



The challenges of modern Slovenian paediatric palliative care

Izzivi sodobne slovenske pediatrične paliativne oskrbe

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Abstract

Paediatric palliative care is specialized palliative care that differs from palliative care for adults. Equating the principles of modern palliative care of a child with protocols of restrictions or discontinuation of treatment is a reflection of ignorance and is completely wrong. A multidisciplinary team for palliative care of children, consisting of the child's treating subspecialists, nurses, psychologist and, if necessary, other health professionals, including those with additional knowledge of palliative medicine and nursing, develops a plan for further treatment and constantly adjusts care procedures to growth and child development, disease progression, deterioration of the patient's condition and the emergence of new problems. The content of the palliative care plan depends on the moment of the patient's and his family's involvement, the type of disease and the identified and expressed needs. In pediatrics, the need to introduce a palliative approach by health professionals is often recognized too late or not at all. By spreading knowledge and awareness of the benefits of modern paediatric palliative care for patients and their relatives, we try to make the palliative approach in sick children with advanced incurable diseases a part of everyday clinical practice. It is not only one of the inalienable rights of the most seriously ill children, but it is also a matter of working by following a code of ethics.

Izvleček

Paliativna oskrba otrok je specializirana paliativna oskrba z določenimi posebnostmi v primerjavi s paliativno oskrbo odraslih. Enačenje načel sodobne paliativne oskrbe otroka s protokoli opustitve in odtegnitve zdravljenja je odraz nepoučenosti in je povsem napačno. Multidisciplinarni tim za paliativno oskrbo otrok na terciarni ravni sestavljajo otrokovi lečeči subspecialisti, medicinske sestre, psiholog in po potrebi drugi zdravstveni sodelavci, med njimi tudi tisti z dodatnim znanjem iz paliativne medicine in zdravstvene nege. Tim oblikuje načrt nadaljnega zdravljenja ter postopke oskrbe nenehno prilagaja rasti in razvoju otroka, napredovanju bolezni, slabšanju bolnikovega stanja in pojavljanju novih težav. Vsebina načrta paliativne oskrbe je odvisna od trenutka vključitve bolnika in njegove družine, vrste bolezni ter prepoznanih in izraženih potreb. V pediatriji se potreba po uvedbi paliativnega pristopa pri zdravstvenih delavcih pogosto prepozna prepozno ali pa se sploh ne. S širjenjem znanja in zavedanja koristi sodobne pediatrične paliativne oskrbe za bolnike in njihove svojce poskušamo doseči, da postane paliativni pristop pri bolnih otrocih z neozdravljivo napredujočo boleznijo del vsakodnevne

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klinične prakse. Ne gre le za eno neodtujljivih pravic najtežje bolnih otrok, gre tudi za delo zdravstvenih delavcev v skladu z etičnim kodeksom, da zdravljenje, ki je bolniku bolj v breme kot v korist, etično ni utemeljeno.

1 Introduction

Palliative care includes active holistic treatment of patients with an incurable disease and support for their close relatives. The term “palliative” comes from the Latin word “palliare”, which means “covered with a cloak, to shield”. In a figurative sense, therefore, the patient is “provided with relief”. According to the World Health Organisation (WHO), palliative care is an approach that improves the quality of life of patients and their families while facing problems related to incurable disease. It includes prevention and alleviation of suffering through early detection, professional judgement and the treatment of pain and other problems, both physical and social, and mental and spiritual (1,2). Palliative medicine and palliative care are integral parts of the palliative care service. Supportive care means relieving symptoms when treating a patient with any disease, while the term palliative care is used exclusively for patients with an incurable disease. Palliative care is a fundamental right of every terminally ill person, as stated in the Declaration of Helsinki, the Code of Medical Ethics, the Patients’ Rights Act, and other documents. Numerous untruths are spreading among the modern lay and professional public, which are the consequences of ignorance, and limit the exercising of the right of the terminally ill patient to palliative care (Table 1).

2 Historical overview of the development of paediatric palliative care

The development of paediatric palliative care began in the United States with the work of Professor Ida Martinson of the University of Minnesota School of Nursing, who, after caring for a dying patient at home, began providing care for dying children. Together with other nurses, she cared for eight dying children in the home environment. The first children’s hospice was founded in Suffolk, Virginia, in 1977 by Allan and Joan Hogge, the parents of a boy named Marcus, who was dying of a rare, progressive neurological disease. Dr. L. Joseph Butterfield founded the first neonatal hospice program in 1979 in Denver.

