INTRODUCTION

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Congenital and developmental eyelid abnormalities are among the most challenging problems encountered by the reconstructive surgeon. Eyelid abnormalities in children may present at birth as a result of abnormal embryogenesis (congenital) or they may occur at later stages as the child matures (developmental).[1]

Embryology of eyelids

Eyelids development is characterized by three main stages. *The first stage* is the initial development in which the optic vesicle is covered by a thin layer of surface ectoderm during the first month of embryonic development. During the second month, active cellular proliferation of the adjacent mesoderm results in the formation of a circular fold of mesoderm that is lined by ectoderm. This fold constitutes the rudiments of the eyelid, which gradually elongates over the eye. The mesodermal portion of the upper lid arises from the frontal nasal process, the lower lid from the maxillary process. The covering layer of ectoderm becomes skin on the outside, conjunctiva on the inside. Tarsal plate, connective, and muscles of the eyelids are derived from the mesodermal core. *The second stage* is fusion of the eyelids by an epithelial seal begins at the two extremities at 8 weeks and is soon complete, covering the corneal epithelium. The eyelids remain adherent to each other until the end of the fifth to the seventh month. The third stage is final reopening in which a separation begins from the nasal side, and is usually completed during the sixth or seventh month of development. Very rarely, this process is incomplete at birth in a full-term infant .The specialized structures in the lids develop between 8 weeks and 7 months. By term, the lid is fully developed.[2]

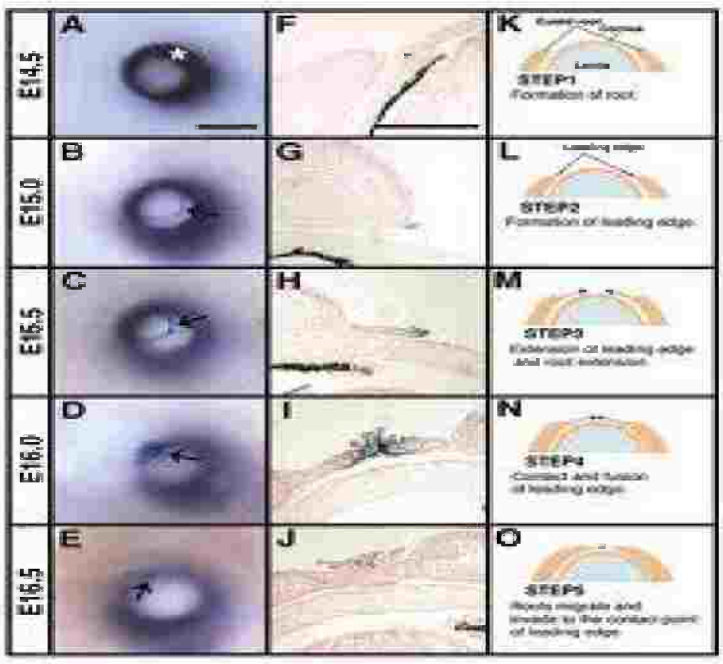

Figure (1): Showing epithelial cells migration during eyelid development.[3]

Anatomy of eyelids

The eyelids have several characteristic horizontal and vertical folds. The most conspicuous is a well-demarcated horizontal skin crease 3–4 mm above the upper lid margin, which flattens out on depression and becomes deeply recessed when the upper lid is elevated. It divides each lid into an orbital and tarsal portion. The orbital portion lies between the margin of the orbit and the crease; the tarsal portion lies in direct relationship to the globe. A tarsal plate composed of dense connective tissue is found in both the upper and lower eyelids. The upper lid tarsal plate has a marginal length of 29 mm and is 10–12 mm wide. The lower lid tarsal plate is 4 mm wide. The palpebral fissure is the entrance into the conjunctival sac bounded by the margins of the eyelids; it forms an asymmetrical ellipse that undergoes complex changes during infancy. After birth, the upper lid has its lowest position with the lower eyelid margin close to the pupil center. Between ages 3 and 6 months, the position of the upper lid reaches its maximum. The distance between the pupil center and the lower eyelid margin increases linearly until age 18 months.[4]

The principal muscle involved in opening the upper lid and in maintaining normal lid position is the levator palpebrae superioris. Müller's muscle and the frontalis muscle play accessory roles. *The levator palpebrae superioris* arises as a short tendon blended with the origin of the superior rectus from the undersurface of the lesser wing of the sphenoid bone. The levator palpebrae superioris is innervated by branches from the superior division of the oculomotor nerve. *Müller's muscle* is a thin band of smooth muscle fibers 10 mm in width that arise on the inferior surface of the levator palpebrae superioris. It courses anteriorly, between the levator aponeurosis and the conjunctiva of the upper eyelid to insert into the superior margin of the tarsus. Branches of the sympathetic nerve innervate Müller's muscle. The eyelid is indirectly elevated by attachment of the frontalis muscle into the superior orbital portions of the orbicularis oculi muscle. The frontalis muscle is innervated by the temporal branch of the facial nerve.[5]

