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Management of congenital dacryocystocele: report of 3 clinical cases

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Abstract. *Management of congenital dacryocystocele: report of 3 clinical cases*. This case study describes three newborns referred to our oto-laryngologic service for investigating and treating a cystic dilatation of the lacrimal duct. These dilatations corresponded to unilateral or bilateral dacryocystoceles, with or without complications. The first newborn exhibited respiratory distress at birth and received early surgery and endoscopic marsupialization of intranasal and bilateral cysts. The second newborn did not show any signs of complications, and after conservative treatment for a week, the cyst spontaneously resolved. The third newborn was diagnosed *in utero* with ultrasonography, and the cyst resolved spontaneously during childbirth.

These cases provided an opportunity to review the pathophysiology of this rare congenital lacrimal anomaly and to note responses to different therapeutic approaches. Indeed, these three cases illustrated three different management approaches, and allowed us to address the issue of prenatal diagnosis.

Introduction

Congenital dacryocystocele is a rare, benign variant of the congenital nasolacrimal duct cyst. This condition accounts for 0.1%[1] of nasolacrimal duct obstructions that occur during prenatal development. This benign phenomenon is revealed mostly by the formation of a bluish, uni- or bilateral cyst, located at the internal canthus. The diagnosis can be established *in utero*, with ultrasonography, or at birth during examination of the newborn. It is important to make a distinction between congenital dacryocystocele and other diseases with similar symptoms associated with an intra- or paranasal mass.

The dacryocystocele may resolve spontaneously or after application of a prophylactic, well-conducted, conservative treatment.[1,2] Nevertheless, it may be necessary to intervene surgically to ensure the opening of the tear duct, and thus, to prevent rare, but potentially serious complications, such as dacryocystitis or respiratory distress.[1,3,4] Congenital dacryocystocele is an example of a disease that can have multiple scalable modes, which require management adaptations, based on the conditions encountered.

This study describes three patients with congenital dacryocystocele diagnosed in our oto-rhino-laryngological department over the period of one month. We also provided an update on approaches for managing this rare congenital lacrimal anomaly. Indeed, the proposed management of a congenital dacryocystocele has varied widely in the ophthalmic, otolaryngological, and pediatric literature. We took advantage of the fact that the patients presented in this paper illustrated three different treatments, and we highlighted the value of a prenatal diagnosis.

Case histories

Patient 1

At 10 min of life, this boy presented with mild respiratory distress that led to monitoring in the neonatology department. At one day old, he had marked nasal congestion and hypertelorism. Nevertheless, the choanae were permeable to the passage of the suction probe. Next, an examination of the nasal cavity and nasal endoscopy were conducted. These exploratory tests revealed the presence of an intranasal cystic mass, located bilaterally in the inferior meatus. These masses were obstructive, and they were combined with a bluish, cystic dilatation of the internal canthus, predominantly on the left side. These observations indicated a diagnosis of bilateral congenital dacryocystocele.

We performed ultrasound and computed tomography (CT) of the face to assess the masses,

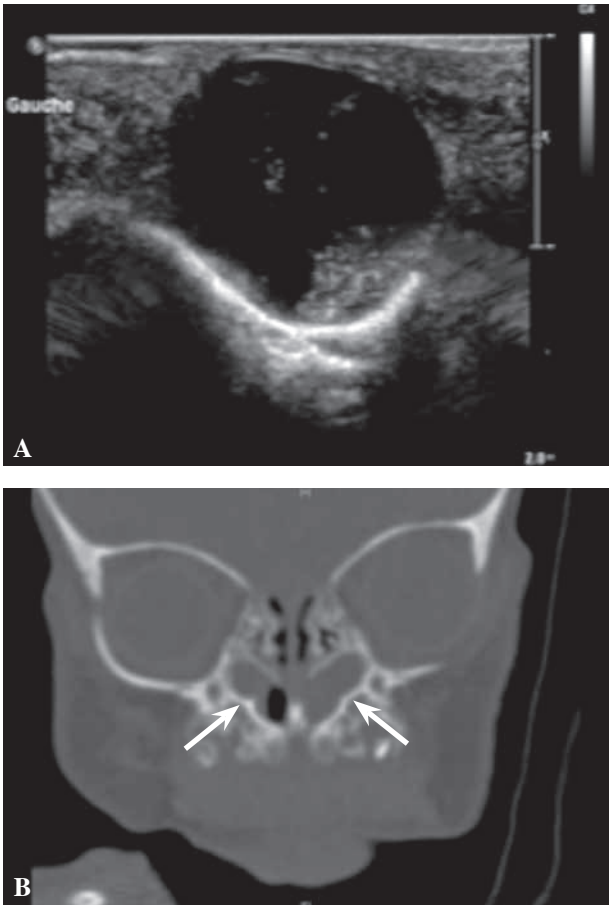


Figure 1
Imaging results for patient 1.
A: The ultrasound examination on post-natal day 2 confirms the cystic nature of the mass.
B-C: CT-scans on post natal day 2 show the two-sided involvement of the lacrimal duct; a voluminous cystic mass on the left propagates into the nasal cavity (white arrows). The tear ducts are widened and no bony obstacle is present.

