An Overview of Paediatric Oculoplastics
Part II: Lacrimal

Introduction

Lacrimal problems, particularly epiphora, are a common reason for children to be referred to eye clinics. All ophthalmologists should be able to distinguish benign, self-limiting epiphora from conditions requiring specialist lacrimal attention. The lacrimal surgeon must then tailor their management to the child's age, bearing in mind that pathology may change as the patient grows. On the other hand, severe dry eye (eg. in Riley Day Syndrome) and lacrimal gland tumours (to be covered in Part III) also form a rare but important facet of lacrimal problems in childhood.

Embryology of the lacrimal system

The nasolacrimal system initially arises during the first trimester when two of the facial prominences derived from the first branchial arch fuse. Fusion of the lateral nasal prominence with the maxillary prominence entraps a double layer of surface ectoderm to form a solid epithelial lacrimal cord in the rudimentary naso-optic fissure. This cord grows during foetal life and becomes surrounded by mesenchyme. It moves from a horizontal to a vertical position with midface growth. The cephalic end buds to form the canaliculi and sac and the caudal end grows towards the nasal cavity. Canalisation of the sac and duct proceeds inferonasally during months four and five of gestation. The puncta open when the eyelids separate (around month five). The final portion to become patent is the point where the lower end of the duct coalesces with the mucosa of the inferior meatus; this membranous valve of Hasner ruptures late in gestation or after birth.[1]

Evaluation of epiphora in children

A thorough history from the child's carer is crucial, since only limited examination may be possible in children in the outpatient department. Tearing is the usual presenting complaint in nasolacrimal abnormalities, but discharge, crusting and red eye may also feature. Frequency of symptoms should be established, as well as aggravating factors such as cold, windy weather in a congenital nasolacrimal duct obstruction or worsening of epiphora in spring if due to allergy rather than obstruction. Age of onset helps to establish whether symptoms are congenital or acquired. Tear production begins a few weeks into neonatal life, so congenital obstruction may remain asymptomatic for up to month after birth, or longer in premature babies. Note the laterality of symptoms, which may correlate with facial or lacrimal anomalies apparent on inspection. A history of trauma is relevant, as the presence of canalicular or bony damage will influence management. Enquire about eye rubbing, photophobia and nystagmus, since infantile glaucoma is a rare cause of epiphora. Other medical history may be important; craniofacial abnormalities such as Goldenhar's syndrome and hemifacial microsomia may underlie nasolacrimal obstruction, and occasionally systemic conditions can cause obstruction (eg. mucus plugging of the duct in cystic fibrosis, altered bone growth in fibrous dysplasia).

A swift, focused examination in outpatients can glean useful information with minimal distress to the child.

Simple observation allows inspection for a high tear meniscus and epiphora at rest. Adequate tear drainage requires both a patent nasolacrimal system and a functioning tear pump, so examine for any lid abnormality that could cause tearing in the presence of a patent system. Palpation over the medial canthus may reveal reflux from the puncta if blockage is distal to the sac. The puncta themselves may be absent, imperforate or duplicated. A mucocoele or dacryocoele may be palpable, but a swelling above the medial canthal tendon may be an anterior ethmoidal mucocoele or nasofrontal encephalocoele and as such requires further investigation and imaging. The dye disappearance test can be performed by instilling fluorescein and examining with a pen torch fitted with a cobalt blue filter if the child is too young to sit at the slit lamp. With normal tear outflow the meniscus is relatively unstained by five minutes.[2] Fluorescein also allows assessment for ocular surface disorders that could increase reflex tearing. Finally a general ocular examination must be made to check for associated pathology and establish baseline visual function.

Imaging tests have limited usefulness in children; they may require general anaesthesia and radiation exposure and add little diagnostic information above that obtained during examination under anaesthetic. Lacrimal scintigraphy has been successfully performed without anaesthesia in children as young as two years and might be considered useful if history and examination suggest an atypical situation.[3]

