The auditory phenotype of behavioural variant and right temporal variant frontotemporal dementia

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INTRODUCTION

Patients with behavioural variant and right temporal variant frontotemporal dementia (bvFTD and rtvFTD) commonly exhibit abnormal hedonic and other behavioural responses to sound, such as musicophilia or aversion to certain sounds [1]. Although hearing impairment has emerged as a potent association of cognitive decline in dementia syndromes and a promising treatment target, the hearing changes in bvFTD and rtvFTD which lead to auditory dysfunction are poorly characterised [2].

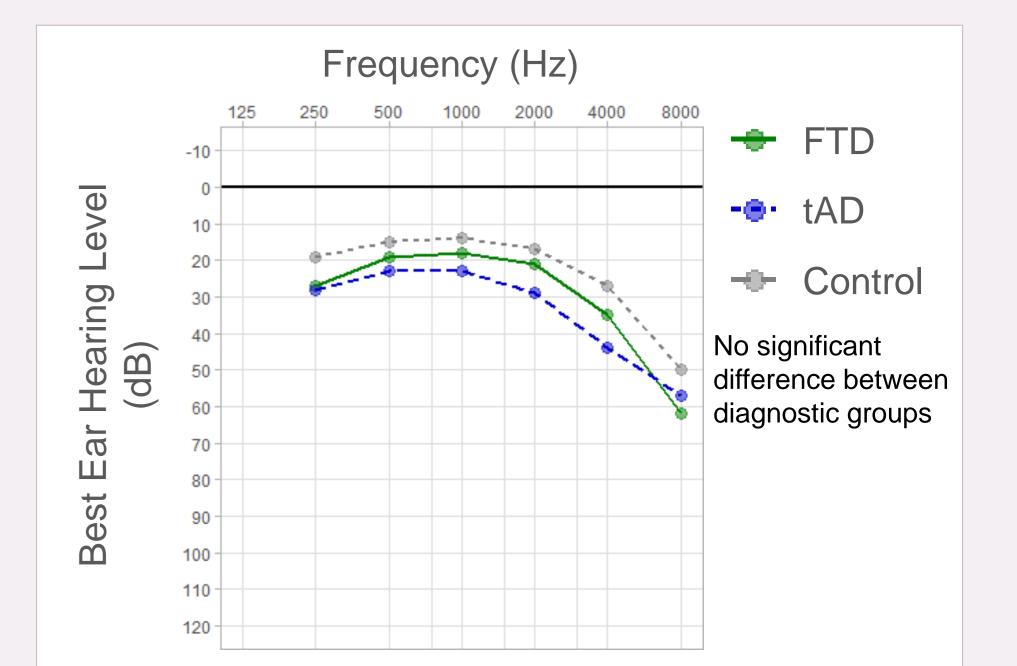
Here we addressed this issue using the Queen Square Tests of Auditory Cognition (QSTAC) – a novel psychoacoustic battery for the systematic assessment of peripheral and central auditory functions (including pitch pattern perception, environmental sound recognition, sound localisation and emotion processing) to characterise discrete 'auditory phenotypes' in dementia syndromes [3].

METHODS

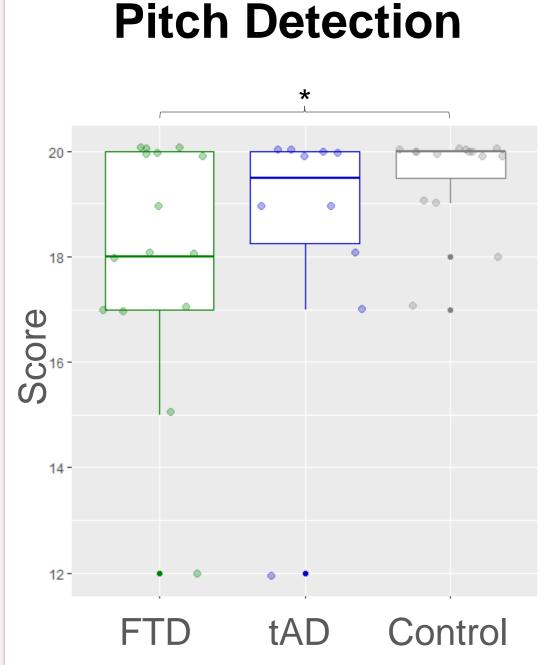
The QSTAC was administered to patients with bvFTD or rtvFTD (n = 15), typical Alzheimer's Disease (tAD, n = 10) and healthy age matched controls (n = 15). All patient groups met diagnostic consensus criteria [4,5,6]. QSTAC stimuli were administered through headphones at a comfortable listening level.

Pure tone audiometry was administered to all participants to assess peripheral hearing function.

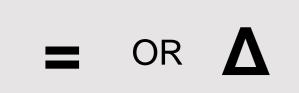
PURE TONE AUDIOGRAM

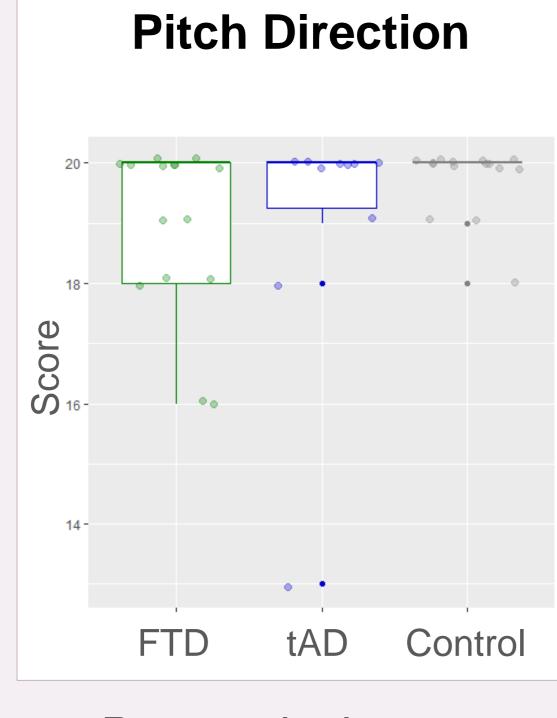


RESULTS



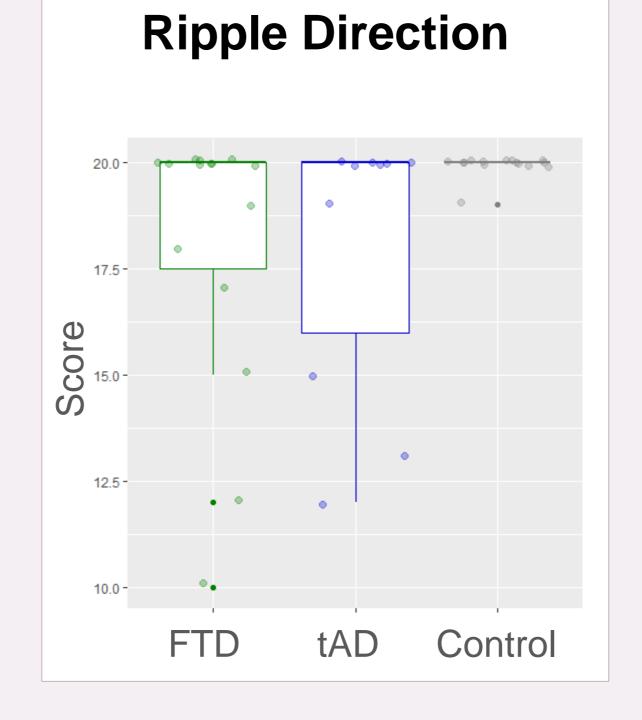
Prototypical source identity



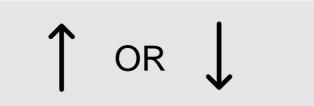


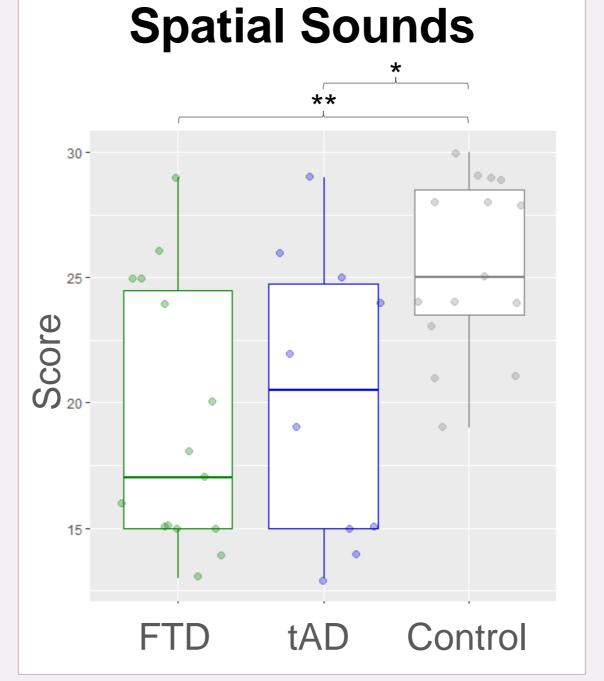
Prototypical pattern identity

OR



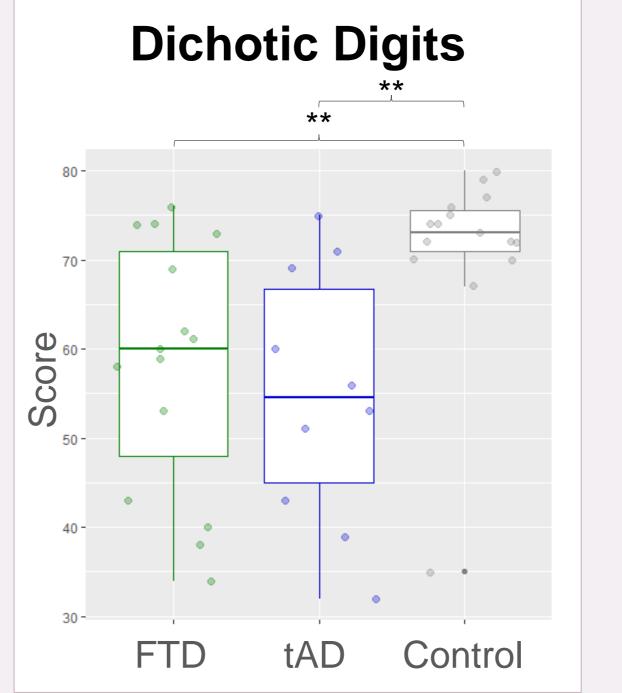
Auditory apperception





Auditory spatial processing



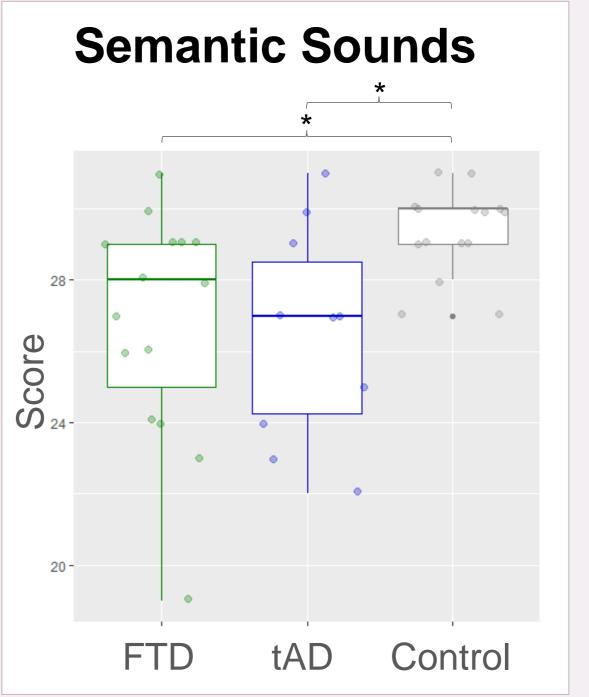


Auditory scene analysis

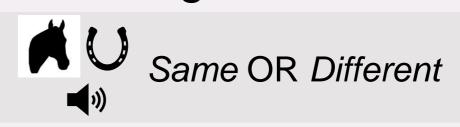


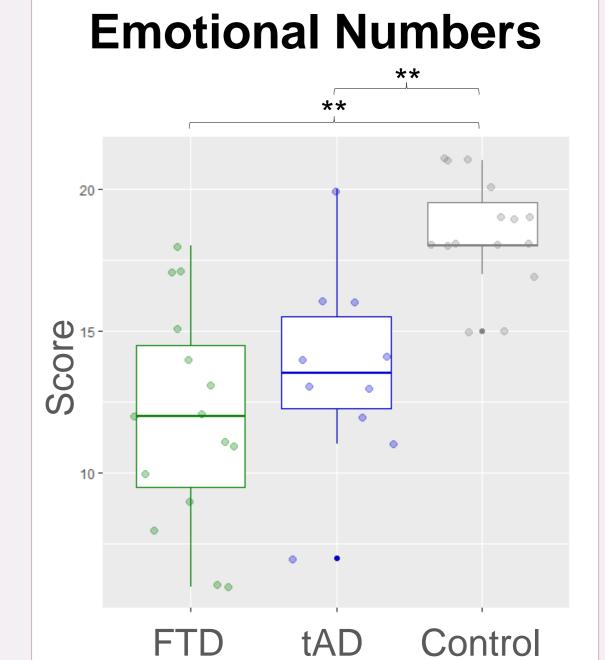






Auditory object recognition





* = p < 0.05; ** = p < 0.01

Auditory affective processing



CONCLUSION

bvFTD and rtvFTD have distinct phenotypes of auditory cognitive dysfunction which likely contributes to the hearing alterations that many patients with these diagnoses experience in daily life. This group demonstrated impairment compared to healthy controls in auditory source identification, spatial processing, scene analysis, object recognition and affective processing.

Our findings call attention to an under-recognised issue in frontotemporal dementia that warrants further clinical interpretation and the development of management strategies tailored to real-world acoustic environments. Peripheral hearing deficits should be corrected using hearing aids, however, real-world hearing disability in bvFTD and rtvFTD is likely to reflect auditory brain dysfunction and will require additional management strategies.

REFERENCES

[1] Fletcher et al. 2015. Cortex 67, 95-105. [2] Johnson et al. 2021. Brain, 144(2), 391-401. [3] Johnson et al. 2022. Journal of Neurology, Neurosurgery & Psychiatry 93(6), A48. [4] Dubois et al. 2014. The Lancet Neurology 13(6), 614-629. [5] Gorno-Tempini et al. 2011. Neurology 76(11), 1006-1014. [6] Rascovsky et al. 2011. Brain 134(9), 2456–2477.















