# Surgical management of congenital cataract

**Essay**

**Submitted for partial fulfillment of master degree in ophthalmology**

**BY**

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# List of abbreviation

| | |
|---|---|
| **5-FU** | 5- Fluro uracil |
| **AAPOS** | American association for pediatric ophthalmology and srabismus |
| **Ac IOL** | Anterior chamber intraocular lens |
| **AMD** | Age related macular degeneration |
| **ASCRS** | American society of cataract and refractive surgery |
| **ASD** | Aquas shunt device |
| **BCVA** | Best corrected visual acuity |
| **BSS** | Balanced Salt Solution |
| **CALOs** | Congenital anterior lens opacities |
| **CCC** | Continuous Curvilinear Capsulorehxis |
| **CME** | Cystoid macular edema |
| **CRYAA** | Crystalline alpha A |
| **CRYBA1** | Crystalline beta A1 |
| **CT** | Computerized tomography |
| **ECCE** | Exstracapsular cataract exstraction |
| **ERG** | Electro retinogram |
| **FDA** | Food and drug administration |
| **I / A** | Irrigation / Aspiration |
| **ILO** | Intra lenticular opacfication |
| **ICG** | Indocyanine green |
| **LAL** | Light adjustable lens |
| **LECs** | Lens Epithelial cells |
| **LP** | Lens placode |
| **LV** | Lens vesicle |
| **MVR** | Microvitreoretinal |
| **NE** | Neural ectoderm |
| **OP** | Optic vesicle |
| **PAK** | Portable Autorefractometer |
| **PCCC** | Posterior Continuous Curvilinear Capsulorehxis |
| **PCO** | Posterior capsular opacification |
| **PCP IOL** | Posterior chamber phakic intraocular lens |
| **PFV** | Persistent fetal vasculature |
| **PHPV** | Persistent hyperplastic primary vitreous |
| **PMMA** | Polymethyl meta Acrylate |
| **RD** | Retinal detachment |
| **RGP** | Rigid gas permeable |
| **SE** | Surface ectoderm |
| **TMMC** | Trabeculoectomy with mitomycin-c |
| **TORCH** | Toxoplasmosis,Rubella,Cytomegalovirus,Herpes |
| **VAO** | Visual axis opacification |

# List of Figures

According to the most recent epidemiologic studies, the prevalence of visually significant cataracts diagnosed within the first year of life ranges from tow to three per ten thousands births **(Bhatti, et al., 2003).**

Cataract surgery in children younger than 2 years should be considered a 2-stage procedure in view of the higher incidence of PCO. Secondary glaucoma decreased significantly when surgery was performed after 30 days of age and the eye was left pseudophakic after surgery. Further improvements in IOL design, surgical instrumentation, and implantation techniques will continue to improve the ability to visually rehabilitate children **(Astle, et al., 2009).**

Surgery for congenital cataract at an early age increases the risk of glaucoma development, regardless of whether the eye is aphakic or pseudophakic. Intraocular pressure control with Ahmed valves is frequently required. Glaucomatous damage and dense amblyopia contribute to poor visual outcome in these eyes **(Kirwan, et al., 2009).**

Reasonably good postoperative visual acuity was achieved in the bilateral cataracts group, but relatively poor acuity in the unilateral cataract group. Poor stereopsis was recorded in both groups. One child developed glaucoma. None of the children developed retinal detachment. The expected myopic shift in the unilateral group compared with the bilateral group was demonstrated. Elective primary capsulotomy and elective anterior vitrectomy are mandatory to keep a clear visual axis **(Hussin, and, Markham, 2009).**

In spite of optimized care and surgery before 9 months, the BCVA was subnormal in our population compared to healthy children.

The long-term visual outcome can be predicted at 7 years of age. Screening with early detection followed by surgery before the end of the third month is important to decrease the risk of marked acuity loss **(Sjöstrand, et al., 2009).**

Preoperative examination with fully dilated pupils if necessary under anesthesia is mandatory in both eyes. This should include examination under operating microscope or slit lamp biomicroscope to assess the cataract, tonometry to rule out any association of glaucoma, measurement of corneal diameter, posterior segment evaluation, keratometry and biometry. The surgeon should look for a preexisting posterior capsule defect, which may turn out to be a camouflaged catastrophe **(Vasavada, et al., 2004).**

Options for optical correction following pediatric cataract surgery are primary IOL implantation, aphakic glasses and contact lenses. Primary IOL implantation has become a preferred approach in children above two years . IOL implantation is still questioned in children under two years as these eyes are most susceptible to intense PCO and excessive uveal inflammation **(Dahan, et al., 2000).**

The three piece acrylic IOLs may be preferred in order to decrease the rate of Nd:YAG capsulotomy after IOL implantation. one piece IOLs were developed more recently and are not as well studied as the three piece IOLs. Further study of the one piece IOLs, with longer follow up, may help identify the lens of choice. This study shows a higher incidence of Nd:YAG capsulotomy in patients who receive the one piece acrylic AcrySof lens compared to patients who receive the three piece acrylic AcrySof lens **(Mian, et al., 2005).**

Posterior capsular opacification (PCO) is by far the most common complication of cataract surgery in children. Various surgical techniques to reduce the incidence of PCO that have been described in the literature include primary posterior capsulotomy, posterior capsulorhexis with anterior vitrectomy despite the use of these techniques, PCO does occur, necessitating Nd:YAG laser capsulotomy **(Mitra, et al., 2003).**

# Aim of the work

To review the recent literature on the surgical management Of congenital cataract and to evaluate efficacy, complications, And visual rehabilitation.

# Embryology of the lens

The crystalline lens is derived embryologically from the surface ectoderm. Early in embryonic life, at about 2 weeks of gestation, the surface ectoderm overlying the optic vesicle (Figure 1.1A) thickens, forming the lens plate or placode (Figure 1.1B). The placode invaginates, forming a hollow ball of cells called the lens vesicle as the neuroectodermal optic vesicle concurrently invaginates to form the optic cup (Figure 1.1C, D) **(Tasman, et al., 2001).**

The lens vesicle is initially attached to the surface ectoderm, but it separates at about 4 weeks to lie within the anterior portion of the optic cup. The lens vesicle consists of a single layer of cells surrounded by a basal lamina, which becomes the lens capsule.

At about 5 weeks of gestation, the epithelial cells lining the posterior portion of the lens vesicle elongate anteriorly, forming the primary lens fibers. The formation of the primary lens fibers obliterates the cavity of the lens vesicle (Figure 1.1E, F) **(Tasman, et al., 2001).**

The proliferating equatorial epithelial cells are stimulated to differentiate terminally into mature, highly specialized lens cells, known as secondary lens fibers (Figure 2). The secondary lens fibers begin to form at about 7 weeks of gestation. Seven or eight mm long and hexagonal in cross section, these elongated, strap-like cells are joined together in a remarkably regular array of complex, interdigitating intercellular connections. Mature lens fibers lack nuclei and other organelles, and their homogeneous cytoplasm is filled with a concentrated solution of ordered crystalline proteins. The absence of vessels, lymphatics, nerves, and connective tissue, and the paucity of extracellular space in the lens also contribute to its transparency **(Tasman, et al., 2001).**

As new secondary lens fibers form, they elongate anteriorly beneath the lens epithelium and posteriorly beneath the capsule to envelope the embryonic nucleus. In the fetal nucleus, the tips of the fibers meet anteriorly and posteriorly, forming the Y-shaped lens sutures. The anterior Y is upright, and the posterior Y is inverted (Figure 2). The Y sutures lie within the fetal nucleus, just beneath the capsule at birth. As new concentric lamellae of secondary lens fibers form after birth, the sutural pattern becomes increasingly complex. A nine-branched stellate pattern is found in the adult nucleus at 20 years of age **(Tasman, et al., 2001).**

