

Hydrocephalus: Cognitive and Behavioural Interventions

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Overview

- Aetiology and classification
- Outcome improvements
- Cognitive and behavioural difficulties
- Cognitive assessment
- Cognitive profiles
- Triangulating data: ASBAH funded project
- Implications
- Interventions with emphasis on insight
- Need for further research



Aetiology and classification

Hydrocephalus (communicating/non-communicating)

Congenital

- Spina Bifida
- Arnold-Chiari/Dandy-Walker malformations
- Stenosis of the aqueduct of Sylvius

Acquired

- Intraventricular haemorrhage
- Infections (meningitis, encephalitis)
- Vascular abnormalities
- Tumours or cysts
- Traumatic brain injury

Spina bifida (co-occurrence major confounding factor)

Neural tube defects (failure to close) associated with homocysteine-folate pathway - with large variation in lesion location and severity affecting mobility, renal and bowel function

Neuroanatomical consequences

Accumulation of CSF and ventromegaly:

- Compresses brain tissue in a characteristic posterior to frontal pattern
- Stretches and damages neural fibres
- Distorts and disrupts cerebral vascular system
- **Disrupts CSF production and absorption**

Concentration of important chemicals and cerebrospinal fluid is altered dependent upon location of blockage - implicating CSF composition as a major contributory factor for cognitive function

(Miyan et al, 2003; Cains et al, 2009)

Neuroanatomical links to
cognitive and behavioural function

(Biopsychosocial approach)

Neuronal transmission

Damaged neuronal transmission associated with:

- reduced processing speed
- impaired input and output from the hippocampus (linked to memory)
- impairments in information transmission between hemispheres

Posterior to frontal pattern

- Posterior cortex more vulnerable, particularly on the right side affecting attentional system
- Reorganisation of frontal areas can result in cortical thickening and can affect executive functioning and result in organisational difficulties, impulsivity and mood swings

CSF disruption

Concentration of important chemicals and cerebrospinal fluid is altered dependent upon location of blockage - implicating CSF composition as a major contributory factor for cognitive function

(Miyan et al, 2003; Cains et al, 2009)

Creates unifying theory of hydrocephalus and spina bifida whereby circulating CSF around the outer cortical layers is disrupted, associated with impairments in cognitive functioning (Williams, 2005)

All neuroanatomical consequences are dependent on numerous factors including age, magnitude, aetiology, duration, intervention (Brewer et al, 2001)

Incidence

- Estimated prevalence of neonatal HC is 1 in 1000 births (Buxton, 2007) Rates can vary between (0.41-0.81)

Up until 1950s outcome poor

- with less than 50% of individuals surviving
- Less than a third reaching adult life
- Two thirds with impairments in intellectual functioning

Improved outcomes

Associated with:

- Early diagnosis and early intervention
- Advances in treatments
- Antibiotic - impregnated shunt materials
- Heightened awareness

Results in:

- Improvements in intellectual functioning

However - subtle cognitive difficulties can remain

Cognitive assessment

Assess:

- processing speed
- different components of attention e.g. sustained attention, selective attention
- learning ability
- different components of memory e.g long term and short term memory
- executive functioning
- language production **and** comprehension

Note - wide variability - impact for education

Cognitive tasks

- Processing speed and attentional flexibility and assessed using Trail making task
- Language understanding assessed using WAIS vocabulary and DKEFS Proverbs
- Executive functioning assessed with verbal fluency tasks
- Learning and memory functioning assessed using Hopkins list learning and WMS story recall

Intriguing modal cognitive profile

Strengths

- Elaborate use of language
- Verbal IQ
- Working memory performance
- Long term memory performance
- Forthright manner

Weaknesses

- Poor comprehension
- Poor calculation
- Poor learning
- Poor short term memory
- Poor planning
- Poor strategy use
- Attentional deficits

Implications

- Self - perception
- Perception of carers and professionals
- Essential for education programme
- Essential for goals and outcomes

Cognitive assessment provides baseline and can be used to monitor change associated with shunt blockage

Behavioural assessment

Assess:

- Anxiety and/or depression
- Anger, frustration, irritability
- Emotional regulation
- Empathy
- Insight
- Situational difficulties e.g. novelty
- Coping strategies
- Self-esteem and self-perception
- Fears, hopes and dreams

Behavioural assessment

- Hospital Anxiety and Depression Scale
 - Behavioural assessment questionnaire
 - Observation
 - Self report
 - Carer report
-
- Reveals need for more detailed assessment tools

Psychosocial consequences

- Mood disorders including depression, anxiety, mood swings, poor emotional regulation
- Lowered self-esteem
- Perceived difficulties
- Actual behavioural difficulties
- Disinhibition
- Agitation/frustration
- Apathy
- Impact on family, friends, peers, educators and carers (esp. throughout child development and adolescence)

Rationale for our study

- Individuals with HC often have cognitive impairments which range from subtle to disabling
- Anecdotally, insight varies as does recognition of difficulties by family, friends and employers
- Focus on
 - cognitive and behavioural difficulties and more crucially their impact on day-to-day life
 - information regarding effective cognitive and social strategies
- 3-stage clinical project funded by ASBAH (Iddon, Loveday, Edginton, Morgan & Pickard)
- Aims to formulate useful guidelines for dissemination to clients, their family, their employers and their clinicians

Stage 1

Development of a new questionnaire to assess:

- the impact of cognitive and behavioural difficulties on everyday lives in adults with hydrocephalus
- the insight/perception of individuals with HC and how this links to others' perceptions

Plan to make questionnaire freely available to use as a standardised clinical tool for adults with HC

CONFIDENTIAL SURVEY- SERVICE USER

Date today: _____

We are conducting a survey to find out more about certain aspects of hydrocephalus and spina bifida. We want to get a better understanding of some of the difficulties you may have and in particular we want to know whether you are receiving the right kind of support and guidance. For section 1 to 4 we would like you to read the statements given and decide how true they are of you. There are no right or wrong answers and it is important that you answer all questions. If you feel that no answers are quite right then please just choose the answer that you think is closest. For section 5 to 8 we are asking some general questions. Please do your best to answer all of these. If you would like any help with writing some of the answers or would prefer not to complete the questionnaire please speak to the person who gave you the questionnaire.

This questionnaire is entirely confidential and will not directly affect your treatment at Chelsea & Westminster. However, we hope it will provide us with the information we need to continue improving services here and throughout the country and that ultimately you and others will benefit.

Please FILL IN THE CIRCLE that best represents your answer to HOW TRUE the following statements are about you.

	Not True	Partly true	Quite true	Very true
SECTION 1: MEMORY AND CONCENTRATION				
1. I am forgetful	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
2 I find it easy to recall passwords and PIN numbers	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
3. I am not easily distracted	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
4. I am a quick learner	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
5. I feel that my memory is no worse than most other peoples	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
6. I have difficulties with calculations, such as adding up or subtracting	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
7. I have difficulty concentrating (E.G. on a TV program)	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
8. I have no difficulty following a conversation	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
9. I get distracted and lose my train of thought	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
10. I generally need things to be explained to me very carefully and sometimes more than once	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
11. I enjoyed learning at school	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
12. At school/college I found studying frustrating and stressful	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
13. At school/college/work I often feel/felt that my achievements did not match my efforts	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
14. I am a slow learner	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>

Questionnaire

Sections

1. Memory & attention/concentration
2. Planning & organization
3. Behaviour
4. Emotional & social
5. Strategies*
6. Service provision*
7. Psychological assessment & treatment*
8. General Questions*

(* indicates qualitative section)

