

Introduction and Background

Achondroplasia, the most common form of dwarfism and skeletal dysplasia, results from a mutation in the FGFR3 gene, leading to impaired long bone growth. Although its incidence is low, achondroplasia frequently requires surgical intervention, meaning anesthesia providers will likely encounter affected patients during their careers.

Key features of achondroplasia that present challenges in anesthesia care include short stature and abnormal body proportions, requiring adjustments in medication dosing and specialized equipment for positioning. Comorbidities such as lordosis, spinal stenosis, and macrocephaly complicate these issues further. Additionally, achondroplastic patients often have non-skeletal conditions like obesity, obstructive sleep apnea, and hydrocephalus, which increase peri-operative risks, particularly in airway management and pharmacological considerations.

This review explores existing literature pertaining to peri-operative complications in achondroplastic patients, offering a systematic approach to anesthetic management. Key considerations include customized medication dosing, patient positioning, and airway management techniques. The goal is to reduce post-anesthetic morbidity and mortality, ensuring better outcomes and recovery for achondroplastic patients. By understanding these considerations, anesthesia providers can enhance the safety and outcome of procedures for these patients.

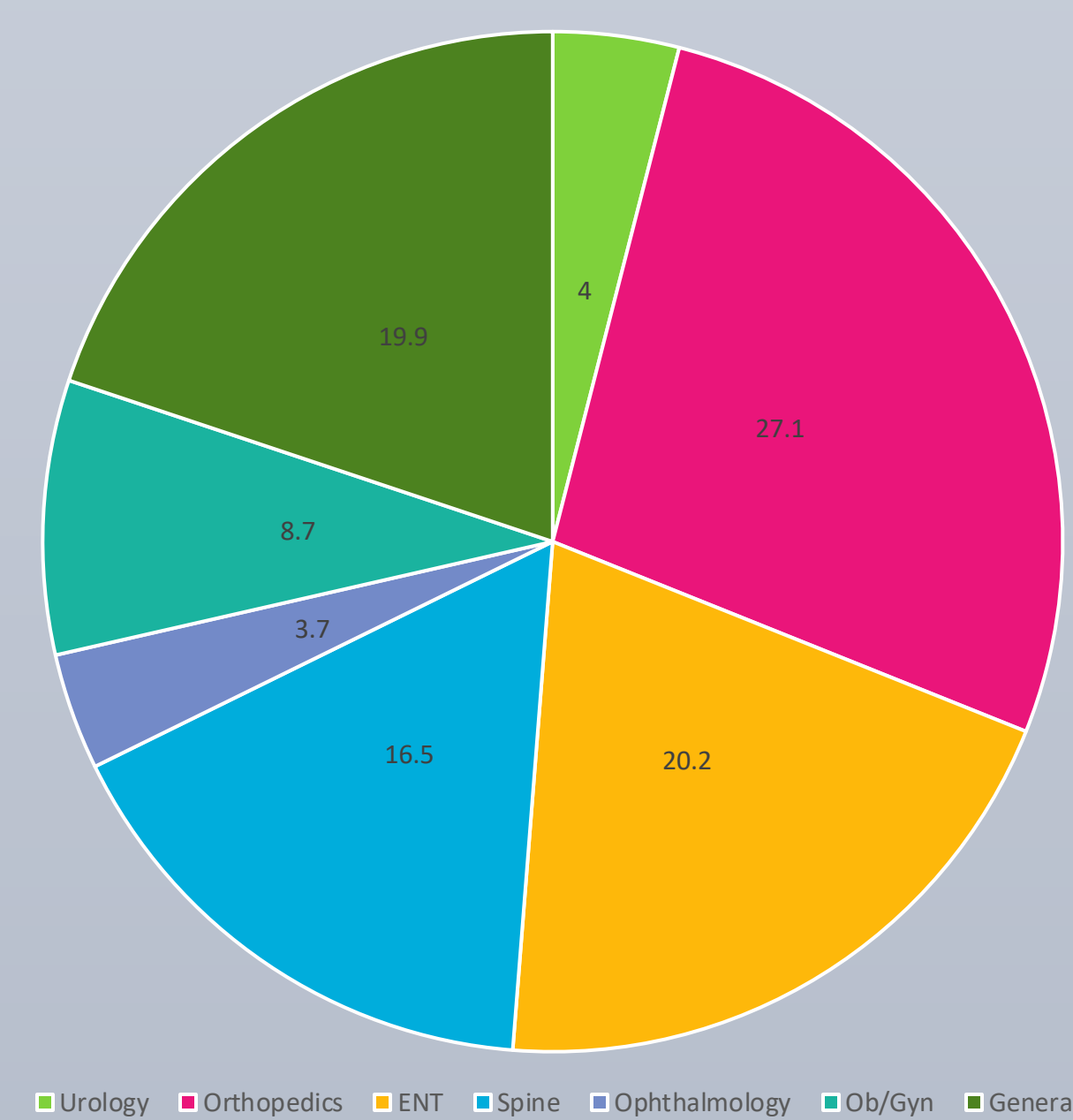


Figure 1. Ambulatory surgery type by surgical diagnosis in a study of 123 achondroplastic adults (Broder et. al., 2022.).

Preoperative Considerations

Preoperative planning for anesthesia care of achondroplastic patients is crucial to minimizing complications. The patient’s medical history and comorbidities should be carefully examined, alongside other preoperative preparations as indicated, such as the following tests:

- Lung function: incentive spirometry,; chest radiographic imaging
- Cardiac function: echocardiogram; arterial blood gas
- Neurologic function: somatosensory evoked potential
- Airway: airway evaluation; neck exam; obstructive sleep apnea questionnaire

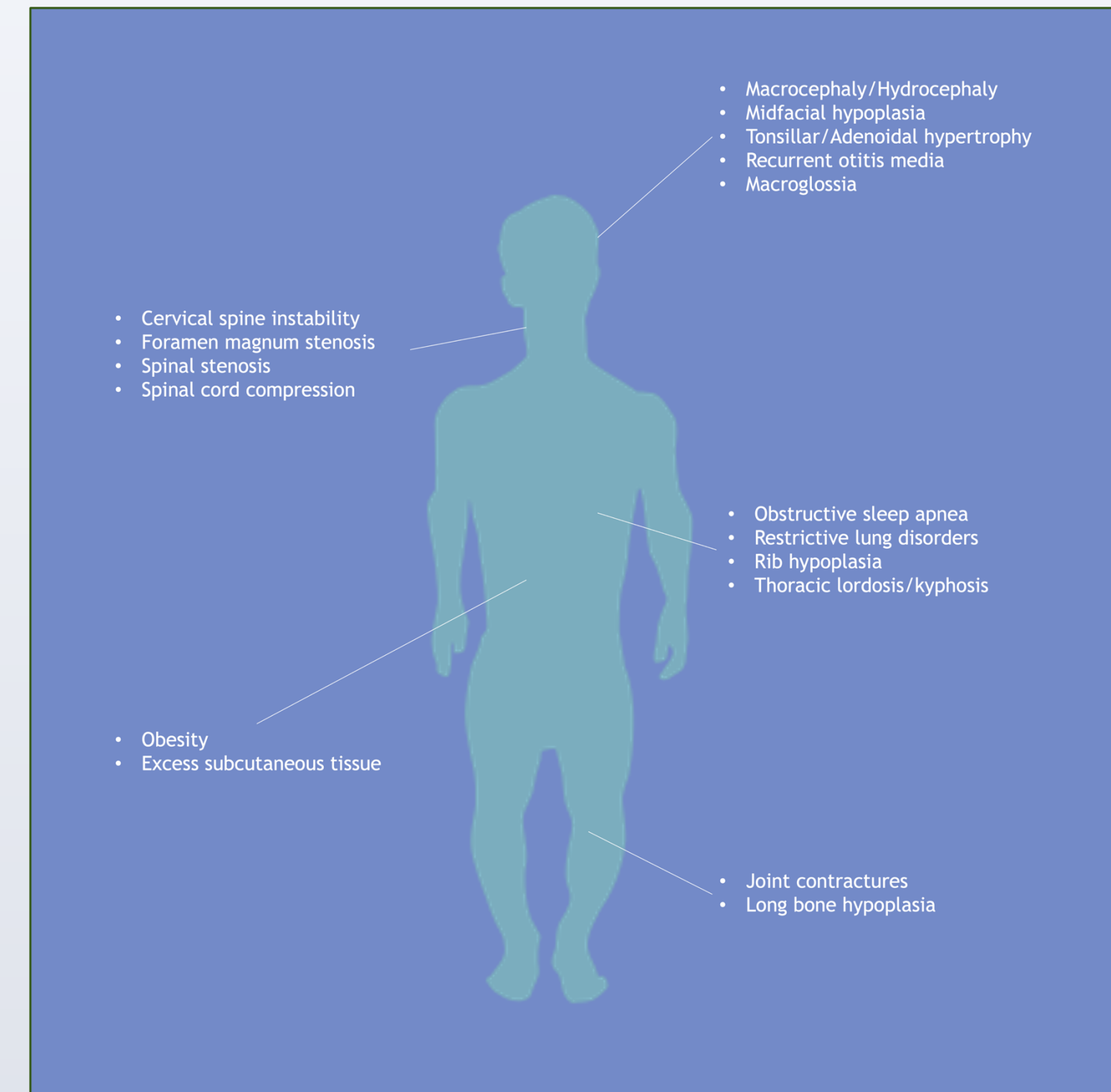


Figure 2. Common achondroplasia manifestations and comorbid conditions.

Intraoperatively, many anesthesia complications may arise due to physical manifestations and comorbid conditions of achondroplasia. The anesthesia provider should anticipate these complications, alter their anesthesia plan accordingly, and treat unforeseen problems as they arise. Proper mitigation and detection of unsafe patient conditions results in the greatest patient outcome.

Intraoperative Considerations

	Presenting Concern	Management Technique
Positioning	<ul style="list-style-type: none"> Increased head size to body size ratio Joint contractures and stenoses Increased subcutaneous tissue 	<ul style="list-style-type: none"> Use a shoulder roll for atlantooccipital support Monitor for nerve compression Use properly-sized equipment
Drug Administration	<ul style="list-style-type: none"> Increased organ mass to skeletal muscle mass ratio Macrocephalic and hydrocephalic conditions Obesity 	<ul style="list-style-type: none"> Increase doses of lipophilic drugs Avoid drugs that increase intracranial pressure Adjust doses per weight as recommended
Fluid Management	<ul style="list-style-type: none"> Macrocephalic and hydrocephalic conditions Increased subcutaneous tissue 	<ul style="list-style-type: none"> Avoid fluid overload Use diuretics as needed Ensure proper IV placement
Airway	<ul style="list-style-type: none"> Maxillary and pharyngeal hypoplasia Macroglossia Tonsillar and adenoidal hypertrophy Limited neck range of motion 	<ul style="list-style-type: none"> Utilize awake fiberoptic intubation Prepare for emergency difficult airway management scenarios Avoid neck manipulation
Ventilation	<ul style="list-style-type: none"> Reduced residual volume and functional residual capacity Obstructive sleep apnea Hypoxemia 	<ul style="list-style-type: none"> Increase inspired oxygen concentration Use optimal positive end expiratory pressure Utilize arterial cannulation
