presented in the chapters should help you to think about how you would approach the provision of care within the case discussed. There is some duplication in different chapters, as many of the chapters will be read as stand-alone chapters; however, we have tried to keep this to the minimum. Due to the format of this book, we hope that it will be useful to a wide range of professionals including clinicians, researchers, educators and students.

We hope that you find this case-based manual helpful, and I would like to thank all of the authors who have contributed to it and the children and families who have permitted us to use their stories.

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# About the Editor

**Prof. Julia Downing** is an experienced palliative care nurse, educationalist and researcher with a PhD in palliative care education. She has been working within palliative care for 29 years, with 19 of those working internationally in low- and middle-income countries, including in Uganda, across Africa, Eastern Europe and globally, developing palliative care services for children and adults. She is recognised as a global leader and expert in the field and sits on various technical and advisory groups. She is the Chief Executive of the International Children's Palliative Care Network (ICPCN), an Honorary Professor at Makerere University, Kampala, a Visiting Professor at the University of Belgrade in Serbia, Edge Hill University and the University of South Wales a, an Honorary Senior Research Fellow at the Cicely Saunders Institute, King's College London and an Honorary Senior Fellow at the University of Edinburgh.

She has extensive experience in research, presenting at conferences and writing for publication, and is on the editorial board of the International Journal of Palliative Nursing (IJPN), ecancer and Annals of Palliative Medicine. Professor Downing serves on the boards of several NGOs including the Worldwide Hospice Palliative Care Alliance, the International Association of Hospice & Palliative Care, the African Palliative Care Association UK and the Palliative Care Research Society. She was the recipient of the IJPN's Development Award in 2006, the Robert Tiffany lectureship from the ISNCC in 2014 and the Pearl Moore 'Making a Difference' International Award for Contributions to Cancer Care from the Oncology Nursing Society in 2015. She was also recognised as a leading change agent in oncology in the 'Women as Change Agents' publication for International Women's Day in 2016.

# List of Abbreviations

| | |
|---|---|
| AAHPM | American Academy of Hospice and Palliative Medicine |
| AAP | American Academy of Pediatrics |
| ACP | Advance care plan |
| ACT | Association of Children's Palliative Care |
| ADC | Average daily census |
| AIDS | Acquired Immune Deficiency Syndrome |
| ALCP | Asociación Latinoamericana de Cuidados Paliativos (Latin American Association of Palliative Care) |
| APCA | African Palliative Care Association |
| APHN | Asia Pacific Hospice and Palliative Care Network |
| ART | Antiretroviral therapy |
| AS | Asthma score |
| ASPEN | American Society for Parenteral and Enteral Nutrition |
| AVSD | Atrioventricular septal defect |
| AYA | Adolescents and young adults |
| BAPM | British Association of Perinatal Medicine |
| BID | Bis in die (twice a day) |
| BIP | Bleomycin interstitial pneumonitis |
| BMT | Bone marrow transplant |
| BOS | Bronchiolitis obliterans syndrome |
| CASP | Critical Appraisal Skills Programme |
| CBD | Cannabidiol |
| CBT | Cognitive behavioural therapy |
| CCC | Complex chronic conditions |
| CINHAL | Cumulative Index to Nursing and Allied Health Literature |
| COPD | Chronic obstructive pulmonary disease |
| COX-2 | Cyclooxygenase-2 |
| CME | Continuing Medical Education |
| CPAP | Continuous positive airway pressure |
| CPC | Children's palliative care |
| CPR | Cardiopulmonary resuscitation |
| C-POS | Children's Palliative care Outcome Scale |
| CRIES | Crying; Requires increased oxygen administration; Increased vital signs; Expression; Sleeplessness |

| | |
|---|---|
| DH | Department of Health |
| DIPG | Diffuse Intrinsic Pontine Glioma |
| DMD | Duchenne muscular dystrophy |
| DNR | Do not resuscitate |
| DVT | Deep vein thrombosis |
| EAPC | European Association for Palliative Care |
| ECG | Electrocardiogram |
| EEG | Electroencephalogram |
| EPEC | Education in Palliative and End-of-Life Care |
| ESPGHAN | European Society for Paediatric Gastroenterology, Hepatology and Nutrition |
| EoL | End-of-life |
| FDA | Food and Drug Administration |
| FLACC | Face, legs, activity, cry, consolability |
| GMI | Graded motor imagery |
| Hb | Haemaglobin |
| HCPs | Health care professionals |
| HCT | Haematopoietic cell transplantation |
| HDI | Human Development Index |
| HICs | High-income countries |
| HIE | Hypoxic-ischemic encephalopathy |
| HINARI | Hinari Access to Research in Health Programme |
| HIV | Human immunodeficiency virus |
| HPCA | Hospice Palliative Care Association of South Africa |
| hr | Hourly or hours |
| IAHPC | International Association of Hospice & Palliative Care |
| ICD | Intercostal drain |
| ICD10 | 10th revision of the International Statistical Classification of Diseases and Related Health Problems |
| ICPCN | International Children's Palliative Care Network |
| IHME | Institute of Health Metrics and Evaluation |
| INCB | International Narcotics Control Board |
| IOELC | International Observatory on End of Life Care |
| IPS | Idiopathic pneumonia syndrome |
| IQ | Intelligence quotient |
| IV | Intravenous |
| Kg | Kilogram |
| KGF | Keratinocyte growth factor |
| L | Litre |
| LL | Life limiting |
| LLCs | Life-limiting conditions |
| LLLT | Low-level laser therapy |
| LMICs | Low- and-middle income countries |
| LST | Life Sustaining Treatment |
| LTCs | Life-threatening conditions |

| | |
|---|---|
| LVAD | Left ventricular assist device |
| Max | Maximum |
| MDT | Multi-disciplinary team |
| Mg | Milligram |
| mo | Months |
| MoH | Ministry of Health |
| MMRC | Modified Medical Research Council |
| MRI | Magnetic Resonance Imaging |
| MSAS | Memorial symptom assessment scale |
| N/A | Not applicable |
| NCCPC-R | Non-communicating children's pain checklist-Revised |
| n.d. | No date |
| NRS | Numerical rating scale |
| NSAIDs | Non-steroidal anti-inflammatory drugs |
| PC | Palliative care |
| PCA | Patient-controlled analgesia |
| PCEP | Palliative Care Education and Practice |
| PCRA | Patient-controlled regional anaesthesia |
| PDE | Principle of Double Effect |
| PHC | Primary health care |
| PICU | Paediatric Intensive Care Unit |
| PO | Per Os (by mouth) |
| PMLD | Profound and multiple learning disabilities |
| PNET | Primitive neuroectodermal tumour |
| PQRS | Preview, Question, Read and Summarise |
| PR | Per rectum |
| PRN | Pro re nata (as required) |
| PRAM | Paediatric Respiratory Assessment Measure |
| PPP | Paediatric pain profile |
| Q6hr | Every 6 hours |
| QDS | Quarter die sumendus (four times a day) |
| QHS | Quague hora somni (every night at bedtime) |
| QoD | Quaque altera die (every other day) |
| QoL | Quality of life |
| r-FLACC | Revised-face, legs, activity, cry, consolability |
| RCPCH | Royal College of Paediatrics and Child Health |
| RCTs | Randomised Controlled Trials |
| RDOS | Respiratory distress observatory score |
| SC | Subcutaneous |
| SCC | Spinal Cord Compression |
| S-COS | Simplified concrete ordinal scale |
| S-FPS | Simplified faces pain scale |
| SDGs | The Sustainable Development Goals |
| SHS | Serious Health-related Suffering |
| SIG | Special interest group |

| | |
|---|---|
| SL | Sublingual |
| SMA | Spinal muscular atrophy |
| SSRIs | Selective serotonin re-uptake inhibitors |
| TB | Tuberculosis |
| TCA | Tricyclic antidepressants |
| TfSL | Together for Short Lives |
| TENS | Transcutaneous electrical nerve stimulation |
| THC | Tetrahydrocannabinol |
| TID | Ter in die (three times a day) |
| UHC | Universal health coverage |
| UK | United Kingdom |
| UN | United Nations |
| UNAIDS | The Joint United Nations Programme on HIV/AIDS |
| UNCRC | United Nations Convention on the Rights of the Child |
| UNDP | United Nations Development Programme |
| UNHCR | United Nations High Commissioner for Refugees |
| UNICEF | United Nations Children's Fund |
| USA | United States of America |
| VAS | Visual analogue scale |
| VDS | Verbal descriptor scale |
| VP | Ventriculoperitoneal |
| WHA | World Health Assembly |
| WHO | World Health Organization |
| WHPCA | The Worldwide Hospice Palliative Care Alliance |
| WPC | Warn, Pause, Check |
| Yrs | Years |
| ZPD | Zone of Parental Discretion |

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# The Need for and Gaps in Provision of Children's Palliative Care Globally

1

Julia Downing, Stephen Connor, and Lorna Fraser

**Key Learning Points**

1. The need for and accessibility to children's palliative care (CPC) varies globally, with differences seen between countries and regions.
2. There are a variety of challenges to assessing the need for CPC, which need to be addressed in order to get a clear understanding of the need as well as existing service provision.
3. It is not enough just to know what CPC services exist, but it is also important to be able to assess the quality and outcomes of the service being provided.
4. Once the needs and gaps have been identified, there are challenges to the development and implementation of CPC services that need to be overcome.

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**Case Study**

*At a meeting about Global Children's Health Care a member of the Ministry of Health from The Gambia heard about the importance of palliative care for children, how it can improve quality of life for children and their families, and the need to ensure that all children have access to palliative care regardless of where they live. He recognised that children in The Gambia do not currently have access to palliative care services. He was put in contact with the International Children's Palliative Care Network to explore ways of starting up children's palliative care services in The Gambia. Whilst his colleagues in the Ministry are supportive, he needs to be able to put a strong case to them for the development of children's palliative care and therefore asked for information on: the global need for children's palliative care; how he can assess the need in The Gambia; how it compares to other countries, and what they can learn from what other countries have done to develop children's palliative care.*

## 1.1 Question 1. Why Is It Important That We Know the Need for Children's Palliative Care?

The need for, and accessibility to, CPC varies widely across the world and there is now a growing awareness of the unmet need for CPC worldwide. Often emphasis in child health globally is focused on the reduction of mortality, which is essential, but we also need to look at the need for, and quality of, CPC, which has been a neglected area.

Understanding what services are currently available and any issues in accessing or implementing these services, along with the need for CPC, is important when developing services. For example: How many children need CPC? What diagnoses/conditions do they have? What are their needs? Where do they live? What services exist, if any? What is the quality of the services? and What are the gaps? Often the need for CPC is under-estimated, and there is a lack of awareness as to which children need palliative care, although this may vary in different countries due to the resources available and the model of providing healthcare and CPC. When advocating for the development of CPC, and in order to influence policy makers, it helps if we can demonstrate that there is a need and where the gaps are. Therefore, assessing the need for CPC is important so that we can describe how big the need is and why reducing any unnecessary suffering of children and their families is so important.

## 1.2 Question 2. What Is the Global Need for Children's Palliative Care?

Until recently the need for CPC globally has been unknown, thus work has been undertaken on developing estimates for the need for CPC. Several studies have been undertaken, with three main studies being referred to on a regular basis: The global atlas of palliative care (PC) at the end of life (EoL) (Connor and Sepulveda 2014),

**Table 1.1** Summary of studies estimating the global need for CPC

| Study | Data source | Age range | Results |
|---|---|---|---|
| • Global atlas of PC at the EoL (Connor and Sepulveda 2014) | • Utilised mortality data for conditions requiring palliative care, adjusted by the estimate pain prevalence for each disease category. | • 0–14 years | • Global number of children in need of PC at the EoL is >2.5 million. |
| • Estimate of the global need for CPC (Connor et al. 2017) | • Disease prevalence data was used instead of mortality for the majority of conditions requiring PC.<br>• Representative sample of countries covering 59.5% of the world' population stratified by region and World Bank income groups. | • 0–19 years | • The estimated global number of children in need of PC was identified as 21.1 million, with greater than eight million requiring specialised service provision. |
| • Lancet Commission: Alleviating the access abyss in palliative care and pain relief—an imperative of universal health coverage (Knaul et al. 2017) | • Utilised the concept of Serious Health-related Suffering (SHS) associated with illness or injury that could be helped by PC and pain treatment.<br>• Utilised WHO mortality data and did not include prevalence. | • 0–14 years | • >5.3 million children aged under 15 years experience SHS each year worldwide, with 1/3rd of children who died in 2015 experiencing SHS. |

estimates of the global need for CPC (Connor et al. 2017) and the Lancet Commission on Alleviating the access abyss in palliative care and pain relief—an imperative of universal health coverage (Knaul et al. 2017). Two of these studies utilise the WHO definition of CPC (WHO 2002) and the other looks at SHS (Table 1.1).

The global atlas of PC at the EoL (Connor and Sepulveda 2014) utilised a framework published by the WHO in 2007 (WHO 2007) to estimate the number of children in need of PC at the EoL through considering mortality data from conditions requiring PC, and adjusted by the estimated pain prevalence for each disease category as an indicator of symptom burden (Higginson 1997). The atlas focuses on EoL and identified diseases requiring PC for children between the ages of 0 and 14 years. It estimates the global number of children in need of PC at the end of life (EoL) as being >2.5 million, including half at the EoL and half prior to EoL. The greatest number of children needing CPC was for congenital anomalies (25.06%), neonatal conditions (14.64%), malnutrition (14.12%), meningitis (12.62%), Human Immunodeficiency Virus (HIV)/Acquired Immune Deficiency Syndrome (AIDS) (10.23%) and cancer (5.85%). Whilst overall children needing PC for progressive non-malignant disease constitute the highest proportion of all children, with HIV/AIDS and cancer being a small proportion, this will vary by country and region, for example, countries in sub-Saharan Africa have high numbers of children with HIV/AIDS.

The study estimating the global need for CPC (Connor et al. 2017) aimed to create an accurate global estimate of the need for CPC, based on a representative sample of countries covering 59.5% of the world's population stratified by region and World Bank Income Groups. This was done using prevalence and mortality data for children between 0 and 19 years of age. In many high-income countries, children are living longer with life-threatening conditions (LTCs), and so it was felt to be important to include all those up to at least 19 years of age. Based on the Together for Short Lives Categories of children requiring PC (Together for Short Lives 2018) along with expert opinion, the study applied similar assumptions as the global atlas, but covered the whole spectrum of PC provision and not just at the end of life. Estimate of the need for specialist CPC was calculated at 37.5% of the total need and an average daily census (ADC) figure was used to account for survival in the group of children who will need CPC in any given year—an ADC of 100 days was used based on work in Australia (Australian Institute of Health and Welfare 2011). The estimated global number of children in need of PC was identified as 21.1 million children, with greater than eight million requiring specialised service provision. This is higher than the estimate in the global atlas due to the age range being higher, the fact that disease prevalence data was mainly used instead of mortality, and it did not focus purely on the EoL. This was the first attempt to measure the global need for CPC for the majority of the world's population and was thought to be a minimum estimate of need because it did not include many severely disabled children whose lives may be shortened.

The Lancet Commission on Alleviating the access abyss in palliative care and pain relief—an imperative of universal health coverage (Knaul et al. 2017) developed and utilised the concept of SHS, which is associated with illness or injury that could be helped by PC and pain treatment. They looked at children aged 15 years or younger and estimated that >5.3 million children aged 15 years or under experience SHS each year worldwide with 1/3rd of children who died in 2015 experiencing SHS. Of the estimated nearly 2.5 million children dying each year needing PC and pain relief >98% are from low-and-middle income countries (LMICs). They also found that establishing a special annual fund for purchasing medicines for children living in LMICs who are in need of opioids for the relief of pain and PC would cost only US$ one million. These estimates, like those in the global atlas, were derived from WHO mortality data and did not include prevalence, accounting for the lesser numbers than the Connor et al. study.

Whilst each of these studies uses different age ranges, methods, and are thought to be under-estimates, they help us begin to see the scale of the challenge for CPC globally.

## 1.3 Question 3. Are There Differences in the Need for Children's Palliative Care Between Countries, Regions, or World Bank Income Group Countries?

The rate of need for CPC varies between regions, countries and low-middle-high income groups. There are clear differences globally—the global atlas of palliative care at the EoL showed the greatest need for CPC in the African region (49% of total

need), followed by the South East Asian region (24%), followed by the Eastern Mediterranean (12%), with the African region having the highest number of children dying from HIV/AIDS (Connor and Sepulveda 2014). Generally, the higher the human development index (HDI) for a country, the lower the need for CPC despite the children with highest need, e.g., those on ventilators, being in high-income countries. For example, in low-income countries, childhood deaths account for >30% of all deaths associated with SHS, compared with <1% in high-income countries, >50% of the burden of SHS in children in low-income countries is associated with HIV and >90% of all child deaths associated with SHS in low-income countries are avoidable (Knaul et al. 2017).

## 1.4 Question 4. How Do We Estimate the Need for Children's Palliative Care?

There are different ways to estimate the need for CPC as has been seen in the studies discussed above. One way is through a cross-sectional mixed methods approach, using both quantitative and qualitative data from primary and secondary sources looking at both mortality and disease prevalence. Country estimates have utilised country-level disease prevalence data from the Institute of Health Metrics and Evaluation (IHME), mortality data from the WHO Global Health Estimates, the Joint United Nations Programme on HIV/AIDS (UNAIDS) data on HIV prevalence and country-level data collection, interviews and surveys. (Connor and Sepulveda 2014). Exactly how it will be done in an individual country will depend on a variety of issues including resources, availability and quality of country-level data, e.g., disease statistics, age ranges used, understanding of PC, etc. It will also vary if you are looking at country-level data, or community level needs. At the community level you could do a household survey, whereas at the national level this would not be possible. Key questions to be addressed include:

(a) What are the clinical and demographic characteristics of the population?
(b) What services do they need access to?
(c) How long do they need these services for?

## 1.5 Question 5. Are There Examples of Countries Where the Need for Children's Palliative Care Have Been Assessed and How Was This Done?

There are examples where the need for CPC has been assessed in different countries. Two examples are the United Kingdom (UK) and South Africa.

In the UK children with life-limiting (LLC) or life-threatening conditions (LTC) were identified and the prevalence of LLC calculated utilising individual level hospital admission data available on a national basis (Fraser et al. 2012). The diagnoses with the highest prevalence were the life-limiting or life-threatening congenital anomalies. Through the work done in the UK it has also been possible to see the rise

in need for CPC, e.g., from 25:10,000 children with a LLC in 2000 to 32:10,000 children in 2010 (Fraser et al. 2012). A more detailed study undertaken in Scotland (Fraser et al. 2015) identified that although the number and prevalence of children with a LLC was rising, many of these children are stable each year (around 80%) with a smaller percentage unstable, deteriorating or dying. A higher percentage of babies under 1 were unstable, deteriorating or dying (around 40%) (Jarvis et al. 2017). These data have been used, in combination with other work, to get commitment for extended funding for CPC in Scotland.

South Africa (Connor et al. 2014) utilised country-level prevalence data from the IHME, mortality data from the WHO Global Health Estimates, UNAIDS Data on HIV prevalence and country-level data collection, interviews and surveys. Results suggested that 549,778 need PC with 207,909 requiring specialist PC, a rate of 102.5 per 10,000 children aged 0–19 years (Connor et al. 2017). The study also looked at service provision and it was estimated that in 2012 <5% of those needing specialist PC received it. The need is high in South Africa due to the HIV prevalence—almost four times that in the UK.

## 1.6 Question 6. What Are the Challenges in Assessing the Need for Children's Palliative Care?

There are a number of challenges in assessing the need for CPC, these include:

(a) Which children and when they should receive PC:
- Assessment is complex due to uncertainty about which children need PC, and the nature of PC for children (Hain et al. 2011), which may vary by countries as their healthcare systems and population needs differ.
- Need is generally based on mortality statistics and does not accurately capture the CPC need for children before the EoL and last year of life.
- A lack of clarity with regards to the conditions and diagnostic groups needing CPC—e.g., in the global atlas, where 75% of children with neonatal encephalopathy were discounted by the WHO (Connor and Sepulveda 2014).
- There is variability in severity and outcomes for many of the diagnoses, e.g., severe cerebral palsy or hypoxic ischemic encephalopathy (HIE).
- How do you estimate need when the length of time children receive PC varies considerably, e.g., from 1 day to several years—hence the use of an ADC. An early study looking into survival and mortality in a hospice in the UK found that some 'children' were still under the care of the hospice 21 years after first being referred for PC (Taylor et al. 2010).
- A child who may need PC in a low-income country may not in a high-income country due to the availability of different resources for care and cure, late presentation, lack of understanding of PC, etc.

(b) Challenges with the data:
- Many countries, particularly LMICs, do not have accurate statistics on prevalence or mortality.

- The main diagnostic coding system, ICD10, does not have individual codes for each diagnosis so diagnoses are grouped together based on pathology not prognosis, so specificity of these codes is an issue.
- Some of the presumptions behind the data are based on adult data, e.g., the estimation for the symptom burden of pain used is 67%, which may be an underestimation as pain may not be the best indicator for need for PC in children as there are many other troubling symptoms affecting children alongside pain.
- Reliance on secondary data due to a lack of in-country data.

## 1.7 Question 7. Once We Have Estimated the Need, How Do We Assess the Gap Between the Need and Provision?

Mapping CPC provision can be challenging, for example: what constitutes CPC provision? how do we define specialist CPC? how do we measure access to generalist as well as specialist CPC? how do we get accurate information about existing services? who decides if they are providing palliative care and against which criteria? Also, many services provide CPC as part of a wider service and are often missed in mapping exercises.

A systematic review of CPC provision around the world (Knapp et al. 2011) was undertaken to assess the level of CPC provision and provide a baseline for future assessments. Each country was assigned a level of provision based on a four-part typology developed by the International Observatory on End of Life Care (IOELC) (Wright et al. 2008). They found that: 65.6% of countries had no known CPC activity; 18.8% had capacity building activities; 9.9% had localised provision and only 5.7% had provision reaching mainstream providers. They acknowledged the need to expand the levels of care provision as some countries, e.g., the United States of America (USA) were assigned to level 4 with services reaching mainstream providers and yet widespread implementation and accessibility was lacking. The review identified large gaps and showed disparities between regions, e.g., North America and Europe, when compared to Africa, Asia and Latin America. However, they also identified the lack of published information on CPC globally, impacting on the findings of the review.

The global atlas of palliative care at the EoL (Connor and Sepulveda 2014) identified that there was no central repository to track CPC progress and provision. They also noted that provision is an ever-changing concept, which impacts on tracking progress. A new edition of the global atlas is under development that will utilise a new six level 'mapping' of PC development for adults and separately for children.

Since its inception in 2005, the International Children's Palliative Care Network (ICPCN) has held a directory of CPC services globally and estimates the provision of CPC (Marston et al. 2018). From this ICPCN has been able to track the development of CPC globally. This highlights the ongoing gaps and need for urgent and ongoing developments within CPC. As developments are ongoing, ICPCN tries its best to keep the maps up to date; however, this is dependent on receiving regular updates from its members. Thus, an exercise is currently ongoing to review the status in each country.

Over the past 8 years several regional atlases of PC have been developed, e.g., the African Palliative Care Association (APCA) Atlas of Palliative Care in Africa (Rhee et al. 2017), the Atlas of Palliative Care in Latin America (Pastrana et al. 2012) and the European Atlas (Arias-Casais et al. 2019). These utilise the WHO Public Health framework for PC development (Stjernsward et al. 2007). Alongside this a series of indicators were developed to map PC development with there being an indicator on *'Number of hospices or palliative care services in the country with palliative care programmes specific to paediatrics'* (Arias et al. 2019). An example of the use of this indicator is for Africa, with limited numbers of services identified, with no known services in many countries (Fig. 1.1).

## 1.8 Question 8. Why Is It Important to Assess the Gap in Children's Palliative Care Provision?

Assessment of the gap in CPC provision between need and existing service provision is important as it is needed for policy makers to help them understand the need and be able to justify the allocation of resources. By identifying existing CPC services, it may also help in the ongoing development of models of CPC provision within countries through the identification of where it is working well, where it can be improved, what are the lessons learnt, etc.

It is important that any mapping does not just look at stand-alone services but takes into account community and hospital-based services—especially with the ongoing importance of integration, e.g., through the WHA resolution on PC (WHO 2014), Universal Health Coverage (UHC) and the Astana Declaration (WHO and UNICEF 2018). In most countries there are no specific indicators being collected routinely at the country level to help ascertain service provision, and often where there are indicators, they are proxy ones such as the per capita use of morphine. Although now that indicators have been developed to help with the mapping of PC which include CPC (Arias et al. 2019), it is hoped that these may be used in the future to help show and map in-country CPC development.

## 1.9 Question 9. It Is Also Important to Measure the Outcome of Children's Palliative Care Service Provision, How Might This Be Done?

It is not enough just to know what CPC services exist, but it is also important to be able to assess the quality and outcomes of the service being provided. However, there is limited evidence of the quality of the services being provided, and one reason for this has been the lack of appropriate validated outcome measures or indicators for children (Downing et al. 2018a, b; Harding et al. 2014). Over the past decade outcome measurement, including the use of patient reported outcome measurement, has been used to assess the effectiveness, efficiency and availability of PC; however, until recently the majority of outcome measures for palliative care have been for use in adults, with a lack of an appropriate multi-dimensional

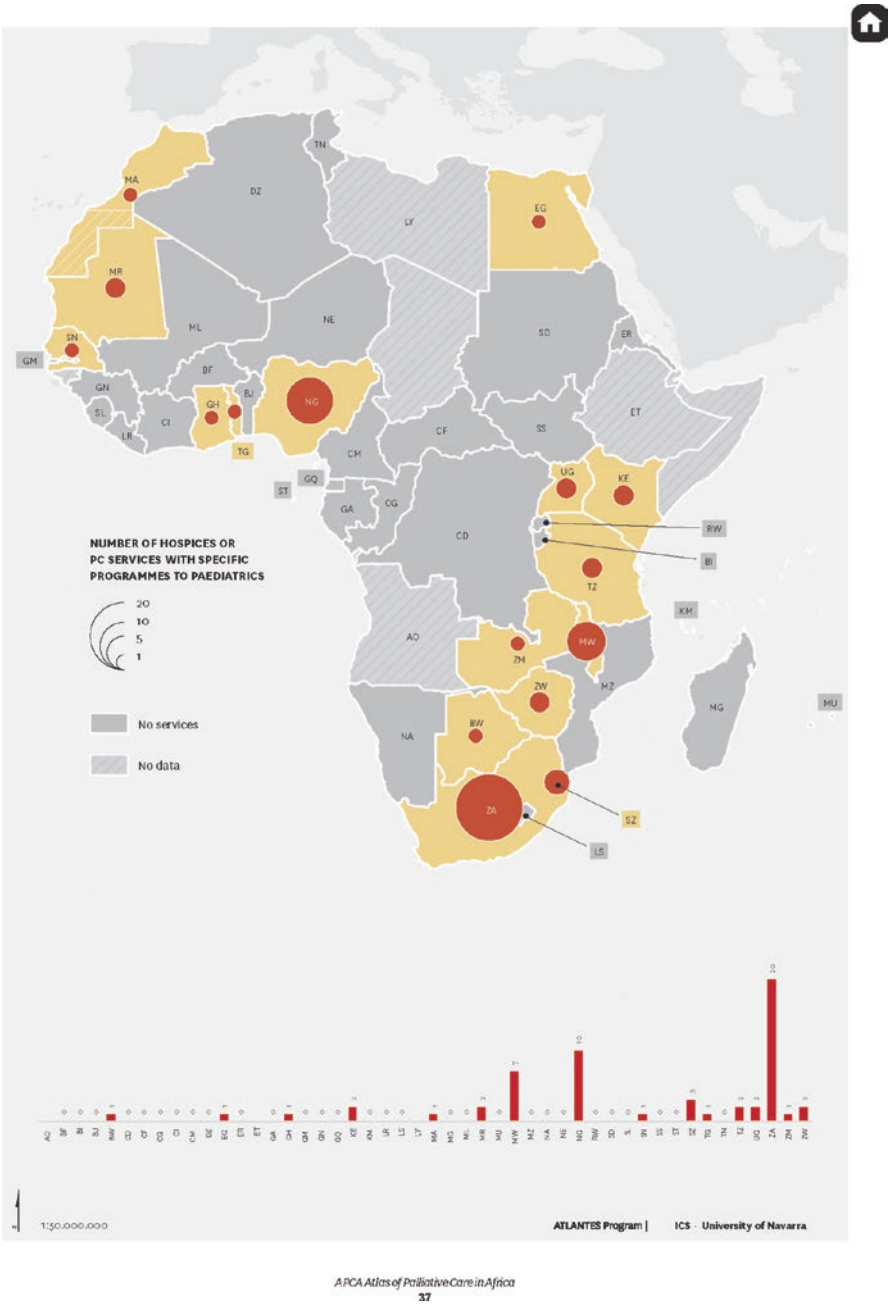


**Fig. 1.1** Number of hospice or PC services in the country with PC programmes specific to paediatrics (Rhee et al. 2017, p. 37 Reproduced with permission)

outcome measure for use in CPC (Downing et al. 2018a, b; Coombes et al. 2016). Work is ongoing on the development of appropriate outcome measures for children, such as the APCA Children's Palliative Outcome Scale (APCA C-POS) (Downing et al. 2012, 2018a, b) which is being used in a variety of countries within sub-Saharan Africa. Similar tools are also currently being developed for the use in other parts of the world, e.g., in the UK. Outcome measures such as the C-POS can then be used at the service level in order to assess the outcomes of the care being provided. Whilst use of this outcome measure is in its early stages, it is anticipated that it will become more widely used and accessible for use within CPC, thus enabling us to look not only at what CPC services exist but also the quality of the services being provided.

## 1.10 Question 10. What Are the Challenges to the Development of Children's Palliative Care Within a Country and What Can We Learn from Other Countries?

Estimation of need is an important step in the development of CPC within any country and is often the first step that is needed to gain support for and recognition of CPC. Further work is then needed to understand the need in more detail, e.g., where the children are, at what point in the continuum of care, etc. Challenges exist to the development of CPC and estimating need and gaps is the first part in trying to dispel some of these challenges such as (Downing et al. 2016, 2018a, b):

- A lack of understanding of CPC.
- Denial of the state of the problem and that children need PC—therefore lack of recognition of the need.
- Lack of policies to support CPC.
- Lack of funding to develop and integrate CPC into existing health care systems.
- Lack of integration of PC into health services across the board.
- Lack of access to education for health care professionals (HCPs), treatment, trained professionals, medicines, etc.
- Lack of resources to develop services.

However, there are various global priorities that will help in overcoming the challenges which will be discussed in the next chapter.

## 1.11 Conclusion

Having an understanding of the need for CPC globally and within country, along with the gaps in service provision, will help to strengthen advocacy to governments to support the development of CPC and improve the quality of life of children and their families globally.

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# Universal Health Coverage and Serious Health-related Suffering: A Case for Children and Young People

2

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**Key Learning Points**

1. There are many policies, documents, platforms and resolution statements published each year, some of which have a direct impact on strengthening the global imperative for children's palliative care (CPC).
2. Ensuring equitable access to, availability of and usage of quality palliative care (PC) services is a fundamental and definitional component of Universal Health Coverage (UHC).
3. The Lancet Commission report on 'Alleviating the access abyss in PC and pain relief—an imperative of universal health coverage', recognises that children and their families have specific PC needs, with children accounting for 9% of those experiencing serious health-related suffering (SHS).

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4. The relief of SHS in children is not always seen as a priority, with existing measures of care focusing on extending life and productivity as opposed to quality of life (QoL). In the absence of PC, inappropriate disease-focused care can worsen the QoL of children.
5. The International Association of Hospice and Palliative Care (IAHPC) essential practices for PC in Primary Care, along with the Lancet Commission report, set out essential packages of PC and pain relief that should be available for CPC.

**Case Study: Advocacy**

*An advocate for CPC in a low-income country has been given the opportunity to speak to the Ministry of Health (MoH) about CPC. They have previously carried out a needs assessment and have local information but have also been asked to position the need for CPC within existing global health priorities.*

## 2.1 Question 1: What Are Some of the Most Current Global Health Platforms and Priorities and How Do They Strengthen the Global Imperative for Children's Palliative Care?

There is a need for ongoing development of CPC globally, with some countries providing little or no access to PC services for children (Knapp et al. 2011; Downing et al. 2018). In the past decade several global policies, documents and resolution statements have been published that could have a direct impact on strengthening the global imperative for CPC. Key platforms include: The Sustainable Development Goals (SDGs) (UNDP 2019), UHC (WHOa) and the Astana Declaration of Primary Care (World Health Organization and UNICEF 2018). It is important that we link CPC to these developments in order to increase access for children and young people in need around the world.

Seventeen SDGs were adopted in 2015 to be achieved by 2030. SDG3 focuses on 'Good Health and Well-Being' in order to ensure healthy lives and promote well-being for all. Access to health coverage, including safe and effective medicines, is key, focusing on the individual. It is a person-centred goal, aiming to provide person-centred health care. To achieve SDG3 it will be necessary to be creative and innovative so that services are provided across the care continuum, including PC for children.

UHC, a key strategy for achieving SDG3, means that all people and communities can access and use the "*promotive*, *preventive*, *curative*, *rehabilitative and palliative health services they need*, *of sufficient quality to be effective*, *while also ensuring that the use of these services does not expose the user to financial hardship*" (WHO n.d.). There are three core components of UHC: (a) equity in access to health services so that everyone who needs them can access them, regardless of where they live, age, gender, condition, financial status, etc., (b) quality of health services, (c)

protection against financial-risk, ensuring that the cost of access to health care does not put individuals and their families at risk of financial harm (WHO 2019). WHO clearly recognises PC as an essential health care service and that ensuring equitable access to, availability of and usage of quality PC services is a fundamental and definitional component of UHC (ICPCN et al. 2018).

UHC cannot be achieved without access to PC, including CPC (ICPCN et al. 2018). Governments in most countries are working towards implementing/piloting schemes to ensure UHC. Central to UHC is reaching those who need it most, i.e., the particularly vulnerable, the poor, the young, the old, etc. All children deserve access to quality, affordable health services, which includes PC, and children who need PC have some of the greatest needs; they are some of the most vulnerable in society and CPC provision must be included in UHC. The implementation of UHC is in its early stages in most low- and middle-income countries (LLMICs), with pilot schemes being carried out and feasibility studies undertaken. In a statement on the 12th December 2018, the WHO reported that Africa is making strides to ensuring everyone, everywhere, has access to quality health care services without financial hardship.

The Astana Declaration on Primary Health Care (PHC) was passed in 2018. Within the declaration it states: "*We are convinced that strengthening PHC is the most inclusive, effective and efficient approach to enhance people's physical and mental health, as well as social well-being, and that PHC is a cornerstone of a sustainable health system for UHC and health-related Sustainable Development Goals*" (WHO and UNICEF 2018, p. 5). It is important that CPC, along with other services, is accessible to all across the continuum of care, and across a range of services. With families wanting to care for their children at home whenever possible, access to PC within the primary care setting is essential. Much of CPC can be provided within the PHC system as long as the systems are available and there is a good, ethical system for the provision of PHC, something which is lacking in many countries. The Lancet Commission report proposed a package of PC services that can be delivered at all levels of care—including primary care—as part of UHC (Knaul et al. 2017).

Thus, all three of these global health platforms, the SDGs, UHC and PHC, are interlinked and important in the ongoing development of CPC globally.

## 2.2 Question 2: What Key Palliative Care Platforms and Tools Are Available to Help Achieve the Global Health Priorities Inclusive of Children's Palliative Care?

In 2014 the World Health Assembly (WHA) passed a resolution on PC (WHO 2014). The resolution affirms that access to PC contributes to the realisation of the right to the enjoyment of the highest attainable standard of health and well-being (WHO 2014). It clearly recognises the need for PC across all disease groups, and all ages, including children, along with its inclusion within the provision of UHC. Essential to the resolution are: the need to support the strengthening of health systems including the integration of PC, with an emphasis on primary care, community and home-based care; adequate funding and human resources; inclusion of

PC as an integral part of education and training; ensuring access to medicines and the need to work in partnership to achieve this. As a resolution from the WHA, all WHO member states have to report back on the developments on PC within their country. This offers an excellent opportunity to hold countries accountable for increasing access to CPC.

A similar resolution was passed at the WHA in 2017 (WHO 2017), this time on cancer. It recognises that appropriate treatment, including pain relief and PC, can improve both patient outcomes and quality of life. Recommendations include working towards the implementation of the WHA resolution on PC as well as developing and implementing evidence-based protocols, including PC for children. Similar to the PC resolution, all WHO member states have to report back on the developments of cancer care within their country, again providing opportunity to hold governments accountable.

The WHO Model List of Essential Medicines for Children was initially developed in 2007, and has been revised and updated every few years since then with the 7th Edition published in 2019, being the latest (WHO 2019). This list recommends the essential medicines that should be available for children up to the age of 12 years. It is divided into a core and a complementary list, with the core identifying the essential medicines for all children, and the complementary list being for specific conditions for which specialised diagnostic facilities or training is needed. Within the EML there is a section for pain and PC, clearly showing the minimum requirements that should be available for all, including opioids. Alongside this guidelines were published by the WHO on the pharmacological management of persisting pain in children (WHO 2012) which are currently under review.

The Lancet, one of the world's leading medical journals with huge global impact and reach, hosts commissions on issues of worldwide importance that have often been neglected. Cognisant of the need to expand the evidence base and awareness of the global unmet need for PC, The Lancet launched the Commission on Global Access to Palliative Care and Pain Relief in 2014 (Harvard Global Equity Initiative 2014). The Commission report 'Alleviating the access abyss in palliative care and pain relief—an imperative of universal health coverage' (Knaul et al. 2017) was published in October 2017 and represents a milestone in the development of PC globally. The report acknowledges that people in many parts of the world are dying with little or no PC or pain relief and introduces the concept of SHS. It recognises that children and their families have specific PC needs that can easily be overlooked as the absolute number of those needing PC is low compared to adults, with children accounting for 9% of those who experience SHS. Whilst this includes one-third of all children who died in 2015, 98% of the 2.5 million children that die annually needing PC and pain relief are from low- and middle-income countries (LMICs). Whilst the report highlighted the unmet needs of children, the authors recognise and recommend that much more specific work is required with regards to CPC globally.

**Case Study: Navin**

*Navin lay struggling with both arms splinted and tied to the side of the bed, one of them with an intravenous line on it, when a PC doctor heard his screams of pain.*

*The weary-looking young surgeon behind the desk grimaced and said "Burns. Pain", hesitated for a moment, not sure if this was appropriate or not, but asked whether the PC doctor would like to see Navin. He had sustained 20% burns in an accident in the kitchen and the only analgesic that he had been given was an occasional infusion of paracetamol which the family could ill afford—they had begged and borrowed and bought the medicine. Whilst health care was supposed to be free for those below the poverty line, in reality the services were very limited and anything expensive such as infusions had to be bought. Thus Navin remained in pain, experiencing debridement and dressing changes with no analgesia, such that his mother prayed fervently that both herself and Navin be killed by lightning and the monsoon rains, rather than her child having to go through this agony.*

*With tiny bits of fentanyl going into the vein, 3 micrograms every 3 minutes, Navin became visibly quiet in about half an hour and subsided to sleep. With the reluctant agreement of the surgeon, he was started on 2.5 mg of oral morphine every 4 hours along with oral paracetamol. In addition, a scheme of analgesia was planned for every change of dressing. The treatment was paid for by the PC team, who had a comfort fund available for such situations.*

*For burns and other conditions, hardly any effective analgesia is used in most developing countries. Some PC teams are willing to offer support in these situations and then are often asked 'Why are you treating this person? This [pain] is not a life-threatening disease'. Life is not mere existence; it is something that carries meaning and if that meaning is threatened by suffering, we would consider that a life-threatening situation. Fortunately, the Lancet Commission of 2017, by giving us the phrase 'SHS' has made the issue easier for us to explain.*

## 2.3 Question 3: What Is the Definition of Serious Health-related Suffering?

SHS was developed by the Lancet Commission as a new concept for measuring the global burden of suffering that is not captured in mortality or morbidity statistics. SHS describes physical, psychological, social and spiritual suffering which cannot be relieved without medical intervention but can be reduced by PC and pain relief. SHS cannot go untreated and where health-care resources are inadequate, health conditions that would and should not be serious or life-threatening become so and will need PC. PC focuses on relieving the SHS that is associated with life-limiting or life-threatening conditions, or end-of-life (EoL) (Knaul et al. 2017).

## 2.4 Question 4: What Are the Challenges in Trying to Relieve Serious Health-related Suffering in Children?

The relief of SHS in children is not always seen as a priority, with existing measures of care focusing on extending life and productivity as opposed to QoL. There are many challenges to achieving universal access to PC to relieve SHS, and these are often particularly intense for children—see Table 2.1.

**Table 2.1** Challenges to achieving access to PC to relieve SHS (Knaul et al. 2017; Downing et al. 2018)

| |
|---|
| Opiophobia—there are even more myths about the use of opioids to treat pain in children than in adults, thus even if they are available they are not always used |
| A lack of trained and experienced personnel |
| A lack of attention to relieving SHS and PC—doctors want to cure, especially in children, and not to palliate |
| It is not always easy to get the voice of the child and their family heard with regards to advocacy etc. for CPC and relieving SHS in children |

In addition, parental pressure may come out of over-protectiveness. Particularly in the absence of adequate emotional support from the medical system, parents find it difficult to accept incurability and often put pressure on oncologists to pursue chemotherapy or other life-prolonging treatment. The combined effect of the misguided efforts of the medical system and the parental pressure is needless suffering. A study in India found that 38 out of 44 children with cancer had received tumour-specific treatment in the last month of life. Twenty-five of the 44 children received chemotherapy in the last week of life (Sinha et al. 2018).

## 2.5 Question 5: Does the Concept of Serious Health-related Suffering Have Implications for the Definition of Children's Palliative Care?

The Lancet Commission report recommends that the definition of PC and therefore also of CPC be refined "*to encompass health system advances and low-income settings where medical professionals often have the difficult task of caring for patients without necessary medicines*, *equipment*, *or training*" (Knaul et al. 2017, p. 10). The concept of SHS was developed within the context of EoL and also chronic or acute life-threatening or life-limiting disease, ill health and injury; therefore within the context of PC. CPC should be responsive to suffering of any kind and should seek to prevent and relieve physical, psychological, social and spiritual suffering.

During 2018 the IAHPC developed a consensus-based definition of PC. The objective was to develop a definition that focuses on the relief of suffering, and one that is also timely and applicable to all patients regardless of diagnosis, prognosis, geographic location, point of care or income level. The resulting definition follows a similar structure to that of the existing WHO definition (WHO 2002), comprising a concise introductory statement, the definition and recommendations to governments on how to achieve integration of PC. In summary PC is seen as the active holistic care of individuals across all ages with serious health related suffering due to severe illness, and especially of those near the end of life. It aims to improve the quality of life of patients, their families and their caregivers (IAHPC 2020).

## 2.6 Question 6: Is There an Essential Package of Palliative Care and Pain Relief That Should Be Available for Children's Palliative and Part of Universal Health Coverage?

Various examples exist of essential packages for PC, some of which address the packages needed for children as well as adults. The IAHPC essential practices for PC in Primary Care addresses the core components needed for provision of care in four domains: (a) Physical care needs; (b) Psychological/emotional/spiritual care needs; (c) Care planning and co-ordination issues and (d) Communication issues (De Lima et al. 2012). The European Association for Palliative Care (EAPC) primary PC taskforce have also developed a 'Toolkit for Primary PC' which is aimed at helping develop PC services at the community level for both adults and children (Murray et al. 2015).

As mentioned previously the Lancet Commission developed an essential package, which is a comprehensive package of cost-effective and affordable PC that should be included within UHC. It contains what is needed for safe and effective provision of essential PC interventions including those at the primary care level, and it also covers are range of domains including: (a) medicines, based on the WHO list of essential medicines it considers their doses and routes for both adults and children; (b) medical equipment; (c) human resources, which are dependent on the setting; (d) the need to complement health services by interventions for the relief of psychological, social and spiritual suffering: and (e) ensuring that families do not sacrifice basic needs in order to care for their loved ones. The package is not all inclusive, and is focused on people of all ages. There is a strong and decisive call to develop a package that is specific to the needs of children; however, it builds on the basic essentials that should be available to everyone (Knaul et al. 2017).

The essential package should be available to all children and families needing PC. The cost of implementing such packages varies although, according to the Lancet Commission, such a package could be provided in LMICs for as little as $3 per capita. As a crucial part of the spectrum of essential health services of UHC, it is important that an essential package of PC services is publicly funded as a component of UHC packages, such that PC services are universally available for free to promote equitable access for all children and their families. In order for this to become a reality the provision of CPC requires a multi-sectoral and multi-professional approach including a range of factors covering health, social, community and family care.

## 2.7 Question 7: Is Child Serious Health-related Suffering and Hence Children's Palliative Care Different in Low, Middle and High Income Countries?

Whilst the concept and definition of SHS and resultant need for PC is similar across different countries, the provision of care for the relief of SHS varies tremendously and hence the focus on PC as part of UHC.

It was clear that in Navin's case, the only way that his pain was going to be managed was when the PC team became involved, as they had the appropriate knowledge and skills to help him. However, had Navin been cared for in a high-income country (HIC), it is likely that his care will have been provided through the burns specialists and/or a pain team, both of which were not available to Navin. Due to limited resources in LMICs, health professionals may see a broader range of SHS in primary health care than in high-income countries (HICs) and the resources available to relieve this SHS will be different. However, despite this, it is still possible to manage SHS in LMICs as well as HICs, though the resources used may vary.

**Case Study: Anmol**

*A pleased warm smile on 15-year-old Anmol's face lit up the whole room when she saw the PC team, and replaced the perpetual frown which had taken hold of her face and her life. She had Juvenile Rheumatoid Arthritis and the team had been seeing her since the age of 10. In many countries, someone with her problem—pain and swelling in multiple joints—would not have to see a PC team. But for Anmol, she was seeing different doctors each time, and getting medication which would bring some relief for a few days but there never was any treatment plan and no protocol had been followed.*

*A phone call from the PC doctor to one rheumatologist resulted in a treatment plan. Anmol and her mother were eager for information and lapped up every word as the nature of the disease and plan of treatment were explained to her, as there had been a lack of access to information and shared decision-making for Anmol and her family.*

*With a combination of two disease-modifying medicines, her pain was brought under reasonable control. For a couple of years she was then able to attend school regularly and could also pursue her passion of painting flowers and trees. However, her pain seemed to be worsening and the nature of it had changed. It was not consistent, and the doctors wondered whether there was more to her reports of pain than rheumatoid arthritis. On exploration Anmol shared some of the challenges at home and eventually, following the social worker's interventions and legal help, her step-father left home. The PC team provided psychological and spiritual support which helped her and alongside this review of her pain medications, brought her pain under control again. Her mother said she was relieved and Anmol's pain could now be controlled and her smile 'reached her eyes'.*

*Anmol had been helped by the PC team since the age of 10 with regards to managing the pain associated with her condition. If they had not stepped in to manage her pain, it would for all practical purposes be like telling her 'Your condition is not life threatening enough yet. We shall wait a while till you are bed-bound, possibly with pressure sores and urinary infections killing you, when we will offer you tender loving care'. Even by the WHO definition* (WHO 2002), *prevention of suffering is part of PC.*

## 2.8 Question 8: What Is So Important About Universal Health Coverage and Children's Palliative Care?

As the need for CPC is recognised, the global priority to achieve UHC provides great opportunities for us to expand access. As mentioned earlier in the chapter PC, and therefore CPC is a core component of UHC. It alleviates suffering and

improves the QoL of children with life-limiting conditions (LLCs) and supports family members. The provision of CPC can reduce unnecessary hospitalisation and expenditure on ineffective health procedures which may cause more needless suffering. It supports some of the most vulnerable children and their families with extreme health needs. These needs affect QoL as well as their families' ability to bring in income. It can also reduce out-of-pocket expenditure. Where countries have made the political decision to include CPC within their health care packages, the outcomes are positive both for those living with and affected by life-limiting communicable and non-communicable conditions and for the health system. People suffer less and health systems can be more efficient (ICPCN et al. 2018)

## 2.9 Question 9: How Does Universal Health Coverage Vary Between Low- and High-Income Countries?

The provision of UHC will be dependent on the resources available and whilst there is a minimum standard which all countries, including LMICs, will have to attain, there is no maximum level of service provision. Some LMICs may struggle to provide the essentials of UHC and PC, whereas HICs may be able to provide over and above. Likewise, within LMICs there may be limited medicines available, whereas the spectrum provided in HICs may be a lot broader—therefore more complicated to use and the need for more training. PC cannot just be provided by specialist services but should be integrated into primary health care to ensure equitable access for adults and children. Within LMICs generalist services may not have the support of a specialist service as such services may not exist or have limited capacity; therefore, it is important that all social and health professionals have generalist PC training and can provide CPC—in LMICs this may be the only PC that the children can access.

## 2.10 Question 10: Are There Any Globally Accepted Indicators for Universal Health Coverage or Palliative Care That Are Linked to Children's Palliative Care?

The measurement of the impact of PC remains challenging. Indeed even developing indicators with regards to structure and process has taken time and there is ongoing work globally with regards to the development of PC indicators. The Lancet Commission report highlighted that indicators to accurately track the development of PC for both adults and children are needed at the national level around the world (Knaul et al. 2017).

In 2019, a 'Brief Manual on Health indicators to Monitor Global PC Development' (Arias et al. 2019; Arias-Casais et al. 2019) was published in order to try and inform developments and increase access to PC. In a systematic review published in 2019, over 165 indicators were identified that have been used within PC. In an attempt to narrow this down and identify a core range of indicators, the 25 best indicators to assess PC at the national level were assessed by a panel of experts. Whilst this

manual is a welcome step forward in the development of indicators, they do not address the issue of integration of PC within the health system, which needs to be addressed and is essential for CPC. The indicators are based around the public health framework (Stjernsward et al. 2007)—and come under the headings: Policy, Education, Use of Medicines, Service Provision, and Professional Activity. There is only one specific indicator within the manual for CPC and this is the "Number and type of PC programmes for children (estimate)"—however, many have implications for CPC, e.g., policy, national standards, allocation of funds, etc. Globally, the accepted indicator for PC is around opioid consumption of morphine equivalence, excluding methadone, per cancer death as reported to the International Narcotics Control Board (INCB). Much more work is needed not only on defining specific CPC indicators but also, even more importantly on their application, using them as tools for change.

## 2.11 Conclusion

Despite the ongoing need, we know that there are many children suffering needlessly around the world who have no access to PC services. Utilisation of global concepts such as SHS and of global platforms such as UHC can help to focus advocacy efforts, and ensure that they are centred around key and tractable issues. Not only can this offer support to governments in the roll out of UHC, but also in incorporating CPC as an essential component.

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# Children's Palliative Care Across a Range of Conditions, Settings and Resources

3

Megan Doherty, Regina Okhuysen-Cawley, and Lizzie Chambers

**Key Learning Points**

1. Palliative care (PC) should be provided for children with a variety of life-limiting conditions (LLCs) or life-threatening conditions (LTCs), which will vary depending on available resources.
2. Providing home-based PC is an important component of PC for children, feasible in high- and low-resource settings.
3. Hospital and hospice-based PC can provide critical support for children.
4. Prognostic uncertainty and continuation of life-sustaining treatment (called concurrent care in some countries) should not preclude a child from receiving PC.

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**Case 1: A Child with a Degenerative Incurable Illness in a Low-Income Country**

*Ali is a 14-year-old boy with a neurogenerative illness which has slowly deteriorated with acute complications such as aspiration pneumonia. Each episode of pneumonia is more severe than the last, but his condition is improved with antibiotics and supplemental oxygen. Several days after being discharged from hospital, Ali develops respiratory distress and his family rushes him to the local district government hospital. He has difficulty breathing and signs of poor cardiac perfusion (cool extremities and weak pulses). He is admitted to the intensive care unit and is provided with supplemental oxygen and IV antibiotics. There is no mechanical ventilation available at this hospital and Ali's parents cannot afford a private hospital. After several days, it becomes clear that Ali's respiratory function is deteriorating, and the intensive care team explains to his family he is expected to die in the next few days. The intensive care team consults with the children's palliative care (CPC) team about how to support Ali's family at this stage of his illness.*

## 3.1 Question 1. What Are the Possible Locations of Care for a Child in the Last Days of Life?

In both high- and low-resource settings, PC can be provided in a variety of locations, including home, hospice and in-patient health care facilities, and should be based on the child and family's preference as much as possible. This provision will depend on the resources available, but there are common principles that should be applied in all settings and resource levels. The World Health Organization (WHO) provides detailed guidance for health care planners and managers about the implementation of PC for children (WHO 2018).

### 3.1.1 Home

Previous studies report that parents often prefer to be at home for their child's death (Coyne et al. 2014). Home-based care generally involves visits from skilled nurses and other health care providers, with 24-hr phone access to a member of the care team (Weaver et al. 2016; Bona et al. 2011). Community-based PC teams have been implemented successfully in a number of resource-limited settings in the Americas, Asia and Africa (Kumar 2007; Harding et al. 2013). As a lower cost model of care, these may be well suited to resource-limited settings. Teams may also include trained volunteers and community health workers who provide regular visits to provide emotional, spiritual and practical support. Community health workers should be trained to screen for more severe symptoms and alert clinicians to the need for a visit from health care professionals.

### 3.1.2 Hospice

Countries from all income levels provide PC for children through free-standing hospices. These facilities offer end-of-life (EoL) care, respite, symptom management and bereavement support. Hospices generally have in-patient facilities where children can be admitted under the care of a dedicated team of PC nurses with oversight by a doctor (paediatrician/physician/general practitioner). They may also provide day and home-based care services. More of these facilities are needed throughout the world.

### 3.1.3 Hospital

In many children's hospitals, a paediatric PC team provides in-patient and outpatient consultation services. These teams provide multidisciplinary support and often include nurses, physicians, psychosocial professionals (counsellors, psychologists or social workers) as well as a variety of other therapists such as physiotherapists, occupational therapists, and play therapists.

PC can be provided throughout the health care system (Fig. 3.1). At a foundational level, all health care staff can be trained in basic PC principles, to

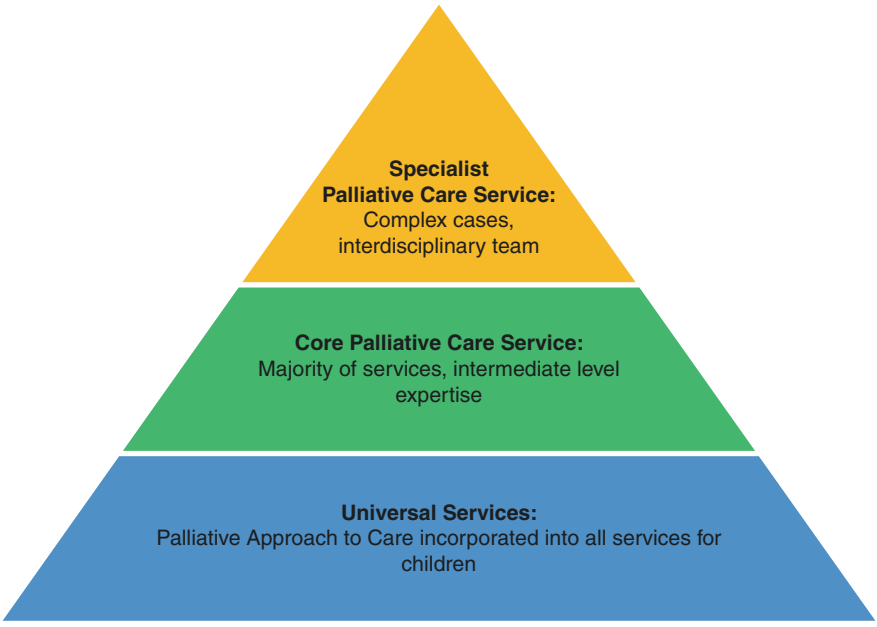


**Fig. 3.1** Different levels of PC service in paediatrics (adapted from McCulloch et al. 2008)

ensure that a palliative approach to care is adopted universally throughout the health care system. Core PC services form the majority of services for children. At this level, health care professionals will have intermediate levels of expertise. Complex cases may be referred to a specialist PC service, where more staff, training and resources are available. Specialist teams are generally inter-disciplinary, under the direction of a specialist PC physician and can be found in hospitals, hospices and primary care settings (McCulloch et al. 2008; WHO 2014, 2018). Within low- and middle-income countries (LMICs) there are however, few CPC specialist doctors or nurses, and limited specialist CPC training, thus those working within specialist teams may be developing their specialist role, whilst not actually having been trained at the specialist level (WHO 2018).

**Case Study**

*Ali's family has exhausted their limited financial resources during this hospitalisation and they share Ali's wish to go home for EoL care. The hospital's PC team develops a plan in coordination with a local home-based PC team.*

*Ali goes home by ambulance with oxygen therapy. He receives oral morphine and other medications to manage his symptoms. A nurse visits him daily and the team provides 24-hr telephone support. Two days later, Ali's mother calls the team at night, when he develops what she describes as wet and raspy breathing. The physician explains how to treat this respiratory congestion. The next day, Ali dies peacefully surrounded by his family.*

## 3.2 Question 2. What Are the Benefits and Challenges of Providing Home-Based Palliative Care?

Hospitals can be frightening for children, and home is often preferred as a familiar and comforting place. At home children can continue with their normal family activities, socialise with friends and may be able to go to school. In LMICs, the high out-of-pocket costs of hospitalisation mean that many families choose to return home once they learn that their child cannot be cured. Hospitalisation can mean that families have to bear the direct cost of medications, investigations and supplies, but also transportation, food and childcare at the same time as income and livelihood may be lost.

In resource-limited settings, seriously ill children return home from hospital after their parents are told that there is "nothing more that can be done". In reality, there is always something that can be done. Home-based care teams can provide basic resources and support families to feel less isolated and helpless in caring for their child. Whilst not everywhere has access to a home-based care team, families may still take their child home, due to cultural and/or financial reasons. Having a child in hospital can cause additional financial challenges for many reasons such as needing to have a parent caring for the child in hospital; parents being unable to work as they are at the hospital; the cost of being away from home and child care for the remaining children at home.

In high income settings, supporting families to plan for home-based care is associated with fewer hospital admissions and less intensive care usage, which generally correlates with improved quality of life (QoL) and reduced suffering in the terminal phase of illness.

When discharging a child home from hospital, it is important to establish contact with the local home-care team prior to discharge, to enable a smooth transition and ensure that there are adequate services to support the child's needs. The hospital team should develop an EoL care plan which incorporates the family's goals and wishes and ensures access to an emergency symptom kit to ease distress if symptoms develop. Emergency kits should contain the following medications:

- Morphine (or other opioid) for pain and dyspnea.
- Haloperidol for nausea and delirium.
- Hyoscine butylbromide (or other agent for oral secretions/congestion) for secretions.
- Midazolam (or other benzodiazepine) for seizures, catastrophic bleeding and acute respiratory distress.

Ensuring 24-hr phone access to a member of the care team is important to ensure that the family can manage symptoms and to enable the child to die at home if this is the family's wish.

It is important to ensure that PC services in these settings are integrated with other social and community-based care organisations to enable families to have access to comprehensive support.

**Case 2: A Child with a Life-Threatening Critical Illness in a High Income Country**

*Zara is a 13-year-old adolescent girl with myocarditis, placed on heart-lung bypass after suffering a cardiac arrest. Her heart has not recovered, and she has developed gastrointestinal and pulmonary haemorrhage as complications of bypass therapy. The intensive care team consults with the children's PC team to support Zara's family and the team as they decide how to proceed.*

*Since Zara is awake, a collaborative decision between the family and the medical team is made to proceed with insertion of a left ventricular assist device (LVAD), which attaches to the main pumping chamber in her heart and is connected to a console at Zara's bedside until a heart transplant can be performed.*

## 3.3 Question 3. Does Zara Have a Condition Where Palliative Care Is Appropriate?

PC should be provided in situations where a child's illness is life-threatening (where cure may be possible and PC interventions can help the child stabilise and tolerate treatment) or life-limiting (LL) (where the illness has no realistic hope of cure) (Wolfe et al. 2011).

In Zara's case, her condition appears to be life-threatening due to poor cardiac function, with the need for high-level invasive support. Despite the potential for significant improvement with heart transplantation, Zara has a substantial risk of death prior to transplantation due to organ shortages and declining health status which may make her ineligible for transplantation. Zara's intensive treatment has a significant impact on her physical health, as well as her and her family's psychological, social and spiritual health. PC can provide support in all of these domains, ensuring that suffering is addressed, and QoL is optimised.

Globally, the most common types of conditions where children need PC include neonatal conditions and congenital anomalies (Connor and Sepulveda 2014). Figure 3.2 illustrates the global distribution of disease groups where PC at EoL is needed (Connor and Sepulveda 2014).

In high income settings, children who need PC often have complex chronic conditions (CCC). CCC are defined as those conditions which are expected to last for at least 12 months, and involve either several organ systems or one organ system so severely that specialty tertiary paediatric care is expected to be required (Feudtner et al. 2001). These conditions are frequently associated with congenital and chromosomal anomalies, often requiring complex and costly treatments. Cancer, which is often strongly associated with PC, constitutes only about 20% of

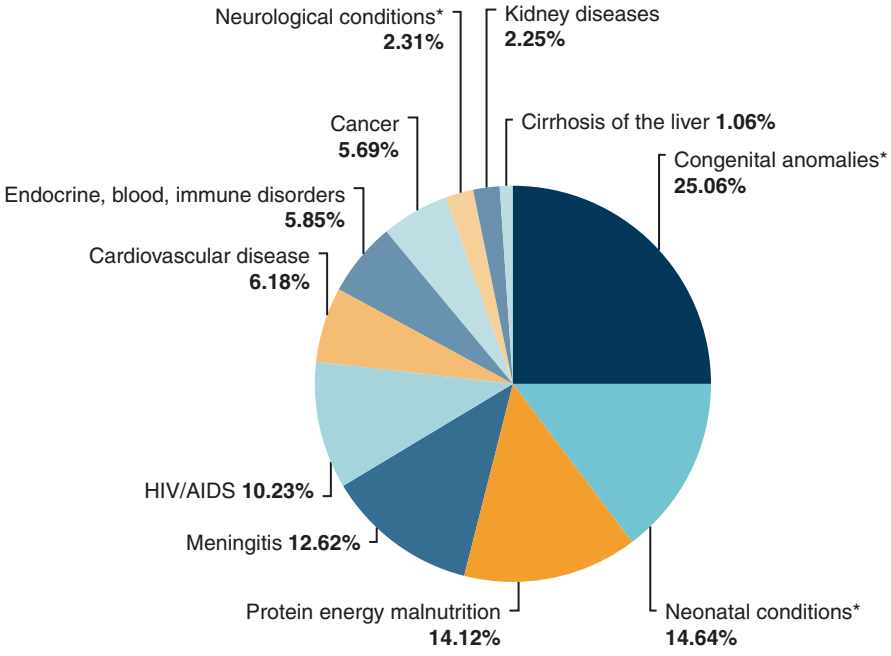


**Fig. 3.2** Distribution of children in need of PC at EoL. From Global Atlas of PC at EOL (Connor and Sepulveda 2014, p. 20)

children in high income settings, each Canada, who receive PC due to advances in curative treatment (Widger et al. 2007).

