Clinical Medicine Insights: Case Reports

CASE REPORT

Balloon Dilation in Sporadic Inclusion Body Myositis Patients with Dysphagia

Ken-ya Murata[1], Ken Kouda[2], Fumihiro Tajima[2] and Tomoyoshi Kondo[1]

[1]Department of Neurology, [2]Department of Rehabilitation Medicine, Wakayama Medical University, Wakayama, Japan.
Corresponding author email: kemurata@wakayama-med.ac.jp

Abstract: Here, we describe balloon catheter dilation at the upper esophageal sphincter (UES) in three sporadic inclusion body myositis (s-IBM) patients with dysphagia. Initially, we performed IVIg therapy, and, three months later, switched to balloon dilation therapy. A 12-Fr balloon catheter was inserted from the mouth under fluoroscopy and the balloon inflated at the UES. The catheter was pulled back and re-inserted several times. We examined videofluoroscopy (VF) and pressure at the oropharynx, hypopharynx and UES using computed pharyngoesophageal manometry (CPM). Before both therapies, the VF study revealed a very small amount of barium paste passing through the UES. After balloon dilation therapy, as well as IVIg, subjective complaints of dysphagia disappeared and the VF study revealed an increased amount of barium paste passing through the UES. We conclude that balloon dilation therapy is a complementary method for conventional dysphagia therapies in s-IBM patients with dysphagia.

Keywords: balloon dilation, computed pharyngoesophageal manometry, cricopharyngeal achalasia, dysphagia, sporadic inclusion body myositis

Clinical Medicine Insights: Case Reports 2013:6 1–7

doi: 10.4137/CCRep.S10200

This article is available from http://www.la-press.com.

Introduction

Sporadic inclusion body myositis (s-IBM) is an inflammatory myopathy characterized by selectivity of muscle involvement, finger flexor and/or quadriceps femoris involvement, and a chronic progressive corticosteroid-resistant course.[1] Dysphagia has been reported in approximately one-third of s-IBM patients,[2] and patients with progressive dysphagia have a significantly poorer functional rating than patients with non-progressive dysphagia.

In the current study, we performed dilation of the upper esophageal sphincter (UES) using a balloon catheter (balloon dilation) in s-IBM patients with dysphagia. We compared the efficacy of this therapy with intravenous immunoglobulin (IVIg) therapy using videofluoroscopy (VF) and computed pharyngoesophageal manometry (CPM).

Materials and Methods

This study was performed using 3 s-IBM patients with dysphagia (2 males and 1 female; average age, 78.3 ± 4.5 years; average disease duration, 10.0 ± 5.3 years) at the Department of Neurology in Wakayama University between January 2008 and December 2010 (Table 1). These patients fulfilled the proposed diagnostic criteria for s-IBM.[3] The study was approved by the ethics committee of the Faculty of Medicine at Wakayama Medical University. All patients provided written informed consent in order to be included in the study.

First, we performed IVIg therapy (400 mg/kg/day for 5 days) on the three patients. Three months after IVIg treatment, we performed balloon dilation therapy. A 12-Fr balloon catheter for bladder placement was inserted from the mouth to the region of the UES including the cricopharyngeal muscle. The insertion position of the catheter was confirmed using fluoroscopy. The UES was then dilated by balloon inflation to 18 mm and then 20 mm (Fig. 1B). The catheter was pulled back, re-inserted and dilated several times during dry swallowing. Once the insersion position was determined, the patients themselves were able to insert the catheter at the best position without fluroscopy. This procedure was straightforward, and patients performed balloon therapy alone at home.

Swallowing was assessed before and after the two therapies using videofluoroscopy (VF)[4] and computed pharyngoesophageal manometry (CPM).[5] For VF, patients were placed upright and the oropharynx was viewed in lateral and anterior-posterior projections. Three mL of liquid barium and paste barium were administered by teaspoon. Swallowing examinations were repeated in different upright positions. For CPM, a sequential computer manometry system (PC polygraph; Medtronic, Medtronic Parkway, Minneapolis) with a 4-intraluminal pressure transducer assembly (Mui Scientific, Mississauga, Ontario) was used with the recording sites set 5 cm apart. The assembly was placed transnasally and recording sites were selected at the oropharynx, hypopharynx, UES, and proximal esophagus (Fig. 2A). We evaluated UES pressure, along with pharyngeal and esophageal peristalsis, during barium swallowing. We also performed VF and CPM studies on ten aged-matched healthy volunteers without neuromuscular disease as controls.

Results

Clinical symptoms

Prior to both therapies, it was difficult for the three subjects to eat even half-solid meals. Following IVIg therapy, status improved and the three patients were able to eat regular meals. Effectiveness endured for two months following single dose therapy. We were able to successfully perform balloon dilation in only 2 (patients 2 and 3) out of the 3 subjects, because a severe pharyngeal reflex in patient 1 prevented insertion of the catheter into the pharyx. In the two patients receiving balloon dilation, dysphagia improved and

Table 1. s-IBM patient profiles.

Patient	Sex	Complaint of dysphagia	Age	Duration of disease (years)	Aid for walking	CK (IU/L)	Complications
1	M	(+)	83	4	Cane	186	Hepatitis C
2	F	(+)	79	12	Walker	482	HT, LS
3	M	(+)	74	14	Cane	691	HT

Abbreviations: HT, Hypertension; LS, Lumbar spondylosis.

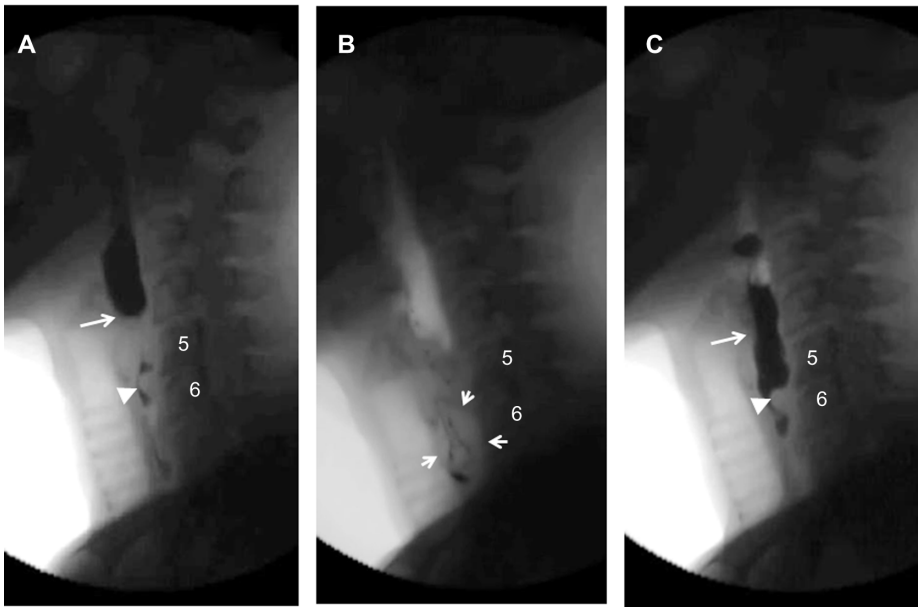

Figure 1. Videofluoroscopy in an s-IBM patient (Patient 2) using barium paste (**A** and **C**) and during balloon dilation therapy (**B**).
Notes: The barium paste is concentrated at the UES opening sites before balloon therapy (A, arrow) and is passed through the UES after balloon therapy (C, arrow). The expanded balloon (B, surrounded by arrows) pushes against the crycopharyngeal muscles. Pharyngeal muscle propulsions (PP; arrow head) were observed in the UES at the position of the crycopharyngeal muscle (A and C). The sites and shapes of PP remained unchanged before (A) and after (C) balloon therapy.

it became possible to eat regular meals. During the first two weeks, patients performed balloon dilation therapy once a day to eat regular meals. However, the effect of balloon therapy weakened and it became necessary to perform this therapy several times before each meal every day. Once balloon dilation therapy was performed, patients were easily able to eat meals for 30 minutes on each occasion. These two patients could eat regular meals for at least one year by simply performing balloon dilation therapy. No adverse effects were recorded in relation to performing balloon dilation.

