



# 12

## Palliative and End-of-Life Care for Children with Intellectual Disabilities

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### Understanding Palliative and End-of-Life Care for Children with Intellectual Disabilities

The death of a child at any age is not frequently considered due to the profound sadness it brings to families. Often the difficulties experienced are hidden from view and families can struggle to gain support and understanding regarding their needs and those of their child. The concept of paediatric palliative care focuses on the relief of suffering of the child and family by controlling pain and other distressing symptoms, integrating psychological and spiritual aspects of care, offering a support system to allow children and their families to live as active and full a life

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as possible, and supporting families to help them cope into bereavement (Health Service Executive & Faculty of Paediatrics, RCPI, 2016).

Children's palliative care is an active and total approach to care, from diagnosis or recognition, throughout the child's life, death and beyond. It embraces physical, emotional, social and spiritual elements and focuses on the enhancement of quality of life for the child or young person and support for the family (Together for Short Lives, 2018). This means that children's palliative care seeks to provide a child with the best quality of life possible from the time they are diagnosed with a palliative care need until the point where specialist palliative services are no longer required or until the end of life. Palliative care provides not just physical care such as pain relief and symptom management but also emotional, social and spiritual support encompassing the family in the process (Together for Short Lives, 2018). A common focus of palliative care is the preparation for death that is anticipated (Grinyer, 2012).

## **The Specific Groups of Children with Intellectual Disabilities Requiring Palliative Care**

Although understanding of and reference to children with intellectual disabilities may seem evident, much that is written regarding this cohort suggests a variety of potentially ambiguous terms. These include children with life-limiting conditions (Together for Short Lives, 2018), children with complex care needs (Brenner, Kidston et al., 2018), children who are technology dependent (Brenner et al., 2021), and children with life-limiting neurodevelopmental disabilities (Guerin et al., 2020). Children with intellectual disabilities may have conditions with limitations in the functioning of the brain or neuromuscular system encompassing congenital or acquired conditions, and a variety of neurological, genetic or metabolic aetiologies that can limit the lifespan (Goldstein & Reynolds, 2011; Health Service Executive & Faculty of Paediatrics, RCPI, 2016; Hudson, 2014). Many children with intellectual disabilities have no

formal diagnosis, while some will have more common conditions such as chromosomal abnormalities, severe cerebral palsy and epilepsy.

In determining who might be expected to require children's palliative or end-of-life care, Together for Short Lives (2019) identified four typical groups (Table 12.1).

Group 4 includes those children likely to have intellectual disability. The vast majority of these children live at home with their families and may periodically require hospital and hospice-based care where available (Guerin et al., 2020). Many of these children will undergo frequent hospital and intensive care unit admissions, receive care from multiple subspecialty teams, and often suffer from distressing symptoms (Cohen et al., 2011; Schwantes & O'Brien, 2014; Walter et al., 2013). Care can be fraught with complexity of medical regimes, technology dependence, medication management and frequent health instability (Brenner et al., 2021; Doyle, 2020, 2021; Guerin et al., 2020). As a result, interaction and supports are sought regularly with many different healthcare providers (Courtney et al., 2018; Doyle, 2020).

**Table 12.1** Four groups of children most likely to have palliative care needs by summary of condition

Group	Summary of condition
1	Life-threatening conditions for which curative treatment may be feasible but can fail. Where access to palliative care services may be necessary when treatment fails, children in long-term remission or following successful curative treatment are not included (Examples: cancer, irreversible organ failures of heart, liver, kidney)
2	Conditions where premature death is inevitable, where there may be long periods of intensive treatment aimed at prolonging life and allowing participation in normal activities (Example: cystic fibrosis)
3	Progressive conditions without curative treatment options, where treatment is exclusively palliative and may commonly extend over many years (Examples: Batten disease, mucopolysaccharidoses, muscular dystrophy)
4	Irreversible, but non-progressive conditions causing severe disability leading to susceptibility to health complications and likelihood of premature death (Examples: severe cerebral palsy, multiple disabilities such as those following brain or spinal cord insult)

While the number of children with intellectual disability can be difficult to ascertain, it is estimated that there are 45,325 children between the ages of 0–19 years with complex disabilities (prevalence of 4%) requiring services in Ireland (Health Service Executive & Faculty of Paediatrics, RCPI, 2016). Approximately 2.5% (351,000) of children in the UK are believed to have an intellectual disability (Office for National Statistics, 2019), while Zablotsky et al. (2019) determined that in the US about 1 in 6 (17%) children aged 3–17 years were diagnosed with a developmental disability, with a portion of these specifically identified as intellectual disability (1.2%).

Notably, in contrast to adult palliative care, children with cancer and malignancies constitute a minority of those referred to palliative care services. In Ireland, there are in the region of 490 childhood deaths per year, with approximately 350 deaths per year from life-limiting conditions (Department of Health & Children, 2010). Of these childhood deaths due to life-limiting conditions, the majority occur in the first year of life. Based on the most recent epidemiological data from England, the prevalence of children with life-limiting illnesses has risen over the last 17 years and is predicted to continue to rise in the future (Fraser et al., 2021). It is estimated that there are 8,311 children (0–18 years of age) in Ireland with a life-limiting condition. Of these, 19% (1,579 children) are said to be either unstable (15%), deteriorating (2.2%) or dying in any one year (Fraser et al., 2015). Approximately, 5,000 children die each year in England and Wales, 2,500 as a result of a life-limiting or life-threatening condition (Fraser et al., 2013).

Internationally, malignancies constitute approximately only 22% of referrals to paediatric palliative care, with diseases of the nervous system (39.1%), congenital conditions or illnesses originating in the perinatal period (22.1%) or other illnesses (16.7%) making up the majority (Widger et al., 2007). McMahon et al. (2018) received 83 perinatal referrals over a 4-year period, with chromosomal abnormalities accounting for 35% of diagnoses, congenital heart disease 25%, complex neurological abnormalities 11% and renal agenesis 4%. Children with intellectual disabilities may have specialist palliative care needs at different stages

during the illness trajectory. Referrals are likely to increase as the care of children with complex disability improves and awareness of paediatric palliative care intensifies.

## Children's Palliative Care Differs from Adult Palliative Care

While adult palliative care is well established, palliative care for children is developing. It is not yet widely recognised but gaining some momentum in research and policy in recent years. There are many differences in caring for the dying child versus the dying adult, including:

- The number of children dying is small.
- The conditions are extremely rare with diagnoses specific to childhood.
- Predicting a prognosis can be difficult.
- The palliative phase is often much longer and can be episodic and unpredictable.
- Children may experience several apparently terminal phases.
- Care embraces the whole family and uses a model of family centred care.
- Parents are typically involved in care and require adequate resources to support them with the responsibility of personal and nursing care.
- Siblings are vulnerable and parents must continue to care for them while often providing 24-hour care to a sick child.
- Conditions are sometimes familial—other children in the family may be living with, or have died from, the same condition.
- Children's ability to communicate and understand varies according to their age or stage of development.
- The provision of education and play when a child is sick is essential.
- Grief over the death of a child is known to be more intense, long lasting and complicated.
- Professionals caring for children need specific training and skills to communicate effectively with the child.
- Ethical and legal issues of self-determination (Department of Health & Children, 2010).

For children with intellectual disabilities, due to the complexity of care often arising from rare conditions and unknown trajectories, the palliative phase is often much longer and more unpredictable (Guerin et al., 2020). The provision of palliative and end-of-life care to children with intellectual disabilities can present challenges. Children born with complex care needs may access palliative care for their whole life, however long that trajectory may be with little or no emphasis on a curative phase, unlike children who might present with cancer or malignancies who undergo treatment aimed at cure. The provision of paediatric palliative care and access to adequate services can also be fragmented and inconsistent, whereas models of adult palliative care tend to be well established. This may be due to fewer children requiring these services. The profile of children requiring palliative care has also changed in recent decades due to technological advances and increased survival rates of preterm babies (Department of Health, 2008; Myrhaug et al., 2019).

## Family Centred Approaches to Delivery of Palliative Care

While disability services provide most support to children with intellectual disability and their families, services tend to be chronically under resourced and, as previously noted, palliative care services are also less well developed (Guerin et al., 2020). The aim of support is to provide maximum levels of care as close to home as possible and ensure access to specialist supports when required. Optimal care should be facilitated through integrated service networks with supports from tertiary and quaternary specialties as required (Guerin et al., 2020). The preferred care setting is within the home with minimal interruption of normal routines (Carter et al., 2016; Olsen & Maslin-Prothero, 2001). While this has positive benefits for both the child and family, recognition of the care burden on parents needs to be acknowledged (McCann et al., 2015; Scott, 2000). Approximately 98% of children with intellectual disabilities live at home with their parents (Hourigan et al., 2018). It is assumed that children with disabilities and possible technology dependence will be cared for at home with little or no alternative afforded to parents

(Carnevale et al., 2008; Lindahl & Lindblad, 2011; Mendes, 2013). However, needs can shift over time with the birth of new siblings, the illness of another sibling with the same condition and other life events (Grinyer, 2012), therefore flexibility is an imperative. A range of care settings should be available for delivery of palliative and end-of-life care.

The interface between disability and palliative care services can be problematic and unclear. Universal services such as the general practitioner provide a good foundation with core palliative care services delivered mainly in the community, encompassing a range of services including community nursing and hospice care (Harrop & Edwards, 2013). Specialist palliative care services tend to be delivered through hospice and tertiary specialist paediatric services and are more concerned with symptom control (Harrop & Edwards, 2013).

Palliative care benefits children and families by providing expert symptom management, care coordination and supportive counselling, including advance care planning (Heckford & Beringer, 2014; Miller et al., 2012; Schwantes & O'Brien, 2014). However, identifying children most likely to benefit from paediatric palliative care can pose a challenge. Sometimes referrals are made too late in the illness trajectory (Harrop & Edwards, 2013). This may be due in part to healthcare professionals associating palliative care with dying and death, thereby failing to recognise its role in enhancing quality of life (Twamley et al., 2014). Early integration of paediatric palliative care is recommended by national organisations for a variety of paediatric illnesses and has been shown to be feasible and acceptable (American Academy of Paediatrics, 2013; Berlinger et al., 2013; Department of Health, 2004; Lafond et al., 2015; Mack & Wolfe, 2006). While gaining some momentum in adult palliative care, rarely is advance care planning considered for children as it requires initiation of difficult conversations. However, Liberman et al. (2016) reported that while challenging, early conversations in the outpatient setting can increase access to palliative care for children and often improve knowledge and comfort with the topic.

Approaches to deciding who requires palliative care are important, with one of these being the use of the Paediatric Palliative Screening Scale (PaPaS; Song et al., 2021) (see Resources at the end of this chapter), a tool that can be used with children and their families irrespective of the

origin of their condition. Such an instrument is important in capturing the child's needs at an early stage and assists healthcare professionals to accurately identify children with palliative care needs. This type of assessment captures detail about the illness trajectory and its impact on daily activities, expected outcome of the illness and burden of treatment, symptom burden, preferences of the child and family, and lastly, life expectancy. This tool may be helpful in supporting timely referrals and ultimately be of benefit to the child and family.

The process of referral to palliative care services may initially commence as an information gathering and wayfinding exercise. Referrals may be received as early as the antenatal period where there is identification of life-limiting conditions. Usually, the groupings identified in Table 12.1 assist in categorising those requiring palliative care. However, with palliative care services under resourced, not all children receive optimal services. Therefore, providers need to focus on those children and families that are likely to benefit most from the available paediatric palliative care services, including symptom control and end-of-life care (Table 12.2).

As children with intellectual disabilities may have a long palliative trajectory with many surviving longer than possibly predicted, transition from paediatric to adult palliative care must be considered. This would ideally be seamless in nature, however, can be difficult to achieve. Often strong relationships have been formed through paediatric services meaning a daunting transition for the child and family. Brown et al. (2021) acknowledged that the experience of transition continues to be negative for people with intellectual disabilities, and parents are often forced to drive the fragmented and poorly managed transition process. A good transition requires coordination with a strategic focus but should encompass key elements: start early, offer flexibility and individually tailored to meet the needs of the child and family (Brown et al., 2021).