In resource-limited settings, there are conditions for which it is not possible to provide the potentially curative treatments that would be available in high-income countries (HICs). This may be due to the high cost of treatments, a lack of health care system capacity and the complexity and toxicity of the treatment regimens themselves (Amery 2009). In many parts of the world there are significant numbers of children with Human Immunodeficiency Virus (HIV)/Acquired Immune Deficiency Syndrome (AIDS), and PC is an essential component of their treatment, it may be provided alone or in combination with anti-retroviral therapy (ART). Severe malnutrition also contributes significantly to the global need for PC, with malnutrition estimated to contribute to 45% of all child deaths (WHO 2019).

**Case Study**

*Five days after an LVAD is placed, Zara suddenly develops an intense headache and becomes unconscious. Imaging reveals an extensive haemorrhagic stroke. Zara is placed on a respirator to support her breathing, but no longer has normal brainstem reflexes. The team sadly informs Zara's parents that she has had a devastating neurological injury and a joint decision is made to discontinue ventilatory and LVAD support. Zara's parents spend the night at her bedside. Zara remains comfortable as these interventions are withdrawn. The chaplain leads family, friends and staff in prayer as she dies.*

*After Zara dies, her family and friends spend time in her room. The chaplain and PC team counsellor stay with them, providing emotional support and helping with arrangements for the funeral. They continue to provide bereavement support to Zara's family, including her siblings, over the coming years.*

## 3.4 Question 4. What Are the Challenges in Determining Which Children Can Benefit from Palliative Care and How Can This Be Addressed?

Health care providers commonly cite prognostic uncertainty, clinician or the family's unwillingness to acknowledge the child's condition is incurable, and preference for life-sustaining treatment as barriers which limit access (Davies et al. 2008).

Prognostic uncertainty is common in children with life-threatening or life-limiting conditions, but should not preclude referral, since all children and families in this situation can benefit from the support provided by a PC team. Children's illness trajectories are typically much less predictable than adults with advanced incurable illness, due to lower incidence of cancer, the higher incidence of rare diseases and the relative health of unaffected organ systems (Wolfe et al. 2011).

CCCs include a broad range of conditions, including those caused by prematurity, congenital disorders of every organ system and neurodegenerative conditions. Prognostication is difficult with many of these rare conditions; and unlike adults,

children receiving PC often receive services for several years. In Canada, more than half of all children who received PC were alive after 1 year (Widger et al. 2007).

Zara's case illustrates the complexity of prognostication for children with serious illnesses. Children with LTCs may die suddenly when the child is still receiving active or intensive treatments aimed at cure or life-prolongation. Indeed, the use of life-sustaining treatment is not a contraindication to PC, and PC will support families facing the challenges of providing intensive treatment for their child. Clinicians often view PC as a distinct model of care which must be separate from aggressive medical management, but this is not the case.

Given the challenges faced by clinicians in determining prognosis and life expectancy, clinicians should present PC to families early, as an important component of their child's treatment which will enhance the care being provided, ensure comfort and provide holistic support for the whole family. Clinicians should not expect families to acknowledge the incurability of their child's condition to begin receiving PC.

In many cases, PC consultation occurs very close to the EoL, once clinicians feel certain that death is imminent. This can lead to a loss of opportunities for palliative intervention for the child and family (Hays et al. 2006), so the recommendation is for referral at the time when a child is diagnosed with a life-threatening or life-limiting condition to ensure that the child and family can be supported throughout the illness trajectory, whether the outcome ends in cure or death (American Academy of Pediatrics 2000). Zara received a PC referral early, when her condition was deemed to be life-threatening, allowing the team to develop a relationship with the family and provide support, even though at that stage Zara was receiving intensive cure-oriented treatment and her family was hoping for her to return to good health after heart transplantation.

Together for Short Lives have defined four broad groups of life-threatening or life-limiting conditions which identify the wide range of conditions likely to benefit from a PC approach. The groups are intended to be a helpful tool in determining which children could benefit from PC. These are not fixed and a child with one condition may be in more than one group, or in a different group depending on the resources available where they live (Together for Short Lives 2018). However it is a guide and can be useful in helping determine which children would benefit from PC (Table 3.1).

## 3.5 Question 5. What Are the Key Considerations of Providing Hospital-Based End-of-Life Palliative Care?

Hospital-based care, including intensive care, is becoming more frequent as children live with CCC for longer. Children may have prolonged and repeated hospital admissions, and in the United States, more than half (56%) of all child deaths occur in hospital with the majority in the ICU setting (IoM et al. 2003; Feudtner et al. 2007). In LMICs, deaths are much more likely to occur at home; in Africa, an estimated 80% of all child deaths occur at home (Amery 2009).

**Table 3.1** Four groups of life-limiting and life-threatening conditions (Together for Short Lives 2018, p. 11)

| | |
|---|---|
| Category 1 | *Life-threatening conditions for which curative treatment may be feasible but can fail,* where access to PC services may be necessary when treatment fails, irrespective of the duration of that threat to life. On reaching long-term remission or following successful curative treatment there is no longer a need for PC services. |
| | *Examples: cancer, organ failures of heart, liver, kidney, transplant and children on long-term ventilation.* |
| Category 2 | *Conditions where premature death is inevitable,* these may involve long periods of intensive disease-directed treatment aimed at prolonging life and allowing participation in normal activities. Children and young people in this category may be significantly disabled but have long periods of relatively good health. |
| | *Examples: cystic fibrosis, Duchenne muscular dystrophy and spinal muscular atrophy (SMA) Type 1.* |
| Category 3 | *Progressive conditions without curative treatment options,* where treatment is exclusively palliative and may commonly extend over many years. |
| | *Examples: Batten disease, mucopolysaccharidoses and other severe metabolic conditions.* |
| Category 4 | *Irreversible but non-progressive conditions causing severe disability leading to susceptibility to health complications and likelihood of premature death.* PC may be required at any stage and there may be unpredictable and periodic episodes of care. |
| | *Examples: severe cerebral palsy complex disabilities such as following brain or spinal cord injury.* |

For children who are seriously ill for a long time, hospitals often become safe and familiar locations, and for this reason, parents may wish to come to hospital when their child is at EoL. Hospitals, however, separate children from normal life and can be challenging environments in which to provide comfort-focused care. Staff are typically more familiar with providing all possible medical care measures to extend life. Also, many countries severely restrict visiting hours—even when death is imminent—which is upsetting to families.

Providing cardio-pulmonary resuscitation (CPR) in acutely and terminally ill children is not as helpful to seriously ill children as it is in previously healthy adults, with less than 27% of children with an in-hospital cardiac arrest surviving until discharge, a fact many healthcare professionals and families do not understand (Wolfe et al. 2011). For families who decide to forgo resuscitation, it is important that clinicians assure them that this does not mean that less care will be given, but instead that the focus of care will shift to comfort-focused treatments.

In situations where a continued life-sustaining treatment will not provide a meaningful QoL and may prolong suffering, it is ethically appropriate to withhold or withdraw these interventions. Although confusion is very common, this is very different from euthanasia or medical assistance in dying. It is important to involve the family in the decision-making process and it may also be necessary to involve religious or cultural leaders and a medical ethicist. After withdrawal of ventilatory support, children may die immediately or live for a brief period of time (minutes or hours, a few days, or longer); clinicians should plan for both possible outcomes to ensure appropriate symptom control and minimise distress.

**Case 3: A Child with an Incurable Condition in a Middle-Income Country**

*Ram was a 15-year-old young man who had been diagnosed with Duchenne muscular dystrophy (DMD). Ram's older brother, Joseph, died from DMD five years ago, at the age of 14. Ram had some learning difficulties which complicated his care. By the age of 12, he was no longer able to walk, and he had developed progressive heart failure from cardiomyopathy, a feature of DMD.*

*Ram's mother noticed that he snored loudly at night and had morning headaches. A sleep study was recommended by his neurologist, but the family was unable to afford it. Instead a friend loaned Ram a pulse oximeter which showed that his oxygen saturation dropped into the 70's when he fell asleep. Armed with this information, his mother was able to get a Continuous Positive Airway Pressure (CPAP) machine from the local district hospital. Ram hated the CPAP machine and would argue endlessly with his mother saying "Let me be! I know I am going to die, just like Joseph did!".*

*His mother provided all of Ram's personal care and was physically exhausted, so his neurologist referred Ram to the Paediatric PC Team at a local children's hospice. The team helped Ram and his mother to talk about the CPAP machine and explained to Ram's mother how QoL should be emphasised over quantity of life and the importance of respecting Ram's wishes. The hospice had a community-based care team who visited Ram at home, initially monthly and then more frequently when Ram's respiratory status declined. This improved QoL for Ram and his mother. Brief hospice admissions were arranged for respite, and the team engaged Ram and his family in discussions about his preferred location for EoL care.*

## 3.6 Question 6. What Support Can Palliative Care Teams Provide for Children with Progressive, Incurable Conditions in This Setting?

Children like Ram with incurable illnesses, which are progressive from the time of diagnosis should be referred for PC early in their disease trajectory to ensure that PC can provide the maximal benefit for the child and family. For some, especially in LMICs, they may never receive a definitive diagnosis, in which case they should still be referred for PC early.

Especially in resource-limited settings, where physicians may lack confidence and skill in explaining complex medical conditions to patients and families, PC teams are particularly valued for their communication abilities. Skilled communication can help families to fully understand their child's illness and treatment options, which allows them to make properly informed treatment decisions which match their wishes for their child. This is particularly relevant as families may go into significant debt to pay for costly and unnecessary treatments, leaving them unable to afford basics such as food or school for siblings. In Ram's case, his mother struggled to access a sleep study or CPAP machine. If Ram had been able to share his wish not to use CPAP earlier, these tests and interventions may have been avoided.

PC teams in all locations have expertise in the management of pain and other physical symptoms, using pharmacological and non-pharmacological approaches. Depending on locally available resources, the team may provide free or subsidised medications and medical equipment to patients.

Other community support services, such as respite care and education, can be organised. Respite, where available, provides a short break for parents or caregivers and can be provided in hospices, other health care facilities or in the child's home depending on the types of services available locally. PC teams can support children to continue attending school, by helping schools understand the child's condition and develop appropriate emergency treatment plans.

**Case Study**

*Following discussions, Ram's mother felt that providing EoL care at home was more than she could handle. A plan was made for Ram to remain at home as long as he was comfortable, with home visits by the PC team and to move to the hospice inpatient unit when he was reaching the end of his life.*

*A few days later, Ram was brought to the hospice with laboured breathing and low oxygen levels. He was offered an oxygen mask but refused it. He found it helpful to have a fan at his bedside and he received oral morphine which helped his breathing feel more comfortable. Ram's mother stayed in the hospice with him until he passed away the next day.*

*The CPC team did not have dedicated bereavement resources but kept in touch with Ram's mother via telephone and home visits. She expressed how helpful the team had been, in terms of practical support, helping her resolve her feelings of guilt, and empowering Ram to participate in decisions about his care.*

## 3.7 Question 7. What Are the Benefits of Hospice-Based Palliative Care?

Hospices offer an alternative to hospitalisation, which may be more appropriate for some children and families. Hospice staff are more likely to be familiar and experienced in the use of morphine and other essential PC medications, which may improve symptom management for children in these settings. In contrast, hospital clinicians in LMICs are often unfamiliar with the use of morphine.

Many hospices provide additional support services, including counselling, spiritual support, education and a range of therapies (e.g. physio, occupational, speech, art, music, play) depending on local resources and the needs of the children whom they serve. Through the course of the child's illness, families may have already developed trusting relationships with hospice providers, and thus hospices are often well placed to provide bereavement care in the form of counselling and peer support groups.

Similar to Ram's case, many hospices have expanded to provide out-patient care and home-care services to support children to remain at home as long as possible. Sustainable models of hospice care have been described in resource-limited

settings; most notably, South Africa which has a network of 60 hospices which support children and families. Many of these hospices have in-patient care units, as well as day care and "drop-in" centres (Drenth et al. 2018).

## 3.8 Conclusion

CPC should be available for children with a wide range of conditions and in a range of settings e.g. at home, in hospital and in a children's hospice where they exist. Often within CPC there is an uncertainty about prognosis, but this should not prevent a child and their family from receiving care. It is important to try and support the child and their families wherever they are and to integrate CPC into existing programmes such as community-based care organisations.

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# Holistic Assessment

4

Emma Al-Khabbaz, Busisiwe Nkosi, and Jane S. Nakawesi

**Key Learning Points**

1. A holistic needs assessment is vital in order to care for the child with a palliative diagnosis.
2. Holistic assessment is not a linear process but an evolution of reflection and continuous review parallel with the illness trajectory.
3. The history should include detailed information about their disease journey from diagnosis to fully understand the impact of illness on the child as an individual.
4. It is important to build rapport with the child. It will be difficult to gather information without first building a relationship with the child so keep this in mind when allocating time for the holistic needs assessment.
5. There are a variety of different elements to a holistic assessment and tools exist to help with these.

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**Case Study**

*Omar is a 9-year-old boy diagnosed with acute lymphoblastic leukaemia 3 years before. Having relapsed post chemotherapy he had a bone marrow transplant (BMT) in 2018 but unfortunately relapsed following BMT. Omar was referred to the PC unit for symptom control as he was starting to experience pain. The pain was mainly located in the testicular area, the site of local recurrence. Omar and his parents have been referred to the PC service where a holistic needs assessment will be conducted at the first meeting.*

## 4.1 Question 1. What Is a Holistic Assessment in the Context of Children's Palliative Care?

The term holistic is philosophically defined as the belief that the parts of something are intimately interconnected and explicable only by reference to the whole. Medically this encompasses the treatment of the whole person, taking into account psychological and social factors, rather than just the symptoms of a disease (Oxford Dictionary 2019).

The WHO's definition of PC for children categorically includes the holistic approach: 'The active total care of the child's body, mind and spirit, and also involves giving support to the family. It begins when illness is diagnosed, and continues regardless of whether or not a child receives treatment directed at the disease. Health providers must evaluate and alleviate a child's physical, psychological, and social distress' (WHO 2002).

In order to provide PC as outlined in the WHO definition, the health care provider is required to complete a holistic assessment of individual need which is multifaceted encompassing all elements of the physical, psychosocial and spiritual needs of the child (WHO 2002). This requires an exchange of information from the patient, their parents/caregivers with the clinical team, ideally a keyworker or person nominated to be taking a lead role in their PC (Contro et al. 2002).

The holistic assessment is not a linear process but an evolution of reflection and continuous review parallel with the illness trajectory (Baker et al. 2008). The child and family's need for information will develop and change through the course of the illness. This may relate to phases of physical decline such as an increased symptom burden or when entering the end-of-life (EoL) phase, with differing phases of the illness journey requiring additional support from the PC team.

## 4.2 Question 2. Are There Any Paediatric Tools Available to Help Complete a Holistic Needs Assessment?

Whilst there are assessment tools available for adult PC populations when conducting a holistic assessment, there is no standardised tool yet available in paediatrics (Ahmed et al. 2014). Encouragingly Together for Short Lives, a CPC organisation in the United Kingdom (UK), has piloted a holistic needs assessment tool (Hartley et al. 2016). We can however collate expert experience of the best approach to complete an assessment.

**Case Study**

*Omar was one of 4 boys, he was the third child. His oldest brother Yousef was 21 years old, Abdullah was 15 years old and he had a younger brother Nasser who was 8 years old. His family heritage was his mother was originally Egyptian and had come to the UK as a young girl and his Father was originally Sudanese. The family lived in central London. Omar was a very bright child and excelled in his school subjects. Omar enjoyed cooking and computer games. He really enjoyed school and had many friends. Omar's mother Nermeen had been studying nutrition and had a particular interest in natural remedies and food as an agent for healing. His father was a chef and had variable hours as a result. Omar was very close to his family particularly his younger brother Nasser who he used to play computer games with. At the weekends Omar would have his friends over and they would play computer games together and play football. They had a supportive local community. They had extended family in Australia, Egypt and Sudan. Omar enjoyed going out to local London cafes to watch football games with his father and spending time at home cooking meals with his mother.*

## 4.3 Question 3. How Would You Start Your Holistic Assessment?

As a starting point explain to the child and family the purpose of the holistic assessment and the length of time this may take. Gain consent for sharing this information with other health care providers involved in the care of the child.

### 4.3.1 Understanding the Child

It is essential that the child is known and understood as an individual person. This contextual information supports the health care professionals to perceive aspects of the child's life before their illness and understand their holistic needs. Questions about their personality, interests and what is important to them and their family before the diagnosis will gain perspective. The child and family will appreciate this genuine enquiry; taking the time to understand their child demonstrates empathy whilst enhancing the care provider's understanding of what the care needs are of the child and the family (James and Johnson 1997).

To understand better how to frame this enquiry it may be helpful to visualise the child in a multi-dimensional way. The child is at the centre of a network of relationships or relationship circle (Rolls 2004) (Fig. 4.1).

The child will have links to immediate family, more extended family, the community including school peers, and perhaps spiritual/religious communities. These relationships may be influenced by the social construct within which the child lives, their ethnicity and culture, socio-economic status will likely also have an influential role.

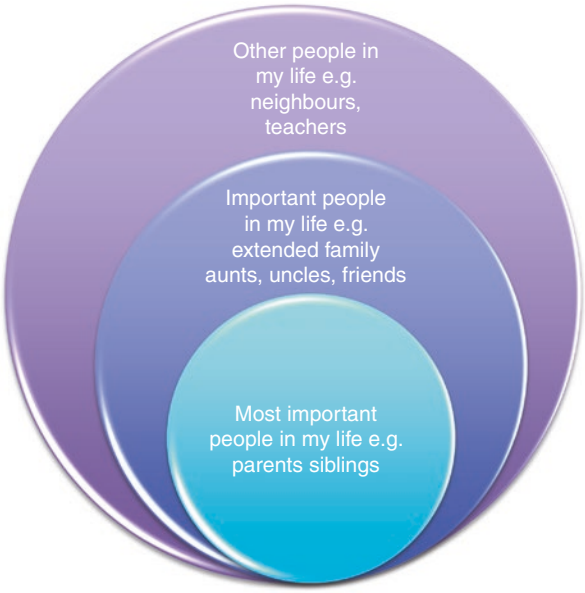


**Fig. 4.1** A child's relationship circle

### 4.3.2 Learning About the Family

The child lives and is dependent on the family for survival and the family is often the provider of care and source of information. The structure of the family should be understood, e.g., is it a nuclear family, extended family, single parent, same sex family, child headed family, etc. It is important to know the family structure as this may assist decision-making regarding treatment and care. Different tools are used to assess families, one such is a genogram. First conceptualised by Murray Bowen, genograms were later developed and popularised in the clinical setting by Monica McGoldrick and Randy Gerson. It is a pictorial display of a person's family relationships and medical history. It goes beyond a traditional family tree by allowing the user to visualise hereditary patterns and psychological factors that punctuate relationships. Information that can be recorded on a genogram includes family medical history, ethnic background, migration date, religion, education and employment status, etc. (Fig. 4.2).

The main elements that should be assessed in a family are as follows (Tindyebwa et al. 2017):

- The family's knowledge of the diagnosis and their reaction to it.
- How the illness has affected the siblings of the child.
- Beliefs, attitudes and expectations regarding treatment outcomes.
- Socio-economic status of the family.
- History of previous losses.
- Health status of all family members.

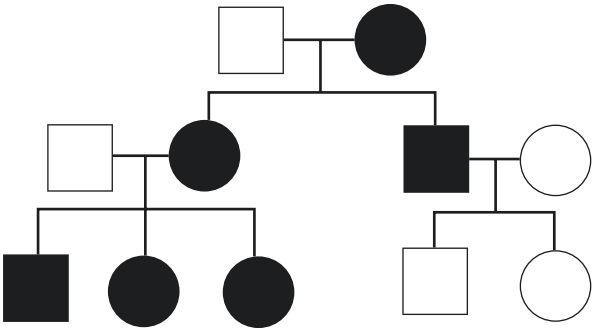

**Fig. 4.2** Genogram showing the occurrence of inherited diseases (123rf.com Image ID: 6414123)

### 4.3.3 What Is the Child's Role in His/Her Community?

Children are participants within their communities which may play an important role in their lives. It is important to assess the community and services to know what resources the family can access in caring for their sick child and to whom health care workers can refer to for extended support. These resources are often helpful in times of family crises. Holistic enquiry may include:

- Community groups working in the area.
- Health care facilities closer to family home.
- Places of worship, e.g., churches or mosques.

**Case Study**

*The diagnosis came after Omar presented with a feverish illness. He underwent a high-risk chemotherapy protocol. Parents were unsure as to whether to have a BMT but felt since this was the only curative option left available to them they had to try. The BMT came at a physical and psychological cost. Prolonged admissions and the physical side effects of treatments, namely pain and nausea had significantly affected Omar. He had become more fearful, angry and withdrawn. Helpfully Omar did have a trusting relationship with the specialist cancer nurse at the local hospital, who was seen as separate from the difficult experience of the BMT in the tertiary hospital. Omar and his parents were not sure if they wanted the PC team to be involved as they were seen as a part of the BMT/tertiary team.*

## 4.4 Question 4. Why Is It Important to Know the Child's Disease Journey?

The history should include detailed information about their disease journey from diagnosis to fully understand the impact of illness on the child as an individual. What is the child and family's understanding of the illness and prognosis? What is

important to achieve for the child and family at this point in the disease journey? What are their hopes and wishes? Gaining contextual understanding of how the disease has affected the child, their family and community may highlight the need for particular elements of support.

Relationships may already exist with healthcare providers such as community nurses, or primary paediatricians, who have already been a part of their disease journey and may have an important supportive role for the child and their family (Steinhorn et al. 2011). Information can also be taken from the relationships with the clinical team already involved and any challenges that exist.

Ascertaining from the patient/care provider what was helpful or less helpful through these interactions with the clinical team gives an indication of what communication patterns may best suit the family and how to make plans moving forward.

## 4.5 Question 5. How Would You Conduct Omar's Symptom Assessment and Physical Examination?

It is important to build rapport with the child. It will be difficult to gather information without first building a relationship with the child so keep this in mind when allocating time for the holistic needs assessment. When conducting the holistic needs assessment be cognitive of the well-being and energy level of the child. Listen and observe for verbal and non-verbal cues and always clarify information by summarising back to ensure you understand what the child has communicated (Rose and Amery 2009).

A full symptom assessment of the child should be carried out along with the impact these symptoms are having on their quality of life. As well as a detailed history of each symptom, various tools can be employed to complete these assessments. Symptom assessment will be discussed in other sections of the book so for further detail please refer to these chapters.

The child may require investigations to understand the cause of their symptoms and to guide management. Utilisation of investigations and certain therapies will be dependant, however, on resources available. For example, a child who gave a history suggestive of spinal cord compression in a high-income country would automatically have spinal imaging and likely go on to receive radiotherapy. In resource constrained settings, some of the diagnostic procedures are not available due to costs involved and the child may die even before an appropriate diagnosis was made. Management would therefore be mainly directed at symptom management.

**Case Study**

*Nermeen was finding it hard to keep on top of the housework and look after her 4 boys as she would like due to the demands of caring for Omar. Nermeen was particularly worried about her 15-year-old son Abdullah who had become more withdrawn. Nermeen felt this behaviour change was related to Omar's diagnosis. Omar and all of the children except the youngest Nasser (who understood Omar was very*

*ill), knew that there were no further curative therapies. The family were open in talking about this together. Omar's mood was often very low and he would become angry and depressed at times. He was particularly distressed about his appearance due to significant weight loss and some swelling of the left side of his face. He did not want to look in mirrors as a result.*

## 4.6 Question 6. How Would You Approach a Psychological Assessment in Omar?

The impact of the diseases in terms of the emotional and psychological health of the child and their family should be continually ascertained in order to understand better what the child and family need in terms of emotional support (Kane et al. 2004). Probing questions such as 'what worries you most?' can be helpful in gaining this information. Screening questions to understand the resilience of the child and family and how they cope should be discussed. This can alert the practitioner as to whether there is further need of support from the multi-disciplinary team (MDT) or specific therapeutic interventions from other professionals.

The psychological assessment should take into account the developmental age of the child. In the young child or infant the parent or carer can be asked about sleep disturbances, feeding issues, inability to be soothed or irritability. In the older child explore:

- Fears and anxieties associated with his illness.
- Separation anxiety.
- If the child is angry, withdrawn or depressed.
- Disclosure issues about his illness.
- Relationship with close family members and extended family.
- Presence of stigma in the community.
- Participation in sport/cultural activities.
- Neglect and/or abuse.

Children with psychosocial problems may also present with emotional disorders, e.g., anxiety, depression, personality changes that may include mood swings, poor interpersonal relationships and poor impulse control. Manifestation of behavioural disorders are also common, e.g., delinquency, disobedience. There may also be existing psychiatric conditions such as depression and anxiety (Tindyebwa et al. 2017).

Tools that can be used for this assessment in children include world charts, drawings/paintings, play dough, emotion chart, house and community plan, self-mapping, sentence completion, messages, monster technique and others.

**Case Study**

*Omar followed an Islamic faith which had an important influence on how he lived his life within his family and within his community. He enjoyed rituals around his religion such as fasting in Ramadan and daily prayer. Omar would go to the nearby*

*mosque for his daily prayers with his brothers until a time when he was unable to leave the house. It was important for Omar to be able to make a religious pilgrimage to Mecca which was facilitated by achieving symptom control with analgesics and providing a symptom management plan and medications for the journey. He would talk about God with his family trying to make sense of why he was going through all this suffering. At times, especially when his pain was severe, he would verbalise that he was angry for the suffering he was going through, and he worked through this anger by talking about it with both his parents. His parents stated soon after meeting the PC team that they would like to have a quick burial in line with their Muslim tradition and had already made arrangements for him to be buried in a Muslim cemetery when the time came of his death.*

## 4.7 Question 7. How Would You Approach a Spiritual Assessment in Omar and His Family?

Spirituality is a very individual concept that is experienced uniquely by each person. A screening question should be asked as to whether the child and/or parents are spiritual and if they have a religious belief. The child's spiritual needs may be very different to that of the family so it is important to make this distinction during your assessment. Support around spirituality is as important to a child's well-being as physical and emotional support. Spirituality can contribute considerably to how the patient/family perceive death and may inform their decision-making in terms of advance care planning and wishes and/or cultural practices around burial (Hexem et al. 2011). Please refer to Chap. 12 on spirituality for more information.

**Case Study**

*Omar's father was able to work flexibly which was of great help to the family as it meant they did not have financial concerns. Omar lived in a 4 bedroom house. This was set out over 3 floors. His bedroom was on the ground floor and the living room and kitchen were in the basement. When Omar became more fatigued as his illness progressed his father would carry him downstairs to the living room and his mother would sponge bath him in the bed. He declined having a medical bed in his bedroom. Omar did not wish to return to school as he felt his appearance had changed too much but was interested in continuing his learning, particularly home schooling. Towards the end of his life Omar and the family opted for EoL care in the hospice. This was due to increased symptom burden and care needs and they found the hospice to be a supportive environment which promoted family centred activities which improved his quality of life (QoL).*

## 4.8 Question 8. What Would a Social Assessment Consist of?

Financial or practical concerns should be openly discussed. It is common during a child's illness that there may be a financial consequence as parents often need to stop working or reduce hours in order to support their child (Cadell et al. 2012;

Bona et al. 2014). Social services may be offered by the state or charities, e.g., availability of grants for vulnerable people to help the family financially where necessary. Access to practical issues such as availability of transport, whether public or private, should also be included in the holistic assessment. In resource-limited settings where grants for vulnerable people are usually not available, such resources are drawn from the extended family. This is where drawing the genogram is very important as one can explore the social and financial abilities of the different family members and how they can help with the different social aspects of care.

The living environment should be discussed, i.e., the general layout of the accommodation. Discuss whether activities of daily living are hindered by disability related to illness. This will direct the health care provider as to whether a change needs to be made to accommodate the physical disabilities.

An educational assessment should also be performed, e.g.:

- Has the child been able to attend school?
- Is the school within reasonable distance to keep the child in school?
- What is important to the child, would they like to complete the exams for their year for instance if unable to attend school full time?
- What is their relationship with teachers/other children?

An example of a tool that can be used to assess both the child and family status is the CARES score (Meiring et al. n.d.). This tool is particularly useful in low- and middle-income countries (LMICs). It is a practical tool to assess the basic and overall functioning of a child according to Comfort, Access, Resources, Emotional functioning and Safety (CARES). The aspects for evaluation are divided into *Green* which means all is well, *Orange* which means there could be problems if one element is missing, therefore be ready to act, and *Red* which means danger and immediate action should be taken. A score of *two* is given for green, *one* for orange and *zero* for red. At the end of each aspect, scores are added together to compare against the total of each aspect requiring evaluation. A plan is then drawn up to best address areas that require support/management from the MDT (Fig. 4.3).

## 4.9 Question 9. How Do You Conclude Your Assessment?

After you have completed your detailed assessment, summarise the information you have collated back to the child and family as appropriate to ensure understanding. Ask if there are any outstanding concerns or questions. If actions were decided on in terms of investigations, referrals or recommendations for therapies these should also be summarised. A second meeting should then be arranged and contact details given of the PC team and relevant healthcare services. Consent should also be ascertained to share this information with other healthcare professionals (HCPs).

| Aspects requiring evaluation | | Green: Class I | Orange: Class II | Red: Class III |
|---|---|---|---|---|
| C-Comfort | C1 Basic needs: food, shelter, warmth (clothing) | Completely met. | Adequately met but at risk of not being adequate if challenged by stressor (e.g. mother hospitalised, winter weather, etc.). | Not met (child often misses meals, clothing or shelter inadequate, homeless, etc.) |
| | C2:Pain | None. | Mild – moderate. | Severe. |
| | C3: Symptoms other than pain | None. | Mild - moderate. | Severe. |
| | C4: Compliance with treatment (e.g. Antiretroviral therapy (ART), Tuberculosis (TB) medication, epilepsy, medication etc.) | Compliance with all treatment and clinic/hospital visits. | Generally compliant but compliance dependant on several factors (e.g. transport) which may not always be available. | Not compliant. |
| A-Access | A1: Transport | Own vehicle/transport always available. | Reliant on public transport but would be able to access transport in an emergency. | No transport services, no money for transport. |
| | A2: Healthcare | Easily accessible, good level of care. | Average access, reasonable level of care. | Not accessible (too far or very poor healthcare facilities). |
| R-Resources | R1: Primary caregiver | Good caregiver, loving, caring. | Satisfactory caregiver but may need extra help in a crisis. | Not satisfactory, caregiver not coping, elderly grandparent, child headed household. |
| | R2: Financial resources | Well resourced. | Adequate but could become a problem if challenged by an unforeseen crisis. | Inadequate. |
| E-Emotional | E1: Child (the patient) | Happy, content. | Coping but elements of stress anxiety or depression observed. | Uncontained, suicidal. |
| | E2: Caregiver | Happy, content. | Coping but elements of stress anxiety or depression observed. | Uncontained, suicidal. |
| S-Safety | S1: Abuse/ neglect | None. | Suspicion of abuse/ neglect/exploitation. | Confirmed abuse/ neglect/exploitation. |
| | S2: Environment | Safe. | Elements of concern but not life threatening. | Unsafe living environment posing a threat to survival. |

**Fig. 4.3** CARES Scale (Meiring et al. n.d. reproduced with permission)

## 4.10 Conclusion

A holistic needs assessment is an important tool to be utilised by healthcare providers in children with a palliative diagnoses. It includes a detailed history of the child and their relationship to their family, community and healthcare services. It assesses the physical, psychological and social aspects of care. It is a continual assessment

that evolves with the illness trajectory. The holistic needs assessment is vital to understand the child within their environment. It informs care needs and appropriate PC management plans.

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# Communication with Children and Their Families

5

Suzanne Boucher, Maha Atout, and Katrina McNamara-Goodger

**Key Learning Points**

1. Thoughtful and honest communication is central to the provision of good children's palliative care.
2. Being able to communicate well with co-workers, children, young people and families requires appropriate skills, practice and critical reflection.
3. Good communication requires mutual respect and deep listening to understand what is being said.
4. Barriers to good communication require the development of strategies for overcoming the same.
5. Considered use of electronic communication can enhance opportunities to communicate with children and families.

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**Case Study**

*Hala is a 12-year-old female diagnosed with nephrotic syndrome at the age of 8. Her father suffers from a severe chronic illness that prevents him working and has led to financial hardship for the family consisting of two parents, five sisters and one brother. Hala and her family live in Jordan. No extended family support is nearby.*

*Hala's doctor, Dr Nihad, reports being honest with the parents about the deteriorating nature of Hala's diagnosis and probable complications and not giving false reassurances. She adopted a gradual, step-by-step approach to communicate this news to the parents, focusing on short-term outcomes. She avoided anticipating events due to the unpredictable nature of children's diseases.*

*Hala's mother reports feeling confused by the lack of certainty. Dr Nihad did not talk about the possibility of Hala dying because she felt that providing hope for parents is essential. She explains that the Jordanian culture precludes 'hopelessness' but acknowledges that her conversations with parents differ according to the stage of a child's illness.*

*A nurse caring for Hala, Nurse Alaa, agrees that hope is essential for parents saying it would be 'too painful' for them to be told that death is a possibility. In her experience, parents do not want doctors who are too direct (honest) to treat their children. Hala's mother reported finding Nurse Alaa more approachable and asks her questions she is not comfortable asking the doctor.*

*On first hearing the diagnosis, Hala's parents experienced a period of denial. They asked a great many questions and wanted a second opinion. Dr Nihad understood this response and provided contact details for other physicians. Her mother also spoke to a parent who encouraged her to take Hala to a doctor who had cured their child. She did not disclose this to Dr Nihad for fear of her being annoyed.*

*In conversations with Hala neither Dr Nihad, Nurse Alaa nor her parents have been completely honest about her diagnosis, citing concerns that it will affect her treatment compliance. Despite this, Hala displays an excellent understanding of her condition, has had many interactions with other children with the same disease and was traumatised on being told of the death of her best friend, Sami, from the same illness. She asked her mother 'Am I going to be like him?' She also has free access to the internet and social media platforms through her smart phone.*

*Hala protects the adults around her from feeling sad by suppressing her own concerns about her illness but is more open when talking to her father. She was overheard describing her disease in detail to her father, who is not well educated.*

## 5.1 Question 1. Why Is Communication So Important in Children's Palliative Care?

Good communication is essential in children's palliative care and is a skill that must be learnt and practised. When honest two-way communication occurs, trust is built and good relationships are formed. This in turn reduces levels of anxiety in situations that are often traumatising, saves time that can be wasted when misunderstandings

arise and leads to better outcomes and ultimately an improved quality of life for children and families.

Hsiao et al. (2007) point out, the exchange of adequate information is crucial in communication between healthcare workers and parents. Families often report that being given adequate information is very helpful in alleviating their fears and doubts regarding their child's illness. Parents who receive relevant and adequate information about their child's ailment tend to feel more secure, even if they remain uncertain about a prognosis (Ringner et al. 2011).

Families should also be given information regarding their child's illness regularly and this is deemed to be a fundamental right of children suffering with critical illnesses (Bartel et al. 2000). Ringner et al. (2011) point out that parents are very appreciative of healthcare workers who give them honest and reliable information.

In addition, it is important to be sensitive to the unique and individual needs of families. Several researchers (including McGrath et al. 2007) have found that the amount of information that relatives are given largely varies between cases.

## 5.2 Question 2. Who Talks to Whom in Children's Palliative Care?

A person's role within the multi-disciplinary team dictates avenues of communication. Hala's case study provides examples of discussions between:

- Dr Nihad and her parents.
- Dr Nihad and Hala.
- Nurse Alaa and Hala.
- Nurse Alaa and Hala's parents.
- Hala and her parents, and
- Hala and her friends.

Additional stakeholders could include other siblings and grandparents, fellow team members, school teachers and community support groups. To ensure the best possible care for the child, principles of good communication should be applied in every conversation with every stakeholder.

## 5.3 Question 3. What Are the Principles of Effective Communication?

Communication is improved by employing certain skills which include:

- Giving the speaker your undivided attention.
- Listening deeply and responding appropriately to what is being said—not only with words but also through body language and tone of voice.

- Asking open ended questions.
- Paraphrasing and summarising what you have heard to ensure clarity and providing an opportunity to identify what may not have been said.

## 5.4 Question 4. What Needs to Be Communicated?

Remembering that communication is always a two-way process of talking and listening, much of what needs to be communicated will be built on the questions asked or concerns raised by the child and family.

In addition, it is important for the family to know what support there may be available to them, and, where appropriate and at the right time, the child and parents should be guided through advanced care planning. Failure to do so can have serious and unintended harmful outcomes where an emergency situation could arise and care given, or not given, without their expressed wishes being taken into account (Gauvin and Cyr 2015).

In our case study Dr Nihad provided Hala's parents with all the relevant information regarding her disease, its trajectory, possible treatment protocols and options, but was not completely straightforward about the prognosis, preferring to focus on short-term goals.

## 5.5 Question 5. What Can Happen When Information Is Purposely Withheld?

The family's cultural background often affects how much information they prefer to share with their children about the incurable nature of their diseases (Papadatou et al. 2002). There is often collusion of behaviours in conservative cultures which results in parents not revealing the true nature of their child's diagnosis. It is typically believed in such cases that the truth will take away any hope from the patient and enhance their suffering (Seth 2010).

The full extent of her disease trajectory was not fully disclosed to Hala leading to collusion behaviours from her mother, her nurse and Dr Nihad. In turn, Hala protected her loved ones by not communicating her concerns with them.

## 5.6 Question 6. What Factors Can Hinder Good Communication in Children's Palliative Care?

- *The use of medical jargon:* Parents in many studies have stressed that they wish the physicians and nurses would not use medical language and jargon (Contro et al. 2002; Davies et al. 2003; Kästel et al. 2011; Abib El Halal et al. 2013). They also emphasise the importance of not using English in non-English speaking countries for minority populations who do not speak English as their mother tongue (Majdalani et al. 2014).

- *Receiving mixed information:* Several studies have revealed that providing parents with inconsistent information regarding the illnesses of their child from different members of the care team worsened their distress and made them confused (Kästel et al. 2011).
- *Answering difficult questions:* Nurses reported that they found it hard to respond to family questions regarding their child's prognosis when they themselves have insufficient information about their condition. This largely shows communication issues between staff and between nurses and families (Papadatou et al. 2002).
- *Poor communication skills:* A number of studies have explored nurses' desires to improve their communication skills so that they can effectively manage families and their questions, particularly in cases of critical illness. This includes skills such as informing families about the sudden death of their relative, the transition from curative to palliative care, and talking about the 'Don't Resuscitate status' (Tubbs-Cooley et al. 2011; Peng et al. 2013).
- *Conflict with families:* Many researchers have explored conflict between nurses and patients' families, and it has been highlighted as a key factor hindering communication amongst nurses employed in child healthcare and looking after children in PC (Price et al. 2013). Health and social professionals have often felt the parents' grief and anger (Price et al. 2013).
- *Inability to uphold professional boundaries:* A further cause of conflict has been highlighted in research by Citak et al. (2013), namely the inability to uphold professionalism due to the extended time periods for which patients and their families remain in the hospital (Citak et al. 2013).

## 5.7 Question 7. How Do You Break Difficult or Significant News in Children's Palliative Care?

Most health care providers indicate a discomfort in delivering any painful or hopeless information to parents, since it largely affects their own emotions and well-being. It is thus recognised as one of the most challenging tasks facing health care professionals (Stein et al. 2019). However, this is an essential skill in providing holistic PC to children and families.

The following key steps have been identified in the process of breaking difficult news to parents and families (Amery 2016, p. 11):

1. Prepare for breaking significant news. Know what you are going to say and try to predict what questions may be asked.
2. Assess the awareness of everyone involved.
3. Find out how much the child and family know.
4. Find out how much the child and family want to know.
5. Break the news using the 'Warn Pause Check' (WPC) approach. This involves **w**arning that you have bad news to share, **p**ausing to give the listener/s time to prepare to hear the news and then providing the information in small pieces or 'chunks' at a time. Lastly, **c**heck to make sure they have correctly understood what you have said.

6. Respond appropriately to the child's and family's feelings.
7. Allow time for silence.
8. Manage denial and collusion.
9. Plan the next steps and ensure there is follow through.

Carefully consider where you plan to break the news to families and children, trying to provide a relaxed environment, with complete privacy and ensuring that the right people are present.

## 5.8 Question 8. How Did Dr. Nihad Communicate the News of Hala's Illness to Her Family?

The way professionals give bad news to families has an enormous effect on their acceptance of that news. Hala's doctor used several techniques to deliver bad news to her parents.

Dr. Nihad attempted to keep relatives informed about what was happening with Hala's treatment believing it was their right to be informed of everything. Furthermore, she attempted to avoid the parents blaming her if they were dealt bad news after being given false hope.

Dr. Nihad believed it was crucial to use a gradual approach to inform bad news, particularly when giving the initial diagnosis to eliminate any subsequent anger or denial. It is important to be aware that parent's emotions in the immediate aftermath of being given negative news can temporarily block or minimise their understanding of important information about the illness.

Dr. Nihad predominantly focused on explaining short-term outcomes to Hala's parents and refrained from speculating on possible future events, particularly given that Hala was still in the early stages of the illness:

**Case Study**
*This means that I do not discuss with them [Hala's parents] about these issues [long-term expectations] because she is still a long way from dialysis … so… I tell them instead about the transplant (Dr. Nihad).*

A gradual approach is often used by physicians when discussing the expected path of their child's disease, and this is particularly the case for conditions that are unpredictable. Nonetheless, Hala's mother indicated confusion and feelings of uncertainty following the discussion with Dr. Nihad.

**Case Study**
*She [Dr. Nihad] told me that it could be difficult to anticipate what could happen, she [Hala] could improve, but…she could be moved to dialysis (Hala's mother).*

Although Hala's mother indicated being given honest information by physicians regarding her daughter's illness, she did not approve of the direct, blunt, words that were used, regardless of the stage of the disease.

## 5.9 Question 9. Is It Possible to Combine 'Hope' and Honesty in Children's Palliative Care?

Many mothers have reported that hope has helped them to deal with the suffering that has been caused by their children's illness (Atout 2017). They believed hope to be a fundamental part of life, and without hope, they indicated that they would have found it harder to cope. Health care providers agreed with this, highlighting the significance of hope for parents.

Hala's parents stated that they wished doctors would give them more hope. Even though they continually insisted that the doctors were honest with them, they still deemed hope to be crucial and found it significantly hard to manage without having any. The issue of telling the truth about their diagnosis and prognosis is approached differently in various countries, balancing hope and honesty in communication is important in building trust between the family and their child's care team. It was stated by Hala's mother that she wanted to be given hopeful information, and she displayed communicative behaviours in which she constantly sought hope and reassurance.

Some parents ask lots of questions regarding their child's illness. Health care providers sometimes can feel irritated by this and fail to understand why the parents ask the same questions many times. Dr. Nihad reported that Hala's family asked a great deal of questions following the initial diagnosis.

**Case Study**

*Dr. Nihad provided the parents with optimistic information, highlighting the significance of upholding kidney function levels as much as possible. After informing the family that there was a significantly low likelihood that Hala could be fully cured, she continued to provide hope that Hala could be kept off dialysis for as long as possible.*

Dr. Nihad and Hala's parents agreed that the provision of realistic hope was crucial. It helped to promote compliance with treatment and to safeguard their feelings. Parents hold on to any hope following their child's initial diagnosis.

## 5.10 Question 10. How Do You Communicate Well Across Cultural and Language Barriers?

Cultural and language differences between the child, the family and the health care provider may be a barrier to good communication, negatively influencing the making of important decisions. This can leave families feeling ignored and isolated.

When these barriers exist, use interpreters for every important conversation and inform the interpreter about the nature of the conversation before speaking with the family. Remember that language as well as cultural differences can make it difficult to reach a shared understanding of the important issues, so finding an interpreter that is also culturally sensitive will help to navigate the conversation about the child's care in a way that respects the family's culture.

## 5.11 Question 11. What Do You Need to Know to Communicate Well with Children?

Being comfortable communicating with children does not come naturally to all and can become even more intimidating when talking to a child who is sick and may be dying. As with all communication, it is vital to show respect, to take time and to listen. Making the effort to build a relationship of trust between you and the child is paramount.

Key points when interacting with children include:

- Look critically at your own clothing and the location, especially when talking to younger children who may be frightened by white coats and clinical environments.
- Show respect for a child's personal space and warn them before doing anything invasive. Where possible, explain what you are doing and why in a way that they can understand.
- Familiarise yourself with stages of cognitive and language development for guidance on appropriate approaches and language use.

In our case study, 12-year-old Hala is approaching adolescence. Adolescents are interested in talking about themselves and their relationships with others, they are keen to assert their independence and prefer the company of friends to their parents. They can act in a more negative way and will often engage in conflict.

Adults in Hala's life should remember that, in addition to her illness, she is transitioning to adolescence, with all the hormonal changes, social anxiety and physical insecurities that this entails. Gentle questioning and seeking explanations for any difficult behaviour would be appropriate (Gable 2019).

## 5.12 Question 12. How Do Children Communicate?

Spoken language is not always possible. A child may be non-verbal, too young or simply unwilling to discuss certain topics. Careful observation of a child's body language during conversations gives clues to how they are feeling.

Taking time to observe children at play can also provide insights into their feelings, concerns and fears. Older children, like Hala, may also benefit from art and music therapy to assist them in releasing anxiety and expressing their thoughts.

**Case Study**

*Hala's mother shared how Hala once described a dream to her father in which she was with her dead grandmother who had 'come to take her and fly into the sky.' In her dream she also saw her friends Hosam and Raed who had died from the same condition as hers. They played together in the sky using stars. This conversation proved upsetting to her father.*

Hala's description of her dream is very revealing. She talks of her dead relatives and friends and describes them playing amongst the stars, showing that she is thinking and has anxieties about death and what will happen if she dies.

**Case Study**
*On occasion Hala wakes up in pain and shouts and screams very loudly. She can be demanding and become very angry. She asks questions that her mother and her doctor find difficult to answer. For example, she asks why her sisters' bodies are changing and hers is 'like a boy' and why their skin is white while hers is yellow. Dr Nihad answered these questions honestly, which caused Hala a great deal of sadness.*

Hala is clearly expressing her anxieties and her fears through these negative emotions and is asking questions that the adults in her life find difficult to answer.

## 5.13 Question 13. How Would You Communicate Difficult Information to Hala?

You know that Hala shows a tendency to protect the adults around her from feeling sad by suppressing her own concerns and anxiety about her illness, so she may find it hard to talk about her worries or concerns. Build in time to develop a trusting relationship with her before having the more difficult conversations.

It is also important to prepare yourself in advance, to find out more about her condition and management to date. Try to anticipate the kind of questions she may ask and prepare your answers. Think about the words Hala uses and try to integrate these into your conversation, using clear, jargon-free and readily understandable language, appropriate to her age.

Where possible, find a quiet, private space to have the conversation and make plans to ensure you are not interrupted during the conversation. Be honest, reassure her, give her permission to come back with questions and concerns, take your lead from Hala, do not make promises you cannot keep. Do not be afraid to say you do not know the answer to questions, but try to find out the answer and go back to Hala later.

Hala shows that she understands her condition in discussions with her father, she knows other children with similar conditions and has access to the internet to seek other information. It would be important to find out exactly how much Hala knows, and to dispel any misconceptions she has.

Ensure that you have enough time to talk and check out Hala's understanding of what she has been told or may have found out before moving on. Ask her how much she wants to know and do not force information on her that she does not want.

It has been proved that activity-based communication often helps young people feel more comfortable expressing their feelings and helps build trust. Think about any tools you may need to be available to help communicate effectively with Hala. For younger children, it may be toys, coloured pencils and flashcards. You know that Hala has a smart phone, so you could discuss which sites she uses to find out information and visit reliable sites together.

Remember to pay close attention not only to what Hala says, but also to what she is not saying and how she is behaving.

Communicating with children such as Hala about difficult issues, including their diagnosis and prognosis, is challenging. Thus some evidence based guidelines have

been developed for use in high-income countries (Stein et al. 2019), which are in the process of being adapted for low-and middle-income countries (LMICs). These can be helpful in preparing yourself to have these difficult conversations.

## 5.14 Question 14. How Do You Communicate Well with Bereaved Families?

Many people believe that the death of a child is more traumatic than any other bereavement (McLaren 2004). Remember that everyone experiences grief differently; there is no 'normal' or 'right' way to grieve. How people react will be influenced by many different things, including the age and personality of the individual, their cultural background and religious beliefs, previous experiences of bereavement and how they cope with loss.

The death of a child is a difficult, complex, emotional, physical and practical thing to deal with. You may find it helpful to prepare to support families by referencing the theories of bereavement and grief (see Chap. 16). Ensure you understand the process of grieving and understand the spiritual aspects of death and dying and the mourning ceremonies and traditions of the family's culture.

You may find it helpful to explore how the child's death has affected the family and to encourage the family to share their feelings and thoughts and to recognise that by remembering their child, the memories will help maintain a connection. Families may also find comfort in talking to others who have been bereaved.

Where there are siblings, each child needs the opportunity to explore their grief and for support as they mourn, while supporting the wider family to provide the child with love and support. It can help to keep the bereaved child's routine as normal as possible and to reassure them that they are not to blame for what has happened. Think about using age-appropriate information about what has happened and what is going to happen, especially in taking part in opportunities to say goodbye and commemorate the person who has died.

Children express their grief in a variety of ways. Death can be hard to understand at any age, but children probably will not have the experience or information they need to understand what has happened. They may have many questions about what happens when someone dies, whether the person is coming back, why they have died, what will happen to their body, whether other people are going to die and whether it is their fault.

## 5.15 Question 15. Is There a Role for Electronic Communication in Children's Palliative Care?

Electronic communication can include a wide range of approaches including email, electronic prescribing, telehealth, text messaging and video conferencing. It provides opportunities for sharing information immediately between families and professionals and enables access to up-to-date information on which to base

clinical decisions. It can help overcome the challenges of reaching out to children and families, especially those living in remote areas. It can help families reduce the time and costs of travelling to specialist centres. Such approaches can include, for example, electronic sharing of patient centred information about family preferences via email, improved efficiency such as electronic transmission of prescriptions direct to the pharmacy and videoconferencing, offering opportunities to enhance communication and sharing of good practice between specialist and local teams.

There is potential for children's palliative care (CPC) services to build on the experience of other specialities and countries such as Australia (Bursell et al. 2013), Canada and the United States of America (USA) (Bashshur et al. 2014) in using this technology. Project ECHO is a telehealth approach which links specialists with clinicians in local communities (Zhou et al. 2016) to allow knowledge sharing and can help overcome the isolation often experienced when delivering CPC.

In Hala's case there may have been an opportunity to link Hala with young people with similar conditions, or for Hala's parents to link with other families going through similar experiences, providing networks of support and helping to reduce isolation. The care team could also maintain contact through video calls, which can help capture non-verbal communications and the camera can also provide a diagnostic tool and assist with symptom management.

For professionals, being able to see as well as talk to patients and their families is often preferable to using the phone, but families may find the use of telehealth impersonal and sometimes unreliable, or difficult to use.

Technology is a useful support but is not a substitute for human skills. It can also be unreliable, and it should not replace face to face communication, especially when sharing significant news such as unexpected test results, or changes in the child's condition or prognosis.

While electronic communication can provide opportunities for new ways of working, this may require professionals to develop new skills and knowledge to work effectively with families and care must be taken to ensure that confidentiality is maintained through the system and that the use of electronic communication fits with local data protection legislation, policies and professional guidance.

## 5.16 Conclusion

Good communication is key within the provision of CPC wherever you live in the world. This requires mutual respect and understanding, as well as the need for sensitivity and recognising the unique needs of children and their families. It is important to have an understanding of how children communicate and how we can respond to that. We all have a role to play in communication and need to develop and practise our skills. There are a range of factors that can hinder good communication within CPC but these can be overcome, and there are guidelines to help with regards to communicating difficult topics e.g. breaking bad news.

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# Assessment, Prevention, and Treatment of Pain in Children with Serious Illness

6

Stefan J. Friedrichsdorf and Wendy Cristhyna Gómez García

**Key Learning Points**

1. Multi-modal analgesia acts synergistically for more effective paediatric pain and symptom control with fewer side effects than a single analgesic or modality in children with serious illness.
2. For complex pain situations at the end-of-life period "medications only" approaches are often insufficient, and just adjusting the choice, route, and dose of analgesics alone is not effective to provide excellent analgesia without over-sedation.
3. Integrative ("non-pharmacological"), psychological, and rehabilitative pain management strategies complement and enhance the analgesic outcome of patients suffering from pain.
4. It is considered unethical and inappropriate to perform elective painful procedures (such as wound dressing changes, blood draws, intravenous cannulations, injections, lumbar punctures, etc.) in children without providing evidence-based treatments to avoid or minimise pain
5. Offering a bundle of evidence-based modalities to all children every time prevents or reduces pain and anxiety caused by needles and procedures.

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**Case Study**
*Maria is a 9-year-old girl recently diagnosed with a bone tumour (Ewing Sarcoma), who is now undergoing cancer-directed chemotherapy, surgery, and radiation. On admission she appears to be in severe pain, and she needs to have blood taken but is reluctant to let any health professional near her in case they make her pain worse. She also requires a lumbar puncture and bone marrow aspirate.*

## 6.1 Question 1. What Is the Prevalence of Pain in Children Requiring Palliative Care?

More than 21 million children would benefit annually from a PC approach worldwide, and around eight million children 0–19 years would require specialised children's palliative care (CPC) (Connor et al. 2017). Children living with serious illnesses commonly experience pain, which is among the most distressing and prevalent symptoms (Goldman 2000; Hongo et al. 2003; Feudtner et al. 2011). Nearly all studies of pain and other distressing symptoms in CPC were undertaken in children with malignancies and show a significant symptom burden to this population (Wolfe et al. 2000, 2008, 2015; Friedrichsdorf et al. 2015). A prospective study describing patient-reported outcomes in paediatric patients with advanced cancer showed that 39% of all children were self-reporting high distress from pain, increasing to 58% at end-of-life (EoL) (Wolfe et al. 2015).

Pain in the largest group of CPC patients, children with progressive neurologic, metabolic, or chromosomal conditions with impairment of the central nervous system, has shown to be common, under-recognised, and undertreated (Friedrichsdorf et al. 2017). In this group of children, the majority experience daily pain, with nearly 22% experiencing pain nearly all the time (Friedrichsdorf et al. 2017).

The majority of children with serious illnesses experience different distinct and at times overlapping entities of pain pathophysiology concurrently and/or subsequently, explaining the need of advanced protocols providing multi-modal analgesia. The most common pain entities PC patients experience include acute somatic pain, procedural pain, neuropathic pain, total (psycho-spiritual-emotional) pain, chronic post-surgical pain, and/or chronic persistent pain.

## 6.2 Question 2. How Can We Assess Pain in Children?

The measurement of pain intensity is important and required to titrate and evaluate analgesics; however, it is a necessary oversimplification. To quote Carl L von Baeyer "*Measuring pain by its intensity alone is like describing music only in terms of its loudness*" (von Beyer 2006, p. 157).

For example, a child scoring 8 out of 10 on a pain scale would be treated very differently according to the underlying pain pathology (see Fig. 6.1). While for severe cancer pain morphine is likely to be appropriate, it would be contraindicated

**(1) Nociceptive Pain:** arises from the activation of peripheral nerve endings (nociceptors) that respond to noxious stimulation *[e.g. localised, sharp, squeesing, stabbing, or throbbing]*

- **Somatic**(for example, muscles, joints)
- **Chronic somatic** pain typically well localised & often results from degenerative processes (such as arthritis)

**(2) Visceral** (internal organs) *[poorly localised, dull, crampy, or achy]*

**(3) Neuropathic Pain:** resulting from injury to, or dysfunction of, the somatosensory system. *[burning, shooting, electric, or tingling]*

- **Central pain:** caused by a lesion or disease of the central somatosensory nervous system

**(4) Total Pain** suffering that encompasses all of a child's physical, psychological, social, spiritual, and practical struggles (Richmond 2005)

**(5) Persistent (Chronic) Pain:** Pain beyond the expected time of healing

**Fig. 6.1** Examples of pain pathophysiologies (Reproduced with permission from Stefan Friedrichsdorf)

for chronic pain or psycho-social pain. Of course, many children experience different kinds of pain at the same time.

A recent comprehensive review consistently reported an underestimation of paediatric pain by nurses and physicians, and the extent of underestimation tended to increase with pain severity (Seers et al. 2018). There are more than 800 validated paediatric pain scales so it can be hard for professionals to know which tool to use. Examples of commonly used tools are as follows:

*Multi-dimensional observational rating scales.*

For non-verbal and/or children younger than 4 years, pain is measured using observation rating scales. Examples include:

- *Infants*: e.g., CRIES (*C*rying; *R*equires increased oxygen administration; *I*ncreased vital signs; *E*xpression; *S*leeplessness) (Krechel and Bildner 1995) or Infant FLACC scale (Merkel et al. 2002).
- *Toddlers*: FLACC (*F*ace, *L*eg, *A*ctivity, *C*ry, *C*onsolability) pain scale [0–10] (Willis et al. 2003).
- *Intubated*: COMFORT (van Dijk et al. 2000) or FLACC (Willis et al. 2003).
- *Non-verbal, intellectually impaired*: r-FLACC (Malviya et al. 2006), Paediatric Pain Profile (PPP) (Hunt et al. 2004), or Non-communicating Children's Pain Checklist-revised (NCCPC-R) (Breau et al. 2002; Merkel et al. 1997, p. 294).

*Self-Assessment*

- *4–6-year-olds*: Simplified Faces Pain Scale (S-FPS) or Simplified Concrete Ordinal Scale (S-COS) (Emmott et al. 2017).
- *6–12 years*: Faces Pain Scale-revised (Hicks et al. 2001).
- *>10 years*: Visual Analogue Scale (Bailey et al. 2012) or Numerical Rating Scale (NRS) (von Baeyer et al. 2009) (Figs. 6.2, 6.3, 6.4, and 6.5).

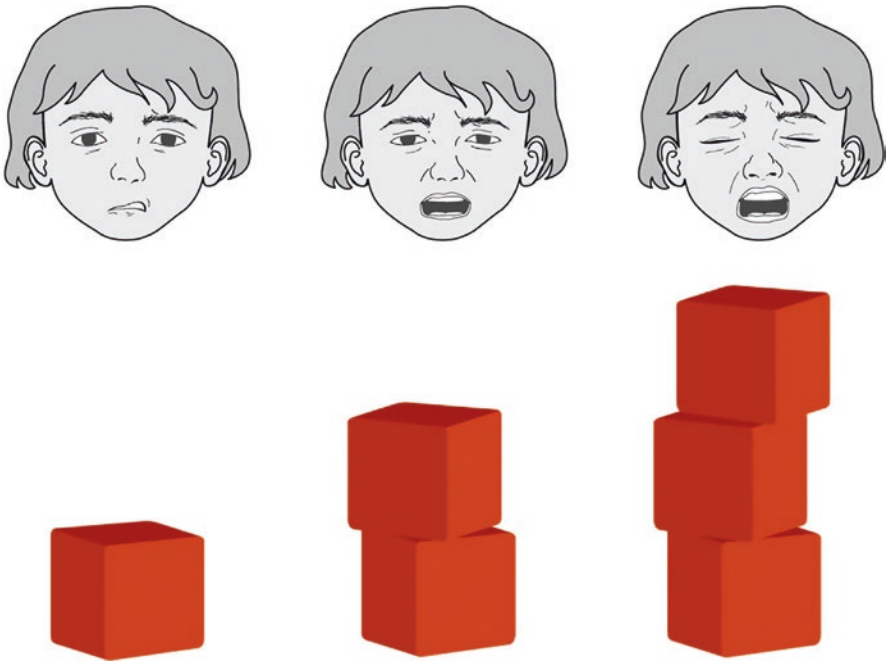

**Fig. 6.2** Simplified Faces Pain Scale (S-FPS) or Simplified Concrete Ordinal Scale (S-COS) for 4–6-year-old children (Emmott et al. 2017, p. 565; Reprinted from J Pain 18(5), Emmott AS, West N, Zhou G, Dunsmuir D, Montgomery CJ, Lauder GR, von Baeyer CL, *Validity of Simplified* Versus *Standard Self-Report Measures of Pain Intensity in Preschool-Aged Children Undergoing Venipuncture. 565, 2017, with permission from Elsevier*). Instruction: Ask child whether or not in pain. If yes, show Faces or building blocks to evaluate for "mild", "medium", or "severe" pain

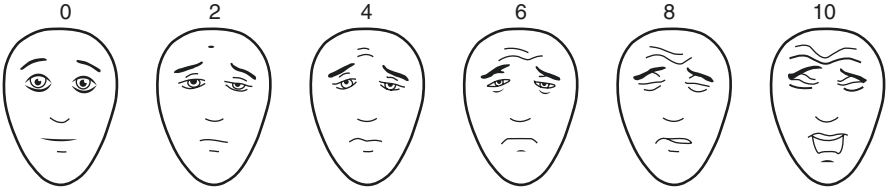


**Fig. 6.3** Faces Pain Scale—Revised (FPS-R) (Emmott et al. 2017, p. 565; Reproduced with permission from Stefan Friedrichsdorf) for children 7 year and older

**Fig. 6.4** Visual analogue scale (VAS)

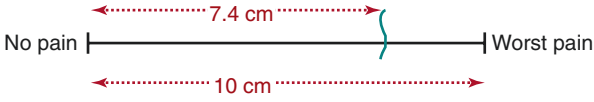



**Fig. 6.5** Numerical rating scale (NRS)

## 6.3 Question 3. How Can We Prevent and Treat Procedural Pain?

Untreated needle pain in children with serious illness, caused by procedures such as taking blood, injections, venous cannulation, etc., can have long-term consequences including needle phobia, pre-procedural anxiety, hyperalgesia, and avoidance of health care, resulting in increased morbidity and mortality (Taddio et al. 2009, 2010a). Current evidence strongly suggests that four modalities (3 for children >1 year) should be offered for elective needle procedures in order to reduce or eliminate pain experienced by children (Taddio et al. 2010a, b, 2015a, b; Friedrichsdorf et al. 2018a, b) (See Table 6.1). Offering four simple steps (and not just some of them) for all needle procedures for all children has now been implemented system wide in children's hospitals and paediatricians' offices on several continents (Friedrichsdorf et al. 2018a, b; Postier et al. 2018). However, topical anaesthetics may not be available in all countries, particularly in low-and middle-income countries (LMICs), but should be used as available.

Other modalities, including vapocoolants, ice, cool/cold packs, vibrating devices, etc., might be helpful, but currently have insufficient evidence for or against their use to reduce pain at time of injection and could be considered in places where numbing cream is not available or in addition to topical anaesthetics (Taddio et al. 2010b).

If adequate procedural analgesia and anxiolysis is not feasible with the above bundled modalities alone (e.g., because the child has been held down in the past and has become too anxious, or it was difficult to draw blood in the past, etc.), consider adding a child life specialist, referral to a child psychologist to offer cognitive behavioural therapy (CBT) to overcome needle phobia. For more invasive procedures, such as the lumbar puncture, she will require nitrous gas (Friedrichsdorf 2017) or propofol or ketamine, and for the bone marrow aspiration, propofol or ketamine. Where possible these two procedures should be done during the same period of sedation.

*Nitrous Gas Analgesia and Sedation:* Data reveals that children receiving nitrous gas before and during painful procedures have lower levels of distress, lower pain scores, were more relaxed, and many have no recollection of the procedure afterwards (Hockenberry et al. 2011; Pedersen et al. 2013; Tobias 2013). Nitrous gas concentrations between 40 and 70% can be can be titrated to achieve minimal sedation only, avoiding moderate sedation (Zier and Liu 2011; Livingston et al. 2017). Children receiving minimal sedation are able to respond to verbal commands, maintain and protect their airway, spontaneous ventilation, and cardiovascular functions are unaffected (American Society of Anesthesiologists 2002).

**Case Study**

*Maria rates her pain on the Faces Scale-revised as 8/10. The pain appears to be acute, nociceptive cancer and post-operative pain. She weighs 18 kg on admission.*

**Table 6.1** Prevention and treatment of needle pain (Friedrichsdorf et al. 2018a, b; Goubert and Friedrichsdorf 2019)

| *Offer a bundle of 4 evidence-based modalities to all children*: |
|---|
| 1. *Topical Anaesthesia "Numb the skin"* (for children 36 weeks corrected gestational age and older). Topical anaesthetic include 4% lidocaine cream (Taddio et al. 2017), EMLA-cream or needle-less lidocaine application via a J-tip® (sterile, single-use, disposable injector that uses pressurised gas to propel medication through the skin) (Lunoe et al. 2015a, b). |
| 2. *Sucrose* (Stevens et al. 2016, Gao et al. 2016) *or breastfeeding* (Shah et al. 2012) for infants 0–12 months (CHEO 2014). |
| 3. *Comfort positioning "Do not hold children down"*. Restraining children for procedures is never supportive, creates a negative experience, and increases anxiety and pain (Karlson et al. 2016). For infants consider swaddling, warmth, skin-to-skin contact, or facilitated tucking. For children six months and older offer sitting upright, with parents holding them on their laps or sitting nearby. |
| 4. *Age-appropriate distraction* (Uman et al. 2013), such as toys, books, blowing bubbles or pinwheels, stress balls, and using apps, videos or games on electronic devices. |

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## 6.4 Question 4. Can We Use a Variety of Approaches to Treat Pain in Children (Multi-Modal)?

Advanced pain management for children with serious illness often requires multimodal analgesia. This describes an approach of utilising:

- Multiple analgesic agents (such as basic analgesia, opioids, adjuvant analgesia).
- Regional anaesthesia (such as nerve blocks or neuraxial analgesia).
- Rehabilitation (such as physical therapy, motor graded imagery).
- Psychological (such as cognitive behavioural therapy).
- Integrative (formally known as "non-pharmacological") therapies (such as massage, hypnosis).

Using multiple modes of managing pain enables them to act synergistically for more effective paediatric pain control with fewer side effects than a single analgesic or modality (Friedrichsdorf et al. 2011; Friedrichsdorf 2016), see Fig. 6.6.

## 6.5 Question 5. What Are the WHO Step-1 Basic Analgesics That Could Be Used to Manage Maria's Pain?

The WHO Step-1 basic analgesia (World Health Organization 2012) includes the non-opioid analgesics paracetamol (acetaminophen), non-steroidal-anti-inflammatory drugs (NSAIDs)/cyclooxygenase-2 (COX-2) inhibitors. Of note, ibuprofen sodium (available over the counter in many countries) requires only half the dose, has analgesic effect within 10 min, and lasts longer (Moore et al. 2014). In

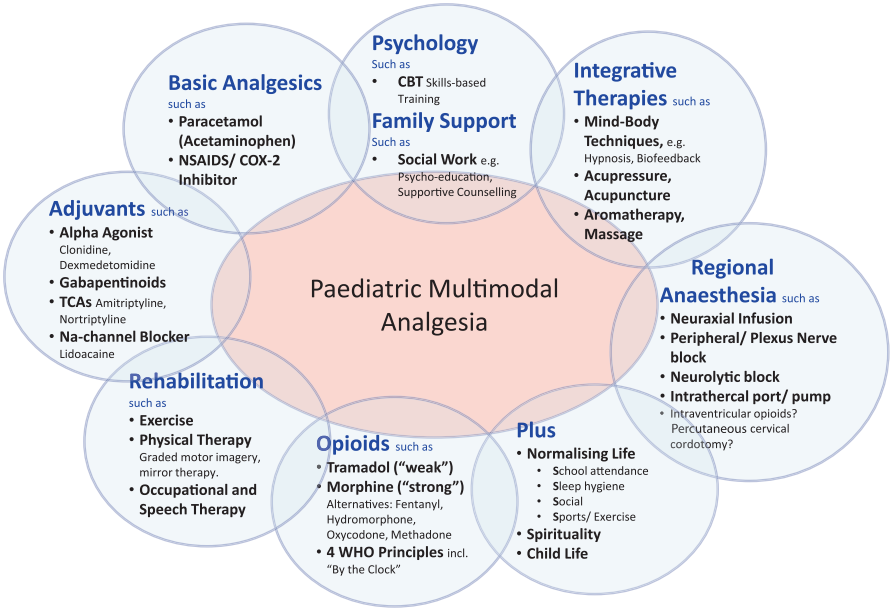


**Fig. 6.6** Multi-modal analgesia for children in PC (Reproduced with permission from Stefan Friedrichsdorf)

some countries, dipyrone (metamizole, not available in the United States of America (USA)) is included as a basic analgesia (de Leeuw et al. 2017).

- *Paracetamol (acetaminophen)* (10–15 mg/kg po/pr/iv every 4–6 hr; dose limit: <2 years: 40 mg/kg/day, >2 years: 75 mg/kg/day) is generally well-tolerated by children and lacks gastrointestinal and haematological side effects. Significant hepatoxicity (Heubi et al. 1998) is rare, but careful attention to dosing is paramount.
- *Ibuprofen* (5–10 mg/kg po every 6 hr; dose limit 2400 mg/day) has the least gastrointestinal side effects among NSAIDs that are nonselective for COX-2. It should be used with caution in individuals with hepatic or renal impairment, or a history of gastrointestinal bleeding or ulcers, and it inhibits platelet aggregation.
- *Ketorolac* has the advantage of IV administration, but it should be rotated to oral ibuprofen, as soon as tolerated (< 2 years: 0.25 mg/kg every 6 hr; >2 years: 0.5 mg/kg every 6 hr; max. 30 mg/dose; recommended dosing no longer than 3–5 days).
- *Celecoxib* (a COX-2 inhibitor) might be considered if classical NSAIDs are contraindicated (e.g., owing to bleeding risks, or gastrointestinal side effects). It does not display less renal toxicity compared to classic NSAIDs. Safety and efficacy have been established only in children 2 years of age or older and for a maximum of 6 months of treatment in juvenile rheumatoid arthritis (1–2 mg/dose; max. 100 mg every 12–24 hr) (Fig. 6.7).

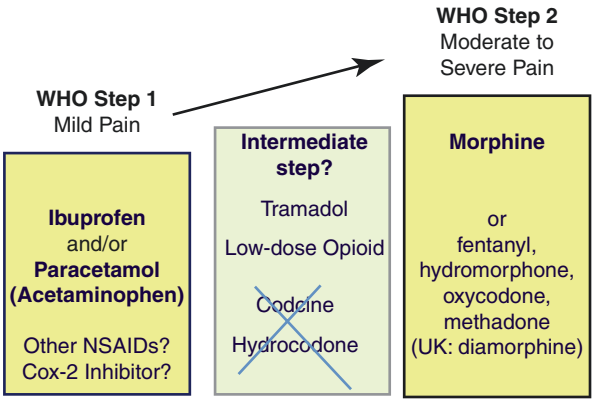


**Fig. 6.7** WHO pain ladder (World Health Organization 2012), with addition of possible intermediate step (Reproduced with permission from Stefan Friedrichsdorf)

## 6.6 Question 6. As Maria's Acute Pain Is Severe, What Other Analgesics, For Example, Opioids Could Be Used?

The WHO guidelines for managing persisting pain in children with serious illness recommended a two step analgesic ladder for use in children (WHO 2012). These guidelines were withdrawn in 2019 and are currently being revised, and there is ongoing discussion with regards to the use of a two or three step ladder. For medium-severe acute pain: WHO-Step 2, i.e., morphine will be added (WHO 2012). Other equally effective "strong" opioids include fentanyl, hydromorphone, and oxycodone (in the United Kingdom (UK) only: diamorphine). A switch from one opioid to another is often accompanied by a change in the balance between analgesia and side effects (Drake et al. 2004). The most frequently used opioid and gold standard in paediatrics for moderate to severe pain remains morphine. Opioid-associated side effects (e.g., constipation, pruritus, and nausea) should be anticipated and treated accordingly. For recommended starting doses, see Tables 6.2, 6.3, and 6.4.

Two potentially particularly effective multi-mechanistic opioids include the "weak" tramadol (for mild to medium pain) and "strong" methadone (for medium-severe pain) (Friedrichsdorf 2019). Tramadol appears to play a key role not only in outpatient surgery (due to its relative respiratory safety, more than 6000 paediatric tramadol scripts were filled at Children's Minnesota in 2018) but especially in treating episodes of inconsolability in children with progressive neurologic, metabolic, or chromosomally based conditions with impairment of the central nervous system (Friedrichsdorf 2019). However, the recent 2017 U.S. Food and Drug Administration (FDA) warning against paediatric use of tramadol does not seem to be based on clinical evidence (3 children died worldwide in 49 years, and therefore it appears far safer than any other opioid), and therefore unfortunately puts children at risk for unrelieved pain or distressing symptoms (Friedrichsdorf 2019).

Methadone, due to its multi-mechanistic action profile, is possibly among the most effective and most under-utilised opioid analgesics in children with severe unrelieved pain, especially in children receiving PC. *However, methadone should not be prescribed by those unfamiliar with its use: Its effects should be closely monitored for several days, particularly when it is first started and after any dose changes* (Friedrichsdorf 2019; Fife et al. 2016; Madden and Bruera 2017; Mercadante and Bruera 2018).

**Table 6.2** Usual starting doses for opioid analgesics (Doses for children >6 months of age are capped at 50 kg body weight)

| Drug (Route of administration) | Equianalgesic dose (parenteral) | Starting dose IV | IV:PO ratio | Starting dose PO (transdermal) | Starting dose controlled release |
|---|---|---|---|---|---|
| *Morphine* (PO, SL, IV, SC, PR) | 10 mg | Bolus dose: 50–100 mcg/kg every 2–4 hr Continuous infusion: 10–30 mcg/kg/hr | 1:3 | 0.15–0.3 mg/kg every 4 hr | 0.45–0.9 mg every 12 hr |
| *Fentanyl* (IV, SC, SL, transdermal, buccal) | 100–250 mcg | Bolus dose: 1–3 mcg/kg (slowly over 3–5 min—fast bolus may cause thorax rigidity) Continuous infusion: 1–2 mcg/kg/hr | 1:1 (IV to transdermal) | 12 mcg/hr patch (must be on the equivalent of at least 30 mg oral morphine/24 hr, before switched to patch) | n/a |
| *Hydromorphone* (PO, SL, IV, SC, PR) | 1.5 mg | Bolus dose: 15-20 mcg/kg every 4 hr Continuous infusion: 5 mcg/kg/hr | 1:5 | 60 mcg/kg every 3–4 hr | 180 mcg/kg every 12 hr—Currently not available in USA |
| *Oxycodone* (PO, SL, PR) | 5–10 mg | n/a | n/a | 0.1–0.2 mg/kg every 4–6 hr | 0.3–0.9 mg/kg every 12 hr |
| *Codeine* (not recommended) | 120 mg | n/a | n/a | 0.5–1 mg/kg every 3–4 hr | n/a |
| *Tramadol* (PO, PR) | 100 mg | IV not available in USA *[Bolus dose: 1 mg/kg every 3–4 hr Continuous infusion: 0.25 mg/kg/hr]* | 1:1 | 1–2 mg/kg every 3–4 hr, max. of 8 mg/kg/day (>50 kg: max. of 400 mg/day) | 2–4 mg/kg every 12 hr |

Reproduced with permission from Stefan Friedrichsdorf
Calculated rescue (breakthrough) dose: 10–16% of 24-hr opioid dose to be given every 1–2 hr as needed
*IV* intravenous, *PO* by mouth, *SL* sublingual, *SC* subcutaneous, *PR* rectal, *n/a* not applicable

**Table 6.3** Opioid analgesia for neonates and infants 0–6 months of age (Adapted from Hockenberry et al. 2011; WHO 2012)

| Opioid | Dose | Route | Dosing Interval (h) |
|---|---|---|---|
| Morphine | 0.075–0.15 mg (neonates 0–30 days)<br>0.08–0.2 mg (infants 1–12 months) | PO/PR/ SL | 6<br>4–6 |
| Morphine[a] | 0.025–0.05 mg/kg (neonates 0–30 days)<br>0.1 mg/kg (infants 1–6 months)<br>*Infusion (with PCA bolus of same dose):*<br>0.005–0.01 mg/kg/hr (neonates 0–30 days)<br>0.01–0.03 mg/kg/hr (infants 1–6 months) | IV/SC | 6<br>6 |
| Fentanyl[a] | 1–2 mcg/kg (neonates and infants 0–12 months)<br>*Infusion (with PCA bolus of same dose):*<br>0.5–1 mcg/kg/hr (neonates and infants 0–6 months) | IV/SC | 2–4 |

[a]The intravenous doses for neonates are based on acute pain management and sedation dosing information. Lower doses are required for non-ventilated neonates

**Table 6.4** Usual starting doses for patient (or nurse)-controlled analgesia (PCA) pumps—dose escalation usually in 50% increments both for continuous and PCA bolus dose (Department of Pain Medicine, PC and Integrative Medicine, Children's Hospitals and Clinics of Minnesota, USA)

| | Continuous Infusion [mcg/kg/hr] | PCA bolus [mcg] | Lock-out time [min] | Maximum number of boluses/hr |
|---|---|---|---|---|
| Morphine | 20 (max. 1000) | 20 (max. 1000) | 5–10 | 4–6 |
| Hydromorphone | 3–5 (max. 250) | 3–5 (max. 250) | 5–10 | 4–6 |
| Fentanyl | 1 (max. 50) | 1 (max. 50) | 5 | 4–6 |

Doses for children >6 months of age and are capped at 50 kg body weight (Reproduced with permission from Stefan Friedrichsdorf)

## Case Study

*You start Maria on the following medications according the WHO pain ladder:*

(a) *Paracetamol (acetaminophen): 15 mg × 18 kg = 270 mg Q6hr PO*

(b) *Ibuprofen 10 mg × 18 kg = 180 mg Q6hr PO*

*If NSAIDs are contraindicated* (e.g., *due to bleeding risk or neutropenia), consider COX-2 inhibitor celecoxib instead 2 mg × 18 kg = 36 mg PO BID*

*Since she is in severe acute cancer and post-operative pain, a strong opioid will be started immediately (and we will NOT wait on the effect of the basic analgesia)*

(c) *Oral morphine 0.3 mg × 18 kg = 5.4 mg Q4hr PO [=32.4 mg/day] plus 10% of total daily dose = 3.2 mg PO Q1-2hr PRN for breakthrough pain or*

(d) *Intravenous morphine 0.1 mg × 18 kg = 1.8 mg Q4hr IV [=10.8 mg/day] plus 10% of total daily dose = 1.1 mg IV Q1-2hr PRN for breakthrough pain or*

(e) *Morphine continuous IV or SC infusion plus PCA: 20 mcg × 18 kg = 0.36 mg/hr IV plus 0.36 mg PCA bolus [lock-out time 5–10 min, max. 4–6 boluses/hr]*

*If inadequate analgesia (and no severe opioid-induced side effects), titrate to effect in 50% steps (*e.g., *from 5.4 mg to 8.1 mg morphine Q4hr PO plus 4.9 mg Q1-2hr PRN) carefully observing for over-sedation or respiratory depression*

*Of note: Intravenous (IV) morphine can be swallowed by mouth*

*BID = twice per day*
*IV = intravenous*
*PCA = Patient (or nurse) controlled analgesia*
*PO = by mouth*
*PRN = as needed*
*Q6hr = every 6 hr*
*SC = subcutaneous*

*Three years later: Maria is now a 12-year girl with a 3-year history of her Ewing Sarcoma, which unfortunately did not respond well to cancer-directed chemotherapy, surgery, and radiation. Most recent imaging revealed several metastases in her lung, liver, and a large primary tumour in her left hip infiltrating the spinal cord and compressing her left sciatic nerve. For the last few days she reported constant acute background pain of up to 5/10 adequately controlled on oral morphine.*

*When asked in more detail, she reports "shooting pain hundreds of times per day from my buttock into my feet". She displays significant allodynia on examination in parts of her left lower extremity. She presents to clinic today with a sudden loss of some bladder control and sudden inability to walk on both legs.*

## 6.7 Question 7. How Would We Manage Maria at This Stage?

As always, when a child presents with a distressing symptom, we need to try to identify and treat the underlying disease process if this is feasible and within the goals of care for this particular child and family. Taking a medical examination and history is paramount.

Symptomatically, Maria's presentation is suggestive of metastatic spinal cord compression (SCC), a true PC emergency, which requires prompt action within hours. This diagnosis would be confirmed by imagery, where available although in places where this is not possible, her symptoms would confirm a clinical diagnosis. The new symptoms of lower extremity weakness and urinary incontinence could be resolved by repeated palliative radiation therapy aimed at the spinal metastasis and the administration of dexamethasone. Her neuropathic leg pain (likely due to her sciatic nerve compression) continued, but was improved by using a corticosteroid.

**Case Study**

*For Maria it was most important to learn active strategies and "to be in control", although she enjoyed the passive modalities (massage, aromatherapy) as well. Physical therapy and occupational therapy started working with her. She and her parents were able to follow the daily exercises as spelled out by her therapist, whom she saw once per week. Maria enthusiastically learned deep-breathing, biofeedback and self-hypnosis, and also used some relaxation "Apps" on her smartphone. She felt all of these modalities helped to reduce her pain, and they helped her feel more in control. After a few days she would usually go daily "to her happy place" in her imagination using deep-breathing as an induction technique.*

## 6.8 Question 8. How Can Integrative (Non-Pharmacological) Treatment Modalities Help Manage Pain?

Integrative modalities (sometimes referred to as complementary or alternative medicine) that have been described as effective in the management of paediatric pain include hypnosis, yoga, acupuncture, and massage (Bussing et al. 2012; Evans et al. 2008, 2010; Vas et al. 2016; Verkamp et al. 2013; Friedrichsdorf et al. 2009; Kuttner and Friedrichsdorf 2014; Hunt and Ernst 2011; Richardson et al. 2006; Friedrichsdorf and Kohen 2018). Active mind–body techniques, such as guided imagery, hypnosis, biofeedback, yoga, and distraction, impact pain modulation by engaging a number of mechanisms within the analgesic neuraxis. These techniques may have heightened importance in LMICs where there is a lack of diversity of pain management medications or the costs do not allow access to them and clinicians must be even more creative to fight against pain.

- *Relaxation therapy* might include progressive muscle relaxation and is used to help patients recognise and lower body tension associated with pain and anxiety. Children can be taught how to tense and relax different muscle groups in a relaxed and quiet setting, or to visualise a happy or peaceful scene and reduce body tensions that way.
- *Hypnosis* involves the cultivation of an altered state of awareness, leading to heightened suggestibility that allows for changes in a child's perception and experience, bypassing conscious effort. In hypnosis the clinician enters the child's world, engaging the child's imagination as the agent of change and creating alternate experiences to promote therapeutic change. In trance, the child addresses distressing symptoms utilising suggestions by the clinician for altering sensations, perceptions, and increasing comfort (Kohen and Olness 2011). Teaching hypnosis to children and adolescents is an extremely versatile skill which can be acquired by paediatric clinicians through formalised training workshops and practice (National Pediatric Hypnosis Training Institute 2019).

- *Cognitive and behavioural methods* include comfort measures such as pacifiers, massage, touch, music; distraction methods such as bubbles, counting, toys, video games; suggestion methods such as magic glove or magic blanket techniques; breathing techniques such as patterned, shallow, or deep-breathing; guided imagery; muscle relaxation; and hypnosis. A counselling relationship that supports coaching and problem-solving is an important component of these approaches.
- *Transcutaneous electrical nerve stimulation (TENS)* can be used independently or with other pain treatment modalities. TENS uses stimulation pulses delivered to skin electrodes around the area of pain. It is thought that electrical stimulation delivered by a TENS unit reduces pain through nociceptive inhibition at the presynaptic level in the dorsal horn, thus limiting its central transmission. The electrical stimuli on the skin preferentially activate low-threshold, myelinated nerve fibres. The afferent input from these fibres inhibits propagation of nociception carried in the small, unmyelinated C fibres by blocking transmission along these fibres to the target or T cells located in the dorsal horn. The TENS unit is compact, portable, and easily managed by many children. However, it may not be available in some LMICs.
- *Acupressure or Acupuncture* is an approach based on traditional Chinese medicine and has been used for thousands of years. Its utility in reducing post-operative nausea and vomiting is well documented. There are few studies in children.
- *Biofeedback* is a method of treatment that uses monitors to feed back to patients' physiological information of which they are normally unaware. By watching the monitor, patients can learn by trial and error to adjust their thinking and other mental processes in order to control "involuntary" bodily processes such as blood pressure, temperature, gastrointestinal functioning, and brain wave activity. Biofeedback requires trained instructors and specialised equipment.

**Case Study**

*Maria (current weight: 30 kg) has been on scheduled oral morphine 6 mg every 4 h plus 3.6 mg (10% of daily dose) once every 1–2 hr as needed. She required five as-needed doses of the morphine in the last 24 hr. However, she also continued to experience medium to severe neuropathic leg pain (VAS 6–8/10). The dose was increased by 50% to scheduled oral morphine 9 mg every 4 hr plus 5.4 mg once every 1–2 hr as needed. This improved her analgesia somewhat and she did not display any opioid-induced side effects. Therefore, the dose was increased by another 50% to 13.5 mg q4hr. On the following day, she reported being sick of taking so many tablets and liquids. She said "It takes too long for the extra dose to work" and complained they made her sleepy. Her parents were distressed to see her*

*in pain, and also expressed worries that she might get nauseous if she was taking too much oral medication.*

*A decision was made to rotate her opioid and route of administration to intravenous hydromorphone to achieve a better effect/side effect profile. Since she had displayed pruritus with fentanyl in the past, hydromorphone was chosen. She went home with a portable pump delivering hydromorphone 0.15 mg/hr plus 0.15 mg PCA bolus every 10 min with a maximum of five boluses per hour. On the following day she reported an average pain of 4–6/10, which was an improvement, and the PCA boluses to be helpful "for a short while". She initially used about 2 PCA boluses per hour (thereby tripling the continuous infusion) over 24 hr without any side effects, so the dose was escalated by 50% to 0.22 mg/hr plus 0.22 mg PCA bolus. She now reported improved analgesia. She stated "for me 4–5/10 is ok, because then I can sleep". Maria then used about 4–8 PCA boluses/day.*

*Maria started seeing a physical therapist and daily exercises inpatient, and then at home increased her activity level significantly. She started seeing a psychologist once a week who was experienced in cognitive behavioural therapy. The hospital chaplain connected with the spiritual leader of her faith community to arrange for home visits.*

## 6.9 Question 9. What Do We Mean by "Rotation of Opioids" and When Is It Used?

A switch from one opioid to another is often accompanied by a change in the balance between analgesia and side effects (Drake et al. 2004). Opioid rotation has been shown to be effective for improving analgesia management and lessening side effects (e.g., constipation, nausea, vomiting, pruritus, urinary retention, hallucinations, oversedation, etc.) in children (Drake et al. 2004; Fine et al. 2009). Differences between opioids in the balance between analgesic cross-tolerance level and the level of cross-tolerance to adverse effects can be exploited to clinical advantage. Switching opioids may achieve a more favourable balance between analgesia and adverse effects, hence the rationale for trial of a different opioid in the event of toxicity or inadequate analgesia (Lawlor et al. 2001).

## 6.10 Question 10. What Psychological Interventions Could Be Used to Help Manage Maria's Pain?

Anxiety, depressive, and behavioural disorders are early risk factors of chronic and complex pain in children and adolescents (rather than vice versa) (Tegethoff et al. 2015). At low levels of anxiety, higher pain is predictive of greater disability; however, highly anxious adolescents tend to function poorly regardless of their

level of pain (Cohen et al. 2010). Psychological treatments significantly reduce pain intensity that is reported by children and adolescents with headache, abdominal pain, and musculoskeletal/joint pain (Palermo et al. 2010; Eccleston et al. 2014). CBT led to significant improvements in pain coping, catastrophising, and efficacy that were sustained over time in adolescents with chronic pain (Kashikar-Zuck et al. 2013). CBT has been shown to increase grey matter in the prefrontal cortex of patients with chronic pain, and this increase in prefrontal cortical grey matter has been associated with reduced pain catastrophising (Seminowicz et al. 2013).

## 6.11 Question 11. What Is the Role of Physical Therapy/Rehabilitation in Managing Maria's Pain?

Physical therapy and exercise are key modalities in the treatment of patients with chronic pain and primary pain disorder, including in children with serious illness (Logan et al. 2012; Eccleston et al. 2003; Maynard et al. 2010; Palermo and Scher 2001; Lynch-Jordan et al. 2014; Sherry et al. 1999; Odell and Logan 2013; Lee et al. 2002). Patients with chronic pain usually have a lower physical activity level (Wilson and Palermo 2012) and physical activity has been shown to reduce the risk for depression (Jerstad et al. 2010). In patients participating in a rehabilitative pain programme, the rate of improvement in function was significantly more rapid than the decrease in pain (Lynch-Jordan et al. 2014). Particularly helpful as a rehabilitative approach, because it is not causing pain, is graded motor imagery (GMI), including mirror therapy, i.e., the process of thinking about moving without actually moving (Ramsey et al. 2017).

## 6.12 Question 12. How Can Our Spirituality Impact on Pain Management?

Religion, spirituality or life philosophy play an important role in the life of most parents whose children are receiving PC (Hexem et al. 2011) and screenings tools, such as FICA, have been successfully implemented into CPC (Borneman et al. 2010). A link between spiritual coping and quality of life (QoL) in adolescents with serious illness has been described (Grossoehme et al. 2013; Reynolds et al. 2013). More on spirituality can be found in Chap. 12.

**Case Study**

*Although Maria had better analgesia since her opioid hydromorphone was titrated to effect, she continued to display significant neuropathic pain in her leg and buttock. An adjuvant analgesic was added. Her weight was 30 kg and she started with amitriptyline 3 mg once at night, slowly increasing every second day by 3 mg to a*

*maximum of 12 mg once at night. She immediately reported improved sleep initiation, and felt more rested the next day. Her average pain score in her leg decreased over the next 2 weeks to about 2–3/10. No clear anticholinergic side effects were noted: She already reported underlying constipation (possibly worsened by the increase in opioid), which did require increases in laxatives, both stool softener "mush" and stimulants "push" plus occasional suppository. She occasionally displayed some urinary retention. However, this was not daily and might have been more attributed to her primary tumour. A rotation from amitriptyline to nortriptyline was not necessary.*

*Maria enjoyed a fairly good QoL, much better sleep and analgesia utilising her integrated therapies. She especially enjoyed deep-breathing and self-hypnosis. The scheduled opioid (hydromorphone) and amitriptyline for the last 4 weeks proved very helpful. Her dexamethasone was successfully weaned off two weeks ago. Regular assessment of her distressing symptoms with the paediatric Memorial Symptom Assessment Scale (MSAS 10–18) revealed some constipation, some worry, and increased pain.*

*Evaluating this further, in the last 2–3 days, she experienced recurrence of significant neuropathic pain in her left leg but did not mention it in order to not worry her parents and her younger brother. In addition to an increase of the hydromorphone by 50%, the decision was made to start her on gabapentin. The initial dose was 200 mg TID. Due to the acute worsening of her pain, her dose was escalated rapidly over 3 days from 200 mg QHS, to 200 mg BID and then 200 mg TID. In a less acute situation this titration may take 6–9 days. About a week later, she did not display significant side effects, but she also experienced only somewhat improved analgesia. The dose was escalated slowly to 400 mg TID and finally 600 mg TID, which was a dose where she seemed to have much better analgesia.*

*Although Maria's leg pain improved, as the gabapentin was increased, the maximum pain seemed to be localised over her left gluteus maximus muscle. A lidocaine patch (cut to fit) was administered 12 hr on- 12 hr off. Maria described it as "pretty helpful".*

## 6.13 Question 13. Which Are the Adjuvant Medications That Can Be Used in Managing Pain?

Adjuvant analgesics are *medications that, when added to primary analgesics, further improve pain control* (Table 6.5). Occasionally they may also be primary analgesics. They can be added at any step in the WHO ladder.

**Table 6.5** Adjuvant analgesia in neuropathic paediatric pain management (Pain Medicine and PC, Children's Hospitals and Clinics of Minnesota)

| Class | Medication | Dose | Route of Administration | Comments/side effects (see text for further details) |
|---|---|---|---|---|
| Tricyclic antidepressants (TCA) | Amitriptyline | Starting dose 0.1 mg/kg QHS, usually slowly titrated up to 0.5 mg/kg (max 1–2 mg/kg). | PO | Tertiary amine TCA; stronger anticholinergic side effects (incl. sedation) than nortriptyline. |
| | Nortriptyline | Starting dose 0.1 mg/kg QHS, usually titrated up to 0.5 mg/kg (max 1 mg/kg). | PO | Secondary amine TCA; anticholinergic side effects. |
| Gabapentenoids | Gabapentin | Starting dose 2 mg/kg QHS, usually slowly titrated up to initial target dose of 6 mg/kg/dose TID (max 300 mg/dose TID). Max. Dose escalation to 24 mg/kg/dose TID (max. 1200 mg/dose TID). | PO | Slow dose increase required; side effects: ataxia, nystagmus, myalgia, hallucination, dizziness, somnolence, aggressive behaviours, hyperactivity, thought disorder, peripheral edema. |
| | Pregabalin | Starting dose 0.3 mg/kg QHS, usually slowly titrated up to initial target dose of 1.5 mg/kg/dose BID (max 75 mg/dose BID). Max. Dose escalation to 6 mg/kg/dose BID (max. 300 mg/dose BID). | PO | Switch from gabapentin if distressing side effects or inadequate analgesia. Side effects: ataxia, nystagmus, myalgia, hallucination, dizziness, somnolence, aggressive behaviours, hyperactivity, thought disorder, peripheral edema; associated with weight gain. |

(continued)

**Table 6.5** (continued)

| Class | Medication | Dose | Route of Administration | Comments/side effects (see text for further details) |
|---|---|---|---|---|
| Sodium-channel blocker/local anaesthetic | Lidocaine 5% | Max. of 4 patches (in patients >50 kg) 12 hr on/12 hr off. | Transdermal patch | Not for severe hepatic dysfunction. |
| Glucocorticoid | Dexamethasone | 0.1–1.5 mg/kg (max. 10 mg) starting dose, then 0.1–0.25 mg/kg × 2/day (for <14 days) [Malignant spinal cord compression (adult dose): Dexamethasone 16–96 mg/day or equivalent]. | PO, IV | Add gastro-protective agent. |
| NMDA-receptor Antagonist | Ketamine (racemic mixture of S(+)/R(−) enantiomers) | IV: 0.06–0.3 mg/kg/hr<br>PO: 0.2–0.5 mg/kg TID-QID and PRN. | IV, PO, (sc, sl, intranasal, pr, spinally) | Typical side effects rare at low-dose, but would require benzodiazepine administration. |
| Alpha-agonist | Dexmedetomidine | Infusion: 0.3 mcg/kg/hr; titrate to max. 2 mcg/kg/hr. | IV | |
| | Clonidine | 1–3 mcg/kg Q4-6hr. | PO | |

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*Corticosteroids* are potent anti-inflammatory agents useful in both nociceptive and neuropathic pain. Reducing inflammation and peri-tumour oedema can relieve pressure on a nerve or the spinal cord, decreasing intracranial pressure from a brain tumour or decreasing obstruction of a hollow viscus.

In advanced disease, dexamethasone is the corticosteroid of choice because of its minimal mineralocorticoid effects and thus decreased risk of salt and fluid retention. Corticosteroids may also enhance pain control through the creation of a sense of euphoria. Most of the complications of steroid use (proximal muscle weakness, osteoporosis, and immuno-suppression) are long-term sequelae and are therefore not a concern at the end-of-life. Steroid psychosis is occasionally a problem that may require either rotation to another corticosteroid, cessation of the drug, or treatment with neuroleptics.

Dexamethasone has a long half-life and need only be dosed once a day. Typical doses start at 0.1–1.5 mg/kg (max 4–10 mg) daily.

*Tricyclic antidepressants (TCAs):* There is no data that TCAs provide better analgesia than gabapentinoids. The reason that TCAs are often the first adjuvant include once at night dosing options (versus three times per day for gabapentin) and the significant sedative effect at night, often improving insomnia. However, some centres choose gabapentinoids first, possibly followed by a TCA later in the treatment trajectory (especially in the setting of QTc-prolongation, already significant underlying constipation/urinary retention, etc.). It may take days or weeks for tricyclic antidepressants and/or gabapentinoids to show its analgesic effect, so in select cases this time period might be bridged by (or combined with) low-dose ketamine.

Adverse effects of all TCA include arrhythmia and anticholinergic/antihistamine effects, such as dry mouth, constipation, urinary retention, blurred vision, and sedation. Nortriptyline (a secondary amine) may be better tolerated than amitriptyline (a tertiary amine) because it has fewer anticholinergic side effects. The side effect of inducing sleepiness can be particularly helpful for patients with concomitant insomnia when the medication is given at nighttime. Occasionally, the anticholinergic effects of dry mouth, sedation, constipation, and urinary retention limit the usefulness of this medication. However, a slow increase over several days generally reduces the onset of side effects. A rotation from amitriptyline to nortriptyline might decrease the anticholinergic side effects as well.

Both agents, which also come as a liquid, are usually started at 0.1 mg/kg by mouth at bedtime (adult dose 5 mg) and increased to a max of 0.4–0.5 mg (max. 20–25 mg QHS). There is anaecdotal evidence that increasing beyond that dose (to 1–2 mg/kg (50–100 mg) does not result in increased analgesic effect. It may take 1–2 weeks to titrate up to an effective dose, and to determine if the analgesic therapy is working, although the induction of sleep will start much sooner. An electrocardiogram (ECG) to rule out QTc-prolongation/WPW-syndrome prior to initiation should be considered; however, administered doses for neuropathic pain are usually low compared to "antidepressant" dosing.

Of note, TCAs, despite being called "antidepressants", do not seem to improve anxiety/depression in children (as opposed to selective serotonin re-uptake inhibitors [SSRIs]).

*Gabapentin* (alternative: pregabalin) is commonly used in paediatric pain management. An initial low starting dose is 2 mg/kg/dose (max 100 mg/dose) once at night titrated to 6 mg/kg/dose (max. 300 mg/dose) three times/day (TID). For mild to medium neuropathic pain, the titration may take up to two weeks to avoid side effects. For severe pain, the titration may be significantly faster (1–3 days). If analgesia is inadequate, the dose may be titrated in steps up to 12 mg/kg/dose (max. 600 mg/dose) TID, then up to 18 mg/kg/dose (max. 900 mg/dose) TID, and finally up to 24 mg/kg/dose (max. 1200 mg/dose) TID. Side effects include lethargy, ataxia, nystagmus, dizziness, thought disorder, hallucinations, headache, peripheral oedema and myalgia; these side effects appear to be mitigated by slow dose escalation. Gabapentin, and to a lesser degree pregabalin, is commonly used in paediatric chronic pain management (Hauer and Solodiuk 2015; Edwards et al. 2016).

*Lidocaine:* Sodium-channel blocker, such as intravenous or subcutaneous (Peixoto and Hawley 2015; Seah et al. 2017) continuous lidocaine infusion appear to be effective in some children. A case series ($n = 5$) showed it to be effective after anti-GD2 antibody therapy in children with neuroblastoma at a dose of 1 mg/kg/hr (Wallace et al. 1997). Published dose recommendation for lidocaine for neuropathic pain includes 1 mg/kg over 5 min or 2 mg/kg over 30 min, then 1 mg/hr—target: 2–5 mcg/mL (Massey et al. 2002).

Side effects of intravenous lidocaine include allergic reaction (serious, but rare), and are dose related: numbness around mouth, dizziness, slurring of speech, hallucinations, muscle twitches, and seizures (Ferrini and Paice 2004). Lidocaine patches are for localised pain only. The patch can be cut to fit, and about 12 hr on/12 hr off. A contraindication would be severe hepatic dysfunction. Side effects include skin problems, such as irritation and redness.

*Ketamine* has been found to be effective for paediatric neuropathic and acute pain in low (sub-anaesthetic) doses both alone or in combination with opioids. Ketamine is unique among anaesthetic agents in that it does not depress respiratory and cardiovascular systems.

In low analgesic doses, the typical anaesthetic-dose side effects of ketamine (nystagmus, lacrimation, tachycardia, altered sensorium) are not usually seen, though there is limited paediatric data. Some paediatric centres schedule a low-dose benzodiazepine during the ketamine administration to avoid the rare potential psycho tomimetic side effects. There is evidence of significant opioid reductions in end-of-life paediatric cancer care after the initiation of low-dose ketamine. The advantage of ketamine in comparison to other frequently used adjuvant analgesia, such as anticonvulsants or antidepressants, is its rather immediate onset of action. Ketamine is often available in LMICs when other medications are not and may therefore be a medication of choice.

**Case Study**

*About 5 weeks later, after good QoL at home, Maria had a sudden pain crisis, which resulted in hospitalisation. Imagery showed a significant increase of her primary tumour and metastases. Her hydromorphone had been slowly titrated to effect over the weeks to the rather high dose of 7 mg/hr intravenously with 7 mg PCA boluses. It was now escalated to 10 mg/hr plus 10 mg PCA bolus. In weighing treatment options her oncology and PC teams decided to trial low-dose ketamine prior to a rotation of hydromorphone to methadone.*

*She was started on 30 mcg/min [1.8 mg/hr] and slowly titrated over 36 hr up to 150 mcg/min [9 mg/hr] of intravenous ketamine. She did not display any psychotomimetic side effects, so no benzodiazepine (which is ordered on an as-needed basis) was eventually given. Within about 6 hr Maria reported significant reduction of discomfort.*

*Over the next 24 hr her use of the hydromorphone PCA decreased from 55 boluses per day to 12 boluses per day resulting in an opioid decrease of over 60%.*

*She did not show any signs of opioid over-sedation, so the continuous infusion of hydromorphone was not decreased as it would have been, if any signs of over-sedation had emerged.*

*Three days later her hydromorphone was rotated to methadone. Six days later her intravenous ketamine of 4 mg/hr was switched to 25 mg oral ketamine q4hr. After discharge to home, 7 days later, Maria decided to change the ketamine to as-needed only, about twice per day, and discontinued the ketamine another week later, stating "I don't need it anymore".*

*Alpha-2-adrenergic* agonists such as *clonidine* or *dexmedetomidine* can be particularly effective adjuvant analgesics for both nociceptive and neuropathic pain. Clonidine can be given orally or transdermally or delivered intraspinally (some centres administer it intravenously). Side effects include lethargy, dry mouth, and hypotension. Dexmedetomidine can also be an effective adjuvant, leading to opioid-sparing. It has the advantage of not affecting respirations. However, its use is occasionally limited by side effects of hypotension and bradycardia, leading most institutions to restrict its use to intensive care units.

**Case Study**

*During Maria's hospitalisation, after initiation of low-dose ketamine her high-dose hydromorphone was rotated to methadone. She started to display renal dysfunction with a creatinine rise to 2.9, so the clinical team worried about accumulation of "the bad guy",* i.e., *the metabolite hydromorphone-3-glucoronide which acts in a nociceptive (*e.g., *increased pain) fashion and needs to be renally cleared. Some team members expressed worry about opioid-induced hyperalgesia, although Maria did not seem to display this. Additional hydromorphone PCA boluses decreased her pain, rather than increased it. However, her constipation had worsened. Since it was unclear how much of this was opioid-induced* versus *caused by her primary tumour which by this time extended well into her abdomen, rotating methadone (due to its significantly lower dose) with an NMDA-receptor blocking mechanism appeared a good choice.*

*Her very high dose of hydromorphone 10 mg/hr was rotated to, initially, 30 mg PO methadone/day. [Conversion range between 30 and 60 mg/day]. On day 1 she received 5 mg methadone q4hr PO; on day 2: 7.5 mg q6hr; and on day 3: 10 mg q8hr. The continuous infusion of hydromorphone was discontinued after the second methadone dose, and she continued on a PCA bolus only of 10 mg hydromorphone. On day 5, the methadone dose was increased to 13 mg q8hr—which was her final dose for the next 6 weeks. She continued initially with hydromorphone PCA. However, 2 weeks later she requested to be discontinued from all pumps (she had only required about 3–4 PCA boluses/day) and the PRN dose was switched to 4 mg methadone q4hr PRN PO in addition to her scheduled methadone 13 mg q8hr to a maximum of 3 doses per day. She had daily home*

*visits to ensure the safety of the regimen and Maria's parents were instructed to hold any methadone administration if she were to show any signs of opioid oversedation. Her constipation and her analgesia improved significantly over the next few days.*

*During Maria's last hospitalisation 6 weeks prior she was assessed by a paediatric anaesthesiologist and an epidural catheter was considered. If the above regimen would not have been successful, she would have been sent home with a tunneled epidural catheter. These often last for weeks at home and have been shown to be very effective. However, the team worked hard to keep her care regimen as simple, as least restrictive, and as effective as possible for ease of management at home. Maria continued to receive PC visits at home, occasional oncology clinic visits, and had a fairly good QoL. As her condition declined, her parents were able to coach her in using her well-practiced creative visualisation skills, which helped her focus on comfort and relax. She celebrated her 13th birthday at home with her family and friends and died peacefully 2 days later, at home.*

*Of Note: methadone should not be prescribed by those unfamiliar with its use: Its effects should be closely monitored for several days, particularly when it is first started and after any dose changes.*

## 6.14 Question 14. Where Available, What Role Does Regional Anaesthesia Have in Paediatric Pain Management

The majority of CPC patients with pain due to tissue injury to date unfortunately do not receive one of the most effective analgesic modalities, which would prevent and treat unrelieved pain with the least amount of side effects: Regional or neuraxial anaesthesia (Sen and Sen 2012; Cuignet et al. 2005; Bussolin et al. 2003; Hernandez et al. 2013; Shank et al. 2016; Cuignet et al. 2004). If tissue injury, such as tumour pain, of an extremity or the trunk requires hospitalisation, it must now be expected standard of care to ensure assessment of the infant, child, adolescent, by an anaesthesiologist for potential regional anaesthesia. Blocking pain nociception using a local anaesthetic such as bupivacaine, in some cases in conjunction with an opioid and/or alpha-agonist, can provide complete analgesia, without any of the opioid-induced side effects. Pain pathways can be blocked when anaesthesiologists trained in regional anaesthesia utilise central neuraxial infusions, peripheral nerve and plexus blocks or infusions, or neurolytic blocks (Rork et al. 2013). Occasionally implanted intrathecal ports and pumps for baclofen, opioids, local anaesthetics, and other adjuvants might be considered.

Benefits of regional anaesthesia include (Burns 2017):

- Significantly reduce or eliminate need for opioids.
- No systemic side effects.

- No sedation.
- No nausea.
- Minimal side effects with epidural (itching, urinary retention).
- Improved gastrointestinal motility.
- Less post-operative cardiac arrhythmias.
- Significantly reduced pulmonary complications.
- Significantly reduced delirium.
- Improved mobility that reduces rates of deep vein thrombosis (DVTs).
- Extremely high patient satisfaction.
- Patient is awake and can remember conversations with clinicians and family.
- Evidence for reduction of development of chronic pain and phantom pain.

Because the nociceptive nerves cannot be numbed independent of all the other nerves that receive local anaesthesia, there are side effects such as motor weakness, hypotension, pruritus, or urinary retention (Burns 2017). If the patient has breakthrough pain that breaks through a low continuous infusion of the local anaesthetic, a PCA bolus allows the patient to give him- or herself additional medication as needed, called patient-controlled regional analgesia (PCRA). Similar to an opioid PCA, the patient can use their PCRA button for breakthrough pain, but without the side effects caused by opioids. Patients can be sent home with a nerve block catheter, connected to a disposable pump or one that is returned to the hospital. There are no opioids in the infusion, eliminating misuse potential. That may lead to less adverse events, including sedation, delirium, sleep disturbances, and opioid-induced hyperalgesia.

Unfortunately, regional anaesthesia is not always available, particularly in LMICs, and where it is available, it may not be appropriate for children to be sent home while receiving such anaesthesia, e.g., with an epidural catheter.

## 6.15 Question 15. There Has Been a Lot of Talk About the Use of Medical Marijuana, How Does This Fit into Paediatric Pain Management?

Cannabis and medical marijuana (including Cannabidiol (CBD) and Tetrahydroxannabinol (THC)) lack any evidence to support its use for acute or chronic pain and cannot be recommended (Hill 2015; Deshpande et al. 2015). The updated American Academy of Pediatrics policy opposes marijuana use (Ammerman 2015), citing lack of research and potential harms including correlation with mental illness (Casadio et al. 2011), testicular cancer (Daling et al. 2009; Trabert et al. 2011; Lacson et al. 2012), decline in IQ (Meier et al. 2012; Moffitt et al. 2013), and increase risk of addiction (Meier et al. 2016). In our clinical practice, we do not support the use of marijuana (or medical cannabis) for a child with a primary pain disorder and a normal life expectancy. However, in children with life-limiting conditions, the

administration of medical cannabis is often requested by patients and their parents, and certainly might be considered on a case-by-case basis. Careful watching for frequent side effects (including pancreatitis, psychosis, etc.) is required.

## 6.16 Conclusion

The effective prevention and treatment of pain in children and teenagers with life-limiting conditions (LLCs) often requires intensive "multi-modal" pain control. Safe multi-modal analgesia (Friedrichsdorf 2016) may include one, several, or all of the following approaches: Pharmacology (e.g., simple analgesia and/or opioids and/or adjuvant analgesia), anaesthetic interventions (e.g., neuraxial analgesia, nerve blocks), rehabilitation (e.g., physical therapy, occupational therapy, sleep hygiene), psychology (e.g., cognitive behavioural therapy), and age-appropriate positioning and integrative ("non-pharmacological") therapies, such as breathing techniques, self-hypnosis, and distraction.

Also, it is inappropriate to perform elective painful procedures (such as wound dressing changes, blood draws, intravenous cannulations, injections, lumbar punctures, etc.) in children without evidence-based treatments to avoid or minimise pain.

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# Respiratory Care

7

Mary Ann Muckaden and Satbir Singh Jassal

**Key Learning Points**

1. Dyspnoea is a common symptom associated with advanced cancer and the level of dyspnoea is subjective and does not correlate with the condition.
2. There is a systematic methodology to assess dyspnoea and there is a strong correlation between psychological distress and dyspnoea.
3. Causes of dyspnoea should be reversed when there is a benefit, e.g., pleurocentesis and pleurodesis.
4. For terminal dyspnoea, morphine is the drug of choice, along with anti-anxiolytics.
5. End-of-life care (EoL) should be optimal to provide a peaceful death.

**Case Study**

*Elina is a 17-year-old unmarried girl who is in the 10th standard at school. She presented with complaints of breathlessness with a Modified Medical Research Council (MMRC) dyspnoea scale Grade 3 and excessive fatigue for the past 4 days.*

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## 7.1 Question 1. How Do You Assess Dyspnoea and What Are Some of the Different Scales That Can Be Used?

'*Dyspnoea is an uncomfortable abnormal awareness of breathing*' (Bass 1990, p. 200) and is a complex symptom which is very subjective. Patients perceive the symptom and its effect on their activities of daily living differently. It is important for health professionals to turn this information into something which can be quantified, so as to decide on different management options and assess improvement or deterioration. To this end, experts have created a number of scales to measure dyspnoea.

Measuring breathlessness in a young child below the age of 8 relies heavily on the report of the mother. The same is true for a child with a cognitive impairment. Some commonly used complex scales include the asthma score (AS) and paediatric respiratory assessment measure (PRAM) which are thought to be the most accurate. However, Eggink et al. (2016) have compared the various scales and found them to be inadequate in terms of validity.

More commonly used with children and adolescents are the single-item patient-reported scales, the verbal descriptive scale (VDS), numerical rating scale (NRS, e.g., 0—no breathlessness to 10—worst breathlessness possible) and the modified Borg scale (Fig. 7.1) (Wilson and Jones 1989). Wysham et al. (2015) have compared the NRS with the VDS and conclude that these scales are practical and clinically intuitive and can be easily integrated into routine clinical practice and provide cut off points that can be used to screen for inclusion in clinical dyspnoea management trials. The Dalhousie scale (Fig. 7.2) (Pianosi et al. 2014) is also used and takes into consideration three common themes: (a) throat closing, (b) chest tightness and (c)

| | |
|---|---|
| 0 | NOTHING AT ALL (just noticeable) |
| 0.5 | VERY VERY SLIGHT |
| 1 | VERY SLIGHT |
| 2 | SLIGHT |
| 3 | MODERATE |
| 4 | SOMEWHAT SEVERE |
| 5 | SEVERE |
| 6 | |
| 7 | VERY SEVERE |
| 8 | |
| 9 | VERY VERY SEVERE (almost maximal) |
| 10 | MAXIMAL |

**Fig. 7.1** The modified Borg scale (Wilson and Jones 1989, p. 278; Awaiting permissions to reproduce)

effort. These themes were chosen because they were identified by individuals with dyspnoea when the scale was being developed.

The most widely used scale, however, in children and adolescents is the MMRC dyspnoea scale (Fig. 7.3) and was used when assessing Elina's breathlessness. It is an activity tool which grades the severity of dyspnoea in

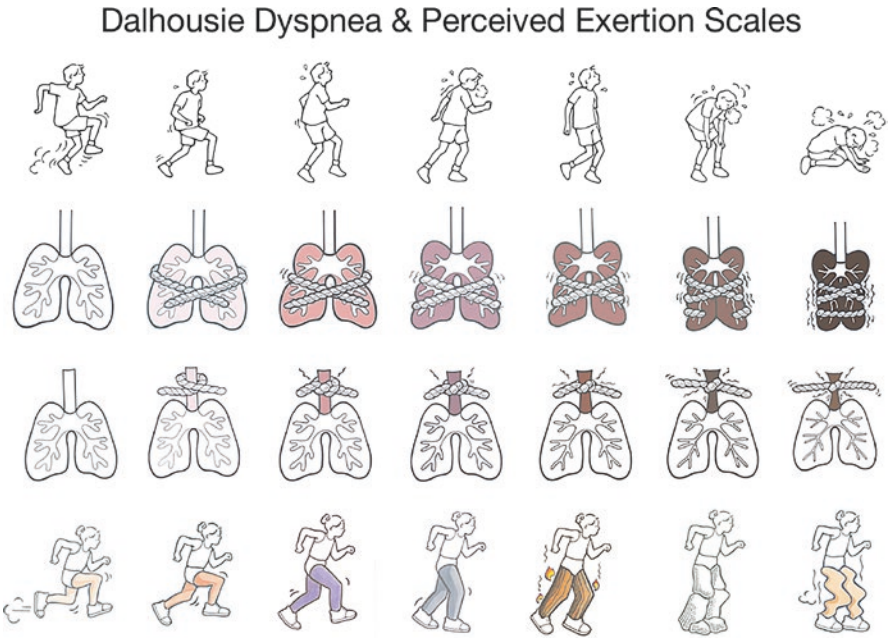


**Fig. 7.2** The Dalhousie scale (Pianosi et al. 2014, p. 35; Reprinted from Respir Physiol Neurobiol, 199, Pianosi PT, Huebner M, Zhang Z, McGrath PJ, Dalhousie Dyspnea and Perceived Exertion Scales: Psychophysical properties in children and adolescents, 34–40, 2014, with permission from Elsevier)

| Grade | **Description of Breathlessness** |
|---|---|
| 0 | I only get breathless with strenuous exercise. |
| 1 | I get short of breath when hurrying on level ground or walking up a slight hill. |
| 2 | On level ground, I walk slower than people of the same age because of breathlessness, or have to stop for breath when walking at my own pace. |
| 3 | I stop for breath after walking about 100 yards or after a few minutes on level ground. |
| 4 | I am too breathless to leave the house or I am breathless when dressing. |

**Fig. 7.3** The Modified Medical Research Council breathlessness scale (Stenton 2008, p. 226; Stenton C, The MRC breathlessness scale, Occupational Medicine, 2008, 226–227 by permission of Oxford University Press)

respiratory diseases. Other important parts of a child's history related to their dyspnoea include: the time period, quality or descriptor that they associate with their breathing discomfort, precipitating and alleviating factors, history of asthma, association of cough, fatigue or palpitations (Lands 2017). Clinical signs also include the level of airway obstruction, the presence of rales or rhonchi, results of spirometry and levels of oxygen saturation. However, it is important to remember that these do not always correlate to the level of dyspnoea experienced.

## 7.2 Question 2. Elina Is Experiencing Both Fatigue and Breathlessness. Is Her Fatigue Caused by Breathlessness?

Fatigue and breathlessness are almost always co-stressors and have to be managed accordingly. Paulus (2013) studied the contribution of distress and anxiety to the development of breathlessness and Chin and Booth (2016) explored how episodes of breathlessness can trigger anxiety. They showed that '*strategies to break the cognitive-behavioural cycle of breathlessness and anxiety, form part of complex non-pharmacological interventions for the symptom'* (p. 396). Therefore many strategies that do help to reduce dyspnoea in patients with malignant and nonmalignant disease incorporate anxiety management.

**Case Study**

*Elina was initially diagnosed with a metastatic Primitive neuro-ectodermal tumour (PNET) of the scalp, with mediastinal metastasis. At diagnosis she had symptoms of pain, shortness of breath, mild bleeding and a foul smelling discharge from the fungating wound. She was extremely anxious about what was happening to her.*

## 7.3 Question 3. What Is the Differential Diagnosis of Her Current Dyspnoea Knowing This Information?

Dyspnoea refers to a subjective sensation that breathing has become unpleasant, rather than an objective observation that it is become fast or difficult. An understanding of this is required when trying to differentiate between physical and psychological causes. The management of dyspnoea requires a good understanding of the differential diagnosis of this symptom. This allows the doctor to take an accurate history with specific questions, as well as an understanding of what is required in terms of examination and subsequent investigations (Collins and Fitzgerald 2006).

In Elina's case we needed to consider the following (Cachia and Ahmedzai 2008):

a. Anaemia.
   - This may be associated with the malignancy and metastasis themselves or linked with secondary factors such as a poor diet or dysfunctional uterine bleeding.
   - The cancer may produce a normocytic normochromic chronic anaemia of malignancy or iron deficiency anaemia secondary to chronic bleeding.
   - It is important to understand that in many resource limited settings nutrition is a major consideration in symptoms such as this one.
   - Although we are a paediatric specialty we must appreciate that we are dealing with a young woman here and as such may have to manage gynaecological issues.

b. Anxiety.
   - As dyspnoea is a subjective sensation it has a very strong association with fear and therefore in a young adult feels very frightening.
   - The anxiety may therefore be directly associated with the sensation of not being able to breathe.
   - We should also consider that at the age of 17 years she may also understand that this represents recurrence of her cancer and therefore she may be fearful of the effects of the cancer as well as any treatment she may require.

c. Ascites.
   - We already know that her tumour has metastasised to her lungs. It would therefore be reasonable to expect it to have spread to other parts of the body including her abdomen.
   - The development of ascites is a common problem with certain types of tumours.
   - The amount of fluid buildup can be large.
   - The pressure effect of the ascites can cause direct mechanical problems to the diaphragm to limit lung expansion.
   - This could then lead to her development of dyspnoea.

d. Cerebral tumours.
   - Cerebral metastasis can have a direct effect on respiratory drive and therefore directly cause dyspnoea.

e. Hepatic involvement.
   - Liver metastasis can develop into very large masses.
   - This would then have a direct pressure effect on the diaphragm or in severe cases push into the right lung cavity which can therefore have a direct effect on the lungs being able to be ventilated by the muscles as well as a reduction in lung space and capacity.

f. Infection.
   - Any form of respiratory infection can present as breathlessness.
   - It is important to be aware that with oncology patients atypical infections may also occur.
   - Always consider atypical bacterial, viral and fungal infections.

g. Metabolic.
   - There is a long list of metabolic conditions that may directly or indirectly affect respiration, including diabetes, pituitary dysfunction, pseudohypoaldosteronism, hypothyroidism, hyperthyroidism, hypoparathyroidism and lysosomal storage disorders (Milla and Zirbes 2012).
   - A full series of biochemical investigations are required to rule out these conditions.

h. Pain.
   - Pain associated with direct lung involvement, whether in the lung tissue itself, pleura or musculoskeletal will all have an effect on respiration leading on to dyspnoea.
   - In addition pain associated with metastasis elsewhere in the body will have an indirect effect on the patient's respiration and can also lead on to a sensation of dyspnoea.

i. Lung pathology.
   - We already know that Elina has had mediastinal metastasis and therefore she may well have a recurrence of tumour in her lungs.
   - The tumour may well cause the development of a pleural effusion or even a pneumothorax.
   - It is also important to recognise that the heart and lung are very closely related both physically in the body but also in how they work together.
   - Metastasis in the lung can lead on to heart strain and subsequent cardiac failure. Left ventricular failure particularly can lead on to distressing dyspnoea.
   - A combination of factors from the cancer itself and general debility can lead to the development of pulmonary embolism.

j. Respiratory muscle dysfunction.
   - Although we have touched upon other conditions that may affect the respiratory muscles, we must also consider that poor diet, nutritional deficiencies and cachexia can contribute towards weakening of her lung muscles.

**Case Study**

*Elina also expressed issues of fear of breathlessness, altered body image and the possibility of losing her hair with prospective chemotherapy. This caused untold anxiety to her parents, her primary caregivers and other members of her family.*

## 7.4 Question 4. How Do Interfamily Relationships Affect Psychological Issues and Therefore Impact Upon Dyspnoea?

There is evidence in the literature that family dynamics have a large impact on child and family coping when faced with a life-limiting condition (LLC). Kissane et al. (2003) found that 71% of families with a child or adolescent with a LLC

were at some risk of morbid outcomes. Significantly greater levels of psychosocial morbidity were found in families where group functioning was poorer, suggesting the need to assess families as a whole rather than individuals, in order to predict those at high risk. Thus they developed the 'family focused grief therapy' protocol. Results showed significant improvements in distress and depression and many studies have aimed at refining the original protocol. Mondia et al. (2012) looked at its impact on Asian American value systems. They found that: family closeness; respect for hierarchy; gender-determined roles; intergenerational tensions; preoccupation with shame and limited emotional expressiveness impacted on how they coped. Therefore it is important to pay thoughtful attention to ethnic issues when dealing with culturally diverse groups of families and working in different countries or regions of the world.

When children with LLCs are hospitalised, family resilience and the need to stay strong for their child are indicators of good coping. The role of siblings as protectors, helpers and companions is beneficial, thus the importance of trying to ensure that siblings are included in care provision whenever possible (Nabors et al. 2017). Uncertainty with prognosis can lead to stress in parents, which in turn is passed on to the sick child; however, psychotherapy interventions can provide a protective or buffering function over time, thereby promoting resilience (Mullins et al. 2015). Previous experiences, along with cultural expectations, are important and Woodson et al. (2015) raised the question of extended family support, longitudinal hardiness and the need to study various ethnic groups as future research topics within this topic of coping.

**Case Study**

*Elina was placed on a standard protocol which entailed induction chemotherapy for 12 weeks, followed by local treatment in the form of a wide excision of the scalp lesion and reconstruction along with adjuvant external beam radiation to the scalp. This was followed by maintenance chemotherapy.*

## 7.5 Question 5. Which Drugs Used in Oncology and Palliative Care Can Cause Issues with Breathlessness?

Many drugs which are routinely used as anti-cancer treatment in children and adolescents have the capacity for causing breathlessness. These include:

a. Bleomycin which is commonly used in the management of Hodgkin's lymphoma and germ cell tumours. Toxicity has been reported, that usually starts after 6 months, with bleomycin interstitial pneumonitis (BIP) which can progress to pulmonary fibrosis in a small percentage of individuals (Uzel et al. 2005). Thus, most oncologists try to avoid using bleomycin if possible.

b. Radiation can cause lung fibrosis (Abid et al. 2001).
c. Monoclonal antibodies such as dasatinib and bortezomib can cause pulmonary toxicity.
d. Bronchiolitis obliterans syndrome (BOS) and idiopathic pneumonia syndrome (IPS) are a significant source of morbidity and mortality in hematopoietic cell transplantation (HCT) (Dudek et al. 2003).

**Case Study**
*While still on therapy, Elina presented with new lesions in her thorax. Ultrasonography of the thorax revealed gross right-sided pleural effusion with collapse of underlying lung, with multiple pleural-based masses on the right side, along with a grossly enlarged liver full of metastases. Further tests also showed pulmonary embolism with super added infection. She had symptoms of moderate breathlessness even at rest.*

## 7.6 Question 6. How Can One Manage Elina's Breathlessness and What Treatment Should You Give Her?

It is important to consider the various causes of Elina's breathlessness. The pleural effusion can be drained and this may also help the collapsed lung to re-inflate. Unfortunately malignant pleural effusions have a tendency to re-accumulate relatively quickly (Hoffer et al. 2007). The multiple pleural masses and liver full of metastases may be treatable through chemotherapy or radiotherapy dependent on the type of cancer.

