Leicestershire Partnership
NHS Trust

Psychological support for people with Huntington's Disease (HD)

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Leicestershire Partnership NHS Trust

(With thanks to Jordan Rainbird, Trainee Clinical Psychologist and Prof Jane Simpson)

Aims of this session

01 To understand psychological difficulties among people with HD (pwHD)

02 To highlight the research base for psychological interventions for pwHD

03 An overview of psychological approaches across HD stages

04 Discuss provision of psychology resources for pwHD in UK

01

Psychological difficulties among pwHD

Psychological difficulties in pwHD

Low Mood (depression)

Perseveration

Mood swings

Increased risk of suicide

Psychosis

‘Irritability’ and aggressiveness

Anxiety

Obsessions/ compulsions

Apathy

Cognitive difficulties

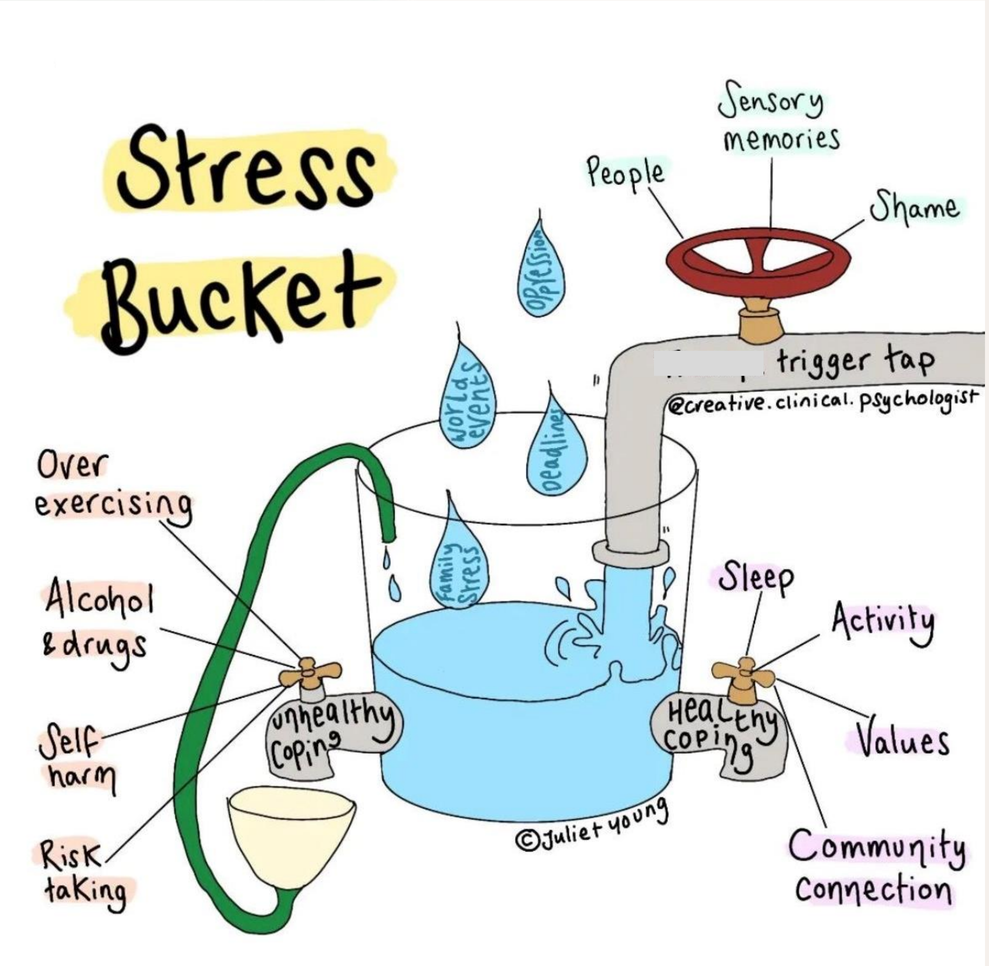
A psychological approach to HD

- ***Zarotti, Dale, Eccles & Simpson (2022)***

  In ***More than Just a Brain Disorder: A Five-Point Manifesto for Psychological Care for People with Huntington's Disease***, highlighted that HD literature has predominantly used a biomedical approach to understand psychological difficulties in HD, often linking symptoms like depression and anxiety to brain-based pathophysiological changes.

- ***Snow et al. (2024)***

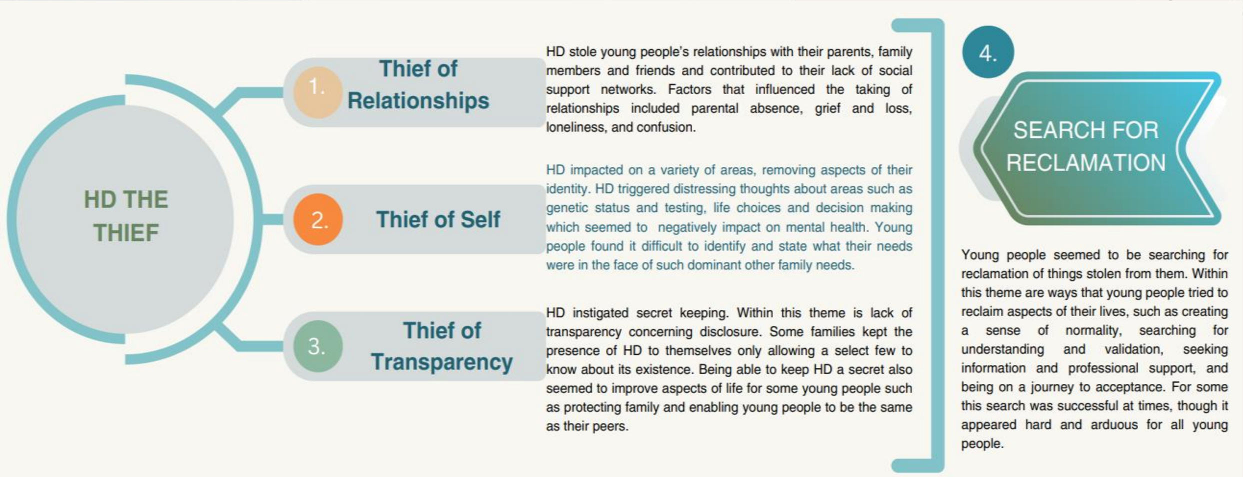
  \- suggest that ***psychological challenges in HD are multifactorial***, significantly influenced by cognitive functioning and stress, and may also be shaped by individuals' coping strategies when dealing with life stressors.


