

Congenital lacrimal fistula

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PURPOSE. *Congenital lacrimal fistula is a rare abnormality of the lacrimal system. The patients may be asymptomatic but treatment of those with symptoms such as epiphora and dacryocystitis is still controversial.*

METHODS. *The clinical findings, methods of surgical treatment and results were evaluated in seven patients (four male, three female) with congenital lacrimal fistula. Three were asymptomatic, types of surgical treatment were fistula excision with dacryocystorhinostomy (DCR) (two cases), fistula excision with conjunctival DCR (one case) and closed fistula excision alone (one case).*

RESULTS. *Three of the four patients who underwent surgery had relief of symptoms. The fistula recurred in the case that had undergone fistula excision alone.*

CONCLUSIONS. *It is difficult to design controlled studies to detect which method is superior for the treatment of lacrimal fistula, considering the small number of cases in the literature. We recommend treating each case according to its own characteristics. (Eur J Ophthalmol 2000; 10: 22-6)*

KEY WORDS: *Congenital, Lacrimal fistula, Surgical treatment*

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INTRODUCTION

Congenital lacrimal fistula is a rare developmental abnormality first reported by Rasor and later by Von Amon (1, 2). Congenital lacrimal fistula is estimated to occur in one in 2000 births (3). Family history is positive occasionally and autosomal dominant inheritance has been shown (4). The fistula, surrounded by epithelium, is commonly located between the skin and common canaliculi and lacrimal sac. In some cases the orifice of the fistula ends in the subcutaneous tissue near the sac. Most cases are unilateral and are located inferonasally to the medial canthus.

These fistulas are usually asymptomatic at birth and may be overlooked for some time. Their smallness and the lack of contrasting skin pigmentation make it easy for them to remain unnoticed. Mucoid secretion can be detected by pressing over the lacrimal sac in some cases. There may be epiphora from the eye, secretion from the fistula or recurrent dacryocystitis

in symptomatic cases.

We present the first series of patients with congenital lacrimal fistula in Turkey. The clinical findings, method of surgical treatment and results are described.

MATERIALS AND METHODS

Congenital lacrimal fistula was detected in seven patients referred to the lacrimal unit between 1990 and 1998. All patients underwent a routine ophthalmologic examination. Syringing from the punctum and dacryocystography were done in four symptomatic cases. Two had canalicular obstruction (cases 2 and 4), and one had nasolacrimal duct obstruction (case 7). The nasolacrimal passage was patent in case 5. Patients who were going to be operated were referred to the ENT clinic for nose examination to check that the nasal passages were suitable for dacryocystorhinostomy.

Surgery was done under general anesthesia in three cases and local anesthesia in one. The mode of treatment was fistula excision with dacryocystorhinostomy in the three cases with nasolacrimal duct obstruction and simple fistula excision (closed fistula excision) in the case with patent nasolacrimal canal.

A dacryocystorhinostomy incision was made vertically 11 mm medially to the medial canthus. The lacrimal sac and common canaliculi were reached after the skin and subcutaneous incisions by severing the anterior part of the medial canthal tendon. A Bowman number 0 probe was inserted from the fistula and passed through the lacrimal sac to establish the origin of the lacrimal fistula. The periosteum was separated from the lacrimal bone and an 8 x 8 (in children) or 10 x 10 mm (in the 30 year-old case) osteotomy including the anterior lacrimal crest was opened with a striker. The medial wall of the lacrimal sac was incised to form anterior and posterior flaps. Anterior and posterior nasal mucosal flaps were also formed by a horizontal incision opposite those in the lacrimal sac. The posterior flaps were sutured to each other. The fistula tract was then dissected from the surrounding tissues under the guidance of the lacrimal probe within the tract. The origin of the fistula that was connected to the lacrimal sac was closed with a 6/0 Vicryl suture and the stump was excised. In two cases the fistula was opened to the common canaliculi; one of them had a bicanalicular silicone intubation (case 4). After dilatation of both puncta a silicone tube was passed through the lacrimal sac and its ends were tied together in the nose cavity. Then the anterior flaps were anastomosed. Subcutaneous and cutaneous tissues were closed in the usual fashion.

Conjunctival DCR was performed in the other case whose lacrimal fistula originated from the common canaliculi (case 2). After anastomosis of the posterior flaps, the lacrimal sac was entered from the caruncle with a cataract knife. The new passage was dilated with gold dilator probes. A Jones tube was placed in the passage and fixed with a 5/0 silk suture from its neck to the skin in the medial canthal region. Then the anterior flaps were sutured together. Subcutaneous and cutaneous tissues were closed as usual.

Closed fistula excision was done in one case (case 5). The fistula tract was dissected entirely by an incision surrounding the fistula and excised out. The root of the fistula in the lacrimal sac was sutured with a 6/0 Vicryl. After closure of subcutaneous and cu-

taneous tissues, probing and irrigation were done to ensure the nasolacrimal passage was patent.

RESULTS

There were three female and four male patients. The fistula was present on the right eye in five cases and on the left in two. The families had discovered it in the first month to four years after birth in the four cases that were operated. These patients were between 6 and 30 years old (mean 13.7) at the time of surgery. We had three asymptomatic cases whose fistulas had been detected at 7 months, 6 years and 9 years. There was no family history. No systemic abnormality was found in any case.

The orifice of the fistula was inferonasal to the medial canthus in all cases (Fig. 1). The origin of the fistula was the lacrimal sac in two cases and the common canaliculi in two cases. It could not be detected in the three asymptomatic cases that were not operated.

The symptoms were chronic dacryocystitis in two cases and epiphora in two. The epiphora also occurred from the fistula region (Fig. 2). One patient also had chronic dacryocystitis in the other eye (case 7). The clinical features and type of surgical treatment are shown in Table I.

The follow-up period is 4-36 months (mean 20 months) for the four cases operated. The symptoms were cured in two cases that had DCR with fistula excision and in one that had conjunctival DCR with fistula excision (Fig. 3). The tube extruded two weeks after the operation in the case that had silicone intubation together with DCR. The epiphora recurred from the fistula region four months after the operation in the case that had closed fistula excision alone. The parents refused further treatment.

DISCUSSION

During the sixth week of embryological development, neuroectodermal cells, which are in the naso-optic groove between the lateral nasal and maxillary processes, form a solid epithelial cord. After canalization, the upper part of the cord forms the canaliculi and the lower part forms the nasolacrimal canal (5). In the middle part forming the lacrimal sac, the

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Fig. 1 - The congenital lacrimal fistula at the inferonasal part of the medial canthus (arrow).



Fig. 2 - Epiphora from the congenital lacrimal fistula (arrow).

epithelial cords are thicker, and therefore when the sac canalizes it has a larger diameter than the canaliculi or the nasolacrimal canal.

The pathogenesis of congenital lacrimal sac fistula is uncertain. An overdevelopment of the lacrimal duct, incomplete closure of the embryonic facial fissure, an abnormality of amniotic bands, and a primary developmental arrest may cause the fistula (6). Some authors suggest it is analogous to fistula formation elsewhere in the body and that the surface ectoderm failed to fuse fully after invagination (7). Welham et al (8) proposed that the fistula is really an extra canaliculus extending from the common canaliculus to the skin in the medial canthal area. They observed a large number of canalicular abnormalities in their patients, with stratified squamous epithelium lin-



Fig. 3 - Postoperative appearance of the patients three years after conjunctival DCR. The fistula is closed and Jones tube is in place.

TABLE I - CLINICAL FINDINGS AND SURGICAL TREATMENT OF THE PATIENTS WITH CONGENITAL LACRIMAL FISTULA

Case No.	Age*	Sex	Eye	Punctum lavage	Origin of fistula	Surgical treatment
1	7 mo	Male	Left	-	Unknown	-
2	6 y	Male	Right	Obstructed	Common canaliculi	Fistula excision + CDCR
3	9 y	Female	Right	-	Unknown	-
4	11 y	Male	Right	Obstructed	Common canaliculi	Fistula excision + DCR+ silicone intubation
5	8 y	Male	Left	Open	Lacrimal sac	Fistula excision
6	6 y	Female	Right	-	Unknown	-
7	30 y	Female	Right	Obstructed	Lacrimal sac	Fistula excision + DCR

* Age at first visit to our clinic. DCR: Dacryocystorhinostomy, CDCR: Conjunctival dacryocystorhinostomy

ing the fistula tract, which support the hypothesis. Sullivan et al. (9) believed that a defect interfering with the invagination, burial, and later tissue remodeling of the surface ectoderm cord was responsible.

There is no agreement on the treatment of congenital lacrimal fistula. Cauterization and simple skin closure are historical methods, but have not been successful. The closed excision in which only the fistula is repaired (9), dacryocystorhinostomy with fistula excision (8), and fistula excision with nasolacrimal intubation (9, 10) are reported to be successful.

The authors suggesting fistula excision with dacryocystorhinostomy stated that opening the lacrimal sac facilitated surgical dissection and accurate ligation and removal of the fistula from its origin, minimizing the risk of damage to the common canaliculus (8). Those who favor a closed incision sustain that a properly performed closed incision is safe and effective (9, 10). With a generous elliptical skin incision anatomical landmarks can be identified and there is minimal damage to the common canaliculus for the experienced lacrimal surgeon (9). It is agreed that if there are associated nasolacrimal drainage abnormalities, intubation of the lacrimal system should also be performed (8-10).

We had success in two cases that had DCR with fistula excision. The case operated by closed fistula excision had a recurrence after four months. We performed conjunctival DCR in one case with an obstruction at the level of the common canaliculi. We have not encountered this method in previous series. Conjunctival DCR is useful to relieve the proximal nasolacrimal obstruction but acceptance of the Jones tube is a problem in children.

Our limited experience prevents us commenting on

one of the main treatment types. Fistula excision with DCR provides a wide passage, which may avoid the consequences of possible damage to the lower lacrimal system during the operation. On the other hand, DCR may be unnecessary with a delicate surgical approach. Silicone intubation is needed when repairing a fistula involving the common canaliculi.

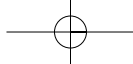
The incidence of congenital lacrimal fistula is reported to be 1 in 2000 births (3), but the limited number of reports suggests an even lower incidence. Cases may easily be overlooked because so many are asymptomatic, as in 42.8% (3/7) of our series. No sex nor race predominance has been reported (10). Some congenital systemic abnormalities were reported to be associated with lacrimal fistula, the most common being preauricular fistula (11). Down's syndrome, meningocele, facial clefts, and umbilical hernia were other abnormalities associated with lacrimal fistula (9, 12). A generalized defect in the ability of the surface ectoderm to close fully after invagination was suggested (9).

The closed excision and fistula excision with DCR are both successful methods for the treatment of congenital lacrimal fistula. With such limited numbers of cases it is difficult to design controlled trials to establish which method is superior. We suggest evaluating each case individually and choosing the method of treatment in the light of the main characteristics.

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