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November 6, 1998

Robert Steiner, M.D.
OHSU-Metabolics

RE: **Matthew A. Wulf**
MR#: 01-38-94-88

Dear Bob:

On November 4, 1998, I saw Matthew Wulf, the 11-month old child with infantile Refsum's disease. As you know, I

On examination, we were initially unable to elicit any fixation or following with toys or light but later on after the eyes were dilated, he actually would fix a light for brief moments for cycloplegic retinoscopy. At this point, his eyes looked to be straight, although earlier prior to dilation at times he looked to be slightly exodeviated. Cycloplegic retinoscopy was +1.50 sphere

OU. Prior to dilation of the pupils, they were definitely responsive to light but only weakly so. The anterior segments appeared to be essentially unremarkable. The optic nerve heads appeared to be pink with a very slight peripapillary pallor, the retinal vessels were possibly slightly narrowed, and the peripheral retina was hypopigmented. The macular regions showed no gross atrophy or discernible lesions but there were no foveal reflexes in either eye evident. Later in the morning, Matthew underwent an electroretinogram under propofol sedation. This study disclosed severely subnormal responses of both rods and cones that were typical for disorders of peroxisomal biogenesis. Matthew's responses were considerably better than those of his brother, Adam; however, Adam was first tested by ERG when he was 4-years of age.

I recommended that we re-examine Matthew in one year. The ERG could be repeated in the future as an objective measure of progression versus stability on his DHA supplementation.

Thanks very much for asking me to participate in Matthew's evaluation and care. Please let me know if I can be of any further assistance.

Sincerely yours,

Richard G. Weleber, M.D.
Professor of Ophthalmology and
Molecular and Medical Genetics

RGW/sg

encl: copy of ERG report
cc: Brian and Joyce Wulf