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# Bilateral Dislocation of the Crystalline Lens in a Patient with Presumed Syphilitic Uveitis

Jeffrey S. Rapkin, MD,\* and David D. Bogorad, MD†

*Dislocation of the crystalline lens has been described in association with numerous systemic disorders and as a complication of ocular trauma. We present a case of bilateral lens dislocation in a patient with presumed syphilitic uveitis. The mechanism of lens dislocation is related to humoral and cellular effects of inflammation, although the role of trauma cannot be ignored. Persistent uveitis should be considered as a sign of active infection. (Henry Ford Hosp Med J 1986;34:207-10)*

Dislocation of the crystalline lens is a well-recognized complication of ocular trauma (1-3). In the absence of trauma this condition can occur in association with several recognized systemic disorders (1). With the exception of syphilis and syphilis-induced uveitis, dislocation of the crystalline lens has not been reported as a complication of uveitis. This case report presents a patient with a long history of nongranulomatous uveitis who developed bilateral dislocation of the crystalline lens with secondary acute glaucoma over a short period of time and in the absence of admitted trauma.

## Case Report

The patient is a 44-year-old woman, known to this clinic with a past history of uveitis. Her past medical history is significant for alcohol abuse and medical noncompliance. At the time of presentation her chief complaint was vision loss in the right eye.

On examination, the best vision in the right eye was 20/300 with an aphakic correction. The intraocular tension (IOT) was 12 mm Hg, and evidence of nongranulomatous uveitis without formation of synechiae was present. The crystalline lens was dislocated inferiorly (Fig 1). There was a hazy view of the fundus secondary to the inflammation.

Laboratory studies revealed normal electrolytes and urinalysis. The hemoglobin was 11.0 g/dL. The white blood cell count was 4,800, and the differential count was unremarkable. The Westergren erythrocyte sedimentation rate was 43 mm/hr. The chest X ray revealed no findings suggestive of sarcoidosis. The antinuclear antibody was negative, and HLA typing was negative for HLA B-27. The VDRL was negative. The treponeme hemagglutination was reactive.

The patient's uveitis was treated with topical steroids and cycloplegic agents. An infectious disease consultation was obtained. The patient was known to their clinic, since she had been treated for secondary syphilis in May 1983. At that time, she had received treatment with 18 million units of parenteral penicillin daily for two days and 12 g of parenteral ampicillin daily for six days. Against medical advice, she left the hospital before completing the prescribed ten-day regimen. At the time of this consultation, it was felt that the patient's syphilis had been adequately treated, and no further intervention was indicated.

Three weeks after presentation to our clinic, the patient returned complaining of a painful right eye. The visual acuity in the right eye was counting fingers, and the IOT was 56 mm Hg. The lens had become dislocated into the anterior chamber (Fig 2).

Intravenous mannitol was used to reduce the IOT. An intracapsular lens extraction and anterior vitrectomy were performed. A peripheral iridectomy was performed at the time of surgery. Postoperatively, the best vision in the right eye was 20/200. The IOT was 10 mm Hg, and there was persistence of a smoldering nongranulomatous uveitis. The vision was limited primarily by the presence of chronic cystoid macular edema with macular hole formation.

On initial presentation the vision in the left eye was 20/200. A minimal posterior subcapsular cataract was present; this had been noted previously and was unchanged from previous description. There was no evidence of dislocation of the lens nor of phakodonesis. Three months after presentation, during the patient's routine postoperative visit for the right eye, the left lens appeared minimally subluxed posteriorly along the superior border (Fig 3). There was minimal phakodonesis present. As in the right eye, the left eye showed evidence of nongranulomatous uveitis. The fundusoscopic examination revealed evidence of chronic macular edema.

