Appendix A: Details of the studies included in this exceptional surveillance review of motor neurone disease NG42

Studies on cognitive assessments

Study Details	Population	Assessment tool	Reference Standard	Outcomes
Gosselt et al. 2020 (qualitative overview/comparison of ALS-specific screening tools)	ALS	Cognition: ACE-R, ALS-BCA, ALS-CBS, ECAS, FAB, MMSE, MoCA, PSSFTS, UCSF-SB Behaviour: ALS-FTD-Q, AES, BBI, DAS, FBI, FrSBe, MiND-B, NPI	Not reported	**Validated tools identified and outcomes per study identified against e.g., FrSBe, AES, ALS-FTD-Q):** BBI cut-off 22.5: sensitivity: 90%, specificity: 96% (moderate changes) (Elamin et al. 2017) MiND-B cut-off 35/36 or 33/36: sensitivity: 81-90%, specificity: 50-75% (Mioshi et al. 2014) ALS-CBS cut-off 35: sensitivity: 83%, specificity: 69% (Turon-Sans et al. 2016)
Gray et al. 2022 (Exploratory survey)	MND	ECAS	-	Clinician responses regarding use of cognitive assessments in MND clinics. Themes: identify and validate changes in cognition and behaviour, aid understanding of clinical impact of disease, inform/direct care, patient decision-making abilities. Factors affecting implementation/administration of cognitive assessments in clinics
Hodgins et al. 2020 (Survey of current clinical practices)	ALS	ECAS	-	Most health care service responses used ECAS. Themes: awareness about cognitive and behavioural changes, validation/reassurance, aids understanding of patients' presentation and informs clinical-decision-making (e.g., suitability

				of interventions, MDT adaptation, discussions about EOL, referral to other services, identifying carers support needs)
Radakovic et al. 2020 (modified Delphi method	MND	MiNDToolkit	-	Developed structured guidance/recommendations/techniques for non-pharmacological management of cognitive/behavioural impairment
Simon et al. 2019 Systematic review of validation studies	14 observational studies assessing validity of screening tools	ECAS, ALS-CBS, ALS-FTD-Q-J, ALS-FTD-Q, MiND-B, BBI	Standardised batteries of neuropsychological function, questionnaires, current diagnostic criteria, recommended cut-offs from previous literature	**<u>Results reported for individual studies in the SR:</u>** **Pinto-Grau et al. 2017*** (varied cut-offs): ECAS Total: sensitivity: 100%, specificity:80%, PPV: 37.5%, NPV: 100% ECAS Specific: sensitivity: 100%, specificity: 85%, PPV: 43%, NPV: 100% ECAS non-specific: sensitivity: 100%, specificity: 79%, PPV: 44%, NPV: 100% **Niven et al. 2015*** (ECAS total score cut-off): ECAS ≤ 105: sensitivity:69%, specificity:89%, PPV:75%, NPV:86% ECAS ≤ 107: sensitivity:77%, specificity:81%, PPV:67%, NPV:88% ECAS ≤ 108: sensitivity: 85%, specificity: 81%, PPV:69%, NPV: 92% ECAS ≤ 110: sensitivity:92%, specificity: 81%, PPV: 71%, NPV:96%

				ECAS ≤ 115: sensitivity: 100%, specificity: 52%, PPV: 50%, NPV: 100% **Lulé et al. 2015*** ECAS memory: sensitivity: 33%, specificity: 92% ECAS language: sensitivity:33%, specificity:75% ECAS fluency: sensitivity: 50%, specificity:91% ECAS executive function: 43%, specificity 100% **Turon-Sans et al. 2016*** ALS-CBScog, cut-ff: 8 no FTD vs FTD: sensitivity: 83.3%, specificity: 75%, PPV: 31.3%, NPV: 97.1% ALS-CBScog: cut-off: 15 normal vs CI: sensitivity: 86.2%, specificity: 62%, PPV: 75.8%, NPV: 76.5% ALS-CBSbv, cut-off: 35 No FTD vs FTD: sensitivity: 83.3%, specificity: 69%, PPV: 25%, NPV: 96.7 ALS-CBSbv, cut-off: 36 normal vs CI: sensitivity: 93.3%, specificity: 74.3%, PPV: 61%, NPV: 96.3% **Branco et al. 2017** ALS-CBS: cutoff 10 ALS with executive impairment vs. ALS without executive impairment: sensitivity: 90%, specificity: 87.2% **Woolley et al. 2010***

