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Case Report

Performance of an interdisciplinary hospital team in pediatric palliative care: case report

Atuação de uma equipe interdisciplinar hospitalar nos cuidados paliativos pediátricos: relato de caso

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ABSTRACT

The article aims to report a rare case of a patient diagnosed with Thanatophoric Dysplasia admitted to the Neonatal Intensive Care Unit of a tertiary hospital in a city in the inner state of São Paulo. The female infant has been hospitalized since birth, with history, physical, and radiological examination suggestive of Thanatophoric Dysplasia and an unaltered karyotype test, whose evolution and severity made it necessary to institute minimally invasive care. She remained in the Neonatal Intensive Care Unit for five months and 23 days requiring invasive mechanical ventilation through a tracheostomy and gastrostomy feeding, until the irreversible nature of the condition was determined. The purpose of this case report aims to promote reflection on the importance of a multidisciplinary approach to patient care, providing quality of life, orthothanasia, and the importance of family-centered care practices.

Keywords: Thanatophoric Dysplasia; Palliative Care; Congenital Anomalies.

RESUMO

O artigo tem por objetivo relatar um caso raro de uma paciente com diagnóstico de Displasia Tanatofórica internada na Unidade de Tratamento Intensivo Neonatal de um hospital terciário de uma cidade do interior paulista. A lactente do sexo feminino, internada desde o nascimento, com história, exame físico e radiológico sugestivos de Displasia Tanatofórica e exame de cariótipo sem alterações, cuja evolução e gravidade fez-se necessário instituir cuidados minimamente invasivos. Permaneceu internada na Unidade de Tratamento Intensivo Neonatal por cinco meses e 23 dias com necessidade de ventilação mecânica invasiva por meio de traqueostomia e alimentação por gastrostomia, até definição do caráter irreversível da patologia. Este relato de caso tem o propósito de promover uma reflexão da importância de uma abordagem multidisciplinar com acolhimento ao paciente, oportunizando qualidade de vida, proporcionando a ortotanásia e a importância das práticas de cuidados centrados na família.

Palavras-chaves: Displasia Tanatofórica; Cuidados Paliativos; Malformações Fetais.

INTRODUCTION

Pediatric Palliative Care (PPC) goes beyond prolonging the life of children with chronic and progressive pathologies, offering quality of life, relieving suffering, whether physical, spiritual, emotional or social[1], and sharing decisions and therapeutic goals with the child and their family. Multidisciplinary care involves the technical and emotional preparation of the team[2]. It is a unique approach, providing safety, respect and autonomy for the family group and excellence in patient-centered care. In the Intensive Care Unit (ICU), care for children is at a tenuous limit, especially at times when life cannot be preserved or maintained. In this case, total pain management (i.e., physical, emotional, social,

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and spiritual) is important and should be started as early as possible, enabling comfort by planning interventions aimed at the child and their guardians[3].

There are situations in the perinatal period where palliative care is expected, such as extreme premature infants, congenital malformations and some pathologies that can have the patient's survival increased due to technological advances[2]. The focus is on providing a better quality of life for the newborn and their family or guardian, transcending health care and promoting a welcoming environment that is consistent with their values.

Considering palliative care at home, management strategies and adaptations should be discussed before and after discharge and shared with the family or guardian, setting goals that fit into family dynamics[4]. "*As important as caring for those who are sick is caring for those who care*", as Vattimo[5] describes. Therefore, planning, and dividing tasks in a family environment improves the quality of life of everyone involved.

In the event of death, the grieving process should be anticipated and elaborated. The anxieties should be recognized, farewell and tribute rituals should be prepared and discussed, and it is also important to provide mental health support, so that the family is welcomed during and after the death[6]. Grief is not defined only by death, but by the losses that the disease can cause, and it is essential that the professional is prepared to assist in this adaptation process that the patient and their family members will go through[7].

In 2018, the World Health Organization (WHO) launched a complement to the indication of PPC, suggested by the European Association for Palliative Care (EAPC) in 2009. The classification contains six groups, with Thanatophoric Dysplasia (TD) being an expected condition that requires palliative care, as it is classified in WHO group 5[4].

