ABSTRACT

Dacryocystocele is an uncommon congenital obliteration of the nasolacrimal drainage system. Based on a case diagnosed at 30 weeks gestation using two-dimensional (2D) and three-dimensional (3D), its ultrasound characteristics as well as the evolution and therapeutic options applied in the scarce existing literature are described.

Keywords: Dacryocystocele, 2D and 3D ultrasound, Nasolacrimal duct cyst, Prenatal diagnosis.

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INTRODUCTION

Dacryocystocele, amniocele, amniotocele, lacrimal gland cyst and mucocele are congenital stricture in, or obliteration of, the nasolacrimal drainage system, resulting in a fluid-filled closed sac (mucus, debris and/or amniotic fluid). It is a rare and benign condition that occurs quite commonly in neonates.1

ORIGIN AND ETIOLOGY

There is always a membrane obstruction at the opening of the duct in the nasal cavity (membrane of Hasner) which, if accompanied by proximal stenosis, dilates and forms a dacryocystocele (Fig. 1).

Diagram of the development of the nasolacrimal duct during intrauterine life. The membrane of Hasner depicted in red. To the left, still imperforated (month 7), to the right it is perforated (month 8). Congenital dacryocystocele only appears when there is stenosis or atresia of this membrane. When there is atresia or stenosis of the duct at the orbital level, the so called canaliculi lacrimalis communis, congenital dacryocystocele appears (blue arrows; modified from 2).

The nasolacrimal duct starts to develop toward the 6th week from the epiblast of the basal plaque. As the surface ectoderm in the naso-optic fissure thickens an epithelial cord detaches from it and buries itself between the lateral nasal and maxillary processes. Cephalical and caudal growth of this epithelial cord will give rise to the lacrimal canaliculi, sac and duct.3

Canalization commences around week 12 and is not complete until week 24; however, the distal area (nasal) is sometimes only perforated at around the time of birth or even afterward. This permeabilization first starts at the proximal portion of the eye. The opening in the nose takes place between the 7th and 9th months, but the membrane that keeps it closed (Hasner’s) continues until the time of birth in 30 to 70% of cases.2,4

ULTRASOUND DIAGNOSIS

Two-dimensional Image

Fetal ultrasound scans before 27 weeks are normal and appropriate because the canalization commences around week 12 and is not complete until week 24.

At week 30 practically all described cases showed with 2D one or two echo-negative round cystic formations, just below the medial canthal area under the orbits, on the inner lower medial line of one or both eyes. The size oscillates between millimeters, in our case measuring 7 and 6 mm, well defined and with no Doppler flow.

In summary their 2D ultrasound characteristics are:

- Hypoechoic periorbital mass
- Located in the lower and medial area of the orbits
- Rounded, with no vascular flows or calcifications, that can be uni- or bilateral
- The eyeball is symmetric, normal, it is not out of place
- Synchronous eye movements can be observed
- Exhibiting normal brain and facial structures.
All these characteristics make difficult to confuse this anomaly with other facial masses distorting the fetal face 2D image left. The white arrows show the orbits and dacryocystoceles from a frontal (left) and cranial (right) view. On the right-hand side DIO = internal interorbitary diameter; DBO = external or biorbital interorbitary diameter. The white arrow shows a small but evident swelling of the subcutaneous tissue. The yellow arrows show the communication between the cysts paraocular mass and the orbit.

In view of the diagnosis of dacryocystocele, we resorted to surface three-dimensional (3D).

3D Image

When using 3D, the following advantages where observed:

- Clearly depicted and identified the anomaly, and allowed to determine both the precise location of the cysts and the degree of intranasal extension.5,6
- Both tumors were clearly observed under the eyes, next to the nose. The cysts were sharply defined when region of interest (ROI) was applied to the tumor site (Fig. 3).
- Showed the degree of intranasal extension and the communication with the orbits (Figs 1 and 3, above left)
- Surface-rendering imaging showed swelling below the medial canthal area.6
- Make much clearer parents understanding of the anomaly.5

3D surface image. Above, frontal view of the face showing eyes closed and two tumors. Below, side view of the surface of the nose, eye and dacryocystocele (red arrow). Also, the face of the newborn showing two blue tumors (red arrows)

and the symptoms appear between 10 and 30 days after birth.4 In most cases perforation is spontaneous.

- Dacryocystoceles are much less usual; there is always an additional functional obstruction in the lacrimal sac near the orbit (Rosenmüller’s membrane; Fig. 1, blue arrows). They are practically always symptomatic at birth or shortly afterward.

Newborns show no symptoms (only 6%)7 or such symptoms are very slight, and because of spontaneous resolution (78% at 3 months and 91% at 6 months) it is not considered as being a serious condition.8

Only few references have resulted in neonatal respiratory distress requiring surgical intervention.9,10

About Ultrasound

The majority of prenatal reported cases using 2D ultrasound (US) have been isolated cases.4,9,11-21 Nevertheless, two series with six (1) and ten cases (6) respectively have been published.

Although some other cases have been reported as prenatal diagnosis, many belong to postnatal controlled series (ref. 10: 54 cases, ref. 2: 3 cases, ref. 7: 21 cases, ref. 8: 4 cases, ref. 22: 4 cases, ref. 4: 5 cases).

Cases using 3D US are very scarce, appeared more recently and showed the diagnostic importance of this new technology5,6,23 which includes simultaneous orthogonal views, multiplanar plane slicing and rotation and surface rendering.

Prevalence and Clinical Evolution

As most cases are mild, or more commonly resolve spontaneously in intrauterine life at the end of the pregnancy or during the immediate neonatal period, their prevalence
is unknown and is probably very much greater than reported by the literature. Table 1 summarizes the 34 cases diagnosed prenatally with US and show their clinical evolution.

All the cases were diagnosed after week 27 (one case); and usually they are not described before 30 weeks because the canalization may not be complete.

Some were transitory (18 cases) and disappeared during intrauterine life, others persisted.

The size is always small and ranged between 3 and 13 mm. They were all hypoechoic (32 cases) or gray echoes (2 cases gray or with debris 6); and the shape were always rounded or oval.

It was reported that they appeared alone, unilaterally (90%), on either side, only 13% were bilateral and predominantly in female fetuses.1,4,6,10,12,15

Our review shows that 47% were bilateral and only five out of 34 were male newborns.

It has been reported that they may accompany other malformation syndromes, such as ectrodactyly-ectodermal dysplasia-clefting (EEC), down, craniofacial anomalies, multicystic kidney disease,2 microphthalmia, palatoschisis, spheroidal meningocoele and hydrocele.10 However, we only encountered one case, a baby that died 6 months after birth from Canavan syndrome1 and three cases were described with polyhydramnios,11, 17 but two of them11 did not show an excess of amniotic fluid at the time of delivery. Hence, reports on accompanying syndromes have not been proven, at least in fetal life.

### Table 1: Description of published cases

<table>
<thead>
<tr>
<th>Authors</th>
<th>Years</th>
<th>Cases</th>
<th>Week at diagnosis</th>
<th>Maternal age</th>
<th>Gravida</th>
<th>Para</th>
<th>Lesion</th>
<th>Outcome</th>
<th>T/P</th>
<th>Comments</th>
</tr>
</thead>
<tbody>
<tr>
<td>Davis 1987</td>
<td>1,32</td>
<td>1,2</td>
<td>30,25</td>
<td>1,1</td>
<td>0,0</td>
<td>8,13</td>
<td>Healthy,T, polyhydramnios</td>
<td>Healthy,P, polyhydramnios</td>
<td>Female</td>
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<tr>
<td>Schaub 1992</td>
<td>1,32</td>
<td>3,2</td>
<td>1,4</td>
<td>2,2</td>
<td>10/8,13</td>
<td>Healthy,T</td>
<td>Healthy,P</td>
<td>Twins healthy,P</td>
<td>Surgery</td>
<td></td>
</tr>
<tr>
<td>Walsh 1994</td>
<td>1,36</td>
<td>3,4</td>
<td>0</td>
<td>1,11/5,13</td>
<td>Healthy,P</td>
<td>Female</td>
<td>Female</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Batiglia 1994</td>
<td>1,33</td>
<td>2,2</td>
<td>0</td>
<td>11,11</td>
<td>Healthy,T</td>
<td>Female</td>
<td>Female</td>
<td></td>
<td></td>
<td></td>
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<tr>
<td>Shipp 1995</td>
<td>1,32</td>
<td>?</td>
<td>?</td>
<td>B</td>
<td>Healthy,P</td>
<td>Female</td>
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<tr>
<td>Sherer 1997</td>
<td>1,32</td>
<td>3,1</td>
<td>0</td>
<td>9,9</td>
<td>Healthy,P</td>
<td>Male</td>
<td></td>
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</tr>
<tr>
<td>Kivikoski 1997</td>
<td>1,32</td>
<td>7,5</td>
<td>7/5,5</td>
<td>Healthy,P, polyhydramnios</td>
<td>Female</td>
<td></td>
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<td></td>
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<td></td>
</tr>
<tr>
<td>Sharony 1999</td>
<td>4,8</td>
<td>6</td>
<td>3</td>
<td>5/3,5</td>
<td>Healthy,P</td>
<td>Diabetes,?</td>
<td>Diabetes,?</td>
<td>Male</td>
<td></td>
<td></td>
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<tr>
<td>Suman 1999</td>
<td>5,36</td>
<td>24,1</td>
<td>0</td>
<td>10/4,8</td>
<td>Healthy,P</td>
<td>MCKD,?</td>
<td>MCKD,?</td>
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<tr>
<td>Salvetat 1999</td>
<td>1,38</td>
<td>2,2</td>
<td>8,8</td>
<td>Healthy,P</td>
<td>?</td>
<td>Female</td>
<td>Female</td>
<td>Female</td>
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<tr>
<td>Goldberg 2000</td>
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<td>3</td>
<td>8,5</td>
<td>Healthy,P</td>
<td>Female</td>
<td>Female</td>
<td>Female</td>
<td>Female</td>
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<tr>
<td>D’Addario 2001</td>
<td>1,34</td>
<td>4</td>
<td>3</td>
<td>Healthy,P</td>
<td>?</td>
<td>8 months</td>
<td>Female</td>
<td>Female</td>
<td></td>
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<tr>
<td>Petrikovsky 2003</td>
<td>1,27</td>
<td>34</td>
<td>7</td>
<td>Healthy,T</td>
<td>Female</td>
<td></td>
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<tr>
<td>Sepulveda 2005</td>
<td>5,30</td>
<td>2,6</td>
<td>7/4,7</td>
<td>Healthy,T</td>
<td>Female</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
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<tr>
<td>Brown 2011</td>
<td>1,28</td>
<td>30,1</td>
<td>5/7</td>
<td>Healthy,T</td>
<td>3D, male</td>
<td>3D, female</td>
<td>3D, female</td>
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<tr>
<td>Total 34 27-38</td>
<td>17-40</td>
<td>1-7</td>
<td>0-5</td>
<td>3-13</td>
<td>16</td>
<td>5</td>
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</table>
The progressive accumulation of mucus secretion leads to cystic dilation of the duct and as the nasal bones are still very soft, a bulge may appear which, in extreme cases, can hinder or obstruct breathing.\(^2\)

All with 3D-reported articles [ref. 5 (one case), ref. 6 (three cases), ref. 23 (one case)], showed that the image was much better, with greater definition, more exact localization, volumetric calculation, determination of size, spatial demarcation and orientation and, therefore, it was much better for giving advice to the parents.

This was exactly the same in our case; 3D vision defined much better and made all differential diagnoses easier.

The literature stresses that differential diagnoses should be performed with inflammations (canaliculitis, dacryocystitis, rhinitis), obstructions (atresia or choanal stenosis), anterior encephalocele, meningocoele, teratomas, hemangiomas, gliomas and rhabdomyosarcoma, neurofibromatosis and lymphangiomas.\(^1,2,15,22\)

The complications described have been dacryocystitis, orbital cellulitis and respiratory distress due to extension of the cyst in the nasal cavities.

**Treatment:** Those that persist do not usually pose a problem and at birth present taut, firm, gray-blue mass just below the lower ocular angle and medial to the orbits. The impatency of the nasolacrimal duct is a result of the thin mucosa that obstructs and usually perforates spontaneously, or with gentle massage, at birth. Treatment options include only observation and monitoring for 6 months (sufficient in the vast majority of cases). Hot compresses and/or topical antibiotics, anterograde massage and nasal probing and drainage, if necessary, are further options.\(^2\,4,10\)

After 6 months all the subjects, except for the case with Chavanan syndrome, were alive and free from tumors.

In our review four cases needed massage, two cases drainage and in only one case surgery was required. After birth or 6 months later 77% of all described cases were healthy without any treatment.

**REFERENCES**


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