

<https://helda.helsinki.fi>

---

## ALS patients in otorhinolaryngology : A retrospective study

Rautiola, Jesper

2022-08

---

Rautiola , J , Laaksovirta , H , Geneid , A , Ilmarinen , T , Pietarinen , P & Kinnari , T J 2022 ,  
' ALS patients in otorhinolaryngology : A retrospective study ' , Laryngoscope Investigative  
Otolaryngology , vol. 7 , no. 4 , pp. 1071-1077 . <https://doi.org/10.1002/lio2.859>

---

<http://hdl.handle.net/10138/347515>

<https://doi.org/10.1002/lio2.859>

---

cc\_by

publishedVersion

---


*Downloaded from Helda, University of Helsinki institutional repository.*

*This is an electronic reprint of the original article.*

*This reprint may differ from the original in pagination and typographic detail.*

*Please cite the original version.*

# ALS patients in otorhinolaryngology: A retrospective study

Jesper Rautiola MD<sup>1</sup> | Hannu Laaksovirta MD<sup>2,3</sup> | Ahmed Geneid MD, PhD<sup>1</sup>  |  
Taru Ilmarinen MD, PhD<sup>1</sup> | Petra Pietarinen MD, PhD<sup>1</sup> | Teemu J. Kinnari MD, PhD<sup>1</sup> 

<sup>1</sup>Department of Otorhinolaryngology – Head and Neck Surgery, University of Helsinki and Helsinki University Hospital, Helsinki, Finland

<sup>2</sup>Department of Neurology, University of Helsinki and Helsinki University Hospital, Helsinki, Finland

<sup>3</sup>Research Program Unit, University of Helsinki, Helsinki, Finland

## Correspondence

Teemu J. Kinnari, Helsinki University Hospital, PO Box 263, FI-0029, HUS, Helsinki, Finland.  
Email: [teemu.j.kinnari@helsinki.fi](mailto:teemu.j.kinnari@helsinki.fi)

## Abstract

**Objectives:** Given its rarity and the lack of clear clinical markers, amyotrophic lateral sclerosis (ALS) remains a diagnostic challenge. Because bulbar-onset ALS (buALS) presents with impaired speech or swallowing, patients are often primarily referred to an otolaryngologist (ORL) or phoniatician. The objectives of this retrospective cohort study were to analyze the role of ORLs and phoniaticians in ALS diagnostics and treatment and the potential diagnostic delay related to initial visit to aforementioned specialists.

**Methods:** We reviewed data for all 327 patients treated for ALS through the Hospital District of Helsinki and Uusimaa (HUS) between 2010 and 2014, focusing specifically on 110 (34%) patients presenting with bulbar nerve onset (buALS). Their presenting symptoms, referral to specialized care, and delay in referral to a neurology clinic were assessed. Indications and findings from swallowing studies were reviewed as well as the incidence of percutaneous endoscopic gastrostomy (PEG) and tracheostomy.

**Results:** Among the 110 patients with buALS, 64 (58%) were primarily referred to a neurologist, 28 (25%) to an ORL, and five (5%) to a phoniatician. The most common presenting symptom was dysarthria in 89 patients, (81%), followed by dysphagia in 26 (24%). In most cases, an ORL or phoniatician suspected a neuromuscular disease; however, in eight (24%) cases, the neurological etiology of symptoms was missed. Overall, 49 (45%) patients underwent a swallowing study and 86 (78%) patients underwent PEG placement.

**Conclusions:** Among buALS patients, 30% initially consulted an ORL or phoniatician and 45% underwent a swallowing study. Based on our results, swallowing studies rarely lead to immediate PEG placement. An initial visit to other specialists had no impact on diagnostic delays or survival.

## KEYWORDS

amyotrophic lateral sclerosis, dysphagia, fiberoptic endoscopic examination of swallowing, motor neuron disease, percutaneous endoscopic gastrostomy

This is an open access article under the terms of the [Creative Commons Attribution-NonCommercial-NoDerivs](https://creativecommons.org/licenses/by-nc-nd/4.0/) License, which permits use and distribution in any medium, provided the original work is properly cited, the use is non-commercial and no modifications or adaptations are made.

© 2022 The Authors. *Laryngoscope Investigative Otolaryngology* published by Wiley Periodicals LLC on behalf of The Triological Society.

## 1 | INTRODUCTION

Amyotrophic lateral sclerosis (ALS), a progressive, neurodegenerative disease, affects the motor neurons in the cerebral cortex, brainstem, and spinal cord, resulting in weakness, spasticity, and the progressive loss of muscular control and function.<sup>1</sup> In Europe, the estimated annual incidence of ALS is 2.1–3.8 with a prevalence of 4.1–8.4 per 100,000 person-years.<sup>2</sup>

Up to 70% of ALS patients experience spinal nerve onset affecting the extremities (extALS).<sup>1</sup> By contrast, 25%–30% of ALS patients present with impaired speech and/or swallowing, or bulbar nerve onset ALS (buALS).<sup>1,3–6</sup> The clinical presentation, disease course, and prognosis in these subtypes differ. Furthermore, several other motor neuron diseases closely related to ALS fall within the disease spectrum in the El Escorial revised version, the most commonly used classification system.<sup>7,8</sup>

The etiology of ALS remains primarily unknown<sup>5</sup> with the exception of genetically determined ALS variants, which affect 5%–10% of patients.<sup>1</sup> Remaining ALS patients with no evident family history of motor neuron disease, referred to as “Sporadic ALS,” make up 90%–95% of all ALS patients.<sup>1</sup>

As yet, no curative treatment for ALS is available, although subtle slowing of disease progression has been reported with riluzole, and some short-term alleviation of weakness occurs with edaravone.<sup>9</sup> Assisted ventilation is used by a minority of patients.<sup>10</sup> The exact number of tracheostomies among ALS patients at present is unclear, although in Finland from 2017 to 2020, the incidence of tracheostomy-assisted ventilation might have decreased.<sup>11,12</sup>

Prognosis remains poor, with an expected 3-year survival of 40% and an 8-year survival of 10%.<sup>1</sup> The progression of symptoms ultimately leads to respiratory failure, the leading cause of death among ALS patients.<sup>5</sup> Those receiving tracheostomy-assisted mechanical ventilation eventually develop a complete locked-in state with increasing oculomotor affixion.<sup>5</sup> Survival can vary, partly depending on the genotype, from less than 1 year to decades.<sup>13</sup> BuALS is associated with higher all-cause mortality, greater co-morbidities, and a lower quality of life.<sup>1,2,4</sup>

Given the rarity and the lack of clear clinical markers, ALS and buALS specifically represent diagnostic challenges. The delay from symptom onset to a definitive diagnosis is often lengthy.<sup>1</sup> Because buALS presents with dysarthria and dysphagia,<sup>3</sup> patients may be primarily referred to an otorhinolaryngologist (ORL) or a phoniatician. Thus, awareness of the typical clinical signs and symptoms of ALS are necessary.

During the course of disease, patients with extALS can also develop bulbar symptoms, especially dysphagia,<sup>1</sup> requiring referral to an ORL, phoniatician, or a speech-language pathologist (SLP). The evaluation of dysphagia is crucial to provide recommendations for dietary changes, compensatory maneuvers, and supplemental forms of feeding such as a percutaneous endoscopic gastrostomy (PEG) tube. Early screening and intervention can prevent complications such as aspiration pneumonia and malnutrition, which lead to rapid patient decline. A videofluoroscopic swallow

study (VFSS) and a fiberoptic endoscopic examination of swallowing (FEES) are often employed, although no protocols exist regarding the timing of dysphagia assessment in neurodegenerative diseases.<sup>6,7,14</sup>

This retrospective cohort study aims to characterize ALS patients treated in the Helsinki metropolitan region during a 5-year period (2010–2014). Specifically, we focused on patients with buALS, their presenting symptoms, diagnostic delays, and referrals to specialized care. We analyzed the feasibility of VFSS and FEES in patients with buALS, as well as the number of patients undergoing PEG and tracheostomy.

