

# **Infant with Pentalogy of Cantrell and Tetralogy of Fallot requiring Omphalocele Repair**

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## **Objectives:**

- Discuss preoperative evaluation and anesthetic implications for a neonate with post natively diagnosed omphalocele requiring emergent abdominal surgery.
- Discuss the anatomy and physiology in patients with unrepaired tetralogy of Fallot.
- Describe appropriate consultation and perioperative care for a neonate with a history of unrepaired tetralogy of Fallot scheduled for omphalocele repair.

## **Case history:**

7 day old boy with post natal diagnosis of omphalocele who presented with respiratory distress at an OSH.

Born at 39 weeks 6/7 week to a 19 yo G1P0 mother, whose pregnancy was complicated by hypertension and obesity. At delivery, he is noted to have a moderate sized omphalocele. Despite APGARs of 7 and 8, respiratory distress is noted at delivery and he receives positive pressure ventilation x 2min. His oxygen saturations are 80-83% on 50% O<sub>2</sub>. His first capillary gas is 7.18/60.2/29.3/22.2/-7.1 on 70% FiO<sub>2</sub>. He is intubated, sedated, and placed on high frequency oscillatory ventilatory (HFOV) and inhaled Nitric Oxide and transferred to your hospital for further management and workup.

- 1) What is normal fetal circulation? What is transitional circulation? How does this affect your assessment of a neonate with respiratory distress at birth?
- 2) For a neonate with respiratory distress and circulatory collapse would you start IV Antibiotics? Prostaglandins? Why?
- 3) What are you concerned about? What is the difference between gastroschisis and omphalocele?
- 4) What are common abnormalities associated with the presence of omphalocele? What other syndromes are associated with omphalocele?

## **Case Presentation and Exam:**

His physical exam demonstrates a 3.1kg baby. Vital signs are HR 120 BP 53/39 SpO<sub>2</sub> 85%. His physical exam demonstrates an intubated sedated neonate. His extremities are warm and well perfused. His cardiac exam is significant for a loud systolic ejection murmur heard along the left

sternal border with. His lungs sounds are significant for ventilated breath sounds. The abdominal exam demonstrates a dressing over his abdomen.

- 1) What studies do you want to further assess the patient prior to going to the OR?

The baby is diagnosed with Pentalogy of Cantrell and Tetralogy of Fallot.

- 1) What is Pentalogy of Cantrell? How does this affect your preoperative evaluation of this patient?
- 2) In a critically ill neonate, what are you looking for on the babygram?
- 3) What is a normal Hct in a neonate? What is the circulating blood volume of a 3.1kg neonate? A premature neonate? Would you have the patient typed and crossed before starting the procedure? Blood in the room?
- 4) What is Tetralogy of Fallot? How does this affect your preoperative assessment and management of this patient? What does the term “Pink Tet” mean?

### **Preoperative preparation**

The patient is scheduled for omphalocele repair, possible anterior diaphragmatic repair, and sternal cleft repair. He is intubated and sedated. He currently has a 24g PIV.

- 5) Would you proceed with the case? What is your anesthetic plan?
- 6) What preoperative considerations do you have for a neonate coming for surgery?
- 7) What vascular access do you want? What monitors do you want?
- 8) What type of fluids would you run intraoperatively? How does neonatal renal and liver function affect your choice of intraoperative fluids?

### **Case Progression**

Prior to coming to the OR, you sedate and paralyze the patient with 5 mcg/kg of Fentanyl and 1mcg/kg of Cisatracurium and are able to easily ventilate the patient. Transport from the ICU to the OR is uneventful. While the surgeons are attempting to replace the abdominal contents, the patient becomes profoundly hypotensive and his oxygen saturations drop dramatically.

- 1) What are the physiological changes you anticipate occurring during omphalocele repair?2) What is the differential diagnosis? Could the patient be having a “tet spell”?
- 3) How are you going to treat the patient?
- 4) Despite your treatment, the patient continues to have low oxygen saturations and hypotension. What will you do now? Can you change ventilation strategy? What inotropes/vasopressors could you use?
- 5) Are you going to continue the case? What parameters will help you decide?

### **Post-operative care**

After a discussion with the surgeons, the decision is made to stop the case and transfer the patient to the ICU. The abdomen is closed and the patient is transported to the cardiac ICU. He is on Dopamine 7.5mcg/kg/min in a critical condition. During sign out, the respiratory therapist

suctions the patient who becomes hypotensive and his oxygen saturations began to drift down.

