Surgical Improvement of Speech Disorder Caused by Amyotrophic Lateral Sclerosis

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Amyotrophic lateral sclerosis (ALS) is a progressive debilitating neurological disease. ALS disturbs the quality of life by affecting speech, swallowing and free mobility of the arms without affecting intellectual function. It is therefore of significance to improve intelligibility and quality of speech sounds, especially for ALS patients with slowly progressive courses. Currently, however, there is no effective or established approach to improve speech disorder caused by ALS. We investigated a surgical procedure to improve speech disorder for some patients with neuromuscular diseases with velopharyngeal closure incompetence. In this study, we performed the surgical procedure for two patients suffering from severe speech disorder caused by slowly progressing ALS. The patients suffered from speech disorder with hypernasality and imprecise and weak articulation during a 6-year course (patient 1) and a 3-year course (patient 2) of slowly progressing ALS. We narrowed bilateral lateral palatopharyngeal wall at velopharyngeal port, and performed this surgery under general anesthesia without muscle relaxant for the two patients. Postoperatively, intelligibility and quality of their speech sounds were greatly improved within one month without any speech therapy. The patients were also able to generate longer speech phrases after the surgery. Importantly, there was no serious complication during or after the surgery. In summary, we performed bilateral narrowing of lateral palatopharyngeal wall as a speech surgery for two patients suffering from severe speech disorder associated with ALS. With this technique, improved intelligibility and quality of speech can be maintained for longer duration for the patients with slowly progressing ALS.

Keywords: amyotrophic lateral sclerosis (ALS); narrowing of lateral palatopharyngeal wall surgery; speech disorder; speech surgery; velopharyngeal closure incompetence


Speech disorder inevitably develops during the course of amyotrophic lateral sclerosis (ALS). Most of the patients with a bulbar onset had the worse prognosis. The median survival is considered as approximately 20 months from the onset of bulbar symptoms, with only 5 per cent survival after 5 years for the patients without the use of artificial ventilation (Donaghy 2001). It was noted that some female patients had slowly progressive course (Nakano 2007). Recent researches reported that some Cu/Zn superoxide dismutase gene mutations might be related to the slow progression or longer survival of ALS (Radunovic and Leigh 1996; Georgoulopoulou et al. 2010). Although ALS is a progressive disease, it may be highly beneficial to improve intelligibility and quality of speech, especially for the patients with slowly progressive course and with fewer disabilities. The patho-physiological features of speech disorder of the patient with ALS were reported as the result of reduced range and velocity of articulatory movement and slowing in the rate of speech synthesis (Darley 1975; Hirose 1986; Duffy 2005). These features of speech disorder are worsened by the progression of respiratory dysfunction. Reduced range of articulatory muscle movement could lead to velopharyngeal closure inadequacy and the reduced lingual dysfunction with atrophic change of the tongue muscles. Limited range of articulatory movement, including velopharyngeal closure and lingual muscle activities could lead to hypernasality, vowel distortion, and imprecise weak articulation during speech. Slow rate of speech synthesis consists of abnormally slow segment and sentence duration with prolonged phonemes, prolonged interval, and inappropriate silence (Darley 1975; Hirose 1986; Duffy 2005). These elements of speech disorder will progress along with

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the progression of ALS within an average of two or three years in many cases. Physical and speech therapies including behavioral modification has not been effective in improving the speech intelligibility of these patients (Duffy 2005; Kühnlein et al. 2008). Surgical or prosthetic interventions for velopharyngeal insufficiency were considered to significantly improve intelligibility and quality of speech by limiting air flow to the nasal cavity (McGuirt 1980). However, the surgical interventions including pharyngeal flap surgery and Teflon injection into the posterior pharyngeal wall are not recommended due to insufficient data to support the efficacy to treat speech disorder caused by ALS (Duffy 2005). In addition, it was previously reported that pharyngeal flap surgery is less favorable than palatal lift prosthesis for dysarthria caused by ALS (Roth et al. 2000; Duffy 2005). The palatal lift prosthesis cannot be worn for the patients who have insufficient supporting dentition, have significant gag reflex, or could not insert and remove the prosthesis by themselves (Roth et al. 2000; Duffy 2005).

The prosthetic management for speech disorder of the patient with ALS is often difficult in many cases, caused by the irritation or the discomfort of the prosthesis due to preserved sensation. In addition, many patients with speech disorder caused by ALS are attributed by reduced movements of the lateral pharyngeal wall along with poor velum elevation. Therefore, the prosthetic and pharyngeal flap surgery alone may be insufficient (Kelsey et al. 1972). ALS is a progressive disease, thus, the efficiency of the treatment should be certain without prolonged post-operative rehabilitation. Here, we performed a simple and effective surgical technique of narrowing of lateral palatopharyngeal wall (Saigusa et al. 1996, 2010) for two patients suffering from severe speech disorder caused by slowly progressing ALS. After the surgery, hypernasality of their speech was decreased, intelligibility and quality of the speech sounds were improved, and the length of generated phrases was longer compared to the ones preoperatively.