After a painful personal experience, Ann Armstrong-Dailey founded the Children’s Hospice International (CHI) in 1983 with the vision that palliative care

should be an integral part of paediatric treatment and not a separate activity in special institutions. In San Francisco, they started a series of annual international conferences to bring together everyone involved in children’s hospice and palliative care. The first palliative care unit in the United States was opened in 1984 at St. Mary’s Hospital in New York.

UK- and world-recognised professor Dr. Myra Bluebond-Langner published the first, but very influential research into *The Private Worlds of Dying Children* back in 1978. Until 2010, when she took on the position of the chair of The True Colours Trust, established at the Louis Dundas Centre for Children’s Palliative Care under the auspices of Great Ormond Street Hospital (GOSH) and the UCL (Institute of Child Health), she had published a number of studies and important contributions in the treatment of pain and other symptoms and neonatal palliative care and paediatric palliative care education.

The development of paediatric palliative care in the UK, however, began with the support for children and families facing childhood cancer. The first children’s hospice, Helen House, was set up in Oxford in 1982 by Frances Dominica Ritchie, a nun and nurse, when she met a girl with a brain tumour who had suffered a severe brain injury during surgery. In 1986, Dr. Ann Goldman recognised the need for a hospital team of palliative care professionals at London’s Great Ormond Street Hospital for Children. In addition to numerous professional publications, she is also the co-editor of the *Oxford Textbook of Palliative Care for Children* (10,11).

Organised education on paediatric palliative care was introduced by Dr. Charles A. Corr, who founded the first American-accredited course on children and dying in 1984. He is the co-editor of the CHiPPS E-Journal (Children’s Project on Palliative/Hospice Services) web portal, released in collaboration with the American National Hospice and Palliative Care Organisation (11).

Prof. Susan Fowler-Kerry and Dr. Pat McGrath developed the first guidelines on paediatric palliative care in 1998, namely, guidelines for helping children with cancer. Early innovative guidelines on basic symptom control in paediatric palliative care were developed by Dr. Satbir Jassal from Rainbows Children’s Hospice in the UK and first published in 1999 (12). In 2000, the first

Table 1: Misconceptions and truths about paediatric palliative care. Adapted from the International Children's Palliative Care Network, ICPCN (3).

Misconception	Truth
Palliative care for adults and children is the same, only slightly adjusted for children.	Although there are similarities and the basic principles of palliative care for adults and children do not differ, palliative care for children has certain peculiarities.
Paediatric palliative care is only for children with cancer.	Every child with an advanced incurable disease has the right to palliative care.
Palliative care is necessary and appropriate only at the end of life.	Palliative care begins depending on the needs of the child and the family: at the time of diagnosing the disease as incurable and progressive or at any time during the progression of the disease. It can last for several years, simultaneously with the treatment of the underlying disease, and continue for the rest of the child's life.
Paediatric palliative care can only be provided in well-equipped environments.	Palliative care can be provided in different settings and at all levels of healthcare (primary, secondary and tertiary). The levels are interconnected in the care of the child and work together.
Palliative care is a plan to discontinue treatment.	Palliative care is a continuation of treatment that benefits the patient. In the absence of unnecessary laborious investigations, referrals and procedures, a number of additional measures are introduced to ensure the best possible quality of life until death.
Working in palliative care is sad and depressing.	In addition to the grief that healthcare professionals face in palliative care, there is also joy and satisfaction. It alleviates the suffering of the child and the family and improves the last period of life.
The introduction of palliative care means giving up hope and stopping treatment.	Not true. Treatment is not stopped, but adjusted. Despite the fact that the child will not recover, the child and the family are supported in finding new and different goals that can be achieved through additional palliative care measures.
Children must always be protected from the awareness that they are dying.	Children over the age of three intuitively know they are ill and dying, so they want to know the truth. If the truth is concealed, they are in distress. To reduce distress, it is beneficial to offer them the opportunity to express their thoughts and ask questions that need to be answered with clear but personalised answers.
Children do not want to talk about their death.	Children do not talk about their death in front of their parents because they want to protect them from grief. That doesn't mean they don't want to talk about it.

