

Perinatal palliative care following prenatal diagnosis of severe fetal anomaly: a new family-centered approach in a level III Portuguese hospital

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Abstract

Perinatal palliative care (PNPC) is an emerging field that aims to improve the quality of life of families affected by an antenatal diagnosis incompatible with long-term survival through a specialized, multidisciplinary approach and holistic and emotional support. An advance care planning begins at the moment of diagnosis and continues in the postpartum period addressing comfort measures to alleviate pain, psychological support for family members and the collection of mementos that allow parents to make lasting memories of their child.

PNPC is also an integral part of the Newborn Individualized Developmental Care and Assessment Program (NIDCAP) in Obstetrics and Neonatology. Since 1997, there are more than 300 programs described, tailored to the newborn's best interest.

Up to now, providers often feel difficulties in the implementation of PNPC programs, given the lack of evidence-based quantitative empirical studies that are necessary to establish the best model of care. Obstacles to palliative care include diagnostic and prognostic uncertainty and logistic obstacles related to interdisciplinary collaboration. Literature also suggests a great discrepancy involving end-of-life decisions between different countries.

This paper presents an overview of the first PNPC program implemented in a level III Portuguese Neonatal Intensive Care Unit (NICU) with two

case studies providing a distinctive perspective on the support and care needed. It also seeks to serve as a resource for other institutions since there are no well-established and published guidelines regarding PNPC in Portugal. It is also important to include obstetric residents, midwives, medical students and nursing students in the provision of palliative care so this model of care can be incorporated into future practices.

Keywords

Perinatal palliative care, end-of-life care, perinatal loss, bereavement, multidisciplinary team.

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Introduction

Over the last twenty years, the field of perinatal palliative care (PNPC) has been burgeoning all over the world [1, 2]. Since the first recognition of a perinatal hospice in 1997 [3], in which the concept of PNPC was first introduced in the literature, there are now almost 300 PNPC programs described in the USA. In Europe, ten countries have already implemented the concept, including France, Germany, Italy, Spain and the Czech Republic [4].

Technological support and medical expertise made possible to screen for a wide range of conditions at early gestational age [5]. Nonetheless, life-threatening conditions, defined as those with “no reasonable hope of cure that will ultimately be fatal”, are still one of the leading causes of death in Portugal’s Neonatal Intensive Care Units (NICUs) [6]. Recently, Moura et al. showed that despite an increase in the withholding and withdrawing of medical interventions and improvements in palliative care in the last three decades, a large number of neonates with life-limiting conditions still receive aggressive treatments towards end-of-

life [7]. Later, Parravicini et al. reported that the provision of intensive care did not prevent death nor prolong life of these infants compared with that of infants treated with individualized comfort measures [8].

PNPC should be considered in three main categories: 1) newborns at the threshold of viability; 2) newborns with complex congenital anomalies considered to be incompatible with long-term survival; and 3) newborns with severe clinical conditions not responding to aggressive cure-oriented treatments, for whom continuation of intensive care is no longer helpful [12, 13].

Facing a prenatal diagnosis of an incompatible with prolonged life anomaly, parents often experience feelings of shock and anticipatory grief [9]. Although the majority of these parents choose elective termination of pregnancy, research shows that the choice of continuing a pregnancy, whether for personal, cultural or religious reasons, is on the rise, ranging from 20% to 87% [9-11]. PNPC can be a valuable option in these cases focusing on the holistic and multidisciplinary approach for families affected by a poor antenatal diagnosis. It is a carefully planned intervention to minimize the physical, psychological, social, emotional and spiritual suffering these families face throughout the pregnancy and the birth of the newborn [12]. An advance care planning begins at the moment of diagnosis and continues in the postpartum period addressing comfort measures to alleviate pain and discomfort, psychological support for family members and the collection of mementos that allow parents to make lasting memories of their child [10, 12]. In 2012, a systematic review of the literature by Balaguer et al. concluded that the various aspects of PNPC should include: “pain relief, comfort, maternal bonding, family-centered care, comprehensiveness and integrative care started early” [10].

