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Palliative Care and Intellectual Disability

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Abstract

People with intellectual disabilities make up an estimated 1–3% of the population. This is an aging population, with an associated increasing need for palliative care provision.

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However, many do not have equitable access to palliative care services, for a variety of reasons. They often have issues, challenges, and circumstances that make it particularly difficult to meet their palliative care needs. This includes communication difficulties which affect all the aspects of palliative care provision; difficulties around insight and the ability to participate in decision making; unconventional ways of expressing signs and symptoms of ill health and distress; multiple comorbidities; complex family and social circumstances; and higher levels of behavioral or psychiatric problems.

This chapter describes the unique challenges in meeting the needs of people with intellectual disabilities at the end of life. It is based on the White Paper on Intellectual Disabilities, published by the European Association of Palliative Care in 2015. It addresses the following key areas: equity of access; communication; recognizing the need for palliative care; assessment of total needs; symptom management; end of life decision-making; involving those who matter; collaboration; support for families and carers; preparing for death; bereavement support; education and training; developing and managing services. This provides a comprehensive overview of the current state of the art.

1 Introduction

There has been a growing recognition in recent years of the importance of focusing attention on the palliative care needs of people with intellectual disabilities. The life expectancy of people with intellectual disabilities has increased significantly over the past 50 years (Patja et al. 2000). The increase in life expectancy for people with Down syndrome has been particularly marked, from 12 in 1949 to nearly 60 in 2004 (Bittles and Glasson 2004). This dramatic shift has been attributed to reduced childhood mortality and to better knowledge, healthcare, advocacy, and services (Yang et al. 2002; Haveman et al. 2009).

As people with intellectual disabilities are living longer, they are more likely to die of illnesses usually associated with old age, and they more likely to need a period of palliative care (Tuffrey-Wijne 2003). The aim of this chapter is to describe the unique challenges in meeting the needs of people with intellectual disabilities who require palliative care, as well as important considerations in addressing those challenges. It is based on the White Paper published by the European Association for Palliative Care (EAPC) in 2015, which sets out aspirational norms in 13 areas of practice. These norms were reached through using the Delphi method to reach consensus, involving 92 professionals in 15 countries who had expertise in the fields of palliative care, intellectual disabilities, or both. The full methods and detailed norms can be found in the White Paper itself (Tuffrey-Wijne and McLaughlin 2015) and an accompanying open access paper (Tuffrey-Wijne et al. 2015). Here, the focus is on describing the relevant issues under the following headings, in line with the 13 norms:

1. Equity of access
2. Communication
3. Recognizing the need for palliative care
4. Assessment of total needs
5. Symptom management
6. End-of-life decision making
7. Involving those who matter: families, friends, and carers
8. Collaboration
9. Support for families and carers
10. Preparing for death
11. Bereavement support
12. Education and training
13. Developing and managing services

First, the context of these issues will be set out through describing the prevalence of intellectual disabilities and the profile of illness and dying among this population.

2 Background

2.1 Definition

Intellectual disability is characterized by significantly impaired intellectual and adaptive functioning. Someone has intellectual disabilities if the following three aspects are present simultaneously: (1) a significantly reduced ability to understand new or complex information and to learn and apply new skills (impaired intelligence); (2) a significantly reduced ability to cope independently, expressed in conceptual, social, and practical adaptive skills (impaired adaptive functioning); and (3) onset before the age of 18, with a lasting effect on development (American Association on Intellectual and Developmental Disabilities 2013a).

This definition covers a large and heterogeneous group of people with a wide range of skills and limitations. On one end of the spectrum, it includes people with mild intellectual disabilities who may be able to function in society with little or no support and may have good communication skills. Sometimes, it is only when the equilibrium of life is disturbed (e.g., when their health fails) that their independent coping is challenged. On the other end of the spectrum, people with profound intellectual disabilities have significant and multiple impairments, usually including physical impairments; they will need 24 h support.

The term "intellectual disability" is currently most widely accepted across the world, replacing earlier terms including "mental retardation" (Schalock et al. 2010). In the UK, the term "learning disabilities" is used synonymously with "intellectual disabilities," but this can be confusing internationally. In the USA, for example, "learning disabilities" refers simply to weaknesses in certain academic skills, such as reading or writing. "Developmental disabilities" is an umbrella term that includes intellectual disabilities but also includes other disabilities that are apparent during childhood, such as cerebral palsy or epilepsy; they are severe chronic disabilities that can be physical, cognitive, or both (American Association on Intellectual and Developmental Disabilities 2013b).

An estimated 20–30% of adults with intellectual disabilities also have an autistic spectrum disorder (ASD). ASD is a lifelong condition that affects how a person communicates with, and relates to, other people. It is characterized by difficulties with social communication, social interaction, and social imagination (Emerson and Baines 2010). Not all people with ASD have intellectual disabilities. For example, people with Asperger's syndrome (a form of autism) have average or above-average intelligence, and therefore do not have intellectual disabilities.

2.2 Prevalence

Intellectual disability affects an estimated 1–3% of the population (Mash and Wolfe 2004). The exact prevalence is unknown, as there is little standardization of definitions or methods of data collection and there is a general lack of statistical information.

People with mild intellectual disabilities make up around 85% of the total population of people with intellectual disabilities (Department of Health 2001). There is a higher incidence of mild to moderate intellectual disabilities in deprived areas. It is difficult to establish causal effects with certainty. Exposure to socioeconomic adversity prenatally and in the early years of development is likely to increase the incidence of intellectual disability. It is also possible that the heritability of intellectual ability, and the link between low intellectual ability and social position, contributes to a higher incidence of intellectual disabilities in the areas of social and economic deprivation (Emerson 2012).

Many people with intellectual disabilities, especially those with mild and moderate intellectual disabilities, are not known to specialist services and may never have been diagnosed as having intellectual disabilities (Learning Disabilities Observatory 2016). This is therefore a largely hidden population. The fact that people's disabilities may not be recorded, supported, or even recognized creates particular challenges for services trying to meet their needs at the end of life. Some risk being labelled as "difficult" or "uncooperative," whereas their behavior or coping strategies may simply be due to undiagnosed intellectual disabilities.

