Characterising congenital double punctum anomalies: Clinical, endoscopic and imaging findings

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Running Head: Epiphora with coexistent duplex punctum

Precis: Double puncta co-exist with a range of other nasolacrimal pathologies. OCT suggests that the medial of the two double puncta is the abnormal one, with DCG and endoscopy demonstrating a range of accessory canaliculus lengths.

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Abbreviations:

OCT – optical coherence tomography

FDDT - fluorescein dye disappearance test

NLDO – nasolacrimal duct obstruction

DCG – dacryocystography

DCR – dacryocystorhinostomy

Keywords: Lacrimal punctum, epiphora, optical coherence tomography**,** malformation

ABSTRACT

Purpose: To characterise the anatomical anomaly of congenital double puncta, identify factors contributing to epiphora and its treatment.

Methods: Retrospective review of patients presenting with epiphora and a double lacrimal punctum over a 6-month period.

Results: 5 consecutive patients (3 female) were identified. The median age was 50 years (range 34-76).

Investigations included punctum Optical Coherence Tomography (OCT), Dacryocystography and canalicular endoscopy. The medial punctum was more morphologically abnormal when compared to healthy puncta, with OCT showing no vertical canalicular component in 80% (4/5) of patients; and a more superficial than usual horizontal canaliculus in 80% (4/5). Dacryocystography and canalicular endoscopy showed the junction of the two inferior canaliculi, which was very proximal in one, mid canalicular in 2 and at the level of the common canaliculus in 2.

In one patient, the only abnormality identified was the double lower punctum, with a high tear meniscus and delayed fluorescence dye disappearance test. They underwent endonasal Dacryocystorhinostomy with improvement of epiphora.

In the other four patients, causes of epiphora were: Nasolacrimal duct stenosis with mucocoele, punctal stenosis, common canalicular stenosis with nasolacrimal duct stenosis, upper canaliculitis, and blepharitis. Their treatments included endonasal Dacryocystorhinostomy, punctoplasty, canaliculotomy and blepharitis treatment. All had improvement of epiphora following treatment.

Conclusion: These findings suggest that the underlying canalicular anomalies are varied and that the clinical manifestation is also heterogenous. OCT imaging suggests that the medial of the two puncta is the more morphologically abnormal one, with Dacryocystography and canalicular endoscopy demonstrating different accessory

canaliculi lengths.

INTRODUCTION

A double punctum is a rare congenital abnormality of the lacrimal drainage system. These patients can present with epiphora later in adult life and the underlying mechanism remains undefined.

This series aims to characterize the double punctum using novel Optical Coherence Tomography (OCT) imaging and endoscopy techniques in order to better understand the contributing factors to epiphora and guide management.

MATERIALS AND METHODS

Subjects and ethics

5 consecutive patients presenting to the Lacrimal Clinic in Moorfields Eye Hospital with epiphora and a double punctum were included. Application to access the previously collected data was made to, and granted by the Moorfields NHS Trust Audit committee (Ref no CA17/AD/01). This study adhered to the tenets of the Declaration of Helsinki.

Epiphora score

Patients were asked to grade the degree of epiphora on a typical day in each eye, using the Munk scoring system -- with a Munk score of “0” for no epiphora, “1” for epiphora requiring dabbing with a tissue less than twice a day, “2” for dabbing 2-4 times daily, “3” for dabbing 5-10 times daily, and “4” for epiphora requiring dabbing more than ten times a day.[(1)]

OCT of the lacrimal punctum

OCT of the lacrimal puncta was performed using the Heidelberg Spectralis with the anterior segment module and has been described previously by the authors.([2-4])

Dacryocystography

Dacryocystography (DCG) is performed using a 24G Rabinov Sialography Set filled with Lipiodol Ultra fluid. The canaliculus is canulated to the mid canaliculus. The tubing is taped to the cheek. Video fluoroscopy is performed as the doctor introduces the dye until the point when the dye has reached the nasal space or significant reflux is observed. The procedure is repeated through the second lower lid punctum. An erect x-ray is then taken 10 minutes later.