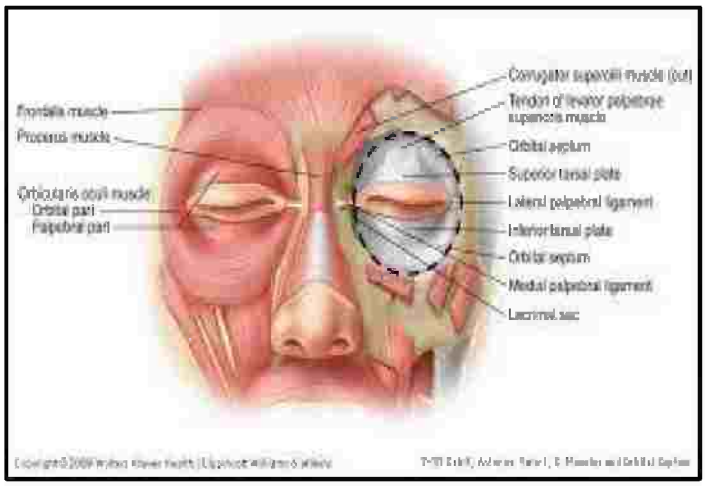

Figure (2): Showing eyelid muscles.[6]

The lacrimal glands are formed from epithelial buds arising from the basal epithelial layer of the conjunctiva. Thus canalization of the lacrimal duct begins at the third month of gestation and is usually completed by the sixth month, but a persistent closure of the lower portion of the duct at birth may result in congenital nasolacrimal duct obstruction.[7]

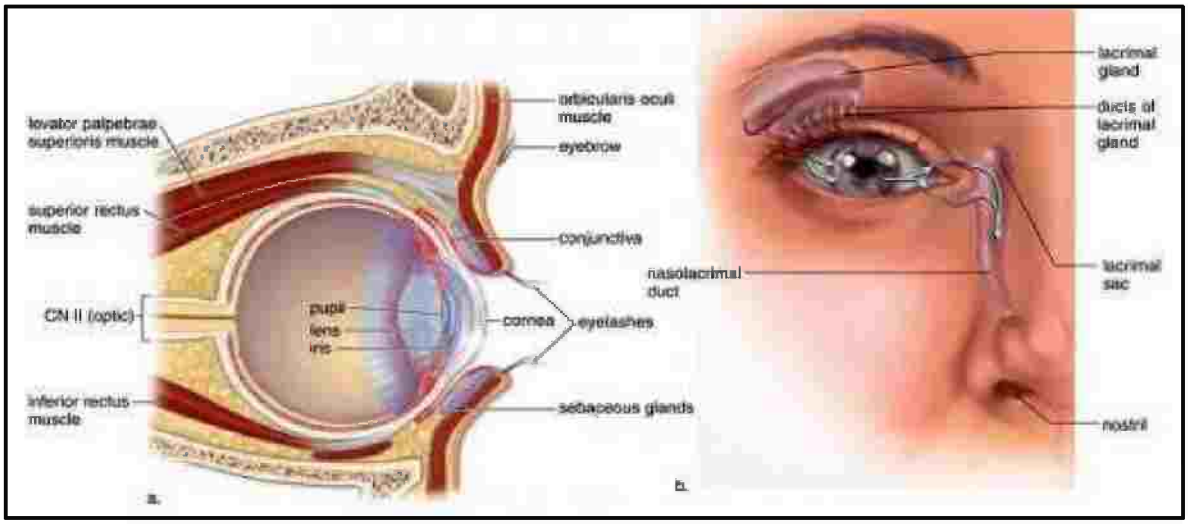

Figure (3): Showing lacrimal gland, lacrimal sac &duct. Sebaceous gland also seen on the left.[8]

Categories of congenital anomalies

Four categories of congenital anomalies have been proposed, including those resulting from:

1. Single morphogenetic defects.
2. Intrauterine mechanical constraint on an otherwise normal embryo or fetus.
3. Destruction of a normal structure.
4. Dysplasia, defined as a defect in the differentiation and organization of a tissue. Many of these dysplasias are caused by a single morphogenetic anomaly in development that leads to a subsequent series of defects, defining a sequence or syndrome.[9]

Anomalies

Tearing (Nasolacrimal duct obstruction)

Excess tearing from the eye is not uncommon in children. Unfortunately about 5% of infants are born with a congenital blockage of the tear drain. The tear drain is located in the inner corner of the eyelids and runs into the nose. When this is present, babies will develop excess tears dripping from the eye along with occasional discharge form the eye and crusting along the lashes. The good news is 90% of these blockages open spontaneously during the first year of life. During this time period conservative treatment with massaging the tear system and topical antibiotics are the mainstay of treatment. If the problem continues beyond the first year of life or if a severe infection occurs during the first year of life, a surgical procedure can be performed to open up the tear drain, which is highly successful. Sometimes extra procedures are needed like placing a stent or stretching the tear drain with a balloon to help establish normal flow down the drain.[10]

Centurion syndrome

Deriving its name from the resemblance of the patients to the Roman centurions in their helmets, Centurion syndrome is an uncommon, idiopathic medial canthal anomaly that causes epiphora due to the forward displacement of the lacrimal punctum out of the tear lake associated with the abnormal anterior insertion of the medial canthal tendon and enophthalmos. The facial appearance is typical, with a sharp medial canthus, long

horizontal palpebral fissure and square eyebrows. Lacrimal syringing comes patient but dye disappearance is delayed. The treatment is surgical, anterior medial canthal tendon release with medial canthoplasty or punctoplasty.[11]

Congenital ptosis

A drooping of the upper eyelid may be unilateral or bilateral, with more severe forms involving hypoplasia of the levator palpebrae superioris muscle or tendon with a minimal or absent eyelid crease. Symptoms occur on a spectrum, from no visual or cosmetic disturbance with mild ptosis, to visual impairment resulting in compensatory positioning of the head, difficulty in daily activities, and significant cosmetic deformity with severe ptosis. If the lower margin of the upper eyelid falls to or below the center of the pupil, vision may be affected. This often causes children to adopt a chin-up posture for viewing objects straight ahead, with resultant difficulty ambulating, or they may arch their eyebrows using forehead muscles to partially elevate their eyelids. Early surgical correction of ptosis is often indicated to prevent amblyopia, but in less severe cases surgery may be deferred to affect a more favorable outcome. Some children who have congenital ptosis may exhibit Marcus Gunn jaw winking, in which jaw movement results in simultaneous elevation of the upper eyelid. The majority of congenital ptosis cases represent an isolated eyelid malposition, absent other ocular or systemic associations. The ptosis can be unilateral (75% of known cases, it affects one eye) or bilateral and is typically noticed shortly after birth. Incidence rate worldwide is unknown. In the United States, the frequency of congenital ptosis has not been officially reported. Sometimes a familial element is present, but a gene responsible for bilateral congenital ptosis has yet to be identified. It occurs equally among different races and equally between male and female.[12]

In children with congenital ptosis, amblyopia is common. When present, amblyopia is usually refractive and is caused by "induced with-the-rule" astigmatism. Occlusion amblyopia is much less common. Surgical repair of congenital ptosis and other pediatric eyelid abnormalities involves procedures that require special considerations and techniques that may differ from oculoplastic surgical procedure performed on adults.[13]

Ptosis associated with congenital syndromes

Congenital ptosis may be associated with several congenital syndromes (although it is rarely the hallmark), especially those involving disorders of eye movement, such as congenital fibrosis of the extra ocular muscles.

1. Smith-Lemli-Opitz syndrome
   Commonly present in association with a defect in cholesterol biosynthesis
2. Noonan syndrome
   An autosomal dominant condition characterized by congenital heart disease, short stature, abnormal faces, and the somatic features of Turner syndrome.
3. Saethre-Chotzen syndrome
   An autosomal dominant craniosynostosis syndrome with minor limb anomalies and characteristic faces mapped to chromosome 7p.
4. Cornelia de Lange syndrome
   A rare multisystem malformation disorder with characteristic facial features, growth and cognitive retardation, and abnormalities of the limbs, gastrointestinal system, respiratory system, genitourinary system, auditory and ocular systems, kidneys, heart, blood cells, and hair.[14]

Ptosis Associated with Ocular and Systemic Abnormalities:

- Marcus-Gunn Jaw Winking unilateral upper eyelid
  Ptosis with eyelid retraction associated with activation of the pterygoid muscle.
- Congenital Myasthenic Syndromes (CMS)
  Are heterogeneous group of syndromes characterized by defects in neuromuscular transmission the acetylcholine receptor, CMS is caused by genetic presynaptic, synaptic, or postsynaptic defects at the neuromuscular junction.
- Congenital Fibrosis of the Extraocular Muscles (CFEOM)
  Is a rare, non-progressive condition that results in restrictive globalophthalmoplegia and congenital ptosis.
- Chronic Progressive External Ophthalmoplegia (CPEO)
  Is a mitochondrially inherited disorder that is characterized by ptosis and ophthalmoplegia secondary to progressive weakness of the extraocular muscles.
- Congenital Third Nerve Palsy
  It can be due either to a developmental abnormality or intrauterine/birth trauma. They represent nearly half of third-nerve palsies seen in children.
- Congenital Horner's Syndrome
  A defect in the sympathetic innervation to the eye and adnexal structures and causes an ipsilateral ptosis, miosis of the pupil and anhydrosis of the affected side of the face.
- Blepharophimosis-Ptosis-Epicanthus Inversus Syndrome (BPES)
  Is an autosomal-dominant disorder, rare condition; with unknown worldwide prevalence but can be estimated to be less than 1/50,000.[15]

Characterized by small horizontal palpebral fissures (blepharophimosis), folds of skin extending from a flat nasal bridge inferolaterally (epicanthus inversus), and congenital ptosis. Although the distance between the medial canthi is usually increased (telecanthus), the interpupillary distance is usually normal. The major concern in children with BPES is amblyopia and head posture from ptosis. Because the epicanthus inversus and telecanthus do not generally improve with age, corrective surgery may be undertaken in a staged fashion or as a single procedure.

It is characterized by four features that are present at birth.

Patients have:

- ✓ Severe bilateral, symmetric ptosis.
- ✓ Telecanthus (an abnormally wide intracanthal distance with normal interpupillary distance).
- ✓ Epicanthus inversus (skin fold arising from the lower eyelid that covers the medial canthus).
- ✓ Blepharophimosis (profound narrowing of the palpebral fissure).[16]

Epicanthus

Epicanthus tarsalis is typical of East Asians with normal facies (observed in 45% of Asian male subjects and approximately 62% of Asian female subjects). It is a common mild eyelid anomaly with rare visual or syndromic significance that mostly appears between 3 and 6 months' gestation in the fetus in every race, and disappears before birth.[17]

Types of abnormal epicanthal folds

1. Epicanthus inversus
   A redundant fold arising from the lower lid, is the only epicanthal fold with syndromic significance, and is seen in blepharophimosis.
2. Epicanthus tarsalis
3. Epicanthus palpebralis
   A fold arising from the nasal root and directed toward the medial upper eyelids is the most common type seen.
4. Epicanthus supraciliaris
   Has its origin in the upper eyelid close to the eyebrows.[18]

Ectropion and euryblepharon

Ectropion refers to the eversion of the entire length of the lower eyelid. It can result in lagophthalmos and exposure keratopathy if left untreated. It is typically seen as a consequence of aging, but may also be seen as a congenital anomaly. Congenital ectropion is very rare and usually associated with other developmental anomalies of eyelid as in several autosomal recessive skin disorders such as congenital cutis laxa with looseness of the lid or as cicatricial ectropion in harlequin ichthyosis. It has also been associated with macrostomia, ectropion, atrophic skin hypertrichosis syndrome, and blepharocheilodontic syndrome.[19]

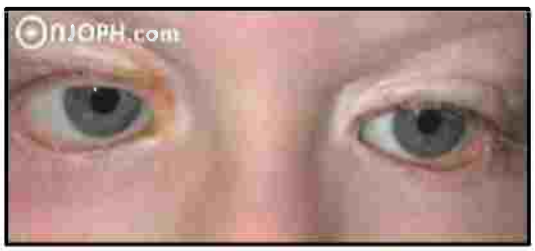

Figure (4): Showing lower lid ectropion.[20]

Euryblepharon is a congenital defect of increased horizontal length of the eyelids of unknown origin. The increased horizontal length of the palpebral fissure coupled with decreased eyelid skin in the vertical dimension causes the lateral portion of the eyelid to become everted (lateral ectropion). It is typically, but not necessarily, symmetric, and more commonly involves the lower eyelids. It may be seen in isolation, or with a host of other ocular anomalies, such as displacement of the proximal lacrimal drainage system, a double row of meibomian gland orifices, telecanthus, and strabismus. As is the case with ectropion, in severe cases it may result in lagophthalmos and exposure keratopathy.[21]

It is characteristic of Kabuki make-up syndrome, a constellation that also includes mental and growth retardation, large and protruding ears, and characteristic facies.[22]

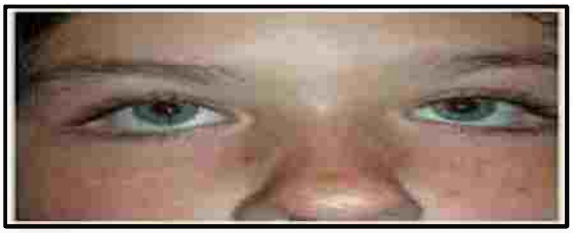

Figure (5): Showing bilateral euryplepharon.[23]