				ALS-CBS (cognitive): Cut off 10 for FTD: sensitivity: 100%, specificity: 100%, PPV, 100%, NPV: 100% ALS-CBS (cognitive): cut-off 17 for any cognitive deficit: sensitivity: 85%, specificity: 86%, PPV: 69%, NPV: 71% ALS-CBS behaviour: cut-off 32 for FTD: sensitivity: 88%, specificity: 80%, PPV: 94%, NPV: 67% ALS-CBS behaviour: cut-ff 36 for any behavioural deficit: sensitivity: 90%, specificity: 86%, PPV: 82%, NPV: 92% **Hsieh et al. 2016** MiND-B, cut-off: 33: sensitivity: 90%, specificity: 79%, PPV:73%, NPV:78%, LR+: 4.32 **Mioshi et al. 2014** MiND-B, cut-off 35: sensitivity: 90%, specificity: 50%, PPV: 77.8%, NPV: 72.7%. Cut-off 33, sensitivity: 81%, specificity:75% **Elamin et al 2017** BBI cut-off 7: sensitivity: 87.9%, specificity: 78.9%, PPV: 72.5%, NPV: 91.1% BBI cut-off 22.5: sensitivity: 90%, specificity: 96%, PPV: 75%, NPV: 98.7% **Pinto-Grau et al. 2017** BBI cut-off 7: sensitivity: 50%, specificity: 76%

				BBI cut-off 23: sensitivity: 100%, specificity: 92% *these studies were also identified from literature search
Aiello et al. 2022 (observational)	ALS (n=89)	ECAS	Neuropsychological assessment	ECAS and subscales: sensitivity: ALS-specific and executive: 73.9-78.3%, ALS-non-specific subscales: 8.7-17.4%, specificity: 81.8-100%, PPV: 60-100%, NPV: 75.9-91.5%
Aiello et al. 2023 (observational)	ALS (n=348)	FAB	ECAS	FAB cut off ≤ 15: sensitivity: 78.8%, specificity: 71.6%. Cut-off <15.6: sensitivity: 81.8%, specificity: 68.6%
Beeldman et al.2021 (observational)	ALS (n=72), bvFTD (n=5 with concurrent ALS), healthy controls (n=34). A subset of AL patients (n=29) age-matched and education-matched with healthy controls were administered the ECAS	ALS-FTD-cog, ECAS total, ECAS ALS-specific	Neuropsychological examination	ALS-FTD-cog: cut-off ≥ 1 (impairment): sensitivity: 65%, specificity: 63.5%. Cut-off 46.9: sensitivity: 65%, specificity: 75% ALS-bvFTD: sensitivity: 94.4%, specificity: 100% ECAS total and ECAS specific: sensitivity: 83.3% (ALS), specificity: 82.6% and 91.3% respectively
Greco et al. 2022 (observational)	Definite or probable ALS diagnosis (n=154)	ECAS	ALS-CBS	The distribution of patients according to Strong criteria was different for ECAS and ALS-CBS and the degree of agreement between the two tests in terms of Cohen's Kappa coefficient resulted equal to 0.2047 (95% CI 0.1122 to 0.2973)
Lazzonling et al. 2022 (observational)	ALS (n=90), healthy participants (n=100)	BBI	FRSBe	BBI: sensitivity: 85.7%, specificity: (79.7%) (cut-off 10.5)
Saxon et al. 2020 (observational)	ALS-FTD (n=20), bvALS (n=23), and healthy controls (n=30)	ECAS	Standard full length neuropsychological test directly comparable to ECAS	Frequency of impairment on ECAS: ALS-specific domain in detecting ALS-FTD: sensitivity 90%, specificity: 20% (non-ALS-specific impairment); bvFTD: sensitivity:78%, specificity: 35% (non-ALS-specific impairment)

				ECAS vs neuropsychological tests: ECAS subdomain sensitivity: naming: 56%, graded naming 68%, spelling:61%, PALPA spelling: 64%, sentence completion: 55%, Hayling test: 76%, social cognition: 47%, judgement of preference: 55%. Specificity: 100%, with exception of PALPA spelling (93%), sentence completion (97%), judgement of preference (97%)
Tiokrowikoto et al. 2023 (observational)	MND (n=64), healthy controls (n=45)	ECAS	Standard neuropsychology tests of executive function and social cognition	ECAS: ALS-specific score: low/moderate sensitivity (social cognition, inhibition, working memory deficit), high specificity. High sensitivity and specificity for alternation deficits. ECAS: executive function domain: low sensitivity and high specificity for all four executive function subdomains. ECAS individual subtests: good sensitivity, high specificity. Sensitivity was low for social cognition.

