Ophthalmological Surgical Conditions in the Newborn and Neonate

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Siddharth Madan and Sarita Beri

6.1 Introduction

Eye is aptly labeled as a window to the body. It is among the most important of the basic senses that an individual possesses [1]. Loss of vision can have huge effects on the quality of life of a person. Therefore, tackling these diseases in the neonatal period is of utmost importance [2, 3]. The eye of a newborn baby differs from that in infants and adults in various aspects (Table 6.1).

Any intervention in a neonate and newborn raises a call for general anesthesia and vigilant monitoring. A preanesthetic evaluation (PAE) for medical and surgical fitness and clearance for administration of anesthesia is indispensible. Some ophthalmic conditions may have accompanying systemic comorbidities that make administration of anesthesia more challenging and difficult. This involves a battery of investigations as advised by the attending anesthesiologist, which may be time consuming. Moreover, various drugs used for anesthesia may have potential effects on the IOP and other toxicities that should be kept in mind.

6.1.1 Ophthalmic Conditions of Importance in the Neonatal Period that Require Anesthetic Attention, Include

1. Congenital cataract
2. Congenital glaucoma
3. Retinopathy of prematurity (ROP)

S. Madan
Department of Ophthalmology, UCMS and GTB Hospital, New Delhi, India

S. Beri (✉)
Department of Ophthalmology, Lady Hardinge Medical College, SSK and Kalawati Saran Children's Hospitals, New Delhi, India

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Table 6.1 Comparison of the Eye of a Newborn, Infant and Adult

	At Birth	Infancy	Adult
Orbital Volume	7 cc	16 cc	30 cc
Ocular Volume	2.8 cc	3.9 cc	6.8 cc
Palpebral fissure (Horizontal)	17 mm	23.5 mm	27 mm
Corneal thickness **(Mean Keratometry)**	581 microns 55 D	530 microns 47.5 D	510 microns 43 D
Lens Thickness	3.2 mm	3.4 mm	4 mm
Optic nerve length	24 mm		30 mm
Refraction	−4 to +6 D	−3 to +5 D	Emmetropization
Axial Length	16.8 mm	20.19 mm	23 mm
IOP	7.8–11.4 mmHg	10–15 mmHg	12–21 mmHg

4. Retinoblastoma
5. Child abuse and shaken baby syndrome and vitreous hemorrhage
6. Coloboma of the eyelid
7. Congenital dacryocystitis
8. Cryptophthalmos and Ankyloblehpheron
9. Congenital entropion
10. Keratomalacia, corneal ulceration, and
11. Trauma

6.2 Congenital Cataract

Congenital cataract may occur in isolation or associated with other congenital conditions. Childhood cataract accounts for 7.4–15.3% of childhood blindness and a significant amount of preventable disability-adjusted life years, with a prevalence of 1.03/10,000 children (0.32–22.9/10,000) and annual incidence of 1.8–3.6/10,000 [4]. In high-income economies' prevalence rate is 0.42–2.05 compared to 0.63–13.6 in low-income economies per 10,000 children. There is no difference in prevalence of cataract based on laterality or gender [5]. India has around 280,000–320,000 visually impaired children [6, 7]. It is associated with ocular abnormalities in 27% of cases and with systemic abnormalities in 22% of cases. The diagnosis is incidental, made on routine screening in 41% of cases, whereas leukocoria and strabismus are pointers in 24% and 19% cases, respectively. Management of pediatric cataract has changed in the past decade and there is an increasing trend toward surgical removal and implantation of an intraocular lens (IOL) in the neonatal period.

The most common cause of congenital cataract is genetic mutation with autosomal dominant pattern of inheritance in nearly 25% of cases. Other causes include chromosomal abnormalities [Down's syndrome (trisomy 21) and Edwards Syndrome (trisomy 18), Lowe syndrome], metabolic disorders [Galactosaemia (central oil-droplet like morphology of cataract), Wilson's disease, hypocalcemia, hypo/

hyperglycemia] and as part of syndromes associated with congenital infections (rubella, toxoplasmosis, cytomegalovirus, syphilis, and varicella zoster virus) [8, 9].