TD accompanies bone abnormalities, most of the time lethal ones, shortening of the upper and lower limbs, small rib cage, reduced height and flattened vertebral bodies, and macrocephaly[8]. Subdivided into types I and II, differentiated by the characteristic of the femur and the autosomal dominant mutation, genes Lys650Glu and Gly380Arg[9]. Because the rib cage is restrictive, and the marked pulmonary hypoplasia is present, infants and newborns end up developing respiratory failure followed by cardiorespiratory arrest[10].

The objective of this study is to share the evolution of a rare case, following the natural history of the disease and thus contributing to the management of new similar cases. The case report was approved by the Research Ethics Committee of the *Faculdade de Medicina São Leopoldo Mandic*, Campinas, SP, Brazil, under CAAE 63224922.0.0000.5374; the parents signed a free and informed consent form (TCLE) authorizing the report of the case.

CASE REPORT

A 32-year-old mother G3PC2A0 (3 pregnancies, 2 cesarean deliveries, 0 abortion), two previous children who were 14 and 10 years old, with no apparent comorbidities. She started prenatal care at 12 weeks of gestation and had a total of seven consultations during her pregnancy. VDRL, HIV, and Hepatitis B serology were negative, rubella was susceptible and group B Streptococcus testing was not performed.

The morphological ultrasound (US) on April 20, 2021, at 24 weeks of gestation, as well as confirmed with two other US performed on June 10, 2021, and July 1, 2021, showed a narrow chest, upper and lower limb deformities (severe micromelia) and relative cardiomegaly.

Vaginal delivery was on July 3, 2021, at 34 weeks and four days gestation, with an 18-hour bag rotation. Resuscitation was carried out in the delivery room with two cycles of positive pressure ventilation, followed by orotracheal intubation. There was an unsuccessful attempt at extubation when she was admitted to the neonatal ICU and she was promptly reintubated, requiring moderate ventilator settings.

Newborn data was Capurro, 34 weeks gestational age, birth weight 1,880 g, length 33 cm, head circumference 33 cm and chest circumference 23 cm. Apgar score of 6 in the 1st minute and 8 in the 5th minute. Typical female genitalia. Infectious screening was carried out at 12 hours of life due to the prolonged time the amniotic sac had been ruptured, but there were no significant alterations.

An echocardiogram was carried out on July 7, 2021, showing normal cardiac cavities and a patent foramen ovale, with no hemodynamic repercussions. Transfontanelle ultrasound on September 1, 2021, with grade I hydrocephalus, without compression of the parenchyma, and abdominal ultrasound, carried out on the same day, without alterations. Karyotype without abnormalities,46 XX. Chest X-ray showing reduced dimensions of the rib cage, relative reduction of lung fields, shortened and flattened limbs.

She presented with prematurity jaundice, requiring phototherapy, and late-onset sepsis caused by *Klebsiella pneumoniae*, *Enterococcus faecalis* and *Staphylococcus epidermidis*, the latter from a difficult-to-access phlebotomy. Contact precautions were taken and treatment with systemic antibiotics was recommended by the infectious disease specialist.

Over the days, the infant did not tolerate the progression of the oroenteral tube diet and required a nasogastric tube to decompress her abdomen. The decision was made to perform a tracheostomy and gastrostomy. The growth of the chest did not keep pace with the growth of the rest of the trunk and the patient had frequent episodes of bronchospasm and respiratory discomfort. The gastrostomy was replaced with a nasoenteral tube, due to a false route.

Sedation and analgesia were maintained, with the aim of keeping her at Ramsay Scale 4[11] for most of the time. In this way, the mother was able to provide care and monitor the child's development, which was neurologically adequate for the age group.

As her diagnosis had a guarded prognosis, she opted to follow up with a PPC involving the entire neonatal ICU team (doctors, nurses, nursing technicians, speech therapists, psychologists and social services), drawing up a care plan aimed at preventing and relieving physical, spiritual, social, and emotional suffering, with the aim of providing a favorable quality of life for the child and her parents.

