Case Report

Open wide: Anesthetic management of a child with Hecht-Beals syndrome

ABSTRACT

Hecht–Beals syndrome (HBS) is a rare disorder characterized by trismus and deformity of the extremities. The etiology of trismus is unknown; theories suggest invasion of enlarged coronoid processes into the zygomatic bone. Of primary concern is the limited mouth opening and possible difficult airway. Since the syndrome was first described in 1969, there have been several articles in the pediatrics and dental literature but only 6 case reports describing the anesthetic management of these patients. Successful airway approaches have utilized various techniques including blind nasal intubation, fiberoptic intubation, and tracheal tube introducer guidance. In this case report, we discuss a multidisciplinary approach to the anesthetic management of a child with HBS undergoing MRI and outpatient surgery.

Key words: Congenital trismus; Hecht-Beals syndrome; pseudocamptodactyly

Introduction

We describe the anesthetic management of a child with Hecht–Beals syndrome (HBS) undergoing sedated magnetic resonance imaging (MRI) and outpatient surgery. We will also explain the importance of using a multidisciplinary approach in the coordination and surgical planning of patients with HBS.

Also known as trismus pseudocamptodactyly syndrome or Dutch–Kentucky syndrome, HBS is a rare autosomal dominant disorder with variable expressivity due to mutations in the gene MYH8, which encodes a perinatal myosin (myosin heavy chain 8) that has a role in early skeletal development.[1] First described by Drs. Hecht and Beals in 1969, HBS is characterized by limited excursion of the mandible and flexion deformity of fingers with wrist extension. It is also associated with foot deformities and short stature, although these findings are not specific. Congenital trismus is present at birth and persists throughout life.[2] The degree of trismus may vary among family members, or may vary at different stages of life. The etiology of trismus is unknown; however, theories have suggested enlarged coronoid processes and fibrous changes around the masseter muscles. If severe, this may require surgical release via bilateral coronoidectomies. However, surgical release may not permanently eliminate trismus nor the potential for a difficult airway in subsequent procedures.[3,4]

Case History

Our patient was a 6-year-old male who was scheduled for sedated MRI of his head and subsequent outpatient

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surgery. Due to his diagnosis, the patient was referred to the preoperative care clinic in order to facilitate involvement of the various subspecialties needed for his care. Prior anesthesia records from another institution noted that during elective Achilles tendon lengthening surgery, a laryngeal mask airway could not be inserted due to severity of trismus. Instead, the child had to be intubated via a nasal fiberoptic approach. On physical exam at the preoperative care clinic, he was a well-nourished (18.4 kg [9%ile], 117 cm [38%ile]), sociable young boy who was developmentally appropriate for age. There was no sign of micrognathia or any other facial anomalies. However, on further exam, he had very limited mouth opening with an interincisional opening of 8 mm [Figure 1]. The remainder of the physical exam was unremarkable. The patient's mother reported that his food needed to be pureed. Family history was positive for maternal involvement of the disease but in our patient's case, disease presented earlier in age and with more severe trismus. The possibility of tracheostomy was also discussed in the preoperative visit.

Due to his limited mouth opening and known difficult airway, the plan was to perform MRI under general anesthesia with tracheal intubation. At our institution, the MRI suite is located in a separate wing of the hospital some distance away from the operating room (OR). Because of this, the patient was brought to the OR for induction of anesthesia with a pediatric otolaryngologist experienced in tracheostomy present. The necessary safety checks and metal screening for MRI were done prior to induction of anesthesia.