The need for a multidisciplinary integrative approach to paediatric palliative care is essential in the provision of seamless care. This type of approach encompasses the child and family and is centred on the needs of the individual with the family, often including wider family members and not just the child, siblings and parents. Clear, concise communication is a fundamental component of multidisciplinary working,



**Table 12.2** Paediatric palliative care services and their function

Service type	Function
Respite support	Usually for a defined period /number of nights and may be delivered in the child's own home or in a specified respite facility. Nursing care is usually provided by suitably qualified/experienced staff
Acute admission to hospital or hospice facility	For the management of poorly controlled symptoms usually at short notice. Sometimes initiated for specialist treatment such as syringe drivers, buccal medication, second line opioids or patient controlled analgesia. May be able to return home with support from community health
Supported discharge from hospital	Children already known to palliative care services may be discharged earlier following elective admission for procedures. This can allow for more normality in a less medicalised environment
End-of-life care	This may involve a new referral or emergency referral when a child's status has changed. The child and family should be supported in the environment of their choice (e.g. home, hospital, hospice)
Compassionate withdrawal of life-sustaining treatment	Paediatric palliative care can often support withdrawal of life-sustaining treatment (e.g. ventilatory support) when families and the multidisciplinary team have made a decision that further treatment is futile and not in the best interests of the child
Sibling support	Specific support programmes may be run for siblings/bereaved siblings. This may involve weekend activities/camps aimed at normalising the experience of having a chronically ill sibling

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**Table 12.2** (continued)

Service type	Function
Child and adult bereavement services	Specialist bereavement support may be offered as individual or group sessions. There may also be a remembrance day offered

Adapted from Harrop and Edwards (2013)

with each member holding distinct roles and sharing common goals for the child and family. Working in multidisciplinary teams requires specialist training and education (Bergsträsser et al., 2017). Paediatric palliative care approaches require agreement on the elements of caring and avoidance of conflicting information for the child and family. Each multidisciplinary team should appoint an identified key worker, often a nurse, as the liaison person irrespective of where care delivery takes place (Health Services Executive, 2020).

The composition of the multidisciplinary team varies according to the child's needs, but might include any, or all, of the following:

- The child and family
- Social worker
- Dietician
- General practitioner
- Palliative care consultant and team
- Pharmacist
- Physiotherapist
- Occupational therapist
- Psychologist
- Chaplain
- Teacher
- Play specialist
- Nurses—community/hospital/hospice
- Voluntary organisations

The provision of integrated care across Europe has been widely emphasised in recent years, culminating in the provision of principles to be

adopted for effective, personalised care of children with complex care needs (Brenner, O'Shea, et al., 2018). The three main principles underpinned by a child-centred focus are (1) access to care, (2) co-creation of care with parents and (3) effective integrated governance. These guiding principles offer a benchmark for existing services which providers and policymakers can adopt and use to assess current and future service developments. More recently, Ireland has developed a national framework for clinical governance and operational arrangements for supporting a model of care for children with life-limiting conditions towards the end of life (Health Services Executive, 2020). This marks a fundamental milestone in its explicit recognition of children with intellectual disabilities and their need for paediatric palliative and end-of-life care.

Specialist paediatric palliative care is a relatively new area, with evidence suggesting the benefits of early engagement (Harrop & Edwards, 2013). Specialist roles see nurses in particular working across a range of settings delivering family centred palliative and end-of-life care. Settings include disability services, schools, respite facilities, voluntary homecare providers and tertiary hospital settings. In providing care for the family, the delicate nature of the task must be emphasised including the navigation of key ethical issues, when decisions might be required about life-sustaining treatment or provision of palliative and end-of-life care (Barone & Unguru, 2018; Furingsten et al., 2015). This makes skilled and sensitive communication with the child and family of paramount importance (Marsac et al., 2018; Sharkey et al., 2016; Ulrich et al., 2018).

## Symptom Management

The assessment of symptoms related to palliative or end of life and the evaluation of the outcomes of symptom management require considerable attention to detail and dedication to the task. The single most important skill in assessment is the ability to engage fully and secure trust with the child and their family (Hain et al., 2021; McCrate Protus et al., 2014; Together for Short Lives, 2018). Without this, all other aspects of care are diluted, and the goals of palliative care difficult to achieve.

The professional cannot begin to respond to the experience of the child without meaningful engagement that facilitates a deep understanding of the impact of the illness on the child and family. This is particularly challenging for the child with an intellectual disability where communication and goal setting, in particular end-of-life care wishes, may go undisclosed, unknown or unheard (Feudtner, 2007; Quinn, in press). Parents are recognised as the key advocate for children with a significant disability and require collaborative opportunities to participate in advance care planning about the end of life (Kuo et al., 2011; Quinn, in press; Simon et al., 2010; Vollenbroich et al., 2016).

Assessment is a constant thread in care for individuals receiving palliative care and is an all-important element of care with children with intellectual disabilities. The symptom assessment process is often guided by tools which examine the child and family caregiver experience with a view to addressing current needs and anticipating potential future needs, irrespective of the length of the expected trajectory (Hain et al., 2021).

Key principles to guide symptom assessment include:

- Identify a suitable time and private space to take a history and conduct an assessment.
- Create an unrushed environment. Adapt and use available technology to assist with communication where appropriate.
- Recognise the essential importance of the therapeutic relationship in building rapport and trust with the child and family.
- Do not make assumptions or jump to conclusions. Listen, see, sense and feel all that is told to you (and not told to you) and make a clear, informed, unbiased clinical judgement.
- Ask the child/parent/caregiver to describe the symptom in detail. Parent/caregiver reporting about the child with intellectual disability is key (Jassal & Hain, 2016; McCrate Protus et al., 2014).

In assessing symptoms, it is important to consider both qualitative and quantitative aspects of the symptom. Some aspects of symptoms can only be assessed through a qualitative approach. For example, how pain impacts on the child's sleep, diet, mood etc. can be explored with the

child and their parents/ caregivers, but cannot be quantified or measured. Other aspects of symptoms are quantifiable. For example, the severity of pain can be measured using pain intensity rating scales. A variety of assessment tools and computer-based mobile apps have also been developed in recent years and may assist the identification and experience of symptoms (Brock et al., 2018). When assessing symptoms that are indicative of the end of life it is important the following considerations are addressed:

- Time of onset of symptom.
- Associations with the symptom, e.g. movement, eating etc.
- Duration of symptom, e.g. is it constant or episodic?
- Frequency and severity of symptom.
- Symptom associations: What exacerbates the symptom? What relieves the symptom? Impact on quality of life, e.g. sleep, mood, function, social interaction.
- Believe the child or parent/caregivers responses about the symptom.
- What meaning do the child and family attach to the symptom.
- A full physical examination.
- Current medications/treatments noted.
- Whether any new medications or treatments have been recently started.
- Consider the use of diversional or alternative approaches to assessment e.g. through play.
- Consider 'break periods' for the child and family if required.
- Make allowances for communication challenges or non-English speaking children and families. Consider the use specific assessment tools that can assist with identification of symptoms.
- Document all findings (McCrate Protus et al., 2014).

The assessment of specific symptoms should result in the identification of possible cause and approach to treatment that can be implemented. The most common symptoms in palliative and end-of-life care for children with intellectual disabilities are outlined in Table 12.3, including their cause and associated treatment approach.

**Table 12.3** Common symptoms by cause by palliative and end-of-life care treatment approaches

Symptom	Cause	Treatment approaches
Pain	Between 25 and 46% of children experience persistent chronic pain, incidence is likely higher in the palliative child. Numerous and multifactorial in causation	Team approach Non-pharmacological—physical, complementary and cognitive behavioural interventions may reduce the perception of pain, e.g. acupuncture, art therapy, aromatherapy, heat/cold application, humour clown therapy, storytelling oral motor stimulation (sucking) Pharmacological—wide range of approaches based on the child's understood pain level; by the clock; less invasive route; tailored to child's unique circumstances and needs
Agitation and delirium	Life-threatening illness can cause widespread cerebral dysfunction (biochemical, disease). Terminal agitation, also known as terminal restlessness, is agitated delirium at the end of life caused by impaired consciousness. Other causes are medication related, infection, organ failure or psychosocial	Pharmacological—medications to relieve anxiety Non-pharmacological—e.g. acupressure, acupuncture, massage, music/diversion therapies
Anorexia and cachexia	Dietary, disease related, gastrointestinal, iatrogenic (chemotherapy, radiation, medications, opioid induced constipation)	Team approach Family/carer education Pharmacological approaches are limited in children but may be considered, e.g. anti-emetic, appetite stimulant. Steroids may also assist in a limited capacity

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Table 12.3 (continued)

