**Hadassah University Medical Center**

**The Center for Retinal and Macular Degeneration**

**The Unit for Clinical Electrophysiology and Visual System Function**

**Dept. of Ophthalmology, Hebrew University and Hadassah Hospital**

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**Referring physician:** Dr. Oranit Kristal-Shalit Patient: Yehonatan Einhorn

 Ophthalmologist Pinchas Rozen 90

 Tel Aviv

**ID no.: 34046553-3** **Age: 6 yr.** **Yr. of birth:** 2015 **Examination date:** Mar. 24 2021

Below are the results of the electrophysiological and psychophysical visual exams performed at our institution:

**Full Field ERG**: performed under anesthesia with sevoflurane gas and proceeded for a short time only.

The retinal responses of both eyes, immediately at the start of darkness, contain the characteristic components but their size is severely inferior to the norm for a short protocol exam. Noted is the tendency for a negative pattern of the responses.

**Cone Flicker 30 Hz:**

Rt. eye: amplitude 66 µV, within normal range. Response time – 35 msec, lengthened.

Lt. eye: amplitude 78 µV, within normal range. Response time – 35 msec, lengthened.

(Amplitude: lower threshold of the norm is 60 µV. Response time in adults is up to 33 msec.)

**Flash VEP**: (performed while alert)

After light flashes to each eye separately, consistent and clear responses to stimuli were received in the occipital brain, with both eyes having similar patterns. The responses are within the normal range for each eye.

Rt. eye: PW wave, peak response time to wave – 96 msec, amplitude -17 µV, within normal range.

Lt. eye: PW wave, peak response time to wave – 93 msec, amplitude -13 µV, within normal range.

(Amplitude: lower threshold of the norm in adults 7.0 µV. Response time: upper threshold of the norm 120 msec.)

**Conclusion:**

Mixed cone-rod response according to full-field ERG is severely inferior to the short-term protocol in both eyes, with a tendency towards a negative pattern of the responses. In the short-term protocol exam, the rod response cannot be accurately quantified, but the impression is that the rod response is inferior, and in fact even missing in each eye. The cone response is within normal range in each eye with a slightly lengthened response in cone flicker.

According to the FVEP exam, there is nerve conductivity from each eye to the occipital brain within normal range. During stimulation with flashes of light to each eye, separately, consistent and clear responses were registered in the occipital brain with similar patterns in both eyes. It is important to emphasize that the FVEP exam may be within normal range even in the presence of considerable impairment in the conducting fibers.

**Summary:**

Full-field ERG responses demonstrating severe inferiority of the mixed cone-rod response with a tendency towards a negative pattern and lengthened response time in some of the cone flicker responses may be consistent with one of the main following diagnostic possibilities:

1. Progressive disease from the group of retinitis pigmentosa in initial stages (sometimes in these stages, there is a tendency towards negative patterns of the responses). The fundoscopic findings reported in the referral letter may support this diagnosis.
2. Static disease from the CSNB group.

In order to try to progress with the diagnosis, referral is recommended for genetic investigation. This may be done by genetic counseling through the HMO or with the aid of the Michaelson Institute at Hadassah, and may be done as well in research studies in coordination with Prof. Dror Sharon, tel.: 02-6777112.

In addition, repeating the FFERG exam is recommended using a full protocol including darkness adaptation, where Yehonatan may participate in such an exam. The exam helps quantify the rods branch function as well as helps to examine if this is a static or progressive disease.

Referral recommended to the treating physician to assess the clinical significance of the findings of the ancillary tests detailed above.

Sincerely,

Prof. Eyal Benin M.D., PhD

Center Director

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