Wilson’s disease (WD) is characterized by excessive accumulation of copper in the central nervous system, liver, kidneys, cornea, and other organs, leading to damage of the liver and brain. A Kayser–Fleischer (KF) ring, a brownish coloration of the outer margin of the cornea in the Descemet’s membrane, is characteristic of WD. Another rare ophthalmic manifestation of WD is sunflower cataract. Both KF rings and sunflower cataracts are best observed with an ophthalmic microscope.

An 8-year-old girl with WD presented with lethargy, abdominal distension, and bilateral edema that had been present for 6 months. On examination, we noted jaundice, abnormal liver enzymes, and prolonged coagulation time. The total and conjugated serum bilirubin levels were 125 μmol/L and 98 μmol/L, respectively. Alanine transaminase was 71 IU/L, aspartate transferase 96 IU/L, and the international normalized ratio 1.6. Liver biopsy showed excessive accumulation of copper, confirming the diagnosis of WD. Eye examination revealed KF rings on the corneas of both eyes, superiorly and inferiorly (Figure 1), and early sunflower cataracts in both eyes (Figure 2). Her vision was 6/6 in both eyes. The neurologic examination was normal. She was started on penicillamine 250 mg twice a day and was scheduled for follow-up.

KF rings and sunflower cataracts are both distinctive and diagnostically valuable signs of WD. KF rings are seen almost universally in adult patients with neuropsychiatric presentation; reports indicate that it is present in 98% of such patients. However, the frequency of KF rings in asymptomatic patients is low. Importantly, in children with no neurologic involvement, but who present with hepatic manifestations, KF rings are often absent. Sunflower cataract is a late manifestation of WD and should alert the clinician of possibly severe underlying disease. KF rings do not interfere with visual function. Similarly, sunflower cataract rarely interferes with vision. Both KF rings and
sunflower cataracts resolve with adequate chelation of copper.\textsuperscript{2,3}

Despite having an awareness of these signs, many clinicians may never have actually seen them, and may not note their presence when reviewing a patient having a possible diagnosis of WD. The images in Figures 1 and 2 demonstrate that these two signs are easily recognized on routine eye examination, even without the aid of ophthalmic microscopes. All that are needed to look for sunflower cataracts are a good light source and adequate dilation of the pupil. KF rings develop first in the superior cornea followed by in the inferior cornea, and subsequently in the 3 o’clock and 9 o’clock positions of the cornea.\textsuperscript{2}

References