Symptom	Cause	Treatment approaches
Bowel patterns changes	Mechanical— physiological changes i.e. behavioural, dehydration, disease and medication related, metabolic, hyper/hypocalcaemia, hypothyroidism, neurologic with damage to nerve pathways, extension of disease tumours Non-mechanical— changes in normal intestinal muscle activity	Team approach requiring dietary review Pharmacological and non-pharmacological treatments may be considered
Cough/excessive secretions	Allergens, cardiac (heart failure), infection, malignancy, medications (ACE inhibitors, antibiotics, inhaled steroids, radiation therapy), pulmonary. Psychosocial (psychogenic cough), Tourettes and other (aspiration, reflux, secretions, seizures, rheumatic disease) Excessive secretions can be caused by over production of saliva, inability to retain saliva in the mouth and difficulty swallowing	Non-pharmacological treatments—e.g. fluids, honey, saline nasal washes, humidified air, lozenges (if appropriate), upright position, eliminate antigens with air filters, vapour rubs, acupuncture or acupuncture, chest physiotherapy, airway clearance (with training). Fluid intake requires assessment Pharmacological approaches can assist/inhibit cough reflex or reduce sensitivity Medication to thin or dry up secretions can be considered (anticholinergic)
Gastroesophageal reflux and dyspepsia	Anatomical, diet, medications, motility, other (smoke related)	Team approach Positioning Bland diet Small frequent meals

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Table 12.3 (continued)

Symptom	Cause	Treatment approaches
Nausea and vomiting	Anxiety, anatomical, bowel obstruction, constipation, gastrostasis, infection, increased intracranial pressure, pain, medications, metabolic disorders (hypercalcaemia, renal failure), radiation, reflux	Non-pharmacological—e.g. acupuncture, avoid triggers, eliminate medications if possible, play/distraction, good oral care, clear liquids, positioning, relaxation techniques Pharmacological approaches
Seizures	Nervous system anomalies (known/deterioration), comprehensive system decline (chronic or acute), metabolic deterioration (hypoglycaemia, hypocalcaemia, hyper/hyponatremia)	Team approach Assessment is key Non-pharmacological—e.g., positioning, monitoring, education and reassessment Pharmacological—e.g., advance planning for emergency medication application, titration and effect
Dysnoea	Physical, psychosocial, spiritual factors may contribute Causes are extensive and have multiple contributing factors Underlying causes may be treated successfully, others cannot be reversed and treatment is palliative	Non-pharmacological approaches can have significant success, eg., breathing strategies. Use of fans or open windows, repositioning, relaxation, minimise triggers (strong perfume, smoke) Ensure companionship Pharmacological management is aimed at decreasing the perception of dyspnoea and considers the use of benzodiazepines and opioids Oxygen therapy may be helpful but is rarely beneficial at the end of life and hinders communication and connection

(continued)



**Table 12.3** (continued)

Symptom	Cause	Treatment approaches
Communication	May already have difficulty in communication and require additional supports	Ensure all staff have knowledge of and are familiar with communication method

Hain and Jassal (2010), Friedrichsdorf et al. (2015), McCrate Protus et al. (2014), National Institute for Health and Care Excellence (NICE) (2019), and Zadeh et al. (2015)

To conclude this section, it is essential symptoms are reassessed regularly. Reassessment includes attention to symptoms already assessed, responses to treatment, screening for new symptoms and medication side effects. In sum:

- Listen to both the child and their parent/carer.
- Work with the parent/carer as experts in their child's care and be open to new approaches/assessment.
- Keep approaches simple and child/family focussed.
- Pay close attention to detail.
- Work as a team.
- Keep all lines of communication open at all times.
- Know your own limits, scope of practice and ask for help.

## Ethical Considerations in End-of-Life Decision-Making

A great deal of stress can be experienced by the family when discussions concerning possible need for palliative and end-of-life care occur. Decision-making where all stakeholders are in agreement is the ideal. Ethical concerns can arise concerning treatment options or withholding treatment and resuscitation, or other conflicts between parents and with health professionals (Hain et al., 2021). Families likely have significant knowledge and experiences that influence their decision-making

processes (Mitchell et al., 2019), but most remain unprepared for end-of-life decision-making. Listening skills by health professionals, including awareness of both their own verbal and non-verbal communication as well as others, cannot be overemphasised (Inglin et al., 2011; Mitchell et al., 2019). Information exchange should be treated as an honest and reciprocal process, while being sensitive to the child and family needs (Weidner et al., 2011; Winger et al., 2020). While end-of-life care decision-making for children with intellectual disabilities is complex and emotionally overwhelming, preparation becomes possible if parents reach a place of acceptance (Mitchell et al., 2019). Amy's story demonstrates the importance of addressing key symptoms, with an appropriate management approach co-created and agreed with parents.