Clinical features include the absence of red fundal glow, nystagmus, and inability to fix and follow light. Evaluation of an underlying cardiac disease is important in newborns suffering from congenital rubella syndrome (CRS). Other manifestations of CRS include microphthalmia, glaucoma, retinopathy, iris atrophy, keratitis, and uveitis [10]. With the introduction of the measles, mumps and rubella (MMR) vaccine in 1988, the incidence of CRS has decreased significantly.

Congenital varicella syndrome due to maternal infection with varicella zoster virus in the 1st or 2nd trimester can cause cataract in about 2% babies [11]. Other features include skin lesions, neurological defects, skeletal limb deformities, microphthalmia, chorioretinitis, and optic atrophy [12]. Newborn is kept isolated from the mother until all her lesions have crusted and dried. When infection occurs near the time of delivery (5 days prior–2 days after delivery), risk of development of varicella in the newborn is considerable. Immediate treatment involves Varicella Zoster immunoglobulin and antiviral therapy (acyclovir). Cataract surgery can be undertaken once the disease becomes quiet.

Cataract may be associated with **microphthalmos**, where total axial length is 2 SD below similar age controls. In Nanophthalmos, a subtype of microphthalmos, both anterior and posterior segments are shortened, lens is enlarged and sclera is thickened. These have implications during cataract surgery as babies may have to be left aphakic with IOL implantation planned later.

Aniridia (hypoplasia or absence of iris) usually involves both eyes and is often associated with other ocular abnormalities including macular/optic nerve hypoplasia, cataract, glaucoma, and corneal opacification [13]. Sporadic aniridia exposes babies to increased risk of developing Wilm's tumor and, therefore, must be screened using serial ultrasounds of the abdomen. Treatment is in the form of lubricants, opaque contact lenses (to create an artificial pupil), and cataract surgery.

Cataract in microphthalmia is usually associated with persistent hyperplastic primary vitreous (PHPV) and requires intraoperative diathermy, need for pars plana vitrectomy along with lens extraction. In continentia pigmenti, underlying toxocariasis needs to be ruled out before proceeding with surgery as both conditions present with leukocoria (white pupillary reflex). Fundal screening is imperative under sedation or anesthesia. Other causes of leukocoria are retinoblastoma, ROP, vitreous hemorrhage, retinal detachment (RD), and persistent fetal vasculature.

Dislocation of the crystalline lens is associated with Marfan's syndrome. These babies usually have underlying cardiac disease, requiring 2-D Echo.

6.2.1 Management

Surgery is not necessary if the cataract does not obscure the visual axis. Unilateral cataracts need surgery at an earlier date as it has significant risk of development of recalcitrant amblyopia compared to bilateral cataract. Prognosis is better if cataract is diagnosed and treated before the age of 2 months. Intraoperatively in cataract

surgery because of the low scleral rigidity in Paediatric age group, the anterior chamber shallows and Vitreous pressure increases, making surgery difficult. Hyperventilation during General Anesthesia helps reduce the Vitreous pressure and allows easier anterior chamber manipulation.

Staphylomas may be congenital or comorbid in high myopia, although their actual incidence in pediatric population is unknown [14]. Intraconal eye blocks in congenital cataracts or when a staphyloma defect is suspected requires extreme caution. Posterior staphyloma (defect in the scleral wall) causes outward pouching of the pigmented retina, and this increases the risk of scleral perforation during needle blocks [15]. Extraconal or sub-Tenon block are safer options.

6.3 Congenital Glaucoma

Isolated trabeculo-dysgenesis (obstruction to the pathway for aqueous outflow) is the underlying cause for primary congenital glaucoma (PCG). This rare disorder contributes to 0.01–0.04% of total blindness. Incidence of PCG is 1:10 000 to 1:20 000 in western countries. The disease usually manifests at birth or early under the age of 3 years [16]. Gene mutation in PCG shows a recessive pattern of inheritance, with CYP1B1, MYOC, and FOXC1 genes being the commonest. The risk of developing PCG is more in consanguineous marriages. A classical triad of epiphora, photophobia, and blepharospasm is observed but seldom evident in neonates. Other features are buphthalmos (large eyeball) and corneal haze or red eye. A comprehensive ophthalmic EUA is done to confirm the diagnosis and IOP measurement. Most anaesthetics reduce IOP, and injudicious anaesthesia will reduce the IOP to an extent to mask raised IOP, affecting management. Ketamine is frequently used. A slight increase in IOP may occur, but this does not much interfere with management. Sevoflurane-based anesthesia is a good alternative, but IOP reading should be taken at the beginning of the induction of anesthesia as higher concentrations (>5%) and prolonged exposure may result in underestimation of IOP.

