

## **Title: Hadju-Cheney Syndrome with Airway Collapse Complicating Anesthetic Management**

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### **ABSTRACT BODY:**

**Introduction:** Hadju-Cheney syndrome is defined by the findings of acroosteolysis, wormian bones, facial hypoplasia, bathrocephaly and osteoporosis. We report on a patient with Hadju-Cheney syndrome with a review of the published literature. We noted significant airway anatomical distortion despite an apparently mild expression of this syndrome. In addition, as patients with congenital abnormalities reach adulthood, the pediatric anesthesiologist is often consulted in the anesthetic management due to experience and knowledge of issues relevant to the congenital syndrome.

**Case Presentation:** Our patient was a 48 kg, 25 year-old female scheduled to undergo an elective teeth extraction procedure due to Hadju-Cheney syndrome. The patient had a history of multiple surgeries during infancy and childhood as a result of her syndrome. Most notable procedures included cervical spinal fusion at age 15 and life-long, multiple orthopedic procedures the repair spontaneous fractures and limb abnormalities. Remaining medical history included GERD, osteoporosis, penicillin allergy, difficult intubation, and awareness during anesthesia. Upon questioning during her pre-operative anesthetic evaluation, the patient denied respiratory symptoms of both her upper and lower airway. Due to her history of cervical fusion and cranio-facial abnormalities including microretrognathia with limited mouth opening, we chose to secure the airway using a fiberoptic intubation. The patient received 2 mg of IV midazolam and 0.2 mg glycopyrrolate. After topicalization the airway with 3cc of nebulized 4% lidocaine and 2% lidocaine jelly on a tongue depressor, we used a 1 cc volume of phenylephrine/Lidocaine (0.25/3%) nasal spray to topicalize her right naris. The fiberoptic scope preloaded with a 6.0 ID Mallinckrodt cuffed nasal RAE ETT was advanced to the level just above the vocal folds and 3cc of 2% lidocaine was instilled in that area. As the scope was advanced through the trachea, there was noted to be collapse of the distal trachea in a transverse plane that extended from the carina superiorly about 2.5cm. On either side of the collapsed trachea, there was a small patent passageway leading to each primary bronchus. This tracheal collapse significantly lessened as controlled ventilation was instituted. The surgery proceeded in an uneventful fashion and the patient was extubated awake and recovered from anesthesia without any complication.

**Discussion:** Hadju & Kauntze in 1948 and Cheney in 1965 were first to independently describe syndromes of acroosteolysis, wormian bones, facial hypoplasia, bathrocephaly and osteoporosis that define Hadju-Cheney Syndrome. (1, 2) Since then at least fifty-seven cases of the syndrome have been reported in the literature under various terms. Tracheomalacia-like symptoms as was found in our patient has not been reported. The precise path physiology of the syndrome remains uncertain. Cheney proposed vascular changes with focal ischemia contributing to bone resumption. (2) Definitive treatment attempts have only occurred in the twenty-first century with evidence of increases in bone mineral density and content after anabolic and anti-remodeling therapies in patients with the syndrome (3).

Airway management a patient with Hadju-Cheney syndrome is one challenge, but having the potential for tracheal collapse distally is another very significant issue as well. The tracheal abnormality may have been diagnosed preoperatively by obtaining flow volume loops or MRI/CT imaging of the chest. The use of these two modalities pre-operatively to assess the trachea in this population of patients is currently not performed. We believe, due to the possibility of such severe distal tracheomalacia, these pre-operative evaluations should be considered in patients with this syndrome.

1. Hajdu N, Kauntze R. Cranioskeletal dysplasia. *Br J Radiol* 1948;21:42-8.
2. Cheney WD. Acro-osteolysis. *AJR* 1965;94:595-607.
3. Gass M, Dawson-Hughes B, Preventing osteoporosis-related fractures: an overview. *Am J Med.* 2006 Apr;119(4 Suppl 1):S3-S11. Review.