American guidelines on paediatric palliative care were published (4). In 2006, a group of health professionals from Europe, Canada, Lebanon and the United States met in Trento, Italy, to discuss the state of paediatric palliative care in Europe at the time. Based on the findings and conclusions of the International Meeting for Palliative Care in Children, Trento (IMPCCCT), the first European guidelines for paediatric palliative care were developed and published in German in 2008 (13).

In our country, the principles of paediatric palliative care in the Division of Paediatrics in Ljubljana were introduced in patients with cancer more than 20 years ago by paediatric haematologists and oncologists Prim. Jožica Anžič and Prim. Majda Dolničar. Prim. Dolničar and her colleagues expanded their knowledge of palliative care to other subspecialist fields, such as neonatology, neurology and pulmonology. It began with palliative care being provided to a dying child with advanced

cancer at home in the form of visits by a mobile palliative team consisting of a paediatrician and a nurse from the Department of Oncology of the Paediatric Clinic and, if necessary, a psychologist of the department (14). At the end of 2018, a multidisciplinary team was formed at the Paediatric Clinic in Ljubljana, which started providing tertiary level paediatric palliative care and began educating health professionals in paediatrics at all levels of healthcare.

3 Characteristics of paediatric palliative care

Palliative care:

1. Provides the patient with relief from physical, mental, spiritual and social symptoms and problems caused by incurable disease. At the same time, it neither accelerates nor delays death, but offers a system

of support with procedures and measures that help the patient to live as actively as possible until death. It is essential that it takes into account the patient's wishes. It improves the quality of life, has a beneficial effect on the course of the disease and can even prolong life.

2. Offers a support system that also helps the patient's family cope with a relative's illness and with accompanying grief.
3. Uses a team approach to address the needs of patients and their families.
4. Makes sense in the early stages of the disease while implementing other types of treatment intended to prolong life.

Regardless of when and where the patient is cared for, the treatment should be continuous. The palliative approach in the treatment of the patient should begin early after the diagnosis of an incurable disease and should initially intertwine with treatment aimed at slowing the progression of the underlying disease. As the underlying disease progresses, palliative care gradually prevails and treatment continues in the process of dying, death, and mourning. Providing care for a dying patient is only part of a holistic palliative care regime (2).

Specificities of paediatric palliative care compared to palliative care for adults:

1. These patients are fewer in number, but they are more complex than most adult patients.
2. The spectrum of diseases that cause the death of a child before adulthood is very wide, many of which are rare diseases.
3. Some genetic diseases can also affect more than one child in the family.
4. As a child develops during the course of the disease, physically, emotionally and cognitively, his health and social needs also change, as does his understanding of illness and death.
5. Child care takes place in different environments, including school, for example. Despite the incurable disease, the provision of education for an ill child is also his/her basic right and continues in the form adapted to his/her abilities.

Similar to the process of palliative care for adult patients, palliative care for children is family-oriented and includes siblings and grandparents, possibly friends. It takes place in accordance with family values, traditions, and culture. The period of illness until the death of a child can be long and can last several years. Care is appropriate at any stage of the disease and may be accompanied

by treatment of the underlying disease if it is not burdensome and is beneficial to the child. Although formal training on palliative care for children is not yet widely available, the staff providing palliative care must be specially trained and experienced in child care (3,4,5).

The most common medical conditions in paediatrics that require palliative care are divided into four groups (adapted from 6,7):

1. Diseases for which targeted treatment is possible but may fail:
 - advanced or progressive cancer or cancer with poor prognosis;
 - complex and severe congenital or acquired heart disease.
2. Chronically ongoing potentially progressive disease conditions that require intensive and long-term treatment to maintain quality of life:
 - severe epidermolysis bullosa;
 - severe forms of immune deficiency;
 - renal failure if dialysis and/or kidney transplant is not possible;
 - chronic severe respiratory failure;
 - spinal muscular atrophy type 1;
 - cystic fibrosis;
 - severe inoperable gastrointestinal malformations, severe gastrointestinal conditions (e.g. short bowel syndrome).
3. Progressive diseases for which targeted treatment is not possible:
 - progressive metabolic disorders;
 - certain chromosomal abnormalities (e.g. Trisomy 13 or Trisomy 18);
 - severe forms of osteogenesis imperfecta.
4. Non-progressive but irreversible conditions with a high probability of health complications:
 - severe cerebral palsy and other conditions with severe developmental disorders;
 - conditions with severe neurological consequences due to infectious diseases, extensive hypoxic brain damage, severe brain malformations.

