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Difficult Airway Management in a Patient with Achondroplasia

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INTRODUCTION

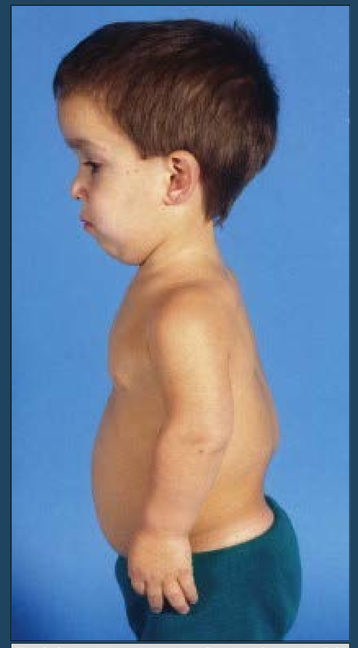
Achondroplasia is the most common type of disproportionate dwarfism.[1] It is caused by a mutation in the Fibroblast Growth Factor Receptor 3 gene.[2] The resultant decrease in the rate of endochondral ossification coupled with normal membranous ossification produces short tubular bones.[1] Transmission occurs in an autosomal dominant pattern, though up to 80% of cases occur through spontaneous mutations.[1]

CASE DESCRIPTION

A sixteen-year-old girl with achondroplasia presents for tibial and fibular osteotomies as treatment for genu varum deformities. She stood 118.6 cm tall and had a body mass index of 49. Preoperative intravenous access was difficult and unsuccessful, so inhalation induction was performed with sevoflurane. Shortly after induction, the patient's airway became obstructed, and mask ventilation was inadequate despite the insertion of an oral airway and two-person ventilation. While intravenous access was obtained, a laryngeal mask airway was placed with subsequent easy ventilation confirmed by the return of end tidal CO_2. A secured airway was necessary so the patient received fentanyl and propofol to facilitate intubation. Due to limited neck extension along with concerns for possible undiagnosed atlantoaxial instability, laryngoscopy was obtained using a GlideScope #4 with successful endotracheal intubation. The remainder of the general anesthetic course was uneventful, with no postoperative complications.

Table 1
CLINICAL FEATURES OF ACHONDROPLASIA

Macrocephaly
Protruding forehead
Saddle nose
Short maxilla
Large tongue
Prominent mandible
Shortened skull base
Foramen magnum stenosis
Atlantoaxial instability
Kyphoscoliosis
Short stature
Spinal stenosis
Rhizomelia
Genu varum
Ligamentous laxity
Excess skin and subcutaneous tissue



Facial appearance of a boy with achondroplasia. Note the frontal bossing and apparent enlargement of the head, especially in relation to limb size.[3] Copyright © 2014 by Saunders / Elsevier Inc.

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DISCUSSION

Patients with achondroplasia have characteristic physical features and deformities that have important anesthetic implications (Table 1). General considerations include airway management, recognition of altered respiratory function, ease of peripheral access, and proper patient positioning.

Airway management is challenging due to craniofacial abnormalities, which can lead to inadequate face mask ventilation from suboptimal mask fit and limit airway axis alignment for endotracheal intubation.[4] Neck hyperextension is avoided because the risk of compressing the medulla or upper cervical spinal cord through a stenotic foramen magnum may have disastrous outcomes.[5] Atlantoaxial instability from a ligamentous or bony abnormality can cause spinal cord or nerve root impingement.[6] Obstructive sleep apnea is a common complication in achondroplasia which predisposes patients to upper airway obstruction upon induction of anesthesia.[1]

Respiratory function is altered due to a small ribcage, obesity, and kyphoscoliosis.[5] The decreased chest wall compliance and diaphragmatic excursion may impair lung expansion; patients are at risk for a number of complications during the perioperative period including atelectasis, pneumonia, and airway obstruction.

Other considerations for patients with achondroplasia include proper positioning and peripheral venous access. Ligamentous laxity leads to loose joints, especially in the knees, that requires care in patient positioning.[5] At the same time, some patients will have flexion or extension deformities warranting extra padding on the operating table.[2] Excess skin and subcutaneous tissue may cause difficulty in obtaining intravascular access which can be distressing for patients preoperatively.[1]

CONCLUSION

Anesthetic management of patients with achondroplasia requires recognition of common clinical features and anticipation of potential airway difficulties.