Congenital Entropion

Congenital entropion is inward rotation of an eyelid toward the globe. Eyelashes directed toward the cornea can lead to irritation, epithelial defects, and secondary scarring and infection. Congenital entropion may be sporadic or an autosomal-dominant trait. It occurs frequently among Asians but is rare among people of European descent in whom spastic and cicatricial forms are commonly encountered. The Asian race has a high prevalence of congenital entropion. It was reported that over 20% of Japanese children have congenital entropion at the age of 1 year. Congenital entropion may occur alone or in association with epiblepharon—a redundant fold of skin and orbicularis that rotates the lid inward. Congenital "kinking" of the tarsus or disinsertion of lid retractors are proposed mechanisms. If corneal injury cannot be prevented with conservative measures such as lubrication or lid taping, then surgery is indicated. Advancement of eyelid retractors with excision of a small ellipse of skin is helpful in cases caused by congenital retractor disinsertion.[24]

Entropion temporary treatments

- ✓ **Artificial teardrops** are a temporary, first-line treatment. The drops help to lubricate the cornea and relieve dryness, Medline Plus explains.
- ✓ **Stitches that turn the eyelid outward** using two or three stitches in certain spots along the base of the eyelid.
- ✓ **Botulinum toxin injection** into the lower eyelid to reverse the condition. A July 2004 case study in the "American Journal of Ophthalmology" suggests that it is beneficial for treating babies. According to Gregory Christiansen and colleagues, a 3-week-old infant treated with 5 units of botulinum toxin for entropion showed no recurrence of the condition seven months later.
- ✓ **Surgical correction** it usually requires surgery. Several different types of surgery may be used to treat entropion.[25]

Figure (6): Showing bilateral congenital entropion.[26]

Coloboma of the eyelid

Congenital notching of the upper or lower eyelid margin is referred to as eyelid coloboma. It is usually triangular with the base at the lid margin and varies from a small notch to a major defect when present on the lower eyelid. It is more commonly medial or central with a rectangular shape when it is present in the upper eyelid, and is commonly associated with lid-to-globe attachments. Although upper lid coloboma does not usually have a negative impact on vision; colobomata of the lower eyelid may result in ulceration or desiccation of the inferior cornea. It has a prevalence of 0.5-0.7/10000 birth. It may be unilateral or bilateral with approximately equal frequency. The exact cause of eyelid coloboma is unknown; although intrauterine factors are believed to contribute. It can be seen in isolation or as part of congenital malformation syndromes. It has been described as a result of the amnion rupture sequence caused by mechanical disruption.[27]

Upper eyelid colobomas are commonly found in the previously mentioned Goldenhar syndrome (oculoauricularvertebral spectrum). Lower lid colobomas, especially with a typical downsweeping with an upsweep to the lateral canthus and an absence of the lower eyelashes, are typical of autosomal dominant Treacher Collins syndrome (mandibulofacial dysostosis); this is a syndrome of aberrant development of the first branchial arch also characterized by auditory anomalies with bilateral conductive hearing loss, hypoplasia of the facial bones, cleft palate, and down-slanting palpebral fissures.[28]

A "wave shape" of the palpebral fissure is characteristic of the Cohen syndrome, with microcephaly; developmental delay; slim, tapering, extremities with truncal obesity; hypotonia; joint laxity; neutropenia; retinal degeneration or myopia; and characteristic facies.[29]

Management

- Surgery to the lids may be required, depending on the size of the defect.
- Protection of the cornea is essential. Non-surgical treatment (lubricants and patches worn at night) may be used to defer surgery for very young babies.[30]

Figure (7): Showing upper lid coloboma in Left eye.[31]

Epiblepharon

A common mild congenital malformation involving a redundant skin fold below the lid margin of one or both eyelids, resulting in direction of the eyelashes toward the cornea, occasionally causing trichiasis and corneal damage. It is more common in Asians of the Far East. With advancing age, the eyelashes tend to assume a more normal position. The upper eyelid may be involved in rare instances.[32]

