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NeuroImages

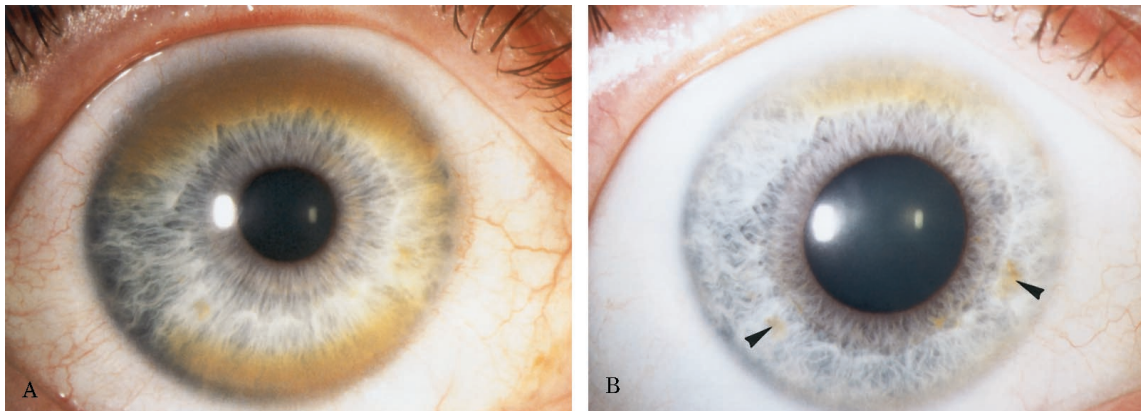


Figure. (A) Marked Kayser-Fleischer corneal ring superiorly and inferiorly. (B) Same eye 12 years after successful treatment with D-penicillamine. Note marked regression of Kayser-Fleischer corneal ring, which has virtually disappeared inferiorly. Note two unrelated small iris nevi of similar color at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

Kayser-Fleischer corneal ring

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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 decoppering.

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Kayser-Fleischer corneal ring

Neurology 2000;55;280
DOI 10.1212/WNL.55.2.280

This information is current as of July 25, 2000

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