Videofluoroscopy (VF)

Prior to IVIg and balloon dilation therapies, VF results indicated that all patients exhibited a normal oral phase of swallowing but possessed abnormalities in the pharyngeal phase. The UES opening was impaired and aspiration was confirmed due to an influx of remaining food from the piriform recess (Fig. 1A).

After IVIg therapy, the amount of barium paste passing through the entrance of the UES increased (data not shown). Following both balloon and IVIg therapy, the amount of barium passing through the entrance of the UES increased (Fig. 1C). We could confirm these effects at least one year later, following introduction of of the first balloon dilation thearpy. As a consequence, food residues in the piriform recess were reduced, and aspiration of food residues was alleviated by both therapies.

In all cases, the cricopharyngeal prominence (CP) was confirmed at the UES including the cricopharyngeal muscle (C5–6; Fig. 1A and C arrow head). CP was also observed at the inferior pharyngeal constrictor muscle (C2–3) in patient 3 (data not shown). The size and site of the CP did not differ when compared before and after IVIg and balloon therapies (Fig. 1A and C arrow head).

Computed pharyngoesophageal manometry (CPM)

In normal healthy control subjects, the pharyngeal peak pressure at the oropharynx and hypopharynx was elevated simultaneously (Fig. 2B, 1 and 2). Contrary to the high pharyngeal pressure at the oropharynx and hypopharynx, the pressure at the UES decreased until the UES opened (nadir deglutitive UES pressure; Fig. 2B-3, arrow). After the barium paste passed through the entrance of the UES, pharyngeal pressure at the UES increased (Fig. 2B-3, asterisk).

