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The association of neonatal dacryocystoceles and infantile dacryocystitis with nasolacrimal duct cysts (an American Ophthalmogical Society Thesis)

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THE ASSOCIATION OF NEONATAL DACRYOCYSTOCELES AND INFANTILE DACRYOCYSTITIS WITH NASOLACRIMAL DUCT CYSTS (AN AMERICAN OPHTHALMOLOGICAL SOCIETY THESIS)

By Gregg T. Lueder MD

ABSTRACT

Purpose: To investigate whether neonatal dacryocystoceles and dacryocystitis are associated with nasolacrimal duct cysts, and to report the outcomes of treatment of these disorders.

Methods: This was a retrospective medical record review of two groups of infants with nasolacrimal duct (NLD) obstruction. The first group had dacryocystoceles with or without dacryocystitis. The second group had NLD obstruction with symptoms severe enough to require early NLD probing. All of the patients underwent NLD probing and nasal endoscopy. When present, NLD cysts were removed.

Results: In the first group, 33 infants had dacryocystoceles. Acute dacryocystitis was present in 16 patients, 12 had noninfected dacryocystoceles that did not resolve, and 5 had dacryocystoceles that resolved but severe symptoms persisted. All of the patients had NLD cysts that were surgically removed. The symptoms resolved after surgery in 31 patients (94%). In the second group, 27 infants less than 6 months old without dacryocystoceles underwent early NLD probing and endoscopy due to severity of symptoms. Twelve (44%) of these patients had NLD cysts. The symptoms resolved in 11 (92%) of 12 patients following NLD probing and cyst removal.

Conclusions: Neonatal dacryocystoceles are almost always associated with NLD cysts. The success rate of NLD probing and endoscopic cyst removal in these patients is excellent. Nasolacrimal duct cysts also are present in many young infants with severe symptoms of NLD obstruction. Nasal endoscopy is an important adjunct to the management of these infants.

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INTRODUCTION

Nasolacrimal duct obstruction (NLDO) is one of the most common problems encountered in pediatric ophthalmology. Approximately 6% to 20% of infants develop some symptoms of this disorder.[1,2] Normally, the tears drain through the lacrimal system into the nose. In NLDO, infants usually have a membranous obstruction of the distal nasolacrimal duct. This typically causes two problems, which are reflected in the clinical presentation of NLDO. First, the obstruction creates a mechanical blockage to outflow of tears into the nares. The fluid backs up through the lacrimal system, causing an increased tear lake. If there is enough blockage, the tears overflow the eyelids and the children present with epiphora. The second problem is infection due to stasis of fluid within the lacrimal system. Normal flora bacteria are usually present within the tears, but they do not cause problems because they are flushed readily into the nares.[3] In NLDO, stasis of fluid within the lacrimal sac creates a moist, warm environment that is conducive to bacterial growth.[3] The infection associated with NLDO is typically low-grade and chronic, with clinical symptoms of intermittent eyelid crusting and mucopurulent discharge.

A small number of infants with NLDO present within the first few weeks of life with a more severe infection: acute dacryocystitis. Acute dacryocystitis in neonates differs in several respects from the chronic low-grade dacryocystitis that occurs in typical NLDO. First, acute dacryocystitis appears earlier than chronic dacryocystitis, usually within the first 1 to 2 weeks of life. Second, the infection is more severe. Acute dacryocystitis presents with swelling and erythema overlying the lacrimal sac, whereas children with chronic dacryocystitis have no visible external abnormalities in this area. In addition, patients with acute dacryocystitis may develop progressive cellulitis and may form abscesses in the lacrimal sac due to accumulation of purulent material (Figure 1).

The third important difference between acute and chronic dacryocystitis in infants is that acute neonatal dacryocystitis is often associated with palpable masses that arise from the lacrimal sac. Various names have been used to describe these lesions, including dacryocystocele, dacryocele, mucocele, and amniotocele. The terms *amniotocele* and *mucocele* imply that the fluid within the lacrimal sac derives from amniotic fluid or mucus secretions. Because the origin of the fluid is often not identifiable, these terms may be inaccurate. The term *dacryocystocele* anatomically describes the lesion without implying an etiology.[4] Therefore, this is the term that will be used for these lesions in this thesis.

Dacryocystoceles typically are noticed at or within a few days after birth. If they are not infected, they usually have a bluish color and are located medial to the eye beneath the medial canthus. The lesions may resolve spontaneously,[5] but they frequently become infected and progress rapidly to acute dacryocystitis and lacrimal sac abscesses.

Dacryocystoceles are associated with nasolacrimal duct cysts. These are outpouchings of tissue that extend from the distal nasolacrimal duct beneath the inferior turbinate into the nasal cavity. These cystic lesions are almost always present in infants with dacryocystoceles. They also may be present in older children with NLDO. A previous study by the author[6] found intranasal lesions at the distal lacrimal duct in 6% of children with NLDO who were older than 18 months at the time of their initial probing and 9% of children who had persistent symptoms of NLDO following previous probing. To our knowledge, the association of NLD cysts in young infants who have symptoms of NLDO that are more severe than normal, but who do not have dacryocystoceles, has not been systematically studied.

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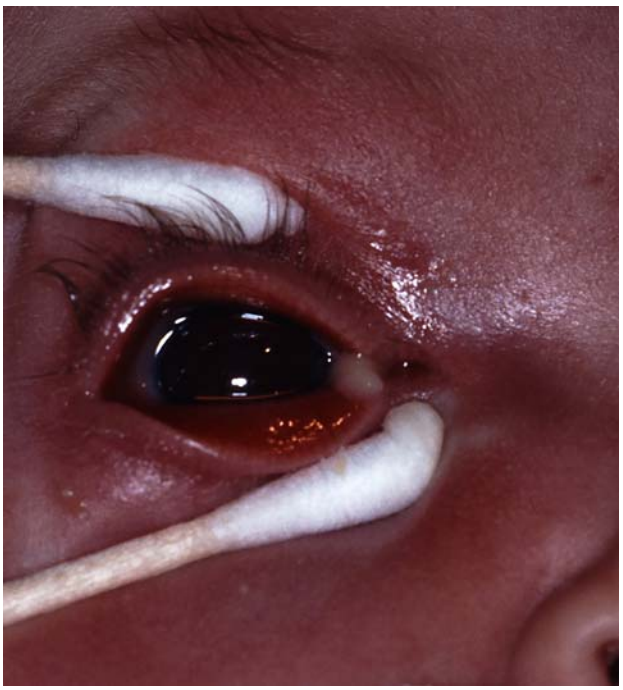

FIGURE 1

Infant with acute dacryocystitis and dacryocystocele. Note reflux of purulent material due to application of pressure over lacrimal sac abscess.

The purpose of this thesis is to describe the clinical findings and management of patients with neonatal dacryocystoceles and nasolacrimal duct cysts. The primary hypothesis is to test whether neonatal dacrycystoceles are associated with nasolacrimal duct cysts, and whether treatment directed at removal of the cysts is the most effective method of managing this disorder. In addition, the association of nasolacrimal duct cysts in older infants without dacryocystoceles, but with more severe symptoms than those seen in typical NLDO, will be reported.

METHODS

The Washington University School of Medicine Institutional Review Board approved the retrospective review of patient data, and the study was compliant with the Health Insurance Portability and Accountability Act regulations. The records of patients treated by the author at St Louis Children's Hospital from 1993 through October 2011 were reviewed to identify all infants with dacryocystoceles and all infants who underwent NLD probing at age 6 months or younger.

The first group of patients in the study consisted of infants who underwent surgery for treatment of neonatal dacryocystoceles, or who had histories of dacryocystoceles that had resolved but the patients had persistent severe symptoms that required surgery. In this group, infants who had acute dacryocystitis underwent immediate surgery (described below). Young infants with noninfected dacryocystoceles were initially treated conservatively with lacrimal massage and topical antibiotics. If they did not improve by 2 weeks of age, or if the patients initially presented beyond 2 weeks of age, surgery was performed. The patients with histories of dacryocystoceles and persistent severe symptoms also underwent surgery.

The second group of infants had histories of NLD obstruction, but not dacryocystoceles. The author's normal practice for patients with typical NLDO is to treat conservatively until approximately 9 months of age, then perform NLD probing. The infants in this second group underwent early surgery due to severity of symptoms.[7] The indications for early probing were (1) recurrent marked mucopurulent discharge and breakdown of the periocular skin despite treatment with topical antibiotics and lacrimal massage, (2) symptoms of NLDO associated with respiratory symptoms (rasping or snorting respirations and/or difficulty with feeding due to respiratory problems), or (3) acute dacryocystitis with swelling and erythema overlying the lacrimal sac.

Infants in both groups who had acute dacryocystitis were treated with parenteral antibiotics prior to surgery and a 1-week course of oral antibiotics postoperatively.

Patients were excluded from the study if they had craniofacial anomalies affecting the lacrimal system, other ocular conditions (glaucoma or cataracts), atresia of the lacrimal canaliculi or puncta, or trisomy 21. Patients were also excluded if NLD probing was performed before 6 months of age because the infants were undergoing a separate surgical procedure and the probing was performed during the same general anesthesia, rather than because they had severe symptoms of NLD obstruction.

The surgical procedure was performed under general anesthesia. Oxymetazoline-soaked pledgets were placed beneath the inferior turbinates. The lacrimal puncta were dilated, and size 00 through 2 Bowman probes were passed through the upper and lower puncta

into the distal NLD. In some infants the size 2 probe was not used because of the small size of the lacrimal canaliculi. The inferior turbinate was infractured using a Freer periosteal elevator. The nasal endoscope was used to visualize the space beneath the inferior turbinate. When NLD cysts were present, they were removed using alligator forceps under endoscopic guidance. The Bowman probes were used to move the cyst tissue to verify the presence of the NLD cyst (by noting movement of the cyst tissue as the probe was moved) and to assist in the removal by holding the cyst tissue in place as the forceps grabbed it. After the cyst was removed, fluid was irrigated through the lacrimal system and recovered from the nares using suction. In two patients lacrimal stents were placed because of poor irrigation.

Surgical results were assessed by either office examination or a telephone call to the family. The procedure was considered successful if the dacryocystocele and/or dacryocystitis resolved and there were no clinical signs of recurrent lacrimal obstruction at office examination or no symptoms of recurrent excess tearing or periocular crusting reported by the parents during a phone interview.

RESULTS

GROUP 1 (PATIENTS WITH DACRYOCYSTOCELES)

In the first group of patients, 33 infants had dacryocystoceles or histories of dacryocystoceles. Eighteen (54%) were female and 15 (45%) were male. The dacryocystoceles were present at birth in 24 infants (73%) and were noted during the first 1 to 2 weeks of life in the other patients.