Since 2010, the Portuguese Neonatal Society has been focusing on perinatal and neonatal care with several workshops and educational lectures around the country. However, there are no well-established and published guidelines regarding PNPC in Portugal [14, 15]. Providers often feel they have little training in end-of-life care and feel difficulties in the implementation of such programs, given the lack of evidence-based quantitative empirical studies that are necessary to establish the best model of care [1, 10, 16].

The following cases seek to illustrate the development and implementation of a PNPC

protocol (**Appendix A**) following prenatal diagnosis of severe fetal anomalies in a Portuguese level III hospital. To this end, a multidisciplinary team of an obstetrician, a neonatologist, a psychologist and a nurse was set up. Considering the large number of professionals involved, a key-point person was designated, keeping an open line of communication to address parents' needs. This is the first report published about creating a suitable birth plan tailored to the newborn's best interest and the expectations and bereavement needs of the parents in this hospital. It also seeks to serve as a resource for other institutions, as the literature suggests a great discrepancy involving end-of-life decisions between different countries [17].

Case 1

A 40-year-old Gravida 2 Para 1, with a previous iatrogenic preterm delivery at 28 weeks gestation with severe pre-eclampsia. The combined screening test was negative, with a risk of 1:800 for trisomy 21. The second-trimester ultrasound at 21 weeks gestation showed multiple anomalies: agenesis of corpus callosum, agenesis of cerebellar vermis, bilateral microphthalmia, bilateral cleft lip and palate, left diaphragmatic hernia, ventricular septal defect, bilateral postaxial polydactyly and hypospadias. The provider performed an amniocentesis and a karyotype 47,XY,+13 was found. Parents were informed about the poor prognosis related to this genetic condition. Despite the possibility of pregnancy termination, the couple decided to continue the journey until its natural end.

After a short period of time, a meeting with the palliative care team took place. During this meeting, a birth plan (**Appendix A**) was delicately elaborated through shared decision-making. It was gently decided that active resuscitation would not be in the baby's best interests and so the focus of care would be to provide comfort.

At 37 weeks of gestation, the labor was induced using a Foley catheter and oxytocin. No fetal heart monitoring was used during labor. Baby L. was born alive with 2,950 g, 37 cm of head circumference and 47 cm of body length with an APGAR score of 1/1/1.

After birth, Baby L.'s parents cut the umbilical cord. No vitamin K injection or antibiotic eye ointment was made.

Comfort care took place in the delivery room where he had routine care and was then

wrapped in hot blankets and kept in his mother's lap with skin-to-skin contact. During L.'s short two hours of life, the team made arrangements for him to be baptized in the delivery room by a hospital chaplain chosen by the parents. Although the birth plan contemplated the possibility of breastfeeding, as long as it was not a source of discomfort either for the mother or the baby, Baby L. did not breastfeed during his short time of life. Mementos as footprint were collected. No pharmacological pain relief was necessary as Baby L. never demonstrated signs of discomfort or pain, only with sporadic gasping and his short two hours of life were spent comfortably with his mother.

After he died, the team reunited with his parents explained all the emotional and bereavement support that was at their disposal. Baby L.'s parents declined psychological support. Autopsy was not authorized by the parents. They performed a funeral.

Case 2

A 42-year-old Gravida 2 Para 1, with an unremarkable medical history except for asthma (with SOS medication) and a first uncomplicated pregnancy. The combined screening test was positive with a risk of 1:7 for trisomy 18. An amniocentesis was performed at 16 weeks, and the karyotype was 47,XX,+18. The ultrasound performed at 18 weeks gestation noted several abnormalities: strawberry-shaped head, hypertelorism, myelomeningocele, clenched hands, club feet and hypoplastic left heart syndrome.

Parents of Baby P. were informed about the poor prognosis of this condition and, after all options discussed, the couple decided for palliative care and were referred to our institution. They agreed that intensive resuscitation measures would not be in their child's best interest, taking into account her life-threatening condition.

A meeting with the PNPC team took place. The team reassured parents that withholding of life-sustaining treatment did not mean that care of their baby girl would be withdrawn, only there would be a change in the focus of care, always making sure Baby P. would not experience any distress or pain. Through shared decision, a birth plan (**Appendix A**) was tailored to the parents' needs.

At 40 weeks of gestation, the labor was induced using a Foley catheter and oxytocin. No fetal heart monitoring was used during labor. Baby

P. was born alive with 2,445 g, 30.5 cm of head circumference and 45.5 cm of body length with an APGAR score of 7/7/7. No vitamin K injection or antibiotic eye ointment was conducted.

