



Neonate with Omphalocele and Dextrocardia: Anaesthetic Goals and Challenges

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Omphalocele and gastroschisis are the two common congenital malformations of the anterior abdominal wall. Omphalocele can be associated with other congenital anomalies such as cardiac anomalies. Association of omphalocele with dextrocardia has been reported in few literatures previously. We describe here such a rare association of omphalocele, dextrocardia with patent ductus arteriosus with a brief review on the anesthetic challenges in the perioperative period.

Key words: Anesthetic management, dextrocardia, Neonatal Intensive Care Unit, omphalocele, patent ductus arteriosus

INTRODUCTION

Omphalocele and gastroschisis are the two common congenital malformations of the anterior abdominal wall. Omphalocele or exomphalos is a midline abdominal wall defect containing herniated viscera covered by a membrane. This membrane consists of amnion on the outer surface and peritoneum on the inner surface.¹ Omphalocele can be associated with other congenital anomalies such as cardiac malformations ranging at the rate of 15–54%.^{2,3} Among cardiac anomalies, association of omphalocele with dextrocardia has been reported in few literatures.^{3,4} We describe here a rare coexistence of omphalocele, dextrocardia with patent ductus arteriosus (PDA) along with a brief review on the anesthetic challenges.

CASE REPORT

A 3-day-old male child weighing 2.8 kg was admitted for repair of an abdominal wall defect [Figure 1]. The baby was born vaginally after 37 weeks of gestation with Apgar scores of 9 at both 1 and 5 min after birth. Physical examination revealed a heart rate of 144 beats/min; blood pressure 53/36 mm Hg; SpO₂ 96% on oxygen hood; respiratory rate of 48 breaths/min;

and temperature 36.5°C. On inspection in the supine position, the patient showed no apparent respiratory distress. On auscultation, the lungs were clear, and a pansystolic murmur was heard over the right side of the precordium. Chest X-ray and echocardiography revealed a dextrocardia and PDA measuring 3 mm [Figure 2]. The abdominal wall defect measured 7 cm in size. A peripheral intravenous (I.V.) cannula was placed, and a maintenance infusion of isolyte P at 12 ml/h was started. Prophylactic antibiotics were given. Preoperative laboratory results included a glucose level of 79 mg/dL and hemoglobin of 15.5 g%. The patient was posted for surgery the following day. Informed consent was taken from the parents, and all associated risks were explained including the need for postoperative ventilation. In preparation for surgery, the room was warmed. Forced-air warming was placed, and intraoperative fluids were warmed. The standard American Society of Anesthesiologists monitors was attached. Invasive monitors were not placed as they were thought not to be necessary. Ringers lactate with 1% dextrose was started in the drip for replacing intraoperative losses. Fentanyl 6 mcg was given intravenously. The patient was intubated awake after giving propofol 10 mg bolus. The trachea was secured with a 3.0-mm uncuffed endotracheal tube. The patient was then relaxed using atracurium 1.5 mg

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Figure 1: Anterior abdominal wall defect painted with antiseptic solution to prevent abdominal sepsis



Figure 2: Chest X-ray revealing dextrocardia

intravenous and put on the ventilator in pressure mode to maintain normocapnia. Anesthesia was maintained with a mixture of oxygen, air, and isoflurane. Nitrous oxide was avoided. An infant feeding tube was passed to suck the gastric contents. The surgical procedure was smooth and uneventful lasting for around 90 min. On the return of the contents into the abdominal cavity peak, airway pressures increased but <10 cm H_2O , and hemodynamic stability was maintained throughout without any intervention. Primary closure was achieved easily. Paracetamol rectal suppository and local wound infiltration with 3 ml of 0.2% bupivacaine were given for postoperative pain relief. The baby with its trachea intubated was returned to the Neonatal Intensive Care Unit (NICU) where he was sedated and ventilated. The baby was extubated on the third postoperative day; orogastric feeding was resumed on the fourth postoperative day, and the baby was discharged in 3 weeks after ensuring he was able to suckle well.

DISCUSSION

Exomphalos and gastroschisis are common congenital anterior abdominal wall defects. Exomphalos has an incidence of 1 in 13000, whereas gastroschisis has an incidence of 1 in 6000–10,000. Up to 72% of neonates born with exomphalos have associated anomalies and cardiac anomalies comprise 20% of these. Tetralogy of fallot and atrial septal defects are the most common cardiac anomalies associated with exomphalos.^{3,5} Some syndromes associated with exomphalos are nonchromosomal disorders such as Beckwith–Wiedemann syndrome, pentalogy of Cantrell, Charge syndrome, Marshall–Smith syndrome, Goltz syndrome, fetal valproate syndrome, and chromosomal genetic disorders such as trisomies 13, 14, 15, 18, or 21.^{6,7}

These anomalies are routinely detected on antenatal ultrasound. Once exomphalos is identified search for other anomalies should be carried out. Exomphalos is not considered as an emergency unless the abdominal contents are exposed. Hence, such babies should be managed conservatively in the NICU with main emphasis on fluid resuscitation, care of the herniated bowel and its blood supply, bowel decompression using a nasogastric tube, and temperature regulation. Complete blood count and capillary blood gas analysis along with abdominal and chest X-ray must be performed. In neonates with exomphalos, an echocardiogram, kidney ultrasound, chromosomal analysis, and computed tomography head are performed to ascertain the presence of other associated anomalies. Cross-matched blood should be arranged for intraoperative transfusion if required.