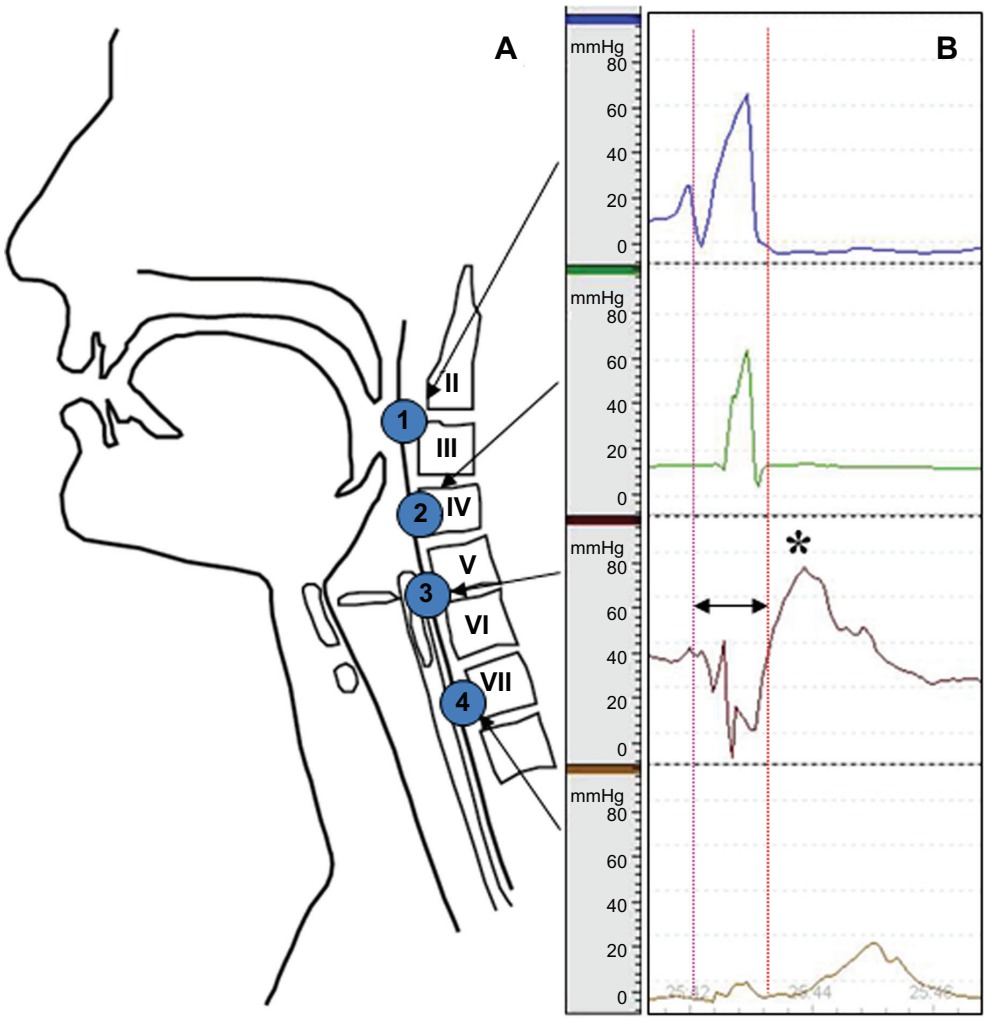

Figure 2. Manometry study of normal control. (**A**) Manometry study using simultaneous 4-channel pressure recording during 3 ml barium swallowing. channel 1 = oropharynx; channel 2 = hypopharynx; channel 3= UES; channel 4 = proximal esophagus. (**B**) Manometric findings in a healthy control subject. **Notes:** The pharyngeal peak pressure at the oropharynx and hypopharynx elevated simultaneously (1 and 2). Contrary to the high pharyngeal pressure at the oropharynx and hypopharynx, the pressure at the UES decreased until the UES opened (nadir deglutitive UES pressure; 3, arrow). After the barium paste passed through the entrance of the UES, pharyngeal pressure at the UES increased (3, asterisk).

Before IVIg and balloon dilation, the peak pressure of the oropharynx, hypopharynx and UES in s-IBM patients was very low compared with that of normal controls (Fig. 3A-1–3 and 3C-1–3). Nadir deglutitive UES pressure was not observed in s-IBM patients (Fig. 3A-3 and 3C-3). Following IVIg therapy, although nadir deglutitive UES pressure was not observed (Fig. 3B-3), the peak pressure of the hypopharynx and UES became elevated (Fig. 3B-2 and 3).

After balloon dilation, although nadir deglutitive UES pressure was not observed (Fig. 3C-3), the peak pressure of the oropharynx and hypopharynx was elevated (Fig. 3D-1 and 2). The contraction muscle power of the UES increased to the same level as that of normal controls (Fig. 3D-3, asterisk). Thirty minutes later, internal pressure of the deglutition was re-determined and found to be the same as that immediately following balloon dilation. Consequently, we concluded that sustainability duration of the balloon dilation procedure was at least 30 minutes.

Discussion

Cricopharyngeal achalasia is defined as when the opening of the UES is not adequate during the pharyngeal phase, and a food mass is hard to pass.[6] Incomplete UES opening during swallowing has a number of different causes. Causes include impaired relaxation or spasm of the UES, such as hyperplasia and hypertrophy. They can also include fibrosis of the cricopharyngeal muscle, weakness of the suprahyoid

