Anaesthesia for general surgery emergencies in children with severe unpalliated congenital heart defects-

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Introduction: Children with severe heart defects present a major challenge to anaesthetists. This is compounded if they present with non-cardiac surgical emergencies requiring correction before repair of their heart defects. We report the anaesthetic management of children with hypoplastic left heart syndrome (HLHS), unbalanced atrioventricular septal defects (AVSD), unstable Tetralogy of Fallot (TOF) and Truncus Arteriosus (TA) presenting to us for treatment of general surgical emergencies.

Methods: We reviewed the medical notes of neonates who presented for major thoraco/abdominal surgery in whom there was an untreated complex cardiac anomaly between November 2003 and November 2006. These patients were identified using the anaesthetic audit program of Birmingham Children’s Hospital NHS Trust. The following clinical details were noted; birth weight, gestational age, co-existing congenital anomalies, surgical diagnosis, management of anaesthesia, postoperative analgesia, and the place where postoperative care was provided.

Results: We identified 16 children; 7 had (HLHS), 5 (AVSD), 3 TOF and 1 had TA. All presented with clinically serious general surgical emergencies requiring operation in the first week of life. Fourteen underwent laparotomies for either necrotising enterocolitis (NEC), intestinal atresias or imperforate anus, and 3 had repair of tracheoesophageal atresia (TEF). Induction of anaesthesia was provided by intravenous (ketamine) or gaseous (sevoflurane) routes. Maintenance was with volatile agents supplemented with varying doses of opioids, commonly high dose fentanyl. Post-operative analgesia was provided by either parenteral administration of opioids and transfer to the intensive care unit or regional analgesia in specific instances, thus allowing the patient to return to a specialized ward.

Discussion: Neonates with untreated and unpalliated congenital heart disease present a challenge to anaesthetists in all circumstances. We have described a group in whom a general surgical emergency made anaesthesia and surgery a necessity in the first week of life. In no case could it be expected that the surgery would improve the child’s underlying cardiac condition. We were prompted to review the anaesthetic management of these patients having perceived an increase in the frequency of presentation, perhaps as a result of the changes in the referral of neonates with complex and univentricular cardiac lesions, as surgical outcomes have improved. These children may benefit from a prolonged period of stabilisation on the PICU before transfer to theatre, but this needs to be balanced against the requirement for early surgical intervention, and worsening of the clinical condition resulting in further abdominal distension, sepsis or respiratory failure. It is possible however, that the anaesthetic and surgical management of such general surgical emergencies may need to take place in the referring centre before arrangements can be made for transfer for cardiac care. The principles of anaesthesia are to minimise pathophysiological changes which may upset the delicate balance of blood flow between pulmonary and systemic systems. In Fallot’s tetralogy the aim is to prevent peripheral vasodilation or pulmonary hypertension increasing the right R to left (L) shunt. In AVSD, HLHS and TA the shunt can worsen in either directions, and efforts are made to avoid this by balancing the pulmonary and systemic vascular resistances. In anaesthesia this is primarily done by controlling PaCO$_2$ and PaO$_2$, and by judicious use of inotropic support.

Our significant experience of these children in our institution has allowed us to provide conditions where complete repair of the underlying surgical lesions can occur in contrast with described case reports (1,2). Children who had underlying HLHS were all anaesthetized by consultants with significant experience of this condition, none of these children required hypoxic gas mixtures (in fact in our series of 486 patients for primary correction of HLHS we have never needed to use a hypoxic gas mixture).

Other factors which are paramount in these cases are timing of surgery, monitors, placement of intravenous monitoring devices (avoiding the internal jugular veins in HLHS), level and experience of the anaesthetic practitioner and the possible need for transfer of these patients to major centres.

Refs: