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Prevalence and clinical characteristics of congenital dacryocystocele

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Abstract

Purpose—To report the prevalence, clinical findings, and outcomes in children diagnosed with congenital dacryocystocele in a well-defined population over a 20-year period.

Methods—The medical records of all Olmsted County, Minnesota, patients diagnosed with congenital dacryocystocele from January 1, 1988, through December 31, 2007, were retrospectively reviewed.

Results—A total of 9 children were diagnosed with dacryocystocele during the 20-year period, yielding an incidence of 1 in 3884 live births. The median age at diagnosis was 12 days (range, birth to 40 days); 7 (78%) were female. Eight patients (89%) had unilateral disease. Clinical findings included a cystic mass in all 9, dacryocystitis in 3 (33%), intranasal cysts in 3 (33%), and 1 (11%) each with facial cellulitis and dacryocystocele-induced astigmatism. Conservative treatment resolved the obstruction in 3 (33%), while the remaining 6 (67%) each underwent one surgery. Complete resolution was observed in all 9 patients.

Conclusions—Congenital dacryocystocele is an uncommon unilateral condition of predominantly neonatal females. Clinical findings support a relatively high incidence of complications such as dacryocystitis and intranasal cysts as well as a less common finding of astigmatism that resolved without evidence of amblyopia. One-third of the cases were successfully managed with conservative treatment alone; most patients required surgical intervention.

Congenital dacryocystoceles, a variant of nasolacrimal duct obstruction, are characterized by the appearance of a cystic blue mass over the region of the lacrimal sac soon after birth.¹ Dacryocystoceles are thought to be the result of a persistent membrane at the level of the valve of Hasner and a functional obstruction of the common canaliculus or valve of Rosenmuller.² The resulting lacrimal sac distention has been reported to be more common in females^{3–7} and non-Hispanic whites,³ while familial cases have been described only sporadically.³ Common presenting signs include dacryocystitis, facial cellulitis and

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respiratory distress,⁴ while the development of astigmatism has been observed only rarely.^{1,3} Controversy exists regarding to the optimal management of dacryocystoceles: some authors advocate conservative medical management,^{1,4} whereas others recommend early surgical intervention, such as nasolacrimal duct probing or nasal cyst marsupialization.^{5,8}

The prevalence of congenital dacryoceles, to the best of our knowledge, has not been reported in any population. The purpose of this study is to describe the prevalence, clinical findings, and outcomes among children diagnosed with congenital dacryoceles while residing in Olmsted County, Minnesota, over a 20-year period.

Subjects and Methods

The medical records of all patients younger than 5 years of age who were residents of Olmsted County, Minnesota, when diagnosed with a dacryocystocele between January 1, 1988, and December 31, 2007, were retrospectively reviewed. Institutional review board approval at Mayo Clinic and Olmsted Medical Group was obtained for this study. The resources of the Rochester Epidemiology Project, a medical records-linkage system documenting medical care received in Olmsted County, Minnesota, were used to identify cases of dacryocystocele.⁹ Virtually all medical care is provided to the residents of Olmsted County by Mayo Clinic, Olmsted Medical Group, and affiliated hospitals. Patients who were not residents of Olmsted County at the time of their diagnosis were excluded from the study. Patients were identified by computerized diagnosis code retrieval methods. We reviewed the gender, age at presentation, common presenting signs and symptoms, treatment modalities, and final outcome.

To determine the incidence of dacryocystocele in Olmsted County, annual age- and sex-specific incidence rates were constructed using the age- and sex-specific population figures for this county from the U.S. Census. Because dacryocystocele is typically diagnosed within the first year of life, a birth prevalence was calculated from the number of births occurring from January 1, 1988, through December 31, 2007, using the annual birth incidence for this county.

Results

A total of 9 children were diagnosed with congenital dacryocystocele in Olmsted County, Minnesota, during the 20-year study period, corresponding to a birth prevalence of 1 in 3,884 live births. Pertinent historical and initial clinical characteristics of the 9 study patients are shown in Table 1. Of the 9 dacryocystocele cases, 8 (89%) were unilateral. The left nasolacrimal system was affected in 5 cases (56%), the right in 3 cases (33%), and both in 1 (11%). There were 7 girls and 2 boys with the following ethnicities: 8 non-Hispanic whites (89%), and 1 Hispanic (11%), diagnosed at a median age of 12 days (range, birth to 40 days). Six of the 9 patients were the result of full-term pregnancies, 2 were born prematurely, and the final patient's pregnancy was complicated by gestational diabetes and an elevated core temperature at the time of delivery. None of the patients had a family history of dacryocystocele. Clinical findings included a cystic mass in all 9 (100%), epiphora in 4 (44%), dacryocystitis in 3 (33%), intranasal cysts in 3 (33%), 1 (11%) with facial cellulitis, and 1 (11%) with dacryocystocele-induced astigmatism. One patient had a right dacryocystocele and a left congenital nasolacrimal duct obstruction. There were no associated medical conditions other than prominent hypertelorism in a patient with bilateral nasal cysts and dacryocystocele-induced astigmatism. The patient's face was slightly dysmorphic, with wide-set eyes, down-slanted palpebral fissures, and at the time of examination, periorbital edema. Evaluations for chromosomal abnormalities, Smith-Lemli-Opitz syndrome, and 22q deletion syndrome were normal.

