Perioperative management of a neonate with Cantrell's pentalogy

Sir,

Pentalogy of Cantrell (POC) is a rare congenital anomaly with multiple defects involving sternum, pericardium, diaphragm, and anterior abdominal wall.[1] Survival of these babies depends on associated ectopia cordis and intracardiac anomalies.[2] Here, we report the perioperative management of a newborn with POC for surgical repair of omphalocoele.

A 5-day-old 2.5 kg male child with thoraco-abdominal defect was referred to our institute for surgical management. Parents deferred termination of pregnancy when the antenatal scan was suggestive of POC, and the baby was delivered at term by Caesarean section. The child had midline defect extending from the mid-sternum to umbilicus with the heart outside the thoracic cavity and bowel loops prolapsing through the defect [Figure 1]. However, there was no respiratory distress or cyanosis at birth. The child was referred to our institute on day four of life and was found to have dehydration, hypothermia, and elevated renal parameters (Urea – 110 mg/dl, creatinine – 2.5 mg/dl). The child was resuscitated with intravenous fluids, dopamine, and dobutamine infusions at 5 μg/kg/min. Echocardiography revealed pulmonary atresia, patent ductus arteriosus (PDA), and major aortopulmonary collateral arteries (MAPCAs). Expert opinion was sought from nephrologist and cardiothoracic surgeons.

The child was scheduled for omphalocoele repair under general anaesthesia. In the operating room, the child had a baseline heart rate of 145/min, blood pressure of 60/36 mmHg, and room air saturation was 94%. Anaesthesia was induced with intravenous fentanyl 5 μg, ketamine 6 mg, and atracurium 1.5 mg. Trachea was intubated with 3.5 mm inner diameter uncuffed endotracheal tube. The right femoral artery was cannulated for continuous blood pressure monitoring. Anaesthesia was maintained with sevoflurane in O_2/air with FiO_2 50–60%. The

lungs were ventilated with peak inspiratory pressure of 16 cmH_2O. The diaphragmatic defect was repaired and omphalocoele was covered with mobilized skin. There was no change in peak airway pressure. Total duration of surgery was 40 min; blood loss was minimal, urine output was 3 ml, and the child received 15 ml of crystalloid. The child was shifted to neonatal intensive care for further management. Eventually, the sepsis and kidney injury worsened and the child expired on the fourth postoperative day.

Thoracoabdominal syndrome or POC has five characteristic features – omphalocoele, diaphragmatic hernia, sternal cleft with or without ectopia cordis, diaphragmatic pericardial defect, and intracardiac defects. Toyama *et al*. classified the syndrome into three types based on the severity.[3] Class-1 or severe form of the syndrome has all five components whereas Class-2 has four defects including ventral wall and intracardiac anomalies. Class-3 or incomplete form consists of sternal cleft with other combination of defects.

In this case, the child had all five components, i.e., complete or severe form [Video 1, Supplement data]. Management of this syndrome requires a multidisciplinary team involving neonatologist, paediatric anaesthesiologist, cardiothoracic, and paediatric surgeons. Most complications such as dehydration, hypothermia, sepsis, and renal injury could have been prevented if the child was referred to a higher center early after diagnosis.

The complexity of the surgery depends on the extent of defect and the physiological reserve of the child.

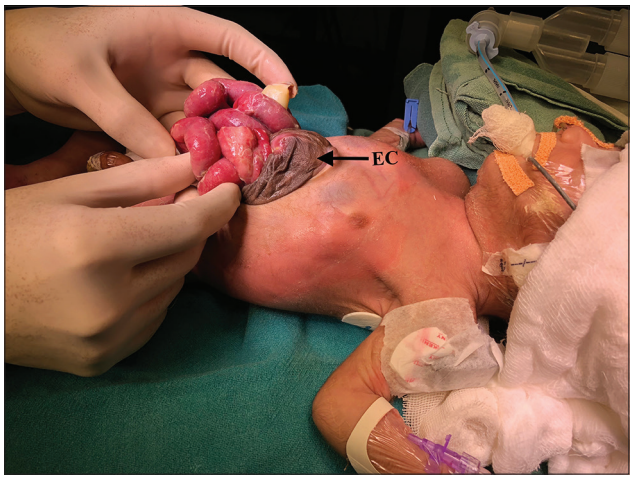

Figure 1: Cantrell's pentalogy with omphalocoele and ectopia cardis

Hence, a staged surgical procedure is preferred.[4] Our primary aim was to cover the exposed bowel to prevent further infection, heat and fluid loss, and to close the diaphragmatic defect to prevent herniation of abdominal contents into thorax.

The anaesthetic challenges were chances of cyanosis, hypoxia, and right ventricular failure due to pulmonary atresia and haemodynamic effects due to compression of ectopic heart or kinking of major vessels. There was no significant hypoxia/cyanosis; possibly due to adequate pulmonary blood flow by PDA and MAPCAs. The intravenous fluid was meticulously titrated in the newborn with kidney injury as peritoneal or haemodialysis could not be possible. Above all our main concern was to prevent abdominal compartment syndrome. Increase in intra-abdominal pressure due to reduction of omphalocoele may lead to further decrease in renal perfusion and diaphragmatic splinting and can have a cumulative effect on the already compromised cardiac and pulmonary function.[5] However, in our case there was no rise in intra-abdominal pressure and we opted to monitor abdominal pressure in the postoperative period. This article discussed the various anomalies associated with POC and its anaesthetic significance. Furthermore, it highlights the complications due to delayed referral and their effect on already deranged cardiac and pulmonary function.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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