

Pediatric Cataract Surgery: Current Concepts.

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Abstract: Recent reports have shown a trend towards posterior chamber intraocular lens implantation for management of pediatric cataract. Surgery in very young children is best performed in a referral center by ophthalmologists with reasonable experience in this field. Decreased scleral rigidity and increased vitreous upthrust make surgical manipulations within these eyes more complex. The anterior chamber is often unstable; capsule management requires special technique, and the tendency for postoperative inflammation is more. Advances in technology has made surgery in children faster and safer. Modern vitrector units, viscosurgical devices with high viscosity, flexible single-piece acrylic IOLs, synthetic absorbable sutures, and newer radiofrequency diathermy and plasma blades have allowed delicate surgical maneuvers to be performed with greatest precision. Innovative surgical maneuvers such as optic capture and pars plana capsulectomy techniques have also added to the approaches that can be used in these complex cases. The consensus towards early surgery is becoming more popular. Visually significant cataracts may be successfully operated in newborns in order to prevent amblyopia. Careful planning of surgical procedure, selection of IOL and use of appropriate IOL power is must for pediatric cataract surgery. Primary posterior CCC with anterior vitrectomy helps to decrease the incidence of PCO and is particularly useful for pediatric cataract in children younger than 8 years. Most importantly, the words of Ellis¹⁰² are worth remembering, "Young children with an IOL in place are a unique clinical responsibility of an ophthalmologist. Long-term follow-up is especially important."

INCIDENCE

There are 1.5 million blind children (corrected visual acuity <20/400 in the better eye) in the world and one million of these live in Asia¹⁻³. The prevalence of childhood cataract has been reported as 1 to 15 cases in 10,000 children. It is estimated that globally, there are 200,000 children blind from bilateral cataract³.

ETIOLOGY OF CHILDHOOD CATARACT

The main causes of infantile cataract are genetic, metabolic disorders, prematurity and intrauterine infections^{4,6}. Other causes of childhood cataract in older children include trauma, drug-induced cataract, radiation therapy and laser therapy for ROP. Trauma is one of the commonest cause of unilateral cataract in the developing countries^{5,6}. Bilateral cataracts occur commonly due to the long-term use of topical or systemic steroid therapy. In industrialized countries, in approximately 50% of bilateral cases and virtually all of the unilateral cases, the underlying cause usually can not be determined^{1,3}. (Figure 1) Zonular cataract is the commonest type of congenital cataract.

OCULAR EXAMINATION

A thorough ocular and systemic examination is must in every child. Ocular examination should include visual acuity assessment, pupillary response and ocular motility. Biomicroscopic examination should be carried out in each case to evaluate the size, density, and location of cataract to plan the surgical procedure. Fundus examination should be carried out after pupillary dilatation. A-scan helps to measure the axial length for calculating IOL power and monitoring the globe elongation postoperatively. A history from the parents is useful to understand whether the cataract is congenital, developmental or traumatic in origin. One must ascertain if there is any history of maternal drug use, infection or exposure during pregnancy. Each child should be examined by a pediatrician for thorough systemic work up to rule out systemic associations, anomalies or congenital rubella.

LABORATORY WORK-UP

The work-up includes fasting blood sugar, urine for reducing substance for galactosemia after milk feeding and urine amino acids for Lowe's syndrome. Plasma phosphorus, red blood cells transferase



Figure 1.: Bilateral congenital cataract in children. These are one of the common causes of childhood blindness which is treatable

and galactokinase levels and calcium evaluation for hypothyroidism should be done. The titres for toxoplasma, rubella, cytomegalovirus and herpes simplex (TORCH titers) to rule out these disorders should be carried out. Genetic testing should be carried out for children with congenital cataract.

