VENTRICULO-PERITONEAL SHUNT SURGERY
IN AN INFANT WITH DOUBLE AORTIC ARCH,
PATENT DUCTUS ARTERIOSUS
AND ATRIAL SEPTAL DEFECT

- Case Report -
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Abstract
Double aortic arch with patent ductus arteriosus and atrial septal defect is an uncommon association. Such complex cardiac lesions may complicate an otherwise normal anesthetic course. We came across a case with aqueductal stenosis and hydrocephalus, scheduled for ventriculo-peritoneal shunt surgery, on an emergent basis. The child was managed successfully. The anesthetic implications of resultant left-to-right shunt with increased intracranial pressure have been described.

Key Words: Double aortic arch, Patent ductus arteriosus, Atrial septal defect, Anesthesia.

Introduction
Double aortic arch is a rare but life threatening condition, if misdiagnosed¹. Its association with patent ductus arteriosus (PDA) and atrial septal defect (ASD) has been mentioned in association with few genetic abnormalities²,³. However, the anesthetic implications of this association in patients undergoing neurosurgical procedures have never been described in literature.

We present a case of congenital hydrocephalus due to aqueductal stenosis, with associated double aortic arch, PDA, and ASD, scheduled for an emergency ventriculoperitoneal (VP) shunt surgery.

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Case Report

A 3-months-old male baby weighing 4 kg was admitted to our hospital with complaints of increasing size of head and upward gazing of eyeballs, for the previous 15 days. This was associated with vomiting and decreased feeding for 2 days. There was no history of seizures or focal neurological deficit. The antenatal history was remarkable for breach presentation and cesarean section delivery from a polyhydramnios mother, at 38 weeks of gestation. He developed bronchopneumonia during the first month of life which improved following antibiotic treatment. On examination, the child was pale but, there was no cyanosis, jaundice, clubbing, or pedal edema. The heart rate was 116 beats.min⁻¹, and respiratory rate 28 breaths.min⁻¹. He had features of hydrocephalus with increased head circumference (42 cm), open and tense fontanelles, and presence of sunset sign. It was noticed that the baby developed bluish discoloration of mouth and lips while crying. On cardiac auscultation systolic murmur was audible. Chest X-ray showed a globular cardiac silhouette with cardiomegaly, bilateral hilar prominence and thickening of bronchovascular markings. ECG showed normal sinus rhythm with no evidence of ventricular hypertrophy. Echocardiography revealed a complex congenital heart disease with presence of ASD (ostium secundum type), PDA, and double aortic arch. There was no evidence of pulmonary hypertension. Peripheral arterial pulsations were feeble bilaterally in the upper limb, whereas they were well felt in the lower limbs. The oxygen saturation on room air was 99%. The child had a reducible umbilical hernia and hypospadiasis as well. Routine blood tests were within normal limits with a hemoglobin level of 14 g/dl. MRI of brain showed gross hydrocephalus with aqueductal stenosis and corpus callosum agenesis. Non-contrast CT scan head showed hydrocephalus and dilated lateral ventricles with periventricular ooze. This child was posted for an emergency ventriculoperitoneal shunt surgery in view of the raised intracranial tension and deterioration of neurological condition.

In the operation theatre, inhalational induction was performed using sevoﬂurane in oxygen and air mixture. After securing an intravenous (IV) access, fentanyl 2 mcg.kg⁻¹ and rocuronium 1 mg.kg⁻¹ was given. Tracheal intubation was facilitated using a 3.5 mm ID uncuffed endotracheal tube. Anesthesia was maintained with sevoﬂurane in oxygen (FiO₂ -0.5)-air mixture, fentanyl, rocuronium, and intermittent positive pressure ventilation. Monitoring parameters included ECG, non-invasive BP, SpO₂, end-tidal CO₂, and inspired concentration of inhalational agents. The end-tidal CO₂ was kept between 33 ± 2 mmHg. Posterior tibial artery was cannulated for continuous BP monitoring. Intraoperatively, the mean BP was targeted to keep within 10% of the baseline values. However, there was no episode of desaturation or hemodynamic instability. Serial arterial blood gas analysis revealed metabolic acidosis (Table 1), which was managed with IV supplementation of sodium bicarbonate (4.2%) solution.

