

Study/Type	Quality Rating	Sample Description	Method	Outcomes	Key Findings
<p>31. Rooney et al 2015</p> <p>Population-based cohort study</p>	<p>Class II</p>	<p>719 incident ALS cases over 25 years of age, diagnosed between January 1, 2005, and December 31, 2010. Cases were identified in population-based ALS registries from Ireland and Northern Ireland.</p>	<p>Proportions of gastrostomy and NIV use were compared between groups.</p> <p>Kaplan-Meier (KM) estimator with log-rank test and multivariable Cox Proportional Hazard models were performed to estimate survival.</p> <p>Predictor: multidisciplinary care in Ireland;</p> <p>Comparator: non-specialty care in Ireland or Northern Ireland.</p>	<p>Unadjusted and adjusted survival;</p> <p>NIV use;</p> <p>Gastrostomy use;</p>	<p>In the adjusted Cox model, ALS care by a multidisciplinary team was associated with prolonged survival from diagnosis (hazard ratio 0.59, 95% confidence interval 0.49 to 0.71, $p < 0.001$).</p> <p>Larger proportions of NIV use and combined NIV and gastrostomy use were demonstrated in patients attending multidisciplinary clinic vs non-specialty clinics (38% vs 15.4%, $p < 0.001$, and 15.9% vs 6.4%, $p < 0.001$, respectively).</p>
<p>32. Traynor et al 2003</p> <p>Population-based cohort study</p>	<p>Class II</p>	<p>345 Irish patients, diagnosed with ALS between January 1, 1996, and December 31, 2000.</p>	<p>Kaplan-Meier (KM) estimator with log-rank test and multivariable Cox Proportional Hazard models were performed to estimate survival.</p> <p>Riluzole and NIV use was compared between groups.</p> <p>Predictor: attendance to multidisciplinary ALS clinic;</p> <p>Comparator: attendance to general neurology clinics.</p>	<p>Unadjusted and adjusted tracheostomy-free survival;</p> <p>Riluzole use;</p> <p>NIV use.</p>	<p>In the adjusted Cox model, patients followed by general neurology clinics had shorter tracheostomy-free survival from diagnosis compared to patients followed at a multidisciplinary clinic (hazard ratio 1.47, 95% confidence interval 1.06 to 2.06, $p = 0.002$).</p> <p>The proportion of patients using Riluzole was greater in the multidisciplinary clinic vs. general neurology clinics (99% vs 61%, $p < 0.001$).</p> <p>The proportion of NIV use was similar</p>

					between the groups (6.1% vs. 2.7%, p=0.14).
33. Van den Berg et al 2005 Cross-sectional study	Class II	208 patients with ALS and their caregivers were interviewed between June 2001 and March 2004.	In-person interview using a structured questionnaire and validated quality of life and functional scales. Comparison of interventions and quality of life between groups. Predictor: multidisciplinary ALS care; Comparator: general care.	Use of aids and adaptive equipment. Quality of life: Medical Outcome Study 36-item Short Form Health Survey (SF-36) and visual analogue scales (VAS) on life satisfaction and well-being; Number of visits to neurologists, physiatrists, physical therapists, occupational therapists, speech pathologists, dieticians, and social worker;	The proportion of patients who received adequate adaptive equipment was higher in the multidisciplinary ALS care compared to general care (93% vs 81%, p=0.008). When adjusted for confounders, the summary scale scores of the SF-36 mental health were improved in patients followed by multidisciplinary care compared to general care (p=0.01). There were no significant differences in the SF-36 physical functioning scales, life satisfaction or well-being.
34. Chiò et al 2006 Population-based cohort study	Class II	221 patients diagnosed with ALS between 1995 and 1996.	Prospective data collection from The Piemonte and Valle d'Aosta Register for amyotrophic lateral sclerosis (PARALS); Kaplan-Meier (KM) estimator with log-rank test and multivariable Cox Proportional Hazard models were performed to estimate tracheostomy-free survival.	Unadjusted and adjusted tracheostomy-free survival; Hospital admissions and length of stay. Gastrostomy use; NIV use; Riluzole use.	In the adjusted Cox model, patients followed by general neurology clinics had shorter tracheostomy-free survival compared to patients followed at tertiary ALS clinics (hazard ratio 1.61, p=0.006); There were fewer hospital admissions per patient attending tertiary ALS clinics than patients not attending such clinics (mean 1.2 and SD 0.9 vs. mean