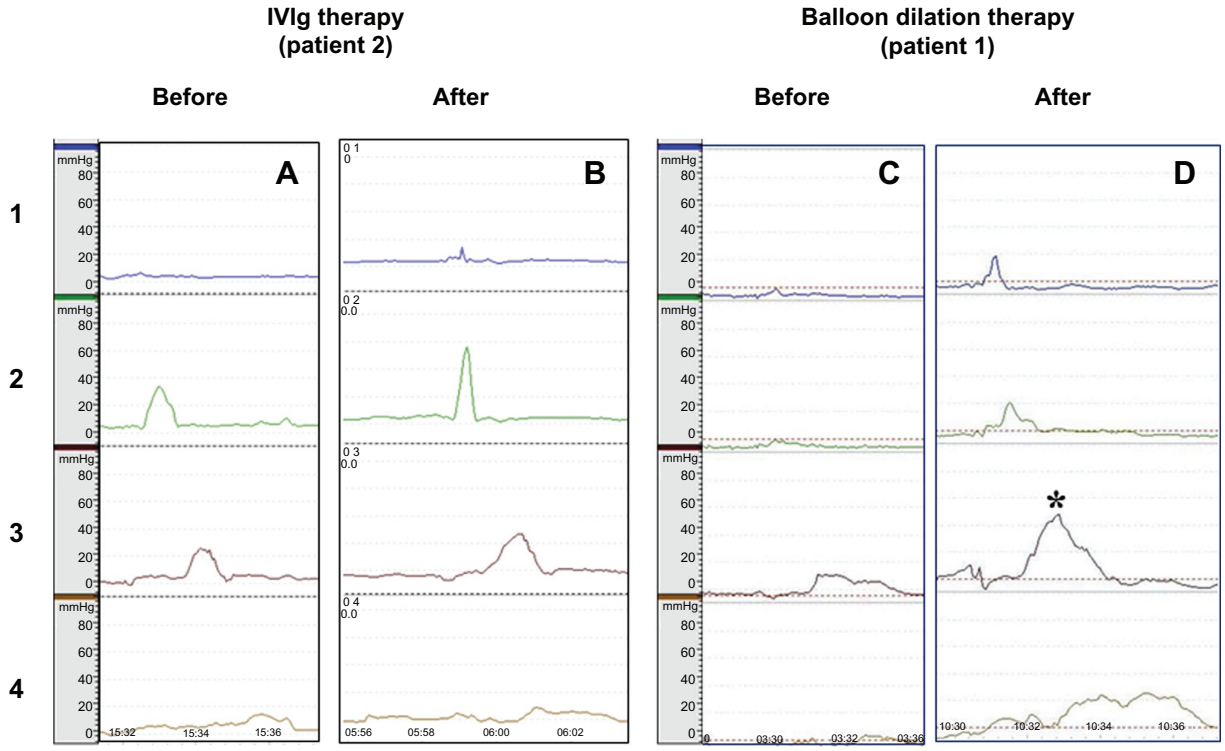

Figure 3. Manometry study of s-IBM patients. (**A**) Manometry during swallowing solid barium paste before IVIg therapy (patient 2). (**B**) Manometry during swallowing solid barium paste after IVIg therapy (patient 2). (**C**) Manometry during swallowing solid barium paste before balloon dilation therapy (patient 1) (**D**) Manometry during swallowing solid barium paste after balloon dilation therapy (patient 1) 1 = oropharynx; 2 = hypopharynx; 3 = UES; 4 = proximal esophagus.
Notes: Although the peak pressure at the oropharynx (A-1), hypopharynx (A-2) and UES (A-3) in patient 2 was low before therapy, the hypopharynx (B-2) and UES (B-3) pressures elevated after IVIg therapy. Nadir deglutitive UES pressure, which was observed in normal controls, was not observed both before (A-3) and after (B-3) therapy. The peak pressure at the oropharynx (C-1), hypopharynx (C-2) and UES (C-3) is almost 0 before balloon therapy, and is elevated remarkably after therapy (D-1–3). In particular, the peak pressure at the UES is similar to that of normal controls (D-3, asterisk). Nadir deglutitive UES pressure was not observed both before (C-3) and after (D-3) balloon therapy.

muscles, and diminished distending forces imparted by the bolus.[7] Because cricopharyngeal achalasia is considered a main reason for dysphagia in s-IBM patients, cricopharyngeal myotomy and botulinum toxin (BTX) therapy have been employed for dysphagia in s-IBM patients.[8] On the other hand, IVIg therapy has also been shown to be effective for s-IBM patients with dysphagia, although this treatment does not lead to significant improvement for limb muscle weaknesses.[9,10] Prior to IVIg therapy, our s-IBM patients suffered dysphagia. In addition, peak pressure of the oropharynx, hypopharynx and UES was low. Following IVIg therapies, the subjective complaint of dysphagia disappeared, and both the peak pressure of hypopharyngeal muscles and the contraction muscle power of the UES increased. Therefore, IVIg therapy improved the contraction power of pharyngeal and UES muscles.

