Cricopharyngeal (CP) achalasia is a rare condition of unknown etiology in which the upper esophageal sphincter (UES) fails to open properly during deglutition. The normal UES is tonically closed to prevent the inflow of air into the esophagus during inspiration and the regurgitation of food from the esophagus back into the pharynx. The UES briefly relaxes during deglutition, belching, or emesis.

Cricopharyngeal achalasia is a well-described condition in adults, with symptoms including dysphagia, expectoration of excess saliva, hoarseness, and regurgitation. In infants, however, CP achalasia is exceedingly rare and potentially life-threatening. Presenting signs include difficulty in feeding, regurgitation, coughing, nasal reflux, and failure to thrive. We describe a neonate in whom botulinum toxin was used for the temporary resolution of CP achalasia.

REPORT OF A CASE

A 3210-g boy was delivered after 38½ weeks of gestation to a 22-year-old gravida 2, para 2 woman. The pregnancy was complicated by polyhydramnios requiring therapeutic reduction. Failure to progress during delivery prompted an uncomplicated cesarean section delivery with Apgar scores of 7 at 1 minute and 8 at 5 minutes.

The neonate, who was a poor feeder with inadequate sucking and swallowing reflexes, developed inspiratory stridor and desaturations with feedings. An oropharyngeal motility study demonstrated a web-like partial obstruction at the C2-C3 vertebral level, just inferior to the pyriform sinuses, and at the level of the UES (Figure 1). Esophageal motility was noticeably impaired at this level. Marked pooling of feedings allowed laryngeal penetration and frank tracheal aspiration.

Despite the radiographic findings, an anatomical obstruction was not evident on esophagoscopy. The mucosa of the UES was redundant (Figure 2) but easily accommodated a rigid 6-mm esophagoscope. In the absence of an anatomical obstruction, the neonate was diagnosed as having CP achalasia. The findings of computed tomography of the head and magnetic resonance imaging of the brain and brainstem were normal, showing no evidence of a compressive brainstem lesion. Proton pump inhibitor therapy (omeprazole, 1 mg/kg) was initiated, and the patient was discharged to home with nasogastric feedings in an attempt to allow him to grow and to determine whether his condition would resolve without surgical intervention.

The patient underwent an endoscopic botulinum toxin injection at 3½ months of age, when his condition failed to improve. The UES was visualized with suspension laryngoscopy using a pediatric Benjamin operating laryngoscope (Karl Storz Endoscopy, Culver City, Calif). The absence of an anatomical obstruction was again confirmed, and a total of 10 U of botulinum toxin was injected into the posterior midline, right posterolateral, and left posterolateral portions of the cricopharyngeus muscle (0.13 mL of 25.0 U/mL at each site) under direct visualization (Figure 3). Botulinum toxin was not injected into the
region of the thyroarytenoid muscles. The patient had no stridor on awakening from anesthesia. The postoperative oropharyngeal motility study showed mild persistence of the CP bar but marked improvement in esophageal flow, with only trace aspiration (Figure 4). The patient tolerated thickened feedings well and gained weight appropriately.

Stridor and dysphagia returned 2 months later, and the parents opted for another botulinum toxin injection because of the success of the initial procedure. A total of 11 U of botulinum toxin was injected into the region of 3 original sites (0.15-0.20 mL of 25.0 U/mL at each site). Improvement was short-lived, and a CP myotomy was recommended when the patient’s dysphagia returned. This procedure was performed through a cervical incision, and the patient was discharged 3 days later on a regimen of oral feedings, which were well tolerated. He was still doing well 14 months after the myotomy.

**COMMENT**

Utian and Thomas described the first 2 cases of pediatric CP achalasia in 1969 in 4- and 7-week-old patients. Since then, approximately 30 pediatric cases have been documented in the literature. According to the reports that had an identifiable date of diagnosis, the majority (87%) of diagnoses were made by the time the patients were 1 year of age. Only 15% of the cases, however, were diagnosed by 1 month of age, making CP achalasia in a neonate a very rare condition.