			<p>Comparison of interventions between groups.</p> <p>Multivariable analyses on hospital admissions were carried out with a stepwise linear regression model.</p> <p>Predictor: care in tertiary ALS centres;</p> <p>Comparator: care in general neurology clinics.</p>		<p>3.3 and SD 1.8, $p < 0.001$);</p> <p>The mean duration of hospital stay was 10.3 (SD 27.6) days shorter for patients followed by tertiary ALS centres (mean 5.8, SD 9.5) than in general neurology clinics (mean 12.4, SD 31.6; $p = 0.001$).</p> <p>Higher percentage of patients undergoing gastrostomy and NIV in tertiary ALS clinics compared to general neurology clinics (32% vs 9%, $p = 0.01$, and 15% vs 7%, $p = 0.04$, respectively).</p>
35. Stephens et al 2015 Cross-sectional, online survey	Class III	295 patients with ALS in the United States.	<p>Online questionnaire and validated scales (ALS Specific Quality of Life Instrument-Revised (ALSSQoL-R), the ALS Functional Rating Scale-Revised (ALSFRRS-R));</p> <p>Comparison of interventions between groups.</p> <p>Predictor: patients attending multidisciplinary clinic;</p> <p>Comparator: patients attending non-multidisciplinary clinics.</p>	<p>Riluzole use;</p> <p>Gastrostomy use;</p> <p>NIV use;</p> <p>Quality of life.</p>	<p>There was no difference in Riluzole or gastrostomy use between the groups;</p> <p>NIV use was more common in patients in multidisciplinary clinics ($p = 0.03$)</p> <p>There were no differences between the groups in global quality of life, physical function, or social problem-solving skills.</p>
36. Aridegbe et al 2013	Class II	417 patients with ALS, diagnosed and followed up between 1998 and 2002 and 2006 and 2010.	<p>Kaplan-Meier (KM) estimator with log-rank test and multivariable Cox Proportional</p>	<p>Unadjusted and adjusted survival from symptom onset;</p>	<p>A higher proportion of multidisciplinary clinic patients used Riluzole and NIV than general</p>

Clinic-based retrospective cohort study			<p>Hazard models were performed to estimate survival.</p> <p>Comparison of interventions between groups.</p> <p>Predictor: multidisciplinary clinic patient group;</p> <p>Comparator: general neurology clinic patient group.</p>	<p>Riluzole use;</p> <p>NIV use;</p> <p>Gastrostomy use.</p>	<p>neurology clinic patients (89% vs. 55%, $p<0.001$, and 29% vs. 5%, $p<0.001$).</p> <p>Survival from symptom onset for patients who were followed in a multidisciplinary clinic was longer compared to general neurology clinic attenders (hazard ratio 0.58; 95% confidence interval 0.46 to 0.73, $p<0.001$).</p>
37. Zoccolella et al 2007 Population-based cohort study	Class II	126 incident ALS cases from a registry established in Puglia, Italy, in 1997.	<p>Kaplan-Meier (KM) estimator with log-rank test and multivariable Cox Proportional Hazard models were performed to estimate survival from onset and diagnosis.</p> <p>Comparison of interventions between groups.</p> <p>Predictor: multidisciplinary ALS care;</p> <p>Comparator: general neurology clinic care.</p>	<p>Unadjusted and adjusted survival from symptom onset and diagnosis;</p> <p>Riluzole use;</p> <p>NIV use;</p> <p>Gastrostomy use.</p>	<p>There was no difference in NIV or gastrostomy use between the groups;</p> <p>Riluzole use was more frequent among patients followed by multidisciplinary care (66% vs 43%, $p=0.02$).</p> <p>In the adjusted Cox model, there was no difference in the probability of death between patients followed by ALS multidisciplinary and general clinics (hazard ratio 0.9; 95% confidence interval 0.44 to 1.89; $p=0.9$).</p>
38. Martin et al 2017 Register-based cohort study	Class III	547 patients with ALS from the SEALS Register, diagnosed between 1995 to 1998 and 2008 to 2011.	<p>Kaplan-Meier (KM) estimator with log-rank test and multivariable Cox Proportional Hazard models were performed to estimate survival.</p>	<p>Unadjusted and adjusted survival;</p>	<p>In the adjusted Cox model, the 2008-2011 multidisciplinary care cohort was a significant predictor of prolonged survival (hazard ratio 0.79, 95% confidence interval 0.64 to 0.97, $p=0.023$).</p>