## 2 | MATERIALS AND METHODS

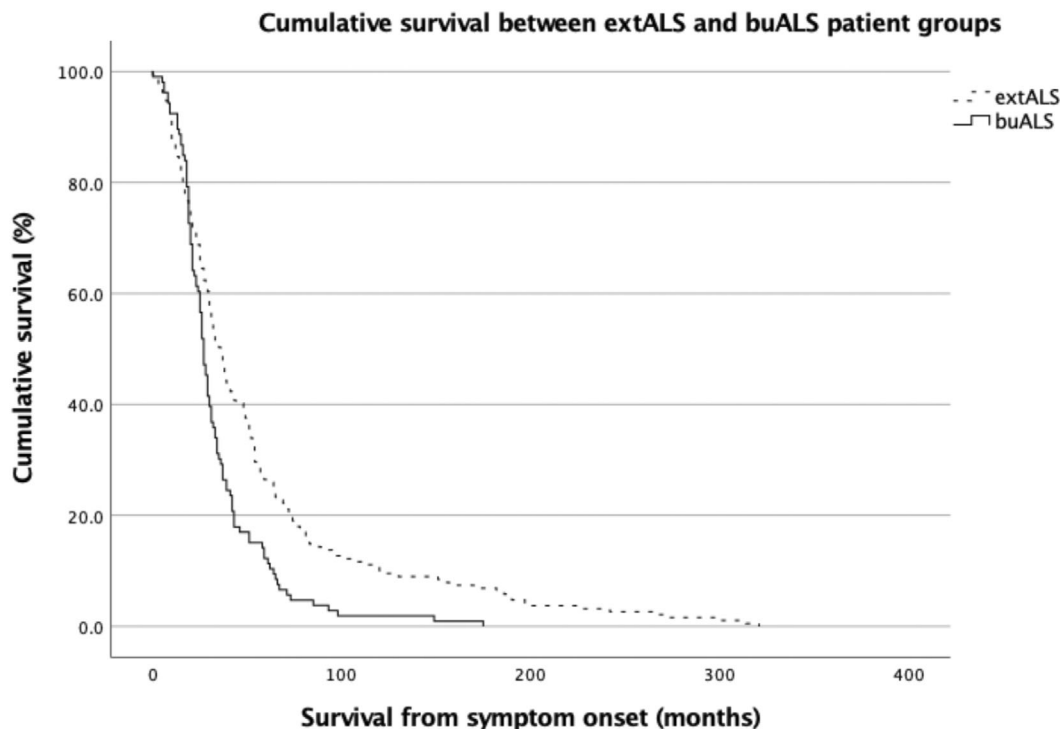
### 2.1 | Patients

We included all patients treated for ALS in the Hospital District of Helsinki and Uusimaa (HUS) between 2010 and 2014. The study period was chosen approximately 5 years prior to the data review, allowing for sufficient follow-up. HUS provides health care services to 1.7 million people. Data retrieval was completed with assistance from HUS's and the Helsinki municipality's statistics departments. ALS diagnosis was confirmed by a senior neurologist (HL), using the El Escorial revised version.<sup>8</sup> Among 388 patients for whom data were available, nine (2.3%) patients were lost to follow-up and 52 (13.4%) had been erroneously diagnosed with ALS. Thus, our cohort included a total of 327 patients. Approval was issued by the institutional review board of HUS.

Hospital patient records were reviewed for age at onset (defined as the age at which the first symptoms of motor dysfunction appeared), gender, time from the presentation of first symptoms until death, and the incidence of tracheostomy and PEG. In buALS, we also reported the date of the first visit to a health care provider, to any medical doctor, to specialized health care, and to a neurologist. The presenting symptoms (dysarthria, dysphagia, breathing symptoms, and any combination of these) and findings during clinical and endoscopic examination (presence of vocal nasality, vocal hoarseness, dysphagia, and/or fasciculations of the tongue and laryngeal findings assessed by either ORL, phoniatician, or SLP) were reported. The first specialty a patient was referred to (neurology, otolaryngology, phoniatics, or internal medicine), referrals to these specialties later during follow-up, and the date of onset of dysphagia and breathing symptoms were also reported. Furthermore, we collected the incidence of instrumental swallowing tests, FEES, or VFSS, as well as their indications, findings, and conclusions.

### 2.2 | Statistical analysis

We performed all statistical analyses using the SPSS statistical software package version 27.0 (IBM Corp.). Correlations were calculated



**FIGURE 1** Comparison of survival among extALS and buALS patients ( $p < .005$ )

**TABLE 1** Clinical characteristics of study patients, overall and within groups

Characteristic	All patients	Bulbar-onset	Limb-onset	<i>p</i>
Age at onset				<.001
Mean	61.5	65.9	59.2	
Range	23–88	23–88	27–85	
Sex, <i>n</i> (%)				<.005
Male	138 (42%)	37 (34%)	101 (47%)	
Female	189 (58%)	73 (66%) $p < .005$	116 (53%)	
PEG, <i>n</i> (%)	192 (59%)	86 (78%)	106 (49%)	<.005
Tracheostomy, <i>n</i> (%)	8 (2%)	3 (3%)	5 (2%)	
Survival in months				<.001
Mean	47	33	55	
95% CI	41.5–53.3	28.9–38.7	46.5–63.7	
Range	0–321	0–175	0–321	

Note: The *p* values are provided in the right-hand column for the difference between groups, and in the respective cells for the intragroup difference. *p* Values are provided only for statistically significant differences.

Abbreviations: CI, confidence interval; PEG, percutaneous endoscopic gastrostomy.

using Pearson's correlation analysis, survival rates were assessed using the Kaplan–Meier method with the log-rank test, and means were compared using the independent *t* test and two-way analysis of variance (ANOVA), where applicable. We report two-tailed *p* values whenever possible, and present our findings with 95% confidence intervals (CIs) when applicable. The homogeneity and normality of data between groups were assessed with the Levene's test and the Shapiro–Wilk's test, respectively.

### 3 | RESULTS

#### 3.1 | Demographic data and clinical characteristics

From 327 study patients, 217 (66.4%) had extALS and 110 (33.6%) buALS. We observed a female predominance among ALS patients (189/327, 58%). The proportion of women with buALS (73/110, 66%;  $p < .005$ ) was significantly higher than for extALS (116/217, 53%;

$p < 0.005$ ). Among all 327 ALS patients, 296 (91%) had died by the time of data retrieval. In the buALS group, 106 of 110 (96%) patients had died, compared to 190 of 217 (88%) patients in the extALS group. A total of 176 new ALS diagnoses were made during the 5-year study period. Thus, 151 patients were diagnosed with ALS before 2010.

The mean age at symptom onset in the buALS group (66 years) was significantly higher compared with the extALS group (59 years;  $p < .001$ ). The mean age at symptom onset in neither group was associated with gender. A higher age at symptom onset was significantly associated with a shorter overall survival time ( $R = -.333$ ;  $p < .001$ ). Mean survival was 47 months among all patients (95% CI: 41.5–53.3), climbing to 55 months (95% CI: 46.5–63.7) in the extALS group, and falling to 34 months (95% CI: 28.9–38.7) in the buALS group. Thus, survival in the extALS group was significantly longer ( $p < .005$ ; Figure 1). The clinical characteristics for all study patients as well as a comparison of buALS and extALS patients appear in Table 1.

### 3.2 | Presenting symptoms and referral to specialized health care among patients with bulbar-onset ALS

Among the 110 patients with buALS, 64 (58%) were primarily referred to a neurologist, 28 (25%) to an ORL, 12 (11%) to internal medicine,

and five (5%) to a phoniatician. Among the 33 patients initially examined by an ORL or phoniatician, the mean delay in referral to an academic neurology center was 56 days (median 26, standard deviation [SD]  $\pm 78$ , range 0–337). In 25 (76%) patients, a clear suspicion of a neuromuscular disease existed and patients were referred to a neurologist primarily within 1 month. Among the remaining eight (24%) patients, the neuromuscular etiology was missed and referral was delayed for up to 1 year. Several reasons explain these difficulties in understanding the neurological nature of the disease symptoms and signs. A tumor was suspected in two patients presenting with constant supraglottic spasticity, obstruction, and a tense voice. In six patients, mild symptoms were linked to misleading findings such as a loose denture, Reinke's edema, vocal fold atrophy, or laryngeal reflux, whereas the incipient dysarthria or dysphagia was ignored. In several of these patients, the examination was completed by a less experienced specialist, including two co-authors, 10 years ago.

Among all buALS patients, the most common presenting symptom was dysarthria, observed in 89 (81%) patients, followed by dysphagia in 26 (24%) patients, and respiratory difficulties in seven (6.6%) patients. A combination of symptoms was reported in a total of 12 (11%) patients.

In buALS, the mean time from symptom onset to the first health care provider visit was 4.4 months (range 0–23, SD = 4.8), 7.1 months (range 0–55, SD = 8.4) to a specialist visit, and 8.5 months (range 0–56, SD = 9.7) to a neurologist referral. In some non-sporadic ALS patients, disease progression was extremely slow resulting in a long diagnostic delay. To capture the typical course of disease for buALS, we omitted from our analysis outliers with a delay exceeding 2 years from symptom onset to the first health care provider visit.

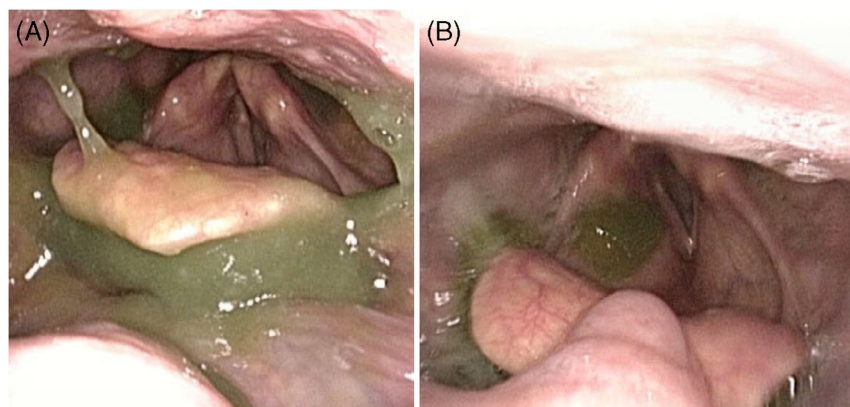
At some point during follow-up, 48 of 110 (44%) patients were referred to an ORL, 17 (16%) to a phoniatician, and 102 (94%) to an SLP. Among all buALS patients, three (3%) were not referred to any such specialist. All patients were ultimately referred to an academic neurology center, excluding one patient who presented with clear clinical symptoms of ALS who refused the referral.

