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

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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).

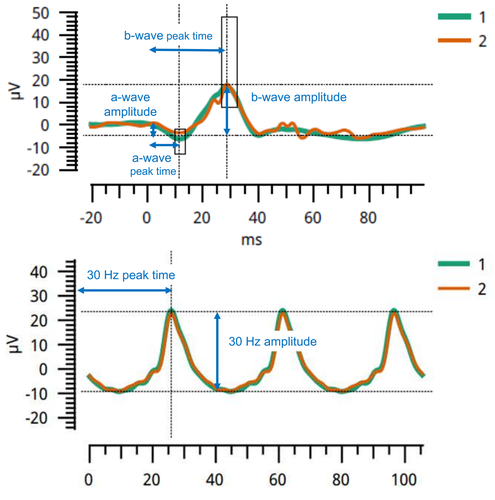


Figure 1 (Left): ERG photopic flash (upper panel) and flicker responses (lower panel) from a healthy subject. Green and orange traces represent averaged recordings from two consecutive stimulus presentation (each lasting <1 minute in each eye) demonstrating high intrasession repeatability.

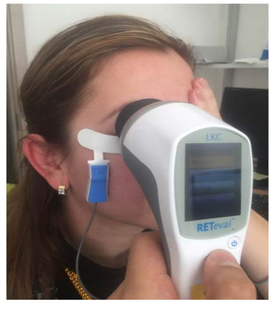
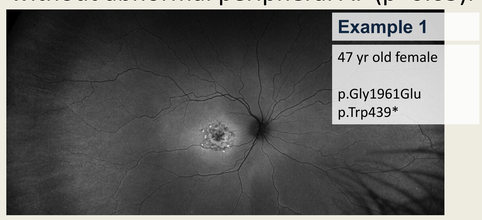
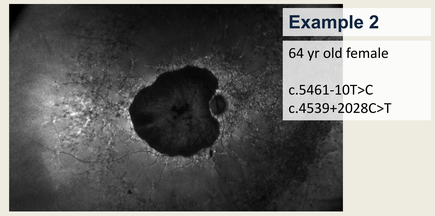


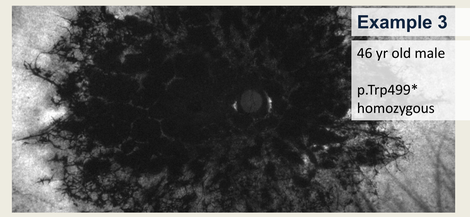
Figure 2 (Right): A participant undergoing ERG recordings with the portable device

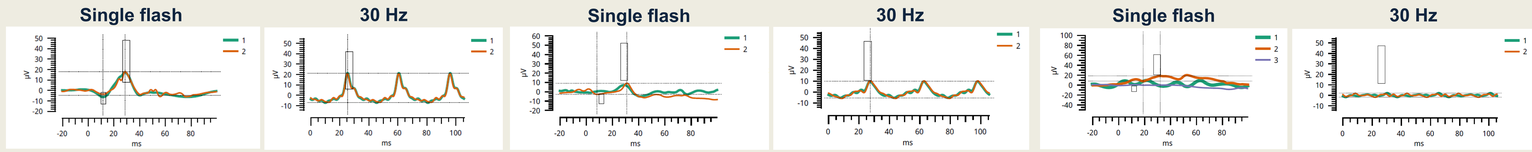
Results

- 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).

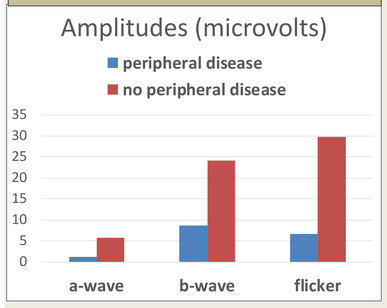






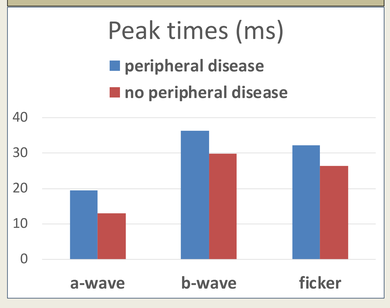


Graphic 1: Mean photopic and flicker response amplitudes using hand-held ERG device



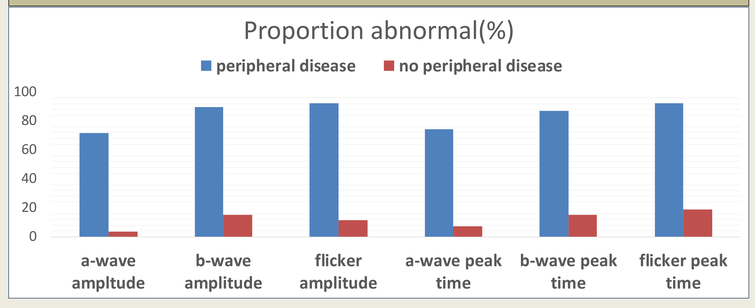
*p<0.05 unpaired t test

Graphic 2: Mean photopic and flicker response implicit time using hand-held ERG device



*p<0.05 unpaired t test

Graphic 3: Percentage of abnormal ERG parameters in patients with or without peripheral retinal involvement



Patients with peripheral disease had greater prevalence of abnormal ERG parameters (p<0.05)

Comparison with conventional ERGs

- Of **32** patients with previously **abnormal conventional** ERGs, **31** (97%) had **abnormal** parameters with the **hand-held device**.
- 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

1. Lois N et al. Arch Ophthalmol. 2001 Mar;119(3):359-69.
2. Fujinami K et al. Am J Ophthalmol. 2013 Jun;155(6):1075-1088.e13.
3. Soorma T et al. Invest. Ophthalmol. Vis. Sci.. 2017; 58(8):4889.

Acknowledgements

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