Anaesthetic management in a patient of uncorrected double outlet right ventricle for emergency surgery

Sir,

With increasing diagnostic and therapeutic modalities, more and more patients with congenital heart disease (CHD) survive till adulthood and may present for coincident elective or emergent noncardiac surgery.[1] Such patients, especially with unrepaired or unpalliated cardiac defects, offer a great challenge because of their complex physiological modifications secondary to chronic hypoxia and excessive or reduced pulmonary blood flow. Double outlet right ventricle (DORV) is a complex cardiac defect that comes under the broad category of single ventricle physiology. We present the successful management of a patient with DORV for emergent noncardiac surgery.

A 20-year-old male presented with Fournier's gangrene involving the entire inguino-scrotal region and the anterior abdominal wall up to the umbilicus. On presentation, the patient appeared malnourished, dehydrated and tachypnoeic. He had central cyanosis and Grade IV clubbing. His heart rate was 122/min, blood pressure (BP) was 90/56 mmHg, central venous pressure (CVP) was 6 mmHg (as measured through a central line inserted preoperatively under local anaesthesia) and arterial oxygen saturation (SpO_2) while breathing room air was 85%. Cardiovascular examination revealed loud P_2 and a Grade IV pansystolic murmur in the left parasternal region. Airway examination showed retrognathia and anterior larynx. Laboratory investigations revealed haematocrit of 54.5%, leucocyte count 18,700/mm^3, platelet count of 74,000/mm^3 and INR of 1.6. Arterial blood gas revealed pH 7.26, PaO_2 49.3 mmHg, $PaCO_2$ 44.9 mmHg and SaO_2 88.3%. Echocardiogram revealed "DORV, right ventricular hypertrophy, large subpulmonic ventricular septal defect (VSD), with predominantly right to left shunt and moderately severe pulmonary artery hypertension."

Informed American Society of Anesthesiologists (ASA) III_E consent was taken. Lactated Ringer's solution was infused to correct his dehydration and build the CVP up to 12 mmHg. Preoperatively, he received two units of random donor platelets and four units of fresh frozen plasma. Since the systolic BP was persistently around 90 mmHg in spite of fluid resuscitation, noradrenaline infusion was started and titrated up to a rate of 0.3 mcg/kg/min started before induction of anaesthesia. Premedication in the form of glycopyrrolate 0.2 mg, midazolam 2 mg and fentanyl 100 mcg were administered intravenously. General anaesthesia was induced with graded doses of intravenous ketamine and sevoflurane in oxygen. Intravenous suxamethonium was given, and the trachea intubated with the help of gum elastic bougie. The patient was ventilated limiting peak airway pressures to below 25 cm H_2O. Anaesthesia was maintained with oxygen, air and sevoflurane and atracurium for muscle relaxation. Perioperative monitoring included electrocardiogram, SpO_2, BP, CVP, end-tidal carbon dioxide, airway pressures and urine output. Noradrenaline infusion was titrated to maintain the systolic BP between 110 and 130 mmHg. At the end of the procedure, neuromuscular blockade was reversed, patient was extubated and was shifted to the intensive care unit.

DORV is a heterogeneous group of congenital cardiac malformations, in which both the aorta and the pulmonary artery arise from the right ventricle and the only outlet to the left ventricle is a VSD which is almost always present. Both the systemic and pulmonary circulations are in parallel, and the circuit with lower resistance would preferentially be perfused.[2]

Goals of anaesthetic management in a patient with double outlet right ventricle include:

Maintenance of cardiac output by maintaining preload, contractility and heart rate, and maintaining systemic vascular resistance (SVR) higher than pulmonary vascular resistance (PVR) to avoid increased recirculation of systemic venous blood. A fall in SVR leads to further reduction in pulmonary blood flow, which impedes oxygenation; PVR needs to be lower to ensure adequate blood flow to the lungs.

PVR can be reduced by increasing inspired oxygen concentration (FiO_2), elective hyperventilation to achieve moderate metabolic alkalosis and avoiding hypothermia, hypoxemia and acidosis. Cyanosis produces polycythemia which increases viscosity

and thus SVR and PVR. Adequate hydration must be ensured to prevent intravascular thrombosis.

As our patient presented with features of septic shock (elevated total leucocyte counts, persistent hypotension, thrombocytopenia and altered coagulation profile), noradrenaline was the vasopressor of the first choice. Ketamine, because of its sympathomimetic properties which lead to rise in SVR, was used for induction. Nitrous oxide was avoided because of preexisting pulmonary hypertension. We did not opt for regional anaesthesia because of the preexisting coagulopathy and to avoid the hypotension that would result from sympathectomy. This, on the background of septic shock, would have completely jeopardized systemic oxygenation by diverting blood flow away from the pulmonary circulation.

There are no evidence-based recommendations to guide the anaesthetic management of patients with CHD undergoing noncardiac surgery.[3] It is imperative for anaesthesiologists to understand the underlying pathophysiology of each CHD patient and to tailor the intraoperative anaesthetic management according to the specific needs and to improve the patient outcome.

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