



Twenty Months to a Diagnosis of Amyotrophic Lateral Sclerosis: A Narrative Review of Diagnostic Delay and Communicating the Diagnosis

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Abstract

Amyotrophic lateral sclerosis takes about a year to diagnose in a disease that kills within three to five. The delay therefore consumes a substantial fraction of the illness, and unlike most diagnostic delays it cannot be recovered by treating sooner, because there is little treatment to bring forward.

Objectives: *To review the published evidence on diagnostic delay in amyotrophic lateral sclerosis: how long it is, where in the pathway it accrues, why it has not improved, what is actually lost by it, and what is known about communicating the diagnosis once it is made.*

Material and Methods: *Narrative review of multicentre cohorts, retrospective analyses of diagnostic pathways, literature reviews and studies of diagnosis disclosure indexed in PubMed and the major neurology and neuromuscular journals. No new patient data were generated; every estimate quoted is one published by the original investigators.*

Results: *Median time from symptom onset to diagnosis was 11 months in a tertiary cohort of 260 patients and 11 months in a four-country cohort of 1405 patients, with literature reviews reporting a range of 10 to 16 months. In a French centre cohort of 90 patients, mean delay was 17 months and median 12, comprising 2.7 months from first symptom to first consultation with a general practitioner and a further 6.5 months before the first neurologist visit. Only 38.8 per cent of patients had seen a neurologist before diagnosis in one series.*

Conclusion: *The delay is stable across countries and decades and is concentrated in the interval before neurological assessment. Because disease-modifying options are limited, the argument for shortening it rests on access to multidisciplinary care, respiratory support, trial eligibility and time to plan, rather than on treatment that works better when started earlier.*

Abbreviations: *ALS – Amyotrophic Lateral Sclerosis, DiDe – Diagnostic Delay, EMG – Electromyography, GP – General Practitioner, MND – Motor Neuron Disease, NIV – Non-Invasive Ventilation.*

Keywords: *Amyotrophic Lateral Sclerosis; Diagnostic Delay; Referral Pathway; Breaking Bad News; Multidisciplinary Care*

Introduction

Amyotrophic lateral sclerosis is a progressive degeneration of upper and lower motor neurons producing weakness, spasticity and wasting, with most patients dying of respiratory complications within three to five years of symptom onset [1,2,3]. Onset is typically focal and insidious, spreading to other regions over months.

Early presentation mimics several other conditions, some treatable such as multifocal motor neuropathy, and some far commoner such as cervical myelopathy [4,5]. Diagnosis rests on clinical criteria supported by electromyography, and there is no confirmatory biomarker [6,7].

The consequence is a diagnostic interval of about a year, which in this disease represents a large share of the total illness [4,8]. This review examines the size and distribution of that interval, why it has proved resistant to change, and what follows once the diagnosis is finally given.

Material and Methods

Search strategy: PubMed and the archives of the major neurology, neuromuscular and palliative care journals were searched for studies of diagnostic delay and diagnosis disclosure in motor neuron disease. Search terms combined amyotrophic lateral sclerosis and motor neuron disease with diagnostic delay, time to diagnosis, misdiagnosis, referral and breaking bad news. Reference lists of reviews [8,9] were screened by hand for primary sources.

Eligibility: Studies were included if they quantified the interval from symptom onset to diagnosis, disaggregated that interval, examined determinants of delay, or reported patient and carer experience of receiving the diagnosis. Therapeutic trials are cited to establish what earlier diagnosis would make available and are identified as such.

Data extraction and handling: Design, setting, sample size, whether mean or median was reported and the interval definition were extracted as published. Whether the figure is a mean or a median is reported alongside every estimate, because the distribution is strongly right-skewed in this disease and the two differ by several months in the same cohort. No pooling or re-analysis was performed. Figures 1 to 4 and Tables 1 and 2 reproduce published values only. This is a narrative synthesis and was not registered as a systematic review.

Results

How long the delay is: A retrospective analysis of 260 patients at a tertiary referral centre found a median time from onset to diagnosis of 11 months (range 6 to 21), with 10 months in bulbar-onset and 12 months in spinal-onset disease, a difference that was not significant [4]. A cohort of 1405 patients across five centres in four countries reported the same median of 11 months, similar between centres [10]. Literature review across 21 studies from multiple continents places the delay at 10 to 16 months [8]. A French centre reported a mean of 17 months against a median of 12, illustrating the skew (Figure 1) [11].