Choristomas

Choristomas are benign, congenital tumors consisting of normal tissue in an abnormal location. Epibulbar and orbital choristomas are the most common epibulbar and orbital tumors in children. Choristomas are divided into four histologic types: (1) dermoids, which contain collagenous connective tissue surrounded by epidermis; (2) lipodermoids, which are similar to dermoids but also contain fatty tissue; (3) single-tissue choristomas, which contain either dermis-like or meso/ectodermal tissue of one type; and (4) complex choristomas, which contain tissues of several types. A teratoma is a choristoma that contains tissue from all three embryonic layers, ie, ectoderm, mesoderm, and endoderm. A teratoid tumor contains tissue from two embryonic layers. Dermoid cysts frequently contain a keratinized epithelium with adnexal structures. Epibulbar choristomas vary in appearance and visual significance. Their color may be white, yellow, or pink. They may be unilateral or bilateral and range in size from relatively small and flat lesions to large, bulky masses that fill the interpalpebral space and displace the globe. Epibulbar choristomas can cause astigmatism that, if untreated, can lead to amblyopia. The appearance of the tumor may be a cosmetic problem that affects a child's psychosocial development. Treatment involves dissection and excision, along with treatment of refractive error and amblyopia. Excision of any choristoma that involves the cornea will leave a residual opacity. It is therefore important to warn parents that surgery will not completely remove the opacity but will remove the mass. Occasionally, if the choristoma creates an opacity that extends into the visual axis, a lamellar keratoplasty may be indicated.[33]

Conjunctival or episcleral osseous choristomas contain compact bone. They may be freely mobile or may adhere to sclera or other extra ocular muscles. Choristomas may also occur in the orbit, with a predilection for the supratemporal quadrant and the nasal area above the lacrimal sac. Orbital choristomas may present as a bulging mass or as proptosis in childhood. Sometimes they are asymptomatic and present, after years of growth, in adulthood. The spontaneous rupture of an orbital choristoma may cause acute proptosis and inflammation, mimicking an orbital cellulitis. Orbital imaging with CT or MRI is helpful in diagnosis and surgical excision.[34]

Distichiasis

Distichiasis refers to the presence of a second row of eyelashes that arise from the meibomian glands. This is in distinction to trichiasis in which eyelashes arise from their normal location but are misdirected. Distichiasis can be inherited either alone or, more commonly, as part of the lymphedema-distichiasis syndrome. The aberrant lashes in distichiasis may cause ocular irritation and photophobia.[35]

Infantile hemangiomas

Infantile hemangiomas are the most common eyelid tumors in infancy. They have a bright red or purple appearance. Superficial ones typically blanch with pressure. At birth, they may be clinically undetected. However, they typically enlarge in the first 12 months followed by a slow involution during the first decade. Vision loss is related to amblyopia because of induced astigmatism or visual deprivation due to ptosis.[36]

At times these benign vascular tumors may hemorrhage. This blood accumulation gradually subsides and usually requires no treatment. Yet, treatment is indicated if the hemangioma is large -preventing the child from developing formed vision, causing amblyopia or significant astigmatism. Treatment is usually centered around steroid injections into the hemangioma (intralesional steroids). Systemic steroids, surgical debulking, and radiation therapy have a negligible role as they provide limited benefit and high risk.[37]

Treatment is necessary when ulceration, facial disfigurement and risk of amblyopia are present. Corticosteroids, in various forms are the mainstay of treatment. Local injection of corticosteroids is the most common route of administration; however it is associated with serious adverse effects such as occlusion of the ophthalmic artery or central retinal vein, retinal embolization, adrenal suppression and hypopigmentation at the site of injection. Topical application of corticosteroids has been used for superficial lesions only.[38]

There are some reports on the dramatic effect of oral propranolol on the size and volume of vascular masses. Beneficial effects of propranolol are probably due to reduction of the expression of genes for vascular endothelial growth factor, basic fibroblast growth factor and matrix metalloproteinase 9, in addition to induction of apoptosis in capillary endothelial cells.[39]

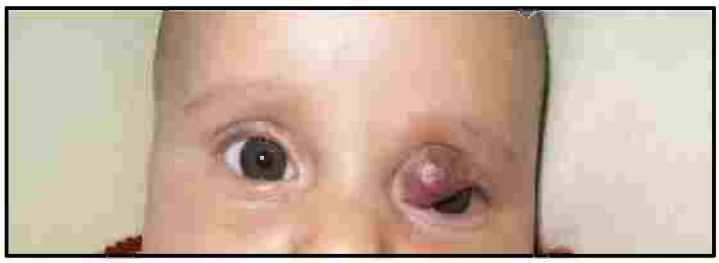

Figure (8): Showing left upper eyelid hemangioma.[40]

Ankyloblepharon (ankyloblepharon filiforme adnatum)

Ankyloblepharon (ankyloblepharon filiforme adnatum) is a rare defect, typically seen in isolation, characterized by a persistent connection between the ciliary edges of the upper and lower eyelid margins. Ankyloblepharon does not usually affect vision, because the connections tend to be small, but larger connections may occur with partial or full obstruction of vision. Surgical excision is curative. Ankyloblepharon has been described in the autosomal dominant Hay-Wells syndrome with ectodermal dysplasia and cleft lip or palate, and in neonates who have Edwards syndrome.[41,42]

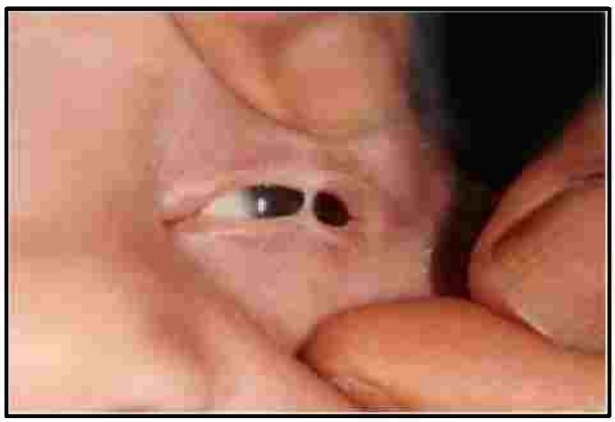

Figure (9): Showing left ankyloblepharon.[43]

Lagophthalmos (lid retraction)

Lagophthalmos is the inability to totally close the eyelids. Typically lagophthalmos is noted during sleep. It may be an isolated finding or a part of a syndrome. Lagophthalmos can be associated with ectropion. Occasionally infants present with a history of one or both

eyelid appearing to be retracted. Treatment depends upon the etiology. For primary congenital eyelid retraction, initial management should consist of observation and lubrication. Indications for surgical interventions include corneal exposure and cosmesis.[44]

Ablepharon

An exceedingly rare condition that describes a total absence of the eyelids and is the defining feature of the autosomal recessive ablepharon-macrostomia syndrome, also associated with auricular deformity, nasal alar deformity, absence of lanugo hair, ichthyotic skin, ambiguous genitalia, and absence of the zygomatic arches.[45]

Cryptophthalmos

A rare congenital malformation, refers to complete failure of development of the eyelids, with skin continuity from the forehead to the cheek. It is a common feature of Fraser syndrome, a rare autosomal recessive disorder associated with hypoplastic genitalia, laryngeal stenosis, and renal anomalies. Typically, no conjunctival fornices are present and microphthalmia may be present. Cryptophthalmos may be unilateral or bilateral, isolated, or syndromic. The phenotypic continuum in this disorder ranges from a complete, uninterrupted layer of skin from brow to cheek, through partial formation of lids and adnexal structures, to adherence of lids directly to the globe.[46]

The surgical management of cryptophthalmos is complex and challenging. Reconstruction is made difficult in many cases by the absence of normal corneal and anterior segment tissue and the absence of conjunctival fornices. Some success has been achieved with amniotic membrane grafts in the reconstruction of conjunctival fornices in patients with partial cryptophthalmos. Advances in keratoprostheses may someday help these patients.[47]

Amblyopia with eyelid anomalies

Amblyopia occurs in approximately 2% of the general population and is the most common cause of decreased vision in childhood. The term amblyopia is derived from the Greek language and means “amblys=dull” “ops=eye”. Other terms for this type of amblyopia include functional amblyopia and amblyopia ex anopsia. Children are susceptible to amblyopia between birth and age of 7 years. The earlier the onset of abnormal stimulation, the greater is visual deficit.[48]

The critical period for visual development is somewhat controversial but probably ranges from 1 week to 3 months of age. For practical purposes, amblyopia is defined as at least 2 snellen lines difference in visual acuity between the eyes, but amblyopia is truly a spectrum of visual loss, ranging from missing a few letters on 20\20 line to hand motion vision. Functional amblyopia should be distinguished from organic one in which the poor vision is caused by structural abnormalities of the eye or brain. Functional amblyopia is reversible when treated with appropriate visual stimulation during early childhood, whereas organic amblyopia does not improve by visual stimulation.[49]

Challenges that are intrinsic to the detection of the eye disease in the pediatric population include the vast number of children and the inability of children to describe symptoms that indicate pathology. These difficulties highlight the importance of vision screening in the pediatric population.[50]

Visual loss is mainly due to amblyopia.[51,52] This arises principally through stimulus deprivation that prevents normal retinal images forming and being transmitted to the visual cortex.[53] In industrialized countries the most common visually disabling disorders of children are present or become manifest in early childhood.[54]

The prevalence of amblyopia is 3-5%. Its onset is in the first decade of life, but its effect can last a lifetime.[55] Amblyopia is best diagnosed and treated as early as possible, but data from clinical trials have challenged the notion of a significant age effect of treatment.[56,57]

