

No good choices, only hard decisions: the delicate nature of team family communication in pompe disease

Abstract

Objective: To describe the experience of a family going through the palliative care decision-making for a child with Pompe disease.

Method: Qualitative research, case study design, approved by Ethics Committee (CAAE: 5864381640005327), conducted between 2017/2018. The setting was the Pediatric Intensive Care Unit of a university hospital in Brazil. Participants were family members of children admitted to the Intensive Care Unit and referred for palliative care.

Results: The case study relates the decision-making on palliative care for a child with Pompe disease. The parents, surrounded by the palliative care available for the situation, experience the existential reality of the decision-making process to adopt palliative care for their child, together with the multi-professional team.

Conclusions: Understanding the concept of quality of life that each family develops when faced with the seriousness of their child's illness, when the possibilities of cure have been exhausted, is fundamental to the decision making process for adopting palliative care. It is essential to include the participation of the child's or adolescent's parents and other family members in the palliative care decision-making process, in order to give the family a voice, minimising their doubts and suffering, as well as offering shelter in a situation of extreme existential vulnerability.

Keywords: child, family, glycogen storage disease type II, integrative palliative care, pediatric nursing

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Abbreviations: PPC, pediatric palliative care; PICU, pediatric intensive care unit; FCC, family-centered care; AAP, American academy of paediatrics; SDM, shared decision making

Introduction

Palliative care is a philosophy of care that focuses on the quality of life and well-being of the person, regardless of their clinical condition, whether their pathological condition can be cured or not, or how long they have left to live. As a philosophy, it proposes the acceptance of death as a natural part of existence, the non-use of invasive technologies to prolong life at any costs, the proper control of unpleasant symptoms such as pain in the final process of a person's existence.^{1,2}

One of the key principles of this philosophy is to develop a holistic care plan that takes into account all dimensions of being - physical, psychological, social and spiritual - in order to care for the person and their family, and not just the pathological situation itself. In this way, it is indicated for the most varied situations of chronic and complex illnesses, including genetic pathologies for which there is no curative treatment.^{3,4}

The philosophy of palliative care places the person as a whole at the center of care, prioritizing his or her well-being and quality of life, proposing a natural acceptance of death and contraindicating the use of invasive technologies to sustain life. This philosophy is also a counterpoint to the cold practices of the biotechnological model, which focuses its attention on the disease and the healing process, which is often no longer possible, disregarding the integrity of the

human being. In essence, the adoption of comprehensive care is the keynote of this proposal, which aims above all to control and reduce the negative effects of the disease, promoting a state of life that is as active as possible as long as it lasts, with emphasis also on psychosocial and spiritual care.^{1,5}

In Pediatrics, palliative care was defined in 1998 as the care of children with chronic and/or life-threatening illness. This care should begin at the time of diagnosis, irrespective of the treatment of the underlying disease. Pediatric Palliative Care (PPC) involves the whole multi-professional team and aims to provide physical (symptom control), emotional, spiritual and social support for the child, including the needs of the family.⁶ In addition, PPC should address ethical issues, advance care planning, shared decision-making processes, end-of-life care and bereavement support for families. This care should be progressive, adapted to the needs of the illness and its treatment, and individualized for each child, taking into account the values and wishes of the family and the child.⁷

According to the World Health Organization, indications for PPC include children with acute life-threatening conditions from which recovery may or may not be possible; children with chronic life-threatening conditions that can be cured or controlled for a long period of time, but who may also die; children with progressive life-threatening conditions for which there is no curative treatment; children with severe neurological conditions that are not progressive but may lead to deterioration and death; newborns who are severely premature or have severe congenital anomalies; and family members of a fetus or child who dies unexpectedly.⁸

The importance of palliative care lies in the early identification, assessment and appropriate management of complaints, improving quality of life and promoting dignity and comfort. It is important to emphasize that PPC does not hasten or delay death, but it can have a positive influence on the course of the disease, which is an important aspect of prognosis. As they do not exclude other treatments, whether curative or supportive, PPCs should be integrated and complementary to the prevention, early diagnosis and treatment of serious, complex or life-limiting health problems, and should be used from the time of diagnosis and in conjunction with other therapies aimed at prolonging life with quality.⁹