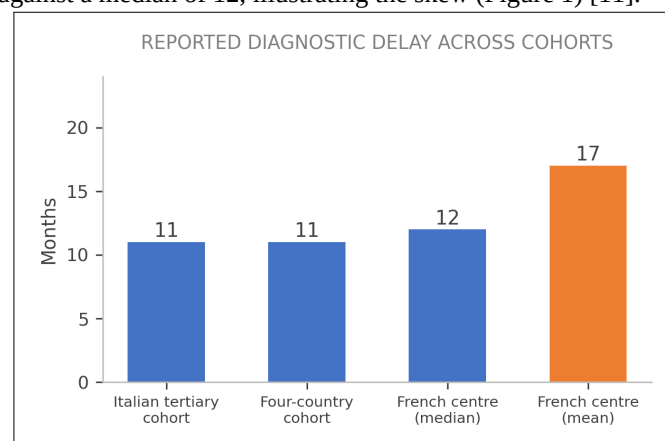


Figure 1: Reported diagnostic delay across cohorts, showing mean against median in the same series [4,10,11]

Where it accrues: The French cohort disaggregated the pathway. Mean delay was 2.7 months between first symptoms and the first complaint to a general practitioner, followed by a further 6.5 months before the first visit to a neurologist, with the remainder elapsing before diagnosis was reached (Figure 2) [11]. The largest single block therefore sits between primary care contact and neurological assessment.

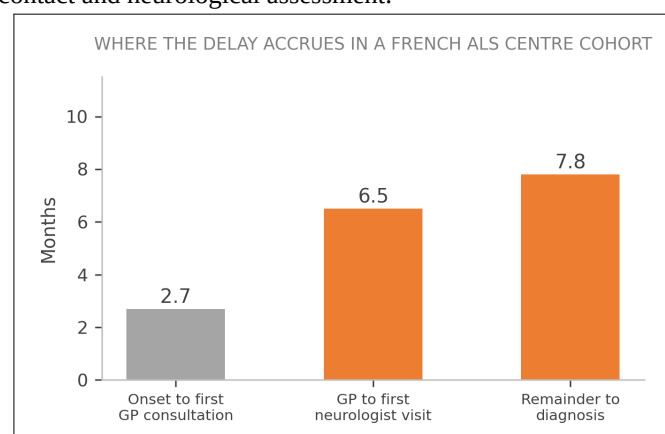


Figure 2: Distribution of the diagnostic interval in a French motor neuron disease centre cohort [11]

Who has been seen: In a large natural language processing analysis of clinical records, overall diagnostic delay from symptom onset was 11 months (interquartile range 6 to 18), and only 38.8 per cent of patients had visited a neurologist before diagnosis (Figure 3) [12]. Nearly half of patients are initially

misdiagnosed, attributed to limited familiarity with the disease in primary care and to genuine clinical difficulty distinguishing it from mimics [5,13].

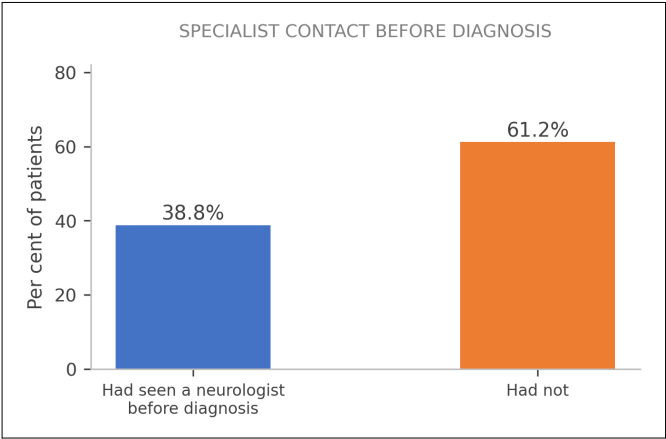


Figure 3: Proportion of patients who had seen a neurologist before diagnosis [12]

What the delay represents: Against a median survival of about three years from symptom onset, an 11-month diagnostic interval consumes roughly a quarter to a third of the illness (Figure 4) [3,4,14]. In one description the delay amounts to as much as two-thirds of total disease duration in some patients [11]. This is the feature that distinguishes delay in this disease from delay in most others: the denominator is small.

