Anesthetic management of a 2-day old neonate with double outlet right ventricle associated with anorectal malformation posted for emergency colostomy surgery

Double outlet right ventricle (DORV) is a rare congenital heart disease in which both aorta and pulmonary artery arise from the right ventricle.[1] It is always associated with ventricular septal defect (VSD), the only outlet to the left ventricle.[2] Being a cyanotic heart disease, it becomes challenging when associated with other congenital heart diseases. Here we present a case of a 2-day-old child with associated congenital anomalies (VACTRAL anomalies) posted for emergency surgery.

A 2-day-old preterm baby, weighing 2.2 kg, born to a diabetic mother, presented with abdominal distention due to anorectal malformation (AVM) and was planned for emergency laparotomy and colostomy. His birth history revealed that he did not cry and was cyanotic at the time of birth with room air oxygen saturation (SpO_2) 25%–40%. Despite providing positive pressure ventilation, he did not improve and was intubated and put on mechanical ventilation in the neonatal intensive care unit (NICU). He was diagnosed with DORV with significant ventricular septal defect and pulmonary stenosis. He had associated AVM, bilateral pelvic-ureteric junction obstruction, and sacral agenesis. Two days after birth, he developed abdominal distension and was planned for emergency surgery. His vitals include SpO_2 60%–66% on 100% fiO_2, blood pressure (BP) 80/50 mm Hg, and heart rate (HR) 160/min. His blood reports were normal. Despite being on dextrose infusion, he had persistent hypoglycemia with blood sugar ranging 30–35 mg/dl. His blood gas analysis showed pH 7.194, pCO_2 62.5, pO_2 27.2, bicarbonates 18.6, and base deficit −4.6.

After taking informed written consent, he was taken for surgery. He was induced with injection ketamine 2 mg/kg, injection fentanyl 4 mg/kg, and injection atracurium 0.5 mg/kg. The patient was put on pressure control ventilatory mode with pressure support (PS) 18, respiratory rate 30/min, fiO_2 80% maintaining saturation of 66%. He was started with 10% dextrose in ringer lactate solution at 20 ml/h. Maintenance of anesthesia was done with ketamine infusion

at 0.5 mg/min and atracurium injection as intermittent boluses. The surgery lasted for 1 h. The baby was shifted to the NICU.

Our case is different from previous case reports mentioned in the literature as we altogether avoided inhalational agents, even sevoflurane, and maintained the patient on ketamine infusion. Our main aim was to maintain systemic vascular resistance (SVR) and decrease pulmonary vascular resistance (PVR) to prevent further deterioration of oxygen saturation by preventing further increase in the venous blood flowing into the systemic circulation. This goal was achieved with ketamine infusion at 0.5 mg/min as ketamine has a sympathomimetic effect.[3] The main reason for avoiding sevoflurane was to prevent a fall in SVR.[4] Intraoperative management of these neonates with multiple cardiac malformations is very challenging. The primary learning objective in our case is that intravenous ketamine should be the preferred intravenous anesthetic agent in such cases. It can be used both as an induction and maintenance agent in such cases, as we did in our case. Inhalation agents decrease systemic vascular resistance; thus, they should be used judiciously.

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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