

# Congenital Lacrimal Sac Fistula: A Case Report and Review

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## GOAL

To discuss a case of congenital lacrimal sac fistula

## OBJECTIVES

Upon completion of this activity, dermatologists and general practitioners should be able to:

1. Describe the epidemiology and embryologic development of a congenital lacrimal sac fistula.
2. Outline the clinical features of a lacrimal sac fistula.
3. Delineate complications and surgical management of a lacrimal sac fistula.

**CME** Test on page 130.

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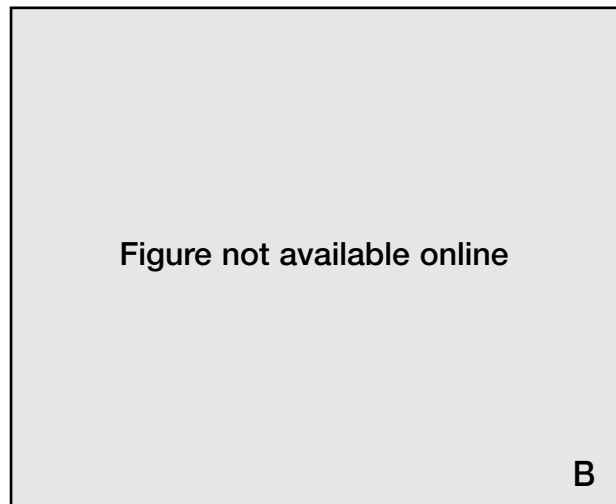
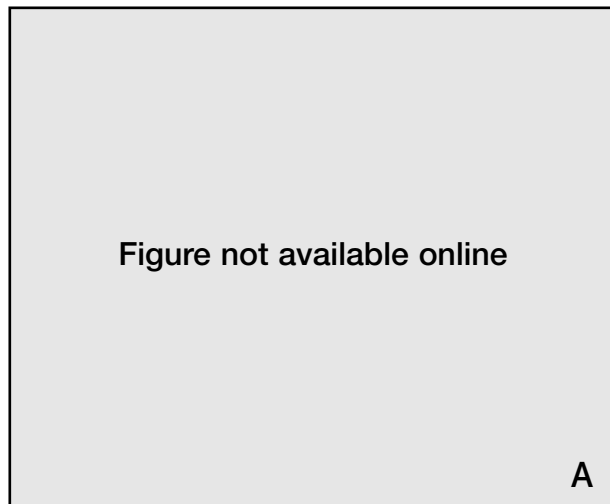
*The lacrimal system is crucial for vision and comprises the lacrimal glands, which produce tears, and the lacrimal drainage system, which drains tears away from the eyes. While various disease processes affect the lacrimal glands, the lacrimal drainage system is commonly prone to congenital abnormalities such as lacrimal sac fistula. We describe a case with classic findings and review dermatologic manifestations of this entity.*

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**T**he lacrimal system is essential for vision because continuous tear production maintains the integrity of the cornea, the refracting surface of the eye. Decreased tear production can occur when viral and bacterial pathogens enter the lacrimal gland, resulting in acute infection and many other disease processes that inhibit or destroy the glands. Proper function of the lacrimal glands depends on a patent lacrimal drainage system. Tears produced by the lacrimal glands bathe the surface of the eye and are discharged through the lacrimal drainage system into the nose. Fistulae of the lacrimal drainage system have been described in the ophthalmology literature,<sup>1</sup> but many dermatologists are unaware of this finding. We describe a case with classic clinical findings and discuss this entity because timely recognition of this abnormality is important for maintaining the integrity of the eye.



Congenital lacrimal sac fistula. Bilateral raised orifices inferior to the medial canthal angle (A). Close-up view of flesh-colored and asymptomatic fistula's ostium (B).