Balloon dilation has been performed in patients with cricopharyngeal achalasia due to Wallenberg syndrome,[11] and in newborns with primary cricopharyngeal achalasia.[12,13] Prior to balloon dilation, it was difficult for the three s-IBM patients to eat even half-solid meals. The UES opening was impaired and aspiration due to an influx of food residue in the piriform recess was confirmed. After balloon therapy, the subjects were sucessfully able to eat regular meals, and the passage of a food mass through the UES was facilitated. In addition, the amount of barium paste passing through the UES increased, the amount of food residue in the piriform recess decreased, and aspiration with an influx of barium paste into the larynx disappeared.

An examination of cricopharyngeal muscle biopsies from s-IBM patients with severe dysphagia previously showed a marked increase in the replacement of endomysial connective tissue by fat, along with numerous small and round atrophic muscle fibers.[14] In the present study, UES contraction power was recovered by dilation of the crycopharyngeal muscles. These findings suggest that the number of UES muscle fibers

were preserved and that muscle fiber atrophy was not severe. Since reduced mobility by replacement of connective tissue prevented UES muscle contraction, only the improvement of UES muscle elasticity by balloon dillation promoted UES contraction power. These observations suggested that the UES muscle was in a condition of disuse. Increased UES muscle power induced coordination with the oropharyngeal and hypopharyngeal sphincter muscles, and contraction movement at the pharyngeal muscles improved. Elevated hypopharyngeal intrabolus pressure has previously been reported to be a good predictor of favorable symptomatic outcomes following sphincter disruption, whereas a recovery of the UES opening was not.[15] The recovered sphincter muscle power induced propulsion of the bolus through the UES. The balloon dilation method is effective not only for the alleviation of UES mechanical dilation, but also for the improvement of contraction power of the UES in coordination with other sphincter muscles. Based on these results, we conclude that balloon dilation therapy, as well as IVIg therapy, is effective in the treatment of s-IBM patients with dysphagia.

CP was observed at the UES and inferior pharyngeal constrictor muscle, and the size and shape of the CP did not change following balloon dilation and IVIg therapy. CP, however, is not a direct cause of dysphagia, but represents only the relaxation disorders of the pharyngeal constrictor muscle and UES.[16] CP has therefore proven to be a good marker to detect dilation points of sphincter musculature.

Both of the two patients rejected the option of cricopharyngeal myotomy and BTX. Neither patient experienced difficulty performing the daily balloon dilation procedure, and both were satisfied with this straightforward and non-invasive method for one year. Balloon dilation is a simple and low-cost therapeutic method. However, the improvement of dysphagia by balloon dilation is finite in nature. This procedure is not likely to be helpful in ongoing management, or as an alternative treatment to established procedures such as cricopharyngeal myotomy and BTX. We propose balloon dilation therapy as a temporary or complementary method for conventional dysphagia therapy in s-IBM patients with dysphagia. We recommend that this method be used for several days until the effect of IVIg begins, or while the patient is awaiting surgery or BTX.

Author Contributions

KM was involved in the conception of the report, the literature review, and manuscript preparation and editing. KM and KK were involved in the clinical care of the patient. FT and TK were involved in manuscript drafting, editing, review and submission. All authors proofread, reviewed and approved of the final manuscript.

Funding

This work was supported by Grants-in-Aid for the Scientific Research from the Ministry of Health, Labor and Welfare of Japan.

Competing Interests

Author(s) disclose no potential conflicts of interest.