Circulation	<ul style="list-style-type: none"> Pulmonary hypertension 	<ul style="list-style-type: none"> Utilize permissive respiratory alkalosis Avoid pulmonary vasoconstricting drugs Administer inotropic drugs

Table 1. Anticipated anesthesia concerns presenting in the achondroplastic patient and associated risk management techniques.

Postoperative Considerations

Postoperative care for achondroplastic patients should focus on managing comorbidities like respiratory and cardiac issues. Ventilator weaning should be gradual, and opioids should be avoided to prevent respiratory depression, with non-opioid analgesics used for pain control. Common postoperative complications include atlantoaxial dislocation, spinal cord injury, high spinal anesthesia, and ventilatory depression. Careful attention to patient positioning, neuraxial anesthesia, and optimizing oxygenation and ventilation is essential to reduce these risks.

Conclusion

Achondroplasia presents a great number of potential difficulties as the patient undergoes anesthesia, and common comorbidities increase these risks. Anesthesia providers should be aware of these risk factors when reviewing the achondroplastic patient’s medical history and conducting the preoperative interview. Special care must be taken in crafting the anesthesia plan, with emphasis on an appropriate airway management technique with backup options prepared to anticipate any deviation from the expected plan. The anesthesia provider should be attentive in managing the patient’s organ function during anesthesia, especially oxygenation status and hemostasis, to ensure the best postoperative outcome for the achondroplastic patient.

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