2.3 Death and People with Intellectual Disabilities

2.3.1 Life Expectancy

Although life expectancy for people with intellectual disabilities has increased dramatically over the past century, it is still significantly below that of the general population. A recent government inquiry in England investigated the deaths of 247 people with intellectual disabilities between 2010 and 2012 (the CIPOLD study: Heslop et al. 2013). This important study, which was comprehensive and methodologically sound, has informed several parts of the EAPC White Paper and this chapter. A key finding was that the median age of death for people with intellectual disabilities (65 years for men; 63 years for women) was, on average, 16 years younger than the general population. Similar statistics have been found in the USA, where there have been mortality reviews for people with intellectual disabilities since 2004. The average age of death in 2014 was 59 (Connecticut State Department of Developmental Services 2015).

This shorter life expectancy may be partly due to factors related to the intellectual disability itself. For example, some conditions that cause intellectual disabilities can also cause significant physical health issues, which may be life-shortening. Some are related to the premature birth of babies that would not have survived in the past, but are now living into childhood or beyond. A significant proportion of the population of people with intellectual disabilities have a specific syndrome, which can be genetic (Down syndrome is the most common chromosomal disorder) or caused by toxins, injuries, infections, and genetic/metabolic disorders which can affect the central nervous system or other organ systems during the developmental period. These effects can become evident during the person's life (Evenhuis et al. 2001). For example, there are high rates of cardiovascular disease and diabetes among adults with Prader-Willi syndrome, arising from morbid obesity (Greenswag 1987).

However, there is sound evidence that the shorter life expectancy of people with intellectual disabilities is not just related to factors inherent in the presence of intellectual disabilities itself. This population experiences substantial health inequalities, leading to poorer outcomes (Emerson and Hatton 2013). There are inequalities in healthcare provision, including poorer access to palliative care services. People with intellectual disabilities, therefore, are at risk of premature death that could be amenable to better healthcare provision (Heslop et al. 2013).

2.3.2 Causes of Death

Leading causes of death among people with intellectual disabilities are respiratory disease, heart disease, and cancer. International data on cancer deaths among people with intellectual disabilities are lacking. In the CIPOLD study, cancer accounted for 20% of deaths among people with intellectual disabilities. The cancer profile is slightly different from the general population, with a higher than average incidence of gastrointestinal cancers (Hogg and Tuffrey-Wijne 2008). People with Down syndrome have a significantly increased risk of leukemia and a lower risk of many solid tumors (Satgé and Vekemans 2011).

The incidence of dementia is higher among people with intellectual disabilities (Strydom et al. 2010). In particular, the incidence of Alzheimer's disease is high among people with Down syndrome, with incidence rising sharply between the ages of 40 and 60. Around 40% of people with Down syndrome aged 60 and over suffer from the condition (although exact prevalence estimates vary). It is thought that there is an association between the presence of the third chromosome 21 and the production of the beta-amyloid protein which is involved in Alzheimer's disease.

3 Palliative Care for People with Intellectual Disabilities

Palliative care sets out to preserve the best possible quality of life until death. This involves management of pain and other symptoms, and of social, psychological, and spiritual problems. It requires an approach that encompasses the patient, the family, and the community in its scope (European Association for Palliative Care 1998).

The palliative care needs of people with intellectual disabilities are, on the face of it, no different from those of the general population. However, they often present with unique issues, challenges, and circumstances that make it much more difficult to meet those needs. This includes, for example: communication difficulties which affect all aspects of palliative care provision; difficulties around insight and the ability to participate in decision-making; unconventional ways of expressing signs and symptoms of ill health and distress; multiple comorbidities; complex family and social circumstances; and higher levels of behavioral or psychiatric problems.

In order to promote best practice, it is worth considering the 13 areas of practice and service delivery that are set out below.

The scope of the White Paper on which this chapter is based includes patients who are *adults* (children require a particular and additional focus); across *the entire spectrum of intellectual disabilities* (ranging from mild to profound); and *in a wide range of settings*, including the family home, independent living arrangements, residential care settings, nursing homes, hospitals, and specialist palliative care settings.

3.1 Equity of Access

3.1.1 Barriers

The barriers people with intellectual disabilities face in accessing health services, including palliative care services, arise from a number of different sources (Emerson and Hatton 2013). Some of these are related to late diagnosis of life-limiting illness such as cancer. People with intellectual disabilities themselves may not recognize the signs and symptoms of ill health, or they may be less able to communicate these signs effectively to others. They may also not appreciate the importance of taking up health screening.

It has often been asserted that late diagnosis is due, in large part, to family members or paid caregivers not realizing that something was wrong (Tuffrey-Wijne et al. 2007b). However, the CIPOLD study found that the majority of people with intellectual disabilities who died had been identified as being unwell prior to the diagnosis and treatment of their final illness, either by themselves, a family member or a paid carer; in most cases, medical attention had been sought in a timely way. However, there were significant problems with making a correct diagnosis. Frequently, the investigations that were needed to diagnose the problem were not done or posed difficulties. Physicians were more likely to take a "wait and see" approach. In a quarter of those identified as being unwell and who responded appropriately, the concerns of the person with intellectual disabilities, their family, or paid care staff were reportedly not taken seriously enough by medical professionals. Families of people with intellectual disabilities were significantly more likely than those of people without intellectual disabilities to not feel listened to; this finding echoed previous reports (Michael 2008).

Barriers can be created by attitudes and a lack of knowledge of clinicians and carers. Those working in generic health or social care settings may lack training and knowledge of intellectual disabilities. There is a risk of professionals attributing the signs and symptoms of ill health (which may take uncharacteristic forms of expression) to the intellectual disability itself rather than to the underlying illness – a phenomenon known as "diagnostic overshadowing" (Reiss and Syzszko 1983).

Another barrier may simply be that those working in palliative care services do not know the population of people with intellectual disabilities in their catchment areas and are therefore unlikely to reach out to them. Among those working with people with intellectual disabilities, there may be a misconception about hospice and palliative care services as being concerned only with the final stages of dying, rather than with helping people to live and cope with the life they have left. It may not be known to families and support staff that palliative care can be provided within people's own homes.