Following this logic, serious and life-limiting genetic diseases such as Pompe disease are included in the indications for PPC. Pompe disease is a type II glycogen accumulation or acid alpha-glucosidase deficiency with a prevalence of 1/40,000 to 1/300,000.¹⁰ The main symptoms are cardiomyopathy, hypotonia and muscle weakness, dyspnoea and progressive loss of independent ventilation.¹¹ There are two types of presentation: infantile onset and late onset, depending on the age of onset, the organs affected and the rate of progression of the disease. Onset of the disease in children under 1 year of age is associated with delayed motor development, mainly affecting skeletal and cardiac muscles, and is characterized by a generalized decrease in muscle strength and tone, combined with cardiomyopathy. These manifestations often progress rapidly and, without timely and effective supportive treatment, often lead to cardiac and respiratory failure by the age of one year. Later-onset Pompe disease, on the other hand, affects children after the age of 1 year and mainly affects the muscles of the trunk, limbs and respiratory muscles, but involvement of the heart is relatively rare.¹²

The diagnosis is made by detecting a deficiency of the lysosomal acid alpha-glucosidase enzyme, which can be analyzed in dried or liquid blood samples, together with supportive biomarker tests, and further confirmed by molecular genetic analysis. Screening for this condition, targeting at-risk populations and universal newborn screening can lead to early diagnosis and allow immediate initiation of enzyme replacement therapy - the current option for this condition - contributing to improved clinical outcomes.¹² Although still incurable, patients who receive enzyme replacement therapy benefit in terms of survival and quality of life.^{13,10}

It should also be noted that screening and early diagnosis may allow patients with Pompe disease to receive enzyme replacement at an early stage of the disease, which has a positive impact on their prognosis. Therefore, due to its diverse clinical manifestations, multidisciplinary assessment during screening is essential for early detection and diagnosis.^{14,13,10}

The multi-system nature of the disease also requires comprehensive multidisciplinary management throughout treatment to improve patient outcomes.^{13,15} However, as it is an irreversible chronic disease with high morbidity and mortality and a low life expectancy, the introduction of palliative care from the moment of diagnosis is considered fundamental to promote comprehensive care at every stage of the child's development. It is also essential to support the family in the processes of preparation, planning and coping with the illness throughout the child's life.^{13,4,16}

The inclusion of PPC is currently considered the gold standard in the management of critically ill children and their families.¹⁷ Despite this, the timely referral and integration of PPC into ICUs remains variable, with a high incidence of late referral of children who would be indicated for this type of care.¹⁸

From this perspective, the decision to implement PPC is not an easy task and represents a considerable challenge for healthcare teams, given the lack of models to support this practice and, consequently, gaps in the training of professionals in this field.¹⁹ When there is space for clear dialogue and quality information between the patient and their family and all other members of the healthcare team about this type of decision, the suffering not only of the children but of all those involved in their care is reduced and a space of comfort care is created within the hospital environment.²⁰

The aim of this case study is to describe the experience of caring for a family going through the palliative care decision-making process for a child diagnosed with Pompe disease in the Pediatric Intensive Care Unit (PICU) of a university hospital in Southern Brazil.

