Management of behavioural symptoms in the MNDFTD spectrum
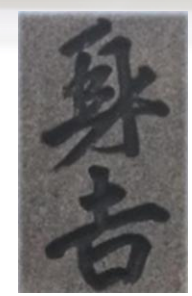
Eneida Mioshi

Professor of Dementia Care Research
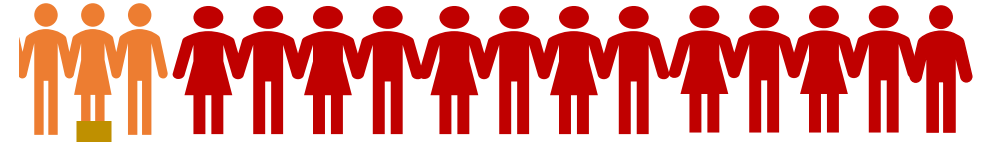
University of East Anglia

田川後藤寺駅
TAGAWAGOTOJI STATION

身吉

Norwich, UK

United Kingdom

Newcastle upon Tyne
Durham

Leeds

Blackpool   Great Britain

Manchester

Liverpool

Sheffield

Birmingham

Cambridge

ENGLAND

Oxford

London

Bristol

Cardiff

Exeter

Bournemouth

Gron

Amsterdam

Rotterdam   Netherlands

Antwerp

Brussels

Lille

Liège

35%

Mild to severe FTD-like symptoms

15% ALSFTD

?

50%
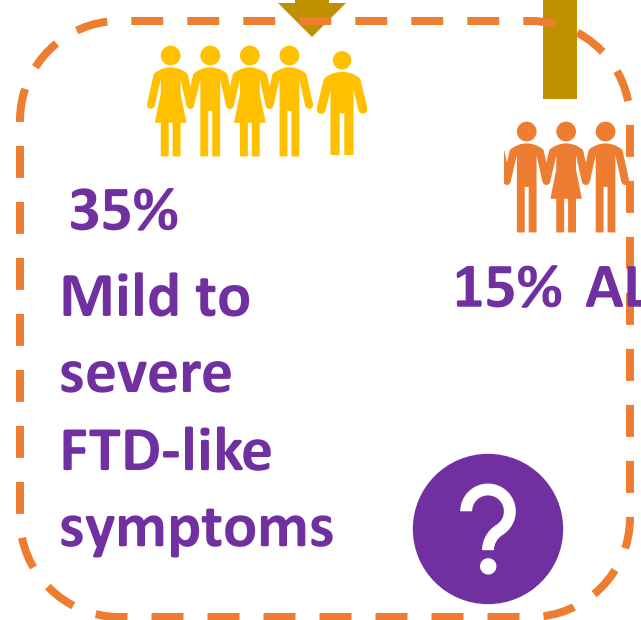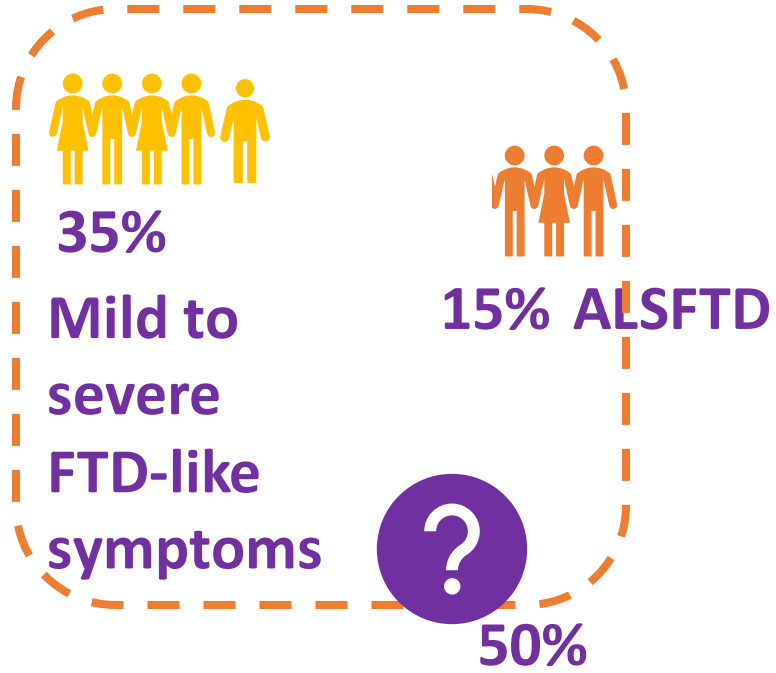
Apathy

"But yes, regards going out, he doesn't want to go out. He won't even go, you know, I'm sitting here listening to the rain, but you know, if we have a nice day, he won't go out in the car for a drive just, you know, for pleasure. He just sits inside all day and listens to the radio or listens to his iPad or, but he won't go out.

People come to visit us which is nice but obviously exhausting for him because he has to now type everything. But I would say, yes, he has no interest in much at all outside the house" *Carer 09*

35% **Mild to severe FTD-like symptoms**

15% ALSFTD

?

50%

"This was quite a while before he was diagnosed [with PLS]. He started having an obsession that we had bedbugs in the house and that he had been bitten. Anyway, I spoke to his GP about it and I'll keep my voice down a little bit, and the GP arranged to come up and see him. They had given him some [name of cream] which takes away the itching and the sensation that he's got, bugs crawling on him, and that seems to have done the trick" *Carer 02*

Hallucination

35%

Mild to severe FTD-like symptoms

15%

?

