

ALS Diagnostic Criteria: The Gold Standard

Scientia Professor Matthew C. Kiernan



Matthew.Kiernan@neura.edu.au

Disclosure/Conflict of Interest



World Federation of Neurology

Fostering quality neurology and brain health worldwide



International Federation of
CLINICAL NEUROPHYSIOLOGY



Australian and New Zealand
Association of Neurologists

Learning Objectives

1. Understand the investigations necessary to make a diagnosis of ALS
2. Understand the concepts about ALS consensus criteria
3. Discuss the background and validation of the Gold Coast Criteria for ALS
4. Anticipate the utility of biomarkers in ALS Clinical Trials



World Federation of Neurology

Fostering quality neurology and brain health worldwide



Key Message

Accurate and early diagnosis is critical to enabling ALS patients and their carers to plan for the disease course, optimising disease management, and giving therapies the best chance of success.

A more streamlined approach to diagnosis enhances recruitment into clinical trials, which are key to tackling ALS and ultimately improving patient outcomes.



Opinion Paper

A proposal for new diagnostic criteria for ALS

Jeremy M. Shefner^{a,*}, Ammar Al-Chalabi^b, Mark R. Baker^c, Li-Ying Cui^d, Mamede de Carvalho^e, Andrew Eisen^f, Julian Grosskreutz^g, Orla Hardiman^h, Robert Hendersonⁱ, Jose Manuel Matamala^j, Hiroshi Mitsumoto^k, Walter Paulus^l, Neil Simon^m, Michael Swashⁿ, Kevin Talbot^o, Martin R. Turner^p, Yoshikazu Ugawa^p, Leonard H. van den Berg^q, Renato Verdugo^r, Steven Vucic^s, Ryuji Kaji^t, David Burke^u, Matthew C. Kiernan^v



References

1. Shefner JM, Al-Chalabi A, Baker MR, Cui LY, de Carvalho M, Eisen A, Grosskreutz J, Hardiman O, Henderson R, Matamala JM, Mitsumoto H, Paulus W, Simon N, Swash M, Talbot K, Turner MR, Ugawa Y, van den Berg LH, Verdugo R, Vucic S, Kaji R, Burke D, Kiernan MC. A proposal for new diagnostic criteria for ALS. *Clin Neurophysiol*. 2020;131(8):1975-8.
2. Hannaford A, Pavey N, van den Bos M, Geevasinga N, Menon P, Shefner JM, ... Kiernan MC, et al. Diagnostic Utility of Gold Coast Criteria in Amyotrophic Lateral Sclerosis. *Ann Neurol*. 2021;89(5):979-86.
3. Vucic S, Ferguson TA, Cummings C, Hotchkin MT, Genge A, Glanzman R, et al, ... Kiernan MC. Gold Coast diagnostic criteria: Implications for ALS diagnosis and clinical trial enrollment. *Muscle Nerve*. 2021;64(5):532-7.
4. Kiernan MC, Shefner JM, Kaji R, Burke D. Amyotrophic lateral sclerosis: a new diagnostic paradigm. *J Neurol Neurosurg Psychiatry*. 2020;91(9):903-4.
5. Jewett, G., S. Khayambashi, G. S. Frost, B. Beland, A. Lee, V. Hodgkinson, L. Korngut, S. Chhibber. Gold Coast criteria expand clinical trial eligibility in amyotrophic lateral sclerosis. *Muscle Nerve* 2022;66:397-403.
6. Pugdahl K, Camdessanché JP, Cengiz B, de Carvalho M, Liguori R, Rossatto C, Oliveira Santos M, Vacchiano V, Johnsen B. Gold Coast diagnostic criteria increase sensitivity in amyotrophic lateral sclerosis. *Clin Neurophysiol* 2021;132:3183-3189.
7. Odat RM, Alomari O, Elgenidy A. Evaluation of the Gold Coast criteria as a diagnostic criteria of amyotrophic lateral sclerosis. *eNeurologicalSci*. 2024 Aug 30;37:100524.
8. Shen DX, Yang Y, Wang, He X, Sun Z, Cai J, Li, Liu M, Cui L. The Gold Coast criteria increases the diagnostic sensitivity for amyotrophic lateral sclerosis in a Chinese population. *Transl Neurodegener* 2021;10:28.
9. Kiernan MC, Timmins HC. Utility of the Gold Coast criteria for amyotrophic lateral sclerosis: Experience with fast progressors. *J Neurol Sci*. 2025 Mar 15;470:123417.
10. Timmins HC, Thompson AE, Kiernan MC. Diagnostic criteria for amyotrophic lateral sclerosis. *Curr Opin Neurol*. 2024 Oct 1;37(5):570-576.
11. Kiernan MC. Recent developments in consensus diagnostic criteria for amyotrophic lateral sclerosis. *eNeurologicalSci*. 2025 Feb 1;38:100559. doi: 10.1016/j.ensci.2025.100559.