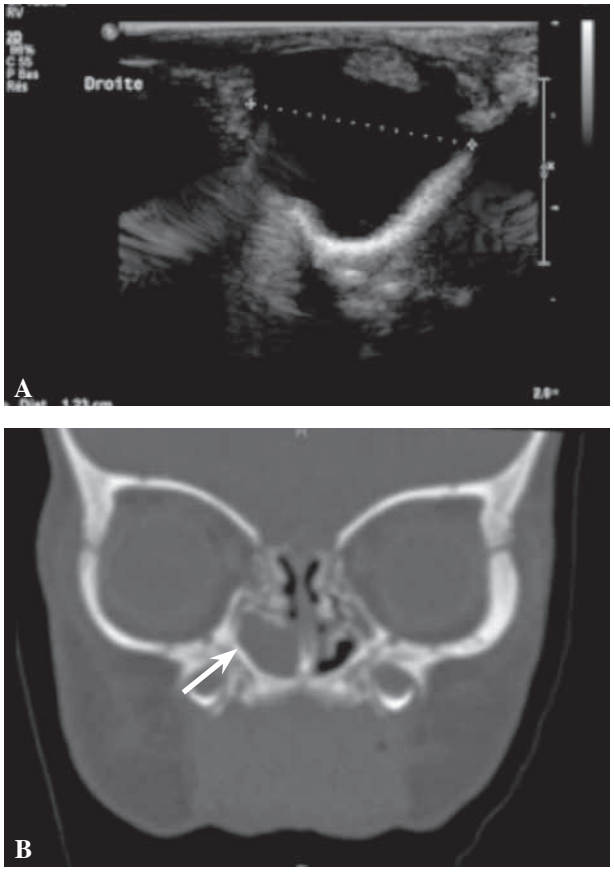
to identify the bony contours, to determine the nature of the cysts, and to achieve a more accurate differential diagnosis (Figure 1). These examinations confirmed the diagnosis of bilateral dacryocystocele, predominant on the left side, propagating into the nasal cavity, and expanding to the tear ducts, but without any bony barriers. In light of this diagnosis and the risk of complications, we immediately scheduled cyst marsupialization with intranasal endoscopy, under general anesthesia at five days of life. In this context, we used a fine needle and a pediatric set of optics.

The post-operative course was uneventful. Nose washes with saline and the application of intranasal antibiotic drops (a mixture of *bacitracin* and *polymyxin B sulfate*) were prescribed. These measures were associated with regular massages of the internal canthus. The newborn continued to receive cardiopulmonary monitoring after the immediate post-surgical period, for another 24 hours. The newborn was allowed to leave the neonatal intensive care department when nasal congestion had resolved and oral refeeding was well tolerated.

Further examination and endoscopy were performed 10 days post-operation (15 days old) in a follow-up visit. This exam showed positive signs of recovery; the obstructive symptoms had disappeared and intranasal cysts were absent. The child was reviewed one month later and was totally asymptomatic.

Patient 2

This boy exhibited a bluish cystic dilatation of the right medial canthus accompanied by abnormal respiratory sounds and nasal congestion. He also showed discomfort during feeding, but no apparent associated signs of respiratory distress. Nasal tube passage was achievable bilaterally, but it was more difficult on the right side. An examination of the nasal cavity and nasal endoscopy were performed at 24 h of life. The results showed an obstructive polypoid mass in the right lower meatus. An ultrasound and a CT scan of the face were also performed (Figure 2). The results confirmed the liquid nature of this right-side, para-nasal swelling and revealed a cystic formation that extended into the right nasal cavity, continuous with the lacrimal duct, which appeared enlarged compared to the opposite side. No bony obstacle was found along the nasal lacrimal duct. Thus, we diagnosed a right congenital dacryocystocele.



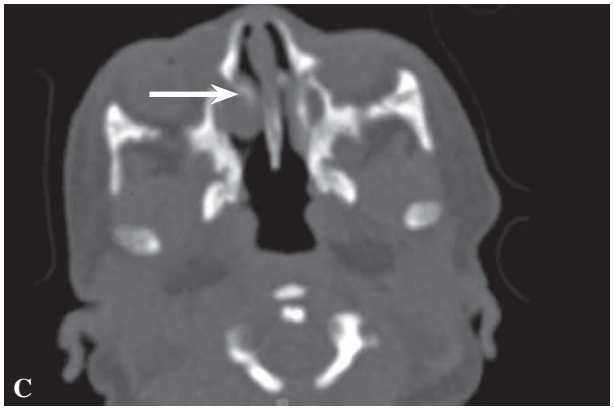


Figure 2
Imaging results for patient 2.
A: The ultrasound examination performed on post natal day 2 confirms the cystic nature of the right para-nasal swelling.
B-C: The CT-scans performed on post-natal day 2 show a cystic formation continuous with the right tear duct, which is enlarged compared to the opposite side (white arrows). The cystic formation extends into the right nostril. There are no bony obstacles between the ocular origin and nasal destination of the lacrimal duct.

Because there were no apparent complications, conservative measures were applied. These included regular massage of the internal canthus and intranasal antibiotic drops (*bacitracin* and *polymyxin B sulfate*) associated with saline nose washes. A follow-up visit at 14 days old showed spontaneous recovery of oral feeding, comfortable nasal breathing, and restored facial symmetry. Endoscopy showed no further evidence of the intranasal cyst. Thus, spontaneous resolution had been achieved. The child was also reviewed one month later, and no recurrence was observed.

Patient 3

In this case, an *in utero* bilateral dacryocystocele was diagnosed in a female fetus during an ultrasound exam of a 25-year-old mother (Gravida 1 Parity 0) with no significant medical history. This exam was performed at 33 weeks gestational age. It showed, on transversal cuts, bilateral cystic para-lateronasal dilatations (Figure 3). These images strongly suggested the presence of a cystic dilation of the lacrimal ducts that had extended into the nasal cavity; thus we diagnosed bilateral dacryocystocele. No other morphological abnormalities were observed. The girl was born at 37 weeks, and she was in good health. She showed no respiratory distress or infectious syndrome of the airway during the neonatal period. She also had no problem feeding. Ophthalmologic and nasal cavity examinations, which included a nasal endoscopy, were performed at 48 hours of life and showed no anomaly. Therefore, we had witnessed a peri-partale spontaneous resolution of a bilateral dacryocystocele. The child was reviewed at 30 days of life and showed good evolution with normal nasal breathing.

Discussion

Pathology

In the normal outflow system, tears drain into the lacrimal *puncta*. Tears flow through the upper and lower *canaliculi*, into a common *canaliculus*, which empties into the lacrimal sac through the Rosenmuller valve. The nasolacrimal duct extends from the sac to its mouth in the inferior nasal meatus through the *ostium* lacrimale, below the inferior turbinate, where it meets the Hasner valve.[4] Dacryocystoceles are believed to develop due to a persistent membrane at the valve of Hasner. This

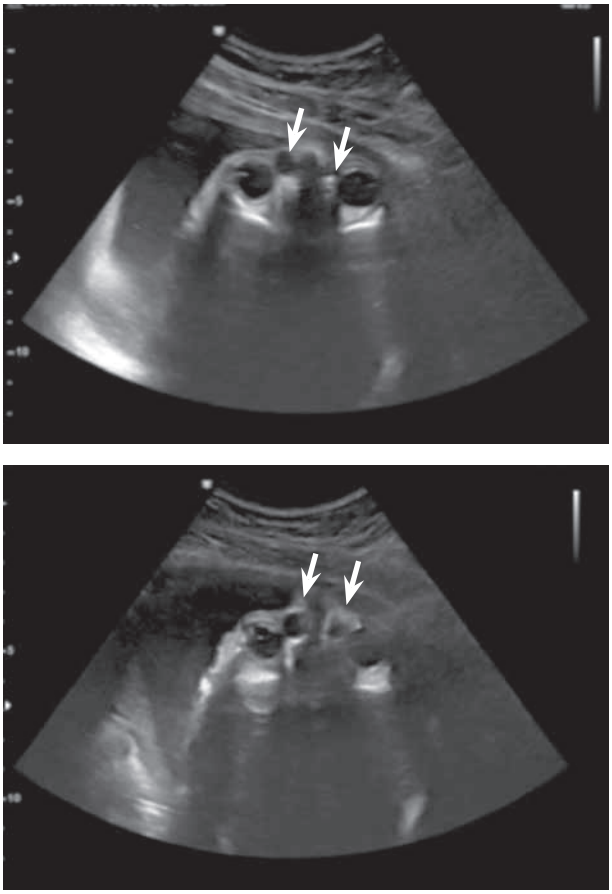


Figure 3
Imaging results for patient 3.
Echographic exam at 33 weeks gestation clearly demonstrates bilateral para- and latero-nasal cyst formation on transversal cuts, which correspond to bilateral dacryocystoceles (white arrows).

blocks passage, which leads to an accumulation of amniotic fluid, mucoid secretions (from the early onset of mucus production by goblet cells), tears, and bacteria. Bacterial colonization also causes distension of the lacrimal system.[2] Fluid accumulation may also result from a functional obstruction, due to secondary changes in the anatomic orientation of the proximal end of the lacrimal sac or the valve of Rosenmüller.[5] This valve acts as a check-valve system.[5] Furthermore, cystic expansion from the nasolacrimal duct mucosa into the nose may result in an intranasal cyst.[6]

The nasolacrimal duct develops from an epithelial cord, which appears at week 6 of gestation. Canalization of this cord starts from the ocular end and progresses to the nasal end by 12 weeks gestation. Canalization of the system is completed by the sixth month of embryonic age.[2,4,7] Failure of canalization occurs most frequently at the lower end of the nasolacrimal duct, at the valve of Hasner. However, canalization may be completed at birth or even later.[3,8] This explains why majority of cases are not diagnosed early in pregnancy, because lesions are not present at 18 to 22 weeks of gestation, when the anatomic survey is routinely performed.[5] Indeed, majority of reported dacryocystocele cases were diagnosed at a gestational age of 30 to 32 weeks.[2,7,8]

Some authors have performed postmortem examinations of infants with dacryocystoceles. They showed that there was no connection between the *canaliculus* and the lacrimal sac.[2] These observations suggested that a proximal obstruction might be formed by remnants, similar to the way a distal obstruction forms. Thus, a dacryocystocele may be a cyst that develops in the early canalization stages of the lacrimal passage.[3] Also, fluid accumulation within the closed lacrimal drainage system may force a perforation in the membrane at the valve of Hasner, which can potentially cause a spontaneous resolution of the dacryocystocele.[1]

In the literature, some reports found that dacryocystoceles occurred with preponderance in Caucasians and females. That finding has been attributed to the genetic trait of narrow nasolacrimal ducts, which suggested a potentially inherited predisposition to nasolacrimal system impotency. However, this theory remains to be proven.[1,5,9,10]

Clinical symptoms

The classic presentation of a dacryocystocele is a bluish, cystic, firm mass located in the medial canthus that appears shortly after birth.[1,7,9] Clinical symptoms appear during the first week of life; these include ocular symptoms, breathing difficulty, and breastfeeding difficulty. It is important for the physician to identify congenital dacryocystoceles, because life-threatening complications can arise. Indeed, patients with bilateral involvement and substantial intranasal extensions may develop a respiratory distress syndrome that may require emergency surgical treatment.[3,4] A dacryocystocele associated with an intranasal cyst located in the inferior meatus area is the rarest, most symptomatic clinical presentation of congenital nasal obstruction. This is, however, the most frequent presentation observed in our otolaryngologic department.[9] The patient may develop dacryocystitis and facial

cellulitis, which may lead to sepsis.[1,3,4] Two studies have shown that most cases of acute pediatric dacryocystitis were caused by α-hemolytic *Streptococcus* and *Staphylococcus epidermidis*, which are among the bacterial flora normally present in the nasal and lacrimal systems.[2,4,7]

Management

Imaging

Ultrasonography, which eliminates the need for sedation, is a rapid, reliable method for the differential diagnosis of dacryocystocele. Additionally, because fluids and debris are visible with ultrasonography, it is possible to examine the content of the lacrimal sac.[2,6,8,11] Alternatively, CT and magnetic resonance imaging (MRI) scans are equally effective for diagnosing cystic lesions. However, these methods are typically employed only when an ultrasonographic diagnosis is inconclusive. Although CT and MRI require sedation, these methods allow a more functional analysis of the lacrimal ducts. An MRI provides excellent characterization of the lacrimal sac content, and a CT scan offers the advantage of a detailed evaluation of the lacrimal bone structure and determination of bone components in lacrimal obstructions. However, CT involves radiation.[2,9,12,13]

A differential diagnosis includes benign and malignant lesions that may cause a mass to form in the medial canthus region, such as encephaloceles, hemangiomas, dermoid cysts, and nasal gliomas.[1,3] Radiological imaging can reveal intranasal cysts, when present.