Care of the newborn with abdominal wall defects begins with preoperative assessment and optimization. After stabilization of the newborn, the exposed abdominal contents should be covered with a warm sterile moist dressing to prevent temperature and fluid losses. These babies are prone to hypoglycemia; therefore, dextrose containing fluids should be given for maintenance. Hypoglycemia can be treated with 1–2 ml/kg of 10% dextrose. An orogastric tube should be passed to decompress the bowel. Even if these babies have not fed, they should be considered as full stomach because of the inflammatory exudates in the bowel secondary to exposure to the amniotic fluid in utero and air after birth the bowel may be distended and functioning abnormally. I.V. access should be obtained in the upper limbs as venous return maybe hampered after the return of bowel. All babies should receive appropriate antibiotics preoperatively to reduce the chances of intra-abdominal sepsis. Capillary blood gas analysis must be done. Serum electrolytes should be tested, and electrolyte imbalances should be corrected before surgery. Care must be

taken during manipulation to avoid trauma to the abdomen and its contents.

All neonates have a tendency for hypothermia more so in neonates with exomphalos because of the exposed bowel. Therefore, much emphasis should be on maintaining normothermia in these babies such as transfer to the operating theater in a warm incubator, ensuring theater temperature at least 27°C, placing the neonate on a warming mattress, covering the head to prevent loss of heat, infusing warmed fluids, and using forced-air warming devices intraoperatively.

Some anesthetic considerations of isolated dextrocardia include placing defibrillation paddles in reverse in case of cardiopulmonary resuscitation. The chest leads of electrocardiogram (ECG) should be reversed but not the limb leads if not the ECG tracings can erroneously display readings of intraoperative myocardial ischemia.⁸ Central venous catheterization if required should be inserted in the left internal jugular vein to gain direct access to the right atrium and avoid injury to the thoracic duct.

Routine use of invasive blood pressure monitoring is not required; however, patients with PDA are prone for coronary ischemia due to pulmonary runoff, especially in the diastolic phase of the cardiac cycle. Therefore, careful monitoring to prevent diastolic hypotension and adequate steps to maintain systemic vascular resistance in case of hypotension should be carried out which would include the use of vasopressors.

Neonates with exomphalos are prone to aspiration and regurgitation. Therefore, these babies should be intubated either awake or by rapid sequence induction. Nitrous oxide should be avoided during the maintenance to avoid bowel distention which can increase intra-abdominal pressure on returning bowel contents to the abdomen during closure. An oxygen air mixture should be used to maintain saturation around 90% to avoid the risk of retinopathy in neonates.

As the bowel is returned to the abdominal cavity, the rise in intragastric pressure may result in hypotension. Compression on the inferior vena cava can decrease preload. As the abdominal contents are pushed up into the thoracic cavity, the pulmonary compliance, and diaphragmatic excursion decreases thus oxygenation and ventilation may be compromised. Peak airway pressure should be monitored closely during this part of the surgery, especially when the hernial defect is larger than 4 cm. The increase in the intra-abdominal pressure can reduce renal blood flow; therefore, a close check on the intraoperative urine output should be ensured.

Main priorities in the postoperative period include monitoring peak airway pressure, looking for signs of increased abdominal pressure, infection control, temperature regulation, glucose monitoring, and fluid-electrolyte homeostasis regulation. Elective postoperative ventilation shall improve outcome in such babies as such babies often have some degree

of compromised respiratory function secondary to raised intra-abdominal pressure postoperatively. The patient will have some degree of postoperative ileus therefore, should be kept fasting till orogastric tube aspirate becomes minimal with the return of bowel sounds. Till then the patient can receive supplements through the parenteral route.

Next challenge is the high incidence of malnutrition and subsequent complications in the late postoperative period among such neonates. Therefore, it has to be ensured that the baby can suckle well before discharging them from the hospital. As most of the patients with these complaints belong to lower socioeconomic status and most of them find it financially difficult to come for regular follow-ups postoperatively, the parents have to be taught basic health care for a successful long-term outcome. We have lost this patient as they have not returned for follow-up after discharge. However, our team wishes to track this patient in the coming time to see the outcome of our efforts.

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Conflicts of interest

There are no conflicts of interest.

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