CATARACT SURGERY IN CHILDREN

Management of congenital and childhood cataracts remains complex. The physiology and anatomy of the growing eyes of children are so different from those of elderly adults that unique applications of modern technology are needed to maximize outcomes. Low scleral rigidity, increased elasticity of the anterior capsule, and high vitreous pressure are among the major obstacles that interfere with the highly demanding surgery. Even the best of surgeons can be humbled by the challenges of the small, soft, poorly developed eyes that require cataract surgery. Intraoperative problems coupled with a risk for

increased postoperative inflammation, a changing refractive state, higher re-surgery rate and an inherent risk of amblyopia make cataract surgery in children more complex and thus have an effect on final visual outcome. Historically, several primitive surgical techniques such as surgical iridectomy^{7,11}, needling and needling and aspiration were used for cataract surgery in children and a few of them are even today in practice in many parts of the developing world. As IOL designs and biomaterials have evolved, implantation in children has become safer and more acceptable, even for children in the first 2 years of life¹². Phacoaspiration with primary posterior capsulotomy with or without anterior vitrectomy and capsular bag implantation/optic capture of intraocular lens¹³⁻⁴¹ is today the most accepted technique in handling pediatric cataract surgery. The timing of the surgery is most important. The first 6 months of life, however, remains a controversial timeframe for IOL implantation. Preliminary results of a study of unilateral congenital cataracts indicate that IOL implantation in the first 6 months of life may produce better visual acuity, but at the expense of a higher complication rates^{42,43}. Unilateral cataract should be operated with in first 6 weeks of life to prevent development of deprivation amblyopia.

Visually significant cataracts not only produce a blurred image on the retina but also affect the development of visual pathways. In the 1970s, it was customary to defer infantile cataract surgery until at least 6 months of age. In sharp contrast, presently more and more surgeons recommend that visually significant cataract should be removed at the earliest possible time as sensory deprivation due to cataract in the first few months of life is critical^{42,43}.

HOW DOES PEDIATRIC CATARACT SURGERY DIFFER FROM ADULT?

The specific characteristics of the pediatric eye especially in children less than two years of age are: (1) intra-operative: scleral collapse, high intraocular pressure, elasticity of capsule for CCC, miosis, fibrin release, etc.; (2) post-operative: uveitis, PCO, secondary membrane formation, amblyopia; (3) long-term: growth of the eye and induced myopia.

A factor of important concern after the birth is the changing axial length of the globe. The eye of an adult is 40 to 50% larger than that of a child⁴⁴. The mean axial length of a newborn's eye is 17.0mm compared to 23 to 24mm in adult. The mean diameter of the crystalline lens is 6.0mm at birth and 9.3mm at 16 years⁴⁵. There is also change in the size of the capsular bag from 7mm at birth to 9.0 mm at two years⁴⁶. The resultant major problem is IOL power calculation particularly for infantile cataract. Most of the authors suggest under correction to prevent myopic shift⁴⁷⁻⁴⁹. In the growing eyes of monkeys and rabbits, lensectomy decreases the ocular growth as measured by axial length. In children less than 14 months of age with unilateral cataracts, the cataractous eye is shorter than the non-cataractous eye. In older children, (3-10 years) lensectomy and IOL accelerates the growth of the eye.

Use of currently available adult size rigid IOLs, particularly in small eyes in the first two years of life is associated with the problems of over-sizing. The incidence of post-operative complications such as uveitis, secondary glaucoma and posterior capsular opacification is also much higher in the pediatric age groups⁵⁰⁻⁵².

IOL POWER CALCULATION

Most reports have recommended under-correction of the IOL power for pediatric cataract, anticipating the myopic shift following IOL implantation^{47,48}. The axial length and keratometry readings should be measured for IOL power calculation in children. Dahan, et al⁴⁷

have suggested to aim for under-correction in children between 2 to 8 years by performing biometry and under-correcting by 10%. For children younger than 2 years, perform biometry and under-correct by 20% or use the axial length only. IOL power suggested for 21mm is (22.00D), 20mm (24.00D), 19mm (26.00D), 18mm (27.00D) and for 17mm axial length 28.00D.