3.1.2 Reasonable Adjustments

Equitable access to health care is an internationally recognized human right (United Nations 2006). In Great Britain, the requirement to make "reasonable adjustments" to healthcare services, in order to make them accessible to people with disabilities, is enshrined in law (Disability Discrimination Act 2005). The underlying principle of equality is not usually disputed, but it can be difficult for palliative care services to know what changes they have to make in order to provide equal access to all patients. The need to remove physical barriers (such as providing lifts and ramps) may be easily understood, but it is important also to include changes to the ways in which services are delivered, so they work well for people with intellectual disabilities. In order to do so, services will have to recognize the specific additional needs of people with intellectual disabilities. Examples of reasonable adjustments for people with intellectual disabilities, which can be made by generic healthcare services or specialist palliative care services, include:

- Giving people information that is tailored to their communication needs (e.g., providing easy-read materials and pictures, or opportunities to see clinical areas or equipment beforehand)
- Allowing more time
- Involving family and other care givers
- Providing staff training about the needs of people with intellectual disabilities
- Accessing expertise about intellectual disability when needed (e.g., by engaging with intellectual disability nurses)

It is important to acknowledge that different countries have different ways in which health services are delivered to people with intellectual disabilities, which may affect the kinds of adjustments that may be needed. In the UK, Public Health England (2016) has an online database of reasonable adjustments provided by healthcare services, including tools and resources (Public Health England 2016).

3.2 Communication

Most people with intellectual disabilities, even at the mild or moderate end of the spectrum, will have some difficulty with communication. This can include any or a combination of the following (Iacono and Johnson 2004):

- Speech that is difficult to understand
- Problems in understanding what is said
- Problems in expressing themselves because of limited (or even absent) vocabulary and sentence formulation skills

These problems need to be recognized and taken into consideration. It is not surprising that difficulties with communication are often highlighted as one of the main reasons why palliative care provision for people with intellectual disabilities is so difficult (Tuffrey-Wijne and McEnhill 2008). It affects assessment of pain and other symptoms; the provision of emotional, social, and spiritual support; truth disclosure; and issues around consent and decision making.

Many people with intellectual disabilities benefit from communication aids to augment their spoken language, such as *objects of reference* (e.g., being shown a cup to signify drinks), *signs* (there are some specific sign languages used by people with intellectual disabilities, such as Makaton and Signalong), or *symbol-based systems* (including photograph and line drawings). Picture books, such as *Am I Going To Die?* from the *Books Beyond Words* series (books designed to help adults with intellectual disabilities understand and talk about difficult issues, see www.booksbeyondwords.org) can be useful.

However, some people with intellectual disabilities, especially those at the severe and profound end of the spectrum, do not easily understand either words or pictures. They have high individual communication needs, and it is imperative to involve family and other care givers in interpreting their behavior. As Thurman et al. (2005) describe:

> They may be unable to ask for things that are not actually present and are dependent on others to present them with the real tangible items... [they] can only react to situations as they arise. Such reactive communicative behaviour is often interpreted as challenging (for example, "He spits his food out on purpose").

It is important, therefore, to see any unconventional or "challenging" behavior as a possible message that the person is trying to communicate

– and to become a “detective,” trying to interpret this unconventional communication correctly, together with those who know the person well.

3.2.1 Truth Disclosure

Many people with intellectual disabilities are being protected from knowing that their illness is expected to lead to their death. In one study, staff and families gave the following reasons for non-disclosure: “He will get upset”; “I will get upset”; “He can’t understand”; “He has no concept of time”; and “Others don’t want him told.” Reasons for disclosure were related to the person’s rights (“He has a right to know”), their coping (“Understanding will help him cope”), and involvement (“He needs to be able to plan and make decisions”) (Tuffrey-Wijne et al. 2013). Similarly, people with intellectual disabilities are often not prepared for the death of someone close to them. Staff who work with people with intellectual disabilities usually talk to them about death *after* the death of someone close to them has occurred, but not beforehand (Ryan et al. 2011). This is especially poignant for people with intellectual disabilities who are themselves dying; they are not offered opportunities to engage with the topic of death unless they themselves initiate the conversation (Wiese et al. 2013).

It is important to make no assumptions about how much someone has understood. It is important to take people’s life experiences into account, which will affect how someone makes sense of new information. This is illustrated by the following example:

> Dale, living with and caring for his remaining terminally ill parent told me in response to the question ‘What is cancer?’ that he had learned about it at school and that it was ‘a disease the grows in your body, in your lungs and other places’. When I asked him whether it a serious illness he said ‘Yes, very serious’ but when I asked him whether he had then expected that his father would die of the disease, he said ‘No I never expected that, no one told me’. Now faced with his mother’s illness I asked him what he had thought when he had been told that she had cancer, he said ‘I just froze, I thought, I am going to be on my own’. (McEnhill 2008).

There is little evidence within the literature that truth disclosure can be harmful for people with intellectual disabilities who are at the end of life, but research in this area is very limited. One study has suggested that for some people, full knowledge of what will happen in the future could be overwhelming, particularly if they are unable to put the information into the perspective of a time frame. The concepts of illness, treatments, and deaths might be too abstract to understand, which could cause severe distress for some people. Some people have high levels of anxiety, which makes it difficult to cope with distressing information. Any decision *not* to disclose the truth needs to be taken in the person’s best interest, after careful consideration by everyone involved (especially those who know the person well), and reviewed regularly (Tuffrey-Wijne et al. 2013).

In recent years, a new model has been developed for breaking bad news to people with intellectual disabilities (Tuffrey-Wijne et al. 2012; www.breakingbadnews.org). This is based on evidence that the widely taught step-by-step approach to breaking bad news (Kaye 1996; Baile et al. 2000) doesn’t work well for people with intellectual disabilities. For example, “finding out how much the patient already knows” can be difficult. “Warning shots” preceding disclosure of bad news can be confusing or even alarming for people with intellectual disabilities. Traditional models for breaking bad news do not take into consideration that people with intellectual disabilities usually begin to make sense of their situation (and the bad news) in their own environment, rather than in a doctor’s office. Families and other care givers are often involved in disclosure of bad news, and they may find this particularly challenging. The new model takes account of the person’s understanding and capacity, the people involved in the situation, and everyone’s support needs. It is based on the premise that news needs to be broken down in very small chunks and added gradually, in order to build someone’s understanding. This is different from “warning shots,” even if it looks similar. Warning shots tend to be given in order to make the person aware that the news is bad. Telling someone “Dad is not going to get better” as a way of getting someone to ask or understand “Dad is going to die” is a warning shot – it’s much better, in that case, simply to say “Dad is going to die.” However, “Dad is not going to get better” could also be used in order to help someone understand what is happening with Dad’s illness. It may be too early to tell someone “Dad is going to die” (especially if they have a poor sense of time), but when that time comes, this earlier bit of information will help the person to make sense of the situation.

