A Challenging Case Report of an Obstetric Patient with Morquio Syndrome and Polyarticular Onset Juvenile Rheumatoid Arthritis

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Introduction: Morquio syndrome is a rare autosomal recessive disease resulting in the accumulation of mucopolysaccharide in the soft tissue of the airway. It is clinically manifested by prognathism, a broad mouth, macrocephaly, short stature and trunk dwarfism creating the potential for a challenging airway (1). Polyarticular juvenile rheumatoid arthritis (PJRA) can complicate airway management because it may result in limited mouth opening, micrognathia, atlantodens subluxation and even cervical spine instability (2). We present a successful anesthetic in a parturient with both Morquio syndrome and PJRA for cesarean section (C/S).

Case: A 39 year old primigravid patient with history of Morquio syndrome and PJRA presented for C/S at 37 weeks gestation. Her examination revealed a 30.8kg and 125cm tall woman with truncal dwarfism, macrocephaly, a short neck, maxillary prognathism, micrognathia, limited TMJ mobility, decreased incisor distance and thyromental space and a Mallampati class IV airway. Her history was significant for a hypocoagulable state which precluded neuraxial anesthesia. At her preoperative anesthetic evaluation at 28 weeks gestation, a plan for an awake fiberoptic intubation was decided upon. This plan was followed, and after adequate airway topicalization the fiberoptic bronchoscope was used to place a 5.5mm endotracheal tube. General anesthesia was then induced followed by the delivery of her child. She was extubated without incident at the end of the case.

Discussion: The anesthetic management of this parturient presented several challenges given her complex medical history and difficult airway. An awake fiberoptic intubation avoiding hyperextension with a 5.5mm endotracheal tube was the safest method given her size, facial features, possible cervical instability and mucopolysaccharide deposition in her airway. This case highlights the complexities of anesthetic care in a patient with both Morquio syndrome and PJRA and also provides an example of how appropriate peripartum planning between the obstetric and anesthesia care teams can result in optimal patient care.

References: