Care of a baby born with a cleft palate: A case study

Abstract

Orofacial clefts are the most common facial birth defect in humans. Their management is complex due to a range of immediate and ongoing difficulties. These include breathing, feeding, speech, hearing and dental problems. This article uses a case study approach to illustrate these difficulties for baby 'Esther' (a pseudonym) who was born with a cleft palate. Her case is presented followed by a discussion of the contemporary evidence-based care provided for Esther and her family. The piece focuses on family centred care and multi-disciplinary teamwork. The central role of the children's nurse is highlighted throughout.

Key words

- Cleft lip/palate
- Feeding difficulties
- Children's nurses role
- Family centred care
- Multi-disciplinary team

Key points

- Babies with clefts are often admitted to neonatal units due to co-morbidities such as breathing and feeding difficulties
- Parents/carers of babies with cleft lip and/or palate require a great deal of support, especially when diagnosed post-natally
- Management and treatment of a baby with a cleft lip and/or palate requires the input and support of a wide MDT
- Prone or lateral placement of babies with clefts while sleeping is recommended to ease breathing difficulties, but there is a lack of conclusive evidence on which position is best
- Hydration and electrolyte balance are key to effective and safe management of babies with clefts
- Long-term, ongoing support from SALT and cleft lip/palate Clinical Nurse Specialist are vital

- Family centred care and empowerment from the outset are essential to enable parents/carers to look after their baby after discharge, whilst awaiting surgical repair and beyond.

Introduction

A cleft palate is a congenital abnormality that results from the failure of fusion of the palatine processes of the maxilla bone and the nasal septum in the uterus, resulting in the palate containing an opening into the nose (Lissauer and Carroll 2018). Clefting occurs in two potential patterns: isolated soft tissue cleft palate which affects the back of the palate, or a combined soft and hard palate cleft (Bishop and Ebach 2019). In 70% of cases, it is seen in association with a cleft lip. Clefts affect approximately 1 in 700 babies, a prevalence of 1.2 in every 1000 live births, making orofacial clefts the most common facial birth defect in humans (Rahimov et al 2012, Kati 2018). Clefts have a strong genetic component, and children with affected first-degree relatives are most at risk, although there is no single gene inheritance, (Rudolf et al 2011). Surgical closure of the palate usually takes place between 6-12 months of age (Kliegman and Marcdante 2019). Due to the complex nature of the condition, babies with cleft palate may initially face difficulties in establishing milk feeding. Muhammad et al (2023) identified a high incidence of aspiration pneumonia (64.53%) in babies with clefts. Breathing difficulties, speech, hearing and dental problems are also common (Leslie and Marazita 2013).

This case study aims to give an overview of care for Esther (pseudonym) who was born with a cleft palate. A description of Esther's condition and difficulties are presented. This is followed by a discussion of the evidence-based care provided by the children's nurse and other members of the team. The discussion focuses on Family centred care (FCC) and multidisciplinary team (MDT) working.

Esther

Esther was a 3-week-old infant, born at 38 weeks and 2 days gestation via an emergency C-Section. Esther had no respiratory effort at birth so inflation breaths and positive end-expiratory pressure (PEEP) were applied until her oxygen saturation level was above 95% and her tone and colour improved. After 15 minutes, Esther was alert, active, and crying. At seven hours of life, Esther was

examined due to feeding concerns. She was having episodes of nasal regurgitation, aponea and was unable to breastfeed. Following a physical examination of her nose and mouth, Esther was discovered to have a U-shaped unilateral midline posterior hard and soft cleft palate, which is visible to inspection using a torch and depressing the tongue (Bennett and Meier 2019). Esther's upper lip and gums were intact.