Abbreviations: Amyotrophic lateral sclerosis (ALS); ALS Cognitive Behavioural Screen (ALS-CBS); Amyotrophic Lateral Sclerosis-Frontotemporal Dementia-Questionnaire (ALS-FTD-Q); Apathy Evaluation Scale (AES); Beaumont Behavioral Inventory (BBI); bv: behavioural variant; cog: cognitive; Dimensional Apathy Scale (DAS); Edinburgh Cognitive and Behavioural ALS Screen (ECAS); Frontal Assessment Battery (FAB);Frontal Behavioral Inventory (FBI); Frontal Systems Behavior scale (FrSBe); Frontotemporal Dementia (FTD); Multidisciplinary team (MDT); Motor Neurone Disease (MND); Motor Neurone Disease Behavior scale (MiND-B); Neuropsychiatric Inventory (NPI); Not reported (NR)

Studies on prognostic factors

Study Details	Population	Assessment tool	Reference Standard	Outcomes
Caga 2016 (observational)	ALS (n=76), caregivers	Apathy on the CBI-R	Neurological examination and assessment, ALSFRS-R	CBI-R cut-offs: 0%–25%, mild; 26%–50%, moderate; 51%–75%, severe; 76%+, very severe. Kaplan-Meier graph for survival in population with no apathy, mild apathy, and moderate/severe apathy from time of diagnosis. Apathy was associated with mortality, with those with moderate/severe apathy surviving for a shorter time (21.7 months) than people with mild apathy (46.9 months) and no apathy (51.9 months) (P=0.0001). Apathy remained significantly associated with survival time (HR 3.8, 95% CI 1.9-7.5; P=0.0001) after adjusting for cognitive status, disease status and symptom duration. Cognitive status was the only confounder that differed between the groups
Nguyen 2021 (observational)	ALS (n=134)	MiND-B (behaviour), ACE (cognition)	-	Multivariate regression analysis of survival with prognostic risk factors including site of onset, age, pre-diagnostic duration, and baseline ALSFRS-R score: Behavioural impairment in MiND-B cut-off >33 vs MiND-B ≤ 33: HR 2.53 (95% CI 1.3-4.6), P=0.003. Those without behavioural impairment had a median survival of 46months compared to 19 months in those with impairment.

Study Details	Population	Assessment tool	Reference Standard	Outcomes
				Multivariate regression analysis of survival with prognostic risk factors including site of onset, age, pre-diagnostic duration, and baseline ALSFRS-R score. Cognitive impairment in ACE < 88 vs ACE ≥ 88: HR 2.07 (95% CI 1.04-3.3), P 0.042. Those without cognitive impairment had a median survival of 46 months compared to 20 months in those without impairment

Abbreviations: Addenbrooke's Cognitive Examination (ACE); Amyotrophic Lateral Sclerosis (ALS); Amyotrophic Lateral Sclerosis Functioning Rating Scale-Revised (ALSFRS-R); Cambridge Behavioural Inventory-Revised (CBI-R); Hazard Ratio (HR), Motor Neurone Disease Behavioural Instrument (MiND-B)

Studies on organisation of care

Abbreviations: Amyotrophic Lateral Sclerosis (ALS); Motor Neurone Disease (MND)

Study Details	Population	Outcomes
Ando et al. 2022 (qualitative: semi-structured interviews)	MND (n=26)	Thematic analysis: four themes were important for quality of life of participants: perceived illness prognosis, sense of self, concerns for significant others, life to enjoy. These factors reflected psychological stress from MND, participants value system, and beliefs about life. Regulatory flexibility (mindful approach) and psychological flexibility (savouring positive experiences) were important to maintain and enhance quality of life.
Young et al. 2019 (Trajectories of Outcomes in Neurological Conditions (TONiC)) (questionnaire on demographic factors and patient-reported outcome measures)	ALS/MND (n=636)	Wilson and Cleary modelling showed importance of physical functioning and anxiety upon quality of life. Breathlessness and fatigue had indirect effects. The model was invariant for gender and onset type
Zarotti et al 2019 (qualitative: focus groups)	People with MND (n=51), health care professionals involved with MND care (n=47), service user representatives (n=4)	Themes from qualitative synthesis: psychological adjustment, patient engagement, nutrition and the need for control, knowledge of nutrition and complexity of MND, psychological nature of eating.

Studies on psychological support

Study Details	Population	Intervention	Controls	Outcomes
Lapin et al. 2022 (observational)	ALS (n=578) The study aimed to test bio-psycho-social model of QOL by investigating direct effects of physical, psychological variables and estimate whether the relationships differ in early vs late disease stage	NA	NA	Association/correlation of patient-reported outcomes and disease stage: Early disease stage: ALSFRS-R and EQ-5D utility: Pearson r= 0.56 (95% CI 0.49-0.65; P<0.001) ALSFRS-R and EQ-VAS: Pearson r=0.41 (95% CI 0.33-0.49, P<0.001) ALSFRS-R and PHQ-9: Pearson r=-0.37 (95%CI-0.45—0.28, P<0.001) Late disease stage: ALSFRS-R and EQ-5D utility Pearson r=0.57 (95% CI0.46-0.62, P<0.001) ALSFRS-R and EQ-VAS Pearson r= 0.24 (95% CI 0.09-0.39, P<0.05) ALSFRS-R and PHQ-9 Pearson r=-0.05 (95% CI -0.20-0.11)
Weeks et al. 2019 (qualitative)	MND (semi-structured interviews, n=22; workshops, with pwMND, n=15, informal	NA	NA	Overarching themes identified: Unfamiliar territory: participants felt that there is a lack of understanding and

	caregivers, n=10, and MND healthcare professionals, n=12)			knowledge about the disease among the general population and particularly non-MND health professionals. Participants and their caregivers said that health professionals did not provide adequate information regarding the condition or support at diagnosis, however, some participants felt that information provided was adequate. A series of losses: recurring theme; sense of loss experienced by people with MND related to physical functioning, mobility, independence, future hopes and dreams, social relationships and identity. Associations included anger, frustration, uncertainty, and hopelessness. Variability and difficulty meeting individual needs: recurring theme; people with MND have different symptoms, rates of progression, experiences and needs. Any psychological intervention would need to be flexible due to the nature of the disease. Informal support: health professionals highlighted different types of support including peer, family members, and formal caregivers. They also reported that support from other people with MND was important. Caregivers and health professionals discussed benefits of engaging family members or caregivers in sessions as they would be able to help with equipment, communication, and encouraging overall

				engagement. Fear of burden among people with MND was reported (attending appointments, taking medication, and day-to-day personal care). People with MND had anxiety about how carers would cope after their death, familial MND.
Groenestijn et al 2015 (single blind multicentre RCT, 43 weeks)	ALS (N=15), and caregivers People with HADS score of ≥ 8 randomised Mean age: (years, SD): CBT: 57.4 (50.9); UC:54.8 (31.9) Male (%): CBT: 60; UC: 60 ALSFRS-R (n, %): severe: CBT: 0 (0%); UC: 1 (20%) Moderate: CBT: 1 (10%); UC: 0 (0%) Mild: CBT: 9 (90%); UC: 4 (80%)	CBT+ usual care (n=10) CBT: stress coping model; sessions were provided by three trained and instructed psychologists. Participants received individualised or as patient/caregiver, 5-10 sessions within 16 weeks, each session lasted for 1 hour. Six modules focused on coming to terms with ALS diagnosis, coping with emotional instability, maintaining autonomy, mobilising social support, coping with fear of the future, maintaining activity levels. The psychologist tailored the intervention to the	Usual care (not described) (n=5)	Due to low patient numbers, the researchers reduced the number of outcomes as reported in the study protocol Health Survey Short Form (QOL): outcome reported in graph format; mental health deteriorated less in the CBT group compared to the UC group (P 0.05) Mental Component Summary (SF-36 MCS) (patient and caregiver measures): caregiver mental health improved in the CBT group compared to UC but decreased in UC during follow up (P<0.05) Emotional Functioning subscale of the ALS Assessment Questionnaire (ALSAQ-40): not reported Hospital Anxiety and Depression Scale (HADS) as a measure of psychological distress (patient and caregiver measures): no significant difference over time in patients (P=0.26) nor caregivers (P=0.18)

		needs of patient/caregiver.		Caregiver Strain Index (CSI): remained stable over time in the CBT group but increased in the UC group (P<0.05).
De Wit et al. 2020 (RCT)	ALS or PMA (N=148) and caregivers Patients: Mean age (SD): CBT: 62 (11); waitlist: 63 (8.9) % female: CBT: 35.5; waitlist: 36.5 Level of functioning ALSFRS-R (mean, SD): CBT: 31.7 (9.8); waitlist: 31.0 (9.5) ALS-FTD-Q (mean, SD): CBT: 16.3 (11.8); waitlist: 17.2 (13.9) Caregivers: Mean age (years, SD): CBT: 62 (10.6); waitlist: 61 (9.8) % female: CBT: 48; waitlist: 48	Psychological support programme (face-to-face and online) (n=74) The intervention consisted of 1 face-to-face contact, 6 online guided modules (each module lasted 1 hour 30 minutes), and 1 closing telephone contact, guided by a psychologist. The intervention was an adapted format of an Acceptance and Commitment Therapy (ACT) based programme for patients with cancer and their caregivers. Modules focused on a specific theme (e.g., coping with emotions and thoughts). The sessions included psychoeducation, psychological, mindfulness exercises. Patients were also given information and	Waitlist (n=74) (not reported)	Psychological distress (HADS, mean, SD) (26 weeks): CBT (n=48): 11 (7.71); waitlist (n=60): 11.48 (7.42) Caregiver burden (Zarit Burden Interview, mean, SD, 26 weeks): CBT (n=48): 13.89 (7.76); waitlist (n=60): 14.20 (7.58) Caregiver quality of life (Care Related Quality of Life, mean, SD, 26 weeks): CBT (n=48): 10.09 (2.23); waitlist (n=60): 9.97 (2.09) Patients quality of life (McGill Quality of Life Questionnaire, mean, SD, 26 weeks): CBT (n=24): 6.63 (1.86); waitlist (n=31): 6.52 (2.10) Patients distress (HADS, mean, SD, 26 weeks): CBT (n=24): 9.33 (7.75): waitlist (n=31): 11.87 (6.98) Satisfaction with blended support (CSQ-8): CBT: 25.57 (3.8). Participants were least satisfied with peer contact and mindfulness exercises. The support programme had no effect on primary and secondary outcomes. Adherence: 19% caregivers dropped out of the study, the most common reason being the death of the patient. Drop-outs had

		references for relevant websites. Patients received feedback from the psychologist after each module. Patients could contact other patients via private messaging to share advice using a forum. In the closing telephone call (30 minutes) the caregiver could ask for advice.		partners with significantly lower physical functioning compared to completers
Paganini et al. 2022 (RCT)	ALS (n=47), caregivers (n=27)	Mindfulness programme (n=not reported)	Waitlist (n=not reported)	Patient's QOL 5 weeks post-treatment, and 3, 6 months follow-up: higher levels of QOL observed in the intervention group after treatment compared to waitlist. Anxiety and depression: lower scores observed in patients compared to waitlist over time. Care burden: lower scores in caregivers, as well as lower scores for depression and anxiety, with higher scores for energy and emotional well-being.
Averill et al 2013 (RCT)	ALS (N=48)	Expressive disclosure (n=27). This group was asked to either write or talk into a tape recorder about their deepest thoughts and feelings related their	Control (n=27). No disclosure exercise but did include completion of study measures. A telephone call was	Measures at 3 and 6 months after the intervention: Psychological wellbeing outcomes: Affects Balance Scale (positive affect, negative affect): No short-term effects of the

		ALS experience (20 minutes every day for 3 days) over 1 week. Participants were provided with written instructions on how to complete the exercise, suggestions and paper to write. They were asked to record start and stop times for writing and speaking. A telephone call towards the end of the last scheduled day of disclosure was conducted by an investigator	scheduled following week 1 after time 1	intervention on negative effects were observed. However, long term there was a significant group and timing interaction (P<0.05). Control group: significant decrease in wellbeing from baseline at 3 months post-intervention (P<0.05) Geriatric Depression Scale (depression) McGill Quality of Life Questionnaire (psychological quality of life, existential quality of life) Ambivalence over Emotional Expression (extent to which participants felt uncomfortable or regret expressing their emotions): associated with lower wellbeing across whole study Emotional Approach Coping (emotional processing) Social Constraints Scale (perceived inadequacy of social support) Multi-level modelling showed that participants with disclosed thoughts and feelings about ALS had higher wellbeing compared to control group at 3 months post-intervention (1 SD improvement) but not at 6 months. The control group experienced 0.15 SD decrease in wellbeing at 3 months. Participants with low ambivalence over emotional expression

				(AEE) had higher wellbeing compared to those with high AEE regardless of condition.
Raglio et al 2016 (RCT)	ALS (N=30)	Active music therapy (12 sessions 3 times a week) (n=not reported)	Standard care/control (physical and speech rehabilitation sessions, occupational therapy, psychological support) (n=not reported)	Outcome measures: ALSFRS-R, HADS, McGill Quality of Life Questionnaire, Music Therapy Rating Scale The active music therapy group improved in McGill Quality of Life Questionnaire (P=0.035), and there was a positive trend in nonverbal and sonorous music relationship during the treatment