50%
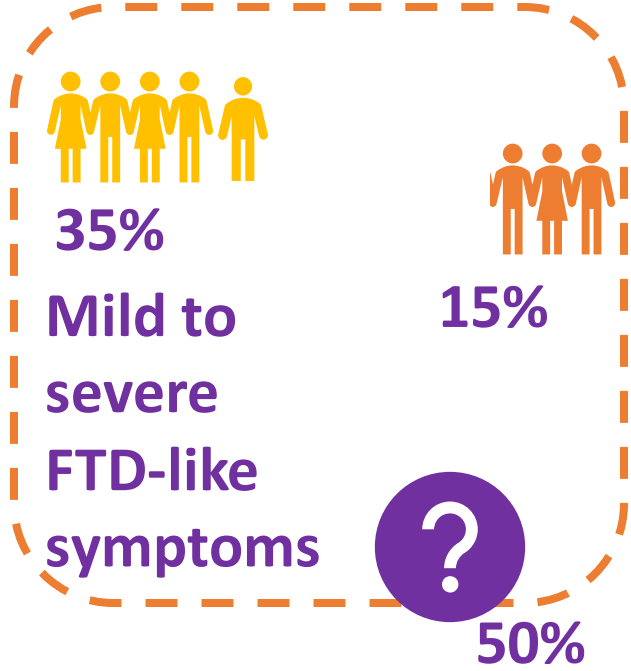
Compulsive behaviour

"And there were other things such as he's still eating the same food, he would eat the same food day after day after day. He would take himself to a supermarket and come back with exactly the same six or seven items every time. And so things like that, which I had noticed and thought, "This is just really weird," you know, it suddenly fell into place when she said they become fixated on things and you get this sort of behaviour."

Carer 09

Strong et al, ALSFTD 2017

NICE National Institute for Health and Care Excellence
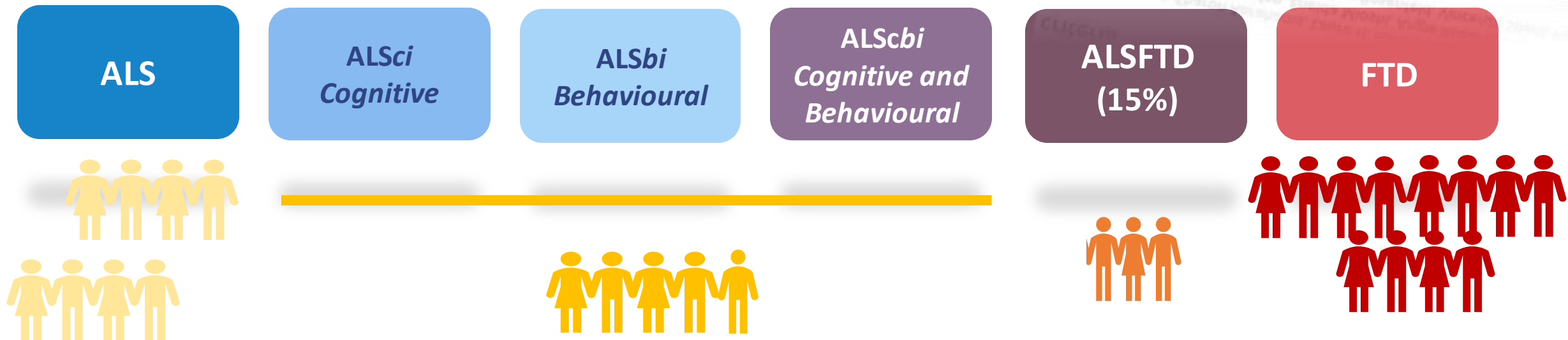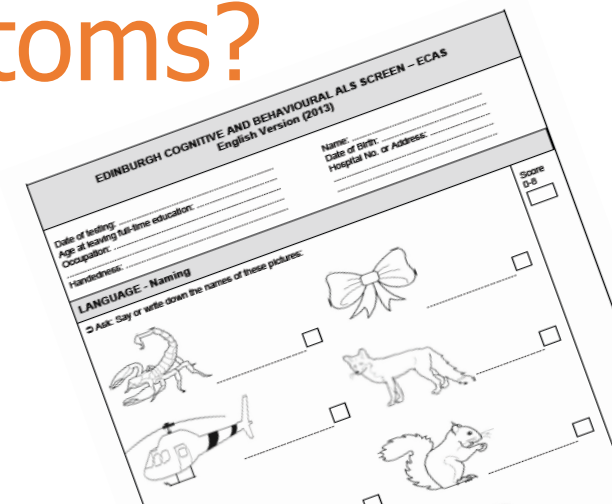
NICE

guideline

Motor neurone disease: assessment and management

NICE guideline
Published: 24 February 2016
Last updated: 23 July 2019

www.nice.org.uk/guidance/ng42

How do we professionals handle cognitive and behavioural symptoms?

EDINBURGH COGNITIVE AND BEHAVIOURAL ALS SCREEN – ECAS
English Version (2013)

MND Connect

MND Info Line
1800 777 175
9am to 4.30pm Monday to Friday

Information & Support > For health professionals & service providers > Managing symptoms > Cognitive change

🖨 PDF/Print this page

Cognitive change

This information is for health professionals and service providers.

More than 50% of people with MND will experience changes in cognition, language, behaviour and personality. These changes may be classified as ALS with cognitive impairment, ALSci, ALS with behavioural impairment, ALSbi, and ALS with cognitive and behavioural impairment, ALScbi.

Most people experience relatively mild changes. However, a small proportion will show more significant changes and will receive a diagnosis of 'motor neurone disease with frontotemporal dementia' or ALS-FTD.

mnd Australia   Information & Support   Research   Advocacy   Get Involved   About Us   🔍

MND Connect

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Factsheet

Information & Support > Information resources > Cognitive and behaviour change in MND

🖨 PDF/Print thi

Cognitive and behaviour change in MND

helping you today help others tomorrow

Best Prac
Guideline
Care Prof

Cognitive and behavioural change
Prevalence, detection, and implica

What can I do after I detect these symptoms?

