Total Laryngectomy in Patients with Advanced Bulbar Symptoms of Amyotrophic Lateral Sclerosis

MAYO CLINIC

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Abstract

Objectives: To (i) identify patients with amyotrophic lateral sclerosis (ALS) who would benefit from a total laryngectomy (TL) and outline specific surgical indications; (ii) educate physicians about the surgical procedure, peri-operative course and benefits from having a TL; and (iii) retrospectively review the clinical course of Mayo Clinic-Jacksonville patients with ALS who had a TL.

Methods: A retrospective review of four patients who underwent TL for advanced bulbar symptoms related to ALS at the Mayo Clinic in Jacksonville, Florida.

Results: Between January 1999 and July 2008, fourteen patients with severe bulbar symptoms associated with ALS were recommended to undergo TL. Only four patients (28.6%) opted for the surgery. All patients were aphonic at time of surgery with a multitude of bulbar symptoms. Average surgical time was 102 minutes (range 87-116 minutes). No intra-operative complications were reported. The median hospital stay was thirteen days (range 5-28 days). All patients and caregivers were pleased with the results of the TL. Benefits reported were: (i) elimination of choking spells and pulmonary infections, (ii) improved comfort with mechanical ventilation, (iii) safe and easy tracheostomy tube changing and cleaning, (iv) less hospitalizations, (v) increased oral intake, (vi) reduced anxiety of aspiration, and (vii) less coughing.

Conclusion: TL is a relatively safe, quick and uncomplicated surgical procedure that should be considered earlier and more frequently in the treatment plan of patients with advanced bulbar symptoms of ALS. Patients with pre-operative respiratory insufficiency may experience prolonged recovery times.

Introduction

Amyotrophic Lateral Sclerosis is the most common adult motor neuron disease with an incidence between 1 and 5 per 100,000 of the general population with a slight male preponderance.[1,2,3] It usually follows a rapidly progressive course with median time of onset of symptoms to death ranging from 23-48 months.[4] Five and ten year survival rates range from 9-40% and 8-16%, respectively.[4,5]

20-30% of ALS patients will present with bulbar dysfunction, typically involving muscles of the face, tongue, pharynx and larynx. Patients with advanced bulbar symptoms suffer from severe dysphonia, dysphagia, drooling and are at significant risk for malnutrition, chronic cough, recurrent choking episodes, and aspiration pneumonias.

The ALSAQ-40 and the ALS-Specific Quality of Life Questionnaire (ALSSQOL) are two validated instruments designed to measure QOL parameters in patients with ALS.[4,6,7] Numerous studies have demonstrated that patients with advanced ALS, even when they are ventilator dependent with extreme physical weakness, can still maintain high QOL standards.[4,8-11]

The purpose of this article is to review the surgical indications, timing and post-operative course of patients who underwent TL at our institution for advanced bulbar ALS symptoms. Additionally, we will argue that despite a typically rapid, progressive disease that is ultimately fatal, ALS patients can still maintain a relatively high QOL and TL can be a safe surgical alternative to alleviate some of the physical and emotional discomfort experienced by both patient and caregiver.

Methods

A retrospective chart review was performed from January 1999 to July 2008 on ALS patients at the Mayo Clinic in Jacksonville, Florida. In this timeframe, fourteen patients with advanced bulbar symptoms were identified as appropriate candidates for TL. Indications for TL were unintelligible speech with chronic aspiration. As of July 2008, only four patients had opted for this surgery.

Results

Between January 1999 and July 2008, fourteen patients with advanced symptoms of ALS at the Mayo Clinic in Jacksonville, Florida were deemed appropriate candidates for TL. Table 1 shows a significant increase in recommended TLs over the past 5 years. Ten patients refused to undergo the recommended TL. Many of these patients gave no specific reason for the rejection. One patient chose the less invasive tracheostomy option and another was afraid of having any surgery. Three patients wanted no mechanical ventilation, refused all interventions and died soon thereafter. Two patients are alive and still considering TL.

Four patients had TLs for amelioration of their bulbar symptoms. Table 2 outlines their demographics, duration of disease prior to TL, and pre-operative symptoms and morbidities.

Operative times, peri-operative complications, hospital stays, and times from TL until death are shown in Table 3. The average surgical time was 102 minutes with no intra-operative complications. Patient A, who was discharged in five days, required no immediate post-operative ventilatory support. Patients B, C and D were mechanically ventilated through their tracheostomas post-operatively. Patient B developed pneumonia post-operatively, likely due to his underlying respiratory insufficiency, requiring an additional three weeks in the hospital. Patient C and D's extended hospital stays were not medically related and due to placement issues with hospice care.

Patient B experienced recurrent stomal granulation tissue formation requiring cauterization approximately every three months by his primary physician.

All four patients and their caregivers were pleased with the overall outcome of the surgery. Patient A reported complete resolution of his choking spells and cough. He was able to increase his oral intake without fear of aspiration. His spouse, the primary caregiver, was extremely grateful. She found suctioning him much easier. Patient B was also able to increase his oral intake. He experienced no further aspiration and found mechanical ventilation through his stoma more comfortable than with the BiPAP mask. Patient C was transferred to a skilled hospice nursing facility post-operatively as planned. She died there 6 months later. Post-mortem, her son was contacted. He expressed gratitude for his mother having the TL. It eliminated her coughing, aspiration and frequent respiratory infections. She was able to resume some oral intake which gave her great pleasure. Patient D and family members were overall satisfied with the surgery. Aspiration was eliminated, coughing reduced, handling of secretions and dyspnea improved, and oral intake resumed.

Table 1

	1999 - 2002	2003-2008
# of patients recommended to undergo TL	2	12
# of patients who had TL	2	2

Table 2

Patient	Gender	Age	Disease duration prior to TL	Pre-operative symptoms and medical problems
A	Male	44	52 months	Aphonic, dyspnea, dysphagia, drooling, choking spells, aspiration pneumonias, severe lower extremity weakness, weak cough
B	Male	41	17 months	Aphonic, respiratory insufficiency (on BiPAP), dysphagia (PEG), aspiration, severe lower extremity weakness
C	Female	62	24 months	Aphonic, dyspnea (unable to tolerate BiPAP), cough, choking spells, dysphagia (PEG), aspiration pneumonias, drooling, COPD, hypertension, OSA
D	Female	61	11 months	Aphonic, dyspnea (unable to tolerate BiPAP), dysphagia (PEG), biphasic stridor, aspiration, cough, drooling, COPD, CAD, HTN

COPD=chronic obstructive pulmonary disease, OSA=obstructive sleep apnea, **CAD**=coronary artery disease, HTN=hypertension

Table 3

Patient	Operative Time	Intra-op complications	ST post-op complications	Hospital Stay	LT post-op complications	Time from TL until Death
A	116 min	None	None	5 days	None	30 months
B	102 min	None	Pneumonia	28 days	Recurrent stomal granulation tissue	36 months
C	101 min	None	None	14 days*	None	6 months
D	87 min	None	None	6 days*	None	Living

ST= short term, LT= long term, * denotes extended stay due to placement issues

Discussion

Among non-otolaryngologists, there is a misconception that TL is a long complex surgery fraught with many potentially serious complications. Most clinicians will instead recommend tracheostomy tube placement instead of TL for patients with advanced bulbar symptoms of ALS. This is often a very reasonable and necessary procedure to secure the airway and provide a comfortable means of mechanical ventilation. It is faster, and less complicated than TL. However, tracheostomy tubes do not prevent aspiration, and generally make aspiration worse.

Our review identified no intraoperative and one short term and long term post-operative complications. The short term complication was pneumonia requiring an extended three week hospitalization to treat. The patient's underlying respiratory compromise was a major contributing factor to this complication emphasizing the necessity for aggressive pulmonary therapy peri-operatively. The long term complication was recurrent stomal granulation tissue formation requiring several cauterizations and local stomal care.

In patients with advanced bulbar symptoms of ALS, we consider the surgical indications and timing of performing TL to be straightforward. As soon as speech becomes unintelligible, the larynx serves no useful purpose and should be removed. Removing the larynx (i) eliminates the risk of aspiration, cough, and choking spells (ii) facilitates tracheobronchial suctioning, (iii) provides a safe and comfortable means for mechanical ventilation, (iv) alleviates the anxiety and potential complications found with maintaining a tracheostomy, and (v) allows resumption of oral intake. There may also be a subset of patients with preserved speech and severe aspiration who would choose a TL to eliminate their aspiration at the expense of speaking again.

Our retrospective analysis of hundreds of ALS patients over nearly a decade identified only fourteen patients who were recommended to undergo TL for their severe bulbar symptoms. So, if TLs can provide such a benefit for these patients, why were so few recommended? Did the vast majority of these patients have such a short life expectancy when their bulbar symptoms developed that a TL was not justified? Did the medical team not fully understand the surgical procedure and the benefits it could provide these patients? Were tracheostomies recommended over TL to solve the obvious mechanical ventilation problem, but overlook the aspiration, choking and coughing issues? Were the QOL studies on ALS patients not available yet or did we not fully comprehend the impact a TL could have to justify the recommendation? These are important questions outside the scope of this paper that certainly warrant further study.

Another interesting finding was that only 4 of 14 patients (28.6%) opted to undergo the recommended TL. In most cases, no specific reason was reported on the chart for rejecting TL. Why do such a small percentage of patients refuse a recommended therapy that will almost certainly improve their QOL? Frontotemporal cognitive impairment has been reported in up to 75% of ALS patients.12-14 Future studies tracking cognitive function will help determine whether this is a factor in ALS patients' ability to make informed judgments regarding TL.

Educating physicians and health care providers of ALS patients about the benefits of TL and the overall improvement in QOL is essential. As Table 1 shows, a disproportionately higher number of TLs were recommended over the past five years, perhaps signifying a better understanding of QOL issues and the benefits a TL can provide these patients.

The medical team caring for ALS patients must be unified in its recommendation for TL. Lack of unity instills doubt, confusion, and reluctance by patients to undergo an elective procedure. Understanding the goals of surgery, the amelioration of multiple symptoms, and the overall improvement in QOL will better equip ALS patients and their family members to make an informed decision to improve their remaining days.

Conclusion

We feel TL is a safe and worthwhile procedure to improve the QOL in ALS patients with severe bulbar symptoms, and should be considered more frequently and earlier in their treatment plans. Educating the medical team caring for these patients about the benefits of TL is essential. All patients and their caregivers were overall pleased with the benefits of TL. The one post-operative respiratory complication is notable and emphasizes the need to manage their compromised pulmonary function peri-operatively. Since our recommendations are based on a small group of patients, a larger study is required to support our findings.

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