

Posterior Spinal Fusion Procedure in a Pediatric Patient with Congenital Heart Disease

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Introduction: Posterior spinal fusion and instrumentation procedures are indicated for pediatric patients with severe spinal curvature who may exhibit restrictive respiratory physiology and compromise secondary to the scoliosis. These patients are usually healthy apart from their underlying skeletal abnormality, the majority of which are idiopathic in nature. Anesthetic considerations for these patients include the implications of prone positioning, the anticipation of large intraoperative blood loss and the role of deliberate hypotension, neuromonitoring SSEPs and MEPs, and the anticipation of the intraoperative wake-up test. On the other hand, in a pediatric patient with severe cardiac and pulmonary pathology, the anesthetic implications differ significantly.

Case Report: A 15 year old 38 kg female with severe S-shaped congenital scoliosis and resultant restrictive lung disease and chronic respiratory insufficiency requiring nasal cannula oxygen at baseline and BiPAP overnight presented for posterior thoracolumbar spinal fusion and instrumentation. Other significant past medical history included a history of cor triatriatum status post repair, left pulmonary artery hypoplasia, left pulmonary vein stenosis, moderate pulmonary hypertension, anomalous left superior vena cava to coronary sinus connection, status post tethered cord release six months previously, reactive airway disease, and a left solitary malrotated kidney.

Two months prior to surgery, the patient was admitted for increased work of breathing despite her very limited activity level. A multidisciplinary care conference was held, presenting to the patient and her family the substantial risks involved and the potential limited benefits of spine surgery. Because the patient felt that her quality of life was deteriorating and was highly motivated to attempt anything that would reverse her progression towards cardio-respiratory failure, the patient and her family opted to proceed. The day prior to surgery, the patient was admitted for preoperative preparation, including lab work, a preoperative ECG, and echocardiography. On physical exam, the patient had decreased breath sounds on the left compared to the right lung, occasional expiratory wheezing, and minimal chest excursion on the left. Heart sounds were regular rate and rhythm, normal S1 and S2, and no murmurs were auscultated. Of note, the patient had a short neck and her lateral range of motion was significantly limited.

On the day of surgery, the patient was effectively premedicated with intravenous midazolam. In anticipation of an acute pulmonary hypertensive crisis, nitric oxide and epoprostenol were available. With standard monitors in place, and after proper positioning and preoxygenation, the patient was induced with fentanyl, etomidate, and rocuronium, and intubated without difficulty. A right femoral arterial line was placed, as well as two large bore peripheral IVs. In addition, an introducer was placed in the right internal jugular vein under SiteRite guidance, and a pulmonary artery (PA) catheter was placed by the cardiologist under direct fluoroscopy. The patient was turned prone and positioned appropriately. Anesthesia was maintained on remifentanyl and propofol infusions, titrating to mean arterial pressures (MAP) in the 60s, within 20% of baseline. Systolic PA pressures remained 50-60% of systemic BP in the 50s/30s for the duration of the case. There were no difficulties with ventilation or oxygenation. Renal dose dopamine was started early to aid in perfusion to her solitary kidney. MEPs and SSEPs were intact throughout the case. Because the patient had been on sildenafil, bosentan, and aspirin for treatment of her pulmonary hypertension, blood loss was more than usual. Acute normovolemic hemodilution and deliberate hypotension were not undertaken because of her cardio-respiratory disease, and the hematocrit was maintained at 40-45%. At the completion of the procedure, the patient was turned supine and emerged from general anesthesia for the wake-up test. After moving bilateral lower extremities, the patient was reseeded and brought to the intensive care unit intubated and paralyzed, BP 120/65, PAP 60/30, HR 105, SpO2 100%. During her admission to the intensive care unit, the patient's MAP and PAP were stable and the patient was weaned to BiPAP and extubated on post-operative day #5 and transferred to the ward. On post-operative day #9, the patient was discharged home in stable condition. However, at two months post surgery, she still requires BiPAP.

Discussion: Anesthesia for scoliosis surgery can be a real challenge when caring for the healthy patient. Add concomitant respiratory and cardiac problems, and the anesthetic management is significantly modified. In this patient with a complex medical history, the anticipation of an acute crisis precipitated multidisciplinary involvement and early planning. Although there exists various modifications to anesthetic management for this patient, the plan presented illustrates one option such that the outcome was favorable for all parties involved, most importantly for the patient.