Feeding, growth, and nutrition in children with congenitally malformed hearts

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Abstract In the United States of America, approximately 40,000 infants are born annually with congenitally malformed hearts. Children with defects that require complex surgical palliation, or definitive repair, face many challenges in achieving optimal short-term and long-term growth. The presence of associated chromosomal abnormalities, cyanosis, and cardiac failure adds to the complexity and challenge. In this review, we address three themes related to feeding, growth, and nutrition of infants after neonatal cardiac surgery: nutritional challenges after chylothorax; breastfeeding after surgery; and the challenges of feeding after discharge. Chylothorax is a rare complication following cardiothoracic surgery in children. Children with chylothorax have nutritional depletion secondary to protein losses in chylous fluid, hypovolaemia, and electrolyte losses. In spite of the evidence supporting the use of human milk and breastfeeding in preterm infants, barriers to its use appear to persist in infants with critical cardiac disease. Yet, human milk is the preferred form of nutrition for well, preterm, or ill infants. It is well documented that after complex neonatal cardiac surgery medical teams and families struggle with infant feeding problems. Parents have described feeding their children as difficult, time consuming, and anxiety producing. Medical complications such as chylothorax, limited access to human milk, and parental concerns and stress about feeding are but three of the myriad of factors that may contribute to poor outcomes regarding nutrition and growth. Compelling evidence exists that this multi-factorial problem must be addressed with both physiological and behavioural strategies.

Keywords: Infant; congenital cardiac disease; feeding; human milk; chylothorax

In the United States of America, approximately 40,000 infants are born annually with congenitally malformed hearts.1 Children with defects that require complex surgical palliation, or definitive repair, face many challenges to achieve optimal short-term and long-term growth. The presence of associated chromosomal abnormalities, cyanosis, and cardiac failure adds to the complexity and challenge. Optimal nutrition increases metabolic reserves of such children, albeit that the robust metabolic demand and stress from cardiac surgery can potentially deplete their limited metabolic reserves. As such, the potential for perioperative morbidity increases. Decrease in perioperative metabolic stress, while delivering optimal nutrition early in the perioperative course, is key to improving appropriate weight gain. Achievement of optimal nutrition may be a modifiable variable that can positively impact outcome. In this review, we address several themes related to feeding, growth, and nutrition of infants after neonatal cardiac surgery, including nutritional challenges after chylothorax, breastfeeding after surgery, and the challenges of feeding after discharge.

Nutritional implications of chylothorax

Chylothorax is a rare complication following cardiothoracic surgery in children, with an estimated incidence in recently reported series ranging from 0.85% to 3.8%.5, 4 There are three main aetiologies of...
post-operative chylothorax. First, this complication can occur secondary to direct injury to the thoracic duct following procedures that occur in the vicinity of the thoracic duct, such as repair of coarctation of the aorta, ligation of a patent arterial duct, or placement of extracardiac systemic-to-pulmonary arterial shunts. Second, chylothorax can occur as a result of thrombotic occlusion of the superior caval vein. Third, it can occur in settings of high central venous pressures, such as following operations involving cavo-pulmonary anastomosis for the palliation of hypoplastic left cardiac syndrome. The most common procedures associated with post-operative chylothorax include complete repair of the tetralogy of Fallot, the Fontan operation, the Glenn operation, and orthotopic cardiac transplantation.\textsuperscript{2-3}

The consequences of chylothorax are manifold. Patients with chylothorax have nutritional depletion secondary to protein losses in chylous fluid, hypovolaemia, and electrolyte losses including loss of sodium leading to hyponatraemia, loss of sodium bicarbonate leading to acidosis, and loss of calcium leading to hypocalcaemia. Immunodeficiency occurs secondary to the depletion of lymphocytes and hypogammaglobulinaemia.\textsuperscript{4,5}

Management

Conservative strategies for management include drainage of fluid with tube thoracostomy, institution of diets enriched with medium-chain triglycerides, or enteric rest with total parenteral nutrition. The enriched diets contain medium-chain triglycerides that bypass the lymphatics, and enter directly into portal circulation.\textsuperscript{6,7} Recent reports have suggested the use of skimmed breast milk for enteral nutrition for infants with chylothorax.\textsuperscript{10} The process of creating skimmed milk includes centrifugation of breast milk cooled to 2\textdegree C, with the removal of the solid fat layer formed on top of the milk, and analysis of the lower liquid portion.\textsuperscript{11} The benefit of the use of breast milk includes the immunological properties that formula milk cannot provide. Enteric rest and total parenteral nutrition are recommended for high-volume chylothorax that does not decrease with medium-chain triglyceride feeds. The goal of parenteral nutrition is not to only achieve a positive nitrogen balance, but to also provide electrolyte replacement for ongoing losses in chylous fluid. Adjuvant therapy includes reduction of central venous pressures with diuretics, albumin supplementation, electrolyte replacement including sodium, calcium, and bicarbonate, immunoglobulin replacement, anti-thrombin III, and fresh frozen plasma replacement. Given the increased incidence of central venous thrombosis, we should consider the prophylactic anti-coagulation with heparin or enoxaparin. Success with corticosteroids has been reported following the Fontan operation.\textsuperscript{12}

