Difficult Airway Management in a Patient with Achondroplasia

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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 CO2. A secured airway was necessary so the patient received fentanyl and propofol to facilitate induction. The remainder of the general anesthetic course was uneventful, with no postoperative complications.

### Introduction

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

### Case Description

**Achondroplasia** is a genetic disorder that affects growth and development. It is characterized by short stature, short arms and legs, and several other physical features, such as a prominent forehead, saddle nose, and large tongue. Patients with achondroplasia may also have other conditions, such as spinal stenosis, which can cause pressure on the spinal cord and lead to nerve damage.

**Clinical Features**

- **Facial appearance:** Prominent forehead, saddle nose, and large tongue.
- **Skin and subcutaneous tissue:** Excess skin and subcutaneous tissue may cause difficulty in obtaining intravascular access.
- **Ligamentous laxity:** Loose joints, especially in the knees, that requires care in patient positioning.
- **Other considerations:** Proper positioning and peripheral venous access. Ligamentous laxity leads to loose joints, especially in the knees, that requires care in patient positioning.

**References**


**Table 1**

<table>
<thead>
<tr>
<th>ACHONDROPLASIA</th>
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<tbody>
<tr>
<td>Macrocephaly</td>
</tr>
<tr>
<td>Protruding forehead</td>
</tr>
<tr>
<td>Saddle nose</td>
</tr>
<tr>
<td>Short maxilla</td>
</tr>
<tr>
<td>Short mandible</td>
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<td>Ectodermal dysplasia</td>
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<td>Hypertelorism</td>
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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. 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. Atlantoaxial instability from a ligamentous or bony abnormality can cause spinal cord or nerve root impingement. Obstructive sleep apnea is a common complication in achondroplasia which predisposes patients to upper airway obstruction upon induction of anesthesia.

Respiratory function is altered due to a small ribcage, obesity, and kyphoscoliosis. 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. At the same time, some patients will have flexion or extension deformities warranting extra padding on the operating table. Excess skin and subcutaneous tissue may cause difficulty in obtaining intravascular access which can be distressing for patients preoperatively.

**CONCLUSION**

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