

# GENERAL ANESTHESIA IN A PATIENT WITH CLEIDOCRANIAL DYSPLASIA

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## Introduction

Cleidocranial dysplasia (CD) is a rare disorder that involves developmental abnormalities of bony structures. CD is characterized as an autosomal dominant skeletal dysplasia with a variety of clinical manifestations; most commonly supernumerary teeth, brachycephalic skull, short stature, and hypoplastic or aplastic clavicles. In a systematic review of a study population comparing CD individuals with non-affected relatives as controls, scoliosis and upper respiratory complications (including recurrent sinus infections, collapsing nasal passages, sleep apnea, and wheezing) were noted to be significantly increased in CD individuals in comparison with controls<sup>1</sup>. Other possible manifestations of CD that involve upper airway structures include maxillary hypoplasia, high vaulted palate, and palatal clefting<sup>2</sup>. The variety of CD associated structural abnormalities can potentially pose challenges to anesthetic management; however, there is limited literature describing anesthetic implications of CD patients undergoing surgery. Potential difficulties for airway management and neuraxial placement are described in the anesthetic management of a patient with CD who underwent a variety of obstetrical procedures<sup>3</sup>. We present our approach to airway assessment and anesthetic management of a patient with CD undergoing a general surgical procedure.

## Case Description

A 24 year-old male with CD presented to our operating room for repair of pelvic prolapse. He had complaints of worsening urinary and fecal incontinence over the past several years and was scheduled for repair of both rectal and bladder prolapses (low anterior resection with rectopexy and urethral sling). It was unknown if his pelvic prolapse was related to cleidocranial dysplasia. Past medical history was significant for prior surgeries at an outside hospital related to developmental abnormalities from cleidocranial dysplasia. He had three prior back surgeries related to scoliosis (mainly in the thoracic region) and jaw surgery 3 years prior. Family history was significant for a mother with CD.

During pre-operative evaluation on the day of surgery, our patient was noted to have a limited mouth opening (Mallampati Class III), brachycephaly, frontal bossing, and short stature (the patient's height and weight were 165 cm and 57 kg respectively). He had full flexion, extension, and range of motion of his neck. Since his prior surgeries were at an outside

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hospital, no history was available on the ease of prior intubations. Upon assessment of the airway, it was felt that airway management could potentially be challenging and a video laryngoscope was made available in the operating room. Upon entering the operating room, ASA monitors were placed and our patient was pre-medicated with midazolam. After pre-oxygenation and intravenous induction with propofol and fentanyl, mask ventilation was uncomplicated and a neuromuscular blocking agent was given. We decided to first assess our patient's airway with direct laryngoscopy. With a Macintosh 3 blade and external laryngeal manipulation, a limited Cormack-Lehane Grade 2 view was obtained. The video laryngoscope (GlideScope, Verathon, Bothell, WA) was then used to facilitate the endotracheal intubation. The GlideScope provided full visualization of the vocal cords allowing for atraumatic intubation with a 7.0 standard cuffed endotracheal tube with the assistance of the GlideScope rigid stylet. Our patient was maintained on inhaled anesthetic and the case proceeded uneventfully with successful extubation at the end of the case. He recovered fully with no anesthetic complications.

## References

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## Discussion

There is very little information available regarding anesthetic care for patients with CD presenting for surgery. In particular, patients with CD have a variety of structural abnormalities that can potentially interfere with the patency of their airway requiring careful assessment by the anesthesiologist involved in their care. Anatomic abnormalities of the skull and facial structures (including dentition) may impede with mask ventilation and endotracheal intubation while spinal abnormalities may pose challenges to neuraxial techniques. In addition to performing a standard history and physical, there should be preparation to deal with a potentially difficult airway. Additional equipment such as a video laryngoscope or fiberoptic scope should be readily available. In a recent analysis of intubations at two academic medical centers, the GlideScope video laryngoscope assisted in successful intubation in 94% of cases after a failed direct laryngoscopy attempt<sup>4</sup>. Our patient did have a difficult intubation that was successfully managed with the use of the GlideScope. Our case report demonstrates how patients with CD may have skeletal abnormalities that require careful assessment of the airway and preparation with appropriate equipment to manage a potentially difficult airway.

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