Rehabilitation after Treatment for Jugular Foramen Lesions

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ABSTRACT

Swallowing problems following jugular foramen surgery are more common than is often acknowledged and affect up to a third of our patients. They have a significant effect on quality of life. We have become more proactive in this respect and anticipate these problems before they become established. In this article we present our management protocol that has evolved over the past 30 years as a result of our experience treating 134 glomus jugulare tumors. Our current protocol involves a thorough preoperative assessment of swallowing. After jugular foramen surgery, patients undergo further evaluation using fiberoptic endoscopic evaluation of swallowing (FEES), videofluoroscopy, and manometry. Those with prolonged or poorly compensated dysphagia are offered rehabilitation surgery. We describe this technique, which has proved beneficial to our patients. Guidelines for management are proposed.

KEYWORDS: Skull base, paraganglioma, dysphagia, surgical rehabilitation of swallowing

Disturbance of swallowing following lateral skull base surgery is widely under-recognized. It is well known that even the most experienced clinicians can fail to identify aspiration in 40% of patients.^{1,2} The literature regarding complications of lateral skull base surgery tends to focus on hearing and facial nerve function. Many authors^{3–14} mention the morbidity that can result from damage to the lower cranial nerves, but remarkably little outcome data exist from the patient's point of the view.

There are several reasons for these apparent discrepancies. Facial weakness and deafness are very obvious complications to both the doctor

and the patient. They are the traditional concerns of most otologically based surgeons. Dysphagia is generally an under-recognized complication unless it is severe enough to cause aspiration and subsequent aspiration pneumonia. Furthermore, disturbed nutrition and weight loss tend to develop after discharge from the hospital and may be accepted by patients as the inevitable result of the major surgery they have undergone. Consequently, they may be reluctant to complain. The usual questions asked by most doctors regarding "any other problems" may fail to elicit the very significant deterioration in quality of life suffered by the patient.

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Briner et al,⁴ in reporting the long-term results of the Zurich series, noted a 33% increase in dysphagia and a 19% increase in hoarseness immediately after surgery. Two years after surgery, 97% of these patients considered their neurological deficit acceptable, but, when asked for their subjective views, 36% complained of difficulty in swallowing. This was by far the major subjective complaint. Such discrepancies between the responses to medical questionnaires and more appropriate quality-of-life questionnaires are common.

The senior author (A.C.) has experience with lateral skull base surgery extending more than 30 years. In the early part of his career, he was only aware of swallowing complications when they were severe enough as to warrant a tracheostomy to protect the airway. The occasional use of nasogastric feeding in the postoperative period was seen to be a temporary problem that would soon be overcome by neurological compensation for what was only a unilateral neurological deficit. Some 10 to 15 years later, he developed a separate interest in laryngopharyngeal rehabilitation and started to work very closely with colleagues in speech and language therapy. This different experience radically altered his approach to the assessment and management of lateral skull base problems that involved the jugular foramen. This approach is the subject of this article.

PATIENT SERIES, EVALUATION, AND MANAGEMENT

Patient Data

The patients studied were drawn from a heterogeneous group of 135 tumors involving the jugular foramen, with 116 glomus jugulare tumors, 16 neuromas, and 3 meningiomas. In addition, 12 other patients who had swallowing problems unrelated to jugular foramen pathology contributed to the development of our approach to the rehabilitation of voice and swallowing.

The glomus jugulare tumors were managed using three different strategies. The first group of 31 glomus jugulare tumors were treated between 1974 and 1989.¹⁵ The majority were advanced tumors (67% Fisch class C and D tumors).¹⁶ Swallowing and voice problems were symptomatic in 10%. Preoperative neuropathies of the jugular foramen occurred in 32%, and this increased to 67% postoperatively. In the immediate postoperative period, nasogastric tubes were necessary in only two patients and only one tracheostomy was necessary. Subsequently, three vocal fold medializations and one cricopharyngeal myotomy were performed. The quality of life assessment was very basic, and we reported 85% as having a good result with acceptable disability (no specific data were recorded regarding voice and swallowing.)

A second group of 61 glomus tumors was added in 1994,¹⁷ and by this period we were more aware of the importance of patient outcomes. Preoperative jugular neuropathies had decreased to 16%, which reflected a greater number of smaller tumors. Similarly, the postoperative neuropathies decreased to 30%.

The third group of glomus jugulare tumors (24 patients) was managed according to our current treatment protocol, which aims to preserve jugular foramen function. In this group, five patients had jugular neuropathies preoperatively but only three agreed to a surgical resection that included total tumor clearance from the jugular foramen. The other two cases were treated with primary radiotherapy. Surgical rehabilitation was necessary in 9 of 24 patients, including one patient whose tumor was treated by primary radiation. In five patients, the tumor was found to involve the neural compartment, and a conservative resection was performed with postoperative radiation. None of these five patients suffered swallowing or voice problems during the time course of our follow-up.

In patients with jugular foramen schwannomas,¹⁸ 35% complained of swallowing and voice problems preoperatively and 70% had clinical neuropathies. Postoperatively there were no increased neuropathies and remarkably little morbidity.

Evaluation of Swallowing

With the help of speech and language therapists, the assessment of swallowing has become more comprehensive. As well as taking a detailed history and conducting a neurological examination, a clinical assessment of swallowing is performed. This is then generally supplemented by an instrumental assessment.

METHODS OF ASSESSMENT¹⁹

History A detailed history of dysphagia symptoms and oral intake is essential and must be acquired before examination of swallowing. The patient may report significant dysphagia before surgery, including the spontaneous use of compensatory strategies such as altered head positioning during swallowing or modified oral consistencies. This information aids the construction of postoperative strategies.

The patient's postoperative perception of swallowing—such as ability to swallow and clear secretions, normality of voice, and strength of cough—may provide early indicators of the level of swallowing impairment in the days following surgery. This information can be used to guide the timing of clinical and instrumental swallowing investigations.

Clinical Clinical assessment of swallowing is an important prerequisite to instrumental examination and should include assessment of the following:

- Saliva and secretion management (noting drooling, wet voice, expectoration of saliva, persistent cough, or throat clearing).
- Screening of cranial nerve function by assessment of the appearance and movement of the jaw, face, tongue, and palate. The range, strength, symmetry, and coordination of movements are noted. Orofacial sensation is also assessed.
- Perceptual voice assessment provides information about vocal function and strength of cough.
- Coordination of laryngeal movements by palpating the suprahyoid musculature, hyoid and thyroid cartilage during swallowing of saliva.

- Oral food and fluid trials if saliva clearance, airway protection and alertness are assessed as adequate.

Fiberoptic Endoscopic Evaluation of Swallowing

Fiberoptic endoscopic evaluation of swallowing (FEES) is normally performed at the bedside and provides information about palatal and laryngopharyngeal physiology, sensation, secretion management, pharyngeal clearance, and aspiration.²⁰ It is used to assess the benefit of compensatory strategies (such as head rotation during swallowing and airway protection maneuvers) and as a biofeedback tool during therapy. Although invasive, it is safe when undertaken by trained clinicians. This technique does not require ingesting contrast agents or exposing the patient to radiation.

Videofluoroscopy Videofluoroscopy¹⁹ is used to assess the oral, pharyngeal, and esophageal stages of swallowing. It provides more detailed information about swallowing pathophysiology and the impact of disordered swallow physiology on bolus flow and airway protection. This information enables the speech and language therapist to implement appropriate therapy and to recommend compensatory strategies to improve bolus clearance and airway protection. The oropharyngeal swallow efficiency (OPSE) scores were calculated from the image data.

Manometry Pharyngeal and upper esophageal manometry enabled objective measurement of pressure generated during swallowing in a select group of patients. A catheter containing solid-state sensors is passed transnasally, enabling measurement of pressure at the base of the tongue, hypopharynx, and upper esophageal sphincter during swallowing.²¹ This information is used to quantify the amount of pressure generated, to identify the sequencing of muscle activity within the pharynx during swallowing, and to determine appropriate treatment strategies. Pharyngeal manometry is usually performed concurrently with videofluoroscopy as it facilitates analysis of the relationship between pressure generation, structural movements, and bolus flow.²²

QUALITY OF LIFE

The patient's perception of his or her swallowing function may not always correlate with the clinician's assessment of the patient's dysphagia severity. Documentation of the patient's perception of his or her swallowing function and involvement of the patient in oral intake and dysphagia treatment decisions is critical.