The primary embolism would normally require anti-coagulation. However, the damage to her liver may well cause portal hypertension with oesophageal varices. There is therefore the risk of severe haemorrhage in this case. The use of anti-coagulation would increase the risk of catastrophic haemorrhage and Elina bleeding to death. She was therefore not anti-coagulated.

The ethical issues around treating a chest infection are complex. On the one hand, she has a terminal cancer which is likely to cause significant distress in its latter stages through dyspnoea, pain and haemorrhage. On the other hand, at this stage, she is well enough to have a limited quality of life. With the correct management of her case we should be able to control some of these distressing symptoms. Sometimes the decision to use antibiotics is governed by the desire of the child and family to stay at home and avoid intravenous antibiotics. In these cases it can be helpful to do a trial of oral antibiotics at home. It was decided to treat Elina's infection with antibiotics.

**Case Study**
*Elina experienced frequent panic attacks, which restarted the anxiety in her, adding to her breathlessness.*

## 7.7 Question 7. How Can We Manage Her Anxiety and Panic Attacks to Help Her Breathlessness

There is a close relationship between dyspnoea and anxiety (Craig et al. 2015). Any child or adolescent who cannot breathe will become more anxious. Anxiety will cause hyperventilation which in turn will exacerbate the sensation of dyspnoea. Management would require both pharmacological and non-pharmacological measures to be effective. Studies in adults with cancer and chronic obstructive pulmonary disease (COPD) have not shown any added benefit of midazolam (buccal or nasal spray) over placebos in the management, and thus should be considered on an individual basis. These anxiolytics can be used safely in children and intranasal midazolam or oral haloperidol is often used. Along with the medication, calming both Elina and her parents will help (Jassal 2017). A propped up sitting position, with nasal prongs for oxygen, along with individual counselling, will relax the situation and promote some control over the symptoms. Breathing and relaxation techniques will also help, as will a quiet room, with familiar people (although in many places this is may not be possible). Diversion therapy, yoga, meditation and reiki are also measures used very frequently to help individuals like Elina to control their anxiety and panic attacks due to their dyspnoea.

**Case Study**

*In view of her panic attacks and dyspnoea, an intercostal drain (ICD) was inserted for the gross right-sided pleural effusion and Elina reported improvement in her symptoms and she was started on incentive spirometry.*

## 7.8 Question 8. How Can Malignant Pleural Effusions Be Managed?

Malignant pleural effusions occur due to the infiltration of cancer cells into pleural tissue which causes a reactionary effusion. This may result in positive fluid cytology and/or on a pleural biopsy. If left untreated, the effusion will grow causing collapse of the underlying lung. Both will contribute to the symptom of dyspnoea (Herrera et al. 2017).

Elina should initially undergo therapeutic thoracentesis to drain the fluid, the needle is inserted carefully into the pleural cavity, preferably under the guidance of ultrasound. Ideally the aspiration should not exceed 1.5 L, as there is a potential low risk of the development of re-expansion pulmonary oedema. Potential complications of thoracentesis include pain, pneumothorax, shortness of breath, cough and vasovagal reaction. The recommended location for the needle insertion is the mid-axillary line in the sixth, seventh or eighth intercostal space. It is critical that Elina holds her breath to avoid piercing of the lung. Chest radiography is performed immediately following the procedure to rule out pneumothorax. If feasible, large volume thoracentesis with manometry is advised to ascertain the response of Elina's symptoms to pleural fluid removal and to assess whether or not the lung is expandable.

Repeated drainage is decided by time to re-accumulation, the individual's prognosis and severity of their symptoms. Where the malignant plural effusion re-accumulates slowly (e.g., more than 1 month), the child may be managed with repeat therapeutic thoracentesis, especially if life expectancy is short (<2–3 months) and there is poor performance status (Feller-Kopman et al. 2018; Kalifatidis et al. 2015). Where it recurs rapidly (e.g., less than 1 month), options include inserting an indwelling pleural catheter drainage, pleurodesis by various techniques, for patients with an expected survival of more than 6 months. Another option is a pleuroperitoneal shunt. Where there are loculations, the instillation of an intrapleural fibrinolytic agent (tissue-plasminogen activator, urokinase or streptokinase) may be of benefit (Mehta et al. 2015; Shaw and Agarwal 2013). Rarely systemic antitumour therapy and/or radiation may be considered when estimated time to survival is relatively long. Ideally patient preferences should guide the subsequent management choices.

**Case Study**

*The drain was removed after 2 weeks, as there was minimal drainage and Elina remained asymptomatic besides mild pain at the site of the drain. She remained asymptomatic and was restarted on oral chemotherapy/palliative radiation. She later presented with increasing dyspnoea, MMRC Grade 4, excessive fatigue, heart failure and bilateral lower limb oedema.*

## 7.9 Question 9. What Might Have Caused Her Symptoms and How Could They Be Managed?

Elina's excessive fatigue may well exacerbate her dyspnoea (Ullrich and Mayer 2007). The fatigue may be secondary to cachexia, anaemia or biochemical imbalance. Cachexia itself can be very difficult to treat. Anaemia and biochemical imbalances can be corrected with appropriate therapies. There is once again an ethical dilemma in the benefit against burden of blood transfusion in her case.

Her heart failure could certainly cause her significant dyspnoea. The pulmonary embolism can cause significant heart strain. This can then lead to left ventricular failure. This could be managed with the use of loop diuretics as well as the addition of ACE inhibitors.

The numerous lung tumours can cause compression on key blood vessels which can also lead to heart strain. Vena caval obstruction can cause very distressing symptoms with associated oedma. The use of palliative radiotherapy in these situations can be particularly helpful.

The bilateral lower limb oedema may be due to right ventricular failure, low-protein/albumin or orthostatic oedema secondary to her generalised weakness and inability to move around. The right ventricular failure can be managed in the same way as all heart failures. Her nutrition could be improved with the use of supplementary feeds to help raise her protein and albumin levels. The orthostatic oedema can be managed with elevation, massage and gentle movement of her lower limbs through walking or with the help of physiotherapy.

**Case Study**
*The ultrasound of Elina's thorax revealed a mild right-sided pleural effusion with extensive lung and pleural metastasis. Patient's Respiratory Distress Observatory Score (RDOS score) at this time was 14.*

## 7.10 Question 10. What Is the Differential Diagnosis of Her Current Dyspnoea Knowing This Information?

Some of the earlier causes of her symptoms could also be causing the dyspnoea on this occasion with anaemia and anxiety being the main differential diagnosis. She has pleural-based metastasis which has caused a mild pleural effusion which would add to the feeling of breathlessness, although would not be the primary cause. The metastasis in the lung parenchyma is taking away part of the lung tissue which is needed for oxygen exchange and in an already compromised situation, the dyspnoea gets triggered. Heart failure will cause left ventricular failure leading to accumulation of fluid in the lungs and increasing dyspnoea. Finally her bilateral lower limb oedema could be caused by hypoproteinemia or right ventricular failure, both of which will contribute to dyspnoea.

## 7.11 Question 11. Can Any of the Causes of Elina's Dyspnoea Be Reversed and If So, How?

Anaemia can be treated with judicious use of blood or packed cell transfusions. This should be done only when the doctor decides that the symptom will be relieved; rather than only on the basis of blood counts. Anxiety in an adolescent such as Elina needs specialised counselling techniques. Counsellors and psychologists use a range of diversional techniques, which may need to be combined with a minimum dose of anti-anxiolytic agents, for optimum benefit. At this stage of the disease her lung metastasis, heart failure and lower limb oedema are not reversible. Attempting to correct these imbalances would cause more toxicity than benefit. A good balance between both has to be maintained in all decision-making.

**Case Study**
*Elina was started on morphine 2.5 mg 4 hr, dexamethasone 4 mg 8 hr and lorazepam 1 mg at night along with appropriate laxatives. Her RDOS improved to around ten with these measures. The importance of sitting in a propped up position and pacing any activities were explained. Morphine was escalated as per requirement to 10 mg 4 hr. Her subjective experience of breathlessness was better. She died peacefully a week later in the hospital. Her family was with her and no resuscitation was attempted.*

## 7.12 Question 12. How Do We Manage Terminal Dyspnoea?

The management of terminal dyspnoea when all reversible causes have been addressed requires an empirical approach. The first issue is around the overwhelming fear and anxiety linked with shortness of breath and often the feeling of suffocation or drowning. The use of low-dose benzodiazepines can be most helpful (Duval and Wood 2002). Sublingual lorazepam or buccal midazolam can be very effective due to their speed of onset. Lorazepam has the benefit of being a tablet that dissolves quickly in the mouth and so can be easily carried by the patient to use when required. Low-dose buccal midazolam given at a quarter strength of that required for seizure management can be given through preloaded syringes or drawn up from ampoules. In the terminal stages the midazolam can be given through a syringe driver as a continual infusion, with the dose gently increased to control dyspnoea and allow a level of sedation.

Opioids used at half the analgesic dose can be very effective in helping with dyspnoea (Viola et al. 2008). They appear to work by reducing anxiety and pain. They settle down the respiratory centres and reduce pulmonary artery pressure. The type of opioid that one can use is dependent on the availability within different parts of the world. We would use oral morphine or subcutaneous morphine/diamorphine. Combining the opioid with midazolam through a subcutaneous syringe driver is a very effective combination. Sometimes the use of nebulised salbutamol or ipratropium can also be helpful. The use of hyoscine hydrobromide or glycopyrronium can be helpful in managing death rattle (Lawrey 2005) although the evidence for this is poor (Wee and Hillier 2008).

The role of oxygen in dyspnoea can be controversial (Currow et al. 2009; Quinn-Lee et al. 2012). There is a school of thought that says that oxygen should only be used in hypoxia when the oxygen saturation levels are low. However, clinical experiences suggest that many children function very well with low oxygen saturation figures so it can be an unreliable measure. Many studies have shown that the use of oxygen is often of little benefit and that opening a window or using a fan to push air across the child's face is equally effective. In reality both parents and nurses will initiate the use of oxygen at the onset of dyspnoea within the hospital or hospice environment. From a practical point of view it is best to avoid using gadgetry and look at the child instead. If the child is given oxygen and appears to benefit from that, then leave the oxygen on. If there is no obvious benefit, then stop the oxygen (Halpern and Hansen-Flaschen 2006). Always remember that when giving oxygen the use of a face mask is a barrier between the family and the child's face. If a child is going to die, it may be better for the family to remember them without a plastic mask in the way. Children tend to die through respiratory rather than cardiac arrests. It is therefore helpful to pre-warn the family about apnoea episodes and Cheyne Stokes breathing patterns.

## 7.13 Question 13. What Procedures Were in Place for End-of-Life Care for Elina and to Provide as Peaceful and Dignified Death as Possible?

When an individual centre has an end-of-life (EoL) care policy it helps when managing a child who has approached the EoL. The care can be simplified more when an advance directive is in place. Ideally the family has a chance to choose the most appropriate place for EoL care, often this is home, hospice and hospital in that order. Taken for granted is the quality of care available in all these situations. In India, home has been cited as the preferred place and all teams work synergistically to make this happen. Community doctors are primarily involved in the care and provide the death certificate. Sadly India does not have a legalised EoL care policy; this makes a peaceful and dignified death in a hospital, not always a reality. The National Accreditation Board (2015) for hospitals has made an EoL hospital plan a necessity while applying for accreditation. This was made possible through extensive work undertaken by the Indian Association of PC. Many children die in the hospital as the preferred place of care, as families are familiar with it, thus the family need to be provided with adequate privacy on the wards and the ability for key carers to be present till the end. There should also be a 'Do Not Resuscitate' (DNR) order in place which is respected by all hospital personnel. This has become possible in many centres, where palliative care units are in place. Bereavement care and grief counselling is provided by the same teams till the family can adjust and move on. Recently in 2018, the Supreme Court of India issued a directive allowing advance directives for patients suffering with life-limiting conditions in India. In high-income countries, adequate policies for EoL care are often in place, e.g., in the United States of America (USA), the United Kingdom (UK) and Australia.

## 7.14 Conclusion

Elina went through phases of dyspnoea management from diagnosis to terminal care. In each phase, the management was different, multi-professional and aimed at best possible quality of life (QoL), an aim which was managed well.

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# Gastrointestinal Symptoms

8

Mercedes Bernadá, Julia Ambler, and Leticia Fuentes

**Key Learning Points**

1. Gastrointestinal symptoms are a common source of suffering in children receiving palliative care (PC).
2. There are a variety of gastrointestinal symptoms experienced in children receiving PC including, but not limited to: nausea and vomiting, diarrhoea, constipation and mucositis.
3. Using a structured approach including comprehensive evaluation, treating the underlying cause (if possible), the use of non-pharmacological as well as pharmacological therapies will help in the management of gastrointestinal symptoms.
4. Non-pharmacological therapies should be used alongside pharmacological therapies, and have an important role to play.
5. There is a need for close clinical monitoring of symptoms and the side effects of medication to enable good symptom control.

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**Case Study**

*Martin was a healthy 10-year-old boy who loved playing football until the beginning of his illness. Martin is currently in grade 5 and loves school. He lives with his mother and his sister.*

*In January 2019, after a football match Martin presented with pain and swelling of the left lower leg. A diagnosis of non-metastatic proximal tibia osteosarcoma was made. A weekly regimen of chemotherapy was commenced (cisplatin and doxorubicin). During the chemotherapy he developed nausea and frequent vomiting.*

## 8.1 Question 1. What Is the Most Likely Cause of Martin's Nausea and Vomiting?

Even though, in children, there are many causes of nausea and vomiting the most likely cause, in this case, is the side effect of the chemotherapy.

## 8.2 Question 2. How Can We Manage Martin's Nausea and Vomiting?

### 8.2.1 Comprehensive Evaluation

Nausea and vomiting may be triggered by different stimuli acting in different areas (brain's vomiting centre, vestibular apparatus, vagal afferent gut receptors, etc.), thus a complete history and physical examination is needed. It is important to consider that anxiety, fear and pain may also cause or increase nausea and vomiting. The complete history should include:

- Timing, frequency, consistency and volume of the vomiting.
- Is there associated constipation or diarrhoea?
- Does anything make it worse or better?
- Does the child have any contributing disorder: swallowing problems, gastro-oesophageal reflux or poor gut motility?
- Is the child in pain?
- Is the child anxious?
- Is nausea and vomiting related to positioning or movement?

### 8.2.2 Treat the Underlying Cause (if Possible)

Ideally nausea and vomiting caused by antineoplastic drugs should be prevented (Dupuis et al. 2013). Although, sometimes it may be necessary to transiently interrupt or even withdraw chemotherapy to allow recovery.

### 8.2.3 Non-pharmacological (Integrative) Therapies

- As nausea and vomiting can cause and result in anxiety, a good explanation to the child and family about the source and management is very important.
- The child should be offered small meals frequently, avoiding strong smells or foods that may worsen the nausea and vomiting: those that are very fatty, spicy or sweetened.
- Ensure adequate hydration by giving small amounts of fluids and rehydration solution.
- Feed the child in an upright position if reflux is a concern.
- Encourage the child to gently rinse out the mouth and brush teeth after vomiting.
- If vomiting is associated with a certain medication, split the dosages where possible.
- Address psychological issues such as anxiety or fear.
- Massage (Mazlum et al. 2013), acupressure (Garcia et al. 2013) or hypnosis (Richardson et al. 2007) can be used to reduce nausea.

### 8.2.4 Pharmacological Therapies

A combination of medications, according to the presumed aetiological mechanism involved, may be required to achieve the best symptom control (Dupuis et al. 2013) (Table 8.1).

**Case Study**
*Martin was given dexamethasone and ondansetron which helped, and his nausea and vomiting improved. At the end of the 4th cycle of chemotherapy, he developed diarrhoea, diffuse abdominal pain, refusal to eat due to oral pain and fever. On assessment he was alert, well hydrated with pale mucous membranes, slimmed down with globally diminished muscular mass. He was also found to have tooth cavities and oral ulceration. His abdomen was soft without visceromegaly.*

**Table 8.1** Medication useful for nausea and vomiting

| Condition | First-line medication |
|---|---|
| Drug-induced (e.g. morphine) | Ondansetron, haloperidol |
| Delayed gastric emptying | Domperidone |
| Chemotherapy or radiotherapy | Ondansetron/granisetron<br>Dexamethasone, aprepitant |
| Bowel obstruction | Cyclizine, hyoscine butylbromide, dexamethasone |
| Raised intracranial pressure | Cyclizine, dexamethasone |
| Metabolic causes | Haloperidol |

## 8.3 Question 3. What Are the Possible Causes of Martin's Diarrhoea?

While diarrhoea is a known side effect of the chemotherapy he is receiving, the most likely diagnosis of Martin's diarrhoea is gastrointestinal mucositis or neutropenic enterocolitis (typhlitis). Gastrointestinal mucositis causes diarrhoea as a consequence of enteral inflammatory mucosal damage and malabsorption. Typhlitis is a life-threatening necrotising enterocolitis occurring in immunosuppressed patients causing diarrhoea, pain and fever. It is a medical emergency (Rodrigues et al. 2017). Other possible causes of diarrhoea include:

- Infections: bacteria, viruses and parasites.
- Food intolerance: a pre-existing food intolerance or temporary intolerance caused by chemotherapy.
- Drug side effects: antibiotics.

## 8.4 Question 4. How Should We Manage Martin's Diarrhoea?

### 8.4.1 Comprehensive Evaluation

- Are there signs of acute infection? (mucous/blood in the stool, fever, abdominal pain)
- What is the hydration status of the child?
- What is the nutrition status of the child?
- How long has the diarrhoea persisted?
- Is the diarrhoea made worse by specific drugs or foods?

### 8.4.2 Treat the Underlying Cause (If Possible)

Given that diarrhoea is frequently a consequence of chemotherapy, in some cases treatment needs to be transiently withdrawn. In addition, when infection is suspected, intravenous antibiotics must be given.

### 8.4.3 Non-pharmacological (Integrative) Therapies

- The child and family should be counselled about the condition, risks of cross-infection and correct handwashing.
- Feeding should be continued during acute diarrhoea.
- If the child has neutropenic enterocolitis, his gut should be rested using total parenteral nutrition if available (Rodrigues et al. 2017).

- Food and medicines that might be aggravating diarrhoea should be stopped when feasible.
- Ensure regular nappy changes (where wearing nappies) and barrier creams to protect the skin.

### 8.4.4 Pharmacological Therapies

- Hydration is the mainstay of acute diarrhoea treatment.
- Zinc supplementation for 14 days is advised (WHO 1979).
- Acute diarrhoea does not usually require anti-diarrhoeal drugs, although there is some evidence to suggest that diosmectite, racecadotril and probiotics may help (Faure 2013).
- When dysentery is suspected (blood/mucous in the stool), an antibiotic should be used.
- If the child has stomach cramps, hyoscine butylbromide may help.
- For chronic persistent diarrhoea, loperamide, bismuth subsalicylate, colestyramine and octreotide could be used with caution, in special circumstances (Friedrichsdorf et al. 2011).
- The constipating side effect of morphine can be beneficial in this patient group.

## 8.5 Question 5. What Are the Possible Causes of Martin's Oral Pain?

The appearance of oral ulceration in a child undergoing their 4th cycle of chemotherapy leads to the diagnosis of oral mucositis, which is one of the most common symptomatic complications of antineoplastic therapy. It is a hugely painful condition, associated with a significant increase in morbidity, pain, functional limitation and deterioration in quality of life (Molina and Estupiñán 2010). Chemotherapy causes inflammation and ulceration through tissue damage along the entire digestive tract, frequently resulting, as in Martin's case, in oral mucositis and diarrhoea. Most common oral lesions include: mucositis, xerostomia, tooth decay, dysgeusia, trismus, mucosal ulcerations, sores, gingival bleeding, periodontitis, viral, bacterial or fungal infections and necrosis (Molina and Estupiñán 2010; Carreón et al. 2018; Fekrazad and Chiniforush 2014).

Many of these lesions may be reversible, while others produce permanent complications. The earliest signs and symptoms of oral mucositis appear in the oral mucosa after the 5th to 10th day of chemotherapy or radiotherapy and include: erythema, oedema and swelling of the gums, burning sensations and an increase in sensitivity to hot or spicy food. The erythematous areas progress to white scaly raised patches and subsequently to painful ulcers, which can be secondarily infected (Fekrazad and Chiniforush 2014).

**Table 8.2** WHO grades of oral mucositis

| Grade | Description |
|---|---|
| 0 | None |
| 1 | Oral soreness, erythema |
| 2 | Oral erythema, ulcers, solid diet tolerated |
| 3 | Oral ulcers, liquid diet only |
| 4 | Oral alimentation impossible |

## 8.6 Question 6. How Severe Is Martin's Mucositis?

The most commonly used scale to measure mucositis is the World Health Organization (WHO) scale, which classifies the severity of lesions into 4 grades (WHO 1979; Sonis et al. 2004) (Table 8.2).

**Case Study**
*Martín had grade 4 lesions with very intense pain and discomfort preventing him from swallowing solids and liquids. It had affected his quality of life and ultimately leads to malnutrition.*

## 8.7 Question 7. How Can We Manage Martin's Oral Pain?

### 8.7.1 Comprehensive Evaluation

It is very important to carry out a complete assessment and identify: difficulty in swallowing, food refusing or mouth pain and a complete physical examination that includes the entire mouth to identify possible factors that require specific treatment: oral candidiasis and tooth caries (Fekrazad and Chiniforush 2014).

### 8.7.2 Treat Underlying Causes (If Possible)

Martin has previously had tooth cavities. It is important to identify and treat this, and other oral co-morbidities, such as oral candidiasis, xerostomia and bleeding gums, because mucositis may get worse or develop due to poor oral health. The participation of a dentist/stomatologist to help maintain excellent oral health is important to prevent oral mucositis or to reduce its severity and symptoms should it occur (Carreón et al. 2018).

### 8.7.3 Non-pharmacological (Integrative) Therapies

The management of oral mucositis includes: prevention, pain control and nutritional support (Molina and Estupiñán 2010). Regardless of the grade, it can be controlled or cured either with treatment or by dose reduction, interruption and/or unscheduled

withdrawal of antineoplastic treatment, as new cell proliferation, immunological recovery, and adequate control of the bacterial flora improve the condition (Carreón et al. 2018).

- *Good oral hygiene:* it is recommended to brush gently with a soft toothbrush, dental floss and non-medicinal mouth rinses with saline solution or bicarbonate water. Mouthwashes that contain alcohol should be avoided (Molina and Estupiñán 2010).
- *Good hydration:* keep the mouth moist by drinking plenty of fluids. Try soft foods that are easy to swallow and drink through a straw. Use lip balm to keep the child's lips from peeling or cracking. Avoid food with extreme temperatures, both hot and cold.
- *Cryotherapy:* may be offered to cooperative children receiving chemotherapy or regimens associated with a high rate of mucositis as prevention. Regimens appropriate for cryotherapy are restricted to agents with a short infusion time and short half-life. It is an attractive intervention because of its low cost and universal access. Oral cryotherapy involves placing ice cubes or ice chips in the mouth and continually replenishing fresh ice during the period of cytotoxic treatment (typically 30–60 min). Flavoured ice popsicles, ice slushy drinks or "freezies" are likely to be more acceptable to children than plain ice. Be careful if ice chips are to be used, as they may be a choking hazard in very young children (Fekrazad and Chiniforush 2014; Sung et al. 2017).
- *Low level laser therapy (LLLT)* is a new modality for managing mucositis via photo biomodulation (PBM). It is a safe, non-pharmacological method which can modulate various metabolic processes. It requires expertise and specialised equipment (Sung et al. 2017). LLLT is administered intraorally and is performed by diode lasers including red and infra-red wavelengths. Shorter wavelengths are the most effective. The main effects are anti-inflammatory, analgesic and it is favourable for wound healing. LLLT effects are: reduction of the severity of mucositis, reduction of the incidence, severity and duration of pain. There is no consensus regarding intensity of wavelength, energy density, time of exposure and ideal time of starting laser therapy (Fekrazad and Chiniforush 2014; Sung et al. 2017; Anschau et al. 2019). Unfortunately, LLLT is not available in many countries.

### 8.7.4 Pharmacological Therapies

Pain management is an important part of severe oral mucositis: local anaesthetics (lidocaine) combined with oral or parenteral analgesics according to severity of pain should be prescribed (Costa et al. 2018).

- Several *"magic" solutions* with compounds like: lidocaine, diphenhydramine, aluminium hydroxide, saline, nystatin, dexamethasone, morphine, sucralfate, glutamine and B complex are used in different countries. Although, many of this strategies

and products have been studied, so far there is no consistent evidence about their usefulness (Molina and Estupiñán 2010; Sung et al. 2017; Costa et al. 2018).

- Palifermin is a keratinocyte growth factor (KGF), available in some countries, for children receiving regimens associated with a high rate of severe mucositis. It significantly reduces severe oral mucositis. KGF should be administered at a dose of 60–90 mg/kg/day for 3 days prior to conditioning and 3 days following stem cell infusion. KGF should be used carefully in individual patients after weighing risks and benefits (Molina and Estupiñán 2010; Sung et al. 2017).

**Case Study**

*Martin was admitted with strict isolation measures, and intravenous cefepime, fluconazole and neupogen were started. Analgesia with Non-steroidal anti-inflammatory drugs (NSAIDs) and regular intravenous morphine every 4 h plus rescue doses were given. He received red blood cell transfusion, and mucositis treatment was initiated with bicarbonate water rinse and laser therapy. In the hospital his diarrhoea stopped and all stool cultures were negative.*

*Over the next few days his oral mucosal lesions worsened, with bleeding and uncontrolled pain that hindered oral feeding and speaking. Analgesia was titrated and parenteral feeding started. He remained admitted for 15 days with partial pain control. After 10 days of parenteral feeding and treatment, the mucosal lesions improved and he started eating again.*

*On day 14, he complained of moderate abdominal pain without stools for 5 days. At examination, on abdominal palpation, a large faecal mass in the colon and rectum was found.*

## 8.8 Question 8. What Could Be the Cause of Martin's Abdominal Pain and Reduced Stools?

In a child with cancer, any pain and appearance of a new tumour must alert you to the presence of metastasis. Nevertheless, in this case, the lack of stools for 5 days, the presence of a large faecal mass in the colon and rectum, in a child receiving opioids, indicate a diagnosis of constipation with faecal impaction. Children receiving PC commonly experience constipation as a result of: use of medicines (e.g. morphine), disease, dietary and mobility factors (Rabia and Turgay 2016).

*Rome III* consensus for healthy children with *functional constipation* defines it when a child at age 4 years or older presents with at least two of the following criteria for 2 months or more: two or fewer defaecations per week, at least one episode of incontinence per week, history of excessive stool retention, painful or hard bowel movements, presence of large faecal mass in the rectum, large-diameter stools that may obstruct the toilet (Tabbers et al. 2014).

Similar definitions for children living with LT and life-limiting conditions (LLCs) do not exist. So, even though *Rome III* definitions may be used as a baseline guide in children receiving PC, a systematic assessment should always be done in order to promptly identify children at risk of constipation (Table 8.3).

**Table 8.3** Risk factors for constipation in children with LTC and LLC

| |
|---|
| • Chronic neurological conditions |
| • Intra-abdominal cancer |
| • Inactivity |
| • Low liquid and/or food intake |
| • Medication: opioids, antiemetics, tricyclic antidepressants, phenothiazines, diuretics, antihistamines, anticholinergics, vincristine |

## 8.9 Question 9. How Could We Manage Martin's Constipation?

### 8.9.1 Comprehensive Evaluation

- Complete clinical history: previous intestinal patterns, food preferences and habits, defaecation routines, privacy and environmental conditions. Identification of the above risk factors.
- Thorough physical examination, particularly an abdominal exam to identify stool mass in colon or rectum, inspection of the perianal region, looking for skin tags, fissures or dermatitis. The digital anorectal examination may give important information (presence and characteristics of stools). Nevertheless, it was not done in Martin because in a child with neutropenia or thrombocytopenia, invasive examination is contraindicated because it can cause bleeding and/or infection.

### 8.9.2 Treat the Underlying Cause (If Possible)

Review the indication and doses of medications contributing to the constipation. Give special attention to privacy and environmental factors, particularly in admitted adolescents.

### 8.9.3 Non-pharmacological (Integrative) Therapies

- Educate the parent and child about the cause of constipation and discuss the treatment options. A common myth is that laxative use leads to dependence or addiction, which may result in parental under-dosing or "*only as needed*" administration.
- Whenever possible, encourage regular bowel routine: scheduled toilet time for 5–10 min after meals.
- If appropriate, increase activity or passive movements, increase intake of fluids and carbohydrates, especially sorbitol (found in juices) and increase the fruit, whole grains and vegetables in the diet.
- Clockwise abdominal massage may help.

### 8.9.4 Pharmacological Therapies

There is no evidence-based pharmacological protocol for the management of constipation in children with LTC and LLC. Most protocols are based on expert opinions for functional constipation and on adult research. The last Cochrane systematic review ("*Laxatives for the management of constipation in people (adults) receiving palliative care*") did not show any difference in effectiveness of three laxatives: senna, docusate and lactulose. None of the studies evaluated the effectiveness of polyethylene glycol which is strongly recommended in children (Candy et al. 2015).

The pharmacological treatment of constipation in children includes disimpaction, maintenance and specific treatment of anal fissures when present (Madani et al. 2016) (Table 8.4). Particular treatment for opioid-induced constipation is described.

### 8.9.5 Disimpaction

The presence of a palpable faecal mass on abdominal examination or hard stool in a dilated rectum on a digital rectal exam justifies disimpaction. This can be done via oral or rectal therapy or a combination of both. Although rectal disimpaction has been shown to be faster, children prefer and tolerate better the oral route, because it is not invasive and it gives them a sense of control. The method of choice should be determined with a discussion among the physician, parent and child.

- *Stool softener laxatives:* They are the first-line therapy for oral faecal disimpaction. *Polyethylene glycol 3350, lactulose or sorbitol* can be used according to availability, cost or child preference and tolerance. All of them are best

**Table 8.4** Medication dosages for treatment of constipation in children (Friedrichsdorf et al. 2011)

| | | |
|---|---|---|
| Lactulose | Infants: 0.8–3.5 ml (not per kg)<br>Children: 10–30 ml (not per kg) | bd–td/PO<br>bd–td/PO |
| Polyethylene glycol 3350 | 8.5–17 g in 120–240 ml fluid | qd/PO |
| Senna | Sennoside:<br>• 6 mo–2 yrs: 3.75 mg (not per kg)<br>• 3–10 yrs: 7.5–15 mg (not per kg)<br>• >10 yrs: 15–30 mg (not per kg) | *qhs*/PO |
| Bisacodyl | 6–11 yrs: 5 mg<br>>12 yrs: 5–15 mg<br>1 suppository daily | qd/PO<br>qd/PO<br>qd/PR |
| Surfactant laxatives | Docusate sodium oral:<br>0.5–1 mg/kg/dose | 1–4 dos/day/PO |
| | Docusate enema:<br>>3 yrs: 200–283 mg | Qd/PR |
| Methylnaltrexone | 0.15 mg/kg/dose | qod/SC |

*PO* oral, *PR* per rectum, *SC* subcutaneous, *qd* once a day, *bd* twice a day, *td* three times a day, *qhs quaque hora somni* (every bed time), *qod* every other day, *mo* months, *yrs* years

given in water/juice/milk in the morning when the child is most likely to finish it at once.
- *Stimulant laxatives*: The most frequently used are oral *senna and bisacodyl*. They should be given intermittently or for a short period if the child has a full rectum with soft faeces or when *stool softener laxatives* are ineffective.
- *Rectal disimpaction:* In cases of severe impaction, a child may need a rectal suppository or enema for resolution. In communicative children and adolescents, it always should be explained and discussed with them. In children with cancer, when neutropenia or thrombocytopenia is present, the risks (infection/bleeding) should be balanced with the potential benefits.
- *Bisacodyl or glycerine suppositories* are effective and safe, the latter especially in infants.
- *Enemas* may be required when *stool softener and stimulant laxatives* have been adequately used, but proven to be ineffective. *Phosphate enema* may be used in children older than 2 years of age, but *surfactant rectal laxatives* are preferred in paediatrics because of the smaller volume required.

### 8.9.6 Maintenance Therapy

When faecal disimpaction has been accomplished, the goal is to prevent recurrences with maintenance therapy. It consists of:

- Continued application of non-pharmacological measures.
- Laxatives: Polyethylene glycol 3350 or lactulose on a scheduled regimen.

### 8.9.7 Anal Fissure Treatment

An anal fissure is a tear in the skin of the anal canal, usually as a result of trauma caused by the expulsion of hard stool. It is a very painful injury that causes suffering and/or bleeding at the time of defaecation, thus children hold the stool in, increasing constipation. With appropriate treatment, most heal in the short term, but can sometimes transform into chronic lesions requiring specific treatments. Initial treatment of anal fissures includes:

- Softening stools.
- Delicate hygiene of the area without rubbing.
- Application of anaesthetic and corticosteroid creams.

### 8.9.8 Opioid-Induced Constipation

Methylnaltrexone is an antagonist of peripheral receptors without central nervous system action. Its efficacy for opioid-induced constipation has been largely demonstrated in adults, and some evidence of its benefit in children is appearing. It may be used when enteral or rectal measures have been unsuccessful, or when clinical deterioration prevents enteral medication administration (Flerlage and Baker 2015).

**Case Study**

*Martin's constipation responded very well to PEG 3350. Throughout his antineoplastic therapy he continued to have gastrointestinal problems that were well managed by his team.*

## 8.10 Conclusion

Gastrointestinal symptoms are a common source of suffering in children receiving PC. Using a structured approach which includes: comprehensive evaluation, treating the underlying source, offering non-pharmacological-integrative measures and pharmacological treatment, contributes to managing them. Close clinical monitoring of symptom control and medication's side effects should be taken in order to provide optimum symptom control.

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# Fatigue

9

Christina Vadeboncoeur and Chi-Kong Li

**Key Learning Points**

1. Fatigue is a subjective feeling which may be measured by a fatigue severity scale.
2. Fatigue is common in children with cancer and non-cancer conditions, in particular during the advanced phase.
3. Causes of fatigue should be explored and managed accordingly, such as transfusion for anaemia, respecting the overall goals of care.
4. Pharmacological agents are not proven to be of value in relieving fatigue, and non-pharmacological methods may be tried.

**Case Study**

*Hugh is a 5-year-old boy diagnosed with Duchenne Muscular Dystrophy (DMD) at age 3 in Canada. He is enrolled in a trial for a steroid medication to try to preserve his ability to walk. He is currently able to walk but cannot keep up with his classmates during gym class.*

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## 9.1 Question 1. What Is Fatigue?

The European Association for Palliative Care (EAPC) defines fatigue as '*a subjective feeling of tiredness, weakness or lack of energy*' (Radbruch et al. 2008, p. 13). Fatigue is the most common symptom reported in children with advanced cancer.

## 9.2 Question 2. What Is the Impact of Fatigue on a Child, and How Can It Be Measured?

DMD is a neuromuscular disorder affecting approximately 1 in 3500 newborn boys (NORD 2016). It is characterised by progressive muscle loss resulting in profound physical disability. Most boys die before the age of 30 years due to respiratory or cardiac failure. Recent reports identify increasing fatigue in children with neuromuscular disorders, such as DMD as their disease progresses.

The standard of care for DMD involves corticosteroids to prolong ambulation and ventilation to extend life expectancy (Eagle et al. 2002). Paediatric health related quality of life (QoL) research utilises child and parent proxy reporting together to provide the most comprehensive view of the child's health-related QoL. In a Canadian study (Wei et al. 2016), greater fatigue and use of wheelchairs were consistently associated with worse health-related QoL, independent of other factors. Fatigue was associated with every measure of health-related QoL and explained more of the variability in health-related QoL than any other variable.

There are no discrete tools for the measurement of fatigue in children. There is a fatigue severity scale validated in adults with a variety of conditions (Krupp et al. 1989). It asks individuals to use a 7-point scale to report their experience of fatigue in the past week. The Memorial Symptom Assessment Scale (MSAS) (Collins et al. 2002) used in children with cancer, includes one item 'Lack of Energy' measured on a 5-point Likert like scale which is based on experience of the symptom over the last week. Crichton et al. (2015) have reviewed scales measuring fatigue in children younger than 18 years with specific health conditions. No scale was found that was generalisable, and there was little evidence for assessment in younger children.

**Case Study**

*Hugh developed daytime fatigue at age 12. He was having difficulty attending school full time and needed to nap when he returned home from school in the afternoon. A sleep study was arranged, as well as a number of other investigations, and he was started on night time non-invasive ventilator support. This allowed him to attend school for full days and keep up with his peers academically. He was no longer able to participate in gym class.*

## 9.3 Question 3. What Is the Prevalence of Fatigue in Children with Serious Illness?

Fatigue has most commonly been measured in children with cancer (Ullrich and Mayer 2007). Reports of the number of children affected range from 46% (Wolfe et al. 2015) in children living with advanced cancer to 86% (Jamsell et al. 2006) in children in their last month of life. Most studies done in high income countries report lethargy as the most common symptom in children with advanced cancer (Ullrich and Mayer 2007). In low-and middle-income countries (LMICs), 68% of children being treated for cancer reported lack of energy on the MSAS (Olagunju et al. 2016). Causes of fatigue in children with advanced cancer include inactivity, poor nutrition, dehydration, anaemia, pain, depression, insomnia, medication side effects and radiation side effects (Poltorak and Benore 2006). A study in 2000 by Wolfe et al. showed that 89% of children who died of cancer suffered 'a lot' or 'a great deal' from at least one symptom in their last month of life, most commonly pain, fatigue or dyspnoea. Fatigue was reported as not treated in this study. Also in 2000, in 160 children with cancer who completed the MSAS, lack of energy was the most prevalent symptom (Collins et al. 2000).

**Case Study**

*Hugh is now 17 and remains on non-invasive ventilator support at night. Despite this, his daytime energy level has been decreasing and he was started on daytime assistance with a sip and puff ventilator 8 months ago. This has allowed him to continue to attend school part time and he hopes to graduate from high school with his peers this year*

## 9.4 Question 4. Are There Measures of the Symptom Burden of Fatigue in Children?

Fatigue accounts for the greatest degree of suffering in children with cancer (Ullrich and Mayer 2007). It can cause a burden physically, psychologically and socially. The child may experience a decrease in activity, a loss of control in a situation where he or she is already feeling a lack of control and loneliness and isolation from his or her peers. All of these can lead to a decrease in QoL. In children with neuromuscular disorders, fatigue is a progressive, insidious manifestation of the disorder and can become disabling. In a US study by Hendricks-Ferguson (2008), it was shown that in the last week of life, loss of motor function and changes in energy level were the most concerning symptoms noted by parents. There is one study (Rodgers et al. 2013) looking at symptom clusters in various paediatric diseases. Pain and fatigue cluster together and are most studied in children with juvenile idiopathic arthritis, cancer and children at the end-of-life (EoL). In children dying of cancer, the most frequent physical symptoms reported by parents were pain, poor appetite, fatigue, lack of mobility and vomiting (Theunissen et al. 2007).

## 9.5 Question 5. What Are the Non-Pharmacological Ways to Treat Fatigue?

As fatigue can have multiple causes, there can be several strategies to improve the child's QoL. Transfusion may improve fatigue (Munzer 2010), especially in settings where transfusion can occur at home. It can help to perform essential functions in the morning when the child may have more energy (Poltorak and Benore 2006). Scheduling a regular afternoon nap, avoiding complete inactivity, engaging in very mild activity may all help with fatigue.

One study looked at interventions in a hospitalised paediatric population approaching the EoL. No statistically significant benefit was shown with the use of complementary and alternative medicines, exercise based interventions or nursing based interventions (Bhardwaj and Koffman 2017).

**Case 2: A Child with Cancer**

*David is a 14-year-old boy diagnosed with osteosarcoma of his right distal femur. He was started on pre-operative chemotherapy including cisplatin and doxorubicin. He had repeated vomiting for a couple of days after completion of each course of chemotherapy. He always felt very tired at home for 1–2 weeks following treatment and a loss of energy.*

## 9.6 Question 6. What Are the Possible Causes for the Fatigue and How We May Help David?

Cisplatin and doxorubicin is one of the most emetogenic combinations of chemotherapy. The repeated vomiting will make an individual dehydrated which can lead to significant fatigue. Prevention of vomiting by potent anti-emetics such as aprepitant and dexamethasone should be given with the chemotherapy. Delayed emesis is common and individuals should be given adequate fluid hydration, sometimes intravenous rehydration is necessary. Another cause of fatigue is related to anaemia after several courses of marrow suppressive chemotherapy such as doxorubicin. Haemoglobin should be checked if his fatigue symptom is significant, and top up blood transfusions may help to relieve the fatigue. For a previously healthy teenager, the pain and immobility caused by the bone tumour can be very distressing. He cannot maintain his usual active school life and may lose contact with his peers. The psychological cause of fatigue should be addressed and child life specialists (where available) can help to support the adolescent.

**Case Study**

*After surgical resection of the tumour with placement of a prosthesis and completion of chemotherapy, David remained in remission for one year and returned back to school. Unfortunately he returned to clinic with chest discomfort and a chest-X-ray showed multiple bilateral lung nodules compatible with pulmonary metastasis.*

*Due to the extensive involvement of virtually all the lobes in both lungs, surgical resection was not possible. Second line chemotherapy was also considered not of benefit to the extensive lung metastasis, palliative care was offered to David and he accepted. In the following few months, he stayed at home most of the time due to shortness of breath and fatigue.*

## 9.7 Question 7. What Is the Prevalence of Fatigue in Children with Advanced Cancer and How Does It Present?

Fatigue is one of the commonest symptoms in children with advanced cancer. It is present in nearly all children with cancer in the last months of life. Self-reporting of suffering from fatigue was 58% to 93.7%, and it is similar in different ethnic groups (Wolfe et al. 2000; Ye et al. 2019). Fatigue is a multidimensional symptom and may result from physical conditions such as weakness and somnolence. In older children cognitive and emotional manifestations may be predominant, including diminished concentration and depressed mood. Fatigue is not an isolated symptom and always comes with other discomfort, commonly breathlessness, pain and vomiting. Fatigue is more common in cancer conditions with systemic involvement, e.g., leukaemia. Cardiopulmonary involvement by cancer will also make the child get tired easily. Fatigue is also associated with distress from other symptoms such as anorexia, nausea, sleep disturbance, sadness and irritability (Ullrich et al. 2018). The possible associated medical causes that may contribute to fatigue should be managed accordingly, e.g., pain, muscle weakness, dehydration, endocrine abnormalities and infection. Like David with extensive lung metastasis, hypoxia will certainly add to the burden of fatigue.

## 9.8 Question 8. What Non-Pharmacological Measures May Benefit David?

The possible causes leading to fatigue should be explored and addressed. As anaemia is correctable, haemoglobin (Hb) should always be checked and top up transfusions provided if necessary (Munzer 2010). There is no rigid threshold for starting transfusion as some children may feel very tired with higher Hb levels, whilst others may tolerate a low Hb without fatigue. The individual should be assessed for all possible causes that may lead to fatigue, sometimes giving trial treatment to see if there is any response. Hypoxia is also a contributing factor for fatigue, and oxygen (where available) may provide some relief. Disturbed sleep may aggravate fatigue and intervention with sleep hygiene and regular day time activities may help (Ekti Genc and Conk 2008). Non-pharmacological interventions for fatigue have been studied but the result is not convincing (Munzer 2010). Likewise, a study of the direct treatment of fatigue in children with cancer was also shown not to be successful in most cases (Ullrich et al. 2010).

## 9.9 Question 9. What Pharmacological Measures May Benefit David?

Since David also has pain, morphine or other opioid analgesics are commonly prescribed. Some children may be sensitive to one type of opioid analgesic and this can cause significant fatigue. Switching from one type of opioid to another type, such as from morphine to fentanyl may occasionally help in reducing fatigue, but in LMICs switching opioids may not be possible due to lack of access to opioids. Reducing the dose of one opioid and adding on another opioid as a combination may also be tried. Drugs that may cause fatigue should be reviewed and sometimes have to be stopped. On the other hand stimulants may be tried in some children, methylphenidate and modafinil have been reported, but the value of stimulants in children with cancer is not proven. A systematic review on pharmacological treatment of cancer-related fatigue did not show definite benefits (Mücke et al. 2015). Some children with significant fatigue may have a major depressive disorder, and anti-depressants such as selective serotonin re-uptake inhibitors (SSRIs) may be considered.

## 9.10 Conclusion

Fatigue is a distressing symptom and is often seen as a result of progressive disease or treatments such as chemotherapy. It is the most common symptom reported in children with advanced cancer and is increasingly being seen in the range of children requiring PC. Whilst it is important to try and understand the causes of fatigue, more importantly is equipping the child and their family with ways that they can cope with it, and minimise its impact.

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# Nutritional Support

# 10

Ana Forjaz de Lacerda and Gayatri Palat

**Key Learning Points**

1. Malnutrition is often present in children with life-limiting conditions (LLCs) or life-threatening conditions (LTCs), either due to illness or therapy.
2. Early identification and management are essential for preventing deterioration, improving quality of life (QoL) and maybe even survival.
3. The approach should be multidisciplinary and individualised to needs, available resources and child/family preferences. It may change over time.
4. At the end-of-life (EoL), nutritional goals should be reassessed.
5. In the terminal phase it is often in the best interest of the child to recommend minimal feeding and/or hydration.

**Case Study**

*Maria is a previously healthy 13-year-old girl, who was recently diagnosed with high-risk acute lymphoblastic leukaemia. She has been ill for several months and has lost 5 kg. She now weighs 32 kg and measures 150 cm. After a central line is inserted, treatment is started—chemotherapy and steroids.*

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## 10.1 Question 1. What Is Malnutrition?

Achieving a longer survival is not the clinician's only target for children with LLCs or LTCs. Appropriate growth, development and QoL are also critical. Children are recognised to be at higher nutritional risk than adults, since they have less reserves and additional needs. This is especially true in low-and middle-income countries (LMICs), with a high incidence of childhood malnutrition, where a severe illness can rapidly aggravate a child's condition.

Malnutrition can be of two types. The most common is *undernutrition* (the one we usually mean by "malnutrition"); on the other extreme there is *overnutrition*, ranging from excess weight to obesity. The American Society for Parenteral and Enteral Nutrition (ASPEN) defined paediatric malnutrition (undernutrition) as "*an imbalance between nutrient requirement and intake, resulting in cumulative deficits of energy, protein, or micronutrients that may negatively affect growth, development, and other relevant outcomes*" (Mehta et al. 2013, p. 462).

Early identification of patients at risk for malnutrition and timely intervention has the potential to decrease morbidity and mortality, as well as improve QoL. However, nutritional support and management are often a challenge for healthcare providers and parents alike. The former must recognise that when parents face their child's severe illness often the only thing left for them to control is how and what they feed them. Moreover, food carries a great symbolic value. It is thus crucial to reduce parental stress and guilt by being open, creative and building trust, so parents will feel comfortable to ask questions and make suggestions.

Additionally, and particularly in LMIC settings, any nutritional intervention will only be successful if available, affordable and based on the local culture. In traditional medicine, food is often used for a therapeutic effect. Families may have unfounded beliefs (e.g. in India pulses are avoided after surgery for belief they cause "pus"), or religious dietary restrictions (such as fasting, food taboos or special ways of preparing food). Thus, working with the highly respected traditional health practitioners is advised (WHO 2013; Alonso 2015).

## 10.2 Question 2. How Would You Assess Maria's Nutritional Status?

To adequately assess Maria's nutritional status various information must be gathered. To start, *gender, age, height and weight*—these will allow us to evaluate the child's:

- *Percentile* (how it compares to other children of the same gender and age) of height and weight; in infants the *head circumference* should also be obtained;

- *Body mass index*—a relationship between height and weight (by gender and age) that allows us to classify a child according to their percentile (Kidshealth 2015):
  - <5th = underweight.
  - 5th–85th = healthy weight.
  - 85th–95th = overweight.
  - ≥95th = obese.

However, weight may be misleading (a large tumour or significant oedema may artificially increase it) and height may be difficult to assess (as in neurologically impaired children). Thus, where the resources exist, more sensitive assessments should be performed (Romano et al. 2017):

- Measurement of the *triceps skin fold* (correlated to fat mass) and of the *mid-upper arm circumference* (correlated to lean mass).
- *Micronutrient* status.
- *Bone mass densitometry*.

Moreover, any food allergies, restrictions, aversions and preferences should be noted, as well as the current use of medications or supplements (traditional, complementary or alternative). Increasingly used in clinical practice, the *STRONGkids survey* (Hulst et al. 2010) is an easy, practical and reliable tool to evaluate nutritional risk (Table 10.1). Furthermore, interventions are suggested, and reassessment is encouraged.

Maria is currently underweight (<1st percentile), with a BMI of 14.2. According to *STRONGkids* (Hulst et al. 2010), Maria's score would be 5 (cancer + poor nutritional status + decreased food intake + weight loss), indicating a high risk for malnutrition. It is suggested to get a full nutritional evaluation and intervention, check weight and follow-up twice a week, evaluating the nutritional risk weekly.

**Table 10.1** Nutritional risk score and recommendations for nutritional intervention (Hulst et al. 2010, p. 110)[a]

| | Risk from malnutrition and need for intervention | |
|---|---|---|
| Score | Risk | Intervention and follow-up |
| 4–5 points | High risk | Consult doctor and dietician for full diagnosis and individual nutritional advice and follow-up. Start prescribing sip feeds until further diagnosis. |
| 1–3 points | Medium risk | Consult doctor for full diagnosis; consider nutritional intervention with dietician.<br>Check weight twice a week and evaluate the nutritional risk after 1 week. |
| 0 points | Low risk | No intervention necessary.<br>Check weight regularly confirm hospital policy and evaluate the nutritional risk after 1 week. |

[a]Reprinted from Clin Nutr 29(1), Hulst JM, Zwart H, Hop WC, Joosten KF, Dutch national survey to test the STRONGkids nutritional risk screening tool in hospitalized children: 106–111., Copyright 2010, with permission from Elsevier

## 10.3 Question 3. Does Nutritional Status Have Any Implications on Treatment Success?

Malnutrition results in poorer clinical outcomes for many children with severe illness (Visca et al. 2015; McKean et al. 2017). The lack of nutrients leads to several organ or system dysfunctions, which may influence the incidence of complications, treatment tolerance, ability to achieve remission and performance/QoL.

For instance, it has been demonstrated that being obese or underweight significantly decreases event-free survival of children with acute lymphoblastic leukaemia (Orgel et al. 2014). The success of rehabilitation and physical therapy may also be compromised by inadequate nutrition. Children with motor impairment, not able to move adequately for long periods of time, may develop pressure ulcers (associated with pain and infection), muscle wasting and fatigue—adequate protein and micronutrient intake is essential for healing.

## 10.4 Question 4. What Nutritional Related Complications Could Be Expected from Treatment?

In children with cancer, it is estimated that up to 60% are malnourished during treatment (Montgomery et al. 2013), more commonly if diagnosed with solid or brain tumours (Brinksma et al. 2012). When a severe illness is diagnosed, often we are "caught between a rock and a hard place": you must start disease-directed treatments, which may cause adverse side effects and further deteriorate the clinical condition, which may already have caused malnutrition…but if you do not undergo treatment, one cannot alleviate the disease… Therefore, we must be prepared to deal with treatment-related complications and advise the child and family how to best deal with them.

Children who are receiving steroids for a prolonged time, as will happen with Maria, are likely to develop an increased appetite and be prone to some metabolic effects: salt and water retention, increase in body fat, high blood sugar.

Chemotherapy agents used to treat cancer, such as Maria's leukaemia, act by killing rapidly dividing cells. These include mucosal cells lining the digestive tract, from the mouth to the rectum. Therefore, we may expect oral and/or enteral mucositis; she may suffer from diarrhoea, either from the mucositis or from infections. Maria will also be nauseous or may even vomit despite the use of prophylactic antiemetics; she will also develop anaemia (low red blood cells), thrombocytopenia (low platelet counts, leading to bleeds), fatigue, constipation, all of which impair her ability to eat, but also to digest and absorb food.

If Maria were to receive radiotherapy, and depending on the irradiated field, she might also suffer from post-inflammatory damage to mucosal cells, again causing mucositis, but also possibly dysphagia (inability to swallow) and diarrhoea. Since the enzyme lactase (which digests lactose, the sugar in milk) is the first to be lost when there is severe intestinal mucosal damage it is recommended to avoid milk-containing products, and probably also fatty acids (Kuiken et al. 2015)—labels must

be read! Yoghurt, as a fermented food, is usually better tolerated. Grain or nut "milks" are another alternative.

## 10.5 Question 5. Which Nutritional Interventions Would You Recommend to Support Maria Through Her Treatment?

For nutritional interventions a multidisciplinary approach is recommended, involving physicians, nurses and dieticians, but also often psychologists and speech and/or occupational therapists where available (Sharp et al. 2017).

Whenever there is an intact oral route, with the ability to chew, swallow, digest and absorb food, this should be the preferred feeding method (*oral nutrition*). The first step must be to evaluate the child's food preferences and aversions. Nonetheless, some "incentives" may still be needed for children to eat: asking and allowing them to help choose and prepare food, inventing child-friendly plate presentations, avoiding strong smells and flavours, creating reward systems, etc. For some picky eaters it may be useful to prescribe a multivitamin and mineral formula, but care must be taken to read labels and avoid over intake of some nutrients. For example, children with cancer related anaemia *DO NOT* need iron supplements—it may even be harmful. Parents often also ask about appetite stimulants. Remaining active and sharing meals with family and friends is helpful. In severe cases, a short course of steroids may be tried. Some children may benefit from behavioural interventions and/or oral-motor therapy (Sharp et al. 2017).

There are situations when even optimal oral intake is not enough to achieve the energy and nutritional needs, as in children with cardiac or respiratory conditions. Then, and also if chewing and/or swallowing are impaired (as in the case of oral mucositis or neurological disabilities), an alternate route must be sought for *enteric nutrition* (Hauer 2018; Yi 2018). The choice will depend on clinical considerations and child/family preferences—nasogastric tube (the most common, inserted through one of the nostrils into the stomach), orogastric tube, nasojejunal tube (past the stomach, into the proximal intestine) or even a percutaneous gastro or jejunostomy (a surgically inserted tube, through the abdominal wall) if the horizon is that of long-term need. The experience of receiving tube feedings varies widely; it impacts on the child, parents, family and surroundings, both in positive and negative ways (Nelson et al. 2015). Beforehand it is essential to discuss values, past experiences and expectations. In fact, if concerns are not properly addressed, many children and families will choose parenteral (see below) over enteral nutrition (Montgomery et al. 2013).

Sometimes a "regular" diet is enough, with guidance from a dietician. For tube feedings a homemade blended diet is not recommended as first choice, except if that is the parent's wish (Brown 2014) or the only available, as may occur in LMIC settings. Here, nutritional counselling should not only adopt the local culture but also include education about safe food handling (buying and storing) and preparation (cooking and serving). Instructions must be simple and given in the


**Fig. 10.1** An example of an illustration re nutrition and Cancer (Lauler and Doherty, World Child Cancer-Bangladesh Twinning Project. Printed with Permission)

local language; it is advantageous to use illustrations, since people may not be able to read (Fig. 10.1).

Nonetheless, very often supplements are needed—a dietician, according to the setting, should lead the recommendations. Where resources are low, food-enriching techniques may be used, like adding extra fat, sugar or flour. Commercially available supplements can be of individual *macronutrients* (carbohydrates, a.k.a. sugars, lipids, a.k.a. fats, and proteins), usually powders or liquids that can be mixed into regular foods. They may also be offered as formulas with a combination of macro and *micronutrients* (vitamins, minerals, etc.), as porridges, puddings or drinks.

Occasionally the medical team or the parents will suggest starting a ketogenic diet, more commonly in children with neurological conditions. Although it may help with symptom control, there is a concern about the long-term effects on bone health (Simm et al. 2017). Families may seek other restrictive diets, usually "something-free". They should feel enough opening and trust in their healthcare providers to allow for discussion of these ideas. Some may be interesting and

harmless, while others are a non-sense that only contributes towards the child feeling miserable and unloved.

Sometimes children are able to eat but not to digest the nutrients so the intestine can absorb them. In these cases, the dietician will prescribe an "elemental diet"—a commercial formula with pre-digested nutrients. These formulas are expensive and unpalatable, usually requiring the use of a tube.

When children have gastric reflux or vomiting, they may still be fed by the oral or enteral route, but will need some additional measures:

- Use of food thickeners—commercial formulas; xanthan gum; rice, corn or carob flour; over soaked flax or chia seeds.
- Smaller and more frequent meals.
- Positioning after meals—not to lie down for at least 30 min.
- Medication: ondansetron (decreases vomit reflex), metoclopramide (promotes movement), anti-acids (decrease heartburn).
- More severe cases may need surgery.

On the other hand, in some situations food is not properly absorbed through the intestines. Then we must resort to *parenteral nutrition*, where the child receives all the nutrients directly in the blood stream, usually through a central line (peripheral lines cannot stand the volume and/or concentration which are needed). Receiving parenteral nutrition does not mean the child must stay in the hospital, since in many countries there are home programmes; however, the rate of complications, mostly infections, is very high (Raphael et al. 2019), and this may not be an option in LMICs.

It must be stressed that even if a child is receiving enteral or parenteral nutrition, she may still be able to enjoy at least small amounts of food by mouth—but only after checking with the responsible physician.

Many families, both in high and low resource settings, will seek the use of complementary or alternative medicine, which many times takes the form of nutritional supplements. While it is recognised that some may help to reduce selected toxicities of traditional medicine interventions (as chemotherapy or radiotherapy), most have not been tested adequately. As such, little is known about their true efficacy, safety and potential interactions with conventional interventions (Kelly 2008).

When a child is exposed to prolonged steroid use, families should pay special attention to the intake of:

- Salt: avoid salty foods (snacks!), use less salt while cooking.
- Carbohydrate: favour whole grains (slowly absorbed sugars), vegetable and legumes, avoid sweets and sugary drinks.
- Fat: avoid saturated fats (the solid ones, of animal origin).

Some conditions deserve particular considerations. In chronic renal disease, it may be needed to restrict foods rich in phosphorus, sodium, potassium or highly acidic; simultaneously, the child may receive alkaline supplements or ion-binding powders (Akchurin 2019). Oral glutathione supplements have been tried in children

with cystic fibrosis, with encouraging growth and clinical outcomes (Visca et al. 2015). For a long time in Oncology a "neutropenic diet" has been recommended during periods of low blood counts and heightened immunosuppression: it is highly restrictive, lacking in raw fruits and vegetables. In recent years the trend has been to substitute for a well-balanced diet, with carefully chosen and safely handled foods. For children with neurological impairment we recommend reading the European Society of Pediatric Gastroenterology, Hepatology and Nutrition (ESPGHAN) guidelines (Romano et al. 2017).

It must be stressed that when children have a poor appetite (anorexia) we may have to compromise on some of our goals and allow a few "guilty pleasures" if that is the only food the child will have. And a soda or chocolate once a week will do no harm! In fact, many of the foods children *LOVE* are highly caloric and nutritionally interesting: full-fat ice-creams, sundaes, puddings (especially ones made with rice), etc.

**Case Study**

*While on treatment, Maria's leukaemia relapses. There is no suitable bone marrow donor to undergo a transplant. After remission re-induction treatment fails, her clinical condition is very poor (profound bone marrow aplasia, severe oral and enteral mucositis). Her parents agree with the team that aggressive therapy will not be pursued—the focus will be on QoL and on restoring Maria's ability to go home.*

## 10.6 Question 6. What Should Be the Focus with Regards to a Nutritional Approach with Maria, Now That Aggressive Therapy Is Not Being Pursued?

At this turning point, the team's focus should shift to Maria's QoL, protecting her from adverse events and keeping her away from the hospital for the longest time possible. Even if Maria had not had any major nutritional issues so far, it is very likely that she will now. Her appetite may increase (if doctors prescribe steroids to slow disease progression—see above), but most likely it will decrease. Fatigue now also plays a major role, as well as disease-related factors which increase needs ("consumption").

Cachexia ("bad condition", from the Greek "kakos" and "hexis") has been extensively studied in adult cancer patients, where it is defined as *"a multifactorial syndrome characterised by an on-going loss of skeletal muscle mass (with or without loss of fat mass) that cannot be fully reversed by conventional nutritional support and leads to progressive functional impairment"* (Fearon et al. 2011, p. 490). It is classified in three stages (Fig. 10.2), the last corresponding to a low performance status and short life expectancy.

In children the first two stages are commonly described as "malnutrition", and only the last as "cachexia". The hallmark of advanced paediatric cachexia is growth impairment, in the absence of endocrine disorders. In chronic renal disease it has

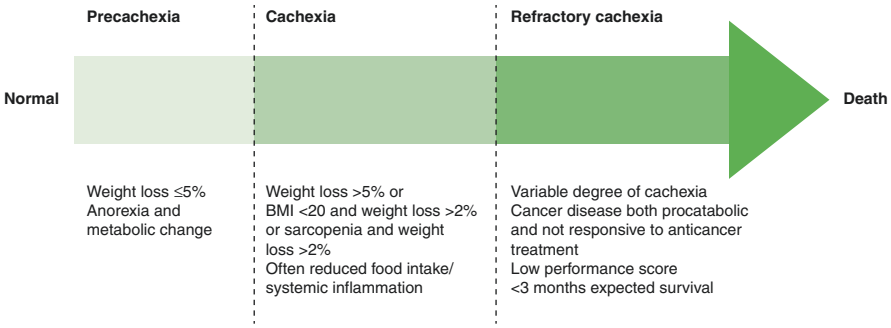


**Fig. 10.2** Stages of cancer cachexia (Fearon et al. 2011, p. 491) (Reprinted from Lancet Oncol 12(5), Fearon K, Strassser F, Anker SD, Bosaeus I, Bruera E, Fainsinger RL, Jatoi A, Loprinzi C, MacDonald N, Mantovani G, Davis M, Muscaritoli M, Ottery F, Radbruch L, Ravasco P, Walsh D, Wilcock A, Kaasa S, Baracos VE. Definition and classification of cancer cachexia: an international consensus 489–495., Copyright 2011, with permission from Elsevier)

been suggested that lack of expected weight gain should also be classified as cachexia (Mak 2016); the same should probably be true in other LLC or LTCs.

