

## An Overview of Paediatric Oculoplastics Part I: Eyelids

### Introduction

Care of the paediatric patient poses unique challenges for the oculoplastic surgeon. Conditions of the ocular adnexa may threaten life and vision in children: preservation of life is the priority in orbital cellulitis and tumours; preservation of vision in cases where the eyelids do not function adequately to protect the cornea, such as colobomas and craniofacial syntosis, or where obstruction of vision might prove amblyogenic, as in the child with ptosis. Cosmetic considerations are also important to children and their parents, and a holistic approach to a disfiguring periocular condition can be critical to a child's psychological development. Patient co-operation during assessment and treatment raises challenges in paediatric practice. Careful planning of interventions is required, as pathology may change as the child grows. Techniques used to fine-tune surgical outcome in adults, eg. use of local anaesthetic or adjustable sutures, are often not possible in children.

### Embryology of the eyelid<sup>1</sup>

The eyelids are derived from ectodermal neural crest tissue. At gestational week six the developing eye induces differentiation of surface ectoderm overlying the cornea into cranial and caudal folds with a core of mesenchyme. Normal lid development is therefore intimately dependent on successful formation of the eye from the optic vesicle, as well as being influenced by development of the facial structures from the branchial arches. The embryonic lid folds grow towards each other and fuse between weeks eight and ten. The mesenchymal core gives rise to the orbicularis muscle and tarsal plate. Separation into upper and lower lids usually occurs by the seventh month of gestation.

Cryptophthalmos results when the eyelid folds fail to advance over the developing globe. The underlying eye is usually incompletely developed and visual potential is poor. Systemic associations include facial abnormalities, renal anomalies and syndactyly (Fraser's syndrome).<sup>2</sup> In cases of incomplete cryptophthalmos, lid-sharing procedures may be used to protect the cornea.

### Abnormalities of lid shape

Colobomas occur when eyelid margin formation is incomplete, due either to failure of migration of lid ectoderm to fuse the lid folds, or to mechanical forces such as amniotic bands. The most frequent location is the medial half of the upper eyelid. Associations include clefting phenomena, dermoids and Goldenhars syndrome, which is a sporadically occurring triad of facial, vertebral and auricular defects. Colobomata of up to 25% of the lid are repairable by direct closure. Larger defects require tissue supplementation from other locations such as hard palate mucosa.

Lid sharing procedures can be used but carry a risk of amblyopia as the palpebral aperture is obscured for a period. Ankyloblepharon results from incomplete separation of the fused fetal lid folds, leaving tissue connections between the lids. Division of this tissue is usually sufficient to separate otherwise normal lids.

Distichiasis is the formation of accessory eyelashes in association with the meibomian glands. Thirty percent of congenital cases are associated with lymphoedema, and mutations in the FOXC2 gene have been identified in familial cases.<sup>3</sup> Small areas of distichiasis can be treated with excision of individual follicles, electrolysis or cryotherapy. Larger areas of ectopic follicles can be excised in a lid splitting procedure.

### Abnormalities of lid position

Three main aspects of lid position can be identified when considering the shape of a child's eyelids: the position of the lids relative to the corneal reflexes (ptosis and lid retraction), the position of the lid margin and associated eyelash direction (eyelid margin malpositions) and the morphology of the medial and lateral canthal angles.

### Eyelid margin malpositions

Epiblepharon, particularly common in oriental children, is a congenital condition in which an extra fold of skin across the lower lid margin causes the lash line to roll in towards the globe. It usually improves with age as the growth of the nasal bridge flattens the fold, and treatment is rarely necessary.<sup>4</sup> The less common congenital entropion results when preseptal orbicularis overrides pretarsal orbicularis. If corneal defects or chronic discharge arise, tightening the lower lid retractors, excising a strip of skin and orbicularis, and creating a scar between preseptal and pretarsal orbicularis can surgically treat both these conditions.

Ectropion in the neonatal upper lid may be due to inflammatory changes in the conjunctiva, such as Chlamydia trachomatis infection, and the conjunctiva should be scraped for culture in cases of apparently congenital ectropion.<sup>5</sup> Ichthyosis can produce ectropion by shortening the anterior lamellae due to chronic skin changes. True congenital upper lid ectropion may occur as an isolated problem or associated with Down syndrome. It can be treated by decreasing the size of the internal lamella by wedge excision, or augmenting the anterior lamella with skin grafts, although skin grafts to lids often give poor cosmetic results in young children.

### Canthal morphology

Medial epicanthal folds are redundant folds of tissue in the region of the medial canthus. They may be a normal



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finding, especially in Asian races, or can be associated with Down syndrome or blepharophimosis syndrome. Treatment is rarely required, as the folds tend to become less prominent with age. If treatment is requested for cosmetic reasons, procedures such as a Y-to-V plasty or Mustarde 'jumping man' flap are effective. If associated telecanthus is present then repositioning of the medial canthus by transnasal wiring may be required.

