Nutrition in neonatal congenital heart disease

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Abstract: There are 40,000 infants born in the USA with congenital heart disease annually. Achievement of adequate oral nutrition is difficult in this population. Malnutrition is common. Single ventricle physiology, the risk of necrotizing enterocolitis, and cardiopulmonary bypass prevent the establishment of normal oral feeding patterns. Improved nutrition results in improved surgical outcomes, lower mortality, and shorter hospital stay. In this review, we discuss the challenges this population faces.

Keywords: necrotizing enterocolitis, malnutrition, growth failure, hypoplastic left heart

Introduction

Approximately 40,000 infants are born in the USA annually with congenital heart disease.¹ The postoperative mortality rate for neonates with congenital heart disease who undergo cardiac surgery has reduced significantly in the last number of years.² Neonates with congenital heart disease face considerable challenges when it comes to achieving optimal growth rates and the ability to orally feed. In addition, it has been suggested that the ability to orally feed may be a marker of future neurodevelopmental outcomes.³ Neonates face challenges both preoperatively and postoperatively in achieving adequate enteral nutrition that lead to problems with growth, wound healing, and immune function. Infants with congenital heart disease are known to be at risk of growth failure, with up to 40% of neonates being undernourished prior to cardiac surgery.⁴ Infants who feed less orally have a longer inpatient hospital stay and nearly half of infants with congenital heart disease are discharged home with feeding tubes following cardiac surgery.⁵ Postoperative complications such as chylothorax and vocal cord palsy add considerable morbidity to this population. Controversy remains regarding the practice of preoperative enteral feeding in prostaglandin E1-dependent lesions; clinicians cite the rare but potentially catastrophic complication of necrotizing enterocolitis (NEC) the main reason for this. However, there is considerable variation in practice between the USA and Europe and indeed between different institutions, with Europe tending to adopt a more liberal approach to enteral feeding in the pre- and immediately postoperative settings.

In this review, we examine the metabolic stress of cardiothoracic surgery and cardiopulmonary bypass, the controversy surrounding the role of enteral feeding in the ductal-dependent congenital heart lesions, and the nutritional consequence of the potential complications following cardiothoracic surgery.
Metabolic requirements in congenital heart disease

Failure to thrive or growth failure is a common finding in the congenital heart disease population. Approximately 6% of infants with congenital heart disease are growth retarded in utero. Increased metabolic demand combined with reduced energy intake leads to an imbalance of energy. Weight tends to be affected more than height; however, at 2 years of age, almost half of children with congenital heart disease are stunted. There are a large number of factors affecting growth in this population, including the type of congenital heart disease, disturbance in energy metabolism, decreased energy intake, disturbance in gastrointestinal function and the presence of chromosomal disorders such as trisomy 21. An infant with acyanotic heart disease with a large left to right shunt (patent ductus arteriosus, ventricular septal defect, atrial septal defect) in the presence of elevated pulmonary arterial pressures is at high risk of severe growth failure. The same applies to cyanotic heart lesions, where growth failure is common and is found in up to 70% of patients in that population. Decreased levels of insulin-like growth factor-1 (IGF-1) with elevated levels of growth hormone are also seen in these patients, further contributing to malnutrition in this population. Interestingly, the leptin regulating axis is normal in this patient group, suggesting that leptin does not play a role in growth failure. There is no general consensus on the calorific requirements of an infant with congenital heart disease; however, gross energy intake of up to 140 kcal/kg/day has been suggested by some. Nevertheless, achieving this nutritional requirement is only realistically possible with the help of nasogastric feeding. Nasogastric feeding tubes are ideal for short-term aids to enteral feeding, as they can be placed easily by nursing staff and parents and provide physiologic feeding into the stomach, but they are not without their disadvantages. They may exacerbate respiratory distress by occluding a nare, contribute to gastroesophageal reflux, and have the potential for misplacement into the bronchus, leading to a devastating chemical pneumonitis.

Many studies have looked at total daily energy expenditure (TDEE) in infants with congenital heart disease using the doubly labeled water method. TDEE is significantly greater in this population when compared with that of healthy infants. Healthy infants would be expected to have a mean TDEE of 280 kJ/kg/day at 3 months of age. In infants with congenital heart disease, the TDEE is increased, with a mean TDEE of 341 kJ/kg/day. Basal metabolic rate (BMR) in infants is almost twice that of adults, reflecting the higher proportion of body weight that vital organs such as the brain, liver, and heart contribute to overall weight. Similarly to TDEE, infants with congenital heart disease have a higher BMR than healthy infants. Hypertrophied myocardium, increased sympathetic nervous system activity, increased respiratory effort, and increased risk of infection contribute to a higher BMR. Resting energy expenditure (REE) is also increased in infants with congenital heart disease when compared with healthy infants. In addition, infants with congenital heart disease who are undernourished have a higher REE than well-nourished infants with congenital heart disease. REE does not seem to be affected by the presence or absence of cyanosis.