Disorders of the lacrimal system
Punctal and canalicular abnormalities

Punctal atresia may result from failure of dehiscence of the conjunctiva overlying a canaliculus during eyelid separation, leaving a persistent membrane across the punctum.[4] If a papilla with a visible membrane is present at EUA, perforation of the membrane with a fine needle followed by dilation and irrigation may restore patency if the distal system is normal. If no papilla is evident, canalicular agenesis must be suspected; developmental failure of lacrimal budding or proximal canalisation may lead to an absent canaliculus. Impatency of a single punctum or canaliculus may be asymptomatic, especially if it is the upper system which is affected. If epiphora is associated with atresia of a single punctum, accompanying nasolacrimal duct obstruction must be suspected and probing carried out via the patent punctum. If both puncta are absent, treatment is more difficult. Patients may have only mild symptoms and it may be appropriate to wait until adulthood before operating. If surgery is required, external dacryocystorhinostomy (DCR) with retrograde intubation via the internal opening of the common canaliculus may be possible. If the common canalicular opening cannot be identified, Jones bypass tubes are required; in this situation it is preferable to perform the rhinostomy but wait until the child is older and more able to co-operate prior to insertion of the glass Jones tube. Extra buds from the rudimentary lacrimal cord can produce duplicated canaliculi opening onto the lid or skin and partially canalised accessory buds result in lacrimal fistulae; these supernumerary

Rebecca Ford, MA, MRCP, MROphth
is a Clinical Fellow at the Central Middlesex Hospital. She is currently working on research on adnexal trauma and diabetic retinopathy.

Vickie Lee, MA, FRCOphth
is a Consultant Ophthalmic & Oculoplastic Surgeon at the Central Middlesex Hospital, London. Her special interests include adnexal trauma and paediatric oculoplastics.

Correspondence:
Miss Vickie Lee,
Central Eye Service,
Central Middlesex Hospital,
Acton Lane,
Park Royal,
London,
NW10 7NS, UK.
Email: vickielee@mac.com

channels most frequently arise from the lacrimal sac. Treatment is by surgical excision and ligation. This can be facilitated by injection of methylene blue dye into the channel to outline the mucosa.

Punctal and canalicular stenosis may also be acquired, usually secondary to viral infections such as herpes simplex and chickenpox, or due to trauma. Simple dilatation may be sufficient treatment, or if necessary a two-snip procedure may relieve symptoms without compromising the lacrimal pump mechanism. Allergies can obstruct the lacrimal system due to mucosal oedema during flare-ups.

Disorders of the lacrimal sac

Dacryocoeles (mucocoeles / amniotocoeles) are sterile accumulations of mucus or amniotic fluid within the lacrimal sac which present within four weeks of birth. Obstruction of the sac is thought to arise from nasolacrimal duct obstruction at the valve of Hasner distally and the valve of Rosenmuller (at the junction of the common canaliculus and sac) proximally. Approximately one quarter of patients have bilateral obstruction. The dilated sac appears as a tense, blue-tinged mass inferior to the medial canthal tendon which may resemble a haemangioma. If there are no signs of infection, gentle massage over the sac may relieve the obstruction; vigorous massage must be avoided as it can rupture the sac leading to secondary cellulitis. More than 75% resolve with conservative treatment, but in cases where massage is unsuccessful, syringing and probing will release the obstruction.[5] This procedure may be required as early as six weeks of age, so dacryocoeles are the exception to the general rule of waiting until at least one year of age before probing. Neonatal dacryocystitis should be regarded as an emergency, as it may rapidly progress into orbital cellulitis or septicaemia. The infant will have an inflamed, tense mass at the position of the lacrimal sac, usually accompanied by fever, malaise and leucocytosis. Treatment is with broad-spectrum intravenous antibiotics and early syringing and probing to drain the collection into the nose once the acute inflammation settles. Dacryocystitis should not be drained percutaneously in children since fistulae often result. Some authors have, however, reported good results from decompressing the sac with a fine insulin needle and syringe during the acute period.[6] This relieves symptoms and provides material for microbiological analysis and the opportunity to introduce antibiotic directly into the sac. The resulting tiny puncture does not seem to fistulise. Tumours of the lacrimal sac are extremely rare in children. They should be suspected in cases of masses above the medial canthal tendon or blood stained tears. Treatment is dacryocystectomy plus oncologic care.

Figure 1: A second lacrimal probe is passed along the nasal floor for metal on metal touch to confirm patency of naso-lacrimal system on probing. Some lacrimal surgeons recommend direct visualisation with a nasal endoscope to minimise trauma to the nasal mucosa.

Figure 2: This young girl presented with a bluish swelling above the medial canthal tendon present since infancy. Because of its location it is definitely not a dacryocele/ lacrimal mucocele. After CT scanning to exclude sinus mucocele/encephalocele, she underwent excision of a dermoid cyst.

Congenital nasolacrimal duct obstruction (CNLDO)

CNLDO is the commonest cause of epiphora in early childhood. Up to 50% of nasolacrimal ducts are not fully patent at birth, although less than 20% are blocked by one month. The typical cause is membranous obstruction of the opening into the inferior meatus at the valve of Hasner, but dacryostenosis may be due to failure of canalisation anywhere along the duct or to bony abnormalities such as deviation of the nasal septum or hypertrophied inferior turbinate. CNLDO usually presents within one month of birth with epiphora, mattering of the eye, mucous discharge and periocular excoriation. It is more commonly unilateral but may be bilateral. Spontaneous resolution is the rule, with up to 96% resolving in the first year and a further 60% in the second year. Some children improve spontaneously at up to five years old. Conservative management is therefore recommended wherever possible. The parents should be encouraged to perform lid hygiene and lacrimal sac massage using firm strokes with the little finger starting over the inferior

lacrimal crest in the medial canthal region and moving downwards. Antibiotics are unnecessary unless true conjunctivitis develops; parents and carers may require reassurance that sticky secretions do not necessarily indicate infection.

Timing of probing in CNLDO is controversial, since the procedure requires general anaesthetic with its attendant risks.[7] Most authors recommend waiting until at least one year of age, and there is an argument for waiting until age two depending on parental preference, since spontaneous resolution is quite likely during the second year of life. Mannor et al found that success of nasolacrimal probing was negatively correlated with increasing age (92%, 89%, 80%, 71% and 42% at age 12, 24, 36, 48 and 60 months respectively). Factors predictive of failure of probing in CNLDO include dilated lacrimal sac, age greater than 36 months, bilateral affectation, daily or severe epiphora, previous failed probing and firm rather than membranous obstruction on probing.[8] Some groups have observed higher success rates with early probing, but their studies were not sufficiently controlled to establish whether the 'successful' early probings included cases which would have resolved without intervention, therefore evidence that early probing is beneficial is not clear-cut. A reasonable policy is therefore to carry out a first probing at age two years in most cases but to consider earlier probing in those with dacryocoeles or severe or bilateral involvement. Probing should be carried out under general anaesthetic with a protected airway. A laryngeal mask is preferable to endotracheal intubation as it allows free access to the nose for retrieval of dye and irrigation fluid. Examination for accessory canaliculi, fistulae, punctal abnormalities etc. should be carried out first. A Nettleship dilator is then used to gently dilate the punctum. Some ophthalmologists advocate the use of the upper punctum only to avoid trauma to the lower punctum during the procedure. The proximal canaliculus can also be dilated by angling the dilator towards the nasal bridge whilst putting the lid on stretch by pulling it gently laterally to straighten the canaliculus. The system can then be syringed with saline or dilute fluorescein via a 26G lacrimal cannula. Any reflux should be noted. If the cannula is advanced into the sac mucus may regurgitate. Probing is then carried out with a size 00 Bowman probe. The probe is introduced into the punctum and then advanced initially horizontally with the lid on stretch until the hard stop of the medial wall is felt. The probe is then withdrawn slightly before angling inferiorly to probe vertically down the nasolacrimal duct. The probe should slide easily into the duct; it should never be forced as this may create a false passage. A scraping sensation rather than smooth sliding of the probe should raise

the suspicion of a false passage. Some resistance may be encountered at the membrane of the valve of Hasner at the lower end of the duct. Firm but gentle pressure should pass the probe through into the nasal cavity with a slight 'popping' sensation. Patency of the system can then be confirmed by repeat syringing. Concurrent nasal endoscopy, if available, can assist in confirming the correct passage of the probe or dye. It also allows examination of the inferior turbinate; if this is impacted over the nasolacrimal duct it can be infractured medially towards the septum with a Freer elevator. Post-operatively a steroid-antibiotic combination drop for about two weeks is suggested.

Failed probing with persistent symptoms can be tackled in various ways. If the lacrimal system was patent immediately after first probing, repeat probing with or without infracture of the inferior turbinate may be successful. The success of repeat probing is increased if combined with balloon dilatation of the nasolacrimal duct or bi-canalicular intubation with silastic tubing.[9] Several introducer systems exist for silicone intubation. Crawford tubes are attached to flexible metal bodkins at each end. One bodkin is introduced through the upper and one through the lower canaliculus. They are retrieved from the nose with a hook, the bodkins removed and the tubes knotted in the inferior meatus. Ritleng tubes are similar in design but have a rigid introducing probe rather than bodkins.

Dacryocystorhinostomy (DCR) is indicated when CNLDO remains symptomatic after repeat probing or intubation, or when an abnormal bony canal is detected on probing or intra-operative DCG. It may also be required in canalicular atresia, traumatic obstructions and obstructions associated with craniofacial abnormalities. A lower success rate might be expected in paediatric than adult DCR due to less clearly defined anatomy, post-operative facial bone growth and increased scarring potential. However, series examining DCRs performed for unresolved CNLDO have shown results comparable to adult series with success rates of 88-94%. DCR before two years of age is not routinely suggested, but has been performed in cases such as severe persistent dacryocystitis without disturbing bone growth.

Anatomical differences between children and adults necessitate subtle modifications of DCR technique. The lacrimal fossa and anterior lacrimal crest are poorly defined in children, making selection of the proper osteotomy site more difficult. The medial canthal tendon insertion provides a landmark to identify the superior portion of the anterior lacrimal crest. In children it is usually easier to initiate the osteotomy through the thin lacrimal bone than the relatively thicker nasal process of the maxilla. The ethmoid sinuses are not prominent in childhood. Haemostasis is particularly

important in children due to their smaller blood volume; mildly hypotensive anaesthesia may minimise blood loss when administered by an experienced paediatric anaesthetist. Infiltration of the medial canthus with local anaesthetic mixed with epinephrine also helps haemostasis by providing vasoconstriction. Packs for the middle meatus may be soaked in oxymetazoline or cocaine; cocaine must be used with caution in young children due to potential toxic or tachyarrhythmic side effects. Severe haemorrhage is uncommon during DCR but transfusion may rarely be necessary, so it is important to discuss this possibility with the parents before surgery. Post-operative care should include cold compresses, adequate analgesia, antiemetics and sleeping with the head raised to minimise bleeding. Topical steroid-antibiotic combination drops plus oral antibiotics are suggested for one week. The silastic tubing is left in situ for 8-12 weeks and can usually be removed without any anaesthetic.

Conclusions

Epiphora is a common complaint in infancy and early childhood, so it is important that general ophthalmologists have a good understanding of their basic management. The specialist lacrimal surgeon must in turn be able to judge not only which surgery is most appropriate but also which timing is best for a particular individual in order to offer the best care. The development of new technologies such as endoscopic and laser surgery suggest that paediatric lacrimal surgery will remain an exciting and evolving field in years to come. ■

Suggested Further Reading
Jane M Olver. Colour Atlas of Lacrimal Surgery. Butterworth-Heinemann 2001

References

1. Moore KL, Persaud TVN. *The Developing Human, 5th ed*. WB Saunders; Philadelphia, USA; 1993.

2. MacEwen C, Young J. The fluorescein dye disappearance test (FDT): An evaluation of its use in infants. *J Paediatr Ophthalmol Strab* 1991;**28**:302.

3. Heyman S, Katowitz J, Smoger B. Dacryocystography in children. *Ophthalmic Surg* 1985;**16**:703.

4. Lyons CJ, Rosser PM, Welham RAN. The management of punctal agenesis. *Ophthalmology* 1993;**12**:1851-5.

5. Schnall BM, Christian CJ. Conservative treatment of congenital dacryocoele. *J Paed Ophthalmol Strab* 1996;**33**:219-21.

6. Smith B, Tenzel RR, Buffam FB, Boynton JR. Acute dacryocystic retention. *Arch Ophthalmol* 1976;**94**:1903.

7. Katowitz JA, Welsh MG. Timing of initial probing and irrigation in congenital nasolacrimal duct obstruction. *Ophthalmology* 1987;**94**:698-705.

8. Mannor GE, Rose GE, Frimpong-Ansah K, Ezra E. Factors affecting the success of nasolacrimal duct probing for congenital nasolacrimal duct obstruction. *Am J Ophthalmol* 1999;**127**:616-7.

9. Aggarwal RK, Misson GP, Donaldson I, Willshaw HE. The role of nasolacrimal intubationin the management of childhood epiphora. *Eye* 1993;**7**:760-2.

10. Hakin KN, Sullivan TJ, Sharma A, Welham RAN. Paediatric dacryocystorhinostomy. *Aust NZ J Ophthalmol* 1994;**22**:231-5.