Several relevant quality-of-life tools are available, including the SWAL-QOL and the Performance Status Scale for Head and Neck (PSS-HN).²³ The SWAL-QOL is a validated tool that provides detailed information about the patient's perception of his or her swallowing function and the impact of dysphagia on quality of life. This is the most comprehensive swallowing-specific quality-of-life tool available; however, the time required for completion and data analysis precludes its repeated use during routine clinical examinations. The PSS-HN is quick to complete and therefore easily repeated within the clinic. It provides information about consistencies of food in swallowing, eating environments, and the intelligibility of speech.

Combining clinical and instrumental assessments of swallowing with selective use of quality-of-life tools enables integration of patients' perceptions of their swallowing function and treatment preferences into the dysphagia rehabilitation program.

We have also used our own Dysphagia Index (Table 1) to assess progress and record outcome. Although it has not been fully validated, we find it useful in the clinical situation.

Management of Dysphagia

PRINCIPLES OF SWALLOWING THERAPY

The several aspects of our swallowing therapy principles can be summarized as follows:

- Educating the patient about the nature of the dysphagia and the benefits of compensatory strategies and therapy. Biofeedback is an extremely effective method of increasing patient awareness and cooperation.

Table 1 Dysphagia Index

0 Normal

- Eats full range of boluses with variable sizes and consistency
- Normal meal duration (similar to rest of family)

1 Problematic with compensation

- Adequate nutrition to maintain body weight
- Acceptable meal duration (< 30% longer than rest of family)
- Occasional aspiration (< 1 x per week)
- Uses modified diet and postural strategies

2 Problematic noncompensated

- Inadequate oral nutrition to maintain body weight
- Unacceptable meal duration (> 30% longer than rest of family)
- Frequent aspiration uncontrolled by postural strategies
- Nutrition maintenance with PEG feeds
- Informed and alert patient can take some oral feeds for QOL purposes

3 Unsafe swallowing

- Overt aspiration of all consistencies
- History of recurrent chest infections
- Need total PEG nutrition

PEG, percutaneous endoscopic gastrostomy; QOL, quality of life.

- Recommending the most appropriate food and fluid consistencies. This is designed to maximize swallowing efficiency (and hence oral intake) and to protect the airway.
- Teaching compensatory strategies to improve bolus flow and protect the airway.
- Implementing a program of exercises aimed at compensating for the pathophysiology (e.g., reduced laryngeal elevation and closure, impaired movement of the tongue and pharyngeal musculature, and reduced opening of the upper esophageal sphincter).

SURGICAL REHABILITATION

Surgical rehabilitation is limited to the correction of morphological changes that result from the various neuropathies. The potential morphological changes resulting from dysfunction of the various nerves related to the jugular foramen are listed in Table 2.

Table 2 Laryngopharyngeal Changes Following Total Vagus Palsy

Palate
<ul style="list-style-type: none"> • Incompetence of velopharyngeal sphincter
Larynx
<ul style="list-style-type: none"> • Failure in laryngeal elevation • Paralysis of vocal fold in paramedian position • Anterior subluxation of arytenoid with lowering of vocal fold • Wasting of vocal fold • Loss of laryngeal sensation
Pharynx
<ul style="list-style-type: none"> • Constrictor paralysis • Ballooning of pyriform fossa • Incoordination of upper esophageal sphincter

With incomplete neuropathies, isolated changes may exist; however, with extensive involvement of the foramen, multiple changes are generally present.

Many surgical procedures have been described (Table 3) to rehabilitate swallowing where swallowing therapy has had limited success in remedying severe dysphagia. None of our patients have required palatal surgery. We have tried most forms of vocal fold medialization, and most gave suboptimal results. As a result, we prefer to medialize and elevate the paralyzed fold to its normal adducted position by performing an arytenoidopexy, originally described by Montgomery.²⁴ Laryngeal elevation was used in patients where videofluoroscopy showed that it was deficient. In such cases, we invariably performed an upper esophageal myotomy, incising the full length of the upper esophageal sphincter rather than the more com-

monly described, but limited, cricopharyngeal myotomy.

Over the last few years we have tended to use a single-stage rehabilitative procedure to correct the different morphological problems commonly found with a complete jugular foramen syndrome.

