Case Report

Emergency laparotomy for necrotising enterocolitis in a newborn with hypoplastic left heart syndrome

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ABSTRACT

Necrotising enterocolitis (NEC) and congenital heart disease are two distinct disease processes, but they appear to be inter-related, particularly in patients with the congenital heart condition known as hypoplastic left heart syndrome (HLHS). Both NEC and HLHS are causes of significant morbidity and mortality in the neonatal population. As medical and surgical advances allow for the palliation and correction of complex heart lesions at an earlier gestational age and lower birth weight, the already high-risk of NEC in this population is likely to increase. In this article, we report a case of a neonate with unpalliated HLHS who underwent emergency laparotomy for NEC. We also discuss the pathophysiology of these two diseases and the perioperative care of this rare group of patients.

Key words: Hypoplastic left heart syndrome, laparotomy, necrotising enterocolitis, non-cardiac surgery, unpalliated

INTRODUCTION

Neonatal surgical emergencies carry the risk of mortality and co-existing unpalliated congenital heart defects make the anaesthetic management a formidable challenge. Until the surgical palliation by Norwood and allograft cardiac transplantation by Bailey in the 1980s, mortality of hypoplastic left heart syndrome (HLHS) was 100%. HLHS is the abnormal development of the left-sided cardiac structures, resulting in obstruction to blood flow from the left ventricle. Necrotising enterocolitis (NEC), one of the common neonatal gastrointestinal emergencies, is characterised by ischaemic necrosis of the intestinal mucosa, invasion of organisms and dissection of gas into the muscularis and portal system.

HLHS neonates who develop NEC belong to a unique group of term babies with reasonable birth weights, unlike the common NEC patient.[1] These children require meticulous preoperative evaluation and with thorough preparation they tolerate general anaesthesia well.[2] The surgical management may need to take place before transfer to cardiac centres.

We have reported a neonate with unpalliated HLHS who underwent an emergency laparotomy for NEC.

CASE REPORT

A 4 kg term male with normal Apgar scores and blood investigations at birth, developed respiratory distress, acidosis and abdominal distention following initiation of oral feeds. X-ray abdomen showed bowel perforation. Echocardiogram reported HLHS with atrial septal defect (ASD). With a diagnosis of NEC, the baby was scheduled for emergency laparotomy. In the neonatal intensive care unit (NICU), the baby was intubated, ventilated, umbilical vessels were catheterised and infusion of dopamine and prostaglandin E1 was started. Anaesthesia was induced and maintained with fentanyl and vecuronium. Nitrous oxide was avoided. Fluids were administered based on central venous pressures. SpO_2 was maintained at 95%. Resection of the affected bowel was done along with ileostomy. The baby was shifted back to NICU and ventilated. The post-operative course was marked with cardiovascular instability following sepsis. On day seven, the fourth post-operative day, the baby died following a cardiac arrest.

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DISCUSSION

Children with severe uncorrected heart defects presenting with non-cardiac surgical emergencies are a major challenge to the anaesthetist and only few case reports are available in the medical literature.[2]

NEC and HLHS though inter-related, are two distinct entities and to understand the anaesthetic implications of both, we need to know the pathophysiology of each.

HLHS is the most common form of functional single ventricle heart disease with a prevalence rate of 2-3 cases per 1000 live births.[3,4] It is characterised by underdevelopment of the left heart with hypoplasia of the left ventricle and ascending aorta with atresia, stenosis or hypoplasia of the aortic or mitral valve; resulting in the right ventricle supporting the pulmonary and systemic circulation. Survival depends on a patent ductus arteriosus (PDA) for systemic perfusion and an adequate ASD for mixing of oxygenated and deoxygenated blood. Diagnosis is by echocardiographic imaging. Neonates with a non-restrictive ASD, are asymptomatic after delivery and become symptomatic with PDA closure. They have cyanosis, respiratory distress and decreased peripheral pulses. Initial management is providing adequate systemic perfusion by maintaining a PDA with prostaglandin E1 (0.01-0.05 ug/kg/min) and if necessary, creating a transcatheter atrial septoplasty. Later, palliative surgical repair is performed in three stages. With these interventions, the 5 year survival rate is approaching 65-70%.[5]

The incidence of NEC varies between 1 and 8% with a mortality rate between 20% and 40%.[6] NEC primarily occurs in premature infants. They have respiratory failure, abdominal distension, vomiting and diarrhoea. The Bell's criteria define the stages of NEC. Thrombocytopenia, acidosis and a heme-positive stool are associated with NEC. The radiographic finding of pneumatosis intestinalis [Figure 1] supports the diagnosis. Treatment is by nasogastric decompression, antibiotics, fluids, parenteral nutrition and insertion of a peritoneal drain. Surgery is indicated for perforation.