Proactive carers also report feeling unsupported and frustrated with services' lack of knowledge of ALSFTD, and related symptoms

The state of play of HCPs doing research

Current numbers are low

Registered HCPs represent the largest single workforce in the NHS. But the proportion that facilitates, delivers or leads research remains low. In 2017 clinical academics represented 4.6% of NHS medical consultants. This is less than 0.1% of the nursing, midwifery and allied health professions (AHPs).

NIHR and NHS England are working in partnership to overcome the barriers to research. The collaboration is focused on increasing the awareness of research careers at early career stages. It also includes providing a clear framework for post-doctoral clinical or practitioner academics.

0.1%

ALS/MND Specialist Services

Frontotem...
Early onset...

✅ 50%

35% Mild to severe FTD-like symptoms

❓ 50%

15% ALSFTD

Photo by Jonny Gios on Unsplash

Intervention development

Develop intervention

Either developing a new intervention, or adapting an existing intervention for a new context, based on research evidence and theory of the problem

OR

Identify intervention

Choosing an intervention that already exists (or is planned), either via policy or practice, and exploring its options for evaluation (evaluability assessment)

Feasibility

Assessing feasibility and acceptability of intervention and evaluation design in order to make decisions about progression to next stage of evaluation

Core elements

- Consider context
- Develop, refine, and (re)test programme theory
- Engage stakeholders
- Identify key uncertainties
- Refine intervention
- Economic considerations

Evaluation

Assessing an intervention using the most appropriate method to address research questions

Implementation

Deliberate efforts to increase impact and uptake of successfully tested health innovations

Psychoeducational intervention

Compulsory modules	Modules on symptoms	Modules on strategies
What is MND? How common are non-motor changes?	What is apathy?	Encourage and Prompt
Which are the brain changes in MND?	What is disinhibition?	Adapt and Accommodate
Top Tips from carers, to carers	What is rigidity?	Simplify and Clarify
Looking after your wellbeing	What are deficits in social cognition?	Prepare and Increase Awareness
	What are hallucinations?	Support and Share Decisions
	What is lack of insight into own changes?	
	What are eating changes?	

Welcome to MiNDToolkit

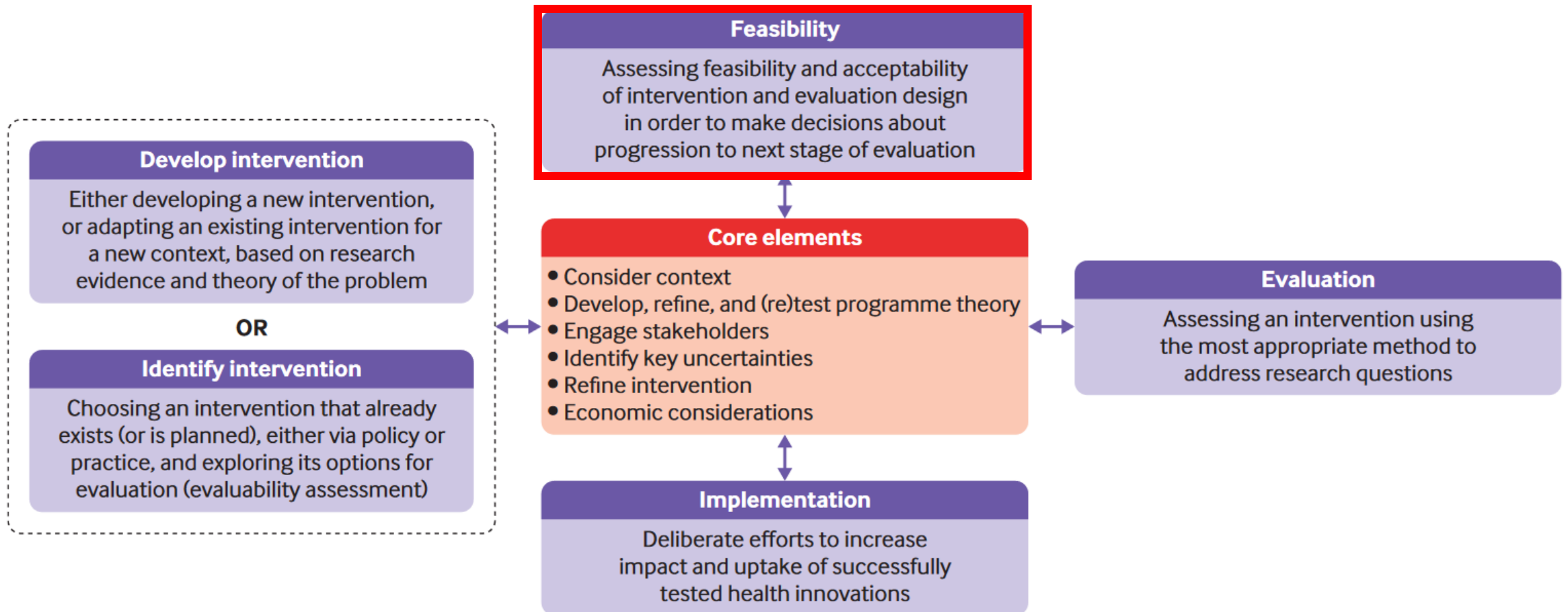A novel online platform for management of non-motor symptoms in Motor Neurone Disease