**Lid height: upper lid retraction and ptosis**

Eyelid retraction may be congenital, eg. associated with shallow orbits or vertical lid shortening. If it is acquired, thyroid eye disease and neurological disorders must be considered. Careful examination for contralateral ptosis is required to exclude pseudoretraction. Ptosis is one of the commonest paediatric oculoplastic conditions. Children may present with ptosis at any age, depending on functional status, aetiology and carers' perceptions of cosmetic acceptability. Around 20% of cases are bilateral and 20% have a family history of ptosis.<sup>6</sup> Some studies also suggest a slight male preponderance. Myogenic ptosis is the commonest group in children, accounting for about 70% of presentations. Abnormal embryonic development of the levator palpebrae leads to a dystrophic muscle, resulting in a ptotic lid that shows poor elevation in upgaze, but also lags behind the movements of the globe ('hangup') on downgaze. In a lesser proportion of children, abnormal formation of the levator aponeurosis leads to aponeurotic ptosis. Dehiscence or disinsertion of the aponeurosis leads to a high skin crease with a ptosis characterised by normal levator function. Neurogenic causes of ptosis should not be forgotten in the paediatric age group. Marcus-Gunn jaw-winking syndrome is the most common neurological association (about 8% of all childhood ptoses), in which abnormal connections between the third and fifth cranial nerves produces synkinesis of the levator palpebrae and pterygoid muscles. This is a sporadic congenital condition, which is usually noticed by carers but should be routinely sought in young ptosis patients due to its implications for successful treatment.

Spontaneous improvement of jaw winking with age has been reported, but may be due to learned behaviours rather than true resolution. The treatment approach depends on the aims. Amelioration of the ptosis may be achieved with a levator shortening procedure as the levator function is often reasonable. Abolition of the 'wink' requires more complex surgery including the lemagne levator transposition procedure or bilateral frontalis suspension with disinsertion of contralateral/both levators. Ptosis also occurs in congenital Horner's syndrome, in which a 1-2mm ptosis is associated with ipsilateral miosis and decreased iris pigmentation. Third nerve palsies may be congenital. These are usually isolated, but may be asso-

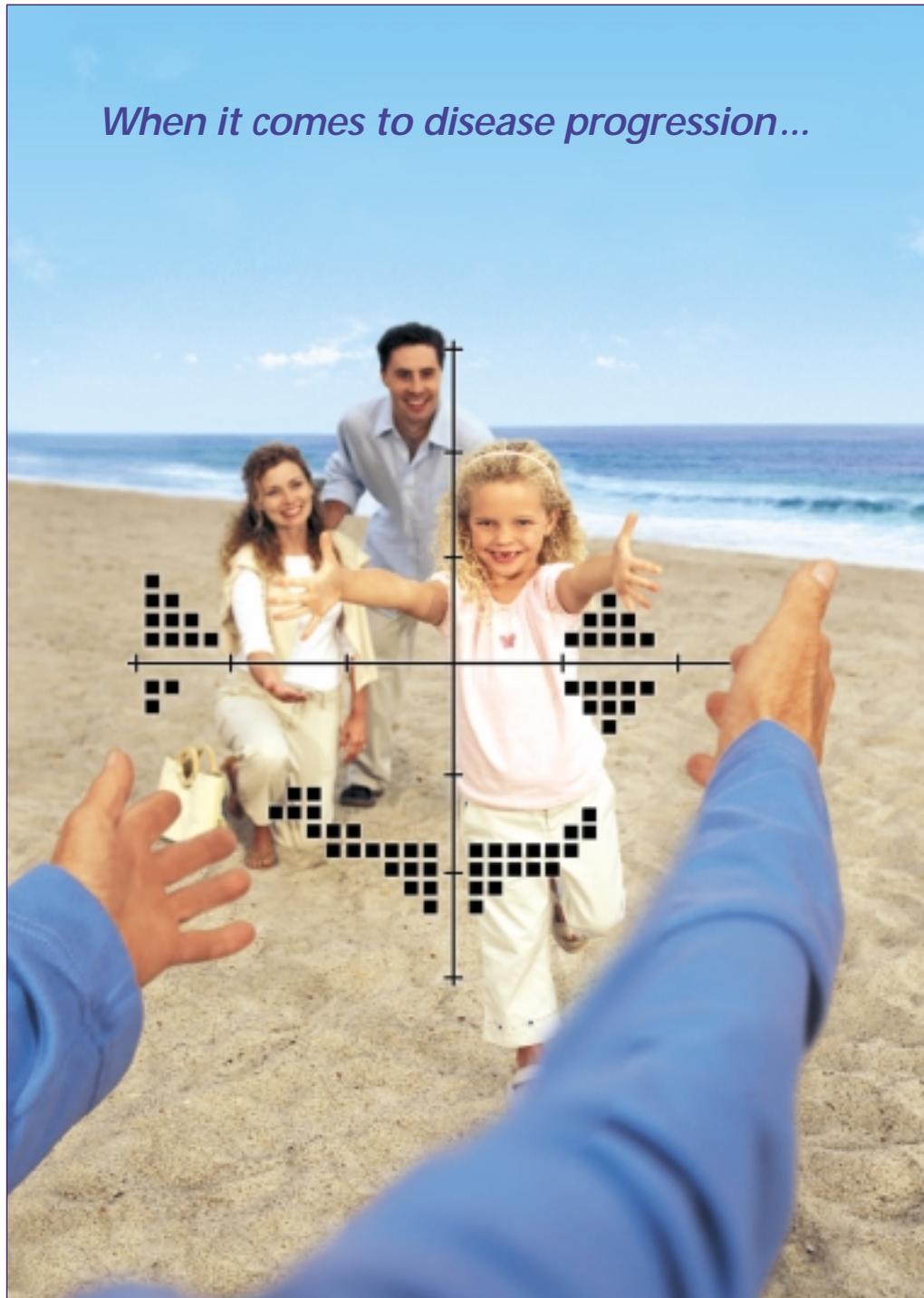
ciated with aberrant regeneration phenomena. It must be noted that, whilst Horner's syndrome and third nerve palsy in childhood are often congenital, an acquired case may be a marker of underlying tumour or trauma. Mechanical ptosis may result from lesions such as neurofibromas and capillary haemangiomas. These hamartomas often diffusely infiltrate the levator muscle complex, so debulking procedures may only partially correct the ptosis.

