

A CROSS-SECTIONAL STUDY OF VESTIBULAR CHARACTERISTICS OF INDIVIDUALS WITH INFRATENTORIAL (CLASSICAL) SUPERFICIAL SIDEROSIS

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INTRODUCTION

Infratentorial superficial siderosis (iSS) is a rare neuro-otological disorder. It results from chronic extravasation of blood into cerebrospinal fluid (often due to dural defects) and deposition of iron-degradation product haemosiderin on the surfaces of CNS structures (commonly: cerebellum, and brainstem, but also the 8th cranial nerves).^{1,2} Susceptibility-weighted MRI is the reference standard diagnostic modality (**Figure 1**). Slowly progressive impairment of hearing and balance are the most common features of iSS. Vestibular dysfunction is described as mixed, of central (cerebellar or brainstem) and peripheral origin³. It is difficult to ascertain the site of vestibular involvement in iSS due to small cohort numbers or limited test battery used, as well as paucity of published reports⁴.

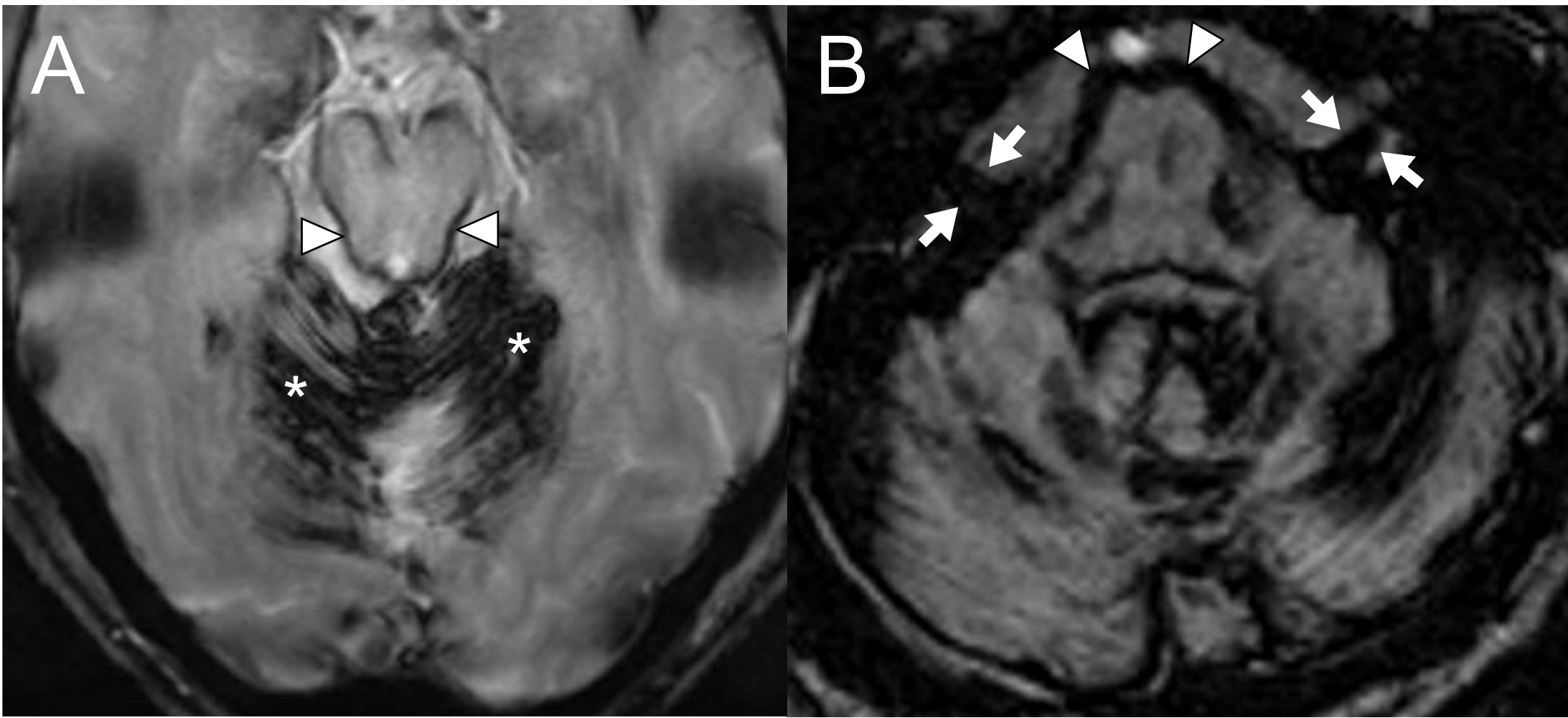


Figure 1. Axial susceptibility-weighted MR images with signal loss typical for iSS, involving cerebellum (asterisks, **A**), brainstem (arrowheads: midbrain, **A**; pons, **B**), and 8th cranial nerves (arrows, **B**).

AIM

To characterise vestibular function in a large cohort of individuals with iSS and attempt to localise the affected segment of vestibular pathway