Materials and Methods

Prior to surgery, each patient was evaluated by pushing the posterior palatine arch and the soft palate to the pharyngeal wall postero-superiorly during the phonation of non-nasal sound /a/. If the hypernasality of the sound is decreased, the patient is considered for this surgery to correct the velopharyngeal closure inadequacy (compression test) (Saigusa et al. 1996). During the evaluation, the most effective regions for surgical narrowing from the posterior palatine arch area to the soft palate where determined.

This surgery was performed under general anesthesia with no muscle relaxant. The patient’s mouth was held open and the body of the tongue was held depressed, similar fashion to the tonsillectomy. The posterior palatine arch and the pharyngeal wall are injected with 1% lidocaine with 1:100,000 epinephrine. Then, mucosal incisions were made in the posterior palatine arch bilaterally (Fig. 1A). Then, next incisions were made in the posterior pharyngeal wall in parallel with mucosal incisions in the posterior palatine arch bilaterally. The incisions were slightly extended to higher level by hooking the uvula up superior to the level of the posterior palatine arch which are determine by compression test preoperatively. The mucosa of the regions between the incisions in the posterior palatine arch and the posterior pharyngeal wall were resected from the pharyngeal wall bilaterally (Fig. 1B). The pharyngeal constrictor muscle was sectioned along the incision on the pharyngeal wall, and the muscle was elevated from the deep fascia bilaterally. The stripped flaps of the posterior palatine arch were inserted high under the pharyngeal constrictor muscle and sutured with 4-0 vicryl sutures bilaterally (Fig. 1C).

We performed this surgical procedure for two patients with severe speech disorder caused by slowly progressing ALS. We reported clinical course and the condition of speech of each patient before and after each surgery in detail.

Results

Patient 1

A 69-year-old woman with a 6-year course of slowly progressing ALS was referred by a neurologist to improve her speech disorder. Speech disorder was her initial symptom of ALS. Her speech disorder was characterized by
hypernasality, imprecise and weak articulation, reduced volume and rate, and short generating phrases, consistent with mixed flaccid-spastic dysarthria. The degree of her intelligibility rating scale for motor speech disorder (Duffy 2005) was estimated as two. The spirometry revealed the respiratory weakness with 58.4% of normal vital capacity. The oropharyngeal examination revealed velopharyngeal closure incompetence (Fig. 2A, B) and weakness of the tongue with fasciculation and atrophic change. Compression test was positive preoperatively. Lateral fluorography for the velopharyngeal region during speech showed disturbed elevation of the velum (Fig. 3A, B). After performing bilateral lateral palatopharyngeal wall surgery, hypernasality of her speech sound was decreased, and the length of speaking phrase was elongated. She was able to speak with increased volume and clarity after the surgery. The degree of her intelligibility rating scale for motor speech disorder (Duffy 2005) at one month postoperatively improved to five. Postoperative fibrescopic examination revealed the whole velopharyngeal port narrowing and the significantly improved velopharyngeal closure during speech (Fig. 2C, D). Lateral fluorography for the oropharyngeal region during speech showed that the velum was fixed to the posterior pharyngeal wall at high position, and the velopharyngeal closure was improved during speech (Fig. 3C, D). Acoustic analysis during speech production showed that the formants above the frequency of 2000 Hz could be seen more clearly postoperatively than those of preoperative state (Fig. 4). Swallowing function was maintained, with actual improvement of drinking was noted by the patient. After the surgery, the degree of intelligibility rating scale of her speech sounds was preserved for one and a half years even with gradual deterioration of respiratory function until the introduction of artificial ventilation system with tracheostomy.

Patient 2

A 70-year-old man with a 3-year course of slow-progressing ALS was referred by a neurologist to improve his speech disorder. Speech disorder was his initial symptom of ALS with reduced mobility of the hands. His speech disorder was characterized by hypernasality, imprecise and weak articulation, decreased speaking rate, and strangled voice, as spastic dysarthria. The degree of intelligibility rating scale for motor speech disorder (Duffy 2005) of his
speech was estimated as two. The spirometry revealed his respiratory weakness with 76.84% of normal vital capacity. The oropharyngeal examination revealed velopharyngeal closure incompetence (Fig. 5A, B) and weakness of the tongue with fasciculation and atrophic changes. Compression test was positive preoperatively. After surgically narrowing bilateral lateral palatopharyngeal wall, hypernasality of his speech was considerably decreased, and he could articulate more precisely with ease than preoperatively. The degree of intelligibility rating scale for motor speech disorder (Duffy 2005) of his speech at one month after the surgery improved to four (“usually reduced under ideal conditions even when content is unrestricted but adequate with repairs”). Postoperative fibrescopic examination showed that the whole velopharyngeal port was narrowing and the degree of velopharyngeal closure inadequacy has improved during speech (Fig. 5C, D) Swallowing function was not disturbed. Though his speech duration gradually shortened with the progression of respiratory weakness, the degree of intelligibility rating scale of his speech was preserved for one year.

