initiated the treatment of these patients with DHA ethyl ester (DHA-EE is the most pure form of DHA available nowdays, and it was provided to me by the NIH).

I want to make clear that the main rationale for the DHA treatment is the correction of DHA deficiency. Thus, it is in fact a physiological treatment and, as such, it must be given with the aim of normalizing things as much as possible. That is why I give the DHA-EE in the context of a normal diet and watch carefully the fatty acid changes that may be produced. If given with a low-fat diet, a relative excess of DHA may result in imbalance with other polyunsaturated fatty acids (PUFA). Furthermore, I have proved that a complete diet does not produce any increase in the VLCFA when accompanied by DHA-EE. On the contrary, a reduction is obtained. The same can be said of steatorrhea. For example, Mathew's steatorrhea passed from ++++ to + in just three weeks taking DHA-EE! Liver tests also improved dramatically. Since this was coincident with an increase (rather than with a decrease) in the fat intake, a probable role of DHA deficiency in the pathogenesis of the disease is suggested.

A most importan factor that has to be taken into account when speaking of DHA therapy is the time of initiation of the treatment. In the brain, DHA accretion starts to increase abruptly at 32 weeks of gestation, and continues at lower speed after the first months of life (see J. Pediat. 1992; 120: S129-138). In the retina, the time of DHA maximal accretion is even earlier (J. Neusci. Res. 1988; 20: 484-490). This means that the sooner we start the treatment the better. Unfortunately, this has been ignored by other doctors now giving DHA therapy to peroxisomal patients.

In the case of Matthew, he was almost 5-month-old when we started to give him DHA-EE. Fortunately for him, he was being breast-fed and his mother had started to give him another form of DHA since one month before he came here. So Matthew started to receive DHA therapy quite early (although not optimally so), and I think this has been the main reason why he has made a very good progress, at least until now. His general status, liver and weight improved dramatically. For the moment, his muscle tone and vision have improved frankly and I hope he will continue to improve neurologically. He is deaf but his hearing improves a great deal with hearing-aids. So I think it is important that he wears them am receives auditory stimulation.

In other children, we have found improvement in brain myelination during the treatment with DHA-EE (see Neurology 1998; 51: 26-32). Although it is still delayed, I hope myelination is also progressing in Matthew. If we compare with the MRI taken in June1998, more myelin has been formed since then. Relative to his age, a 4-month delay is less significant now than it was before, so it is possible that Matthew's MRI normalizes with time, as we have seen in other patients.

As for the protocol, I basically give DHA-EE plus a normal diet. Vitamin E is added as an antioxidant. Vitamin K is also given because these