RESULTS

5 patients were identified. 3 were female. The median age was 50 years (range 34-76). All had a Munk score of 4 on first presentation. 40% (2/5) of patients had sticky discharge along with thier epiphora. The double puncta were unilateral and on the lower lid in all cases.

Patient 1

A 51-year-old man presented with left sided epiphora for 5 years and a Munk score of 4. Abnormal examination findings on the left side were: two lower puncta (Figure 1A), a narrow lateral lower punctum, a high tear meniscus and delayed fluorescein dye disappearance test (FDDT). Lacrimal syringing required dilation of the lateral lower punctum with a nettleship dilator to allow the introduction of a cannula. Partial reflux of saline from the upper punctum was observed when irrigating at the mid canalicular level. When advancing the irrigation cannula, a narrowing at the common canaliculus was encountered which was overcome. The cannula was then passed into the lacrimal sac, confirmed with a hard stop. Irrigation then showed no reflux, with all the saline entering the nasal space. The patient was investigated with OCT of the punctal anomaly. This identified

a normal vertical canaliculus beyond the lateral punctum, but only a horizontal canaliculus beyond the medial punctum (Figure 1B). A DCG showed that the two lower canaliculi fused half way between the puncta and the common canaliculus. DCG contrast confirmed narrowing of the common canaliculus, and a distended lacrimal sac. No passage of contrast through the nasolacrimal duct into the nasal space was observed (Figure 1C, D). The patient underwent punctoplasty of the lateral lower lid punctum, which reduced his epiphora to a Munk score of 2 at his two month post operative review, which he felt did not interfere with his daily life sufficiently enough to warrant any further surgery.

Patient 2

A 46-year-old lady presented with left sided watering and stickiness for 1 year and a Munk score of 4. Abnormal examination findings were; two left lower lid puncta (Figure 2A) and a red pouting upper punctum with mucous expressed on upper canalicular massage (Figure 2D), suggesting canaliculitis. The tear film was high but FDDT normal. Lacrimal syringing through both lower puncta did not identify reflux or resistance. Punctum OCT showed both lower canaliculi to have vertical components, with the horizontal components or junction not visualised. An upper punctum canaliculotomy was recommended to treat the infection. Several stones and mucoid material were expressed from the upper canaliculus. Canalicular endoscopy did not show any anatomical abnormality of the upper canaliculus that could have predisposed to the canaliculitis. The mid canalicular junction of the lower canaliculi was visualised through canalicular endoscopy through the lateral lower lid punctum by passing a Bowman probe through the medial lower lid punctum (Figure 2D). At her 1-month postoperative review she had no epiphora or discharge.

Patient 3

A 50-year-old man presented with left sided epiphora for 1 year and a Munk score of 4. Abnormal examination findings were: two left lower lid puncta, bilateral blepharitis, normal tear meniscus height and FDDT. The nasolacrimal duct was patent to syringing through the lateral lower punctum. Punctum OCT showed a vertical canaliculus beyond the lateral punctum, but only a horizontal canaliculus beyond the medial punctum (Figure 3B). DCG showed passage of dye through the NLD and into the nasal space, with dye filling the medial lower canaliculus with an origin close to the common canaliculus. The erect x-ray showed retention of some dye within the NLD. He was then treated conservatively for blepharitis with daily lid cleaning, hot massage and lubricants. At two-month review, his epiphora had reduced to a Munk score of 1 and he did not wish further intervention.

Patient 4

A 34-year-old lady presented with left sided epiphora for 3-4 years and a Munk score of 4. Two left lower puncta with a delayed FDDT was identified on examination but all other findings were normal, with the NLD patent on syringing. OCT (Figure 4B) and lacrimal probing (Figure 4C) identified the junction of the two lower canaliculi to be very proximal to the lid margin, with their one horizontal canaliculus visible on OCT. She underwent endonasal DCR surgery with O'Donaghue intubation and her Munk score was 1 at her 6-week post operative review (2 weeks after tube removal).