			<p>Comparison of interventions between groups.</p> <p>Predictor: multidisciplinary ALS team care (2008 to 2011 cohort);</p> <p>Comparator: no multidisciplinary ALS team care (1995 to 1998 cohort);</p>		
39. Hobson et al 2016 Systematic review	Class II	16 studies were included in qualitative synthesis. No studies were included in quantitative synthesis (meta-analysis)	Systematic review of the current technologies used to facilitate access to specialist care for patients with ALS.	<p>Clinical and health resource impacts of telehealth;</p> <p>Feasibility and acceptability of the intervention and impact on burden and quality of life.</p>	<p>Telemedicine as part of ALS care was feasible and largely accepted by patients.</p> <p>Telehealth monitoring of respiratory failure and non-invasive ventilation in ALS is also feasible.</p>
40. Geronimo et al 2017 Mixed-method survey study	Class IV	11 patients with ALS, 12 caregivers, and 15 health care professionals.	Quantitative and qualitative (mixed-method) approach for synthesis of feasibility and acceptability of telehealth as part of multidisciplinary ALS care.	<p>Satisfaction with the telehealth visit;</p> <p>Quality of care</p> <p>Confidence with the virtual interface.</p>	<p>All patients and caregivers, and most health care providers agreed that the system allowed for good communication, description of concerns, and provision of care recommendations.</p> <p>Most participants felt that telehealth removed travel burden, resulting in lower stress and more comfortable interactions.</p>
41. Selkirk et al 2017 Clinic-based retrospective cohort study	Class II	68 patients with ALS followed between March 1, 2008, and January 31, 2015.	Logistic regression to provide adjusted odds ratios of Riluzole use, NIV use, home health care, communication	Adjusted odds ratios of Riluzole use, NIV use, home health care, communication device use, gastrostomy	There was no difference in the likelihood of receiving Riluzole, NIV, home health care, communication device use, and gastrostomy between patients

			<p>device use, gastrostomy insertion.</p> <p>Kaplan-Meier (KM) estimator with log-rank test and multivariable Cox Proportional Hazard models were performed to estimate survival from onset and diagnosis.</p> <p>Comparison of interventions between groups.</p> <p>Predictor: telemedicine care;</p> <p>Comparator: tertiary in-person ALS clinic care.</p>	<p>insertion between groups;</p> <p>Unadjusted and adjusted Survival.</p>	<p>followed by telemedicine or in-person.</p> <p>In the adjusted Cox model, there was no difference in the probability of death between patients followed by telemedicine and in-person clinical care (hazard ratio 0.48, 95% confidence interval 0.21 to 1.07, p=0.07).</p>
<p>42. Van De Rijn et al 2018</p> <p>Case series</p>	Class IV	<p>97 ALS patients assessed by video televisits between September 2014 and January 2016</p>	<p>Descriptive experience with telemedicine.</p>	<p>Feasibility.</p>	<p>Video televisits are feasible, particularly to address medication management, goals of care, research, and equipment use.</p>