- 1) Now what is your differential? Do you think these are “tet” spells or something else?
- 2) Would you open the abdomen at the bedside? What other treatments would you consider?
- 3) Should the patient have gone for heart repair prior to abdominal surgery? Should this have been done at Day 7 of life? Earlier? Later? How would this potentially affect your anesthetic management or plan?

## Discussion:

Pentalogy of Cantrell is a rare congenital malformation with an incidence of 5.5 per 1 million live births. The syndrome is characterized by midline defects resulting from defective development in the septum transversum. The constellation of defects usually includes: 1) a midline, supraumbilical abdominal wall defect or omphalocele; 2) a defect of the lower sternum; 3) an anterior (intrapericardial) diaphragmatic hernia; 4) a defect in the diaphragmatic pericardium, and 5) congenital intracardiac defects. In severe cases, the heart may herniate through the diaphragmatic defect causing ectopia cordis. In a review by Vazquez-Jimenez et al of 153 cases, heart defects were present in 83%, abdominal wall defects in 74.5%, sternal malformations in 59.4%, diaphragmatic defects in 56.8%, and pericardial defects in 41.8% of patients. The intracardiac lesions may include ASDs (34.7%), VSDs (72%), Tetralogy of Fallot (17.3%) (TOF), double outlet right ventricles (DORV) (1.2%), anomalous pulmonary venous connection, transposition of the great arteries (TGA) (6.3%), tricuspid atresia (5.5%), and truncus arteriosus (3.9%). Pentalogy of Cantrell is associated with a low survival rate, less than 40% depending on the severity of the defects, with only 5-10% survival in patient with severe ectopia cordis.

Tetralogy of Fallot (TOF) is characterized by four components 1) larger ventricular septal defect (VSD), 2) right ventricular outflow obstruction, 3) overriding aorta, and 4) right ventricular hypertrophy (RVH). The RVOT obstruction can have a range of mildly hypoplastic pulmonary valve to complete atresia. The valvular obstruction is a *fixed* obstruction, but the subvalvular obstruction is *dynamic*. The dynamic subvalvular obstruction is what causes the hypoxic spells or “tet spells” in patients with TOF/PS and infundibular obstruction. In brief, hypoxia during feeding, crying or anxiety induces an increase in pulmonary vascular resistance, which worsens the right-to-left shunt through the VSD. As cardiac output and preload decrease, tachycardia and an underfilled RV result in worsening infundibular spasm and worsening of the right-to-left shunt. The goal of treatment is to decrease PVR and increase SVR so as to induce a left-to-right shunt.

Treatment includes:

- Administration of 100% O<sub>2</sub>
- Increase RV filling and preload with fluid boluses or by placing the patient in a knee-to-chest position (traditionally seen as squatting in older unrepaired Tetralogy of Fallot patients)
- Administration of morphine or deepening the anesthetic to relax the patient and infundibular spasm
- Phenylephrine (5-10mcg/kg IV) to increase SVR and reduce R-to-L shunting
- Propranolol or esmolol may reduce infundibular spasm by depressing contractility and decreasing heart rate
- Sodium Bicarbonate to treat severe metabolic acidosis during a tet spell

Omphalocele is a midline abdominal wall defect with extrusion of abdominal viscera, covered by a membranous sac, into the base of the umbilical cord. Gastroschisis is a defect in the anterior

abdominal wall typically located to the right of the umbilical ring and resulting in the herniation of the gut and occasionally of the urogenital tract, without a surrounding membrane. While both abnormalities are associated with other congenital malformations, the frequency of other abnormalities associated with omphalocele is 27-63% and 5-27% for gastroschisis.

Management for omphalocele repair includes meticulous attention to volume replacement, covering the mucosal surfaces with sterile, saline-soaked dressings to minimize evaporative and heat losses, and a rapid sequence induction for intubation. As abdominal closure is attempted, it is important to monitor for 1) decreased perfusion to abdominal organs, 2) decreased ventilation/oxygenation, and 3) decreased venous return. Impaired organ function/damage may lead to decreased drug metabolism, lactic acidosis, and renal congestion. It is important to monitor for UOP, lactic acidosis, and ventilator changes as well as electrolyte abnormalities that may develop as the abdomen is closed. In patients who have large omphaloceles, the replacement of abdominal contents may lead to mechanical obstruction to the IVC and subsequent decreased venous return, lower body edema and lactic acidosis. In these cases, the reduction of abdominal contents may be done in a staged procedure to allow for the body to adapt.

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