Learn more
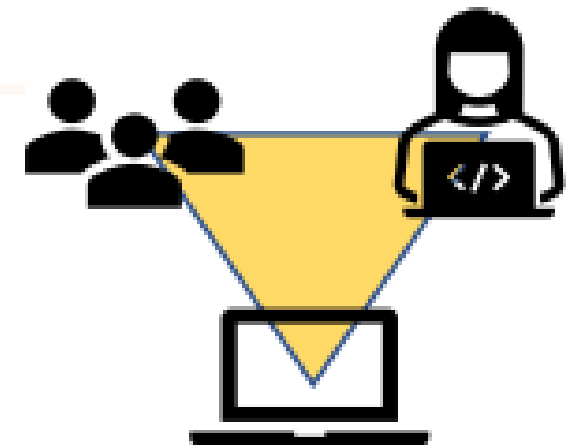
Feasibility study

MiNDToolkit feasibility trial design
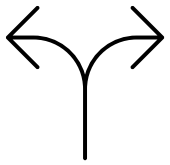
Two-arm, randomised controlled multi-centre trial
Arm 1: treatment plus usual care
Arm 2: usual care only (wait list)

Sample size: 20-30 carers
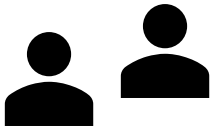
Sites: 10. HCPs: 20 (2 per site)

Trial registration: ISRCTN 15746123

Mioshi et al, under review

What is the MiNDToolkit for?

To support carers to manage people with MND with behavioural symptoms such as apathy, rigidity, disinhibition and other ALSFTD related symptoms

Who delivers the intervention?

The new online platform. HCPs are trained and can re-enforce the MiNDToolkit content, during consultations and home visits

Research question

Before conducting any large randomized control trials of the MiNDToolkit as an intervention it is important to answer the question "Can this study be done?"

Study aims

To test the feasibility and acceptability of the MiNDToolkit online intervention in MND Specialist Settings

Feasibility
Acceptability

Results of the MiNDToolkit feasibility trial

MiNDToolkit participating UK sites (n=11)
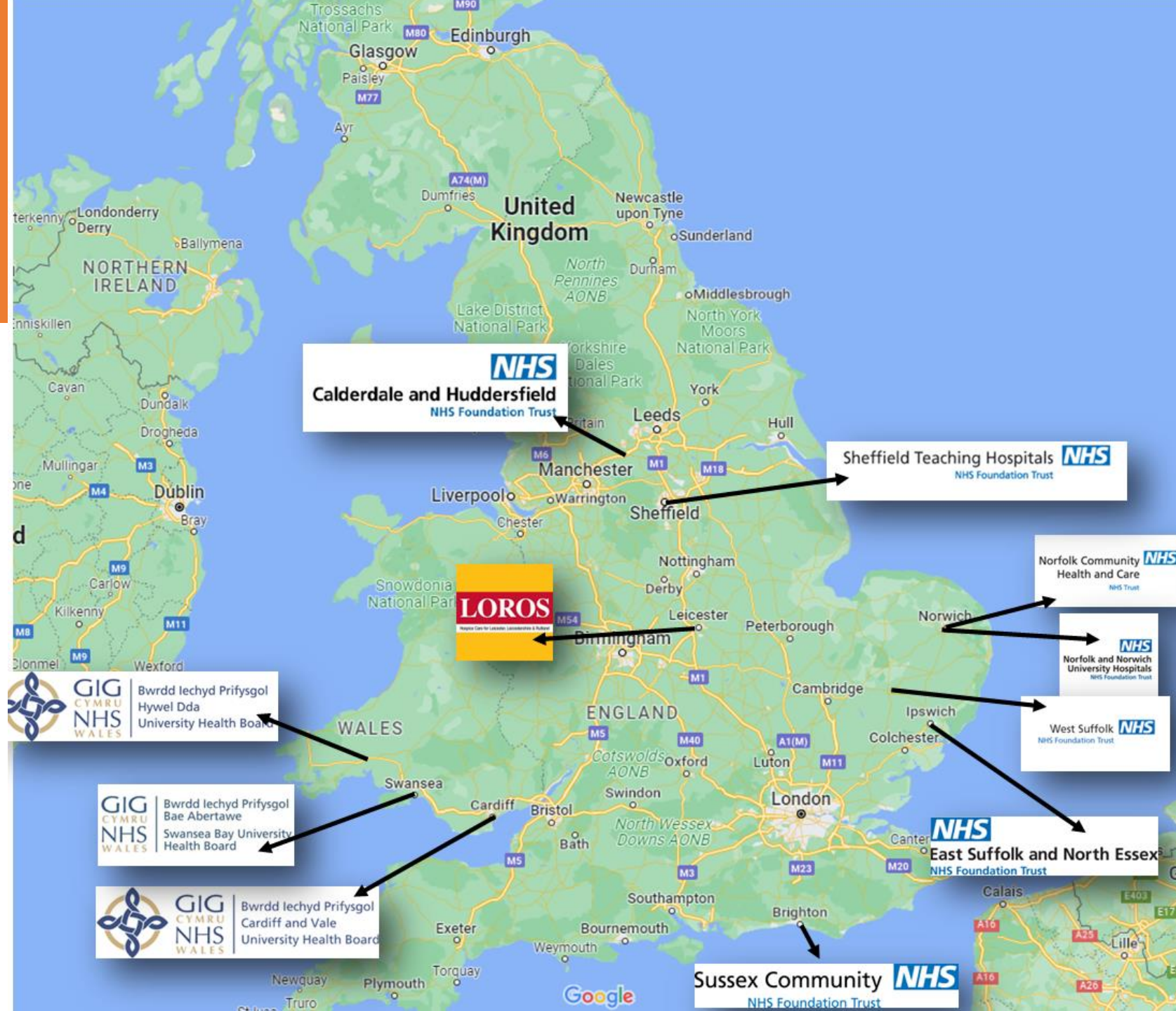
Feasibility results

