

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

Ephraim P. Hochberg, M.D.

Massachusetts General Hospital
Boston, MA 02115

Steven J. Morgan, M.D.

72 Highland Ave.
Salem, MA 01970

Judith A. Ferry, M.D.

Massachusetts General Hospital
Boston, MA 02115

1. Nishi Y, Suzuki S, Otsubo Y, et al. B-cell-type malignant lymphoma with placental involvement. *J Obstet Gynaecol Res* 2000;26:39-43.
2. Meguerian-Bedoyan Z, Lamant L, Hopfner C, Pulford K, Chittal S, Delsol G. Anaplastic large cell lymphoma of maternal origin involving the placenta: case report and literature survey. *Am J Surg Pathol* 1997;21:1236-41.
3. Kurtin PJ, Gaffey TA, Habermann TM. Peripheral T-cell lymphoma involving the placenta. *Cancer* 1992;70:2963-8.
4. Catlin EA, Roberts JD, Erana R, et al. Transplacental transmission of natural-killer-cell lymphoma. *N Engl J Med* 1999;341:85-91.
5. Maruko K, Maeda T, Kamitomo M, Hatae M, Sueyoshi K. Transplacental transmission of maternal B-cell lymphoma. *Am J Obstet Gynecol* 2004;191:380-1.

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 μ mol 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-

Giovanni Montini, M.D.

Azienda Ospedaliera
35128 Padua, Italy
montini@pediatria.unipd.it

Cristina Malaventura, M.D.

University of Ferrara
44100 Ferrara, Italy

Leonardo Salviati, M.D., Ph.D.

University of Padua
35128 Padua, Italy

Supported by a grant (005151 UBIGENES) from the European Union, a Telethon Italy grant (GGP06256), and a grant from Association Française contre les Myopathies (to Dr. Salviati).

1. Salviati L, Sacconi S, Murer L, et al. Infantile encephalomyopathy and nephropathy with CoQ10 deficiency: a CoQ10-responsive condition. *Neurology* 2005;65:606-8.
2. Rötig A, Appelkvist E-L, Geromel V, et al. Quinone-responsive multiple respiratory-chain dysfunction due to widespread coenzyme Q10 deficiency. *Lancet* 2000;356:391-5.
3. Quinzii C, Naini A, Salviati L, et al. A mutation in parahydroxybenzoate-polyprenyl transferase (COQ2) causes primary coenzyme Q10 deficiency. *Am J Hum Genet* 2006;78:345-9.
4. López-Martín JM, Salviati L, Trevisson E, et al. Missense mutation of the COQ2 gene causes defects of bioenergetics and de novo pyrimidine synthesis. *Hum Mol Genet* 2007;16:1091-7.
5. Diomedì-Camassei F, Di Giandomenico S, Santorelli FM, et al. COQ2 nephropathy: a newly described inherited mitochondrialopathy with primary renal involvement. *J Am Soc Nephrol* 2007;18:2773-80.

Correspondence Copyright © 2008 Massachusetts Medical Society.

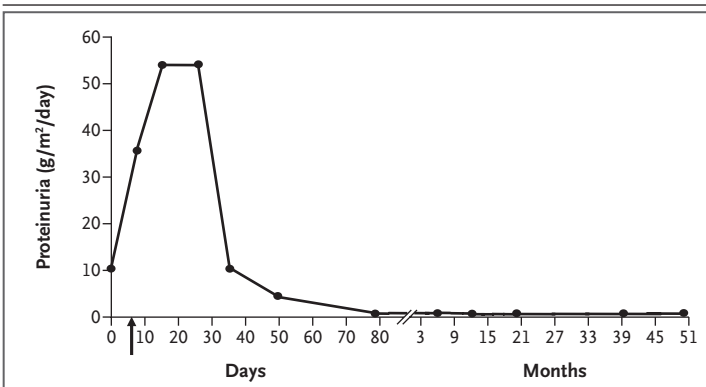


Figure 1. Proteinuria in Patient 2 during the 50-Month Follow-up Period.
The arrow indicates the initiation of coenzyme Q10 supplementation.

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.

INSTRUCTIONS FOR LETTERS TO THE EDITOR

Letters to the Editor are considered for publication, subject to editing and abridgment, provided they do not contain material that has been submitted or published elsewhere. Please note the following: •Letters in reference to a *Journal* article must not exceed 175 words (excluding references) and must be received within 3 weeks after publication of the article. Letters not related to a *Journal* article must not exceed 400 words. All letters must be submitted over the Internet at <http://authors.nejm.org>. •A letter can have no more than five references and one figure or table. •A letter can be signed by no more than three authors. •Financial associations or other possible conflicts of interest must be disclosed. (Such disclosures will be published with the letters. For authors of *Journal* articles who are responding to letters, this information appears in the published articles.) •Include your full mailing address, telephone number, fax number, and e-mail address with your letter.

Our Web site: <http://authors.nejm.org>

We cannot acknowledge receipt of your letter, but we will notify you when we have made a decision about publication. Letters that do not adhere to these instructions will not be considered. Rejected letters and figures will not be returned. We are unable to provide prepublication proofs. Submission of a letter constitutes permission for the Massachusetts Medical Society, its licensees, and its assignees to use it in the *Journal*'s various print and electronic publications and in collections, revisions, and any other form or medium.