Patient 5

A 76-year-old lady presented with right-sided epiphora and stickiness for 3 years and a Munk score of 4. Examination revealed two right lower lid puncta, and a right mucocoele with a blocked NLD on syringing. Punctum OCT showed a vertical canaliculus beyond the lateral punctum, but a horizontal canaliculus beyond the medial punctum (Figure 1B). DCG showed a blocked NLD, with the two lower canaliculi fusing at the common canaliculus.

She underwent endonasal DCR surgery without intubation. At one-month post operative follow up her Munk score was 0 with no discharge.

DISCUSSION

Association of double punctum anomaly with epiphora

This case series suggests that the congenital double punctum is associated with symptoms of watering, as all five patients had ipsilateral epiphora. Interestingly, none of the patients complained of epiphora from birth despite the presence of this congenital abnormality, which suggests that there is significant redundancy in the capacity of the lacrimal system in these patients. Additionally all double puncta were unilateral and in the lower lid, as described by Satchi et al in a case series of 23 patients.[5]

This study's findings of a variety of adnexal abnormalities co-existing with the double punctum anomaly emphasises the importance of a comprehensive nasolacrimal assessment to exclude other pathologies such as canaliculitis and nasolacrimal duct obstruction. Coexistent double punctum and canaliculitis has been described in the same eyelid[6], with infection postulated to have occurred due to a cul-de-sac lateral canaliculus. However, our patient had canaliculitis in the fellow ipsilateral upper punctum, without a cul-de-sac canaliculus identifiable on canalicular endoscopy.

Identification of a double punctum can be an incidental finding, as illustrated by patients 2 and 3, whose epiphora mostly settled after treatment of upper punctum canaliculitis and blepharitis.

Novel anatomical findings

<u>Range of anatomical locations of the junction of the two lower canaliculi</u>

We identified a range of anatomical locations of the junction of the two lower canaliculi with one at the proximal ocular surface end, two mid-canalicular and two at the common

canaliculus suggesting abnormalities of embryogenesis at slightly different timings. A junction at the proximal ocular surface end was also noted in 2 patients from a case series of 23 and described as a defect in the canalicular roof of a single canaliculus [5].

Superficial horizontal canaliculus

Punctum OCT in these patients allows visualisation of the horizontal canaliculus attached to the medial punctum in the majority (80% 4/5), which has not been visible in patients with a single punctum with or without epiphora.[2-4] This may suggest a more superficial location of their canaliculi during embryogenesis.

Lack of a vertical canalicular portion

The lack of a vertical canalicular portion attached to the medial punctum was seen on OCT in the same 4 patients who had a superficial horizontal canaliculus. This suggests that the lid surface was encountered prior to commencement of vertical canalicular growth, due to the superficial location of the horizontal canaliculus.

Patient 2, who had upper lid canaliculitis, was the only patient with a vertical canalicular component and without a superficial horizontal canaliculus leading to their medial punctum on OCT. They achieved full resolution with canaliculotomy of the upper lid alone. This suggests that a medial punctum that has a similar morphology to a normal punctum (with a vertical canalicular component and deep horizontal canaliculus) will cause less severe tear drainage disruption than in other double punctum patients.

Potential causes of poor drainage

Potential theories to consider how the double punctum contributes to reduced tear drainage includes;

Reduced lacrimal pump function

It is possible that the double punctum affects the lacrimal pump mechanism[7] of drawing tears into the lacrimal sac through contraction of Horner's Muscle[8]. OCT examination

suggests that the lateral of the two puncta is the more physiological with a morphology more consistent with that of healthy volunteers[2]. The vertical component of the canaliculus was present in all of the lateral puncta. Additionally, characterising the horizontal component of its canaliculus was not possible as the depth of this structure was beyond the maximum penetration depth of the OCT, as is the case in normal subjects. Accordingly, OCT imaging suggests that the canalicular element related to the medial of the two puncta has a more abnormal morphology [2-4] with no vertical canalicular component in 80% (4/5) of patients and an abnormally superficial horizontal canaliculus visible on OCT in 80% (4/5) of patients.