Methods

This is a qualitative case study, focusing on data and testimonies from an ethics committee-approved study (CAAE: 58643816400005327) conducted between 2017 and 2018 in a university hospital in Southern Brazil. Case studies are chosen because they allow the analysis of holistic representations of knowledge, dependent on the context of practice, in its natural environment, generating data from multiple sources of conversation.²¹ Data for this study were collected using field notes, observations and semi-structured interviews with families of children admitted to the PICU and referred for exclusive palliative care in 2017. Therefore, the inclusion criteria were: to be a family member who experienced a conversation with health care professionals about palliative care referral in the PICU. The research participants were selected by convenience, and 11 family members responsible for patients admitted to the PICU with a clinical situation that justified the adoption of palliative care were invited to attend a meeting with the healthcare team for this proposal in the PICU between January and November 2017. All invitations to participate in the research were accepted. The invitations were made personally by the researcher – who, in the meantime, was a female nurse at the hospital and a master's student - in the hospital environment. Participants were given information about the aims of the research and a guarantee of anonymity, and expressed their agreement by signing the informed consent form. The semi-structured interviews were conducted by the researcher in a reserved room within the PICU between January and November 2017, and lasted an average of 45 minutes. Data saturation was reached after the 11th interview.²²

Data collection took place in audio-recorded dialogue sessions with the following guiding questions: What do you know about the child's illness? What was discussed about the child? Which professionals were present at the meeting? What was decided about the child's treatment as a result of the meeting with the health team? What do you feel about what was discussed?²²

The audio-recorded dialogues were transcribed by the researcher, stored on computer and subjected to thematic content analysis. The following steps were followed: 'floating reading', in order to immerse and imbibe the impressions and orientations contained in the qualitative materials; then the identification of 'units of meaning', consisting of the nuclei of meaning in each particular statement, in relation to the problem in focus; this continued with the "categorization and subcategorization" stage, which made it possible to form the set of descriptive categories by classifying the constituent elements of each participant's discourse through differentiation; and in the final stage, regrouping was carried out by recognising the common characteristics of these elements.²²

Ethical requirements were respected in accordance with the norms established by Resolution 466/2012 and 510/2016 of the National Health Council, Ministry of Health of Brazil. Participants' names were changed to pseudonyms to protect data confidentiality.

This study takes as its point of reference the experience of a mother and her five-month-old baby with a poor prognosis when faced with the process of deciding on palliative care, recorded during the research that led to the Master's thesis "Decision-making process about palliative care in a pediatric intensive care unit: communication, experiences and feelings".²²

Findings

The case described relates to the process of making a decision to provide exclusive palliative care for a five-month-old white male baby who had been transferred from another hospital to the University Hospital PICU in 2017 for investigation of hypotonia and extubation failure. He was followed up by the medical genetics service, and while awaiting tests reports and a primary diagnosis, new attempts at ventilator weaning and extubation were made in the PICU, without success. When the tests reports were finalized, in addition to the confirmation of Pompe disease, it was found that the child already had cardiomegaly and severe and irreversible impairment of his heart function. As a result, the process of deciding with the family on palliative care began.

The first approach to the subject was made only with representatives of the PICU medical team and the child's mother, without the knowledge and subsequent support of the other members of the team (nurses, psychologist, physiotherapist, nursing technicians). In this meeting, the situation of therapeutic exhaustion in relation to the child's progressive heart failure was discussed and three proposals were made to the family: stopping enzyme replacement (as it was not having a satisfactory effect), palliative extubation or a tracheostomy to allow the child to have a better quality of movement (without mechanical restraint because of the risk of accidental extubation). They also discussed the contraindication of resuscitation maneuvers in the event of a cardiac arrest.

The child's mother was quite shaken and stunned by the topics discussed and only expressed tears and sadness. She was unable to make a decision that day. Her emotional speech revealed:

Why didn't I say nothing? Well... I somehow felt... I can't say. But now I'm going to say it on Tuesday. I went home this weekend and I thought a lot, I wrote down some things on a piece of paper, everything I'm going to ask them [the medical team], everything I want to know. I know that Floriano is in danger. They want to say that Floriano is at risk if he has a tracheostomy. He's at risk because he's like that. So I think we have to try. Or do you want to leave your child on a bed with a tube up to...? You don't! Try to put yourself in my shoes, see your child there - as I explained to them - I'm not selfish! But I see that my son - it may be in the eyes of a mother, right? - but I see that Floriano is fine, you know? I don't think he's too sick to die, that doesn't even cross my mind! (Maria das Flores)

Faced with the philosophical-existential questions that arise in moments of extreme emotional fragility, spirituality emerges as a form of support for what is being experienced. Turning to a higher being proves to be a facilitating resource in these moments, as can be seen in the extract below:

I believe deeply in God, I have great faith in God, that God will do a miracle, I don't know... I have already asked God: may your will be done, but I don't want to see my son suffering in an intensive care bed!