METHODS

Permission for the study was obtained from the departmental clinical governance team (as part of a clinical audit). Patients were also invited to participate in a dedicated research study; permission from the NHS Research Ethics Committee was granted (REC 19/LO/1162AM01). We reviewed results of vestibular assessments of patients with radiologically confirmed diagnosis of iSS between 30/6/2004 and 01/09/2023. Vestibular assessments were undertaken at the UCLH NHS Foundation Trust, in line with the BSA guidelines⁵⁻⁷; the results were compared to departmental or published or equipment manufacturers' norms.⁸⁻¹¹ Relevant anatomy, tests performed, equipment used and the departmental norms are described elsewhere.¹¹ Data were anonymised at extraction; each case was reviewed separately for evidence of central, or peripheral, or mixed involvement where:

Vestibular dysfunction of central origin was considered if involving vestibular pathway proximal to Scarpa's ganglion; peripheral vestibular involvement was considered in the presence of end-organ dysfunction or if involving distal (to Scarpa's ganglion) vestibular pathway.¹²

RESULTS

N = 31 patients had vestibular assessments; **22 (71%) males**
Bedside findings were included in 2 cases

	Mean	Median	Standard deviation	Interquartile range
Age at test, years	58.2	63.0	14.5	19.0
Causative event to test (n=28), years	25.0	23.0	12.2	14.0

Vestibular tests

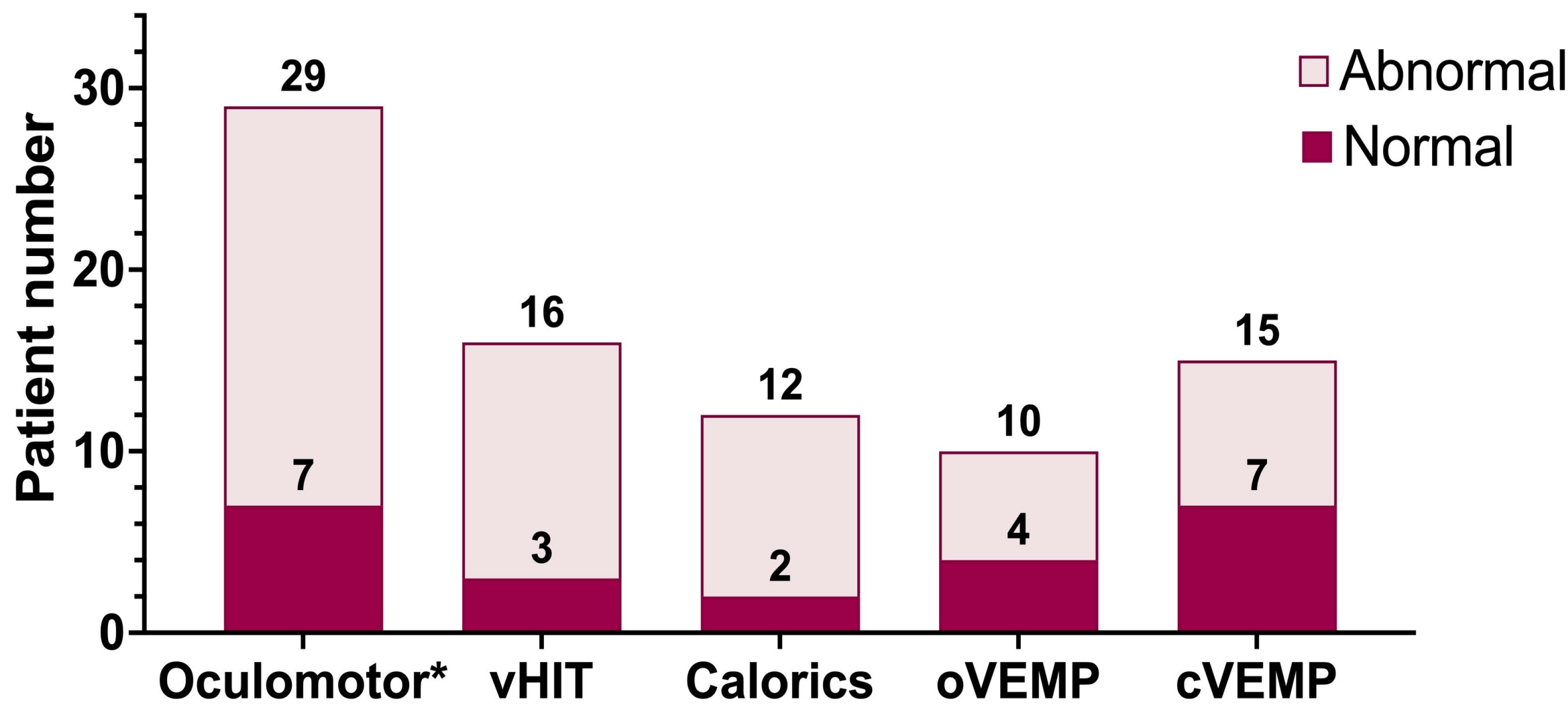


Figure 2. Vestibular tests performed in the cohort. *Abnormal central vestibular oculomotor findings (abnormal findings in at least one: gaze, saccades, smooth pursuit; optokinetic nystagmus or vestibulo-ocular reflex suppression). Legend: vHIT video head impulse test, o/cVEMP ocular/cervical vestibular evoked myogenic potentials.

Oculomotor findings (n=29)

