# An Ultrasound Diagnosis of a Congenital Periorbital Rare Condition

**Rita Ladeiras*, Joana Sousa Nunes, Sílvia Torres, Carla Duarte, Elsa Pereira, Adosinda Rosmaninho**
Department of Gynecology and Obstetrics, Hospital Senhora da Oliveira, Guimarães, Braga, Portugal

## Section 2 – Answer

### Case description

A 38-year-old pregnant woman, gravida 2 para 1 (cesarean delivery for active phase arrest) with asthma medicated with a combination of inhaled glucocorticoid (fluticasone) plus a long-acting beta agonist (salmeterol) twice a day, no history of congenital malformations in the family, was referred to our obstetric department in the first trimester for pregnancy surveillance.

The combined first trimester screen reported a low risk for major chromosomal aneuploidies (trisomies 21, 18, and 13). The second trimester scan showed a female fetus, apparently without any anatomical abnormality. The placenta was anterior, the umbilical cord had three vessels and the amniotic fluid volume was normal. At 28 weeks of pregnancy, an ultrasound showed a hypoechogenic and circular image located on the anterior and medial left orbital surface, without blood flow in or around it, with approximately 8 mm × 7 mm [Figure 1]. A complementary magnetic resonance study was requested, which identified a small cystic aspect formation with hyposignal on T1 and hypersignal on T2 [Figure 2].

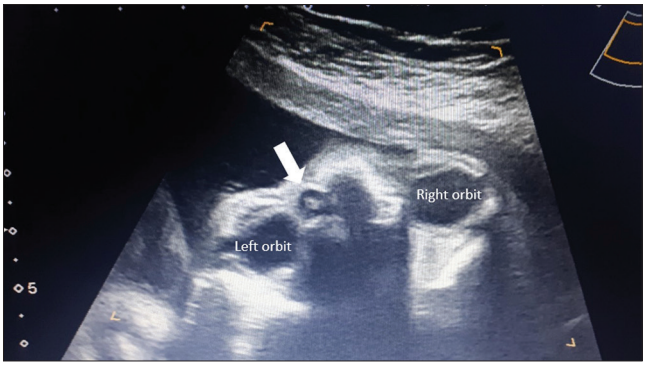


**Figure 1:** Ultrasound image of the orbital transverse plan. A hypoechogenic and circular image at the anterior and medial left orbital surface is pointed by a white arrow

### Interpretation

In the orbital transverse plan, both fetal orbits appear symmetrical and intact. However, at the anterior and medial left orbital surface, a hypoechogenic and circular image without blood flow in or around it, with approximately 8 mm × 7 mm, is shown. Due to the fetal position, it was not possible to assure the integrity of the nasal bone contiguous to the lesion and the differential diagnosis of encephalocele frontoethmoidal was proposed. In order to exclude the presence of brain tissue in the lesion image, a complementary magnetic resonance study was requested. In Figure 2, a small cystic aspect formation

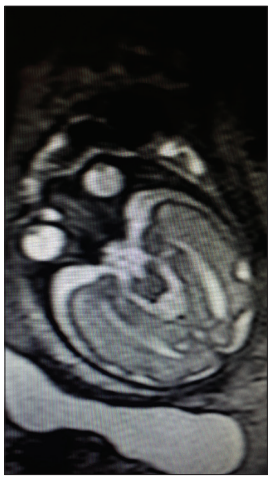

**Figure 2:** Magnetic resonance imaging of the orbital transverse plan, T2-weighted image

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**Address for correspondence:** Dr. Rita Ladeiras, Rua dos Cutileiros, Creixomil, 4835-044 Guimarães, Portugal.
E-mail: ritaladeiras218@gmail.com



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with hyposignal on T1 and hypersignal on T2 is identified. The nasal bone integrity was confirmed and the presence of brain tissue was excluded. The cystic image was located in the nasolacrimal duct trajectory confirming the diagnosis of congenital dacryocystocele.

## Discussion

Congenital dacryocystocele is an uncommon variant of congenital nasolacrimal duct obstruction, corresponding for 0.1%–0.3% of all such cases.[1] It occurs when both the proximal and distal parts of the nasolacrimal system are obstructed, usually at the proximal valve of Rosenmuller, or at the common canaliculus, and at the distal valve of Hasner.[2] This double blockage creates a complete obstruction of the lacrimal drainage with a fluid accumulation within the lacrimal sac, which appears as a cystic bluish swelling below the medial canthus.[2-4] The diagnosis is usually made by physical examination at the 1st day of life, with a mean age at presentation at 7 days, however prenatal diagnosis by ultrasound is possible, mostly in the third trimester.[1,5] The tears accumulation within the lacrimal sac may infect, and then complicate as a dacryocystitis, a medical emergency in the newborn. On the other hand, the dacryocystocele may extend into the nasal cavity and create a respiratory distress by nasal obstruction.[6] Due to these potentially serious complications, congenital dacryocystocele diagnosis requires urgent referral to an ophthalmologist.[2]

Dacryocystoceles are more common in females, Caucasians and are predominantly unilateral.[3] This malformation is considered benign, with spontaneous prenatal resolution in 50% of all cases.[3,7] However, more severe malformations, like hemangioma, teratoma, venous lymphatic malformation, dermoid cyst, rhabdomyosarcoma, and encephalocele can present as facial cysts.[8] Therefore, prenatal exclusion of these more serious situations is important and, sometimes, it is not possible to assure the malformation extension and nasal permeability by ultrasound. Magnetic resonance imaging (MRI) allows more accurate images, providing a better contrast between tissues and spatial relations.[8] For these reasons, the authors recommend performing MRI in cases of difficult visualization of the extent of the malformation, as described in this case, in order to exclude these differential diagnoses, allow better parental counseling and to identify postnatal treatment options.

The treatment is often conservative and consists of lacrimal sac massage. When complicated by dacryocystitis, it should be treated promptly with systemic antibiotics, once it can be complicated by orbital or preseptal cellulitis, meningitis, or sepsis. The surgical approach is not usually necessary, but when required, the lacrimal duct probing is the preferred procedure.[1,2]

In conclusion, it is important to know the ultrasound findings of congenital dacryocystocele to do prenatal diagnosis and to exclude more severe malformations with similar presentations, which may avoid additional postnatal diagnostic techniques and potentially serious complications by allowing early referral to an ophthalmologist.

### Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understand that her name and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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Nil.

### Conflicts of interest

There are no conflict of interest.

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