I don't want that! Then, Lord please take him! Then, Lord, take him! Because we're getting more and more attached to him! I have spoken to God, I have been honest. If the Lord is going to take him, then take him once and for all! So don't leave me with this agony any longer! (Maria das Flores)

The mother's anguish was highlighted by her disagreement with the end-of-life diagnosis and the lack of therapeutic options for the child's illness, as described below:

And then I asked the doctors? They say, say, say that Floriano is going to die! All they can tell me is that Floriano is going to die. I know they're doctors, they have to tell me. But I want them to do their job! I want them to give me solutions! I won't rest until I see that they have done EVERYTHING, everything, everything. I want them to give him the enzyme, to give it every two weeks, because that's Floriano's rights. That's what we're going to fight for! Everything I'm saying now, I don't care if they find out, you know why? On Tuesday I'll be ready for that meeting and I'll tell them everything I want them to do! (Maria das Flores)

Following the mother's reaction to this initial discussion with the doctors, the members of the nursing team were deeply moved by the handling of the case. The nurses expressed feelings and perceptions of incomprehension and disagreement with the way in which the decision making had been carried out, stating that there had been no prior planning by the environment, the team or even the family for this moment. In light of this, a new meeting was arranged, this time with representatives of the whole healthcare team and other family members.

In the second approach, the issue of therapeutic exhaustion was revisited and the same three previous options were presented: stopping enzyme replacement, palliative extubation and tracheostomy, emphasizing that this last option involved the risk of the child dying during the procedure due to his weak heart. The mother was able to express her concerns about the child's discomfort with the endotracheal tube and the mechanical restraint required, as well as her position on the options presented by the team. Her comments were:

I was told there are two options: either a tracheostomy or taking him off the tube. Then they'll decide whether to do a tracheostomy to see if he'll prolong it if he stays with us a bit longer! I told them I wanted them to do it and make Floriano comfortable. The way he's in the tube, he can't stay there much longer. And I want them to do their job to the end. Do the necessary procedures. Everything with Floriano has risks, every procedure with Floriano has risks! If we don't try, he won't be able to stay like this. He doesn't feel well, he gets very agitated. I know he's on a lot of medication. So it's not easy for us, for me as a mother! (Maria das Flores)

The family opted for a tracheostomy because, despite the risk to the child during the surgical procedure, it was very painful to watch the baby suffer from the need to be restrained. As his neurological development was unaffected by the disease and his limb movements were quite active despite the hypotonia, by five months he was showing signs of annoyance at being restrained and was unable to perform simple activities such as putting his hands in his mouth or picking up objects.

What's more, as time passed and more experiences were made - such as the appearance of communication by looking at the mother, the first smile and other experiences related to the mother-baby relationship - the bond with the child became stronger, making it very difficult for her to cope with the grief of a disease with no chance of cure and a very short life expectancy. From the parents' point of view,

the aim of choosing a tracheostomy was to reduce the child's suffering caused by the endotracheal tube and to allow him to have a better quality of life and relationship for as long as his days lasted, because even with the cardiac dysfunction he didn't show that he was nearing the end of his life, being active and smiling.

The members of the nursing team involved in this case also saw the tracheostomy as the best option because, having lived closely with this family throughout their hospitalization, they had witnessed the quality of the bond between the child and his parents. They also saw the tracheostomy as comfort care, as it would allow the child to be more mobile, to be held by his parents, and perhaps to be discharged from the PICU with oxygen support for comfort.