Discussion

Velopharyngeal closure inadequacy could not only contribute hypernasality of speech sound and weak articulation with nasal emission, but also worsened respiratory and laryngeal functions with air wastage to the nasal cavity during speech. Thus, management of velopharyngeal function is beneficial for the treatment of speech disorder with velopharyngeal inadequacy. For the patient suffering from speech disorder induced by ALS, velopharyngeal closure incompetence is caused by the dysfunction of both the velum, and the pharyngeal wall (Darley 1975; Duffy 2005). Thus, pharyngeal flap surgery or the palatal lift prosthesis

Fig. 3. The lateral X-ray imaging for the velopharyngeal port before and after the surgery for Patient 1. Before the surgery, velopharyngeal closure was inadequate during the phonation of /a/ with insufficient elevation of the velum (arrow). After the surgery, the velum was fixed to the posterior pharyngeal wall at the high position (*), and the velopharyngeal closure was improved during the phonation of /a/. A: on rest before the surgery, B: during the phonation of /a/ before the surgery, C: on rest after the surgery, D: during the phonation of /a/ after the surgery.
Fig. 4. Acoustic analysis during the production of vowel /e/, for Patient 1. After the surgery, the formants above the frequency of 2000 Hz could be seen more clearly than those of preoperative state.

Fig. 5. Nasopharyngeal fiberscopic findings before and after the surgery for Patient 2. Before the surgery, velopharyngeal closure was inadequate on the phonation of /a/ with insufficient both elevation of the velum (*) and lateral movement the pharyngeal wall. After the surgery, the whole velopharyngeal port was narrowed, and the velopharyngeal closure was improved on the phonation of /a/. A: on rest before the surgery, B: during the phonation of /a/ before the surgery, C: on rest after the surgery, D: during the phonation of /a/ after the surgery.
which was managed to reinforce the velum dysfunction does not fully improve speech disorder of the patient with ALS (Duffy 2005). Additionally, efficiency of the pharyngeal flap surgery may be greatly dependent upon skill and experience of surgeons (Duffy 2005). Considering reduced range and velocity of the movement of the articulatory organs including both the velum and the pharyngeal wall, reducing whole velopharyngeal port by a surgical intervention is reasonable solution to improve the velopharyngeal inadequacy. This surgical intervention for speech disorder caused by ALS should be indicated for the patients only with slowly progressive course with positive compression test for velopharyngeal closure inadequacy. Though the patients who qualify for the surgery may be limited, it still provides significant improvement of the quality of speech for qualified patients until the severe progression of respiratory weakness develops.

Narrowing of lateral palatopharyngeal wall surgery was first reported for acquired cases with velopharyngeal closure incompetence by Saigusa et al. (1996). The method is not very difficult and non-invasive. And the efficiency of the surgery may be estimated preoperatively by “compression test” with pushing the posterior palatine arch to the posterior pharyngeal wall postero-superiorly. Potential complications of the surgery are bleeding, pharyngalgia, odynophagia, occlusion of nasal air flow. These problems are usually temporally, and they all resolve within about two weeks without any serious long-term complication (Saigusa et al. 1996, 2010). The usefulness of this surgery had been reported for the case with peripheral palsy of the soft palate, bulbar palsy, pseudo-bulbar palsy, and refractory state of myasthenia gravis (Saigusa et al. 1996, 2010). In this report, bilateral narrowing of lateral palatopharyngeal wall surgery can improve speech disorder with velopharyngeal closure inadequacy for ALS patients without major complication. As the disease progressed, the surgical efficacy for the intelligibility of speech sounds was preserved longer, even with the progression of respiratory weakness. And swallowing function was not disturbed by this surgery. For the patient 1, the surgery even had positive effect for swallowing function. Therefore, this surgery is considered to be effective to treat such speech disorder associated with ALS, especially for the patient having slowly progressive course. And the improvement of speech disorder could be accomplished rapidly without any post-operative speech therapy.

Conclusion

We performed bilateral narrowing of lateral palatopharyngeal wall surgery as a speech surgery for two patients suffering from severe speech disorder caused by slowly progressing ALS. Postoperatively, they both showed improved intelligibility and quality of speech sounds with elongated speech phrases without long post-surgical therapy. Although ALS is a progressive disease, improved intelligibility and quality of speech can be maintained with this simple surgical technique with minimum complication and post-operative rehabilitation.

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Conflict of Interest

The authors have no conflict of interest to declare in relation to this article.

References