Case report:

At the age of 12 hours, the third child of healthy parents, born on due date after a normal pregnancy, was transferred from the regional maternity hospital to the Division of Paediatrics due to hypotonia and pallor, where he was then dealt with by various subspecialists. The following was found: macrocytic non-haemolytic anaemia, lactic acidosis and low-grade proteinuria, small cysts in the renal cortex revealed by ultrasound, feeding problems. At 4 months of age, suspicion of mitochondrial disease

with predicted multiorgan dysfunction was confirmed in a provenly known but rare mutation. This was a case of an incurable progressive disease that the parents were made aware of.

By the age of five months, when the child overcame pneumococcal sepsis, he had already needed an erythrocyte transfusion three times. Proteinuria increased, hypoproteinaemia and oedema occurred. In addition to vitamin supplements, he needed immunoglobulin replacement. At the age of six months, he was given a nasogastric tube due to growth failure, through which he received a hypercaloric diet. There was a marked developmental delay and the head grew insufficiently. At the age of eight months, tonic spasms of the upper limbs began to appear as infantile spasms, and several antiepileptic drugs were introduced. As a side effect, hyperglycaemia developed, so insulin was introduced. In the following months, recurrent urinary retention, electrolyte disorders and an increase in nitrogen retention, myocardial hypertrophy, and hypertension occurred. A gastrostomy tube was inserted. The child needed a prokinetic; he often vomited. Chronic kidney disease progressed to their final shut-down at the age of one year.

By that time, he had been treated in hospital several times a month, occasionally for longer periods. The mother confided her concerns about dialysis to his doctor. The medical team of all subspecialists weighed the burdens and benefits and included the child's parents in the consideration. They concluded that with the multiorgan progression of the disease the need for dialysis would not be limited to the transitional period until kidney transplantation, and that it would not result in a miracle cure, and therefore does not benefit the child. A diuretic, electrolyte replacement, and frequent monitoring of laboratory results were prescribed instead. During the check-ups, the parents reported restlessness and pain.

A palliative care plan was developed together with the parents and the visiting healthcare team. A number of drugs that were not necessary and that caused vomiting were discontinued. Regular painkiller treatment was introduced. The parents wanted as few stressful hospital visits as possible, so the boy's condition was monitored in the home environment regularly by phone. Parents learned to catheterise the bladder and all the measures for faecal elimination. They were acquainted with the symptoms of dying, measures for alleviating respiratory distress, convulsions and pain, and the necessary procedures after death. The child died peacefully after about three weeks in the arms of his parents and in the presence of both siblings.

Later, during one of the supportive psychological interventions for siblings, the family visited the hospital ward

where the child was treated most often. The parents expressed their gratitude for the period of the last month, when all members of the family, closely connected with each other, spent time together with the youngest child. They described it as one of the most beautiful periods in the life of their family.

4 Treating a child with palliative needs in our country

4.1 Patient identification and approach

The beginning of palliative care, which takes place at all levels of a patient's medical treatment, can be initiated by a healthcare professional at any level. Basic care is provided by the child's personal paediatrician in cooperation with the visiting nursing service at the primary level. Secondary and tertiary care with a multidisciplinary approach can make an important contribution to the holistic treatment of complex patients with multiorgan dysfunction. The multidisciplinary team for paediatric palliative care at the tertiary level consists of the subspecialists, nurses, psychologists and, if necessary, other health professionals, treating the child, including those with additional knowledge of palliative medicine and healthcare.

Palliative care is effective if it is included into the treatment early enough. It is beneficial that physical, mental, spiritual and social problems are anticipated in advance in order to prevent them and thus even prolong the child's life (3,5,6). A positive answer to the question "Would the survival of a child for more than a year surprise you?" is a "call" for health professionals to introduce palliative care as soon as possible. End-of-life palliative care should be introduced at least six months before expected death (13). In practice, we manage to introduce palliative care in paediatrics as soon as the disease is diagnosed as incurable and progressive, mostly only in new-borns or in the event of a sudden onset of the disease, e.g. in severe injury in a previously healthy child. More often it is initiated when the physical problems are already severe or even only when the child's parents start questioning the benefits of the many measures for improving the condition.

The authors Bergstraesser et al. named the list of criteria that determine whether a child would benefit from palliative care the Paediatric Palliative Screening Scale (PaPaS Scale). It is useful for children older than one year (19). Patients in the neonatal period and up to one year of age are classified according to specially tailored criteria (20,21).

The paediatric palliative screening scale of the authors Bergstraesser et al. includes questions about the duration of expected survival, the expected outcome of treatment of the underlying disease, and the child's clinical condition. The patient falls into one of four possible groups (listed in the "Introduction" section). Impairment of normal life activities, frequency and duration of necessary hospitalisations, frequency of deteriorations without the condition returning to its previous level, severity and duration of symptoms and problems, and mental distress of the patient and family members are assessed. The priorities of the patient and the family are also evaluated. The intention is to always try to identify, determine and take into account all the side effects of the treatment of the underlying disease from the perspective of the patient and the family (19).

Many authors state that even in environments with an otherwise developed healthcare system, the need for palliative care is recognised too late (15-17). There are many reasons for this. It is a matter of not knowing the basics of palliative care with the prevailing opinion that it is unnecessary in one way or another, equating palliative care with "treatment failure" and thus avoiding its introduction. The term "palliative" is often interpreted as the fact that a child will soon die. There is often a general opinion that a child and family do not want palliative care. At the same time, the equating of the principles of modern paediatric palliative care with protocols of abandonment and withdrawal of treatment are often encountered, which is a reflection of ignorance and therefore completely wrong. The patient is still actively treated for the rest of his/her life, but only with those procedures that are exclusively for his/her benefit, but do not burden him/her unnecessarily in accordance with the professional recommendation by palliative medicine.

In adult patients with incurable disease, health professionals have fewer reservations about open palliative measures than in paediatrics. Younger adult and young senior patients are more likely to need a clear prognosis of the outcome of the disease due to a number of social factors (e.g. concerns for the survival of the children). In paediatrics, most healthcare professionals avoid unfavourable information for parents if at all possible. They want to instil hope in the parents for their child's recovery and therefore refuse to provide open information about the child's actual condition, but at the same time to include them in palliative care. With an emotional attachment to a chronically ill child who is being treated for a long time in a particular ward, the staff are reluctant to admit that treatment is no longer beneficial to the child. Nevertheless, the patient and relatives become

aware of the poor condition sooner or later and feel distressed. As they are not given the opportunity to ask questions about further deterioration, dying and death, they do not get any answers. The professional approach of aware and educated staff is therefore extremely important (15-17).

In clinical practice, it is a healthcare professional in the team treating the child, such as a doctor, nurse or psychologist, who brings up the discussion about changing the treatment in the case of terminally ill patients who are not treated in the intensive care unit. The plan of further treatment is rightly first drawn up by a professional team, then, through a conversation, presented to the child's parents in an appropriate way by the attending physician. It is best if a good relationship has already been established between the patient, relatives and the doctor, if the parents trust the medical team, are kept informed of the child's condition and if the doctor knows the family. The doctor listens to the parents and coordinates the ultimate goals of healthcare in accordance with the goals of the parents and the child. It is always necessary to reach a consensus with relatives, and not leave the decision to them, e.g. in intensive care in case of deterioration and resuscitation. If, at the first conversation about the change of treatment, the medical team and the parents are not coordinated, the treatment that burdens the child as little as possible is continued, and at the same time the team regularly communicates with the parents in a proper manner about the burden and benefits of all measures. The members of the medical team always represent a unified professional position and cooperate with the palliative care team, whose members are additionally trained in this field. In paediatrics, a psychologist is involved in the medical team from the very beginning, providing psychological support to the child and relatives and helping them communicate with the medical team.

In older children with a serious illness from birth, at least one parent often completely adjusts their life to the care of the child and finds it most difficult to accept the gradual deterioration of the condition. In these cases, it usually takes more time to coordinate the plan if it also contains, for example, limiting invasive treatment in the intensive care unit in case of respiratory failure, which would not benefit the child. The most suitable and effective approach is to establish an appropriate, respectful, non-judgmental and non-patronising attitude of the doctor towards the parents, the ability to listen to the parents and understand their distress, and to build trust and take enough time for numerous conversations. In practice, it is in these cases that we see great relief and

satisfaction from parents when they realise that we are not stopping the treatment of their child, but are offering him/her individually tailored care that makes him/her feel better and suffer less.