Abbreviations: Amyotrophic Lateral Sclerosis (ALS); Amyotrophic Lateral Sclerosis-Functional Rating Scale-Revised (ALSFRS-R); Amyotrophic Lateral Sclerosis-Frontotemporal Dementia-Questionnaire (ALS-FTD-Q); Cognitive Behavioural Therapy (CBT); Hospital Anxiety and Depression Scale (HADS); Not applicable (NA); Progressive Muscular Atrophy (PMA); Randomised controlled Trial (RCT); Standard Deviation (SD); Quality of Life (QOL)

Studies on cough effectiveness

Study Details	Population	Intervention	Controls	Outcomes
Nicolini et al. (2022) (preliminary RCT)	ALS (N=30)	Mechanical insufflation/exsufflation (MI/E) and expiratory flow accelerator (EFA) technology (n=15)	Mechanical insufflation/exsufflation technology only (n=15)	Outcome measures at 1, 6 and 12 months: Respiratory function, respiratory muscle strength, peak cough flow decreased in both groups but significantly less in the combined intervention group. Gas exchanged decreased over time in both groups (no difference between groups). No differences were observed in other outcomes: number of exacerbations, comfort and distress. No AEs were reported. Perceived cough efficacy increased in both groups.

Abbreviations: Adverse event (AE); Randomised controlled trial (RCT)

References

Aiello EN, Iazzolino B, Pain D, Peotta L, Palumbo F, Radici A et al. The diagnostic value of the Italian version of the Edinburgh Cognitive and Behavioural ALS Screen (ECAS). Amyotrophic Lateral Sclerosis and Frontotemporal Degeneration. 2022; 23 (7-8): 527-531

Aiello EN Solca F, Torre S, Carelli L, Ferrucci R, Priori A et al. Feasibility and diagnostics of the frontal assessment battery (FAB) in amyotrophic lateral sclerosis. Neurological Sciences. 2023; 44 (2): 587-592

Andersen PM, Borasio GD, Dengler R et al. (2005) EFNS task force on management of amyotrophic lateral sclerosis: guidelines for diagnosing and clinical care of patients and relatives. European Journal of Neurology 12: 921–38

Aridegbe T, Kandler R, Walters SJ, Walsh T, Shaw PJ, McDermott CJ.The natural history of motor neuron disease: assessing the impact of specialist care, Amyotrophic Lateral Sclerosis and Frontotemporal Degeneration. 2013; 14:1, 13-19

Ando H, Cousins R, Young CA. Flexibility to manage and enhance quality of life among people with motor neurone disease. Disability and Rehabilitation. 2022; 44(12):2752-2762

Averill AJ, Kasarskis EJ, Segerstrom SC. Expressive disclosure to improve wellbeing in patients with amyotrophic lateral sclerosis: a randomised controlled trial. Psychology and Health. 2013; 28 (6): 701-13

Beeldman E, Govaarts R, de Visser M, van Es MA, Pijnenburg YAL, Schmand BA et al. Screening for cognition in amyotrophic lateral sclerosis: test characteristics of a new screen. Journal of Neurology. 2021; 268 (7): 2533-2540

Bersano E, Sarnelli MF, Solara V, Iazzolino B, Peotta L, De Marchi F, et al. Decline of cognitive and behavioural functions in amyotrophic lateral sclerosis: a longitudinal study. Amyotrophic Lateral Sclerosis and Frontotemporal Degeneration. 2020; 21(5-6):373-9

Caga J, Turner MR, Hsieh S, Ahmed RM, Devenney E, Ramsey E, et al. Apathy is associated with poor prognosis in amyotrophic lateral sclerosis. European journal of neurology. 2016; 23(5):891-7

Cerutti P, Marconi A, Pozzi S, Gragnano G, Pain D et al. Psychological intervention in amyotrophic lateral sclerosis: suggestions for clinical practice. J Psychology and Clinical Psychiatry. 2017; 8(2): 00480

De Wit J, Beelan A, Drossaert CHC, Kolijn R, Van Den Berg LH, Schroder CD et al. Blended psychosocial support for partners of patients with ALS and PMA: results of a randomized controlled trial. Amyotrophic Lateral Sclerosis and Frontotemporal Degeneration. 2020; 21 (56): 344-54

Elamin M, Pinto-Grau M, Burke T, Bede P, Rooney J, O'Sullivan M, Lonergan K, Kirby E, Quinlan E, Breen N, Vajda A, Heverin M, Pender N, Hardiman O. Identifying behavioural changes in ALS: Validation of the Beaumont Behavioural Inventory (BBI). Amyotrophic Lateral Sclerosis Frontotemporal Degeneration. 2017;18(1-2):68-73.

Gosselt IK, Nijboer TCW, van Es MA. An overview of screening instruments for cognition and behaviour in patients with ALS: selecting the appropriate tool for clinical practice. Amyotrophic Lateral Sclerosis and Frontotemporal Degeneration. 2020; 21(5-6):324-36

Gould RL, Thompson BJ, Rawlinson C, Kumar P, White D, Serfaty MA et al. A randomised controlled trial of acceptance and commitment therapy plus usual care compared to usual care alone for improving psychological health in people with motor neuron disease (COMMEND): study protocol. BMC Neurology. 2022; 22 (1): 431

Gray D, Abrahams S. International evaluation of current practices in cognitive assessment for motor neurone disease. British Journal of Neuroscience Nursing. 2022; 18(1):38-44.

Greco LC, Lizio A, Casiraghi J, Sansone VA, Tremolizzo L, Riva N, et al. A preliminary comparison between ECAS and ALS-CBS in classifying cognitive-behavioural phenotypes in a cohort of non-demented amyotrophic lateral sclerosis patients. Journal of Neurology. 2022; 269(4):1899-1904

Hodgins F, Mulhern S, Abrahams S. The clinical impact of the Edinburgh Cognitive and Behavioural ALS Screen (ECAS) and neuropsychological intervention in routine ALS care. Amyotrophic Lateral Sclerosis and Frontotemporal Degeneration. 2020; 21(1-2):92-9.

Iazzolino B, Pain D, Laura P, Aiello EN, Gallucci M, Radici A et al. Italian adaptation of the Beaumont Behavioural Inventory (BBI): psychometric properties and clinical usability. Amyotrophic Lateral Sclerosis Frontotemporal Degeneration. 2022; 23(1-2):81-86.

Lapin B, Mate K, Li Y, Thakore N. Subjective health perception prioritizes psychological well-being over physical function in advanced ALS: A multigroup structural equation modelling analysis. Journal of the Neurological Sciences. 2022; 442:120442

Lulé D, Burkhardt C, Abdulla S, Böhm S, Kollewe K, Uttner I et al. The Edinburgh Cognitive and Behavioural Amyotrophic Lateral Sclerosis Screen: a cross-sectional comparison of established screening tools in a German-Swiss population. Amyotrophic Lateral Sclerosis Frontotemporal Degeneration. 2015 ;16(1-2):16-23

Macpherson CE, Bassile CC. Pulmonary physical therapy techniques to enhance survival in amyotrophic lateral sclerosis: a systematic review. Journal of Neurologic Physical Therapy. 2016; 40(3):165-75

Nicolini A, Prato P, Beccarelli L, Grecchi B, Garuti G, Banfi P, et al. Comparison of two mechanical insufflation-exsufflation devices in patients with amyotrophic lateral sclerosis: a preliminary study. Panminerva Medica. 2022; 64 (4): 525-531

Niven E, Newton J, Foley J, Colville S, Swingler R, Chandran S et al. Validation of the Edinburgh Cognitive and Behavioural Amyotrophic Lateral Sclerosis Screen (ECAS): A cognitive tool for motor disorders. Amyotrophic Lateral Sclerosis Frontotemporal Degeneration. 2015; 16(3-4):172-9

Nguyen C, Caga J, Mahoney CJ, Kiernan MC, Huynh W. Behavioural changes predict poorer survival in amyotrophic lateral sclerosis. Brain and Cognition. 2021; 150: 105710

Pagnini F, Marconi A, Tagliaferri A, Manzoni GM, Gatto R, Fabiani V et al. Meditation training for people with amyotrophic lateral sclerosis: a randomised clinical trial. European Journal of Neurology. 2017 Apr; 24 (4): 578-586

Pagnini F, Phillips D, Haulman A, Bankert M, Simmons Z, Langer E. An online non-meditative mindfulness intervention for people with ALS and their caregivers: a randomised controlled trial. Amyotroph Lateral Sclerosis and Frontotemporal Degeneration. 2022; 23 (1-2): 116-127

Pinto-Grau M, Costello E, O'Connor S, Elamin M, Burke T, Heverin M, et al. Assessing behavioural changes in ALS: cross-validation of ALS-specific measures. Journal of Neurology. 2017;264(7):1397-1401

