the skin and sclera occur. This disorder was thought to occur in a specific geographic area that extended from the Mediterranean through the Middle East, India, and into Southeast Asia. Although a high gene frequency occurs in those areas, today thalassemia is known to have a worldwide distribution.

Because of prenatal hemoglobin F production, clinical manifestations do not appear until about the sixth month of life. Without treatment, 80% of the patients with the classic form die within 5 years. Among involved infants, the most common clinical features are pallor, irritability, growth retardation, abdominal swelling, and jaundice. Transfusions are used to prevent the associated anemia, and that has extended life expectancy. Now, a common cause of death is heart failure from myocardial hemosiderosis, related to the iron overload of repeated transfusions. More recently, chelating agents, such as deferoxamine, have had some influence on this.

Ophthalmic manifestations involve many parts of the eye. In the conjunctiva, multiple focal regions of dilated and tortuous vessels can be found. Often scleral darkening occurs secondary to hemosiderosis. Cataracts are not common but a few reports of posterior capsular opacities exist. Whether these lens changes were just coincidental, or disease related, remains unknown. Retinal abnormalities have been described most often in patients that have co-existing sickle cell disease. Nevertheless, the literature describes patients with thalassemia alone that have retinal vascular tortuosity, pigmented chorioretinal scars (black sunburst pattern), iridescent intraretinal deposits, focal retinal arterial occlusions, central retinal vein occlusion, retinal neovascularization, intraretinal hemorrhages, angiod streaks associated subretinal neovascularization, macular pucker, and macular ischemia. Intra-vitreal hemorrhages have been reported to occur secondary to the retinal vascular abnormalities. In addition to the above, patients requiring blood transfusions may develop retinal toxicity from iron accumulation or from the chelating agents, such as desferrioxamine. These retinal toxicities cause night blindness, visual field defects, and ERG changes.

The primary treatment of thalassemia is systemic therapy. Transfusions to prevent anemia are standard. However, such therapy causes an iron overload, hemosiderosis, and death. With the use of iron chelators, such as deferoxamine and desferrioxamine, this danger is lessened. Nevertheless, splenomegaly can be a problem and require splenectomy.

The most serious visual threats happen in patients that have both thalassemia and sickle cell trait. Such patients develop multifocal areas of peripheral retinal neovascularization and secondary vitreous hemorrhages. Angiography can identify the neovascular growth, and laser treatment can prevent such hemorrhages. Treatment of the surrounding ischemic retina can reduce the stimulus for recurrence.

It is important to be aware that ocular complications may indicate the presence of an additional hemoglobin abnormality. Treating that other hemoglobin abnormality may offer a significant benefit to such patients. When an intraocular hemorrhage, visual field defect, or retinal neovascular change is identified, a search for an additional co-existing hemoglobin abnormality is needed. If found, treatment directed to the manifestations of the other disorder can prevent additional visual loss.

Regular ophthalmic examinations are recommended for thalassemia patients to detect and treat possible ocular complications and to monitor iron toxicity.

**SUGGESTED READINGS**


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**PHAKIC INTRAOCULAR LENS FOR MYOPIA**

**Question: What Clinical Circumstances Would Lead You to Consider a Phakic Intraocular Lens for Myopia?**

**Answer:**

Phakic intraocular lens (IOL) for myopes is a hot issue these days. Lenses are available for implantation in the posterior chamber, fixation in the angle, and fixation to the iris. The ease of implantation, long-term safety and stability, early and late major and minor problems and their management, as well as the possibility of reversal of the procedure are important considerations.
Furthermore, tissue reaction can cause inflammation, degeneration, change in position, and raise intraocular pressure.

The earliest phakic lenses were implanted by Strampelli and Barraquer in the late 1950s. Serious corneal and angle related problems led to their abandonment. The issue was revived in the mid-1980s by Dveli (angle support), Baikoff (angle support), Fyodorov (posterior chamber), and Fechner-Worst (iris support). In the past two decades, the lenses have been redesigned and the surgeons worldwide have reported short follow-up results for all types of lenses.