Stress Bucket
Sensory memories
People
Shame
oppression
world events
deadlines
family stress
[redacted] trigger tap
@creative.clinical.psychologist
Over exercising
Alcohol & drugs
Self harm
Risk taking
unhealthy coping
Healthy coping
Sleep
Activity
Values
Community connection
©Juliet young

Growing up in an HD family

Cooper, H., Simpson, J., Dale, M., & Eccles, F. J. R. (2024). **Experiences of young people growing up in a family with Huntington's disease: A meta-ethnography of qualitative research.**

13 papers , four themes were identified highlighting aspects of childhood that were stolen and fought for:



HD THE THIEF

1. Thief of Relationships

HD stole young people's relationships with their parents, family members and friends and contributed to their lack of social support networks. Factors that influenced the taking of relationships included parental absence, grief and loss, loneliness, and confusion.

2. Thief of Self

HD impacted on a variety of areas, removing aspects of their identity. HD triggered distressing thoughts about areas such as genetic status and testing, life choices and decision making which seemed to negatively impact on mental health. Young people found it difficult to identify and state what their needs were in the face of such dominant other family needs.

3. Thief of Transparency

HD instigated secret keeping. Within this theme is lack of transparency concerning disclosure. Some families kept the presence of HD to themselves only allowing a select few to know about its existence. Being able to keep HD a secret also seemed to improve aspects of life for some young people such as protecting family and enabling young people to be the same as their peers.

4. SEARCH FOR RECLAMATION

Young people seemed to be searching for reclamation of things stolen from them. Within this theme are ways that young people tried to reclaim aspects of their lives, such as creating a sense of normality, searching for understanding and validation, seeking information and professional support, and being on a journey to acceptance. For some this search was successful at times, though it appeared hard and arduous for all young people.

Within the themes, the **complex challenges young people** faced when growing up in a HD family were explored such as the **impact of adverse childhood experiences** and the possible effects of HD on **attachment and social relationships**.

Living at-risk

Maintaining psychological wellbeing when living at risk of Huntington's disease: and interpretative phenomenological analysis

Cooper, H. Simpson, J. Dale M. and Eccles (2024)

This resulted in three themes:

(1) "you're constantly in limbo": living in two worlds;

(2) "I have to live, just bloody live": managing the possibility of a time-limited lifespan; and

(3) "I try and try my hardest to look past the disease": the exhausting quest to keep living well.

Stress affects distress among pwHD

A recent study by Snow et al. (2024) found that HD-related stressors are:

•Highly prevalent among both HD patients and at-risk individuals.
•Heterogeneous in nature, encompassing a wide range of personal and interpersonal stressors.
•Examples of common stressors include:
Interpersonal stressors, such as:
  -**The impact of HD on family members.**
  -Taking on increased responsibilities.
Personal stressors, such as:
  -**Fears or concerns about the future.**
•Higher stress levels are associated with increased symptoms of depression and anxiety for both HD patients and at-risk individuals.

  -The effect size was large for HD patients.
  -The effect size was medium for at-risk individuals.

A reflection about living at-risk

- Pause for a moment… if you were at risk of HD, would you want to know?

- What would the reasons be for your decision?

- What would the implications be of testing?

- What would the pros and cons be of testing and knowing, or living with uncertainty?

Psychological difficulties in pwHD

Other **difficult emotions** and/or **stressors** often include:

- Loss – of roles, identity, independence, planned future, other family members
- Shame
- Guilt
- Attachment issues
- Self-esteem
- Lack of control/uncertainty
- Family secrets

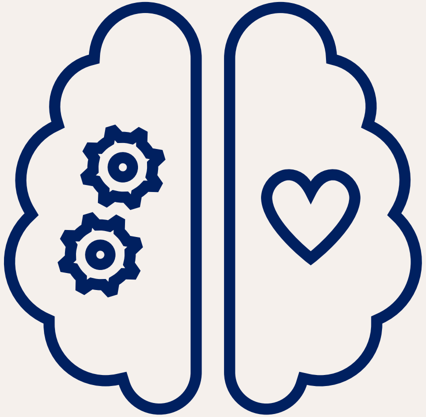
- Parenting
- Stigma/discrimination
- Loneliness
- Family dynamics e.g. communication, testing, differences of opinion, change in roles
- Decisions about having children

Stress and depression

Bilal et al (2023) - depression among **both premanifest and manifest groups** was associated with stress regarding numerous psychosocial aspects of HD

1. Difficulties with movement	11. Not feeling in control of my life
2. Difficulty with thinking and communication skills	12. Feeling isolated from my friends and/or family
3. Personality changes, aggression, and/or mood swings	13. Grieving the death of a loved one (past or future)
4. Sleeping difficulties (e.g., not getting enough sleep)	14. Not getting enough help or social support
5. Not being able to socialise as much with family and/or friends	15. Passing the HD gene on to my child (or children)
6. Not being able to work or do leisure activities as much as before	16. Losing a sense of meaning or purpose for my life
7. Depending on others as my symptoms get worse	17. Financial concerns
8. Concern about what others think of me (now, or as my symptoms get worse)	18. The effect of HD on my current/future relationships
9. Concerns about the future and/or death	19. Not knowing how, or if, to tell others about my diagnosis
10. Unstable or chaotic home life due to HD	

Psychological impact of cognitive changes in pwHD



Cognitive changes among pwHD can impact emotion regulation, leading to agitation, fear, memory loss, decreased information processing, and communication difficulties. Individuals may experience apathy, emotional blunting, disinterest in activities previously enjoyed, and distress.

On top of this, difficulties with executive function in individuals with HD can lead to reliance on strict routines for reassurance and predictability, causing distress when routines are disrupted.

How people cope seems to matter

- According to Snow et al. (2024), coping strategies that involve **cognitive reframing, acceptance, and distraction** (referred to as secondary control coping) are significantly linked to reduced symptoms of depression and anxiety in **both individuals with HD and those at risk**.

- Primary control coping strategies, which focus on addressing or changing the stressor or one's emotional response through methods like problem-solving, emotional expression, and emotion regulation, may offer some benefits,

- **The use of disengagement coping strategies could have negative effects**. These strategies involve attempts to avoid a stressor either behaviourally or psychologically and include avoidance, denial, and wishful thinking.

- However, further research is needed to confirm these findings.

People in HD families without the gene are affected too

- According to Osawa et al. (2024), noncarriers who received negative results experienced a range of emotions, including relief, surprise, doubt, release from tension, and even regret. The study revealed that noncarriers often felt survivor's guilt, not just as a feeling of guilt but also as a sense of duty or responsibility towards others. Additionally, some participants expressed concerns about disclosing their results to their siblings. These findings highlight that noncarriers for Huntington's disease can go through complex psychological experiences.

- Carers with no genetic risk (e.g. unaffected parents, partners, spouses etc) – high levels of “burden” compared to other neuro conditions, potentially have to care for multiple family members, complexity of the disease.

Impact on Families

- Carers often struggle with the change in their loved ones (e. g. changes in personality), and the changing roles in the relationship, e. g. from being partners to being a carer for their loved one.

- Anticipatory grief as well as guilt can be common, particularly around times such as the loved one receiving the HD diagnosis, once their loved one starts experiencing symptoms, or once they move from home to a nursing facility/hospital. Families may grieve losing the person as they knew them before the HD symptoms started, or feel like they let their loved one down by not being able to keep them home, spend more time with them, etc.