The metabolic response to cardiac surgery

When compared with children and adults, neonates experience a greater metabolic response to cardiopulmonary bypass. Infants with cyanotic heart disease requiring cardiopulmonary bypass experience longer inpatient stay, delay in first feed, delay in gavage feeding, and maximal nipple feeds compared with the non-bypass infants. In addition, neonates have less metabolic reserves compared with older children, and, as a result, this leaves the neonate with congenital heart disease at greater risk of growth failure, poor wound healing, and prolonged inpatient stay. Once short-term glucose stores are depleted in the immediate postoperative period, skeletal muscle breakdown is employed to sustain a prolonged catabolic state. Usually, by 72 hours following surgery, anabolic metabolism returns; however, this change in metabolism is delayed in those infants who have cyanotic heart lesions. REE returns to preoperative levels by day 5 following surgery. Optimal postoperative control of pain aids reduces the degree and duration of catabolic metabolism. Enteral nutrition should be introduced as soon as it is deemed medically safe to do so in the immediate postoperative phase. This reduces the risk of gut atrophy and prevents the loss of the intestinal mucosal barrier. Enteral nutrition in the intensive care unit (ICU) setting is often preferentially given via a nasogastric tube.

Duct-dependent lesions

Perhaps the most controversial aspect of feeding practice in neonates with congenital heart disease is whether to allow enteral feeding in neonates with duct-dependent circulation on prostaglandin E1 infusions, especially in the setting of hypoplastic left heart syndrome. The fear of hemodynamic instability, gut hypoperfusion, and subsequent
development of NEC leaves many pediatric cardiologists and neonatologists reluctant to commence enteral feeds in the preoperative setting.

The occurrence of NEC is a complex multifactorial process that is yet to be fully understood. It is a rare yet potentially catastrophic complication of the management of congenital heart disease. There are, however, some important differences between the cohort of preterm infants that develop NEC and the cohort of term infants in the setting of congenital heart disease. First, NEC occurs only rarely before the introduction of enteral feeding in the preterm population. In contrast, Iannucci et al reported that up to 27% of infants studied with congenital heart disease developed NEC before the introduction of enteral feeds. Secondly, complications from NEC such as stricture formation are less common in the congenital heart disease population compared with in the preterm infant population.

The metabolic demands of open cardiac surgery and cardiopulmonary bypass in the presence of single ventricle physiology lead to a higher prevalence of NEC in infants with congenital heart disease. The prevalence of NEC is ten to 100 times higher in term infants with congenital heart disease than in term infants without congenital heart disease. Factors associated with an elevated risk of NEC in infants with heart disease include premature birth, hypoplastic left heart syndrome, truncus arteriosus, aortopulmonary window, and episodes of poor systemic perfusion or shock.

Howley et al surveyed the feeding practices of over 700 neonatologists and pediatric cardiologists. There was a stark variation in practice when Europe was compared with the USA. Routine preoperative enteral feeding in prostaglandin E1-dependent lesions was reported by 93% of clinicians outside the USA, whereas only 56% of US-based clinicians reported the same. Intestinal hypoperfusion and the presence of a right to left or bidirectional shunt being the main reason cited on both sides of the Atlantic when deciding whether to use enteral feeds. Over two-thirds of practitioners were willing to feed with an umbilical venous catheter in place.

Reported mortality rates in preterm infants with NEC are similar to those in infants with congenital heart disease, ranging from 15% to 30%.

The presence of umbilical venous or arterial catheters has not been shown to increase the risk of NEC in the congenital heart disease population.

We would advise caution in any enteral feeding in children with prostaglandin-dependent lesions where ductal flow is entirely right to left. We support the development of feeding protocols in the immediate perioperative period to assist with the early introduction of enteral feeding in the ICU setting. In the setting of low cardiac output state or shock, enteral feeding should be withheld to reduce the risk of NEC.

**Chylothorax**

Chylothorax is accumulation of chyle in the pleural space. It can occur following cardiac surgery and is commoner in the pediatric population than in the adult. Reported incidences vary, but can be as high as 4.7%. It can occur due to occlusion of the superior vena cava secondary to thrombus, direct injury to the thoracic duct, or conditions involving high central venous pressures such as the Fontan procedure for hypoplastic left heart syndrome. Chylothorax is commonest after the Fontan procedure, bidirectional Glenn, tetralogy of Fallot repair, atroventricular septal defect in the setting of trisomy 21, and orthotopic heart transplantation. The consequences of chylothorax include electrolyte disturbance, immunodeficiency secondary to lymphocyte depletion and hypogammaglobulinemia, prolonged inpatient stay, and, ultimately, nutritional compromise secondary to protein loss in the chyle.

