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Electrophysiology

When physicians first realized that the patterns of electrical current through the heart correlated with different forms of heart disease, a new era was born not just in cardiology but in all of medicine. Today, electrocardiograms, commonly called EKGs, are a staple of medical student education and routinely performed on almost every patient admitted to the hospital. The retina, itself a neural structure, also produces an electrical signal with distinct physiological and pathophysiological patterns. Although it does not have the wide utility of the EKG, the electroretinogram (ERG) is a key diagnostic tool in inherited retinal degenerations, toxic maculopathies, occult retinopathies, and cases of vision loss of unclear etiology.

The ERG is unique in ophthalmology because it is the only test in the entire field that measures photoreceptor function rather than just structure. Other retinal diagnostic tests, like optical coherence tomography (OCT), OCT angiography, and adaptive optics, tell us about the structure of the retina, retinal vasculature, or the photoreceptors themselves, from which we clinicians attempt to extrapolate conclusions about functions, but not strictly about their function - thus the uniqueness of the ERG. It comes essentially in two flavors: a full-field ERG and a multifocal ERG (mfERG). The full-field test, the first one developed, measures the mass electrical response from the

entire retina in response to a single flash of light. Because the majority of the retina consists of rods, this signal is dominated by the rod photoreceptor. Thankfully, rod and cone responses can be teased out by varying ambient lighting conditions. The use of "dark adaptation", which involved surrounding a patient by darkness for an extended period of time before exposing them to a flash of light, isolates the rods and is called the scotopic response. Similarly, the photopic response is more specific to cones. Cones can be further isolated by the use of a high-frequency. flickering light (30 Hz response), as the rods cannot repolarize fast enough to respond to such a rapid cycle of flashes. The full-field ERG is profoundly affected in inherited retinal degenerations like retinitis pigmentosa, and many of these patients have almost no measureable response to a flash of light. Other rare acquired diseases like autoimmune retinopathy, cancer-associated retinopathy, melanoma-associated retinopathy, and acute zonal occult outer retinopathy are also diagnosed by full-field ERG.

The multifocal ERG measures the response from multiple small foci of retina. Commonly, this is applied to the posterior pole in order to measure the response of photoreceptors in the macula. This test represented a major advancement in retinal electrophysiology because the full-field ERG typically drowns out the

cone response from the macula. Thus, a patient could have a severe maculopathy but a normal full-field ERG, making the test in such cases useless at best and confusing at worst.

The multifocal ERG is commonly used in assessing macular function in cases of suspected toxicity, such as from plaquenil. Patients on plaquenil should undergo full assessment by a retinal specialist annually. The mfERG is also used in cases of central vision loss in the setting of a normal exam and negative results of other more conventional tests. This can distinguish between a macular or optic nerve origin to unilateral or bilateral central vision loss of unclear etiology.

In short, while these tests are not common, they are critical in the diagnosis of several important and serious retinal diseases. The full-field and multifocal ERGs, both of which are offered at the Kentucky Lions Eye Center, allow us to isolate rod and cone responses and to focally test the macula. The test requires strict control of conditions and calibration of instrumentation, which is why few centers are able to match the rigorous standards set by the International Society for Clinical Electrophysiology of Vision. However, when performed correctly, it provides insight into the retina that no other test can.

By: Harpal Sandhu, MD

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