The phakic IOL-containing eye has to be monitored at regular intervals throughout life. This is to prevent or treat any adverse development. A phakic IOL is not thought to be in conflict with corneal refractive surgery because it treats high to very high refractive errors, an area that is said to be out of reach for the latter. Sometimes corneal refractive surgery is used as an additional procedure to clear the remaining spherical and astigmatic errors.

Myopia is a common problem that starts at a young age. Unilateral myopia with or without amblyopia is a severe challenge for the parents as well as the surgeon. The young patients usually do not cooperate with the use of glasses or contact lenses, as they prefer to use the normal or better eye. There are cosmetic problems with the thick glasses as well as from the development of strabismus. Regular retina check-up is also essential.

The suitability of a patient for phakic lens implant has to be determined. The important questions are the minimum age of the patient, the maximum age of the patient, the maximum refractive error, the smallest size of the cornea, and the minimum depth of the anterior chamber permissible. Also, the surgeon must evaluate how accurate the white-to-white diameter corresponds to the required IOL diameter. It is essential that the surgeon explain to the patient the various types of lenses available, the various types of complications that can occur, and how they can be minimized or managed.

It is important that the anatomy of the tissues and the spaces that are concerned with phakic IOLs are understood clearly by the surgeon.

The pupillary margin normally rests on the crystalline lens. The contact is maximal in a mid-dilated condition. The posterior chamber, which has a mean volume of 65 μL, is a triangular space on cut section. The base of this triangle is toward the periphery and the apex is toward the pupillary margin (where the chamber depth is zero or near zero). Very little is known about the variations of this space and its relationship with age, the depth of the anterior chamber, or with the refractive status of the patient.

Anteriorly, there is a floppy iris about 0.5 mm in thickness; the thickness does not change throughout life. Posteriorly, there is the firm crystalline lens, which has a mean volume of 140 μL at birth. The mean volume increases to 163 μL in the 30s and to 240 μL in the 80s, a change of 100 μL.

The anterior chamber depth decreases by 7% every decade. It is the increasing size of the crystalline with age that causes shallowing of the anterior chamber. At the same time, it is not unreasonable to assume that the restricted posterior chamber space is progressively encroached upon by the increasing volume of the crystalline lens. By the time the patient is in the seventh decade, the space is nothing more than a mere slit. In the case of a posterior chamber lens, the thickest part of the implant, the optic, occupies the shallowest space in the posterior chamber.

If a plate haptic lens has to stay clear of the crystalline lens, it should vault in the central area. This can happen only if the haptic rests strongly on the ciliary processes and bows forward. Staying clear of the crystalline lens can lead to rubbing against the posterior pigment of the iris. It also means impinging on the ciliary epithelium of the ciliary processes. If the ciliary epithelium abrades, then the implanted lens will further touch the ciliary vasculature. The ciliary capillary endothelium has fenestrations of 30–100 μm, which are permeable to plasma proteins and tracer elements. The zonular fibers are also put under stress and might actually result in zonular lysis with time.

Lying between the crystalline lens and the iris, the phakic posterior chamber lens produces resistance to the flow of aqueous. The increasing volume of the crystalline lens with age encroaches on the posterior chamber volume. The average anterior chamber depth is 3.15 mm (2.6–4.4 mm). The mean volume of the anterior chamber is 250 μL, which decreases by 7.5% per decade.

The pathophysiology of the three types of IOLs differs. A short-angle supported lens will move. A large lens will impinge against the canal of Schlemm and corneoscleral trabeculae that overlie the scleral spur. It also might impinge nerve endings and blood vessels. Pressure on the segmental blood supply of the iris causes ischemia, atrophy, and progressive ovalization of the pupil.

An iris claw lens compresses iris tissue in its claw, which might cause subclinical inflammation and atrophy of the iris tissue inside the claw. In my experience, there are no angle-related or pupil-related complications.