Congenital Nasolacrimal Duct Obstruction: Variant based Approach

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Abstract: *Congenital nasolacrimal duct obstruction (CNLDO) is one of the common lacrimal disease but also one of the commonly miss-managed entity. Be it related to method of sac compression, impractical use of eye drops, age of probing or blanket treatment with silicone intubation and balloon dacryoplasty. Different specialties right from pediatricians to general ophthalmologist, pediatric ophthalmologist or oculoplastic surgeon may be involved, therefore the protocol for management of CNLDO is not standard and uniform. For children above 4 years straightforward dacryocystorhinostomy is no more a treatment of choice. Current review aims to highlight the recent advances in understanding of different types of CNLDO and a case based approach.*

Congenital nasolacrimal duct obstruction (CNLDO) is the most common cause of epiphora in infants and children[1]. Actual incidence in newborns is 50% but due to spontaneous perforation of membrane (Hasner valve) at lower nasolacrimal duct (NLD) after birth, clinical symptoms of CNLDO are seen in about 6-30% children[2]. Embryological basis of CNLDO is that the lower NLD is last to canalize during development, and therefore its failure leads to CNLDO. During crying or respiration, spontaneous rupture of membrane may occur over 3-4 weeks after birth but if it doesn't occur, CNLDO symptoms are manifested. This review aims to make you understand about the basics as well recent paradigm shifts in understanding and management of CNLDO. Over the recent years, nasal endoscopy guided (NEG) syringing and probing (S&P) is being accepted as gold standard for the management of CNLDO. NEG is not only needed for repeat or failed cases, which have undergone previous S & P, but also during primary attempt itself. Many cases can be saved from dacryocystorhinostomy (DCR) if done properly through NEG. Even older cases up to 10-12 years of age can be successfully managed with NEG S & P although with decreased success rate. Such older age children should always be given trial of S & P, before considering them for DCR.

Another recent change in the management of CNLDO is the approach based on the type of variant. CNLDO is broadly of two types: simple and complex[3,4]. Complex CNLDO encompasses various variants. Management of these variants not only need NEG but also customized treatment as discussed in this review. It is important to understand that some of the widely practiced methods of primary silicone intubation and balloon dacryoplasty may be entirely unnecessary.

SYMPTOMS/SIGNS OF CNLDO

Symptoms include classic triad of watering, discharge and matting of eyelashes. Three important signs are: positive regurgitation on pressure over lacrimal sac (ROPLAS), increase tear meniscus height and positive Fluorescein dye Retention test (FDRT) (Figure 1). Sometimes acute dacryocystitis may occur which can complicate into orbital cellulitis, orbital abscess or cavernous sinus thrombosis. Dacryocele is a bluish-purple dilated lacrimal sac seen in infants with CNDLO and functional common canalicular obstruction. It is important to carefully follow up and treat Dacryocele early as it is associated with intranasal cyst in NLD which can cause respiratory problems. Other findings may be lacrimal fistula (congenital/acquired), mucocele, incomplete punctal canalization and canalicular stenosis.

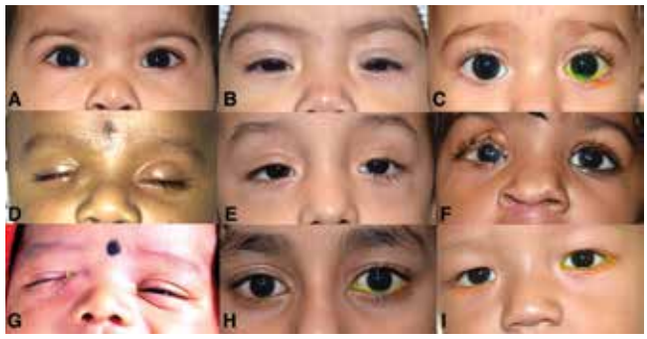

Figure 1: *Panel of photographs showing spectrum of clinical manifestation of CNLDO* ***(A)*** *A 5-month old child presenting with bilateral epiphora* ***(B)*** *A 6-month old child having classic triad of left eye epiphora, discharge and matting of eyelashes* ***(C)*** *Left eye showing increased tear meniscus with positive fluorescein dye retention test (FDRT)* ***(D)*** *A 2-month old child presenting with right dacryocele* ***(E)*** *A child with down syndrome presenting with symptoms of left eye complex CNLDO* ***(F)*** *A 6-month old child presenting with right eye watering and discharge along with Goldenhar syndrome and operated cleft lip/palate suggesting complex CNLDO* ***(G)*** *A 23 days old child presenting with right acute dacryocystitis* ***(H)*** *A 5-year old child presenting with persistent symptoms of CNLDO after twice failed previous attempts of probing suggesting complex CNLDO* ***(I)*** *A 8-months child presenting with recurrent symptoms of watering left eye associated with upper respiratory tract infection suggesting diffuse stenosis of nasolacrimal duct.*

TYPES OF CNLDO (SIMPLE AND COMPLEX)

All cases with soft membranous obstruction at lower NLD with rest of lacrimal system being normal and no intranasal abnormality constitutes simple CNLDO (Figure 2). All others are complex CNLDO. First study comparing simple and complex

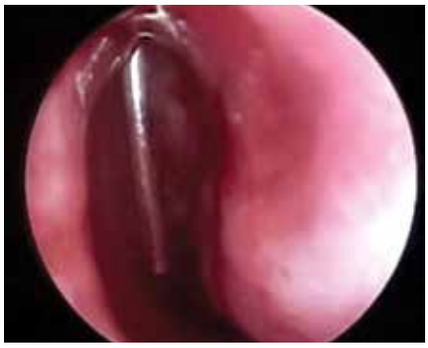

Figure 2: *Nasal endoscopy view of right nostril showing Bowman lacrimal probe coming through nasolacrimal duct opening without any nasal abnormality i.e. Simple CNLDO.*

CNLDO was published by Ali MJ et al reporting demography, clinical features and management outcomes at a tertiary eye care center. Table 1 shows the overview of difference between the two types.

Table 1: Modified table adapted from Ali MJ, Kamal S, Gupta A et al.

Parameter	Simple CNLDO	Complex CNLDO
Average age at presentation	Younger (mean 17.6 months)	Older (mean 45.6 months)
History of prior failed intervention	none	Positive in one-third cases
Intervention needed	Only NEG with S & P	Adjunctive procedures like intubation, balloon catheter needed in one-third cases
Success rate Anatomical Functional	97.8% 94.7%	58% 51%

CNLDO – Congenital nasolacrimal duct obstruction, NEG – Nasal endoscopic guidance, S & P – Syringing and Probing

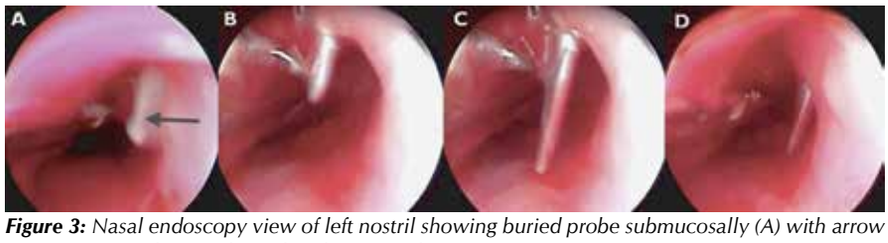

Figure 3: *Nasal endoscopy view of left nostril showing buried probe submucosally (A) with arrow pointing. (B-D) showing buried probe externalization.*

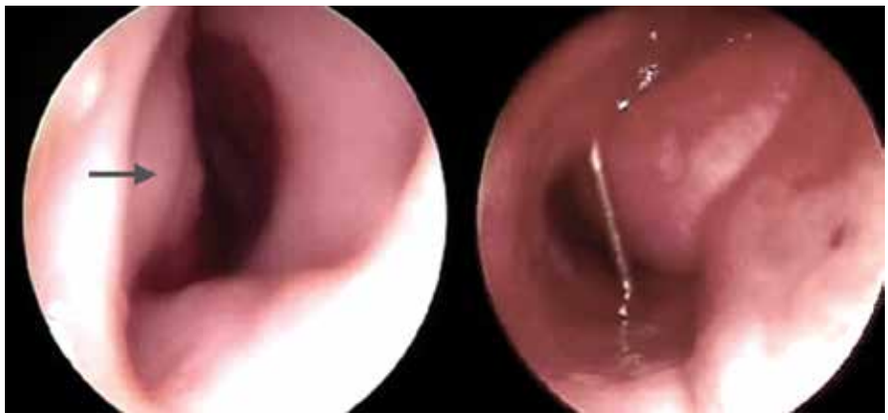

Figure 4: *Nasal endoscopy view of right nostril showing lateralized inferior turbinate (left Picture). Probe seen after medialization of inferior turbinate (right picture). True impaction of inferior turbinate is very rarely seen.*

Complex CNLDO encompasses following

1. *Bony obstruction* – This is due to complete absence of NLD formation or NLD directed into lateral maxillary bone.
2. *Craniofacial syndromes* – Such as Downs syndrome, Crouzon syndrome, Treacher Collins syndrome, Cleft lip/palate, hypertelorism.
3. *Buried probe*[5] – In this variant probe lies sub-mucosally along lateral wall of nose and fails to come out of the NLD opening. It needs tilting of probe to create and enlarge the opening along entire length (Figure 3).
4. *Lateralized/Impacted inferior turbinate* – This means the inferior turbinate lies in close approximation to the lateral nasal wall giving no access to the inferior meatus and visualization of NLD (Figure 4).
5. Dacryocele without intranasal cyst
6. *Anlage duct* – Lacrimal fistula (Anlage duct) may be associated with CNLDO. If congenital it needs fistulectomy.
7. *Multiple blocks* – Stenosis at valvular sites in canaliculus, sac and NLD or sometimes diffuse stenosis of NLD can lead to multiple level NLD blocks.
8. *Dacryocele with intranasal cyst* – Pressure inside Dacryocele can be transmitted to NLD and lead to its dilation and nasal cyst formation (Figure 5).
9. *Atonic sac* – Longstanding CNLDO can lead to atonicity of lacrimal sac.
10. *NLD upto the floor* – Variant in which NLD extends within the bone till nasal floor
11. *NLD into the inferior turbinate* – Caused by misdirection of inferior NLD.
12. *Lateral nasal wall hypoplasia* – Lead to non-differentiation of lateral nasal wall structures such as inferior turbinate, inferior meatus or NLD opening.

NATURAL HISTORY

It is very important to be aware of natural history of CNLDO since the management decision solely rests upon the chances of self resolution at that particular age. Macewen and Young followed children with symptoms of CNLDO till one year of age and calculated the chances of spontaneous resolution (Table 2) and suggested that probing should be performed at one year of age. Subsequently same authors published spontaneous resolution between 12-24 months age and observed resolution rate

Table 2: Predicted spontaneous resolution of CNLDO till one-year age depending upon presenting age (Adapted from Macewen and Young)

Age (months)	Resolution rate
1	96%
3	90%
6	75%
9	36%
12	0%

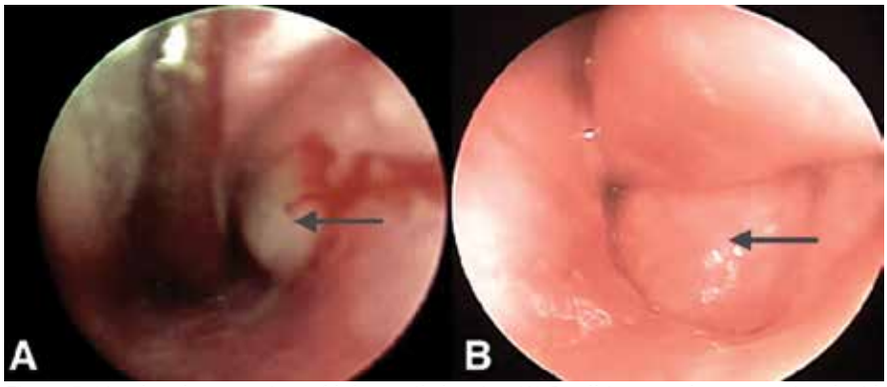

Figure 5: *Nasal endoscopy view of left nostril showing* ***(A)*** *Small intranasal cyst associated with dacryocele* ***(B)*** *A very large intranasal cyst which may cause respiratory difficulties in infants.*

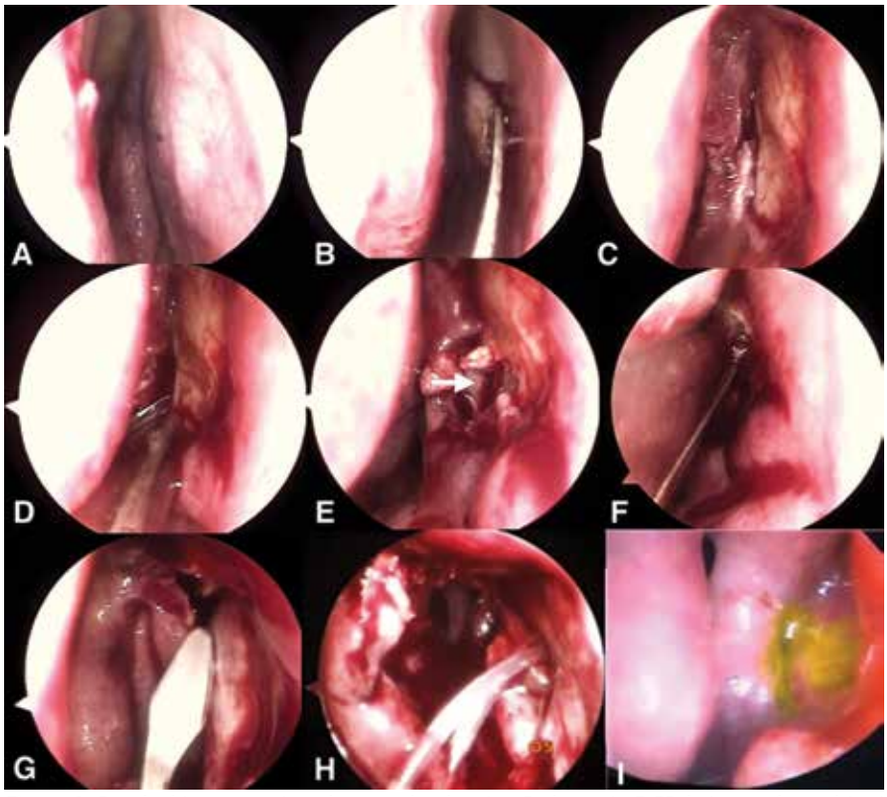

Figure 6: *Panel of intraoperative photographs showing Endoscopic DCR being performed in 4 year-old children with persistent CNLDO with bony obstruction.* ***(A)*** *Left nasal cavity showing septum on left side and lateral maxillary wall on right side* ***(B)*** *Nasal mucosal incision being given with No 15 BP blade* ***(C)*** *Frontal process of maxilla seen after elevation of nasal mucosal flap* ***(D)*** *Initiation of Osteotomy with punch at sac-duct junction* ***(E)*** *Bluish color sac seen after initial osteotomy (arrow)* ***(F)*** *Enlargement of osteotomy To expose lacrimal sac completely* ***(G)*** *Crescent knife for making anterior and posterior lacrimal sac flaps* ***(H)*** *Lacrimal sac opened in book-like fashion with bicanalicular silicone tube in situ* ***(I)*** *Evaluation of ostium at 4 weeks follow up showing positive functional endoscopic dye test (FEDT).*

of 50% between 13-18 months and 23% between 18-24 months age[6]. Appropriate time of probing recommended was 18 months. More recent data from PEDIG (Pediatric eye disease investigator group) observed that 66% of cases are likely to resolve if age is between 6 to less than 10 months[7] Author consider 9-12 months of age appropriate for S & P in non-resolving CNLDO.

MANAGEMENT OF CNLDO

Conservative treatment and Lacrimal sac compression - Compression over the sac increases hydrostatic pressure and can cause membrane rupture at lower NLD. It was described by Criggler in 1923. It can be done 3-4 times a day, 10 times each. It is important to demonstrate and then ask the parents to perform in clinics.

Probing - Timing of probing is controversial. Spontaneous resolution can occur beyond one year age, but success of probing decreases with increasing age. Katowitz and Welsh recommended probing before 13 months of age because they found that probing before 13 months of age was associated with a cure rate of 97%, which dropped to 54.7% after 13 months of age 9. Most important is to explain to the parents about chances of spontaneous resolution versus success rate of probing at particular age. Most clinicians prefer 9-12 months age for first probing.

Balloon catheter dilation and silicone intubation – These measures are not necessary for simple CNDLO but may be needed for complex CNLDO, persistent simple CNLDO, recurrent CNLDO or associated canalicular stenosis. Cost of the procedure is the main factor which has limited its use in recent times.

Nasal endoscopy - Use of rigid nasal endoscope is now becoming gold standard during probing, balloon catheter dilation or silicone intubation. It allows the visualization of NLD opening, helps in proper evaluation and treatment of complex CNLDO, marsupialization of intranasal cyst and correction of associated nasal abnormalities[1]. Figure 2 shows its utilization during management of CNLDO allowing confirmation of probe. Older children should undergo a trial of probing with endoscopy before considering DCR. A rigid 2.7 mm endoscope is used for nasal endoscopy in children.

Dacryocystorhinostomy (DCR) – Is considered for cases of failed probing, absent NLD or failed cases even when adjunctive measures have been used. Both external and endoscopic DCR can be done, and studies have shown that pediatric endoscopic DCR can be performed with good success rate even in children of one-year age. Figure 6 shows primary endoscopy DCR being performed in a 4-year-old child. Early need for endoscopic DCR is required in cases with recurrent acute dacryocystitis with failed probing or need for intraocular surgery.

CONCLUSION

CNLDO is a common cause of epiphora in children. Usually most of the cases tend to resolve spontaneously. For cases persisting beyond 9-12 months of age nasal endoscopic guided irrigation and probing is becoming gold standard. Use of nasal endoscopy not only helps in identification of complex CNLDO but also its management, thus improving the success rate of probing. This is especially important for cases with previously failed probing where endoscopy is not done.

For cases with atonic sac, mucocele, persistent CNLDO and bony obstruction, endonasal endoscopic DCR is considered.

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