3.3 Recognizing the Need for Palliative Care

Poor access to palliative care services may be due to a lack of recognition by those that support people with intellectual disabilities that palliative care is needed – or even, that palliative care services exist.

Predicting a need for palliative care can be particularly difficult when someone has intellectual disabilities (Vrijmoeth et al. 2016). This is complicated by the fact that prognostication can be challenging, as many people with intellectual disabilities have a range of comorbidities, such as epilepsy. Those with congenital conditions may have had complex health problems throughout their lives, so it can be hard to know when lifelong and ongoing management of these problems turns into a need for palliative and end-of-life care.

It may be much more important, therefore, to take an approach that does not rely too heavily on prognostic indicators. Commonly used indicators for identifying those in need of palliative care can still be very useful in predicting mortality, including the “Surprise Question” (“Would you be surprised if this person were to die in the next 6–12 months?” (Moss et al. 2010). General and specific indicators can all lead to the answer being “No, I wouldn’t be surprised”: general physical decline, decreasing activity, progressive weight loss, repeated hospital admissions; cancer, organ failure, dementia). But more important is the anticipation and meeting of likely needs, “hoping for the best but preparing for the worst.” There should be a proactive, even instinctive prediction of the rate and course of decline, and a regular review of the situation (Thomas et al. 2011).

3.4 Assessment of Total Needs

The unconventional way in which many people with intellectual disabilities express their emotional, social, spiritual, and physical needs means that their needs can be easily overlooked. In addition, their emotional capacities – including the capacity to cope with illness, death, and loss – are often underestimated.

In assessing someone’s needs, it is essential to have an understanding of that person’s experience of life. Here are some examples of relevant past life experience:

- Many people with intellectual disabilities have a life-long experience of being dependent on others. For some, this can lead to resilience and an ability to accept the need for increased care. Others, particularly those on the autistic spectrum, may find a change of circumstances and routines much more difficult to cope with.
- Many people with intellectual disabilities have not had extensive opportunities to make even the most basic of choices. Discussing different care or treatment options may not make much sense to people who have never been involved in deciding what to have for dinner.

There is evidence that spirituality plays a significant role in the lives of people with intellectual disabilities (Swinton 2001) and, therefore, they may need to be facilitated in expressing their spiritual needs at the end of life, like anyone else.

It is always worth remembering that challenging behavior in someone with intellectual disabilities may be a way of communicating pain. It is also worth paying attention to comorbidities that may be painful, especially if these are longstanding (e.g., contractures, sensory or motor impairments and postural problems). People who have experienced persistent and chronic pain throughout their lives may have been conditioned not to express their pain, or may express pain in unconventional ways.

There are some specific tools available. The Disability Assessment and Distress Tool (DisDAT) (Regnard et al. 2007) is particularly useful for people with intellectual disabilities, including those with severe and profound disabilities. The DisDAT is intended to help identify distress cues people who have severely limited communication. It is designed to describe a person's usual content cues, thus enabling distress cues to be identified more clearly. For example, a hospice nurse may not realize that someone who sits calmly and quietly in her chair is actually severely distressed; but her carers will know that this person usually rocks backward and forward, and therefore carers will realize her stillness is a cause for concern. Documenting this will help all professionals. Identification of the distress is only the beginning of the assessment; unless the person is able to tell you clearly what is causing the distress, this still needs to be determined and can often be no more than an "educated guess" (See also Sect. 3.5).

3.4.1 Tips for Effective Assessment

The following may be useful in assessing the needs of someone with intellectual disabilities (see also Tuffrey-Wijne and McEnhill 2008).

- Get to know the person. The earlier palliative care professionals can be involved, the better, as this provides opportunities to build a relationship of trust, which will be crucial in future needs assessment.
- Involve families and others who know the person well. They can be effective "interpreters" of the person's verbal and nonverbal communication, and should be part of your team.
- Take plenty of time, and accept that this is an ongoing process, to be refined over the coming days, weeks, or even months.
- Always speak to the person with intellectual disabilities first (even if they don't use verbal communication), and only then refer to the person's carer. Even if most of the assessment will need to be through the carer (e.g., if the person's communication is only understood by the carer, or if the person is too anxious to speak to those they don't know well), refer to the person frequently. This will build trust and confidence, not only for the person with intellectual disabilities but also for their carer.
- Use simple and straightforward questions. Never use more than one concept per sentence. Don't ask: "How are you, do you have pain today?", but rather, "How are you?" (wait for response), "Do you have pain today?" (wait for response).
- Allow the person plenty of time to respond. Do not fill necessary silence with another question.
- Many people with intellectual disabilities are eager to please and will tell you what they think you want to hear.
  - It is not unusual for people with intellectual disabilities to answer "yes" regardless of the question. Closed questions ("Do you have pain today?") may be important in assessments, but should be used with care. It is worth asking the opposite question as well, to see if you get a similar response ("Has the pain gone away?")
  - When presented with different options, some people with intellectual disabilities tend to repeat the final option ("Is the pain there all the time or only sometimes?" "Sometimes," so try repeating the question with the options the other way round, to see if you get the same response.
- Abstract concepts are much more difficult to understand than concrete ones. Concepts of time can be particularly difficult. Therefore, try to be as specific and concrete as you can. Instead of "How long have you had the pain," you could ask, "Did you have the pain when you went to church?"
- Do not assume that the person understands the connection between the symptoms and the illness.

3.5 Symptom Management

Pain and symptom management can be particularly complex in people with intellectual disabilities, many of whom have a range of chronic medical conditions and comorbidities; multi-pharmacy is not uncommon (Symons et al. 2008).

Pain is often not recognized, validated, or treated in people with intellectual disabilities. The CIPOLD study (Heslop et al. 2013) found that they receive less opioid analgesia in their final illness than the general population. The belief still exists that people with intellectual disabilities feel less pain than the general population. Pain assessment is complicated by the fact that self-reporting of pain can be difficult, and conventional pain assessment tools may not work well for this population.