The 9 study patients were followed for a mean of 20 months (range, 7 days to 8 years). Of the 9, 3 (33%) were successfully managed with external massage combined with either topical or systemic antibiotics. Massage and topical antibiotics resolved the dacryocystocele within one week in 2 of the 3 patients, while the third developed dacryocystitis that responded successfully to systemic antibiotics within 6 days. There was no association between outcome and the type of antibiotic used. The remaining 6 patients required nasolacrimal duct probing and irrigation following a trial of conservative treatment lasting an average of 12 days (range, 2 to 33 days). Two of the 3 patients with intranasal cysts underwent marsupialization of the nasal cysts along with nasolacrimal duct probing, while the third patient was successfully managed with duct probing alone. Two (22%) patients developed dacryocystitis while receiving conservative therapy, including 1 patient with bilateral cysts who also developed periorbital cellulitis. This patient required probing and irrigation as well as marsupialization of their nasal cysts, while the second patient who developed dacryocystitis was one of the 3 patients who were successfully treated with antibiotics alone. Complete resolution was observed in all 9 patients and no recurrence was noted upon follow-up.

Discussion

Congenital dacryocystocele was diagnosed in 1 in 3,884 births in this population. The majority of patients were female with unilateral disease who presented within the first two weeks of life. Associated conditions such as dacryocystitis or nasal cysts occurred in 56%, while two-thirds required surgical intervention to resolve the obstruction.

Congenital dacryocystoceles are an uncommon variant of nasolacrimal duct obstruction, typically seen at birth or shortly after birth as a blue-colored cystic mass over the lacrimal sac. The valve of Hasner is the most frequent site of a distal obstruction due to incomplete canalization, a process that starts in the third fetal month and is completed by the sixth month.⁵ An incidence of 35% to 73% has been reported for imperforate nasolacrimal ducts in term infants, with the majority opening spontaneously during the first few weeks of life.^{10,11} A congenital dacryocystocele accompanies nasolacrimal duct obstruction in approximately 0.1% of infants.¹²

The clinical characteristics of the study patients were consistent with prior reports. Unilateral involvement has been reported to range from 60%¹³ to 100%,^{1,6,8,14} similar to the 89% reported in this cohort. Likewise, a large proportion of the study patients (89%) were non-Hispanic whites, as has been reported in prior studies (83%³ to 100%⁶). Three-fourths of the study patients were female, consistent with prior reports ranging from 20%¹³ to 100%,¹ with the majority being above 57%.³⁻⁷ The female preponderance is thought to be associated with young girls having a narrower nasolacrimal duct than boys.¹⁵

Secondary infection is common in nasolacrimal duct obstruction due to the stasis of lacrimal sac contents, proximity of the sinuses, and an abundance of lymphatic and vascular systems within the submucosa of the lacrimal sac.³ Dacryocystitis was a presenting sign in 3 (33%) of the study patients, including one patient who also had facial cellulitis. During the course of conservative management of their dacryocystoceles, 2 (22%) additional patients developed dacryocystitis or both dacryocystitis and cellulitis. This finding is similar to the high rate of infection demonstrated in prior studies, which ranges from 18%⁴ to 43%³ for dacryocystitis alone, and 20%⁵ to 38%⁸ for both dacryocystitis and cellulitis on initial presentation. Fittingly, studies with higher rates of dacryocystitis describe higher rates of surgical intervention and vice versa.⁸ Wong and VanderVeen⁷ reported that 28 of 42 patients (62%) presented with cellulitis or dacryocystitis and 36 of 46 affected eyes (78%) required surgical intervention. However, Schnall and Christian,⁴ reporting a prevalence of

only 18% with dacryocystitis on initial presentation, found that, of 21 dacryocystoceles, 16 (76%) resolved with conservative therapy. The development of dacryocystitis during the course of medical management for dacryocystocele has been reported to range up to 25%.^{1,4}

An intranasal mass located in the anterior inferior meatus, arising from the nasolacrimal duct was found in 3 patients (33%). Mansour and colleagues³ described 6 of 54 cases (11%) with nasal cysts that communicated with the lacrimal sac cysts, adding that there were likely 7 (13%) additional cases with nasal cysts that could not be visualized. Shashy and coinvestigators⁵ identified intranasal cysts in 7 of 10 patients (70%), and Cavazza¹⁴ found 1 in 5 cases (20%). Thomas and colleagues¹⁶ reported intranasal cysts in 6 of 8 patients (75%). Since neonates are obligate nasal breathers, a nasal obstruction in the form of an intranasal cyst can cause significant respiratory distress.