CURRENT SURGICAL TECHNIQUES

The aim of the surgical technique is to provide a long term clear axis by preventing development of PCO or secondary membrane. Wound configurations that are self-sealing in adults often leak when used in children because of elastic sclera. Even the corneal tissue is less likely to self-seal in children. Suture closure of tunnel wounds and paracentesis openings is highly recommended. A cohesive viscoelastic like Healon GV is recommended for pediatric cataract surgery to facilitate anterior capsulorrhexis as they maintain anterior chamber stability, and help offset the low scleral rigidity and increased vitreous upthrust found in pediatric eyes. Runaway capsulorrhexis are more common in children as the capsule is very elastic but if completed uneventfully it resists tearing⁵³. In such situations comes the use of cohesive viscoelastics. Coupled to it is the centripetal force required while performing this maneuver. Capsular dyes, 0.1% trypan blue, can provide better visualization of the anterior capsule in children with total, white cataracts⁵⁴. The completion rate of successful anterior and posterior CCC has been shown to be higher when stained with trypan blue dye⁵⁵. Two other options are available for cutting the anterior capsule. The Kloti radio frequency endodiathermy has uses high-frequency (500 kHz) current to heat the probe tip to about 160 degrees and cuts the capsule using thermal energy⁵⁶⁻⁵⁹. The Fugo plasma blade which has been approved for anterior capsulotomy by the U.S. Food and Drug Administration (FDA).

is another new tool for performing an anterior capsulotomy in children^{60,61}. The Fugo blade has the advantage of not tearing or stretching the capsule without requiring a red reflex.

Hydrodissection: Hydrodissection is essential to ensure maximum removal of lens cortex and lens epithelial cells from the equatorial region. It may be a single site or multiple site hydrodissection⁶². It is performed by injecting ringer lactate or balanced salt solution in 2ml disposable syringe with 27-30 G cannula under the capsulorhexis margin. It should be avoided in cataract with posterior lenticulus or posterior polar cataract.

Cataract removal: Most of the pediatric cataract can be aspirated using two-way irrigation-aspiration (IA) canula or automated IA; however membranous or calcified cataract may need phacoemulsification. The best current techniques for pediatric implantation is basically those perfected over the years for adult surgery, but modified to meet the specific characteristic of the infantile eye. The cortical material is aspirated using two port irrigation-aspiration (IA). Two port IA helps to remove the cortex completely and it also maintains the anterior chamber during the procedure

Posterior continuous curvilinear capsulorhexis (PCCC): Posterior capsule opacification (PCO) is the most common complication after a successful cataract surgery in children. In younger children PCO is almost inevitable if posterior capsule management is not performed at the time of primary surgery. The PCO is amblyogenic and the purpose of surgery is defeated if long-term clear visual axis is not achieved. The general consensus is to perform a posterior capsulotomy especially in younger children⁶³⁻⁶⁵. We perform PCCC in children undergoing cataract surgery at age less than 8 years. When using acrylic IOLs with square edge with capsular bag implantation, some surgeons opt for a primary posterior capsulotomy only in

children younger than 4 years of age and do not perform it in children who are older. Technically a posterior capsulotomy can be achieved in several ways. Primary posterior capsulorhexis (PCCC) makes a capsulotomy safe because the smooth margin created at the opening resists peripheral extension of tears. Manual PCCC with the help of cystitome and forceps is preferable over other methods. Vitrector assisted posterior capsulotomy is also done in selected situation. Use of high viscosity viscoelastic helps to achieve PCCC. The desirable size of posterior rhexis is 3-3.5mm.

Anterior vitrectomy: Most surgeons prefer to perform anterior vitrectomy along with primary PCCC to decrease the incidence of PCO⁶⁶⁻⁶⁹. Anterior vitreous acts as a scaffold and helps in LEC migration and proliferation. The vitrectomy may be performed using limbal or pars plana route. We perform anterior vitrectomy using limbal route. In children the aim is to remove only central anterior vitreous in the posterior capsulotomy opening. Two port vitrectomy is ideal. It is performed from the side port incisions. Posterior capsulotomy (CCC) with anterior vitrectomy or posterior CCC with optic capture have revolutionized pediatric cataract surgery by providing clear visual axis.

Intraocular lens implantation: Capsular bag implantation of IOL is the best choice to reduce the contact of IOL with uveal tissue and to achieve IOL centration. The capsular bag is filled with viscoelastic agent and IOL is implanted into the capsular bag. Viscoelastic material is finally removed from the capsular bag and anterior chamber. Other options of IOL fixation which are equally effective in reducing PCO formation and achieving IOL centration are optic capture of IOL⁷⁰⁻⁷² with the haptics either in the ciliary sulcus or in the bag. Two techniques may be employed- one with the haptics in the sulcus and the optic captured either into both the anterior as well as posterior rhexis and secondly placing the haptics within the capsular bag and capturing the optic into the posterior rhexis. These maneuvers are difficult and should be performed only by experienced surgeons. A point of importance when considering optic capture of IOL is an adequate sizing of rhexis vis-a-vis the optic size. Too large a rhexis may lead to a lightly captured optic which may get de-captured later. Too small a rhexis may make the maneuver of optic capture very difficult as well as dangerous.

IOL selection and design: Presently, highly refined and perfected microsurgical techniques that have made lens implantation one of the most successful surgical techniques in history for adult cataract, are now providing highly favorable results in the field of pediatric cataract surgery. There is a clear swing towards implantation of IOLs over contact lenses for management of cataracts among children¹³⁻³⁵. Primary IOL implantation in young children becoming more acceptable as surgical techniques and biocompatible IOL materials and designs improve. Until recently, implanting PMMA IOLs were the only choice. (Figure 2) However, in one of the surveys, 69.0% of the respondents reported they were using hydrophobic acrylic IOLs⁷³. The trend away from PMMA in favor of acrylic for childhood implantation has been driven by the desire for a proven biocompatible material that could be inserted via a smaller incision. Three piece acrylic IOL are now routinely used for pediatric cataract surgery. Limitations with the three-piece design include kinking of the haptics and inadvertent sulcus fixation, which can be minimized with the use of the newer single-piece, AcrySof IOLs. The single-piece acrylic AcrySof SA series lenses ideal for implantation into the small capsular bag of children (Figure 3). The flexible haptics can be to have them in the desired position. However the single piece IOL's are not suitable for sulcus fixation. Multifocal IOL implantation is gaining ground for pediatric pseudophakia⁷⁴. Also, newer-generation multifocal IOLs



Figure 2.: PMMA IOL implanted in a child. Note the clear visual axis.

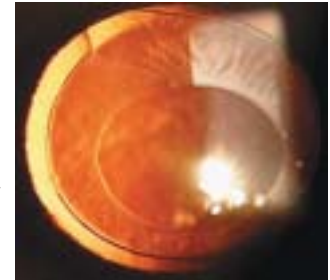


Figure 3.: New generation AcrySof SA 60 AT single piece IOL. The square edge design prevents formation of PCO.

and accommodative IOLs are the pipeline that will eventually provide the child with a functional alternative to the natural accommodation.

Incision closure: Because of lower scleral rigidity in children with a consequently greater risk of fish mouthing of the incision with resultant anterior chamber collapse, all incisions should be closed with a suture especially the main incision⁷⁵. Some surgeons advocate suturing of even the side port incisions. Wound closure may be performed using 10-0 nylon suture or a 10-0 polyglactin suture.

Secondary intraocular lens implantation

Since IOLs are now commonly implanted in infants during the first two years of life prior to the reaching of the adult size axial length of the eye, dioptric adjustment by the use of a secondary or exchange IOLs will become more common in the future.

