WELCOME!

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ALS ASSOCIATION

Evaluating Cognitive Changes in ALS

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COGNITIVE AND BEHAVIORAL IMPAIRMENT IN MND/ALS:

AN UPDATE

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Living with ALS Educational Webinar Series
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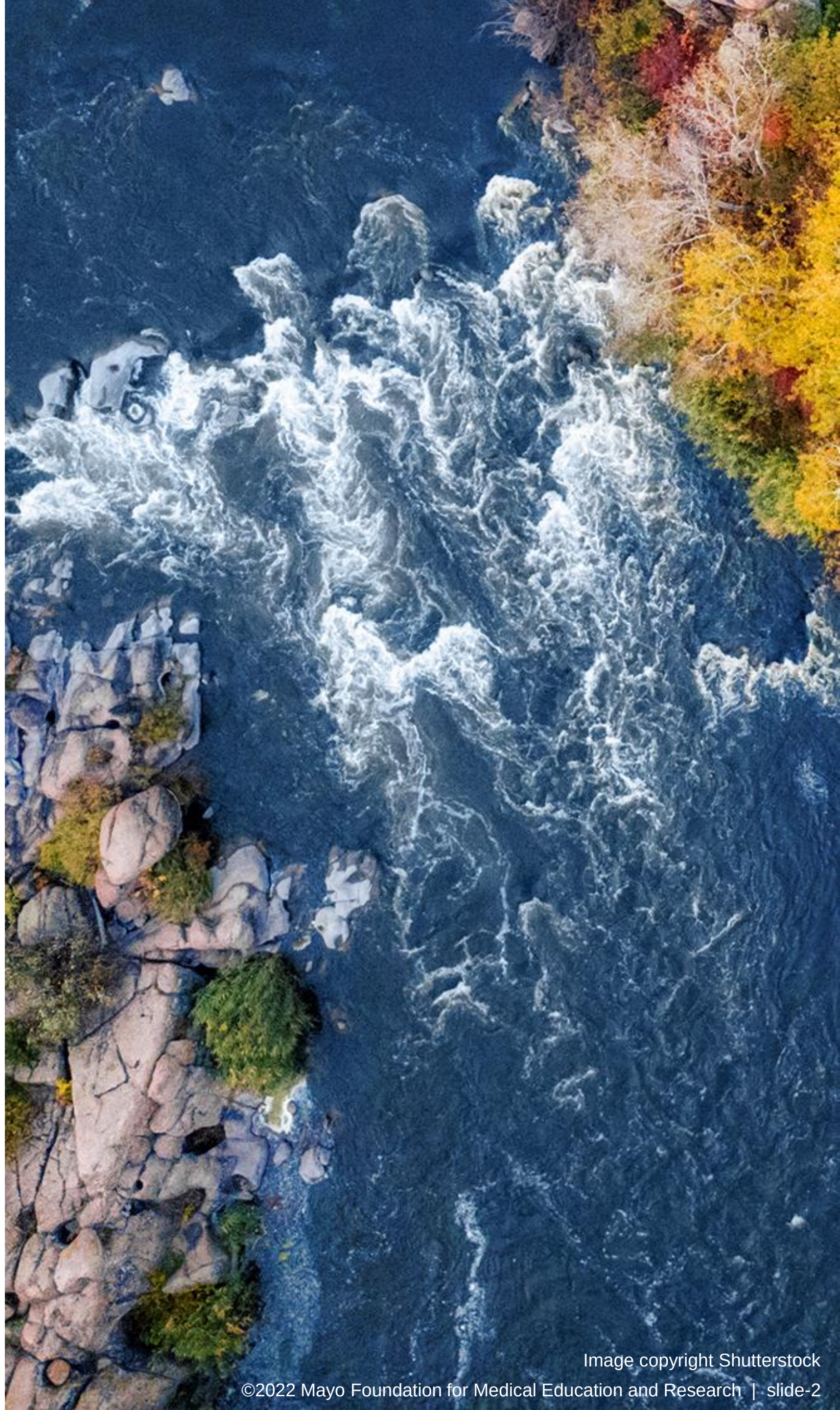

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LEARNER OBJECTIVES

- Name 2 cognitive and 2 behavioral symptoms that can be observed in MND/ALS
- Differentiate diagnostic categories of cognitive and behavioral impairment in MND/ALS
- Name at least 2 measures that are used to screen for cognitive and behavioral impairment
- Describe factors that influence the presentation of impairment
- Define the impact of cognitive and behavioral impairment on symptom management and caregiver stress

WHEN IS IT DEMENTIA?