Patients With Acute Dacryocystitis

Sixteen patients with dacryocystoceles had acute dacryocystitis. Ten of these were first evaluated during the first 2 weeks of life, five at age 2 to 4 weeks, and one at age 2.5 months. The latter patient had a dacryocystocele at birth. Seven of the patients were being treated by their pediatricians with intravenous antibiotics. Referring physicians had manually decompressed the dacryocystocele in one patient and percutaneously incised the lesions in two others, but they persisted. One of the patients had undergone endoscopy by an otolaryngologist during the first day of life because of respiratory distress, but no lesions were identified. The patients who had not been previously treated were given intravenous antibiotics either preoperatively or at the time of surgery. Surgery was performed within 1 to 2 days of presentation in 14 patients.

All of the patients had intranasal cysts that were removed at the time of NLD probing. Surgery was successful in 15 patients (94%). One patient's family reported recurrent periocular crusting, although on examination in the office 3 months postoperatively, neither increased tear lakes nor periocular discharge were noted.

Patients With Nonresolving Dacryocystoceles

Twelve infants had dacryocystoceles not associated with acute dacryocystitis. The lesions were present at birth in 10 of these patients. Infants who presented within the first week of life were treated with topical antibiotics and lacrimal massage. Surgery was performed if the lesions did not resolve by 2 weeks of age with conservative treatment, or if the patients presented after 2 weeks of age. Age at surgery ranged from 2 weeks to 3 months. All of these patients had nasolacrimal duct cysts that were removed at the time of NLD probing. Surgery was successful in all of the patients.

Patients With Histories of Dacryocystoceles

Five patients had histories of dacryocystoceles during the first week of life that subsequently resolved. One patient developed acute dacryocystitis at 2.5 months of age, three had persistent severe symptoms of NLDO (but not acute dacryocystitis), and one had respiratory symptoms (see Case 1 below). All of these patients had NLDO cysts that were removed at the time of NLD probing. One patient had lacrimal stents placed because of poor irrigation following the probing. This patient had persistent moderate symptoms of NLDO. Surgery was successful in the other four patients.

Laterality

Symptoms were bilateral in 13 patients (39%) and unilateral in 20 patients (61%). Nasolacrimal duct cysts were found unilaterally in 21 patients (64%) and bilaterally in 12 patients (36%). Endoscopy was performed on the affected side only in 10 patients and bilaterally in 23 patients. Of the latter 23 patients, 11 patients had bilateral symptoms and 12 patients had unilateral symptoms. In the group of 12 infants who had unilateral symptoms and bilateral endoscopy, NLD cysts were found in 4 (ie, bilateral cysts were found in 4 [33%] of 12 infants who had only unilateral symptoms).

Pathology

Tissue samples of the NLD cysts were sent for histopathologic examination for 21 patients. Ten lesions were lined by epithelium, 4 were described as respiratory mucosa, and 5 had nonspecific chronic inflammatory changes (Figure 2).

GROUP 2 (YOUNG INFANTS WITH SEVERE SYMPTOMS)

NLD probing was performed in 27 infants 6 months of age or younger who had symptoms that warranted early intervention. None of the patients had histories of dacryocystoceles. The age at presentation ranged from 1 to 6 months (mean, 3.5 months). Eight patients (30%) were female and 19 (70%) were male.

Indications for treatment were as follws: 17 patients (63%) had excessive discharge and periocular skin breakdown despite treatment with topical antibiotics and massage (Figure 3), 6 patients (22%) had respiratory symptoms associated with NLDO, and 4 patients (15%) had acute dacryocystitis with swelling and erythema overlying the lacrimal sac. One of the patients with acute

dacryocystitis had group B streptococcal sepsis (see Case 2 below).

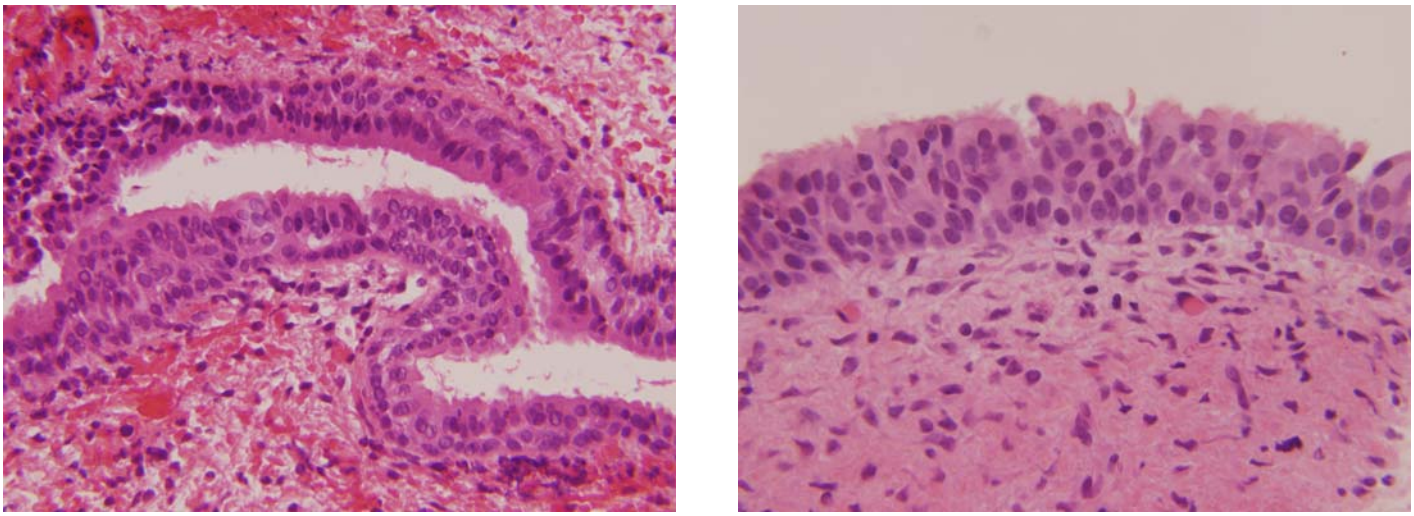

FIGURE 2

Histological sections of nasolacrimal duct cysts. Left, Cyst lined by nasal mucosal-type pseudostratified columnar ciliated epithelium (hematoxylin and eosin, ×400). Some chronic inflammatory changes are present within the stroma. Right, Similar findings in different patient with cyst (hematoxylin and eosin, ×600).

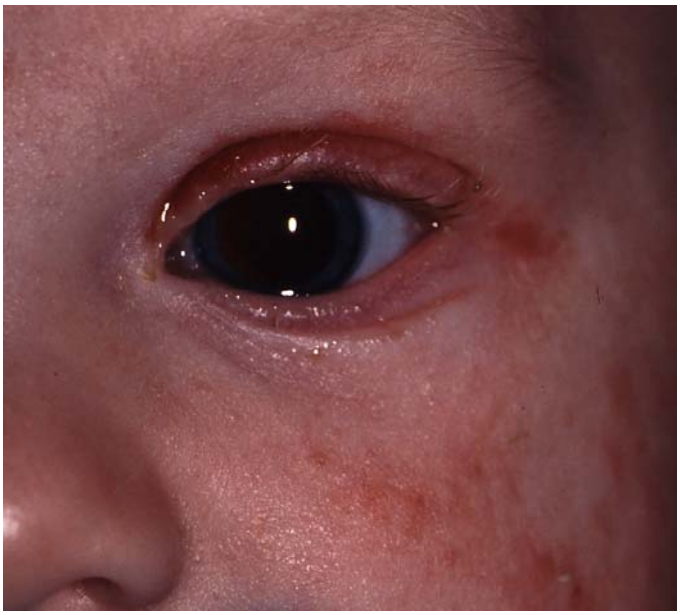

FIGURE 3

Patient with history of nasolacrimal duct obstruction with marked erythema of upper and lower eyelids, with lesser involvement of periocular skin. The patient did not have a history of a dacryocystocele.

NLD cysts were found and removed in 12 patients (44%). The numbers of patients with NLD cysts by indication are listed in Table 1. One patient with acute dacryocystitis had nonfixated stents placed at the time of the NLD probing because fluid irrigated poorly after the probing.

TABLE 1. INDICATIONS FOR NASOLACRIMAL DUCT PROBING IN GROUP 2 (PATIENTS WITHOUT DACRYOCYSTOCELES)

CATEGORY	MARKED DISCHARGE AND PERIOCULAR SKIN BREAKDOWN (N=17)	RESPIRATORY SYMPTOMS ASSOCIATED WITH NLDO (N=6)	ACUTE DACRYOCYSTITIS (N=4)
No NLD cyst	13	1	1
NLD cyst	4	5	3
% of patients with NLD cyst	31%	83%	75%

Overall, 11 of the 12 patients with NLD cysts had successful outcomes and one had mild recurrent symptoms of periocular crusting. In the 15 patients without cysts, 13 had good outcomes, one had persistent moderate symptoms, and one was later found to have a lacrimal sac diverticulum (see Case 4 in "Differential Diagnosis" section).

CASE REPORTS

Case 1 (History of Dacryocystocele With Persistent Respiratory Symptoms)

The infant was born at 35 weeks gestation and was noted to have a right-sided dacryocystocele at birth. He was hospitalized for management of a mesoblastic nephroma, which was removed successfully on day of life 2. He had no respiratory distress in the newborn period, and the dacryocystocele was not infected. Magnetic resonance imaging (MRI) revealed bilateral dacryocystoceles (Figure 4). An ophthalmology consultation on day of life 8 revealed a noninfected dacryocystocele on the right. The patient was treated with massage and topical antibiotics, and the dacryocystocele resolved by day of life 14. The author evaluated the patient at 2 months of age. The baby had persistent symptoms of NLDO and a history of snorting respirations and inability to lie on his back on account of respiratory distress. The patient did not have dacryocystoceles, but cysts were visible in the nares. He underwent nasolacrimal duct probing with bilateral removal of nasolacrimal duct cysts. His respiratory and NLDO symptoms resolved after surgery.

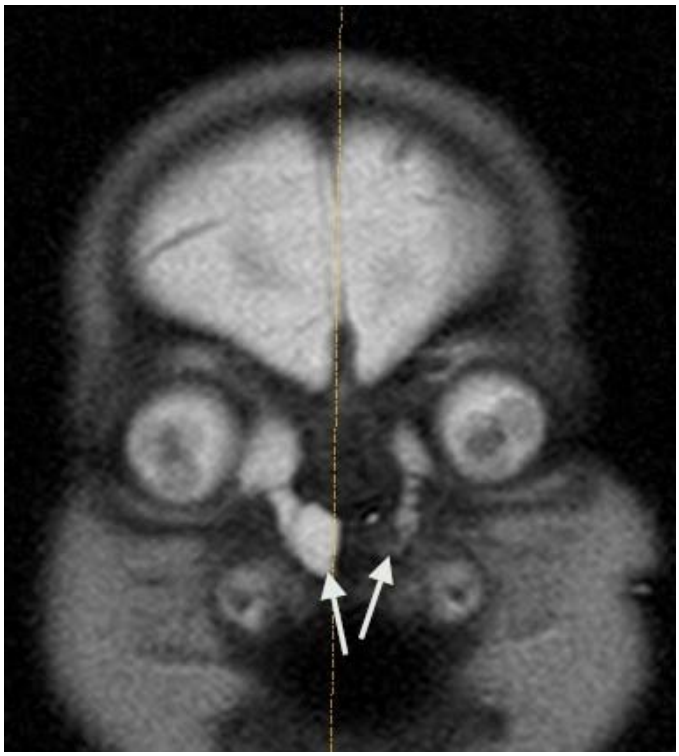

FIGURE 4

Case 1. Magnetic resonance image showing bilateral enlargement of the lacrimal sacs and lacrimal ducts, and nasolacrimal duct cysts (arrows), right greater than left.

Case 2 (Acute Dacryocystitis With Group B Streptococcal Sepsis; No dacryocystocele, No NLD cyst)

The infant was evaluated at age 3 weeks with a 2-day history of progressive erythema and swelling in the left medial canthal area. There was no history of a dacryocystocele. Examination revealed preseptal swelling and cellulitis overlying the lacrimal sac. The infant was febrile, and an evaluation for sepsis revealed positive blood cultures for group B *Streptococcus*. The patient was treated with intravenous antibiotics for 10 days. The patient underwent nasolacrimal probing and endoscopy. Nasolacrimal cysts were not present. The patient's symptoms resolved.

DISCUSSION

EMBRYOLOGY

Knowledge of the embryologic development of the lacrimal system helps in understanding congenital abnormalities of these structures. Most periocular structures begin to develop during the fourth week of gestation from the frontonasal and maxillary processes, which are composed of neural crest cells. During the fifth gestational week, surface ectoderm proliferates and the eyelids begin to form, covering the developing eye. The upper and lower eyelids form separately, eventually fusing at approximately 10 weeks gestation.

The lacrimal drainage structures begin to form during the fifth week of gestation as a crease between the frontonasal and maxillary processes, known as the nasolacrimal groove or naso-optic fissure.[8] A solid cord of ectodermal tissues separates from the surface and enters this groove. This tissue eventually canalizes and forms the lacrimal sac and nasolacrimal duct. A similar process results in the formation of the lacrimal canaliculi. Canalization begins by the eighth week of gestation and continues until birth. Sevel[9] demonstrated that this canalization occurs along the entire system contemporaneously. He noted that partial membranes, which could create a valvelike effect, rarely were found. When present, these most commonly were located at the junction between the canaliculi and lacrimal sac and the junction between the lacrimal sac and nasolacrimal duct.

It is not unusual for the opening between the nasolacrimal duct and the nasal cavity to be incomplete at birth.[10] In an autopsy and histological study of 15 stillborn infants, Cassady[11] found that 73% of ducts were imperforate. In these patients, the membranous tissue between the lacrimal duct and the nasal cavity consisted of epithelial tissue from the lacrimal duct on one side and nasal mucosa on the other side. Prior to sectioning the specimens, Cassady injected fluid into the upper and lower canaliculi. In those patients with imperforate systems, distention of the lacrimal sac at the distal end of the lacrimal duct developed, creating a balloonlike expansion beneath the inferior turbinate (Figure 5). This corresponds to the clinical appearance of nasolacrimal duct cysts that are associated with dacryocystoceles.

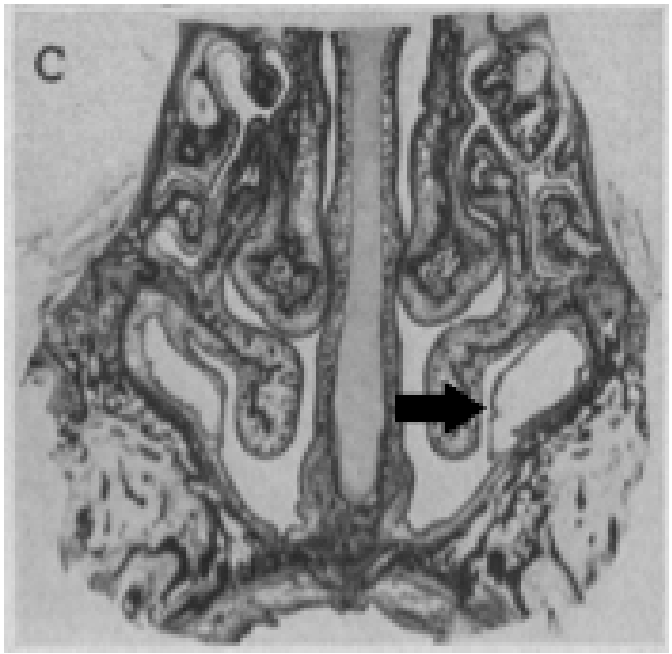

FIGURE 5

Section from cadaver of infant showing cystic protrusion of distal nasolacrimal duct extending beneath the inferior turbinate (arrow on left cyst), following injection of fluid into lacrimal system. (Reprinted with permission. Archives of Ophthalmology, Volume 47, Page 156. Copyright © 1952 American Medical Association. All rights reserved.)

At birth, most infants produce tears.[12] Studies of preterm infants show decreased tear secretion compared to full-term infants.[13] At term, tear production is near normal in most infants. Toker and associates[14] found that approximately two-thirds of full-term infants had normal secretion and that this increased during the first 2 to 4 weeks of life.

Why Do Dacryocystoceles Form?

In order for a fluid-filled mass to develop, material must be able to enter the structure but not be able (or have a limited ability) to exit. The nature of the fluid within dacryocystoceles is not known. Possibilities include amniotic fluid, normal tears, and mucus. It is unlikely that any of these are present in all dacryocystoceles, for the following reasons. Amniotic fluid could be present in lesions that are present at birth, but cannot explain lesions that develop after birth. Conversely, normal tears could not be present in lesions that are present prenatally (as has been documented in several reports of prenatal ultrasonography; see "Imaging" section below). Mucoid material produced by the lining of the lacrimal sac could be present in both prenatal and postnatal lesions, without requiring the entrance of external fluid for dilation of the lacrimal sac to occur. However, at least some material must be able to enter the lacrimal system. If it could not, then infections would not develop, because bacteria could not gain access to the lacrimal sac.

The most likely etiology for dacryocystocele formation combines two elements (Figure 6). First, material must be able to enter the lacrimal system, but not leave it readily by the same route through which it entered. This implies a one-way valve effect between the canaliculi and the lacrimal sac. Jones and Wobig[15] proposed that patients without a common canaliculus might develop this condition because the canaliculi drain into the sinus of Maier, the posterior margin of which represents the valve of Rosenmuller. The valve effect is created because the sinus enters the lacrimal sac at an acute angle, creating an obstruction when the sac is distended. Cibis and associates[16] noted a snapping sensation when a lacrimal syringe was withdrawn across this area, which they attributed to closing of the valve. The inability of fluid to exit the sac through the canaliculi is also demonstrated by the observation that the lacrimal sac contents frequently cannot be expressed in patients with dacryocystoceles despite manual pressure over the sac. For instance, Levin and associates[17] reported that "despite an aggressive initial attempt at manual decompression in all infants, only 40% showed any reflux discharge via the puncta and none showed externally visible nasal drainage."

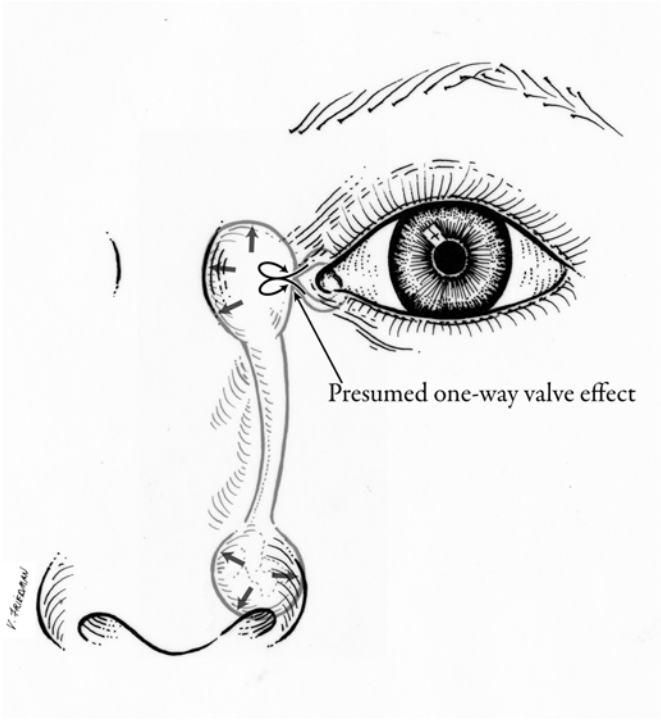

FIGURE 6

In order for a nasolacrimal duct cyst to form, fluid must be able to enter, but not exit, the lacrimal sac. This presumed one-way valve effect creates pressure within the lacrimal system (arrows), causing distention of the lacrimal sac and formation of a nasolacrimal duct cyst due to protrusion of the imperforate membrane occluding the distal lacrimal duct.

The second element necessary for dacryocystocele formation is obstruction of the distal duct, which allows pressure to rise within the lacrimal system. This can be readily explained by the presence of a persistent membrane between the lacrimal duct and nasal mucosal epithelium, which is present in many newborns.[11] If this membrane does not perforate as fluid accumulates in the lacrimal sac, pressure may rise and the lacrimal sac may expand, creating the dacryocystocele. The membrane itself also expands beneath the inferior turbinate to produce an NLD cyst. This process was illustrated in the study by Cassady,[11] described previously, in which

cystic structures developed in infant cadaver specimens as fluid was injected into the lacrimal system (Figure 5). Increased pressure may also cause distention of the nasolacrimal duct. This is illustrated by Rand and associates' report[18] of computed tomography (CT) findings in four patients with dacryocystoceles and NLD cysts, in which the size of the normal nasolacrimal ducts were 1 to 2 mm, as compared to 4 to 5 mm on the affected side. Distention of the lacrimal duct and sac does not occur in all cases of NLD cysts. This is illustrated by cysts that are sometimes present on the contralateral side of affected infants, but which are asymptomatic. In these cases, the membrane itself may be less rigid and therefore easily distensible, such that pressure within the remainder of the lacrimal system does not increase.

EPIDEMIOLOGY

Cassady[11] found that 73% of infants examined at autopsy had imperforate distal nasolacrimal ducts at birth. However, only approximately 6% to 20% of infants develop symptoms of NLDO.[1,2] Many patients with NLDO have relatively mild symptoms, and they often spontaneously improve. Therefore, treatment normally involves conservative measures such as nasolacrimal massage and topical antibiotics. In an observational study, Petersen and Robb[19] found that 89% of NLDO resolved with conservative treatment, with most patients improving by 9 months of age. MacEwen and Young[2] reported similar results, with a resolution rate of 96% by 1 year of age.

Acute dacryocystitis associated with erythema and swelling overlying the lacrimal sac is uncommon in infants. This presentation was present in 7 (2%) of 440 children with NLDO reported by Ffooks,[20] 2 (2%) of 100 children reported by Cassady,[21] and 3% of patients reported by Pollard.[22]

Dacryocystoceles are also uncommon. Becker[23] reported two cases in a series of 250 infants with NLDO, for an incidence of 1%. Shekunov and associates,[24] in a review of patients in Olmsted County, Minnesota, reported a birth prevalence of 1 in 3,884 live births. Although Shekunov did not report the number of patients with typical NLDO in their population, an expected incidence of 6% would yield 233 patients. The nine children they found with dacryocystoceles would therefore represent 4% of the patients with NLDO.

CLINICAL PRESENTATION OF NEONATAL DACRYOCYSTOCELES

Dacryocystoceles are usually present at or within a few days of birth. They are characterized by a bluish-tinged mass medial to and below the medial canthus (Figure 7). They may present as isolated abnormalities or may be associated with acute dacryocystitis or respiratory distress.

Acute Dacryocystitis

Dacryocystoceles are often, but not always, associated with acute dacryocystitis. If untreated, there is a relatively high risk of infections developing in these lesions. Acute dacryocystitis is characterized by swelling and erythema overlying the lacrimal sac. More severe infections may be complicated by progressive cellulitis and formation of an abscess within the lacrimal sac (Figure 8).

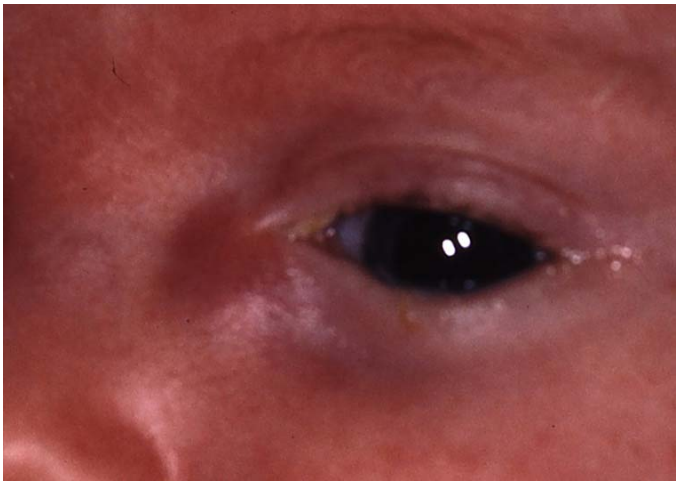

FIGURE 7

Dacryocystocele in an infant. Note violet-hued mass overlying left lacrimal sac, beneath the medial canthus.

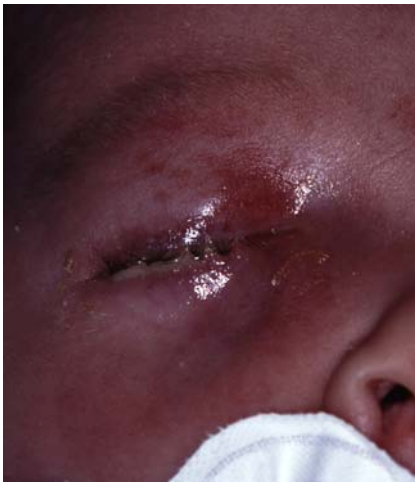

FIGURE 8

Infant with infected dacryocystocele, with erythema and distention of tissue overlying the lacrimal sac, cellulitis extending onto upper eyelid, and purulent discharge.

Respiratory Distress

Because infants primarily breathe through their noses, any mass that obstructs the nasal cavity may cause respiratory difficulties. The symptoms may vary in severity from frank respiratory failure associated with cyanosis to problems during feeding (due to

simultaneous obstruction of the mouth and nares) or difficulty sleeping (due to the mouth being closed). Partial obstruction may manifest as chronic snorting respirations.

Multiple reports have described nasolacrimal duct cysts as a cause of this phenomenon. These have been primarily in the otolaryngologic literature, owing to the nature of this presentation.[25-30] Most patients have had large bilateral intranasal cysts. They present with symptoms including poor feeding, cyanosis, and labored breathing. The symptoms develop shortly after birth or with progressive problems during the first few weeks of life. Most patients have had associated dacryocystoceles, but in some patients there were no visible external masses despite radiographic evidence of lacrimal sac distention.[28] Surgical treatment, with either resection of the intranasal cysts, lacrimal probing, or a combination of both, has been successful in relieving the respiratory symptoms in these patients.

DIFFERENTIAL DIAGNOSIS OF NEONATAL DACRYOCYSTOCELES AND ACUTE DACRYOCYSTITIS

The presence of an enlarged erythematous mass overlying the lacrimal sac in a newborn is usually adequate to establish the diagnosis of an infected dacryocystocele. If the dacryocystocele is not infected or inflamed, the typical location supports the diagnosis. However, other possible diagnoses need to be considered. The presence of purulent periocular discharge supports the diagnosis of an infected dacryocystocele, but its absence does not rule it out. This is because there is resistance to retrograde flow in some patients that prevents backflow of the purulent material through the canaliculi, even in the presence of a distended lacrimal sac.

An important anatomic consideration that is helpful in establishing the diagnosis is the precise location of the lesion. Dacryocystoceles present beneath the medial canthus. They may extend onto the medial lower eyelid, but they do not arise directly on the eyelid. Lesions that present above the medial canthus or on the medial lower eyelid, unless they are extensions of larger lesions centered beneath the medial canthus, are very unlikely to be dacryocystoceles.

Other features that help establish the diagnosis include the absence of globe displacement, the absence of nasal deformities, and the presentation of dacryocystoceles at birth or within the first 1 to 2 weeks after birth.

Lesions Included in the Differential Diagnosis of Dacryocystocele

In the descriptions that follow, many of the patients are beyond the neonatal period when dacryocystoceles appear. Although these patients are older, those with lesions that could have presented early in life (for example, ruptured dermoid cysts) are included for the sake of completeness.

Encephaloceles. Encephaloceles are protrusions of central nervous system (CNS) tissue through defects in the skull bone. If the anterior skull is involved, orbital and periocular masses may be present. They usually present above the medial canthus, but some lesions have been described in the area of the lacrimal sac.

Esila and associates[31] reported a 5-month-old infant who presented with symptoms of NLDO and an enlarging lesion in the area of the right lacrimal sac. The mass involved the lower eyelid, and the right globe was displaced laterally and superiorly. The patient had symptoms of NLDO that did not resolve with probing. Pneumoencephalography revealed an encephalocele. The lesion was surgically removed and the bony defect was repaired.

Chohan and associates[32] reported a 6-month-old female with a medial left orbital lesion that was present shortly after birth and which gradually enlarged to involve the upper and lower eyelids. The lesion transilluminated. The patient had undergone several lacrimal probings for presumed NLDO. At surgery the patient was found to have an encephalocele communicating with the lacrimal sac.

Rashid and colleagues[33] reported a 3-year-old male with a history of NLDO who had lacrimal probing performed in infancy. The patient had a midline mass with an underdeveloped nasal dorsum and extension of a mass into both medial canthi, with hypertelorism. The diagnosis of encephalocele was established by CT, which showed CNS tissue extending through a defect at the juncture of the frontal and nasal bones. The encephalocele was surgically excised and the bony defect repaired.

Tumor. Lazaridou and associates[34] reported a very unusual case of a 17-day-old infant who presented with a history of left ocular discharge and epiphora, with swelling beneath the left medial canthus. The patient was thought to have a dacryocystocele, and probing was performed at age 1 month. The probe could not be passed into the lacrimal sac. The symptoms persisted, and 2 weeks later rapid enlargement of the lesion developed. Imaging studies revealed a solid, vascular mass in the inferior left orbit causing superior displacement of the globe. Biopsy revealed an alveolar rhabdomyosarcoma.

Hemangioma. Infantile capillary hemangiomas may present anywhere on the skin, including the periocular area. They may be present at birth, but typically are first noted after 1 to 2 weeks. They grow rapidly over the next few months, then gradually regress over several years. Superficial lesions have a vascular appearance, either bright red or purplish. Purple-tinged lesions centered over the lacrimal sac may have an appearance similar to dacryocystoceles (Figure 9).

Features that distinguish hemangiomas from dacryocystoceles include their later appearance (Figure 10), the presence of vascular markings on hemangiomas, the frequent presence of hemangiomas elsewhere on the body, and the absence of symptoms of NLDO.

Dermoid and epidermoid cysts. Dermoid cysts arise from entrapment of dermal and epidermal tissue within bony sutures during development of the skull. These most commonly occur in the superolateral orbit but may also develop superonasally. They are usually present at birth and present as cystic masses that are not inflamed unless they rupture.

Lelli and Levy[35] reported a 21-month-old infant who presented with a mass in the right superior medial canthal region that had been gradually enlarging since birth. The patient had no symptoms of lacrimal obstruction, and the lesion was not inflamed. Computed tomography revealed a cystic lesion filled with fluid that was the same density as water, which was in close proximity to the lacrimal sac. The radiologic interpretation was a dacryocystocele. Excisional biopsy revealed an epidermoid cyst. Epidermoid

cysts are similar to dermoid cysts, but are more likely to present medially. Histology shows only epidermal elements without adnexal structures. The investigators emphasized that the location of the lesion above the medial canthus and the absence of symptoms of NLDO were the most important clinical factors that distinguished the lesion from a dacryocystocele.

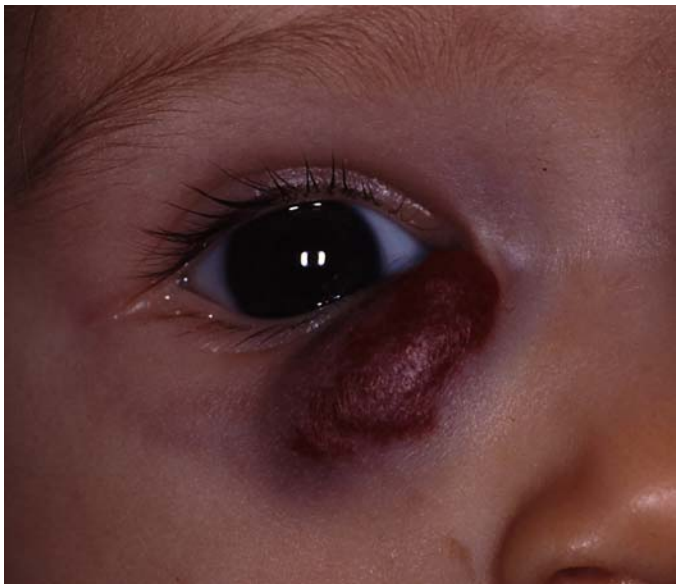

FIGURE 9

Hemangioma overlying right lacrimal sac in a 7-month-old infant. The lesion has a vascular appearance on the surface.