It is important to try and determine the most likely cause of someone's distress. This is an imprecise art, but it is unacceptable to leave pain and other symptoms untreated because of uncertainty. Professionals need to use their clinical expertise and judgement to make an "educated guess" about the most likely cause. For example, is the symptom or the distress cue caused by the disease itself? The treatment of the disease? Debility or comorbidities? The impact of the symptom or illness on the person's life?

Treatment should be instigated accordingly and the result should be monitored, to see if the distress signs diminish over time. If they don't, then the situation needs to be reconsidered and another possible cause may be treated or managed. Consider both pharmacological and nonpharmacological treatments, including complementary therapies, emotional and spiritual support, the use of life stories and reminiscence therapy, relaxation exercises, etc.

The DisDAT assessment tool (See Sect. 3.4) includes a useful clinical decision checklist to help decide the cause of the distress. This is not an exhaustive list, and there is a strong emphasis on physical causes of distress; but it is important to exclude underlying physical causes, especially in this group where there is a risk of "diagnostic overshadowing."

IS THE NEW SIGN OR BEHAVIOR. . ..

- **Repeated rapidly?**

Consider pleuritic pain (in time with breathing); colic (comes and goes every few minutes); repetitive movement due to boredom or fear.

- **Associated with breathing?**

Consider: infection, COPD, pleural effusion, tumor.

- **Worsened or precipitated by movement?**

Consider: movement-related pains.

- **Related to eating?**

Consider: food refusal through illness, fear or depression; food refusal because of swallowing problems; upper GI problems (oral hygiene, peptic ulcer, dyspepsia) or abdominal problems.

- **Related to a specific situation?**

Consider: frightening or painful situations.

- **Associated with vomiting?**

Consider: causes of nausea and vomiting.

- **Associated with elimination (urine or fecal)?**

Consider: urinary problems (infection, retention); GI problems (diarrhea, constipation).

- **Present in a normally comfortable position or situation?**

Consider: anxiety, depression, pains at rest (e.g., colic, neuralgia), infection, nausea.

Taken from **DisDAT** © 2006 Northumberland Tyne & Wear NHS Trust and St. Oswald's Hospice.

3.6 End of Life Decision-Making

People with intellectual disabilities have a right to be facilitated in making choices about care and treatment, where possible. People with intellectual disabilities are particularly vulnerable and can be excluded from conversations that they may be able to have which could help plan the palliative and end-of-life care that they wish to receive. There is evidence that medical decision making is sometimes based on misguided assumptions about the quality of life of people with intellectual disabilities, their ability to comply and cope with treatments, or their ability to consent to treatment and be involved in the decision making process. This can lead to people with intellectual disabilities not receiving potentially lifesaving treatment (Mencap 2007; Michael 2008; Wagemans et al. 2010). There should be no assumptions about their capacity to make decisions due to the label "intellectual disability" (Johnson 2010).

Professionals should be aware of the fact that capacity may be an issue and needs to be assessed. They should also be aware of, and adhere to, national and local laws and regulations around capacity, consent, and advance decision making.

3.6.1 Assessing Capacity

With the right support, many people with intellectual disabilities are able to make at least some decisions. A person's capacity needs to be assessed for each situation. Capacity is “decision specific,” and some decisions are easier than others. A decision to start on opioid analgesia for pain may be easier than a decision to start (or continue with) chemotherapy for an invisible cancer. In the UK, a person is deemed to lack capacity if he or she is unable to do at least one of the following:

(a) Understand the information relevant to the decision
(b) Retain the information (for long enough to be able to make the decision)
(c) Balance the information (in other words, use the information to weigh up the options)
(d) Communicate the decision

It is important that people are given relevant information in a format that they can understand. Professionals must consider, therefore, what information is needed to enable informed decision making. It may be necessary to restrict information to the most essential. It is also important to remember that people have a right to make a decision that others may perceive as “unwise.” In order to assess whether the person has been able to use the information to weigh up the options, it can be useful to ask them how they have come to their decision. This could show that someone does indeed have capacity to make the decision, but it could also demonstrate that someone who makes a seemingly clear decision has not, in fact, understood the full implications of the decision. It is not unusual for someone to be clear that he doesn’t want surgery, and for this choice to be respected; but it may be that he has not understood that surgery could be life-saving and not having the surgery will eventually lead to his death – and therefore, he either has not been adequately informed, or he did not have the capacity to make this decision due to an inability to weigh up the information.

If someone lacks capacity, then someone else needs to make the decision for them. Who the surrogate decision maker is will depend on national laws, but an important general principle is that decisions are made *in the person’s best interest*. All relevant circumstances, as wells as the person’s wishes, feelings and values, must be taken into consideration. Even if it is decided that complying with their wishes is not in their best interest, people’s wishes clearly matter. The important question to ask is: “If this person had capacity, and could understand all the relevant issues, what do we think he or she would choose?”

3.7 Involving those Who Matter: Families, Friends, and Carers

Involving families, friends, and carers is particularly important for people with intellectual disabilities. Families and carers are often effective advocates and can play an important role in reassuring the person, providing communication support, contributing expert knowledge, and participating in decision-making. Studies that have included the voices of people with intellectual disabilities themselves, ascertaining their views on support at the end of life, have shown how important it is for them to have familiar people around (Tuffrey-Wijne et al. 2007a; McLaughlin et al. 2015). Furthermore, it has been shown that a lack of effective carer involvement leads to poorer outcomes for people with intellectual disabilities (Heslop et al. 2013; Tuffrey-Wijne et al. 2016b).

The important relationships of people with intellectual disabilities (“significant others”) should therefore be identified, with the help of the people themselves if at all possible. This could include family, partners, friends, informal (unpaid) carers, paid support staff, and professionals. The profile of this social network is likely to be different from that of the general population. Those in the general population often rely on the support of partners and children when they develop a serious illness, but for people with intellectual disabilities, family bonds tend to consist mostly of siblings and elderly parents (Tuffrey-Wijne 2010). It is often much more difficult for people with intellectual disabilities to create new bonds, including new family bonds, as they get older. It is also worth noting that many people with intellectual disabilities consider their professional support staff as their friends.

