The management of watery eye in an infant with facial dysmorphism

A six-month-old child with facial dysmorphism is brought to the eye clinic with history of watery right eye since birth. How will you manage this child?

Causes for watery eye in an infant

1. Overproduction of tears
   a. Infections
   b. Corneal abrasions
   c. Foreign bodies
   d. Crocodile tears (aberrant innervation)
   e. Exposure kertopathy (associated with dysmorphism)
   f. Epiblepharon with eye lashes touching the cornea
   g. Congenital glaucoma
2. Abnormalities of the lacrimal outflow system
   a. Membranous / bony obstruction of the nasolacrimal duct
   b. Stenosis, atresia, agenesis or duplication of any part of the nasolcarimal system
   c. Dacryocoele

History pertaining to the watery eye

- When was the onset of watering? Majority of congenital nasolacrimal duct obstruction (CNLDO) patients become symptomatic during the first month.
- Was it initially bilateral or always unilateral / when did the left watering resolve?
- Were the eyes sticky in the neonatal period?
- Is there any associated redness or mucopurulent discharge?
- Is there any photophobia?
- Are there any concerns regarding the vision?
- Has there been redness or swelling on the side of the nose, near the medial canthus?
- Are there any breathing problems or recurrent upper respiratory tract infections or nasal obstruction (suggestive of acquired NLDO)?

Ocular examination

- Vision assessment
- Detailed anterior segment examination with a handheld slit-lamp
  - Upper and lower lid puncta (present, absent, duplication, stenosed)
  - Look for conjunctival papillary changes
  - Look for corneal abrasions, erosisons or exposure keratopathy
  - Look for globe enlargement and increased corneal diameter
- Assess for squint
- Check refraction
- Dilated fundus examination

Examination for CNLDO

- Stickiness / crusting on lashes
- Lacrimal sac for a mucocoele, inflammation, mucoid regurge into conjunctival sac with pressure
- Instill Proxymetacaine 0.5% and Fluorescein 2% eye drops. Keeping the infant upright observe with cobalt blue light and measure the tear meniscus (graded 0 to 2) at five minutes and 10 minutes (Dye disappearance) and look for fluorescein appearance from the nostril (Dye appearance):
  - Profuse with patent lacrimal outflow system
  - Minimal with partial NLDO
  - Absent with persistent grade 2 tear meniscus implies NLDO.

Systemic examination

- General developmental milestones
- Facial and general features
- Nasal asymmetry
- Refer to a paediatrician for further assessment

Dysmorphism associated with CNLDO

- Down's syndrome
- Ectrodactyly, ectodermal dysplasia, clefting syndrome (EEC)
- Branchio-oculo-facial syndrome
- Craniometaphyseal dysplasia syndrome
- Craniodiaphyseal dysplasia syndrome
- Lacrimo-auriculo-dento-digital (LADD)syndrome
- CHARGE syndrome
- Goldenhar syndrome

Management

There is a high rate of spontaneous resolution within in the first year which can continue at a slower rate beyond 12 months.

- Explain the aetiology and natural history
- Manage conservatively up to age of 12-18 months
- Clear mucoid regurge with boiled cooled water
- Instruct parent / carer on the correct technique of sac massage
- Do not prescribe antibiotics unless there is overt conjunctivitis
- Discourage conjunctival swabs
- If the symptom persist at the age of 12 months then offer probing under general anaesthetic

Surgical intervention after 12 months of age

Children with craniofacial abnormalities may have complex anatomical variations that may require investigating with digital subtraction dacryocystography (DCG) at the time of intervention and multidisciplinary approach with the ENT surgeons is advisable.

- Syringing and probing from the upper punctum is both diagnostic and therapeutic

- Direct endoscopic visualisation of the Bowman probe at the inferior meatus is important or metal to metal sounding of the tip of the probe
- Consider in-fracture of the inferior turbinate to open the narrow inferior meatus and stretches the stenotic ostium
- Syringing with fluorescein stained saline and recovery of dye from nose with suction confirms patency
- Paediatric Otrivine nasal drops twice a day for two weeks at the discretion of the surgeon, especially if difficult probing with repeated passages
- If symptoms persist three to six months after the procedure, consider:
  - Reprobing
  - Intubation with monalicular or bicanalicular Ritleng tubes
  - Balloon dacryoplasty (catheter dilation of lacrimal system)
  - External / endoscopic dacryocystorhinoplasty (DCR)

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TAKE HOME MESSAGE

The management requires a methodical and multidisciplinary approach.

1. The key is to understand the aetiology.
2. Generally the commonest reason for persistent congenital epiphora is persistence of Hasner's membrane.
3. It is appropriate to manage epiphora here as one would in the absence of facial dysmorphism but have a high suspicion of an alternative aetiology which would indicate alternative management.
4. The systemic diagnosis can point to the aetiology. Facial dysmorphism is present; cast a wide diagnostic net:
   a. *Hyper-secretory epiphora:*
   - Exposure
     - Extreme shallow orbits
       - Syndromal forms of craniosynostosis especially Pfeiffer
       - Osteopetrosis
     - Inadequate lid coverage
       - Treacher-Collins
       - Facial clefting
     - Proptosis
       - Orbital tumours
       - Infantile haemangioma, vascular malformations, absent greater wing sphenoid in NF1
   - Lid mal-position (e.g. entropion related keratopathy)
     - Infantile haemangioma
     - Plexiform neurofibromas
     - Cherubism
   - Lid margin disease. Rare at this age. Commoner in Trisomy 21. Dramatic in pseudohypoaldosteronism
   - Glaucoma; associated with Sturge-Weber, NF1 (rare), neonatal Marfans. Naevus flammus is easily missed in neonates with dark skin

   b. *Outflow problems:*
   - Pre-punctal: Pump failure, lid mal-position (consider CNVII palsy including Moebius, facial clefting syndromes), extorted orbits and 'uphill' lower lids toward punctum (Treacher Collins, craniosynostosis)
   - Punctal / canalicular: dysgenesis, agenesis, stenosis, ectopia
   - Post-punctal: nasolacrimal duct stenosis or non-patency, anteriorly displaced inferior turbinates, congested nasal mucosa
5. The cornerstones of the outpatient paediatric assessment of lacrimal outflow are:
   - Fluorescein dye disappearance test (FDDT)
   - Primary Jones dye test

   If dye is in the nose on Primary Jones testing, this shows anatomical and functional integrity and is evidence against intervention; the greatest value of the test. Absence of dye in the nose generally supports appropriately timed intervention. Examine with indirect ophthalmoscope, cobolt blue illumination, child supine on parents' lap, no cotton bud into nose.
6. Intervention is generally reserved until 12 months of age unless there is recurrent dacryrocystits, secondary dermatitis or amblyogenic stickiness, since there is a reasonable chance of spontaneous improvement.
7. Intervention is indicated without delay if sight is threatened.
8. Management options include observation, probing, intubation (usually mono-canalicular), DCR, Coleman fat transfer, malar augmentation, mid-face advancement, lid reconstructive surgery. Lateral tarsorrhaphy is useful if globe prolapse has occurred or imminent. Oculoplastic interventions are better later. The principal problem is achieving therapeutic permanence. The skills of a multidisciplinary craniofacial team are helpful to deliver these interventions.
9. Life threatening aetiology or sequelae to consider (rare):
   - Congenital dacryocystocoeles, if bilateral, compromise the airway; suggested by bluish swellings in the medial canthal area and noisy breathing, occasionally with facial dysmorphism
   - Encephalocoeles suggested by hypertelorism, midline nasal furrow, mass in nasal cavity and midline central nervous system (CNS) anomalies. Require neurosurgical management
10. Trisomy 21 is relatively high prevalence and causes multi-factorial epiphora. Consider lid margin disease, punctal agenesis, canalicular stenosis and anteriorly displaced inferior turbinates. Have a low threshold for intubation with a mono-canalicular stent and inferior turbinate medilisation.
11. Rhinology expertise is useful. Depending on nasendoscopy findings, medialisation of the inferior turbinate, endoscopic DCR, adenoidectomy and medical treatments can be indicated.
12. Where co-morbidites are present combine procedures to minimise anaesthetic exposure.

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