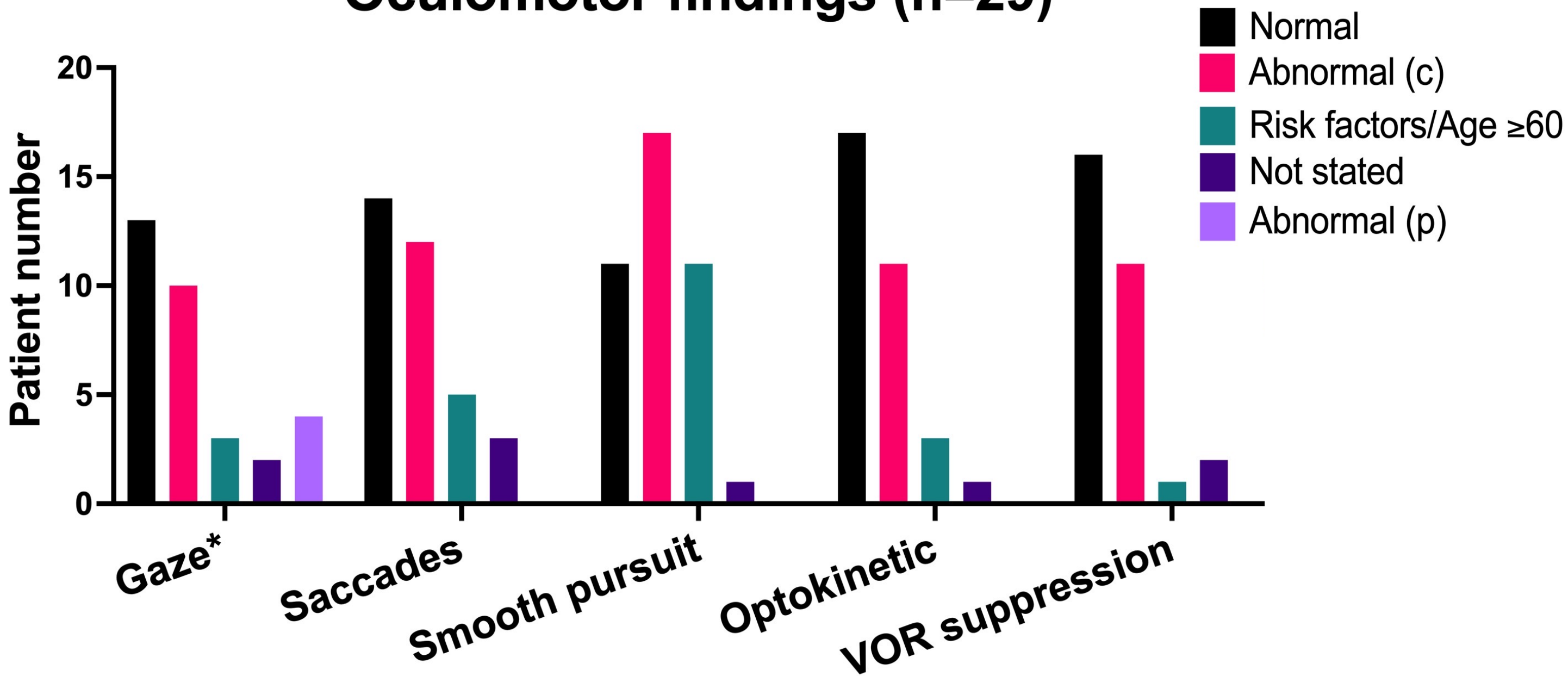


Figure 3. Oculomotor findings. *Abnormal gaze findings suggestive of (c) central and (p) peripheral vestibular involvement.