Cachexia is associated with increased rates of hospitalisation, duration of hospital stay and mortality (Van Doren et al. 2015). Additionally to considering oral or enteral feeds and/or supplements, a trial of megestrol acetate (an appetite stimulant) may be prescribed if available; some encouraging results, with minimal side effects, have been seen in children with cancer (Cuvelier et al. 2014) and chronic renal disease (Mak 2016). In the latter, long acting ghrelin (the "hunger hormone") mimetics seem to improve appetite and nutritional status (Mak 2016). It may also be useful to initiate omega-3 fatty acids, if survival for longer than 8 weeks is expected (Garcia and Shamliyan 2017).

Australian colleagues recently developed an excellent practical online guide of nutrition in the palliative and EoL care setting, with tips for carers and children (NEMO 2017) and recommendations for several clinical situations (NEMO 2019).

**Case Study**

*After 2 months at home, Maria is now terminally ill. She does not have mucositis, but sometimes has bleeding gums, since her platelet count is very low. She no longer wants to leave her room and play.*

## 10.7 Question 7. How Would Your Nutritional Approach Change Now That Maria Is Terminally Ill and How Would You Explain Any Changes to Her Parents?

In the terminal phase administering enteral or parenteral (intravenous or subcutaneous) nutrients and fluids may cause more harm than good. There may be difficulties in oral feeding, pain and discomfort related to digestion, increased flatulence or bowel movements, etc. A decreased intake of food and fluids will mean less trips to

the bathroom or diaper changes. It will also lead to a certain degree of dehydration, causing a natural sedation, and to a decrease in respiratory secretions (the "death rattle"). Additionally, as the body is shutting down and the child is less active, the caloric needs are much lower; moreover, we are no longer concerned about the long-term effects of malnutrition.

All this should be explained to the family, since feeding a child is a basic parental instinct. Most parents will understand and agree to decrease or even suspend feedings and fluids. A recent study with parents showed that none regretted this decision, the child's death being peaceful and comfortable (Rapoport et al. 2013). The need to nurture by feeding may then be fulfilled by appropriate mouth care—washing the mouth (lips, gums, tongue, teeth) with cold water or a saline solution, applying ointments, offering ice chips, etc.

If the parents cannot come to terms with suspending, or if they agree to decrease feedings and fluids but Maria does not have the ability to chew and/or swallow, then she may need to have a tube or a line placed. This may also need to be considered if she needs medication that she is not able to take by other routes (see Chap. 14 on EoL). Having a tube or a line placed may be tricky at this stage—a central intravenous line may not be feasible or appropriate; peripheral intravenous or subcutaneous lines do not last longer than 5–7 days and may cause pain, discomfort and infection. Continuing to act in Maria's best interest, we must, maybe for the last time, openly address with Maria and her family the benefits and burdens of our interventions.

## 10.8 Conclusion

Malnutrition is often present in children with LLC or LTC, resulting in poorer clinical outcomes. It should be addressed early through a multidisciplinary approach, and the solutions must be worked out with the child and family, considering the disease trajectory, the setting and the available resources.

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# Psychological Care and Distress

# 11

Tracey Brand and Tamara Klikovac

**Key Learning Points**

1. The establishment of a therapeutic relationship between the children's palliative care (CPC) team and the family is important to provide good psychological care and support.
2. Understanding the family and the child's psychological distress and how to manage it is essential.
3. It is important to assist the family and child through the disease trajectory, utilising appropriate skills and knowledge such that they feel supported and know where to go if they need help.
4. Assisting the family at end-of-life (EoL) and into bereavement can be challenging, but an important part of palliative care (PC) provision.

## Case Study

*Jeremy, age 5, had been unwell, lethargic and "not his usual self" for several months. In September 2016 his mom, Natasha, took him to the local clinic. The clinic felt that he was anaemic and suggested a course of Vitamin B12. This continued for 2 months with no affect. Finally they were referred to their base hospital. While there, several tests were conducted and the team decided to send Jeremy to the quaternary hospital for the province 100 km away. Communication had been very poor from the outset and Natasha was incredibly frustrated and angry by the*

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*time they arrived at the quaternary hospital. More tests were conducted and in January 2017 it was confirmed that Jeremy had acute myeloid leukaemia (AML) with central nervous system (CNS) involvement.*

*By the time the doctors informed Natasha about Jeremy's diagnosis she was very angry. In addition to being pushed around for months without any kind of intervention, she is now 100 km away from her husband and other children as well as at risk of losing her job. Natasha responded with a high level of anger to the doctors when Jeremy's condition was first explained to her. She felt that it was due to negligence that it had taken so long for the diagnosis to be made and now he was even sicker. The doctors tried to justify the length of time by explaining how the health care system worked. This further infuriated Natasha.*

*Given Jeremy's high-risk diagnosis and the level of anger that Natasha is exhibiting, the PC social worker was called in. The social worker allowed Natasha to "vent". She gave her the space to express her anger and frustration at the delay in the system and the manner in which she had been treated. The social worker did not try to justify the system, instead she apologised that it had been such a terrible time and that Natasha had felt let down. The social worker also gave Natasha the opportunity to tell her story. Throughout this, empathy was shown as well as minimal encouragers and active listening. By acknowledging Natasha's frustrations and anger, the social worker was able to give her the space to explore how she really felt about Jeremy's diagnosis. Up until now, her thoughts and energy had been on what the system had done wrong and not on the reality of Jeremy's diagnosis.*

## 11.1 Question 1: What Are the Psychosocial Issues at the Point of Diagnosis?

It is important to conduct a psychological and social assessment at the point of diagnosis, to help get an understanding of where the child and their carers are at, what they understand with regard to the child's condition, and how they may be coping with what is happening to them. It is important to ascertain what crises the family has previously dealt with, help them identify their previous coping strategies and help them build resilience. Jeremy's case is all too familiar, with many families being "pushed around" the healthcare system for months prior to diagnosis. Alternatively you have families where a child becomes acutely ill and they are "suddenly" diagnosed with a life-threatening condition.

It will be essential to show a range of skills when undertaking a psychological and social assessment. Skills that were shown to Jeremy's family include the following:

- Empathy.
- Active listening.
- Allowing Natasha to be angry but not justifying or defending what makes her angry, rather apologising for it.

- Being present—not just talking to Natasha but also giving her the space to tell her story and explore her feelings.
- Helping Natasha understand more about Jeremy's diagnosis and how the treatments are going to work.

Some common reactions to the diagnosis of a life-threatening illness include:

- Shock.
- Denial.
- A sense of being overwhelmed.
- Information overload.
- Questioning—Why my child? Why now? Who is to blame?
- Fear.
- Financial concerns, e.g. the impact on job, cost of caring for Jeremy.
- Anticipatory grief.
- Coping with the sudden change in lifestyle.

## 11.2 Question 2: How Would You Undertake a Psychological and Social Assessment?

Children's reactions to illness depend on the child's age and cognitive development. Knowing how children of different ages and stages of cognitive development understand concepts, such as illness, hospital, treatment and death is crucial to effective communication.

Natural egocentricity, magical thinking and associative logic all characterise preschool children's thinking (ages 3–5). They are therefore prone to interpret illness and suffering in terms of their own thoughts and actions and to have misconceptions about the cause of the illness or the reasons for medical treatment (Wolfe et al. 2011). It is important to:

- Observe body language—what is it telling you about the child?
- Observe the language of play because preschool children communicate through play. Play is a regulative mechanism. Through play the child expresses traumatic fixations, conflicts and hostilities. Medical play can also be useful.

Assessment techniques for children include:

- *Drawing and painting*—Get the child to draw a picture of themselves and their family. As children draw ask questions about the person/people in the drawing. Look at size, colours, space, emphasis or omission, shadows, content, environment, etc.

- *Story telling*—This is an easy way for establishing contact with children. Stimulate children's fantasy and creativity. Use known stories or make up stories with the children: who are you now? what do you want to be? are you a little mouse? or a cat? what is the mouse/cat doing now? where is the mouse/cat going? what is he/she afraid of? are they hiding from something?
- *World chart; body mapping.*
- *Using a puppet as an object of transference*—Get the child to tell the puppet their story. Children find it easier to talk to someone or something other than an adult.
- *Play dough*—this can be used as relaxation and a way to express feelings.
- *Faces*—similar to the pain assessment scales.

The best way to assess a family is through a genogram. A genogram is a pictorial display of a person's family relationships and medical history. It goes beyond a traditional family tree. It helps us find out what the family has been through and how they have coped previously. It also helps with understanding family dynamics and relationships (Fig. 11.1).

Genograms can be used to map out and help understand the following:

- Who is in the family, e.g., father, brother, sister, etc.
- Health status of all family members.
- Child and family's knowledge of and reaction to the illness.
- Beliefs, attitudes and expectations.
- Coping ability through previous crises.
- History of psychological illness and drug or alcohol abuse.
- Nature and stability of residential and occupational arrangements.
- Quality of relationships.
- Socioeconomic status.
- Socio-cultural factors or religious beliefs that might affect treatment decisions.
- History of previous losses.
- Sources of emotional and financial support.

**Case Study**

*During the induction phase of Jeremy's treatment Natasha stayed with him. This caused great stress on the family as she is the main breadwinner. Calvin, Jeremy's dad, worked part-time and due to lack of funds could not visit Jeremy as often as he would have liked. Natasha's stress as well as watching Jeremy get sick with the chemotherapy led to an often volatile relationship between Natasha and the treating team. Natasha was angry and frustrated that the treating team felt the need to continually tell her that Jeremy's diagnosis is high risk and that prognosis is poor. She felt that they were insensitive and that they were taking away her hope. The PC social worker became a buffer between the two, trying to convey each other's concerns and queries.*

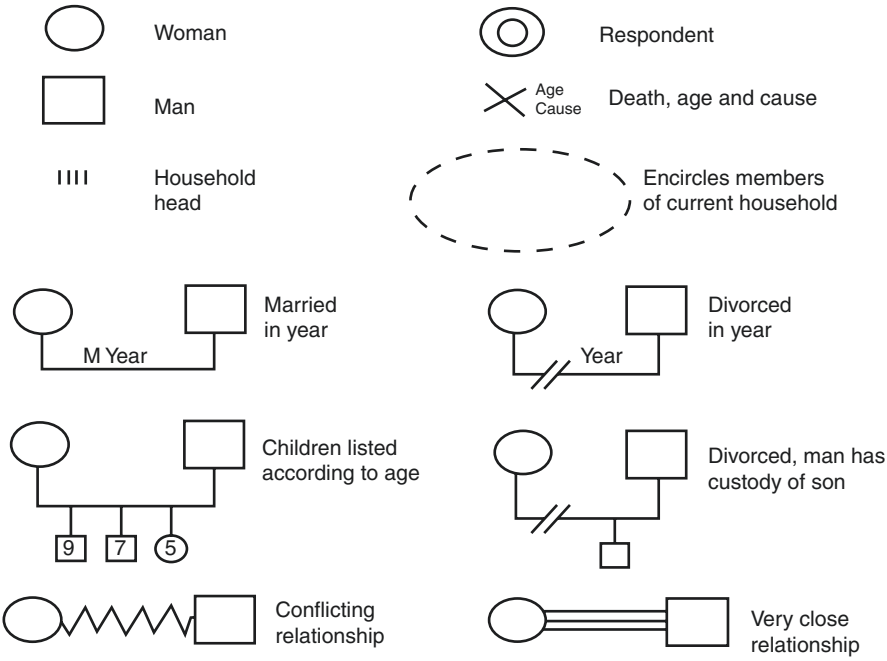


**Fig. 11.1** Recognised genogram symbols (Watts and Shrader 1998, p. 460, by permission of Oxford University Press)

## 11.3 Question 3: What Is Required of the Psychosocial Team During Treatment?

During the treatment phase it is important that the psychosocial team are available to the family. Their responsibilities at this time include:

- Continued psychosocial support.
- Assisting with information sharing between the family and the treating team.
- Assisting in ensuring understanding.
- Assisting with effective communication between family members.
- Acting as a mediator between the family members, and assisting the family to understand each other's feelings and concerns and assist them in finding their own way forward.

It is important to remember that families such as Jeremy's will find their own solutions in their own time. We cannot "force" our beliefs, solutions or desires on to the family. We need to facilitate discussion and support their decisions. A critical role is acting as a mediator and assisting the family to understand each other's feelings

and concerns and assist them in finding their own way forward. It is important to walk beside the family providing support at a difficult time. Try and avoid giving concrete advice. Families will often ask what we would do. It is important to refer back to them—"I'm not sure. What do you think? What do you want to happen?"

Skills utilised by the social worker and care team at this time include the following:

- Empathy.
- Active listening.
- Mediation.
- Facilitating communication.
- Supportive counselling.

During this time, as with the time of diagnosis, many families and the child themselves search for meaning and purpose; they try to "make sense" of the situation. While suffering is seen as a "normal" component of illness, the experience, meaning and purpose are less apparent. This is often experienced more acutely during the treatment phase as families witness their child going through a torrid time with hospitalisations, side effects from the chemotherapy and many scary moments.

Within this context meaning is seen as *the process of making sense of life situations*. This has both an existential component and a cognitive component. Deriving purpose *relates to the aspects of a person's life that are most significant to the person* (Parsonnet and Lethborg 2011). A person's worldview will determine how they view the diagnosis and how they are able to manage. Often this is turned upside down and the family search for stability which will lead to their finding meaning. An inability to find meaning can lead to profound despair, loss of joy and a disconnectedness with life. The psychosocial team's role is therefore imperative, as they assist the family and the child renegotiate their understanding of the world and accepting their present situation.

**Case Study**

*Jeremy was an incredibly intelligent 5 year old. He realised that there was something serious going on and demanded to know what was happening. Natasha was very anxious to tell him as she did not want him to be scared or to think that he was going to die. The social worker and Natasha spent time together discussing the benefits of disclosure. They had the choice to tell him themselves at a pace that suited him, before he was disclosed to accidentally during a ward round. Disclosure is the process of revealing or relaying information to the child about their illness. It must be noted that this is a process and not a once-off event. As Jeremy's condition changed and as he matured, information had to be given to him again in less child-like terms. It was agreed that Natasha and the social worker would tell Jeremy together. They discussed with Jeremy what was happening to him, the treatment regime, the different tests that he would have during his treatment and how long he would be in hospital.*

## 11.4 Question 4: What Is Our Responsibility in the Case of Jeremy?

Throughout his treatment, it will be important to provide psychological and social support to both him and his family. As is often the case, Jeremy is receiving treatment a long way from his home and, apart from his mum who is caring for him, he is a long way from the rest of his family. This has implications not only for Jeremy and his mother but also the rest of the family, including any siblings who may be feeling neglected by their mother as she has had to stay with Jeremy. Thus, providing psychological care and support to the whole family may be a challenge.

During their discussions with Jeremy, it was important that Jeremy's physical and cognitive ages were taken into account by his mother and the social worker. Thought needs to be given as to the best way of discussing diagnosis and prognosis with Jeremy. Guidelines exist, such as those published in the Lancet (Stein et al. 2019), to support professionals telling children, or those by Sara Gable on guidelines for communicating effectively with children (Traub 2016). Through using these guidelines it was possible to identify the best way to talk to Jeremy about what was happening to him.

In addition to that, objects of transference were used. As much as Jeremy wanted answers, children have difficulty expressing themselves verbally; therefore, play is very important. Play is a meaningful and significant activity that allows children to express themselves, and children often use toys to say what they feel they cannot say aloud (Boucher et al. 2014). For Jeremy, he chose two puppets. He gave the one to the social worker and he used the other. Changing his voice he asked all the questions he needed answers for through his puppet. The social worker answered him through her puppet. According to Landreth play is the child's symbolic language of self-expression and can reveal:

a. What the child has experienced.
b. Reactions to what was experienced.
c. Feelings about what was experienced.
d. What the child wishes, wants or needs.
e. The child's perception of self (Landreth 2012).

Skills utilised by the social worker and care team at this time include the following:

- Cognitive age assessment.
- Active listening.
- Facilitating communication through play.
- Disclosure
- Supportive counselling.

**Case Study**

*Through the 3 months of intensive induction therapy Jeremy became very ill. Natasha's stress and anger levels increased significantly and her relationship with the treating team was even more strained. The PC social worker spent a lot of time exploring Natasha's fears, stressors and anger as well as mediating between the treating team and Natasha.*

*After the 3 months induction Jeremy's results still came back positive. The chemotherapy had not had any impact on the cancer cells. A decision had to be made whether to try more invasive therapy or to look at palliative chemotherapy with the aim of controlling his symptoms and giving him time at home.*

## 11.5 Question 5: What Are the Psychosocial Issues When Treatment Fails?

When treatment fails, it is a time of denial, grief and anger for many, their hope of a cure and that "everything will turn out ok" has been dashed. In situations such as with Jeremy, when one of his parents is busy working far from the hospital, it can also be hard to get both parents together to discuss the options, yet this is so important, especially due to the conflict between Natasha and the treating team. In Jeremy's case, the PC social worker sat with Natasha and Calvin together with one of the treating team. Before the issues relating to Jeremy could be discussed the conflict between Natasha and the treating team had to be addressed. Unresolved conflict can be detrimental to and potentially derail the goals of care (Jacobs 2011). Until Natasha felt she was on the "same page" as the treating team any discussions about future care were futile. Through a constructive debriefing session Natasha was able to express her feelings and their causes. The representative of the treating team listened to all her grievances without interrupting with assistance from the social worker. The social worker then mediated a way forward that took into account all party's needs, feelings and desires. Through effective conflict resolution common ground was found and Natasha was able to move forward (Jacobs 2011). It must be noted that while some of her anger was justified, a large portion was linked to her denial of Jeremy's diagnosis and her inability to help him, a natural reaction by a parent to their child's illness.

Decisions needed to be made about Jeremy's future care and these were discussed with Natasha. Natasha felt that she could no longer put Jeremy through the trauma of chemotherapy. He had not responded well physically to it and being away from home was very difficult on both of them. By this stage Natasha was back at work and a caregiver was staying with Jeremy. This was not ideal as the separation anxiety made Jeremy's hospital stays even more difficult.

For decisions such as these to be made there needs to be a foundation of good communication. Communication skills in paediatric PC are imperative and "*because communication is both verbal and nonverbal, it is an art and a science that is not*

*static but rather an ongoing and dynamic process requiring robust collaboration among the interdisciplinary team*" (Jacobs 2011, p. 643).

Too often healthcare professionals shy away from prognostic information as they feel it will cause more harm than good. Research has shown, however, that parents have specific needs when it comes to decision-making in the face of EoL and PC. These include:

- Honest and complete information.
- Access to staff.
- Communication.
- Care coordination and continuity.
- Presence.
- Emotional expression.
- Support by staff.
- Preservation of the integrity of the parent–child relationship.
- Faith.
- Cultural sensitivity (Jacobs 2011).

## 11.6 Question 6: What Are the Decisions that Need to Be Made?

As a team and as a family, decisions about Jeremy's care at this stage need to be made. This is done in the form of an "advance care plan" (ACP). An ACP looks at possible scenarios and how each of these can or should be managed. It is important that this is done with the family as they are the ones that need to ensure its implementation. Issues that need to be explored include, but are not limited to:

- Withholding or withdrawing non-beneficial medical interventions.
- Pain and symptom management.
- Artificial feeding and hydration.
- Resuscitation or AND (Allow Natural Death).
- Place of care (Jacobs 2011).

During the teams' discussions with Natasha and Calvin the following was decided:

1. They would continue with monthly clinic visits where Jeremy could be assessed by the doctors. Part of these visits would include assessing pain management. They would also continue with the low-dose oral chemotherapy to help slow the cancer's progression. This would continue until such time as the treatment was causing more harm than good.
2. Pain and symptoms to be managed by the PC team. Natasha had telephonic contact with them and any issues relating to Jeremy's physical symptoms would be

discussed. For Jeremy's spiritual and psychological pain, the PC social worker was available at the clinic visits but also telephonically.

3. They decided that they want Jeremy at home as much as possible and want to avoid unnecessary hospitalisations. They also felt that they would like to, as far as possible, manage his EoL care at home with the assistance of the PC team and the local hospice. If it got too difficult he would be admitted to the hospice. In agreeing to this they were informing the team that they would like to allow natural death and therefore no resuscitation.
4. Both were adamant that they did not want him to have a nasogastric tube for feeding or an IV line for fluids. They wanted to limit the number of painful procedures he had to go through.

The plan also included possible EoL events and how Natasha and Calvin could manage each of these at home.

Both Natasha and Calvin were anxious as to how to discuss all this with Jeremy. They asked the PC social worker to have the discussion with Jeremy in their presence. Before this could take place it was important to ascertain Natasha and Calvin's feelings about death, illness and bereavement. *"The seriously ill school-age child needs to be able to express fears and concerns in order to decrease anxiety and reduce feelings of isolation and alienation"* (Gibbons 2009). Without addressing Natasha and Calvin's beliefs they would not be able to hear Jeremy's and this would isolate him from them.

Jeremy was very aware that his treatment had not been going well and he wanted to know what the plan was. The social worker discussed with him that his parents did not want him to have to be in hospital any more, that they did not want him to receive any more of the strong chemotherapy and that they wanted to have him home with them. Jeremy was happy with this but he needed to know more. He needed to know how long he would have at home. This was a huge shock for Natasha and Calvin as they did not think that Jeremy knew the severity of his diagnosis nor the treatment failure. According to Gibbons (2009, p. 56) "*terminally ill school-age children are often aware of their fatal prognosis without being told. They are acutely aware of nonverbal clues and often understand much more than their parents or caregivers realise.*" When talking to children it is important to ascertain that you understand them correctly. Therefore answering a question with a question is a good strategy. Jeremy could have been asking how long he had at home before he had to return to clinic, not necessarily before he dies. When asked what he meant Jeremy stated that he knew he is going to die, he just wanted to have a bit of time at home because he has been away so long.

Through the social worker's interventions Natasha and Calvin were able to discuss all their concerns with Jeremy as well as address his fears and understand what he wanted to do before he dies.

Skills utilised by the social worker and care team at this time include the following:

- Empathy.
- Conflict resolution/mediation.
- Breaking bad news to parents and Jeremy.
- Facilitating communication.
- Prognostic counselling.
- Supportive counselling.
- Decision-making.
- Advance care planning.

**Case Study**

*As Jeremy's condition worsened his parents were in contact with the PC team frequently. They were supported in the decisions they were making as well as assisted in keeping Jeremy as comfortable as possible. They were encouraged to help Jeremy with his legacy planning and Jeremy planned his own funeral and wrote a letter to any future siblings he might have.*

*Jeremy died peacefully at home with his family around him. Everyone had a chance to say goodbye to him and honour his last wishes. Through effective PC intervention both Natasha and Calvin were able to grieve effectively.*

## 11.7 Question 7: How Can You Support the Family at the End-of-Life and Beyond?

At the point of EoL, it is imperative that Jeremy's family are given choices and control. This is a time where the family and the child have little control over what happens and little choice. Giving them choice and control in an uncontrollable situation empowers them and assists them in making good decisions for themselves.

A family can be helped by the following:

- Try to do everything you can to give the child and the family choice and control.
- Talk through in detail what might happen (best and worst case).
- For each scenario, explain and agree exactly what the plan is.
- Try to involve them in any plans so that they will not become passive and fearful.

After death the family may not want to have contact with the team as they are a reminder of what happened to their child. As a psychosocial team it is important that we are respectful of this but also that the family know that, if they need anything, the team are still available to them. Sending regular messages of support, especially on the day of the funeral, is often welcomed.

If the family would like to remain in contact, it is recommended that two or three bereavement sessions are held. These are an opportunity for the family to have any doubts or unanswered questions dealt with but also an opportunity to start processing

their grief. It is important for the psychosocial team to be aware of any unhealthy grieving patterns and/or complicated grief and to refer. Further information about supporting the family at the EoL and beyond can be found in Chaps. 14–16.

Skills utilised by the social worker and care team at this time included the following:

- Empathy.
- Supportive counselling.
- Bereavement support.

## 11.8 Conclusion

Throughout the provision of PC, it is important to provide psychological care. There are a range of skills that should be utilised in doing this such as: empathy; active listening; being present; allowing the expression of feelings; mediation; facilitating communication and counselling. Children and their families will experience a range of emotions throughout the disease trajectory and will need support at different times. It is therefore important to assess the families' social situation using tools such as a genogram, in order to ascertain where and who they can get their support from. Psychological support should continue at the EoL and into bereavement, and whilst challenging, this is an important aspect of PC.

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# Spiritual Care

# 12

Joan Marston, Giovanna Abbiati Fogliati, and Richard W. Bauer

**Key Learning Points**

1. Spirituality is a unique construct within children's palliative care (CPC) service provision.
2. Religion and spirituality both need to be assessed and be part of the documented treatment and care plan.
3. All members of the multi-disciplinary team (MDT) should have basic training in spiritual assessment and care of children.
4. The role of a CPC chaplain requires specialised education and skills.
5. Children may express their spiritual needs and spirituality through verbal and non-verbal means.

**Case Study**

*Ming was born in Campo San Pietro, Italy, to Chinese parents. As a young child he was sent to live with his grandparents in China, while his parents remained in Italy for work, together with a younger sister. His grandparents were very religious and brought him up in the Chinese culture.*

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*When he was 6 years old, he was diagnosed with cancer of the bone in his leg (osteosarcoma) and was frequently admitted to hospital. Eventually the doctors in China said they "could do no more" for him and at the age of 11 he returned to his parents in Italy, seriously ill.*

*He received radiotherapy and chemotherapy in a hospital in Padua. Even though he spoke Chinese and very little Italian he was very observant, intelligent, and inquisitive, frequently looking up information on his tablet which was always nearby. He learned from the many people he interacted with in the hospital about the "mix of humanity". His caregivers commented, "His eyes tell everything, even the way he embraces wonder". Not wanting to die, he is willing to accept any treatment for his cancer.*

*Understanding that his son is suffering both physically and spiritually his father, who was also religious and a pragmatist, gave him a Bible and said, "Read this book, see how it can help you".*

*On his own Ming began to read the gospels and became fascinated with the person of Jesus, doing even more research on his tablet. He was able to identify that just as Jesus' mother suffered, his own mother suffers as well. Wanting to understand more, he asked for an Italian language teacher and for someone to help him understand more.*

*As his condition deteriorated he was transferred to the Padua Children's Hospice. His PC physician asked him "What makes you happy?" and he replied he would like to be baptised.*

*Despite being calm and peaceful most of the time, he went through times of anger and asked questions such as, "Why my parents, my little sister Sofia will stay and I have to go? Why? Why?" At times he expressed anger with God, when he became seriously ill after visiting a church. "Why, I came to know You and I feel so bad now, not even you can help".*

*Sr Tania, a Catholic nun met regularly with him and prepared him for baptism. When he met her he said "Finally I have someone who can explain to me more about what I choose". He was baptised when he was 12. He chose Angelo as his baptismal name as he had heard about the guardian angels and felt that when he died guardian angels would come to hold his hand. He smiled, concentrated on every word said, his eyes shone during the ritual and he asked for his ears to also be anointed with holy oil so he could listen carefully to the words.*

*Angelo died when he was 13, and was buried according to Catholic tradition. His parents supported and respected their son's wishes and his spiritual journey.*

## 12.1 Question 1: What Is the Difference Between "Religion" and "Spirituality"?

When the WHO defined palliative care (PC), they stated that it is "*the prevention and relief of suffering by means of early identification and impeccable assessment and treatment of pain and other problems, physical, psychosocial and spiritual*"

(WHO 2002). Care of the spirit is often seen as only belonging to the role of a religious leader or chaplain when PC practitioners lack a clear understanding of the distinction between spirituality and religion. While both religion and spirituality are based on meaning and connectedness, religion is often described as the external manifestation of these realities, while spirituality is the internal experience (Hyman and Handal 2006).

Emile Durkheim defined Religion as "*the shared reverence for the supernatural, sacred, or spiritual as well as the symbols, rituals, and worship that are associated with it*" (Taves 2009, p. 176). He gave names to religions such as Christianity, Judaism, Hinduism, Islam, or Buddhism and recognised they have different viewpoints, structures, guidelines, and practices for their adherents.

Spirituality, however, is a much broader concept and speaks of the essence of the human person and is understood as that which gives meaning, purpose, and connectedness for each unique individual at any age. A child may express their spirituality within a religious framework and through religious rituals such as in Ming's case, or the child may have a multitude of experiences and thoughts outside of traditional religious practices that give and create meaning. For some individuals, religion and spirituality may be interrelated. In CPC, we must understand these concepts and constructs not only for the child, but also for their siblings, parents, and other persons who provide support in their life. While there are various definitions of spirituality, one commonly accepted consensus definition that has been developed from the National Consensus Project earlier definitions is one that we propose can be used practically in CPC: *"Spirituality is a dynamic and intrinsic aspect of humanity through which persons seek ultimate meaning, purpose and transcendence, and experience relationship to self, family, others, community, society, nature and the significant or sacred. Spirituality is expressed through beliefs, values, traditions and practices*" (Puchalski et al. 2014; National Coalition for Hospice and Palliative Care 2018).

Spirituality entails the child's search for meaning resulting from their experiences and questions such as "Who am I?" "What does my life mean?" and "what is happening to me and my family?" The child and family may not be looking for precise answers to these existential questions, but rather the experience of comfort, hope, and safety to ask the questions. Mystery and paradox are at the heart of spirituality and the search for meaning.

## 12.2 Question 2: How Is a Child's Understanding of the Concept of Spirituality Linked to Child Development?

Amery et al. (2009) suggest a modification of the table of spiritual development in children proposed by Himelstein et al. (2004) that includes an age range, characteristics of those ages, and spiritual development interventions (Table 12.1).

**Table 12.1** Spiritual development in children (reprinted from Amery et al. 2009, p. 276)

| Age range | Characteristics | Spiritual development | Interventions |
|---|---|---|---|
| 0–2 | Has sensory and motor relationship with environment. Has limited language skills. Achieves object permanence. May sense that something is wrong. | Faith reflects trust and hope in others. Need for sense of self-worth and love. | Provide maximal physical comfort, familiar persons and transitional objects (e.g., favourite toys) and consistency. Use simple physical communication. |
| 2–6 | Uses magical and animistic thinking. Is egocentric. Thinking is irreversible. Developing language skills. Engages in symbolic play. | Faith is magical and imaginative. Participation in ritual becomes important. Need for courage. | Minimise separation from parents. Correct perceptions of illness as punishment. Evaluate for sense of grief and assuage if present. Use precise, non-metaphorical language (e.g., dying, dead). |
| 6–12 | Has concrete thoughts. | Faith concerns right and wrong. May accept external interpretations as the truth. Connects ritual with personal identity. | Evaluate children's fears of abandonment. Be truthful. Provide concrete details if requested. Support child's effort to achieve control and mastery. Maintain access to peers. Allow child to participate in decision-making. |
| 12–18 | Generality of thinking. Reality becomes objective. Capable of self-reflection. Body image and self-esteem paramount. | Begins to accept internal interpretations as truth. Evolution of relationship with God or higher power. Searches for meaning, purpose, hope and value of life. | Reinforce child's self-esteem. Allow child to express strong feelings. Allow child privacy. Promote child's independence. Promote access to peers. Be truthful. Allow child to participate. |

## 12.3 Question 3: How Would You Assess Ming According to the Elements of the Consensus Definition of Spirituality?

### 12.3.1 Searching for Meaning and Purpose

Although he was raised in traditional Chinese culture by his grandparents, he was searching for something different. Ming needed to know and understand more, and the Bible, given to him by his father who recognised his son's search for meaning, helped him create meaning within his illness and gave him a goal—the ritual of baptism.

He continued to examine and explore through his discussions and interactions with Sr. Tania and his research on the internet through his tablet. His questioning, even when angry, displayed his ongoing search for the meaning, purpose, and connectedness in his own life. Why? Why? He asked many times. He continually

needed to know why this illness was happening to him. Rather than attempt to answer this question, Sr. Tania provided him the sacred space and time to ask these questions. She remained present with him during his moments of suffering and anger so that the spiritual care provided to him would meet his own unique needs.

### 12.3.2 Connectedness

(a) *To the moment*
Ming did not want to die. He was willing to undergo any treatment and continued to research his illness using his tablet. When he conversed with Sr. Tania, his hospice doctor and other clinical staff, and when he was baptised, he concentrated fully on the present moment and what was happening to him. He was aware of the changes in his health and the myriad of feelings that accompanied these changes and his disease progression. The spiritual care provided to Ming was person-centred—focused on his needs, beliefs and questions, and not those of his health care providers.

(b) *To himself*
Ming's words, his search for meaning and purpose, and his need to belong to the "family" of Jesus revealed his understanding of his own needs. He realised that all he had learned and understood before was not meeting his own personal and unique spiritual needs. Needing to know more as to why he suffered, he frequently challenged Sr. Tania with questions. His emotions on the approaching separation from his parents and his sister Sofia poignantly expressed in his question on why it was happening to him and not to them. His thirst for knowledge and understanding, and the openness of the staff, led him to openly question others and continue his internet research. His internet tablet became an important tool for his sense of independence. While Ming was aware of his impending death, he also accessed spiritual care and assistance in the Padua hospice to hope for a miracle and pray for a miracle. This too became part of his own spiritual journey and spiritual care.

(c) *To others*
During many interactions with his caregivers and staff, Ming revealed the beauty and wonder of his own spiritual journey. Many staff remarked that "the beauty of his nature shines through every moment". Much of the time, Ming was calm, peaceful, smiled often, with a gentle serenity beyond his young years. This was particularly poignant during the ritual of his baptism as he smiled through his shining eyes.

Despite language and cultural challenges, Ming was able to connect to others and to observe, learn and interact from those he met in hospitals in both China and in Italy. His connection with others was also seen in his dream of becoming a teacher and wanting his future students to remember his teaching as he remembered his own teachers. Creating this deep sense of connection and remembering, across time, is a key element in the provision of spiritual care.

His interactions and connection with his parents were strong and mutual. Their understanding of his suffering and the willingness to help him explore and find his own spiritual path, even though it was different from their own, was exceptional. They continued to respect his wishes, allowing him to be baptised into the Catholic faith and after his death to be buried within the rituals of his embraced Catholic spirituality. Ming commented, through his readings of the sufferings of Jesus' mother, that his own mother was suffering as well. He said, "My mother suffers too".

(d) *To nature*
In this case study there was no specific indication of his connection with nature. However, for many children, there is a connectedness to nature that they may feel at home in a particular environment, e.g., the forest, on the farm, in the garden. It may be particular flowers or smells which have meaning for them. If this is the case it is important where possible, to enable children to spend time in these environments, connecting to what is important to them. Where they are unable to go outside, items from nature could be brought to them; they could use visualisation technology to experience nature; videos can be shown or they can be exposed to pet therapy.

(e) *To the significant or sacred*
Where and when did Ming's search for the significant or sacred originate? We do not know but this may have come through his upbringing in Chinese beliefs and traditions and a realisation that he needed and wanted something more. He needed to understand and for that he wanted to connect to something or someone greater than himself. He needed a spirituality where he could ask "Why?" and be angry and still experience an unconditional love. His father's gift of a bible was a key for him to find the sacred in the form of Jesus and connect with him. When Sr. Tania asked him why he wanted baptism, Ming replied "To know the Bible, to know more about Jesus, but the story is not enough. I want to be part of his family, I want to become his son". By creating the respect and space for Ming to explore these beliefs, he expressed the sense of the significant and sacred that was most meaningful for him.

## 12.4 Question 4: How Does Ming's Sense of Spirituality Develop Over Time?

When we examine the story of Ming and use Himelstein et al.'s (2004) stages of spiritual development, we realised that he was spiritually, emotionally, and intellectually mature beyond his chronological years. When staff, particularly Sr. Tania, created the respect and space for him to explore, he was able to internally reflect on his illness, express hope, and find meaning and purpose in his life, while developing a deeper relationship and connectedness with the sacred (God) and with others.

We also saw that those around him, family, health, and spiritual carers, responded effectively to his own needs, by allowing Ming to guide the process. His father identified his searching and his suffering and opened the door to a belief system or faith. His parents allowed him to make autonomous decisions and fully supported

his wishes. His hospice doctor asked him a simple spiritual assessment question "What makes you happy?" (Steinhauser et al. 2006) and the response was practical—ensuring that his son had the support necessary to achieve his desire for baptism, the Catholic Christian faith, and to be part of the family of God.

## 12.5 Question 5: How Does Ming Express His Spirituality and Spiritual Pain or Needs?

His health care providers allowed Ming to develop his unique spirituality and helped him find meaning in his illness and suffering through a framework of religion and faith. He felt safe in his changing environment to be able to express spiritual pain, questioning, and explosions of anger. The religious ritual of baptism was exceptionally important to him that allowed a "gateway" for spiritual peace and a sense of belonging before his death. Ming continued to hope for a miracle and the staff respected this hope as a part of his spiritual care. He was fortunate enough to be admitted to a children's hospice that understood the need for spiritual assessment and spiritual care. His hospice doctor knew the right questions to ask and could be present as Ming expressed his own needs and desires. Developmentally, Ming could use words to express himself and his needs, even when translation was required.

Other children who cannot express themselves verbally, may use different ways to seek to find meaning and purpose and to express this seeking. This could be through play, art, music, dance, poetry, words, or writings. Those caring for non-verbal children need the skills, experience, and understanding to discover the child's hope for meaning, unique to his or her own way of communicating. CPC staff members need the communication and empathetic skills to ask the right questions, create the appropriate space, and allow each individual child's unique response.

Very young children, non-verbal children and those with severe physical or cognitive limitations may not be able to easily express their needs in a way understood by their caregivers. However, emotions, thoughts, and basic spiritual needs and desires can be revealed through facial expressions, body language, or sounds. Those providing holistic, comprehensive care, which must include spiritual assessment and care, need the skills to identify when a child is suffering or seeking, and staff may provide spiritual support through touch, gentle comfort care, and providing a caring presence to ensure the child feels love, care, and respect for their individual needs.

## 12.6 Question 6: For Ming, Religion Was an Important Part of His Spirituality, How Might a Child, with No Religious Input, Express His Spirituality and Spiritual Needs?

Some children and their families may not come from a formal religious background or express the importance of religion in their lives or coping strategies. Nonetheless, if spirituality is understood in a broader sense, assessment is still critical and essential. A spiritual assessment would explore themes of meaning, values, hope, trust, and love for both the child and their parents (Robinson et al. 2006).

Spirituality responds to the meaning of life and gives horizons of meaning. It concerns the most intimate questions, the need for health, the need to be healed, especially when faced with the suffering of the illness. Questions individuals may ask include: why me? why my sister, my family? will live and will I die? what will happen to me when/if I die? God, where are you? will I be remembered? who will heal my mother's suffering? Ming had questions such as these, demonstrating his spiritual distress.

## 12.7 Question 7: Ming Was Born in a High-Income Country and Lived in a Middle-Income Country: How Might a Child's Sense of Spirituality Differ, If at All, in a Low or Middle-Income Country to a High-Income Country?

Integrating screening for spiritual distress, a spiritual history and, when indicated a spiritual assessment is important for children and their parents in both low-and middle-income countries (LMICs) (Nascimento et al. 2016) and middle/high-income countries (Elkins and Cavendish 2004). Children are often influenced in their spirituality and beliefs by their family's beliefs. In many LMICs children and their parents tend to combine both traditional religious values with their indigenous spirituality and values, e.g., the concept of Ubuntu (Marston 2015). Their ways of expressing spiritual needs are similar to children everywhere—through art, play, music, connectedness. Rituals may differ, such as the wearing of amulets may be important, and traditional healing and cleansing rituals. Traditional healers who connect to the spirits of the ancestors may be important role-players in spiritual care and in leading ritual activities.

## 12.8 Conclusion

Children are spiritual beings. Every child consists of a body, mind, and spirit, within a family, society and culture. Every member of the CPC multi-disciplinary team must be trained and comfortable in spiritual screening, spiritual history (Puchalski 2010) and basic spiritual care. Just as we have specialists in other aspect of PC for children, specially trained and qualified paediatric chaplains or spiritual caregivers, working as part of the paediatric multi-disciplinary team, where available, can coordinate spiritual assessment and care. This will help children, and their families, address spiritual pain and respond to the unique needs of every individual as they guide the child's search for meaning, purpose, and connectedness in their lives.

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# Social and Cultural Support

# 13

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**Key Learning Points**

1. Social and cultural support is an essential component of children's palliative care (CPC), without it the holistic needs of children and their families cannot be addressed.
2. Social issues impact the families of a child with a life-limiting condition (LLC) or life-threatening condition (LTC) greatly, and often, particularly in low- and middle-income countries (LMIC), they are hard to address.
3. Cultural sensitivity is important when working with children and families and in our multicultural society this is something that we all need to be aware of.
4. Assessing social and cultural issues in CPC is paramount to the holistic management of children.
5. It is important to recognise and understand different cultures of children and families as culture impacts on the children's health and quality of life (QoL).

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**Case Study**

*Emily is a 4-year-old girl recently diagnosed with Wilms tumour, a cancer of the kidney usually found in children below the age of 5 years. For the past 2 months she has been receiving chemotherapy in Uganda's national specialist hospital. She is a refugee from the Congo and with her brother, sister, mother and father live in a refugee settlement 200 km from the hospital in Kampala. Emily has had to stay alone in hospital for long periods of time as her mother had to take care of the family at home. In hospital a volunteer, Mrs T, visited Emily often and was very caring and supportive to both her and her mother. Emily's oldest brother died 4 months previously during a stabbing incident in the refugee settlement.*

*The PC team met Emily and her mother prior to her discharge. Hospital staff had raised concerns regarding both cultural and social issues which could affect her ongoing care once she returned to the settlement.*

## 13.1 Question 1: Why Is It Important to Consider Culture in the Provision of Children's Palliative Care?

The definition of CPC promotes the provision of holistic care for children, i.e., the care of "*mind, body and spirit"* (WHO 2002)—thus we should strive to provide culturally sensitive care. Different cultures impact on children's health, healthcare practices, patterns of nutrition, family relationships and ways of communicating.

A child is born as a member of a family, community (including tribe) and society, as well as into a culture. Different cultural practices influence a child's health and play an important role in their socialisation and development. Cultural background holds a significant place in children's social and emotional development, although not all people from the same cultural or social background share the same behaviours and views. It is important not to assume that all members of any one culture act and behave in the same way. Never stereotype people!

People tend to perceive common behaviours and key health care issues such as personal space, eye contact, time and punctuality, according to their own cultures, experiences and learning. When doing an assessment of Emily and her family it is important to capture their individual perceptions of touch, communication, civil and religious holidays, rituals, diet, biological and environmental variations, etc. in order to ensure that appropriate plans are developed in accordance with the family's cultural background and needs.

A lack of understanding of CPC in many cultures, especially where there is a developing health care service, may lead to refusal of care which would enable a child, such as Emily, to have a better QoL, e.g., pain and other distressing symptom relief. In certain cultures many parents have a fear of being perceived as "giving up" on active treatment because the child is having pain relief. This often combines with the perception that "suffering makes you stronger" and belief (by both public and healthcare professionals) in the myths around the use of opioids (Annon 2005).

Health practices are often the outcome of beliefs originating from the culture of an individual. In many cultures, it is observed that individuals resort to traditional

healthcare practices before modern practices of care (Yildiz et al. 2018). Where there is a culture of "filial piety" many young parents will be under pressure from the older generation to avoid "modern" or western health care and medicine and consult traditional medical practitioners, often using medicines with no proven benefit (Beinempaka et al. 2014). This is a challenge and, whilst it is important to be respectful to the traditions we can advocate the use of other (proven) modes of treatment and care. Exploring the possibility of using both modes of treatment concurrently or a short "trial" of the "new" way could prove acceptable.

In countries where the medical model persists it can be difficult for the multidisciplinary team (MDT) to flourish and if the doctor has no acceptance of the need for holistic care, families will find it hard to access PC and discuss aims of care and treatment options. Along with this model there are many families who will not trust medical care, particularly where money has to change hands and corruption has been rife (Rosenberg et al. 2019).

Cultural differences between caregivers and care recipients can be challenging. The concept of "cultural humility" (the process of building relationships through discovery of another's culture) (Tervalon and Murray-Garcia 1998) has entered into our vocabulary, care delivery and paediatric training priorities. However, it is reported that 40% of paediatric healthcare providers have suggested that cultural differences are a frequent barrier to adequate paediatric PC (Rosenberg et al. 2019).

There are multiple reasons why clinical scenarios may feel additionally challenging to navigate when cultural differences exist—frequently including potential implicit and explicit biases and corresponding assumptions about a given patient, family or cultural group. In the "medical culture" the language used by healthcare professionals when communicating with each other and their clients can be laced through with implicit bias and judgemental connotations, often unrecognised by the speaker. Stereotyping and assumptions based on ethnicity, gender, sexuality, colour, income, religion, social status and employment all perpetuate stigmatisation and frequently lead to conflict and misunderstanding (Annon 2005).

The importance of including multicultural competence and cultural humility in all clinician education programmes is becoming more widely recognised. Such education aimed at building trust and mutual understanding should include tools, which heighten emotional intelligence, awareness of implicit bias and resolution of conflict.

In a workshop on CPC in the multicultural context (Rosenberg et al. 2019) participants identified ways to provide sensitive and respectful CPC through the practice of:

- Maintaining and expressing compassion even in the face of family hostility.
- Approaching all clinical interactions from a place of humility.
- Developing awareness and management skills of one's own emotions. Here, suggestions included cultivating "safe" spaces for clinicians to express their own feelings and regroup to provide compassionate care to patients and families.
- Emphasising the role of interdisciplinary teams to support individual clinicians, provide key family-support roles and fill specific communication needs.

How information is communicated and received can have a big impact on treatment choices and outcomes. In a study on the impact of race and ethnicity in communicating prognosis of children with cancer it was found that *"Most parents, regardless of racial and ethnic background, want detailed prognostic information about their child's cancer. However, physicians underestimate the information needs of black and Hispanic parents. To meet parents' information needs, physicians should ask about parents' information preferences before prognosis discussions"* (Ilowite et al. 2017, p. 3995).

**Case Study**

*Emily and her family's social history revealed the following issues: (a) the family were displaced from their extended family and now living in a refugee settlement; (b) finances were limited due to their refugee status which meant lack of employment opportunities; (c) Emily's father is unemployed, emotionally distant and did not join his wife and child whilst in hospital. He blames Emily's mother for her condition, citing it was "a problem with her womb" and that she must have done something to upset the ancestors and bring this sickness upon Emily along with the death of their oldest son; (d) Emily's parents questioned whether their daughter would ever get cured and how they would be able to care for her in the refugee settlement.*

## 13.2 Question 2: Why Is It Important to Consider Social Issues in the Provision of Children's Palliative Care?

Chronic disease in childhood is well recognised as a stressor that affects the child's normal development and social relationships within the family and community. It can have implications such as physical dysfunction, developmental disabilities and learning difficulties (Torpy et al. 2010).

Social support allows children to engage in important coping processes such as discussing their feelings, developing enjoyable memories such as having birthday parties and being able to temporarily take a break from the stresses in the home. Friends and relevant others are important sources of social support, being mentioned in all significant aspects of everyday life. The family plays a fundamental role in the management of chronic conditions in childhood. It is through this that individuals learn to live, to love, to feel, to take care of and to care for the other, therefore, it is the primary socialising agent.

## 13.3 Question 3: What Challenges May Be Faced by Families Caring for a Child with a Life-Threatening Condition Such as Emily?

Families caring for a child with chronic and life-threatening illnesses face many struggles. For example:

- The family routine changes through frequent consultations, and hospitalisations, affecting all family members, to varying degrees.

- Not knowing who or what to tell others when faced with a child's life-limiting illness thus the need for support outside of the family is vital.
- Extra caregiving tasks have to be learned and carried out, often for an extended period of time, affecting the accomplishment of tasks and the performance of various roles.
- Families may face additional expenses and an inability to work regularly adversely affecting their financial resources, resulting in financial difficulties and an uncertain future for the family.
- Children with a greater number of impairments also have more functional limitations and these impairments impact on family functioning. As far back as in 1993 Crowe found that mothers of children with multiple disabilities spend significantly more hours on child-related activities and less time on socialising.

It is therefore important that the social support network that the family has should be taken into account (Amery et al. 2009). This network will comprise a group of people with whom Emily and her family have bonds. Among these are the relatives, neighbourhood, work colleagues and the circle of friends (Amery et al. 2009), which for Emily and her family may be limited due to their refugee status. The family also needs emotional support to feel capable and motivated to continue caring for Emily. This support should be constant and appropriate for each situation experienced, since at each stage of the disease, the family has special needs. Therefore, the support must be individualised and unique, as each individual has their own characteristics and ways of coping.

## 13.4 Question 4: What Are the Important Considerations When Taking a Cultural and Social History of Emily and Her Family?

Some of the issues that need to be considered include:

- Communication—potential "language" barriers can exist, such as lengthy eye contact, which can be seen as an expression of anger or disrespect.
- Attitudes and beliefs regarding health care and illness.
- Vulnerabilities, e.g.:
  - The basic needs of the family (food, shelter, etc.).
  - Safety (abuse, neglect and unsafe environment).
  - Access (transport, etc.).
  - Resources (financial, care, etc.).
- Cultural sensitivities.
- Social network and support.
- Employment status.
- Religious background and beliefs.
- Their refugee status.

## 13.5 Question 5: How Might We Assess Emily and Her Family's Social Issues?

Use of guidelines can be helpful and constructive when assessing social issues and should include the family's financial and employment status, home and environment, safety issues, family dynamics, possible abuse, etc. Assessing social issues would include identifying vulnerabilities in the family's functioning but also identifying strengths in the family as well as their support system (Fig. 13.1).

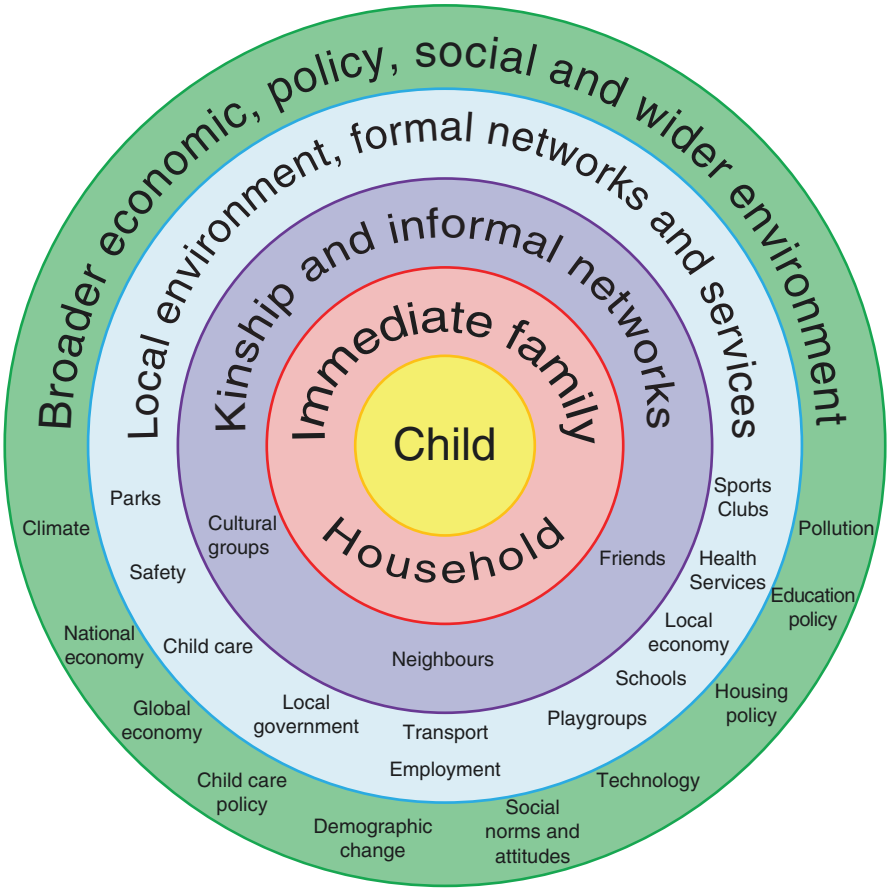


**Fig. 13.1** Bronfenbrenner's ecological model. Diagram by Joel Gibbs based on Bronfenbrenner's 1979 ecological model (Scott et al. 2016, p. 7). Reprinted with permission

## 13.6 Question 6: How Can We Recognise and Understand Different Cultural Beliefs When Providing Palliative Care for Emily and Her Family?

Traditional healthcare practices and cultural beliefs have a significant place and are widely used in all societies. According to current medical knowledge, traditional methods applied to the child may be classified as practices that adversely affect the child's health and practices that have no adverse effect on the child's health (Çınar et al. 2015).

As health care workers we need to have access to good knowledge of different tribes, ethnic, cultural and religious groups. To ask appropriate questions regarding wishes, beliefs, routine and rituals will help to understand the child and family's cultural background and behaviours. Active listening and good observation combined with a non-judgemental and unbiased approach are essential to making an accurate and helpful assessment. This is particularly important in situations such as Emily's where she and her family are refugees and may have been through some difficult experiences, such as their elder son dying in the refugee settlement.

To have information of the child and family's norms and values, religion, language, environment and spiritual thinking will also assist us in how to approach the child and family (Fig. 13.2).

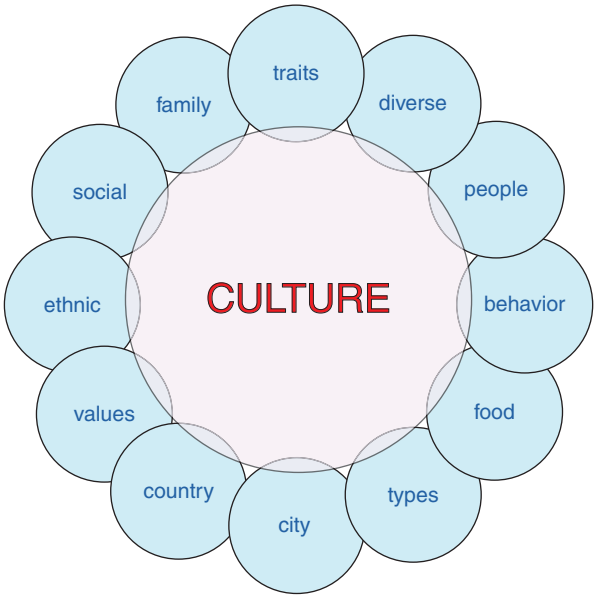


**Fig. 13.2** Concept of culture (123rf.com 2019)

## 13.7 Question 7: How Might the Cultural and Social Background of this Family in Relation to the Assessed Needs, Affect the Ongoing Care of Emily and Her Family?

The health beliefs of the family have been shaped by the cultural reliance upon traditional medicine, affecting their understanding of the cause of their daughter's condition. Children from their culture are not seen as autonomous within the family unit and are not typically involved in discussion or decision making—making it hard for communication which might enable Emily to understand, express her feelings and needs and cope with her situation better.

The family is living 200 km away from the referral hospital and in a refugee settlement. They struggle financially and this, along with distance, could mean that Emily would not be able to access the hospital as needed for clinical care. In refugee settings it often happens that different tribes and cultural groups live in one area. This could cause further tension around and within the family already having to deal with the stress and tension having a child with a life-limiting/threatening condition. There may also be exclusion and discrimination within their community due to Emily's physical condition and fear of the unknown. There is a risk of Emily's condition being attributed to witchcraft or cursing from ancestors, particularly in light of the recent death of their eldest son. Emily and her family may then be perceived as "bad luck" to be associated with.

Alongside being a refugee, her illness also means that Emily is unable to have a "normal" childhood, for example socialising with other children, attending school, etc. Emily's siblings have also experienced separation from their parents for long periods. Even when Emily's parents have been physically available they have been distracted and unable to offer the emotional support needed by their children. Emily's mother has also experienced a lot of pressure, having to travel with Emily and leaving her for long periods at the hospital in order to take care of the other children, whilst not getting a lot of support from Emily's father.

The religious beliefs and accusations of Emily's father have had a very negative impact on the functioning of this family and created tension and conflict, impacting on her care. The family is still grieving the loss of the oldest son, resulting in additional stress and turmoil in the family and impacting Emily emotionally. The family do not feel safe in the refugee settlement as there is a high incidence of violence which led to the violent death of Emily's brother.

## 13.8 Question 8: How Can the Family Be Supported with These Challenges?

It is important to develop a clear management and care plan with Emily and her family. The hospital should refer Emily and her family to the home-based care organisation involved in the settlement for ongoing care and support. Alongside social support it will be important to manager her symptoms as best as possible.

Clear and open communication with the family including Emily, her siblings and both parents is essential. For Emily and her siblings, it is important to explain things

in simple language appropriate to their developmental level and cognitive ability (keep in mind that the social and cultural background and circumstances of the children could have an effect on their development). Listening and observing are very important in order to support this family according to their culture, beliefs and behaviour. Where their behaviour is detrimental to the best interest of Emily, guide in a respectful and sensitive way.

Ongoing counselling and support, based on a social assessment and according to a care plan drawn up with the family, is vital. This family is experiencing a lot of tension due to their environment along with challenges and conflict within the family. Identify the multi-disciplinary team in the refugee settlement and hospital that can support them. This could consist of a religious leader, social worker, caregiver, oncologist, medical doctor and nurse.

Where possible set achievable and context-driven life goals in conjunction with Emily, i.e., addressing what is important to her and supporting her in achieving these goals whilst being mindful of the socio-economic constraints associated with her refugee status. Bereavement counselling and support is needed for all three siblings and both the parents, with regard to what happened to Emily's older brother and what Emily is going through.

Encourage Emily to interact with other children as this may help her with expressing herself and relating to other children, as well as reducing the possibility of stigmatisation. Caring for the siblings is important along with permitting and encouraging them to continue living their lives as normally as possible.

CPC is as concerned with supporting the parents and family as it is about caring for Emily. However, it is essential that the support provided be tailored to the individual needs of her family at any given time. This could be given by a home-based care organisation within the refugee settlement and, where available, include intervention from a social worker. However, in many low-and middle-income countries (LMICs) there is a lack of social workers to support such families.

## 13.9 Conclusion

In order to meet the holistic needs of children and their families within the context of CPC social and culture support needs to be integrated into the care that we provide. Children and their families live within a cultural context, and gaining an understanding of this context will help in the provision of CPC. Likewise social issues will impact the children and families greatly, and even though they can be hard to address it is important that they are recognised and acknowledged.

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# End-of-Life Care

# 14

Nahla Gafer and Poh Heng Chong

**Key Learning Points**

1. Recognising that a child is approaching the end-of-life (EoL) is an essential skill.
2. Principles of good EoL support include meticulous assessment, communication and holistic care.
3. The management of pain and other emergent or anticipated symptoms is critical whilst, at the same time, discussions around hydration, nutrition and review of medication is often needed.
4. As the course of the illness may be sudden or protracted, family caregivers should be actively engaged, to address all questions about EoL and supported as appropriate.
5. The family may need immediate guidance with logistics after the child dies, and follow-up bereavement support.

**Case 1: A Child with Cancer**

*Akifa was carrying her 8-year-old daughter, Yuliana, who had been diagnosed with a neuroblastoma which had metastasised to her jaw and had grown to the size of a 20 cm watermelon, hugely displacing Yuliana's nose and mouth. She appeared to be in a lot of pain. When asked by the nurse whether Yuliana had received any*

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*analgesia, Akifa replied that she had not as they were still waiting to be seen. There had been delays in getting Yuliana seen and started on pain medication.*

*Curative treatment options for Yuliana had been exhausted and she was not expected to live long. Akifa knew that Yuliana had not responded well to treatment and that she was very sick. She had many questions for the doctors and nurses, and eventually asked whether her daughter was dying?*

## 14.1 Question 1: How Do We Know If Yuliana Is at the End-of-Life?

In order to provide good EoL care, you need to know when a child, such as Yuliana, has reached the end of their life and needs EoL care. Having an awareness that a child is dying will help anticipate needs and draw up plans for their care. However recognising the EoL can be difficult.

After rounds of aggressive treatments aimed at controlling a child or young person's cancer, the disease can still prove refractory. Even if response has been consistently favourable, children may unexpectedly succumb to complications of treatments and die. However, prognostication for a child with cancer such as Yuliana, is likely to follow a recognised pattern (Fig. 14.1) with her symptoms getting worse and her condition deteriorating (Meiring and Amery 2009).

## 14.2 Question 2: How Do We Assess and Manage Pain at the End-of-Life?

Pain control at the EoL follows the general principle of pain assessment and management as seen in Chap. 6. Yuliana's pain was assessed using the hand scale, a scale commonly used across Africa (Blum et al. 2014). The hand scale ranges from a clenched hand (which represents "No hurt") to five extended digits (which represents "Hurts worst"), with each extended digit indicating increasing levels of pain. Note: it is important to explain this to the child as a closed fist could be interpreted as worst possible pain in some cultures (Downing et al. 2010) (Fig. 14.2).

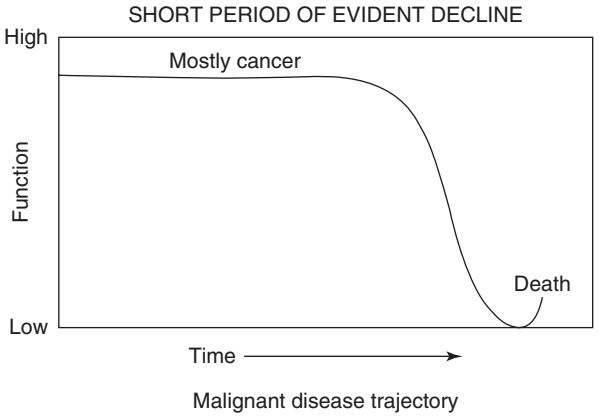


**Fig. 14.1** Malignant disease trajectory (Meiring and Amery 2009, p. 329) (Reprinted from Amery J (ed) Children's Palliative Care in Africa, 329, 2009, with permission from Oxford University Press)

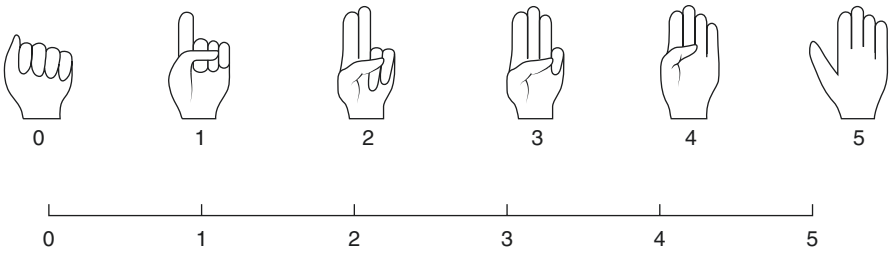


**Fig. 14.2** The hand pain scale (Blum et al. 2014, p. 3)

There are still a lot of myths about managing pain in children including the use of morphine; however, if used correctly morphine is a versatile medication that can be helpful in managing pain in children at the EoL (Meiring 2009), so that they can die as peacefully as possible.

