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Anesthetic management of a patient with Fontan circulation undergoing surgery for correction of spinal stenosis

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Keypoints

- The importance of understanding hemodynamics of a fontan patient for non-cardiac surgery.
- The special considerations for spine surgery in congenital heart disease children.
- The importance of intraoperative monitoring and management of fontan patients going for major spine surgery.

Abstract

Patients with Fontan circulation have increased cardiac and pulmonary risks. In spinal surgery these risks are increased given the nature of the surgery and patient positioning. In this case report, we describe the anesthetic technique used in an 8-year-old female with congenital heart disease and restrictive lung disease requiring spinal surgery for severe scoliosis. We will discuss our management of this complex patient including choice of general anesthetic, hemodynamic goals, ventilator settings, and inherent risks of this procedure.

Keywords

Fontan Procedure; congenital heart defect; congenital scoliosis; Intravenous anesthetics; hemodynamic monitoring, respiratory function testing

Introduction

The link between congenital heart disease (CHD) and scoliosis is currently well known, with idiopathic scoliosis occurring in 2-4% of the general population (1, 2). Spinal fusion with instrumentation is used to stop progression of deformities. Advances in pediatric anesthesia and surgery have led to increased survival of CHD patients. Surgery is indicated for severe back pain or cardiopulmonary restriction despite adequate medical therapy (1). These patients pose an interesting challenge to anesthesiologists due to their limited cardiac and pulmonic reserve.

Fontan circulation is the final stage of palliative surgery in patients with single-ventricle lesions. Fontan physiology is complex and requires meticulous planning to decrease complications. In Fontan circulation, deoxygenated venous blood returning from the IVC and SVC is directed to the pulmonary artery by means of a cavopulmonary shunt, thus bypassing the right ventricle which is functioning as a single ventricle (3). For proper flow, pulmonary vascular resistance must be low for blood to flow through the Fontan conduit and pulmonary arteries; thus, this transpulmonary flow determines cardiac output. This cardiac physiology along with a multitude of other factors make these patients a challenge to manage in the

operating room. Herein, we discuss the anesthetic management of a patient with a Fontan conduit requiring posterior spinal fusion for idiopathic scoliosis.

Case report

An 8-year-old female, with multiple medical co-morbidities, presented for posterior spinal fusion with segmental instrumentation, shilla technique and autograft and allograft of bone. Cardiac history included heterotaxy syndrome with right atrial isomerization, unbalanced complete AV septal defect, double outlet right ventricle, D-malposed great arteries, pulmonary stenosis, and total anomalous pulmonary venous return (TAPVR). She also had scoliosis with thoracic lumbar curvature and right lumbar curvature. Patient had undergone several cardiac surgeries including the blalock procedure, bilateral Glenn shunts with Blalock-Tausig Shunt takedown, Fontan procedure at 4 years with TAPVR connection repair and left sided AV valvuloplasty 3 months after Fontan. Preoperatively, stress testing, pulmonary function testing, and echocardiogram testing were all normal. Additionally, there was no obstruction across the neoaorta, Fontan or Glenn pathways. Laboratory studies showed a hematocrit of 32.8%, hemoglobin 11g/dl, and platelets 167 000/ uL.

In the operating room, standard ASA monitoring devices were used. She was induced with sevoflurane and intubated with a 5.0 endotracheal tube. PEEP was not applied. Peripheral IVs, arterial line, and an internal jugular central line were placed for access and central venous pressure (CVP) monitoring. She was positioned in prone position without respiratory compromise. Total intravenous anesthesia (TIVA) with dexmedetomidine, remifentanil, and propofol were used. No muscle relaxation was used to allow for neuromonitoring. Transaxemic acid, cefazolin and gentamycin were given. Fentanyl, ketamine, and intrathecal morphine were used for analgesia. She was hemodynamically stable with CVP fluctuating from 10-20 mmHg. A unit of packed red blood cells was administered due to blood loss. She was extubated with intact neurological function. Post-operative chest x-ray revealed a small right sided pneumothorax; further, labs were within normal limits. She was weaned to room air from 2L nasal canula without intervention for the pneumothorax.

Discussion

The surgical treatment of scoliosis after a Fontan procedure is challenging due to the risk of various complications. The surgery allows for several post-surgical complications including pleural effusions, pneumothorax and coagulopathies. Maintaining blood volume is required during these surgeries. Herein, the combination of dexmedotomidine, remifentanyl, and propofol provided adequate anesthesia without hemodynamic compromise with MAPS greater than 70s.

Another variable that was considered was pulmonic physiology. Impairment of respiratory function due to restrictive pulmonary disease has been reported in normal children with early scoliosis (4, 5). The patients may eventually have pathophysiological changes leading to increased pulmonary vascular resistance, increased arterial pressures, and cor pulmonale (5) with negative effects on cardiac output and systemic circulation. Intraoperatively, one must avoid causes of increased pulmonary vascular resistance including hypoxia, hypercarbia, acidosis, hypothermia, and PEEP. Our patient was hyperventilated prior to incision and hypothermia was avoided by using bair huggers.

Decreasing the duration of the surgery is paramount in decreasing complications. The lengthier the case, with prolonged corrections, the greater the blood loss, and increased adverse outcomes. Our patient had blood loss requiring transfusion of packed red blood cells, FFP, and crystalloid. Factors correlating with increased blood loss include surgical technique, duration of surgery, number of vertebral levels fused, site of autologous bone graft harvest, MAP, and positioning. Patient had a small right lung pneumothorax complication from surgery; however, she was asymptomatic without intervention. Although she had restrictive lung disease, she was medically optimized prior to surgery with improvement in her pulmonary function tests. This helped prevent an otherwise

prolonged hospital course with the pneumothorax, or risk of severe respiratory complications.

Conclusion

Patient presented challenges from hemodynamic and respiratory perspectives. The Fontan circulation, lung disease, and scoliosis surgery necessitated tailored anesthesia to avoid end-organ hypoperfusion while ventilation was set to avoid adverse events. The combination of careful surgical technique planning, and anesthetic considerations was key in decreasing surgical complications and increase success.

List of abbreviations used in the manuscript:

AV Septum: Arterio-Ventricular Septum
CHD: Congenital Heart Disease
CVP: Central Venous Pressure
FVC: Forced Vital Capacity
FEV1: Forced Expiratory Volume 1
FFP: Fresh Frozen Plasma
ICU: Intensive Care Unit
IVC and SVC: Inferior Vena Cava and Superior Vena Cava
TAPVR: total anomalous pulmonary venous return
MAP: Mean Arterial Pressure
PEEP: Positive End-Expiratory Pressure
TIVA: Total Intravenous Anesthesia

Ethics: There was no ethical approval needed for this case report.

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