Complications due to the mass effect of a dacryocystocele can temporarily result in the development of corneal astigmatism. Anisometropic amblyopia,^{1,17} however, is unlikely due to the characteristically successful response to early treatment and resolution within the first several months of life.¹⁸ A case series by Harris and DiClementi¹ described a patient with a substantial degree of lacrimal sac distention and corneal astigmatism and anisometropia that resolved following nasolacrimal duct probing and silicone intubation. Mansour and coauthors³ also reported cycloplegic refraction results in 10 of 54 patients, reporting that 6 (60%) had unilateral astigmatism between 2 and 3 D, and 1 patient (10%) had 4 D of astigmatism that resolved 10 days following conclusion of the dacryocystocele. Similarly, in the present study 1 patient (11%) had hypertelorism and dacryocystocele-induced astigmatism that resolved without evidence of amblyopia following nasolacrimal duct probing, irrigation and marsupialization of bilateral nasal cysts.

Due to the variability in the natural course of dacryocystoceles, controversy exists regarding the most appropriate management of this condition. Harris and DiClementi¹ reported that 3 of 4 cases (75%) resolved with medical management, and in a prospective study by Schnall and Christian,⁴ of 21 noninfected dacryocystoceles, 16 (76%) resolved within 6 days with medical management alone, with the remaining 5 (24%) requiring probing following 2 weeks of medical treatment. Wong and VanderVeen⁷ described 10 of 46 cases (22%) that resolved with medical management, with the remaining 36 (78%) requiring surgical intervention and Paysee and colleagues¹⁹ found that the majority (97%) of dacryocystoceles in their study were surgically managed with all but 2 procedures being successful. Similarly, the majority of patients in the current study required surgery after failing to resolve with conservative management alone. In all 9 cases, conservative treatment was initially employed. Three cases (33%) resolved while the remaining 6 required surgical intervention after an average of 12 days (range, 2 to 33 days) of medical management. Dacryocystocele recurrence has also been reported in the literature although none was demonstrated upon follow-up in the present study. Mansour and colleagues³ found recurrence in 22%, up to 6 months following nasolacrimal duct probing, and Harris and DiClementi¹ described recurrence in 1 case (25%). Most investigators recommend that uncomplicated dacryocystoceles, those that are noninfected and lacking intranasal cysts, should be managed conservatively; otherwise, a combined approach should be performed, employing massage, warm compresses, antibiotics and surgical intervention as needed.⁵

There are several limitations to the findings in this study. The retrospective design is limited by nonstandardized data and incomplete follow-up. Furthermore, as a rare condition, it is possible that small dacryocystoceles may go undetected by a pediatrician, particularly since neonates are often bruised or have swelling following delivery, resulting in an underestimation of the true incidence. Although a relatively isolated county, it is also possible that some Olmsted County residents with dacryocystoceles may have sought care

outside the region, potentially further underestimating the incidence of disease in this population. Finally, due to the demographic limitations of Olmsted County, the results of this study are best extrapolated to other relatively homogeneous semiurban non-Hispanic white populations.

This study provides population-based incidence rates for congenital dacryocystoceles diagnosed over a 20-year period. Dacryocystoceles were diagnosed in 1 in 3,884 births in this midwestern population of the United States. Clinical findings were consistent with prior reports of dacryocystocele, supporting a high incidence of complications such as dacryocystitis or intranasal cysts. Uncomplicated dacryocystoceles may respond to medical management alone, as in one-third of the cases. However, consistent with the majority of reports, most patients required surgical intervention.

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Table 1

Historical and initial clinical characteristics of 9 children diagnosed with dacryocystocele in Olmsted County, Minnesota, 1988 to 2007.

Factor	Number of patients
Female gender (%)	7 (78)
Premature birth [*] (%)	2 (22)
Median age at diagnosis in days (range)	12 (0 – 40)
Laterality (%)	Right, 3 (33) Left, 5 (56) Bilateral, 1 (11)
Ocular Findings (%)	
Mass	9 (100)
Epiphora	4 (44)
Dacryocystitis ⁺	3 (33)
Intranasal cysts ⁻	3 (33)
Astigmatism	1 (11)
Management (%)	
Massage and/or topical or systemic antibiotics	3 (33)
Surgical intervention	6 (67)
Probing/irrigation	3 (33)
Probing/irrigation and marsupialization of cyst or infraction of inferior nasal turbinate	3 (33)

^{*} <37 weeks gestational age

⁺ Including one patient with facial cellulitis

⁻ Including one patient with bilateral cysts