The family's decision, although it disagreed with the opinion of the physician conducting the meeting, was accepted by the healthcare team, who referred the patient for a tracheostomy. The surgery was performed without complications and the child was discharged from the PICU after a few days of recovery with oxygen support. The death occurred in the pediatric inpatient unit, two months after the tracheostomy, in the presence of the parents, surrounded by all the palliative care available in the situation, focused on alleviating the symptoms and distress and providing the best possible quality of life for the child and the family in this last moment of life.

Discussion

Palliative care decision making remains a challenge in pediatric care settings around the world. Inadequate professional training in the ethical and legal aspects of this philosophy, and a lack of professionals and services specializing in PPC, are factors that create gaps in the development of models that can guide professionals towards this practice.^{19,23,8,24}

The PPC decision-making process is a situation that has the potential to create a deep state of sadness, anguish, despair and even anticipatory grief in the families of children with incurable diseases and a poor prognosis.²⁵ It is therefore essential that the palliative care decision-making process is carried out very carefully and in a multidisciplinary manner.²⁶

In this case we can see that the initial contact had created a conflict within the healthcare team because there was no initial discussion about the proposals to be presented to the family. According to a systematic review conducted by a panel of Canadian experts in Pompe disease, it is clear that the medical team's proposal to discontinue enzyme treatment and offer withdrawal and withholding measures was justified by the lack of evidence of improvement in musculoskeletal function after initiation of therapy and by the patient's short life expectancy, given the already extensive impairment in cardiac function. For this kind of case, the guidelines recommend treatments to relieve or prevent suffering and therefore improve quality of life, suggesting follow-up with a palliative care team.²⁷ However, what generated the ethical dilemma among the other healthcare professionals was the fact that the meeting with the mother took place suddenly, without prior discussion, without preparation of the environment and without the presence of other family members who could support her in such a sensitive decision-making situation, created a feeling of mistrust in the family and anxiety in the healthcare team. In order to deal with this situation, a new meeting had to be held, this time with advance notice, to which representatives of the family and the whole team were invited.

Studies carried out in Brazil show that non-medical professionals feel excluded from decision-making processes related to ethical

issues and palliative care, with no space to express their opinions, as if they were mere spectators of the process, since in many situations decision-making meetings are organized by the medical team directly with the parents, and other professionals in the team are not always involved. The impact of this type of situation on carers is personified in the form of moral distress and ambivalent feelings.^{22, 28-30}

This difficulty is overcome by creating a space for dialogue and exchange of quality information about difficult and sensitive decisions, such as the one presented in this case study, involving the child, when possible, his or her primary caregiver, other family members accompanying the child, and the healthcare team caring for the child and his or her family. It is therefore possible to observe not only the reduction in suffering for the child, but also for all those involved in the care, as a space of comfort, compassion and complicity is created in the hospital environment.²⁰

The participation of children and families in the PPC decision-making process is an element that still needs more attention in the health reality of the pediatric population. One of the central elements of the Family-Centered Care (FCC) philosophy focuses on providing opportunities for family members to exercise their autonomy and choose the role they want to play in making decisions about health issues affecting their children. The American Academy of Paediatrics (AAP) recommends the use of Shared Decision Making (SDM), a decision-making model in which decisions are made jointly - by the health care team, the child and the family - based on the best available scientific evidence, but also taking into account the family's values, culture and preferences, in order to consider the best interests of the child.^{3,20,31-33}

The decision about palliative care, shared between the multidisciplinary health care team and the family, is not only a right but also a duty of health professionals to give the family and, where possible, the child/young person, a voice in decisions about their body, their health and their life.³⁴

Conclusion

Trying to understand each family's concept of quality of life, especially in the face of the adversity caused by the severity of a child's illness when the possibilities of cure have been exhausted, is one of the crucial points to prioritize when planning the decision-making process for the adoption of PPC. The study reveals an understanding of the need for sensitive and qualified listening in order to be able to continue the discussion about which therapeutic approach is the most appropriate and which decisions best serve the terminality situation experienced by each child-family binomial. Respecting different points of view, despite one's own, is a sign of maturity, humanism and professionalism, and requires a great deal of compassion and empathy. On the other hand, acting in this way, in harmony with the child and their family, becomes easier as the bonds of the interpersonal relationship with the child and their family become closer. Affective distancing, rather than helping, makes the process even more difficult for both the professional and the family.

