

The Leicester Grading System for Foveal Hypoplasia

BY SOHAIB RUFAI

The University of Leicester Ulverscroft Eye Unit have published the first medical grading system named after the city of Leicester.

Infantile nystagmus is characterised by constant and involuntary eye movements and affects 24 per 10,000 people [1].

Onset is usually in infancy, which can be a worrying time for parents and families because the pre-verbal infant is unable to communicate their level of vision. However, a great variation in visual prognosis exists, because some children are affected more mildly than others [2]. Predicting future vision in affected children could avert anxiety and / or prompt parents to plan appropriate adjustments to support the child in their development and educational attainment. Furthermore, predicting vision can also guide clinical decision-making.

Infantile nystagmus is commonly associated with underdevelopment of

the fovea – the central part of the retina responsible for fine vision. This is termed ‘foveal hypoplasia’ [3]. In 2011, Thomas et al. [3] developed a structural grading system for foveal hypoplasia based on optical coherence tomography (OCT) data from adults and older children. This landmark study did not feature infants as it took place before the handheld OCT was available in Europe.

Handheld OCT is non-invasive, child-friendly and enables ultra-high resolution imaging of an infant's retina, in vivo, within seconds. In 2012, Leicester became the first centre in Europe to obtain the handheld OCT. Under the leadership of Professor Irene Gottlob, Leicester has used handheld OCT to investigate the normal development of the fovea [4] and optic nerve [5] in children, as well as a wide variety of childhood conditions including but not limited to nystagmus [6], retinopathy of prematurity [7], glaucoma [8], anterior segment dysgenesis [9] microcephaly [10] and cerebral malaria [11].

Now, our group has conducted the first study using handheld OCT to predict future vision in infantile nystagmus [2]. In this

study, we have updated and validated our grading system, which we have named the Leicester Grading System for Foveal Hypoplasia [2]. This is the first medical grading system named after the city of Leicester, which can aid in diagnosis, prognosis and clinical decision-making in affected children. We are delighted that our study has been published in *Ophthalmology* and we hope that our findings will be widely cited.

The main conclusions of our study were as follows: i) handheld OCT can predict future vision in infantile nystagmus; ii) structural grading was a better predictor of future vision than quantitative segmentation; iii) preferential looking was a relatively poor predictor of future vision; iv) inter-grader reliability testing demonstrated that our grading system is robust; v) all grades remained stable throughout the study period. Moreover, our grading system can help to guide clinical decision-making. For instance, if VA is poorer than expected according to grading, suspicion should be raised for other pathology limiting the VA. This could include other retinal pathology, such as retinal dystrophy, cortical / neurological disorders, amblyopia, conditions affecting the anterior segment or refractive error. If a patient has poor VA consistent with a high degree of foveal hypoplasia, it is likely that the poor VA is accounted for at the retinal level.

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“A great variation in visual prognosis exists, because some children are affected more mildly than others”



The University of Leicester Ulverscroft Eye Unit and colleagues.

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AUTHOR



Dr Sohaib Rufai,

NIHR Doctoral Fellow ST4 in Ophthalmology, University of Leicester Ulverscroft Eye Unit, Leicester; Clinical and Academic Department of Ophthalmology, Great Ormond Street Hospital, London.

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