Peri-operative management of a neonate with tracheo-oesophageal fistula and anorectal malformation: Survival of the fittest

Sir,

Oesophageal atresia (OA) with tracheo-oesophageal fistula (TOF) is a rare congenital anomaly usually seen in association with the VACTERL spectrum of disorders in neonates. \[^1\] Management of these groups of anomalies is generally an early surgical intervention in most cases. Majority of neonates are managed with a palliative procedure until fit for a definitive surgery. \[^2\] Isolated case reports have been published about management of multiple defects. \[^3,4\]

We describe a case of a neonate with a high anorectal malformation (ARM) along with TOF who was managed surgically in a single setting for both defects. The pathophysiology of TOF gets further accentuated in a setting of ARM as there is absolutely no way to decompress the bowel, either from above or below. This calls for an urgent intervention in the form of a colostomy/gastrostomy or definitive repair, given further compromise of the already susceptible lungs.

A 1-day-old male neonate was referred to our tertiary care hospital with history of frothing at the mouth and difficulty in breathing soon after birth. Nasogastric tube could not be passed into the stomach, and on examination, there was no appreciable anal opening. X-ray of the chest and an invertogram confirmed the diagnosis of OA with TOF and a high ARM [Figure 1].

The patient was optimised transiently in the Neonatal Intensive Care Unit (NICU). Given the poor socio-economic status of the patient and the remote possibility of the parents returning for follow-up procedures, a decision to operate both defects was taken, and the patient was posted for definitive surgery on the 2nd day of birth.

Pre-anaesthetic evaluation revealed no overt signs of respiratory distress with a few occasional crackles in both lung fields and a respiratory rate of 45/min. The blood gas picture pre-operatively revealed partial pressure of oxygen (pO\(_2\)) of 70 mm Hg, partial pressure of carbon dioxide (pCO\(_2\)) of 45 mmHg and pH of 7.30, with normal electrolyte levels. Head-to-toe examination did not reveal any significant finding except a distended, tense abdomen.

After pre-warming in the OT and pre-medicating with injection fentanyl, the neonate was induced with sevoflurane, maintaining spontaneous respiration. The trachea was intubated with a 3.0 mm internal diameter endotracheal tube and advanced inside till a length where it would have become endobronchial. It was then withdrawn while auscultating till a point where bilateral breath sounds were just heard. Manual ventilation was performed using a Jackson-Rees circuit with the aim of avoiding excess ventilation of fistula and also to perceive lung compliance. The child was placed in left lateral position after proper padding of all pressure points. A precordial stethoscope was fixed, for heart and breath sounds assessment.

The fistula was identified and ligated by the surgeon which improved ventilation, followed by the oesophageal anastomosis. The patient was then put in a lithotomy position for the ARM repair by an anterior approach. After confirming adequate ventilation and bilateral breath sounds again after change of position, the rest of the surgery proceeded uneventfully without the requirement of blood products; haemodynamic stability was maintained with infusion of Ringer lactate with 2% dextrose.

The child was ventilated postoperatively (PO) in the NICU. Post-extubation, the chest tube and urinary catheter were removed on the 5th PO day [Figure 2].
and enteral feeds were initiated on the 6th PO day. The child was healthy and was doing well at the time of discharge.

When an anaesthesiologist encounters a case of TOF in a neonate, searching for other VACTERL group of anomalies is of prime importance as it guides management and also for prognostication. Presence of pneumonia or any cardiac anomaly warrants prior optimisation, rather than taking up the patient for surgery on an emergency basis. If the lung condition is not optimal, palliative procedure in the form of a gastrostomy under local anaesthesia is performed or surgery is delayed till the neonate becomes fit to undergo general anaesthesia.

In the present case, the presence of a simultaneous ARM made the matter complicated as an intervention had to be done, either a colostomy or a definitive repair of ARM. Our patient had good prognostic factors in terms of absence of cardiac anomalies, adequate weight and minimal respiratory compromise at the time of surgery. Thus, the child could tolerate the surgery and subsequently be weaned successfully from the ventilator. It was indeed 'survival of the fittest'.

Thus, a successful outcome depends on good pre-operative evaluation and careful selection of choice and timing of surgery based on prognostic factors and associated anomalies.