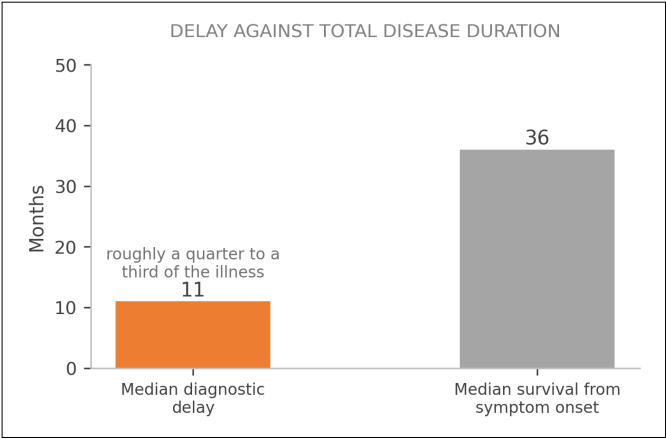


Figure 4: Median diagnostic delay set against median survival from symptom onset [3,4,11]

Study	Design and cohort	Principal reported finding
Cellura et al. 2012 [4]	260 patients, tertiary referral centre	Median delay 11 months; no bulbar versus spinal difference
Four-country cohort [10]	1405 patients, five centres	Median delay 11 months; similar across countries
Richards et al. 2020 [11]	90 patients, French ALS centre	Mean 17 months, median 12; 6.5 months GP to neurologist
Househam and Swash 2000 [15]	57 patients, England and Wales	Mean 16.2 months; incorrect initial diagnosis a major factor
Record-based analysis [12]	Natural language processing of clinical notes	38.8% had seen a neurologist before diagnosis
Literature review [8]	21 studies, multiple countries	Diagnostic delay 10 to 16 months
Brazilian population study [14]	218 patients on riluzole	Mean onset to diagnosis 22.7 to 23.5 months

Table 1: Principal studies reviewed, with the finding as reported by the original investigators

Why it has not improved: An English cohort published in 2000 reported a mean of 16.2 months and noted that the interval had shown no sign of improving despite the introduction of riluzole [15]. Cohorts two decades later report the same figure [4,10]. The identified contributors are consistent: an initial incorrect diagnosis, reluctance to consider the diagnosis when raised by an informed patient, failure to consider a neurological cause, and failure to refer early to a neurologist [15]. Cognitive error based on incorrect use of heuristics has been proposed as a mechanism [4]. One cohort found that earlier first medical evaluation was associated with a longer time to diagnosis, which is not a paradox: patients presenting very early present with less to find [10].

What earlier diagnosis makes available: Riluzole is prescribed to the large majority of patients, and in one cohort 87.6 per cent received it [12]; its effect on survival is modest but established [24,25], and edaravone has randomised support in a defined subgroup [26]. Beyond that, earlier diagnosis provides access to multidisciplinary clinic care, which is associated with improved survival, to non-invasive ventilation, taken up by 64 per cent in one series, to gastrostomy, undergone by 52 per cent, and to clinical trial enrolment, which is frequently precluded by advanced disease [12,16,17]. It also provides time: for advance care planning, for financial and employment decisions, and for the conversations families later say they wish they had been able to have [18,19]. The journey to a specialist multidisciplinary clinic has itself been mapped [31], and the burden carried by family carers is substantial [35].

Communicating the diagnosis: How the diagnosis is delivered is remembered for years and shapes subsequent engagement with services [19,20]. Consensus guidance recommends that it be given by a neurologist familiar with the disease, in person, with adequate time, with a companion present if the patient wishes, and with immediate arrangements for follow-up rather than an unstructured interval [21,22]. Patient

and carer accounts consistently report that being given the diagnosis without a plan, or in a rushed consultation, compounded the harm of the news itself [19,23].