Some people with intellectual disabilities have lived with their parents all their lives and have developed interdependent relationships. Others may have been separated from their families at an early age and spent a lifetime in institutional care. Sometimes, relatives who have had little contact during the person's lifetime would like to be more involved at the end of life, which can at times cause tensions with those who have supported the person on a daily basis. It is important to consider the wishes and perspectives of the people with intellectual disabilities themselves. Family bonds may be crucially important to them, even if there has been a lack of contact (Hubert and Hollins 2006).

The risk that people with intellectual disabilities lose contact with the people that are important to them is heightened during a (final) illness, when they may not be able to organize visits or phone calls independently. Those who need to move into a new care setting are particularly vulnerable to losing contact with friends and familiar care staff.

3.8 Collaboration

Collaboration between services is key to successful provision of palliative care for people with intellectual disabilities. The importance of collaboration has been consistently highlighted in the literature as essential in ensuring that people with intellectual disabilities are well supported at the end of life (Read 2006; Cross et al. 2012; Friedman et al. 2012). Collaborative working should also include family carers and people with intellectual disabilities themselves; in fact, people with intellectual disabilities should be at the center of partnerships at all times.

Collaboration between palliative care services and intellectual disability services is particularly important. Developing a relationship with other services, built on mutual trust and respect for each other's knowledge base and skills, can enable a more robust assessment of the needs of people with intellectual disabilities. This can ensure better outcomes for this population, such as continuity of care and dying peacefully in their place of care with people familiar to them (McLaughlin et al. 2014). Building collaborative links may involve a concerted effort, in particular if professionals are not aware of each other's existence or range of services and expertise. It is important, therefore, that palliative care services and intellectual disability services actively reach out to each other. Often, a particularly complex situation with a person with intellectual disabilities in need of palliative care leads to services getting to know each other and work together. However, it is much better not to wait for a crisis, but to get to know other services within a catchment area in advance. The effectiveness of a proactive approach to collaborative working has been highlighted by the Palliative Care for People with Learning Disabilities Network (PCPLD Network 2016), which encourages an exchange of best practice. One example of a good practice initiative is a group of nurses and social workers from the community intellectual disability teams, who meet monthly with local palliative care specialists, to discuss service users who are known to be at the end of life or suspected to die within a year. The group follows nationally established frameworks and pathways and has developed these to suit the needs of people with intellectual disabilities. Each service user within their catchment area is now offered and of life planning, with a clearly recognizable folder for their health action planning and communication tool (PCPLD Network 2013).

3.9 Support for Families and Carers

3.9.1 Families

Families and carers are usually deeply affected when someone with intellectual disabilities reaches the end of life. This person has often been at the center of their family's or carer's life, sometimes for decades. Their death is a significant and difficult loss. For families of those who have needed active support throughout their lives, the death also signifies a loss of their role and identity as a care giver (Todd 2007; Young et al. 2014). All family carers need considerable and sensitive support, a recognition of their expertise in relation to the cared-for person, and a regular assessment of their needs (Payne and Morbey 2013). For carers of people with intellectual disabilities, whose situation is so much more complex, this is especially important.

The grief of families and carers is sometimes “disenfranchised” (where the relationship is not recognized, the loss is not recognized or the mourner is not recognized) (Doka 2002). Carers (and especially parents) of people with intellectual disabilities can experience their deaths as a painful physical loss of part of themselves. However, families may be given the message that the death of their relative is “for the best” or even a blessing (Young et al. 2014).

3.9.2 Paid Support Staff

The extent to which professional care givers are affected by the death of a person with intellectual disabilities is often under-estimated. Staff can form strong attachments with the people they support, and sometimes see themselves as surrogate family members, building relationships that last many years (Tuffrey-Wijne 2010). The death of a client of resident with intellectual disabilities can have a complex physical and emotional dimension for staff that is seldom recognized (Todd 2013). Many such staff have little experience of death and dying, and are likely to find the situation difficult on both a professional and a personal level. It is important that the grief of all those who loved and supported the person with intellectual disabilities is recognized and validated, including not only family carers but also paid support staff. Staff working with people with intellectual disabilities who are dying will benefit from training on self-care.

3.9.3 People with Intellectual Disabilities

The caring role of people with intellectual disabilities can go unrecognized. Sometimes, the carers of people with intellectual disabilities who need palliative care have intellectual disabilities themselves: they may be partners, friends, housemates, or adult children, for example. They will need a significant amount of support to cope with changing needs and impending losses.

It is also worth noting that people with intellectual disabilities who live at home with elderly parents often become carers within highly interdependent relationships, but they are often invisible to services because of a lack of recognition of mutual caring (Department of Health 2009). In a study of people with intellectual disabilities who were affected by a relative with cancer (usually a parent or partner), most had taken on a caring role (Tuffrey-Wijne et al. 2012). Palliative care services should be alert, therefore, to the possibility that adults with intellectual disabilities who live with a patient (including patients without intellectual disabilities) may need support as carers.

3.10 Preparing for Death

If people with intellectual disabilities are protected from knowledge about death, including their own impending death (See Sect. 3.2), it will be very difficult for them to prepare themselves for the future or be involved in care planning, if they so wish. Giving people opportunities to participate in decision making around their care and treatment, or discuss funeral wishes and make a will, it is necessary to have a culture of openness and inclusion. Conversations about death should happen throughout the life cycle, in order to build a foundation to help prepare people for their own final illness.

Discussions about the person’s preferences could take place as early as is appropriate, even before the need for palliative care arises. Once the need for palliative care has been identified, a care plan should be put into place, taking into consideration any anticipated future needs for treatment and care.

3.10.1 Advance Care Planning

Within the field of palliative care, there is growing emphasis on Advance Care Planning (ACP). This has been described as a process where a patient’s current condition and prognosis is reviewed, and likely dilemmas and options discussed with the patient and their family. It is a structured way of eliciting their wishes and thoughts for the future (goldstandardframework.org.uk). Important elements of ACP include:

- *Deciding what you want* – what care elements are important now and in the future? What is the preferred place of care?
- *Deciding what you don't want* – this can include legally binding statements, such as Do Not Attempt Cardiopulmonary Resuscitation orders
- *Who will speak for you* – e.g., appointing a proxy spokesperson or legal representative (the terminology and powers of others to decide for you will vary in different countries)

With the person's permission, all those involved in their care should be made aware of the patient's wishes and advance decisions.