The mother spent most of the time with her daughter. The team provided care to ease the child's discomfort, breathing and pain, as well as assisting the family emotionally, always sharing their anxieties and decisions with the parents. The pain was alleviated with opioids and non-pharmacological measures such as skin-to-skin contact with the mother, a mild temperature and music from the mobile.

Clinical deterioration and hemodynamic instability worsened in the last week of life, with frequent drops in saturation accompanied by bradycardia, convulsions, and abdominal distension. She required high ventilator settings, reaching an inspired oxygen fraction of 100%, a progressive respiratory rate of up to 47 per minute, inspiratory pressure gradually rising to 32 cmH2O and expiratory pressure between 4 and 6 cmH2O. The condition intensified, and in agreement with the multidisciplinary team and the parents, the decision was made to suspend the nasoenteric tube diet and vasoactive drugs in the last 24 h of life, which evolved into cardiorespiratory arrest.

During hospitalization, the family's faith was encouraged, respecting their rituals and this practice was crucial at the time of death, providing spiritual comfort. The parents invited any professionals they felt comfortable with to take part in a prayer service. The infant then died at the age of five months and 23 days. The family remained emotionally supported during hospitalization and in the dying process. The clear and welcoming approach to diagnosis and prognosis throughout the hospitalization process allowed family members to conceive of finitude as part of the care process. This family intrinsically had internal resources to overcome this phase. Faith was a primary resource for this nucleus, being key for the care and relationship between caregivers and the team[5].

DISCUSSION

The evolution and qualification of neonatal care has brought with it a greater number of surviving children who need special care, requiring qualified and humanized care backed by bioethics to offer a life with dignity[12]. Because they remain with serious sequelae that will accompany them throughout their lives, it is necessary to provide family support whenever the decision is made to start palliative care[13]. The autonomy and complexity of human beings must be respected, even if they are children, although society often consciously fails to respect this right[14].

Involving parents in all stages of care enhances the decisions made and thus later facilitates the grieving process. RBS's parents were present and participative in defining the care proposed for their newborn. They were aware of the processes and experienced every moment alongside their daughter, playing a leading role in palliative care.

When implementing globalized care, caution must be taken to do so at the appropriate and opportune moment, with the aim of serving the patient and their loved ones in a unique and welcoming way[15]. All the professionals involved must be technically and emotionally trained to provide adequate support[2]. In the case reported in this article, the support began before birth, providing dialogue with the family and clarifying doubts. Decisions were discussed and shared with the aim of welcoming the child and her parents, relieving physical and emotional suffering, and providing orthothanasia. The preparation of the entire team involved in the care was necessary, based on respect and responsibility, which must be continuous in supporting grieving. Each case monitored by the team is unique and allows for new experiences and learning. Teamwork allows for knowledge exchange and mutual support in dealing with new situations and emotions. Recognizing weaknesses and receiving support within the team itself was the basis of care during the process[15].

CONCLUSION

Pediatric Palliative Care has brought new perspectives on caring. Besides caring for the child, it expands attention to family members and responsible caregivers. Through multidisciplinarity, person-centered care is provided, with the aim of promoting quality of life and relieving physical, spiritual, emotional, and social suffering.