Management strategies include drainage of chyle using a pleural drain and switching to a medium chain triglyceride (MCT)-enriched formula, which bypasses the lymphatic system and directly enters the portal venous system. MCT formulas resulted in a reduction of chylothorax volume in 84% patients in some single-center studies. If chyle volumes continue to be high despite an MCT diet, recognized therapies include enteral rest and total parenteral nutrition and somatostatin analogs such as octreotide. Thoracic duct ligation is necessary in less than 5% of cases. MCT-enriched formulas are often unpalatable to the infant. Nasogastric feeding tubes are therefore sometimes employed to allow adequate nutrition. This further compounds the inability/delay in oral feeding and contributes to parental anxiety.

**Vocal cord palsy**

Vocal cord palsy is a complication of cardiothoracic surgery. It occurs when the recurrent laryngeal nerve is damaged and can be unilateral or bilateral, with the left cord affected in almost all cases described. Reported incidences vary between 4% and 20% with aortic arch surgery and a higher proportion of cases with the Norwood procedure than with non-arch surgery. It is a cause for extubation failure, which results in prolonged ventilation, adding to the delay in return to oral feeding in the congenital heart disease population. Vocal cord palsy can present with stridor, dysphonia, or...
feeding difficulties and aspiration described in almost half of cases. Diagnosis is made using indirect laryngoscopy, with laryngeal electromyography and ultrasound employed in certain situations. Vocal cord palsy as a postoperative complication ultimately leads to longer hospital stays and significant morbidity related to swallowing and feeding. Spontaneous recovery rates occur in one-third to two-thirds of patients and recovery tends to occur within 6 months. In the small percentage of patients in which recovery does not take place, a number of endoscopic procedures exist to widen the glottic space. Tracheostomy is rarely indicated given the benign natural history of the condition.

The role of breast-feeding
Human breast milk is the preferred form of nutrition for term and preterm infants. In preterm infants, breast milk has been shown to decrease the incidence of NEC and sepsis; however, infants with congenital heart disease frequently encounter obstacles to breast-feeding. Infants with congenital heart disease are often critically unwell in an ICU setting, both preoperatively and in the immediate postoperative period, negating the opportunity for establishing breast-feeding early. Mechanical ventilation, hemodynamic instability, electrolyte imbalance, and sepsis are barriers in preventing breast-feeding in the perioperative period. In addition, the practice of not feeding enterally in the setting of prostaglandin-dependent lesions further compounds this. The American Academy of Pediatrics recommends breast-feeding exclusively until 6 months of life then, with introduction of solid feeds, breast-feeding should continue until 12 months or as long as mutually desired by both mother and baby. Reported breast-feeding rates in infants with congenital heart disease are much lower. Some single-center studies report breast-feeding rates as high as 68% at discharge following a Norwood procedure. Anderson et al retrospectively analyzed infants who were electively admitted for a bidirectional Glenn procedure and found that 25% had been breast-feeding on discharge following the Norwood procedure, but less than 10% were still doing so by the time of the Glenn procedure. Concerns regarding the safety of breast-feeding in this population persist. Reports of desaturations and increased energy expenditure associated with breast-feeding are unfounded. The measurement of oxygen saturations during breast versus bottle feeding has shown that oxygen saturations are in fact lower during bottle feeding. In addition, breast-fed infants do not experience episodes of desaturation as frequently as bottle-fed infants during feeding.

Postoperative oral feeding and discharge home
Neonates with congenital heart disease experience many obstacles to achieving full oral feeding by the time of discharge from hospital. Forty percent of neonates who undergo surgery for their congenital heart lesions are discharged with partial nasogastric feeding. These neonates may require additional procedures such as fundoplication and gastric tube placement to help enteric feeds. Because of the risk of aspiration, not many would want to send these patients home on nasogastric tube feeding. Over two-thirds go home with calorie-enriched formula or breast milk and almost one-third are discharged below their birth weight. The presence of hypoplastic left heart syndrome and/or other ductal-dependent lesions, the need for cardiopulmonary bypass, age at full feeds prior to surgery, and prolonged intubation are risk factors for feeding difficulties and discharge with nasogastric feeding tubes. For infants at high risk of growth failure, such as hypoplastic left heart syndrome following the Norwood procedure, regular weight checks or home surveillance programs are beneficial in monitoring nutritional progress. Regular weight and saturation checks have been shown to reduce interstage mortality and improve nutritional status at the time of surgery for the Glenn procedure.

Disclosure
The authors declare no conflicts of interest in this work.

References