Immaturity of the gut is an important predisposing factor. Reports where NEC has been described in term infants with congenital heart disease have attributed all deaths to NEC. Among patients with HLHS, the incidence of NEC was 7.6%. With decreasing pulmonary vascular resistance (PVR), left to right shunting at the ductal level may decrease mesenteric flow, resulting in bowel ischaemia. Bacteraemia or sepsis seen in congenital heart disease may cause NEC. Patients may present with circulatory collapse and gut ischaemia on ductal closure. Prostaglandin use, indwelling umbilical catheters and cardiac catheterisation have been implicated in NEC, but the underlying circulatory physiology may be more contributory.

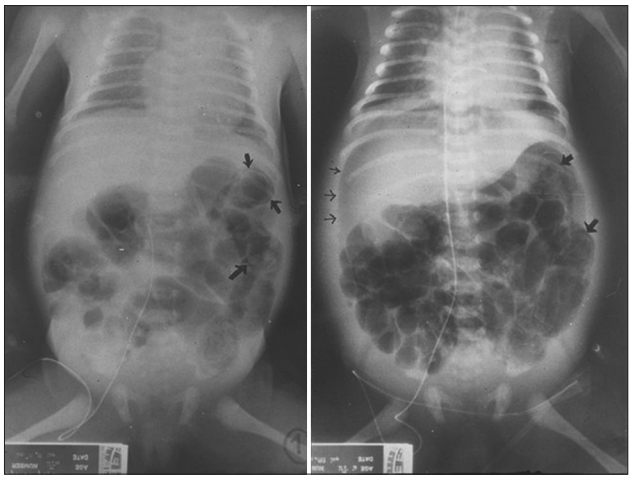

Figure 1: Plain abdominal X-ray on the left shows pneumatosis intestinalis (large arrow) a specific characteristic finding in necrotising enterocolitis. X-ray on the right is a follow-up film, which shows free air indicating perforation of the bowel (small arrow)

After birth, three factors affect the haemodynamic status: Decrease in PVR, size of the ASD and closure of PDA. The ratio of pulmonary blood flow to systemic blood flow (Qp/Qs) describes how the cardiac output from the single ventricle is partitioned. With the ductus open, most infants with HLHS can maintain a balance between PVR and systemic vascular resistance. However, a marked discrepancy in blood flow to the pulmonary and systemic circulations causes rapid onset of haemodynamic instability. An increase in Qp/Qs ratio results in excessive pulmonary flow at the expense of systemic flow. The ductus provides blood to the coronary arteries and the descending aorta and as it constricts, hypoperfusion occurs, resulting in shock. Because of this unique parallel circulation and propensity for systemic hypoperfusion, neonates with HLHS frequently develop NEC. A prolonged period of stabilisation in the NICU before transfer to theatre needs to be balanced against the requirement for early surgical intervention. Anaesthetic management of such infants include a strategy to maintain Qp: Qs slightly <1.[7]

Kang *et al*.[8] described the anaesthetic management of two neonates with HLHS with a tracheoesophageal

fistula in one and meningomyelocele in the other. In both cases, they minimised excessive Qp by prostaglandin E_1 in the first case and by adding nitrogen for a hypoxic mixture in the second case. Both provided stable haemodynamics.

Walker *et al*.[2] reviewed the notes of children with complex cardiac anomalies presenting for non-cardiac surgeries, including laparotomies for NEC. Induction was either by intravenous (IV) ketamine or sevoflurane inhalation. Maintenance was with volatile agents supplemented with high dose fentanyl. None of the children with HLHS required hypoxic gas mixtures. Our patient tolerated general anaesthesia well, but developed sepsis resulting in fatal cardiovascular collapse.

To summarise, the preoperative preparation is crucial. Signs of ductal closure and heart failure should be monitored. Sepsis, acidosis, hypovolemia and hypothermia must be addressed. Usually require invasive monitoring, inotropic support and mechanical ventilation. Consider IV opioid or incremental ketamine for induction. N_2O should be avoided. Volatile anaesthetics are poorly tolerated. Measures that induce pulmonary vasoconstriction, such as permissive hypercarbia or low inspired oxygen concentration, maybe utilised to counteract the presence of high Qp. Enormous fluid losses replaced with isotonic solutions. Transfusion maybe needed to improve the oxygen carrying capacity.

CONCLUSION

As recent advances allow for the palliation of HLHS, the incidence of these patients presenting with NEC is likely to increase and hence, we made an attempt to review the literature about the perioperative management of this rare group patients.

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