Esther was transferred from the maternity unit to the Neonatal Intensive Care Unit (NICU) for monitoring and management of nasal regurgitation, aponeas and feeding difficulties. Following a three-week stay in NICU Esther was admitted to a general medical paediatric ward for further monitoring of her feeding and respiration. This included changing from nasogastric (NG) feeds to bottle feeds and having a nasopharyngeal airway (NPA) inserted. Esthers' healthcare needs were complex and ongoing. The MDT consisted of nurses, consultants, Ear, Nose and Throat (ENT) surgeons, Speech and Language Therapists (SALT), dieticians, Clinical Nurse Specialists (CNS), and orthodontists. These professionals needed to work together in a coordinated fashion to effectively evaluate and treat the difficulties associated with cleft palate, and prevent complications associated with the disorder (Losee and Kirschner 2016).

Once discharged into the community Esther would continue to be under the care of the ENT consultant, community nurses, dietitians, and SALT. She would have speech assessments with the community paediatric SALT team throughout childhood (Vallino 2017) and additional speech therapy if required. A community, outreach, cleft lip and palate CNS would visit regularly (Beaumont 2017, Searle et al 2018, Martin et al 2020) for continued support and feeding management. This would ensure close, continuous monitoring of her condition, provide support for Esther and her parents; particularly during the period between discharge and corrective surgery, which is typically 6-12 months for a cleft palate (Cleft Lip and Palate Association (CLAPA) 2023) and allow problems to be dealt with in a timely manner (Bannister 2017).

The discussion section will centre on the aspects of Esther's nursing care that were of most immediate concern. This includes the insertion of the NPA to aid Esther's breathing and how the MDT worked to address Ether's feeding difficulties. FCC and MDT working are key aspects of this case study as FCC needs particularly sensitive

management in situations where clefts are diagnosed postnatally, and significant MDT support is required (McElroy et al 2017).

Case Discussion

On admission to the general medical ward Esther's vital signs observations were carried out hourly. Esther was initially self-ventilating in air when she arrived from NICU, and as such her oxygen saturation levels were continuously monitored with specific attention to respiratory effort, noting any apnoea's and desaturations. This was documented in line with the guidance given on the Paediatric Early Warning Score (PEWS) Charts, an assessment tool used to detect any early signs of deterioration and initiate a quicker response (Royal College of Nursing (RCN) 2021). Esther was receiving intravenous fluid therapy, so her input and output was monitored on a fluid balance chart, in line with the relevant National Institute for Health and Care Excellence standards (NICE 2016).

Esther was nursed in a prone position when she was sleeping only. When awake she could lie in other positions and be cared for/ cuddled by her parents in the same way as a baby without a cleft palate. Lying in a prone position is a vital aspect of caring for babies with a cleft palate as the defect causes a decreased length and height of the maxilla and mandible bones, resulting in reduction in the size of the airway (Abdel-Aziz 2012). These craniofacial abnormalities create an increased risk of airway problems, such as upper airway obstruction, sleep-related breathing difficulties, including obstructive sleep aponea, and nasal airway impairment (Murray et al 2022). When Esther was first admitted to the ward, she would experience regular oxygen desaturations and aponea, lasting up to 5 seconds, during sleep. It was crucial that nursing staff were alerted to these by the monitor and would record them for the ENT consultants to review alongside her hourly vital sign observations. Supplemental oxygen therapy would be administered if the desaturation or aponea become prolonged (Chandrasekar et al 2022). This helped Esther to maintain her oxygen saturations target of between 91-95%, reducing the risk of morbidity and invasive ventilation (NICE 2020). Furthermore, it was Esther's nurses', paramount responsibility to ensure that everyone on the ward was aware that Esther's sleep position was prone rather than supine (on her back), allowing the tongue and

mandible to fall forward (Lissauer and Carroll 2018). The experience of Esther being nursed prone is in keeping with a quantitative study by Kukkola and Kirjavainen (2022) which concluded that in infants with structural facial or airway abnormalities, such as a cleft palate, episodes of airway obstructions were less frequent when sleeping in the prone position compared with the supine position. Conversely, the findings of a quantitative study by Greenlee et al (2018) found a lack of consistent improvement in oxygenation in the prone/non-supine position. They acknowledge that the prone position may mask breathing difficulties, and suggested caution when recommending this positioning to treat sleep-disordered breathing. Therefore, there is a lack of definitive evidence about the effectiveness of the prone sleep position in infants with cleft palate, and further research is needed to determine the most appropriate sleeping position for an infant with cleft palate to minimise the risk of obstructive sleep aponea (Davies et al 2017).