The lens continues to grow throughout life. New concentric lamellae of secondary lens fibers successively form in the periphery of the lens in an onion-like fashion. As new cells form, the older cells are sequestered centrally. The newer cells comprise the lens cortex, and the older cells constitute the nucleus (Figure 3). Lines of discontinuity form within the lens and represent the interface between layers. These lines create the typical slit lamp appearance of the adult lens (Figure 3) **(Tasman, et al., 2001).**

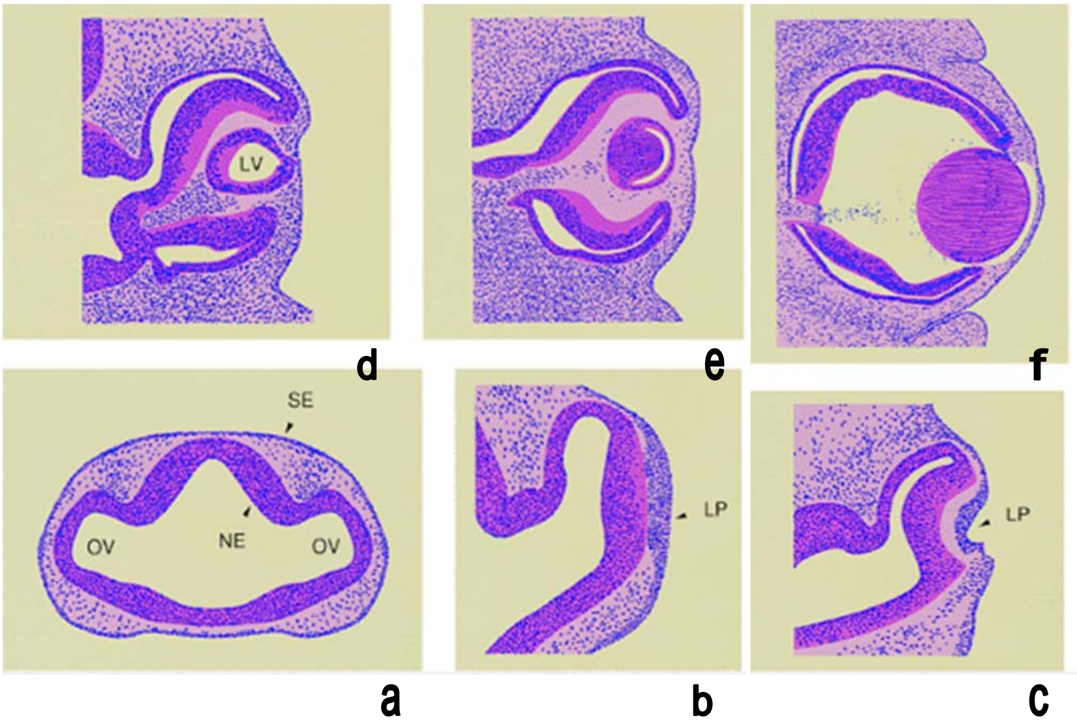


**Fig. (1): A:** Section through the forebrain of a 4-mm human embryo. The neural ectoderm (NE) is completely internalized, and lateral out-pocketings represent the optic vesicles (OV). The surface ectoderm (SE) is undifferentiated. **B:** The surface ectoderm opposite the optic vesicle has thickened to form the lens placode (LP) at the 4.5-mm embryo stage. **C:** The lens placode and the internalized neural ectoderm begin to invaginate at about the 7-mm embryonic stage. **D:** At the 10-mm embryonic stage, the lens vesicle (LV) has closed completely and is beginning to separate from the surface ectoderm. The optic cup has developed, but a space remains between the two layers of neural ectoderm. **E:** The lens vesicle has separated from the surface ectoderm and lies within the optic cup. The epithelial cells lining the posterior portion of the lens vesicle have elongated anteroposteriorly to form the primary lens fibers at the 15-mm embryonic stage. **F:** At the 18-mm embryonic stage, the lens vesicle is completely filled by the primary lens fibers. These fibers denucleate and will form the embryonal nucleus. **(Kleiman, and, Worgul, 1994).**