Total duration of procedure was 90 minutes. The blood loss was approximately 20 ml, and a total of 80 ml Ringer’s lactate was used as maintenance fluid. Temperature was maintained around 36ºC. At the end of procedure, residual neuromuscular blockade was reversed with neostigmine and glycopyrrolate, and trachea extubated, uneventfully. The child was observed in the intensive care unit for 24 hours. Though metabolic acidosis persisted (Table 1), further bicarbonate supplementation was stopped as the child was clinically stable. He was discharged on third postoperative day with an advice for further cardiac evaluation. At 1-month follow-up, the child had improved neurological status.

Table 1
Perioperative arterial blood gas (ABG) analysis

<table>
<thead>
<tr>
<th></th>
<th>pH</th>
<th>pO₂ (mmHg)</th>
<th>pCO₂ (mmHg)</th>
<th>HCO₃⁻ (mmol/L)</th>
<th>BE</th>
<th>S₉₂ (%</th>
<th>Na⁺ (mmol/L)</th>
<th>K⁺ (mmol/L)</th>
<th>Ca⁺⁺ (mmol/L)</th>
</tr>
</thead>
<tbody>
<tr>
<td>After induction</td>
<td>7.22</td>
<td>184</td>
<td>35.9</td>
<td>14.2</td>
<td>-12.1</td>
<td>100</td>
<td>139</td>
<td>3.9</td>
<td>1.21</td>
</tr>
<tr>
<td>Intraoperative</td>
<td>7.254</td>
<td>234</td>
<td>38.0</td>
<td>16.3</td>
<td>-9.6</td>
<td>100</td>
<td>139</td>
<td>3.8</td>
<td>1.05</td>
</tr>
<tr>
<td>Postoperative (ICU)</td>
<td>7.288</td>
<td>190</td>
<td>34.6</td>
<td>16</td>
<td>-9.3</td>
<td>100</td>
<td>139</td>
<td>4.2</td>
<td>1.12</td>
</tr>
</tbody>
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systemic blood flow in the presence of left-to-right shunt as a result of PDA can cause pulmonary over circulation. This results an elevation of left atrial pressure due to augmented pulmonary venous return. Diastolic blood pressure may be compromised if cardiac output is reduced or there is a low resistance vascular bed, as in our case. Infants with greatly increased pulmonary blood flow suffer from congestive heart failure secondary to increased volume overload. Obstruction of the large and small airways results in increased airway resistance and poor compliance. The inspiratory pressure needed for adequate positive-pressure ventilation must be adjusted accordingly.

Hyperoxia and hypocarbia cause pulmonary vasodilatation. Hence, it is advisable to reduce the FiO₂ and to avoid hyperventilation. Normocapnia is helpful in managing the increased pulmonary blood flow resulting from such complex heart disease. However, in this case, mild hypocapnia was maintained without any untoward event, in order to reduce the intracranial pressure (ICP). Nitrous oxide (N₂O) is best avoided, as it causes sympathetic hyperactivity, increased pulmonary vascular resistance, and interferes when FiO₂ requirement is increased. It also causes an increase in cerebral blood flow, which is of concern in patients with raised ICP. Metabolic acidosis in this patient was possibly because of decreased systemic blood flow in the preoperative period, despite adequate efforts to maintain PVR and SVR, intraoperatively. Additional anesthetic issues include avoidance of air bubbles in intravenous lines to prevent paradoxical air embolism.

To conclude, we successfully managed a patient of aqueductal stenosis and hydrocephalus with associated double aortic arch, PDA, and ASD, who underwent VP shunt surgery. A detailed history and complete physical examination along with an adequate investigational work up would help in formulating anesthesia plan in such patients.
References