

Case Series

# Congenital Nasolacrimal Duct Cysts in Infants: Case Series and Literature Review

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## Abstract

**Background:** Congenital obstruction of the nasolacrimal drainage system occurs commonly but rarely causes nasal obstruction. It is usually asymptomatic and resolves spontaneously. It rarely causes respiratory distress.

**Methods:** We describe a case series of infants with congenital unilateral or bilateral nasolacrimal duct cysts (NLDC) and intranasal mucocoeles who presented at different stages of infancy with intermittent respiratory distress and episodic desaturations. We also present a literature review on the topic.

**Results:** All four of our patients required surgical intervention for their symptomatic disease. There were 115 cases of congenital NLDC with significant nasal extension with 93 cases presenting with symptomatic respiratory distress. Majority (87%; n=100) of the cases required some form of surgical intervention.

**Conclusion:** Congenital NLDC causing nasal obstruction are rare but should be considered in the differential diagnosis in infants with symptoms of upper airway obstruction, stertor, dysphonia or feeding difficulties. Surgery is the definitive treatment and results in immediate resolution of symptoms.

**Keywords:** Nasolacrimal duct cyst; Dacryocystocoele; Nasal obstruction; Respiratory distress; Intranasal mucocoele

## Introduction

Congenital obstruction of the nasolacrimal drainage system occurs commonly but rarely causes nasal obstruction [1]. In most cases (94—98%) the obstruction is asymptomatic [2,3] and resolves spontaneously within the first year of life [4].

Nasolacrimal duct cyst (NLDC) is also known as dacryocystocoele, lacrimal mucocoele, lacrimal sac mucocoele, dacryocoele, lacrimal sac cyst, or amniotocoele). It is caused by congenital blockage of both the Rosenmuller valve (proximal NLD valve), where the common canaliculus enters the nasolacrimal sac, and/or the Hasner’s valve (distal NLD valve), where the duct enters the nasal cavity lateral to the inferior turbinate [5-7].

NLDC that expands inferiorly, herniating into the nasal cavity and forming a cystic intranasal swelling if unilateral may present as feeding difficulties and/or sleep disturbances in older infants, while bilateral intranasal mucocoeles present in early infancy with intermittent respiratory distress, apnoeic episodes and episodes of desaturation [8].

We describe a case series of infants with congenital unilateral or bilateral NLDC at different stages of infancy with intermittent respiratory distress and episodic desaturations that ultimately required urgent surgical intervention.

## Case Reports

### Case 1

Patient 1 was a term male born via emergency Caesarean section for foetal distress and was placed on continuous positive airway pressure (CPAP) for three hours post-delivery. The patient presented to the paediatric emergency department (ED) of our institution twice for respiratory distress at the third and 4<sup>th</sup> week of birth with dysphonia, tracheal tug and intercostal recession. At the 4<sup>th</sup> week, he was seen at our institution with worsening respiratory symptoms and associated poor feeding.

A NLDC with nasal extension was observed on flexible nasendoscopy (FNE). The magnetic resonance imaging (MRI) revealed a prominent left-sided dacryocystocoele extending from medial canthus to the level of the superior aspect of the inferior turbinate. There was no lesion on the right side.

A left sided marsupialisation of the cyst and bilateral turbinoplasties were performed with no complications. The

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patient was extubated successfully and discharged home a few days later.

### Case 2

Patient 2, a term male born via vacuum-assisted vaginal delivery was noted to have obstructive nasal symptoms at birth. Attempts to pass a flexible nasal suction through both nostrils were unsuccessful but normal saline was able to be syringed through both nostrils. The patient improved considerably with nasal decongestant sprays and was subsequently discharged. However, he presented to the paediatric ED at our institution on day 19 with worsening respiratory distress, witnessed apnoeic episodes and intermittent desaturations noted in the emergency department.

A thin slice computed tomography (CT) was performed showing bilateral NLDC with intranasal extensions to inferior meatus causing narrowing of anterior and mid-nasal space (Figure 1).