This means that the area between the medial lid margin and canaliculus that is usually filled with the Horner's portion of the orbicularis muscle, which contracts the canaliculus on blinking, is replaced with an accessory canaliculus. Thus canalicular contraction, with subsequent pulsing of tears into the lacrimal sac cannot occur.

Our OCT findings are consistent with clinical observations suggesting that the medial punctum is the abnormal one.[5]

<u>Reflux of tears through the accessory punctum</u>

Tears that enter the lacrimal punctum can return to the tear film through the accessory punctum, particularly if this route provides less resistance to that of the nasolacrimal duct.

<u>Coexistent congenital anomaly of the lower lacrimal drainage system</u>

The lacrimal drainage apparatus develops at 5-10 weeks gestation[9], starting with lacrimal sac formation and followed by simultaneous co-development of the canaliculi and nasolacrimal duct from invaginated ectoderm. This co-development could imply that an environmental or developmental insult to the embryological ectoderm at this point in development leads to a simultaneous abnormality in the nasolacrimal duct development, increasing its resistance to tear flow and predisposing it to obstruction.

DCR surgery

In one patient, patient 4, the only abnormality identified was the double lower punctum. This patient benefited from an endonasal DCR, suggesting that lacrimal bypass surgery may be a possible option for these patients. DCR surgery may increase the flow of tears through the abnormal double punctum by reducing the resistance of the tears through the nasolacrimal duct. Satchi et al 2010 reported 23 patients, 5 of who were asymptomatic. Out of 12 adults, 11 had improvement or resolution of symptoms with DCR surgery,[5] confirming this as a successful management option in these patients.

Punctoplasty

Patient 1 had a narrow lateral punctum. This study has found that the lateral punctum has the more normal morphology. This implies that treating a stenosed lateral punctum may be more beneficial than treating a stenosed medial punctum, in the double punctum patient. There is a guarded prognosis for benefit from punctoplasty due to the likelihood of it being a secondary punctal stenosis from canalicular resistance.

In summary, the double punctum is more common on the lower than the upper lid. OCT imaging suggests that the medial of the two puncta is the abnormal one, with DCG and endoscopy demonstrating that it can be connected to a range of different accessory canaliculus lengths. The duplex punctum can co-exist with a range of other nasolacrimal pathologies, which should be excluded on examination.

LEGENDS

Figure 1.

Patient 1's left lower lid double punctum [1A] infrared image, **[1B]** OCT, **[1C]** DCG through lateral punctum, **[1D]** DCG through medial punctum.

Patient 2's left lower lid double punctum [2A] infrared image, **[2B]** OCT, **[2C]** canalicular endoscopy through lateral punctum with canalicular lumen visible and Bowman probe through medial punctum visible in the mid canaliculus, **[2D]** upper punctum canaliculotomy with release of mucous and stone.

Patient 3's left lower lid double lower punctum [3A] infrared image, **[3B]** OCT, **[3C]** DCG through lateral punctum (unable to cannulate medial punctum) **[3D]** erect x-ray with minimal retained dye in nasolacrimal duct.

Patient 4's left lower lid double punctum [4A] infrared image, **[4B]** OCT, **[4C]** probe revealing medial punctum as a roof opening of the lateral punctum's horizontal canaliculus.

Patient 5's right lower lid double punctum [5A] infrared image, **[5B]** OCT, **[5C]** DCG through lateral punctum, **[5D]** DCG through medial punctum, **[5E]** erect x-ray with retained dye in lacrimal sac

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