- In families with children, parents may often struggle with how to approach conversations about HD with their children.

A brief overview of some psychological therapies…

Psychological therapies

Cognitive Behaviour Therapies (CBTs):
CBT holds that emotional distress and behavioural difficulties arise from ‘maladaptive’ or unhelpful cognitions, which comprise general beliefs about the world, the self and the future. The therapy is predicated on the assumption that changing these cognitions through therapeutic interventions will reduce distress and problematic behaviours.

Third Wave CBTs:

- **Acceptance and commitment therapy** (ACT): aims to improve individuals’ acceptance of distressing thoughts, beliefs, sensations and emotions. There is emphasis on identifying and acting upon values which are most important to the person, designing behavioural changes which direct one towards living these values.

- **Compassion Focused Therapy:** aims to help promote mental and emotional healing by encouraging people in treatment to be **compassionate** toward themselves and other people.

Psychological therapies

Mindfulness-based approaches: focus on bringing the attention to the present moment and accepting feelings, sensations and emotions non-judgementally, including the experience of illness. Approaches may include elements of cognitive therapy (MBCT) or may focus on stress reduction (MBSR).

Relaxation techniques: include interventions that focus on relieving muscle tension (e.g. progressive attention to muscle groups, tensing and releasing in turn while focusing on the sensation of release following tension) or guided imagery (e.g., visualising relaxing scenes). These may be supported by relaxing music or guided narratives to assist the exercises.

Psychological therapies

Psychoeducation: focuses on developing individuals' understanding about their condition with the aim is to empower individuals to manage their condition and their own reactions to it more successfully and consequently improve their well-being. Programmes may include elements of CBT (e.g., cognitive restructuring), behavioural activation, development of stress coping strategies, social skills training, role play, and relaxation skills development.

Narrative therapy: empowers clients to explore and reframe their life stories

Family/systemic therapy: enables family members to express and explore difficult thoughts and emotions safely, to understand each other's experiences and views, appreciate each other's needs, build on family strengths and make useful changes in their relationships and their lives.

Psychological therapies

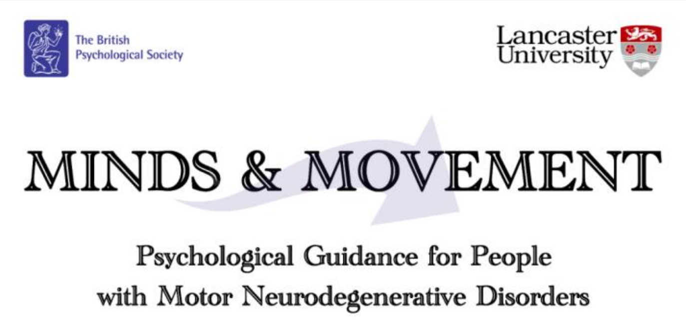
Behaviour therapies – umbrella term for range of approaches based on premise that **behaviours can be learned and subject to change**
e.g., positive behaviour support (promotes pro-active and preventive strategies and includes teaching new skills It may include strategies to avert crisis and keep people safe) **, applied behavioural analysis**

Remotivation therapy - remotivation therapy is a person-centered approach that aims to **rekindle an individual's interest in the world around them**. It's not just about getting people to do things; it's about helping them rediscover the joy and satisfaction that comes from engaging with life.

Life story work – **creating resources about the person with HD**, such as their background, interests, and who and what is important to them (e.g. books, collages, audio/video recordings) for comfort and to enhance knowledge about the person for staff groups

Psychological interventions: What does the research tell us?

02



The British Psychological Society

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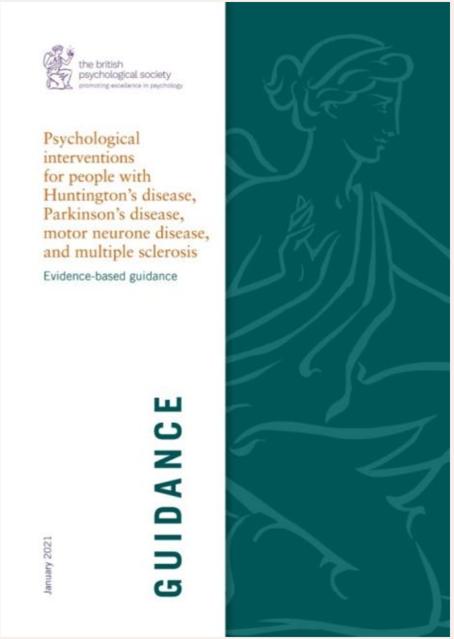
MINDS & MOVEMENT

Psychological Guidance for People
with Motor Neurodegenerative Disorders



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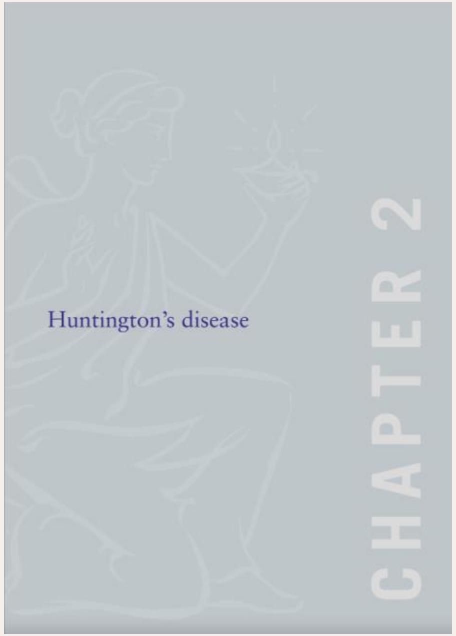
promoting excellence in psychology

Psychological
interventions
for people with
Huntington's disease,
Parkinson's disease,
motor neurone disease,
and multiple sclerosis

Evidence-based guidance

GUIDANCE

January 2021



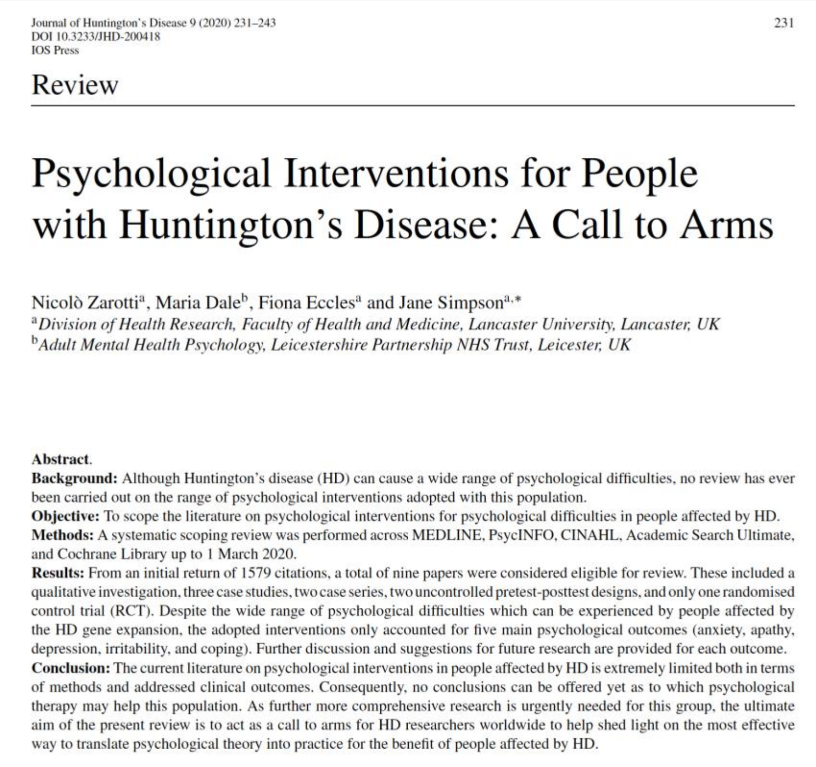
Huntington's disease

CHAPTER 2

Systematic review (2020) of all psychological interventions for HD

- Started with **1579**
- Screened **1062**
- Fully considered **15**
- Included **9**
    - Only **1 RCT (pilot n=12)**

Ref: Zarotti, N., Dale, M., Eccles, F.J.R. & Simpson, J. (2020) Psychological interventions for people with Huntington's disease: a call to arms. *Journal of Huntington's Disease*. 10.3233/JHD-200418



Journal of Huntington's Disease 9 (2020) 231–243
DOI 10.3233/JHD-200418
IOS Press

Review

Psychological Interventions for People with Huntington's Disease: A Call to Arms

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Abstract.
Background: Although Huntington's disease (HD) can cause a wide range of psychological difficulties, no review has ever been carried out on the range of psychological interventions adopted with this population.
Objective: To scope the literature on psychological interventions for psychological difficulties in people affected by HD.
Methods: A systematic scoping review was performed across MEDLINE, PsycINFO, CINAHL, Academic Search Ultimate, and Cochrane Library up to 1 March 2020.
Results: From an initial return of 1579 citations, a total of nine papers were considered eligible for review. These included a qualitative investigation, three case studies, two case series, two uncontrolled pretest-posttest designs, and only one randomised control trial (RCT). Despite the wide range of psychological difficulties which can be experienced by people affected by the HD gene expansion, the adopted interventions only accounted for five main psychological outcomes (anxiety, apathy, depression, irritability, and coping). Further discussion and suggestions for future research are provided for each outcome.
Conclusion: The current literature on psychological interventions in people affected by HD is extremely limited both in terms of methods and addressed clinical outcomes. Consequently, no conclusions can be offered yet as to which psychological therapy may help this population. As further more comprehensive research is urgently needed for this group, the ultimate aim of the present review is to act as a call to arms for HD researchers worldwide to help shed light on the most effective way to translate psychological theory into practice for the benefit of people affected by HD.

What Is Working for HD? Zarotti, Dale, Eccles & Simpson (2020) review

- **Preliminary evidence** for **individual CBT** with both **premanifest** and **manifest** individuals for reducing **anxiety and depression**
- Promising results for **sensory modulation** and **progressive relaxation** in reducing **aggression**
- **No significant difference** in a pilot **RCT** exploring **multisensory stimulation** against simple relaxation **for aggression**
- **Preliminary evidence** for effectiveness **remotivation therapy** (**RmT**) in improving interest, awareness, and overall participation in **manifest** individuals with **apathy**
- Preliminary evidence on effectiveness of **individual CBT** and **narrative approaches** (especially for group genetic counselling) to enhance **coping**

Since our 2020 review…..

Some small (n =12, n=7) uncontrolled feasibility projects show promise for mindfulness-based approaches among premanifest HD participants:

- Eccles, F.J.R., Craufurd, D., Smith, A. *et al.* A feasibility investigation of mindfulness-based cognitive therapy for people with Huntington’s disease. *Pilot Feasibility Stud* **6**, 90 (2020). https://doi.org/10.1186/s40814-020-00631-z

- Eccles, F.J.R., Craufurd, D., Smith, A. *et al.* Experiences of Mindfulness-Based Cognitive Therapy for Premanifest Huntington's Disease 2021;10(2):277-291.
doi: 10.3233/JHD-210471.

A pilot evaluation of an 8-week mindfulness-based stress reduction program for people with pre-symptomatic Huntington's disease
Sarah Velissaris[1], Marie-Claire Davis[2], Fiona Fisher[2], Cathy Gluyas[2], Julie C Stout[3]
•. 2023 Aug;14(4):395-405.

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Further small projects have also been undertaken with the premanifest group and show some promising results:

- **The 'Hold me Tight' program (HmT),** based on emotionally focused **couples' therapy, aims at strengthening the couple bond by targeting attachment needs**. multiple baseline design using three baselines of varying length, **15 couples of presymptomatic HD-carriers and their partners were included.** In three consecutive groups. HmT improved wellbeing and relationship satisfaction of couples facing HD – **Lucienne van der Meer's group in Netherlands**

- Evaluating a genetic counseling narrative group session for people who have tested positive for the Huntington's disease expansion: An interpretative phenomenological analysis

Johanna Spiers, Jonathan A. Smith, Mariangels Ferrer-Duch, Ramona Moldovan, Jay Roche, Rhona MacLeod 2020 https://doi.org/10.1002/jgc4.1229

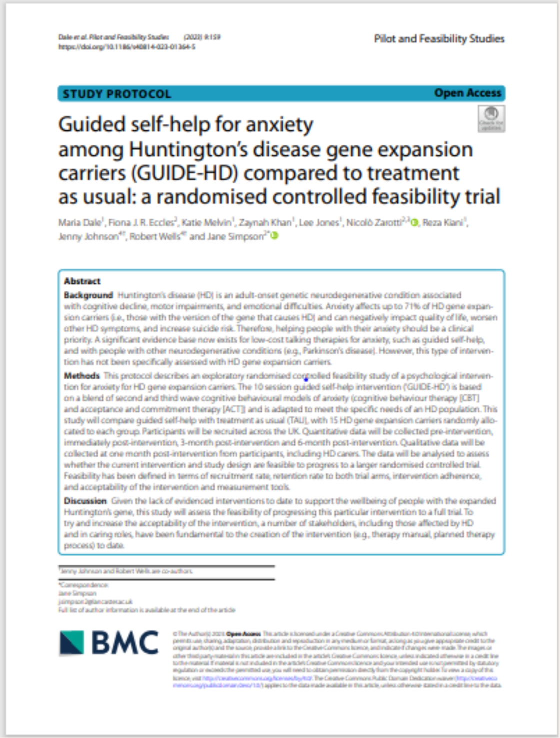
12 people a HD predictive test result had one-off group sessions using a narrative therapy approach

No other RCTs identified , though our group have registered a protocol for a feasibility trial