Stage 2

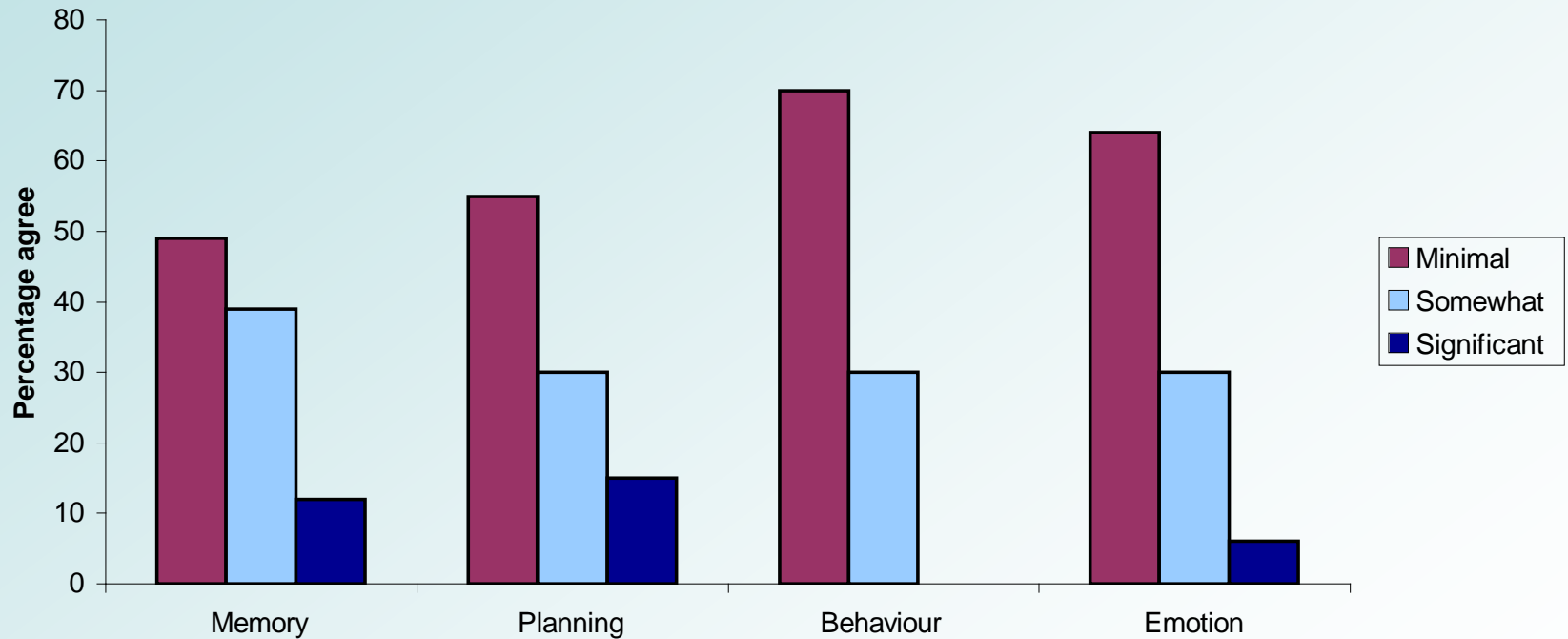
Triangulate data

- Aim to correlate self-reported difficulties with observed difficulties and actual neuropsychological performance on a detailed battery of tasks that assesses memory, attention, planning, language, performance IQ