Disclosures and Ethics

As a requirement of publication author(s) have provided to the publisher signed confirmation of compliance with legal and ethical obligations including but not limited to the following: authorship and contributorship, conflicts of interest, privacy and confidentiality and (where applicable) protection of human and animal research subjects. The authors have read and confirmed their agreement with the ICMJE authorship and conflict of interest criteria. The authors have also confirmed that this article is unique and not under consideration or published in any other publication, and that they have permission from rights holders to reproduce any copyrighted material. Any disclosures are made in this section. The external blind peer reviewers report no conflicts of interest.

References

1. Lotz BP, Engel AG, Nishino H, Stevens JC, Litchy WJ. Inclusion body myositis. Observations in 40 patients. *Brain*. 1989;112(Pt 3):727–47.
2. Wintzen AR, Bots GT, de Bakker HM, Hulshof JH, Padberg GW. Dysphagia in inclusion body myositis. *J Neurol Neurosurg Psychiatry*. 1988;51: 1542–5.
3. Griggs RC, Askanas V, DiMauro S, et al. Inclusion body myositis and myopathies. *Ann Neurol*. 1995;38:705–13.
4. Seaman WB. Cineroentgenographic observations of the cricopharyngeus. *Am J Roentgenol Radium Ther Nucl Med*. 1966;96:922–31.
5. Hurwitz AL, Nelson JA, Haddad JK. Oropharyngeal dysphagia Manometric and cine esophagraphic findings. *Am J Dig Dis*. 1975;20:313–24.
6. Bingham DL. Cricopharyngeal achalasia. *Can Med Assoc J*. 1963;89: 1071–3.
7. Cook IJ, Blumbergs P, Cash K, Jamieson GG, Shearman DJ. Structural abnormalities of the cricopharyngeus muscle in patients with pharyngeal (Zenker's) diverticulum. *J Gastroenterol Hepatol*. 1992;7:556–62.
8. Schneider I, Thumfart WF, Pototschnig C, Eckel HE. Treatment of dysfunction of the cricopharyngeal muscle with botulinum A toxin: introduction of a new, noninvasive method. *Ann Otol Rhinol Laryngol*. 1994;103:31–5.

9. Dalakas MC. Controlled studies with high-dose intravenous immunoglobulin in the treatment of dermatomyositis, inclusion body myositis, and polymyositis. *Neurology*. 1998;51:S37–45.
10. Cherin P, Pelletier S, Teixeira A, et al. Intravenous immunoglobulin for dysphagia of inclusion body myositis. *Neurology*. 2002;58:326.
11. Katoh J, Hayakawa M, Ishihara K, Kazumi T. Swallowing rehabilitation using balloon catheter treatment evaluated by videofluorography in an elderly patient with Wallenberg's syndrome. *Nippon Ronen Igakkai Zasshi*. 2000;37:490–4.
12. Davis D, Nowicki M, Giles H. Cricopharyngeal achalasia responsive to balloon dilation in an infant. *South Med J*. 2005;98:472–4.
13. Erdeve O, Kologlu M, Saygili B, Atasay B, Arsan S. Primary cricopharyngeal achalasia in a newborn treated by balloon dilatation: a case report and review of the literature. *Int J Pediatr Otorhinolaryngol*. 2007;71:165–8.
14. Verma A, Bradley WG, Adesina AM, Sofferman R, Pendlebury WW. Inclusion body myositis with cricopharyngeus muscle involvement and severe dysphagia. *Muscle Nerve*. 1991;14:470–3.
15. Ali GN, Wallace KL, Laundl TM, Hunt DR, deCarle DJ, Cook IJ. Predictors of outcome following cricopharyngeal disruption for pharyngeal dysphagia. *Dysphagia*. 1997;12:133–9.
16. Dantas RO, Cook IJ, Dodds WJ, Kern MK, Lang IM, Brasseur JG. Biomechanics of cricopharyngeal bars. *Gastroenterology*. 1990;99: 1269–74.