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Infantile Systemic Hyalinosis (ISH) is a rare autosomal recessive disease that normally presents itself within the first few months of life. Recent genetic breakthrough has identified the gene that encodes for capillary morphogenesis protein 2 (CMG2) as the affected site of mutation in these patients². Characteristically, patients suffer from thickened and nodular skin, hypertrophic gingiva, hypotonia, and profound limitation of body movement secondary to the diffuse deposition of hyaline^{3,5}. Additional concerns include osteoporosis, contractures of multiple joints, malnutrition and growth failure⁴. Of note these joint contractures may make airway management challenging as a result of cervical spine (c-spine) and TMJ movement limitations. Though physically handicapped, these children are intellectually normal. This case of a 10-month-old male serves to highlight those anesthetic implications that should be of concern for the anesthesiologist presented with the challenge of managing such a patient.

A 10-month-old 5.3 kg male presented for insertion of a tunneled venous catheter for delivery of antibiotics and nutrition. His past medical history included infantile systemic hyalinosis with chronic diarrhea and failure to thrive. On physical exam, he suffered from diffuse contractures and subsequent limited movement of all limbs, micrognathia with limited head, mouth and neck movement secondary to temporo-mandibular joint contracture. With these considerations, the anesthesiologist's choices for management of the airway are critical.

The patient was brought into the operating room, all standard ASA monitors were applied and induction was completed using sevoflurane with 100% oxygen. We avoided the use of nitrous oxide for induction to maintain adequate preoxygenation while maintaining spontaneous respirations. A fiberoptic bronchoscope was in the room and ready for use. The patient was intubated with a Miller 1 laryngoscope successfully and the anesthetic was maintained with sevoflurane. The case was successfully completed and patient was extubated and returned to the PICU for further management.

The options for airway management are dependent on the degree of involvement of head and neck joints in regards to limitation of movement. Other options that have been described in the literature for anesthetic management ISH includes nasal airway use with concomitant oral fiberoptic intubation, nasotracheal intubation when oral intubation is impossible, and use of an LMA if mouth opening allows and if it is appropriate for the surgical procedure⁴. Inhalation induction is considered safest with maintenance of spontaneous respirations.