One week later, the patient returned with "sudden vision loss" in the left eye. The best vision in the left eye was 20/70 with aphakic correction. The lens was significantly dislocated posteriorly, remaining fixed at the 6:00 position. There was hinge-like motion of the lens, whose location varied with head position. The location of the lens ranged from nearly normal to nearly horizontal. Consideration was given to elective lens extraction. However, because of the technical difficulty of such a procedure it was felt that a safer course would be to follow the condition without surgery. Dislocation of the lens into the vitreous cavity was considered the likeliest course of events, and this condition is well tolerated in the presence of an intact lens capsule.

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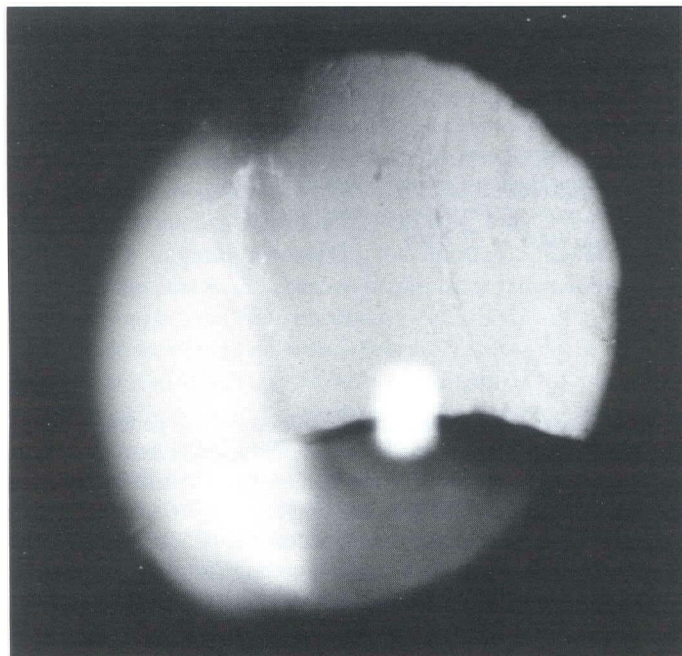
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One week later, the patient presented with a painful left eye. The vision was counting fingers, and the IOT was 42 mm Hg. The lens had become dislocated into the anterior chamber (Fig 4).

The patient was treated with intravenous mannitol to reduce the IOT. An intracapsular lens extraction with anterior vitrectomy and peripheral iridectomy was performed. Postoperatively, the best vision in the left eye was 20/100. Evidence of persistent smoldering nongranulomatous uveitis as well as chronic cystoid macular edema was present.

