

# CASE REPORT

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## Efficacy of endoscopic cricopharyngeal myotomy using a curved rigid laryngoscope in patients with sporadic inclusion body myositis: four retrospective case reviews

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### ABSTRACT

Sporadic inclusion body myositis (s-IBM) is an acquired degenerative inflammatory myopathy that leads to slowly progressive muscle weakness and atrophy of the limbs, face, and pharynx. Owing to the slow progression of the disease, the indications for surgical intervention remain unclear. Herein, we retrospectively reviewed the records of four patients with s-IBM who had undergone cricopharyngeal myotomy for severe dysphagia at our institution between 2016 and 2021. Among these, one patient underwent transcervical cricopharyngeal myotomy and laryngeal suspension, as videofluoroscopic examination of swallowing revealed poor laryngeal elevation. The remaining three patients underwent endoscopic cricopharyngeal myotomy using a curved rigid laryngoscope. Preoperatively, the mean Hyodo score was 8 points (range: 6–10) using a flexible endoscope. The mean surgical duration was 104 min, and no severe complications were observed. Postoperatively, all patients achieved improvement in swallowing function and food intake. Moreover, swallowing function was maintained in all four patients even 6–12 months postoperatively. Cricopharyngeal myotomy may be a safe surgical procedure with the potential to improve swallowing function, and a Hyodo score of 6 may be considered a surgical indication for cricopharyngeal myotomy in patients with s-IBM.

Keywords: sporadic inclusion body myositis, cricopharyngeal myotomy, dysphagia, long-term follow-up, surgical indication

#### Abbreviations:

s-IBM: sporadic inclusion body myositis

CPM: cricopharyngeal myotomy

VE: videoendoscopic examination of swallowing

VF: videofluoroscopic examination of swallowing

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## INTRODUCTION

Sporadic inclusion body myositis (s-IBM) is an acquired degenerative inflammatory myopathy that leads to slowly progressive muscle weakness and atrophy of the limbs, face, and pharynx. A recent study revealed approximately 1000–1500 patients with s-IBM in Japan.<sup>1</sup> Dysphagia is a frequent and life-threatening symptom in these patients; indeed, Lots et al reported the occurrence of clinical dysphagia in 40% of patients with s-IBM as the disease progressed.<sup>2</sup> Aspiration pneumonia is a common cause of death in patients with s-IBM.<sup>3,4</sup> A recent study revealed an increased risk of mortality in patients with s-IBM relative to that of an age-matched comparison population, particularly in those with impaired swallowing and subsequent aspiration pneumonia.<sup>5</sup>

Dysphagia in s-IBM is caused by chronic inflammation of pharyngeal constrictor and cricopharyngeal muscles. Compared to those associated with other inflammatory myopathies, dysfunction of the upper esophageal sphincter occurs frequently in s-IBM. Non-surgical treatments for s-IBM, which are feasible in the early stages of the disease, include intravenous immunoglobulin therapy, speech and language therapy including Mendelsohn's maneuver, balloon dilation, and botulinum toxin injection.<sup>6–8</sup> However, balloon dilation and botulinum toxin injection, although less invasive, are temporary measures that are not sufficient for patients with severe dysphagia.

Cricopharyngeal myotomy (CPM) is a surgical procedure used to treat dysphagia that involves cutting the cricopharyngeal muscle and is often performed in patients with cricopharyngeal achalasia of neurogenetic origin or head and neck resection. Several studies have indicated that CPM is effective in patients with s-IBM with severe dysphagia. However, most patients in previous studies received transcervical CPM. Transcervical CPM has been a traditional surgical approach for cricopharyngeal dysfunction for more than 60 years, but it requires an external neck incision, longer hospital stay, and carries the risk of recurrent laryngeal nerve or great vessel injury, esophageal perforation, and fistula.<sup>9</sup> Considering the slowly progressive neuromuscular nature of s-IBM and risks of transcervical CPM, surgical indications for severe dysphagia in patients with s-IBM remain unclear.

Endoscopic CPM has been introduced as an alternative surgical procedure to transcervical CPM. Although endoscopic CPM is a safe and minimally invasive technique, it has not gained wide usage because of the narrow surgical field and the low usability of surgical instruments using a direct laryngoscope. Recently, we developed a new, safe endoscopic CPM using a curved rigid laryngoscope for patients with severe dysphagia.<sup>10</sup> This procedure allows the wide expansion of the hypopharynx, and can provide a wider surgical field than obtained by using a direct laryngoscope. Herein, we retrospectively review our experience with CPM to evaluate its safety as well as postsurgical changes in swallowing function during the follow-up period in patients with s-IBM. This study was approved by the Ethics Review Committee of the Nagoya University Hospital (2022-0061).

## CASE REPORT

### *Patients*

The clinical data of four consecutive patients with s-IBM who had undergone CPM to treat dysphagia at our institution between 2016 and 2021 were retrospectively reviewed. Table 1 summarizes the clinical characteristics and follow-up periods of the four patients (four women). The average age at disease onset was 57.8 years (range; 46–68 years), and all patients were diagnosed with s-IBM by experienced neurologists after a muscle biopsy. The average duration from disease onset to the first visit was 11.0 years (range; 7–15 years), whereas the average

**Table 1** Patient characteristics and follow-up periods

	Case 1	Case 2	Case 3	Case 4
Sex	Female	Female	Female	Female
Age (years)				
Disease onset	55	62	46	68
Dysphagia onset	68	62	58	78
Diagnosis	70	69	57	73
First visit to otolaryngologist	70	69	58	78
Surgery	76	69	60	79
First symptom	Difficulty in standing up	Swallowing dysfunction	Difficulty in standing up	Weakened grip strength
Duration (years)				
From first symptom to diagnosis	15	7	11	5
From first symptom to surgery	21	7	14	11
From dysphagia to surgery	8	7	2	1
Non-surgical treatment	No	No	Intravenous immunoglobulin therapy	No
Modified Rankin Scale *	4	2	2	3
Serum creatine kinase (U/L) *	266	657	200	120

\*: at the time of surgery

duration from disease onset to surgery was 13.2 years (range; 7–21 years). Among the four patients, one (Case 3) received preoperative intravenous immunoglobulin therapy.

#### *Assessment of swallowing function*

Swallowing function was assessed using videoendoscopic examination of swallowing (VE) and videofluoroscopic examination of swallowing (VF). Comparisons of swallowing function were performed at four different points in this study: the first visit, preoperatively, 1 week, and 6–12 postoperatively.

For VE, the Hyodo scoring system, consisting of four items scored from 0 to 3 (0, normal; 1, mildly impaired; 2, moderately impaired; and 3, severely impaired; range:0–12), was used to assess dysphagia using a flexible endoscope.<sup>11</sup> For VF, AsR scores were used to assess the severity of dysphagia using radiography. The AsR scoring system, comprising VF a 10-point scale (A: aspiration, S: silent aspiration, R: residual; As:4,3,1,0, R:6,3,2,1), is an evaluation tool that was developed to facilitate the assessment of swallowing function in clinical settings. This score has been validated for patients with dysphagia in Japan and is well-correlated with oropharyngeal swallowing efficiency.<sup>12</sup> Scores  $\geq 6$  in the AsR scoring system are a good indicator of safe nutrition from oral intake only.

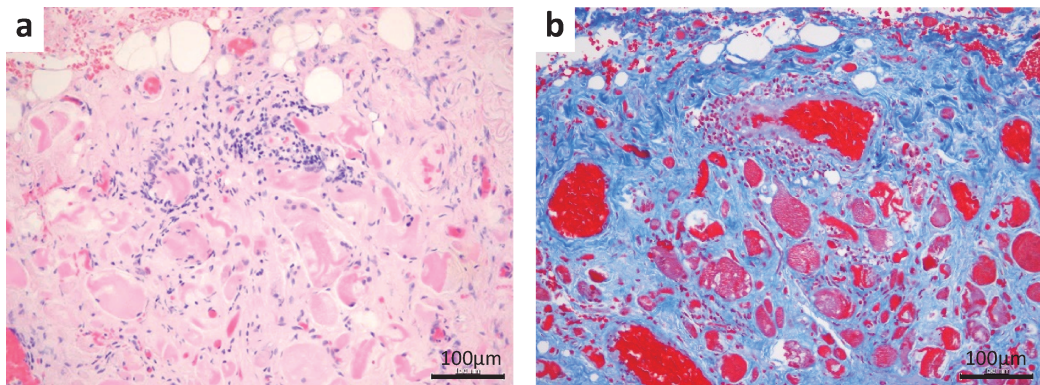
*Surgical intervention and results*

Table 2 shows the surgical interventions and outcomes for the four patients. The mean age at surgery was 71.0 years (range; 60–79 years). Four patients with s-IBM underwent CPM under general anesthesia via an endoscopic or transcervical approach in the operating room. The surgical interventions and perioperative management were performed by the same surgical team, as previously reported by Yokoi et al.<sup>13</sup> One patient (Case 1) underwent transcervical CPM and laryngeal suspension as VF imaging showed poor elevation of the larynx. The remaining three underwent endoscopic CPM using a curved rigid laryngoscope, as reported by Maruo et al.<sup>10</sup> The mean surgical duration was 104 min. No severe complications were observed. One patient

**Table 2** Surgical results

	Case 1	Case 2	Case 3	Case 4
<b>Surgery</b>				
Cricopharyngeal myotomy	Transcervical	Endoscopic	Endoscopic	Endoscopic
Laryngeal suspension	Yes	No	No	No
Operating time (min)	103	111	123	87
Complications	No	No	Minor fistula	No
<b>Duration (days)</b>				
Hospitalization	72 *	16	17	11
From surgery to discharge	15	13	13	9
<b>Food</b>				
Preoperative	Tube feeding	Soft & bite-sized	Minced & moist	Soft & bite-sized
Postoperative	Tube feeding + oral intake	Regular	Regular	Regular

\*: including the preoperative treatment for aspiration pneumonia



**Fig. 1** Biopsy of the cricopharyngeal muscle (Scale bar; 100 µm)

**Fig. 1a:** Hematoxylin and eosin staining reveals atrophic fibers and immune cell infiltration into the muscle endomysium.

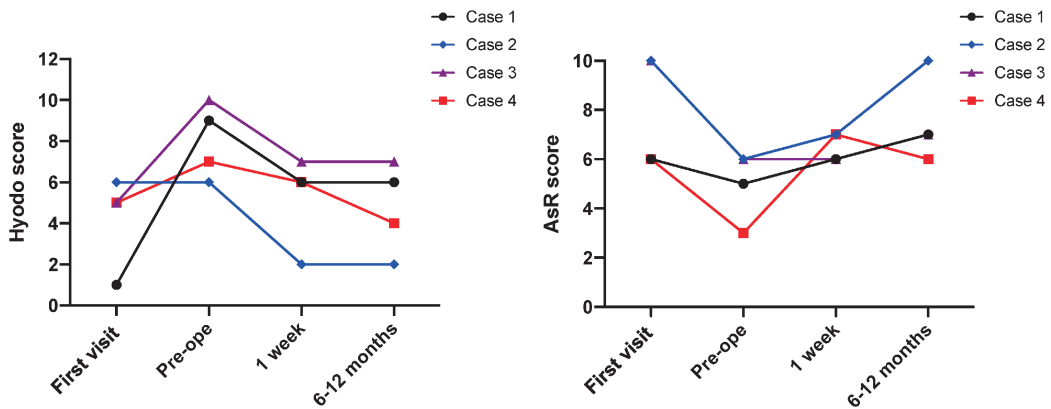
**Fig. 1b:** Masson's trichrome staining reveals endomysial fibrosis and fatty replacement.

(Case 3) developed a postoperative local fistula, but did not require surgical treatment.

Pathological assessments of the resected pharyngeal muscle were performed in two patients (Cases 1 and 4). Hematoxylin and eosin staining revealed atrophic fibers and immune cell infiltration into the muscle endomysium (Fig. 1a). Masson's trichrome staining revealed endomysial fibrosis and fatty replacement (Fig. 1b).

#### *Changes in swallowing function before and after cricopharyngeal myotomy*

Figure 2 shows changes in the Hyodo and AsR scores during the long-term follow-up period. Preoperatively, the average Hyodo and AsR scores were 8 (range: 6–10), and 5 (range: 3–6), respectively. These scores improved postoperatively in all patients. Moreover, swallowing function was maintained in all patients even at 6–12 months postoperatively, indicating the efficacy of CPM in treating severe dysphagia in patients with s-IBM. Two years postoperatively, three patients (Cases 1, 2, and 4) comfortably maintained oral intake, but one (Case 3) required a feeding tube due to rapid disease progression.