Measure[a]	Proportion	Exact[b] lower 95% confidence interval	Exact[b] upper 95% confidence interval
Approached rate (approached/screened)	151/284 0.5317	0.4718	0.5909
Recruitment rate (consented/approached)	30/151 0.1987	0.1382	0.2713
Randomization rate (randomized/consented)	29/30 0.9667	0.8278	0.9992
Attrition rate (to end of FU) (withdrawals/randomized)	1/29 0.0345	0.0009	0.1776
Reaching FU rate	24/29 0.8276	0.6423	0.9415
FU questionnaires abandoning	1/24 0.0417	0.0011	0.2112
FU completion outside 30 days	2/23 0.0870	0.0107	0.2804

53% carers approached

97% randomised

83% Retention rate

Mioshi et al, under review

Randomised (n=29)

Allocation

Allocated to intervention (n=14)
- Received allocated intervention, n=12
- Did not receive allocated intervention, n=02 (carer did not use platform and HCP did not reinforce intervention)

Allocated to usual care/wait list (n=15)
- Received allocated intervention, n=15

- Lost to follow up (n=01)
- Did not interact with the intervention; did not complete FU (n=02)

- Lost to follow up (n=01)
- Withdrawal: discontinued intervention due to patient death (n=01)

Assessment

Assessed for feasibility objectives (n= 11)
Assessed for acceptability objectives (n=11)

Assessed for feasibility objectives (n= 13)
Assessed for acceptability objectives (n=13)

Randomisation was accepted

1/29 carer withdrawn

4 carers lost to follow-up

Mioshi et al, under review

Carer demographics

	Intervention n=14	Control n=15
Age, median (IQR)	61.5 (58, 68)	57 (55, 70)
Sex, female	78.6%	80%
Ethnicity, Caucasian	71.4%	80%
Relationship to PwMND, spouse	85.7%	80%
Employment status, full time	35.7%	20%

Mioshi et al, under review

Acceptability of MiNDToolkit: carers

Process evaluation led by
Thando Katangwe-Chigamba

Carer results: Views and experiences with the MiNDToolkit Platform

MiND TOOLKIT

Carers found bitesize structure of MiNDToolkit accessible, and easy to use

A few carers completed all modules in the first week

Adaptations to the use of the Toolkit included:
- Sharing MiNDToolkit with other family members
- Sharing MiNDToolkit with people with MND

The MiNDToolkit, it was in nice, easy, bite size portions, if you like, to coin a phrase. But yeah, **nothing took more than sort of like 10,15 minutes which was great for me**. So **I could do in between, you know, household jobs or I could even access it at work** and do a couple of modules during my lunch hour or in a break. So it's that kind of short, only about five, 10 minutes long which **makes it more accessible"**
Carer 02

'once I started, I could not stop'
Quote from carer, noted in platform by HCP

27

Mioshi et al, in preparation

Interviewer: […] the explanation of the symptoms, like apathy; how helpful was that?

Carer: they were very helpful because I hadn't – at that stage, the very fact I could talk to you about it, I wouldn't have been able to talk to you about it if I hadn't watched the video because I wouldn't have known.

Learning about behavioral symptoms,
and perceiving an ability to make changes in daily management was empowering and
supported acceptance of these non-motor symptoms.

"Well I didn't actually associate it with MND"

35% Mild to severe FTD-like symptoms

15% ALSFTD

50%

"This was quite a while before he was diagnosed [with PLS]. He started having an obsession that we had bedbugs in the house and that he had been bitten. Anyway, I spoke to his GP about it and I'll keep my voice down a little bit, and the GP arranged to come up and

"Having those explained as symptoms if you like, has been really interesting because I wasn't aware of those.
As I say, I was prepared for the physical side of [MND/ALS] because of his mother, but I had no idea about the cognitive side and so that has been really interesting.
And one thing I sort of remember from the modules is it says you can sort of jolly somebody out of apathy but not out of depression, you know, making a distinction there. So yeah, I would say having those symptoms spelt out, it did like, because I could really identify with those" Carer 09

Apathy

"But yes, regards sitting here listen car for a drive jus or listens to his iP People come to v now type everyth house" *Carer 02*

Carer results: Platform engagement

Engagement with platform was variable, and not dependent on age; employment status or severity of behavioural symptoms

The least engaged carer on the platform, received the most reinforcement i.e., HCP delivered the content

The most engaged carer on the platform did not receive any reinforcement from HCP

Mioshi et al, in preparation

Interviewer: […] the explanation of the symptoms, like apathy; how helpful was that?

Carer: they were very helpful because I hadn't – at that stage, the very fact I could talk to you about it, I wouldn't have been able to talk to you about it if I hadn't watched the video because I wouldn't have known.

Learning about behavioural symptoms, and perceiving an ability to make changes in daily management was empowering and supported acceptance of these non-motor symptoms.

