**Bilateral dacryocystoceles causing respiratory difficulty: a case report and literature review**

A congenital dacryocystocele (also known as a nasolacrimal duct cyst) results from a failure of canalisation of the nasolacrimal duct, forming when amniotic fluid fills the obstructed duct causing distention of the lacrimal sac. This article describes the rare case of a newborn female infant with bilateral congenital dacryocystoceles presenting with respiratory difficulty soon after birth. The diagnosis and management of dacryocystoceles is discussed.

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**Keywords**
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**Key points**
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1. Bilateral nasal obstruction can cause respiratory difficulty which can be life threatening if not treated promptly.
2. Dacryocystoceles are easily treatable yet can be clinically difficult to differentiate from other causes of congenital nasal obstruction.
3. Prompt imaging and expedient surgical management is recommended.

**The case study**
Baby C, a female infant, was born at 41+2 weeks’ gestation by spontaneous vaginal delivery. There had been no antenatal concerns and the anomaly scan at 20 weeks’ gestation was normal. At birth the infant weighed 4.1kg and her Apgar score was 9 at one minute and 10 at five minutes; no breathing difficulties were noted in the immediate postnatal period.

Within an hour of birth it was noted that Baby C was having difficulty in establishing feeding; she was only able to latch on to the mother’s breast for a few seconds before becoming breathless and coughing. By 11 hours, she had significant breathing difficulties and the neonatal team were called to review Baby C. Upon examination she was borderline tachypnoeic with a respiratory rate of 60 breaths per minute, she exhibited nasal flaring and subcostal recession, increased ‘snuffly’ upper airway noise and mouth breathing. She had swollen eyelids with a mucopurulent discharge (FIGURE 1). Oxymetazoline nasal drops were initiated, which improved her upper respiratory obstruction symptoms however she required a nasogastric tube for feeding.

A CT scan was arranged, which was initially reported to be normal. At five days’ old Baby C underwent nasendoscopy under general anaesthetic. She was found to have bilateral cystic swellings originating from underneath the inferior turbinates. Upon their removal, secretions extruded into the eyes, indicating a nasolacrimal duct pathology.

When the CT scans were reviewed with the intraoperative clinical findings, the bilateral dacryocystoceles became obvious (FIGURE 2). Postoperatively the infant had significant improvement of her upper respiratory obstruction; she resumed breastfeeding and was discharged two days after the operation.

Three weeks later, Baby C was admitted with deterioration in feeding and underwent a second examination under
anaesthesia and removal of residual nasolacrimal duct cyst remnants. She was discharged five days later. Baby C remained well and was discharged from follow-up eight weeks later.

Discussion

Onset of respiratory difficulty in the term neonate is a relatively common presentation in the early postnatal period and can be due to a myriad of conditions. Infection is the most frequent cause of neonatal respiratory difficulty and causative organisms include group B Streptococcus, Staphylococcus aureus, Streptococcus pneumoniae, Chlamydia trachomatis and herpes simplex virus. Transient tachypnoea of the newborn, surfactant deficient lung disease and meconium aspiration syndrome are also common causes. Less common aetiologies are pulmonary and cardiac structural abnormalities, haematological derangements and inborn errors of metabolism.

Congenital upper airway obstruction is a rare but important cause of major respiratory problems in the neonate. The differential diagnoses of congenital upper airway obstruction in the newborn are classified according to the anatomical level at which the obstruction occurs.

Nasal obstruction is the most common upper airway obstruction causing respiratory symptoms in the neonate. Neonates are obligate nasal breathers and bilateral nasal obstruction in this population can cause respiratory distress which can be life threatening if not recognised and treated promptly. Clinical presentation can vary but respiratory difficulties often follow a cyclical pattern; they are observed particularly during feeding and sleeping and improve when the baby cries. There are several congenital abnormalities that can cause nasal obstruction. Choanal atresia – blockage, mostly bony, of the superior aspect of the nasal passage due to failed canalisation of the nasal fossae during fetal development – is the commonest, with an incidence of 1 in 7,000 live births. It is twice as common in females than males and more commonly unilateral. Other causes include pyriform aperture stenosis and congenital nasal mass lesions such as haemangioma, lymphangioma, chondroma, neuroblastoma, neurofibroma, meningoencephalocele, nasal glioma, dermoid cyst and dacryocystoceles.

Oral and pharyngeal obstruction can be a result of syndromic craniofacial abnormalities, the most common of which is the Pierre Robin sequence, consisting of micrognathia, glossoptosis and cleft palate. Crouzon, Apert, Pfeiffer, Treacher Collins, craniofacial microsomia and Goldenhar syndromes are also linked with oropharyngeal obstruction in the neonate.

Obstruction in the larynx and trachea can be due to laryngo-tracheal malacia, subglottic stenosis, vocal cord paralysis, vascular rings and laryngeal webs.

Congenital dacryocystoceles are a rare but significant cause of nasal obstruction in the neonate. They can be associated with other congenital facial abnormalities such as Down’s syndrome, craniosynostosis, clefting syndromes and Goldenhar syndrome.

Dacryocystoceles are due to a failure of canalisation of the nasolacrimal duct apparatus during the sixth month in utero, leaving a membranous barrier between the duct and the nasal cavity at the level of the valve of Hasner and a concomitant valve-like obstruction at the junction of the lacrimal canal and sac proximally. A dacryocystocele occurs when amniotic fluid fills the obstructed nasolacrimal duct apparatus at birth, forming a membranous cyst that extends from the end of the lacrimal duct into the nose. Dacryocystoceles may be diagnosed on prenatal ultrasound scans. As well as causing respiratory difficulties, neonates with dacryocystoceles can present with a bluish swelling of the medial canthus, epiphora, mucous or mucopurulent discharge and dacryocystitis.

Early identification of the cause of neonatal upper airway obstruction is important in order to initiate treatment and avoid any hypoxia-related complications. A thorough history should include the nature of the respiratory difficulties; stridor, wheeze, grunting, transient tachypnoea of the newborn, surfactant deficient lung disease and meconium aspiration syndrome are also common causes. Less common aetiologies are pulmonary and cardiac structural abnormalities, haematological derangements and inborn errors of metabolism.

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Early identification of the cause of neonatal upper airway obstruction is important in order to initiate treatment and avoid any hypoxia-related complications. A thorough history should include the nature of the respiratory difficulties; stridor, wheeze, grunting, tachypnoea, apnoeas, cyanosis, abnormal phonation, coughing/vomiting and association with feeding and the position of the neonate. Laryngomalacia, for example, is typically less symptomatic at rest and while asleep, it rarely interferes with feeding but the neonate becomes more symptomatic on crying, and when distressed. A laryngeal mass can cause airway obstruction in the supine position but this is more often due to, at least in part, a degree of supralaryngeal obstruction such as micrognathia and a
posteriorly displaced tongue base causing occlusion. Improvement in the airway with crying occurs in gross nasal obstruction such as bilateral choanal atresia.

Once upper airway obstruction has been identified as the cause of the respiratory difficulty, re-examination of the infant should identify facial dysmorphia and any physical features of congenital syndromes associated with upper airway obstruction (if not already noted). Eye symptoms such as epiphora or mucous discharge can indicate a problem. As a relatively common cause of nasal obstruction, choanal atresia should be excluded by attempting to pass a nasogastric tube down both nostrils.

Nasendoscopy enables the diagnosis of most causes of nasal, pharyngeal and laryngeal obstruction and should be the next stage of investigation as it can be performed without the need for a general anaesthetic and is generally well tolerated by the infant. When nasal cysts are seen on nasendoscopy, CT scanning is a useful preoperative planning aid and can be useful to differentiate between dacryocystoceles, dermoid cysts, meningoencephalocele and nasal glioma. Magnetic resonance imaging can be employed to demonstrate possible intracranial connections in suspected encephalocele, meningocoele and nasal glioma.

With regards to management, dacryocystoceles that are unilateral or not causing nasal obstruction can be managed medically with normal saline drops with or without steroid nasal drops followed by elective surgery at a later date. As demonstrated by the case presented here, when such cysts are bilateral and causing airway obstruction leading to respiratory and feeding difficulties, the authors recommend prompt imaging with this diagnosis in mind and expedient surgical management with marsupialisation of the cyst using a microdebrider.

Conclusion
Dacryocystoceles should be included in the differential diagnosis of respiratory difficulty in the neonate, especially when more common causes of obstruction such as choanal atresia have been excluded.

References