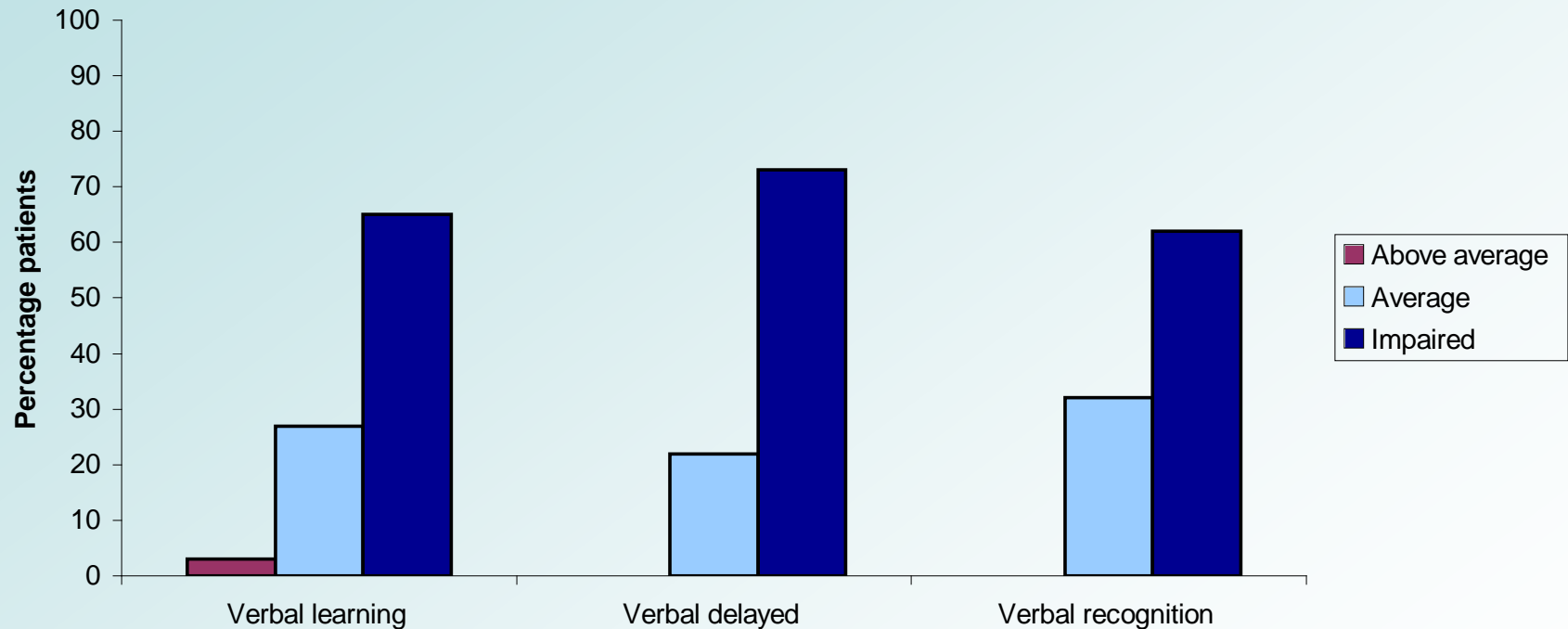
Study design

- Questionnaire data for 80 individuals with HC (with and without SB) and significant other pairs recruited in the multidisciplinary clinic at Chelsea and Westminster
- Neuropsychological data for 30 (14:16 male to female ratio, mean age 36) of these clients to formally assess memory, planning and attention
- More detailed neuropsychological assessments for 30 (14 males; 16 females, mean age 34) of these clients

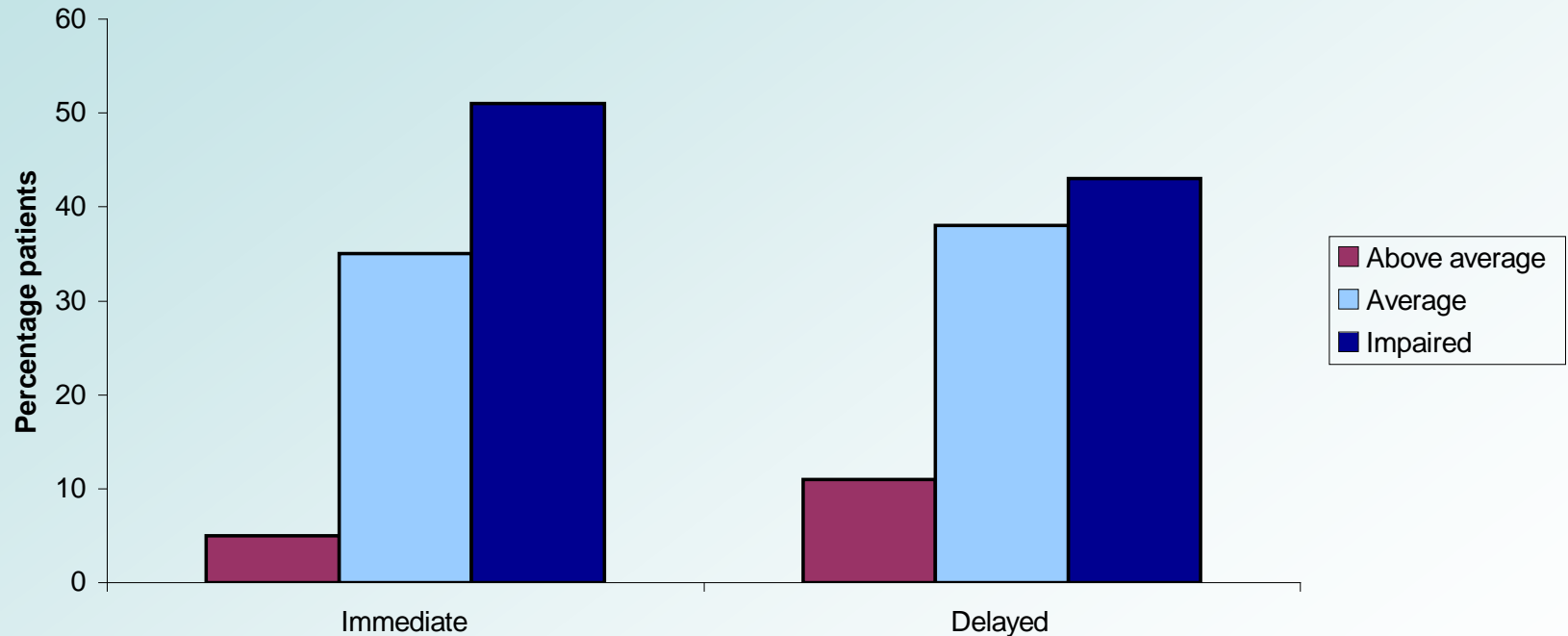
Self-reported difficulties using the Questionnaire (80 participants)



