20. Bonne G, Muchir A, Recan D, et al. Spectrum of mutations in lamin A/C gene implicated in autosomal dominant Emery-Dreifuss muscular dystrophy. *Neuromusc Disord* 1999;9:509.
21. Fisher DZ, Chaudhary N, Blobel G. cDNA sequencing of nuclear lamins A and C reveals primary and secondary structural homology to intermediate filament proteins. *Proc Natl Acad Sci USA* 1986;83:6450–6454.
22. Stuurman N, Heins S, Aebi U. Nuclear lamins: their structure, assembly, and interactions. *J Struct Biol* 1998;122:42–66.
23. Glass CA, Glass JR, Taniura H, et al. The α -helical rod domain of human lamins A and C contains a chromatin binding site. *EMBO J* 1993;12:4413–4424.
24. Hoyer TH, Krohne G, Kleinschmidt JA. Interaction of Xenopus lamins A and LII with chromatin in vitro mediated by a sequence element in the carboxylterminal domain. *Exp Cell Res* 1991;197:280–289.
25. Taniura H, Glass C, Gerace L. A chromatin binding site in the tail domain of nuclear lamins that interacts with core histones. *J Cell Biol* 1995;131:33–44.

NeuroImages

Figure. (A) Marked Kayser-Fleischer corneal ring superiorly and inferiorly. (B) Same eye 12 years after successful treatment with D-penicillamine. Note marked regression of Kayser-Fleischer corneal ring, which has virtually disappeared inferiorly. Note two unrelated small iris nevi of similar color at 4:00 and 7:30 on the iris (arrowheads).

Kayser-Fleischer corneal ring

Josef G. Heckmann, MD, PhD, Christoph J.G. Lang, MD, PhD, Bernhard Neundörfer, MD, PhD, Michael Küchle, MD, PhD, Erlangen, Germany

Editor's Note: This same *NeuroImage* was previously printed in black and white (*Neurology* 2000;54:1839). Because this figure is so instructive, it is being reprinted in color for the benefit of our readers.

A 26-year-old man was diagnosed with Wilson's disease in 1981. D-Penicillamine treatment was started but discontinued because of increased tremor. We first treated the patient in our ICU in 1987 for acute neurologic deterioration after mild brain injury caused by a fall. On admission, he was stuporous and unable to communicate verbally. We observed vertical gaze palsy, an increase in muscle tone, and a prominent circular Kayser-Fleischer corneal ring (figure, A). The laboratory findings revealed low serum

copper (400 $\mu\text{g/L}$), low ceruloplasmin (7 mg/dL), and elevated 24-hour urine copper (1403 $\mu\text{g}/1800\text{ mL}$ urine/day). Treatment with D-penicillamine was reinstated, along with physiotherapy and ergotherapy. The patient, now 44, is ambulatory and was recently readmitted for follow-up. Mild dysarthria, mild bilateral dysdiadochokinesia, and mildly elevated muscle tone of all four limbs were found, and the Kayser-Fleischer corneal ring had markedly regressed (figure, B). Serum copper (203 $\mu\text{g/L}$) and ceruloplasmin (3.9 mg/dL) were diminished. D-Penicillamine will be continued.

The presence of a Kayser-Fleischer corneal ring^{1,2} may correlate with treatment and markedly regress after successful decoupling.

1. Fleischer B. Zwei weitere Fälle von grünlicher Verfärbung der Hornhaut. *Klin Monatsbl Augenheilkd* 1903;41:335–352.
2. Finelli PF. Kayser-Fleischer ring: hepatolenticular degeneration (Wilson's disease). *Neurology* 1995;45:1261–1262.

Neurology®

Kayser-Fleischer corneal ring

Neurology 2000;55;280

DOI 10.1212/WNL.55.2.280

This information is current as of July 25, 2000

Updated Information & Services

including high resolution figures, can be found at:
<http://n.neurology.org/content/55/2/280.full>

Permissions & Licensing

Information about reproducing this article in parts (figures, tables) or in its entirety can be found online at:
http://www.neurology.org/about/about_the_journal#permissions

Reprints

Information about ordering reprints can be found online:
<http://n.neurology.org/subscribers/advertise>

Neurology® is the official journal of the American Academy of Neurology. Published continuously since 1951, it is now a weekly with 48 issues per year. Copyright . All rights reserved. Print ISSN: 0028-3878. Online ISSN: 1526-632X.