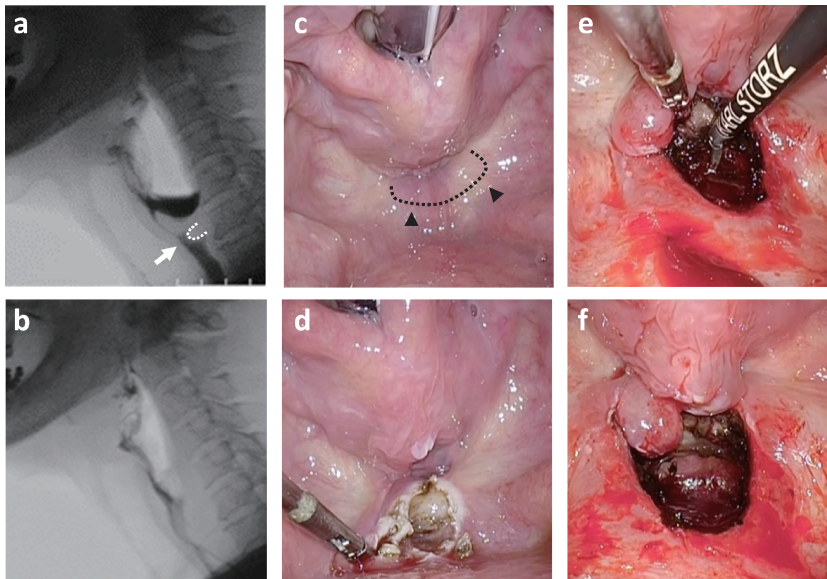


**Fig. 2** Changes in the Hyodo, and AsR scores

All four patients with sporadic inclusion body myositis (s-IBM) achieved improvement in swallowing function and food intake postoperatively. The swallowing function was maintained in all patients, even 6–12 months postoperatively.

#### *Illustrative case (Case 4)*

A 73-year-old woman presented with a 5-year history of weakened handgrip strength and a 2-year history of gait disturbance. Experienced neurologists diagnosed s-IBM based on the clinical characteristics and muscle biopsies. Five years after the diagnosis, the patient developed swallowing dysfunction and visited our department for assessment. VE revealed severe retention of colored water at the vallecula and piriform sinuses, and VF demonstrated pooled barium due to incomplete opening of the upper esophageal sphincter and cricopharyngeal bar (Fig. 3a). The patient had a preoperative Hyodo score of 7 and underwent endoscopic CPM at the age of 79 years (Fig. 3c–f). No surgical complications were observed peri- or preoperatively. VF on the sixth postoperative day showed disappearance of the cricopharyngeal bar and reduced barium residue in the hypopharynx (Fig. 3b). The Hyodo score improved to 6. The patient was able to ingest normal food orally postoperatively and was discharged on the ninth postoperative day. One year postoperatively, the patient had maintained oral intake, and the Hyodo score had improved to 4.



**Fig. 3** Illustrative case in which the patient underwent endoscopic cricopharyngeal myotomy (Case 4) Preoperatively, barium was pooled in the hypopharynx due to the incomplete opening of the upper esophageal sphincter and cricopharyngeal bar (a). Postoperatively, the cricopharyngeal bar disappeared, and barium residue in the hypopharynx had reduced (b). Surgical procedures for endoscopic cricopharyngeal myotomy are shown (c–f). Arrow: cricopharyngeal bar. Arrowheads: upper esophageal sphincter.

## DISCUSSION

Herein, we retrospectively reviewed our experience with using endoscopic and transcervical CPM in four patients with IBM, with a follow-up period of over 10 years. The average duration from disease onset to surgery was 13.2 years in our study. All patients achieved an improvement in swallowing function and enjoyed oral food intake up to 1 year postoperatively, suggesting the safety and efficacy of CPM for swallowing function in patients with s-IBM. Table 3 shows the clinical considerations for performing CPM in patients with s-IBM, based on the nature of s-IBM (ie, slowly progressive disease). To our knowledge, this was the first study to report the efficacy of modified endoscopic CPM using a curved rigid laryngoscope in patients with s-IBM. This approach offers several advantages over conventional techniques, such as the avoidance of incision and no risk of recurrent laryngeal nerve injury. Although general anesthesia and hospitalization are required perioperatively, CPM is considered an essential surgical procedure that improves swallowing function and oral intake in patients with s-IBM. Considering the progression of the disease, preoperative swallowing function, age, general condition, and the patient's thoughts, the surgical indication for CPM should be carefully determined by patients, their families, and a multidisciplinary team.

