COGNITIVE AND BEHAVIOUR CHANGES IN ALS

What Professionals Need to Know

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ALS is a Multisystem Disease

(Hudson 1981)

MND - a multisystem disease

**Clinical**
- dementia
- parkinsonism
- cerebellar signs
- autonomic dysfunction
- sensory abnormalities

**Pathology**
- extramotor neocortex
- striatum
- globus pallidus
- thalamus
- subthalamus
- sub. nigra

Hudson 1981
COGNITIVE IMPAIRMENT AND ALS
How Did We Miss These Changes?
What we always knew.... but couldn’t always see....

- Frontotemporal dementias can develop anterior horn cell degeneration

- Dementia can occur with “classical ALS”

- Occasional descriptions of “childish and credulous behaviour” in some ALS patients (Pierre Marie 1853-1940)
ALS DIAGNOSTIC CRITERIA
El Escorial Diagnostic Criteria

- **Definite ALS:** UMN and LMN signs in three regions.

- **Probable ALS:** UMN & LMN signs in at least two regions with UMN signs rostral to (above) LMN signs.

- **Possible ALS:** UMN & LMN signs in one region, UMN signs alone in two or more regions, or LMN signs above UMN signs.

- **Suspected ALS:** LMN signs only in two or more regions.
EL ESCORIAL CRITERIA IGNORE COGNITIVE STATUS
COGNITIVE /BEHAVIOURAL SYMPTOMS ARE FREQUENTLY NOT RECOGNIZED OR APPRECIATED BY FAMILIES AND MAY BE MISSED BY REFERRING SERVICES
Variants of FTD

- Behavioural Variant
  - Dysexecutive, Apathy, Disinhibition
- Semantic dementia
- Progressive Non-Fluent Aphasia
  - Progressive Non-Fluent Grammatical
  - Progressive Non-Fluent Logopoenic

COMMUNICATING WHAT WE NOW KNOW
Neuropsychological Battery

Executive function
- Verbal fluency (phonological and category)
- Brixton Test
- Stroop Interference Test
- Digit span

Visuo-construction
- Rey Complex Figure (copy)

Language
- Boston Naming Test

Memory
- Logical memory (WMS-III)
- California Verbal Learning Test
- Paired Associate Learning Test (WMS-III)
- Rey Complex Figure Test (immediate and delayed)
CLASSIFICATION OF IMPAIRMENT

- Normal
- ALS with cognitive impairment
  - Executive impairment
    - Single domain
    - Multidomain
  - Non-executive impairment
    - Single domain
    - Multidomain
- ALS with FTD
The syndrome of cognitive impairment in amyotrophic lateral sclerosis: a population-based study

Julie Phukan,1 Marwa Elamin,1 Peter Bede,1 Norah Jordan,2 Laura Gallagher,2 Susan Byrne,1 Catherine Lynch,1 Niall Pender,2 Orla Hardiman1,3

Diagram:
- No Abnormality detected: 49%
- Co-morbid Alzheimer's disease: 1%
- ALS-FTD: 13%
- Executive Dysfunction: 25%
- Non-Executive Cognitive Impairment: 12%
Cognitive Dysfunction in ALS

- No Abnormality detected 49%
- Co-morbid Alzheimer's disease 1%
- ALS-FTD 13%
- Executive Dysfunction 25%
- Non-Executive Cognitive Impairment 12%
- Disinhibition*
- Planning/Reasoning*
- Language Disorder

- Neuropsychological Assessment, and AHP Screening
COGNITIVE IMPAIRMENT IN ALS: UNANSWERED QUESTIONS

Cognitive changes predict functional decline in ALS
A population-based longitudinal study

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Catherine Lynch, MSc
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ABSTRACT

Objective: To determine whether cognitive status in patients with amyotrophic lateral sclerosis (ALS) is a useful predictor of attrition and motor and cognitive decline.

Methods: Cognitive testing was undertaken in a large population-based cohort of incident ALS patients using a longitudinal, case-control study design. Normative data for neuropsychological tests were generated using age-, sex-, and education-matched healthy controls who also underwent repeated assessments. Data were analyzed to generate models for progression/spread.

Results: One hundred eighty-six patients with ALS who had no evidence of C9orf72 hexanucleotide repeat expansion were enrolled. A second and third assessment were undertaken in 98 and 46 of the patients with ALS, respectively. Executive impairment at the initial visit was associated with significantly higher rates of attrition due to disability or death and faster rates of motor functional decline, particularly decline in bulbar function. Decline in cognitive function was faster in patients who were cognitively impaired at baseline. Normal cognition at baseline was associated with tendency to remain cognitively intact, and with slower motor and cognitive progression.

Conclusions: Non-C9orf72-associated ALS is characterized by nonoverlapping cognitive subgroups with different disease trajectories. These findings have important implications for models of ALS pathogenesis, and for future clinical trial design. Neurology 2013;80:1590-1597
Longitudinal Study of Cognition in ALS

![Bar chart showing ALS patients and HC groups at different time points (T1, T2, T3, T4).]
Percentage of ALS Patients with No Cognitive Impairment

- T1: 52% (205 patients)
- T2: 62% (136 patients)
- T3: 86% (103 patients)
- T4: 86% (24 patients)

Legend:
- ALS Patients
- HC
Cognitive Dysfunction in ALS

- The most common cognitive domain affected in ALS is **Executive Function**, implicating *attention/planning/regulation*.

- **Language** can also be impaired in ALS, especially in the form of *word-finding difficulties*, and difficulties with *comprehension*.

- Patients may present with **Memory** difficulties, which may caused by *poor encoding*, or by finding it *linguistically difficult to process* the information.
WHAT DO WE KNOW ABOUT BEHAVIOUR?
Clinical Features of fvFTD

- Violation of interpersonal space
- Increased interest in sex
- Disinterest, withdrawal
- Lack of empathy
- Hoarding
- Over-eating and food fads
- Lack of judgement
- Impulsive buying
- Ignoring social etiquette
- Blunted emotions
- Swearing
- Utilisation behaviour
- New onset criminal behaviour
- Change in personal hygiene
- Repetitive behaviour
ALS AND BEHAVIOUR

- Variable reported rates (25-50%)

- Most common change: Apathy (up to 80%)

- May also be a function of physical disability
BEAUMONT BEHAVIOURAL QUESTIONNAIRE (V2)

Code for Patient __________________________ Informant’s relationship to Patient __________________________ Date --/--/-----

- Your view is very important so please read instructions carefully.
- We would like to ask you a number of questions about changes in behaviour that you may have noticed in the person
  (1) in last 10 years up to start of the motor neuron disease (MND)
  (2) since the start of the symptoms of the motor neuron disease (MND)
  *In each case use a tick (√) to indicate your choice
- If the new behaviour described has been present, then please rate the change as mild, moderate or severe depending on how it has affected your life.
- If the person does not have this behaviour OR has always behaved this way, then select “No/No Change”.

<table>
<thead>
<tr>
<th></th>
<th>IN THE LAST 10 YEARS</th>
<th>SINCE ONSET OF MND</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Has become more irritable than before</td>
<td>No/No change □</td>
</tr>
<tr>
<td></td>
<td>Yes: Mild □</td>
<td>Moderate □</td>
</tr>
<tr>
<td>2</td>
<td>Is much less aware of painful sensations such as hot things, sharp objects etc.</td>
<td>No/No change □</td>
</tr>
<tr>
<td></td>
<td>Yes: Mild □</td>
<td>Moderate □</td>
</tr>
<tr>
<td>3</td>
<td>When talking, often makes more grammatical mistakes than before</td>
<td>No/No change □</td>
</tr>
<tr>
<td></td>
<td>Yes: Mild □</td>
<td>Moderate □</td>
</tr>
<tr>
<td>4</td>
<td>Is generally not as aware of making mistakes as he/she used to be</td>
<td>No/No change □</td>
</tr>
<tr>
<td></td>
<td>Yes: Mild □</td>
<td>Moderate □</td>
</tr>
<tr>
<td>5</td>
<td>Is less able to react to difficulties, plan or foresee problems</td>
<td>No/No change □</td>
</tr>
<tr>
<td></td>
<td>Yes: Mild □</td>
<td>Moderate □</td>
</tr>
<tr>
<td>6</td>
<td>If has an idea to do something, he/she has to do it immediately, often without thinking it through</td>
<td>No/No change □</td>
</tr>
<tr>
<td></td>
<td>Yes: Mild □</td>
<td>Moderate □</td>
</tr>
<tr>
<td>7</td>
<td>Shows much more emotion than before, cries or laughs too easily</td>
<td>No/No change □</td>
</tr>
<tr>
<td></td>
<td>Yes: Mild □</td>
<td>Moderate □</td>
</tr>
</tbody>
</table>
## Behaviour Classifications

<table>
<thead>
<tr>
<th>Factor Loading</th>
<th>Superordinate Classification of Dysfunction</th>
<th>Cognitive/Behavioural Dysfunction</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Initiation (Apathy)</td>
<td>Loss of interest; inability to plan; impulsiveness; decreased sex drive; lack of appropriate embarrassment.</td>
</tr>
<tr>
<td>2</td>
<td>Adherence to social norms</td>
<td>Emotional changes; social disinhibition; social seeking.</td>
</tr>
<tr>
<td>3</td>
<td>Social Engagement</td>
<td>Social withdrawal; distractibility; cognitive rigidity.</td>
</tr>
<tr>
<td>4</td>
<td>Interpersonal Engagement</td>
<td>Aggressiveness; irritability; Increased lability; hypersensitivity to stimuli.</td>
</tr>
<tr>
<td>5</td>
<td>Self-regulation</td>
<td>Reduced concern for hygiene; change in food preferences; new onset repetitious/obsessive behaviour.</td>
</tr>
</tbody>
</table>
Factor Analysis

- Social disinhibition
- Hyper-orality
- Preserveration

- Mental rigidity
- Obsessive-compulsive behaviour
- Aggression
- Language Difficulties

- Apathetic Behaviour
- Repetitive behaviour
- Social withdrawal
- Altered Sensory Perception
All ALS Patients (n=151)

- No Behavioural Impairment: 57.80%
- Mild Behavioural Impairment: 23.20%
- Severe Behavioural Impairment: 19.00%
COMMON BEHAVIOURAL CHANGES

- Inability to plan & to foresee and react to Problems
- Lack of embarrassment
- Impulsiveness
- Grammar Mistakes
- Distractibility
- Mental rigidity
- Same subjects of conversation
- Self-centred behaviour
- Aggressiveness
SEVERE CHANGES

Group 3

- Lack of Insight
- Naming problems
- Obsessive Object/furniture arranging
- Social seeking
- Crams/stores food in mouth
- Social disinhibition
- Increased Libido
Bulbar vs Spinal Onset Disease

Bulbar-Onset
- 54%
  - Behaviourally Impaired
  - No Behavioural Impairment

Spinal Onset
- 36%
  - Behaviourally Impaired
  - No Behavioural Impairment

Bar graph showing:
- Emotional Lability: Bulbar-Onset > Spinal Onset
- Social Withdrawal: Bulbar-Onset > Spinal Onset
Severe Physical Disability

Severe Disability

- Behaviourally Impaired: 64.70%
- No Behavioural Impairment: 35.30%

Mild-Moderate

- Behaviourally Impaired: 40.50%
- No Behavioural Impairment: 59.50%

Bar charts:
- Loss of interest in Previous Hobbies/Interests
  - Severe Disability: 70%
  - Mild-Moderate: 40%
- Repetitive Behaviour
  - Severe Disability: 60%
  - Mild-Moderate: 20%
Summary of Behavioural Dysfunction in ALS

- The most prominent behavioural symptom reported in ALS is **Apathy**, which is defined as loss of motivation, initiative or interest.

- **Disinhibition** can manifest as loss of manners or a new onset of socially inappropriate behaviour.

- Patients can also present with a *reduced awareness or insight* of their deficits and symptoms. This can be a broad lack of insight and reflect poor judgment. *(Dysexecutive Behaviours)*
CLINICAL IMPLICATIONS OF COGNITIVE AND BEHAVIOURAL CHANGE
IS COGNITIVE IMPAIRMENT IMPORTANT?

- Survival
- Decisions: finance, end-of-life decisions
- Compliance with NIV, RIG, multidisciplinary care
- Safety awareness e.g. Fall avoidance, coping with choking episodes
- Critical to our understanding of the biology of ALS
Executive Function predicts Survival

- Executive dysfunction: 27 mths (95% CI 14.2 - 39.7)
- No executive dysfunction: 48 mths (95% CI 30.2 - 65.7)

\[ p < 0.0001 \quad HR \ 3.9 \ (95\% \ CI \ 2.0 \ to \ 7.9) \]
BEHAVIOURAL IMPAIRMENT ASSOCIATED WITH INCREASED CAREGIVER BURDEN
Caregiver burden in amyotrophic lateral sclerosis: a cross-sectional investigation of predictors

Tom Burke\textsuperscript{1,2} · Marwa Elamin\textsuperscript{2} · Miriam Galvin\textsuperscript{2,3} · Orla Hardiman\textsuperscript{2,4} · Niall Pender\textsuperscript{1,2,5}
PREDICTORS OF BURDEN

PATIENT FACTORS
- Apathy
- Disinhibition
- Dysexecutive behaviour

CAREGIVER FACTORS
- Depression
- Anxiety
- Pre-morbid problem solving abilities
WHAT SHOULD WE DO AS HEALTHCARE PROFESSIONALS?
Best Practice Guidelines

Healthcare Professionals

Prevalence
- Cognitive
- Behavioural

Common Phenotypes

Detecting Impairment

Implications
- Caregiver Burden

Management Strategies
DETECTING COGNITIVE AND BEHAVIOURAL CHANGES IN ALS

Edinburgh Cognitive and Behavioural ALS Screen
Screening for Cognitive Impairment: 15mins

**ECAS:** An ALS specific tool designed for
to control for motor impairment.

<table>
<thead>
<tr>
<th>LANGUAGE - Naming</th>
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<tbody>
<tr>
<td><strong>Ask:</strong> Say or write down the names of these pictures:</td>
</tr>
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<table>
<thead>
<tr>
<th>Score</th>
<th>0-8</th>
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<table>
<thead>
<tr>
<th>LANGUAGE - Comprehension</th>
</tr>
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<tbody>
<tr>
<td><strong>Ask:</strong> point to the one which is:</td>
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</table>

1. Something you can fly in
2. Something with webbed feet
3. An animal that climbs trees
4. Something used for chopping
5. A means of transport
6. Something with a sharp edge
7. Something with a sting

**Specific**
- Language
- Executive Function
- Lexical Fluency

**Non-specific**
- Memory
- Visual Processing

**Total**
Assessing Behavioural Dysfunction

Caregiver Behavioural Interview

Key Behavioural Features, including hallucinations:

Disinhibition
Apathy
Dysexecutive Syndrome
Loss of Empathy

<table>
<thead>
<tr>
<th>A. Behavioural disinhibition</th>
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<tbody>
<tr>
<td>1. Socially inappropriate behaviour, e.g. inappropriate behaviour with strangers criminal behaviour</td>
</tr>
<tr>
<td>2. Loss of manners or decorum, e.g. crude or sexually explicit remarks, jokes or opinions that may be offensive to others lack of response to social cues</td>
</tr>
<tr>
<td>3. Impulsive, rash or careless actions, e.g. new onset gambling, or buying or selling property without regard for consequences giving out personal information inappropriately, e.g. credit card numbers</td>
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<tr>
<th>B. Apathy or inertia</th>
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<tr>
<td>4. Loss of interest, drive or motivation, e.g. passivity and lack of spontaneity needs prompting to initiate or continue routine activities</td>
</tr>
<tr>
<td>5. Diminished response to other people’s needs and feelings Positive rating on this feature should be based on specific examples that reflect a lack of understanding or indifference to other people’s feelings, e.g. hurtful comments disregard for others’ pain or distress</td>
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<tr>
<th>C. Loss of sympathy or empathy</th>
</tr>
</thead>
<tbody>
<tr>
<td>6. Diminished social interest, interrelatedness, personal warmth or general closeness in social engagement, e.g. coldness lack of eye contact</td>
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<thead>
<tr>
<th>D. Perseverative, stereotyped, compulsive or ritualistic behaviour</th>
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<tr>
<td>7. Simple repetitive movements, e.g. tapping, clapping scratching, picking skin or clothing repeating words</td>
</tr>
<tr>
<td>8. Complex, compulsive or ritualistic behaviours, e.g. counting, cleaning rituals, checking collecting, hoarding</td>
</tr>
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</table>
ECAS IS A SCREENING TOOL AND REQUIRES TRAINING!

AN “ABNORMAL” ECAS SHOULD TRIGGER A FULL NEUROPSYCHOLOGICAL ASSESSMENT
**Implicated Network**

- **Insula-Orbito-Frontal Circuit**
- **Dorso-lateral**
- **Anterior-Cingulate-Limbic**
- **via Basal Ganglia input/output**
- **Temporal-Limbic/Insular**

**Precipitating Factors**

Processes that serve to impede difficulties:
- Perseveration; apathy; lack of awareness of social norms; hyper-orality; lack of insight; impulsivity.
- Reduced display of emotion; increased please seeking behaviour; increased smoking/alcohol consumption.
- Increased irritability; aggressiveness; altered sensitivity to stimuli; increased repetitive behaviour i.e., tapping etc.

**Predisposing Factors**

- Premorbid personality and function; Systemic factors (family); Social support; Cognitive Reserve; Resilience

**Maintaining Factors**

Self-regulatory skills; dysfunctional attribution style; cognitive distortions; dysfunctional coping strategies; immature defence mechanism.

**Presenting Behaviour**

Psychiatric Dx.; Neurologic Dx.; Distress i.e., denial/low self-esteem; C9orf72 +ive; Genetic vulnerability; Progression of disease; ALSFTD.

**Protective Factors**

Functional gain for behaviour. Undesirable reinforcement.

**Social/Interpersonal Behaviour**

**Functional gain for behaviour. Undesirable reinforcement.**

**Professional Support**

Psychotherapy
- Attention strategies
- Non/Pharmacotherapy
- Managing Mental Fatigue
- Self-regulation awareness
- Coping skills and resilience building

**Coping skills and resilience building**

**Emotional**
**Behavioural**
**Physiological**

**(1) Event**

**(2) Reaction**

(2b) Caregiver response

**(3) Outcome**

Potentially contributing to further events/increased caregiver burden
CLINICAL SCENARIOS
Colleagues have observed that John is much more rigid, unwilling to listen to other’s opinions. John forgets about important meetings and he needs his secretary to organize and supervise his diary.

Notices that when he is at a meeting, he is not as fluent with his speech. Recently, he decided upon an investment which turned out to be detrimental for the company.

- Individualized task steps help patients avoid common hazards at home and to continue independent activities (e.g. cooking). Checklists can also open discourse with family, allowing idea sharing.

- Encourage the patient to use calendars, memory aids or phone alarms to help him remember appointments or recall to take the medication.

- Simplify communication to enhance patient’s comprehension: speak clearly using a simple and straightforward language. Break sentences into short phrases. Avoid providing too much information, and slow down when speaking.

- Families may need to adjust previously held roles for financial activities and health care planning. The patient may not be able to hold a position as the primary decision maker in the family, but may still participate in less demanding ways.
First symptoms stopped Mary playing Volleyball. Continued to support her team and maintained a huge interest.

Mary has **progressively lost interest** in her team, and spectating the sport. She is no longer motivated to converse about the sport, although she listens. Mary is not reporting sadness, or hopelessness, yet has **disengaged with her family**.

- Give the patient enough time to make decisions and offer limited choices and closed ended questions.

- Provide small amounts of information regularly rather than overload them with too much detail or content. The “little and often” rule applies.

- Self-administered checklists and individualized task analyses as prompts

- Participating in activities that are passive, can still be pleasurable, and performed as a group.

**Mary**

39 year old lady
ALS 3 year ago

Athlete
Volleyball Enthusiast
Mild motor disability with evident inappropriate behaviour. Wife notices impulsivity through wreckless spending. His driving has become careless, and he refuses to bathe.

He may interrupt conversation, make inappropriate jokes, sexual comments, or curse. Patrick is unaware of boundaries.

- Potentially dangerous activities such as driving and eating may require direct supervision.
- Patients with FTD may exhibit difficulties in swallowing associated with executive impairment rather than true dysphagia.
- Self-care and hygiene may also need to be supervised.
- In social settings, aim to sit in an area where environmental stimulus won’t be distracting i.e., aim to sit in a quiet area in a restaurant.
- Give a clear structure to the patient’s day and make it as predictable as possible.

**Patrick**

72 year old male
ALS-FTD 6 months ago

Retired
Katherine’s ability to swallow started to deteriorate. She has to avoid certain foods which may cause her to choke.

Recently, she developed a **new food preference** for chocolate with small nuts, despite having a severe choking episode. Katherine **does not seem to be aware** of the risks and is **unwilling to accept advice**.

- Supervise eating, especially in the case of patients with swallowing difficulties.

- When dealing with impulsive patients, items that may be a risk of harm for the patient need to be monitored, including food.

- If food cravings or hyperorality are present, portions need to be limited and cupboards or refrigerators may need to be locked. It is recommendable to keep safe foods visible and handy, and remove the less suitable ones from the patient’s access.

- Environmental modifications can be employed and may include having 1:1 supervision for the patient.

**Katherine**

49 year old female

ALS 18 months ago

Bulbar onset
Margaret is wheelchair-bound and totally dependent for self-care activities. Margaret’s husband is her full time caregiver.

Margaret has become very detached and distant, and never shows her feelings. Her friends stopped visiting her as she seemed indifferent to them and never showed any interest.

- When lack of empathy is present in a patient, the primary intervention is education and support to the family about the nature of FTD brain changes.

- It is important to help prevent the patient and caregiver from becoming socially isolated, by encouraging involvement and participation of understanding friends and family members.

- Make sure objects they desire are within reach so that patients will not feel compelled to get up on their own, finding an acceptable method for the patient to summon help when needed, and adherence to a predictable schedule.

Margaret

67 year old lady

ALS 4 years ago
Wheelchair bound
James was always described by his family and friends as a man with a temper, although for the last couple of years his family have noticed mood swings and bursts of anger with no apparent reason.

It is at dinner time when James’s tends to lose his temper, and he may be verbally aggressive to his wife and son.

- In patients with increased irritability, look for triggers or predictors that may prompt bursts of anger (tiredness, hunger, etc.) and try to prevent them.

- In cases of anger outburst, remain calm and avoid arguments, acknowledging the patient’s irritability.

- Avoid surprises that may create confusion or agitation and keep the environment calm and controlled.

- Eliminate environmental stimuli that may be annoying for the patient such as loud noise, inadequate temperatures, etc.

- If visitors are expected, make sure that the patient is aware of this.

- If large gatherings provoke agitation or irritability in the patient, avoid them.
UNDER CONSTRUCTION
RECOMMENDATIONS

- Cognitive and behavioural changes are important aspects of ALS - this should be recognized
- Recognize that behavioural change is of greater relevance to caregivers
- Recognize that impairment is social cognition is an important and under-recognized behavioural feature
- It is acceptable for healthcare professionals to recognize and help to address behavioural changes, and to recognize the stress that this places on the patient/HCP relationship
RECOMMENDATIONS

- Identify caregivers in need of early, targeted interventions.
- Set realistic expectations for caregivers.
- Recognise that behavioural impairment can affect interpersonal and social relationships, reducing closeness, communication, and shared viewpoints.
RECOMMENDATIONS

- Caregivers should be aware that patients may change their behaviour over time.
- Positive aspect of the caregiver should be emphasised.
- Personality characteristics and problem solving abilities of caregivers are important determinants of burden.
COMMUNICATING WITH CAREGIVERS
Behavioural Changes in MND
What Families Need to Know
WORKSHOPS AND FOCUS GROUPS

CAREGIVERS DO NOT CONSIDER IN THE PRESENCE OF COGNITIVE CHANGES TO BE IMPORTANT OR RELEVANT
Thinking Changes

• Is more forgetful

• Can’t concentrate

• Has difficulty planning and organizing

• Has difficulty solving problems

• Can’t find the words
Inappropriate Behaviour

- Becomes awkward
- Has difficulty with impulse control
- Is much more rigid
- Shows inappropriate social behaviour and does not understand social cues
- Is easily irritated, angry or has temper outbursts
- Is not aware of having any problems
Social Cognition

▶ Is not able to understand the needs and feelings of others

“He does not seem to understand how demands of the illness affect me”

▶ Control

“I have to be there all the time as he refuses other carers”

▶ Lack of acknowledgement

“I never feel appreciated for everything I do for him”
Role and obligation / Lack of freedom

“It’s a constant call on your person, having to be around, having to be available”

“I don’t have much time to myself anymore...”

“I can’t be away from the house for too long and I have my phone always...,

you are tied down”

“You are just constantly on call all the time, you can’t relax”
Guilt

“Making sure he's okay and you're not doing anything wrong, and the worry about it”

“Feeling that you can never quite do enough”

“I am not sure if I am doing the right things…”

“I feel guilty that I can't say no to one of his requests”
Identity and Impact on Relationships

“It's very difficult if you're caring for your husband or wife because the line is so blurred; it's very difficult to pull the husband-wife emotional relationship back”

“The role of a caregiver takes over, then wife. You forget you're not his minder, you're his wife, you're a woman you miss things like holding hands, cuddling. There's a loss of identity”

“Trying to get dad to do the jobs he's supposed to do... It's difficult having to give out to dad, it makes me annoyed”
Emotional Impact

“A sense of frustration, and maybe even anger”

“Not having peace of mind that there's someone else to help”

“I have to motivate myself, push myself to do this”

“He gets cranky and frustrated sometimes so that makes me feel bad” “I find that hard to reconcile: how can I love him when he drives me crazy?”
SUMMARY

- COGNITIVE AND BEHAVIOURAL IMPAIRMENT ARE INTRINSIC ASPECTS OF ALS IN SOME PATIENTS

- 50% OF ALS PATIENTS REMAIN COGNITIVEY AND BEHAVIOURALLY INTACT

- OF THOSE WITH IMPAIRMENT, EARLY RECOGNITION IS IMPORTANT-
  - SCREENING TOOLS ARE HELPFUL BUT MUST BE INTERPRETED WITH CAUTION

- BEHAVIOURAL CHANGE CORRELATES WITH CAREGIVER BURDEN
Team

Clinical
Bernie Corr
Dr.Ger Foley
Dr.Sinéad Maguire
Dr.Deirdre Murray
Lesley Doyle
Kitty McElligott

Imaging & EEG
Dr.Peter Bede
Dr.Ed Lalor
Dr.Bahman Nasseroleslami
Dr.Parames Iyer
Christina Schuster
Kieran Mohr
Michael Broderick

Neuropsychology
Dr.Niall Pender
Dr.Marwa Elamin
Tom Burke
Marta Pinto
Katy Lonnergan

Epidemiology
Dr. James Rooney
Mark Heverin (Register)

Genetics
Prof Dan Bradley
Dr.Russell McLaughlin
Dr. Kevin Kenna

Health Services
Prof.Anthony Staines
Prof.Charles Normand
Dr. Miriam Galvin
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