Following a medical review by the ENT consultants, it was determined that Esther's desaturations and apnoea were so persistent that she would require an airway adjunct. In Esther's case, it was concluded by her MDT that a nasopharyngeal airway (NPA) would be the most appropriate, as her airway obstructions were related to her glossoptosis - the position and displacement of her tongue. An NPA is a non-surgical method of airway management that involves the placement of a hollow plastic endotracheal tube into one nasal passage with the tip entering the distal oropharynx in the throat. This bypasses the upper airway obstructions and prevents the tongue from falling backwards on the pharyngeal wall, thus securing an open airway (Oh et al 2019), which was an important contributing factor in facilitating Esthers safe discharge home. The NPA was inserted under a general anaesthetic in the children's operating theatres, by the ENT surgeons. This was the safest method of insertion for Esther, as her craniofacial condition meant that her nasal passages were narrower than usual (GOSH NHS 2019), therefore her parents were happy to consent to the procedure. Esther's surgery was successful, and her airway difficulties were relieved immediately after the insertion of the NPA. Once back on the ward, it was the nursing team's responsibility to suction and maintain the patency of the tube, and to continuously assess the integrity and position of the NPA (Parhizkar et al 2016). It was fundamental that suction was carried out using the appropriately sized suction catheter for the size of the NPA (3.5mm in Esther's case

- therefore the correct suction catheter was a size 8). Suctioning was required before each feed, when mucus in her nostrils was seen, and whenever any breathing difficulties or increased respiratory effort was noted. This keeps the NPA clean and patent to allow Ester to breathe easily (Chartered Society of Physiotherapy (CSP) 2015). Furthermore, another essential aspect of NPA nursing care is ensuring that it remains secure and safely in situ. Nursing staff checked the position of the NPA and performed a comprehensive integrity assessment of the skin surrounding the NPA every hour following her surgery. The tube position and tapes were changed if any blanching or discolouration of the skin was noted (NICE 2015).

During her time on the medical ward, Esther encountered severe oral feeding difficulties. She was experiencing regular nasal regurgitation and choking episodes. This was causing excessive air intake, and she was struggling to gain weight. Early sucking and swallowing difficulties are associated with structural abnormalities, which can be a barrier to adequate nutrition in infants with a cleft palate (Goswami et al 2016). Esther initially remained in receipt of intravenous fluid therapy of 80ml/kg/day, in line with her weight and daily maintenance fluid requirement, as per NICE guidance (2016). Children with respiratory distress, like Esther, should receive appropriate IV fluid management. In Esthers case she was breathing so irregularly that she could not suck, swallow, and breathe at the same time. Hence, she was not receiving sufficient oral fluid intake and her electrolyte balance requirements were significantly compromised (Ingelse et al 2016). IV fluids kept Esther hydrated and maintained healthy blood glucose levels.

The SALT team reviewed Esther on the ward to determine whether she had a safe swallow. This is the ability to transfer food and fluids from the mouth to the stomach into the lower airways without aspiration (Royal College of Speech and Language Therapists (RCSLT) 2019). SALT carried out a bedside assessment, which involved listening and watching Esther swallow, in line with the Eating, Drinking and Swallowing Competency Framework (RCSLT 2020). It was confirmed that Esther was swallowing safely, and it was decided by her MDT that it would be appropriate to introduce bottle feeds in place of IV fluids.