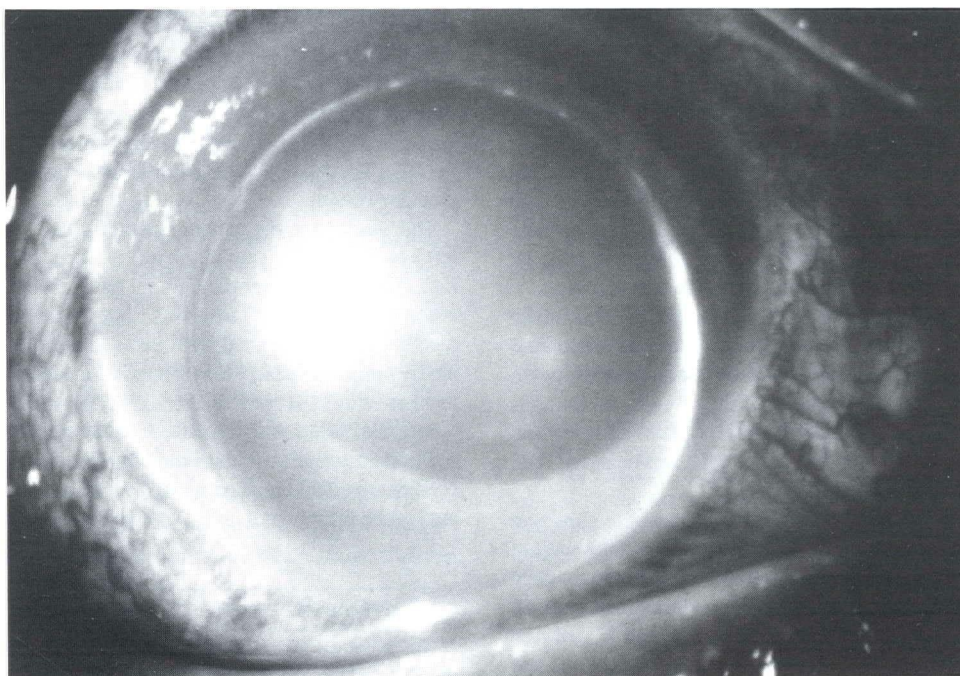


*Fig 1—Inferior dislocation of the crystalline lens in the right eye upon presentation.*

## Discussion

The association of dislocation of the crystalline lens with a number of systemic disorders is well recognized (1). The Table lists a number of the known systemic diseases that have been described in association with this condition. Marfan's syndrome is an inheritable abnormality of collagen biosynthesis. This syndrome includes skeletal and cardiovascular abnormalities as well as ectopia lentis. Homocystinuria is an inborn error of metabolism of the sulfur containing amino acids. The Weill-Marchesani syndrome is a rare familial condition. Both homocystinuria and the Weill-Marchesani syndrome have associated skeletal manifestations, as well as the ocular findings. With the exception of syphilis, the other disorders listed in the Table are either less common or more rarely associated with ectopia lentis.

The association of lens dislocation with syphilis has been retrospective (1,4-7). In patients with lens subluxation in whom other etiologies were ruled out, Smith et al (4) reported that 75% had positive serology for syphilis. However, in a controlled study, Rosenbaum and Podos (3) showed no significant difference in serology between patients with a subluxed lens and those in the control group. They suggest that this association is somewhat less common than is implied in other reviews. The etiologic factors contributing to dislocation of the lens in patients with syphilis is also unclear. It has been postulated that the humoral and cellular effects of uveitis weaken the zonules (7). The role of trauma, though, is not clearly defined. The high incidence of alcohol abuse in patients with positive serology is well known. These patients, therefore, are at higher risk for trauma. Histologic evidence of angle recession has been described in pathologic studies of eyes with dislocated lenses, which supports this theory (2).