Accompanied by a child life specialist, our patient was brought to the OR and standard ASA monitors were applied while preoxygenating via face mask. A slow controlled inhalation induction was performed, using 70% nitrous oxide/30% oxygen and incremental increase of sevoflurane to 8%. Spontaneous ventilation was maintained and easily assisted with bag mask. A peripheral IV was secured and glycopyrrolate was administered to minimize secretions. Dexmedetomidine 1 mcg/kg was infused over 10 min to assist in achieving an adequate plane of anesthesia while still maintaining spontaneous ventilation. Dexamethasone was administered to minimize airway edema. Nasal fiberoptic intubation commenced with 100% oxygen insufflation through the suction port of the scope. View of the airway was improved with manual jaw thrust, bringing the glottic opening into grade 1 view. A cuffed 5.0 endotracheal tube was passed through vocal cords into the trachea atraumatically and appropriate placement was confirmed with bilateral breath sounds, end tidal CO2, and visualization of the carina. Vital signs were stable during induction and intubation. The patient was transported with endotracheal tube *in situ* to the MRI scanner. In MRI anesthesia was maintained with sevoflurane, and the end of the scan the patient was extubated awake without difficulty in the MRI induction bay. He was discharged home the same day from PACU.

The child returned to the OR three months later for bilateral coronoidectomies and release of masseter trismus under the same anesthetic preparation and approach [Figure 2]. This procedure improved mandibular distraction and increased interincisional opening to 23 mm. In our case, the patient was known to have HBS, and we were able to plan accordingly.

Discussion

The primary anesthetic implication of HBS stems from limited mouth opening and the potential for difficult ventilation and intubation. Since first described in 1969, there have been few publications or case reports in the anesthetic

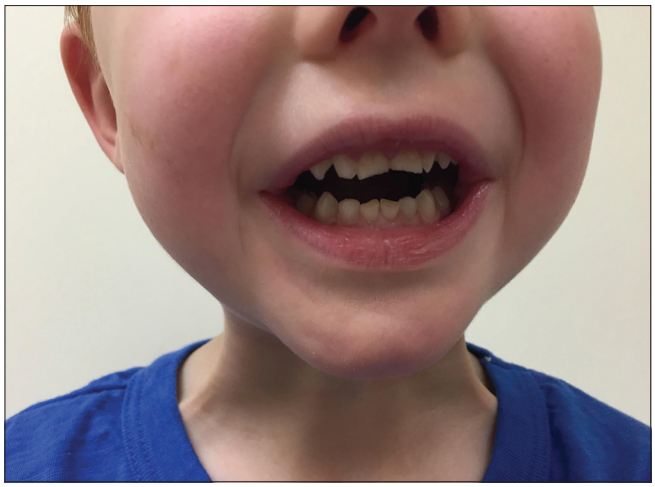

Figure 1: Limited mouth opening with maximal effort. Interincisional opening of 8 mm

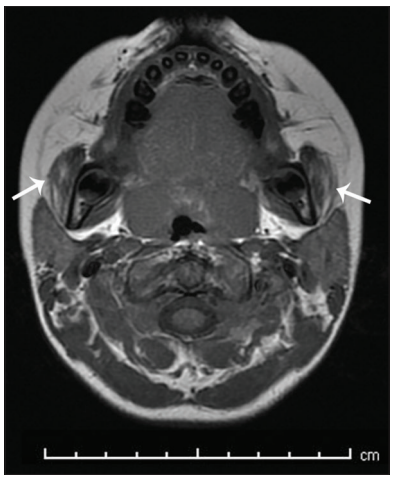

Figure 2: MRI Ax-T1: Atrophied masseter muscles with fatty infiltration

literature describing the anesthetic management of patients with HBS. Successful attempts at securing the airway in HBS patients have utilized various techniques including direct laryngoscopy,[5] blind oral[6] or blind nasal[7] intubation, fiberoptic intubation,[8] retrograde guidewire-assisted fiberoptic intubation,[9] and tracheal tube introducer assisted intubation via laryngeal mask airway.[10] General anesthetics, muscle relaxants, or manual manipulation have not been shown to improve trismus in these cases.

At first glance, patients with HBS generally do not have obvious signs of difficult intubation such as micrognathia, maxillary hypoplasia or other facial anomalies. Moreover, there is variable expressivity in affected families. Rather, the limited mandibular distraction secondary to trismus will be detectable only with careful physical examination. It is important to obtain a detailed history from parents and specifically examine for mouth opening in all children presenting with flexion deformity with wrist extension. Management of this complex patient was facilitated by a multidisciplinary approach. Communication and collaboration among anesthesiology, otolaryngology, child life, and radiology led to the safe care of this 6-year-old boy with HBS.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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