Dale, Maria et al. “Guided self-help for anxiety among Huntington's disease gene expansion carriers (GUIDE-HD) compared to treatment as usual: a randomised controlled feasibility trial.” *Pilot and feasibility studies* vol. 9,1 159. 12 Sep. 2023, doi:10.1186/s40814-023-01364-5

- 25 participants were recruited (22 HD participants, and 3 carers).
- High levels of acceptability

GUIDE-HD
guided self-help for Huntington’s disease



Dale et al. Pilot and Feasibility Studies (2023) 9:159
https://doi.org/10.1186/s40814-023-01364-5

Pilot and Feasibility Studies

STUDY PROTOCOL Open Access

Guided self-help for anxiety among Huntington’s disease gene expansion carriers (GUIDE-HD) compared to treatment as usual: a randomised controlled feasibility trial

Maria Dale[1], Fiona J. R. Eccles[2], Katie Melvin[1], Zaynah Khan[1], Lee Jones[1], Nicolò Zarotti[2,3], Reza Kiani[1], Jenny Johnson[4†], Robert Wells[4†] and Jane Simpson[2*]

Abstract

Background Huntington’s disease (HD) is an adult-onset genetic neurodegenerative condition associated with cognitive decline, motor impairments, and emotional difficulties. Anxiety affects up to 71% of HD gene expansion carriers (i.e., those with the version of the gene that causes HD) and can negatively impact quality of life, worsen other HD symptoms, and increase suicide risk. Therefore, helping people with their anxiety should be a clinical priority. A significant evidence base now exists for low-cost talking therapies for anxiety, such as guided self-help, and with people with other neurodegenerative conditions (e.g., Parkinson’s disease). However, this type of intervention has not been specifically assessed with HD gene expansion carriers.

Methods This protocol describes an exploratory randomised controlled feasibility study of a psychological intervention for anxiety for HD gene expansion carriers. The 10 session guided self-help intervention (‘GUIDE-HD’) is based on a blend of second and third wave cognitive behavioural models of anxiety (cognitive behaviour therapy [CBT] and acceptance and commitment therapy [ACT]) and is adapted to meet the specific needs of an HD population. This study will compare guided self-help with treatment as usual (TAU), with 15 HD gene expansion carriers randomly allocated to each group. Participants will be recruited across the UK. Quantitative data will be collected pre-intervention, immediately post-intervention, 3-month post-intervention and 6-month post-intervention. Qualitative data will be collected at one month post-intervention from participants, including HD carers. The data will be analysed to assess whether the current intervention and study design are feasible to progress to a larger randomised controlled trial. Feasibility has been defined in terms of recruitment rate, retention rate to both trial arms, intervention adherence, and acceptability of the intervention and measurement tools.

Discussion Given the lack of evidenced interventions to date to support the wellbeing of people with the expanded Huntington’s gene, this study will assess the feasibility of progressing this particular intervention to a full trial. To try and increase the acceptability of the intervention, a number of stakeholders, including those affected by HD and in caring roles, have been fundamental to the creation of the intervention (e.g., therapy manual, planned therapy process) to date.

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† Jenny Johnson and Robert Wells are co-authors.

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BMC

03

Psychological interventions across HD Stages (clinical experience)

HD Stages

1

At-risk

Could be 50% through a parent, or 25% through a grandparent

2

Pre-manifest

Who have the gene but don't have symptoms

3

Early manifest

4

Mid-manifest

5

Advanced manifest and end-of life

At-risk stage

Common psychological issues	Considerations and potential approaches
• Concerns about having the gene expansion, but not wishing to be tested • Living as if they have the genetic expansion – HD overshadowing their life and choices • Anxiety and low mood associated with HD threat • Concurrently being in caring role for other family members with HD, while having own risk • Intergenerational impact of HD	• **Giving people space to discuss their concerns** • **Signpost to HDA and HDYO for additional support** • **Liaison with Genetic Counselling** • Psychoeducation • Psychological therapies – aim to increase coping (taps in the stress bucket) • A range of evidence-based psychological approaches could be used, if applied to the circumstances of living at-risk

Pre-manifest stage

Common psychological issues	Considerations and potential approaches
• Adjusting to a positive predictive test. While many cope initially, can be extremely difficult for some people • Anxiety about future decline and dying, worries about impact on children/family and telling others about HD • Grief about familial loss • Hypervigilance to signs of HD starting (vicious cycle of anxiety) • Suicidal thinking, low mood, people raise ideas about euthanasia / taking own life in the future when their symptoms become advanced	• **Giving them space to discuss their concerns** • **Provide support and advice about telling family and children** • **Explore the opportunity of getting involved in research** • **Adapted psychological therapies across a range of approaches** ▪ e.g., CBT, Psychoeducation, ACT, mindfulness family/systemic therapy, narrative therapy, Compassion Focused Therapy (CFT) ▪ Advance decision planning

GUIDE-HD

guided self-help for Huntington's disease

- A blend of traditional CBT and ACT for anxiety
- HD-specific guided self-help workbooks and a therapist manual were developed
- individual preferences, participants will receive either a weekly telephone, video call, or email-guided psychological support across 10 sessions (aimed at weekly sessions lasting one hour).

The GUIDE-HD study: A randomised controlled feasibility trial comparing guided self-help for anxiety among Huntington's disease gene expansion carriers compared to treatment as usual.

Maria Dale[1], Fiona Eccles[2], Katie Melvin[1], Zaynah Khan[1], Lee Jones[1], Katie Newcombe[1], Jordan Rainbird[1], Nicolò Zarotti[2], Reza Kiani[1], Jenny Johnson[1], Robert Wells[1], Hugh Rickards[3] &, Jane Simpson[2]

1 Leicestershire Partnership NHS Trust, 2 Lancaster University, 3 Birmingham and Solihull Mental Health NHS Foundation Trust

BACKGROUND & INTERVENTION

Psychological interventions for people affected by Huntington's disease are in their infancy. People with HD commonly experience anxiety and, within the general population, a significant evidence base exists for talking therapies such as guided self-help. GUIDE-HD is a novel guided self-help programme based on cognitive behavioural and acceptance and commitment therapy (CBT and ACT). GUIIDE-HD involves 10 sessions with a facilitator and accompanying workbooks (Figure 1). Three caregiver sessions are also offered to supplement learning and skills development. GUIDE-HD has been developed in collaboration with people affected by HD and the intervention is designed to meet the specific needs of an HD population. For example, therapeutic materials with examples of HD-specific experiences of anxiety, explanations of how anxiety relates to HD, and tailoring established therapeutic strategies to HD.

OBJECTIVES

The overall aim of is to assess the feasibility of a randomised clinical trial (RCT) evaluating the clinical effectiveness of a psychological intervention, compared to usual care, in reducing anxiety in individuals with pre-manifest and early-stage HD.

METHODS

The study compares guided self-help with treatment as usual. Fifteen HD gene expansion carriers will be randomly allocated to each group. Participants are currently being recruited across the UK. Using both quantitative and qualitative methods, data will be analysed to assess whether the current intervention and study design meets pre-determined criteria that would indicate feasibility to progress to a larger RCT. The trial has been pre-registered.

RESULTS

The study is currently ongoing, and results will be published following the trial completion by the end of 2024.

CONCLUSIONS

Given the current paucity of evidenced psychological interventions for people with the expanded Huntington's gene, GUIDE-HD will assess the feasibility of progressing this current intervention to a full trial.

Figure 1. Example covers of the GUIDE-HD books.

GUIDE-HD
guided self-help for Huntington's disease

Intervention

GUIDE-HD
Welcome to GUIDE-HD for anxiety
Book 1

GUIDE-HD
Understanding anxiety
Book 2
a guide to help those with Huntington's disease

GUIDE-HD
HD and Anxiety
Book 3
a guide to help those with Huntington's disease

GUIDE-HD
Values
Book 4
a guide to help those with Huntington's disease

GUIDE-HD
Relaxation techniques
Book 5
a guide to help those with Huntington's disease

GUIDE-HD
Facing our fears
Book 6
a guide to help those with Huntington's disease

GUIDE-HD
Acceptance and willingness
Book 7
a guide to help those with Huntington's disease

GUIDE-HD
Dealing with anxious thoughts
Book 8
a guide to help those with Huntington's disease

GUIDE-HD
Problem solving
Book 9
a guide to help those with Huntington's disease

GUIDE-HD
Bringing it all together
Book 10

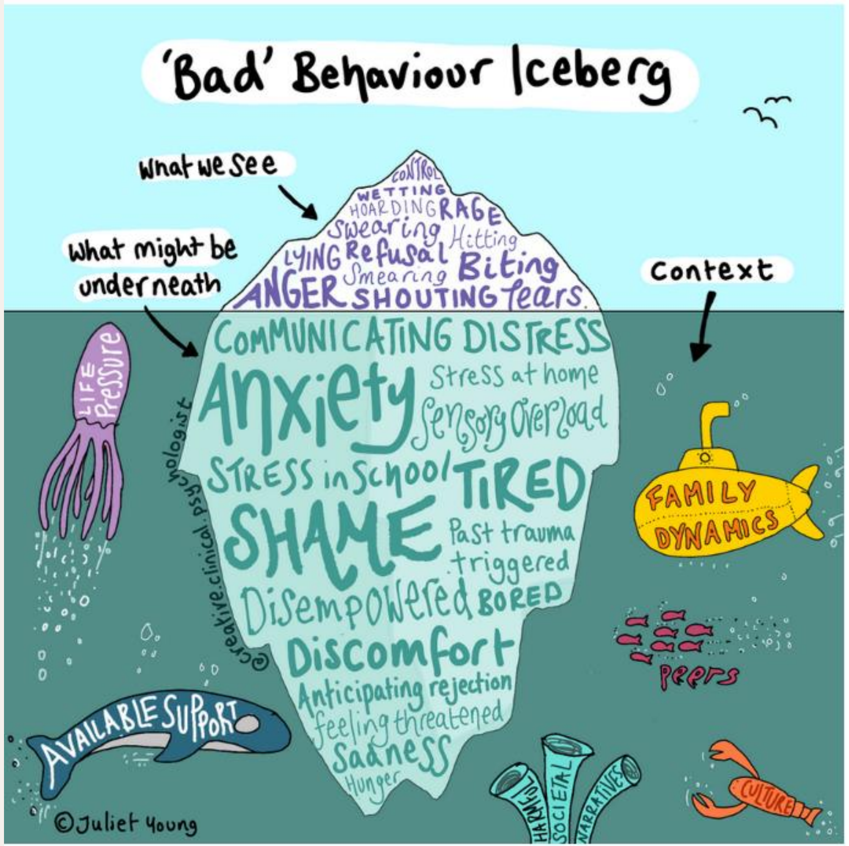
Early stage

Common psychological issues	Psychological considerations and potential approaches
• Coping with the clinical diagnosis • Adjustment to increasing symptoms, loss of roles (and other family members), grief, frustration/ anger (“why me?” thinking) • Guilt – passing on to children, change in family dynamics • Being a burden to others, changes in self-confidence, change in identity • Can become self-conscious/embarrassed about movements (feel more clumsy) • Increasing difficulties with memory and concentration • Mood fluctuations, feeling more short tempered	• Therapeutic approaches mentioned before CBT, Psychoeducation, ACT, mindfulness family/systemic therapy, narrative therapy, Compassion Focused Therapy (CFT) • - may need to start adapting therapy further to meet the needs of those with cognitive decline • **Try to elicit hope** – research activity/engagement – can help at least for guilt about passing on to next generation • Helping people to deal with loss and grief – anticipated as well as past • **Discussions around future planning**