**Amy's\* story**

As Amy's condition started to deteriorate, she began to experience several uncomfortable symptoms requiring careful management to alleviate and ensure comfort. Amy had always experienced seizures (although usually well controlled), but with the physical deterioration in her condition, the seizures became more intense. Seizure management goals were discussed with Amy's parents by the lead paediatrician and community palliative care team with a focus on Amy's comfort, dignity and quality of life during this palliative phase. Medications were reviewed and titration dosages were considered in response to anticipated disease progression and expected management, especially during the end-of-life period. Due to seizure activity, increased irritability was another concern, with Amy showing her frequent distress with a prolonged distressed cry. This was upsetting for her, as well as her parents, wider family and friends. The lead paediatrician and community palliative care team had a further discussion with Amy's parents where anxiolytics and pain management medications were explored, and a plan put in place. Over the next few days, as Amy's condition deteriorated as anticipated, cessation of feeding (diet and hydration) was discussed with Amy's family. This difficult decision had the ultimate goal to reduce an unnecessary burden on Amy's body, especially the management of secretions which became an increasing concern during the last days of life. While this was an

extremely difficult discussion for Amy's parents, together everyone recognised that the delicate balancing of hydration with the challenges of maintaining a good gentle death was paramount. Amy died peacefully with her symptoms well controlled. Amy's parents and a night nurse were in attendance.

\*Assumed names are used in this story.

## Bereavement Support

Bereavement has been referred to as the grief experienced after a loss, especially of a loved one, and includes emotional, psychological, physical, behavioural and functional reactions (Weafer, 2014). This understanding of bereavement allows for a broader consideration of core issues for many families around loss which may have accompanied the initial diagnosis of earlier childhood disability, with the ultimate death of the child being the second loss (Zisook & Shear, 2009). The specific experiences of parents of children with intellectual disability losing their child are not well explored in the literature (Reilly et al., 2008). Grief, if prolonged, can become a pathological condition that may require psychological intervention to promote recovery (Comtesse et al., 2020).

Contrary to earlier views around grief, a bereaved parent's ability to reconstruct their relationship with their deceased child and integrate this memory into their inner and social worlds is now considered central to adjustment following bereavement (McNeil et al., 2020). Possessions and physical objects offer links to the deceased child and evoke memories, assisting to keep their child intact (Papadatou et al., 2021). These memories support families to recount their feelings, thoughts and events following a bereavement, assisting them to adjust while giving meaning to their loss and remaining connected to their loved one (Papadatou et al., 2021). The idea of continuing bonds may require professional support if parents experience difficulty talking about their deceased child to family members and the wider social world. The inability to talk about the deceased child can mean loneliness and isolation (De Clercq et al.,

2017; Papadatou et al., 2021). Healthcare professionals can support parents and families through therapeutic interventions like listening, conversational remembering and referring families to support groups of those who have suffered the same loss. Grief care is not solely allocated to after the death as it is recognised that grief can begin at illness diagnosis, known as anticipatory grief (see also Chapter 2). This is particularly prolonged for families of a child with intellectual disability whereby the trajectory is unknown.

Parental bereavement literature acknowledges that for families losing a child is a tragedy and evokes a significant emotional response (Hannan & Gibson, 2005), implying that those bereaved are at higher risk of ill health when unable to cope with their grief and loss (Johns, 2016; Keegan, 2013; Papadatou et al., 2021; Tatsuno et al., 2012). Additionally, the experience of child bereavement for both parents and other carers can present as emotional distress, reduced life expectancy or reduced overall quality of life and increased anxiety (Eilertsen et al., 2013). However, the experience can also allow for post traumatic growth for families, through creating an opportunity for them to find meaning after their great loss (Barrera et al., 2009). While not all families and caregivers will require professional support, it is recommended that supports and services are available as required. This support can range from a caring listener to requiring more structured assistance. Either way, there is a need to respect and support the family's natural coping mechanisms.