Nasal endoscopy

Large intranasal cysts can also be visualized with endoscopy. Both nostrils should be examined, with greater attention to the inferior meatus. The application of topical xylometazoline will enhance visibility. It is sometimes necessary to dislocate the inferior turbinate medially.[9]

Treatment

Conservative management of dacryocystocele includes the use of gentle pressure over the lacrimal sac, which facilitates decompression and drainage of the content into the nose. Antibiotic drops may be used prophylactically, before infection occurs.[1,11] After a short course of conservative measures, the resolution rate has been reported to be 76%.[11] However, dacryocystitis can develop within a few days and requires intravenous antibiotics to prevent life-threatening sepsis.[1,11] Crying, coughing, and sneezing can generate sufficient force to relieve an obstruction without the need for surgical intervention.[1,3,11]

In cases of infection or respiratory compromise, careful probing and irrigation of the nasolacrimal system[12] has been shown to ameliorate obstructions effectively; however, marsupialization of the cyst wall is often necessary for those with an intranasal component[1,7,9] (with a scalpel or a microdebrider[10]). Probing can be performed in the consulting room, which eliminates the need for general anesthesia. When dacryocystitis occurs, probing of the nasolacrimal system is often delayed for 24 to 48 h, to allow systemic antibiotic therapy to take effect.[12] When the intranasal component of the cyst is not recognized, probing will fail, unless the probe is passed beyond the wall of the cyst. Marsupialization of an intranasal cyst is always necessary to facilitate the more definitive treatment of removing the cyst wall.[12]

Recurrences can be prevented by probing the lacrimal system with silicone intubation.[3,11,12]

Prenatal diagnosis

It is important to identify congenital dacryocystocele correctly with prepartum ultrasonography. The mother should be referred to an otolaryngologist experienced in the treatment of this problem. Moreover, an awareness that the newborn may experience respiratory distress during first days after birth will enhance the quality of nursing and monitoring.[2,4] The hallmark of a dacryocystocele is the sonographic detection of a cystic mass, medial and inferior to the fetal orbit. The mass appears as a hypoechogenic cystic mass with a clear boundary, located on the inferomedial aspect of the orbit. It is relatively easy to visualize on 2D ultrasound with routine coronal and transverse views of the fetal head.[5] A complete assessment of the facial anatomic structures should be performed with particular attention to orbital views. The clinician should search for any lacrimo-auriculo-dento-digital anomaly.[14] Dacryocystoceles can be associated with other structural anomalies, which would suggest an underlying congenital syndrome. Previous studies have reported associations between dacryocystocele and either Canavan's disease or multicystic/polycystic kidney disease.[2,4,5,15] The ultrasonographic diagnosis becomes increasingly difficult with

advanced gestational age, due to the posture or crowding of the fetal body, and the relatively small prominence of the dacryocystocele compared to the enlarging fetal head.[5] Magnetic resonance imaging may be indicated when the ultrasonographic diagnosis is not conclusive, particularly in rare cases, where another malformation is suspected, such as meningocele.[13]

Studies have shown that a prenatally detected dacryocystocele has a 76% chance of resolving spontaneously during the intrauterine period or during birth.[9] The mass may be mechanically decompressed and the Hasner membrane may be ruptured during vaginal delivery.[9] Some authors hypothesize that dacryocystocele formation occurs during the natural course of canalization in nasolacrimal duct development.[1,2,5,11]

Conclusion

This study described three different patients with congenital dacryocystocele, which is a rare, frequently benign entity. Nasal examination in an otolaryngologist's consulting room should be performed in all cases to exclude the coexistence of a nasal cyst. Most clinicians recommend early surgical intervention in cases of respiratory compromise, dacryocystitis, cellulitis, large dacryocystoceles that cause an ophthalmological anomaly, or recurrent dacryocystoceles. Also, surgery is recommended for cases that fail to resolve with conservative measures.[1,9,11,12] Except in cases that involve complicated symptoms, we recommend conservative treatment for dacryocystoceles, because there is a high probability of spontaneous resolution within about two weeks.[1,2,9] However, strict attention should be paid to follow-up exams, because previous studies have shown that dacryocystitis that develops from a dacryocystocele could be more dangerous than congenital nasolacrimal duct obstruction. Therefore, efficient use of antibiotics during the early stage is recommended.[11] A one-year follow-up is also strongly recommended.[9]

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