

*Center for Research in Biochemistry and Molecular Biology  
Laboratory of Lipids and Human Brain Development*

*Dr. Manuela Martinez*

*University Maternity Children's Hospital Vall d'Hebron  
Planta 14, P. Vall d'Hebron 119-129, 08035 Barcelona, Spain  
Tel.: 34-3-4894065; FAX: 34-3-4894064; e-mail: <mmartinez@cinet.fcr.es>*

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TO WHO IT MAY CONCERN:

The patient Matthew Wulf, who has a generalized peroxisomal disorder, has come to this hospital to initiate treatment with docosahexaenoic acid ethyl ester (DHA-EE). On May 5th, 1998, the patient had DHA levels within normal limits, since he had already started to take DHA in triglyceride form (Neuramins), at a dose of about 200 mg/day. However, in an old plasma sample drawn on March 9th, 1998, the DHA levels were low, indicating that the child had a DHA deficiency. On arrival, the patient had moderate axial hypotonia, psychomotor delay and visual deficit, although the infant could see light, bright colors and fix on faces for a short time. Eye fundus examination did not find any alteration but the ERG was extinguished. Visual and brain stem evoked potentials were abnormal. The main clinical problem was failure to thrive and intense steatorrhea.

Treatment with DHA-EE, at increasing doses of 100-300 mg/day, was started on May 5th, 1998. Solid foods have been introduced progressively. The diet now consists of mother's milk supplemented with cereals, some fruit and chicken with rice. When he arrived, Matthew was fed "at libitum" but now he is having 7 meals a day, with fasting periods of 3-4 hours.

The tolerance of the treatment and diet has been excellent. Steatorrhea has virtually disappeared and Matthew has gained one kilogram of body weight in four weeks. Liver tests have improved and the very long chain fatty acids (VLCFA) 26:0 and 26:1 have continued to decrease (they started to fall since Matthew first started to take DHA), despite the marked increase in fat intake.


It is important for Matthew to follow a complete diet, as nutritious as possible for the age. The only restriction recommended is green leaf vegetables and the depot fat in meat. The mother is having a diet very rich in fish. This is providing Matthew with an important extra amount of DHA and should continue for as long as possible.

We did not perform any MRI examination here because this was apparently being planned in Oregon. Spectroscopy, in special, is indicated in Matthew, since MRI alone will not provide enough information at his age. Please, do this examination as soon as possible. When done, I would be grateful if you could send me copies of the slides and comments.

We are growing fibroblasts from Matthew but we did not perform any liver biopsy here. When they are grown I will send the results.

Matthew is on 300 mg/day of DHA ethyl ester just now and I think this dose will be maintained for a while but this will depend on the fatty acid changes. Please, send me a monthly blood sample from Matthew, drawn after 8 hours of fast, in an EDTA tube.

Please, contact me with any doubt you may have.



Manuela Martinez, M.D.