Baby P.'s comfort measures and palliative care began in the delivery room. She was kept in her mother's lap after being dressed in clothes chosen by her parents. As there were no agitation or distress signs, no pharmacological pain medication was needed. In agreement with the birth plan, Baby P. was not breastfed during this time, according to mother's wish.

A priest chosen by her parents, friend of the family, performed the baptism ceremony. She lived for two hours, during which the parents were always with her, collecting memories and keepsakes.

Psychological and emotional support was offered by the palliative care team for as long as required and arrangements were made in order to perform a funeral ceremony. Baby P.'s parents declined psychological support and did not authorize the autopsy.

Discussion

Portugal has one of Europe's lowest perinatal mortality rate, < 0.25/1,000 births as shown in EUROCAT public health indicators for congenital anomalies in Europe [18]. Finding these cases in the NICU of Centro Hospitalar de São João is not an uncommon challenge. From 1st January 2000 to 31st December 2014, 1,222 cases of congenital malformations were notified by this institution, 63% of which were detected through prenatal diagnosis. In 2015, 2,052 consultations of prenatal diagnosis were also held in this hospital [19]. However, the integration of palliative care in the perinatal period is quite recent, and the cases described above are the first ones in which a PNPC program was implemented.

Choosing palliative care

Before delivery, time allows parents to reflect on their baby's condition and goals of care. Choosing the best kind of care for a baby depends on three main variables: the diagnosis, the prognosis, and the meaning that the parents attribute to the prognosis [20, 21]. In both two cases, the confirmation of the diagnosis was a vital step in the process: structural abnormalities were detected by routine ultrasound screening and the karyotype study provided in

both cases the necessary diagnostic confirmation. However, other cases not associated with genetic alterations, such as complex or multiple structural anomalies, may require different diagnostic tests such 3D ultrasound, echocardiography, MRI and consultation by a senior perinatologist, cardiologist or neurologist, depending on the condition [13, 22]. If the prognosis is still uncertain (e.g., severe diaphragmatic hernia) delivery room assessment is needed until a definitive diagnosis or prognosis can be established [13, 21].

The prognosis in both cases was quite certain: life-threatening conditions (Patau's syndrome and Edwards' syndrome) with a very short extrauterine life in most cases, where the provision of intensive treatment could be considered irresponsible and even ethically reprehensible [23]. So, palliative care was the most appropriate option.

Parents usually decide to terminate or continue the pregnancy in 1.5 days, as described by Breeze et al. [24]. This results in the need to address PNPC as early as possible. Also, recent 2016 prenatal testing practice guidelines from the American College of Obstetricians and Gynecologists and the Society for Maternal-Fetal Medicine also state that post-diagnosis counseling should include the option of PNPC. Despite this, the literature suggests a two-month gap between the moment of potential diagnosis and referral for palliative services [2]. Baby L. and Baby P. parents' both chose to take the pregnancy to term after confirmation of the diagnosis and the option of palliative care was promptly promoted.

Creating a birth plan

After the decision for palliative care has been made, during pregnancy, successive meetings between Baby L. and Baby P.'s parents and the palliative care team allowed them to co-create a birth plan that answered their needs. This document also provided a communication tool among the professionals who participated in the care of these families, avoiding burdensome discussions with parents. The birth plan should be clearly documented in the mother's electronic medical record [24, 25], so the team can have access to important aspects at any time.

Prenatal care

While advance planning the babies postnatal care, routine prenatal care for the mothers was

provided and their wishes for labor and birth were discussed. In our institution, vaginal delivery is always performed unless contraindicated. After parental agreement, the option of induction of labor at a viable age was included in the plan which allowed parents to have a live-born child and allowed the designated obstetrician to be present during birth. In the two cases, labor was induced at 37 and 40 weeks of gestation, respectively. It was also agreed that fetal heart monitoring was considered to be stressful, so it was not included in the plan in both cases.

Setting for palliative care

The setting in which comfort care will be delivered should also be tackled: NICUs can often feel intimidating and overwhelming for families, who need maximum privacy and comfort. Thus, consideration should be given to the possibility of moving the mother to a single, quiet room, where she can be visited by family members and friends, if she desires [12, 13, 20, 21]. In the delivery room, instead of in a NICU, both Baby L. and Baby P.'s parents were able to have more privacy and to normalize their parenting experience up to the hilt.

Sometimes, parents wish to take their baby home. Planning for the possibility of home discharge should be considered, since some babies can outlive the immediate time after birth [12, 21, 26]. In these cases, both babies lived only for about two hours, and home discharge was not considered. This could have been contemplated however if the babies survive a period of more than 48 hours, after assessing their mothers' clinical condition, their parents' ability to cope and whether palliative care is accessible in their community and, in this situation, an individualized care plan would be prepared [27]. Home health care professionals should then also be part of the interdisciplinary team and home-based follow-up by pediatric palliative care professionals would be developed. In these cases, had the babies survive for more than 2 hours, the move to a single quiet room in the Maternal-Fetal Medicine Department was considered.

Decisions about interventions

Advance care planning should also include direction and decisions about medical interventions or limitation of interventions to be provided for

the baby. In both two cases, parents chose not to go through intensive resuscitation and limit procedures such as endotracheal intubation, positive pressure ventilation, cardiac massage and chemical resuscitation.

Following this decision, pain and symptom management were discussed. In order to deal with signs of respiratory distress and hypersialorrhea, the option of airway suction and administration of blow-by oxygen was accepted. The use of anticholinergic agents was not considered in this plan as the literature suggests a lack of empirical data [28]. The team also decided to administer morphine 0.1 mg/kg IM or SC, 2-4 h or midazolam (0.25 mg/kg intranasal/sublingual) if the newborns showed signs of discomfort, assessed according to the Echelle Douleur Inconfort Nouveau-né (EDIN pain scale) [29]. In fact, opioids such as morphine are the pharmacological agents most commonly used in the treatment of neonatal pain [30]. The use of IM or SC morphine might sometimes be distressing for the child so, in the event of a prolonged life, administration per os or the positioning of an umbilical venous catheter could be considered in our plans.

In the near future, intranasal fentanyl may be an option to be included in our plans as, compared to morphine, fentanyl is more potent and holds a more rapid onset of action [31]. There is a recognized risk of chest-wall stiffness, but the intranasal administration appears to be effective and safe [31, 32]. Also, respiratory depression is a well-documented side effect with morphine analgesia, but it has been described much less with fentanyl [31].

Intranasal midazolam offers effective control of seizures and benzodiazepines in conjunction with opioids provide the additional effect of alleviating anxiety and agitation [31]. Administering midazolam intranasally can sometimes cause burning thus other routes like the sublingual might be better tolerated.

In the end, none of the two babies received any type of medication. Curiously, in 2015, Garten et al., in consistency with other published studies, showed in a retrospective analysis of neonates who died in the delivery room that the frequency of comfort medication diverges in comparison with those who died in the NICU. Only 2 out of 113 neonates who died in the delivery room were given pharmacological treatment for symptom control [33, 34]. Possible explanations suggested by the authors is the arginine-vasopressin (via vasopressin receptor

1A) release during birth that could act as an effective pain-killer and avoiding invasive procedures thus making pharmacological treatment unnecessary. In addition, it may be difficult to obtain an intravenous line in the delivery room, which also contributes to a less frequent administration of drugs.

Other non-pharmacological measures of comfort should be contemplated and were included in Baby L. and Baby P.'s plan, such as skin-to-skin contact, wrapping and warming.

Nutrition

With regard to newborn nutrition, it is commonly accepted that feeding during end of life is an important measure of comfort [35]. However, parenteral routes of administration are associated with 15% complication rates [36]. With this in mind, parenteral feeding is rarely recommended [21, 22, 37]. Oral nutrition and breastfeeding can be comforting for the baby and should be maintained [22]. Whenever the newborn shows a desire to be fed, the amount of milk needed for its satisfaction should be offered, provided it is not a source of discomfort. The possibility of breastfeeding and the use of a small cup or syringe to deliver colostrum, milk, or sucrose should be included in the birth plan.

Parents of Baby L. chose to have breastfeeding as an option, although it was not performed, in contrast to the parents of the Baby P., who decided not to include this option, as it could be a source of discomfort.