There are several easy-read advance care planning documents available online. Within some of these documents, there tends to be a focus on funeral planning rather than care planning. These resources have not yet been properly evaluated. In order to use advance care planning documents, professionals and carers must have an understanding of the process of advance care planning, including an appreciation of the fact that it is indeed a *process* – it is not a one-off event, but involves discussions over time and should be revisited regularly. How advance care planning with people with intellectual disabilities is best instigated and supported needs further investigation.

3.11 Bereavement Support

The importance of supporting families, carers, and staff through a person's final illness and after their death has already been highlighted (See Sect. 3.9). This section deals specifically with the need for people with intellectual disabilities to receive bereavement support. People with intellectual disabilities often experience more losses than the general population. Most children within the general population will not have experienced the death of a friend; but it is not unusual for children who attend special schools to experience the deaths of their peers, not just once but repeatedly.

The impact of losing a significant person is always enormous, but can be particularly devastating for people with intellectual disabilities who may have been dependent on the deceased person in many ways. If the death of a relative precipitates a move into a care setting, there are multiple losses associated with the bereavement, including the loss of home and all that was familiar.

There is growing recognition of the bereavement support needs of people with intellectual disabilities. This is a relatively recent development; until the 1990s, it was assumed that people with intellectual disabilities did not experience grief (Oswin 1991). Even today, people with intellectual disabilities do not always get recognition for their loss and are not always given opportunities to talk about it or express their feelings (Tuffrey-Wijne et al. 2012). The grief responses of people with intellectual disabilities can be delayed, prolonged, or expressed in atypical ways, so it may not be recognized as a grief reaction (Hollins and Esterhuyzen 1997).

3.11.1 Risk of Complicated Grief

Not all people with intellectual disabilities need specific or specialist bereavement support, but the possibility of difficult grief processes must be borne in mind. A number of risk factors make people with intellectual disabilities more vulnerable than the general population to complicated grief reactions, including (McHale and Carey 2002; Blackman 2008; Blackman 2003):

- Social isolation
- High dependency on a small group (or even a single) significant other(s), with limited opportunities for developing new roles and relationships
- Exclusion from death rituals (such as attending funerals or visiting the grave)
- Difficulties with attachment in early life
- Low self-esteem
- Limited power or control over one's situation
- Associated, often hidden and multiple losses that accompany the death of a parent or close relative (e.g., loss of home)

In assessing the need for bereavement support, is can be useful to focus on how the loss has affected the following three areas of someone's life (Blackman 2008):

- The person's ability to communicate with others (e.g., a parent who dies may have been the only person who could interpret their adult son or daughter's communication)
- The impact on the person's familial network
- The person ability to recognize and express their emotions

3.11.2 Providing Bereavement Support

The following hints and tips can be helpful in providing support for people with intellectual disabilities who have been bereaved or for whom bereavement is anticipated (Blackman 2003; Read 2005; Read 2007).

- Prepare the person for the loss if at all possible. People with intellectual disabilities are often protected from knowing that someone close of them is going to die (perhaps because those around them want to spare them distress), but if they are unprepared, the death will be experienced as an unexpected, sudden death. Sudden death is usually more difficult to cope with and increases the risk of complicated grief (Murray Parkes 1998).
- Ensure that the person participates in death rituals, such as funerals. It can also be very helpful to hold additional rituals, such as memorial events, planting a tree in someone's memory, etc. People with intellectual disabilities are often excluded from active involvement in rituals, including the planning of funerals and memorial events. They are also less likely to have opportunities to share their grief with others, for example, through seeing others cry about the loss or through receiving cards of condolence.
- Provide information about bereavement in a format that the person can understand. This may need to be repeated often. Be open and honest. Often, the truth is easier to cope with than uncertainties.
- Tangible ways of remembering are often helpful. Consider the use of life story books, memory books, or memory boxes. These can also help the person talk about the loss with others; for example, taking photographs at a funeral and showing these to others afterward can help to process what has happened.
- Bereavement counsellors may also need to use a variety of approaches to help someone with an intellectual disability experiencing grief, such as art work, creating family trees, use of pictures, photographs, videos, poetry, and reminiscence work.

Supporting people with profound intellectual disabilities in grief can be particularly complex. They will need to be provided with supportive relationships and sensory experiences in order to increase their sense of safety, enhance a sense of security, and facilitate expression of their grief. The resource created by PAMIS is particularly useful for this group (Young et al. 2014).

For some people with complicated grief responses, specialist bereavement support is indicated. In one randomized controlled trial, bereavement counsellors who worked with the general population received training on intellectual disabilities and then worked with bereaved people in one-to-one sessions; staff working within intellectual disability services received training on providing bereavement support, and worked with bereaved people within their own settings. The study found that the generic bereavement counsellors were able to improve outcomes for people with intellectual disabilities, while many of the staff within intellectual disability services dropped out of the program (Dowling et al. 2006). It seems that engaging with issues of death, dying, and loss is very difficult for staff working with people with intellectual disabilities on a daily basis; a finding that has been confirmed in later studies (Ryan et al. 2011; Tuffrey-Wijne and Rose 2017).

There are very few specialist bereavement services available for people with intellectual disabilities, but it is worth looking for generic bereavement services willing to take on clients with intellectual disabilities. They may need to know how to use different approaches, such as art work, creating family trees, using pictures, photographs, videos, poetry, and reminiscence work.

3.12 Education and Training

3.12.1 Training for Staff Providing Generic Palliative Care Services

Staff working in palliative care services have consistently reported that they lack of confidence, knowledge, and skills in supporting people with intellectual disabilities. They find assessment and communication issues particularly difficult (Tuffrey-Wijne et al. 2008; McLaughlin et al. 2014). Palliative care professionals may see relatively few people with intellectual disabilities, so their knowledge and skills are not being developed. The following areas are not exhaustive, but are important training priorities:

- What are intellectual disabilities and how does it affect people's lives?
- How are people with intellectual disabilities supported within the local area? Where do they live, who provides them with daily support, what specialist intellectual disability services are available? How skilled or experienced are these services or carers in providing end-of-life support, and what help do they need?
- Communication needs; interpreting communication; alternative communication methods; breaking bad news
- Assessment of symptoms and other problems

3.12.2 Training for Staff in Intellectual Disability Services

Staff working in intellectual disability services may not have any experience of death and dying, and may be frightened by it (Todd 2005; Tuffrey-Wijne 2010). Many will be unfamiliar with the needs of people at the end of life. It is easy to assume that people with intellectual disabilities who live within staffed homes or institutions are well supported, but such assumptions may be erroneous. In the UK, for example, support staff for people with intellectual disabilities tend to have very little training; most have limited knowledge of looking after people with failing health. In addition, they may experience anticipatory grief reactions themselves (See Sect. 3.9), making the delivery of support at the end-of-life challenging on many levels – practical as well as emotional. Training and support may be best delivered by outside experts (such as community palliative care nurses or district nurses) on an as-needed basis, showing staff who to support specific individuals. Generally, the following areas are important in training staff in intellectual disability services:

- Thinking about death and dying in general; your own attitudes, issues, reactions, fears, etc. In order for staff to be able to provide good support for others, it is usually helpful for them to think about and articulate these issues.
- What support services are available locally for people who need palliative care? Who is in the multidisciplinary team? (This could include: primary care services including general practitioners and district nurses; specialist services, including hospices and community palliative care services)
- The process of dying: what to expect, how you can help, when to ask for support
- How to communicate about death and dying with people with intellectual disabilities
- Loss and bereavement, and how people with intellectual disabilities can be supported

Cross-fertilization of knowledge and skills between palliative care staff and intellectual disability staff is particularly effective and useful. This could be through formal mutual training sessions and through informal exchange of expertise around a particular individual with intellectual disabilities. It can also be also highly effective to include carers and people with intellectual disabilities themselves, as experts-by-experience. Hearing their stories and perspectives can have a powerful impact on staff.

Training for People with Intellectual Disabilities

People with intellectual disabilities themselves often lack essential and basic knowledge around illness, death, and dying, and will benefit from education in this area. It is possible, and important, to create opportunities for them to learn about death and dying throughout their lives. Families and carers may need help and support in encouraging such discussions. Open discussions at home are particularly important; for example, the death of a celebrity or a soap opera character may prompt conversations about illness, dying, and funerals. There could also be planned sessions at day centers or special educational facilities.

3.13 Developing and Managing Services

In order to ensure that people with intellectual disabilities are adequately supported at the end of life, in the place that is most appropriate to their needs (and that is, ideally, their preferred place of care), it is essential to know where and how they die. Services need to be able to anticipate the likely need. Many people with intellectual disabilities may wish to choose to remain in their existing home environment. If this is a residential care setting, provision will have to be made to make that possible. This is likely to need advance planning, as it will require adequate resources in terms of staff and physical environment. Such services need to anticipate, therefore, the likelihood that their clients reach the end of life with an associated need for increased support. This will reduce the need for hospital admissions or a last minute search for a nursing home able to cater for the person's changed needs.

Palliative care services will need to consider whether they are sufficiently prepared to have patients with intellectual disabilities on their caseload. This is likely to require extra resources. They may need extra staff time, additional resources to help them communicate and additional time to manage their often complex needs. There may be a wide range of carers and professionals involved. Policy makers should commit adequate resources to this.

The following are particularly important in ensuring high-quality care for people with intellectual disabilities at end of life:

- Develop and encourage continuity of care across settings
- Ensure that good basic palliative care skills and knowledge are held within staff teams working in intellectual disability services
- Facilitate collaborative partnerships among palliative care programs, community hospices, and a wide range of other healthcare delivery settings.

4 Conclusion and Summary

Ensuring that people with intellectual disabilities are well supported at the end of life is highly challenging and needs focused attention. One key challenge is the "invisibility" of this population within health and social care services. The vast majority of people with intellectual disabilities are on the mild end of the spectrum, and this group may be particularly difficult to identify. Their needs are largely hidden, but their problems may be significant, and require skilled support. Even if the problems are identified and known, many staff, services, and systems are unprepared for meeting the needs of this population. Across Europe, good practice often depends on the dedication of individual practitioners, rather than effective services and systems (Tuffrey-Wijne and McLaughlin 2015).

4.1 Recommendations

The EAPC White Paper has set out key areas for practice, which have been discussed in this chapter. It also makes the following recommendations.

- **Palliative care services should actively reach out** to *find* the population of people with intellectual disabilities within their catchment areas.
- **Ongoing exchange** of experiences, expertise, and best practice should be encouraged on a range of levels:
  - Locally, between palliative care and intellectual disability services
  - Nationally, between individuals and organizations involved in supporting people with intellectual disabilities at the end of life
  - Internationally within Europe

- **International exchange of expertise,** for example through:
    - An ongoing, regularly updated online multilanguage resource, signposting relevant literature, resources, contacts, etc.
    - A dedicated person or team who can act as a "point of contact" for palliative care provision to people with intellectual disabilities in Europe. Their role could include: collating relevant information and resources (see above online resource); facilitating contact between different services in different countries; organizing exchange visits; signposting training opportunities.

4.2 Future Research

The following areas have been identified as priorities for future research by an international group of academics and practitioners (Tuffrey-Wijne et al. 2016a):

- Investigating issues around end-of-life decision making
- Mapping the scale and scope of the issue (in order to be able to plan adequate care provision)
- Investigating the quality of palliative care for people with intellectual disabilities, including the challenges in achieving best practice
- Developing outcome measures and instruments for palliative care of people with intellectual disabilities.

4.3 Benefits for Everyone

Is it worthwhile spending time and resources on supporting people with intellectual disabilities at the end of life, even for services who may see relatively few such patients? Practitioners, service managers, policy makers, and funders may well raise this question. Clearly, there is an argument for ensuring that the most vulnerable people in society are provided with the same quality of palliative care as the rest of the population. But the benefits of focusing on the needs of people with intellectual disabilities, and ensuring that staff and services are ready to meet those needs, go well beyond this. The skills needed to care for people with intellectual disabilities are transferrable and will benefit all patients. Services that can care for people with very severe communication problems, complex social situations, multiple comorbidities, unconventional ways of expressing symptoms, and perhaps high levels of anxiety, can probably care for all patients, whatever their complexities. Such services need flexibility. Their service delivery needs to be highly adaptable to individual need.

The quality of a palliative care services could be measured by the way in which they are able to support people with intellectual disabilities. It is worth the effort to "get it right."

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