*Fig 2—Dislocation of the lens (right eye) into the anterior chamber.*

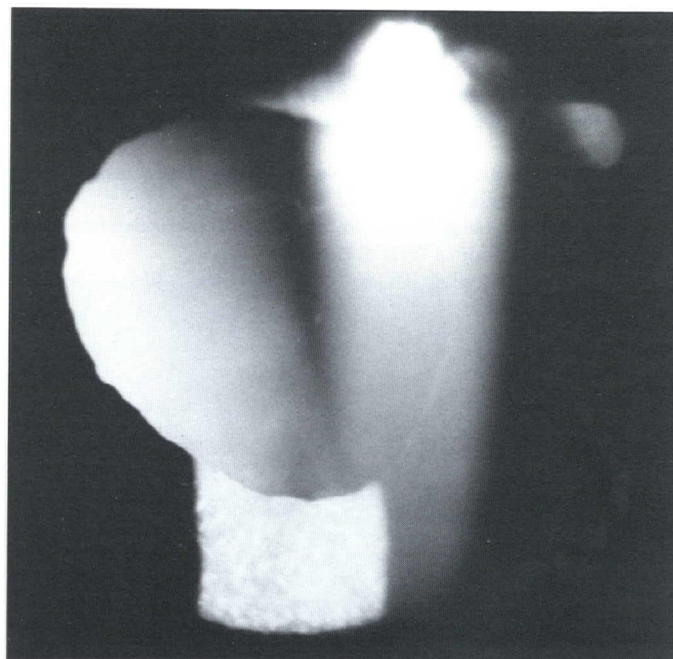


The findings of ocular syphilis have been reported (8-14). The presence of spirochetes in aqueous humor has been identified by various authors (4-7, 15, 16). Their presence has also been noted in patients felt to be adequately treated for syphilis (4, 15) and in patients whose serology (including the FTA-ABS) was negative (4, 6). This finding is considered evidence of active syphilitic infection. However, the sensitivity of this test has not proven to be high when examined in a prospective study (5). Immunofluorescent staining techniques failed to distinguish *Treponema pallidum* from other spirochetes, including the *Borrelia* species, which was felt to be a contaminant in the study by Ryan et al (5). Other, smaller spiral forms identified in this study were thought to be either glass shavings or species of *Vibrio* or *Spirilla* rather than true spirochetes. This test was not performed on our patient, since the decreased volume of aqueous humor present with

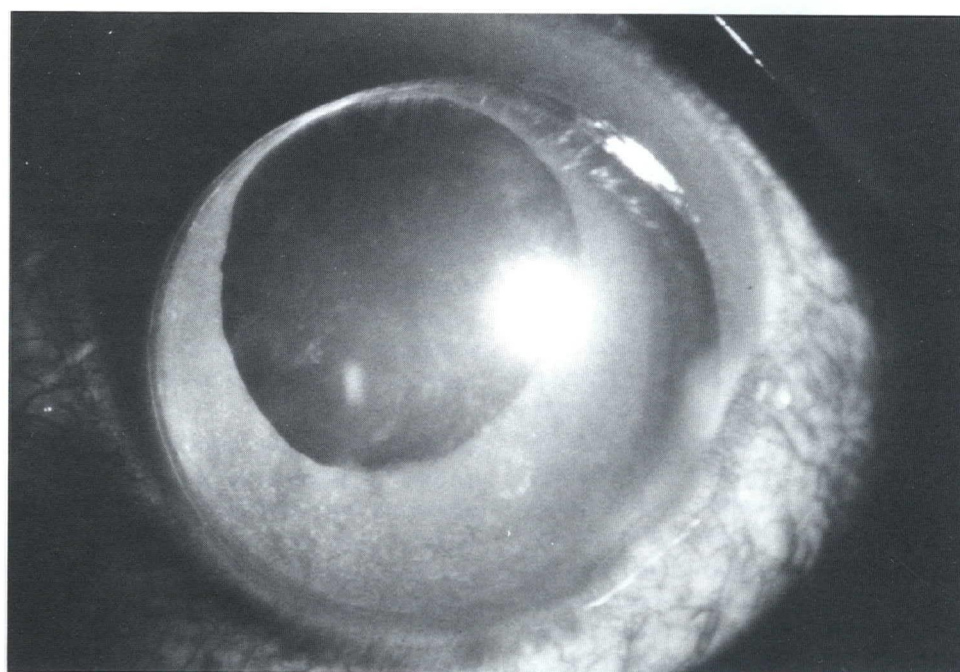
an anterior dislocation of the lens allows too small a sample size and greatly increases the risk of corneal endothelial damage secondary to the maneuver. Immediately following lens extraction, dilution of the aqueous humor would greatly reduce the validity of the test. Also, as indicated by Ryan et al (5), significant false-positive results are often reported in laboratories where the test is not frequently performed.

**Table**  
**Systemic Conditions Associated with Ectopia Lentis**

Marfan's syndrome
Homocystinuria
Weill-Marchesani syndrome
Hyperlysinemia
Sulfite oxidase deficiency
Syphilis
Sturge-Weber syndrome
Ehlers-Danlos syndrome
Crouzon's disease
Scleroderma
Mandibulofacial dysostosis
Refsum's disease



*Fig 3—Minimal posterior subluxation of the superior border of the left crystalline lens noted three months after initial presentation.*



*Fig 4—Dislocation of the lens (left eye) into the anterior chamber.*

We have presented a case of bilateral dislocation of the crystalline lens in a patient with syphilitic uveitis. The two events occurred over a short period of time and in the absence of admitted trauma. The presence of uveitis in a patient with supportive historical findings should raise the clinician's suspicion of active syphilitic infection.

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