

DEPARTMENT OF OPHTHALMOLOGY  
ELECTRORETINOGRAPHY REPORT

Date 11/4/98 Bldg. Fl. Rm.  
Unit No. Test # 5345  
Name 1-38-94-88  
Birthdate WULF, MATTHEW A.  
12/3/97

Referring Physician: **STEINER, DR. ROBERT**  
Referring Diagnosis: **INFANTILE REFSUM DISEASE**

Pupil Size **8.0** mm. O.D. **8.0** mm. O.S Dilation Medications:  
Dark Adaptation Time **45** minute Normal range calculated for **0.3** year old  
ERG Protocol: **ERG SED IV** Preamplifier Gain = 1000 X

**Scotopic Responses:** (Right eye / Left eye)

**2.4 Neutral** Intensity 1  
**Density Filter** b-wave: **3 / 16?**  $\mu\text{V}$  at **116 / 124** msec  
**+0.1 ND** (normal: 15 - 95  $\mu\text{V}$  at 95 - 135 msec)  
**(white)**

Intensity 4:  
b-wave: • / •  $\mu\text{V}$  at • / • msec  
(normal: 55 - 145  $\mu\text{V}$  at 90 - 130 msec)

Intensity 16:  
b-wave: **5 / 4**  $\mu\text{V}$  at **138 / 144** msec  
(normal: 140 - 230  $\mu\text{V}$  at 80 - 120 msec)

**KW 26** Intensity 4:  
**+0.1 ND** x-wave: • / •  $\mu\text{V}$  (normal: 30 - 120  $\mu\text{V}$ )  
**(red)** b-wave: • / •  $\mu\text{V}$  at • / • msec  
(normal: 60 - 200  $\mu\text{V}$  at 96 - 130 msec)

Intensity 16:  
a-wave: **6 / 6**  $\mu\text{V}$  (normal: 20 - 50  $\mu\text{V}$ )  
x-wave: **7 / 8**  $\mu\text{V}$  (normal: 125 - 225  $\mu\text{V}$ )  
b-wave: **12 / 1**  $\mu\text{V}$  at **164 / 163** msec  
(normal: 150 - 240  $\mu\text{V}$  at 80 - 115 msec)

**KW 47, 47A+** Intensity 4:  
**47B & 0.6 ND** b-wave: • / •  $\mu\text{V}$  at • / • msec  
**+ 0.1 ND** (normal: 75 - 220  $\mu\text{V}$  at 90 - 125 msec)  
**(blue)**

Intensity 16:  
b-wave: **16 / 16**  $\mu\text{V}$  at **142 / 128** msec  
(normal: 150 - 250  $\mu\text{V}$  at 80 - 115 msec)

**Dark Adapted** Intensity 4:  
**Cone Response** b-wave: • / •  $\mu\text{V}$  at • / • msec  
**Red minus blue** (normal: 20 - 100  $\mu\text{V}$  at 40 - 50 msec)  
**Subtracted**  
**Waveform**

Intensity 16:  
b-wave: **8 / 9**  $\mu\text{V}$  at **49.5 / 48.5** msec  
(normal: 90 - 200  $\mu\text{V}$  at 45.8 - 48.2 msec)

**Bright White  
Scotopic  
Responses**  
(0.2 ND)  
+0.1 ND

Intensity 1: OP Wavelet Index: 2 / 2  $\mu$ V  
 a-wave: 4 / 3  $\mu$ V (normal: 30 - 60  $\mu$ V)  
 bx-wave: 13 / 7  $\mu$ V at 85 / 83 msec  
 (normal: 190 - 390  $\mu$ V at 40 - 90 msec)

Intensity 4: OP Wavelet Index: 2 / 2  $\mu$ V  
 a-wave: 6 / 4  $\mu$ V (normal: 70 - 130  $\mu$ V)  
 bx-wave: 10 / 11  $\mu$ V at 61 / 59 msec  
 (normal: 210 - 410  $\mu$ V at 40 - 80 msec)

Intensity 16: OP Wavelet Index: 2 / 3  $\mu$ V  
 a-wave: 22 / 23  $\mu$ V (normal: 100 - 180  $\mu$ V)  
 bx-wave: 8 / 9  $\mu$ V at 59 / 57 msec  
 (normal: 240 - 440  $\mu$ V at 40 - 65 msec)

**30 Hertz Flicker**  
**0.2 ND Intensity 8**  
(Background OFF)

(Right eye / Left eye)  
 Amplitude: 3.4 / 3.2  $\mu$ V Normal Subnormal Undetectable  
 Implicit time: 46.1 / 44.4 msec Normal Borderline Prolonged

**Photopic Responses**

**0.2 Neutral  
Density +0.1 ND**

Intensity 1:  
 b-wave: 0 / 3  $\mu$ V at 28.4 / 27.4 msec  
 (normal: 10 - 35  $\mu$ V at 22.3 - 30.0 msec)

Intensity 4:  
 b-wave: 4 / 2  $\mu$ V at 35.2 / 37.4 msec  
 (normal: 35 - 75  $\mu$ V at 21.3 - 30.0 msec)

Intensity 16: OP Wavelet Index: 2 / 2  $\mu$ V  
 a-wave: 3 / 5  $\mu$ V (normal: 15 - 65  $\mu$ V)  
 b-wave: 11 / 15  $\mu$ V at 50.8 / 51.8 msec  
 (normal: 60 - 200  $\mu$ V at 27.0 - 35.0 msec)

**30 Hertz Flicker**  
**0.2 ND Intensity 8**  
(Background ON)

(Right eye / Left eye)  
 Amplitude: 3.4 / 6.3  $\mu$ V Normal Subnormal Undetectable  
 Implicit time: 45.3 / 45.5 msec Normal Borderline Prolonged

**SUMMARY OF INTERPRETATION:**

This ERG, performed using intravenous propofol sedation, demonstrated severely subnormal responses of rods and both dark- and light-adapted cones, with severely prolonged cone implicit times. The maximum-intensity scotopic ERG responses was electronegative. The abnormalities of this study are those of severe rod-cone dysfunction with even greater loss of responses of bipolar cells compared to those of photoreceptors. This study is consistent with the type of ERG seen with infantile Refsum disease.

*Richard G. Weleber*

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