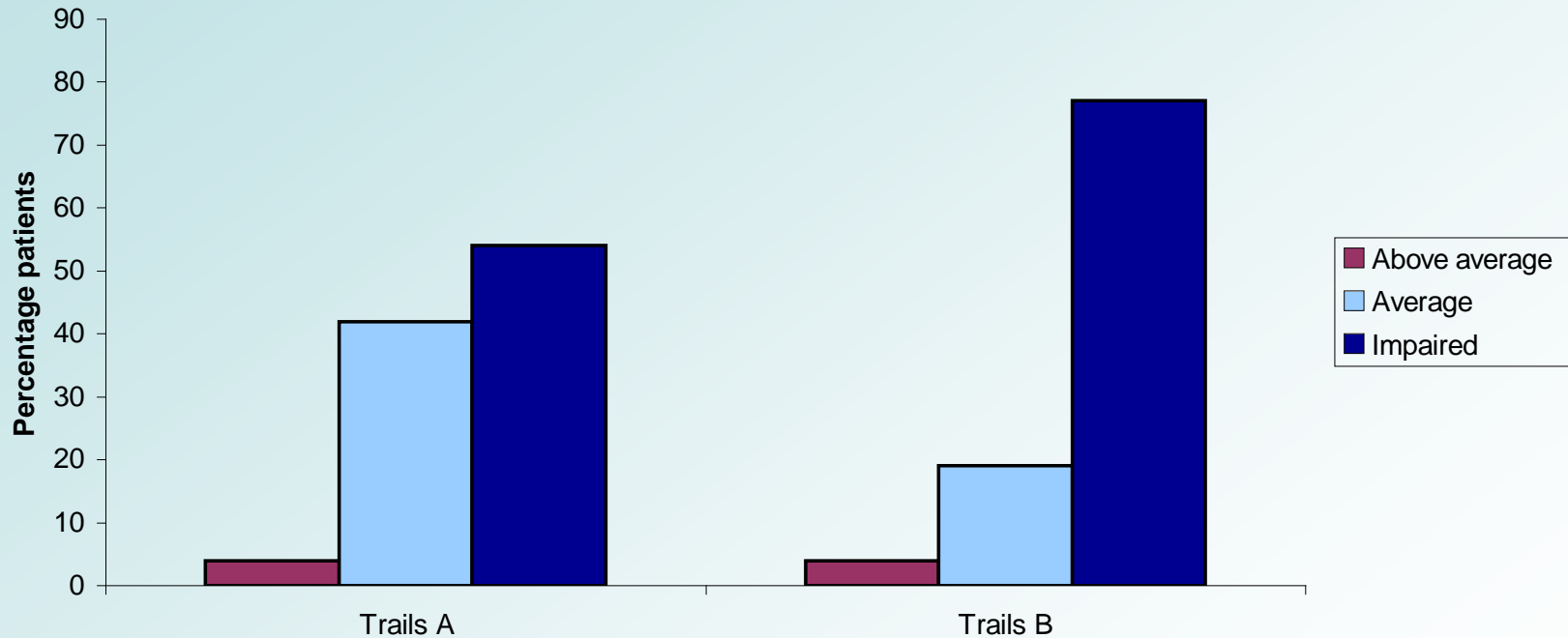
Verbal memory performance on the Hopkins memory task