Impairment: difference in thought/behavior but can still act for his/herself

Dementia: difference in thought/behavior

AND can no longer act for his/herself

Different diseases cause dementia

When dementia is present – family/caregivers must make decisions

2017 REVISED DIAGNOSTIC CRITERIA FOR ALS-FRONTAL TEMPORAL SPECTRUM DISORDER

Update from 2009 initial criteria

Axis I = PLS, PMA, PBP, ALS – type of MND

Axis II = pattern of cognitive or behavioral impairment

C9ORF72

most common pathogenic variant for familial ALS (60 to 70% cases) and familial FTD (18% cases)

Cognitive impairment more likely in ALS/MND that are C9+ versus C9-

2017 UPDATED DIAGNOSTIC CRITERIA

- Cognitive impairment = executive dysfunction OR language dysfunction
- 35 to 40% ALS/MND with no dementia have language impairment
- Some MND/ALS have memory impairment but isolated memory impairment thought unlikely

- Behavioral impairment = Apathy (70% ALS)

OR disinhibition, diminished sympathy/empathy, perseverative behavior, stereotyped behavior

2017 UPDATED DIAGNOSTIC CRITERIA

ALSci = below 5th percentile relative to age and education based normative data on tests of executive function (fluency or 2 other measures), OR language (sentence processing, word-finding, spelling, syntax processing, naming) OR both

//must be impaired on at least 2 measures

ALSbi = Apathy with or without behavioral change or at least 2 of the following: disinhibition, decreased sympathy/empathy, perseveration, compulsive or stereotyped behavior, hyperorality/diet change, dimimished insight, psychotic symptoms

ALScibi = Both features present but no functional activity impairment to meet criteria for dementia

ALS-FTD

SCREENING FOR COGNITIVE/BEHAVIORAL CHANGE

- Should be done at regular clinic visits using a standardized and validated measure
- ALS Cognitive Behavioral Screen (ALS-CBS)
- Edinburgh Cognitive and Behavioral ALS Screen (ECAS)

Preliminary comparison ALS-CBS and ECAS suggest the screens do not measure the same things (doi: 10.1007/s00415-021-10753-w)

SCREENING ADVANCES

Telephone-based versions of ALS-CBS, verbal fluency, and behavioral screens being developed (doi: 10.3109/21678421.2016.1173703)

//make screening and longitudinal tracking more accessible

ARE THERE FACTORS THAT INFLUENCE PRESENTATION OF IMPAIRMENT?

Cognitive reserve = **education, occupational, and physical activity data**

All CR proxies associated with executive function, verbal fluency, and memory domains (doi: 10.1080/21678421.2020.1849306)

Longitudinal evaluation of 189 patients over 16 months

1. High CR at baseline significantly predicted NPSY performance

2. Higher CR performed better through longitudinal evaluation of medium to low CR patients.

(doi: 10.1136/jnnp-2020-324992)

IMPACT OF COGNITIVE AND BEHAVIOR CHANGE

- Presence of cognitive and/or behavioral impairment may decrease survival time (doi: 10.1016/j.bandc.2021.105710)
- Caregiver burden

Impact of COVID19 lockdown on ALS patients and carers (doi: 10.1080/21678421.2021.1912772)

// followed 60 patients and 59 caregivers through a 2 month lockdown due to COVID

// qualty of life and perceived health status did not worsen during lockdown

// caregiver burden increased significantly

WHO IS AFFECTED BY THINKING/BEHAVIOR CHANGE?

People with MND/ALS

Family members of people with MND/ALS

Care providers of people with MND/ALS

WHY LOOK FOR IT?

People with ALS/MND want to know

Caregivers want to know

Family members want to know

We are still learning about ALS

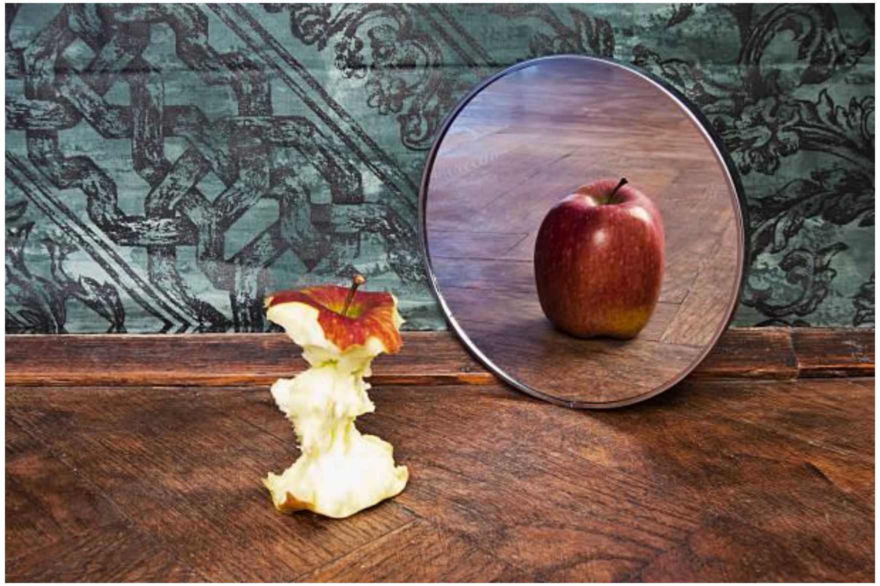
Knowing helps us learn **WHERE** to place expectations

A WORD ABOUT EXPECTATIONS

Expectations for people with ALS/MND, caregivers, care providers

Ideal for expectations to be “just right” and reflective of reality.

Challenges emerge when expectations do not reflect reality



HELPING THE PERSON WITH ALS/MND

Lack of awareness, anticipation, presence of agitation, apathy/indifference, withdrawal, frustration

- AAC (expression)
- Counseling to address anticipatory fear if FMH+ dementia/MND
- Functional communication boards (picture pointing)
- Simplify communication (two word phrases – noun verb, use nonverbals)
- Use of routine and schedule (less demand on memory)
- Audio books (when cannot hold books)
- Limit unfamiliar people/unfamiliar settings
- Physically “flank” person with disinhibition
- Distraction (touch, something the person loves)
- Simple cognitive task (i.e., counting) – counteract PBA
- Acceptance of change (patient’s adjustment is a reality)
- Medication (severe agitation, PBA)
- **Set realistic expectations**

AWARENESS PREDICTS INTERVENTION WITH PATIENT

Critical to evaluate awareness in the person with ALS/MND

Present awareness: Advanced Directives, care decisions, making intentions known, directing team

Absent awareness: Expectations for the person with ALS need to match his/her current ability

Interventions must be directed at family/care team when person with ALS does not have awareness or has restricted awareness.

HELPING FAMILY MEMBERS

Help is needed when there is:

Irritation, resentment, exhaustion, anxiety, physical problems, grief

Anger and anxiety = Responses to threat

Life with ALS is threatening

Life with ALS is an imposition

Life with ALS requires change that may not be wanted

HELPING FAMILY MEMBERS

What does help look like?

- Neuropsychological assessment or cognitive screening of patient
- Durable medical equipment
- Communication devices
- Behavioral management
- ALS Clinic participation
- Education about ALS/MND and different stages of disease
- In-home help (housekeeper >>>>>>>>> home health aide)
- Grief counseling
- Counseling to address anticipatory fear if FMH+ dementia/MND
- Respite time away from the person with ALS
- Giving up perfectionism
- Working with other family members
- Do not wait for a crisis – Advanced Directives, Legal Issues
- Educate providers working with the person with ALS
- Self-care (we are not done here ☺)
- **Set realistic expectations for the person with ALS and YOURSELF**

SELF-CARE FOR CAREGIVERS

You cannot provide care to others when you have nothing left to give.

- Regularly schedule time for yourself
- Tell others specifically what you need
- Schedule and attend YOUR appointments
- Do not aggravate old injuries
- You have feelings!!!! Be honest.
- Faith counseling
- When you feel anger & frustration, it’s a sign!!!
- When you feel avoidant or despondent, it’s a sign!!!

COMMON QUESTIONS FROM FAMILY

Anxiety over making decisions with or for the person with ALS/MND

1. How do I know that’s it is not just psychological?
2. How much do I let her/him do alone vs. assist?
3. When do you invoke advanced directives?

HELPING THE PROVIDER OF A PERSON WITH ALS

- Education (vigilance)
- Choose providers that know the disease and symptoms
- Do not assume all ALS providers know about thinking and behavior change in ALS.
- Share your concerns about yourself and your family
- Make sure your wishes are known
- Do not wait for a crisis to address advance directives.
- Remind providers to recognize “signs” that his/her expectations of the person with ALS or family members may need to change or may warrant additional diagnostic features of ALS presentation

A MANTRA

Caregiving challenges offer opportunity to center/modify expectations.

REFERENCES

Strong MJ, Abrahams S, Goldstein LH, Woolley S, Mclaughlin P, Snowden J, Mioshi E, Roberts-South A, Benatar M, HortobáGyi T, Rosenfeld J, Silani V, Ince PG, Turner MR. Amyotrophic lateral sclerosis - frontotemporal spectrum disorder (ALS-FTSD): Revised diagnostic criteria. Amyotroph Lateral Scler Frontotemporal Degener. 2017 May;18(3-4):153-174. doi: 10.1080/21678421.2016.1267768.

Greco LC, Lizio A, Casiraghi J, Sansone VA, Tremolizzo L, Riva N, Solca F, Torre S, Ticozzi N, Filippi M, Silani V, Poletti B, Lunetta C. A preliminary comparison between ECAS and ALS-CBS in classifying cognitive-behavioural phenotypes in a cohort of non-demented amyotrophic lateral sclerosis patients. J Neurol. 2022 Apr;269(4):1899-1904. doi: 10.1007/s00415-021-10753-w.

Christodoulou G, Gennings C, Hupf J, Factor-Litvak P, Murphy J, Goetz RR, Mitsumoto H. Telephone based cognitive-behavioral screening for frontotemporal changes in patients with amyotrophic lateral sclerosis (ALS). Amyotroph Lateral Scler Frontotemporal Degener. 2016 Oct-Nov;17(7-8):482-488. doi: 10.3109/21678421.2016.1173703.

Consonni M, Dalla Bella E, Bersano E, Telesca A, Lauria G. Cognitive reserve is associated with altered clinical expression in amyotrophic lateral sclerosis. Amyotroph Lateral Scler Frontotemporal Degener. 2021 May;22(3-4):237-247. doi: 10.1080/21678421.2020.1849306.

Costello E, Rooney J, Pinto-Grau M, Burke T, Elamin M, Bede P, McMackin R, Dukic S, Vajda A, Heverin M, Hardiman O, Pender N. Cognitive reserve in amyotrophic lateral sclerosis (ALS): a population-based longitudinal study. J Neurol Neurosurg Psychiatry. 2021 May;92(5):460-465. doi: 10.1136/jnnp-2020-324992.

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

Giusiano S, Peotta L, Iazzolino B, Mastro E, Arcari M, Palumbo F, Torrieri MC, Bombaci A, Grassano M, Cabras S, Di Pede F, DeMattei F, Matteoni E, Solero L, Daviddi M, Salamone P, Fuda G, Manera U, Canosa A, Chiò A, Calvo A, Moglia C, Vasta R. Amyotrophic lateral sclerosis caregiver burden and patients' quality of life during COVID-19 pandemic. Amyotroph Lateral Scler Frontotemporal Degener. 2022 Feb;23(1-2):146-148. doi: 10.1080/21678421.2021.1912772.

QUESTIONS & DISCUSSION