

OREGON HEALTH SCIENCES UNIVERSITY

University Hospitals and Clinics

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March 30, 1998

Robert Steiner, M.D.
Assistant Professor, Pediatrics
Chief of Metabolism

RE: WULF, MATTHEW
MR#: 01-38-94-88

Dear Dr. Steiner:

I had the opportunity to see this three and a half month old back for his infantile Refsum's disease. As you know, since his discharge, he has been getting some supplemental Enfamil 24 calorie per ounce. He takes three to four 3-ounce bottles a day. He nurses every two hours during the daytime except for a three and a half hour nap, and awakens twice at night. His stools are a little on the loose side, averaging three times a day, still with occasional "waxy" appearance which is probably steatorrhea although we have not proven this. His stools now have some odor. He is not vomiting. He had an upper respiratory tract infection for ten or twelve days and had no weight gain during that time but has since gained better, which his family attributes to giving essential fatty acid mix from a health food store 0.8 cc t.i.d.

From reconstructing his growth chart, he has gone from below the fifth percentile to above the fifth although his height by report is at the fiftieth. The family did not want to wake him up and re-weigh and measure him today.

My exam was limited for the same reason. He has some periorbital puffiness. His abdomen was soft and I could not appreciate liver or spleen.

His last liver function tests on March 9 revealed an albumin of 4.2, AST 464, ALT 232, alkaline phosphatase 751. His vitamin E level was a little low. His 25 hydroxy vitamin D was low and his essential fatty acid level is pending. His prothrombin time was normal at 12.8. He is currently on Mephyton 2.5 mg every other day plus a double dose multivitamin.

This young man with hepatic involvement and failure to thrive associated with infantile Refsum's is growing a little bit better but is still extremely thin. I would recommend that we increase the calories in his supplemental formula with MCT oil, adding 0.5 cc per ounce to give him a 28 calorie per ounce formula, and the family was sent to Dietary to get a bottle. I think it might be helpful to add vitamin E in the form of TPGS as liquid E, 1.5 cc q.d. This may enhance the absorption of his other fat soluble vitamins.

Since he has multiple appointments set up in the next couple of months, I will not set up a follow up appointment for Matthew. If his liver disease continues to be a significant problem, I would be happy to