The most common finding reported at an ORL, phoniatician, or SLP consultation was dysarthria in 100 (91%) patients, followed by dysphagia in 94 (85%), fasciculations of the tongue in 77 (70%), vocal

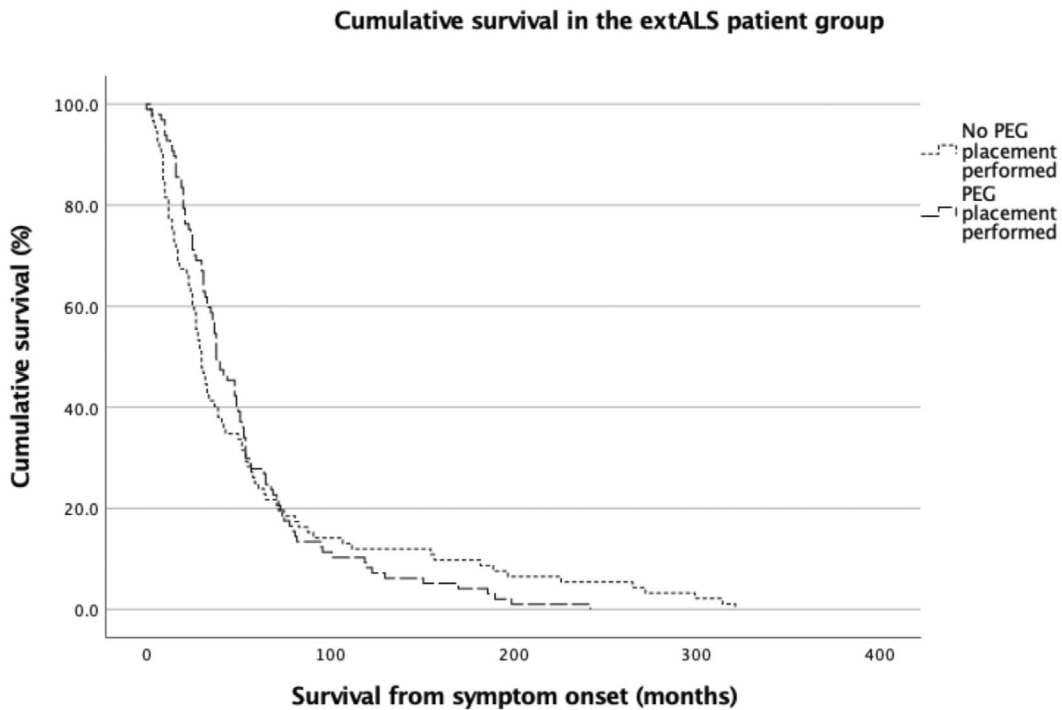
**TABLE 2** Significant findings from the swallowing function tests

Findings	VFSS	FEES
Abnormal/absent laryngeal movement during swallowing, <i>n</i> (%)	34 (87%)	13 (87%)
Difficult initiation of swallowing, <i>n</i> (%)	28 (72%)	7 (47%)
Residua after swallowing, <i>n</i> (%)	27 (69%)	8 (53%)
Penetration to larynx, <i>n</i> (%)	26 (67%)	7 (47%)
Aspiration, <i>n</i> (%)	18 (46%)	5 (33%)
Velopharyngeal insufficiency, <i>n</i> (%)	7 (18%)	2 (13%)

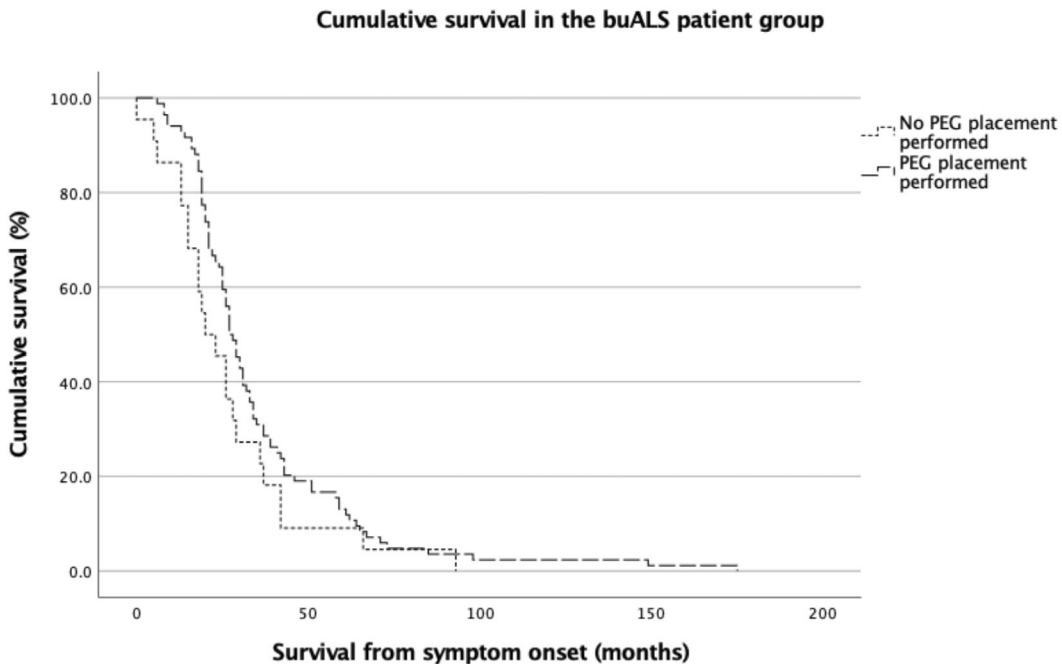
Abbreviations: FEES, fiberoptic endoscopic examination of swallowing; VFSS, videofluoroscopic swallow study.