### Case Report

A 10-month-old boy was referred to dermatology for evaluation of "2 holes by his eyes." The child's mother stated that the holes had been there since birth and that they had been increasing in size as the baby grew. She denied tears, pus, or blood drainage from these fistulae. The infant was otherwise healthy and did not have a history of eye infections. He was born full term of an uncomplicated pregnancy; family history was negative for ocular disease. On physical examination, 2 small raised orifices measuring 1.5 mm in width bilaterally inferior to the medial canthal fold of each eye were seen (Figure). Crying did not elicit any drainage from these sites. Consultation with an ophthalmologist revealed that the visual examination was within normal limits and concurred with our diagnosis.

### Comment

Lacrimal sac fistula was first reported<sup>2</sup> in 1675. Since then, many cases have been described in the ophthalmology literature,<sup>1</sup> but none have appeared in the dermatology literature. The incidence of congenital lacrimal sac fistula is estimated to be 1 in 2000 births and occasionally is familial, appearing to have autosomal dominant inheritance. There does not appear to be a sex or race predilection. Generally, no other congenital anomalies are observed, although an association has been described with preauricular fistulae.<sup>3</sup>

Tears released from the lacrimal glands bathe the eye and are caught by the lid margins, where they enter the lacrimal drainage system at the punctum. Each punctum drains into a canaliculus, which then forms a common canaliculus that empties into the lacrimal sac. The lacrimal sac drains into the con-

cha of the nose by way of the nasolacrimal duct. The embryogenesis<sup>3</sup> of the nasolacrimal system begins as a thickening of ectoderm in the naso-optic fissure in the 32-day-old embryo. This becomes buried in the mesenchyme between the lateral nasal and maxillary process in the 42-day-old embryo. Caudal and cephalic extension of the cord occurs, producing the nasolacrimal duct and canaliculus. Canalization begins in the 60-day-old embryo. The puncta are patent at the seventh fetal month, while the distal nasolacrimal duct is patent in less than 30% of infants at birth.

Congenital lacrimal sac fistula is a developmental condition in which a fistula connects the skin to the common canaliculus or the lacrimal sac. Alternatively, the lacrimal fistula may be incomplete, ending blindly in the subcutaneous tissue near the lacrimal sac.<sup>4</sup> Examination of a lacrimal sac fistula reveals that it is generally lined with stratified squamous epithelium, similar to that of a normal canaliculus.<sup>4</sup> The pathogenesis of congenital lacrimal sac fistula is uncertain.

A congenital lacrimal sac fistula is noticed shortly after birth and is usually asymptomatic and nonprogressive. It can easily be overlooked—especially if the patient is asymptomatic—because it is small and flesh colored. Most fistulae are unilateral but can be bilateral,<sup>5</sup> as in this patient. The location is usually inferior to the medial canthal angle. Although it is true that most fistulae are asymptomatic, some patients present with clear mucoid fluid at the fistula's ostium, or fluid may be expressed by placing pressure on the sac, causing reflux. The 2 main complications are chronic local eczema due to maceration and chronic or acute dacryocystitis (infection of the nasolacrimal duct) due to ascending infection.<sup>5</sup>

The nasolacrimal system is usually patent,<sup>3</sup> but significant complications may occur when it becomes obstructed. Congenital obstruction of the nasolacrimal duct is the most common abnormality of the entire lacrimal system, occurring in up to 6% of neonates. A sign of obstruction is epiphora (tears overflowing without stimulus). There also can be reflux of lacrimal sac material, which can appear mucoid, mucopurulent, or purulent. In more severe cases of ascending infection, there may be conjunctival injection and erythema of the skin near the puncta and lid margins.

The treatment of choice for a symptomatic fistula is surgery, consisting of complete excision of the fistula, sometimes in conjunction with nasolacrimal intubation if there is associated distal nasolacrimal duct obstruction.<sup>1,3,5,6</sup> Before surgery on a congenital lacrimal sac fistula, it is necessary to demonstrate adequate drainage from the lacrimal sac through a patent nasolacrimal duct. This can be done by dye disappearance and irrigation testing or a dacryocystogram. Our patient remains asymptomatic after 1 year and

continues to do well, without intervention therapy. In summary, dermatologists should be made aware of this developmental abnormality because early recognition and referral for appropriate treatment can prevent unnecessary discomfort and infections that could result in the permanent loss of vision.

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