In pediatric patients, CP achalasia can be idiopathic and isolated or associated with other organic conditions, such as prematurity, congenital diseases of the central nervous system, or other neuromuscular disorders. Despite these associations, the exact pathogenesis remains unknown. Spontaneous resolution has been reported in neonates, suggesting that the condition in newborns may be attributable to a relative state of neuromuscular immaturity and may not require surgical intervention. The potential for spontaneous resolution in neonates makes initial management with botulinum toxin a particularly favorable treatment option. Gastroesophageal reflux has been suggested as a cause of achalasia in pediatric patients. In the absence of a reliable diagnostic study for gastroesophageal reflux, an empirical trial of proton pump inhibitor therapy may be indicated for pediatric patients, although reports of resolution with antireflux therapy are lacking in the literature.

Several surgical treatment options have been described for pediatric CP achalasia, with CP myotomy or repeated esophageal dilatations being the most commonly performed procedures. Some authors recommend initial esophageal dilatation, with CP myotomy reserved for patients in whom dilatation has failed or who present with severe symptoms and/or respiratory compromise.

Because we were unsure of the natural history of our patient’s CP achalasia, we decided to administer a botulinum toxin injection before trying more invasive procedures. Botulinum toxin works by preventing the exocytosis of acetylcholine at nerve terminals. The resulting chemodenervation of the affected muscle can last from weeks to months.
effects are reversible, however, as cell death does not occur.11

Botulinum toxin has been used to treat pediatric patients for various conditions, including esophageal achalasia. Studies in which botulinum toxin was used to treat esophageal achalasia have had varying results. Ip et al12 treated 7 patients, all of whom showed immediate improvement. Symptoms returned after a median interval of 4 months, with 43% of the patients having a sustained response beyond 6 months. Hurwitz et al13 found similar results in the treatment of 23 pediatric patients with esophageal achalasia. Nineteen patients responded initially to the treatment, with a mean response of 4.2 months. Seventeen patients eventually required either pneumatic dilatation or surgery to correct the condition, prompting the authors to recommend botulinum toxin injection only for patients who were not candidates for either surgery or dilatation. These results indicate that botulinum toxin injections are effective for the temporary relief of symptoms but that they provide a permanent resolution in only 10% to 40% of cases.

The botulinum toxin injections provided several benefits in the present case. Our patient was able to feed orally, allowing removal of the nasogastric feeding tube and obviating the need for oral stimulation exercises. Also, he was symptom free for several months, during which time we could determine if treatment of gastroesophageal reflux would improve the condition. Most important, the botulinum toxin injections allowed enough time to lapse so that we could determine whether the condition would resolve with further neuromuscular maturation.

The initial botulinum toxin injection was well tolerated and so effective that the parents opted for another injection when their son’s symptoms recurred 3 months later, with the hope that the CP achalasia would resolve with further maturation. A second administration of botulinum toxin again provided immediate relief, but the symptoms recurred and surgical myotomy was performed to provide definitive treatment.

CONCLUSIONS

Cricopharyngeal achalasia is an uncommon condition in pediatric patients. Endoscopic botulinum toxin injection into the cricopharyngeus muscle is an effective treatment strategy that permits neonates and infants to feed orally, while providing time to determine whether the condition will resolve with maturation. Open CP myotomy is used in infants with achalasia whose symptoms persist when the effects of the botulinum toxin wane.

Submitted for Publication: October 25, 2004; accepted January 4, 2005.

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Previous Presentation: This study was presented as a poster at the 31st Annual Meeting of the Society for Ear, Nose, and Throat Advances in Children (SENTAC); Nov 1, 2003; New Orleans, La.

Acknowledgment: We thank Debra (“Kay”) K. Klein for her assistance in preparing the figures for this article.

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