Practice implications

The participation of the child's or adolescent's parents, other family members as a support network, and the multidisciplinary team in the PPC decision-making process is essential to give the child and the family a voice, to minimize their doubts and suffering, and to offer a welcome in a situation of existential vulnerability represented by the imminent finiteness of the child's life, which intensely exposes the fragility of the human condition.

Declaration of Generative AI and AI-assisted technologies in the writing process

During the preparation of this work the author(s) used DeepLWrite in order to improve readability and language. After using this tool, the author(s) reviewed and edited the content as needed and take(s) full responsibility for the content of the publication.

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Declaration of interests

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

References

- Floriani CA, Schramm FR. Palliative care: interfaces, conflicts and needs. *Ciênc saúde coletiva*. 2008;13:2123–2132.
- Schneider AS, Flurin Ludwig MC, Neis M, et al. Perceptions and experiences of the nursing team before the pediatric patient in palliative care. *Cienc Cuid Saude*. 2020;190.
- AAP, American Academy of Pediatrics. Palliative Care for Children. *Pediatrics*. 2000;106(351):351–357.
- Harrop E, Edwards C. How and when to refer a child for specialist paediatric palliative care. *Arch Dis Child Educ Pract*. 2013;98(6):202–208.
- Pinho AAA, Nascimento IRC, Ramos IWS, Alencar VO. Repercussions of paediatric palliative care: an integrative review. *Rev Bioét*. 2020;28(4):710–717.
- Instituto Nacional do Câncer. *[Pediatric Palliative Care]*. 2023.
- Sociedade Brasileira de Pediatria. *[Paediatric Palliative Care: What is it and how important is it? Caring for children at all times]*. 2018.
- World Health Organization. *Integrating palliative care and symptom relief into paediatrics: a WHO guide for health care planners, implementers and managers*. Geneva: World Health Organization; 2018. Licence: CC BY-NC-SA 3.0 IGO.
- Ferreira EAL, Valdez L. *[Pediatric Palliative Care: what is it and why does it matter?] in Seção 40, Medicina da Dor e Cuidados Paliativos*. Tratado de Pediatria. 5a edição. Rio de Janeiro: Manole; 2021. p. 1712.
- Jiao KX, Yue DY, Zhu WH. Advances in the specific treatments of Pompe disease. *Zhonghua Nei Ke Za Zhi*. 2023;62(4):454–459.
- Kohler L, Puertollano R, Raben N. Pompe Disease: From Basic Science to Therapy. *Neurotherapeutics*. 2018;15(4):928–942.
- Grupo de Endocrinologia, Genética e Metabolismo do Ramo de Pediatria da Associação Médica Chinesa, Ramo de Genética Médica da Associação Médica Chinesa, Grupo de Doenças Raras do Ramo de Pediatria da Associação Médica Chinesa, etc. [Chinese expert consensus on the diagnosis and treatment of glycogen storage disease in children]. *Chinese Journal of Pediatrics*. 2021;59(6):439–445.
- Benedetto L, Musumeci O, Giordano A, et al. Assessment of Parental Needs and Quality of Life in Children with a Rare Neuromuscular Disease (Pompe Disease): A Quantitative–Qualitative Study. *Behavioral Sciences*. 2023;13(12):956.
- Sawada T, Kido J, Nakamura K. Newborn Screening for Pompe Disease. *International Journal of Neonatal Screening*. 2020;6(2):31.
- Gonglu L, Zhiying W. Multidisciplinary management of Pompe disease. *Chin J Intern Med*. 2023;62(6):728–732.
- Morrison WE, Gauvin F, Johnson E, et al. Integrating palliative care into the ICU: from core competency to consultative expertise. *Pediatr Crit Care Med*. 2018;19(8S Suppl 2):S86–S91.
- Grunauer M, Mikesell C, Bustamante CG. Primary palliative care integrated model in paediatric ICU: an international cross-sectional study. *Brit Med J Supportive & Palliative Care*. 2021.
- Moynihan KM, Snaman JM, Kaye EC, et al. Integration of pediatric palliative care into cardiac intensive care: a champion-based model. *Pediatrics*. 2019;144(2):1–12.
- Piva JP, Garcia PCR, Lago PM. [Dilemmas and difficulties surrounding end-of-life decisions and the provision of palliative care in paediatrics]. *Rev bras ter intensiva*. 2011;23(1):78–86.
- De Vos MA, Bos AP, Plötz FB, et al. Talking with parents about end-of-life decisions for their children. *Pediatrics*. 2015;135(2):e465–e476.
- Miles R. Complexity, representation and practice: Case study as method and methodology. *Issues in Educational Research*. 2015;25(3):309–318.
- Neis M. *[Decision-making about palliative care in a paediatric intensive care unit: communication, experiences and feelings] [dissertação de Mestrado em Saúde da Criança e do Adolescente]*. Faculdade de Medicina, Universidade Federal do Rio Grande do Sul, Porto Alegre, 2018.
- Gomes ALZ, Othero MB. Cuidados paliativos. *Estud av*. 2016;30(88):155–166.
- D'Alessandro MPS (coord.), Pires CT, Forte DN, et al. *[Palliative Care Manual]*. São Paulo: Hospital Sírio Libanês, Ministério da Saúde, 2020.
- Neis M, Issi HB, Motta M da GC da, et al. The experience of families in face of the finiteness of their children in the process of adopting palliative care. *Rev Gaúcha Enferm*. 2022;43:e20220174.
- Rocha MDS, Silva RS, Amaral JB. [Interdisciplinarity: a path to comprehensive care for people in palliative care]. In: Silva RS, et al., editors. *[Palliative Care Nursing caring for a good death]*. São Paulo: Editora Martinari. 2014;353–363.
- Tarnopolsky M, Katzberg H, Petrof BJ, et al. Pompe Disease: Diagnosis and Management. Evidence-Based Guidelines from a Canadian Expert Panel. *Canadian Journal of Neurological Sciences / Journal Canadien des Sciences Neurologiques*. 2016;43(4):472–485.
- Trotta EA, Scarpa FC, Halal MG, et al. Health professionals' perceptions about the decision-making process in the care of pediatric patients. *Rev Bras Ter Intensiva*. 2016;28(3):335–340.
- Santos RP, Garros D, Carnevale F. Difficult decisions in pediatric practice and moral distress in the intensive care unit. *Rev bras ter intensiva*. 2018;30(2):226–232.
- Nilson C, Vieira AC, Bubadué RM, et al. Decision-Making Process for the Implementation of the Child Therapeutic Support Limitation Plan: Nurses' Experiences. *Inquiry*. 2022;59:469580221100777.
- Dreesens D, Veul L, Westermann J, et al. The clinical practice guideline palliative care for children and other strategies to enhance shared decision-making in pediatric palliative care; pediatricians' critical reflections. *BMC Pediatr*. 2019;19(1):467.
- Bobillo-Perez S, Segura S, Girona-Alarcon M, et al. End-of-life care in a pediatric intensive care unit: the impact of the development of a palliative care unit. *BMC Palliative Care*. 2020;19(1):1–8.
- Eler K, Albuquerque A. *[Human rights of pediatric patients in the context of palliative care]*. In: Ferreira EAL, et al., editors. *[Pediatric Palliative Care]*. Rio de Janeiro: Medbook; 2023, p. 57–61.
- Santoro JD, Bennett M. Ethics of End of Life Decisions in Pediatrics: A Narrative Review of the Roles of Caregivers, Shared Decision-Making, and Patient Centered Values. *Behav Sci (Basel)*. 2018;8(5):42.