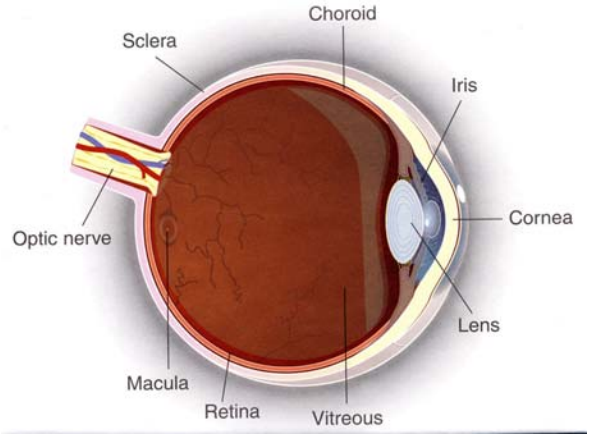
What You Should Know About Ectopia Lentis (Subluxated Crystalline Lens.)

By David J. Browning MD, PhD

Ectopia lentis is a medical term that means that the lens of the eye is out of its normal position. Normally the crystalline lens of the eye is centered behind the pupil and is held in position by radial fibers called zonules that connect the rim of the lens to the ciliary body, a circle of muscular tissue and secretory tissue found just behind the iris. When the muscle of the ciliary body contracts, the zonules relax and the crystalline lens becomes more spherical. When the muscle relaxes, the zonules tighten up and stretch the lens, making it flatter in curvature. In this way, the eye can change its focal length. Figure 1 shows the anatomy.

Figure 1. Eye Anatomy



What Causes Ectopia Lentis?

There are many causes for ectopia lentis. A normal lens may be subluxated after trauma. A blow can break some zonules, causing the lens to shift. Syphilis, a venereal infectious disease, can lead to zonular weakening and ectopia lentis. There are many genetic diseases associated with ectopia lentis. Some of these include:

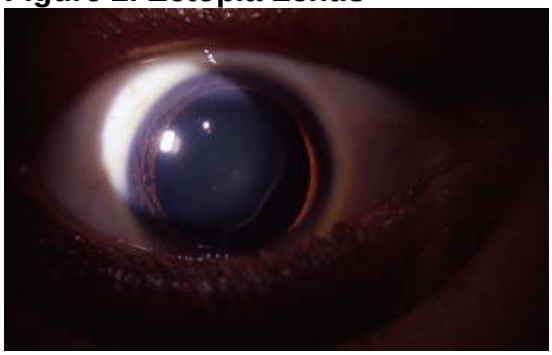
Marfan Syndrome- an autosomal dominant genetic disease in which a fibrillin gene mutation causes weak zonules as well as long fingers, retinal detachment, aortic valvular, and other systemic abnormalities. More information on this condition can be found under Information.

Homocystinuria- an autosomal recessive genetic disease in which a DNA mutation produces defective cystathionine beta synthetase. In addition to ectopia lentis, patients may be mentally retarded and have blood clotting problems.

Isolated Idiopathic Ectopia Lentis- these patients have ectopia lentis and no other abnormalities. Some patients have been discovered to have a fibrillin gene mutation distinct from that causing Marfan syndrome.

Much Less Common Conditions- Weil Marchesani syndrome, hyperlysinemia, sulfite oxidase deficiency, and familial ectopia lentis.

Figure 2. Ectopia Lentis



What Is Done For Ectopia Lentis?

When progressive shifting of the crystalline lens compromises visual acuity, the ectopic lens is surgically removed, most commonly by an operation called vitrectomy. Three one-millimeter incisions are made in the white sclera and instruments are inserted into the eye to remove the ectopic lens. In a person past puberty, most commonly a man-made intraocular lens is simultaneously inserted to focus light on the retina. In infants and children, most commonly no intraocular lens is inserted. Instead, wearing a contact lens or glasses restores focusing. Later, when full growth has been attained, an intraocular lens may be inserted in a secondary operation.

In children with ectopia lentis under the age of eight, it is important to monitor regularly to detect amblyopia, or failure of the brain to develop visual potential associated with a poorly focused retinal image. If amblyopia is detected, the ectopic lens should be promptly, surgically removed. In a large series of cases reported in the medical literature, surgery may be necessary at any age from a few months to the mid teens, with an average age of 5-6 years.

Final Comments

Ectopia lentis is a general term for a displaced lens in the eye. It can imply a systemic disease, and typically investigations are performed to check for this. These eyes can be predisposed to other abnormalities such as later high pressure (glaucoma) or a higher rate of retinal tears and detachments. All patients with ectopia lentis require life long periodic eye examinations both before surgical treatment and afterwards.

After you read this brochure, we encourage you to browse our website. If you have a focused question for which you cannot find an answer, we welcome you to ask Dr. Browning at: contact@retinareference.com. Also, an excellent resource for medical literature is Pubmed, on the National Library of Medicine website, accessible via a link on our website, or directly at www.pubmed.com.

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