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Anesthetic Management of a Patient with Dutch-Kentucky Syndrome

FREDERICK H. BROWDER, M.D.,* DANIEL LEW, D.D.S.,† TIMOTHY S. SHAHBAZIAN, D.D.S.‡

We describe a patient with Dutch-Kentucky syndrome, a relatively rare condition that is inherited as an autosomal dominant trait and is characterized primarily by extremely limited ability to open the mouth and also by flexion deformity of the fingers that occurs with extension of the wrist.

REPORT OF A CASE

A 13.2 kg (less than fifth percentile for age) 4-yr-old boy with trismus-pseudocamptodactyly (Dutch-Kentucky) syndrome was scheduled for bilateral removal of the coronoid processes of the mandible. Past medical history and review of symptoms were unremarkable with the exception of his genetic disorder. The parents stated that the patient had never been able to eat solid foods nor had he been able to put a fork or spoon in his mouth. Family history revealed three relatives (two cousins, one uncle) with the same genetic syndrome, but no one in the patient's immediate family was affected.

On physical examination, he was a pleasant, apparently socially well-adjusted child who appeared small for his age. His maximum intermaxillary opening was 5 mm (fig. 1). The remainder of the external examination of the airway was unremarkable. Preoperative vital signs

included an arterial blood pressure of 100/50 mmHg, heart rate 110 beats/min, respiratory rate of 24 breaths/min, and a temperature of 96.6° F. The preoperative laboratory values, including a clotting profile, were within normal limits.

Following premedication with diazepam, 2.5 mg po (swallowed without difficulty), and atropine, 0.1 mg im, the child was taken to the operating room where blood pressure cuff, precordial stethoscope, and ECG were placed. With a surgeon experienced in pediatric emergency tracheostomy in attendance, an inhalation induction of anesthesia was performed with no problems, using O₂/N₂O and halothane. The patient was easily ventilated via a face mask, and no muscle relaxants were used. When a satisfactory plane of anesthesia was achieved, the anesthetic mixture was changed to halothane in 100% O₂.

After three unsuccessful attempts at blind nasotracheal intubation, fiberoptic nasotracheal intubation was successfully performed with a 5.0 uncuffed Sheridan® endotracheal tube over an Olympus® pediatric flexible bronchoscope. Sixty seconds was the maximum time period allowed for each intubation attempt, at which point the patient was again ventilated for 60 s with 100% O₂/halothane via face mask. At no time during the ventilation/intubation sequence did bradycardia, hypotension, or cyanosis occur. The decision to limit each intubation attempt to 60 s was an arbitrary one.

Successful endotracheal intubation was confirmed by the presence of equal bilateral breath sounds. Bilateral coronoidectomy was performed in conjunction with stretching of the patient's contracted masseter muscles to 30 mm, accompanied by insertion of a bite block. The vital signs remained stable throughout the procedure, which lasted 4 h, the patient being maintained on O₂/N₂O and isoflurane. One hour into the case, dexamethasone, 2 mg iv, was administered in an attempt to decrease any postoperative airway edema.

The decision had been made to keep the trachea intubated overnight; therefore, fentanyl 75 µg incrementally and droperidol 0.625 mg iv were given as the procedure progressed. Subsequently, the patient was taken to the intensive care unit, and 40% O₂ was administered by T-tube. The trachea was extubated the morning after surgery and several hours later the patient was sent to the ward.

* Assistant Professor of Anesthesiology.

† Associate Professor and Chief, Department of Oral Surgery.

‡ Chief Resident, Department of Oral Surgery.

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Address reprint requests to Dr. Browder.

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DISCUSSION

Several articles have appeared during the last 15 yr in the genetics literature delineating the characteristics of trismus-pseudocamptodactyly syndrome (TPS, or Dutch-Kentucky syndrome).^{1,§¶} The syndrome was originally described by Hecht and Beals from Oregon and by Wilson *et al.* from Vanderbilt in 1968.^{§¶} The eponym of Dutch-Kentucky syndrome was proposed in 1974 by Mabry *et al.*¹ from Kentucky after performing an extensive pedigree of a Kentucky family; the earliest affected family member that they were able to trace was a young Dutch girl who immigrated to the southern United States circa 1780, hence, the term Dutch-Kentucky. The syndrome is a relatively rare, autosomal dominant condition that is characterized by two primary features: 1) decreased ability to open the mouth (trismus); and 2) flexion deformity of the fingers that occurs with wrist extension (pseudocamptodactyly). Foot deformities and shorter-than-normal stature are frequently seen, although they are not characteristic.²

Robertson *et al.*,³ in a six-generation family study, determined that TPS has variable expressivity, but that there was no evidence of reduced penetrance. ter Haar and von Hoof⁴ proposed that enlarged coronoid processes were the cause of the trismus. Mercuri⁵ postulated that not all TPSs have enlarged coronoid processes; he proposed that the trismus may also be due to an abnormal ligament extending from the maxilla to the mandible anterior to the masseter muscles. The same author reported that general anesthesia was administered following an awake blind nasotracheal intubation in this adult. Yamashita and Arner⁶ had reported a 4-yr-old child who had an elective tracheostomy performed 4 days prior to coronoid resection.

In our case report, the airway was managed by fiberoptic nasotracheal intubation, a joint decision by both the anesthesia and oral surgery teams to avoid tracheostomy in a child this small, if possible. We would recommend



FIG. 1. Dutch-Kentucky syndrome. Frontal view of patient with mouth opened to fullest extent (5 mm).

that the anesthetist/anesthesiologist choose between blind or fiberoptic tracheal intubation according to their individual skills, with either the oral or nasal route as predicated by accessibility and surgical needs. Although neither pulse oximetry nor transcutaneous oxygen monitoring was available to us, we would recommend their use if available, but do not feel that their absence should indicate postponing surgery or transferring the patient to another institution. An amendment to our technique would be to include insufflation of 100% oxygen during any intubation attempt on such a child. While many approaches to the "difficult airway" have been described, we describe our approach to a relatively rare condition that, because it is inherited as an autosomal dominant trait, may be seen with increasing frequency by anesthesiologists.

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