Minimizing Morbidity and Maintaining Mobility in Morquio

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INTRODUCTION

Morquio syndrome, also known as mucopolysaccharidosis IV, is a rare and progressive lysosomal storage disease. Autosomal recessive inheritance of a deficiency in enzymes that typically catabolize glycosaminoglycans result in abnormal accumulation throughout the body. There are neuromuscular manifestations such as dwarfism, kyphoscoliosis, atlanto-axial instability, and spinal cord compression. Cardiopulmonary considerations include cardiomyopathy, myocardial ischemia, valvular dysfunction, and restrictive lung disease. The airway can be complicated by limited mouth opening, macroglalia, tracheal stenosis, friable tissue, and sleep apnea. Cognitive function is fully intact. These patients often undergo orthopedic, ENT, and cardiac surgeries, and they are considered high risk for general anesthesia. [1,2,3]

PROCEDURE

On the day of surgery, the patient was induced and intubated with video laryngoscopy and minimal cervical manipulation. He was maintained on total intravenous anesthesia – propofol, dexmedetomidine, and remifentanil – throughout the surgery to optimize the evoked potential monitoring. Continuous somatosensory evoked potentials and intermittent motor evoked potentials monitored for any signs of spinal cord ischemia. There was never any change from baseline. A mean arterial pressure of 50 to 60 mmHg was sustained during cardiopulmonary bypass. He was placed on a dopamine infusion at the time of aortic cross clamp, to provide vasopressor support. A milrinone infusion was added prior to transitioning off bypass for inotropic support.

The patient was emerged in the operating room with neuromonitoring, and he demonstrated movement in all four extremities on command. He was weaned off bypass for five. He was ambulating well and experienced no neuropathy. After his surgery, the patient started college as a full-time student.

DISCUSSION

We present this case report of a patient with Morquio syndrome who underwent a successful aortic valve replacement with intraoperative neuromonitoring to shine a spotlight on a rare disease with profound anesthetic implications. Morquio syndrome has an incidence of about one in 200,000 to 300,000 live births. [4]

Although a recently approved enzyme replacement therapy with weekly Vimizim infusions has shown some promise in symptom mitigation, the standard of care continues to include a litany of palliative surgical interventions. [4,5] This case adds to the body of literature that increasingly recommends monitoring for spinal cord integrity during anesthesia.

REFERENCES


PATIENT

An eighteen year old male with Morquio syndrome and aortic valve insufficiency was scheduled for aortic valve replacement. His history was notable for scoliosis and hip dysplasia palliated by thoracolumbar spinal fusion and periactetabular osteotomy. He had experienced an episode of cervical myelopathy from sneezing, with numbness and pain in his hands that persisted for over a week. His risk for spinal cord ischemia would be further compounded by cardiac surgery on cardiopulmonary bypass, and quadriplegia was a possible adverse outcome. We coordinated a multidisciplinary plan to provide intraoperative neurophysiological monitoring from intubation to extubation, and most critically during bypass.