Vestibular findings

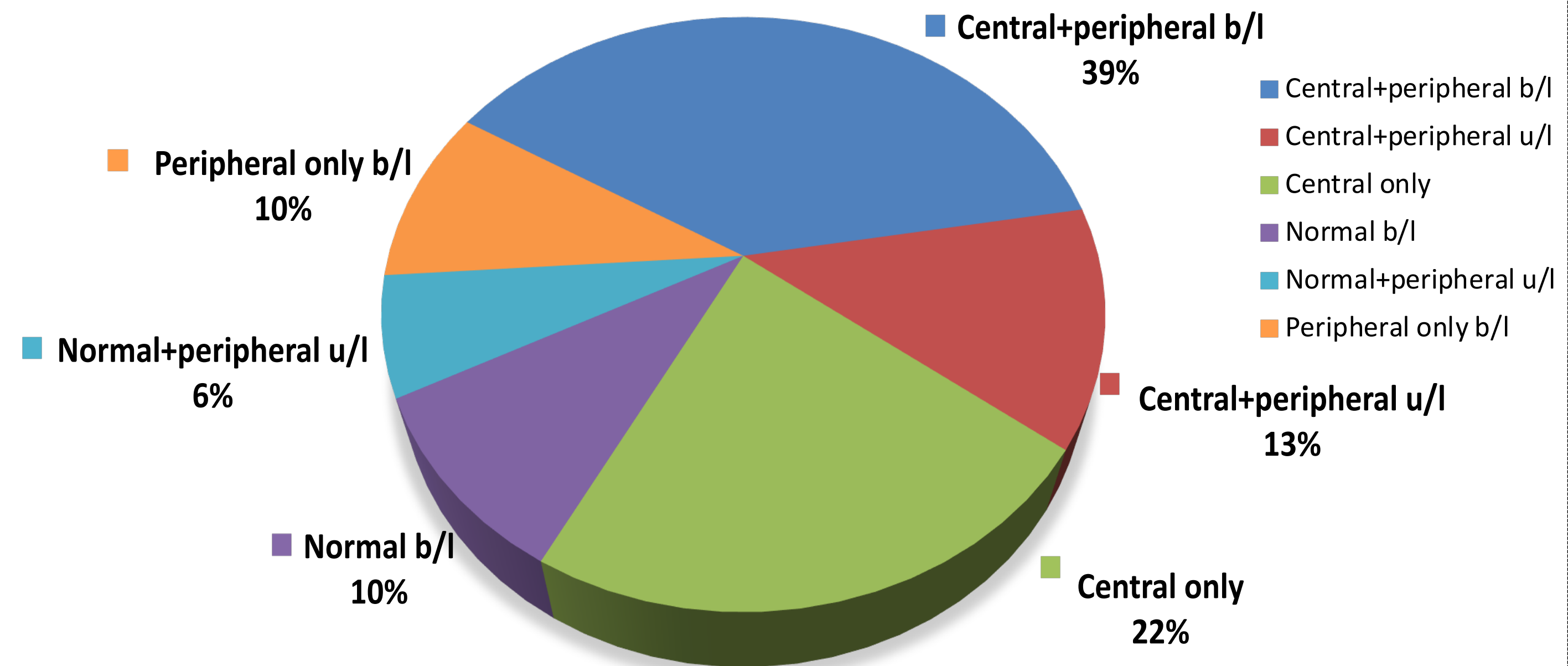


Figure 4. Pie chart of vestibular findings in the cohort. The most common findings were: central with bilateral peripheral vestibular involvement (n=12), followed by only central vestibular involvement (n=7), and central with unilateral peripheral involvement (n=4). Bilaterally normal vestibular function was observed in 3 cases, peripheral findings were unilateral, with normal vestibular function in the other ear, in further 2 cases.

CONCLUSION

First study to analyse vestibular tests from a large cohort of iSS patients. We demonstrate predominantly central vestibular involvement, in keeping with the imaging findings. Presence of additional peripheral involvement may suggest antegrade progression of vestibular dysfunction in iSS. Normal findings suggest the onset of vestibular dysfunction may be preceded by the radiological manifestations of iSS however longitudinal studies are needed.

References

- Fearnley JM, Stevens JM, Rudge P. Superficial siderosis of the central nervous system. *Brain*. 1995;118 (Pt 4):1051-66.
- Wilson D, Chatterjee F, Farmer SF, Rudge P, McCarron MO, Cowley P, et al. Infratentorial superficial siderosis: Classification, diagnostic criteria, and rational investigation pathway. *Ann Neurol*. 2017;81(3):333-43.
- Takeda T, Kawashima Y, Hirai C, Makabe A, Ito T, Fujikawa T, et al. Vestibular Dysfunction in Patients With Superficial Siderosis of the Central Nervous System. *Otol Neurotol*. 2018;39(6):e468-e7.
- Yoo A, Jou J, Klopfenstein JD, Kattah JC. Focused Neuro-Otological Review of Superficial Siderosis of the Central Nervous System. *Front Neurol*. 2018;9:358.
- British Society of Audiology. Recommended Procedures, Position Statements, Minimum Training Guidelines and Practice Guidance: The British Society of Audiology; 2016 [Available: <https://www.thebsa.org.uk/resources/>]
- British Society of Audiology. Recommended procedure for vestibular assessment - eye movement recordings.: The British Society of Audiology; 2015 [Available: <https://www.thebsa.org.uk/resources/>]
- British Society of Audiology. Recommended procedure: The Caloric Test. 2010.
- Halmagyi GM, Chen L, MacDougall HG, Weber KP, McGarvie LA, Curthoys IS. The Video Head Impulse Test. *Front Neurol*. 2017;8:258.
- Interacoustics. Manual Micromedical VisualEyes™ 515/VisualEyes™ 525: Additional Information. 2018.
- GN Otometrics A/S. ICS Impulse USB User Guide. 2017.
- N Kharytaniuk, Clinical and imaging biomarkers of audiovestibular function in infratentorial superficial siderosis. 2023. E-Thesis. [available in UCL Discovery: <https://discovery.ucl.ac.uk/id/eprint/10163608/>]
- Baloh RW, Kerber KA. Clinical neurophysiology of the vestibular system. 4th ed. New York: Oxford University Press; 2011. xxi, 455 p.