August 31, 1998
From: FARAMARZ HIDAJI, M.D.

To: Gregory L. Smith, M.D.
8937 Southpointe Drive
Suite # C-2
Indianapolis, IN 46227-0998
RE: Nathan
Dear Dr. Smith:
I had the pleasure of evaluating your patient, Nathan, on August 24, 1998. Nathan is a four-year-old with a medical history of developmental delay and hypospadias. He is currently on no medications.
Nathan is here to see me because his parents are concerned that he may be having some visual difficulties. They state that they have noticed he has difficulty seeing in the visual periphery. He also has some difficulty seeing in dim light.
On my examination, we could confirm approximately 20/40 vision in Nathan with both his eyes open. Nathan had definite difficulty navigating in dim lighting. His dark adaptation was definitely prolonged. He would not allow us to cover his eyes in order to test each individually. Furthermore, his developmental level was not sufficient to allow testing of his depth perception. An examination of his eye movements showed excellent alignment at distance and near. I saw no nystagmus or abnormal head posture.
His pupils reacted briskly and equally to light. Slit lamp examination was completely normal. External examination was also normal.

After dilating drops were given, I checked a refraction which showed a normal amount of hyperopia for his age. Dilated retinal examination was normal.

In summary, Nathan likely has a retinal degeneration. Although he has no visible signs of such in fundus, at his age there may be no signs other than decreased peripheral vision and poor dark adaptation. As you may know, there are a number of hereditary retinal degenerations that co-exist with systemic findings. Disorders such as Refsum's disease, Spielmeryer-Vogt, Bassen Kornzweig, and Biedl-Bardet are examples of these. Nathan may also have his retinal degeneration as an isolated finding. Hereditary retinitis pigmentosa is such a disease.

I understand that Nathan had a normal hearing test. This effectively rules out Refsum's disease. Furthermore, he has no systemic findings consistent with Biedl-Bardet. Bassen-Kornzweig, or hereditary abetalipoproteinemia, has steatorrhea as an obvious component. Nathan does have diarrhea intermittently, though his parents do not describe it as severe and watery. Nevertheless, I recommended that Nathan take a multivitamin tablet daily.

His parents explained that Nathan is to undergo a full work up at the Riley Child Development Center. I think that this is an excellent idea for this young man, as he may very well have a treatable disorder that is causing his decreased vision, as well as his developmental delay.

Once Nathan is old enough, I would like to perform an electroretinogram to document the nature and extent of his retinal degeneration. Although the test can be performed under general anesthesia, this sometimes skews the results. I would, therefore, perform the test while Nathan is awake, which requires his cooperation.

I plan to see Nathan again in one year. If you have any questions regarding his care, please do not hesitate to contact me. Thank you very much, Dr. Smith, for allowing me to see this delightful young patient.
Faramarz Hidaji, M.D. FH/sjs
copy: 1) Riley Child Development Center

Attn: Barbara J. Weidenbener, B.A.
Riley Hospital for Children
702 Barnhill Drive - Room 5837
Indianapolis, IN 46202-5225