

## Assessment and Management of Speech in Dysarthria Secondary to Young Onset Amyotrophic Lateral Sclerosis: A Case Report

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### ABSTRACT

*Amyotrophic Lateral Sclerosis (ALS) is one of the most common motor neuron diseases affecting the bulbar, limb, and respiratory muscles. To date, only a few studies have been published regarding the nature of speech-swallowing characteristics, assessment, and management. So, the present study highlighted the nature of speech impairment, swallowing difficulties, detailed evaluation, and rehabilitation options recommended in a young-onset ALS individual. Speech evaluation indicated the presence of flaccid spastic mixed dysarthria. His speech was characterized by hypernasality, imprecise consonants, distorted vowels, harshness, short phrases, mono-pitch, mono-loudness, silent pauses, prolonged phonemes, slow rate of speech, leading to poor speech intelligibility. Swallowing assessment revealed a moderate degree of dysphagia. After 40 sessions of speech therapy, there was a slight improvement in the oro-motor strength and control of few articulators increasing the word level intelligibility, but the improvement was not significant enough. As a result, the QOL of the present case did not improve significantly. So, the authors would like to conclude that it is important to focus on total communication along with speech therapy from the very first session for a significantly better QOL.*

**Keywords:** Amyotrophic lateral sclerosis (ALS), dysarthria, dysphagia, motor neurons, degeneration, quality of life (QOL), amyotrophic lateral sclerosis severity scale (ALSSS).

## INTRODUCTION

Amyotrophic lateral sclerosis (ALS) is a progressive motor neuron disease that affects nerve cells in the brain and spinal cord, causing loss of muscle control (Mayo Clinic 1998). It is a rare, idiopathic, fatal disease of insidious onset. The risk of ALS increases with age and commonly occurs between the ages of 40 to 65 years (Kiernan et al. 2011). About 10% of all cases of ALS begin before age 45 (young-onset ALS), 1% of cases begin before age 25 (juvenile ALS) and the rest begins after 45 years (adult-onset ALS) (Kiernan et al. 2011). The onset of ALS can be at the limbs or the bulbar regions which then eventually spreads to respiratory, myotomes (National Institute of Neurological Disorders and Stroke 2013).

Over the last 3 decades, a very few studies have discussed the speech characteristics in ALS and the results have revealed significant problems in respiration – poor respiratory control, poor vital capacity; phonation – poor phonation duration, highly variable phonatory measures (like fundamental frequency, jitter, shimmer, and signal to noise ratio), strained voice, strangled voice, harsh voice; resonance – hypernasality, poor velopharyngeal closure; articulation – slurred speech, imprecise consonant production, poor diadochokinetic rate; fluency – inappropriate frequent pauses, increased effort, slow rate of speech; prosody – mono-pitch and mono-loudness, ultimately reducing the intelligibility of the speech. Acoustic analyses of the voice have revealed deviant fundamental frequency, amplitude, and frequency perturbation (e.g., shimmer, jitter), voice range, vocal quality, and phonatory instability (Darley, Aronson & Brown 1975; Kent et al. 1990; Kent et al. 1991; Mulligan et al. 1994; Ball, Beukelman & Pattee 2002; Lundy, Roy, Xue, Casiano & Jassir 2004).

Besides speech problems, individuals with ALS suffer from mild to moderate dysphagia within the first two years after the incidence as a result of weakness in oro-motor muscles (National Institute of Neurological Disorders and Stroke 2013). The symptoms include the inability to form and hold bolus in the mouth, difficulty in mastication, bolus residue in the oral cavity,

delayed swallow reflex, nasal penetration, cough before or during the swallow, reduced laryngeal elevation, throat clearing, gurgled voice quality after the swallow, and multiple swallows for a single bolus (Grad, Rouleau, Ravits & Cashman 2017).

As the disease progresses, 80-95% of individuals with ALS are unable to meet their daily communication needs. This may prevent individuals with ALS from participating in many activities, affecting their quality of life (QOL). Various studies have evaluated both health-related and non-health-related factors of quality of life in individuals with ALS and reported poor scores as the disease progressed (Feroldi, Ginocchio, Lunetta, Sansone & Schindler 2014).

People with ALS who have difficulty speaking may benefit from speech therapy in the early stages of ALS (Cohen, Elackattu, Noordzij, Walsh & Langmore 2009; Tomik, & Guilloff 2010), especially when the progression of ALS is slow (Borasio, Voltz & Miller 2001; Kühnlein et al. 2008). Lip and tongue exercises may sometimes help individuals with ALS to enunciate words more clearly but there are no credible data on strengthening exercises of the orofacial muscles; whereas in fast progression ALS, neurologists discourage the use of oro-motor exercises (Miller et al. 1999).