SINGLE-STAGE SURGICAL REHABILITATION OF SWALLOWING PROCEDURE

The parapharyngeal space is opened on the paralyzed side using a cervical approach. Dividing the thyropharyngeus muscle just posterior to its insertion into the oblique line exposes the posterior rim of the thyroid cartilage. The dissection is continued round the edge of the thyroid lamina to elevate the mucosa of the pyriform fossa away from the posterolateral aspect of the cricoid cartilage. This exposes the posterior cricoarytenoid muscle so the cricoarytenoid joint space can easily be palpated through the muscle fibers. The muscle fibers are split and the joint space opened to expose the anteriorly prolapsed arytenoid cartilage. A 2-0 Ethibond suture (Ethicon, Somerville, NJ) is then passed through the posterior lamina of the cricoid to enter the joint space. The needle is then passed through the body of the arytenoid. When the suture is tightened, the arytenoid is repositioned and fixed in its normal closed position toward the apex of the cricoid lamina. If wasting of the vocalis muscle has developed, the normal bulk of the vocal fold is restored by creating a subperichondrial tunnel deep to the thyroid lamina at the level of the vocal fold. This tunnel is then packed with an appropriate amount of Gortex sheet to restore the contours of the vocal fold, which is checked by a perioperative fiberoptic endoscope. The freed mucosa of the pyriform fossa is then plicated extramucosally to obliterate the bulging pharyngeal cavity. The thyropharyngeus is resutured into its normal position and then an upper esophageal sphincter myotomy is performed. The larynx is elevated by passing a Gortex suture around the body of the hyoid bone and then through a small tunnel drilled in the mandibular symphysis.

Table 3 Surgical Options for Laryngopharyngeal Rehabilitation

- Palatoplasty
- Medialization of vocal fold
- Arytenoidopexy to medialize and elevate vocal fold
- Augmentation of vocal fold
- Pharyngoplasty
- UES myotomy
- Laryngeal suspension

RESULTS

The results of neural deficits associated with jugular foramen tumors and their surgical resection on the process of swallowing are detailed in Table 4.

Preoperative videofluoroscopy of the nine patients that required surgical rehabilitation showed the following common features:

- Oral tongue weakness secondary to unilateral XII impairment with compensation from the unaffected side. Slow bolus formation and mild oral residue after swallowing were seen.
- Significant weakness of the tongue base with incompetence of posterior movement and, hence, reduced or absent contact against the posterior pharyngeal wall. This resulted in vallecular residue in all cases and significantly reduced pharyngeal clearance of all consistencies.
- Moderate reduction of laryngeal elevation and anterior tilt of the hyoid, resulting in compromised airway protection during swallowing (compounded by the vocal fold palsy) and reduced opening of the upper esophageal sphincter. This contributed to hypopharyngeal clearance problems and the need for multiple swallows to clear each bolus.
- Marked pharyngeal constrictor weakness and reduced pharyngeal shortening during swallowing. In all cases, multiple swallows were required to swallow each bolus. In some cases, this improved with therapy and time, but in one case this has remained a significant impairment.

Table 4 Effects of Cranial Nerve Deficits on Swallowing

VII	Disturbed oral phase Disturbed laryngeal elevation
IX	Loss of sensory input to swallowing reflex Disturbed pharyngeal phase
X	Loss of sensory input to swallowing reflex Loss of sensory input to larynx Disturbed motor function of palate/pharynx/larynx
XII	Disturbed oral phase Poor initiation of pharyngeal phase

Figure 1 Decrease in Dysphagia Index in nine patients undergoing single-stage surgical rehabilitation of swallowing.

- Promptly triggered swallow in all cases, with no pharyngeal delay time.
- Transit times within normal limits in most cases. Low OPSE scores in some of the videofluoroscopies were due to poor clearance of the bolus on the first swallow rather than slow oral or pharyngeal transit. All of the subjects required multiple swallows (up to 11) to clear most of the bolus; hence the overall efficiency of the swallow, in the early postoperative videofluoroscopies, was poor. Postoperative OPSE figures showed improvements, but they did not reach normal.

The results of the patients undergoing single-stage rehabilitation are shown in Fig. 1, which uses the Dysphagia Index to show the resulting improvements to their quality-of-life measures. All patients achieved a worthwhile improvement.