Early identification and treatment of risk factors for ocular congenital anomalies enables prevention of amblyopia. Diagnosis and treatment of amblyopia at an earlier age may lead to a better and more stable final visual result with shorter treatment times, more rapid improvement in visual acuity, the better overall compliance with treatment regimens.[58]

In 1981, the award of Noble prize for medicine for the discovery of the pathophysiology of amblyopia marked a turning point in the management of children with this condition.[59]

Guidelines for screening

The aim of guidelines is to define a screening procedure in order to catch all children whose eyelid anomaly requires treatment. Factors that must also be considered when proposing such guidelines are patient discomfort, the ophthalmologist's time, as well as financial costs, all of which need to be kept as low as possible without endangering the health of the patient or quality of care. The most important issue in pediatric eyelid disorders is to identify whether the lesions affect the visual development or not. If it occludes the visual axis, the pathology must be treated promptly to prevent the development of amblyopia. Entropion, ectropion, distichiasis, epicanthal folds, and telecanthus are common congenital anomalies of the eyelids. Although they are solely cosmetic problems in most cases, they may result in corneal changes secondary to corneal irritation and exposure due to mal-position of the eyelids.[60]

There is inconsistency in the literature regarding the screening protocols. Protocols including more mature babies may be designed for developing countries.[61]

There is no consensus on when the initial eye examination in a healthy child should be performed and how often the examinations should be repeated in the presence of normal eyes. Many congenital ocular abnormalities may be diagnosed by simple observation by a pediatrician, if they are aware of the possible congenital ocular diseases. In developed countries, the initial eye examination by an ophthalmologist is commonly performed at 6 months of age. At this age, the alignment of the eyes and the near focusing of the infant can be checked. An infant should be able to fix and follow faces within 2-3 weeks of age.[62]

The ability of the young children to fixate and follow a small target is an important gross evaluation of vision. Consistent objection from the child to having one eye occluded suggests that the un-occluded eye is amblyopic. Refraction examination and the red reflex test should be performed. Special tests to confirm that, an infant sees, may be performed. Forced choice preferential looking is a popular way of quantifying infant vision. If the child cannot tolerate ophthalmic examination and detailed examination is indicated,

sedation can be required. Eye examinations after 3 years of age are more informative and more easily performed. Visual acuity assessment and the fundamental parts of eye examinations are similar with the adult patients.[63]

Current American Academy of Pediatrics policy recommends eye examinations for infants and children at specified intervals during their development, including an examination to take place sometime during the first 2 years of life, stating: "Vision screening and eye examination are vital for the detection of conditions that distort or suppress the normal visual image, which may lead to inadequate school performance or, at worst, blindness in children. The policy further recommends that an eye evaluation for infants and children from birth to 2 years of age include examination of the following:

1. Eyelids and orbits;
2. External structures of the eyes;
3. Motility;
4. Eye muscle balance;
5. Pupils; and
6. Red reflex.[64]

The World Health Organization has mandated guidelines for a successful screening program as a part of Public Health Program. Generally the guidelines require that the condition being screened for is relatively a common, public health concern, and that the successful detection and treatment exists. These guidelines were designed to be applied to all types of screening.[65]

The WHO guidelines for a successful screening as part of Public Health Program are[66]

1. The condition sought should be an important health problem.
2. There should be an accepted treatment for patients with recognized disease.
3. Facilities for diagnosis and treatment should be available.
4. There should be a suitable latent or early symptomatic stage.
5. There should be a suitable test or examination.
6. The test should be acceptable to the population.
7. The natural history of the condition, including development from latent to declared disease should be adequately understood.
8. There should be an agreed policy on whom to treat as patients.
9. The cost of case finding, including diagnosis and treatment of patients diagnosed, should be economically balanced in relation to expenditures on medical care as a whole.
10. Case finding should be a continuous process and not a "once-for-all" project.

Screening

- Screening the newborn infants is necessary to detect treatable congenital structural abnormalities.[67]
- Vision screening of preschool children is one of the screening programs implemented.[68]
- Screening high risk subpopulation.

Ocular malformations can be isolated or part of complex multisystem syndromes. Certain ocular malformations are harbingers of serious "hidden" abnormalities in the other organs. The management of patients with ocular malformations includes making an

accurate diagnosis identifying any potentially associated syndromes or malformations in other organ systems.[69]

Major congenital anomalies occur in 2-3% of live birth. Causes include single genes, chromosomal anomalies, multifunctional disorders, environmental agents and unknown causes. The last category, unknown causes, accounts for 50% or more of these malformations.[70]