Though low complication rates have been reported with Kelman open loop anterior chamber IOL implantation in adults⁷⁶, long term effect of ACIOLs for pediatric cataract is not known. We do not recommend this technique for pediatric cataract. Other techniques for secondary IOL implantation are placement of IOL in the sulcus-sulcus and bag-bag. There are several reports on the use of secondary IOL implantation for pediatric aphakia⁷⁷. A secondary posterior chamber IOL may be implanted in the sulcus or in the capsular bag if an intact bag can be salvaged and reopened at the time of secondary implants. The newly described piggy back technique, basically adding a new IOL with the appropriate power at the time of secondary implantation into the grown eye represent a future possibility- another example of applying techniques from the adult procedure.

Options in the Management of Pediatric Aphakia

The rehabilitation of pediatric aphakia is must to prevent further amblyopia and changes in the visual pathways. The options in management of pediatric aphakia include aphakic glasses, contact lenses, epikeratophakia and intraocular lens implantation.

In the past, optical correction of pediatric aphakia has been extremely challenging to the ophthalmologists. Aphakic glasses are unsatisfactory for rehabilitation because of several problems associated with their use such as induced magnification, visual field restriction and prismatic effect beside poor compliance. More over, use of aphakic spectacles for unilateral aphakia is impractical. Major goal of visual rehabilitation is to bypass the use of spectacles and strive for a much more practical regimen, the intraocular lens (IOL). Although contact lenses offer several advantages over aphakic

spectacles, such as full visual field and stereopsis, there are several problems associated with their use such as risk of infection, loss of the contact lens, higher cost and difficulty with compliance. Repeated insertion and removal of a contact lens may also be psychologically traumatic to the child.

OPERATIVE PROBLEMS AND COMPLICATIONS IN PEDIATRIC CATARACT SURGERY

There are several operative complications similar to cataract surgery in adults but a few specific complications for pediatric cataract surgery are: 1) difficulty in capsulorhexis formation, 2) positive intravitreal pressure, 3) intraoperative miosis, and 4) wound leak.

Most surgeons perform continuous curvilinear capsulorhexis (CCC) manually using needle cystitome or capsulorhexis forceps or "vitrector rhesis". Can-opener capsulotomy should be avoided to prevent occurrence of IOL decentration.

Constriction of pupil during surgery is a common problem in younger children. Pupillary dilatation is achieved using pre-operatively topical phenyl epinephrine hydrochloride 2.5% eye drops, cyclopentolate (0.5%), tropicamide (0.5%) and flurbiprofen (0.03%). Use of 0.3 ml of adrenaline (1:1000) in 500ml of irrigating fluid also helps to maintain pupillary dilatation. Most surgeons prefer scleral incision to a clear corneal incision in children. Wound closure may be performed using 10-0 nylon.

Postoperative Management

Postoperatively, a child's eye tends to show more tissue reaction. The inflammatory response can be managed with the use of intensive topical steroid (as frequently as six to eight times a day). The steroids are tapered over a period of six weeks. Topical antibiotics are instilled three times a day for 10 to 14 days. Cyclopentolate eye drops 0.5% or atropine eye ointment should be used for about four weeks to prevent posterior synechiae formation.

Postoperative amblyopia therapy should be instituted meticulously. Occlusion therapy for unilateral cataract after surgery should be instituted early as these children are at higher risk of developing amblyopia.

Postoperative Complications in Pediatric Cataract Surgery: The risk of post-operative complications is higher due to greater inflammatory response after pediatric intraocular surgery^{78,79}. Close follow up, early detection and management of complications is must.

Uveitis: Postoperative uveitis (fibrinous or exudative) is a common complication due to increased tissue reactivity in children. As a result of microsurgical techniques, closed chamber surgery and capsular fixation of IOL, severe postoperative inflammation is becoming less significant. (Figure 4) Frequent use of topical steroids and cycloplegics in the postoperative period helps to reduce uveitis-related complications.

