Anesthetic management of a child with neonatal Marfan syndrome

Author(s): Tatiana Kubacki MD, Scott Streater MD, Johanna Schwarzenberger MD, David Roye MD, Thomas Starc MD, Stephanie Levassuer, Andrei Constantinescu MD, Lena Sun MD

Affiliation(s): Children’s Hospital of New York, Department of Anesthesiology and Orthopedic surgery, Columbia University, New York, New York

Introduction: Marfan Syndrome (MS) is an autosomal dominant disorder of fibrous connective tissue due to mutation in fibrinulin-1 gene, located on chromosome 15. The distinctive neonatal phenotype is at the severe end of the clinical spectrum of MS and characterized not only by the facial and skeletal features, but also by the severity of cardiac involvement. We present the perioperative management of a 23 month-old, 10 kg child with neonatal Marfan syndrome (nMS) associated with severe and progressive scoliosis for expansion thoracoplasty. The patient had dilated pulmonic and aortic root with tricuspid and mitral insufficiency. He underwent mitral valve repair at 10 months of age without complications, but continued to have moderate to severe mitral regurgitation. Despite losartan therapy, his aortic root continued to dilate and was measured at 39 mm (normal range 1.16 – 1.79 mm) with Z score of 15.53 preoperatively. His pulmonary function was reduced due to progressive restrictive disease from scoliosis and pectus carinatum, and reactive airway disease. Flexible bronchoscopy performed preoperatively showed significant deformity and narrowing of the left lower lobe bronchus, with thick mucous secretions in the right upper and the right lower lobe, severe distortion and narrowing of right middle lobe and the right lower lobe bronchi. A preoperative CT scan of the chest revealed a complex S-shaped scoliosis of the thoracic spine, severe pectus carinatum, an enlarged heart situated in the right hemithorax, compressive atelectasis of the right middle lobe, and patent central airways. Prior to his scheduled surgery, a multidisciplinary conference was convened between orthopedic surgery, anesthesiology, pediatric cardiology, pulmonology and critical care teams. Specific intra- and post–operative care plans were formulated with detailed discussion of overall perioperative risk.

On the day of surgery the vital signs were BP 90/52, HR 104 and oxygen saturation 94% on room air with a respiratory rate of 32 breaths per minute. Anesthesia was induced with sevoflurane by inhalation, followed by tracheal intubation. Intraoperative monitors included CVP via the femoral vein, radial arterial line and standard ASA monitors. In addition, transesophageal echo (TEE) was used to monitor hemodynamic changes during positioning and surgery; and SSEP/MEP were used as part of routine monitoring during scoliosis surgery. Flexible fiberoptic bronchoscopy was performed to evaluate for distal airway compression and possible mucous plugging prior to positioning. At that point segmental bronchi appeared distorted but otherwise patent without visible secretions. Anesthesia was maintained with continuous infusions of remifentanil, propofol, and intermittent boluses of rocuronium. The patient was carefully padded with custom made foam pads and placed into prone position while being monitored with TEE. The procedure lasted 100 minutes and was uneventful with minimal blood loss. To optimize post-operative pain management, 0.25% bupivacaine with clonidine 1 microgram/kg was injected locally by the surgeon. The patient remained hemodynamically stable throughout surgery with adequate urine output. After meeting extubation criteria, the patient was uneventfully extubated to face mask oxygen. He received additional intravenous morphine and was transferred to the intensive care unit for recovery. Intravenous PCA was prescribed for post-operative pain control. The patient’s postoperative course was complicated by a single episode of respiratory distress requiring BiPAP, persistent right sided atelectasis and difficulties with pain control due to a dislodged lumbar hook. On POD # 7 he presented for revision surgery with additional instrumentation which was uneventful.

Discussion: Affected individuals with nMS often die within the first months of life from cardiopulmonary complications and do not survive beyond 24 months. In this context, our patient with nMS had all of the severe manifestations of cardiac involvement including aortic root dilatation, mitral...
valve prolapse and insufficiency, and he was at the end of life expectancy for this disease. Although expansion thoracoplasty involves only superficial instrumentation and is generally not as extensive as instrumented posterior spinal fusion, we were concerned about potentially serious complications in this patient with a large aortic aneurysm at constant risk for dissection or rupture. It was unclear whether the patient with severe scoliosis, pectus carinatum and enlarged cardiovascular structures could tolerate prone positioning without hemodynamic compromise. Intra-operative TEE allowed online monitoring of possible cardiovascular compression after positioning, assessment of contractility, cardiac output, valvular function and volume status during surgery. Since his pre-operative bronchoscopy demonstrated significant deformity and narrowing of segmental bronchi with mucous retention in the dependent lung fields, it was important to repeat it in the OR setting to prevent any possibility of mucous plugging. We took special care to avoid pressure on the sternum from transverse bolsters by using specially prepared foam pads while positioning this patient with severe chest deformity. A preoperative interdisciplinary meeting was instrumental in defining all perioperative issues including possible catastrophic outcome due to aneurysm rupture.

References: