Prediction of Long-term Visual Outcomes in Infantile Nystagmus using Handheld OCT: A 12-year Longitudinal Study

Sohaib R. Rufai[1,2], Michael Hisaund[1,2], Rebecca J. McLean[1,2], Ravi Purohit[3], Gail Maconachie[4], Viral Sheth[4], Helena Lee[5], Frank A. Proudlock[1], Irene Gottlob[1,6], Mervyn G. Thomas[1,2]

1) University of Leicester Ulverscroft Eye Unit, UK; 2) Leicester Royal Infirmary, UK; 3) Oxford University Hospitals, UK; 4) University of Sheffield, UK; 5) University of Southampton, UK; 6) Cooper University Health Care, New Jersey, USA.

Introduction:

Infantile nystagmus is often associated with foveal hypoplasia.[1,2] It is currently unknown whether foveal hypoplasia grading can predict long-term visual acuity and function.

In this world-first study, we evaluated foveal hypoplasia grading using handheld optical coherence tomography (OCT) as a predictor of long-term visual acuity and function in infantile nystagmus, from pre-verbal to pre-adolescent stages.

Methods:

Single-centre, prospective, longitudinal study from 2012-2024

Dataset

- 19 children with infantile nystagmus
- Recruited as pre-verbal children aged 0-2 years, followed up as pre-adolescents aged 9-12 years
- Bilateral foveal OCTs acquired using Envisu C2300 Handheld OCT, Leica Microsystems (Figure 1)

Main outcome measures

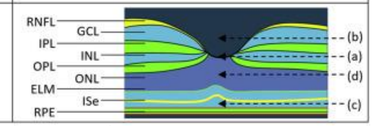
- Leicester Grading System for Foveal Hypoplasia grades (Figure 2)[1,2]
- Distance VA (Thomson Test Chart), near VA (Thomson Test Chart), colour vision (Ishihara), contrast sensitivity (Thomson Test Chart) and stereopsis (Frisby)

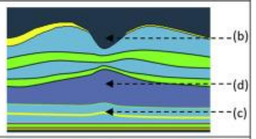
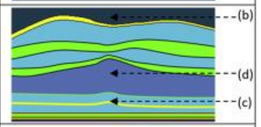
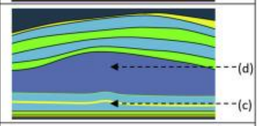
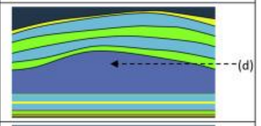
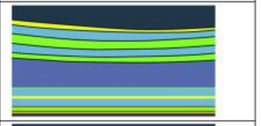
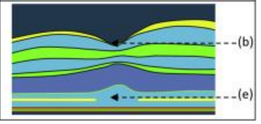
Statistical analysis

- Linear mixed regression model



← Figure 1: Handheld OCT in pre-verbal child
Photograph taken with parental consent

Normal foveal structural features detectable using optical coherence tomography	Illustration
(a) Extrusion of plexiform layers (b) Foveal pit (c) OS lengthening (d) ONL widening	

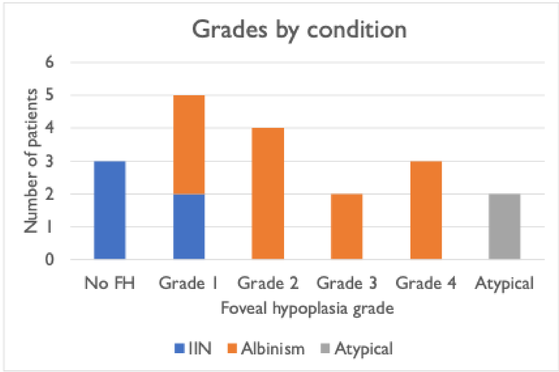
Grade of foveal hypoplasia	Structural features detected on optical coherence tomography	Present or absent	Illustration
1a	**(a) Extrusion of plexiform layers** **(b) Foveal pit – Nearly normal** (c) OS lengthening (d) ONL widening	**(a) Absent** (b) Present (c) Present (d) Present	
1b	**(a) Extrusion of plexiform layers** **(b) Foveal pit – Shallow indent** (c) OS lengthening (d) ONL widening	**(a) Absent** (b) Present (c) Present (d) Present	
2	**(a) Extrusion of plexiform layers** **(b) Foveal pit** (c) OS lengthening (d) ONL widening	**(a) Absent** **(b) Absent** (c) Present (d) Present	
3	**(a) Extrusion of plexiform layers** **(b) Foveal pit** **(c) OS lengthening** (d) ONL widening	**(a) Absent** **(b) Absent** **(c) Absent** (d) Present	
4	**(a) Extrusion of plexiform layers** **(b) Foveal pit** **(c) OS lengthening** **(d) ONL widening**	**(a) Absent** **(b) Absent** **(c) Absent** **(d) Absent**	
Atypical	**(a) Extrusion of plexiform layers** (b) Foveal pit – Shallow (e) ISe disruption	**(a) Absent** (b) Present (e) Present	

← Figure 2: The Leicester Grading System for Foveal Hypoplasia[1,2]
Key: ELM = external limiting membrane; GCL = ganglion cell layer; ILM = internal limiting membrane; INL = inner nuclear layer; IPL = inner plexiform layer; OPL = outer plexiform layer; RNFL = retinal nerve fibre layer; RPE = retinal pigment epithelium.

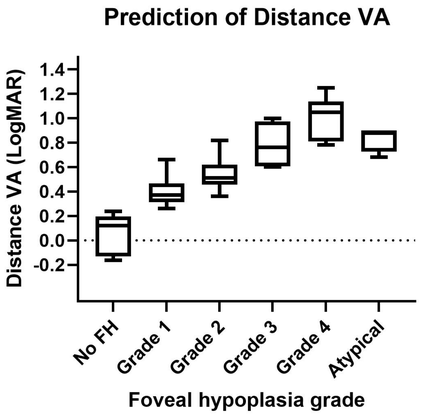
Results:

Nineteen children with infantile nystagmus (idiopathic infantile nystagmus [IIN]: 5; albinism: 12; achromatopsia: 2) were recruited (Figure 3). Median age at first examination was 22 months (range: 1-33; IQR: 13-30). Median age at follow-up was 12 years (range: 9-12; IQR: 11-12).

Foveal hypoplasia grade at first examination was significantly associated with future distance VA (R=0.86, F=98, P<0.0001; Figure 4), near VA (R=0.82, F=74, P<0.0001), contrast sensitivity (R=0.82, F=76, P<0.0001), stereopsis (R=0.70, F=16, P=0.001) and colour vision (R=0.60, F=20, p<0.0001).



↑ Figure 3: Foveal hypoplasia grades by condition
Key: FH = foveal hypoplasia; IIN = idiopathic infantile nystagmus



↑ Figure 4: Prediction of distance VA
Key: FH = foveal hypoplasia; VA = visual acuity,

Conclusions:

Handheld OCT can predict future visual outcomes in infantile nystagmus, from infancy to pre-adolescence. This could help with diagnosis, prognosis, counselling and clinical management.

References:

1. Thomas et al. *Ophthalmology*. 2011 Aug;118(8):1653-60.
2. Rufai et al. *Ophthalmology*. 2020 Apr;127(4):492-500.

FUNDED BY NIHR | National Institute for Health and Care Research

MRC Medical Research Council | Ulverscroft Foundation | Fight for Sight | Nystagmus Network

Financial disclosures: Sohaib Rufai, recipient of Thea medial education grant. Funding: Sohaib Rufai is funded by an NIHR Clinical Lectureship CL-2022-11-001. This poster presents independent research funded by the NIHR, MRC, Ulverscroft Foundation, Fight for Sight and Nystagmus Network. The views expressed are those of the author(s) and not necessarily those of the NHS, the funders or the Department of Health and Social Care. Acknowledgements: Many thanks to all patients and families who took part in this study. Correspondence: