ISSN: 0975-833X

Available online at http://www.journalcra.com

International Journal of Current Research
Vol. 12, Issue, 04, pp.11337-11341, April, 2020

DOI: https://doi.org/10.24941/ijcr.38533.04.2020

INTERNATIONAL JOURNAL OF CURRENT RESEARCH

RESEARCH ARTICLE

APPROACH TO CONGENITAL NASOLACRIMAL DUCT OBSTRUCTION

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ARTICLE INFO

Article History:
Received 08th January, 2020
Received in revised form
24th February, 2020
Accepted 18th March, 2020
Published online 30th April, 2020

Key Words:
Infant,
Congenitalnasolacrimal Duct Obstruction,
Epiphora,
Lacrimal System, Probing.

ABSTRACT

Congenital nasolacrimal duct obstruction (CNDO) is the most common lacrimal system disorder encountered in pediatric ophthalmology, occurring in approximately 5% of infants. It is usually congenital in origin resulting from a failure of canalization of the nasolacrimal duct. The obstruction is most commonly at the valve of Hasner where the nasolacrimal duct would normally enter the nose. It is the most common cause of epiphora in infants, however more specific signs include the presence of high tear meniscus and mucoid discharge that may cause crusting of the eyelid margins and skin erythema. CNDO resolves spontaneously in most cases, but a subset of patients have persistent symptoms that require treatment. Conservative management with lacrimal sac massage is the first line of management within the first year of life. After said time patients unresponsive to conservative management often undergo an intervention to surgically open the obstruction, including probing, balloon dacryocystoplasty, silicone intubation, inferior turbinate fracture, endoscopic intranasal surgery and dacryocystorhinostomy (DCR).However, no consensus has been reached regarding either optimal timing or type of intervention, so the decisionshould be based between patients family and the ophthalmologist.

Citation: ***Daniel Vasquez Valverde and Emily Sanderson Chacón.*** **2020.** "Approach to Congenital Nasolacrimal Duct Obstruction", *International Journal of Current Research*, 12, (4), 11337-11341.

INTRODUCTION

Congenital nasolacrimal duct obstruction (CNDO) is the most common lacrimal system disorder encountered in pediatric ophthalmology, occurring in approximately 5% to 20% of newborns and often resolves without surgery, is more common in patients with Down syndrome (22%) and in those with midfacial abnormalities (American academy of ophthalmology, 2018; Petris, 2017). Its high incidence makes this condition of great importance in order to make a correct diagnosis and treatment. A recent study using high-resolution computed tomography (CT)showed the obstruction to be either a persistence of a membraneat the distal end of the nasolacrimal duct, bony obstruction, or narrowing of the inferior meatus with apposition of the nasal mucosa (Schnall, 2013). What we got to find out is the existence of two schools of thought: the wait-and-see approach (i.e. conservative therapy), and the interventionist approach (i.e. probing). This dichotomy is due to the occurrence of spontaneous resolution of epiphora in the young infant and the good results of early probing (Le Garrec, 2016).

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Recent studies have helped to shed some light on these debates and provide the ophthalmologist with reliable data, which he/she can use when considering their own practice patterns.

Anatomy: The lacrimal system is made up of a secretory system, which produces tears, and an excretory system, which drains the tears. The lacrimal gland is primarily responsible for producing emotional or reflexive tears. As tears are produced, some fluid evaporates between blinks, and some is drained through the lacrimal punctum (Örge, 2014)

The lacrimal system consists of several structures: The lacrimal gland is a bilobed exocrine gland arising from the epithelial cells ofthe superotemporal conjunctiva, located in the lacrimal gland fossa of the frontal bone. The 2 lobes, the lacrimal and the orbital lobes, contain ducts that secrete the aqueous portion of the tear film and are separated anatomically by the lateral horn of the levator aponeurosis. The accessory lacrimal glands of Krause and Wolfring are located within the conjunctival fornices and contribute approximately 10% of the tear secretion.The puncta are located on the upper and lower eyelids in the nasal corner at the junction of the pars ciliaris (lateral five-sixths containing lashes) and the pars lacrimalis (medial nonciliated one-sixth) and rest slightly inverted against the globe.

The puncta serve as the exit point for tears from the conjunctival sac. The ampulla extends vertically from each puncta about 2 mm and the canaliculi run medially about 90° from each ampulla approximately 8 mm and meet to form the common canaliculus, which opens into the lacrimal sac. The valve of Rosenmuller, which is a mucosal flap separating the common canaliculus from the lacrimal sac, prevents tear reflux back into the canaliculi (Örge, 2014; Olitsky, 2014). The nasolacrimal duct empties into the inferior nasal meatus, with proper directional flow facilitated by the valve of Hasner, a mucosal flap separating the nasolacrimal duct from the nasal cavity.

Epidemiology: Approximately 6% of all children will demonstrate signs and symptoms of nasolacrimal duct obstruction at the time of birth or shortly thereafter. Historically the natural history of CNDO has been somewhat unclear due to a number of studies reporting various rates of spontaneous resolution among a small number of patients (Olitsky, 2014). Epidemiological studies report that the prevalence of CNDO ranges from 5% to 20% in the early phase of childhood. Mac Ewen *et al* found that in a cohort of 4792 infants in Britain, the prevalence of epiphora was approximately 20% in the first year of life, and almost 95% of this population showed symptoms at one month of age. The pathogenesis of CNDO lies in a mechanical obstruction located distally in the nasolacrimal duct (NLD) at the valve of Hasner, where this structure enters the nose (Vagge, 2018). In an observational study by Paul, the rate of resolution of CNDO with medical management by 1 year of age was 80% at 3 months old, 70% at 6 months old, and 52% at 9 months of age. Dacryocele is an uncommon manifestation of CNDO in neonates occurring in approximately 1 in 3900 live births (3). Canalization of the nasolacrimal duct is typically complete at birth, full excretory function of the lacrimal glands does not occur until approximately 6 weeks after birth. Therefore, many newborns can be observed crying without tearing (Örge, 2014). Moreover, the higher prevalence of CNDO reported in premature infants compared with ones at full-term suggests the importance of the physiological development of the nasolacrimal drainage system during intrauterine life, in order to ensure the patency of the NLD.

More recently, a higher prevalence of anisometropic amblyopia has been demonstrated in children with CNDO (10–12%); thus in these patients, a complete eye examination, including cycloplegic refraction, should be performed with a follow-up period of at least 3–4 years (Vagge, 2018). There were nearly equal numbers of boys and girls, and 13.1% of the patients were born prematurely. The median age at diagnosis for the 305 consecutive patients was 12.3 months (range, 0.8-57.7 months). The laterality of symptoms at initial presentation was essentially one-third each for bilateral, right, and left eye involvement. Positive symptoms of CNDO included epiphora in 91.1% of patients and discharge in 56.1%. The median spherical equivalent refractive error was 1.8 (range, −6.9 to 9.3) for all the patients. Two hundred two patients (66.2%) underwent surgical intervention for their symptoms (Piotrowski, 2010).

Clinical Features: The most common cause is incomplete canalization at the caudal end of the nasolacrimal duct, leaving an imperforate membrane at the valve of Hasner (Vagge *et al* ., 2018). The out pouching represents the distention of the lacrimal sac and duct caused by an obstruction at both the valve of Hasner and the common canaliculus or valve of Rosenmuller (Örge, 2014). Clinical manifestations of CNDO include persistent epiphora, increased tear lake, and recurrent mucopurulent discharge**.** Distal obstruction at the Hasner valve is more likely to cause a mucopurulent discharge, whereas, when obstruction is near the nasolacrimal sac,valve of Rosenmueller, it is more frequently related to a watery discharge. While usually unilateral, CNDO occurs bilaterally in 20% of cases (Vagge, 2018). Nasolacrimal duct obstruction will result in overflow of the tears with mattering of the lashes. Overgrowth of bacteria in the obstructed nasolacrimal duct will produce crusting on the lashes and may cause excoriation of the skin along the lid margin. The infection is in the nasolacrimal drainage system and therefore there is usually no associated conjunctival injection (Olitsky, 2014). Occasionally, bacterial overgrowth will overflow into the eye and produce conjunctivitis, which may recur after treatment (Vagge, 2018). Chronic epiphora tends to cause the lashes to be singed down to the lower lid skin, and there may be skin changes, mainly in the lateral border of the lower lid because of the chronic irrigation and irritation in this region.

Table 1. Initial Historical and Clinical Characteristics of 305 consecutive children with CNDO (8)

Characteristic	Value
Male sex, No. (%)	145 (47.5)
Premature births (<37 wk), No. (%)	40 (13.1)
Age at CNDO diagnosis, median (range), mo	12.3 (0.8-57.7)
Laterality of CNDO diagnosis, No. (%)	
a)Both eyes	100 (32.8)
b)Right eye	94 (30.8)
c)Left eye	111 (36.4)
Symptoms at CNDO diagnosis, No. (%)	
a)Epiphora	278 (91.1)
b)Discharge	171 (56.1)
Spherical equivalent, median (range)	1.8 (-6.9 to 9.3)

Table 2. Differential diagnosis of Congenital Nasolacrimal Duct Obstruction (1)

Condition	Clinical Features
Congenital Nasolacrimal Duct Obstruction	Increased tear meniscus, reflux of tears
Dacryocystocele	Bluish swelling overlying the lacrimal sac
Glaucoma	Photophobia, increased IOP
Uveitis	Pain, Ciliary Injection, photophobia
Conjunctivitis •Viral/Bacterial/Allergic	Purulent/Watery discharge, respiratory symptoms, allergic stigmata
Ophthalmia neonatorum	Infants younger than one month
Corneal abnormalities	History of trauma, erythema, photophobia
Conjunctivital foreign body	Foreign body viewed with slit lamp
Epiblepharon with trichiasis	Ingrown eyelashes on extra fold of skin

Table 3. Success rate of conservative treatment during the first 12 months of life (11)

Patient age	Kakizaki H (2008)	PEDIG (2012)
1 month	82.9%	-
2 months	82.4%	-
3 months	80%	-
4 months	79.3%	-
5 months	76%	-
6 months	68.4%	-
7 months	66.7%	69%
8 months	64.7%	68%
9 months	57.1%	55%
10 months	33.3%	67%
11 months	14.2%	-

There may be discomfort if the skin becomes red and chafed from chronic rubbing (Örge, 2014). Digital pressure over the nasolacrimal sac can often produce an increase in the mucopurulent discharge. Most parents will note that the tearing is worse in the cold, wind, or when the child has an upper respiratory infection. Unless there is a concurrent infection, the conjunctiva usually shows no sign of injection which helps to differentiate a nasolacrimal duct obstruction from a viral or bacterial conjunctivitis (Olitsky, 2014). Although the clinical picture may look similar to conjunctivitis, patients do not have any discomfort or light sensitivity, and the bulbar conjunctivastays white and quiet (Örge, 2014). These symptoms will usually begin within the first few months of life, a complete evaluation of the eyes by an ophthalmologist is crucial in cases of chronic tearing that presents at an unusual time, does not resolve as expected over time, or has any other associated findings such as conjunctival injection (5). Children with CNDO need to be followed until 3–4 years of age to ensure that anisometropic amblyopia does not develop**.**

Diagnosis: Diagnosis is typically made by history and physical examination alone.Patients with CNDO show signs such as persistent epiphora with high tear meniscus, recurrent mucopurulent discharge and reflux of the lacrimal sac contents by pressure, making the diagnosis typically made by history and physical examination alone. In cases in which we still have the doubt of the diagnosis, because the symptoms are not so clear, we can use the fluorescein dye disappearance test (Takahashi *et al* ., 2010). The fluorescein dye disappearance test, is used to confirm the diagnosis with a sensitivity of 90% and a specificity of 100%(10). The test is performed placing a drop of sterile 2% fluorescein solution or a moistened fluorescein strip, the examiner instills fluorescein into the conjunctival fornices of each eye and then observes the tear film with the cobalt blue fiter of the slit lamp. Persistence of significant dye and, particularly, asymmetric clearance of the dye from the tear meniscus over a 5-minute period indicate an obstruction (1). Although most cases of CNDO can be diagnosed without imaging studies, some require diagnostic imaging for further management. Conditions such as craniofacial malformations or Down's syndrome are associated with a high prevalence of CNDO. In these subgroups, the bony obstruction at the nasolacrimal duct can be confirmed with computed tomography (Takahashiv *et al* ., 2010)

Differential Diagnosis: Excessive tearing due to CNDO must be differentiated from epiphora due to infantile glaucoma, which has additional features, including photophobia, blepharospasm, ocular hypertension, corneal clouding with or without enlargement, and breaks in Descemet membrane. Besides infantile glaucoma, the differential diagnosis of CNDO includes conjunctivitis, uveitis, ophthalmia neonatorum, corneal abnormalities, epiblepharon with irritation due to trichiasis (American academy of ophthalmology, 2018). It is also important to rule out conditions such as encephalocele or meningocele, because these may appear as a bulge in the inner canthal area, but unlike the dacryocystocele, which is always located under the medial canthal tendon, these conditions are found to involve the area above the medial canthal tendon (Örge, 2014).

Management: In the management of CNDO, very complex and noninvasive treatment options can be used: conservative treatment, probing, irrigation, silicon tube intubation, inferior turbinate fracture, balloon dacryocystoplasty, endoscopic intranasal surgery, or dacryocystorhinostomy. It is important that the technique we choose to be in correlation with the etiology of the disease and the complexity of the case (Avram, 2017). Probing may serve to resolve the symptoms by opening the membranous obstruction; however, it may not be successful if the obstruction is due to a bony protrusion of the inferior turbinate into the nasolacrimal duct or when the duct is edematous due to infection such as dacryocystitis (Petris, 2017).

Non-surgical management: Lacrimal saccompression and massage is an essential part of conservative management, rigorous hygiene of the eyelids and, if there is any purulent discharge, antibiotic eye drops. It is generally recommended up to the age of 12 months and then, depending on the severity of the symptoms, other therapeutic options can be discussed. However Recently, Kaplan–Meier analysis was utilized in a large series of infants with CNDO and demonstrated that spontaneous resolution plateaued after 9 months. This observation and a step down in success rate of initial probing after 15 months led them to suggest probing between age 9 and 15 months. We have observed that most parents were taught to massage the bone (nasal bone or frontal process of maxillary bone) rather than lacrimal sac. Thus, the proper way of performing massage on the lacrimal sac must be taught at the commencement of conservative management (Avram, 2017; Kashkouli, 2019). Massaging the nasolacrimal sac in a downward fashion will produce hydrostatic pressure which may rupture the membranous obstruction at the valve of Hasner. Downward message of the nasolacrimal sac is more effective than simple massage and no massage (Schnall, 2013)

Surgical management: Probing: The most common invasive approach treating CNDO. There are several probing techniques, endoscopic assisted probing allows a direct visualization of the nasolacrimal duct and avoids the formation of false routes, its efficiency varying between 92.3% and 100% (Avram, 2017). The right moment of probing remains controversial, the main problem being the possibility of spontaneous resolution during the first 12 months of life. Vagge *et al* ., 2018; Avram, 2017). Early probing, 6 – 9 months of age, allows the procedure to be done in the office, avoiding a general anesthetic, and decreases the duration of the symptoms. Late probing, beyond 1 year of age, allows a greater chance for spontaneous resolution with fewer children being probed in a surgical facility with a general anesthetic (Schnall, 2013). According to the Clinical Outcomes of Initial and Repeated Nasolacrimal Duct Office-Based Probing for Congenital Nasolacrimal Duct Obstruction study, the success rate of the initial probing was 80% (196 of 244 patients) among all subjects, 82% (111 of 136 patients) in the 6 to 12 month age group, 79% (64 of 81 patients) in the 13 to 18 months age group, and 78% (21 of 27 patients) in subjects greater than 19 months of age (Cha *et al* ., 2010). There are various treatment options for CNDO if an initial probing fails. These include further observation, inferior turbinate fracture, repeated probing, balloon catheter dilation, silicone tube intubation and dacryocystorhinostomy (DCR). Further Observation: Spontaneous resolution post-intervention has been reported to occur at 33– 70 months. The mechanism of resolution was thought to depend on the continuing maturation of the lacrimal drainage system as part of the development of the facial structure (Takahashi, 2010). Inferior turbinate fracture:

The use of inferior turbinate fracture usually associated with probing is recommended when there is a narrow space around the nasolacrimal duct ostium. This technique has a controversial efficiency (Avram, 2017). The success rate of inferior turbinate fracture alone is 83%, although a combination of probing with intubation results in good cure rates of 88–100%, however the success rate for a combination of inferior turbinate fracture and probing was no different to that for simple probing (Takahashi, 2010). Repeated probing: In about 20% of cases there are persistent symptoms of CNDO following probing. Probing can be repeated at 1 month after the last attempt, this is a successful treatment for persistent CNDO after a failed probing in about 56% of patients (Takahashi *et al* ., 2010). Balloon catheter dilation: Balloon catheter dilation is a relatively new procedure that has come to be regarded as a viable surgical alternative to intubation. This is performed by inserting a guide wire with a deflated balloon attached through the punctum in the nasolacrimal duct.

The balloon is gently inflated with liquid and the pressure created opens up and expands the blocked duct. The balloon is deflated and removed (9,11). Primary balloon catheter dilation showed slightly less success rates of 86% at ages 12 – 24 months and 75% at 24 – 48 months (Kashkouli, 2019). Balloon catheter dilation is equal to probing alone in children with simple obstruction, but is probably better than probing alone in children with stenotic obstruction (Chen, 2005). Silicone tube intubation: Lacrimal pathways prosthesis with silicone tubes is indicated in ineffective conservative treatment, failed probing, or presence of strictures. According to various authors, the success rate is between 62% and 100% (11). If decided to use it as your primary surgical method after ineffective conservative treatment, primary intubation was successful in 92% at ages 12 – 24 months and 84% at 24 – 45 months (Kashkouli *et al* ., 2019). Dacryocystorhinostomy (DCR): It involves creating an anastomosis between the lacrimal sac and the nasal mucosa by means of a localized bone resection of the nose wing. It represents the last resort when other therapeutic methods have failed, dacryocystitis or dacryocystocele. There are two types of DCR, external and internal (endonasal/ endoscopic). The success rate of the two surgical approaches in children is relatively the same. DCR has high success rates of 88– 96% for external DCR and 82–92% for endoscopic DCR (Takahashi *et al* ., 2010; Avram, 2017).

Complications: Congenital Dacryocystoceles are a relatively rare variant of CNDO, accounting for only 0.1% of infants with CNDO, so an awareness of this condition and its potential complications is important. Dacryocystoceles occur when the mesoderm fails to canalize distally, and along with the distal obstruction there is a functional or mechanical proximal obstruction. It presents with abluish, cystic, firm mass below the medial canthus (the angle formed by the union of the upper and lower eyelids medially) presenting shortly after birth is the classic presentation for a dacryocystocele. Conservative management of dacryocystoceles includes the use of gentle pressure over the lacrimal sac, which can facilitate decompression and drainage of the contents into the nose. Antibiotic drops may also be used prophylacically before infection occurs. The resolution rate after a short course of topical antibiotics, warm compresses, and massage has been reported to be 76%.However, dacryocystitis can develop within a few days or weeks and requires intravenous antibiotics to prevent life-threatening sepsis.

Referral in the early neonatal period can aid in timely intervention before complicationsoccurs (Wong *et al* ., 2008).

Conclusion

The management of CNDO has always been a matter of debate, especially if we talk about the timing and type of surgery to be used. What everyone seems to agree on is a non-surgical management in the first year of life with lacrimal sac compressions and massage. After this time frame, CNDO must be treated according to the severity of the symptoms and not necessarily the age of the patient, this will allow us to have an appropriate approach and choose the most optimal treatment for the patient.

Conflict of interest: The authors declare that there is no conflict of interest.

Glossary of Abbreviations

CNDO: Congenital Nasolacrimal Duct Obstruction
NLD: Nasolacrimal duct
DCR: Dacryocystorhinostomy
CT: Computed tomography

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