**FIGURE 2** (A) Residua after swallowing and (B) penetration to the larynx in fiberoptic endoscopic evaluation of swallowing (FEES)



**FIGURE 3** Cumulative survival in the extALS group, depending on whether PEG placement was performed



**FIGURE 4** Cumulative survival in the buALS group, depending on whether PEG placement was performed

hoarseness in 50 (45%), and vocal nasality in 44 (40%) patients. The most common laryngeal findings were a limitation to the vocal fold abduction movement in 11 (20%) patients, vocal fold atrophy in eight (7.2%), and laryngeal spasm/vocal cord dysfunction in two (1.8%) patients.

### 3.3 | Assessment of swallowing in patients with bulbar-onset ALS

Among all buALS patients, 49 (45%) underwent at least one swallowing function test. A total of 54 tests were performed: FEES in

15 patients and VFSS in 39 patients, with five (4.5%) patients undergoing both tests. Among the 49 patients referred for a swallowing test, 32 (82%) with VFSS and nine (60%) with FEES had either a strongly suspected or confirmed ALS at the time of referral. VFSS was indicated by a previous aspiration in 18 (46%) patients and 18 (46%) patients underwent VFSS to evaluate the safety of oral nutrition, whereas 10 (26%) procedures served to evaluate an esophageal obstruction. Among patients undergoing FEES, the corresponding figures are four (27%), six (40%), and five (33%). The findings of the swallowing function tests appear in Table 2. The most common findings were abnormal laryngeal movement, residua after swallowing, and varying levels of penetration to the larynx and aspiration (Figure 2; Table 2).

Among the VFSS procedures, eight (21%) resulted in a recommendation for PEG placement. None of the FEES procedures resulted in an immediate PEG recommendation. Outright swallowing interdictions were not provided based on either test. However, patients undergoing a swallowing function test more often received PEG at some point ( $R = .207$ ;  $p < .05$ ). The incidence of PEG placement among all buALS patients reached 78% ( $n = 86$ ): 94% ( $n = 46$ ) among those who underwent a swallowing function test and 66% ( $n = 40$ ) among those who did not.

Among all study patients, 192 of 327 (59%) underwent PEG placement, which was more frequent in the buALS group (86/110 patients, 78%;  $r = .281$ ;  $p < .001$ ) compared to the extALS group (106/217, 49%). PEG was associated with a shorter survival time in the extALS group (53 months with PEG [95% CI 44.1–62.1;  $p < .001$ ] vs. 57 months without PEG [95% CI 42.3–72.1;  $p < .05$ ]) and a longer survival time in the buALS group (35.6 months [95% CI 29.9–41.4;  $p < .001$ ] and 26.8 months [95% CI 18.1–35.5;  $P < 0.05$ ]; Figures 3 and 4). The mean age at symptom onset did not associate with PEG placement.

The mean time from symptom onset to death in patients with buALS was not significantly longer among those primarily referred to a neurologist compared to those referred to an ORL or phoniatician. Mean survival in the buALS patient group was 32 months (95% CI 27.3–36.9) compared to 33 months (95% CI 23.0–44.2) and 37 months (95% CI 25.9–47.4), respectively, among those initially referred to an ORL and a phoniatician.

## 4 | DISCUSSION

We characterized 176 newly diagnosed ALS patients in the Helsinki metropolitan region of 1.7 million inhabitants during a 5-year period (2010–2014). The estimated annual ALS incidence of 2.1 per 100,000 mirrored reports from other European countries. Furthermore, the proportion of patients with buALS (34%), the higher mean age at onset in buALS (66 years) compared with extALS (59 years), and the female predominance in buALS (66%) agree with previous studies.<sup>15</sup> The strength of our study setting lies in selecting a retrospective sample, making it possible at the time of data retrieval to trace the complete course of disease for most patients.

In our study, 48 of 110 patients (44%) with buALS were not initially referred to a neurologist. Among 110 patients, 33 (30%) were

initially referred to an ORL/phoniatician, typically because of dysarthria or dysphagia. Practically all patients were referred to a neurologist at some point, however, in eight (24%) patients, an ORL/phoniatician had clear difficulties in understanding the nature of the symptoms. The primary reasons for the diagnostic delay appeared to be the early stage and mild symptoms of disease, physician inexperience, and, in two patients, a constant spasticity or hyperfunction in the supraglottic structures leading to a narrowed airway and tumor suspicion. Chen and Garrett<sup>16</sup> reported similar findings, whereby 42% of buALS patients presenting to an ORL initially received an erroneous diagnosis.

ORLs require awareness of the potential symptoms and signs of bulbar-onset ALS, often presenting with dysarthria or dysphagia. Clinical findings may include weakness in the facial muscles, palate, or tongue, as well as fasciculations of the tongue, and a hypernasal or breathy voice with incomplete vocal fold closure.<sup>16</sup> Dysarthria should not be regarded as a sign of laryngeal pathology. In general, our study confirms Turner et al.'s<sup>15</sup> findings that, although half of all buALS patients being initially referred to other than neurologists, this did not significantly impact diagnostic delays or survival.

Our study also aimed to evaluate the feasibility of VFSS and FEES in buALS patients. A recent systematic review concluded that several assessment strategies are employed in neuromuscular diseases, depending on the center, the country, and local protocols. VFSS is often considered the principal tool to assess swallowing, although interrater reliability appears poor. Substantial variation exists in methodological settings in terms of the thickness, viscosity, and volume of contrast fluids. Generic scales such as the dysphagia outcome severity scale (DOSS) or penetration aspiration score (PAS) are used variably.<sup>17</sup> The feasibility of VFSS in bulbar-onset ALS, as well as the proportion of patients presenting with aspiration, presumably depends on the timing of examination and the stage of disease. In our study, 18 of 39 patients (46%) presented with aspiration in VFSS and 5 of 15 (33%) in FEES.

An obvious advantage of FEES, performed in 14% of buALS patients, is that patients may receive recommendations for modifying food consistency and therapeutic strategies, and compensatory maneuvers can be evaluated during examination with visual feedback. For end-stage ALS, FEES can be modified according to the patient's mobility and the ability to maintain one's posture. In Finland, FEES is increasingly performed independently by SLPs; however, in our study, all FEES procedures were performed under the direct supervision of an ORL. Although most swallowing studies were performed to evaluate the safety of peroral nutrition, a recommendation for complete tube feeding after VFSS or FEES in our study patients remained extremely uncommon. Thus, the usefulness of VFSS, performed on 35% of buALS patients, remains questionable.

In our study, 86 of 110 patients (78%) with buALS underwent PEG placement, similar to reports from previous studies.<sup>15</sup> For instance, the cumulative incidence of PEG during the 2–4-year follow-up period from the Death in USA ALS cohort was 48%.<sup>18</sup> PEG associated with a longer survival period among buALS patients in our study, possibly associated with improvement in nutritional status. This finding was reversed in the extALS patient group, possibly related to the fact that among this ALS subtype, the swallowing symptoms usually

appear late. Those who underwent swallowing studies were also slightly more likely to undergo PEG placement, which may be related to the shared indications in both. However, the driving force in proceeding with PEG placement was rarely a swallowing study. More likely, the decision was forced by increasing aspiration.

Moreover, median survival among our patients—55 months in extALS versus 34 in buALS—agrees with previous studies.<sup>10,15</sup> Survival in extALS is typically longer because bulbar affection represents a major cause of morbidity and mortality in ALS patients and occurs fairly late in the limb-onset disease subgroup.

In our study, eight (2.4%) patients were treated with a tracheostomy. This also agrees with the low overall tracheostomy incidence in Finland.<sup>11</sup> Because invasive mechanical ventilation brings about the risk of entering a complete locked-in state, this option requires a timely consultation with an experienced specialist and a profound consideration of end-of-life treatment options.<sup>1,7,19</sup>

## 5 | CONCLUSIONS

Based on our findings, the diagnosis and treatment of ALS in Finland agree with results from earlier studies. Primary diagnostics are performed quickly and efficiently and do not depend on the specialty of the initial physician a patient consults. Gastric tubes are commonly placed and appear to improve survival among buALS patients, although further study is needed to evaluate the effect of parenteral nutrition on extALS patient survival. Swallowing studies rarely provided clear recommendations when considering feeding, thereby casting doubt on their usefulness. Invasive ventilation remains a rarity among all ALS patients in Finland.

The dominant symptoms among those ALS patients who initially seeking ORL assistance are typical for the buALS group, primarily affecting swallowing and speech.

### CONFLICT OF INTEREST

The authors declare no conflict of interest.