Yuliana's pain was scored as 5 out of 5, using the hand scale. She was started on 5 mg morphine 4 hourly (tablets as no liquid was available), and her pain reassessed after 1 hr, at which stage it had reduced considerably to 3 out of 5. Four hours after the first morphine dose Yuliana was given 10 mg of morphine (increased by 100%). The pain was rescored, and it was 1 out of 5. (NB Usually the increase when pain is not controlled is by one third to one half of the previous dose. However, on this occasion we should have started with a 7.5 mg dose first, but because Yuliana was opioid naïve, we started with 5 mg).

Yuliana was still able to swallow her pain medication—as her condition deteriorates this may no longer be the case, if liquid oral morphine is available then this will help, and can be given buccally if needed (Meiring and Amery 2009). If this is not possible then other options, such as the use of a syringe driver where available, should be explored. A syringe driver will help to ensure steady medication levels over a 24 hr period. Before using a syringe driver it is important that you look at the type of equipment you have, whether you have the correct syringes and needs, and how you should calculate the correct dose. It is also important to remember to continue to prescribe breakthrough medication. Detailed information as to how to manage a child's pain can be found in Chap. 6.

## 14.3 Question 3: Doctor Is My Child Dying?

Behind this question, asked by Akifa, might be other issues. It is important to explore with her what she means by the question and what prompted her to ask it. Clear, sensitive, honest communication and a therapeutic dialogue in which explanation, feedback and further explanation is given is very important in order to explain the situation and how best to manage it (Meiring and Amery 2009). Health professionals can speak of expected death in terms of a range of days, weeks or months, as well as explain the sign and symptoms that accompany the change, and come up with a plan with the family about what to do in those moments.

The aims of our communication with both Akifa and Yuliana at this stage include:

- Ensuring that Akifa understands and accepts that Yuliana is at the EoL.
- Ensuring that both Akifa and Yuliana have the opportunity to say and do what they need to before Yuliana dies.
- Ensuring that plans are in place for all scenarios and that everyone is aware of what to do if and when these scenarios occur (Meiring and Amery 2009).

**Case Study**

*When Yuliana came to the PC clinic, half of her face was covered by a veil and she had multiple complex symptoms including pain, swelling, dehydration and malnutrition, and her mother Akifa used to cover her face so that people would not stare unkindly at her.*

*One of the teams' roles was to manage her different symptoms. Oral morphine was continued for her pain (10 mg 4 hourly), and some dexamethasone (1.5 mg bd) to reduce the swelling. Nutritional advice was given to Akifa because Yuliana could no longer chew food or speak. The work of the PC team extended to reach the refugees offices in order to precipitate the patient's and her mother's travel to another part of the country where some of her family were living. A palliative but non-conventional dose of radiotherapy to the jaw helped to reduce the tumour and therefore the deformity, such that Yuliana was once again able to smile. After her radiotherapy, Yuliana kept repeating the words "istagfaro allah", meaning "I repent to God or may God have mercy on me". This pleased her mother as it was important to Akifa that Yuliana was in a good place spiritually before she dies, however young she might be.*

*Whilst working with the refugee office to enable Akifa and Yuliana to travel to a different part of the country, Yuliana's condition deteriorated. The metastases to her jaw broke through her skin and was oozing and emitting a repugnant smell. The nurses on the ward changed her dressing, and utilised charcoal to try and reduce the smell. Yuliana went downhill quickly and was clearly at the end of her life, and was too ill to travel. She was cared for in the hospital where she died in her mother's arms.*

## 14.4 Question 4: How Do We Provide Holistic Care to Yuliana and Her Mother, Enabling Her to Have the Best Possible Quality of Life?

The availability of a multi-disciplinary team (MDT) of trained staff, who are able to assess Yuliana and her mother's problems: physical, psychological, social and spiritual is very important. Good assessment and management are crucial in all areas, and the following are guides to help:

- Do not panic.
- Immaculate assessment—just as at other times, the key to good management is good assessment.
- Hope for the best, prepare for the worst—think about the different scenarios that could occur and how you would manage these.
- Where possible, offer choices and control to the child and their family. Give them information as to what might happen and the plans that you have for managing different situations.

Fear of the unknown can be greater than fear of the known. The more that Akifa and Yuliana know and understand what is happening the calmer they will be. An aide memoir such as the PEPSI-COLA management plan can be helpful to ensure that all areas are covered (Table 14.1) (Thomas and DoH 2005; Rose and Amery 2009),

**Table 14.1** PEPSI-COLA aide memoir (Rose and Amery 2009, p. 91[a])

| | | |
|---|---|---|
| P | Physical | • Functional problems<br>• Physical symptoms<br>• Medication—regular and PRN<br>• Compliance/stopping non-essentials<br>• Complementary therapies |
| E | Emotional | • Understanding expectations<br>• Fears/security<br>• Relationships |
| P | Personal (for child and family members) | • Family dynamics and relationships<br>• Play<br>• Isolation<br>• Education<br>• Spiritual/religious needs |
| S | Social support | • Nutrition<br>• Financial issues<br>• Housing issues<br>• Care for carers<br>• Practical support |
| I | Information/ communication | Problems that might arise because of some key things not known/ understood by:<br>• The child<br>• Family members<br>• Fellow health workers<br>• Others<br>(for example, factors that might affect drug adherence, place of care or fears about death and dying) |
| C | Control | • Has the child and family had a chance to choose where possible?<br>• Is their dignity protected as much as possible?<br>• Have they been involved in treatment options/management plan?<br>• Have they specified what they want to happen after death?<br>• Place of death |
| O | Out of hours/ emergency | • What will happen if you are not around?<br>• Has cover been planned?<br>• If not, do the family know what to do?<br>• Have they got enough drugs?<br>• Do they have enough information? |
| L | Late | • EoL/terminal management plan<br>• Has non-palliative treatment been stopped?<br>• Are the child and family aware that this is the final stage?<br>• Has spiritual care been addressed?<br>• How often will you review?<br>• Have late stage symptoms been controlled? |
| A | Afterwards | • Bereavement support<br>• Assessment/audit of your performance<br>• Does the team have opportunities for debrief and support? |

[a]Reprinted from Amery J (ed) Children's Palliative Care in Africa, 91, 2009, with permission from Oxford University Press

preparing them for things that might happen, e.g., having noisy secretions ("the death rattle") as Yuliana may be unable to cough or swallow her secretions; how these can be managed, and what it means.

Attending to her symptoms meticulously and maintaining good communication with her and her family is essential. For example, the offensively smelling wounds and the social problems it causes must be addressed, as well as other financial, psychological and spiritual concerns, of her and her family.

## 14.5 Question 5: How Can We Support Yuliana's Spiritually at the End-of-Life?

Spiritual needs are important to address, both in adults and in children, however young they might seem (Foster et al. 2012). Whatever the age of the child, spiritual concerns might be important (See Chap. 12 on Spiritual Care). It is important to learn how to explore such issues and address the spiritual needs, remembering that spirituality and religion are not the same thing, and individuals can be spiritual even if not religious. If not careful, misunderstandings might occur, especially if Yuliana is not talked to, and this might cause distress and might even manifest in physical symptoms. For Yuliana it is important to observe and understand her values and belief system. We can provide support by being physically present, providing quiet time, asking and actively listening and giving her permission to talk about spiritual issues (Meiring and Amery 2009). We can also provide appropriate religious materials, artefacts or music as requested.

**Case 2: A Child with a Chronic Life-Limiting Condition**

*Joy is a 17-year-old girl with Ohtahara Syndrome (early infantile epileptic encephalopathy with burst suppression pattern). The primary condition is associated with general developmental delay and severe epilepsy. It is complicated by scoliosis of the spine and oropharyngeal dysphagia. She has a gastro-jejunostomy tube following previous naso-gastric and gastrostomy tube feeding.*

*Until recently she had been able to attend a special school, which she really enjoyed. However, after frequent admissions (four times within the last 6 months) for high fevers from recurrent chest infections, her parents decided that she should stop activities with large crowds including school, and remain at home.*

*She had required oxygen supplementation and intravenous antibiotics during admissions but not aggressive life sustaining therapy (e.g., invasive ventilation) in the intensive care unit. Frequent physiotherapy with suction was however needed, as she clearly became deconditioned and had poor secretion clearance on her own. She was last discharged stable 2 weeks ago with continuous oxygen at 2 L/min to keep saturations measured on the pulse oximeter above 90%.*

*The hospice home care service have been called to her home in order to review Joy as she is reported by her very concerned mum to have runs of tachycardia at 160/min that were not associated with low saturations and also low saturations of 70–80 s that were not associated with tachycardia. There had not been any fevers,*

*urinary retention, constipation, overt seizures, pain or other signs of distress. Routine feeding has been continued via the continuous feeding pump at usual volumes. Urinary output did not seem different from usual. Since the last discharge from hospital, she has looked tired and did not engage in play as much as before. She also did not tolerate sitting in the wheelchair. Her mum, who is her main caregiver, remarked that she has stopped smiling. Physical examination is unremarkable. Her current medications are:*

1. *Vigabatrin 500 mg twice a day.*
2. *Clobazam 10 mg twice a day.*
3. *Domperidone 10 mg twice a day.*
4. *Omeprazole (mups) 10 mg twice a day.*
5. *Valium suppository 10 mg (as needed for aborting seizures if they last more than 10 min).*

## 14.6 Question 6: What May Be Your Thoughts About Joy's Condition at This Visit?

It is good practice to look for reversible factors that could have resulted in the change of Joy's condition as these may exist despite her chronic life limiting condition (LLC). After all reversible factors have been excluded, progression of primary illness needs to be considered. For young people such as Joy, with chronic LLCs, they can encounter intermittent crises that may be life threatening. The crises often compromise function, mandating external assistance in the form of physical therapies, or dependency on machines or oxygen supplementation. They may or may not return to baseline function afterwards. If the latter happens, the child then survives at a new but lower baseline. To parents or other family caregivers, this has implications on their overall care burden. And worryingly, an acknowledgement that they may be another step closer to "the end". It is an outcome mentioned (and sometimes forgotten) at first diagnosis of the primary condition.

With prognostication universally challenging, especially in a young person such as Joy with a non-cancer diagnosis, the literature has highlighted some ways to help clinicians make better judgements (Brook and Hain 2008; Shaw et al. 2014). With Joy, frequent admissions and increasing technical support are indicators of a poorer outlook. Other markers are her worsening function, including being confined to bed and general lethargy. The "surprise" question that has proven very helpful in the adult setting is equally useful in flagging an at-risk (of dying) child or young person (Burke et al. 2018), i.e., "Would you be surprised if this child or young person has less than 12 months to live?" Joy has a neurodegenerative condition, and early signs of autonomic failure are also a bad prognostic sign.

Recognising that a critically ill child or young person may be close to the dying phase (see Fig. 14.3 for the disease trajectory for non-malignant disease) is vital in good PC. Caregivers may be prepared ahead, minimising panic and regrets. Better support can be organised if emergency care plans are conceived and

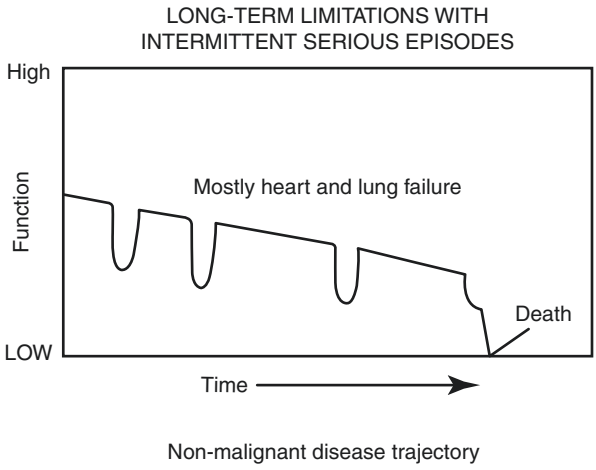


**Fig. 14.3** Non-malignant disease trajectory (simplified and will vary according to the condition) (Meiring and Amery 2009, p. 329) (Reprinted from Amery J (ed) Children's Palliative Care in Africa, 329, 2009, with permission from Oxford University Press)

documented early. This is where children's palliative care (CPC) can make a huge difference. Oftentimes, a family meeting will be organised.

## 14.7 Question 7: Due to Joy's Deteriorating Condition It Is Important to Hold a Family Meeting: How Might This Meeting Be Conducted?

A family meeting to discuss options for EoL care can involve different people depending on the setting. In the hospital, the primary physician/s and nurses, social workers and other allied health specialists are all invited. In Joy's case, where she is being cared for in the home setting, only the PC team and the family are involved. The rest of the healthcare providers will be updated later, although if available the family's general practitioner may also be invited to the meeting. In this instance, since Joy is cognitively impaired with no mental capacity, she does not join the meeting. Other family members such as adult siblings or grandparents may be invited. The healthcare person who has the most rapport with the family can lead the discussion, inviting different colleagues to contribute as relevant. Whilst the same rules of compassionate and respectful communication apply, these family meetings often follow a generic framework:

1. Share updates on the medical condition with a view to "putting everyone on the same page".
2. Solicit family concerns at this time and explore goals of care "if time is short".
3. Set appropriate treatment plans that are congruent with hopes and wishes "whatever happens".

The goal of the meeting is to mutually construct a care plan that is feasible and acceptable to all stakeholders, acknowledging where Joy is on the disease trajectory,

underpinned by love and hope from everyone involved. This process can take a few meetings. Consensus may not be achievable. Emerging differences signpost where more work needs to be done in supporting emotional needs, or addressing a misread of medical facts shared. The fact that families can communicate their ideas, concerns and expectations and give healthcare professionals their recommendations towards appropriate care is in itself remarkable.

In some countries it may be possible to develop an "Advance Care Plan" (ACP) for a young person such as Joy. An Advance Care Plan helps to ensure that an individuals loved ones and doctors know what their health and personal preferences really are, what care and treatment they would like in different situations, and specifically at the end-of-life (Advance Care Planning Australia 2019). Whilst an ACP may not be legally binding, anyone providing care for the individual should take their wishes into account. In Joy's case, she was unable to communicate her wishes, and any care plan would be based on the thoughts and wishes of her parents.

**Case Study**

*A family meeting was convened at home between Joy's parents and the PC team. Her parents were in fact aware that she was doing poorly and feared that she was "close to the end". What has been difficult was frequent admissions. Family routines were disrupted when parents took turns to stay with Joy in hospital (father who works full time in the day took the night shift whilst mom stayed in the ward through the day). The younger children (Joy's siblings who were twelve and eight at the time) were neglected as they had not wanted other relatives to help. The physical toll was bad enough, but what most distressed them was the suffering that they witnessed in hospital as Joy went through rounds of blood draws, chest percussions and suctions. Between themselves, they decided that should Joy deteriorate further or get another bout of chest infection, they wanted care to be provided only at home with no further hospital admissions.*

*A week later, Joy spiked a fever and developed difficulty breathing, associated with thick green secretions. Antibiotics were started together with mucolytics. Her saturations dropped to 85% and she needed supplemental oxygen at 4 L per minute to keep oxygen levels above 90%. Vitals signs that were normal dropped on the third day. Systolic blood pressure became 80 mmHg and her pulse rate was in the 160s. Fever, though persistent, is lower in peaks. Sleep was poor in spite of increased drowsiness, disrupted by frequent coughs and vomiting (mostly half-digested milk).*

## 14.8 Question 8: How Can Joy's Symptoms Be Better Controlled and What Other Medical Aspects Need to Be Managed?

This is most likely another chest infection from recurrent aspiration. With Joy premorbid, already frail and vulnerable, it appears she is succumbing to sepsis. Her parents maintain their original position for maximal management in the home setting, even if this means Joy possibly dying. But they hope that she can be more comfortable.

Joy's main symptom is *breathlessness*, with a rate of around 40 breaths per minute. Even though the respiratory rate is a poor proxy in the assessment of a multidimensional symptom like dyspnoea, it is obvious with her efforts at breathing that Joy is distressed. A low dose opioid such as morphine (half the dose for the treatment of pain in an opioid naïve patient of her age and size) administered round the clock is most appropriate (Craig et al. 2015; Pieper et al. 2018). The route can be either enteral, buccal or parenteral and provision for breakthrough doses should also be made. Simple actions such as sitting Joy up in bed, having a fan lightly blowing in her face, and facilitating a cool and calm environment are examples of helpful non-pharmacological interventions. Careful explanation to her parents fosters compliance and a sense of empowerment. Starting doses may need to be titrated with regular reviews. Her cough and vomiting (most likely post-tussive) should improve with opioid administration. At this point, it is helpful to start a discussion with parents *to stop feeding*.

Hydration and nutrition can be an emotive topic (Tsai 2011; Zaal-Schuller et al. 2016), particularly at the EoL. There must be clarity why fluids or milk ought to be stopped or cut back. Appreciating the meaning of feeding from the caregiver's perspective is critical. In Joy's case, reducing fluids can lead to better symptom control (breathlessness, cough and vomiting). If caregivers are not ready, reducing amounts fed as part of a therapeutic trial is an alternative.

A review of medication is also due. What is essential normally can become unnecessary or even detrimental. Some medication may need to be stopped gradually. The clinician must also be prepared to deal with emerging issues, such as breakthrough seizures after stopping anti-epileptic drugs. When a child or young person such as Joy is on enteral feeds routinely, reconciliation of medications may become secondary, as it does not pose any burden to the dying child who loses the ability to swallow. In Joy's case therefore nothing was changed. However, when a child or young person is dying, drugs may need to be ordered in *anticipation of symptoms common in the dying*. Other than breathlessness and vomiting that are already present in Joy, these may include, pain, agitation, fever or secretions (sometimes called the death rattle). Appropriate medication (available in the formulary of each setting) can be provided in case they surface out of hours or after a dying child or young person leaves the hospital.

**Case Study**

*Joy became much more comfortable after symptoms were controlled. She continued to live on, despite it being predicted that she would die in hours or a few days. Her extremities turned purplish and pressures sores started to develop in some places. She remained comatose and unresponsive. It has been a week since the last review when it was thought that she would die imminently. The tired parents are starting to wonder if they have done the right thing to keep Joy at home and they have asked to speak to the team about this.*

## 14.9 Question 9: Why Has This Happened and How Can We Address the Parents' Doubts at This Time?

It is not uncommon for clinicians to be wrong in their prognostication. There are many factors and no definite answers can be given in the majority of cases. Perhaps, it may be that parents do not really seek a reply but rather want healthcare providers just to listen. This is the time to manage *psycho-emotional issues* and sometimes the spiritual impact of loss.

It can start with a simple open question such as "*how are you making sense of what is happening right now*" and see what is said before proceeding. Often, it may be about making sense of a very difficult situation and meaning making in anticipatory loss. Other times, it may be about seeking validation that they have "left no stones unturned", or they have done everything that all good parents will do.

The multi-disciplinary team that PC is provided through can offer suitably trained professionals such as social workers or chaplains to provide *spiritual support* to caregivers at this time. Clinical workers may need support themselves, where suffering seems most unjust or simply overwhelming.

**Case Study**

*The nurses on the team organised for respite at home with the help of trained volunteers so that the parents could replenish themselves with a break from caregiving. The physician helped reframe the misjudged timing of death as a blessing for the family to be together for longer. The pastor of the family was called to lead prayers. The social worker brought out paints and canvas boards for the family to do hand printing as part of memory making. Joy died the following day.*

## 14.10 Question 10: What Else May Require Attention at This Time?

Depending on local policy, a *death certificate* will need to be issued. Prior arrangements should be made about this such as who will do it and when. After that, the *funeral* director will need to be called to do the rest. Sometimes, parents want to spend more time with their child after death and they may need to be assured that this is alright and normal.

The PC team can decide if they will participate in the proceedings. This can be supportive to family caregivers and often achieves closure for the individual healthcare provider. Plans for *bereavement support* vary between different services, and this can take the form of bereavement cards, phone calls or visitations sometime later at home. If other healthcare providers have not been involved, they may need to be informed soonest. More on this can be found in the following Chaps. 15 and 16.

## 14.11 Conclusion

Meeting children and families' needs with meticulous symptom assessment, an holistic approach (incorporating physical, social, psychological and spiritual dimensions), and good communication are essential for patients with life-limiting diseases, especially when they are facing the end-of-life. The examples set above show how these three principles are applied in two different settings: one of a child with cancer from a LMIC dying in hospital and the second of a child dying from a chronic LLC in the home setting in a high-income country.

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# Supporting the Family at the Time of Death

15

Maiara Rodrigues dos Santos, Zipporah Ali, and Regina Szylit

**Key Learning Points**

1. The sick child's needs and concerns should be at the forefront of both the health care provider and the child's family.
2. When a child is seriously ill, each person in the family is affected differently, hence their specific concerns and needs must be approached individually.
3. It is important that the child's family should get the support and care they need during this difficult time.
4. The healthcare team should listen to family preferences and work with them and their child to plan care of the child (including symptom management) throughout the illness. This will include care for the child's current needs and flexibility for future changes.
5. Discussing how providers can be affected by the end-of-life (EoL) care of a child is a keystone to address grief and bereavement and to prevent moral distress and burnout whilst caring for families.

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**Case Study**

*James is a 13-year-old teenager with a refractory Ewing's sarcoma treated with high-dose chemotherapy. He lives with his 52-year-old mother who is a housewife and his 53-year-old father who is a journalist, in a temporary house near the referral centre for cancer treatment. Since James was diagnosed 2 years ago, the father left his full-time job to stay closer to his son and wife. James's extended family is from a city which has no access to cancer treatment and is 700 miles from the hospital where he is admitted. He has three older sisters and is very close to his uncles and cousins. Despite being away from home, James and his parents feel grateful for the "miracle of God" for being able to get treatment at the cancer centre, considering that there are no medical specialists in their home town. Five months ago, James presented with metastasis in both his skull (bone) and his central nervous system which required palliative chemotherapy that was offered in the outpatient clinic. Since James was diagnosed, his family has been able to return to their hometown only once, as he has needed to be in hospital for prolonged periods during that time. The family expressed that they miss being around the rest of the family and friends. James has many friends who keep in touch with him through social networks.*

*The family had hoped to be able to return home. However, about 2 months ago James was readmitted to the oncology ward with intense pain and rapid progression of paraplegia. The doctors informed the family that James would probably not go back home due to reduced mobility and worsening symptoms. The team believed that due to the worsening general medical condition, James would die at the hospital. Although the doctors initiated conversations with the family on changing the goals of care from curative to EoL care, they sensed that his mother did not want to discuss this. She said the family knew that it was a difficult situation, but they would continue to have hope and belief up to the end that their son would get better.*

## 15.1 Question 1: What Are Some of the Challenges in Transitioning to End-of-Life Care for James and His Family?

The diagnosis of a LTC in a child or adolescent is devastating for all involved: parents, family members, healthcare providers, and the children themselves, as they grieve for the life they had planned and believed they would have. The child, their parents (and sometimes siblings), and clinicians will soon begin the difficult process of talking about death and the options for the child's EoL care.

Koenig and Davies (2003) state that EoL care and PC for children and their families encompass key domains of life that are inevitably shaped by the cultural context and that understandings of the boundary between life and death and the rituals that give meaning to this key transition vary widely. Thus, negotiating the difficult transitions on the path to a child's death can be a daunting challenge and even more difficult. Domains of EoL care are shaped by culture, including the meaning ascribed to illness, the actual language used to discuss sickness and death (including whether

death may be openly acknowledged), the symbolic value placed on a child's life (and death), the lived experience of pain and suffering, the appropriate expression of pain, the styles and background assumptions about family decision-making, the correct role for a healer to assume, the care of the body after death, and appropriate expressions of grief (Koenig and Davies 2003). Challenges arise when the child's family and health care providers do not share fundamental assumptions and goals; in this case, although the health care providers initiated open communication about changing goals of care to EoL care, the foremost goal of culturally appropriate care remained a barrier. Social class and religious background may further have complicated the understanding of transiting to EoL care (Koenig and Davies 2003).

The global unspoken background assumption (across all income levels) is that death in childhood is "unnatural" and unthinkable that morally it should or ought to be controllable (Koenig and Davies 2003). Thus, parents and other family members, and at times, health care professionals see the death of a child as an unspeakable tragedy, almost an obscenity, and may devote unlimited resources to preventing it. This is a core challenge to the transitioning to EoL care since it can lead to asynchronies during the care provided. For families this is a path full of uncertainties, in which they fluctuate between hope and hopelessness as they have their lives changed by the multiple losses along the illness trajectory (Misko et al. 2015).

## 15.2 Question 2: Why Is Good Communication So Important When Caring for James and His Family at This Time?

A significant consequence of this cultural stance is that the death of infants and children in the health care system of countries such as the United States (US) takes place only after all aggressive efforts to prevent death have failed. Often death follows an explicit negotiation about the exact moment, location, and mode of dying. Disparities in families' EoL care worldwide can be noted when we consider the place of children's death. Most deaths occur in inpatient hospital settings, although some countries have higher rates for death at home or in a hospice, which influences how care is delivered during the child's EoL. In many countries, including for years in the US, there are not many hospices for children, thus creating important barriers to care for children from diverse ethnocultural backgrounds (Field and Behrman 2003). As the number of children's hospices increases (Hinson and Rosoff 2016), perspectives must be transformed to include the hospice philosophy into the cultural context worldwide. This makes good communication—the core element of decision-making—a high priority. In an increasingly diverse society, the lack of excellent communication about the realities of EoL care, which is difficult under the best of circumstances, becomes a serious obstacle to care.

Communication is one of the six general principles to consider when delivering PC to children and families. Other important issues include: best interest decision-making, diversity and cultural issues, consent, confidentiality and capacity, continuity and co-ordination of care, and advance care planning (Chambers 2018). There is still a tendency worldwide for families and some healthcare providers to consider

PC as an option after other options have failed, which can lead to low acceptance of families for PC support. Thus, it is important to provide space for honest conversations and make decisions and plans throughout the illness trajectory, introducing the PC approach in a timely way (Chambers 2018). However, for many the reality is that they are not introduced to PC until the EoL.

**Case Study**

*During James's stay on the ward, the ward team realised when discussions on EoL care were initiated his mother would always refer them to her husband. His father, on the other hand, was reserved, and hardly expressed his feelings, or shared his opinions. He trusted his wife with the responsibility to make decisions on their son's care. He understood that James was expecting another "miracle". He did not oppose the treatments proposed by the medical team but said he would support his wife on her decisions.*

*James had developed a very close relationship with everyone on the team. He was very charismatic and affectionate; he would invite some of the team members to play video games or watch movies with him on the ward, thus establishing trust and bonding between him and the team, which was a positive outcome. Although the team realised that James understood the outcome of the treatment very differently from his mother and the father, he supported his mother's decisions on his treatment because he did not want to upset her. The staff noticed that in several different situations James changed his opinion about his treatment after hearing his mother's decision. For example on one occasion, due to paraplegia, the nurses performed intermittent catheterisation every 4 hrs, as they had had to remove his indwelling urethral catheter due to the risk of infection. James reported to the nurse that he felt uncomfortable with intermitted catheterisations that were mostly carried out by female nurses, he also complained that he often had to ask his many visitors to leave the room whenever a procedure needed to be done. James also told the doctor that he felt he had lost his dignity at the hospital. Despite the risk of an infection with the indwelling urethral catheter, the doctor and the nurse agreed that this would be an important measure of comfort to James. This decision gave James a sense of relief; however, his mother expressed serious concerns about the exposure to possible infections; the need to change antibiotics and that all this would make his condition worse. On the next day, the doctor was surprised that James had changed his mind and had decided to continue with the procedure every 4 hrs. The doctor was convinced that his mother influenced his decision. The staff were very disappointed by the fact that James had made a decision not to prioritise his own comfort.*

## 15.3 Question 3: What Are Some of the Issues with Regards to Decision-Making in James's Care?

Most children and adolescents with a LTC rely on their parents or other surrogate carers to make decisions for them, since there are many considerations about appropriate decisional capacity. However, this does not mean that children and

adolescents should not have a say in their future care or have their choices acknowledged and recorded. Health care professionals should aim to encourage early and thorough communication between parents, the child/adolescent, and clinicians. However, in many countries, particularly in low-and middle-income countries (LMICs), this is a challenge as most decisions are made for the child or adolescent by their parents/guardians with no further explorations and discussion about autonomy or what is in the child's best interest.

Engaging children and adolescents in such conversations and decisions requires a developmentally appropriate approach for obtaining the assent of the child. Whilst the "informed consent" for diagnosis and treatment is their parents' or other surrogates' decision, since autonomy in paediatric patients is not legally accepted (Committee on Bioethics 1995) in most countries, it is still important to get the child's assent whenever possible.

In the decision-making process, some tensions can be expected as parents and clinicians try to act in the best interest of the child, and conflicts may arise related to differences of family and providers' wishes (Committee on Bioethics 2016). The model of constrained parental autonomy states that parents must balance the best interest of their child, based on their own understanding of family's best interest, meeting the child's basic and medical needs (Ross 1998). Advance care planning (ACP) is a process to engage with children and families to set goals and plans, respecting medical treatment (IOM 2015). Adolescents should be involved in advanced care planning, since they have shown capability in EoL planning similar to their adult counterparts.

Aarthun et al. (2018) observed that parents' competence and perceived influence and control over their child's health care appeared to affect how they mastered their role of involvement in decision-making. Individually tailored and respectful facilitation of parental involvement in these decisions by health professionals seemed to improve parents' influence, control, and ability to cope with the parental role. Hence, health care providers should strengthen parents' sense of coherence enhancing the quality of health care.

It is important to remember that children who die and their families constitute a very diverse group, not only culturally but also due to different life-limiting conditions (LLCs) and illness trajectories that require multiple resources. Some children will die suddenly whilst others will need care for weeks or even years (Field and Behrman 2003). It is important to outline for the families the options that they have for EoL care in a culturally sensitive manner, considering the right time for each family to provide space for conversations regarding the place of death and other aspects of advanced care planning (Hinson and Rosoff 2016). In James's case, dying at home was not an option due to the lack of resources in his home community. However this should not prevent health care providers from providing good quality EoL care within the hospital setting, taking into account James and his family's priorities. Health providers must engage in conversations in order to ensure that families have access to appropriate resources, where available, although being aware that not all the families will be able to identify their needs, thus the health providers will need to help assess the family's priorities during their child's EoL.

## 15.4 Question 4: What Existing Standard Global Guidelines Could Help James's Family and the Health Care Providers Cope with the Difficult Situation?

A global approach to EoL care is needed in order to produce worldwide standards, including a shared vision and united policies for education, research, and practice, along with other priorities to ensure the provision of high quality EoL care (Hinds and Lafond 2016). Currently there is a challenge due to the lack of international guidelines or laws to ensure high quality of care to all children and their families (Schencker and Arnols 2015). Recent work by Stein et al. (2019) has begun to develop guidelines to communicating some of the challenging issues to children and their families, with guidelines being developed initially for the United Kingdom (UK), but being adapted for use in LMICs (ICPCN 2019).

It is worth noting that whilst standard global guidelines are being tailored and enhanced, patient- and family-centred care is an approach and a philosophy for the planning, delivery, and evaluation of health care that is grounded in a mutually beneficial partnership among patients, families, and health care professionals (IPFCC 2017). Providing patient- and family-centred care to children at the EoL presents many opportunities and challenges.

## 15.5 Question 5: Why Is It So Important at the End-of-Life that the Healthcare Team Have Built Up a Good Relationship with James's Family?

Best practice in EoL care for children and their families includes sharing values of equity in the relationship between parents and providers, as well as practising family-centred care with integrity and commitment to authentic engagement to address the individual needs of children and their families throughout the process (Davies et al. 2017). In James's case, it is fundamental to establish trusting and collaborative relationships to enable interactions where providers and families can express their views. If the child and family have been referred for PC only at the EoL or if the family have developed no confidence in the team throughout their child's treatment, difficult conversations can be hard and ineffective (Santos et al. 2019).

When significant bonds are established with health care providers, parents can feel safe to ask questions and share their deepest concerns. Parents who do not get this support during their child's treatment can develop feelings related to abandonment and negligence due to their lack of a trusting relationship with the care providers. Parents also need to receive adequate information, guidance, and support from healthcare providers to make difficult decisions during the whole of their child's care and treatment and not just at the EoL (Meyer et al. 2006; Price et al. 2011; Aschenbrenner et al. 2012; Santos et al. 2019).

## 15.6 Question 6: How Does Cultural Complexity Impact on the Care of Children Such as James at the End-of-Life?

Terminal illness and the death of a child is always complex and tragic; there is no magic bullet on how to communicate this to a child's parent/family. Campbell and Amin (2013) explored the dilemmas children's palliative care (CPC) providers' experience in sharing bad news with families in rural KwaZulu-Natal, South Africa. Their findings suggest that PC providers experienced four dilemmas with regard to telling bad news: (a) families did not want to be told any bad news; (b) PC providers felt uncomfortable about telling bad news; (c) PC providers and patients shared similar values about telling bad news; and (d) providers were unsure about when to share bad news (Campbell and Amin 2013). They concluded that in the rural areas where their study was conducted, children are not usually given bad news about their illness. Disclosing poor prognosis led to the dilemmas faced by caregivers. The result was that the emotionally charged work of caring for children reaching the end of their lives became more challenging for the caregivers because they were not prepared for the different cultural complexities. It has been suggested that this is a common scenario in many countries, especially in LMICs.

Children may be reluctant to hear bad news because the telling of bad news is not a traditional practice in their culture. There is always hope for a miracle. According to Campbell and Amin (2013) in the presence of a belief system that promises delivery from impending death, it may appear inappropriate to inform the child or their family that the child will die. Sharing bad news may nullify the hope offered by, for example, traditional healers, or miracle/religious healers all of whom do not tell a patient that they will die, as a miracle (such as a cure) is presented as a possibility (Campbell and Amin 2013). PC providers face a choice between their loyalty to their culture (which may or may not also be the culture of their patients) and their role as a PC worker. In this regard, the timing to consider changing the goals for care will differ for each family and may differ between families and healthcare providers. Whilst the team recognises the process of EoL care, families may still be concentrating all their efforts to keep their child alive. In some cultures, talking about death does not happen as it is important to avoid death and dying as a possible outcome and PC teams may struggle to be introduced to the child and their family sooner in order to provide comprehensive care to families.

Allowing the exchange of information is fundamental, as for many parents this is how they can validate that, despite all possible and existing efforts, death was inevitable. Bereaved parents have identified the importance of timing in breaking bad news (Santos et al. 2019). Information needs to be shared in line with families' expectations and time. Parents often feel distressed with the conversations with providers and the information provided is sometimes inadequate or conflicting, and they may have difficulty understanding it (Contro et al. 2004; Aschenbrenner et al. 2012). Conversations with healthcare providers, especially the delivering of bad news, are remembered by parents throughout their lives (Contro et al. 2004; Boles 2015).

Literacy is the state or quality of someone being literate. This concept is broadened in health context, with the term "health literacy", which is defined as "*the degree to which individuals have the capacity to obtain, process, and understand basic health information and services needed to make appropriate health decisions*" (Ratzan and Parker 2000, p. vi). Besides the need to tailor the communication to each person according to their capacity for understanding, there is also the need to understand the moment and limit of each one, regarding the context of distress and vulnerability related to the loss of a child in the family.

## 15.7 Question 7: Why Is It So Important for James that There Is Good Communication with His Family at the End-of-Life?

Family communication at the EoL is important for terminally ill children/adolescents, individuals, family members, and healthcare/PC specialists. When a person experiences the death and loss of loved ones they also face the reality of their own death. Providing space to talk about uncertainties and anguish surrounding illness, death, and dying can help family members, children, and adolescent recognise their needs. For such conversations, providers need to develop sensitive connections with each family member in order, for example, to identify the timing to break bad news. Sensitive connections enable these conversations to be more effective for setting goals for care and aligning expectations at the EoL.

**Case Study**

*James's condition progressed very quickly: his fatigue increased and he had episodes of intense dizziness and hypotension. The staff could see that his condition was worsening rapidly and he was not the "happy boy" that he had been. It was common to hear some nurses say "I hope he does not die on my shift" and at the same time the nurses hoped that when they came back the next day they would find James better.*

*One day, James said that he would like to go out of the room for a little while in his wheelchair. The doctor agreed with this request; however, the head nurse was against it due to the fact that he had severe dizziness and hypotension. His mother was in agreement with the nurse; however, she could see her son's frustration and how he desperately wanted to get out of his bed. The doctor put James in the chair and took him briefly around the ward, and despite his exhaustion James thanked all the professionals he met on the ward. When he was taken back to his bed, he said he felt he needed to say goodbye to everyone. Both his parents asked him to stop saying such things.*

*A few days later, James started to get sleepier and more confused. James's parents requested the medical team to reduce the dose of his medications because they felt that he was sleeping too much and his siblings were coming to visit him. The team agreed to reduce the dosage, but explained that if his symptoms began to intensify it would be necessary to increase the doses again. James's siblings arrived at*

*the hospital and everyone had the chance to talk with him. Hours later, James's symptoms worsened and his medication doses were again adjusted to reduce his distress. He died a short while later in the hospital, with his symptoms controlled and with all his family in the room.*

*The day after his death, the staff expressed that they would miss James. He had become part of their "hospital family". Through his social networks they discovered that he had left several videos for each of the family members and for the hospital staff. He had done this with the help of a friend during his stay at the hospital, but no one knew about these messages. After two weeks his father and mother returned to the hospital to thank the staff for the care that they had all received and for enabling James to have a "good death".*

## 15.8 Question 8: What Constitutes a "Good Death" for a Child Such as James?

Often a *"good death is characterised as one that takes place at home, surrounded by family and/or friends, with pain and symptoms under control, spiritual needs identified and met, and following appropriate goodbyes"* (Koenig and Davies 2003, p. 532). As part of the PC philosophy, the fact that James had the opportunity to say goodbye to his family and the staff, had his pain and symptoms controlled during his last days, and had his family by his side can represent a good pathway for EoL care in many healthcare settings, as long as it provides accommodation for the real demands of the family. However, it is important to note that there is no picture for the "good death", as each family has its own unique meanings, expectations, and beliefs.

Another important aspect to notice from the case is how healthcare professionals provide opportunities to children and families to share appropriate goodbyes. The reluctance to talk about death and dying can prevent sharing meaningful moments, such as the farewell James performed with the staff during his tour around the unit. His age and personality allowed him to come up with this moment, but for many children, adolescents, and family members the lack of such opportunities can result in anxieties, which could serve as a barrier to a "good death".

## 15.9 Question 9: How Might Cultural Variations at the End-of-Life Impact on Providers' Distress When Caring for Families of Children, Such as James, at the End-of-Life?

Healthcare providers can experience suffering when caring for children at the EoL and their families. Some delicate situations during the care of children and their families can result in moral distress for healthcare providers. Moral distress has been defined as negative reactions that emerge when the practitioner cannot act on a morally correct response to a situation (Jameton 1984).

After James shared with the staff that he felt his dignity was threatened by the frequent intermittent bladder catheterisation, the nursing staff felt uncomfortable to perform the procedure every 4 hrs, since his mother did not agree to change the catheter due to the risk of infection. For the health care providers, this conduct might not be the best for James, challenging the practitioner's moral judgement, which can impact negatively on interpersonal relationships with his family. If such reactions are not explored with care providers, many situations go unnoticed and the consequences can result in intense suffering and stress reactions.

Caring for dying patients increases the chances for providers to experience the effects of stress, burnout, grief, and bereavement (Papadatou 2000). Grieving reactions can vary according to the nature of loss. In James's case, a close relationship developed with him along the journey resulted in grief manifestations both before and after his death, resulting in some wanting to avoid caring for him at the time of his death, whilst at the same time fearing not seeing him in the unit on their next shift.

## 15.10 Conclusion

Supporting the family at the time of death requires multiple skills and different dimensions for the care provided to children and families. Introducing EoL care demands an approach in a timely way throughout the illness trajectory, not only when the child is dying. Considering the complexity and diversity of each family's demands, different encounters and relationships must be provided and carefully implicated in caring practices. Families are immersed in a time of deep distress, in which any and all relationships are subject to a huge range of meanings, remaining in families' memories and transforming itself throughout the bereavement process. An understanding of the theoretical components of cultural competence is imperative to provide the best care to children and families in EoL. Changes in health care education are needed to address the growing evidence that provider behaviours and attitudes can affect the health of different cultures.

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# Anticipatory Grief and Bereavement Support

16

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**Key Learning Points**

1. Anticipatory grief and bereavement support for children demands honesty, reassurance and provision of a space for safe expression of emotions.
2. Anticipatory grief is triggered by disclosure of a diagnosis of a life-threatening condition (LTC) and prepares the child and family for dying and death.
3. Bereavement includes a wide range of emotions, experiences, thoughts, physical symptoms, social and spiritual changes over a long period of time, at differing times and with varying intensities and combinations.
4. Bereaved children and young people need to be listened to with empathy and given permission and facilitation to grieve in their own unique way, assurance that there is no right or wrong way to grieve, and given time and support for them to express their feelings.
5. Grief reactions in children and young people will change over time and maturity and vary according to gender, culture, age, previous losses and experiences, family support, the relationship to the deceased and the nature of the death.

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**Case Study**

*Amos is a 15-year-old boy diagnosed with acute lymphocytic leukaemia. He lives with his parents Sybil and John, and his younger sister Pretty (10). His older brother Elijah died in hospital from the same condition 5 years ago, a distressing and frightening time for all the family.*

*The PC team meets Amos, who is suffering a relapse of his condition having received treatment, and his distressed family at their simple home. They ask "what do you think is happening with Amos?" Sybil describes similar signs of deterioration that Elijah manifested before he died, but refuses to acknowledge that Amos will die, preferring to only talk of hope and future plans. She admits to spoiling him with the few luxuries she can afford. She is angry with John who drinks heavily and spends hours out of the house, despite being unemployed. No one seems to know where Pretty is until one of the team finds her hiding with her doll in a tree in the garden.*

*When Amos is asked how he is feeling he stays quiet. His father asks "Are you angry? Are you angry with us? With me?" He refuses to say anything. After a while he looks anxiously at both his parents before asking "Is what happened to Elijah going to happen to me?" Sybil angrily accuses the team of upsetting her son but calms down when she notices that Amos appears eager to talk. Pretty comes quietly into the room accompanied by one of the team, and John asks her to play outside. The team gently asks John's permission to invite her to join them. They answer Amos' questions until he fully understands that his illness will not improve and that he will receive all necessary care until he dies, emphasising how Elijah's care was different. One of the team asks "what do I need to know about you right now so that I can best help you?" Amos cries and responds that he wants his bicycle with him when he dies, the same one that Elijah used to ride, so that the brothers can still enjoy it together.*

*Over the next few months the team assists Sybil, John and Pretty to help Elijah find a way to achieve his dream. Pretty offers to draw a picture of the bicycle that Amos can take with him when he meets Elijah, and Amos agrees this is perhaps the answer. John promises Amos that he will continue to maintain the bicycle in perfect order after he has died and Amos gives permission for Pretty to "inherit" his prized possession. Amos dies peacefully at home some months later surrounded by all his family and his bicycle.*

## 16.1 Question 1: Amos's Family Are Clearly Grieving: Is There One Way of Thinking About Grief and Bereavement?

There is no one correct way to think about dying, death and bereavement. There is a general lack of evidence regarding which type of intervention is the most effective in bereavement care, but a recently published Norwegian study among cancer bereaved parents identified some factors associated with a resilience process:

perception of self, social resources, family cohesion and importance of relations within a "system" (family, village) (Vegsund et al. 2019).

It can be helpful to adopt a non-evaluative stance and remain curious about the child patient's, siblings' and parents' meanings and stories about life and death. While the impact of a child's death is known to be associated with distress, anxiety, depression and sometimes post-traumatic stress disorder (Wikman et al. 2018; Ljungman et al. 2015, 2016), much of managing anticipatory grief and the grief after the death of a child is not necessarily the domain of professionals. By helping families to better listen to, interact and communicate with children, we can support them in not avoiding the child, but remaining close and present, bearing the pain, fear and sadness that comes with this devastating news (Bluebond-Langner 1978). When the child has received formal care, ongoing relationships between providers and bereaved families appear significant (Snaman et al. 2016).

## 16.2 Question 2: What Do We Mean by Anticipatory Grief and Bereavement?

Both anticipatory grief and bereavement of children demand honesty, reassurance and safe expression of emotions. Death shatters a child's illusion that the world is safe and predictable, where bad things happen only to bad people, where if you are good there is reward, and that a God can make things better. After disclosure of a child's life-threatening illness by a clinician, one of the hardest things a parent will ever need to do is continue this discussion with their unwell child and other children in the family. This disclosure and conversation trigger *anticipatory grief*, an important and natural process that occurs during the rest of the child's life, and which prepares him and the family for dying and death. After death, the experiences of grief and the process of bereavement heal those that mourn the deceased child. *Bereavement* is a process whereby the bereaved find ways to take the deceased with them into their futures "not moving on without him, but a moving forward with him" (McInerny 2019; Klass and Walter 2001) to ensure he is never forgotten and that future children and grandchildren will "know" him. Bereavement moreover helps in coming to terms with the reality of a painful world, and is a valuable survival skill.

## 16.3 Question 3: How Do We Manage Disclosure to Amos that He Is Dying?

How to manage disclosure to a child such as Amos of his life-threatening diagnosis is often informed by theories of developmental psychology (Fredman 1997; McKissock 1998). Young children are often thought to be lacking in knowledge in

this area, and most adults will go to great lengths to prevent and protect children and young people of all ages suffering either physical or emotional distress. This often results in parents not wanting, or not knowing how to share the information. Although child developmental understandings of death are useful guides, they may ignore culture, religion, race, class and gender and may make assumptions about what a child or young person may be experiencing.

Literature on dying and death generally encourages an honest explanation about the diagnosis and for family members to talk about the impending death rather than indulge in "mutual pretence", a co-created structure designed to protect but that denies the dying process, "as if" Amos will grow into an adult. This is often associated with a poor dying outcome and complicated bereavement (Kreicsberg et al. 2004; Jalmsell et al. 2015). At the same time most adults find it very difficult to talk openly about death, hence a contradiction of "must talk but cannot talk" (Fredman 1997). Sybil exemplifies the parent that fears that talk about death kills hope, that it is dangerous as it might welcome or invite death and that it might go away if we ignore it. While health professionals often describe patients and family members who are struggling to talk as "in denial" after diagnosis, we suggest this is rather more a persisting for the time being with a version of the information that they can currently manage, rather than resisting the "truth" (Fredman 1997). Children and young people may prefer to talk with friends rather than professionals at times. Failure to converse comfortably about death does not always reflect an inability to do so. Amos' parents may hold a strong belief that it is not good, right, or appropriate at that time, and possibly hold strong views that Pretty is "too young" to hear the conversation. This needs to be respected while sensitively working towards a full and honest disclosure where emotions can be safely expressed.

## 16.4 Question 4: How Can We Talk to Amos About What Is Happening to Him?

Joining the child or young person's language will enable them to describe what they are experiencing, rather than have others attempt to describe their emotions for them, as in Amos being asked "are you angry?" Simple, clear questions can explore what matters to Amos right now (the bicycle, for example). It is vital furthermore to acknowledge Amos and his family's expertise of their own lives and experiences by exploring who knows what, and who wants who to know what? This family is an obvious illustration of a family's previous confrontation with the death of a child and the grief that was associated with that. Resilience and coping strategies of all family members will constantly change and grow, but always an inquiry into their knowledge and experience will provide guidance for the palliative care team. For example, it may be helpful to ask Sybil "What would you want to protect Amos from?" "What does Amos need to know so that he can feel protected?" "Do you think Amos is trying to protect anyone?"

## 16.5 Question 5: How Can We Support the Family in Their Loss of Future Dreams and Help Them Focus on What Is Happening Now?

Childhood development presupposes a future for Amos, comprising dreams, goals and fantasies of what a child or young person may become, so this diagnosis presents confusion for all the family as to their roles and relationships with a child who will not reach adulthood. As difficult as it may be, Pretty and her parents need to move away from a future framework and provide a present focus for interaction while Amos lives. Children such as Amos, who have witnessed the deaths of others from the same or similar conditions (as in HIV or inherited conditions), may have specific fears about dying driven by their personal experience. Recognition and accommodation of these fears is a prerequisite before providing reassurance that all that can be done will be done throughout the illness progression, including living as normally as possible. This includes school attendance and play with peers where possible.

Family stability can be maintained as far as possible by continuing routines, clear rules and boundaries (McKissock 1998). Integral to this is creating a safe environment in which the child feels free to express emotions and talk about preoccupations, with those around able to explore different possible meanings. For example, when Amos asks "Is what happened to Elijah going to happen to me?" the team member can respond: "Are you asking if your illness is the same as Elijah's? Are you also asking if you will die in the same way as Elijah?" Responses to these kinds of questions can clarify meanings as well as generate new questions and guide the team and family towards exploring aspects of anticipatory grief and meeting the needs of the child. Siblings may feel invisible, neglected or abandoned. When we meet Pretty she is on her own, with her own thoughts and feelings and no adult actually knew where she was. Some siblings may feel angry that the sick child receives so much attention, followed by intense guilt for acknowledging these feelings.

## 16.6 Question 6: What Losses Might Amos Be Grieving?

Dying children and adolescents such as Amos may grieve many losses during this time. There may be the immediate shift in health and independence, attractiveness and inclusion in the lives of their peers. Longer term dreams of future romances, sexual experiences, career, travel, marriage and children may also be identified with sensitive inquiry and support (Amery 2016). By providing a context of safety and acceptance, a dying child such as Amos can use his anticipatory grief to construct a uniquely individualised knowledge about his death in relationship with his family, culture, community and hospital. It is helpful during this time for the patient and family to consciously capture memories through development of memory books and boxes, special mementoes and sharing of important conversations. Legacy-making interventions can be powerfully healing, such as Pretty giving Amos a drawing of his bicycle and Amos leaving his bicycle to Pretty as a special inheritance (Akard et al. 2015).

**Case Study**

*When Amos died the family followed their cultural tradition of burying their son next to his brother in the family burial plot. Because Pretty had been able to spend many hours talking and drawing with Amos while he was unwell, she knew that he wanted her picture of his bicycle to be buried with him, and the family fulfilled this promise. Sybil noticed that Pretty sat at the grave almost every day, and heard her crying and talking to Elijah and Amos. Sybil wondered if this was normal but when she asked Pretty how she was doing, her daughter refused to talk, sometimes claiming she had nothing to say and at other times she ran off and climbed a tree. There were also times when Pretty could not sleep and she seemed increasingly anxious. She insisted on sleeping in her parent's bed. At times she refused to leave the house and John had heard from her teachers that she had been reprimanded at school for fighting with her friends.*

*Several months after Amos died John felt depressed and indicated that he found life meaningless and sometimes thought of killing himself. He and Sybil argued a lot and did not mention their sons' names at all. Sybil continued to work with difficulty but felt isolated and alone with her grief, finding it hard to put her agony into words. Pretty became increasingly angry and in an outburst, blamed her parents for giving this illness to both her brothers. She said she wished she could die.*

## 16.7 Question 7: What Emotions Might Amos' Family Be Experiencing?

A child's death feels wrong, out of time, and leads to a liminal space, without anchor, floating "in between" anything that has been previously known (Carter 2017). Members of a grieving family will experience a wide range of emotions, experiences, thoughts, physical symptoms, social and spiritual changes and other reactions over a long period of time, at differing times and with varying intensities and combinations. In a PC context, exposure to the dying process provides valuable preparation for bereavement, with the patient, parents and siblings encouraged to benefit from the healing power of conversations. This can promote resilience and help to prevent post-traumatic stress (Lichtenthal et al. 2015a; Weaver et al. 2015; Jaaniste et al. 2017; Wiener et al. 2018; Waugh et al. 2018). Involvement in this process may result in fewer regrets, knowing that family members have done all they can. When a child dies of a LTC, as in the case of Amos, the family has the chance to discuss and achieve goals, enjoy living to the best of their ability and to say goodbye (Wallin et al. 2019).

Many emotions are experienced in bereavement, not in any particular order, some repeatedly and others a sharp and sudden insight. Grief and its reactions are not completed in stages, but rather as a lengthy, tumultuous roller coaster. While it is commonly believed that the hardest part of grief is when the death occurs, it can take some weeks and months before the body and mind allows the bereaved to fully absorb the reality of the loss. It may be confusing and frightening for Sybil, John and Pretty to feel a lot worse about 6 months after Amos died, despite their anticipatory grief during his illness. It can be some months after the loss that true despair

sets in, a reactive and constructive depression to the loss of a loved one, when there is reflection on what the future holds in the context of unbearable pain. It is usual for the bereaved to allude to or consider suicide at times; this is a measure of how painful grief is for them, and it helps to be able to share these thoughts without judgement or panic.

## 16.8 Question 8: What Are the Specific Challenges for 10-Year-Old Pretty?

For Pretty, there are many important aspects to consider when helping her manage her reactions. Bereaved children in particular need simple, factual information to explain what is happening. They need to be listened to with empathy, and given permission to grieve in their unique way, assured that there is no right or wrong way to grieve, and they need to be given time and support for feelings to be shared or expressed. A simple recognition and explanation of the many emotions experienced, including sadness, may help bereaved children to construct, for themselves, their emotional reactions to and the meaning of this life-changing event (Weaver et al. 2019).

Grief reactions of children such as Pretty will change over time and maturity and will vary according to gender, culture, age, previous losses and experiences, family support, the relationship to the deceased and the nature of the death. It may surprise and confuse parents how quickly young children may switch between emotions, one moment crying for the deceased to return, followed rapidly by laughter and play with friends. Indeed, an initial response may be startlingly practical. Perhaps Pretty asked "Can I have his bicycle? Who will have his toys? Will I still be able to go to school?" This basic need to ensure the child's own continuity possibly assists in making the environment safe enough to allow grief to occur.

Fear is commonly experienced by a grieving child. The death may raise concerns that they caused their loved one to die, (perhaps having had a fight or said things in anger such as "I wish you would die"), that they too may die, or someone they love may die. Fear may manifest in regression to earlier behaviours such as becoming clingy, bedwetting or thumb-sucking which require understanding and sensitive management. Checking on the whereabouts of family members, insisting on not sleeping alone and not wanting to go to school may all indicate separation anxiety.

Sadness is inevitable in grief. Crying can help and afterwards the child may feel tired but peaceful. Modelling of sadness by parents and adults is necessary to normalise this reaction and validate that they too are important and would also be missed if they died. Sadness says "I hurt" and, when it is expressed, elicits compassion, comfort and empathy. Watching sad movies or reading a sad book during their bereavement may cause more crying than usual, and provides "an excuse" for more emotional release. When expression of sadness is difficult or restricted, however, children may show physical, behavioural or social symptoms, such as anger and aggression. Sybil's sense of being unable to share her grief with her family may mean Amos is seldom talked of. Sybil may find it hard to know how to help Pretty and may hope that as a child she will "just get on with things". Sybil's lack of engagement with Pretty and inability for the

moment to demonstrate healthy grieving will make it difficult for Pretty to process the impact of Amos' death on her family. One way she can express her pain and confusion will probably be in her behaviour and angry outbursts. Anger can often be understood as a means of pleading for the situation or the pain to stop, and enquiring as to whether this is what Pretty is feeling, may open up further conversations. In this case we hear that Pretty has been fighting with her school friends. Children, like adults, tend to move away, shut down or become withdrawn when angry to avoid hurting self and others. Understanding this behaviour assists adults in helping bereaved families to distinguish between anger and aggression and to find safe ways to express the pain of grief. "*Grief is an explanation, not an excuse*" (McKissock 1998, p. 149).

Bereaved children may gain understanding through play, enacting death scenes, burial and digging up items to check what has happened to a "body". They may cover themselves in blankets and pillows to experience what it may feel like to be buried. In cultures where cremation is common, preoccupation with burning, what is left in ashes and imagining what happens to the body can be expressed and worked through by drawing or enacting relevant scenes. They may experience vivid dreams and nightmares. Ask them to describe/draw what they remember, what feelings they are left with upon waking and what they understand by them. The family belief system needs to be addressed by exploring the impact of grief upon it. It may be helpful to ask "How does this death change what you believe about life and death?" Integrating spiritual conversations into the bereavement process is known to positively assist bereaved siblings (Lövgren et al. 2019; Eilertsen et al. 2018).

Guilt is commonly experienced during this process, by parents and siblings, for a variety of reasons either real or perceived. Amos was the second child in this family to die from this kind of cancer, although this cancer is seldom an inherited condition, and despite different treatment interventions. It will be helpful for the family to explore their perspectives of guilt, powerful or magical thinking to validate their emotions and help them uncover at their own pace the reassurance that this was not their fault. It is generally unhelpful, however, to simply repeat that they are not guilty when this is what they feel. Where death results from a non-communicable disease siblings may need to be reassured that the illness is not contagious.

To talk or not to talk about the deceased is an individual choice that the child can make in a context where sharing about the deceased is permitted, encouraged and respected. It is relevant to explain to a bereaved child that not mentioning the deceased is to deny his existence, as though he never mattered (Fredman 1997). As we all carry different messages about talking depending on contexts, relationships, timing and culture this will vary over time and situation. Care should therefore be taken not to label a child's expression of grief according to an adult's expectation of what grief should look like.

## 16.9 Question 9: What Support Can We Give to Pretty to Enable Her to Grieve Healthily?

There are many activities that can ensure a safe and nurturing environment for a child to grieve healthily. Where possible, display photographs and memorabilia of the deceased, allocating these to siblings as desired. Inheriting Amos' bicycle will be a

tangible connection between Pretty and both her deceased brothers, something of theirs that she can take into the future. Memory books and boxes that contain special messages, belongings, anecdotes and pictures help to maintain the relationship with the deceased after death (Akard et al. 2015). Special celebrations need to be acknowledged by all in the family, together with events that mark important milestones, memories, future plans and goals.

Parents and other significant adults need to take the initiative to include children of all ages in developing rituals that help to make sense of their loss. By so doing, children such as Pretty feel valued and integrated in the changed family (McKissock 1998). Through a process of sensitive curiosity and compassionate support, children and others in the family can be helped to revisit old selves and evolve new selves by rewriting their personal stories to answer questions such as "Who am I now? Am I still a sister?" These opportunities can also act as active preparation for future death events, including their own. Family sharing helps coordinate memories that enable the bereaved Pretty to create a context from which she can carry on, by incorporating her deceased siblings (Wallin et al. 2019). The bereaved Pretty may find that well informed teachers and the opportunity to interact with peers when returning to school help maintain continuity and routine (Howard Sharp et al. 2018). Where available bereaved siblings and parents can benefit from the camaraderie and sharing that is offered in support groups (Lichtenthal et al. 2015b).

## 16.10 Conclusion

A child's or young person's experience of loss, whether of their own future life or in response to the death of a loved one, is painful on many levels. Recognising that the resulting grief reactions are a healthy and necessary response in order to heal over time is an important step for everyone to take. By honouring the uniqueness of each individual through listening without judgement, providing honest communication and reassurance, children and young people can be enabled to express their grief as they need to over time.

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# Perinatal and Neonatal Palliative Care

# 17

Alexandra Mancini-Smith and Rut Kiman

**Key Learning Points**

1. It is only in recent years that perinatal and neonatal palliative care (PC) have been recognised as an emerging specialty within children's palliative care (CPC).
2. The British Association of Perinatal Medicine (BAPM) has developed five candidate conditions which assist the healthcare professional in identifying the foetus or infant with PC needs.
3. In providing perinatal and neonatal PC it is important to consider: what is important for the parents? what might the parents consider in terms of caring for a baby with complex health needs, or the possibility of the baby dying before it is born or shortly afterwards? what are the challenges in providing choices?
4. Parallel planning is a key component of care and it involves preparing the families for the different scenarios—planning for survival and preparing them for their infants' possible death.
5. In order to deliver high quality perinatal and neonatal PC, it is fundamental to identify, assess and plan care appropriately in partnership with the parents and family.

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**Case Study: Perinatal Palliative Care**
*Jane was pregnant with her 4th child, having had her first three children by caesarean section who are now aged 2 years, 6 years and 15 years old. Antenatally at 22 weeks, the baby was diagnosed with Trisomy 18 (Edwards syndrome) with an additional cardiac condition—an atrioventricular septal defect (AVSD).*

## 17.1 Question 1: Why Would Jane Be Considered for Palliative Care?

It is only in recent years that perinatal and neonatal PC have been recognised as an emerging specialty within CPC. There has been an increasing awareness globally, mainly due to the advancements of technology in detecting in utero conditions, the rise in survival rate of extremely premature infants and also the influence of the media in reporting families' experiences (Wilkinson 2006; Costeloe et al. 2008; Ramnarayan et al. 2007). This is a growing area for CPC, and as such, the focus must be on identifying infants, including those who are not yet born, who may have PC needs at the earliest opportunity possible, so that the appropriate care and planning may be initiated within the multidisciplinary team (Mancini et al. 2014).

The unborn infant or the newborn is a new member of an emerging family, where parents are often learning to be parents for the first time. The mother's health must be integral to the care planning for the infant's uncertain future, where there are high levels of prognostic uncertainty.

High quality perinatal and neonatal PC requires consideration of what the family's wishes are, facilitating the family being together wherever possible, communicating difficult information sensitively, acknowledging differences in language and culture, whilst paying attention to the spiritual needs of both parents and the wider family including siblings.

One cannot underestimate the impact that the quality of care towards the end-of-life (EoL) of an infant can have on families; an impact that can last for many years after the infant has died (Papadatou 2009; Woodroffe 2013).

## 17.2 Question 2: What Is Perinatal Palliative Care?

The BAPM (2010a) defines *palliative care for the fetus, neonate, or infant with life-limiting conditions as "an active and total approach to care, from the point of diagnosis or recognition, throughout the child's life, death, and beyond. It is the holistic management of supportive end-of-life care following multidisciplinary agreement on eligibility" (p. 1)*. The philosophy and principles of PC integrated into routine neonatal care can enhance the quality of care infants and their families receive (Murdoch et al. 2013). Furthermore the integration of relevant services ensures the best possible care and experience for the infant and their family during life and support after death.

1. An antenatal or postnatal diagnosis of a condition which is not compatible with long term survival.
2. An antenatal or postnatal diagnosis of a condition which causes a high risk of significant morbidity or death.
3. Birth at the margins of viability, where intensive support has been deemed inappropriate.
4. Postnatal clinical condition with a high risk of impairment of quality of life (QoL) and the baby is receiving or may require life support.
5. Postnatal conditions which result in the baby experiencing 'unbearable suffering' (BAPM 2010b).

**Fig. 17.1** Candidate conditions eligible for perinatal PC in 5 broad categories (BAPM 2010b)

The British Association (BAPM 2010b) have developed five candidate conditions which assist the healthcare professional in identifying the foetus or infant with PC needs (Fig. 17.1).

