Anesthetic Management of a Patient with Achondroplasia and BMI > 80 Presenting for Nephrolithotripsy

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Introduction

Achondroplasia, a disorder of long bone growth caused by mutations in the fibroblast growth receptor 3 gene, is the most common form of short-limbed dwarfism, and is also commonly associated with:

- obesity and obstructive sleep apnea (OSA)
- facial and laryngeal abnormalities
- neurologic/spinal abnormalities including hydrocephalus, cervical instability, kyphoscoliosis
- cardiopulmonary abnormalities including restrictive lung disease, pulmonary hypertension, cor pulmonale

This condition poses challenges for both general and regional anesthetic techniques. We present the management of a patient with achondroplasia, a body mass index of 84, and severe OSA who required anesthesia in the prone position for nephrolithotripsy.

Patient Presentation

- 18 year old male with achondroplasia, developmental delay, and kyphoscoliosis.
- Scheduled for nephrolithotripsy due to nephrolithiasis.
- Several general anesthetics at another institution many years prior. However, progressive obesity since that time, now 3’11” and 265 lbs.
- Uses a wheelchair except for walking short distances, and typically sleeps in an upright position in a lounge chair.

Preoperative Evaluation

Sleep Study - Severe OSA, with AHI of 38.5 and frequent desaturations to 40-50% even during O2 titration. Additionally, non-apneic nocturnal hypoxemia, average SpO2 89%.

TTE - Not consistent with pulmonary hypertension. Normal biventricular function.

BIPAP titration study, 21/16 and 1/8 LPM O2, demonstrated greatly improved AHI of 2.6 and SpO2 maintained > 95% during sleep.

References


Perioperative Course

Despite acromegalic facial features, our patient demonstrated reasonable mouth opening and neck range of motion. He had previously undergone foramen magnum decompression for hydrocephalus, but MRI revealed a stable cervical spine. We preoxygenated in a sitting position, induced with propofol, demonstrated successful mask ventilation, then administered paralytic and intubated via direct laryngoscopy.

Careful prone positioning included modifying our usual prone pillow. Adequate mechanical ventilation was achieved with a PEEP of 8 cm H2O and frequent recruitment maneuvers. Due to the discordance between limb length and circumference, we found the most reliable blood pressure cuff to be a neonatal cuff placed on the index finger.

As previously planned due to severe OSA, he was brought to the PICU intubated post-operatively to be extubated to BiPAP and closely monitored. He was discharged home in stable condition with continued nightly BiPAP on post-operative day two.