



# WAKE UP SAFE®

*The Pediatric Anesthesia Quality Improvement Initiative*

## **Spinal Cord Infarction or Injury in Patients with Mucopolysaccharidoses (MPS)**

### **Introduction**

Patients with mucopolysaccharidosis present unique challenges to anesthesiologists, of which difficult airway and cervical spine instability are well known. Wake Up Safe has received two reports of paralysis after combined general and epidural anesthesia in a patients with Hurler syndrome. Similarly, a case report of spinal cord infarction in a patient with Morquio syndrome was recently published. This patient also had received combined general and epidural anesthesia. All three patients were having lower extremity orthopedic procedures.

### **Discussion**

There are reports of neurologic deficits in patients with MPS undergoing anesthesia (Drummond, 2015; Pruszczynski, 2015; Tong, 2012). Furthermore, it is likely there are additional cases, which have not been published. In some instances the spinal cord insult is associated with an area of focal kyphoscoliosis. Cadaveric studies have shown that spinal cord intramedullary pressures greatly increase when thoracic kyphosis is  $> 63^\circ$ , increasing the risk for spinal cord ischemia (Farley, 2012). However, given limited case reports it is uncertain what the critical threshold is for the degree of the curve.

### **Conclusion**

Particular care must be taken to ensure spinal cord perfusion in patients with MPS undergoing general anesthesia, as factors that are tolerated by normal patients may be poorly tolerated by MPS patients. Such factors include kyphotic curves of the spine, neuraxial anesthesia, and low MAP. Consider neuromonitoring when patients are to undergo long procedures and also when kyphotic curves are present.

### **Recommendations:**

- Avoid epidurals/neuraxial blocks
- Consider preoperative X-Ray/MRI in patients with kyphosis
- Consider IONM for non-spine procedure when kyphosis is present or if the procedure is of longer duration.
- Special considerations if there is high potential for blood loss during the procedure, or positioning issues.
- Careful positioning and maintain spinal cord perfusion

### **References**

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