In patients with a life expectancy of less than a year and with a number of associated diseases that are still being treated, basic measures are immediately introduced to alleviate the symptoms, while emerging problems are resolved gradually. With an expected survival of less than 6 months and many and varied physical, mental, spiritual, and social symptoms and problems brought on by incurable disease, palliative care is essential. Therefore, not providing it in Slovenian regions is wrong, regardless of the differences in the healthcare conditions.

4.2 Creating a plan for further treatment

After identifying the patient and their needs, further treatment is planned.

The goal of palliative care is to improve the quality of life of patients and their families. It is about adapting the entire medical treatment to the child's benefits, both by abandoning useless burdensome medications, tests or referrals and by introducing various measures to improve the quality of life of the child and family while treating an otherwise incurable underlying disease that leads to earlier death than if the child was healthy. It is not just about humanity that some healthcare professionals display more and others less of; it is the ethical professional work of the physician who decides when the burden of further treatment outweighs its likely usefulness and effectiveness, and a change in treatment should be considered (18). The burdens and benefits of treatment for an individual patient are assessed in accordance with the family's life so far and their goals, which also define the quality of the child's life. This is not called a withdrawal or abandonment of treatment, but an adjustment or change of a patient's treatment.

Healthcare professionals from various fields are involved in the development of the plan. In practice, it is common for those with additional knowledge of palliative care to guide a medical team consisting of the child's treating subspecialists, nurses, psychologists, and other staff members from the primary, secondary, and tertiary levels. The patient will be treated in the home environment, in the regional hospital and the tertiary hospital, so all levels cooperate with each other, know the patient and work according to a single plan.

First, the patient's health, physical and mental development, and understanding of the disease are assessed. In paediatrics, these children mostly suffer from with

multiorgan dysfunction, a developmental disorder, possible visual and hearing impairment, and often an eating disorder. It is necessary to anticipate the development of the disease, possible expected and unexpected complications and deteriorations, and the most likely period of survival. The child's parents are the ones who know the child best and present to us their vision of the situation, fear of deterioration and distress regarding the future. It often happens that with an open conversation we open up areas that they could not or did not dare talk about until that moment. What they want most for their child and what they would like to protect him from becomes clearer. Their greatest distress is often caused by problems in treatment, which are easy to solve, e.g. condensed treatment, discontinuation of many drugs that do not benefit him/her, adjustment of the time of unpleasant interventions, and the like. All parents want their child not to suffer and not to be in pain. When their perception of their child's suffering is subjective and our judgement is different, we try to respectfully understand them, relieve them of the burden and focus their attention on areas that are undoubtedly the most important for the child.

Criteria for quality of life vary greatly with age: it is important for the new-born and the infant not to be hungry or in pain, to be warm in the safe shelter of his/her parents; for a small child, an additional opportunity to play and be in the company of peers is important, as well as, still, the presence of parents; adolescents need to socialise with peers and talk about the hardships they experience. We cannot cure a child, but we can achieve various goals that make the family happy and are important to them.

To facilitate the development of the plan, the areas are divided into prevention and resolution of physical problems (physical symptoms), addressing mental distress and social problems, while taking into account the spiritual orientation of the family.

4.2.1 Treatment of physical symptoms

Physical symptoms treated in palliative care include pain, nausea and vomiting, feeding disorders, digestive problems with constipation or diarrhoea, dyspnoea, epileptic seizures, and other symptoms associated with a specific medical condition (e.g. recurrent lower respiratory tract infections due to recurrent aspirations with saliva and mucus entering the respiratory tract). Knowing the clinical picture of the disease, we predict the most likely deteriorations. By introducing certain measures, these can be prevented and life can be prolonged (e.g.

the introduction of intensive chest physiotherapy, expectorants, saliva and mucus removal, medicines to reduce drooling). The already planned invasive interventions are considered from all aspects and the burden and benefits are assessed, e.g. scoliosis surgery. The types of treatments that do not improve the child's condition are considered, e.g. frequent hearing or vision tests. If frequent neurophysiotherapy sessions are stressful, they can be limited and adjusted. We never discontinue care as long as it keeps parents hopeful of improvement. With further cooperation, most parents see for themselves when certain procedures are not useful and they themselves suggest phasing out, e.g. frequent blood draws for laboratory tests.

As the disease progresses, the physical symptoms change, so the plan is adjusted. With a good knowledge of the patient's health condition and professional knowledge of doctors and the nursing staff, the final period of life is recognised. Some parents of young children want their child to die at home, others, fearing that they will not be able to provide optimal care for their child, want to stay in hospital. We discuss how the child's respiratory distress, restlessness, nausea, and pain will be alleviated. For older children and adolescents, we listen to the child's wishes and support the family to be able to provide adequate care at home. The mobile palliative team conducts a home visit to administer a subcutaneous infusion of painkiller mixture, change the urinary catheter or feeding tube, and is constantly available by phone.

In cases where a chronically ill child is dying in hospital, the space is adjusted and the staff are informed. The family is provided with the possibility of staying together or relatives visiting, such as grandparents; devices that emit alarm sounds are turned off, only people they know enter the room, etc. Palliative care team members also provide support to the staff members.

4.2.2 Psychological care

In practice, it is best if the emotional distress of an ill child and the family members is addressed by a psychologist who is involved in the team and trained in palliative care. However, certain concerns of a child can be well recognised by other team members and the paediatrician or nurse with many years of experience in paediatrics, especially if they take enough time to directly connect with the patient, parents and siblings. It is important for them to be aware that psychological care is not only to comfort the parents, and that its content is much broader in the treatment and conversation with the child at different stages of development, which, for

the most part, only a psychologist is skilled at. The psychologist involved in the team also advises less experienced colleagues on how to communicate with the child and parents. Together, they plan psychotherapeutic steps to alleviate physical symptoms. The distress of siblings is often not recognised and is ignored, although they are also significantly involved in caring for the ill child, they communicate with him/her every day and are significantly disadvantaged in their leisure activities due to lack of parental time as a result of their parents' devotion to the ill child. Therefore, we always include them in our care.

4.2.3 Social care

Severely ill children have the right to education, socialising with peers and the activities they would engage in if they were healthy. Home schooling, occasional visits to the school library and meetings with classmates or just a visit from a teacher at home are easy to arrange, as school staff are usually very open to this. Encouraging parents to take their children to various leisure activities despite the illness and obstacles (e.g. subcutaneous infusion pump for pain control, recommendations to avoid infection, etc.) can make an important contribution to improving the well-being of the child and parents. Parents of children with advanced disease mostly opt to continuously stay together and care for the child during treatment in hospital and at home. Therefore, the financial support of the family must also be taken care of in the event of the loss of the regular income of at least one of the parents, and they should be informed in time about their possibilities and social rights and helped during the execution of the necessary procedures.

4.2.4 Spiritual care

Perception of illness, challenges regarding health and, above all, death, vary from religion to religion. The palliative care team therefore plans activities in accordance with the religious beliefs of the patient and his/her relatives and regardless of the team members' own beliefs (6,13). Everyone needs spiritual support, even those who identify themselves as non-believers. We adapt to the family in the way we talk about death, which we have to inform them of, talk openly about, and, of course, understand their views, although they may be completely different from ours. A visit from a priest can be welcomed earlier and not just before death if that is what the family wishes.

The multidisciplinary team, which develops a plan

Table 2: Contents of a paediatric palliative care plan. Adapted from 6,13,24-31.

