Rapid assessment of cone system function using a hand-held electroretinography device in ABCA4 retinopathy

Zaina Bouzia, 1,3,4 Diana Butu, 2,2 Ana Fakin, 1,3 Talha Soorma, 1,4 April Q Neville, 1 Christopher J Hammond, 4 Michel Michaelides 1-3, Andrew R Webster, 1,3 Omar A Mahroo 1,3,4

1. Moorfields Eye Hospital, London; 2. Royal Free Hospital, London 3. Institute of Ophthalmology, University College London 4. Department of Ophthalmology, King’s College London, St Thomas’ Hospital Campus; 5. Falkirk Community Hospital

Purpose

- Patients with ABCA4 retinopathy (Stargardt disease) can be grouped by electrophysiology,1 with prognostic implications: those with a normal photopic full-field electroretinogram (ERG) are less likely to suffer deterioration in generalised retinal function over subsequent years.2
- We used a portable device to obtain photopic ERGs, investigating tolerability, correlation with ultra-widefield autofluorescence (AF) and with prior conventional ERG testing.

Methods

- Adults with ABCA4 retinopathy underwent photopic recordings with the portable device (RETeval, LKC Technologies Inc., Gaithersburg, MD, USA) using skin electrodes.
- Pupils were undilated; the device adjusts stimulus strength according to pupil diameter to deliver a retinal illuminance equivalent to international standards.
- Right eye recordings were analysed.
- ERG parameters were compared with control recordings from >500 healthy participants from the TwinsUK cohort.3 Amplitudes <5th centile, and peak times >95th centile were deemed abnormal.
- Patients underwent ultra-widefield AF (Optos plc, Dunfermline, UK) imaging the same day; images were grouped by presence of far peripheral involvement. The majority of patients had prior conventional ERG testing (in some cases several years previously).

Results

- 65 patients were included (mean age 46.4 years; 33 females). Recordings were well-tolerated, lasting <5 min.
- 40 patients (60%) had peripheral abnormal AF; the proportion of patients with abnormal ERG parameters was higher in these patients (95% vs 23%, p<0.01), and mean ERG b-wave and flicker amplitudes were lower, and peak times more delayed, compared with those without abnormal peripheral AF (p<0.05).

Comparison with conventional ERGs

- Of 32 patients with previously abnormal conventional ERGs, 31 (97%) had abnormal hand-held device ERGs.
- Of 24 patients with previous normal ERGs, 18 (75%) had normal hand-held device ERGs.

Conclusion

- Recordings were well-tolerated and complete within minutes.
- The majority (95%) of patients with abnormal far peripheral AF had abnormal ERGs, demonstrating strong structure-function correlation.
- Portable ERG findings broadly agreed with prior conventional testing, although the disease progression may have occurred in some patients in the intervening period.

References


Acknowledgements

Wellcome Trust (206619/Z/17/Z); Fight for Sight UK (1409/10); Thomas Pocklington Trust; Moorfields Eye Charity; NIHR Biomedical Research Centre at Moorfields Eye Hospital and the UCL Institute of Ophthalmology; TwinsUK receives support from the Wellcome Trust and Biomedical Research Centre based at Guy’s and St Thomas’ NHS Foundation Trust and King’s College London.