Nutritional implications

Initial nutritional therapy includes medium-chain-enriched feeds for patients with high-volume chylothorax. Enteral rest and initiation of total parenteral nutrition should be considered. If this approach fails, we recommend a trial of octreotide followed by ligation of the thoracic duct. Another new and upcoming practice is the use of skimmed breast milk in infants, which has the added benefit of maternal immunoglobulins.

Breastfeeding after neonatal cardiac surgery

Human milk is the preferred form of nutrition for well, preterm, or ill infants.\textsuperscript{13} In preterm infants admitted to hospital, human milk is reported to decrease the incidence and severity of both nosocomial infections and necrotising enterocolitis,\textsuperscript{14} as well as the incidence and severity of diarrhoea and bacteremia.\textsuperscript{15} In a study of 119 singleton infants born with very low weight,\textsuperscript{16} we noted a protective dose-response relationship between human milk and the prevention of sepsis.

In spite of the evidence supporting the use of human milk and breastfeeding in preterm infants, barriers to its use appear to persist in infants with critical cardiac disease. For the infant with complex cardiac disease, who is at risk for long-term feeding problems and growth failure, human milk might possibly reduce the incidence of feeding intolerance secondary to issues related to the gut. Colostrum has been shown to be highly beneficial to preterm and ill infants, priming the gut with proteins, amino acids, and secretory immunoglobulin A.\textsuperscript{17} Although it is not always possible to put the critically ill infant to the breast, offering colostrum as the first feed mimics the natural feeding process.\textsuperscript{18} At the Children's Hospital of Philadelphia, our lactation nurse scientist conducted an assessment of the use of human milk in infants less than 6 months of age.\textsuperscript{19} This showed that less than half of the eligible infants across the centre were matched with stored human milk. This finding was consistent with what was perceived by the nursing staff as well as the nurse scientist. A retrospective case series in infants admitted for a bidirectional Glenn procedure\textsuperscript{20} showed that one quarter were receiving breast milk at neonatal discharge, but less than one-tenth received breast milk at the time of the bidirectional Glenn procedure, far below the standard set by the American Academy of Pediatrics for the recommended duration of time for infants to be fed breast milk.

Even after an infant is stable, and nipple feedings have begun, there continues to be the concern that putting an infant to the breast will increase the work of feeding. A study using continuously monitored values for the saturation of oxygen during breast and
bottle-feeding revealed a significant interaction between the method of feeding and the changes in saturation of oxygen.\textsuperscript{21} saturations were lowest during bottle-feeding, during sucking, and continued a sustained downward trend. Other investigations continuously measuring saturations of oxygen during breast and bottle-feeding in preterm babies also showed saturations to be significantly higher in breast-fed infants.\textsuperscript{22,23} breast-fed infants do not experience periods of oxygen desaturation,\textsuperscript{24} whereas still others have shown lower incidences of desaturation during breastfeeding when compared to bottle-feeding infants.\textsuperscript{25} it has also been reported that temperatures remain stable during breastfeeding.\textsuperscript{21,22}

A second concern commonly cited by practitioners is the inability to assess intake accurately. Test weighing, however, has been shown to be accurate when performed with an electronic scale using the babyweigh scale by medela to perform test weights.\textsuperscript{18} the accuracy of the babyweigh scale has also been validated in an earlier study.\textsuperscript{26} the infant is weighed twice to ensure accuracy, the weight is locked into the scale, and the scale remains on during the breastfeeding session. once the session is completed, the infant is placed back on the scale with all items of clothing on, including the diaper. the weight subsequent to feeding is subtracted from the earlier measurement, permitting reporting of the milk transfer in grams.\textsuperscript{18}

although the exact aetiology of growth failure in critically ill cardiac infants is unclear, no one has investigated the role of human milk use in this population. previous concerns related to oxygen saturation and inability to access intake accurately should not be a barrier to the use of human milk.

challenges in feeding: transitioning from the hospital to the home. what we have learned from parental experiences?

Feeding problems after complex neonatal cardiac surgery are well documented, but not well understood.\textsuperscript{26} Some infants refuse to suck, whereas others suck for only a short time. Swallowing, which should be present in the foetus by 16 weeks,\textsuperscript{27} is expected to be normally functioning at birth in normal healthy babies. Laryngopharyngeal dysfunction, however, has been reported in up to half of those healthy babies undergoing the norwood procedure, inevitably leading to difficulties in feeding.\textsuperscript{28} Other risk factors for poor feeding include immaturity of the central nervous system,\textsuperscript{29} or injury such as a stroke.\textsuperscript{28} Weight gain is often slow, creating frustration for both parents and caregivers.\textsuperscript{31} Despite valiant efforts of many cardiac centres, no single guideline has emerged as the standard for the introduction of oral nutrition. Cardiac centres vary on which infant is a candidate for continuous versus bolus nasogastric feedings, who is ready for nipple feedings, and when is the appropriate time to begin the discussion about insertion of a gastric tube. Many cardiac centres send infants home on nasogastric tube feeding, either as the sole means of nutritional intake, or in combination with oral feedings, whereas other programmes maintain that if nutritional support is required, then a surgically placed gastric tube is the safest means of delivering nutrients.

All these decisions have an impact on family functioning. Caregivers of children with congenitally malformed hearts feel significantly more stress than caregivers of children with other illnesses, and they identify feeding as one of the key contributors to stress.\textsuperscript{32} parents have described feeding their children as difficult, time consuming, and anxiety producing.\textsuperscript{33} from our own work, both a longitudinal study of infants with functionally univentricular and biventricular circulations, and focus groups, we found that parents reported stress related to feeding to be escalated once there was the suggestion that their infant may need to go home with a nasogastric or gastric tube in place. Given that more than half of the infants with functionally univentricular circulations, and two-thirds of those with biventricular physiology, went home with some feeding device support,\textsuperscript{34} pre-discharge feeding stress was present for a large number of neonates and their families. The parents making up our focus groups described their ambivalence about being responsible for nasogastric or gastric tube feedings. One parent stated, "we had to go to classes on nasogastric feeding, but i was hoping she would be a good eater and we would not have to go that nasogastric route". She went on to say, "we had training in nasogastric feeding ... it was hard, i was scared". For another parent, the concern was related to the plan to insert a feeding tube surgically. She commented, "when we found out, it was very upsetting, he had a very difficult time with the open heart surgery and we were very traumatized ... we never got to the point of thinking about the gastric tube". Learning to accept long-term use of non-oral feeding was a challenge to many mothers. one said, "... physically you don't want to do that to your child, it's hard to feed them that way, but when it's something you have to do ... ". For another mother, inserting a nasogastric tube, or caring for the gastric tube, was something she could not bring herself to do, and the task became the responsibility of the father at night.

Parents identified several themes around the feeding behaviours of their infants, which included frequent vomiting, very long feedings, no hunger
cues, food rejection, oral aversive behaviours, and the removal of the tube by the infant. One mother stated, “Things went well for a while... she worked up to taking 2 ounces by bottle and then whatever she did not take we put down the tube. But then she started vomiting a lot, wasn’t gaining weight, so we had to go back to the hospital for 2 weeks”. The mother goes on to say that, by the time they left the hospital, the child was no longer taking anything by bottle, which caused further anxiety and stress. Over and over again, we heard from mothers that any day their child did not vomit was considered a good day. Other challenges were the synchronisation between medications and times of feeding.

Parents tried various strategies to increase intake of nutrients. One household set alarms every 3 hours around the clock for feedings. Some parents describe letting their infants go for a full hour for some feedings, even though they knew it was not recommended. A few mothers described feeding the infants in an upright position, using frequent feedings with less volume, to achieve increased intake and less vomiting. Even for mothers with infants who were currently nipple-feeding, many of the infants went home with some nasogastric tube support. These mothers talked about feeling traumatised by the initial experience with nasogastric feedings. One mother stated that she would do whatever it takes to prevent her infant from needing to return to a nasogastric tube.

In summary, poor nutritional states resulting from inadequate feeding ability are of great concern, and the source of ongoing stress, for parents of infants who have experienced neonatal surgery for complex congenital cardiac disease. Early planning before discharge, and frequent opportunities to practice, may allay some of the fear surrounding either nasogastric or gastric tube feedings at home.

Conclusions

Medical complications such as chylothorax, limited access to human milk, and parental concerns and stress about feeding are but three of the myriad of factors that may contribute to poor outcomes concerning nutrition and growth. There is compelling evidence that this multi-factorial problem must be addressed with both physiological and behavioural strategies.

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References