Mioshi et al, in preparation
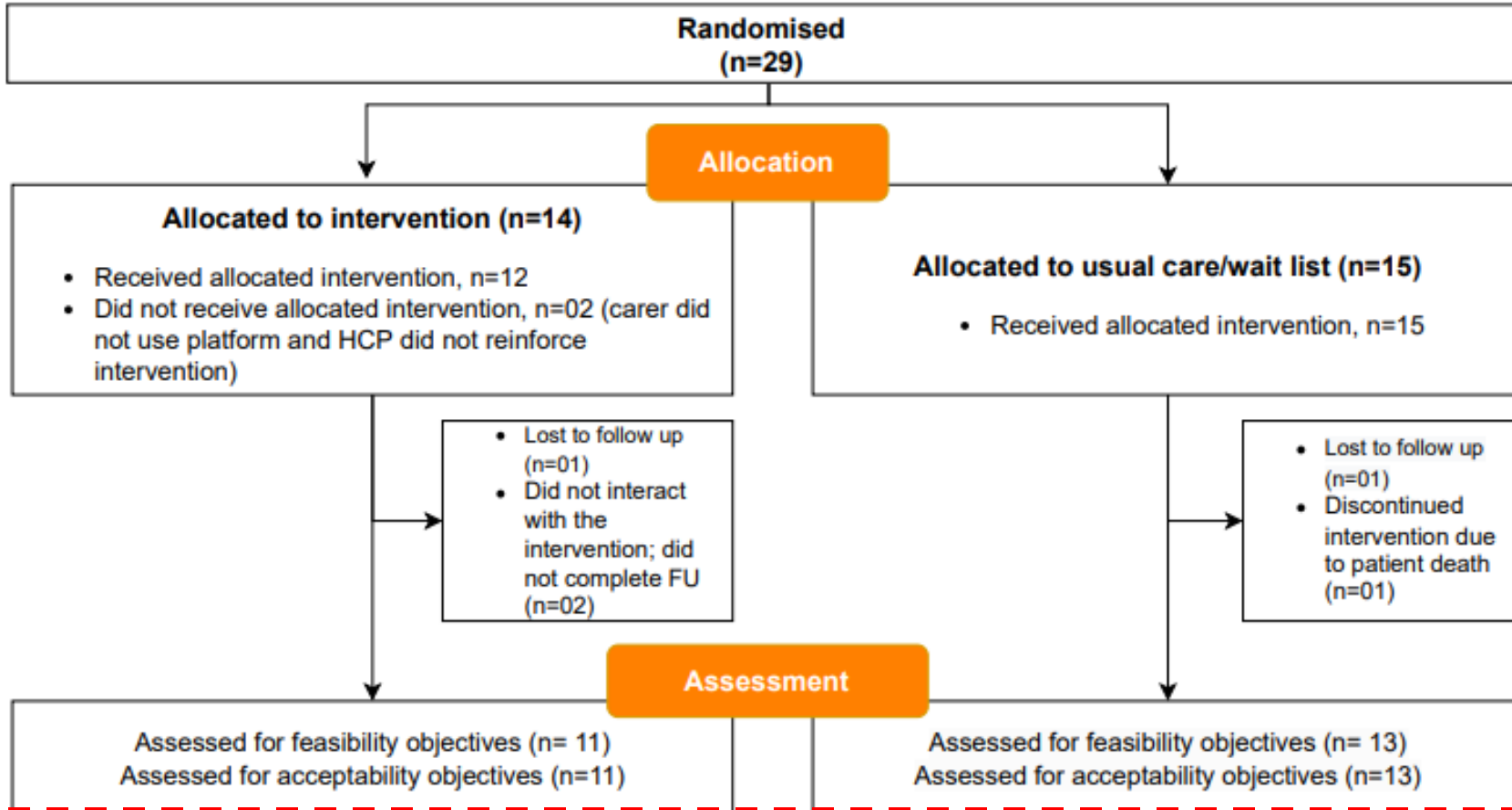
HCP results: the MiNDToolkit Training

HIGHLIGHTED AND ADDRESSED GAPS IN KNOWLEDGE

INCREASED CONFIDENCE TO IDENTIFY AND DISCUSS BEHAVIOURAL SYMPTOMS

HCP EQUIPPED TO DEMONSTRATE AND IDENTIFY STRATEGIES

"**I thought I knew quite a bit about it** [Behavioural symptoms] **but it turns out I didn't, I learnt a lot from doing the training**…I think the quiz at the start and then again at the end is brilliant because to look back and think, "Oh crumbs I got that wrong," and **the presumption of knowledge and experience to then be reminded, well actually I didn't know that and I didn't know that, is really good**…." *HCP_06_MND Coordinator*

ALS/MND Specialist Services
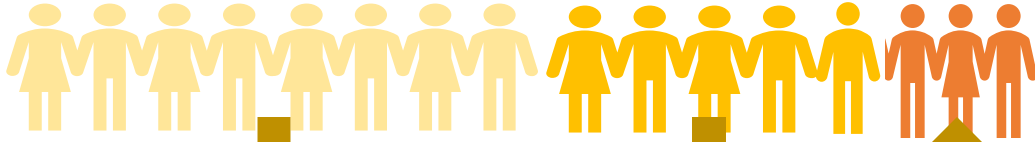
Frontotemporal dementia (FTD) and/or Early onset dementia Specialist Services

50%

35% Mild to severe FTD-like symptoms

15% ALSFTD
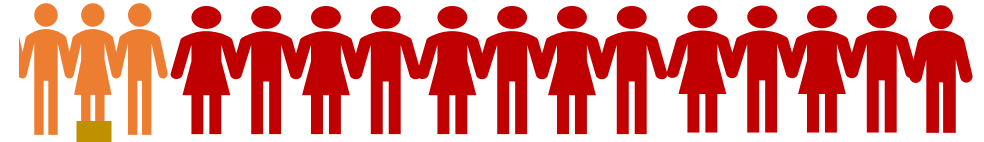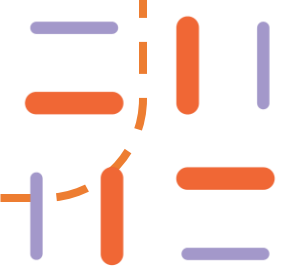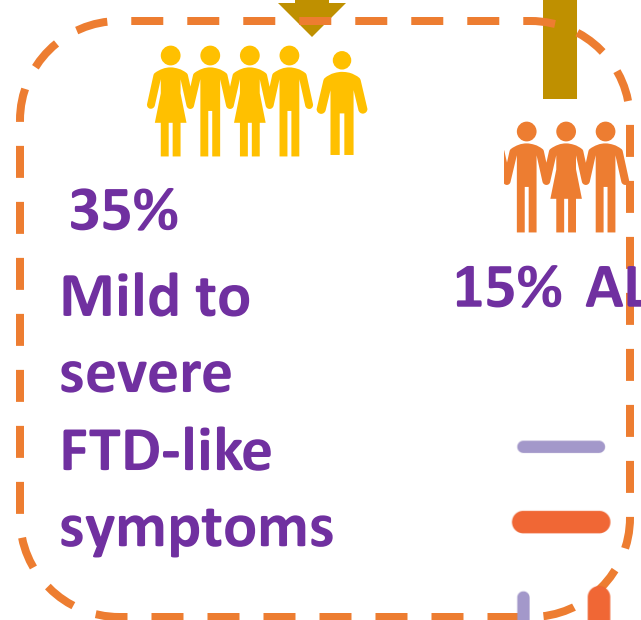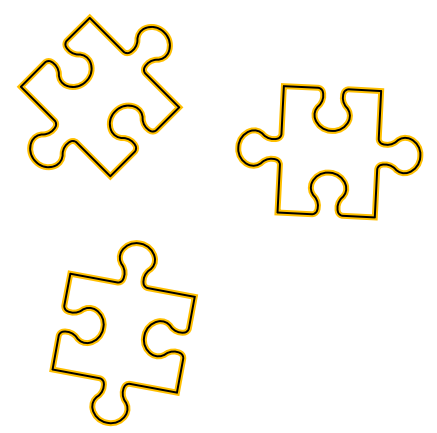
MiND
TOOLKIT