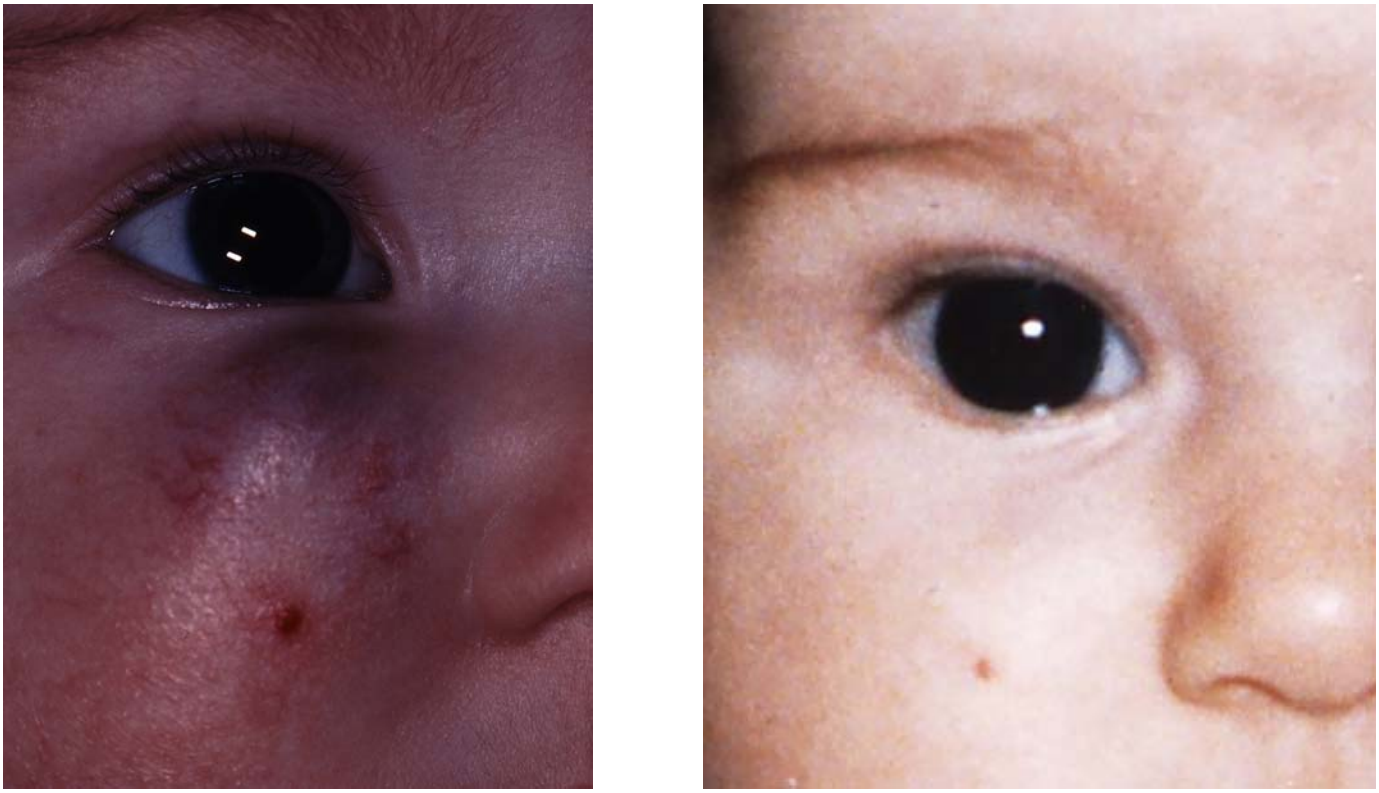

FIGURE 10

Left, Four-month-old female with right periocular hemangioma. Note the vascular markings overlying the lesion and location not directly over the lacrimal sac. Right, Family photograph of patient at age 2 months shows no abnormality.

Hurwitz and associates[36] reported a 27-year-old patient with an inferonasal mass overlying the lacrimal sac that had been present since childhood. The lesion was not inflamed, and the appearance was similar to a dacryocystocele. During surgery, the lesion was noted to be adherent to the lacrimal sac and common canaliculus. Histopathologic examination revealed a dermoid cyst.

If a dermoid cyst ruptures, the contents of the lesion may incite a marked inflammatory response. If a cyst is located in the area of the lacrimal sac and this occurs, it could be mistaken for acute dacryocystitis, as demonstrated in the following case:

Case 3 (ruptured dermoid cyst). The patient was seen at 17 months of age with a 3-week history of swelling over the right medial canthal area that had extended to the left medial canthal area over the previous day. On examination the patient had swelling and erythema in the area overlying the lacrimal sacs and the bridge of the nose (Figure 11). This clinically resembled a lacrimal sac abscess, but the patient had no prior history of NLDO symptoms. Computed tomography revealed a round lesion separating the nasal bones near the glabella, with remodeling of the surrounding bone. The patient was diagnosed with a ruptured dermoid cyst, which was treated successfully by an otolaryngologist.

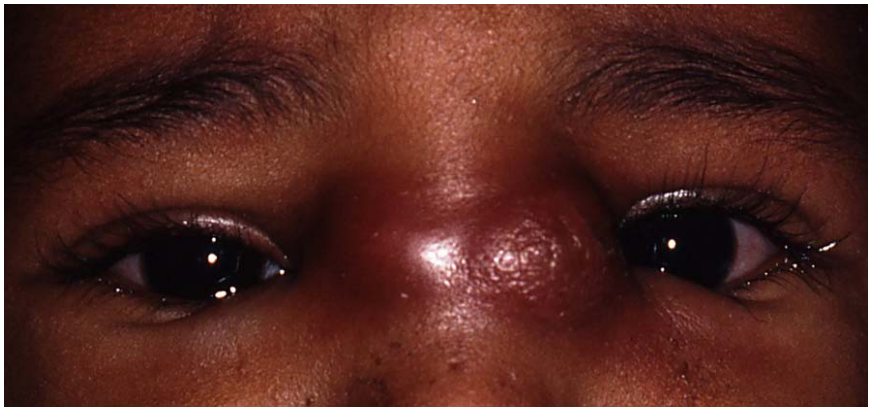

FIGURE 11

Case 3. Seventeen-month-old infant with bilateral swelling and erythema in the area of the lacrimal sacs due to ruptured dermoid cyst.

Lymphangioma. Orbital lymphangiomas may present in early life but are usually not diagnosed until patients are older. They may cause proptosis or a mass within the eyelid. They often enlarge in association with upper respiratory tract infections and may cause rapid proptosis if hemorrhage occurs within the lesions. Although these lesions should be considered in the differential diagnosis of dacryocystoceles, no case reports of this presentation were identified.

Dacryocystitis secondary to lacrimal sac diverticulum. Patients with other anatomic abnormalities of the lacrimal system may present with periocular masses and acute dacryocystitis. The following patient is included in the data for older patients in this report:

Case 4 (lacrimal sac diverticulum with pseudodacryocystocele). The infant was evaluated at 4 months of age with a history of discharge from the right eye 2 weeks earlier that had resolved with topical antibiotics. A cystic mass developed on his medial right lower eyelid after resolution of the infection. On examination he had a firm whitish nodule of the medial right lower eyelid (Figure 12, left). There was no reflux of material with pressure over the lacrimal sac. The lesion was thought to possibly represent a dacryocystocele, and the patient underwent nasolacrimal probing and endoscopy. No nasolacrimal duct cyst was identified, and the firm nodule did not resolve with passage of the lacrimal probes into the distal duct. Postoperatively the lesion resolved, and the patient did well until age 11 months, when he presented with a 4-day history of progressive swelling and erythema of the medial right lower eyelid, in the area of the previous mass (Figure 12, right). Computed tomography revealed distention of the right lacrimal sac along the lower eyelid (Figure 13). The patient underwent dacryocystorhinostomy by an oculoplastic surgeon, during which an infected lacrimal sac diverticulum was identified and excised. The patient subsequently did well.

IMAGING OF DACRYOCYSTOCELES

In the author's experience, imaging of infants with dacryocystoceles is usually not necessary. However, it may be useful in patients with atypical features in whom the diagnosis is in question. Various imaging modalities have been used to demonstrate the characteristic findings associated with neonatal dacryocystoceles and nasolacrimal duct cysts: distention of the lacrimal sac, dilation of the lacrimal duct, and a soft-tissue mass beneath the inferior turbinate.

Computed Tomography

Computed tomography findings have been reported by several investigators. Raflo and associates[37] reported an infant with a dacryocystocele in whom an intranasal lesion was visualized during an office examination. Computed tomography with dacryocystography revealed marked distention of the lacrimal sac, lacrimal ducts, and intranasal extension of the mass. Rand and colleagues[18] described 4 infants with dacryocystoceles who had nasal airway obstruction. All of the patients had enlarged lacrimal sacs, dilated nasolacrimal ducts (4 to 5 mm in width compared to 1 to 2 mm on the normal side), intranasal cysts that obstructed the

nasal passages, and displacement of the inferior turbinate and lacrimal septum. Castillo and associates[38] used CT to evaluate 2 newborns who presented with acute respiratory distress in the first day of life. Both were found to have bilateral soft-tissue masses beneath the inferior turbinates. Neither infant had distention of the lacrimal sac itself.

Dacryocystography

Dacryocystography was used by Cibis and associates[16] to evaluate 3 infants with dacryocystoceles that could not be decompressed with manual massage. Radiographs were performed during nasolacrimal duct probing after injection of radiopaque contrast media. These revealed distention of the lacrimal sacs, widening of the nasolacrimal ducts, and intranasal cysts. Narioka and Ohashi[39] reported an infant with an infected dacryocystocele who failed an initial probing. Subsequent probing using dacryocystography with fluoroscopic guidance revealed dilation of the lacrimal sac and an intranasal cyst. The procedure was successful.