REFERENCES

1. Ferreira EAL, Ferreira FTL, Cardoso MGL, Rocha SR. Princípios de Gestão de Equipe na Saúde da Criança. In: Ferreira EAL, Barbosa SMM, Iglesias SBO (eds). Cuidados Paliativos Pediátricos. Rio de Janeiro: Medbook, 2023. p. 75-78.
2. Lopes CMC (2021) Cuidados paliativos em unidade de tratamento intensivo neonatal / Palliative care in a neonatal intensive care unit. Braz J Hea Rev [Internet]. 2021 Nov [cited 2021 Nov 19];4(6):25731-53. Available from: https://doi.org/10.34119/bjhrv4n6-172.
3. Castro MCF, Fuly PSC, Santos MLSC, Chagas MC. Total pain and comfort theory: implications in the care to patients in oncology palliative care. Rev Gaúcha Enferm [Internet]. 2021 [cited 2024 Mar 18];42:e20200311. Available from: https://doi.org/10.1590/1983-1447.2021.20200311.
4. Ferreira EAL, Iglesias SBO, Lima JS. Cuidados Paliativos Pediátricos. Definição e indicação. In: Ferreira EAL, Barbosa SMM, Iglesias SBO (eds). Cuidados Paliativos Pediátricos. Rio de Janeiro: Medbook, 2023. p. 6-9.
5. Vattimo EFQ. Saúde Mental dos Profissionais de Cuidados Paliativos e dos Cuidadores. Vattimo EFQ, Belfiore EBR, Zen Junior JH, Santana VS (eds). Cuidados paliativos: da clínica à bioética. São Paulo: Cremesp, 2023. vol 2. p. 665-81.
6. D'Alessandro MPS, Pires CT, Forte DN *et al*. Manual de Cuidados Paliativos. São Paulo: Hospital Sírio Libanês; Ministério da Saúde, 2020. Aspectos gerais do luto e peculiares no contexto da pandemia de COVID-19, p. 75-80.
7. Vattimo EFQ. Luto. In: Vattimo EFQ, Belfiore EBR, Zen Junior JH, Santana VS (eds). Cuidados paliativos: da clínica à bioética. São Paulo: Cremesp, 2023. vol 2. p. 563-94.
8. Isaacson G, Blakemore KJ, Chervenak FA. Thanatophoric dysplasia with cloverleaf skull. Am J Dis Child. 1983 Sep [cited 201 Dez 21];137(9):896-8. Available from: https://doi.org/10.1001/archpedi.1983.02140350070017.
9. Wilcox WR, Tavormina PL, Krakow D, Kitoh H, Lachman RS, Wasmuth JJ, Thompson LM, Rimoin DL. Molecular, radiologic, and histopathologic correlations in thanatophoric dysplasia. Am J Med Genet. 1998 Jul [cited 2021 Dez 19];78(3):274-81. Available from: https://doi.org/10.1002/(sici)1096-8628(19980707)78:3<274::aid-ajmg14>3.0.co;2-c.
10. Noronha L, Prevedello LM, Maggio EM, Serapiao MJ, Torres LF. Displasia tanatofórica: Relato de dois casos com estudo neuropatológico [Thanatophoric dysplasia: report of 2 cases with neuropathological study]. Arq Neuropsiquiatr. 2002 Mar [cited 2021 Nov 12];60(1):133-7. Available from: https://doi.org/10.1590/s0004-282x2002000100024.
11. Mendes CL, Vasconcelos LC, Tavares JS, Fontan SB, Ferreira DC, Diniz LA, Alves ES, Villar EJ, Albuquerque Cde F, Silva SL. Ramsay and Richmond's scores are equivalent to assessment sedation level on critical patients. Rev Bras Ter Intensiva. 2008 Dec;20(4):344-8.
12. Gaiva MAM. O cuidar em unidades de cuidados intensivos neonatais: em busca de um cuidado ético e humanizado. Cogitare Enferm. 2006 Jan [cited 2021 Nov 12];11(1):61-6. Available from: https://revistas.ufpr.br/cogitare/article/view/5976/4276
13. Piva JP, Garcia PC, Lago PM. Dilemmas and difficulties involving end-of-life decisions and palliative care in children. Rev Bras Ter Intensiva. 2011 Mar;23(1):78-86.
14. Segre M. Considerações sobre o princípio de autonomia. Rev Medicina. 1995;28:10-5.
15. Fernandes MA, Costa SFG, Morais GSN, Duarte MCS, Zaccara AAL, Batista PSS. Cuidados paliativos e luto: um estudo bibliométrico. Esc Anna Nery. 2016 [cited 2024 Mar 7];20(4):e20160102. Available from: https://doi.org/10.5935/1414-8145.20160102