Segment or factor	What the evidence shows	Implication
Onset to first GP visit	About 2.7 months on average	Insidious onset; symptoms attributed elsewhere [11]
GP to first neurologist	About 6.5 months on average	The largest single interval [11]
Seen a neurologist at all	Only 38.8% before diagnosis	Referral, not neurological assessment, is the bottleneck [12]
Initial misdiagnosis	Nearly half of patients	Mimics are commoner than the disease [5,13]
Change over 20 years	16.2 months in 2000; 11 months now	Little improvement despite awareness [4,10,15]
What is gained by speed	Access to care, ventilation, trials, time	Not disease modification [12,16,17]

Table 2: Segments of the diagnostic pathway and what earlier diagnosis would actually change

Discussion

The stability of this figure is its most striking feature. A cohort published in 2000 reported 16.2 months and observed that the interval had not improved despite riluzole becoming available; cohorts from 2012 and from a four-country collaboration report 11 months [4,10,15]. Two decades of increased awareness, more specialist centres and widespread patient access to information have shortened the interval modestly at best.

The distribution explains why. Roughly 2.7 months elapse before the patient consults anyone, which is unsurprising for insidious focal weakness, and a further 6.5 months before they reach a neurologist [11]. Only 38.8 per cent had seen a neurologist at all before diagnosis in one large record-based analysis [12]. The bottleneck is referral, not neurological assessment, and it is difficult to fix by exhortation because early disease genuinely resembles commoner conditions and because a general practitioner will encounter perhaps one case in a career [5,13,27]. Delay also restricts entry to clinical trials [28], and successive reviews report that the problem persists [29,30].

There is an uncomfortable question underneath all of this, and reviews of diagnostic delay usually avoid it. In most conditions, shortening delay is justified by treatment that works better earlier. Here disease-modifying therapy is limited, so what is gained is real but different: multidisciplinary care associated with improved survival, respiratory support, nutritional intervention, eligibility for trials, and time to arrange a life around a prognosis [12,16,17]. Those are substantial. They are not the same argument, and conflating them overstates what an earlier diagnosis delivers.

A related point is that earlier is not unambiguously better for every patient. Bringing forward the moment at which someone learns they have an untreatable fatal illness extends the period they live with that knowledge, and a minority of patients say

they would not have wanted to know sooner [19,23]. That does not argue for delay, which is currently produced by system failure rather than by choice. It argues that the consultation matters as much as its timing, and the literature on delivering this diagnosis is clear that doing it without time, without a companion and without an immediate follow-up plan causes lasting harm [19,21,22].

Study limitations

The limitations belong to the underlying literature. Almost all estimates depend on retrospective recall of symptom onset in a disease with insidious focal onset, and patients frequently cannot date it [4,11]. Cohorts are drawn from tertiary referral centres, which excludes patients who died before reaching one and those never referred [4,10]. Mean and median are reported inconsistently and differ by several months in the same cohort, so figures quoted without that label are ambiguous [11]. Diagnostic criteria have been revised over the period these cohorts span, altering the point at which diagnosis is recorded [6,7]. Record-based analyses depend on documentation quality, and specialist contact may be under-recorded [12]. Studies of diagnosis disclosure are small, qualitative and subject to recall shaped by subsequent events [19,23]. The association between multidisciplinary care and survival is observational and confounded by referral of fitter patients [16]. And the evidence is drawn almost entirely from high-income countries with organised neurology services [8,10]. Biomarker development that might shorten the interval remains at an early stage [32,33,34].

Conclusion

It takes about eleven months to diagnose amyotrophic lateral sclerosis, most of that spent between a first primary care consultation and a first neurological one, and the figure has barely shifted in twenty years. In a disease with a median survival of three years, that interval is a quarter to a third of everything the patient has left. The honest case for shortening it is not that treatment works better when started sooner, because the available disease-modifying therapy is limited. It is that the patient gains access to multidisciplinary care, respiratory and nutritional support, trial eligibility, and the time to make decisions while still able to make them. That last one is not a clinical endpoint and it is probably the most valuable, which is also why how the diagnosis is delivered deserves as much attention as when.

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Conflict of Interest

The authors declare no competing interests.

Ethics Statement

This article reviews previously published studies only. No new patient data were collected and no participants were recruited, so ethical approval was not required.

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