In the recently published consensus-based care recommendations for adults with myotonic dystrophy type 1 (DM1), visual symptoms caused by retinal dysfunction are not considered despite several studies reporting frequent abnormal electroretinograms (ERGs) in patients with DM1.

Because ERGs provide easily accessible and noninvasive objective information about the electrical activity of cells within the retina in response to light stimulation, they are of considerable interest to assess CNS alterations associated with DM1. Of note, even with no changes in ophthalmoscopy, ERGs in patients with DM1 and asymptomatic parents were shown to be moderately affected like that seen in early retinitis pigmentosa. Importantly, both rod and cones systems can be assessed when testing with scotopic low intensity or dim blue or red stimulation. Assessment of such changes can be crucial as they are consistent with complaints recorded in patient with DM1 support group including longer adaptation to darkness and struggle in dim-light environment such as movie theaters. Such perturbations are often associated with increased risks of falling and driving difficulties, especially at dusk. Altogether, this suggests that ERG screening should be considered as a potential biomarker for patients with DM1 and warrants the interdisciplinary involvement of neurologists and ophthalmologists.

abnormalities in DM1, such as lens opacities. However, we agree that retinopathy could be an objectively measurable biomarker in clinical trials of DM1. Further studies to develop electroretinogram and other retinal functional measures in correlation with the clinical or molecular measures of DM1 need to be considered. Bringing back ophthalmologists to the field of DM is very welcome.


Copyright © 2019 American Academy of Neurology
Reader response: Consensus-based care recommendations for adults with myotonic dystrophy type 1
Tuy Nga Brignol and Patrice E. Fort
*Neurol Clin Pract* 2019;9;366
DOI 10.1212/CPJ.0000000000000733

This information is current as of October 14, 2019

| Updated Information & Services | including high resolution figures, can be found at:  
|                              | http://cp.neurology.org/content/9/5/366.1.full.html |
| References                   | This article cites 5 articles, 1 of which you can access for free at:  
|                              | http://cp.neurology.org/content/9/5/366.1.full.html##ref-list-1 |
| Permissions & Licensing      | Information about reproducing this article in parts (figures, tables) or in its entirety can be found online at:  
|                              | http://cp.neurology.org/misc/about.xhtml#permissions |
| Reprints                     | Information about ordering reprints can be found online:  
|                              | http://cp.neurology.org/misc/addir.xhtml#reprintsus |