Levels of care	Measures
Treatment of physical symptoms	Identifying pain and other symptoms: - planning medications and drug doses, non-pharmacological treatment; - discontinuing medication that does not work.
	Anticipating deterioration and measures, medications: preparation of medication intended for emergencies at home.
	Providing measures to prevent deterioration: giving a subcutaneous infusion with an analgesic mixture.
Psychological support	Recognising the fears and worries of the child, parents, siblings: - having an open and honest conversation with the child and family members; - ensuring that the family in distress is not abandoned; agreement on how to communicate in case of distress.
	Identifying the child's way of expressing distress and coping with it: - adapting the plan to the child's perception of and coping with distress; - communicating with the child in an age and developmentally appropriate manner; - explaining death to the child in an appropriate way, but clearly and openly.
	Talking about difficult life experiences, disappointments, sorrows they've had so far: - adjusting the plan on the basis of information on past experiences; - providing the child with psychotherapeutic or psychiatric treatment.
	Dealing with mourning: - developing a plan for monitoring the family after the child's death; - assuring the parents that they will not feel abandoned after the death of the child.
Social care	Identifying the child's social needs: making arrangements with the school, enabling participation in leisure activities that the child can handle, socialising with friends.
	Identify the social needs of parents and siblings: - informing the family of the possibilities and rights related to sick leave, financial care; - enabling the perhaps overlooked needs of siblings for various activities, socialising; - connecting the family with volunteers if useful and appropriate.
Spiritual care	Getting to know the religion of the family: - talking to the child and family about death in a way that is appropriate for their religion; - allowing visits by the priest if this is what the family wishes; - respecting the differences between the attitudes of the family and the team members.
Important in developing a follow-up plan	Designating care decision-makers, main planners: - including the necessary staff in the team; - transferring agreements to the entire team; - providing necessary and comprehensible information on the course of the disease.
	Talking about the course of the disease: - talking about useful healthcare, agreement with parents; - identifying the impact of the disease on a child's quality of life; - predicting the outcome of the disease and the time of death if possible.
	Defining goals of care: - determining measures for treatment, to prevent deterioration, to ensure the comfort of the child; - acquainting team members with goals and measures, determining their roles.
	Identifying end-of-life needs: - developing a plan according to the anticipated state of health (including the omission of resuscitation, if necessary); - identifying significant changes in the child's condition just before death, informing parents in an appropriate way, taking action; - identifying parents' wishes regarding the place of death (at home, in hospital); - providing parents with appropriate contact with team members in the event of distress upon the child's death; - informing the appropriate family member with the procedures after the child's death..

for further treatment, adjusts care procedures on an ongoing basis, along with the child's growth and development, disease progression, deteriorating health, and the emergence of new problems. Care never ends with the patient's death, as it also includes supporting the family in the grieving process. The care plan depends on the time of patient and family inclusion, the type of illness, and the identified and expressed needs. The professionalism and experience of team members, good communication and mutual respect between colleagues at different levels of healthcare enable quality care with the sole aim of improving the quality of life of patients and families, and indirectly and in many ways relieve the health system. Various studies have shown that quality palliative care can reduce the need for emergency measures and the number of hospital transfers for managing symptoms, which can also be treated in the home environment (24–31). The care of individual problem areas is intertwined; with good pain management, the patient's fear of death will significantly subside and vice versa.

The plan (Table 2) must be familiar to all healthcare professionals who are to care for the patient at home: the selected personal physician, the emergency medical service at the basic level, healthcare professionals in the regional hospital, and healthcare professionals at the tertiary level. It is unacceptable to reject a patient's care at the secondary level, close to the patient's home, simply because the staff is not skilled in palliative care, and the patient could only be admitted to a tertiary care centre. It is useful if the plan contains the contacts of the team staff with additional palliative care knowledge who are available at any time to healthcare professionals who meet the child unexpectedly and are not familiar with the plan for further treatment.

When deciding to change the treatment of patients in the intensive care unit, all patients must be treated in the same way and without discrimination. Professional decisions in the medical team to limit certain treatments,

e.g. to not start dialysis or even withdraw certain treatments, e.g. drugs to support heart function, are very difficult (18). It is right that subspecialists from all fields are involved, who are familiar with the overall condition of the child and the opinion of the family. It is necessary to weigh the expected benefits and burdens of treatment, to consider the course of the disease, possible sudden and unexpected changes, and the consequences of the decision. Parents must be kept informed of the situation. Their opinion is taken into account, but the final decision to abandon various types of treatment is not left up to them.

The decision to change treatment, e.g. abandonment of dialysis, can always be reversed: if the patient's medical condition improves, the assessment of the benefits of dialysis treatment is repeated. Previously accepted treatment changes may be cancelled. The reasons for deciding to change treatment must always be documented (18).

5 Conclusion

Palliative care for children with incurable and progressive disease is provided by paediatricians, paediatric nurses, and other healthcare professionals, including psychologists, at all levels of healthcare. In Slovenia, a broader general identification of needs and additional knowledge in this field is needed. Regardless of the generally unfavourable health situation in our country and the lack of understanding of those who we believe are obliged to regulate working conditions, it is right that palliative care in our environment becomes an integral part of care for all seriously ill children with advanced incurable diseases.

Conflict of interest

None declared.

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