

## Anaesthetic management of a child with unrepaired complete atrioventricular canal defect, double outlet ventricle and pulmonary stenosis for non-cardiac surgery

Sir,

Complete atrioventricular (AV) canal defect with double outlet right ventricle (DORV) and severe pulmonary stenosis (PS) is a rare complex congenital heart disease, requiring surgical correction usually within 6 months of life. Patients with the unrepaired lesion can develop elevated pulmonary venous pressures, which over the time become fixed and they develop reversal of shunt and cyanosis (Eisenmenger syndrome).<sup>[1]</sup> The lifespan of the patient depends on the balance between the pulmonary and systemic circulations.<sup>[2]</sup>

We present a case of a 7-year-old boy weighing 13 kg diagnosed as heterotaxy syndrome with complete

AV canal defect and DORV presenting for emergency exploratory laparotomy for intestinal obstruction. He was not on any medical follow-up. On examination, clubbing and cyanosis were present and oxygen saturation was 75% on pulse oximetry. Jugular venous pressure was not raised and there was no pedal oedema. A pansystolic murmur was audible at the left upper sternal border. The rest of the vital parameters were within normal limits. Blood investigations showed a haemoglobin value of 20 gm/dl. Other investigations including complete blood counts, coagulation profile, liver and kidney function tests, and serum electrolytes were unremarkable. Two-dimensional echocardiography revealed situs ambiguous, interrupted inferior vena cava with azygous communication, complete AV defect Rastelli type C, both great vessels arising from the right ventricle, severe PS with the pulmonary gradient of 60 mmHg, hypoplastic pulmonary artery and normal biventricular function. No abnormality was detected on the chest radiogram and electrocardiogram of the patient.

Rapid sequence general anaesthesia was induced with intravenous (IV) fentanyl 60 µg, ketamine 5 mg and succinylcholine 15 mg. Endotracheal intubation was accomplished and sevoflurane 1–2% in 100% oxygen was used for the maintenance of anaesthesia.

Intraoperatively, an infusion of fentanyl 5 µg/h was administered and atracurium was administered intermittently. Invasive blood pressure and stroke volume variation monitoring was instituted after the induction of anaesthesia. The surgery lasted for about 120 min and the intraoperative blood loss was 200 ml (15 ml/kg). A total of 750 ml of lactated Ringer's solution was administered. After the completion of the surgery, ultrasound-guided bilateral transversus abdominis plane block was given for postoperative analgesia. Neuromuscular blockade was reversed and the child was extubated uneventfully. He was shifted to the intensive care unit for the next 48 h for observation.

The perioperative concerns in the management of these patients with complex cardiac lesions undergoing non-cardiac surgery include identification and optimisation of anomalies associated with heterotaxy syndrome, cardiac risk stratification, maintaining the balance of pulmonary vascular resistance (PVR) and systemic vascular resistance (SVR), and prevention of pulmonary hypertensive crisis.<sup>[3-5]</sup> The goals of anaesthesia include the maintenance of baseline PVR, SVR and biventricular function and avoidance of perioperative hypoxia, hypercarbia, hypothermia and acidosis. Reduction of SVR during induction of anaesthesia can lead to clinical deterioration. Hence, induction agents with minimal changes in SVR and PVR should be selected.<sup>[6]</sup> We chose high-dose fentanyl and ketamine for induction as these agents have minimal effect on the pulmonary vessels, maintain haemodynamic and blunt the response to noxious agents. We used sevoflurane for the maintenance of anaesthesia as it has a relatively lesser effect on SVR and myocardial contractility. Similar to the induction of anaesthesia, catecholamine surge during the emergence of anaesthesia can lead to haemodynamic instability and should hence be carefully managed. Any event of precipitous fall in SVR such as during induction or intraoperatively due to surgical blood loss should be aggressively treated. Inadequate oxygenation and ventilation may lead to increase in PVR and should be optimised. Anaesthesiologists should be aware of the clinical signs of pulmonary hypertensive crisis under anaesthesia and its immediate treatment. Postoperatively, patients should be monitored until they have returned to their preoperative baseline.

To conclude, patients with unrepaired complete AV canal defect and DORV undergoing non-cardiac

surgery have heightened perioperative morbidity and mortality. The anaesthetic management requires a careful approach and is determined by knowledge of the underlying pathophysiology and the effect of anaesthetic agents on the myocardium and pulmonary vasculature.

#### Declaration of patient consent

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

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#### Conflicts of interest

There are no conflicts of interest.

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