Take home message

MiNDToolkit's feasibility results mean we can now take the next step, and test its efficacy

We need to identify ways to support not only the PwMND, but also their carers. Carers need specialist support for management of behavioural symptoms

Supporting carers is everyone's business: multidisciplinary team. MiNDToolkit reached a diverse professional groups, e.g., nurses, OTs, PTs, SLTs, neurologists, palliative care consultants

Acknowledgements

All HCPs
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Gemma O'Brien
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Polly Ashford
Lee Shepstone
Thando Katangwe-Chigamba
Martin Pond
Kelly Grant

Trial Management Group
CTU team
Becky Gould
Sue Heal

Michael Hornberger
Naoko Kishita

All family carers who have given their time to support our research studies

Our funders

MiND TOOLKIT *is powered by* Mantal

e.mioshi@uea.ac.uk
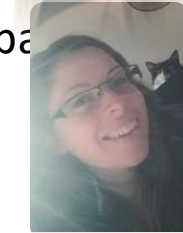
Case studies

Case 1

- Lady
- Family history of MND
- Bulbar onset

- Disinhibited
- Poor insight into own changes

- Lives with one child. Another child lives nearby and is also involved in care
- Referral to dementia team. Waiting list – 3 months

Amyotrophic Lateral Sclerosis - Frontotemporal spectrum disorder (ALSFTD) - International Consensus Revised Diagnostic Criteria (2017)

Amyotrophic Lateral Sclerosis and Frontotemporal Degeneration

ISSN: 2167-8421 (Print) 2167-9223 (Online) Journal homepage: http://www.tandfonline.com/loi/iafd20

Amyotrophic lateral sclerosis - frontotemporal spectrum disorder (ALS-FTSD): Revised diagnostic criteria

Michael J. Strong, Sharon Abrahams, Laura H. Goldstein, Susan Woolley, Paula Mclaughlin, Julie Snowden, Eneida Mioshi, Angie Roberts-South, Michael Benatar, Tibor HortobáGyi, Jeffrey Rosenfeld, Vincenzo Silani, Paul Ince & Martin R. Turner

| ALS | ALSci Cognitive | ALSbi Behavioural | ALScbi Cognitive and Behavioural | ALSFTD (15%) | FTD |

Strong et al, ALSFTD 2017

ALS cognitive impairment - ALSci

| Evidence of executive dysfunction (e.g. verbal letter fluency), including social cognition | OR | Evidence of language dysfunction (two non-overlapping tests) | OR | Evidence of executive dysfunction (e.g. verbal letter fluency), including social cognition | Evidence of language dysfunction (two non-overlapping tests) |

| ALS | ALS*ci* Cognitive | ALS*bi* Behavioural | ALS*cbi* Cognitive and Behavioural | ALSFTD (15%) | FTD |

Strong et al, ALSFTD 2017

ALS behavioural impairment - ALSbi

Identification of apathy with or without other behavioural change

OR

Meeting at least two non-overlapping supportive diagnostic features from the Rascovsky criteria Behaviuoral variant FTD

Apathy

Disinhibition

Loss of sympathy/empathy

Perseverative/ Compulsive behaviours

Hyperorality

ALS

ALS*ci* Cognitive

ALS*bi* Behavioural

ALS*cbi* Cognitive and Behavioural

ALSFTD (15%)