Posterior Capsular Opacification: Posterior capsular opacification is a common cause of visual impairment after successful cataract surgery in adult^{80,81}. PCO is the most common complication after pediatric cataract surgery with or without IOL surgery^{82,83}. (Figure 5,6,7) PCO often occurs during the critical period of visual development and is amblyogenic. In a thick PCO, surgical posterior capsulotomy combined with anterior vitrectomy is required to prevent amblyopia. Nd: YAG laser has also been used to perform posterior capsulotomy in posterior capsule opacification⁸⁴. Use of newer surgical techniques like primary posterior CCC and anterior vitrectomy or posterior capsulotomy with endodathermy of capsule or posterior capsulorhexis with optic capture have shown encouraging

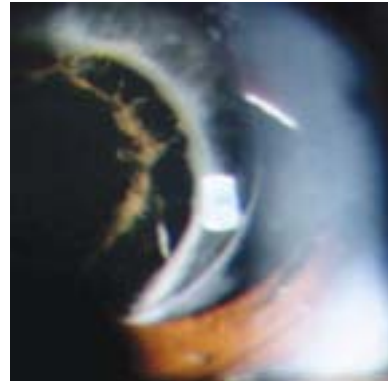


Figure 4. Pigment deposited on IOL surface following uneventful pediatric cataract surgery. The increased inflammatory response should be managed intensively with topical steroids.

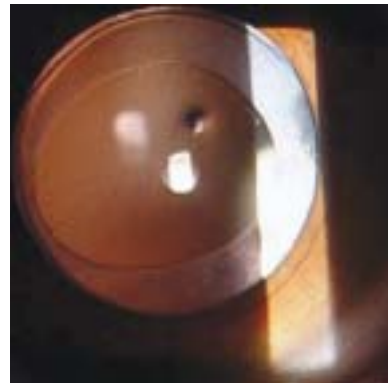


Figure 5. Early PCO formation in a child implanted with acrylic IOL (AcrySof SA60AT) IOL

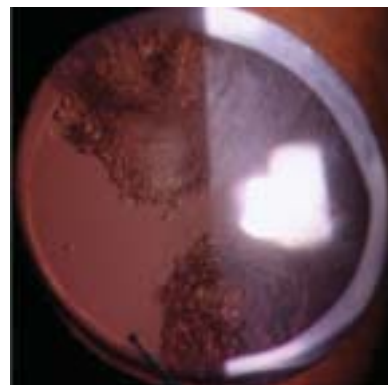


Figure 6. PCO (clump of lens epithelial cells) starting from the upper and lower periphery. The bunch of grapes pattern has spread its wings but is still sparing the centre where the visual axis is clear.

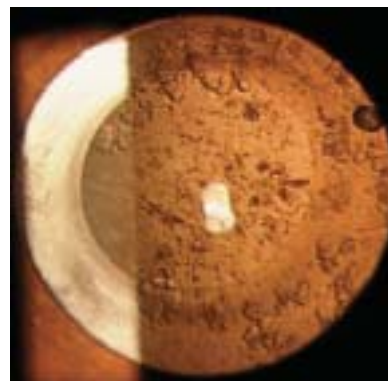


Figure 7. Florid PCO in a child implanted with PMMA IOL

results in maintaining a clear visual axis⁸⁵⁻⁸⁸.

Pupillary Capture: Pupillary capture occurs when a portion of the optic passes anterior to the iris. The incidence of pupillary capture among children is high, varying from 8.5% to 33%^{89,90}. Fixation of posterior chamber IOL in the capsular bag decreases the incidence of this complication⁹¹.

Decentration of IOL: Capsular bag placement of IOL is mandatory to reduce this complication⁹¹. Asymmetrical fixation bag-sulcus is to be avoided in order to minimize IOL decentration⁹¹.