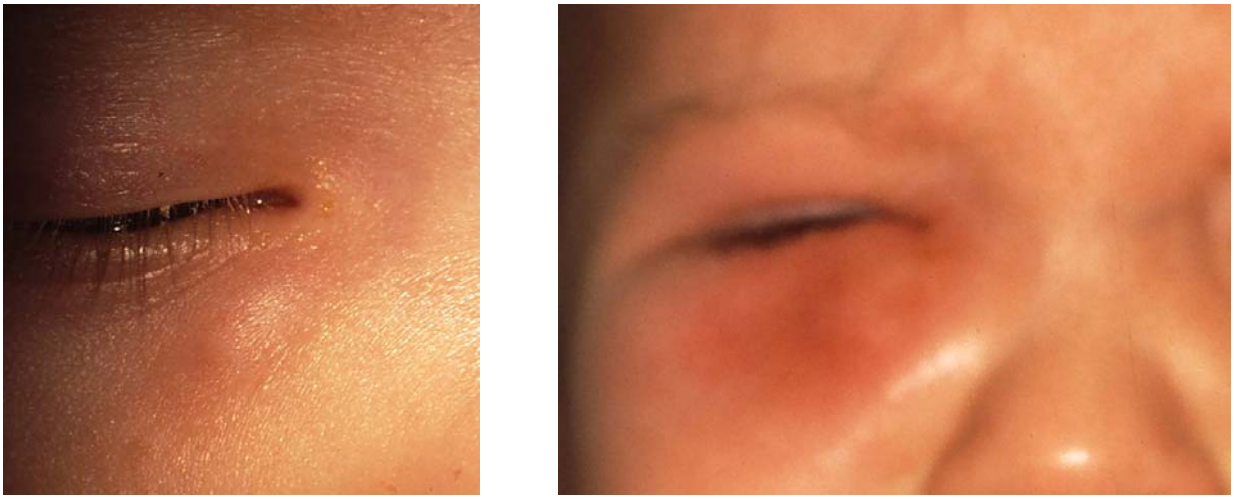

FIGURE 12

Case 4. Left, Four-month-old infant with periocular firm whitish nodule and symptoms of nasolacrimal obstruction. Right, Same patient at age 11 months with marked swelling and erythema overlying same area as previous nodule.

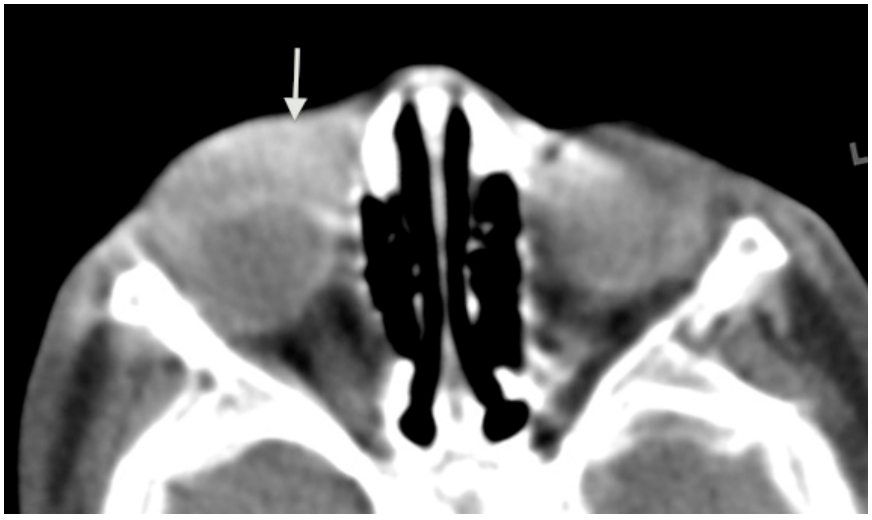

FIGURE 13

Case 4. Computed tomography image showing distention of lacrimal sac (arrow) extending adjacent to lower eyelid.

Echography

Echography has also been used to image dacryocystoceles. Scott and associates[40] described the use of A- and B-scan ultrasonography in 2 infants to demonstrate dilation of the lacrimal sac and lacrimal duct, as well as the presence of fluid within the sac. Divine and associates[41] demonstrated the connection between a dacryocystocele and a nasolacrimal duct cyst using echography in a 10-day-old infant.

Prenatal diagnosis. Several investigators have described prenatal diagnosis of dacryocystoceles by ultrasonography.[42-47] Echography demonstrates hypoechoic masses in the area of the lacrimal sac. In one of the patients reported by Sepulveda and associates,[47] 3-dimensional sonography showed extension of the lesion into the nasal cavity. Most prenatal diagnoses have been made on ultrasound studies performed at 30 to 33 weeks gestation. The earliest reported identification of dacryocystoceles is at 27 weeks gestation.[42,47]

Many of the infants in these reports were found to have dacryocystoceles on examination after birth, but some had no external abnormalities postnatally.[43,45] In a study of 10 infants, Sepulveda and associates[47] found that the lesions resolved prior to birth in 7 of 10 infants.

Magnetic Resonance Imaging

MRI has also been used in the evaluation of dacryocystoceles.[28-30,48] MRI provides better resolution of soft-tissue lesions but is not as good as CT for imaging bone. One of the primary advantages of MRI is the avoidance of ionizing radiation, which may be associated with an increased risk of secondary cancer.[49,50]

TREATMENT OF DACRYOCYSTOCELES AND INFANTILE ACUTE DACRYOCYSTITIS

The recommended treatment of infants with dacryocystoceles is different from that of typical NLDO. In the latter, many infants spontaneously improve during the first several months of life, and conservative treatment is therefore recommended. Children with dacryocystoceles usually require early intervention, either because of the pronounced infection that develops or because of respiratory compromise. These patients often have abscesses in the lacrimal sac that necessitate drainage. Because neonates are relatively immunocompromised, they have an increased risk of local and systemic spread of infection.[51] Prompt surgical treatment and systemic antibiotics are indicated to decrease the risk of this complication.

Literature Review of Treatment

There have been many reports of treatment of infants with dacryocystoceles and infants with acute dacryocystitis. These are described in the following sections and summarized in Table 2.

Acute dacryocystitis. Ffooks[20] reviewed his experience with acute dacryocystitis and lacrimal abscesses in infants with lacrimal obstruction in 1961. He identified seven patients with acute dacryocystitis in a group of 440 infants with NLDO (2%). At least two of the infants had masses overlying the lacrimal sac prior to the onset of infection. All patients improved with NLD probing and antibiotics, but one patient required three procedures before the symptoms resolved.

In Petersen and Robb's study[19] of the natural history of NLD obstruction, they included seven patients with dacryocystoceles, one of whom had acute dacryocystitis. One patient improved with conservative treatment, and the others improved with either canalicular or NLD probing.

Pollard[22] reported 25 infants that developed acute dacryocystitis within the first 3 weeks of life. The patients represented 3% of all patients with NLDO seen during the same period. Six patients had dacryocystoceles and were initially managed conservatively, but four subsequently developed acute dacryocystitis. Treatment consisted of simple NLD probing, which was successful in 24 of 25 patients.

Neonatal dacryocystoceles. The earliest report of a dacryocystocele identified in a review of the English-speaking literature was Mayou's in 1908.[10] The first identified report of a neonatal dacryocystocele associated with a nasolacrimal duct cyst was Raflo and associates' description of a 2-week-old infant with a blue-tinged medial canthal mass.[37] The mass could not be reduced with manual pressure. An intranasal abnormality was visualized, and the presence of a distended lacrimal sac and intranasal lesion was confirmed with CT and dacryocystography. The patient was initially treated with a vertical incision through the cyst, which resulted in immediate release of fluid and decompression of the cyst. However, the dacryocystocele recurred after 2 weeks, necessitating repeated surgery with complete removal of the intranasal cyst, after which the symptoms resolved.

In 1983, Divine and associates[41] described a 10-day-old infant with congenital dacryocystoceles that had recurred and become infected despite two nasolacrimal duct probings. A nasal abnormality was noted, and echography demonstrated a connection between the dacryocystocele and the intranasal lesion. The child's symptoms resolved after excision of the nasal mass.

Weinstein and associates[52] reported 7 infants with dacryocystoceles in 1982. Four of the dacryocystoceles were not infected. One resolved with massage, and three resolved with NLD probing. Three of the infants developed dacryocystitis. Two of these infants required repeated NLD probing to cure the infections.

Mansour and associates[53] reported 54 patients with dacryocystoceles in a collaborative study involving seven centers, which is the largest series identified in the literature. Seventeen patients (31%) had associated cellulitis. The condition resolved without surgical intervention in 9 patients (17%), and NLD probing was performed in 45 patients (83%). In the latter group, 10 (22%) of the 45 patients required more than one procedure. Six of these patients had three or more procedures. Nasal endoscopy was not performed routinely, but NLD cysts were found in 6 patients, 2 by examination and 4 by radiographic studies. The investigators believed that at least 7 additional patients likely had cysts, based on the inability to visualize the probe or irrigate, or because the patients had multiple recurrences.

Sullivan and associates[5] reported seven patients with congenital dacryocystoceles. In four patients the lesions resolved spontaneously. Three patients were treated with NLD probing, and one of these required a second probing to effect a cure. None of the patients had nasal examinations.

TABLE 2. OUTCOMES IN PREVIOUS STUDIES OF DACRYOCYSTOCELE AND/OR INFANTILE ACUTE DACRYOCYSTITIS

STUDY	RESOLUTION WITH CONSERVATIVE TREATMENT	SUCCESS WITH NLD PROBING	SUCCESS WITH ENDOSCOPIC CYST REMOVAL
Ffooks, 1961[20]		6/7 (86%)	
Weinstein et al, 1982[52]	1/7 (14%)	4/6 (67%)	
Raflo et al, 1982[37]			1/1 (after failed NLD probe with slit into cyst)
Harris and DiClementi, 1982[4]	2/4 (50%)		
Divine et al, 1983[41]			1/1 (after failed NLD probe)
Boynton and Drucker, 1989[65]	1/6 (14%)		
Mansour et al, 1991[53]	9/54 (17%)	35/45 (78%)	3/4 (75%) (4th patient success after "wider manipulation"
Grin et al, 1991[66]			7/7
Pollard, 1991[22]		24/25 (96%)	
Sullivan et al, 1992[5]	4/7 (57%)	2/3 (67%)	
Meyer et al, 1993[61]			1/1
Paoli et al, 1993[62]			3/3
O'Keefe et al, 1994[54]	3/22 (14%)	14/16 (88%)	
Schnall and Christian, 1996[55]	16/21 (76%)	4/5 (80%)	
Levin et al, 2003[17]	1/25 (4%)		22/24 (92%)
Becker, 2006[57]	3/29 (10%)	10/10 (100%) noninfected; 10/19 (53%) infected	1/1
Wong and Vanderveen, 2008[58]	10/42 (24%)	28/40 (70%)	3/3
Shekunov et al, 2010[24]	3/9 (33%)	3/3 (100%)	3/3
Present study			31/33 (94%)
Total	53/226 (23%)	140/179 (78%)	76/81 (94%)

NLD, nasolacrimal duct.

O'Keefe and associates[54] reported 22 patients who presented with dacryocystoceles during the first 3 months of life. Six patients did not have infections. Three of these resolved spontaneously, and three resolved after NLDO probing. The other 16 patients presented with acute dacryocystitis and were treated with intravenous antibiotics. Eight of these were treated with percutaneous drainage of the lacrimal sac followed by NLD probing. Eight were treated with NLD probing alone. One patient in each group had persistent symptoms.