Researchers have documented that resistance exercises, oral motility, and strengthening activities, isometric exercises, and loudness activities focusing on intensive vocal fold adduction exercises may result in decreased voice quality and a rapid rate of decline in intelligibility in individuals with ALS (Ramig, Countryman & Thompson 1995; Watts & Vanryckeghem 2001). So, the authors recommend the usage of Augmentative and Alternative Communication (AAC) such as computer-based speech synthesizers that use eye-tracking technology for ALS with fast progression which can improve the overall quality of life (Körner et al. 2013).

Knowledge of the characteristics, detailed evaluation, and management of speech in individuals with ALS is very important as it helps the professionals and the caregivers to give a better service to the individuals with ALS. This knowledge is often used for rehabilitation, employment, participation in the

community, and to inform the role of family members in the betterment of the ALS individual. In India, very few studies have been carried out regarding the nature and severity of speech-swallowing impairment, the severity of ALS, QOL, and management of communication problems in young adults with ALS. The review of the literature also indicates that ALS in young adults is rare (Nusrat, Mahmood, Marsia & Mahmood 2019). Hence, the present study aimed to investigate the nature and severity of speech impairment and checked the QOL in a young adult diagnosed as having ALS, and also documented the effect of therapeutic management that was undertaken.

#### METHOD

A 36 years old male was referred to the department of speech-language pathology by a neurologist in July 2020 with the complaint of unclear speech and difficulty in swallowing. A detailed history revealed that in June 2019, the client noticed changes in his speech following a high-grade fever for 8 days. As the changes were subtle, it was neglected. Four months later, the client developed difficulty in swallowing both solid and liquid food with a significant weight loss. However, the problems gradually progressed, so the client consulted a neurologist, where the Nerve Conduction Study reports revealed anterior horn cell disease involving bulbar segments. CT scan reports revealed a few small T2/FLAIR hyperintense foci in bilateral frontoparietal white matter, few enlarged upper jugular lymph nodes, and no abnormal mass lesions. Later he consulted another neurologist for a second opinion, where the reports revealed signs of both UMN and LMN damage such as left lower limb weakness, weak cough, facial and tongue weakness with fasciculation, and dysphagia. The patient was diagnosed with young-onset ALS by the neurologist.

Speech and language evaluation was done on July 7<sup>th</sup>, 2020 which revealed language was intact, but exhibited severe impairment in all the speech subsystems indicating the presence of severe dysarthria. Frenchay dysarthria assessment (FDA) was administered to know the type of dysarthria and the sub-sections

included reflex, respiration, lips, jaw, palate, laryngeal, tongue intelligibility, and other influencing factors. The examiner scored on a nine-point rating scale (a – normal, e – completely abnormal) based on the patient’s response to the tasks in the test. Perceptual evaluation along with acoustic analysis was done using Praat software (version 5.3) to assess voice. Henningsson’s four-point rating scale (Henningsson et al. 2008) was used to assess the nasality, speech understandability, and speech acceptability.

To assess the severity of the swallowing problem “Sydney swallow questionnaire” was used. The client was rated using the “Amyotrophic Lateral Sclerosis Severity Scale (ALSSS)” on four subsections (upper extremities, lower extremities, speech, and swallowing) to check the degree of functional impairment of ALS (Hillel 1989). “The Dysarthria-Related QOL Questionnaire (QOL-DYS)” (Piacentini, Zuin, Cattaneo & Schindler 2011) was assessed on four subsections (speech characteristics, situational difficulty, compensatory strategies, and the perceived reaction of others) the QOL of the ALS client and it was also administered to check the efficacy of speech therapy. Poorer the scores, better the QOL in those subsections. Forty hours of speech therapy was given for 2 months and post-therapeutic evaluation of speech and swallowing was conducted to track the improvement.

## RESULTS

### *Pre-therapy*

The FDA results revealed impairment in all the subsystems of speech indicating the presence of dysarthria (Figure 1). The speech characteristics observed in the client’s speech had more than one type of dysarthria indicating it to be a mixed type of dysarthria. The client takes frequent breath intake which indicated poor respiratory control. Due to COVID-19 situations, the respiratory system was assessed subjectively but not instrumentally. His maximum phonation duration was about 5 to 6 seconds. He had a weak cough, and the voice assessment done using GRBAS revealed, “Harsh strangled voice” quality with poor control over voice leading to minimal changes in pitch and

loudness. The client's fundamental frequency was 148 Hz with higher F1, F2, F3 and there was a breakdown of formant structure.

Articulation assessment revealed the presence of imprecise consonant productions and vowel distortions. All the classes of sounds were affected, where lingua-palatal and velar sounds were the most affected, followed by bilabials and labiodentals whereas glottal and inter-dental sounds were least affected. Vowel articulation revealed mild distortion for the vowel /a/ whereas vowel /u/ was the most affected followed by vowel /i/. Slightly drooping lips (towards the right), reduced speed, and range of movements during retraction and pursing were observed. At rest, the jaw appeared to be relaxed in a normal position and in speech tasks, minimal deviation of the jaw was noticed. Tongue examination revealed severe weakness where the client was unable to elevate or make lateral movements of the tongue whereas protrusion of the tongue was found to be limited. Alternate motion rate (AMR) and Sequential motion rate (SMR) revealed reduced speed, range, and accuracy of movements. The rate obtained for AMR and SMR were 1.75 and 0.36, respectively.

Resonance assessment revealed hypernasality associated with nasal air emission. So, Henningsson's four-point rating scale was used to check the severity of hypernasality, speech understandability, and speech acceptability. The client scored "three" on all three sections indicating severe hypernasality, as well as severely affected speech understandability and acceptability which shows that the client's speech is hard to understand most or all the time with the least acceptability. Also, both audible, as well as inaudible nasal air emissions, were consistent. Prosodic aspects of speech were also affected which revealed mono-pitch and monoloudness in connected speech. Speech examination also revealed short phrases, inappropriate silences, prolonged phonemes, and prolonged intervals between phonemes. His rate of speech was found to be 1-2 syllables per second indicating a slow rate of speech.

Swallowing assessment revealed that the client had experiences choking occasionally and reported drooling. Sydney

swallow questionnaire was used to assess the severity of the swallowing problem. The results indicated client was having moderate difficulty in swallowing soft, hard, and dry food. He always needs to swallow more than once for the food to go down. The impression obtained from the evaluation showed the presence of dysphagia. However, could not carry out an instrumental evaluation.

The ALSSS rating revealed a score of 4, 5, 9, and 9 out of 10 in each section like speech, swallowing, lower extremities, and upper extremities, respectively. Hence, the bulbar score is 9 (speech + swallowing) and the spinal score is 18 (lower extremities + upper extremities). The results of QOL-DYS revealed a score of 40, 40, 29, and 23 in each section like “speech characteristics,” “situational difficulty,” “compensatory strategies,” and “perceived reaction of others,” respectively.

#### *Post-therapy*

Forty hours of speech therapy was given for two months, through online sessions. The goals taken were: (1) to improve the oromotor skills by using isotonic exercises, (2) To improve the breath support using respiratory exercises, (3) To improve the phonatory and articulatory skill by traditional articulation therapy, and (4) To improve speech understandability and acceptability at the word level. Concerning dysphagia, dietary modifications were done, where he was recommended to take only pureed diet and liquids. Also, head positioning techniques were taught while eating. Post therapeutic assessment revealed considerable improvement in respiratory control where the MPD increased till 10 to 12 seconds. Voice assessment revealed some changes in pitch and loudness but uneven progression.

In articulation, a slight improvement was seen in the spreading, lateralizing, and puckering of the lips. Also, the asymmetry during the spread of the lips was slightly reduced. Very little improvement was seen in tongue protrusion and elevation, whereas, a good improvement was seen in tongue lateralization where the client was able to move his tongue both ways but was laborious or incomplete. At a time, one sound was well articulated, others were distorted. There was a slight

improvement in AMR than SMR. The jaw movements were also improved, where he was able to open the jaw widely and was able to do laterals movement of the jaw. The overall articulation was improved in bilabials than other sounds and had an MLU of 1- 2 words. Resonance assessment revealed that hypernasality was reduced from severe degree to mild to moderate in vowels, in monosyllabic and few bisyllabic words only. There was no improvement noted in the prosodic aspects of speech.

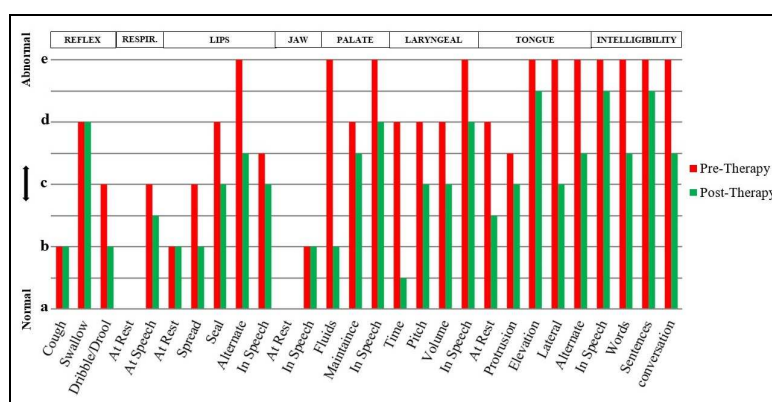


Figure 1. Pre-therapy and post-therapy FDA results.

The ALSSS rating revealed a score of 5, 7, 8, and 8 out of 10 in each section like speech, swallowing, lower extremities, and upper extremities, respectively. Hence, the bulbar score is 12 and the spinal score is 16. The post-therapy results of QOL-DYS revealed a score of 34, 36, 24, and 18 in each section like “speech characteristics,” “situational difficulty,” “compensatory strategies,” and “perceived reaction of others,” respectively.

DISCUSSION

*Pre-therapy*

The present study also shows clusters of abnormal speech characteristics of both flaccid and spastic dysarthria. He had severe hypernasality, nasal air emission and poor respiratory control (a characteristic feature of flaccid dysarthria), strained



and harsh voice quality, slow rate of speech and distorted vowels (spastic component), and imprecise consonants, short phrases, mono-pitch, and monoloudness (characteristic features that are common to both flaccid and spastic dysarthria). These findings agree with the previous findings of Darley et al. (1969) who also reported that ALS with mixed flaccid spastic dysarthria leading to poor speech intelligibility.

The present study results also support the results of several previously done studies on the articulatory abnormalities due to weaknesses in oral speech mechanisms that contribute to reduced intelligibility in ALS (Kent et al. 1990; Duffy, Peach & Strand 2007). Voice quality and prosodic assessment result of the present case is supported by few previous studies who also reported harsh voice quality, prolonged intervals, prolonged phonemes, and inappropriate silences in their study (Darley et al. 1969; Duffy et al. 2007).

Speech or swallowing problems were the initial symptoms noticed in the present case, which can be because of the “bulbar onset” of ALS. Also, the ALSSS score for speech and swallowing was poor compared to the score of upper and lower extremities revealing a poor bulbar score. This proves that the present case is a bulbar onset of ALS and this result supports the previous literature results (Shellikeri et al. 2016). QOL-DYS questionnaire results revealed maximum scoring in the subsections of “speech characteristics” and “situational difficulty,” revealing a drastic impact of ALS on QOL. Whereas “compensatory strategies” and “perceived reaction of others” had fewer scores revealing a better QOL than the other two subsections. This might be because, the subsections “compensatory strategies” and “perceived reaction of others” are not solely dependent on the client, and the family members, friends, and his environment play a major role in these subsections.

#### *Post-therapy*

The therapy goals were taken, to strengthen the subsystems of speech using oro-motor exercises, breathing exercises, traditional articulation techniques, which in turn is believed to improve the

overall speech intelligibility with the usage of the open mouth approach. The post-therapeutic examinations revealed a good improvement in breath support for speech, increase maximum phonation, pitch control, and loudness control. In the articulators, there was a good improvement in lips spread, lip seal, alternating movements of lips, velar movement, tongue lateralization, and word level intelligibility. Very little or no improvement was seen in other aspects of speech subsystems (Figure 1). Concerning swallowing, there was a significant improvement in controlling the drool/dribble, which might be an effect of oro-motor strengthening exercises. Chocking was also significantly reduced because of the diet and head positioning techniques.

The present study supports the results of previous literature which showed improvement in speech after speech therapy in the early stages of ALS (Borasio et al. 2001; Kühnlein et al. 2008; Cohen et al. 2009; Tomik & Guilloff 2010); whereas it contradicts to the results of Ramig et al. (1995), Miller et al. (1999), Watts and Vanryckeghem (2001) who stated that speech therapy decreases voice quality and speech intelligibility in individuals with ALS. The ALSSS revealed not a significant improvement in all the four subsections. So, there was no significant improvement in the post-therapy QOL-DYS results.

#### CONCLUSION

The present case report sheds light on the speech characteristics, assessment, and management of an individual with a rare young-onset ALS. After 40 sessions of speech therapy, there was a slight improvement in the oro-motor strength and control of few articulators increasing the word level intelligibility, as shown by FDA. But there was no significant improvement in the severity of ALS, as shown by ALSSS. As a result, the QOL of the present case did not improve significantly. The authors would like to conclude that it is important to focus on total communication, so we recommend the use of AAC along with speech therapy from the very first session for a significantly better QOL.

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