Glaucoma: The incidence of glaucoma following pediatric cataract surgery varies from 3% to 32%⁵⁰⁻⁵². Glaucoma occurring soon after surgery is usually due to pupillary block or peripheral anterior synechie formation while open-angle glaucoma may occur late, which emphasizes the need for the life-long follow-up of these children. A peripheral iridectomy may prevent pupillary block in pseudophakic glaucoma. Intraocular pressure should be periodically recorded to detect and treat this vision threatening complication.

Secondary Membrane Formation: Secondary membranes are commonly reported after pediatric cataract surgery, particularly after infantile cataract surgery⁸⁹. Nd: YAG laser capsulotomy is sufficient to open them in the early stage⁸⁴. Dense secondary membrane may need membranectomy and anterior vitrectomy.

Retinal complications: Hemorrhagic retinopathy may occur following infantile cataract surgery. These hemorrhages are non-progressive and resolve within a few weeks. Cystoid macular edema is a rare complication following pediatric cataract surgery probably due to healthy retinal vasculature. The incidence of retinal detachment following cataract surgery has been reported between 1 to 1.5%⁹². Retinal detachments are usually a late complication of pediatric cataract surgery.⁹² The significant risk factors for an occurrence of retinal detachment are high myopia and repeated surgeries⁹².

Amblyopia: Pediatric cataract may also be associated with strabismus and/or nystagmus Amblyopia^{93,94} is one of the most important vision-threatening complications. The aphakic or pseudophakic child must be provided with suitable optical correction after surgery. The postoperative occlusion therapy of the normal eye in cases of unilateral congenital or developmental or traumatic cataract is done to achieve binocular vision and stereopsis⁹³⁻⁹⁵.

VISUAL OUTCOME

Several studies^{90, 96, 97} have reported good visual outcome following intraocular lens implantation in children. Pandey et al⁹⁸ found 85.5% of eyes operated for traumatic cataract and having PCIOLs achieved 20/40 or better visual acuity. Birch⁹⁹ compared the visual outcome of unilateral cataract cases operated during the first 6 weeks of life with those operated 2 to 8 years age. The visual acuity of children operated during the first 6 weeks of life was 20/40 compared to 20/100 for children operated at the age between 2 to 8 years. Bradford, et al¹⁰⁰ found visual acuity of 20/80 or more in 61% of children with an average postoperative follow up of 6.3 years in dense bilateral congenital cataract after surgery. They found that preoperative nystagmus; age at the time of surgery and postoperative nystagmus was not prognostically significant in visual outcome. The visual outcome and academic performance is also influenced by the presence of systemic abnormalities and mental status¹⁰¹.

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ETHICAL GUIDELINES FOR BIOMEDICAL RESEARCH

The need for uniform ethical guidelines for research on human subjects is universally recognised. It has acquired a new sense of urgency as the critical issues in the area of biogenetic research involving human subjects have become acute. Apart from the mandatory clinical trials on new drugs, a number of diagnostic procedures, therapeutic interventions and prevention measures including the use of vaccines, are being introduced which involve human subjects. Further the advent of new medical devices and radio-active materials and therapeutic benefits of recombinant DNA products have added a new dimension to the ethical issues that need to be considered before evaluating these for their efficacy, utility and safety.

Any research using the human beings as subjects shall bear in

mind the following principles of : (i) essentiality, (ii) voluntariness, informed consent, (iii) non exploitation, (iv) privacy and confidentiality, (v) precaution and risk minimisation, (vi) professional competence, (vii) accountability & transparency, (viii) maximisation of public interest and distributive justice (ix) institutional arrangements (x) public domain (xi) totality of responsibility and (xii) compliance.

Recent advances in the field of Assisted Reproductive technologies, organ transplantation, Human genome analysis and gene therapy promise unquestionable benefits to mankind. At the same time, they raise many questions of law and ethics, stimulating public interest and concern.

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