Pondofe K, Marcelino AA, Ribeiro TS, Torres-Castro R, Vera-Uribe R, Fregonezi GA et al. Effects of respiratory physiotherapy in patients with amyotrophic lateral sclerosis: protocol for a systematic review of randomised controlled trials. BMJ Open. 2022;12 (5): e061624

Radakovic R, Copsey H, Moore C, Mioshi E. Development of the MiND Toolkit for management of cognitive and behavioural impairment in motor neuron disease. Neurodegenerative Disease Management. 2020; 10 (1):15-25

Raglio A, Giovanazzi E, Pain D, Baiardi P, Imbriani C, Imbriani M et al. Active music therapy approach in amyotrophic lateral sclerosis: a randomised controlled trial. International Journal of Rehabilitation Research. 2016; 39 (4) 365-67

Saxon JA, Thompson JC, Harris JM, Ealing J, Hamdalla H, Chaouch A et al. The Edinburgh Cognitive and Behavioural ALS Screen (ECAS) in frontotemporal dementia. Amyotrophic Lateral Sclerosis Frontotemporal Degeneration. 2020; 21(7-8):606-613.

Simon N, Goldstein LH. Screening for cognitive and behavioural change in amyotrophic lateral sclerosis/motor neuron disease: a systematic review of validated screening methods. Amyotrophic Lateral Sclerosis and Frontotemporal Degeneration. 2019; 20 (1-2): 1-11

Tjokrowijoto P, Phillips M, Ceslis A, Henderson RD, McCombe PA, Robinson GA. Sensitivity and specificity of the ECAS in identifying executive function and social cognition deficits in MND. Amyotrophic Lateral Sclerosis Frontotemporal Degeneration. 2023; 24(5-6):466-474.

Turon-Sans J, Gascon-Bayarri J, Reñé R, Rico I, Gámez C, Paipa A et al. Cognitive impairment in ALS patients and validation of the Spanish version of the ALS-CBS test. Amyotrophic Lateral Sclerosis Frontotemporal Degeneration. 2016;17(3-4):221-7

Toussaint M, Chatwin M, Gonzales J, Berlowitz DJ. The ENMC Respiratory Therapy Consortium. 228th ENMC International Workshop: airway clearance techniques in neuromuscular disorders Naarden, The Netherlands, 3–5 March, 2017 (Report)

Van Gorenestijn AC, Schroder CD, Visser-Meily JMA, van Reenan ETK, Veldink JH, van den Berg LH. Cognitive behavioural therapy and quality of life in psychologically distressed patients with amyotrophic lateral sclerosis and their caregivers: results of a

prematurely stopped randomised controlled trial. Amyotrophic Lateral Sclerosis & Frontotemporal Degeneration. 2015; 16 (56): 309-15

Weeks KR, Gould RL, McDermott C, Lynch J, Goldstein LH, Graham CD et al. Needs and preferences for psychological interventions of people with motor neuron disease. Amyotrophic Lateral Sclerosis and Frontotemporal Degeneration. 2019; 20:7-8, 521-531

Woolley SC, Goetz R, Factor-Litvak P, Murphy J, Hupf J, Lomen-Hoerth C et al. Longitudinal screening detects cognitive stability and behavioural deterioration in ALS patients. Behavioural Neurology. 2018; 5969137-7

Woolley SC, York MK, Moore DH, Strutt AM, Murphy J, Schulz PE, Katz JS. Detecting frontotemporal dysfunction in ALS: utility of the ALS Cognitive Behavioural Screen (ALS-CBS). Amyotrophic Lateral Sclerosis. 2010 ;11(3):303-11

Xu L, He B, Zhang Y, Chen L, Fan D, Zhan S et al. Prognostic models for amyotrophic lateral sclerosis: a systematic review. Journal of Neurology. 2021; 268 (9): 3361-70

Young CA, Ealing J, McDermott C, Williams T, Al-Chalabi A, Majeed T et al. The relationships between symptoms, disability, perceived health and quality of life in amyotrophic lateral sclerosis/motor neuron disease. Amyotrophic Lateral Sclerosis and Frontotemporal Degeneration. 2019; 20 (5-6):317-327

Zarotti N, Coates E, McGeachan A, Williams I, Beever D, Hackney G, Norman P, Stavroulakis T, White D, White S, Halliday V, McDermott C; HighCALS Study Group. Health care professionals' views on psychological factors affecting nutritional behaviour in people with motor neuron disease: A thematic analysis. British Journal of Health Psychology. 2019 Nov;24(4):953-969