What is the neuropsychological impact of Kleine Levin Syndrome?

Two clinical case studies

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Introduction

•Kleine Levin Syndrome (KLS) is a rare sleep disorder with typical onset during adolescence which can profoundly affect cognitive functioning

•characterised by persistent episodic hypersomnia (often sleeping 20 or more hours a day); impaired cognitive and executive functioning; impact on mood including a subjective experience of derealisation; variations in normal appetite and sexual behaviour

•Identified cognitive changes during KLS episodes include confusion and deficits in concentration, attention and memory, though formal cognitive testing has rarely been used.[1] Longer term impact highlights deficits in processing speed and verbal memory with greater impairment in higher frequency, shorter episode KLS[2]

•Aims of project : to generate detailed neuropsychological profiles of young people with KLS 'in' and 'out of' episode to better understand the impact of KLS on their functioning; to identify any specificity of impairments

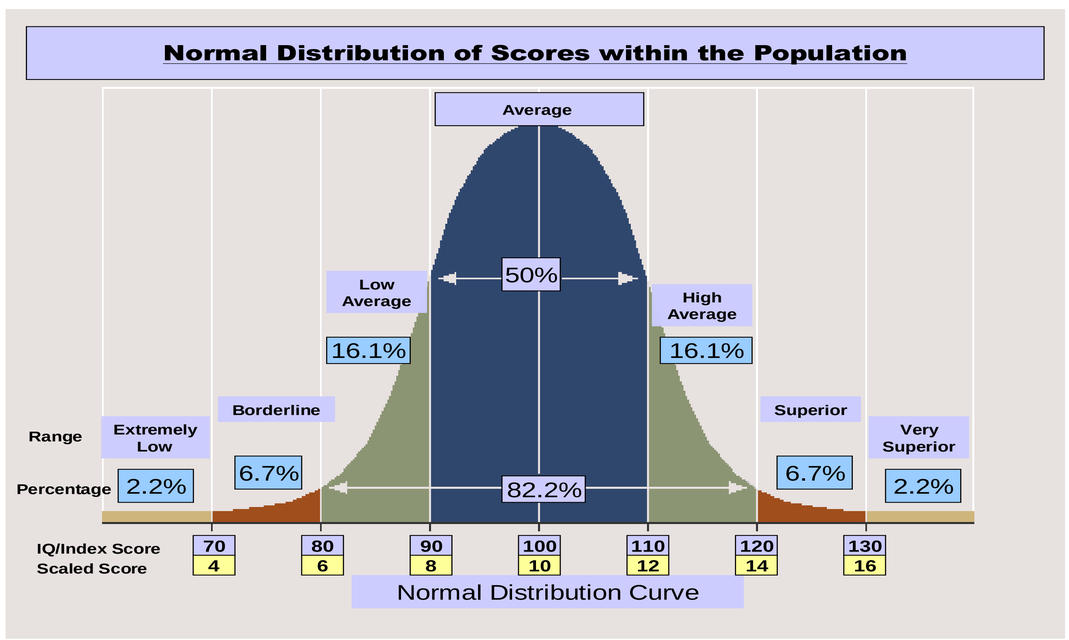
•Here we report the first two cases

Methods

Neuropsychological tasks were selected to assess targeted skills understood to be detrimentally affected during a KLS episode.

- *Processing speed...* coding and symbol search from WAIS
- *Working memory...* digit span from WAIS
- *Executive function...* colour word interference and trail making from D-KEFS
- *Auditory memory...* logical memory and verbal paired associates from WMS
- *Sustained attention...* continuous performance from Conners
- *Motor speed...* pegboard from WRAMVA

Intellectual functioning was assessed out of episode using the WASI; all other tasks were completed both 'in' and 'out of' KLS episode, with a minimum six month interval between assessments.

