

Anesthetic management of a neonate with metatropic dwarfism and severe C-spine instability

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Introduction:

Metatropic dwarfism is a rare genetic disorder characterized by short stature, short extremities, kyphoscoliosis and instability of the cervical spine (CS).¹ The following is a case report of the anesthetic management of a neonate with metatropic dwarfism.

Case Report:

A term newborn with metatropic dwarfism was scheduled for MRI of the CS to assess the severity of the atlantoaxial instability. Associated co-morbidities included severely deformed extremities, kyphoscoliosis and bilateral pulmonary hypoplasia. Immediately after birth the head and shoulder were strapped to a firm board to stabilize the CS. Following intravenous induction and insertion of a #1 Laryngeal Mask Airway (LMA), the MRI revealed severe odontoid hypoplasia with atlantoaxial subluxation and diffuse spinal stenosis. A C-collar was subsequently placed to stabilize the CS. A month later the infant was scheduled for Nissen fundoplication and gastrostomy tube placement secondary to failure to thrive. After placing standard ASA monitors and premedication with atropine and fentanyl and confirmation of easy mask ventilation, anesthesia was induced with a continuous propofol infusion without compromising spontaneous breaths. Multiple attempts to intubate the trachea using fiberoptic bronchoscope (FOB) were unsuccessful due to rapid desaturation in spite of supplemental O₂ while attempts to lighten the depth of anesthesia resulted in the patient gagging and coughing. Hence a decision was made to attempt intubation of the trachea through a #1 LMA.² Following easy placement of the LMA, the FOB could not be maneuvered through the glottis. The LMA was left in place to maintain ventilation. Then the FOB loaded with a 3 mm ID endotracheal tube (ETT) was inserted nasally and navigated to the periglottic area. The LMA was withdrawn slightly without compromising airway patency and ventilation and the tip of the FOB was navigated around the cuff of the LMA into the glottis. The LMA was removed and the ETT tube was advanced over the FOB into the trachea. The stability of the CS was maintained throughout the procedure with the help of the C-collar. Following uncomplicated surgery and mechanical ventilation in the immediate postoperative period, the infant's trachea was extubated the following day. There were no apparent changes in the neurological examination.



Discussion:

Rapid desaturation, in spite of spontaneous respirations (possibly because of altered respiratory mechanics with deep sedation and preexisting pulmonary hypoplasia) resulted in an inadequate time window for successful initial fiberoptic bronchoscopy. The attempt to intubate the trachea through the LMA was unsuccessful due to the acute angle between the point of exit in the LMA and the glottis. We decided to attempt nasal FOB with the LMA in place. Using this innovative method ventilation could be maintained thus ensuring adequate time for successful FOB. Once the tip of the scope was in the periglottic area the LMA was withdrawn just enough to allow the tip of the FOB into the glottis while ventilation was still maintained. In our opinion the decision to maintain assisted ventilation using the LMA ensured adequate time for successful intubation using a FOB.

References:

1. Anesthesiology 1990; 739-759
2. Paediatr Anaesth. 1999; 460-2.