

ALS-ID: Developing a roadmap for early diagnosis and rapid referral of probable ALS cases to specialist care

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Background

Amyotrophic Lateral Sclerosis (ALS) is a fatal progressive neurodegenerative disease. Median survival is 2-3 years from symptom onset¹. Diagnostic delays of 9.1-27 months from symptom onset care are reported².

Delayed referral to ALS multidisciplinary clinics (MDC) can render patients ineligible for some medications (e.g. riluzole and Radicava[®]) and reduce access to specialized ALS care².

Objective: Collaborating with the Physician Learning Program (PLP), we undertook an Edmonton-focused quality improvement program to reduce diagnostic delay for ALS.

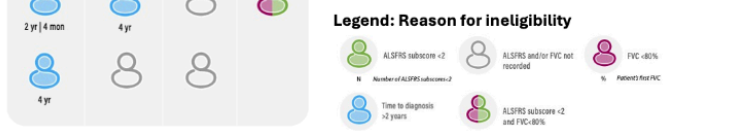
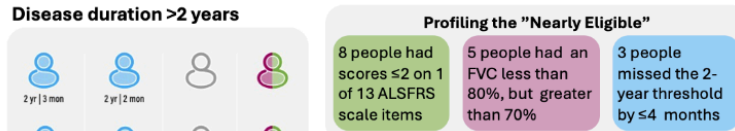
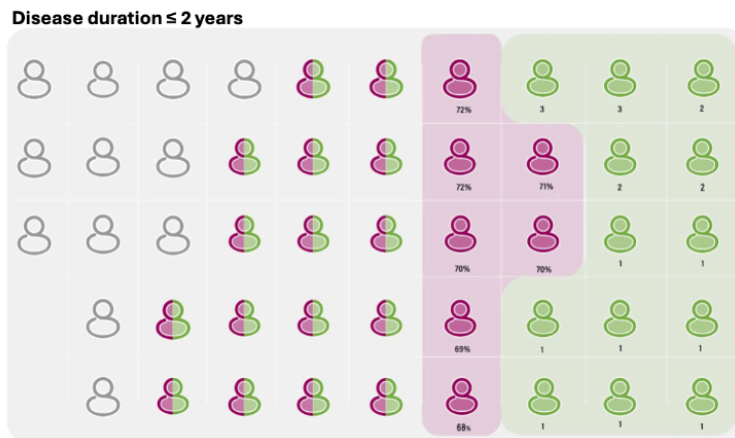
Establishing baseline using a retrospective chart review

Methods: We conducted a retrospective chart review of patients newly diagnosed with ALS at the Kaye Edmonton Clinic (KEC) in Edmonton, Alberta between September 1, 2019 and June 30, 2021. Data collected included patient demographics and clinical features upon which ALS drug eligibility criteria are based.

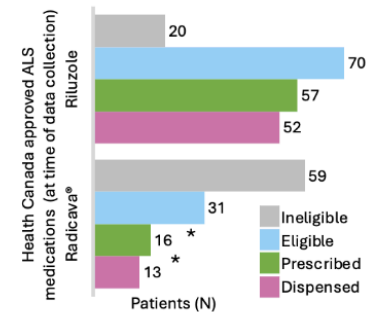
Results: The chart review identified a diagnostic delay of 13.44 months (mean)/10.2 months (median) in 90 patients. 76% of patients were ineligible for a key ALS drug (Radicava[®]). Therefore, while the median diagnostic delay is within the reported range of other jurisdictions, patients are still arriving to the ALS MDC ineligible for some ALS treatments.

Demographics	
N	90
Female N (%)	34 (37.8)
Median Age (IQR)	65.6 (60.3, 70.2)
Site of Onset	
• Limb	61 (67.7%)
• Bulbar	27 (30.0%)
• Generalized	2 (2.2%)
Time between symptom onset & first visit to ALS MDC (Median (IQR; Min, Max)	10.2 months (2.64, 16.44; 2.64, 49.2)
First Forced Vital Capacity (FVC) (Median (IQR; Min, Max) (N=86)	83.5 (69.0, 97.8; 27.0, 125.0)
Overall ALS Functional Rating Scale (Revised) (ALSFRS) score (Median (IQR))	41.0 (36.0, 43.5)

65% of our sample were not eligible for Radicava[®]



Not all eligible patients get medication

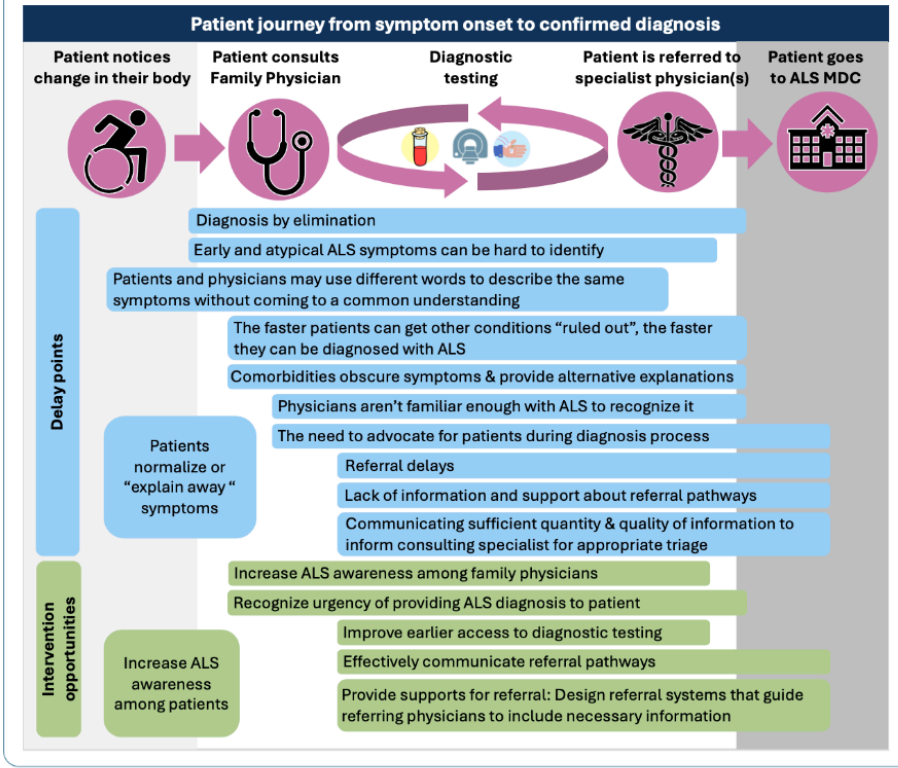


*6 additional people declined IV Radicava[®] due to their interest in participating in a clinical trial for oral Edaravone (Mitsubishi Tanabe Pharma Development America, Inc.)

Journey mapping using human-centred design

Methods: We used human-centred design to develop an ALS diagnosis journey map in Edmonton. 10 ALS patients (4 female; aged 32-78) and 20 practitioners (8 neurologists, 4 neurosurgeons, 1 orthopedic surgeon, 3 ENT specialists, 3 family physicians, and 1 physiatrist) were interviewed between February 2021 and September 2022. Interviews were audio recorded and transcribed. Their experiences and perspectives were qualitatively analyzed identifying common themes.

Results: The journey map identified 2 actionable barriers to timely ALS diagnosis: inefficient EMG referral and physician knowledge about when to expedite referrals for patients suspected of having ALS.



Evidence-based interventions

EMG Form Review: Working with EMG neurologists based at the KEC, a review of the EMG referral form and pathway is underway. Informed by this work and other developments in the Edmonton Zone, KEC EMG triage was centralized in 2024. Currently, a draft EMG referral form is being reviewed by doctors in referring specialties. We are also working with the Alberta Referral Directory to improve the EMG referral information available on their platform.

Physician education: A 4-part interactive webinar series on neuromuscular "red flags" in collaboration with colleagues in ENT, physiatry, and geriatrics was delivered weekly in May 2024. Registration was 160-196 per session. All registrants received presentation recordings and PEARLS summary and resource list. Day-of attendance was 53-67. Feedback via evaluation forms (n=64) was positive overall, with all respondents reporting the sessions were relevant to their practice, met their learning needs, and helped them reflect on their practice.

Conclusions

The multi-pronged, human-centred approach taken by our team, and supported by the PLP, generated evidence for actionable interventions to improve the diagnosis of ALS in the Edmonton Zone. Future work includes evaluating the interventions with a chart review scheduled in 2025 to compare against our baseline, exploring further physician education approaches, and implementing the new EMG referral form in the KEC EMG lab and sharing it with other EMG labs in Northern AB.

Acknowledgements/Reference

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References: [1] Traxinger K, Kelly C, Johnson BA, et al. *Neurol Clin Pract.* 2013; Aug3:313-20; [2] Richards D, Morren JA, Pioro EP. *J Neurol Sci.* 2020; 417: 117054.