exposure from CT imaging would seem to outweigh any potential benefit.

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Early Coenzyme Q10 Supplementation in Primary Coenzyme Q10 Deficiency

TO THE EDITOR: Primary coenzyme Q10 deficiency is considered to be the only treatable mitochondrial disorder, since patients have a response to oral coenzyme Q10 supplementation. The disease usually manifests with nephropathy and encephalomyopathy.[1] It has been shown that oral coenzyme Q10 may stop the progression of encephalopathy, but no benefit from this therapy has been noted with respect to the evolution of renal disease associated with this deficiency.[1,2]

We now describe the results of long-term coenzyme Q10 supplementation in two patients with coenzyme Q10 deficiency caused by a homozygous missense mutation in the *COQ2* gene.[3,4] The clinical history of Patient 1 has been previously reported.[1] Briefly, corticosteroid-resistant nephrotic syndrome developed at 12 months of age, and progressive encephalomyopathy with strokelike episodes developed at 18 months of age. Coenzyme Q10 oral therapy was initiated at 22 months of age, and his neurologic picture improved, but no change in renal function occurred, since advanced chronic renal failure had already developed. He received a renal transplant at 3 years of age. He is now 7 years old, and his kidney-allograft function is normal, but he still has severe neurologic sequelae of encephalopathy, including cognitive impairment, seizures, and hemiplegia.

Patient 2, the sister of Patient 1, received a diagnosis of coenzyme Q10 deficiency before any symptoms developed at 12 months of age. Immediately after the diagnosis, the nephrotic syndrome developed with proteinuria (urinary protein excretion, 55 g per square meter of body surface per day), hypoalbuminemia (13.5 g of albumin per liter), and severe generalized edema. A renal biopsy specimen showed mild focal segmental glomerulosclerosis. Electron microscopy revealed podocytes that were particularly rich in mitochondria.[5] In addition to symptomatic treatment with diuretics, she received supplemental treatment with oral coenzyme Q10 at a dose of 30 mg per kilogram of body weight per day. There was no improvement during the first 2 weeks of treatment; an episode of acute renal failure required continuous hemofiltration for 4 days. However, 20 days after the initiation of coenzyme Q10 supplementation, we observed progressive recovery of renal function and a reduced level of proteinuria (Fig. 1). After 50 months of therapy, her renal function remains normal (creatinine, 41 μmol per liter [0.46 mg per deciliter]). Proteinuria is still present (1.2 g per square meter per day), but the levels of total serum protein (68 g per liter) and albumin (34.9 g per liter) are normal. Neurologic examination is also normal. Her stature is at the 50th percentile for age. The patient's parents declined permission to perform a second renal biopsy.

In this case, no corticosteroids or other immunosuppressive drugs were used. The response to coenzyme Q10 supplementation alone was associated with resolution of the nephrotic syndrome, and it suggests that coenzyme Q10 nephropathy should be considered when mitochon-

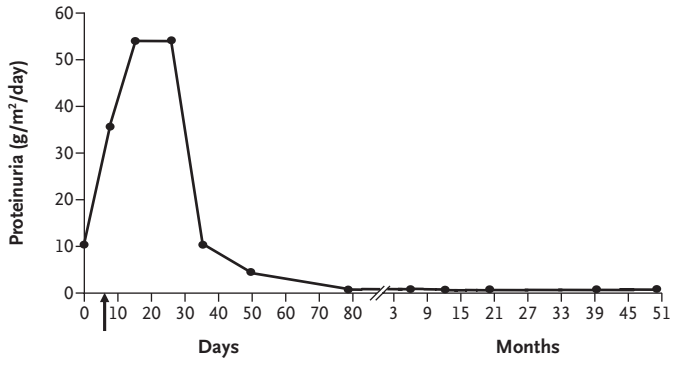

Figure 1. Proteinuria in Patient 2 during the 50-Month Follow-up Period.
The arrow indicates the initiation of coenzyme Q10 supplementation.

drial abnormalities in podocytes are present on electron microscopy. Specimens of the renal cortex, and possibly of cultured skin fibroblasts,[1,4] should be analyzed for coenzyme Q10 content. If coenzyme Q10 deficiency is present, genetic studies should be performed and coenzyme Q10 supplementation should be initiated. It appears that early administration of coenzyme Q10 was important for the resolution of renal symptoms and for preventing neurologic damage in Patient 2.

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