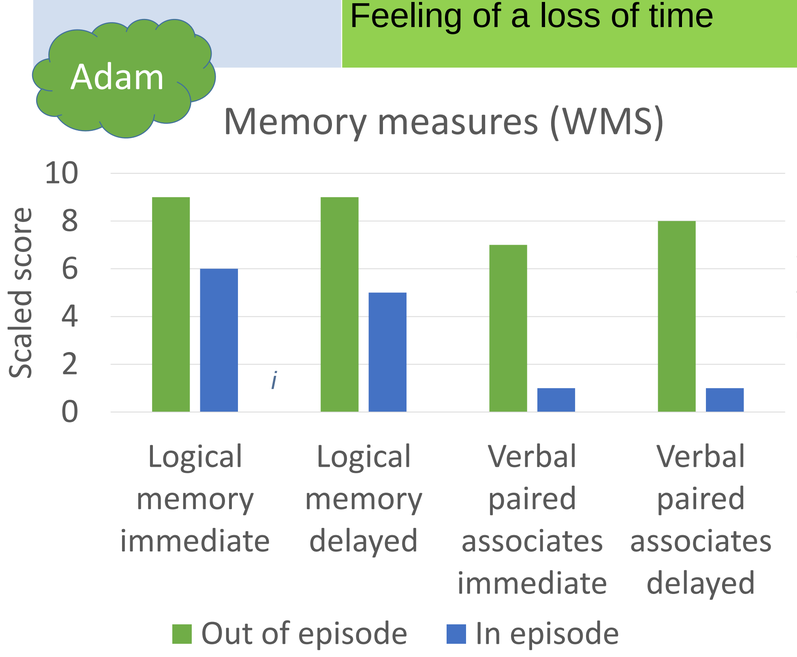


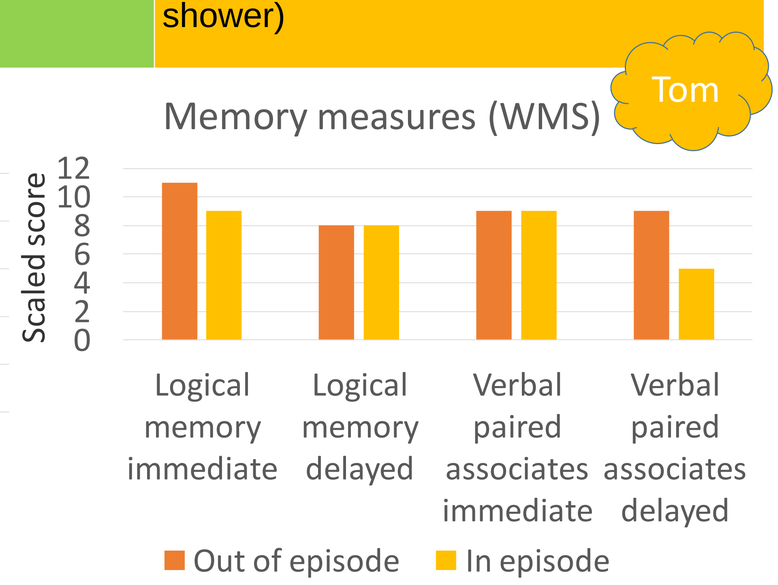
Results

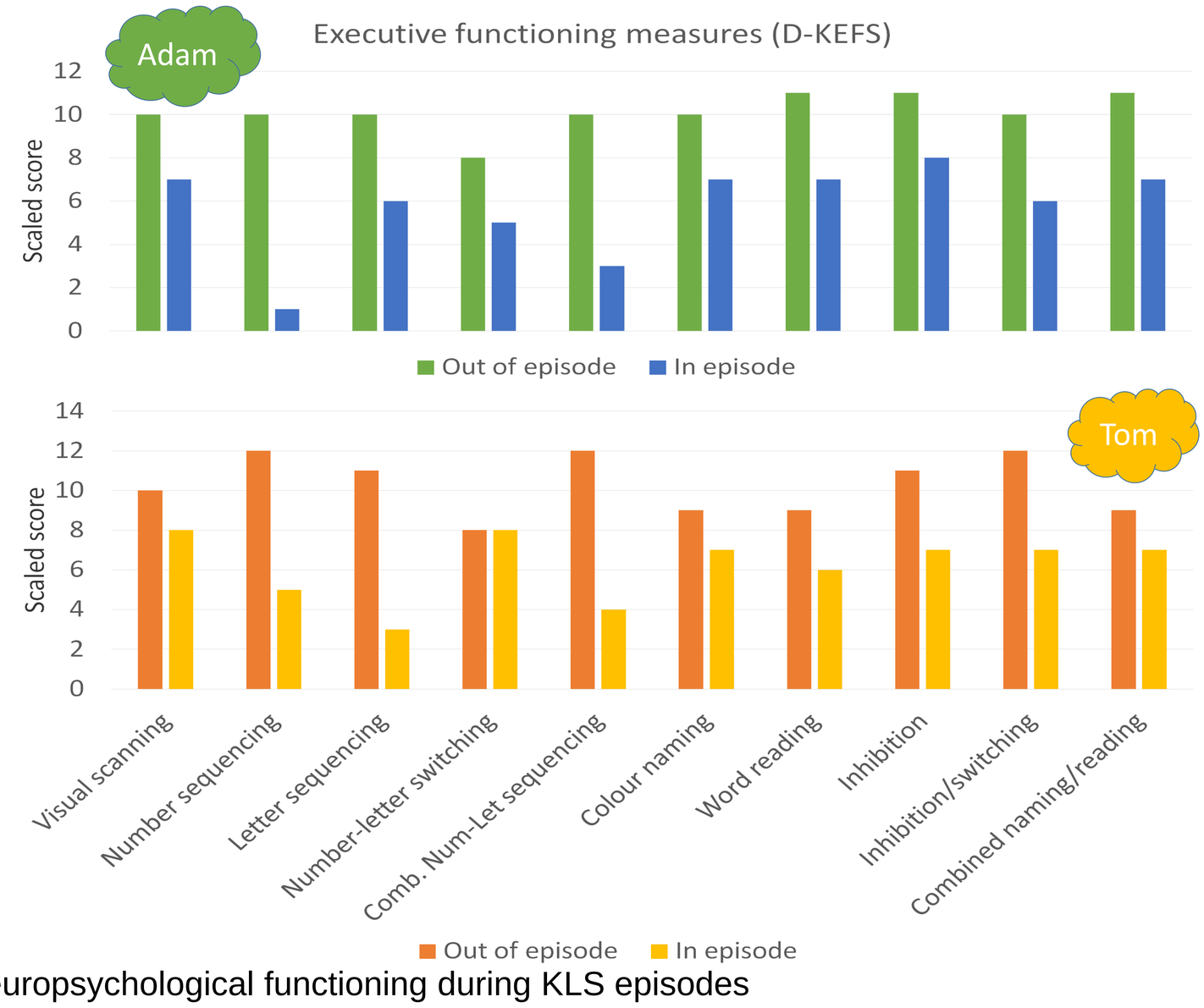
•Both young men were aged 18 and had an IQ in the average range

•Both showed impaired performance in all areas 'in' vs. 'out of' KLS episode

	Adam	Tom
Age of onset	14	15
Mean episode frequency	3-4 monthly	1st Year: 2-3monthly Subsequently: 4-6 monthly
Mean ep duration	6-14 days	7-10 days
De-realisation	Yes	Yes
Sexualised beh'r	No	No
Change in appetite	Increased fluid intake (water) and food intake – craving healthier foods than usual	Presents as being persistent and irritable in his demands for food.
Other features	Lethargy Social anxiety Dislike for noisy/busy environments Reassurance seeking from mother Feeling cold Repetitive behaviours, e.g. Watching a film over and over Feeling of a loss of time	Reduced focus Impaired peripheral vision Dislike of loud noises Changes in temperature perception Seeks out parental presence Feeling inexplicably scared Unable to sense water on skin (in shower)







Discussion and conclusions

•Initial data from two case studies indicate compromised neuropsychological functioning during KLS episodes

•Performance most markedly affected in executive functioning, specifically with weaknesses in sequencing skills and inhibiting impulses

•Deficits also identified in auditory memory (recall and recognition), as well as reduced processing speed and sustained attention

•Variation in profiles in line with clinical report of heterogeneous experience of KLS

•Points to the value of a larger scale research project to corroborate these initial findings

References
1 Arnulf, Zeitzer, File, Farber & Mignot (2005). *Brain*, 128, 2763 - 2776
2 Uguccioni, Lavault, Chaumereuil, Golmard, Gagnon & Arnulf (2015). Sleep, 39, 10.5665/sleep.5458