Partial Cricopharyngeal Myotomy for Treatment of Retrograde Cricopharyngeal Dysfunction

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Retrograde cricopharyngeal dysfunction (R-CPD) is a recently described debilitating syndrome resulting from an inability of the cricopharyngeus muscle to relax for retrograde release of excess swallowed air. This results in the daily experience of the 4 cardinal symptoms of inability to belch; loud and therefore socially awkward gurgling noises; daily discomfort in low neck, chest, and abdomen; and excessive flatulence. Less universal symptoms include postprandial painful hiccups, nausea, hypersalivation in response to the need to belch, and even shortness of breath with exertion due to inability to inhale fully due to esophageal/abdominal air, especially when the latter causes abdominal distention. Importantly, patients with this condition report significant social inhibition with a substantial quality of life impact. Due to their distress from the condition, many patients see several physicians and undergo various procedures, including esophagoscopy, barium studies, and esophageal manometry. None of these tests to date have provided a diagnosis in a caseload numbering 255, and instead, diagnosis is based on the syndrome.

Bastian and Smithson¹ initially described this syndrome in 2019, reporting on their first 51 consecutive patients and the results of botulinum toxin injections into the cricopharyngeus muscle. In a caseload now numbering 255 patients, all but 1 patient has experienced enough relief of symptoms to validate the diagnosis of R-CPD. Approximately 80% seem to be relieved of this problem permanently after a single injection; the remainder lose the benefit some months later. Some patients have achieved “permanent” relief after a subsequent injection, but a few have failed again to achieve permanent relief. We now present the first such patient to undergo partial cricopharyngeal myotomy. Institutional review board exemption was obtained from AspireIRB.

Case Presentation
The patient is a 32-year-old man who initially presented to the first author after learning about retrograde cricopharyngeal dysfunction from material sourced from the Bastian Voice Institute (BVI) but found on the Internet. He had a lifelong history of the symptoms as above, and prior to diagnosis at BVI, he had undergone endoscopy and antireflux therapy without a clear diagnosis or any improvement.

After diagnosis at BVI, the patient underwent injection of 50 U of botulinum toxin into his cricopharyngeus muscle with initial full resolution of his symptoms (Figure 1). However, he developed return of all symptoms by 5 months postinjection and so underwent reinjection with 75 U.

Figure 1. Representative endoscopic view of cricopharyngeus muscle after suspension.
Again, he had complete relief, but by 5 months after his second injection, the problem had again returned.

Both continuing with Botox therapy and partial cricopharyngeal myotomy were offered, and the patient chose endoscopic myotomy. On an outpatient basis, he underwent uncomplicated esophagoscopy and endoscopic partial cricopharyngeal myotomy with division of approximately 80% of the cricopharyngeus muscle using the CO₂ laser, intentionally leaving 20% of the posterior fibers of the cricopharyngeal muscle intact (Figure 2). He was discharged on a soft diet on the same day.

The patient is currently 7 months out from his surgery and was followed up by telephone on the day after, 1 week, 6 weeks, and 6 months. He reported relief of his symptoms and said that he was able to burp, did not have gurgling noises, bloating was gone, and flatulence was diminished to “normal levels.” At his 6-month follow-up, he said that his quality of life was “so much better and [he was] so thankful.”

Discussion

The cricopharyngeus muscle is an important sphincter of the digestive system, which is tonically contracted much of the time and momentarily relaxes during swallowing to allow a food or liquid bolus to enter the esophagus. Antegrade dysfunction of this muscle causing dysphagia is well described, resulting from a variety of neuromuscular, anatomic, inflammatory, neoplastic, or iatrogenic disorders. Typical treatment for the widely known and mostly age-related antegrade dysfunction, a swallowing disorder, has included botulin toxin injection into the muscle, dilation, or myotomy. This type of dysfunction has a typical symptom complex related to solid food dysphagia in particular and is often diagnosed by videofluoroscopic (or preliminarily, videoendoscopic) swallow study after patients complain of solid greater than liquid dysphagia.

Our case describes the first known reported case of myotomy for treatment of retrograde cricopharyngeal dysfunction. Endoscopic cricopharyngeal myotomy carries with it a low but not insignificant risk of subcutaneous air and even mediastinitis. We hypothesized that this risk would be minimized by leaving a thin layer of the posterior fibers of the cricopharyngeal muscle intact. Although further study is needed, we propose that a generous partial myotomy could be considered for the treatment of retrograde cricopharyngeal dysfunction in patients whose R-CPD symptoms have been resolved completely but return after 1 or more injections of botulinum toxin.

Author Contributions

Robert W. Bastian, conception or design of the work, drafting the work and revision, final approval, agree to be accountable; Rebecca C. Hoesli, acquisition, analysis, and interpretation of data, drafting the work and revision, final approval, agree to be accountable.

Disclosures

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