It is important to:

- Identify babies with PC needs.
- Assess what is important and realistic for the family.
- Plan in partnership with the family and the multidisciplinary team.
- Implement care planning with uncertain future.
- Evaluate and review.

## 17.3 Question 3: What Is the Difference Between Perinatal and Neonatal Palliative Care

This is mainly attributed to the time frame or period of recognition, identification, assessment and planning of care. Perinatal PC pertains to care provided during the period of time before the baby is born, at the time of birth and then shortly after birth, when an infant has a life-limiting condition (LLC). The definition of the perinatal period commences at 24 weeks gestation until 7 days post birth. It provides an opportunity for the treating team to give support and care for the parents and family when there is a diagnosis before the baby is born. It is an opportunity for healthcare professionals to help the parents in making decisions and planning care which is unique to their family, for the time of birth, shortly after birth and beyond. A LLC may be confirmed from imaging or genetic testing, but this diagnosis may also require interdisciplinary involvement including maternal foetal medicine specialists and geneticists (ACOG 2019).

Neonatal PC is the term usually used to describe the care of the newborn infant from the time of birth, through the first 28 days of life and then the time the infant is cared for on the neonatal unit when the baby is identified as having PC needs (BAPM 2010a). It is when healthcare professionals can help provide support and assist in care planning after the baby is born. Sometimes there is no diagnosis to support planning, and it is usually initiated when an infant is born prematurely, born with congenital conditions or following a significant insult at the time of delivery. There may be some infants who spend many weeks, even months, on a neonatal

intensive care unit, whilst receiving active treatment alongside PC. Bidegain and Younge (2015) identified six key roles for neonatal PC in the intensive care unit, which include:

1. Pain and symptom management.
2. Prenatal PC consultations.
3. EoL care.
4. Discharge to hospice.
5. Communication and conflict resolution.
6. Collaboration in the care of the medically complex infant.

Internationally, there will be variations in resources, care planning models and differences in treating clinical teams. Some countries may have prioritised advancements in perinatal rather than neonatal PC, for example, there is an abundance of current literature in the United States of America (USA) and Ireland. This may sometimes cause confusion for healthcare professionals and, as previously described, perinatal and neonatal PC are different. However, irrespective of definitions, funding and resources, the key principles of perinatal and neonatal PC may be comparable.

## 17.4 Question 4: What Are Some of the Challenges for Providing Perinatal and Neonatal Palliative Care?

- With the advancements of technological investigations, parents may receive an antenatal diagnosis that their baby has a LLC or a significant surgical condition, which may mean they are eligible/identified for PC support before the baby is born.
- New parents, who have never had their baby at home, may be faced with preparing for the birth of their new baby at the same time as preparing for their baby's death.
- Babies requiring PC are usually cared for in a noisy, busy neonatal intensive care unit, where their clinical condition may deteriorate or may improve suddenly within a short space of time.
- There may have been a multiple pregnancy, where subsequently, there may be one or more babies being cared for in different hospitals. The parents may have to care for a critically unwell baby whilst grieving the death of another baby.
- The father of the baby or the mother's partner may be extremely anxious in worrying about the uncertainty of the future of the baby, whilst the mother may be unwell following delivery requiring medical care of her own.
- There may be a very short amount of time in which families can spend with their baby before death.
- There are limited opportunities to create memories with their baby and other family members, particularly siblings.
- Multiple teams and services may be involved, across professional boundaries, requiring excellent clear communication and documentation.

## 17.5 Question 5: How Would You Conduct an Assessment in Order to Plan for Jane and Her Unborn Child's Care?

As in all PC, assessment is key to the planning and provision of care. You would need to think about what you know about the condition, Trisomy 18, whether it is a LLC and what is the impact on the unborn baby and their family. If looking at the BAPM conditions for perinatal PC (BAPM 2010a) the unborn child would evidently fit into the first group, i.e., have an antenatal or postnatal diagnosis of a condition which is not compatible with long term survival.

In conducting an assessment, it would be important to think about the following:

- Which other healthcare professionals should be involved in the clinical assessment of the unborn baby?
- What impact will the cardiac condition have on the baby? Is it a possibility for the baby to have surgery? What are the risks and benefits for surgery?
- Routine antenatal care for Jane must continue, and it is important to ascertain if she plans to have a caesarean birth, needing specific postnatal care, and this may impact on where the baby is cared for and limit the choices available for the family.
- What will happen regarding the parents' wishes and plans if the baby dies in utero before birth?

## 17.6 Question 6: What Are Some of the Issues to Consider in Providing Care for Jane and Her Family?

In providing perinatal PC it is important to consider the following issues:

- What aspects of care have the parents verbalised are important for them as a family?
- What might the parents consider in terms of caring for a baby with complex health needs, or the possibility of the baby dying before it is born or shortly afterwards.
- What are the challenges in providing choices?
- Is there any new research or evidence that might help healthcare professionals decide what choices may be available for Jane and her family?

Some of the principles of perinatal PC include:

- Ensuring that normal routine antenatal care is continued where possible.
- Make a plan with the family for the birth and what care might look like after the birth.
- If a birth plan has been developed, that it is considered and followed as much as is possible.

- Signpost the family to reliable sources of information, e.g., on the internet or local organisations.
- Manage the parent's expectations of care for their baby and the plan of care.
- Establish a main point of contact for the family, such as a named healthcare professional (midwife or a nurse), include contact details such as telephone number or email address, so that the family may contact that at any time.
- Collaborate closely with multi-professional colleagues and external resources.
- Provide continuing emotional and practical support for the family and health professionals.
- Consider support for the other children in the family—in this case, Jane already has three children, some of whom are old enough to understand what is happening.
- Create opportunities for the family to make memories together.
- Care for the baby and their family with compassion, respect, dignity and kindness.
- Ensure you have a non-judgemental approach to the family and care planning.
- Offer Jane the choice of donating her expressed breast milk, if she wishes.
- Discuss what will happen with regards to a post-mortem and the possibility of organ donation where possible.
- Time……there may be limited time, therefore time is of the essence in arranging meetings and opportunities for the family to be together.
- Ensure there is relevant parallel planning, preparing for different scenarios.

It is also important to think about the following:

- Which other clinical teams need to be informed about Jane's unborn baby?
- What is important for this family?
- Consideration of appropriately skilled healthcare professionals, community teams, geneticists, dietitians, neurologists, etc.
- There may be several teams involved in the care planning, which requires coordination preferably by a dedicated nurse or other healthcare professional.
- An ACP needs to be developed where possible.
- Flexibility of the plan in accordance with the baby's changing clinical condition.
- What are the realistic options available, e.g., can they take their baby home? is there a children's hospice available? is there community service support?
- Consider spiritual and religious needs for the family, remembering that these may differ amongst family members.
- Mother and father to be supported to hold and cuddle the baby at birth, and her brothers to also meet her as soon as possible.
- Non-pharmacological care—warmth, dimmed lights in labour ward theatre, gentle music, baby massage and brothers to meet her with photographs taken.
- Post-mortem for confirmation of the diagnosis and possible further information.
- Consideration for organ donation, explore if this is possible.

## 17.7 Question 7: What Is "Parallel Planning" in the Context of Caring for Jane, Her Family and Her Unborn Child?

Parallel planning is a key component of perinatal PC and it involves preparing the families for the different scenarios—planning for survival and preparing them for their infants' possible death. The unborn child may die before birth in utero, they may die at the time of birth or shortly afterwards, or the baby may survive several days or weeks. The mother will need routine antenatal and postnatal care and needs to be aware that the baby's clinical condition may change suddenly at any time, and that they need to plan for this. Therefore good communication and multiprofessional planning are essential with the plan being continuously reviewed with the family and healthcare professionals.

## 17.8 Question 8: How Can We Support Families Such as Jane's to Make Decisions?

The issues are similar regardless of which country you work in and include the following:

- Through PC, giving the parents the opportunity to share their fears and their hopes.
- Respect.
- Kindness.
- Compassion.
- Being non-judgemental.
- Having all the information possible about the baby's condition, possible options for treatment, resuscitation, etc.
- By providing as normal an experience as possible for the family and supporting any other children, allowing them to be a family and create many good as memories as possible.

**Case Study: Neonatal Palliative Care**

*A baby boy, Peter, was born at 39 weeks following an uneventful pregnancy to married parents. Following a difficult delivery he suffered asphyxia, confirmed as severe hypoxic encephalopathy (HIE). At birth he had a slow heart rate, poor muscle tone and poor respiratory effort. Cardiopulmonary resuscitation was commenced and Peter was intubated. Therapeutic hypothermia was also commenced. A Magnetic Resonance Imaging (MRI) scan confirmed basal ganglia and thalami injury. Peter is now 3 weeks old. The neonatal team are meeting Peter's parents to discuss the plan of care going forward and the consideration of withdrawal of life sustaining treatment. Peter is his parents third child and has 2 brothers aged 2 and 4 years old.*

## 17.9 Question 9: What Are Your Initial Thoughts and Considerations in Talking to the Parents About the Plan of Care for Peter?

Initially it is important for you to consider if Peter has a potentially LLC and requires PC. Referring to the BAPM (2010b) categories could help with this. If so, you need to think about the key principles that you need to consider to ensure that Peter's clinical needs are addressed and his whole family are supported.

## 17.10 Question 10: What Are Some of the Points You Would Consider in Undertaking an Assessment of Peter's Needs Along with His Families?

Having had an uneventful pregnancy, this will come as a shock to Peter's parents, who had no warning that their baby might have a LLC and might not live for very long. For parents and families being given the news that their baby has a LLC and may die is extremely harrowing and this can be a very difficult and confusing time in their lives. Branchett and Stretton (2012) led a study with parents of babies on a neonatal unit who were not expected to survive. Key findings included the way in which healthcare professionals communicate with families at this time is of the utmost importance and must be approached sensitively and compassionately. The focus of care is on maximising the QoL for that baby, in accordance with their family's wishes, encompassing every aspect of the family structure, whilst ensuring families have the relevant information available to support their decision-making process.

Therefore, it will be important to support them through this transition from excitement for having another baby, to the potential loss of their baby. As well as providing clinical care for Peter it is essential to review his mother's health needs, along with the needs of his two brothers and the wider family, some of whom may be needed to take care of his brothers at this difficult time. Peter's specific clinical needs will also impact on where continuing care will be delivered, e.g., in the intensive care unit, in a hospital ward, at home, etc. It may also impact whether he can be cared for near to the family's home, or whether he is in a treatment facility a long way away, therefore potentially splitting the family at what is a difficult time when they need to be together. Therefore, it is important to determine who needs to be involved to ensure that they are provided with the best possible care and support at this time, and where that can be given.

## 17.11 Question 11: What Educational Opportunities Are There Available for Individuals Providing Perinatal and Neonatal Palliative Care?

In recent years, United Kingdom (UK) professional agencies and bodies such as the British Association of Perinatal Medicine (BAPM), Bliss, Together for Short Lives (TfSL), Department of Health (DH) and the Royal College of Paediatrics and Child Health (RCPCH) have supported the growing need for good quality PC during the

perinatal and neonatal period via educational interventions. Several national publications including frameworks of care and pathways are recognised and highlight good standards of practice (BAPM 2010a, b; BLISS 2011; Mancini et al. 2014; Sands 2016; TfSL 2017; Mancini et al. 2020). These publications provide a foundation on which healthcare professionals can develop local education and training initiatives and emphasise the need for individualised care plans and family centred care, where family choice is given a high priority.

Following evidence to support the benefits of PC, the Nuffield Council on Bioethics (2006) introduced recommendations that specified all professionals working within the field of neonatology in the UK must have mandatory training in the principles of PC for infants. More recently, the importance of appropriate and consistent training for professionals within this specialist area has been further supported in national publications (DH 2009; Bliss 2011). Whilst it is important to recognise that there is evidence of effective local training programmes in neonatal PC (Soni et al. 2011; Mancini 2011; Gallagher et al. 2012; Nurse et al. 2020), training and education in the principles of neonatal PC remain variable and fragmented.

Price et al. (2013) reinforce that it is imperative that healthcare professionals receive appropriate training in a highly specialised and complex field such as neonatal PC, where it is rapidly expanding, and professionals need to respond to the demands on an expanding and developing service by focusing on appropriate education for healthcare professionals (Downing and Ling 2012). Argentina is one country where training in perinatal and neonatal PC has been incorporated into the training of paediatric subspecialists in PC in recognition of its importance (Sociedad Argentina de Pediatria 2015). There are also several ongoing training programmes on perinatal and neonatal PC in Argentina producing changes at the local level (Kiman and Doumic 2014).

Education and training are an integral part of the provision of high quality neonatal care which includes complex and palliative care (Mancini et al. 2013, 2014, 2020) and assists professionals in developing and enhancing the skillset required to deliver excellent holistic care. However, education and training on perinatal and neonatal PC is limited, with some individuals having no access to face-to-face training in this area. Thus, efforts have been made to provide online e-learning options for training, such as that provided to its members by the Royal College of Midwives, or through the International Children's Palliative Care Network (ICPCN)s e-learning programme (www.elearnicpcn.org) (Daniels and Downing 2018) where individuals can access online training regardless of where in the world they live. However, wherever possible, formal education and training should begin at undergraduate level for nurses, midwives, medical staff and allied health professionals. Nurse et al. (2020) illustrate the benefits of healthcare professionals accessing relevant training at the inception of their career, which should also continue as lifelong learning.

## 17.12 Conclusion

In order to deliver high quality perinatal and neonatal PC, it is fundamental to identify, assess and plan care appropriately in partnership with the parents and family. In recent years there have been increasing challenges in identifying which infants may

require PC. It can be challenging for healthcare professionals to define and categorise infants who may be eligible for PC as infants do not fit neatly into specific groups or categories. With these challenges in mind, the BAPM in their report (BAPM 2010b) detailed five broad categories to assist identification. Other frameworks are also available internationally and should be used to plan care for the baby and their families. The specialty of perinatal and neonatal PC can be challenging for staff and be a source of distress, so it is therefore essential that care providers are supported emotionally and psychologically to care for babies and their families. This may be via training and education, debriefs, opportunities for reflection and sharing experiences with colleagues.

Wherever you are in the world, despite limited resources, you can always be kind, considerate and inclusive of parents' wishes, warmth, comfort, facilitate memory making opportunities, encourage family moments.

## Useful Links

BLISS: www.bliss.org.uk
CBUK: www.childbereavementuk.org
Child Death Helpline: www.childdeathhelpline.org.uk
Contact a Family: www.contactafamily.org.uk
MBF: www.multiplebirths.org.uk
Newlife: www.newlifecharity.co.uk
NHSBT: www.nhsbt.nhs.uk
Organ Donation: www.organdonation.nhs.uk
Rainbow Trust: www.rainbowtrust.org.uk
Sands: www.sands.org.uk
SIBS: www.sibs.org.uk
TAMBA: www.tamba.org.uk
TFSL: www.togetherforshortlives.org.uk
WellChild: www.wellchild.org.uk
Free Online Perinatal Palliative Care Module: www.elearnicpcn.org

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# Supporting the Adolescent and Young Adult

# 18

Lucy Watts, Julia Downing, Marianne Phillips, and Lizzie Chambers

**Key Learning Points**

1. The needs of this specific population are unique to their age group and services should be tailored to their needs in terms of age and developmental stage.
2. It is important that adolescents and young adults (AYAs) are recognised as individuals, ensuring that their needs are met in a developmentally appropriate way, as well as recognising that they are not yet mature adults and may need additional support to meet their emerging social, emotional, relational and sexual needs.
3. AYAs can often struggle with a high symptom burden and yet want to minimise treatments and health intervention so as to 'get on' with their lives; it is important that they are supported to adhere to treatments and

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attend medical appointments, whilst living life fully and participating in education, employment and social activities.
4. Transitioning young adults from children's to adult-focused services needs to begin early to ensure a smooth transition and continuation of care.

**Case Study**
*Lucy had health problems from birth and regularly saw medical professionals but managed to live a somewhat normal life until she became seriously ill and completely disabled at the age of 14 when it was clear that her condition was going to dramatically shorten her life. Her condition had proven difficult to diagnose as it was complex and rare; however, eventually it was decided she has a rare form of Mitochondrial Disease. At 14 she needed a wheelchair for mobility and soon was completely bed bound, requiring round-the-clock care. Her condition was extremely complex and she depended on a lot of interventions to survive. She was not expected to see her 18th birthday. At the age of 17, Lucy became supported by an age-appropriate young adult hospice (for 16–40 year olds) which respected her for the emerging adult that she was.*

## 18.1 Question 1: How Do You Define an Adolescent and Young Adult Within Palliative Care?

Adolescence is defined as 'the transitional period between puberty and adulthood in human development, extending mainly over the teen years and terminating legally when the age of majority is reached' (Dictionary.com 2019). The WHO describes adolescents as individuals aged 10–19 years. Within the PC context, young adults have been defined as 'individuals in the late teenage to mid-twenties age band: they are "emerging adults" in transition from childhood to adulthood' (Beresford 2013, p. 9). AYAs with PC needs represent a challenging group. It is a time when their specific needs, distinct from those of children and adults, may be unrecognised or unmet. It is a time of paradox—an exciting but frightening time; a time in adolescence when one may be a child in an adult body; a time when starting to be part of an adult world provides time to set roots and establish one's identity (Amery et al. 2009). Added to this, the complexity of living with a life-limiting (LLC) or LTC during this period of change provides a challenge to services to provide appropriate levels of care.

## 18.2 Question 2: Why Has the Care of Adolescents and Young People Such as Lucy Become Such an Important Issue?

There is a new emerging 'unforeseen generation', of young people with life-threatening conditions (LTCs) and PC needs. Many AYAs now survive into adulthood with conditions previously only encountered in childhood (Beresford and

Stuttard 2014). There are a wide range of genetic, congenital and acquired conditions, that dominate the PC field for AYA in developed countries. This unforeseen generation are now outliving children's palliative care (CPC) services, whilst adult PC services are not yet established to appropriately meet their needs.

In some areas of the United Kingdom (UK), young adult-centric services—whether attached to children's hospices, adult hospices or as standalone services—are a recent addition to the PC landscape although there is a lack of consensus around what the upper age limit of 'young adulthood' is, with some services caring only for individuals from 16 to 25 years, whilst others extend their services to those aged 40. Services find these young people have unique needs that neither children's nor adult services are equipped to meet. Some of these young people want to live full and active lives, to participate in society, to continue in education or to get a job; they want to live independently, they may want to have families and they want to explore their sexuality and relationships whilst those with profound and multiple learning disabilities (PMLD) continue to need round-the-clock care.

## 18.3 Question 3: What Is Unique About the Period of Adolescence and Young Adulthood?

The period of AYA represents one of the critical transitions in the life span characterised by a tremendous pace in growth and change, second only to that of infancy. It is the period of development during which the adult identity is formed (Table 18.1). However, the impact of illness may be profound: individuals may regress, delay or even miss critical developmental steps, leading to negative consequences (Craig and Lidstone 2012).

For AYAs there is a wide range of maturity and disease literacy, alongside a range of developmental levels and cognitive abilities. Some AYAs will have been impacted by their LLC or LTC all of their lives, and have grown up with an understanding of what that might mean for them in terms of independence, the need for ongoing care, deterioration, etc. For others, such as Lucy, they will have come to an understanding of their condition as teenagers, at a time of already entering the period of adolescence and young adulthood.

**Case Study**

*Lucy's symptoms were not effectively managed which had a direct impact on her ability to participate in society and family life. Whilst professionals had focused on cure, Lucy's holistic needs were not being met. Lucy was regarded as a patient with conditions to fix, rather than as a person with a life and needs. She wanted to be supported to live well and participate in activities with her peers. Lucy felt her life revolved around her medical needs and treatments, and that she 'existed, rather than lived'. It took time for Lucy and her team to strike a balance between appropriate symptom control and treatments and her quality of life (QoL). It took a few years of trying and testing different treatments before the combination that enabled Lucy to sit up in her wheelchair for long periods and to function coherently was found in order for her to feel she could participate fully. It was then that Lucy's nurse asked 'So what do you want to do now?' that led to Lucy being supported to become*

**Table 18.1** Adolescent development (Chambers 2015, p. 11, reprinted with permission)

| | Early adolescence<br>12–14 years (female)<br>13–15 years (male) | Middle adolescence<br>14–16 years | Late adolescence<br>17–24 years |
|---|---|---|---|
| Key issues and characteristics | • Focus on development of body<br>• Most pubertal changes occur<br>• Rapid physical growth<br>• Acceptance by peers<br>• Idealism<br>• Mood swings, contrariness and temper tantrums<br>• Daydreaming | • Sexual awakening<br>• Emancipation from parents and authority figures<br>• Discovery of limitations by testing boundaries<br>• Role of peer group increases | • Defining and understanding functional roles in life in terms of:<br>– careers<br>– relationships<br>– lifestyles |
| Social, relationships, behaviour | • Improved skills in abstract thought<br>• Foreseeing of consequences and planning for future<br>• Physical mobility prominent<br>• Energy levels high<br>• Appetite increased<br>• Social interaction in groups<br>• Membership of peer group very important | • Relationships very narcissistic<br>• Risk-taking behaviour increases<br>• Intense peer interaction<br>• Most vulnerable to psychological problems | • Increasing financial independence<br>• Planning for the future<br>• Establishment of permanent relationships<br>• Increasing time away from the family |
| Impact of LTC | • Concerns about physical appearance and mobility<br>• Privacy all-important<br>• Possible interference with normal cognitive development and learning (school absence, medication, pain, depression, fatigue)<br>• Comparison with peers hindered, making self-assessment of normality more difficult<br>• Possible lack of acceptance by peers<br>• Reliance on parents and other authority figures in decision-making<br>• Hospitals perceived as very disturbing | • Illness particularly threatening and least well tolerated at this stage<br>• Compromised sense of autonomy<br>• Emancipation from parents and authority figures impeded<br>• Interference with attraction of partner<br>• Fear of rejection by peers<br>• Limited interaction with peers may lead to social withdrawal<br>• Dependence on family for companionship and social support<br>• Hospitalisation, school absences interfere with social relationships and acquisition of social skills<br>• Non-compliance with treatment | • Absences from work, study<br>• Interference with plans for vocation and relationships<br>• Difficulties in securing employment and promotion at work<br>• Unemployment hinders achieving separation from family and financial independence<br>• Discrimination in employment, health cover and life insurance<br>• Loss of financial independence and self-esteem<br>• Concerns about fertility and health of offspring |

*involved in voluntary advocacy and other work. This then became Lucy's life; it kept her going mentally, which significantly helped her medically.*

## 18.4 Question 4: What Are Some of the Unique Needs of Adolescents and Young Adults, Such as Lucy, in Palliative Care?

A key aspect of AYAs is developing independence. Within the context of PC, many AYAs will not be able to reach what is traditionally regarded as maturity into adulthood due to the significant impacts of their condition. For many AYAs needing PC, the definition of independence needs to be what works within the limits of their condition. For example, Lucy has complex needs, is clearly dependent on others for a lot of her care and is often bedbound, so how she can achieve 'independence' within the limits of her condition is vital.

**Case Study**

*As a young person receiving PC, Lucy wants to be viewed as a young person first and as being unwell as a secondary consideration. She wants to retain control over her body and be respected in the decisions she makes. She prefers to be informed even when the prognosis is poor and has an altruistic desire to help others by participating in research. She wants to live until she dies, capturing new life experiences, for life to have meaning and to be remembered.*

AYA is a period of changing relationships with parents and caregivers. Many AYAs with PC needs remain dependent on their parents, some seek their parents' support to make big decisions, while others become relatively independent with organising their life and care. Alongside this there are a range of unique needs for AYA within the context of PC (Fig. 18.1).

- The presence or absence of close friendships and their ability to form a personal identity separate from their family.
- The effect of peer group and peer pressure, particularly as this impacts their condition, and their condition impacts on their ability to socialise independently with their peers.
- Concerns about being different e.g. due to their condition, their lack of independence, their body image etc.
- A perceived loss of control, particularly in relation to an uncertain future.
- A wish to explore and test boundaries, plus the expectation that they can determine and shape their own future with an inherent hope in the future. Yet those with a LLC or LTC will explore this within the limitations of their condition and the foundation of health, on which development into adulthood is built is not there.
- Support to grieve for those things that they may not be able to achieve, for a shortened life, for a lack of independence.
- The development of sexuality, sexual awareness and maturity, and how they are able to express this.
- Mood and behaviour changes linked to deteriorating health as well as adolescent hormones.

**Fig. 18.1** Unique needs of AYA within PC (Amery et al. 2009; Craig and Lidstone 2012)

## 18.5 Question 5: What Are Some of the General Principles for Providing Palliative Care for Adolescents and Young Adults Such as Lucy?

PC should help AYAs to gain some control and perspective to the uncertainties that they face; to develop and work towards realistic goals for the future, whilst at the same time acknowledging and preparing for potential or impending death. This requires flexible, responsive and individualised care embedded in effective communication and partnership with the AYA and their family (Craig and Lidstone 2012). The PC team should aim for an AYA to have the best possible QoL despite their illness; to be clear about their personal identity and gain some freedom from their parents without losing their support through open communication (Weller 1985; Amery et al. 2009).

Finding out what is important to the AYA is key in the provision of PC and enables them to be involved in decision-making, not just for their physical care, but for their lifestyle and decision making. Similarly to asking 'What is important to you?' or 'What does having a good QoL mean to you?', Lucy's PC nurse had asked her 'What do you want to do now?' Through the exploration of what she would like to, and could, do, Lucy was able to get involved in voluntary and advocacy work, specifically supporting the development of PC globally. Despite her limitations, it was this decision that has given Lucy purpose, enabled her to leave a legacy and to feel that, despite everything, she has made a contribution and that her life is valued. Other AYAs may want to continue their education or spend more time with their family. Whilst this might not be possible for all AYAs, trying to explore these areas is essential, and is also a balancing act between being realistic and providing encouragement to explore what is important to them.

Having a social network online can open up a different world to the AYA, as it enables them to access peer support, to make friends and to get involved with things that they would otherwise have not been able to. With the advent of social media and the internet, Lucy now has friends around the world, potentially impacting PC provision internationally.

## 18.6 Question 6: You Are Providing Care for Lucy, What Might Some of the Challenges Be with Regards to Communication?

Whilst recognising that good communication is important in providing PC for AYA, it must also be acknowledged that specific challenges are inherent to communicating with AYAs. Within the PC context there are a wide range of individual AYA, some with full capacity, some unable to communicate verbally although able through other means, and others with severely limited capacities. It is therefore important to individualise the best way of communicating. Establishing rapport and building trust is always core, and this takes time and effort.

A lack of cognitive development can result in reduced understanding of the condition including the consequences of not adhering to treatment. For those with

intellectual capacity, it is hoped that by the time they become AYA they will be aware of their condition and included in decision-making. Supporting carers to discuss some of these issues with the AYA so that they are fully informed is important.

## 18.7 Question 7: What Might Some of the Challenges Be for Providing Palliative Care for a Young Person Such as Lucy?

Providing PC for an AYA such as Lucy can be challenging, and with the growing AYA PC population, health professionals may be '*feeling their way*'. Some of the challenges include:

- Balancing the needs of the AYA and their family members.
- Empowering them to be as independent as possible, whilst they are physically completely reliant on other people.
- Enabling them to be financially independent and/or to make financial decisions.
- Adherence to treatment, medications, physiotherapy, etc. Adolescence is a time of rapid development and uncertainty and it can be hard to follow strict routines, and to understand the implications of not taking the medication.
- A range of psychosocial issues including: dropping out of education, financial problems, accessing health care (particularly in low-and middle-income countries (LMICs)), stigma and adverse changes in body image.
- Sexual challenges, in terms of exploring and understanding their own sexuality.
- They may be more susceptible to physical, sexual or psychological abuse.

## 18.8 Question 8: What Are Some of the Issues Around Decision Making that You Might Experience with Lucy and Her Family?

Decision-making in the provision of PC is complex and the extent of the AYA's involvement varies according to the level of competency they have. For those who are competent, once they have reached the age of 18 years they are legally responsible for making their own decisions, yet may need assistance in understanding available options, consequences of their decisions and implications for the future. In some countries, the ability of an AYA to make an informed decision may be formally assessed, if necessary, through the Mental Capacity Act; in other countries it may be done in a different way or may not even be possible.

Preparation for decision-making by the AYA with regard to treatment and care should ideally start years earlier, with parents encouraged to include them in discussion and decisions from an early age. It is important to identify how the AYA would like to receive information, i.e., individually or as a family group. AYAs may need support to make big decisions about treatment, etc. when some may not be used to

making even the simplest decisions, e.g., what to wear, what time to get up, what time to eat, etc. AYAs may make decisions that go against their parents/carers wishes which may be a way of exercising control: by pushing boundaries. It may also be as a 'punishment' to their parents, or it may be that they no longer want to receive active treatment, e.g., chemotherapy, and really are fully aware of the consequences of their decisions.

**Case Study**

*Lucy's emerging sexuality and sexual identity had not yet been revealed. She had been so removed from the life of her peers that sexuality had not entered her mind, despite being at the age when young people are experimenting, forming relationships,* etc. *It took a researcher entering the family home with support from the hospice to realise there was a part of her life that had been completely neglected. Lucy was supported with age-appropriate information and encouraged to explore this aspect of her life.*

## 18.9 Question 9: Lucy and Her Friends Are at that Stage When They Are Exploring Issues Around Their Sexuality—How Might the Presence of a LLC or LTC Impact Lucy's Expression of Sexuality?

Sexuality is more than just sex, but *'is a central aspect of being human throughout life and encompasses sex, gender identities and roles, sexual orientation, eroticism, pleasure, intimacy and reproduction. Sexuality is experienced and expressed in thoughts, fantasies, desires, beliefs, attitudes, values, behaviours, practices, roles and relationships. …… Sexuality is influenced by the interaction of biological, psychological, social, economic, political, cultural, ethical, legal, historical, religious and spiritual factors'* (WHO 2006). The impact of illness, and possible death, on an AYA's sexuality is vast and many need time away from their parents to discuss issues of sexuality and sexual development (Craig and Lidstone 2012). In some countries, such as in sub-Saharan Africa, AYA may be unable to take part in traditional initiation ceremonies, therefore impacting on their being recognised culturally as adults (Amery et al. 2009).

Many AYAs also have the added pressure that their condition may be degenerative; therefore they have an uncertain future, making it more pressing to ensure that they are supported to feel valued, loved, desired and to have meaningful relationships and/or sexual experiences. They need to be treated as adults and sexual expression is an important part of their overall identify as a human being (Blackburn et al. 2016).

Recent research suggests that there is a gap in the provision of information to AYA with LLCs or LTCs in relation to their sexuality and reproductive choices (Blackburn et al. 2018). Watts and Hameed suggest that '*sexuality is not rocket science, it's part of everyday life…. Yes, talking about sex, intimacy and providing practical support for young people like us can be challenging, but such discussions shouldn't be ignored and swept under the carpet. Staff just need training and*

All young people with a life-limiting or life-threatening condition should expect:

- To have the right to privacy, dignity and confidentiality.
- To be treated in an age-appropriate way, regardless of their developmental stage and mental capacity.
- To be able to address sexuality, intimacy and relationships with freedom from fear, guilt, shame and taboo.
- To be appropriately supported from vulnerability to risk or harm.
- To have the right to discuss, explore and receive relevant information about relationships, intimacy and sexuality, if that is their wish.
- To have their individual needs and views at the centre of care and support, but with information and support provided to their families too.
- To be able to approach professionals to discuss issues of sex, sexuality and intimacy without being judged.
- To have support relating to sex, sexuality and intimacy throughout their life, including early discussions in childhood, as needs change and at the end-of-life (EoL)

**Fig. 18.2** Underlying principles of talking about sex, sexuality and relationships with young people with LLCs or LTCs (Blackburn et al. 2016, p. 4 Reproduced with Permission from the Open University Sexuality Alliance and Together for Short Lives)

*support*' (Blackburn et al. 2016, p. 2). Standards around talking about sex, sexuality and relationships, provide underpinning principles that can help professionals to explore sexuality issues with AYA (Fig. 18.2) (Blackburn et al. 2016, 2017). There are also issues that need to be explored by organisations such as safeguarding and other relevant ethical issues.

**Case Study**

*Over the years, Lucy's condition has progressed and become more fragile and increasingly difficult to manage. Indeed Lucy has repeatedly suffered various complications and battles with sepsis, being lucky to survive. She is dependent on her mother and 24 hour nursing care, which she has been fortunate to receive at home. Whilst still being seen by the young people's hospice, the issue of transitioning to adult services is ongoing and needs to be addressed as does the issue of how to best provide respite care for her mother. Accessing care when her mum was unwell was a challenge, in particular with regard to whether she should receive care from a children's or adult service, with some suggestion that she should be cared for in an old people's home.*

## 18.10 Question 10: How Can Lucy and Her Family Be Supported in the Transition from Children's to Adult Services?

With the increase in the 'unforeseen generation', the need to transition AYA from children's to adult services is growing, and yet services may not be appropriate or available. According to Blum et al. (1993) transition in PC is '*the purposeful, planned movement of adolescents and young adults with chronic physical and medical conditions from child-centred to adult-oriented health-care systems'* (p. 570).

Indicators that an AYA had made a successful transition to adult services include:

1. They feel empowered to take ownership of their lives incorporating health, social, educational, occupational and independent living needs.
2. They feel confident in the team supporting their care, with a single point of contact to address concerns to.
3. They are assured of confidential access to healthcare professionals where they desire it.
4. They understand and take an active role in planning for their future needs including emergency care planning, will writing, advance decisions to refuse treatment, advance care planning and preferred location of care/death where appropriate.
5. They have formalised plans in place for appropriate advocacy/court of protection in line with the Mental Capacity Act when they turn 18.

Key indicators that a family feels that their AYA has made a successful transition to adult services include:

1. They feel confident in the team supporting their young person's care.
2. They are assured of ready access to support in dealing with their young person's condition.
3. They understand and are supported in accepting the changes in their role as the young person enters adulthood and feel supported in 'letting go' and enabling their young person to lead as full and fulfilling an adult life as possible.
4. They have confidence that formalised plans are in place if their son/daughter lacks capacity for decision making which involves them or independent advocacy services.

**Fig. 18.3** Indicators for a successful transition of an AYA from children's to adult services (Chambers 2015, p. 35, reproduced with permission)

*'Transition should be a process, not a single event. When done well it sets the scene for a young person's care in adult services. When it is done wrong, a young patient might not be able to cope and may even disengage from services'* (Watts 2018, p. 1). For many AYAs this can be a scary and confusing time; it can seem daunting and the number of changes overwhelming as they are transitioning from one type of care to another whilst also becoming adults with the responsibilities and expectations that bring.

A systematic review identified a lack of PC specific transition programmes; however, consensus exists on what helps or hinders a successful transition process including the need for multi-disciplinary, individualised care and having a named support worker (Doug et al. 2011). Together for Short Lives has developed a guide to enabling a good transition to adulthood for young people with LLCs and LTCs (Chambers 2015). This aims to provide a generic framework that can be adapted to plan multi-agency services for AYA as they move into adult service provision. The guidelines identify three phases of the transition journey: preparing for adulthood, preparing for moving on and settling into adult services and identifies indicators for a successful transition (Fig. 18.3).

## 18.11 Question 11: What Might Be the Options for Respite Care for Lucy, Other Adolescents and Young Adults and Their Families?

Respite care can be beneficial for the AYA and to their carers, providing opportunities for the AYA to be away from their family, for socialising with other AYAs and giving their families a rest. Ideally this should be available for scheduled respite as

well as for families in emergency situations, such as when Lucy's mother became unwell and was admitted to hospital.

Unfortunately, appropriate respite facilities for AYA are scarce and there is a lack of age-appropriate adult respite services (Mitchell et al. 2016; Knighting et al. 2018). Whilst in the UK some hospices for AYA do exist, these are in the minority (Craig and Lidstone 2012).

**Case Study**

*Lucy's emotions have gone up and down a lot over the past 10 years as she has been living with her condition. One of the challenges she has faced is that of fear—fear of what the future may, or may not hold for her; the fear of ending up trapped in a body where her mind is sharp and she is able to think normally but her body is not working at all and the fear of dying. Yet she has found a way of 'going with the flow', trying to take a day at a time and making the most of it. Several years ago, she decided to try and take some control about the future and, with her family and carers, developed an Advance Care Plan (ACP), so that they would all know what she wants in the future.*

## 18.12 Question 12: Why Is It Important for Lucy to Have an Advance Care Plan?

Goals of care, time frames, parallel planning and discussions around symptoms at EoL and place of death should take place. Many AYAs continue to receive treatments in the last weeks of life. For some this promotes hope and, depending upon the treatments, may result in minimal toxicity and side effects. However, for others, disease directed therapies may prohibit a chance to accomplish activities, spend time with loved ones and visit special places. Lack of communication around anticipated symptoms with AYA and their families can lead to inappropriate and potentially futile invasive interventions. Early engagement with PC service can address anticipated challenging symptoms, provide information and open discussions around ACP and recognise the individual desires of each AYA and their family.

For AYA, such as Lucy, having some control in their care and wishes at the EoL is important, and this can be done through the development of an ACP. This will help to ensure that loved ones and doctors know what the health and personal preferences really are including what care and treatment they would like in different situations, and specifically at the EoL (Advance Care Planning Australia 2019). It is important to plan ahead and introduce this concept early in the care of AYA. The future is uncertain for many AYAs with complex conditions, for whom an infection could cause sudden deterioration in their condition, and therefore being prepared for how to manage this is important. It also helps their families to have discussed the young person's wishes, making it easier to make very difficult decisions.

## 18.13 Conclusion

Providing holistic PC for AYAs is important in order to ensure that they have the best possible QoL, and are able to review their plans, set goals and think about their future. The needs of this emerging population are unique to their age group and services need to be tailored to the differing needs of the population of AYA, in terms of age bracket, and also level of capacity and intellectual ability.

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# Managing Ethical Issues

# 19

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**Key Learning Points**

1. The four common theoretical principles found in principlism are: beneficence; non-maleficence; respect for autonomy; justice.
2. All interventions have more than one outcome. When making a decision to intervene, it is possible to know about all the outcomes without intending all of them, but that only avoids moral responsibility for the outcome if the decision (such as the dose, or the risk of an adverse effect relative to the likelihood of a good one) is proportionate to what was intended.
3. Justice is a universal principle that is equally applicable in countries that are resource rich and those that are not.
4. Some children will always die from cancer. Justice demands that maintaining the quality of a child's life, once cure from cancer is highly unlikely, should be given the same financial priority as attempts to treat it while cure seems possible.

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**Case 1**

*Louie presented to hospital at 6 weeks of age with a seizure. On examination it was noted he had decreased central tone, head lag and nystagmus. Over the following days he started having more seizures which included infantile spasms confirmed with electroencephalogram (EEG) findings. Radiographic imaging was normal. He was started on antiepileptic and steroid medications. One night he had a seizure, which resulted in a prolonged apnoea. He was intubated and ventilated and admitted to paediatric intensive care (PICU).*

*Diagnostic tests including gene panels, and tests looking for mitochondrial disease were reported as normal. Nevertheless, in PICU Louie developed severe respiratory failure, and epileptic encephalopathy. A further EEG showed no normal brain waves and repeat Medical Resonance Imaging (MRI) scans revealed severe progressive neurodegeneration. Eventually the PICU clinicians explained to Louie's parents that, in their view, Louie was no longer benefiting from ventilation. They suggested turning off the ventilator in order to allow Louie to die a 'natural death'. His parents were shocked and angry and expressed the view that all life sustaining treatment (LST) should continue. They wanted 'everything done for him', including a tracheostomy and gastrostomy so they could care for him at home until a diagnosis and possible treatment were found.*

*Social work and psychological services were involved in providing parental support. Louie remained in PICU but the clinicians continued to feel they were doing more harm than good. The bedside nurses in particular found continuing ventilation distressing. The clinicians sought advice from the clinical ethics and legal teams in their hospital and continued to try to work alongside Louie's parents; however, they had lost trust in the clinicians, and felt unable to engage with them. Instead, they turned to social media to gather financial support to take Louie to Mexico where they had found a clinician who was prepared to provide the treatment they were requesting. Unfortunately, in the meantime the clinicians had come to the view that prolonging LST with no prospect of cure was actively harming Louie and they could therefore no longer agree to it. The hospital commenced legal proceedings in order to obtain the Court's permission to turn off the ventilator.*

## 19.1 Question 1: Why Is Medical Ethics Important in the Care of Babies Such as Louie?

Medical ethics underpins our commitment to serve others to the best of our ability in a way that is morally right. It is the backbone of decision-making, offering guidance as to the right decision within the grey areas that medical complexity can cause. Medical ethics addresses morals, principles and values; it has cross-over between philosophy and law. Ethical dilemmas are particularly likely in children's palliative care (CPC) because of the sensitive nature of the cases, the vulnerable population and the intimate relationship with end-of-life (EoL) care. It is therefore important to have a thorough understanding of bioethics and how to approach these issues, should they arise in clinical practice.

Hospital clinical ethics committees support clinical teams in making morally challenging decisions (Whitehead et al. 2009; Johnson et al. 2015; Thomas et al. 2015; McDougall and Notini 2016; Jansen et al. 2018). They represent a valuable resource, but they are not yet available everywhere and those caring for the patient 'at the coalface' often need to be simultaneously clinician and ethicist. This chapter is a practical guide, addressing three issues: withholding and withdrawal of LST, the principle of double effect (PDE) and inequalities of resource allocation, offering the reader the tools needed to analyse bioethical issues when they occur within the reader's own setting.

## 19.2 Question 2: What Are the Ethical Issues in the Care of Louie?

Medical technology and treatments are capable of supporting life far beyond what was possible only a few decades previously. Whilst it is accepted there is great benefit from access to these treatments, having them at our disposal comes with difficult decisions on where to use them and who would benefit, when to start, or stop the technology and the cost (emotional, physical and social) to the child, family, society of providing this level of support.

Louie was given LST in the PICU, but medications, procedures or surgeries that extend life without curing the disease are also considered LST, therefore this ethical dilemma can also be applied to those countries with limited resources.

## 19.3 Question 3: What Are the Principles We Use to Explore the Ethical Issue of Withdrawal and Withholding Life-Sustaining Treatment?

When exploring the ethical issues at play in this case, firstly look at it using an analysis from *principlism*. *Principlism* is the best-known way of considering ethical dilemmas (Beauchamp and Childress 2013). It sets out four common theoretical principles (Table 19.1):

To determine what options are ethically acceptable, the circumstances of Louie's case are explored and weight is given to different aspects of the principles that apply to the case (Ko and Blinderman 2015). Louie clearly lacks autonomy and his

**Table 19.1** The four common theoretical principles found in principlism (Beauchamp and Childress 2013)

| | |
|---|---|
| Beneficence | Duty to act in a way that provides benefit to the patient. |
| Non-Maleficence | Duty to avoid doing harm. |
| Respect for Autonomy | Self-determination, freedom to choose rationally one's own path. |
| Justice | Principle of equity and fairness—in healthcare, usually in resource allocation. |

parents' autonomy is only over their own decision-making; it does not extend to making medical decisions about their child. In the absence of an autonomous decision-maker, the difficult task facing the medical team and Louie's parents is to set aside their own preferences in order to establish in dialogue what is in Louie's own interests (i.e., beneficence and non-maleficence). The clinicians believe ventilation is harming him with no benefit and stopping the ventilator to avoid further suffering is the 'right' thing. They are also considering Louie's future quality of life (QoL), where it is unlikely he will recover any brain function, or come off life support, and be unable to have any interaction with his world, meaning there is little benefit from ongoing technology use while the suffering associated with continued use is doing him harm.[1] His parents, on the other hand, feel that he is benefiting from continuing to exist and from being able to spend time with his family. Furthermore, stopping the ventilator means they will lose him earlier than is necessary, which unacceptably damages their own interests.

The demands of beneficence and non-maleficence to Louie have no room for the needs of other children. Using a PICU bed takes opportunity away from other children and the resources that it takes to keep one child alive could potentially save many children. Parents should not of course be expected to prioritise the needs of other children over those of their own child, but for clinicians, resource allocation is a necessary matter that cannot be ignored (Wilkinson and Savulescu 2019). The demands of justice appear to be potentially in tension with the demands of beneficence and non-maleficence. The principlism approach therefore does not go far enough to tease out such complexities (Fiester 2007; Hain 2015).

Alternatively, it is helpful to have a framework of questions to deconstruct all aspects of the case (Table 19.2), then thoroughly discuss them amongst team members (including ethical committees) in the hope that a considered opinion can be given. Making decisions when it comes to life-sustaining treatments is complex yet essential. It requires hard work and committed deliberation and needs to be done well. Communication and relationships are fundamental. Difficulties arise when there are mismatched perceptions, beliefs and opinions between health professionals and parents.

Unfortunately, this type of case has not been uncommon in recent years (Francis 2017; Alder Hey Children's NHS Foundation Trust 2018; Kings College Hospital NHS Foundation Trust 2018). Disagreements were heavily entrenched and resulted in protracted court proceedings. Honest communication about goals, benefits and burdens (Ray et al. 2018), ethical committee involvement and CPC early in the relationship can help prevent this fallout (Linney et al. 2019; Nuffield Council of Bioethics 2019).

[1] *Predicting future quality of life is fraught with difficulty. The prediction of poor life quality arising from disability by clinicians has led to withdrawal of LST in 20–40% of neonates in the intensive care unit* (Verhagen et al. 2010; Hellmann et al. 2016) *despite the fact that clinicians' perception of disability is known to be considerably worse than that of children and families living with disability.*

**Table 19.2** Framework of questions to deconstruct aspects of each case

| | |
|---|---|
| *Medical facts* (*beneficence/non-maleficence*)<br>– Diagnosis<br>– Goals of treatment—cure/relieve symptoms/stabilise/prevent untimely death/prevent harm/promote health/prevent disease<br>– Prognosis<br>– Treatment options<br>– Are any treatments not indicated<br>– Probability of success—evidence<br>– Risks—physical/emotional/social<br>– Expected clinical course if withheld/withdrawn?<br>– How can patient be benefitted and harm/risks prevented?<br>– Suffering—unrelieved vs. relieved?<br>– Clinicians gut instinct (to check bias) | *Patient preferences* (*Autonomy*)<br>– Have they been involved in decision-making?<br>– Do they have capacity?<br>– Have they stated preferences—do the treatment options align with their wishes and values?<br>– Is there a surrogate—acting on their best interests? |
| *Quality of life (QoL)*<br>– Current QoL?<br>– Anticipated QoL—worse vs. best case scenario<br>– Our own beliefs about their QoL (rule out clinician bias) | *Contextual features* (*justice/fairness*)<br>• Other parties who have interest—stakeholders (patient, doctor, other patients, public, government)<br>• Any other stakeholders have conflict?<br>• Other factors<br>– Confidentiality<br>– Allocation scarce resources<br>– Religion<br>– Legal issues—statutory case law<br>– Research education<br>– Public health safety<br>• Rights<br>– Claim rights<br>– Liberties<br>– What does patient rights require of treating team—obligation vs. duty<br>• Values<br>– Morals<br>– Sanctity of life<br>– Compassion<br>• Literature—any comparable cases to draw conclusions from |

Adapted from Siegler (1982), Doukas (1992), Kaldjian et al. (2005), Ko and Blinderman (2015)

## 19.4 Question 4: How Can the Clinical Team Assess and Manage Parental/Clinician Conflict?

In order to examine conflict, there firstly needs to be consideration about why the decision-makers disagree: parents and clinicians come with their own motives (Table 19.3). Careful inquiry into these may bring you closer to a mutual understanding.

**Table 19.3** Parents and clinicians motives in decision-making (Adapted from Pope and Waldman 2007; White and Pope 2016)

| Parents | Clinicians |
|---|---|
| Distrust. | Avoidance of patient suffering. |
| Cognitive issues—understand fully medical aspects of the case? | Respect for patient autonomy. |
| Psychological/emotional—how is the stressful event impacting on their decision. | Protect integrity of medical profession. |
| Spiritual—religion and miracles. | Avoid moral distress. |
| Invisible decision-makers—social media impact, family? Are those feeding into the present decision? | Promote good stewardship. |

Moral distress is defined as an unpleasant emotional experience arising from a perceived moral tension. This was a large issue in Louie's case in view of his perceived ongoing suffering, especially from the bedside nurses. It is known to lead to burn out and compassion fatigue (Simpson and Knott 2017) and interestingly involving ethical services may be a therapeutic way of reducing moral distress (Jansen et al. 2018).

Once a decision is made, it is helpful to consider whether the decision lies within the 'Zone of Parental Discretion' (ZPD). The ethical idea of the ZPD authorises parents to become key decision-makers over their child's medical treatment, but only within a medico-legal framework in which the ultimate test is the interests of the child (Gillam 2016; McDougall et al. 2016). The ZPD is important as it recognises that, while the preferences of parents must not be allowed to cause serious harm to (or withhold serious benefits from) the child, there might also be harms to the children if doctors do not act on their expressed preference.

Ultimately, using these frameworks and understanding the core beliefs of those involved will go some way to resolve the disagreement or considering the ZPD in order to provide the best moral/ethical/compassionate care to the child and family may avoid the judiciary system which although is necessary at times, can add another level of harm.

**Case 2**

*David is a 17-year-old boy with Combe's Myopathy, a progressive neurological condition which over many years weakens the respiratory muscles, resulting in three unpleasant symptoms of dyspnoea, fatigue and drowsiness. A medication, Siarpamec, helps relieve some of those symptoms. Siarpamec is given via a face mask in the same way as oxygen; its major benefit is that it can therefore be titrated very precisely against the patient's symptoms using the tap on a gas cylinder valve. Siarpamec has several different effects. The drug carries a very high chance of relieving fatigue. Its effectiveness in helping dyspnoea is slightly less, but still extremely likely. Quite often, Siarpamec makes patients drowsy. There is a tiny risk that Siarpamec will stop a patient's breathing.*

*David has been admitted to the children's hospice for EoL care. He is experiencing all the symptoms of Combe's myopathy and the consultant in paediatric palliative medicine at the hospice prescribed Siarpamec 'as needed' for fatigue and dyspnoea. David is too weak to control the valve himself, but he is able to communicate with a member of the care team whom he knows well. Over his last few days, David needs gradually increasing doses of Siarpamec in order to control his symptoms. One day he complains to his carer that he is still feeling short of breath. His carer accordingly turns the Siarpamec up a little higher. David's breathing stops, and he dies.*

## 19.5 Question 5: Was the Carer Responsible for David's Death?

Every action we decide to take has many different outcomes. The PDE expresses the fact that we can intend some of the effects of our action, but that there are also effects we know are possible but hope will not happen. In David's case, it seems intuitively likely that although David's death occurred as a result of his carer's action, she was not to blame for it because that was not what she intended. Her aim was to relieve his suffering.

Medical ethicists who object to the PDE are concerned that it can be used to justify medical decisions that are, in fact, wrong (Glover 1977). They worry that, if clinicians are allowed to excuse their actions on the grounds that they intended something other than what happened, they are not having to take enough responsibility for their actions. Ethicists are right to identify that risk: in the last few years in the United Kingdom (UK) there have been cases where opioids have allegedly been prescribed in order to hasten death, using the excuse that it was impossible to be sure patients were not in pain (Oliver 2018) and cases where fluids seem to have been withheld in order to hasten someone's death, using the excuse that in the last 48 hrs of life fluids might make their symptoms worse (Neuberger et al. 2013).[2]

Nevertheless, there is no getting away from the fact that all actions have multiple effects, and there is no denying that it is possible for someone to intend some of those effects and not others. It is also certainly true that knowing that something *might* happen as a result of your actions is different from setting out with the intention that it *should* happen. Whatever its risks in practice, the PDE is logically unavoidable.

[2] *The problem was not with the Liverpool Care Pathway itself but with the way it was misused by some clinicians. It is true that fluids in the last 48 hrs of life can make some symptoms worse, and it is often right to discontinue fluids for that reason. But that is different from inducing dehydration with the aim of hastening death.*

## 19.6 Question 6: What Additional Information Would You Like to Have in Order to Help Decide Whether the Carer Was to Blame for David's Death or Not?

It was philosopher Thomas Aquinas in the thirteenth century who provided a solution (Fohr 1998). Aquinas recognised that double effect is a reality that cannot be escaped. Like modern bioethicists he recognised the danger that it could be used to justify immoral acts but, unlike some of them, Aquinas also acknowledged that intention is at the heart of what makes action moral. Aquinas clarified that in order for it to be clear that someone intended good even if the outcome was bad, three conditions had to be met:

(a) The *ultimate intention* of the act must be good.
(b) The effect that you *hope will* happen must result just as *directly* from your action as the result you *know might* happen.
(c) The action you take must be *proportionate* to the outcome you intend, rather than to the one you are claiming only to know is possible.

David's carer would have to fulfil all those three conditions in order to show that she did not intend to kill David even though she knew that apnoea was a risk when she increased the Siarpamec.

(a) *The* ultimate intention *of the moral act must be good.* The aim of the carer must, of course, be good in the first place. If it later became clear that his carer had posted on Facebook that she planned to kill David, there would be no point in her invoking the PDE to claim she was trying to help him.
(b) *The effect that you* hope will *happen must result just as directly from your action as the result you* know might *happen.* David's carer can invoke the PDE by saying that her intention was to relieve David's symptoms even though she knew death was possible. She cannot use the PDE to argue that killing David was justifiable because, by causing death, it resulted in resolution of David's symptoms.[3]
(c) *The action you take must be* proportionate *to the outcome you intend, rather than to the one you are claiming only to know is possible.* The golden rule of pharmacological therapeutics in CPC is that 'the right dose of a drug is the lowest dose that will do the job'. That is because the beneficial effects of any medication are always accompanied by the risk of some adverse effects. Once the beneficial effects are achieved, any further increases in dose cannot offer any further benefit, but they do run the risk of adverse effects which harm the patient. That risk cannot be justified if the same benefits can be achieved without it.

[3] Advocates of euthanasia who argue that hastening death is a way of alleviating suffering are making a different argument that does not invoke the PDE at all. They are saying that the intention to kill can be good (Hain 2014).

If it became clear that David's carer had given a higher dose of Siarpamec than was necessary to control his symptoms, it might give rise to some suspicion that she intended to kill him, rather than to help him. If the dose of Siarpamec that David was receiving at the time he died had been reached gradually, and in response to documented changes in his symptoms, then the carer's intention to give only the dose of Siarpamec that is appropriate is clear. If, on the other hand, there is no clear relationship between David's symptoms and the dose of Siarpamec the carer administered, then it becomes hard for the carer to justify taking the risk of stopping his breathing.

## 19.7 Question 7: Can the Principle of Double Effect Be Used as an Excuse for Shortening Lives?

Families often fear that CPC clinicians will use the treatment of symptoms as an excuse for shortening lives. Physicians in CPC constantly find themselves using medications that they intend to relieve suffering, knowing that the medications also carry the risk of harm. They must ensure that their therapeutic reasoning is both well-considered along the lines Aquinas has set out, and that it is well documented:

- Reassure families that hastening death is not part of CPC.
- Set out and record clearly what the intention of an intervention is.
- Never use more of a medication than is justified by the patient's symptoms.

The PDE is an important way of thinking for clinicians in CPC that has important everyday implications for the way we practise. Bioethicists are right to suggest that it is all too easy for the PDE to be misused, but they are quite wrong if they think it is an unimportant 'fudge factor' that logicians can dispense with.

**Case 3**

*Amelia is a 12-year-old girl seen in a paediatric oncology centre in a university hospital in Romania and diagnosed with fibrillary anaplastic astrocytoma localised to the brain stem, with extension in the left cerebellar hemisphere. Due to its location, the risks of surgery were too great and she was treated with chemotherapy. Ten months later she was admitted for radiotherapy but was found to be suffering from raised intracranial pressure and a ventriculoperitoneal (VP) shunt was inserted in order to reduce the symptoms related to this.*

*Two months later, she presents after further deterioration with hemiparesis, multiple cranial nerve palsies (including poor swallow, paresis of the ocular muscles, cerebellar ataxia and hypopnoea). Despite tarsorrhaphy,*[4] *she has injuries to her left cornea as a result of a paralysed eyelid. She has a tracheostomy for respiratory*

[4] *Tarsorrhaphy is a surgical procedure in which the eyelids are partially sewn together to narrow the eyelid opening.*

*failure, naso-gastric tube for feeding and an indwelling urinary catheter. She has painful pressure sores in the right leg.*

*During the admission she develops multiple resistant infections of the chest, urinary tract and septicaemia and is treated aggressively with intravenous antibiotics and antifungals. Due to the degree and complexity of her suffering, the hospital team feels they cannot care for her and decides to refer her to a specialised CPC service, but the closest is 300 km (190 miles) away. There are concerns about the discomfort caused by road transport over such a long distance. Furthermore, the move would take her even further from home. The CPC team accepted the patient, but at the last minute the oncology team withheld approval for ambulance transportation on the grounds that it would be too expensive. Amelia remained in a side room on the oncology ward until her death.*

## 19.8 Question 8: Why Is the Principle of Justice Important in Amelia's Case?

Justice is one of the four pillars of the principlism approach of Beauchamp and Childress (2013). Common to almost all theories of justice is the idea that like cases should be treated alike. In healthcare, the term is usually used to refer to *distributive justice* which refers to fair, equitable and appropriate distribution in society.

The ethical principle of *distributive justice* underpins questions of resource allocation. It presents a challenge to those responsible for allocating healthcare resources globally. Notions of distributive justice vary across cultural, societal and even individual norms, with some definitions allowing for discrimination based on merit or need. Yet the principle of distributive justice itself is universal; it is not specified to any one state, culture or society. Article 27 of the United Nations Convention on the Rights of the Child (UNCRC) states that children everywhere should be treated with dignity and should be offered 'a standard of living adequate for the child's physical, mental, spiritual, moral, and social development' (UN 1989; Ko and Blinderman 2015). That such a right should extend to palliative care globally was set out in 2014, when the World Health Assembly resolution (WHO 2014), called upon Member States to improve access to palliative care as a core component of health systems including CPC (WHO 2018). There is nothing in these claims to indicate that they are rights in some regions of the world but not in others. In practice, however, there are many low-and middle-income countries (LMICs) in which children continue to have no access to adequate PC (Wright et al. 2008; Connor and Sepulveda 2014) despite the fact that over 60% of all cancer deaths worldwide occur in such countries (Hadler and Rosa 2018; WHO 2018).

Nor is there anything in those claims to suggest that a child's right to 'best care' stops once cure is no longer possible. For children whose lives cannot be saved, 'best care' is represented by good quality PC rather than by futile attempts to cure, and that right means that governments have a duty to ensure everyone who needs PC has ready access to it. There is a risk that, in the face of limited resources, palliative and other basic forms of care are seen as lower priority for investment than

technologically advanced interventions that are aimed at cure, even when the equipment needed for the intervention is expensive and the intervention itself of little benefit. That would be a clear transgression of the principle of distributive justice. It is the responsibility of governments to ensure that good quality CPC is accessible to all who need it. The principle of justice demands that in every country there be a coherent national policy framework supporting comprehensive access to CPC so that children such as Amelia can access such care.

## 19.9 Question 9: Whose Responsibility Is It to Offer Children's Palliative Care (CPC) for Children Such as Amelia?

CPC should be an integral part of every Health Committee and should be an element of general health plans, and of specific health programmes such as cancer, Human Immunodeficiency Virus (HIV)/Acquired Immune Deficiency Syndrome (AIDS) and metabolic conditions. The implementation of government programmes should ensure ready access to CPC in any health system where there are children with life-limiting conditions (LLCs) of any kind (Torales 2012). Financial policy reforms may be necessary in order to ensure that CPC programmes are integrated into wider healthcare programmes and are sustainable in the longer term (Williams-Reade et al. 2015). One example of governmental support is exemplified in Albania, where a law was passed in October 2014, asserting that multidisciplinary PC is a basic human right. Among other things, the law allows better access to opioids in the country, seeks to include PC in the state's welfare scheme and enshrines a requirement to provide education and training in specialist PC (Rama et al. 2018). In many LMICs, however, no such laws are yet in place and the clinician can be faced with difficult judgements about resources. The practitioner must be able to make those decisions based on solid knowledge of medicine, their professional code of ethics, an understanding of the respect for autonomy and justice and insight into the influence that personal beliefs and values might exert in the decision-making process (Edwards et al. 2011).

## 19.10 Question 10: When It Comes to Resource Allocation, What Factors Are Relevant in Deciding How to Prioritise Care for Children Who Can Be Cured Against Those for Whom Cure Is No Longer Possible

Resource allocation in LMICs is complex, and throwing into the mix vulnerable children where no further curable options exist means considering a variety of factors: the society in which they live, the population requiring those resources, cost, benefit vs harm of resources, accessibility and cultural expectations. Providing clinical care to children with LLC in resource-poor settings often demands complex trade-offs regarding resource allocation. That is particularly true in conditions such as cancer that are potentially curable, because the cost of improving PC provision for the 20% or more of children who will not survive needs to be weighed against

the cost of potentially increasing the number of children who might be cured. That does not mean, however, that as the disease progresses priority must always be given to the ever-dwindling possibility of cure. For every child who dies from cancer there comes a point in their treatment where it is clear that the chance of cure is so low that some interventions may not change the outcome. Under those circumstances, neither the cost of the intervention nor the harms it will cause are justifiable and the intervention should be withheld in the names of both compassion and justice (in regard to its use be best spent elsewhere). Supporting good quality PC will be the best care that can be offered in this regard.

Additionally, allocating resources requires consideration and sensitivity of cultural needs. It is important to work within cultural norms to improve communication, connectedness and PC outcomes (Marston 2015). Hadler and Rosa (2018) go so far as to suggest that the absence of such sensitivity threatens justice itself: distributive justice *'in the absence of cultural humility or a genuine willingness to understand decision-making priorities in a given culture can contribute to further inequalities'* (p. 1237).

Specialised CPC should be available even if it is not practical for the child to leave the oncology ward, as in Amelia's case. CPC has been delivered very effectively by some oncology hospitals in Romania and other LMICs. Even if specialist CPC is not available on site, potentially the adult PC team can offer support to the oncology staff, and the oncology ward and can set aside one or more rooms in order to offer a child who is dying comfort and privacy in the last days of their life.

## 19.11 Conclusion

Practical paediatric bioethics is an essential component of clinical practice. Its importance is particularly clear for those working in CPC. With growing use of technology and many children with chronic illness living longer than only a decade ago, it seems likely that our specialty will increasingly be faced with these ethical dilemmas.