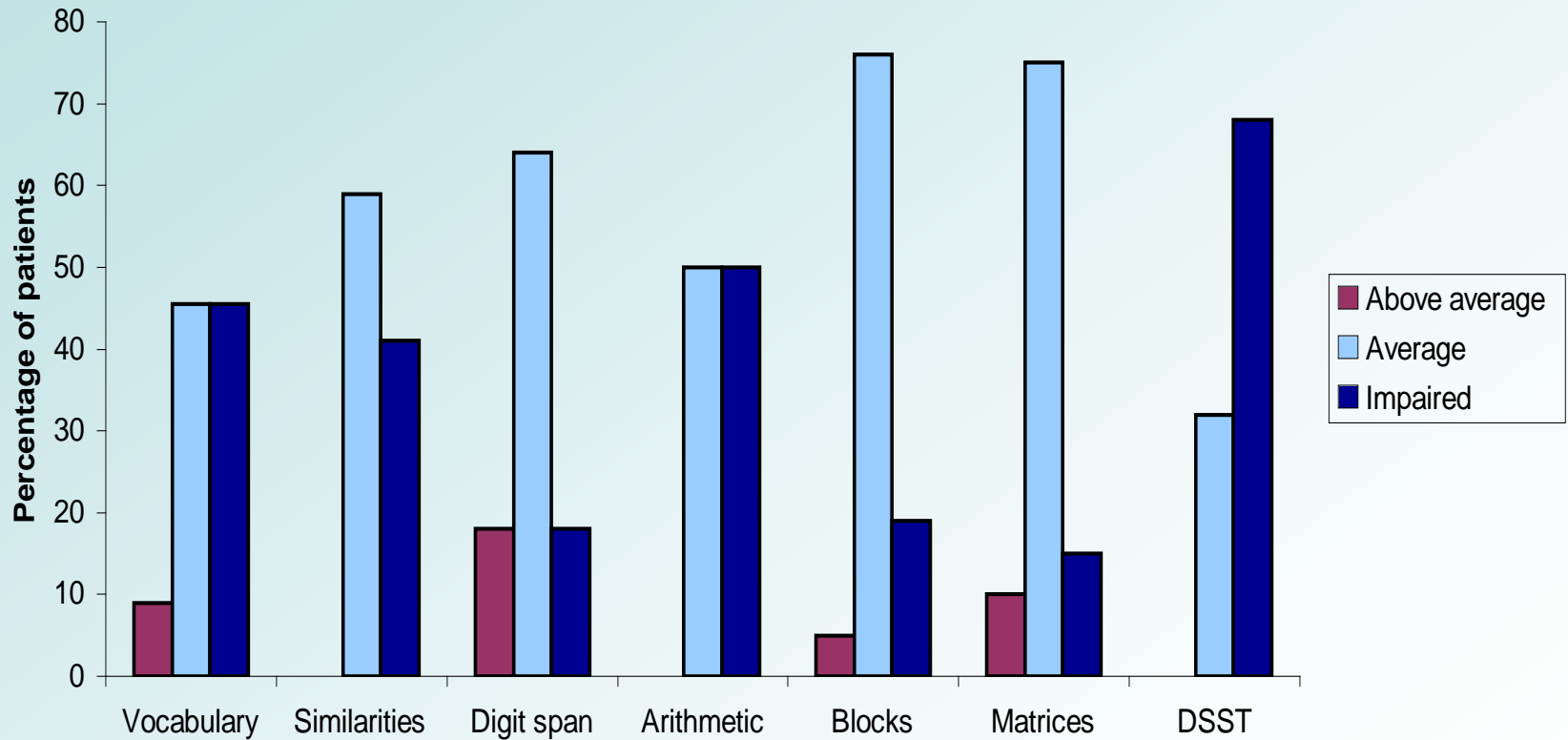
Complex memory performance on Weschler logical story recall (n=37)



Switching performance on the Trail-making task (n=37)