Anticipatory guidance

Anticipatory guidance regarding physical changes when approaching the end of life, such as changes of breathing patterns (gaspings and long periods of apnea), skin color and temperature is crucial as it helps to prepare parents for the imminence of death and guide emotional support [38]. Both parents knew what to expect and were also warned about the uncertainty of duration of life.

Despite clearly written information, it is essential to constantly review the plan's focus on the best interest of the baby, since these babies can live longer than expected and clinical condition may vary over time [22].

End-of-life care

After Baby L. and Baby P. were born, quality time with their parents was promoted as they held

their baby alive and mementos were collected. Typically, parents feel that by doing so their baby's life is honored, cherished and remembered. Thus, they should be allowed to stay with the infant as long as they wish [39]. New technological developments have introduced a new concept: post-death neonatal cooling, which offers a greater opportunity for parents and family members to devote time to their baby [38]. However, such technology still raises ethical issues and is not yet available in our hospital.

Parents' cultural and spiritual needs also need to be tackled [12, 21, 22]. In both cases, parents wanted to hold a baptism ceremony: a hospital chaplain and a chaplain chosen by the parents were then introduced to the program. Nurses and care providers should investigate the parents' wishes and offer access to spiritual or religious services if desired.

Another key element of PNPC is to discuss what will happen after the baby's death. As the literature suggests, a postmortem examination can provide new information in 10% to 76% of the cases, allowing confirmation of the diagnosis and predicting the risk of recurrence and future pregnancy [12, 20-22, 40]. In both cases, autopsies were not performed. Since the definitive diagnosis was already defined by the result of the karyotype study, performing the autopsy would have little impact on the approach and future counseling of these parents. This decision was supported by a senior member of the team, with specific expertise in this area. It should be noted, however, that this decision must always be individualized.

Organ donation options were also not included in our plan as in both cases the babies were not eligible to become donors. Nevertheless, as the Internet has increased parental awareness on this topic and since literature intimates that 75% of organ donations in neonates are initiated after the parents' request, regional organ donation centers should be contacted to see if each individual qualifies as a likely donor [12, 38, 41].

In Portugal, the "Entidade de Verificação da Admissibilidade da Colheita para Transplante (EVA)" is responsible for issuing a binding opinion in case of donation of organs, tissues or cells for therapeutic or transplantation purposes. This is an organism created in each hospital where the collection takes place, and it works in the dependency and as a section of the Ethics Committee for Health of the hospital where the collection takes place.

It should be noted that the possibility of organ donation will involve hospitalization in the NICU and maintenance of advanced life support, which completely changes the type of care to be provided.

Follow-up care during bereavement

Bereavement is part of the entire process precipitated by the loss of the baby that, in case of fatal prenatal diagnosis, starts from the time of diagnosis and last until two years, needing support for a long time. Parents often experience feelings of anticipated loss, and a significant number of women and men report symptoms of depression and psychiatric distress [9, 39]. Baby L. and Baby P.'s parents were offered psychological support, but both parents declined it. It is important to note that coping with the loss of a baby is profoundly personal and will be experienced differently by each person [9]. Nonetheless, resources including regional and national parent support organizations contacts' should be introduced to parents and follow-up care should be established with periodic phone calls and a follow-up bereavement appointment [27]. Although both parents declined psychological help, a follow-up consultation was conducted 30 days after birth, which allowed evaluation of their psychological needs, their coping mechanisms and the impact on their daily lives. It was also assured, to all parents, that they had complete freedom to contact the team whenever needed.

Staff distress and education

Obstacles to palliative care include diagnostic and prognostic uncertainty, logistic obstacles related to interdisciplinary collaboration and lack of time to counsel parents [6, 42, 43]. Furthermore, health providers often highlight having insufficient knowledge regarding certain aspects of PNPC, being the lack of communication skills the most common source of distress [6, 33, 34].

Very often, even where a pathway on PNPC is available, it is not implemented. This aspect and the fact that there are still no guidelines impose the need for specialized groups of professionals on PNPC.

Staff training in communication and decision-making skills, ethical principles, birth plans, neonatal advance care planning and neonatal pain management is an essential step toward providing

compassionate care and support to families [44, 45]. Forty-one percent of the programs in the US have already reported having some sort of formal training [46]. As new candidate cases' arise, appropriate training opportunities to the professionals involved should be given, and regular debriefings should be stimulated.