The congenital phacomatosis, a group of neuro-oculo-cutaneous disorders that include the Sturge–Weber syndrome, neurofibromatosis, tuberous sclerosis, and von Hippel–Lindau disease are all associated with ocular lesions and need surgery at some point in the course of their disease. Seizures, intracranial, and cardiac lesions necessitate appropriate PAE. Pheochromocytoma may rarely be associated.

Regional eye blocks have shown to provide better perioperative hemodynamics and less incidence of OCR and hence beneficial in syndromic babies with comorbid cardiovascular conditions [17, 18].

6.3.1 Management

Medical treatment with topical anti-glaucoma is usually supportive. The definitive treatment is surgery in the form of goniotomy, trabeculotomy, or trabeculectomy with/without anti-fibrotic agents, such as mitomycin C. Deep sclerectomy and

visco-canal-ostomy are alternative methods with satisfactory results. Glaucoma drainage implants and cyclo-destructive procedures have also been tried. The earlier detection and management of glaucoma has a good prognosis. However, the prognosis is worse when the disease presents at birth.

6.4 Retinopathy of Prematurity (ROP)

ROP is one of the most important causes of childhood blindness, worldwide, accounting for 3% of all childhood vision losses. ROP needs to be identified in the neonatal period before development of sight threatening sequel. Prematurity results in a failure of the vessels to reach the temporal periphery and increases the susceptibility of the retina to O_2 damage and halt in retinal vascularization. Newborns with a low birth weight are also susceptible to O_2-related damage as they may have increased periods of stay in the incubator. Increased metabolic demand of the eye stimulates excessive production of vascular endothelial growth factor (VEGF) that causes neovascular complications and blood vessels grow into the vitreous cavity with accompanying fibrovascular proliferation causes contraction of the latter and may cause partial or complete retinal detachment. Birth weight and gestational age are most important risk factors for ROP [19]. Other risk factors are early exposure to high levels of O_2, anaemia, sepsis, intraventricular hemorrhage (IVH), NEC, and mechanical ventilation. Differential diagnosis includes Norrie disease (inherited eye disorder leading to blindness in male babies at birth) and familial exudative vitreo-retinopathy.

6.4.1 Indications for Screening

With increasing number of NICUs and poor neonatal care, the incidence of ROP is on the rise. Early screening (2–3 week age) is recommended to enable early identification of aggressive posterior (AP-ROP), in India as per the national neonatology forum (NNF) [19]:

- Newborns ≤1750 g and/or <34 weeks
- Newborns <2000 g and 36 weeks with a bad postnatal period
- All newborns with birth weight (BW) <2000 g
- All newborns with gestational age <28 weeks or BW <1200 g

Western screening criteria is BW <1500 g or <32 week gestation.

6.4.2 Management

Goal of management is to convert the hypoxic retina into ischemic retina and thereby reduce the aberrant neovascularization from continuing and progressing into RD.

Screening is usually under topical anesthesia, in presence of a pediatrician and anesthesiologist, as babies can go into apnea.

Topical medications used to dilate the pupils vary from place to place. Commonly used combination is of tropicamide (0.4%) and phenylephrine (2.5%), in half dose as in adults. Higher dose can result in a hypertensive episode [19]. The initial dose is given 30–60 min prior to the examination. Atropine, a potent mydriatic, can result in severe gastrointestinal effects, tachycardia, flushing, and fever.

Topical local anesthetic application, just before the examination, helps reduce pain and discomfort of the corneal and conjunctiva from the bright light of the ophthalmoscope. Proparacaine is commonly used but excessive instillation can weaken the intercellular attachments of the corneal epithelium resulting in corneal haze. Administration of paracetamol or oral sucrose provides additional analgesia, but repetitive sucking by the infant can make the procedure difficult.

Treatment depends upon the progression and stage of ROP and zones involved:

i. **Cryosurgery** or cryotherapy was the initial treatment modality, but it was painful and was associated with significant postoperative pain and discomfort from conjunctival swelling. Hence, it was done under general anesthesia. Cryotherapy is not used anymore due to its complications.
ii. **Laser photocoagulation** is the gold standard of treatment today. It is performed under topical anesthesia, sedation or general anesthesia depending on the ophthalmologist, the stability, and cooperativeness of the baby. It is less painful and lasts approximately 30–40 min per eye. Babies are sensitive to the burns themselves, but the procedure also is stressful as it involves manipulation of the globe and discomfort with the bright light of the indirect ophthalmoscope, which predisposes them to development of apnea and bradycardia. Hence, it is important to monitor SpO_2 throughout the procedure [20–22]. General anesthesia reduces the procedure time, but comes with the usual complications. Sedation (e.g., chloral hydrate) and morphine (0.5 mg/kg) may be used.
iii. **Anti-VEGF treatment** promotes rapid regression of acute-phase of ROP, allows potential for retinal vascularization, approaches eyes with a rigid pupil, and minimizes stress of laser to the baby [19, 23, 24]. These potential benefits are a reason for their growing popularity in the management of ROP.
iv. Other treatment options include **vitreoretinal surgery** for severe ROP, under general anesthesia.

6.5 Retinoblastoma

Retinoblastoma, though rare, is the most common intraocular malignancy of childhood, contributing to 3% of all childhood cancers [25]. The peak incidence is in under 1 year age and most cases are seen in under 5 year age. Somatic type of retinoblastomas contributes to 60% of cases and presents at a later age. Remaining 40% are due to autosomal dominant mode of inheritance and retinoblastoma (Rb1) gene

mutation. Most common presentation is a white pupillary reflex, others being strabismus, change in color of the iris, and painful/red eye or orbital cellulitis [26].

6.5.1 Management

The tumor has a very low mortality rate if identified early and if timely treatment is initiated. The available treatment modalities are laser trans-pupillary thermotherapy, trans-scleral cryotherapy, laser photocoagulation, enucleation, external beam radiotherapy, and chemotherapy. General anesthesia is required for treatment in newborns and neonates and thus calls for an anesthesia backup. Eye blockade in the presence of retinoblastoma is relative contraindication. Inadvertent globe penetration during needle block may lead to extraocular seeding, and orbital retinoblastoma with distant metastasis, that necessitates adjuvant orbital radiotherapy after enucleation [27, 28].

6.6 Child Abuse and Shaken Baby Syndrome

Shaken baby syndrome (SBS) is a form of abuse in children under 6 months of age using physical force resulting in damage to the nervous system of the baby. In 78% cases it is associated with retinal hemorrhages, usually bilateral, severity correlating with the intensity of the abusive trauma [29, 30]. The underlying mechanism is acceleration and deceleration forces that cause rigorous movements and displacement of the vitreous, with resultant traction on the retina and its vessels, which eventually rupture and bleed. A classical triad of cerebral damage, subdural/subarachnoid hemorrhage and retinal hemorrhage is observed. The diagnosis requires retinal examination under sedation or general anesthesia.

6.6.1 Management

Pars plana vitrectomy might be required in cases with a non-resolving vitreous hemorrhage, macular hole, or underlying RD. Babies with an underlying intracranial bleed have poor prognosis. Ophthalmic and/or neurosurgical interventions require general anesthesia.

6.7 Coloboma of the Eyelid

Colobomas of the eyelid are rare malformations because of failure of the mesodermal folds of the eyelid to fuse during embryogenesis. This results in a triangular shaped defect usually located at the junction of the medial and middle 1/3 of the upper eyelid [31, 32]. They may be solitary or as a part of Goldenhar or Fraser

syndrome. Large eyelid defects can cause exposure keratopathy and development of corneal ulcers, potentially blinding.

6.7.1 Management

Correction of large upper eyelid defects in the neonatal period present unique challenges, because unilateral procedures on the eyelid may result in development of amblyopia. If possible surgery should be delayed till the age of 3 or 4 years, but in case of larger defects, a prompt, immediate surgery is advocated to prevent exposure keratopathy [33–35].

6.8 Congenital Dacryocystocele

Congenital dacryocystocele presents with a cystic distension of the lacrimal sac, of bluish appearance. The swelling is usually located just inferior to the medial canthus at birth. It can get infected and drainage with a reverse lacrimal sac massage is advocated. If contents of the sac get infected, surgical probing, drainage, and decompression are performed under general anesthesia, along with systemic antibiotics [36].

6.9 Cryptophthalmos and Ankyloblehpheron

Cryptophthalmos that is complete is characterized by complete covering of the eye with the eyelid skin. In incomplete/partial cryptophthalmos, the skin of the eyelid fuses with the conjunctiva or cornea. Bilateral cryptophthalmos is associated with Fraser syndrome [37]. There may be malformation of the underlying ocular structures.

Ankyloblehpheron presents with an adhesion of the edges of the upper and lower eyelids.

6.9.1 Management

Prognosis following a surgical repair has a good prognosis if underlying structures are normal [38].

6.10 Congenital Entropion

Entropion is defined as inversion of the eyelid margin and in-turning of the eyelashes. This requires surgical correction as the in-turned eyelashes can damage the cornea and cause corneal ulcers.

6.11 Keratomalacia, Corneal Ulceration

Vitamin A deficiency (VAD) due to malnutrition is endemic in developing countries in Southeast Asia and sub-Saharan Africa, where it is a leading cause of childhood blindness and accounts for 19–26% of cases of corneal blindness [39–41]. Over 5 million children develop xerophthalmia annually and one-fourth of these become blind. Keratomalacia following severe VAD, seen at 3–4 years is mostly associated with underlying malnutrition. However, VAD in neonates may occur from maternal deficiency of Vitamin A, thus stores are not built-up in the baby. VAD can be precipitated if the neonate develops severe diarrhea [42, 43]. Infection with virulent microorganism such as *Neisseria gonorrhea*, *Pneumococcus*, *H. influenza*, and *listeria* are responsible for penetration of the intact cornea and corneal perforation.

6.11.1 Management

A therapeutic graft may be required as a sight saving measure apart from retrieval of corneal scrapings for isolation of the infecting organism and initiation of appropriate antimicrobial therapy. Cornea transplantation is difficult in pediatric eyes as there are increased chances of graft failure. Optic iridectomy is an option apart from penetrating keratoplasty requiring a donor cornea. These procedures require general anesthesia in the neonate.

6.12 Trauma

Surgery in cases of trauma in children is indicated when the integrity of the globe is compromised with loss of intraocular contents. Excessive crying, coughing, Valsalva, or any maneuver that causes an increase in IOP will further complicate the integrity of the globe, exacerbating risk of extrusion of intraocular contents, and must be avoided. Regional anesthesia may be beneficial by avoiding the need of laryngoscopy, which itself may raise the IOP. Literature supports the fact that no difference in long-term visual outcomes is observed in patients with open globe injuries when comparing regional blockade versus general anesthesia [44, 45]. However, in neonates, general anesthesia remains the technique as it helps to lower IOP for comfortable intraoperative manipulations.

6.13 Conclusion

A multitude of eye disorders affect the newborns, and if not attended to timely, these may lead to vision disturbances, including blindness. Early identification of these conditions is based on a comprehensive history, clinical examination, and necessary ancillary investigations. Screening in newborns and neonates under general

anesthesia might be indicated for establishment of appropriate diagnosis. In neonates, adjunctive regional eye blocks, improve postoperative pain scores [46, 47]. They can reduce many risks of general anesthesia, opioid use, postoperative apnea, desaturation, and bradycardia [48, 49].

Intraconal block is relatively contraindication in neonates, because their extra ocular orbital volume is significantly low. In strabismus surgery or where muscles are being stretched, oculocardiac reflex can cause bradycardia, hypotension, arrhythmias, and asystole. Intraconal or regional blocks can be given to prevent the oculocardiac reflex by blocking the ciliary ganglion.

Extraconal nerve blockade is preferred in neonates. Its safety profile, with regard to adverse events (postoperative apnea and NICU admissions), has been demonstrated in ex-premature neonates undergoing vitreoretinal surgery [50]. Timely management can help preserve the vision of the baby in childhood.

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