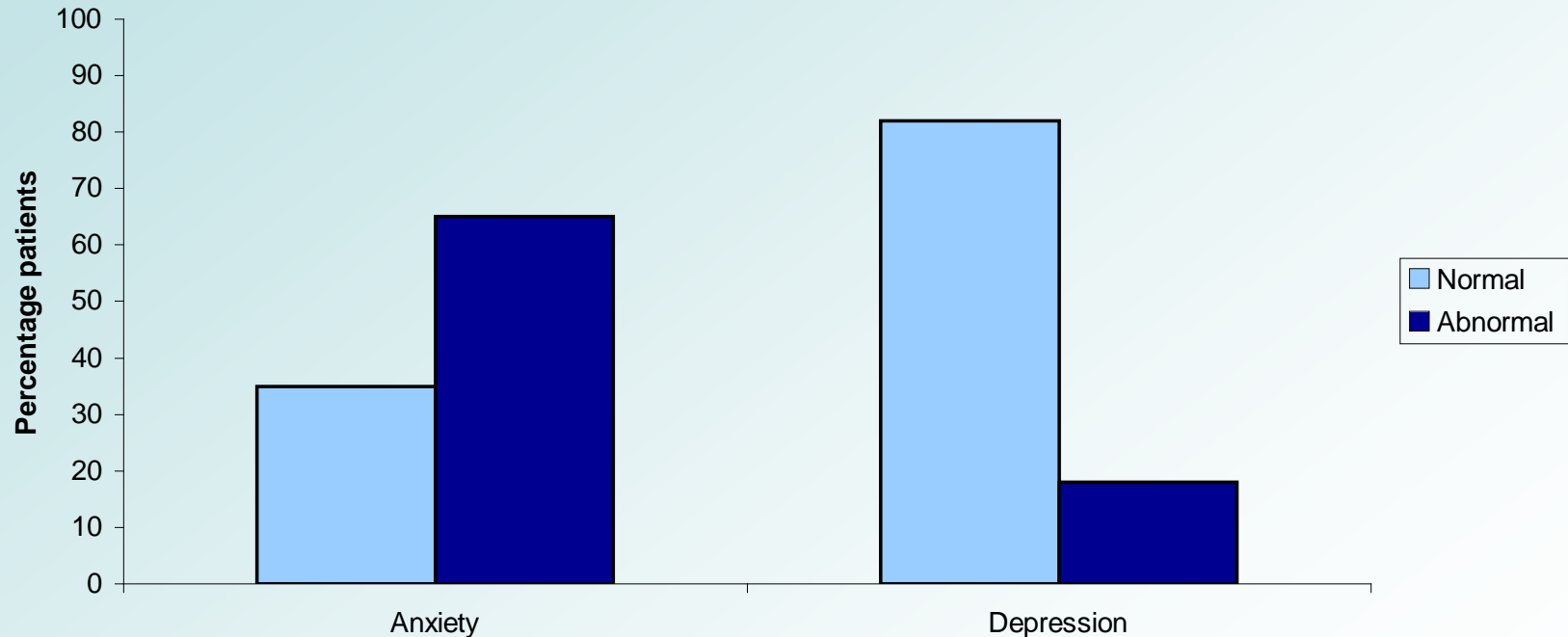
WAIS performance IQ measures (n=24)