Future investigations

In addition, to discuss the program to be implemented, it is necessary that the medical community dedicate interest in the study of its results. Several PNPC programs have been described, mainly in NICUs. However, the results of the implementation of such programs are still insufficient. In addition, quality evaluation parameters of existing programs are still scarce [46, 47]. An additional future direction should aim at the evaluation of the efficiency of our comfort care as well as at studying the impact on parent' satisfaction, given the importance they play.

According to Balaguer et al., "it seems desirable for obstetric and neonatal units to have available an active and efficient Perinatal Palliative Care programme" [10]. Considering the need for education in this area that health professionals from several countries report and the lack of documented programs in our country, it becomes clear the need to establish a consistent PNPC framework in Portugal. Besides, it is important to include obstetric residents, midwives, medical students and nursing students in the provision of palliative care so this model of care can be incorporated into future practices [6, 12, 15].

An existing list of PNPC programs can be found at www.perinatalhospice.com, as well as resources for both caregivers and parents.

Conclusion

PNPC asserts itself as a holistic and caring response to the needs of families affected by early loss. It integrates care focused on the best interests of the baby and the psychological and emotional needs of the parents.

Our knowledge on PNPC has been growing rapidly in the last 20 years. With this paper and the description of these two cases, we aim to foster interest in this area and reinforce the need for a well-structured and individualized plan, consistent among the different professionals involved.

Declaration of interest

The Authors declare that there is no conflict of interest.

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Appendix A. Perinatal palliative care program.**PERINATAL PALLIATIVE CARE PROGRAM
ADVANCE CARE PLANNING – BIRTH PLAN**

Baby's Name: _____

Mother's Name: _____

Father's Name: _____

Obstetrician: _____

Neonatologist: _____

Key-point person: _____

Diagnosis: _____

Goal of care: _____

LABOR AND DELIVERY

Due date: _____

Induction of labor: Yes No

Place of birth: _____

Mode of delivery: Vaginal Cesarean sectionWe want our baby's heartbeat to be monitored: Yes No

We would like these people in attendance: _____

 If conceivable, we would like a family member cut the umbilical cord.**NEWBORN CARE**

Location of care: _____

For our baby's comfort, please suspend routine procedures such as:

 Vitamin K injection, Antibiotic eye ointment.We would like all routine procedures necessary to be executed with our baby in our arms: Yes No

We understand the natural history of our baby's diagnosis and the prognosis associated with the condition. Taking that into account, if our baby is born alive:

 We wish to have all medical interventions to support our baby. We wish all medical interventions, except: _____ We want our baby to receive comfort care: Signs of respiratory distress (Cyanosis, Dyspnea...)/Terminal Secretions: Blow-by oxygen and aspiration of secretions. Pain/Discomfort: Morphine 0.1 mg/kg IM/SC, 2-4 h; Midazolam 0.1 mg/kg IM, 2-4 h or 0.25 mg/kg intranasal or 0.2 mg/kg sublingual.**NUTRITIONAL PLAN**Breastfeeding (if it does not increase the discomfort): Yes No**FAMILY TIME**

While holding our baby, we prefer our baby to be:

 Placed skin-to-skin, Wrapped.

We wish to:

 Dress our baby, Obtain footprints and photos of our baby. We would like to obtain keepsakes like ID bracelet, weight card, lock of hair, blanket and clothes. We would like family members/friends to join us after delivery to spend time with us and our baby.**SPIRITUAL SUPPORT**

We would like a religious ceremony, to be performed according to our beliefs:

 A representative from our faith community will perform the ritual. _____ (Name) We would like support from the hospital chaplain.**END-OF-LIFE CARE**We understand all physical changes that can occur when approaching the end of life, such as changes in breathing patterns, skin color and temperature. We would like to embrace our baby while dying and after death for as long as needed: Yes NoAutopsy: Yes NoGenetic testing: Yes No

Should our baby survive more than 48 hours, consider plans for taking our baby home:

The following changes were made to the plan after admission to the hospital:

Note: If considering organ donation, the infant needs to be hospitalized in the NICU and advanced life support must be maintained.