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Case of Uncorrected and Inoperable Tetralogy of Fallot with Pulmonary Atresia and Major Aorto Pulmonary Collaterals with Anorectal Malformation - Anesthetic Management Mimicking the Pathophysiology in a Limited Cardiac Resource Centre

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Abstract

We report a case of a 7 year old girl with anorectal malformation and uncorrected Tetralogy of Fallot with pulmonary atresia and major aortopulmonary collateral arteries posted for surgery. Based on the pathophysiology of the congenital cardiac lesion and low lying anorectal malformation, anoplasty was performed under general anesthesia with single shot epidural analgesia. We highlight the various pathophysiological mechanisms to be considered while tailoring anesthesia management for a complex rare uncorrected congenital cardiac lesion in a child.

Keywords: Tetralogy of Fallot; Pulmonary atresia; Major aortopulmonary collaterals; Anorectal malformation; Anesthesia; Epidural anesthesia

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Introduction

Children with Anorectal Malformations (ARM) requiring surgical repair are frequently encountered by a pediatric anesthesiologist. The association of cardiac anomalies is well documented in children with ARM [1]. However, associated Tetralogy of Fallot with pulmonary atresia with a ventricular septal defect and major aortopulmonary collateral arteries (PA/VSD/MAPCAs), is a rare entity with varying severity requiring complex anesthetic management [2]. In this report, we discuss specific anesthetic considerations, based on the pathophysiology of an uncorrected complex, congenital cardiac lesion in a child with PA/VSD/MAPCAs presenting for a non-cardiac surgery.

Case Presentation

A 7 year girl weighing 12 kg (weight for age 0.10 percentile) measuring 75 cm (height for age 0.10 percentile) posted for surgical correction of rectovaginal type of anorectal malformation with fecal fistula presented with severe shortness of breath (NYHA class IV). The child was diagnosed at day 15 of life with Tetralogy of Fallot with PA and VSD and MAPCAs. Financial constraints limited early surgical intervention. Her history was consistent with recurrent episodes of cough with expectoration, easy fatigability, limited activity, poor weight gain requiring multiple hospital admissions which was managed symptomatically over the years.

Physical examination revealed an afebrile child with marked peripheral cyanosis, grade 4 clubbing of the fingers and pallor. Her respiratory rate was 38 breaths/min with a room air oxygen saturation of 76%; Heart Rate (HR) 120 beats/min and noninvasive Blood Pressure (BP) of 96/43 mmHg. Cardiac examination revealed a precordial bulge, parasternal heave, systolic thrill and grade 3 systolic murmur in all auscultation fields. 2D Echocardiography showed membranous pulmonary atresia- confluent branch pulmonary artery collaterals, large subaortic VSD with bidirectional shunt, right ventricular hypertrophy and compensatory branches from the start of the descending aorta. Her estimated left ventricular ejection fraction was 57%. Metabolic screening tests and complete blood picture were within normal limits.

After obtaining appropriate consent from the parents, the child was fasted nil per oral for 6 h with administration of infective endocarditis prophylaxis. The intraoperative objectives were to maintain myocardial contractility and systemic vascular resistance with minimal change in volume

status and pulmonary vascular resistance. She was wheeled into the operating theatre after receiving 1 mg of intravenous midazolam. Non-invasive BP, electrocardiogram and SpO_2 were monitored. After preoxygenation, intravenous ketamine 2 mg/kg, fentanyl 2 ug/kg was administered. Anesthesia was maintained with 100% oxygen with a minimum alveolar concentration of 2% sevoflurane with spontaneous mask ventilation to ensure normocarbia. 0.5 ml/kg of 0.2% Bupivacaine with 2 mg/kg of Tramadol was deposited in the lumbar epidural space (L2-3) after adequate sterile precautions. She received adequate intravenous fluid and 15 mg/kg of Paracetamol to supplement intraoperative analgesia. The surgery lasted 30 min with minimum fluctuation in the child's hemodynamic parameters and blood loss. She was shifted to the intensive care unit and was discharged on the 2nd postoperative day uneventfully.

Discussion

TOF/PA/MAPCAs is a rare form of congenital heart disease, with an incidence of 0.7 per 10,000 live births [3]. The pathophysiology of this complex lesion depends upon the pulmonary to systemic blood flow ratio (Qp/Qs) that occur via the MAPCA's [4]. On evaluation, our child belonged to the subset of MAPCA with suprasystemic pressures in the pulmonary vasculature (Qp/Qs >1) and thus was deemed inoperable. For the planned surgical procedure, a simple anesthetic plan was framed with minimal perturbations of the systemic and pulmonary vasculature and avoidance of cardiac and respiratory depression. The basic cardiac grid was followed which included maintaining normal range of heart rate and rhythm with euvolemia and multimodal analgesia, cardiac contractility was ensured with meticulously managing anesthetic agents and volume status. Ketamine with sympathomimetic effect increased SVR, HR and cardiac output. PVR was maintained low to optimal by avoiding triggers such as pain, hypoxia, hypercarbia and positive pressure ventilation [5]. This was achieved with Midazolam used to calm preoperatively. Fentanyl, Paracetamol and Epidural analgesia-complemented the pain management plan. Fraction of inspired Oxygen saturation -0.75-1% was constant.

Spontaneous ventilation was ensured to avoid airway interventions that could increase PVR. Sevoflurane was titrated according to hemodynamic response and expiratory gas concentration. Postoperative care should preferably be provided in the intensive care unit, which can facilitate invasive monitoring and early intervention.

Conclusion

In conclusion understanding the pathophysiological changes of the disease aided management decisions. In our case we found that single shot epidural analgesia supplemental mask anesthesia with careful cardiovascular monitoring was a successful approach in a PA/VSD/MAPCAs patient.

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