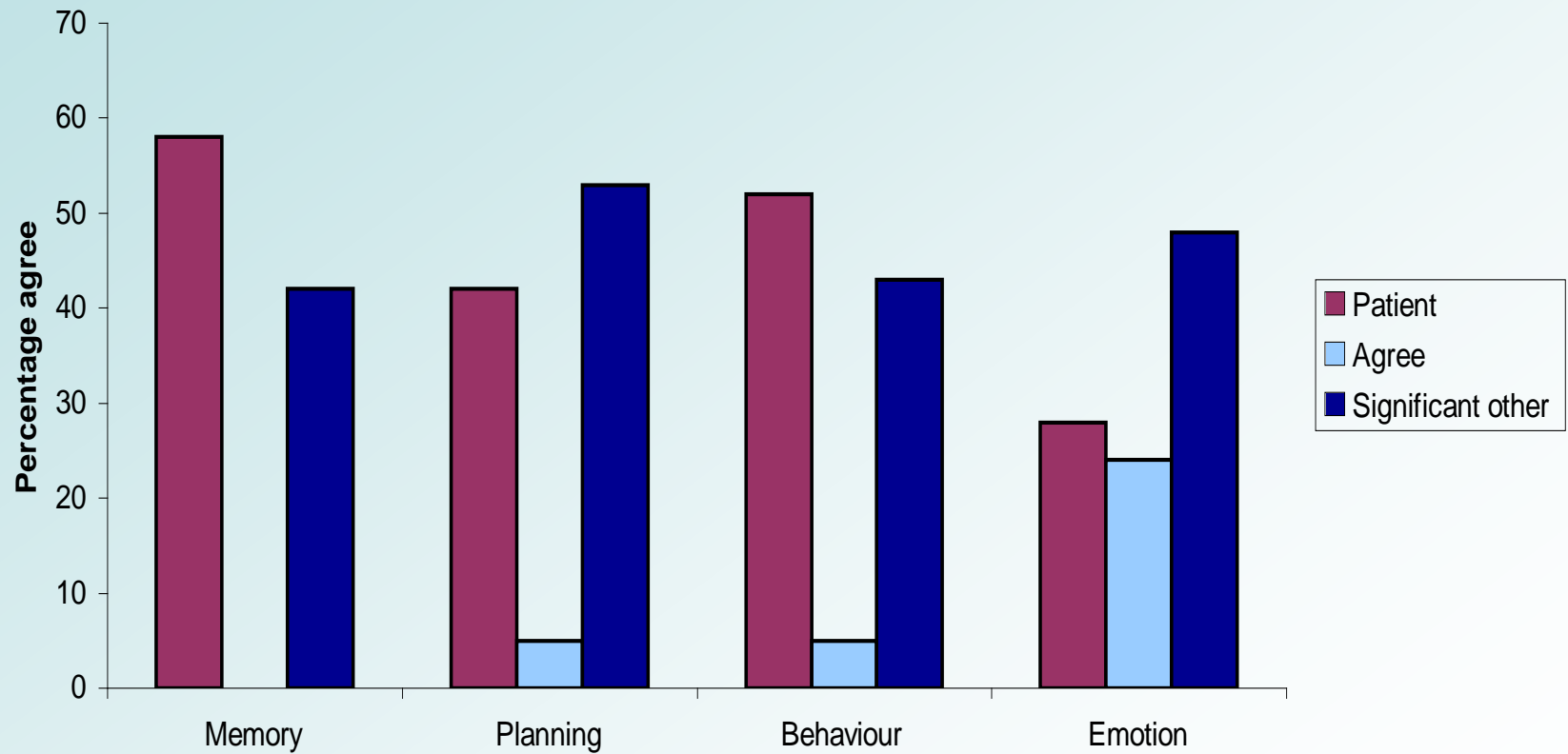
Initial findings

- Minimal memory and attentional difficulties self-reported by individuals with HC
- Contrasts with actual difficulties in verbal memory learning, recall and recognition and attentional switching
- Further contrasts with documented preserved performance IQ measures and working memory performance in the sub group although good use of vocabulary can mask underlying comprehension difficulties

Anxiety and depression levels using the HADS (n=80)



Disparity of insight (n=80)



Summary

- Confirmation of modal profile - strengths and weaknesses in cognitive areas e.g. average working memory in contrast to difficulties with delayed short term memory recall
- Range of difficulties appear to be underestimated by individuals with HC
- Lack of strategy use indicated
- 60% individuals with HC experience increased anxiety
- Bimodal profile of disparity between individuals with HC and their significant others that requires further exploration

Implications

- Cognitive difficulties have wide ranging impact on everyday functioning
- Need to increase understanding of cognitive profile and specific cognitive strengths and weaknesses for individuals with HC, relatives, teachers and employers
- Reduced insight has implications for strategy use and cognitive interventions
- Disparity of insight needs to be further explored
- Questionnaire a useful tool to triangulate questionnaire perspectives with objective neuropsychological data

Interventions

- Primary aim to improve insight
- Individual assessment necessary to identify pattern of strengths and weaknesses
- Assess disparity
- Assess willingness for strategy interventions
- Shared approach to interventions required

- Note lack of systematic evidence-based studies - neuropsychological rehabilitation literature

Cognitive interventions

Insight

- Be aware of strengths and weaknesses, maximise strengths to augment weaknesses
- Encourage self evaluation
- Inform others of cognitive strengths and weaknesses
- Discuss expectations

Information processing strategies

- Provide additional time to allow for slowed processing speed
- Minimise distractions
- Work at own pace and ensure regular breaks are taken
- Ask for material in advance
- Create manageable chunks
- Check understanding
- Awareness of personal strengths and weaknesses

Learning and memory strategies

- Encourage use of external aids e.g. diary, mobile, camera for retrospective & prospective memory
- Use what works best for each individual
- Understand and organise aids effectively
- Use cues, prompts, alarms
- General memory strategies - rehearsal, chunking, imagery, mnemonics

Executive function strategies

- Introduce time management and project management strategies
- Set out structure, set achievable and measurable goals, plan action, evaluate
- Encourage self monitoring and self evaluation
- Ensure provision of support and encouragement

Behavioural interventions

- Insight
- Encourage self-monitoring
- CBT - management of negative perceptions
- Preventive interventions to strengthen coping resources
- Anger management strategies
- Family intervention

Behaviour management

- Ignore negative behaviour if possible
- Reward desired behaviour
- Distraction techniques can be useful
- Maintain consistent approach wherever possible
- Establish appropriate boundaries
- Try to remain calm
- Discussion, negotiation and communication
- Learning process for everyone!

Final thoughts

- Insight for individual, carer and professionals
- Variation requires individualised approach
- Acknowledgement of strengths and weaknesses
- Willingness to incorporate and evaluate strategy
- Dynamic process
- Maximise strengths
- Need for further research and dissemination
- Any other useful strategies?

Acknowledgements

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