Due to air leaks from the mouth through the cleft and into the nose, babies with a cleft palate are unable to create enough suction to draw milk from a bottle (Devi et al

2012). These problems can be solved by using teats and bottles specially designed for babies with a cleft palate (Lissauer and Carroll 2018). When Esther was introduced to bottle feeds, a soft orthodontic teat with a small slit in the side was used. This creates a vent to help prevent the infant from swallowing air when feeding, and its flexible nature is intended to block the continuity of the oral cavity and the nasal cavity (CLAPA 2022). Furthermore, a soft, squeezable plastic slow-flow bottle was used to meet Esther's specific care needs. Through manually squeezing these bottles, the milk flow is mechanically created by generating negative pressure. This was required for Esther to create the necessary suction to draw up milk and swallow without much effort, overcoming the difficulties caused by her cleft (Jindal and Khan 2013). This specialised type of bottle provides control of the milk volume, so Esther's MDT could set a flow rate they knew she could manage without risk of aspiration, and then stop the flow of milk when Esther paused to swallow and breathe. As the nursing staff would feed Esther when her family were not on the ward, it was their responsibility to check that the correct teats and bottles were used and to administer her feeds safely in the correct position with the correct technique. It is recommended that babies with a cleft palate are fed in a semi-upright position, greater than 60°. This position facilitates the transfer of fluids into the stomach via gravity, limits nasal regurgitation and decreases the tendency of nasopharyngeal reflux (Burca et al 2016). In addition, babies with a cleft palate often experience excessive air intake when feeding, and therefore require frequent 'winding' during and after feeds to decrease the amount of air in the stomach (Wijekoon et al 2019). When Esther was transitioned from IV fluids to bottle feeds, it was crucial that the nursing staff continued to accurately document Esther's fluid intake and outtake. This ensured she maintained a correct fluid and electrolyte balance and reduced the risk of adverse events, such as dehydration (NICE 2016).

The nurse demonstrated effective family centred care (FCC) throughout by ensuring all of Esther's family's questions were answered and their needs and preferences were responded to. There is psychological stress and anxiety for parents when making decisions regarding a child with a cleft palate's feeding. This is due to the risk of weight loss and nutritional insufficiency associated with this complex condition, which can lead to poor mother and infant bonding (Hiremath 2016). The nurse ensured that Esther's family developed a method of feeding which suited

them, and she encouraged and empowered them to collaborate and share in the MDT decision about Esther's treatment and care, as outlined in the NMC Code (2018). Breast milk provides the best nutrition for a young baby (World Health Organisation (WHO) 2017), and as such Esther's mother communicated her preference to bottle feed Esther with expressed breast milk (EBM), rather than formula, to support Esther's growth and development. It is widely recognised that FCC in children and young people's (CYP) nursing is integral as it leads to better health outcomes and greater patient and family satisfaction (Glasper et al 2014). Furthermore, a systematic review by Allen et al (2018) found that for children with complex continuing needs, this partnership approach to care and family empowerment and education is essential in supporting families to manage their childs' condition at home (Brimble 2021). They further reported that by encouraging collaborative and effective communication, CYP nurses can support families in establishing themselves as part of the multidisciplinary healthcare team, rather than simply the passive subjects of medical interventions (Allen 2018). Thus, it was important that the nursing staff regularly encouraged Esther's mother to express her breast milk and continue to hold Esther to the breast when feeding to preserve the mother-infant bond, support FCC practice, and ensure effective feeding.

Conclusion

Clefts are the most common facial birth defect in humans. This case study, which focused on managing respiratory distress, NPA insertion and monitoring fluid balance for a child with cleft palate, highlighted the importance of a collaborative MDT approach. It was clear that while the nurse played a fundamental role in Esther's treatment, she invariably worked in close partnership with her family and the MDT, to facilitate a holistic and individualised approach. The nurse respected and supported Esther's family's decision to use EBM to feed her, communicating effectively with other members of the MDT, and encouraging parent-infant attachment, to help reduce the stress and anxiety surrounding this complex conditions. Other elements which contributed to effective FCC were education of the family, being responsive to their needs and encouraging them to participate in MDT discussions and decisions; thus empowering them within the hospital setting and in the future management of Esthers condition at home.

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