Pseudoptosis must be excluded before treatment can be planned: orbital, ocular and adnexal problems such as microphthalmos, hypertropia and phthisis bulbi can produce an

apparent ptosis. Evaluation of paediatric ptosis presents the challenge of gathering enough data to formulate a management plan. Accurate measurements cannot always be obtained, and a careful history from relatives plus observation of the child at play may provide the best assessment.

History of the child's delivery and any previous injuries or illnesses should be obtained. There may be a head posture or associated strabismus. About 25% of children with ptosis have amblyopia, and a baseline visual acuity should be measured wherever possible. Examination of the lids themselves should

*When it comes to disease progression...*



include a measure of the amounts of ptosis and asymmetry, for which corneal light reflexes and margin reflex distance may be the easiest measurement. An effort should be made to check for jaw-winking. Treatment of paediatric ptosis may be indicated for cosmetic or visual reasons. The procedure chosen must be tailored to the individual case. Levator function can guide this choice, with levator shortening being effective where levator function > 4mm. In cases of poor levator function frontalis suspension is indicated. Autogenous fascia lata can only be harvested in children over four years of age, but allogenic materials eg.

silicone rods, mersilene mesh may be used when the operation is indicated to prevent amblyopia in younger patients.

### Infections

Molluscum contagiosum is a pox virus infection common in childhood. It produces 2-5mm umbilicated papules anywhere on the body. Lid margin lesions frequently trigger a follicular conjunctivitis. Molluscum contagiosum is self-limiting in the immunocompetent, though persistent conjunctivitis may require excision or ablation of lid lesions. Children with HIV may develop larger widespread lesions.<sup>7</sup>

Common warts, caused by the human papillomavirus, may occur on the eyelids. If treatment is required, excision, cryotherapy and CO<sub>2</sub> laser can all be used.

### Pigmented Lesions

Congenital melanocytic naevi may occur on the eyelid. Malignant potential is low, though large examples, > 20cm diameter, have a lifetime risk of malignant transformation of 4-6%. Rarely a large divided 'kissing naevus' occurs involving adjacent areas of upper and lower lids. This developmental phenomenon arises before the eyelids separate at around 24 weeks. Surgical excision in early infancy gives the best cosmetic results for these heavily pigmented lesions.

The Naevus of Ota (oculodermal melanocytosis) produces blue-brown pigmentation on the periocular skin and the conjunctiva and sclera. It is most common in the Japanese and affects girls more often than boys. The lesion is composed of dermal melanocytes and has malignant potential, as well as being associated with glaucoma in affected eyes. Treatment with Q switched ruby laser can be used to lighten the cutaneous component.

### Cystic lesions

Dermoid and epidermoid cysts may arise in the lids, but are more commonly orbital and will be dealt with in the orbital review in this series. Milia are 1-2mm yellow-white cysts that occur commonly in the periorbital region in neonates and children. They are accumulations of keratin within pilosebaceous units. Neonatal milia often regress spontaneously, but in older children they may be more persistent.