Establishing a child's interest in the course of sensitive dialogue with parents is key to acting correctly when considering treatments that are potentially life-sustaining. Considering carefully what the aim of an intervention is, and what is the least harmful way in achieving it, are the hallmarks of compassionate medical care. Resource allocation is profoundly inequitable, not only between countries but also often between specialties in the same country. PC is often underfunded because it does not save lives. However, it can transform the lives of those who have to die, and an ethic of justice and compassion demands that the vulnerable should not be condemned to unnecessary suffering.

The 'principlism' structure of beneficence, non-maleficence, autonomy and justice is helpful in considering clinical ethics, but in cases of complexity principlism alone often cannot offer an adequate analysis. Personal values, such as compassion and empathy, harnessed to practical expertise in communication and analytical thinking, are key to preventing conflict. Those are the skills our

colleagues should expect from practitioners in CPC, and it is essential that in cases that are ethically complex at the EoL, PC is amongst the multidisciplinary teams involved.

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# Collaborative Working and Use of National, Regional and International Networks

20

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**Key Learning Points**

1. Despite distinct difference in access to resources, it is evident that irrespective of geographic location, the needs of children and families are similar and palliative care (PC) services face comparable challenges.
2. Communications are critical to the success of any network, including keeping up with new digital technologies. Being consistent in the tone of voice and messaging, having clear policy asks, producing good quality

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evidence-based resources provide a platform from which a national, regional and international network can speak with authority on behalf of its members.
3. Managing mutual patients with colleagues across a region brings positive outcomes and inspires an appreciation for collaborative work.
4. It is important to allow children and families to guide a network's decision-making, ensuring that their needs are at the heart of the organisation. Reaching out to a local, regional or international community of like-minded colleagues can improve the care of children and families, provide solutions, wisdom, understanding and personal and organisational growth.

**Case 1**

*In 2017, 11-year-old Makuta, originally from the Democratic Republic of Congo (DRC), travelled to the United States of America (USA) with his father and brothers while his mother and four other siblings remained in a refugee camp in Zimbabwe.*

*In April of 2018 Makuta was admitted to a hospice unit in Atlanta with end stage cancer and a desperate wish to see his mother and siblings again. The children's palliative care (CPC) team in Atlanta providing PC to Makuta and his family reached out to international colleagues. With the collaboration of the United Nations High Commissioner for Refugees (UNHCR), Island Hospice and Childline in Zimbabwe, his mother was located and transported to Harare where she was able to hold several video calls with her dying son. Sadly, despite interventions by UNHCR and even a Hollywood star, it proved impossible to physically reunite the family.*

*After Makuta's death, Island Hospice provided his mother and siblings with bereavement support and the Atlanta hospice did the same for his family in the USA. A few weeks later, the CPC Nurse leading the team caring for Makuta in Atlanta carried a precious letter from him to the 2018 International Children's Palliative Care Network (ICPCN) conference held in South Africa. Here she passed it on to a social worker from Zimbabwe who was able to hand-deliver the letter to Makuta's mother.*

## 20.1 Question 1: Why Is Collaboration So Important in Children's Palliative Care?

Collaboration is key to the provision of PC for children, not only at the service delivery level but also at national, regional and international levels. Makuta's story illustrates the value of connection and collaboration across all these levels. We live in a time when it has never been easier to access available networks to ensure the best possible quality of care for a child and family. This chapter provides insight into the work and achievements of a national, several regional and an international network. All supporting and promoting the development of PC for children through connection and collaboration, with each providing a case study in their own right.

**Case 2: A National Association—Together for Short Lives**

*In the United Kingdom (UK) there are at least 49,000 children living with a life-limiting (LLC) or life-threatening conditions (LTCs). As a high-income country with some of the longest-standing CPC services, the need at national level is bringing together the broad range of services so that care is planned and delivered effectively and in an integrated way and ensuring that funding for these services can be sustained.*

*Together for Short Lives formed in 2011 as the result of a merger between the Association of Children's Palliative Care (ACT) and Children's Hospices UK. It had become apparent that there was a need for one voice to represent children with LLCs or LTCs and their families and the two charities came together to facilitate this.*

## 20.2 Question 2: How Does Together for Short Lives, as a National Network, Directly Support Families?

Together for Short Lives provides high-quality information to families through printed and online resources and a telephone helpline, offering a listening ear and signposting to services that can help locally. Families are encouraged to engage further with the charity by joining a family 'community' which provides a newsletter and a national peer-support group for families going through the same challenges and heartbreak of caring for a life-limited child. Families can also join a closed Facebook Group, moderated by parents, to share their experiences. Some families choose to become active advocates, with Together for Short Lives providing important opportunities for them to share their stories in the media, in parliament and through public awareness campaigns such as the national Children's Hospice Week. One mother wrote:

> We are blessed in getting support from lots of great organisations. We've found lots of useful information from Together for Short Lives. Their family newsletter, Together for Families, showed me that our family isn't alone and opened up a network of families in a similar position. This is invaluable, and it gives me the time I need to get my head around everything.

## 20.3 Question 3: What Does Together for Short Lives Do at a National Level for Children's Palliative Care Professionals and Services?

A key role of a national network is to bring together a shared vision and a framework for good practice for CPC that is relevant for that country. Together for Short Lives has produced numerous publications, probably the best known being the Guide to Children's Palliative Care (Chambers 2018) which sets out a blueprint for CPC that can be used as an advocacy tool in a range of settings. Several child and family-centred care pathways have been developed to provide frameworks for

services to adapt to make them locally relevant (Widdas et al. 2013; Chambers 2015; Dickson 2017).

Enabling sharing of good practice and knowledge is another key function of a national network, enabling practitioners to come together and share their expertise. This has been achieved through traditional activities such as workshops, seminars and national conferences, but also through online forums such as the 'PaedPalCare' (Together for Short Lives n.d.-a), an email list enabling professionals from all over the world to share and respond to challenging cases. This is especially valuable in an emerging field in which children have a range of rare conditions and unusual symptoms. There is potential now to expand this forum, using new and more effective digital solutions.

There is a key role to play in advancing the development of CPC through building and sharing the evidence base. Together for Short Lives has hosted a Joint Research Group with the Association of Paediatric Palliative Medicine since 2002. The group brings together leading academics and clinical researchers from across the country to address barriers to research in CPC. A collation of all published research is pulled together twice a year in a publication called Synopsis (Together for Short Lives n.d.-b).

Together for Short Lives has recently entered an exciting new programme of work, by running its first major national grant programme, with the theme of transition from children to adult services, to stimulate innovative practice, evaluate and share what works.

## 20.4 Question 4: How Does Together for Short Lives Collaborate Regionally and Internationally?

It has always been an important part of the charity's ethos to work collaboratively, not only with other national bodies in the UK (such as Hospice UK or the Association of Paediatric Palliative Medicine) but also with regional groups such as the European Association for Palliative Care (EAPC) and internationally with the ICPCN (http://www.icpcn.org/about-icpcn/). This not only enables the development of CPC in other countries, but also brings back rich learning to be shared with organisations in the UK.

**Case 3: A Regional Association—Asia Pacific**

*Dr. Lee is a paediatric oncologist in a Chinese city hospital. Even though state-of-the-art modern medicine is available to almost everyone, when Wang Wei's (9-year-old boy) neuroblastoma fails to respond to treatment, there is not much else she can offer as the primary physician. The concept of CPC has only just crept into the system. Structural and psychological barriers that are prevalent mean only very few practitioners provide this support. On recommendation from other parents in their 'Neuroblastoma families' chat group, Wang Wei's parents brought him to Singapore for further treatment. Unfortunately, after one year of intensive second- and third-line treatments, the disease proved refractory and he was referred to a local community hospice supporting children.*

## 20.5 Question 5: Is This Situation Common Across the Asia Pacific and How Can the Regional Network Provide Support?

Countries within the Asia Pacific region are in various stages of administrative and economic development, impacting access to healthcare. The outreach of CPC is oftentimes influenced by how adult PC services develop. A service mapping study found that CPC in this region is mostly provided by adult trained practitioners who support both adult and paediatric patients (Chong et al. 2017), with a lack of dedicated CPC training opportunities. That said, there are also centres classified as 'mainstream providers' (Knapp et al. 2011), e.g. in Australia, so there are great disparities within the region.

The Asia Pacific Hospice Palliative Care Network (APHN) was formed in 2001 to promote the development of hospice and PC in the region. A few practitioners came together in 2015 at one of its biennial scientific conference in Thailand to form a CPC special interest group (SIG). Even though health systems differ, it soon became clear that countries shared almost similar learning needs and practice challenges. The SIG decided that consolidated efforts in advocacy, education and collaboration were needed for CPC within the region to move forward.

**Case Study**

*After Wang Wei's distressing symptoms were controlled, the goals of care were discussed with his parents. The PC physician on the home care team confirmed the estimated prognosis of weeks to short months given by the treating oncologist in Singapore. Wang Wei's grandparents were consulted on video conference. A final decision was eventually made. The family wished to take him back to China. Fortunately, they lived in the city and would be able to access blood transfusions or other interventions if needed, though they were told before that the hospital would hesitate to admit Wang Wei, as his disease was no longer treatable.*

*The CPC physician checked the APHN service directory on its website for contacts in China who provide support to children. He managed to find one in the vicinity (a colleague of Dr. Lee) and made contact for continuation of care. The parents were relieved to know that some assistance at least was available back home. They were initially prepared for Wang Wei to die in a foreign country, as their boy's comfort was of highest priority.*

## 20.6 Question 6: How Can a Regional Network Improve the Care for a Child Such as Wang Wei and His Family?

With the support of a family foundation based in Singapore, APHN started the *Lien Collaborative for Palliative Care*, to develop PC services in countries within the Asia Pacific. Its three prong objectives of training leaders, engaging policy makers and ensuring access to essential medications have already seen results in three developing countries (Goh and Lee 2018). The impact of spill over effects into CPC is obvious.

To provide greater opportunities for education in CPC, the APHN collaborated with colleagues in the States to bring *Education in Palliative and End-of-Life Care* (EPEC)—Pediatrics (Friedrichsdorf et al. 2019) into the Asia Pacific. This is a blended learning programme curated by leaders in the field of CPC consisting of virtual lectures and face-to-face teaching sessions. A few editions have been organised, and a Chinese version is being planned to reach an even wider audience.

The Asia Pacific Hospice Conference has been the most successful platform to bring people together. With the dearth in training opportunities, practitioners in some Asia Pacific countries look forward to each meeting. Experts from around the world are always invited to share best practice and teach fundamentals. This is also where the CPC SIG becomes even more active, consolidating and setting new targets for the next 2 years.

## 20.7 Question 7: What Are the Challenges Facing the APHN?

Language, systems and culture across a region can create numerous challenges, with language being a major obstacle. Partnerships have been difficult to forge due to a varying command of English in different settings. Challenges go beyond introductions at meetings to communication in texts or emails, where much can be lost in translation. Limited success has mostly come through support from members who speak the local vernacular. Within the global economy that is supposed to bring the world closer, governments can impose restraints on the internet that limit communication through social media or chat groups. Experience has proved that culture, in and of itself, can pose unimagined barriers between practitioners, even as they serve the same mission.

## 20.8 Question 8: Is There a Regional Association in Europe and What Is the Situation for Children's Palliative Care in the Region?

The European Association for Palliative Care (EAPC) is the regional PC association and it has a children and young people's task force. Across Europe the availability of, and access to, CPC services varies hugely. Some of the best developed services for children globally exist in Europe. In the UK, for example, PC is recognised as a paediatric medical and nursing specialty and a range of services exist to support children and their families. Paradoxically, many other countries in Europe have very limited provision of PC for children, and in several countries, access to even the most basic medicines to treat pain and other symptoms can be challenging. Where PC services have been developed, this has often been achieved by several CPC pioneers who have been instrumental in establishing services in their country.

A systematic review published in 2011 identified that of 43 European countries, 33% had no known CPC activity. CPC was found to have reached some degree of integration with mainstream healthcare services in only 12% of countries (Knapp

et al. 2011). The EAPC Atlas of PC in Europe, published in 2019 (Arias-Casais et al. 2019) shows that there has been 'vigorous developments' in PC, including CPC, although there is still a way to go to provide integration throughout the region.

## 20.9 Question 9: How Does the EAPC Support Health Care Professionals Providing Children's Palliative Care?

The EAPC is a membership organisation that aims to advance, influence, promote and develop PC in Europe across the life span, thus recognising the importance of CPC. The EAPC supports health care professionals caring for children by providing high-quality guidelines and white papers on a range of relevant aspects. In 2007, members of the EAPC children's task force produced the highly regarded IMPaCCT standards for CPC in Europe (Craig et al. 2007). These European standards identified best practice and agreed minimum standards for the provision of CPC.

In 2014, the EAPC CPC task force developed core competencies for education in CPC (Downing et al. 2014). This white paper provides a framework for multidisciplinary curriculum development for CPC and provides recommendations and guidelines for the ongoing development of initiatives in relevant training and education for healthcare professionals.

## 20.10 Question 10: What Support Does the EAPC Provide for Children's Palliative Care Professionals and Services?

Showing respect for the cultural and political diversities of members across Europe, EAPC aims to speak with 'one voice-one vision' on matters of importance for the practice and development of PC, including the care of children and their families. The EAPC task force on CPC (EAPC 2019a) is an active group of professionals from throughout Europe who aim to drive developments for professionals and organisations working in children and young people's PC across Europe. The group promotes the development of accessible and high-quality children and young people's PC by facilitating and encouraging education and research.

The EAPC world congress takes place on alternate years and is attended by nearly 3000 delegates from Europe and across the globe. In 2019, for the first time, there was an entire day of the congress dedicated to CPC. The task force was instrumental in developing the programme which saw more than 900 delegates attend.

## 20.11 Question 11: How Does the EAPC Collaborate Nationally and Internationally?

The EAPC understands the importance of working collaboratively and the impact of working together and has strong links with the ICPCN and Together for Short Lives. It has several CPC national associations among its members (EAPC 2019b).

## 20.12 Question 12: Is There a Regional Association in the Americas and What Is the Situation for CPC in the Region?

The development of CPC in the Americas, a large and highly populated continent with high, mid and predominantly low-income countries, can be best described as multicentric, with a strong, abiding 'ripple effect'. Pioneers in the USA, Argentina and Costa Rica established clinical centres of excellence which have accrued decades of experience, having a hand, in some capacity, in the training of all the paediatricians/academicians currently practicing in the continent. The disparities within the region can be seen in the Atlas of Palliative Care in Latin America (Pastrana et al. 2012) and the Global Atlas of Palliative Care at the end of life (Connor and Sepulveda 2014). The Asociación Latinoamericana de Cuidados Palliativos (ALCP) promotes the development and implementation of PC in Latin America through education, research, management, policy development and access to medicines for PC.

## 20.13 Question 13: What Options Are There for Education in Children's Palliative Care in the Region?

*PCEP, Palliative Care Education and Practice* (Paediatric Track) was created at Harvard University. Collaboration with Northwestern University and other institutions resulted in *EPEC, Education in Palliative and End-of-life Care. EPEC-Pediatrics Latin America* is taught in Spanish. The *Pediatric Pain Master Class* in Minneapolis provides scholarships to Latin-American clinicians. Internet-based list-servs, including one maintained by the American Academy of Pediatrics (AAP) are widely utilised. WhatsApp is also widely used for real-time clinician, patient and even bereavement support in the absence of other resources in remote locations.

Scholarships created by the American Academy of Hospice and Palliative Medicine (AAHPM), through its Global SIG exist for clinicians in Latin America (and throughout the world) for participation in the annual conference. Similarly, Mercy Children's Center for Bioethics sponsors clinicians, an important consideration in a continent with wide variability in end-of-life (EoL) practice: while many countries prohibit compassionate extubation, euthanasia has been legalised in Colombia. Finally, the ALCP has a Paediatric Commission which interfaces with the continent-wide paediatric oncology and critical care societies, and with ICPCN: a very successful conference in Buenos Aires was held in 2016; clinicians take advantage of the free online PC Spanish-language education resources maintained by the organisation.

## 20.14 Question 14: What About the Situation Globally and Is There a Global Organisation for Children's Palliative Care?

Globally there are more than 21 million children with conditions that will benefit from a PC approach, with eight million needing specialised care. Less than 5% of these children have access to relevant services, with the greatest need and scarcity of services occurring in the developing world (Connor et al. 2017).

ICPCN is the only international charity connecting close to 3000 individual and organisational members working in CPC from 124 countries. Its mission is to achieve the best quality of life (QoL) and care for children and young people with life-threatening or life-limiting conditions, their families and carers worldwide. Key activities of the network include advocacy, service development, expanding the evidence base for CPC through research and sharing expertise, skills and knowledge through information and education (ICPCN n.d.).

## 20.15 Question 15: Does ICPCN as a Global Organisation Have a Direct Impact on Palliative Care Provision for Children?

In June 2018 ICPCN received the following email:

> *I am a registered nurse working in home-based CPC in North Carolina, USA. We have a Maya family from Mexico who receive services through our program. We would love to know if there is anyone with whom we could connect to discuss the Maya people's culture and how their lifestyle and beliefs could be impacting this family's approach to decision-making with regards to medical technologies (tracheostomy, long-term ventilation) and end-of-life conversations. Any connections, resources, or insight you can provide would be greatly appreciated.*

It took a simple act of reaching out to ICPCN members from Mexico and receiving this almost immediate response from a doctor:

> *… of course I will be able to help to discuss Maya people's culture, lifestyle and beliefs concerning the decision-making approach at end of life in Mexican children.*

Contact details were exchanged, a supportive connection between two professionals made and a family and child cared for in a culturally appropriate manner.

This example, and that of Makuta and the young boy and his family in China illustrate the value of international connections. However, the outcomes of much of the collaborative work undertaken by the ICPCN are often more strategic in nature.

## 20.16 Question 16: How Does the ICPCN Support Its Individual Members?

ICPCN is a central hub, connecting members and serving as a repository and signposting agent for relevant information. Specific knowledge, skills or experiences are regularly sought and ICPCN has been instrumental in brokering international partnerships between programmes and co-ordinating numerous international visits for learning purposes.

Indeed, relationships with local and national organisations have emerged as an essential component of ICPCN's core focus. Collaborations for conferences, training purposes and research have to date led to partnerships with over 100 organisations from more than 42 countries across four continents. Several of these collaborations

have developed into strategic partnerships that have served to educate thousands of healthcare workers on CPC through the sharing of knowledge, skills and resources.

## 20.17 Question 17: How Does Collaborating with International Palliative Care Organisations Achieve the ICPCN Vision?

WHO recognises ICPCN as the voice of CPC globally. Working together with the International Association of Hospice and Palliative Care (IAHPC) and the Worldwide Hospice Palliative Care Alliance (WHPCA) to advocate for the inclusion of PC across all ages as part of Universal Health Coverage (UHC) has resulted in major advocacy wins including the passing of the WHA67.19 resolution calling on all member states to 'develop, strengthen and implement, where appropriate, PC policies to support the comprehensive strengthening of health systems…' (Downing 2014).

While the benefit from these advocacy victories can seem difficult to measure, ensuring the absolute right for children and families to access PC will, in time, vastly improve the quality of life for millions of children needing PC. Then, no child or family will feel abandoned by a healthcare system that cannot meet their unique needs nor be uprooted from their homes and in some cases, their countries, to find appropriate care.

## 20.18 Question 18: What Ripple Effect Does Education Have on Developing Children's Palliative Care Globally?

Providing relevant education is essential to the development of the field. As a relatively new specialty, access to training of staff working with children with PC needs is not always accessible to those who need it most (Downing and Ling 2012).

Collaborative partnerships have been critical to ICPCN educating healthcare professionals with diverse training needs, languages, cultures and disparate access to resources. A key educational focus has been providing online learning courses devised by recognised experts and presently available in 11 languages. With this free resource Dr. Lee in China, for example, could access a Chinese course on *EoL Care*, providing the knowledge to care for future patients.

In collaboration with national and local agencies, ICPCN has given training in over 25 countries and hosted or co-hosted ten international conferences. This has resulted in over 1000 people benefiting from face-to-face training workshops and more than 3500 from e-learning. If each of these people provided appropriate care for only ten children, they will have reached over 45,000 children globally (Daniels et al. 2019).

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# Empowering the Team Through Education

21

Alex Daniels, Linda Ganca, and Susie Lapwood

**Key Learning Points**

1. There are opportunities for children's palliative care (CPC) teams to expand their skills and knowledge through ongoing learning and commitment to support the care needs of children living with life-limiting conditions (LLCs) and life-threatening conditions (LTCs) and their families.
2. Each member of the team plays a vital role in the interdependent nature of service delivery which requires that each member assumes responsibility for their own learning and development so as to promote team confidence and competence.
3. There are a range of CPC educational programmes available worldwide: learners may choose from face-to-face, online and blended training options. Clinical placement is recommended.
4. The importance of reflective practice and its role as an intrinsic tool to self-care.
5. There is an essential need for ongoing mentoring and supervision of individuals and teams working in the field of CPC.

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**Case Study**
*Zola Manyana is a bright 11-year-old student who lives with her Mom Zanele, Dad Sipho, sisters, Lulu aged 19 years and Thandi aged 6 years at their family home in the Western Cape, South Africa. Zola presents to her local clinic with a brief history of blurred vision, facial drooping, dysphagia and speech difficulties and is referred to an acute tertiary health care centre. She is diagnosed with a diffuse intrinsic pontine glioma (DIPG). There are no curative treatment options, and she is sent back from the acute tertiary health centre to her local hospital for PC. You are a paediatric nurse working at the local hospital and realise that you and the team caring for Zola will need some education on CPC in order to provide her with PC.*

## 21.1 Question 1: Why Is Education Important in Children's Palliative Care?

Education lies at the heart of PC service delivery and forms a key component of the WHO's public health strategy (Stjernsward et al. 2007; Downing and Ling 2012) and the roadmap for PC development (Callaway et al. 2018). The WHO strategy outlines four essential elements to achieve PC for all: '(1) *appropriate policies*, (2) *adequate drug availability*, (3) *education of health workers and the public and* (4) *implementation of PC services*' (Stjernsward et al. 2007, p. 487) with the addition of a fifth element at a later date—that of research (Harding et al. 2013). Furthermore, working towards the inclusion of palliative care within Universal Health Coverage (UHC), the Worldwide Hospice Palliative Care Alliance (WHPCA) recommends that palliative care training be integrated into the curricula of all undergraduate health workers and be incorporated as a component of continuing professional development (Connor and Sepulveda 2014).

In 2014, the first ever global resolution on palliative care, WHA resolution (WHO 2014) 67.19 called upon the WHO and Member States to improve access to PC as a core component of health systems, with an emphasis on primary health care and community/home-based care. WHO undertook to support member states to strengthen the integration of PC globally by:

- Promoting increased access to PC for children,
- Promoting adequate resources for programmes,
- Developing models for care,
- Supporting the development of PC guidelines and tools,
- Supporting national processes around accessing PC medicines,
- Monitoring and evaluating progress made in global PC programmes, encouraging research especially in low-and middle-income countries (LMICs) (WHO 2014).

From a CPC perspective, a global study released in 2017 estimated that more than 21.1 million children worldwide would benefit from a generalist PC approach with eight million children requiring specialist PC (Connor et al.

2017). The study recommended a move to integrate PC services into primary health care and paediatric sectors in an attempt to address this large unmet need (Connor et al. 2017). Undoubtedly education holds the key to improving the QoL for children living with LLCs or LTCs. Increasing the number of health care professionals who are knowledgeable and skilled in the field is of utmost importance.

## 21.2 Question 2: Who Makes Up the Team in Children's Palliative Care?

The WHO acknowledges the role that health care providers play in evaluating and alleviating the holistic needs of the child and suggests that holistic care be provided by a broad multidisciplinary team (WHO 2002).

The team providing CPC may include nurses, doctors, paediatricians, social workers, child life specialists, allied health professionals, complementary health therapists, spiritual and family counsellors, traditional healers, teachers and the patient's own family. The child's own contribution to the team will depend on their age and level of cognitive development. Teams comprise of members with varied disciplines, whose roles focus on the common goal of optimising care. Health providers with experience in supporting children with PC needs and their families will appreciate both the diversity of skills required and the value of a team approach to care.

Working together as a team provides a range of opportunities for health workers to grow and develop both as individuals and professionals, thus enhancing the team's collective capacity and efficacy. In the quest to provide best quality care, it is important for health workers to recognise these opportunistic educational moments at all points along the child's disease trajectory.

Teams are thus integral to CPC provision and the effectiveness of a service often correlates with how well teams and their members work together. Effective communication is not only crucial to supporting children and families but is a central aspect of team work. If this breaks down, the repercussions are detrimental for all. Amery et al. suggest that '*dysfunctional teams cause dysfunctional communication which causes dysfunctional children's palliative care*' (Amery et al. 2009, p. 11). A dysfunctional team is unlikely to enjoy the culture of mutual respect, trust and honesty that is essential for shared learning.

Team functionality is dependent on individual roles assigned to members, how the team is coordinated and who assumes team leadership. There are several models of teamwork:

- In a *multidisciplinary model* of teamwork, professional roles are specialised with members focused on their own task. Coordination is supervised and leadership is hierarchical, with the professional often contributing expertise towards patient well-being in isolation.
- Although roles remain specialised within *interdisciplinary* teams, there is interaction between roles with members working interdependently and sharing

information from their own expertise. Everyone coordinates their own activities and leadership is task orientated.
- Within *trans-professional* teams, roles are specialised with members expected to replace roles as needed, coordination is interactive and flexible and the team self-regulates leadership (Crawford and Price 2003).

Recognising and integrating opportunities for learning in the daily practice of all teams is essential if we are to optimise the well-being of children and families in our care.

**Case Study**
*You carry out Zola's assessment on admission. You have not cared for a child who is this sick for a long time as they usually stay at the tertiary hospital for care, so you decide that it is important to reflect on Zola's assessment and identify any gaps in your knowledge and skills.*

## 21.3 Question 3: Why Is It Important to Carry Out the Process of Reflection in This Instance?

Reflection is considered a key aspect of undergraduate, postgraduate and continued education and integrating regular reflective practice into professional life supports lifelong learning. Reflective practice facilitates the assimilation of theoretical and clinical components of learning with the ultimate goal of improving clinical practice. Furthermore it is through the process of regular reflective practice that self-awareness is developed and stimulated (Sandars 2009; Salins 2018).

Self-awareness is a key component of emotional intelligence and has an important role to play in a health care professional's relationship with both patients, family members and colleagues. Self-awareness is described as the ability to recognise and understand your emotions, the effect of your actions and moods as well as the emotions of other people (Rasheed 2015). In a CPC context, it is also important to have the ability to distinguish between what one knows and does not know so as to identify gaps in one's own knowledge and skills. Once gaps or challenges in knowledge and skills are recognised, training needs may then be identified and pursued.

Teams reflecting on the experience of caring for their patients may identify numerous gaps in their knowledge and skills. Zola's team is likely to need more support in their understanding of her diagnosis and expected disease trajectory, in identifying and managing distressing symptoms and in communicating with Zola and her family in relation to her illness.

## 21.4 Question 4: How Would You Carry Out the Process of Reflection?

Different models of reflection share common features despite varying format and structure. Reflection commences with a superficial description of a positive, negative or new experience. Reflection proceeds to focus on what is already

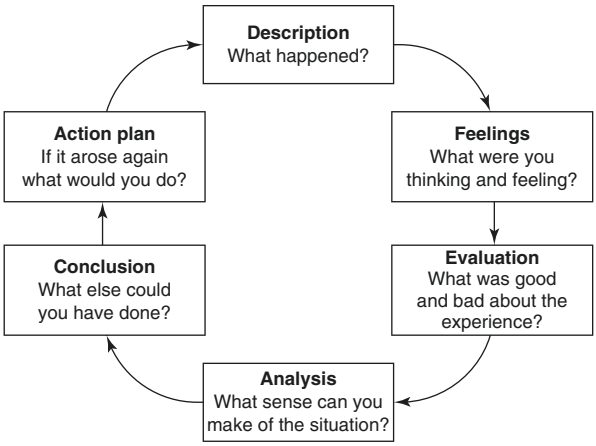


**Fig. 21.1** Gibbs (1988) reflective cycle. Reprinted from Paterson and Chapman 2013. Copyright 2013, with permission from Elsevier

known about the experience—this may integrate the relevance/appropriateness of theory. What follows is an acknowledgement of feelings, assumptions, knowledge or gaps in knowledge in relation to the experience. The final and deepest stage of reflection relates to change—it includes beliefs, views, opinions and actions, how you view yourself and others and what may be needed to change, in attitude, learning or action. Gibbs model of reflection (Gibbs 1988) (Fig. 21.1) is a useful framework for undertaking reflection in your day-to-day work.

## 21.5 Question 5: How Might You Decide Which Learning Needs to Be Prioritised?

Individual and team reflection will help identify the most important and urgent issues for education and training to optimise Zola's care. Children living with LLCs have unique care needs and have a right to access specialised care at any point along their disease trajectory. In order to provide appropriate care, Zola's care team need to be equipped with the following skills and knowledge:

- A clear understanding of the core aspects of CPC.
- An understanding of how to enhance Zola's physical comfort throughout her disease trajectory—this includes assessment and management of pain and other distressing symptoms.
- The ability to identify and respond to Zola's psychosocial, educational and spiritual needs.
- The ability to assess and respond to the needs of Zola's caregivers including her siblings.
- The ability to facilitate communication and decision-making around Zola's care including end-of-life (EoL) care.

**Case Study**
*Whilst you have been a health care professional for some years, on reflection you realise that you do not know much about CPC. You have heard a bit about it, but were not taught anything about this in your initial training, or since. You therefore realise that you have many gaps in your CPC knowledge, but are not sure what you need to know at this stage, or how to go about getting further education or training.*

## 21.6 Question 6: What Are the Different Types of Training Available on Children's Palliative Care and at What Levels?

The WHA resolution 67.19 (WHO 2014) sets out that education and training offered to care providers should include palliative care as an integral element, tailored to their roles and responsibilities as follows:

a. *Basic training and continuing education in PC* should be a routine element of all undergraduate medical and nursing professional education. It should also be part of in-service training of caregivers at the primary care level, including health care workers, caregivers addressing patients' spiritual needs and social workers.
b. *Intermediate PC training* should be offered to all health care workers who routinely work with patients with LTCs, including those working in oncology, infectious diseases, paediatrics and internal medicine.
c. *Specialist PC training* should be available to prepare health care professionals who will manage integrated care for patients with more than routine symptom management needs (p. 4).

The European Association for Palliative Care (EAPC) also outlines a three tier framework for PC education in their guidance concerning both adult and CPC (Gamondi et al. 2013; Downing et al. 2013). This promotes educating all healthcare professionals in the principles and practice of PC at the time of their initial training. Gaining a specialist level of knowledge and skills is recommended for those whose work has a primarily palliative focus. With regard to CPC they (Downing et al. 2013) describe the three levels as:

a. *Basic education in the CPC approach*: This approach focuses on the general principles and practices of CPC and aims to integrate PC methods and procedures in general settings of care, such as paediatrics and family medicine.
b. *General CPC education*: This level of education and training is necessary for professionals who may have frequent involvement in the provision of CPC, or may act as the resource person for PC in their particular setting. This group

includes professionals such as paediatric oncologists, adult PC staff, and nurses who do not provide CPC as the main focus of their clinical work.

c. *Specialist CPC education*: This level of education and training is required for professionals working solely or mainly in the field of CPC, who are frequently dealing with complex problems and may be called upon to give expert advice to wider colleagues. This requires specialised skills, knowledge and competencies in symptom management, facilitated decision-making and communication.

Therefore a multifaceted approach is required to increase CPC education globally. It is important for health care professionals to have access to a range of courses varying in length, intensity and presentation style. Basic, intermediate or specialist CPC training courses are available via public, private or non-governmental organisations. These courses are presented as face-to-face, online or blended learning sessions. In addition to specific courses, there is a need to create and promote accessible resources, guidance and learning modules, in order to ensure health care professionals who may need this material infrequently can access relevant information, training and refreshers at the time and point of need. It would be appropriate for health care professionals caring for Zola to undertake a basic or intermediate course in CPC so as to deepen their understanding of CPC and, in so doing help optimise the quality of care being provided to Zola and her family (Table 21.1).

## 21.7 Question 7: What Is the Place of Clinical Experience in Children's Palliative Care Education?

Clinical placement is recommended alongside the completion of a basic or intermediate course (Downing et al. 2013). Joining an experienced CPC team for a few days allows the learner to actively witness delivery of PC, including, amongst others, symptom management, communication with children, families and team members through interdisciplinary team discussions.

It is important to develop supportive networks during CPC training to enable health care workers to have a platform in which they can feel safe to ask questions, share ideas and seek guidance. Such networks are particularly crucial in settings where there are no experienced PC teams nearby.

**Case Study**

*You have undertaken an initial basic level training, which gives you some understanding of CPC. Zola has remained on the ward and you are concerned at her level of pain management, as you think that her pain should be better managed. You suggest to the clinicians that Zola may benefit from a trial of oral morphine, but they are not keen to do this.*

**Table 21.1** Examples of CPC courses (at both generalist and specialist levels)

| | | | |
|---|---|---|---|
| International Children's Palliative Care Network (ICPCN) | Face-to-face and online training | Basic and intermediate training | Face-to-face workshops are undertaken in response to specific requests and are funding dependent. The ICPCN's e-learning platform offers a variety of short courses on CPC and participants may choose from a range of topics, e.g.: Introduction to CPC, Pain assessment and management for children, Communicating with children and emotional issues, Child development and play, End of life care, Grief and bereavement, Perinatal PC and Symptoms other than pain. The courses are endorsed by the University of South Wales and are freely available in a range of languages including French, Spanish, Portuguese, Russian, Mandarin, Serbian, Czech, Dutch, Vietnamese and English |
| Education in Palliative and End-of-Life Care (EPEC)—Paeds | The course consists of 20 distance learning modules and six in-person conference sessions | Basic and intermediate training | It is a comprehensive training course adapted from the EPEC adult curriculum. The programme, devised in Minnesota, United States of America (USA), is directed at paediatric care providers including oncologists and consists of 24 modules addressing pain and symptom management in CPC |
| Portuguese Catholic University's School of Public Health and Family Medicine | The courses have regular on-site sessions for a duration of 124 hrs | Basic and intermediate training | They run courses annually from October to May. Students receive classes on principles and philosophy of CPC, research, ethics, communication, grief and bereavement, spirituality, family support, service development and organisation and symptom control. Classes consist of formal lectures, practical sessions and group discussions |
| University of Cape Town | This 1 year blended course includes face-to-face training and elements of distance based learning | Specialist training | Postgraduate Diploma in CPC for multidisciplinary team members. Students are required to be working in a paediatric setting where they can engage in work based learning applying the learnt theory to children and families they care for |

## 21.8 Question 8: How Might You Be Able to Help the Clinicians Understand How to Manage Zola's Pain?

It would be important to support the clinical decision makers/prescribers to optimise Zola's pain management. You decide to offer training on better pain management to the team and identify this as an urgent topic to be addressed in the next interdisciplinary team meeting. Other ways to promote ongoing learning include:

- Continuing medical education (CME) sessions on all aspects of paediatric pain assessment and management.
- Sharing resources—circulating a range of pain rating scales such as the FLACC scale, numeric/word scale and ELAND colour scale (see Chap. 7 on pain assessment and management for further information).
- Sharing papers—access to papers such as the Pediatric Pain and Symptom Management Guidelines (Hauer et al. 2014) and the Lancet Commission report (Knaul et al. 2017).
- Journal clubs—starting a journal club or connecting with existing or electronic journal clubs, advice forums and abstract database reviews, such as:
  - Paedpalcare, hosted by Together for Short Lives (UK) https://www.together-forshortlives.org.uk/changing-lives/sharing-learning-networking/digital-care-forum/
  - PedPalASCNET, a Canadian research library https://pedpalascnetlibrary.omeka.net/

**Case Study**

*You realise that Zola needs more PC expertise than you are able to provide, due to your own and the team's lack of knowledge. You are fortunate to have a specialist CPC team nearby, so your team asks for some initial 'in principle' advice by phone and email. You also ask if a member of their team could provide on-site advice as well as some ongoing mentoring and clinical supervision.*

## 21.9 Question 9: Why Is It Important to Provide Ongoing Mentorship and Supervision?

Mentoring and supervision supports the process of learning by integrating what has already been learnt (Downing et al. 2013). Developing and nurturing of a successful relationship between a lesser experienced health care professional (mentee) and more experienced health care professional (mentor) is critical to the mentoring process. An evolving mentoring relationship supports changes to the personal and professional environment with the benefit of enhancing clinical learning and professional development (Ikbal et al. 2017).

The formal process of clinical supervision (individual or group) further helps the individual to develop knowledge and competence in the field of CPC through professional support. These are important elements that require consideration so as to

not only support professional development but also nurture well-being and optimise quality of work (Taylor and Aldridge 2017).

If there are no opportunities for formal mentorship and supervision, it is worth finding out about people with specialist CPC expertise in your setting or region and identifying if they are willing to be contacted by phone, email, etc. for 'in principle' advice, support and signposting, even if they are unable to visit your service. You can also post anonymised clinical queries on CPC advice forums such as PaedPalCare.

**Case Study**

*The PC team is very supportive to you and your team and empower you to provide good PC to Zola and her family. Unfortunately Zola's disease is very advanced and she dies whilst in the hospital. Following her death, you encourage the team to reflect on what you have learnt through caring for Zola and her family, and how you have worked together to provide PC.*

*On reflection you realise that you are really interested in PC and explore intermediate training options. However, there are no PC specialists at your hospital, so you consider undergoing specialist CPC training.*

## 21.10 Question 10: What Are the Competencies That You Would Develop Through Undertaking Specialist Children's Palliative Care Training?

For *specialist* CPC, five broad domains are outlined (Downing et al. 2013), embracing wider issues such as leadership, policy, training and research:

- *The Caregiving Relationship*—an understanding of the practice and principles of CPC, communicating with the child and his/her family, psychosocial and spiritual care, bereavement care and self and team care.
- *Clinical Care*—pain assessment and management, assessment and management of other distressing symptoms and EoL care.
- *Collaboration and Inter-professional Practice*—team work and networking.
- *Leadership*—leading and developing services and advocacy.
- *Professional Practice*—research, evaluation of services, policy and training and education.

## 21.11 Question 11: How Would Specialist Children's Palliative Care Training Empower You for Future Roles?

Health care professionals undertaking specialist CPC training are well placed to optimise their contribution to the field. Development of sustainable CPC initiatives and services requires charismatic leaders to be equipped with skills and knowledge to advocate, collaborate, communicate and network with other role players (Downing et al. 2013).

## 21.12 Question 12: How Could You Best Care for Yourself, to Protect and Enhance Your Own Resilience in This Demanding Work?

It is important to adopt a personal self-care plan that addresses your physical, emotional, spiritual and social needs. Some helpful self-care practices include:

- Maintaining a balanced lifestyle that includes exercise, a healthy diet and rest.
- Making time for relaxation and positive social exchanges.
- Setting clear boundaries and managing time efficiently.
- Creating opportunities for creative expression through movement, music and art.
- Prioritising individual supervision and mentoring.
- Reflective practice and regular team meetings.
- Attending bereavement/memorial services.

## 21.13 Question 13: How Would the Team Have Reflected Together on Their Learning Through Caring for Zola?

Ideally there should be a menu of opportunities to aid individuals and teams in reflecting on and learning from an episode of care. This could be within a team and/or across service boundaries, which can further enhance wider team working and education, including:

- Interdisciplinary team discussions.
- Mortality and morbidity meeting.
- Reflective debriefing sessions.
- Ongoing mentoring and supervision.

## 21.14 Conclusion

This chapter encourages the learner to actively engage with opportunities for personal and professional development within a team context. Learning should be ongoing for every care professional: lifelong, life-deep and life-wide. It has the potential to transform both individuals and teams, impacting positively on CPC practice. Most importantly, developing regular self-care practices is vital to enhance the health care professional's capacity for resilience in this rewarding albeit challenging area of work.

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# Integrating Research into Care

# 22

Jan Aldridge and Julia Downing

**Key Learning Points**

1. There are different types of evidence to consider including systematic reviews, randomised controlled trials (RCTs), case-controlled studies, cohort studies, case series.
2. A thorough, critical review of the existing literature informs thinking about research and practice and should have a clear search and selection strategy, critically evaluate the existing literature, be well-structured and well-referenced.
3. Undertaking a literature search can be divided into four steps: framing the question; choosing a search method; identifying criteria and keywords; narrowing the search results.
4. In developing your own study it is important to get the research question right as this will aid the identification of the appropriate research methodology and the feasibility of the study. It is also important that any tools used to address the research question are reliable and well validated and data analysis is appropriate and transparent.
5. Dissemination of findings of sound research is important. Through dissemination we keep learning and improve the care that is delivered.

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**Case Study**

*Aysha is a nurse who has been working in children's palliative care (CPC) for the past 3 years. She is looking after Seb, a 14-year-old boy with a slow degenerative condition, Duchenne Muscular Dystrophy (DMD).*

*Seb is aware that his condition is progressing and for the past 2 years he has used a wheelchair to get around. He is usually quite lively and outgoing with a strong group of friends. Six months ago his older brother, who had the same condition, suddenly and unexpectedly died. Since then Seb has become increasingly anxious and withdrawn and is now reluctant to leave the house. His doctor has checked him thoroughly physically and tried hard to reassure him…but to no avail. The situation is getting worse and Seb is getting more isolated from his friends.*

*His mother has brought up her two sons on her own and has always had a close relationship with Seb, but since his brother's death he is reluctant to talk to her. She does not know what to do. She is struggling to keep her job.*

*Aysha is wondering how Seb might be helped with his increasingly crippling anxiety. She wonders what helpful information there might be in the research literature to help in her understanding and management of anxiety in children and teenagers with life-limiting conditions (LLCs).*

## 22.1 Question 1: How Would Aysha Go About Finding Evidence with Regard to the Management of Anxiety in Children's Palliative Care?

Ensuring that our practice draws on the available evidence base is important in the provision of CPC. Indeed, research has been proposed as the fifth pillar of the WHO's public health strategy alongside that of policy, education, drug availability and implementation (Stjernsward et al. 2007; Harding et al. 2013). There is a lack of research in CPC (Downing 2016; Harding et al. 2014) and generation of a good evidence base through sound research along with its application is essential in the ongoing development of CPC globally.

There are a variety of ways of gathering evidence on the management of anxiety in CPC and this has become easier now that many journals and books are available online. Sources of evidence will include the following:

a. *Books*—whilst books can be an important source of comprehensive information, some information in books can become out of date quite quickly.
b. *Peer reviewed papers*—i.e., papers that have been published in a journal that subjects all submitted papers for review by experts prior to publishing are an important source of up-to-date evidence.
c. *Grey literature*—this includes reports, policy statements, conference proceedings, theses, newsletters, fact sheets, etc. These need to be read critically as, in terms of research quality, standards can be extremely variable.

Whilst the internet has made it easier to search for information, it offers many challenges with regard to both quality (as noted above) and quantity (you may get many 'hits', i.e., matches to your search and not all will be relevant). There are a variety of

online databases which you can use for your search, such as PubMed, EMBASE, Medline, PsychINFO, Google Scholar, CINHAL (Cumulative Index to Nursing and Allied Health Literature) and HINARI (Hinari Access to Research in Health programme).[1] The database that you use will depend on the topic that you are looking at, but for Aysha, who is looking at issues of understanding more about anxiety in CPC, databases might include PubMed, PsychINFO, Medline and CINHAL. Most of the databases can be searched for free online, although access to papers themselves may only be available for a fee or through individual or institutional membership.

## 22.2 Question 2: How Might Aysha Carry Out a Literature Review on the Understanding and Management of Anxiety in Children's Palliative Care?

A literature review is '*the selection of available documents (both published and unpublished) on the topic, which contain information, ideas, data and evidence written from a particular standpoint to fulfil certain aims or express certain views on the nature of the topic and how it is to be investigated, and the effective evaluation of these documents in relation to the research being proposed*' (Hart 2000, p. 13). Many health professionals find the thought of undertaking a literature review daunting, not knowing where to start, how to select the topic, how many articles they should include, etc. (Cronin et al. 2008). A good literature review is clear and concise and easy to follow (Table 22.1).

There are a variety of different types of literature reviews ranging in complexity, these include (Cronin et al. 2008):

- *Traditional or narrative literature review*—its purpose is to provide a comprehensive understanding of current evidence, highlighting any significant new research.
- *Systematic literature review*—its purpose is to provide as complete a review as possible, within defined criteria, of all the published and unpublished studies in a particular area. Thus, the literature on a particular topic is identified, critically evaluated and synthesised.
- *Meta-analysis*—this looks at quantitative findings from many studies and involves conducting statistical analyses in order to enhance understanding.
- *Meta-synthesis*—this looks at qualitative research findings and integrates, evaluates and interprets the findings.

**Table 22.1** Characteristics of a good literature review (Carnwell and Daly 2001; Colling 2003; Cronin et al. 2008)

| |
|---|
| • Well written and contains few, if any biases. |
| • Contains a clear search and selection strategy. |
| • Is well-structured to enhance the flow and readability. |
| • Uses accurate terminology with limited jargon. |
| • It is well-referenced. |

[1] HINARI enables individuals and organisations in low-and middle-income countries (LMICs) to access biomedical and health literature and is managed by the WHO (Robertson 2014; Sarik 2016).

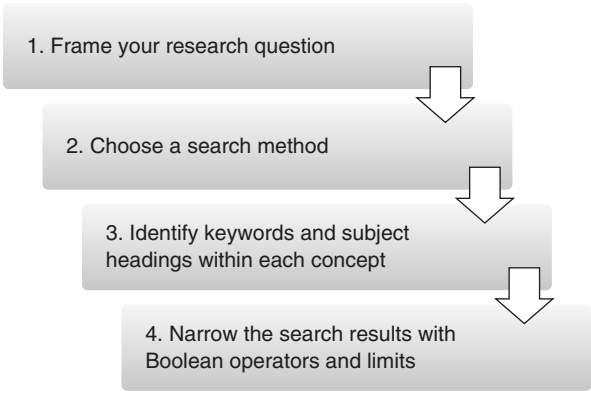


**Fig. 22.1** Literature search process. Reprinted from Aoki et al. (2013). Copyright 2013, with permission from John Wiley & Sons

A literature search process can be divided into four key steps (Fig. 22.1):

a. The first step is to think about the question that you wish to address. Anxiety is a big topic, what is it that Aysha is particularly interested in with regard to anxiety, what is her review topic? For example, she might ask, 'What is the best treatment for managing anxiety in children?' It is likely that this depends on the cause of the anxiety and the child and their situation, so it would be helpful to refine this further and make it more specific, for example: pharmacological and non-pharmacological management of anxiety in teenagers with serious health conditions. The more specific the topic, the more feasible the review.
b. The second step is to think about how you are going to undertake the search and which databases to search? Online databases can be searched, along with the wider internet for the grey literature, as well as any local and national library resources available. You might also want to contact relevant experts in the field.
c. The third step is to think about the keywords and subject headings you are going to use? The clearer your question, the easier it is to identify your keywords. Keywords are words that are used anywhere within a citation, i.e., the title, abstract and subject headings. You may need to think about different forms of the same word and combine these in searches, e.g., paediatric palliative care, paediatric palliative medicine, children's palliative care. The more keywords you can combine and the more specific you can be, the more relevant the papers identified. It is important to keep a record of the keywords that you have used, along with the databases you have searched, as you need to be transparent about these in any subsequent writing.
d. The fourth step is narrowing the search—often it will still be necessary to narrow your search further, otherwise you may have a large sample with many irrelevant articles. You can do this in a number of ways, but you need to think very carefully about your rationale for this and be explicit about it. For example, you might do this by only selecting articles in peer reviewed journals published in the last 20 years, articles only published in English or Spanish. 'Boolean operators' are also useful, e.g., 'OR' (looks for articles that include any of the keywords), 'AND' (looks for articles that include all the keywords), etc.

If Aysha cannot find much robust literature, then she may have used the wrong terms or databases and she may want to check with a colleague or her local librarian. However, as often happens with complex situations in CPC, there may indeed be little robust, directly relevant research upon which to draw. In the case of Seb, she finds that there is some research on anxiety in children and teenagers in general and some on anxiety in children with chronic health conditions. However, she notes that a significant number of these studies have been done in the United States of America (USA), which has a different approach to diagnosis than many other countries. In the existing studies, a higher prevalence of anxiety and young people with LLCs has been found. However, there is little research on the factors that are associated with anxiety in these children, little understanding of the risk and protective factors and little research on the impact of different treatment approaches, pharmacological and non-pharmacological. In addition, there is some research on anxiety in adults in PC, but these studies are mostly with older adults as they approach the end of their lives. Aysha will need to think critically as to what she might take from this existing literature, e.g., how relevant is it to generalise from an older adult age group to a teenage population?

## 22.3 Question 3: Having Identified the Literature How Can Aysha Assess Its Quality?

Once Aysha has gathered together the literature that she thinks is appropriate, it is essential to review and critique it to ensure it is of sufficient relevance and quality. Most articles will have an abstract or summary at the start, which can be useful for an initial reading of the papers. There are different tools available to help with critiquing the literature. For example, the Critical Appraisal Skills Programme (CASP—www.casp-uk.net) provides eight critical appraisal tools to help review and critique different types of papers, e.g., systematic reviews, qualitative research, randomised controlled trials, etc. This is freely available and can be downloaded from the website. A simple way of critiquing the literature is the PQRS system (Cohen 1990), i.e., Preview, Question, Read and Summarise the papers. Evaluating the literature can be challenging, but is a key research skill. Utilising some of the free online tools can help you in learning how to do this, ensuring that the papers upon which you are drawing are credible and of sufficient quality for the information to be meaningful.

## 22.4 Question 4: There Are Different Types of Studies, How Will Aysha Know Which Ones to Look at?

Understanding the different types of studies is important when reviewing the evidence found through a literature review (Table 22.2). The 'hierarchy of evidence' (Fig. 22.2) suggests that not all evidence is the same and there used to be an assumption that systematic reviews were the strongest type of evidence, followed by RCTs, with case series and reports being seen as the weakest (Shaneyfelt 2016). Yet in

**Table 22.2** Examples of different types of studies

| | |
|---|---|
| Case study | A study that explores individual or small, similar accounts of a phenomenon or disease and may be either quantitative or qualitative (Ellis 2013, p. 147). |
| Case-controlled study | A study that identified individuals by outcome status at the outset of the study, e.g., specific type of disease. It also identifies controls, i.e., those without the disease and compares the two groups (Song and Chung 2010). |
| Cohort study | A study that examines a group of people with defined characteristics who are followed up to determine incidence of, or mortality from, some specific disease or outcome. It can be prospective or retrospective (Song and Chung 2010). |
| Cross-sectional study (prevalence study) | Research that examines the data on an issue, e.g., disease and exposure, at one particular time point (Song and Chung 2010). |
| Meta-analysis | A statistical method used to combine the results from multiple studies to provide robust understanding of the effect of an intervention (Ellis 2013, p. 149). |
| Qualitative study | Research that explores attitudes, opinions, experiences or behaviours through interviews, focus groups or observation (Ellis 2013, p. 150). |
| Quantitative study | Research that seeks to discover relationships between variables in a statistical way (Ellis 2013, p. 150). |
| Randomised controlled trial (RCT) | A specific form of study that is used in the clinical setting in order to compare the usefulness of two or more interventions (Ellis 2013, p. 150). |
| Systematic review | A process by which various research papers on a topic are identified and appraised for their quality in order to synthesise a solution to a problem (p. 151). |

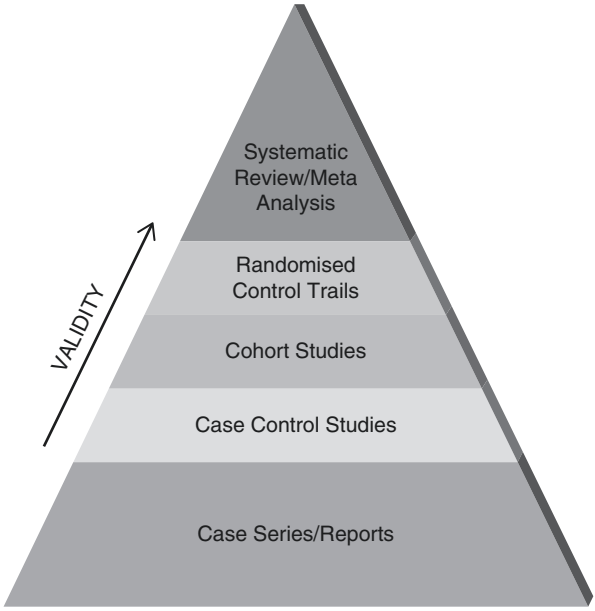


**Fig. 22.2** Traditional hierarchy of evidence. Reprinted from Murad et al. (2016). Copyright 2016, with permission from BMJ Publishing Group Ltd


**Fig. 22.3** Revised hierarchy of evidence with systematic reviews being the lens through which evidence is applied. Reprinted from Murad et al. (2016). Copyright 2016, with permission from BMJ Publishing Group Ltd

reality the line between each type of evidence is not so clear cut, as studies may be of differing quality and a study from a lower level might be more valid than the one above, e.g., a poorly designed RCT vs. a well-designed cohort study. Recently Murad et al. (2016) have suggested that systematic reviews and meta-analyses are tools for appraising, synthesising and applying evidence—they are the lens through which evidence is applied (Fig. 22.3). Whilst the hierarchy of evidence will not fit every situation, it is a useful tool for helping us learn about evidence and to think about the studies we are reading. It might provide a framework to help guide Aysha as she searches to deepen her understanding of anxiety in CPC (Shaneyfelt 2016).

There has been a tendency to rate quantitative data stronger than qualitative data, with the hierarchy of evidence traditionally being based on quantitative studies. It is important to recognise the strengths in both types of evidence. Both have important contributions to make; however it depends on the question being asked.

**Case Study**

*Seb is not at all keen on taking medication or on pharmacological approaches to managing his anxiety. He says he would prefer to manage the cause, rather than just deal with the symptoms. He is particularly interested in helping himself and wants to know more about such approaches as self-hypnosis, mindfulness and meditation.*

*He would also like help for his mother. He knows he is keeping his fears and worries from his mother at the moment, as he does not want to worry her more than she is already. He is concerned about how much she is grieving for his brother and he worries how she will cope as he himself needs more help. He thinks she already has too much to deal with and he fears what will happen to her when he dies.*

*Interestingly Aysha found in the literature that higher anxiety in mothers is linked to higher anxiety in their children. However, she can find no research on this in*

*mothers of children with potentially life-shortening conditions. She wonders about the impact on Seb of his Mum's worries and anxieties. She can find no research on family therapy or systemic working in this context. Given the limited literature she starts to think about doing some research herself in this area.*

## 22.5 Question 5: How Would Aysha Go About Undertaking a Study Looking at Anxiety Issues in Children, Teenagers and Their Families in Palliative Care?

When designing and conducting a study there are several key issues that Aysha would need to think about:

a. *The importance of getting the research question right*: The most important thing for Aysha is to make sure that the research question is right. It is always tempting to ask big questions, but in order to be achievable it needs to be a manageable question. Arriving at a good research question is often a process and takes time. It is worth spending the time thinking, reading, talking to people and refining the question into one that is viable. Aysha might regard deciding on a question as a group process and enlist the help of others to ensure that she has the right question. The question also needs to take into account what resources are available. It is likely that she will start with a small study, addressing just one aspect of the area of interest, or maybe a pilot study, which might lead to her seeking funding for a bigger study at a later date.
b. *What is the best method to use to address Aysha's research question?* Once Aysha has a viable research question it will help her decide upon the best method to use to address the question. The type of question to which Aysha seeks answers will push her towards the choice of research strategy. Aysha will need to read in considerable detail about choosing a research strategy. It is time well spent. There is much written about quantitative and qualitative approaches. Traditionally the suggestion has been that quantitative research is associated with the testing of theories, qualitative research with the generation of theories. In practice, there are situations and topics where a quantitative approach is called for and others where a qualitative, naturalistic approach is appropriate. Qualitative approaches are valued for their sensitivity, flexibility and adaptability. There are situations that are best served by a combination of the two traditions, 'mixed methods'.
c. *The need to think about numbers and trustworthiness*: To ensure that the results are trustworthy Aysha will need to have sufficient numbers for the design she is planning to use. In establishing trustworthiness two key issues are validity and generalisability. Validity is concerned with whether the findings are really about what they appear to be about. Generalisability is the extent to which the findings of the particular study are more generally applicable (Ellis 2013). In a quantitative study if there are not sufficient numbers, the study may well be underpowered and the results not valid. In a qualitative study sufficient participants of the right kind are needed in order to reach 'thematic saturation'. The exact numbers vary, but are generally a lot smaller than for quantitative research, with thematic saturation often being reached before 20 participants. Maybe the best advice for Aysha, as

she has limited experience in designing research proposals, is for her to seek out and work with others with good experience to shape and hone the study.

d. *Importance of using validated instruments*: If Aysha decides to do quantitative research, then the importance of using validated, reliable instruments, relevant to the population she is studying, cannot be overemphasised. If the measures have not been validated in a CPC population, then much caution needs to be exercised in deciding whether or not they are still applicable and also in interpreting any findings.

## 22.6 Question 6: Who Would Aysha Need to Liaise with in Order to Be Able to Undertake Her Study?

Aysha will need to decide if the work is research or service evaluation. If it is research, then she will need to seek ethical approval from the appropriate body at the health facility and/or university. This ensures that the research is worthwhile, participants are not taking part in research that is inadequately designed or subject to unnecessary risk and that their needs are provided for appropriately. If the work falls into service evaluation, then it is still essential for Aysha to think about the ethics of the work and her responsibility to the participants, but she will not usually need to seek formal ethical approval. Aysha will need to check this with her organisation as each will have its own processes that need to be followed.

Recruitment for the study depends on the nature of the study. If it is an internal piece of research, or part of a multi-centre study, Aysha would present it to the relevant people in her organisation and ask for their help in recruiting. It helps to have flyers or letters about the research that they can give out to potential participants, who can then contact Aysha for further information, before they make a decision about whether or not to participate. Increasingly researchers are using social media to advertise their studies and recruit directly. For example, in a recent study in the United Kingdom (UK) on anxiety in young adults with serious health conditions, participants were recruited via social media. It is important to be aware of bias, depending upon your source or sources of recruitment. It is also important that any participants are able to make fully informed decisions about taking part, that they are informed as to what will happen to their data, how it will be stored and for how long and that they have the option to withdraw at any point, without any impact on any care they might be receiving, should they so wish.

## 22.7 Question 7: What Might Be Some of the Challenges for Aysha in Carrying Out Research Within Children's Palliative Care?

Challenges to undertaking good quality research include: a lack of research skills; issues around ethical approval; the numbers of potential participants.

a. *Lack of research skills*: Aysha is interested in research and appreciates its value. She undertook a small-scale study as part of a Master's degree, but she does not consider she has the necessary research skills and experience to design and

conduct the study. There may be research methodology courses available or an opportunity to work with an experienced team. A good, experienced mentor is a wonderful resource.

b. *Issues around ethical approval*: Children and their families who have experience of serious illness, dying and bereavement are often regarded as vulnerable populations. This means that ethical committees are, rightly, going to look at any research proposal very carefully indeed. They will want to be convinced that the research is worthwhile and that it will be well executed. They will want reassurances that any issues stirred up by participation in the research will be dealt with by having appropriate help and support in place. Traditionally, professional gatekeepers have made the decision about whom to approach and when. Interestingly some children and families are now saying they would like to take the decision for themselves about whether or not to take part in the research. This has led to an increase in recruiting directly via other routes for some studies. When recruiting children it is more complicated, as if they are under 16 or 18 years of age (depending on the type of decision to be made) it is the parents who usually have to give their consent to their child taking part. Children can give their assent if they have the capacity to do so.
c. *Numbers of potential participants*: A further challenge to researchers in CPC is the number of children and families who might be available and want to take part. With some conditions the numbers are very small (e.g., Spinal Muscular Atrophy (SMA) Type 1) and so for certain types of studies the research would need to be carried out across a number of different sites. In some areas of research different conditions might be meaningfully grouped together.

**Case Study**

*Aysha decided she would begin with a small study which collected data on anxiety in mothers who had children with life-shortening conditions. Over a 12-month period she invited all new mothers who attended her clinic to take part. She used well validated measures of anxiety and found higher levels overall than would be expected from the population norms. Aysha wonders about the implications of her findings for working with Seb and his mother and also for other mothers and children she sees.*

## 22.8 Question 8: How Does Aysha Incorporate What She Has Found into Practice?

It is important that Aysha does not claim too much from this small-scale study, as it was on a limited sample of mothers, but she did find significantly higher levels of anxiety in these mothers than the general population. It could be that they are a selected group that have been referred to the clinic who are particularly anxious. They have children who are very ill and are concerned about them and may have had other children who have died from similar conditions, as in Seb's case. There

might also be other factors at play, such as more unsupported and isolated mothers in the sample or higher levels of poverty. Aysha thinks the issue needs more research.

In the meantime, Aysha shares and discusses the findings with colleagues. The team decides to look at resources to support these mothers, e.g., to provide drop-in sessions for these mothers to meet up with other mothers in similar situations; to write supportive, accessible materials for these mothers; to offer sessions with a specialist psychologist. Aysha agrees to undertake a relevant literature search and to put in place an evaluation of what they decide to offer, with the intention that this will both feed into practice and also guide further research.

## 22.9 Question 9: How Would Aysha Tell Others About What She Has Found?

Having undertaken research it is important to share the findings. In Aysha's case there was limited literature available on the topic, the small study was well conducted and therefore anything that she can add is potentially relevant. Unfortunately, many studies undertaken remain unpublished and unshared.

There are different ways for Aysha to disseminate her findings. Of course, reporting back to her colleagues and her participants via written materials or face to face meetings is essential. The traditional ways of presenting to wider professional and academic audiences at conferences and publishing in peer reviewed journals remain important. However, alongside these, the internet and social media have expanded opportunities to enhance dissemination (Devitt 2016). These might include: a continuing education session; writing a blog; writing an article for ehospice (www.ehospice.com); tweeting and other forms of social media. Such means of dissemination can help ensure the results get to a wide-ranging audience.

## 22.10 Question 10: How Can We Encourage More People to Utilise and/or Conduct Research?

Not everyone working in CPC will undertake research. Individuals have different roles with regard to research and evidence utilisation and generation. Not everyone's role is to undertake research studies and write papers, or to provide good research supervision or mentorship. It is, however, important that we can all critically evaluate the relevant research that has been done in our professional area and utilise it wisely to help inform the care that we provide.

Good research is so often a team effort. Many of the most practical ideas for research, for example, come from front line workers or from families with the lived experience. These ideas are then developed into strong, viable projects by people with the necessary research expertise. This partnership of working with families and with colleagues, both inside and outside of our own organisation, can make for powerful research and bring enormous satisfaction to us all.

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