Schnall and Christian[55] initially treated 17 infants with dacryocystoceles conservatively. They reported their results as the number of dacryocystoceles (21) rather than the number of patients. Seventeen of 21 dacryocystoceles were not infected at the time of initial presentation, and 12 of these resolved with conservative treatment. Of the five nonresolving dacryocystoceles, four were treated successfully with NLD probing and one required two probings. Three patients had infected dacryocystoceles at the time of presentation, and one developed cellulitis while being treated conservatively. These four patients were treated with intravenous antibiotics, and their dacryocystoceles resolved without NLD probings.

Paysse and associates[56] reported a series of 22 patients with dacryocystoceles. Twelve (55%) of the patients had acute dacryocystitis. Nasal endoscopy was performed in 13 patients, and nasal examination was performed in seven. Nasolacrimal duct cysts were identified in 16 (80%) The investigators did not specify whether these were in the patients who had been examined endoscopically or with a speculum and headlight. One patient's dacryocystocele resolved spontaneously. The other patients were treated with a variety of procedures, including NLD probing, stent placement, cyst removal, or a combination thereof. Two of the patients treated with NLD probing alone required additional surgery, and the other treatments were successful.

Becker[57] reported 27 patients with 29 dacryocystoceles that were present at birth. Cellulitis developed in 11 dacryocystoceles (38%), and these patients were treated with intravenous antibiotics. Three dacryocystoceles resolved with conservative treatment. Initial NLD probing was successful in the seven lacrimal systems that were not infected, but in only 10 (53%) of 19 patients with

dacryocystitis or cellulitis. A variety of additional treatments were used in these patients, including repeated probing, balloon catheter dilation, stent placement, and external incision and drainage of the lacrimal sac. At least three patients had intranasal cysts that were identified endoscopically. Becker postulated that the poor success rate of probing alone in the patients with infection was due to thickening of the cyst wall because of inflammation, which made the opening created during probing more likely to close.

Wong and VanderVeen[58] reviewed 42 patients with dacryocystoceles treated at their institution. Infections with either dacryocystitis or cellulitis were present in 28 patients (67%), who were treated with systemic antibiotics. Symptoms resolved with topical or systemic antibiotics in 10 patients without surgical intervention. Of the remaining patients, outpatient NLD probing was successful in 13 (76%) of 17 lacrimal systems, probing in the operating room was successful in 15 (65%) of 23, and cyst removal was successful in 8 of 8 (100%). The investigators did not specify whether nasal endoscopy was performed routinely in all patients.

Several additional studies with smaller numbers of patients have shown similar findings, with improvement with conservative treatment in some patients and successful surgical treatment that included excision of intranasal cysts in some patients.[4,24,59-65]

Studies with routine endoscopy. Three studies were identified in which endoscopy was used routinely in the evaluation and treatment of infants with dacryocystoceles. The earliest of these was Grin and associates' report[66] of eight infants with dacryocystoceles, in whom NLD cysts were found in seven. Five of the patients had dacryocystitis. All of the patients were successfully treated with NLD probing and endoscopic cyst removal.

In an earlier study by the author, 22 infants with neonatal dacryocystoceles were treated with NLD probing and endoscopic cyst removal, with a success rate of 95%.[6] These patients are included in the current study in addition to patients subsequently treated. The details are described in the "Methods" and "Results" sections.

Levin and associates[17] reported a series of children with dacryocystoceles who underwent NLD probing in conjunction with routine endoscopic NLD cyst removal. In this series of 25 infants with dacryocystoceles or acute dacryocystitis, one patient improved with medical management alone. In the 24 patients who underwent surgery, only one did not have a NLD cyst. This patient had dacryocystitis but no dacryocystocele. Treatment was successful in 23 of 24 infants.

Summary of literature review. Table 2 summarizes the results of these studies. Analysis of these results is complicated by the usual problems inherent in such a review. These include the retrospective nature of the studies; lack of uniformity in patient selection, evaluation, and treatment; a mixture of patients with dacryocystoceles, acute dacryocystitis, or both; and variability in outcome measures. While recognizing these limitations, the following can be concluded from a review of these studies:

First, dacryocystoceles may spontaneously resolve. The reported incidence of resolution is very wide, ranging from 14% to 76%. This variability is presumed due to patient selection, referral patterns, and other biases of ascertainment. Overall, spontaneous improvement occurred in 23% of patients.

Second, NLD probing is reasonably effective in treating dacryocystoceles and infantile acute dacryocystitis, with an overall success rate of 78%.

Third, NLD probing in conjunction with NLD cyst removal is exceptionally effective in treating these disorders. Only four patients were identified who did not respond to this treatment. In the study by Mansour and associates,[53] one patient had persistent problems after initial endoscopic cyst removal, which resolved after a "wider marsupialization" was performed. In Levin's study,[17] one of the patients who had recurrent episodes of dacryocystitis did not have an NLD cyst. This patient may have had another anatomic abnormality of the lacrimal system, such as the lacrimal sac diverticulum described previously in Case 4. A second patient in Levin's study and two patients in the present study had symptoms that did not completely resolve, but all had resolution of the dacryocystoceles and marked improvement, with only mild residual symptoms.

COMPLICATIONS OF DACRYOCYSTOCELES AND ACUTE DACRYOCYSTITIS

Respiratory Distress

As already described, infants with NLD cysts may experience respiratory compromise due to obstruction of the nares. This problem develops because infants are obligate nasal breathers. Respiratory compromise usually occurs in the setting of bilateral cysts, buy may occur even if cysts are present unilaterally.[26] Severe cases may present with cyanosis, intercostal retractions, and stridor requiring intubation.[27-30,38] In most patients the respiratory symptoms develop within the first day of life, but they have also been reported at 2 to 3 weeks of age.[26,28,29] The diagnosis is usually readily identified by the presence of respiratory distress in patients with dacryocystoceles. Treatment that includes removal of the cysts has been uniformly successful in the reported cases.

Bilateral Nasolacrimal Duct Cysts in Patients With Unilateral Dacryocystoceles

Paysse and associates[56] noted that patients with unilateral dacryocystoceles can have bilateral NLD cysts. In their report, 2 (17%) of 12 patients with unilateral dacryocystoceles were found to have bilateral NLD cysts. Wasserman and associates[67] confirmed this phenomenon in two patients who had unilateral dacryocystoceles that persisted despite initial NLD probings. One patient had both nares examined intraoperatively, and bilateral cysts were identified and treated. The second patient had a cyst on the affected side, but endoscopy was not performed on the opposite side. Postoperatively the symptoms resolved on the originally affected side, but the patient developed a dacryocystocele on the opposite side, which resolved with medical treatment. In Petersen and Robb's report,[19] one patient with left dacryocystocele and acute dacryocystitis was successfully treated with intravenous antibiotics and NLD probing at 3 weeks of age. One week later the infant developed swelling over the right lacrimal sac with purulent discharge. The association of dacryocystoceles and NLD cysts had not been recognized at that time, but in retrospect this child likely had bilateral NLD cysts.

Subsequent to Paysse and associates' description of asymptomatic contralateral cysts in 2000,[56] the author has performed bilateral

endoscopy during surgical treatment of patients with unilateral dacryocystoceles and infantile acute dacryocystitis. In these patients, bilateral NLD cysts were found in 4 (33%) of 12 infants who had unilateral symptoms.

Potential for Local Spread of Infection and Bacteremia

As discussed already, the acute dacryocystitis associated with dacryocystoceles is different from the low-grade dacryocystitis associated with typical NLDO for three reasons. First, the infection begins earlier in life, usually within the first 1 to 2 weeks. Second, the infection is more severe. If untreated, patients may develop an abscess within the lacrimal sac, and preseptal cellulitis may occur. Third, the immune system in infants is underdeveloped, and they are at greater risk for spread of infection.[51] Therefore, more aggressive systemic antibiotic treatment is indicated for neonates with infections.

In addition to contiguous spread of infection, bacteremia is a risk in infants with infected lacrimal sacs. Surgical manipulation of an infected lacrimal sac increases the chance of developing bacteremia, but it is not necessary for bacteremia to occur. Two studies have compared preoperative and postoperative blood cultures in children undergoing NLDP. Schaeffer and associates reported the results of cultures taken from material expressed from the lacrimal sac and postoperative blood cultures in 12 infants who underwent nasolacrimal probing (Schaeffer et al., Association for Research in Vision and Ophthalmology, 1990, Abstract 2987-34). Four of the patients developed positive blood cultures, and the organism matched the culture from the lacrimal sac material in three of the patients. In a larger study, Eippert and associates obtained preoperative blood cultures in addition to lacrimal and postoperative blood cultures in 40 infants undergoing NLDP. The children ranged in age from 4 to 15 months. They found lacrimal probe–induced bacteremia in 7 (18%) of the 40 infants.[68]

Surgical manipulation of the lacrimal sac is not required to produce bacteremia in the presence of acute dacryocystitis. In a study of patients less than 6 weeks of age with acute dacryocystitis, Baskin and associates[69] reported positive blood cultures taken prior to administration of antibiotics or NLDP in 5 (23%) of 22 infants.

The combination of decreased immune responses and systemic spread of acute lacrimal infection may cause serious problems in infants. In Case 2 (described previously), an infant with acute dacryocystitis developed group B streptococcal sepsis. Other unpublished complications include an infant with an orbital abscess (Amy Hutchinson, MD, personal communication) and two patients with meningitis (Julia Stevens, MD, and Monte Mills, MD, personal communications). Given these risks, the use of systemic antibiotics in young infants with acute dacryocystitis (with or without dacryocystoceles) is indicated.

Failure to Resolve With NLD Probing

As noted in the literature review, the reported success rate of NLD probing alone in infants with dacryocystoceles and acute dacryocystitis ranges from 53% to 100%, with an overall average of 78%. There are at least two likely reasons that simple probings may fail in these patients.

The first reason for probe failure is that during NLD probing the cyst is penetrated but not removed (Figure 14, left and middle). The patient is left with a flaccid tissue mass that may reapproximate and cause reobstruction of the duct. This is illustrated by Raflo and associates' report[37] of a patient in whom a vertical incision through an NLD cyst resulted in drainage of material from within the cyst and partial collapse of the cyst. The patient's symptoms temporarily improved but recurred within a week. The patient was successfully treated with complete removal of the cyst. Removal of this tissue improves the likelihood of cure, as demonstrated by the excellent success rate of NLD probing combined with endoscopic NLD cyst removal (overall success rate of 94% [Table 2]).

The second reason that probes may fail is that acute infection increases the likelihood of scar formation due to inflammation, which may cause recurrent obstruction of the NLD after probing (Figure 14, right). This reason for probe failure is illustrated by Becker's report, in which 10 of 10 patients without infection improved with probing, whereas only 10 of 19 (53%) with acute infection resolved.[57]

THE USE OF NASAL ENDOSCOPY IN THE MANAGEMENT OF NEONATAL DACRYOCYSTOCELES, INFANTILE ACUTE DACRYOCYSTITIS, AND OLDER INFANTS WITH SEVERE SYMPTOMS OF NLDO

Rigid nasal endoscopy has evolved as a useful adjunct to lacrimal surgery over the past 2 decades. The technology was originally developed for otolaryngologists[70] but has been increasingly used by ophthalmologists, primarily to evaluate tearing disorders.[71,72] Raflo and associates[37] were the first to describe the use of endoscopy in the management of dacryocystocele, and it has been reported by many others since, as detailed in the earlier sections of the manuscript.

The instruments and techniques for endoscopy are straightforward. A rigid endoscope attached to a light source allows direct visualization of the nasal cavity. A suction catheter in the other hand can be used to remove blood and to gently move tissue. Familiarity with nasal anatomy is essential but is also straightforward. The primary intranasal site of interest for patients with dacryocystoceles is beneath the inferior turbinate. The nasolacrimal duct normally enters the nasal cavity near the apex of the inferior turbinate, and this is the site where dacryocystoceles are visualized. Infracture of the inferior turbinate improves visualization of the cyst (Figure 15). Removal of the cysts can be performed with the use of alligator forceps or other grasping instruments.

Endoscopy has been used in older children in an attempt to better understand and manage those patients with complicated NLDO.[6,73-75] In an earlier study by the author,[6] distal nasolacrimal duct anomalies were found in 6% of infants whose initial probing was performed after age 18 months and in 9% of children who had failed previous NLD probing.

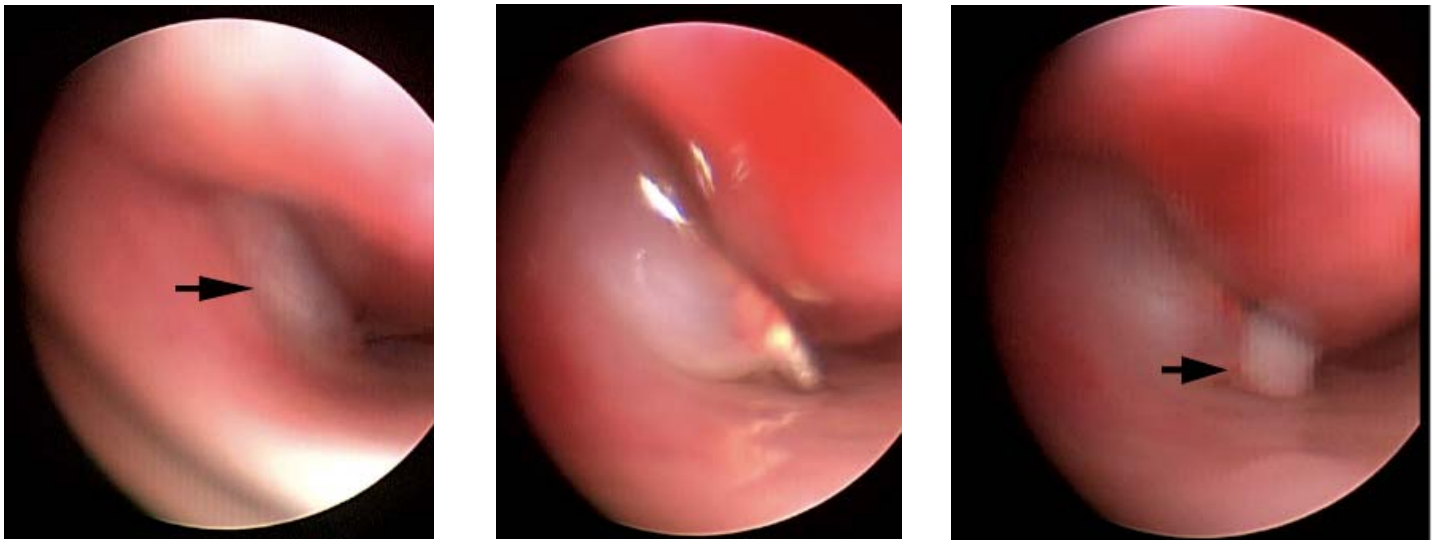

FIGURE 14

Left, Endoscopic view of right nasolacrimal duct cyst (arrow) after infracture of inferior nasal turbinate. Middle, The cyst is penetrated by a Bowman probe. If the cyst is not removed, a large flaccid mass remains at the distal nasolacrimal duct, which may reapproximate to cause recurrent obstruction. Right, After the probe is withdrawn, purulent material escapes from the cyst (arrow), due to presence of abscess within the lacrimal system.

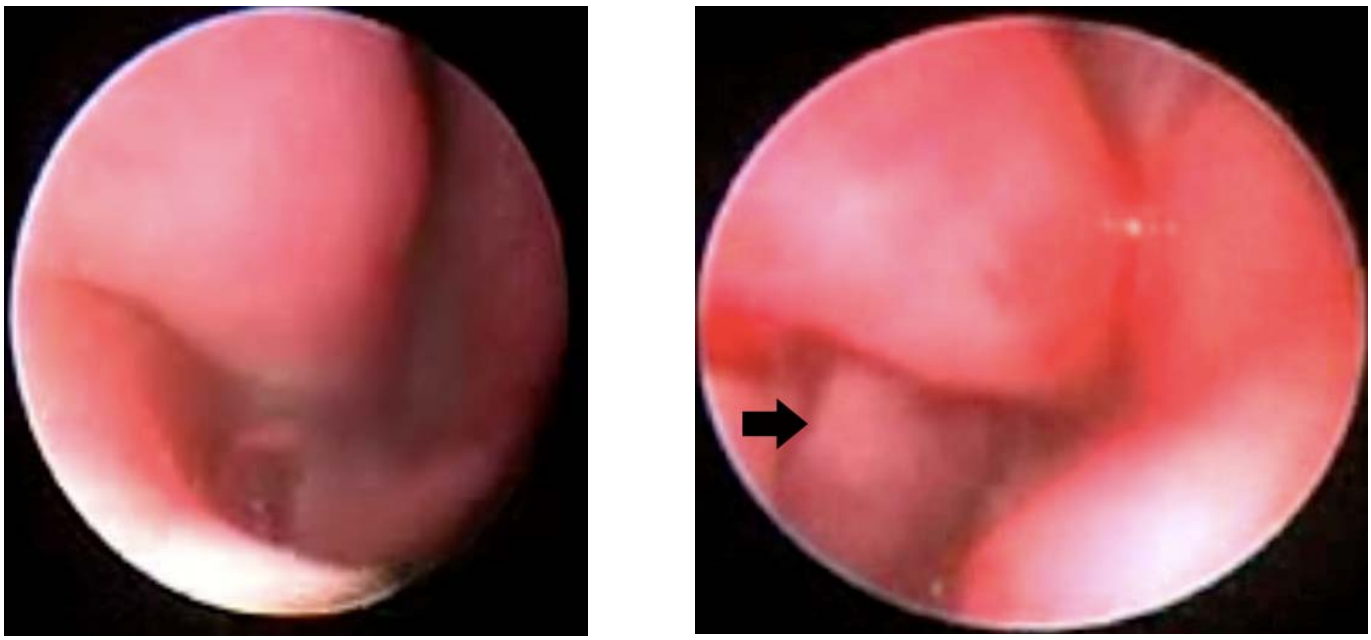

FIGURE 15

Left, Initial endoscopic view of patient with right nasolacrimal duct cyst. The inferior turbinate obstructs the view of the cyst. Right, After infracture of the inferior turbinate, the nasolacrimal duct cyst is easily visualized (arrow).

The current study demonstrates a role of NLD cysts in an additional group of children with NLDO. To our knowledge, the routine use of endoscopy has not been previously studied in young infants who have severe symptoms of NLDO but who do not have dacryocystoceles. The symptoms that warranted early intervention were divided into three categories. The first indication for early intervention was the presence of respiratory symptoms in association with NLDO. Such symptoms suggested that a nasal lacrimal cyst might be present that interfered with breathing, particularly during feeding. It is not surprising that infants with respiratory symptoms in Group 2 were the most likely to have nasal cysts (83% of patients), because the cysts themselves caused the obstruction that led to respiratory difficulties. The second indication for early intervention was acute dacryocystitis. The presence of acute infection with erythema and swelling over the lacrimal sac suggested the presence of an abscess, which required early intervention. In the Group 2 patients with these findings, 3 (75%) of 4 patients with acute dacryocystitis had NLD cysts. The third indication for early

intervention was marked mucopurulent discharge and breakdown of periocular skin. This group of children did not have acute dacryocystitis (with erythema and swelling over the lacrimal sac), but the presence of skin breakdown despite medical treatment warranted early intervention. In the Group 2 infants with these findings, 31% of patients had NLD cysts. The overall success rate of NLD probing and cyst removal in these older patients with severe symptoms was excellent (92%), which supports the use of endoscopy in these patients.

CONCLUSIONS

The current study and the review of the medical literature support a number of conclusions regarding infantile dacryocystoceles, infantile acute dacryocystitis, and nasolacrimal duct cysts:

First, almost all infantile dacryocystoceles and most cases of infantile acute dacryocystitis are associated with nasolacrimal duct cysts. The enlargement of the lacrimal sac appears to be caused by a one-way valve effect that allows fluid to enter, but not exit, the lacrimal system via the canaliculi. The nasolacrimal duct cyst forms due to pressure from above, causing distention of the membrane at the site where the lacrimal duct enters the nares.

Second, early treatment of dacryocystoceles is indicated. Noninfected dacryocystoceles are at risk of progressing to acute dacryocystitis. Because neonates are relatively immunocompromised, infants with acute dacryocystitis are at risk for local or systemic spread of infection. For patients with noninfected mucoceles that do not resolve rapidly with conservative treatment, early NLD probing with or without endoscopic cyst removal may prevent this progression.

Third, patients with acute dacryocystitis should be managed with systemic antibiotics and prompt surgical intervention, because of the risk of spread of infection. The presence of acute infection decreases the success rate of NLD probing alone. Based on the results of this thesis, NLD probing with endoscopic NLD cyst removal is the recommended method of treatment for these patients.

Fourth, patients with unilateral dacryocystoceles and acute dacryocystitis may have bilateral NLD cysts. Patients with unilateral symptoms whose treatment includes nasal endoscopy should have both nares examined for the presence of cysts.

Finally, nasolacrimal duct cysts are also present in many children with NLDO beyond the neonatal period. Endoscopy is a useful adjunct in older infants with severe symptoms of NLDO and in infants whose symptoms persist after NLD probing.

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