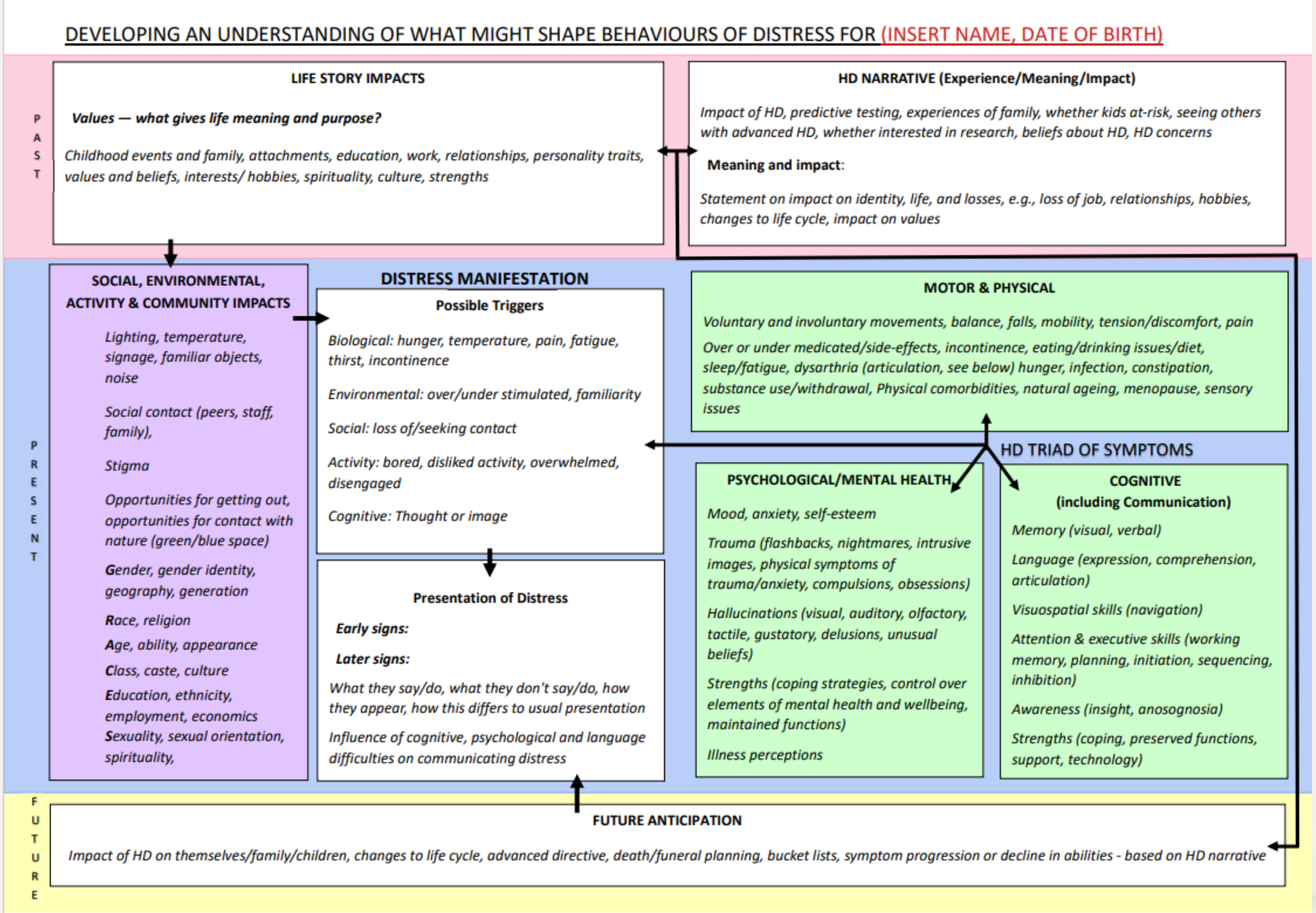
Mid-manifest stage

Common psychological issues	Considerations and potential approaches
• Behaviours that can be distressing or frustrating (to the individual and others) – aggression, fixation/ obsession, lack of initiation, perseveration • Reduced self-awareness of difficulties, risk issues can increase e.g. falls, burning themselves • Perceptions of self – adjusting to new identity – coping with mobility aids, loss of independence, sexual relationship/intimacy • People can often become distressed and embarrassed by physical and cognitive changes; perceived as drunk	• **Formulation-based approach** • **Relaxation methods** • **ACT** informed work – meaningful activity based on person's values – focus on what people can do – help acceptance and adjustment to changes • **Behavioural approaches** e.g., PBS • **Life story** work • **Focus on supporting partners/carers –** psychoeducation, helping carers care for themselves, "living grief", "ambiguious loss" - carers groups

Mid-manifest stage



Clinical Formulation (e.g., Dale et al 2022)



DEVELOPING AN UNDERSTANDING OF WHAT MIGHT SHAPE BEHAVIOURS OF DISTRESS FOR (INSERT NAME, DATE OF BIRTH)

PAST

LIFE STORY IMPACTS

Values — what gives life meaning and purpose?

Childhood events and family, attachments, education, work, relationships, personality traits, values and beliefs, interests/ hobbies, spirituality, culture, strengths

HD NARRATIVE (Experience/Meaning/Impact)

Impact of HD, predictive testing, experiences of family, whether kids at-risk, seeing others with advanced HD, whether interested in research, beliefs about HD, HD concerns

Meaning and impact:

Statement on impact on identity, life, and losses, e.g., loss of job, relationships, hobbies, changes to life cycle, impact on values

PRESENT

SOCIAL, ENVIRONMENTAL, ACTIVITY & COMMUNITY IMPACTS

Lighting, temperature, signage, familiar objects, noise

Social contact (peers, staff, family),

Stigma

Opportunities for getting out, opportunities for contact with nature (green/blue space)

***G**ender, gender identity, geography, generation*

***R**ace, religion*

***A**ge, ability, appearance*

***C**lass, caste, culture*

***E**ducation, ethnicity, employment, economics*

***S**exuality, sexual orientation, spirituality,*

DISTRESS MANIFESTATION

Possible Triggers

Biological: hunger, temperature, pain, fatigue, thirst, incontinence

Environmental: over/under stimulated, familiarity

Social: loss of/seeking contact

Activity: bored, disliked activity, overwhelmed, disengaged

Cognitive: Thought or image

Presentation of Distress

Early signs:

Later signs:

What they say/do, what they don't say/do, how they appear, how this differs to usual presentation

Influence of cognitive, psychological and language difficulties on communicating distress

MOTOR & PHYSICAL

Voluntary and involuntary movements, balance, falls, mobility, tension/discomfort, pain

Over or under medicated/side-effects, incontinence, eating/drinking issues/diet, sleep/fatigue, dysarthria (articulation, see below) hunger, infection, constipation, substance use/withdrawal, Physical comorbidities, natural ageing, menopause, sensory issues

HD TRIAD OF SYMPTOMS

PSYCHOLOGICAL/MENTAL HEALTH

Mood, anxiety, self-esteem

Trauma (flashbacks, nightmares, intrusive images, physical symptoms of trauma/anxiety, compulsions, obsessions)

Hallucinations (visual, auditory, olfactory, tactile, gustatory, delusions, unusual beliefs)

Strengths (coping strategies, control over elements of mental health and wellbeing, maintained functions)

Illness perceptions

COGNITIVE (including Communication)

Memory (visual, verbal)

Language (expression, comprehension, articulation)

Visuospatial skills (navigation)

Attention & executive skills (working memory, planning, initiation, sequencing, inhibition)

Awareness (insight, anosognosia)

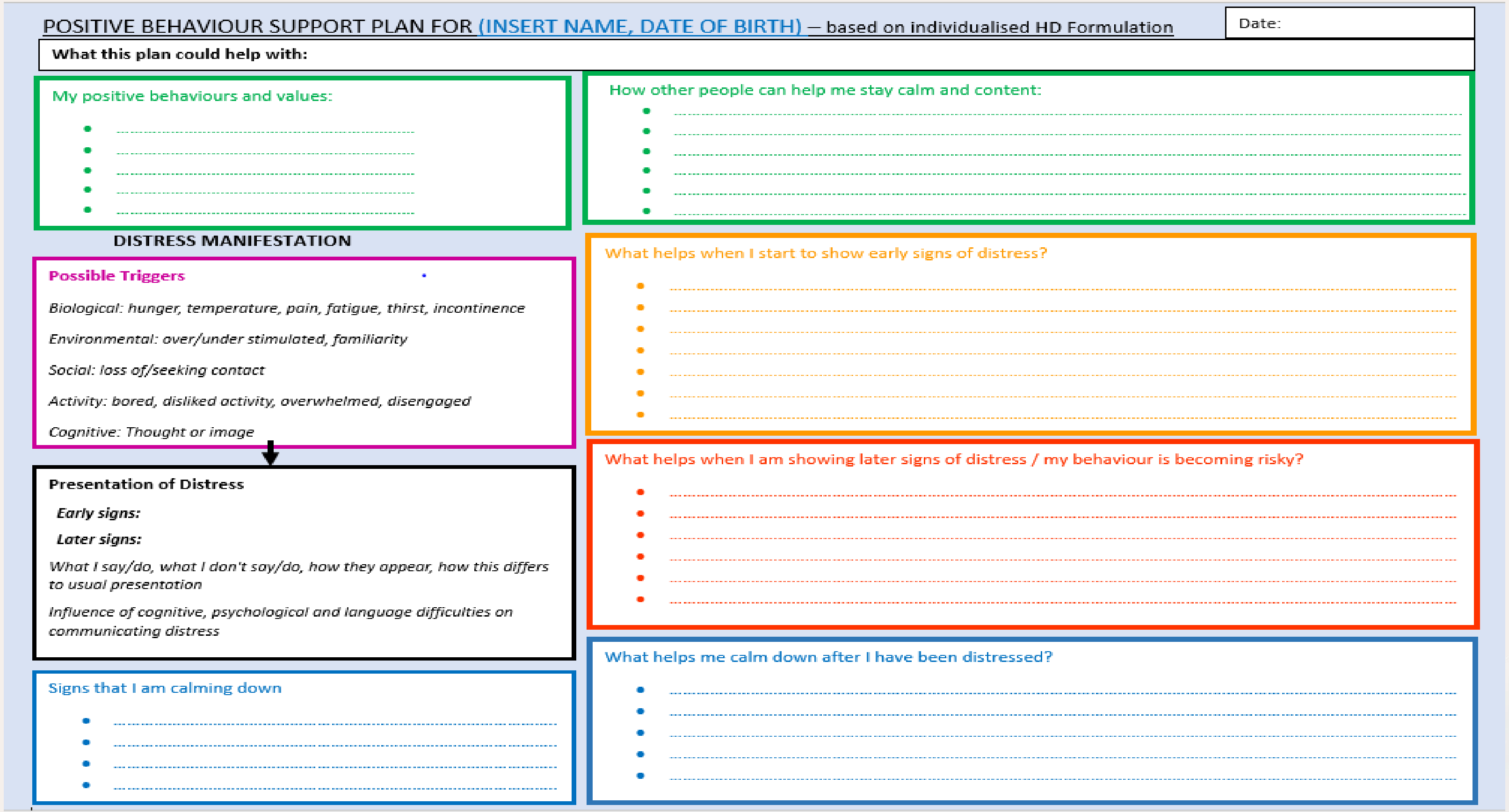
Strengths (coping, preserved functions, support, technology)

FUTURE

FUTURE ANTICIPATION

Impact of HD on themselves/family/children, changes to life cycle, advanced directive, death/funeral planning, bucket lists, symptom progression or decline in abilities - based on HD narrative

Positive Behaviour Support



POSITIVE BEHAVIOUR SUPPORT PLAN FOR (INSERT NAME, DATE OF BIRTH) – based on individualised HD Formulation

Date:

What this plan could help with:

My positive behaviours and values:

- ....
- ....
- ....
- ....
- ....

How other people can help me stay calm and content:

- ....
- ....
- ....
- ....
- ....
- ....

DISTRESS MANIFESTATION

Possible Triggers

Biological: hunger, temperature, pain, fatigue, thirst, incontinence

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↓

Presentation of Distress

Early signs:

Later signs:

What I say/do, what I don't say/do, how they appear, how this differs to usual presentation

Influence of cognitive, psychological and language difficulties on communicating distress

Signs that I am calming down

- ....
- ....
- ....
- ....

What helps when I start to show early signs of distress?

- ....
- ....
- ....
- ....
- ....
- ....
- ....

What helps when I am showing later signs of distress / my behaviour is becoming risky?

- ....
- ....
- ....
- ....
- ....
- ....

What helps me calm down after I have been distressed?

- ....
- ....
- ....
- ....
- ....

Reflective exercise

- Experiential exercise… think about the things you value most in your life, or what you like to do in a typical day.
- Now imagine having moderate to advanced HD – how would those things be affected?
- How would you feel?

Advanced manifest stage and end-of-life

Common psychological issues	Considerations and potential approaches
• Needing 24 hour care, can be periods of confusion, screaming, agitation, behaviours that others find distressing e.g. hitting out during personal care or mealtimes • Often can be physically poorly and in pain, low mood	**• A whole MDT approach** **• Relaxation methods,, behavioural approaches such as Positive Behaviour Support (PBS), and functional analysis, life story work** **• Supporting family members** **• Team formulation sessions, reflective practice, staff consultation** **• Focus on dignity, interests, activities based on values and ensure those are central.** **• End of life discussions**

Adaptations to standard therapy

- Formulate the client’s difficulties in terms of past, present and future
- Understand neurocognitive needs – potentially do a cognitive test before if needed and adapt therapy accordingly
- Break information down into small chunks, allow time for processing information, provide pictures, have written or audio resources to aid memory skills
- 1:1 therapy not suitable for all, some people can mean they struggle to switch to another topic after difficult things being discussed
- Group work can sometimes be tricky – people at different stages, not ready to hear certain information
- Really try to empathise, how would I feel if it was me?
- Pace sessions, keep shorter
- Try using talking mats/pictures
- Consider therapeutic activity that isn’t reliant on verbal skills: e.g., mindfulness, relaxation
- Carer sessions

04

Psychological services for people with HD

Psychological intervention pathways for pwHD

- Not clear and no consistency around the UK. Will largely depend on who the referrer is and the perceived level of complexity. In mainstream services there is a hierarchy of services, but this is not defined in HD
- GP can refer to generic adult mental health services or to neuropsychology (where available), may need to see a neurologist before accessing a neuropsychologist. Neuropsychology services often focus more on undertaking neurocognitive assessments rather than having capacity for delivering mental health interventions
- Referrals to IAPT –NB: not always successful or useful, variable reports

Psychological intervention pathways for pwHD

- Specialist hospital services – more contact at manifest stage. Could refer to hospital psychology services – e.g., health/medical psychology or neuropsychology.
- Support at later stage possibly from young onset dementia service
- Specialist NHS HD centres sometimes have a psychologist as part of the multi-disciplinary team, e.g., Leicester
- Specialist services run by charities sometimes include psychology
- Given difficulties accessing psychology, many pwHD seek private psychotherapy/counselling

Psychological intervention pathways for pwHD

- HDA – psychologically informed 1:1 support and groups - “Keeping Yourself In Mind” – online psychological groups based on an ACT approach
- The Brain Charity (https://www.thebraincharity.org.uk) offers some time-limited free counselling

Lastly - what can we all do?

- Get to know the person, their history, what might be likely to cause them distress, their signs of comfort.

- Get to know what is important to them and bring these into their care whenever possible.

- Get to know HD and don’t assume is it something in the background – throughout the whole disease stage, understanding the implications this has for people will go a long way

Thanks!

Do you have any questions?

Please also feel free to get in touch:
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