## Conclusion

Appropriate healthcare services are vital for the health and wellbeing of children with intellectual disabilities, including for children who are dying. This should incorporate a family centred model of palliative and end-of-life care which responds to all stages of the palliative trajectory and extends from diagnosis to beyond death. Palliative care can be seen as a philosophy of care that stretches across a broad range of services, where the primary goal is to make a child's life as comfortable as possible while experiencing palliative symptoms and end-of-life needs. This care provides supports to the child and family and can range from emotional

support to palliative care services within the home or elsewhere. Care also encompasses support for bereavement and loss and adjusting to life after losing a child. Quality care requires a cohesive, collaborative and fully integrated partnership inclusive of all professionals together with the child and their family.

## Reflection

### **Bella's\* story**

Bella is 4 years old and has profound intellectual disabilities and complex health needs. She lives 3 hour away from the main children's hospital but attends a special needs playschool and is well established within the health care services and early intervention team. On a planned admission, Bella is admitted for a review of her metabolic disorder and deteriorating symptoms. Unfortunately, investigations show that she is doing less well and her condition is no longer amenable to treatment. Her organs are now demonstrating dysfunction and she is significantly more symptomatic. It is expected she will continue to deteriorate in the coming weeks. In view of further treatment not being undertaken, Bella's treating paediatrician discusses the new findings and implications with her family. Her parents are understandably devastated and wish her to be discharged home immediately. A paediatric palliative care assessment is undertaken. Bella and her parents are involved in the plan and wish to engage with the paediatric team at her nearest paediatric hospital unit, seek help from the community Specialist Palliative Care Team (SPCT) and allied colleagues to ensure they can obtain help and support. A teleconference is to be held prior to Bella's return home. Her parents are made aware of the need to seek home nursing supports including alongside Bella's current disability services (for respite and continuity with family support), as well as the (new) contribution of the SPCT and the Paediatrician in symptom management. It is agreed the overall management will be co-ordinated locally by a Clinical Nurse Co-ordinator (CNC) for children with life-limiting illness. This nurse liaises with the family directly to offer information and guide access to voluntary home nursing services, in home respite care and end-of-life nursing hours from the

Cancer Society Night Nursing Services. Bella's death is not expected in the immediate hours or days following this discharge, but it is made clear that her condition may deteriorate suddenly. The CNC, paediatrician, SPCT, other disciplinary colleagues and Bella's family engage in a teleconference to agree a plan of care and relevant documentation is shared securely via email. Communication is accelerated and Bella and her parents leave the tertiary hospital with a hard copy of their referral documentation forwarded to all teams including the general practitioner, the paediatric team in their nearest hospital unit, the community SPCT, the CNC and public health nurse. Voluntary sector service referrals are completed remotely by the CNC and submitted with parents involvement and expressed consent. A team approach to end-of-life care is expedited to ensure that Bella's requirements and the needs of her parents and family are fully met as they journey towards her end of life.

Bella lived for four more months with the coordination managed by the CNC with the focus on her quality of life and making memories, guided by the collaborative effort of both voluntary and statutory providers. This collaborative effort ensured that Bella and her family remained connected to established services and personnel (who had become familiar and valued by the family). These professionals (public health, community nurses and disability providers) had previously attended various educational programmes to ensure they had an awareness and knowledge of the principles of children's palliative care and the competencies required to deliver skilled palliative and end-of-life care. The participation and contribution of previously known professionals (including respite carers and allied health colleagues) had the benefit of consistency in care giving and family support, recognising Bella's individual needs as well as changes or new symptoms as they arose. With this full team effort and excellent interdisciplinary communication Bella was able to remain in her preferred home setting, even as her physical condition deteriorated, with providers contributing their unique skill set with the overall aim to support Bella and her family to her final days.

\*Assumed names are used in this story.

## Questions

1. Consider how this example of paediatric and end-of-life care offers support to the child and family.
2. An integrated family centred home-based plan of care is described. Consider the role of each person involved in the care process.
3. What are the key competencies that professionals working with children with intellectual disabilities can contribute to in the end-of-life period?

## Resources

1. *Paediatric Palliative Screening Scale (PaPaS)*. A tool for screening paediatric patients for palliative care needs. Useful for primary care clinicians unfamiliar with palliative care. Not purposefully designed to accommodate the individual needs of people with intellectual disability. <https://bmcpalliatcare.biomedcentral.com/articles/10.1186/s12904-021-00765-8>
2. *Together for short lives*. A website of resources to support children who are dying and their families. While not purposefully designed for children with intellectual disability, the information is helpful. <https://www.togetherforshortlives.org.uk/>

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