### ORCID

Ahmed Geneid  <https://orcid.org/0000-0003-1617-1181>

Teemu J. Kinnari  <https://orcid.org/0000-0001-9949-7640>

### REFERENCES

- Kiernan MC, Vucic S, Cheah BC, et al. Amyotrophic lateral sclerosis. *The Lancet*. 2011;377(9769):942-955. doi:10.1016/S0140-6736(10)61156-7
- Longinetti E, Fang F. Epidemiology of amyotrophic lateral sclerosis: an update of recent literature. *Curr Opin Neurol*. 2019;32(5):771-776. doi:10.1097/WCO.0000000000000730
- Zarei S, Carr K, Reiley L, et al. A comprehensive review of amyotrophic lateral sclerosis. *Surg Neurol Int*. 2015;6:171. doi:10.4103/2152-7806.169561
- Tsunoda K, Takazawa M, Chong T, Morita Y. Slow, slurred speech as an initial complaint in amyotrophic lateral sclerosis. *Auris Nasus Larynx*. 2019;46(2):193-195. doi:10.1016/j.anl.2018.07.007
- Hardiman O, Al-Chalabi A, Chio A, et al. Amyotrophic lateral sclerosis. *Nat Rev Dis Primer*. 2017;3(1):1-19. doi:10.1038/nrdp.2017.71
- Ruoppolo G, Schettino I, Frasca V, et al. Dysphagia in amyotrophic lateral sclerosis: prevalence and clinical findings. *Acta Neurol Scand*. 2013;128(6):397-401. doi:10.1111/ane.12136
- Andersen PM, Borasio GD, Dengler R, et al. Good practice in the management of amyotrophic lateral sclerosis: clinical guidelines. An evidence-based review with good practice points. EALSC Working Group. *Amyotroph Lateral Scler*. 2007;8(4):195-213. doi:10.1080/17482960701262376
- Brooks BR, Miller RG, Swash M, Munsat TL, World Federation of Neurology Research Group on Motor Neuron Diseases. El Escorial revisited: revised criteria for the diagnosis of amyotrophic lateral sclerosis. *Amyotroph Lateral Scler Mot Neuron Disord*. 2000;1(5):293-299. doi:10.1080/146608200300079536
- Jaiswal MK. Riluzole and edaravone: a tale of two amyotrophic lateral sclerosis drugs. *Med Res Rev*. 2019;39(2):733-748. doi:10.1002/med.21528
- Spittel S, Maier A, Kettemann D, et al. Non-invasive and tracheostomy invasive ventilation in amyotrophic lateral sclerosis: utilization and survival rates in a cohort study over 12 years in Germany. *Eur J Neurol*. 2021;28(4):1160-1171. doi:10.1111/ene.14647
- Kotanan P, Kreivi H-R, Vainionpää A, Laaksovirta H, Brander P, Siirala W. Home invasive mechanical ventilation in Finland in 2015–2019. *ERJ Open Res*. 2020;6(4):00223-2020. doi:10.1183/23120541.00223-2020
- Siirala W, Vainionpää A, Kainu A, Korpela J, Olkkola K, Aantaa R. Prevalence of life-supporting prolonged invasive ventilation support in Finland. *Duodecim*. 2017;133(7):675-682.
- Swinnen B, Robberecht W. The phenotypic variability of amyotrophic lateral sclerosis. *Nat Rev Neurol*. 2014;10(11):661-670. doi:10.1038/nrneurol.2014.184
- Desport JC, Preux PM, Truong CT, Courat L, Vallat JM, Couratier P. Nutritional assessment and survival in ALS patients. *Amyotroph Lateral Scler Other Motor Neuron Disord*. 2000;1(2):91-96. doi:10.1080/14660820050515386
- Turner MR, Scaber J, Goodfellow JA, Lord ME, Marsden R, Talbot K. The diagnostic pathway and prognosis in bulbar-onset amyotrophic lateral sclerosis. *J Neurol Sci*. 2010;294(1):81-85. doi:10.1016/j.jns.2010.03.028
- Chen A, Garrett CG. Otolaryngologic presentations of amyotrophic lateral sclerosis. *Otolaryngol Neck Surg*. 2005;132(3):500-504. doi:10.1016/j.otohns.2004.09.092
- Audag N, Goubau C, Toussaint M, Reyckler G. Screening and evaluation tools of dysphagia in adults with neuromuscular diseases: a systematic review. *Ther Adv Chronic Dis*. 2019;10:2040622318821622. doi:10.1177/2040622318821622
- Heffernan C, Jenkinson C, Holmes T, et al. Nutritional management in MND/ALS patients: an evidence based review. *Amyotroph Lateral Scler Other Motor Neuron Disord*. 2004;5(2):72-83. doi:10.1080/14660820410020349
- Chiò A. ISIS survey: an international study on the diagnostic process and its implications in amyotrophic lateral sclerosis. *J Neurol*. 1999;246(S3):III1-III5. doi:10.1007/BF03161081

**How to cite this article:** Rautiola J, Laaksovirta H, Geneid A, Ilmarinen T, Pietarinen P, Kinnari TJ. ALS patients in otorhinolaryngology: A retrospective study. *Laryngoscope Investigative Otolaryngology*. 2022;7(4):1071-1077. doi:10.1002/lio.2.859