DISCUSSION

The incidence and severity of the swallowing problems that develop from pathology involving the jugular foramen is well documented in the literature. Many groups have studied their patients with videofluoroscopy and their findings are similar to those presented here. Many authors emphasize the importance of active collaboration with speech and language therapists in the management of these patients. There has, however, been little discussion regarding the different problems that exist on presentation and those that follow surgery.

Table 5 Degrees of Nerve Injury Classified by Sunderland²⁵

First degree (neuropraxia)	Physiological conduction block
	No Wallerian degeneration
	Complete recovery
Second degree (axonotmesis)	Axon damage with peripheral Wallerian degeneration
	Intact endoneurial tube allowing axonal regeneration
	Complete recovery
Third degree (neurotmesis)	Axon and endoneurial tube damaged with loss of continuity
	Perineural sheath preserved
	Regeneration incomplete and imperfect
Fourth degree (neurotmesis)	Funicular sheaths ruptured
	Loss of continuity with disorganized scar tissue
	Regeneration very poor
Fifth degree (neurotmesis)	Complete severance of nerve
	Complete loss of continuity
	No regeneration in absence of reconstruction

The difference depends on the type of neural damage sustained by the nerves. Sunderland²⁵ classified nerve damage according to the degree of anatomical damage (Table 5). His first and second degrees of injury, neuropraxia and axonotmesis, respectively, generally follow sustained compression of the nerve such as results from tumor growth. Both of these injuries have good potential for regeneration. Makek et al²⁶ demonstrated four histological grades of neurological infiltration of cranial nerves with glomus temporale tumors. The degree of infiltration increased with the size of the tumor, and they proposed a progressive involvement along the nerve first of the perineurium and subsequently the endoneurium. The nerve axons themselves remain viable. The authors also showed neural infiltration occurred frequently in the absence of preoperative nerve dysfunction. Because glomus tumors are slow growing, the gradual loss of neural function resulting from tumor compression allows neurological compensation in swallowing to develop; such patients are often asymptomatic. This is the common situation seen on presentation. Con-

versely, Sunderland's three degrees of neurotmesis often result from some form of trauma such as resection of tumor from the foramen. There is an acute loss of neural function, absence of neurological compensation, and poor chance of regeneration leading to muscle atrophy.

Another factor to be considered is the number of neurons damaged by slow tumor growth. It is known that a high percentage of neurons may be lost before clinical dysfunction is noted. This explains the acute severe loss of function that sometimes happens following limited tumor dissection of tumor in the neural compartment.

We feel this increased morbidity resulting from the disturbance of swallowing after tumor clearance from the jugular foramen is of prime importance to our patients' quality of life. Accordingly we have elected in the absence of a preoperative neural deficit to leave any tumor involving the neural compartment and to use postoperative irradiation to control the residual tumor. We have now treated 26 patients in this way and have only had significant recurrence in one patient. If the patient has evidence of a swallowing disorder preoperatively, we recommend total surgical resection with subsequent rehabilitation as necessary. In this situation patients will occasionally elect for primary radiation therapy.

Makek et al²⁶ showed that previous irradiation did not protect the nerves from subsequent tumor infiltration; however, increasing reports^{16,27-30} show good tumor control with focused irradiation and low morbidity in terms of swallowing. It is possible that the low morbidity is due to the slowed growth, allowing compensation.

We still feel that glomus tumors should be treated primarily with surgery on sound radiotherapeutic principles. These suggest that with radio-resistant tumors, reduction in tumor size facilitates outcome, and our experience shows that a better neurological outcome also results.

Considering the surgical rehabilitation of swallowing, most authors have concentrated on various cord medialization procedures; however, there is little agreement about the efficacy of any

particular approach. This is not surprising because most cord medializations are performed for paralysis following recurrent laryngeal nerve palsy. In this situation, neither arytenoid prolapse nor wasting of the vocal fold is common and the results in voice and cough restoration are good. Our experience with these procedures in complete vagal palsy shows less than optimal improvements, and this is the reason for recommending arytenoidopexy. Zietels et al³¹ recommend a similar procedure.