Multiple milia are associated with syndromes such as hereditary trichodysplasia, oral-facial-digital syndrome, anhidrotic ectodermal dysplasia, Gorlin syndrome and dystrophic epidermolysis bullosa.

### Tumours

Basal cell carcinoma<sup>8</sup> (BCC) is exceptionally rare in children in the absence of an underlying predisposition, but may occur in those with Gorlin syndrome or Xeroderma pigmentosum. Gorlin syndrome or basal cell naevus syndrome is autosomal dominant and consists of multiple BCCs associated with skeletal and intracranial abnormalities. Xeroderma pigmentosum is an autosomal recessive condition with a DNA repair defect.

Capillary haemangiomas (strawberry naevi) are benign proliferations of endothelial cells. They are not usually present at birth (2.5%), but 90% manifest in the first month of life. Their natural history is to grow for three to six months before stabilising and involuting. Fifty percent resolve spontaneously by five years, and 90% by nine years. Intervention may be required if obstructive amblyopia or strabismus starts to develop. Many favour primary intralesional steroid injection,<sup>9</sup> but pulsed dye



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Figure 1: Right incomplete and left complete cryptophthalmos.



Figure 2: Ptosis from plexiform neurofibroma infiltration of left upper lid. Note the characteristic S-shaped deformity.

lasers, primary surgery, systemic steroids and interferon have also been used.

Mastocytomas are collections of mast cells within the skin. They arise in the eyelids in the context of three patterns of cutaneous mastocytosis. In urticaria pigmentosa, patients develop numerous yellow to red-brown macules with urticaria on touching the lesions. In solitary mastocytoma, which is very rare in the eyelids, and diffuse cutaneous mastocytosis there is diffuse skin infiltration with mastocytes and systemic symptoms of flushing, fainting and diarrhoea.

Pilomatrixomas are hair follicle tumours commonly seen under the age of 20 years. They are 3-30mm and appear as irregular, often calcified, nodules in the dermis, with relatively normal overlying skin. Malignant transformation has been reported, but excision is usually for cosmesis.

Pyogenic granulomas are elevated, bright red to brown, vascular-looking nodules, which often grow rapidly with recurrent bleeding. They are more common on the lower lid and often follow trauma, including incision and curettage of chalazia. They can be curetted or excised, and should be sent for histopathology as carcinomas or sarcomas occasionally mimic pyogenic granulomas. Syringomas are small, skin-toned or yellowish benign tumours arising from eccrine duct structures. They are commoner in females and are usually multiple. Treatment is necessary only if they cause cosmetic concern. Xanthelasma are rare in chil-

dren unless associated with disorders of lipid metabolism, such as familial hypercholesterolaemia, hyperapoprotein B syndrome and phytosterolaemia. Any child with xanthelasma should be investigated for these conditions by a physician.

Plexiform neurofibromas typically arise in the lids in neurofibromatosis type 1 and give a characteristic S-shaped upper lid deformity. Granulomatous lesions of the eyelids are uncommon in children. Granuloma annulare can produce an annular pattern of nodules in the lids, and is unrelated to diabetes when seen in children. Sarcoidosis is rare in children, but can produce granulomatous lid nodules associated with uveitis and polyarthritis.

#### Histiocytic disorders

Juvenile xanthogranuloma is a benign cutaneous histiocytosis of infants and children. It produces orange-red papules which usually present before nine months of age. The child must be examined for intraocular lesions, which occur in about 10% of those with cutaneous papules. Skin lesions can be treated with excision, steroids or radiotherapy.

#### Other dermatological conditions of the paediatric eyelid

Children can present with various lid dermatoses that may occur primarily in the periorbital region or may be related to a more generalised skin disorder. Joint management with a paediatric dermatologist is desirable.

Stevens-Johnson Syndrome (Erythema multiforme major) – this syndrome is an idiosyncratic reaction to certain infectious agents or drugs. It causes a widespread cutaneous bullous eruption with involvement of the mucous membranes. Eyelid complications include symblepharon and cicatricial lid malposition, leading to visual loss from corneal exposure, infections and scarring. Similar manifestations may be seen in children who develop Graft versus Host disease after bone marrow transplantation.

#### Conclusion

Many principles developed for adult patients still apply in the practice of paediatric oculo-plastics. However with the child's natural growth, the oculoplastic surgeon may be battling changing pathology. Amblyogenic conditions may require more aggressive treatment in younger patients, whilst for other conditions a conservative approach may be more desirable in children than in adults. It may be more critical to seek underlying pathology in a child with an eyelid lesion; for example, basal cell carcinoma, whilst common in adults, should raise the possibility of Gorlin syndrome or xeroderma pigmentosum in a child. Joint care with paediatricians will reap rewards, especially in the care of children with systemic conditions. It is also worth remembering that patience, compassion and rapport with the family can be as important as surgical acumen when the patient is a child. ■

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