FTD

Strong et al, ALSFTD 2017

ALS cognitive and behavioural impairment - ALScbi
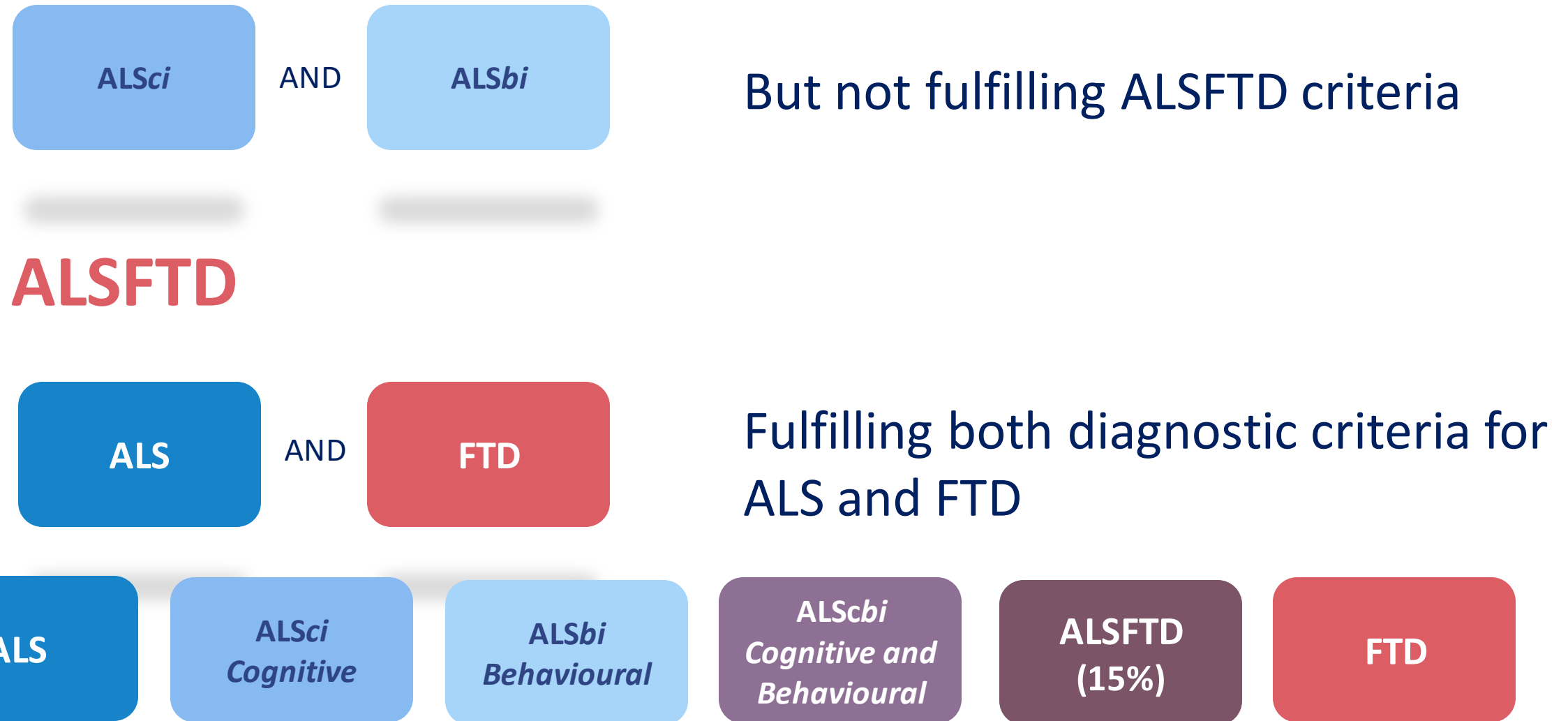
ALS*ci*	AND	ALS*bi*

But not fulfilling ALSFTD criteria

ALSFTD

ALS	AND	FTD

Fulfilling both diagnostic criteria for ALS and FTD

ALS	ALS*ci* Cognitive	ALS*bi* Behavioural	ALS*cbi* Cognitive and Behavioural	ALSFTD (15%)	FTD

Strong et al, ALSFTD 2017

ECAS: Edinburgh Cognitive and Behavioural ALS Screen

Multidomain:

- MND-Specific functions: Executive, Language, Fluency;

- MND Non-specific functions (Memory and Visuospatial functions)

Designed for physical disability: written or spoken response

Carer Behaviour Interview: based on new diagnostic criteria

Niven, Abrahams et al, ALSFTD 2013

MiND-B

ORIGINAL ARTICLE

A novel tool to detect behavioural symptoms in ALS

ENEIDA MIOSHI[1,2,3], SHARPLEY HSIEH[1], JASHELLE CAGA[1], ELEANOR RAMSEY[1], KELLY CHEN[2], PATRICIA LILLO[1], NEIL SIMON[1], STEVE VUCIC[3], MICHAEL HORNBERGER[1,2,4], JOHN R. HODGES[1,2] & MATTHEW C. KIERNAN[1,2,5]

[1]Neuroscience Research Australia, Sydney, Australia, [3]Department of Psychiatry, U Neurosciences, University of Cambridge,

- *Multidomain: 9 items only*
- *Can be completed in waiting area by proxy*
- *5 minutes*

6. Repeatedly uses the same expression or catch phrase
4 – Never; or no change to 15 years ago
3 – A few times per month
2 – A few times per week
1 - Daily
0 – Constantly

Stereotypical Behaviour/Rigidity

7. Shows less enthusiasm for his/her usual interests
4 – Never; or no change to 15 years ago
3 – A few times per month
2 – A few times per week
1 – Daily
0 – Constantly

Apathy

8. Shows little interest in doing new things
4 – Never; or no change to 15 years ago
3 – A few times per month
2 – A few times per week
1 – Daily
0 – Constantly

9. Fails to maintain motivation to keep in contact with friends or family
4 – Never; or no change to 15 years ago
3 – A few times per month
2 – A few times per week
1 – Daily
0 – Constantly

Disinhibition

Case 2

- Gentleman
- Bulbar onset
- Emotional lability

- Does not engage with MDT
- Does not pursue any of his hobbies any longer, e.g. abandoned gardening and does not feed or walk the dog

- Very engaged spouse who is very upset because "he has given up"

MiND-B

To detect behavioural symptoms

- *Multidomain: 9 items only*
- *Can be completed in waiting area by proxy*
- *5 minutes*

PHQ-9

To detect depressive symptoms

Carer interview
+
Clinical observation

The state of play of HCPs doing research

Current numbers are low

Registered HCPs represent the largest single workforce in the NHS. But the proportion that facilitates, delivers or leads research remains low. In 2017 clinical academics represented 4.6% of NHS medical consultants. This is less than 0.1% of the nursing, midwifery and allied health professions (AHPs).

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https://www.nihr.ac.uk/news/extra-30m-a-year-to-boost-research-careers-for-healthcare-professionals/34213

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All family carers who have
given their time to support our
research studies