The work published on both laryngeal elevation³² and pyriform fossa obliteration, although sparse,^{8,33} supports the value of these procedures. Regarding the timing of surgical correction,³⁴ we feel strongly that this should be delayed for 6 months to allow the benefits of neural compensation and swallowing therapy to have an effect. We performed early rehabilitation in one patient because she had significant problems on presentation. Surgery was notably difficult to perform because the muscles had not wasted and it was not easy to judge the right amount of correction to her disturbed morphology. Although she obtained a good voice immediately, her swallowing therapy was prolonged and the end result was considered suboptimal.

Patients that have a high probability of swallowing problems following treatment need the early intervention of speech and language therapists. Their postoperative involvement will improve patient outcome and reduce recovery time in the hospital. We recommend a systematic approach to these problems and include some guidelines for their management.

MANAGEMENT GUIDELINES FOR SWALLOWING PROBLEMS RELATED TO JUGULAR FORAMEN PATHOLOGY

Preoperative Assessment

Preoperatively, it is important to establish whether there is any evidence of disturbed vocal and swallowing function. Clinical assessment of the cranial

nerves alone is not adequate; specific questions concerning usual diet and duration of eating are equally important. For example:

- Are you concerned about your swallowing?
- Do you eat the same food as the rest of your family? Do you avoid certain foods?
- Do you cough or choke when you eat and drink?
- Does food get stuck in your throat? Do you cough up pieces of undigested food?
- Do you finish your meal in the same time as the rest of your family?

If the size of the tumor predicted by imaging is greater than Fisch type C3 and any degree of intracranial tumor is found, there is a high chance of tumor involvement of the lower cranial nerves. A speech and language therapist should assess such patients clinically. If any disturbed function is suspected, instrumental assessment of swallowing (FEES/videofluoroscopy) is essential to allow an informed decision about treatment.

Perioperative Assessment

During dissection of the jugular bulb, care must be taken not to inadvertently damage the medial wall, which is immediately in contact with the neural components. Excessive diathermy in this area or overpacking of the inferior petrosal sinus may also damage the neural elements.

If preoperative evidence of swallowing dysfunction was found or the surgical exploration includes a dissection of the jugular neural compartment, a fine-bore nasogastric tube should be passed at the completion of the operation to facilitate tube feeding in the early postoperative period.

Postoperative Assessment

All patients should have their swallowing assessed postoperatively by a speech and language therapist before commencing oral nutrition. Assessment is appropriate when the patient is alert, stable, and able to sit upright. If the patient experiences

preoperative dysfunction, clinical assessment supplemented by FEES assessment at the bedside is advised. Evidence of significant dysphagia and aspiration requires oral nutrition to be started cautiously under supervision, and, if significant, a fine-bore nasogastric tube should be passed. Tube feeding should then be started (under supervision of the dietician). Swallowing therapy should commence at this stage, and the nasogastric tube can be removed as soon as the patient is capable of meeting nutrition and hydration requirements orally. Careful management of oral and nasogastric tube intake in the early days postoperatively is important to prevent dysphagia-related morbidity (e.g., weight loss, aspiration pneumonia, and dehydration).

Natural compensation will generally be recognized in 7 to 10 days. If at this time no progress has been achieved, a more intensive rehabilitation program must be commenced. The first step is to perform a percutaneous endoscopic gastrostomy (if the gastrostomy is performed under a general anesthetic, we temporarily medialize a paralyzed vocal fold with a Gelfoam injection). This allows removal of the nasogastric tube. There are considerable psychological advantages to this step, and removing the nasogastric tube from the pharynx may facilitate more effective neurological compensation.

With the gastrostomy established, the patient can leave the hospital and begin a supervised program of rehabilitation. A formal videofluoroscopy at this stage is essential for several reasons:

- Identification and documentation of pathophysiology
- Assessment of the efficacy of compensatory strategies (such as head positioning and airway protection maneuvers)
- Identification of therapy approaches to rehabilitate physiology and improve swallow efficiency
- Increasing the patient's insight into the nature of the swallowing dysfunction and the benefits of compensatory strategies and therapy
- Providing a baseline to chart progress
- Planning any surgical rehabilitation, which is limited to correcting abnormal morphology

Continued swallowing therapy is given over the next 3 months, and dietetic assistance is sought to ensure that gastrostomy feeding proceeds without problems.

At 3 months, we expect to see signs of recovering function; further videofluoroscopy studies are performed. In the absence of significant improvement, we warn the patient that surgical rehabilitation may be necessary, but we always wait for at least 6 months before undertaking any surgery.

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