In our study, the swallowing function gradually worsened between the first visit and the preoperative period. Although the surgical indications for CPM in patients with s-IBM remain unclear due to the slow progression of the disease, one study on endoscopic swallowing examinations demonstrated that a Hyodo score of  $> 6$  was a strong predictor of aspiration.<sup>14</sup> Moreover, Kimura et al revealed that a Hyodo score of 6 was suitable for predicting whether patients with aspiration pneumonia required tube feeding.<sup>15</sup> In fact, all four patients in this study often

**Table 3** Surgical strategy for cricopharyngeal myotomy in patients with sporadic inclusion body myositis

Factors	Considerations
Patient and family	Willingness to undergo surgery
	Stable general condition
	Swallowing dysfunction for normal food
	Desire for oral intake
	Repeated aspiration pneumonia
Swallowing assessment	Requirement of frequent suction
	Videoesopic examination of swallowing (VE)
	Videofluoroscopic examination of swallowing (VF)
Staff	Manometry (if possible)
	Sufficient swallowing management training provided to all staff
	Experienced surgical team available

suffered from dysphagia and their average preoperative Hyodo score was 8 (range: 6–10). Taken together, the evidence suggests a Hyodo score of 6 may be a surgical indication for CPM in patients with s-IBM.

CPM is a surgical procedure that involves cutting the muscles from within (via an endoscopic approach) or outside (via a transcervical approach) the pharynx and reducing the pressure during oral intake, and is a useful treatment option for dysphagia in patients with s-IBM.<sup>16</sup> A recent review indicated that approximately 60% of patients with s-IBM who undergo myotomy report benefit.<sup>17</sup> Furthermore, Oh et al reviewed 18 patients with s-IBM who underwent one or more interventional procedures and reported that symptomatic improvement was noted with CPM (63%) and balloon dilation (33%).<sup>4</sup>

The occurrence of dysphagia in s-IBM is attributed to disordered pharyngeal constriction, inability to relax the cricopharyngeal muscle, and disturbances in laryngeal elevation.<sup>18</sup> In our study, three patients (Cases 2, 3, and 4) underwent endoscopic CPM only and one underwent transcervical CPM and laryngeal suspension as VF imaging revealed poor elevation of the larynx. One study demonstrated no significant differences in recurrence rates, complications, hospitalization duration, surgical duration, or return to preoperative diet between transcervical CPM and endoscopic CPM.<sup>19</sup> Although transcervical and endoscopic CPM are considered safe and effective approaches, endoscopic CPM is the recommended first choice of treatment for dysphagia in patients with s-IBM due to being less invasive. In our study, the surgeons took approximately 2 hours to perform transcervical and endoscopic CPM, and no severe perioperative complications occurred, demonstrating its safety in patients with s-IBM.

VF is the gold standard for assessing swallowing function. A cricopharyngeal bar is defined as a marked protrusion lacking relaxation and stricture of the upper esophageal sphincter on videofluoroscopy and is indicative of IBM with high specificity (96%).<sup>20</sup> Moreover, manometry is often performed to assess swallowing function. Murata et al demonstrated the efficacy of a combination test using VF and manometry to assess the preservation of sphincter muscle strength and the dysphagic processes occurring subclinically in patients with s-IBM who may not report swallowing impairments.<sup>18</sup> When planning CPM in patients with s-IBM, clinicians should carefully review the patient's disease history and VF as it is unsuitable for patients with hiatal hernia. A case report presented a patient with s-IBM and an incidental hiatus hernia who underwent CPM, resulting in aspiration pneumonia and assisted ventilation.<sup>21</sup> Thus, in a clinical

setting, preoperative assessment of swallowing function using VF is essential in patients with s-IBM scheduled to undergo CPM. Patients with dysphagia require multidisciplinary management to receive appropriate rehabilitation and treatment for swallowing dysfunction, which can aid in determining the surgical indications for CPM. Further multicenter prospective studies with larger numbers of patients are needed to support our findings.

In conclusion, CPM may be a safe surgical procedure with the potential to improve swallowing function in patients with s-IBM. Further, a Hyodo score of 6 may be used as a surgical indication for CPM in patients with s-IBM.

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## ETHICS APPROVAL

This study was approved by the Ethics Review Committee of the Nagoya University Hospital (2022-0061) and performed in accordance with the ethical standards of the 1964 Declaration of Helsinki and its later amendments.

## DISCLOSURE STATEMENT

The authors have no financial conflicts of interest to declare.

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