Bilateral cysts marsupialisations and inferior turbinoplasties were performed successfully with immediate improvement in the patient's respiratory symptoms and he was discharged home a few days later.

### Case 3

Patient 3 was a term female born via normal vaginal delivery. After discharge, she developed recurrent respiratory distress and stridor which affected her sleep and feeding. At three months of age, she was referred to our institution with worsening respiratory symptoms despite aggressive medical management.

The FNE showed mild laryngomalacia and bilateral NLDC with nasal mucocoele (Figure 2). There was no evidence of choanal atresia or pyriform aperture stenosis. A CT scan was arranged showing bilateral NLDC with the left nasal mucocoele (Figure 3) causing significant nasal obstruction. Bilateral cysts marsupialisations and inferior turbinoplasties were performed successfully with significant improvement in her breathing and feeding post-operatively. She was discharged home after a few days.

### CASE 4

Patient 4 was a term female born via spontaneous vaginal delivery who presented with episodic respiratory distress, feeding difficulties and facial swelling. CT at birth revealed a



Figure 1. Coronal computed tomography of the paranasal sinus showing bilateral NLDC.



Figure 2. Endoscopic view of the NLDC with nasal extension, appearing below inferior turbinate.

right NLDC causing partial nasal obstruction. She was treated successfully with conservative medical management. A repeat CT at one-month of age showed decreased cystocoele size.

The patient's respiratory symptoms worsened progressively a month later and she was referred to our institution. Further CT imaging revealed the right NLDC causing almost complete right nasal obstruction.

At three months of age, a right marsupialisation of the cyst, inferior turbinoplasties and concurrent probing of the right NLD were performed successfully. The patient showed significant improvement and had an uneventful recovery prior to discharge.

### Discussion

A review of the literature from 1979 to 2012 reveals 115 cases of congenital NLDC with significant nasal extension with 93 cases presenting with symptomatic respiratory distress (Table 1). Of the papers which included gender differences, there were 58 females & 22 males. Majority (87%; n=100) of the 115 cases required some form of surgical intervention. Of these 70% (n=70) required marsupialisation and probing; while a minority (13%, n=13) required marsupialisation only, and probing only in 12% (n=12). Most patients with complicated intranasal cysts requiring surgical intervention had further imaging to delineate the location, boundaries and extent of the cyst for surgical planning.

The function of the NLD system is to facilitate the drainage of tears from the eye into the inferior meatus of the nose. This system has two valves whose function is to ensure the unidirectional flow. The nasolacrimal apparatus begins developing in the third month of gestation from a cord of ectodermal tissue found between the maxillary and lateral aspect of the frontonasal processes. Canalization of the cord occurs uniformly and final communication with the inferior nasal meatus is usually complete by the sixth month of gestation. Incomplete canalisation occurs in approximately 30% [6,7,9] of term infants, however rates as low as 6% and as high as 84% have been reported [10,11].

Two distinct pathologic conditions appear clinically as congenital nasolacrimal obstruction. The more common type of obstruction is a result of malfunction of one of the two valves of

Table 1. Summary of the patient characteristics of congenital dacryocystoceles with nasal extension.

Source	No. of cases/sex	Nasal obstruction/ respiratory distress	Surgical Treatment
Guery D et al. [11]	4/F	Yes	Probe & marsupialisation, 3
Raflo et al. [17]	1/F	No	Probe & marsupialisation, 1
Divine et al [6]	1/F	No	Probe & marsupialisation, 1
Lusk et al. [18]	2/F	Yes	Probe & marsupialisation, 2
Berkowitz et al. [19]	1/M	Yes	Nil
Grin et al. [16]	5/F; 2/M	Yes, 2; no, 5	Probe & marsupialisation, 2
Edmond et al. [8]	2/F	Yes	Probe, 2
Righi et al. [20]	1/M	Yes	Nil
Mazzara et al. [21]	2/F	Yes	Probe & marsupialisation, 2
Castillo et al. [22]	2/M	Yes	Marsupialisation, 2
Yee et al. [12]	1/M	Yes	Probe & marsupialisation, 1
Helper et al. [13]	3/F; 1/M	Yes	Marsupialisation, 4
Paoli et al. [23]	5/F; 2/M	Yes, 6; no, 1	Marsupialisation, 7
Paysse et al. [2]	16	Yes, 9; no, 7	Probe & marsupialisation, 9
Fishman et al. [24]	1/M	Yes	Probe & marsupialisation, 1
Roy et al. [14]	6/F; 2/M	Yes	Probe & marsupialisation, 8
Tabor et al. [25]	1/M	Yes	Probe & marsupialisation, 1
Shashy et al. [1]	7/F; 3/M	Yes, 3; no, 7	Probe & marsupialisation, 7; Probe, 2
Teymoortash et al. [26]	1/M	Yes	Probe, 1
Brachlow et al. [27]	9/F; 1/M	Yes	Probe & marsupialisation, 7
Duval et al. [28]	1/F	Yes	Probe & marsupialisation, 1
Cavazza et al. [29]	4/F; 1/M	No	Probe & marsupialisation, 1; probe 4
Leonard et al. [15]	1/M	Yes	Probe & marsupialisation, 1
Wong et al. [31]	1/F	Yes	Probe & marsupialisation, 1
Wong et al. [31]	4	Yes	Probe & marsupialisation, 4
Hain et al. [5]	4/F; 1/M	Yes	Probe & marsupialisation, 4
Dagi et al. [32]	15	Yes	Probe, 3; probe & marsupialisation, 12
Wong et al. [31]	1/F	Yes	Probe & marsupialisation, 1
Total	115	93	100 surgical procedures



Figure 3. CT scan of case 3 showing bilateral NLDC with the left nasal mucocoele.

the nasolacrimal system, namely the distal Hasner’s valve. This causes backflow of tears, with resultant epiphora, which may progress to dacryocystitis secondary to stasis. These cases are solely managed by ophthalmologists and majority resolve with

expectant management [5]. The second less common pathologic condition results from malfunction and blockage of both the Hasner’s and Rosenmuller valves. The distention of the duct with fluid accumulation may lead to infection with suppuration, which in the neonate can lead to complications like cellulitis, orbital involvement, meningitis, and sepsis [5].

In a minority of the cases, NLDC expand inferiorly and herniate into the nasal cavity. Unilateral cysts may present as feeding difficulties, sleep disturbance or intermittent acute respiratory distress in older infants. Bilateral cysts present in early infancy with intermittent respiratory distress and desaturations [8,12], as infants are obligatory nasal breathers [13]. Patients with unilateral NLDC may have respiratory distress due to a combination of prolapsed cyst causing nasal obstruction, and/or alternating obstruction of the nasal airways due to cyclical vascular congestion. The severity of the symptoms vary depending on the degree of obstruction and range from intermittent episodes of desaturation to requiring emergency intubation for airway support [14].

Differential diagnosis of nasal obstruction includes a variety of congenital malformations like choanal atresia, meningo-encephalocoele, dermoid cysts, Thornwald’s cysts, haemangioma and pyriform aperture stenosis [15], as well as inflammatory conditions, tumours and hamartomas.

Appropriate management include a full history and examination at a specialised tertiary institution. Investigations should include a detailed examination of the external nasal anatomy in combination with anterior rhinoscopy and nasendoscopy. As the differential diagnosis of nasal obstruction includes a variety of congenital malformations like choanal atresia, meningo-encephalocele, dermoid cysts, Thornwald's cysts, haemangioma and pyriform aperture stenosis [15], as well as inflammatory conditions, tumours and hamartomas, investigations like CT or MRI are helpful in delineating the extent of the mass and is essential in ruling out any intracranial communication prior to any biopsy or invasive surgery. MRI has excellent soft-tissue detail and is the best imaging modality to delineate any relationship between the mass and the central nervous system without ionizing radiation. However, if the differential diagnosis includes choanal atresia, nasal stenosis or other nasal abnormalities, a CT scan is recommended because of its superior bony detail. NLDC is defined as a triad of radiological findings of medial canthal cystic mass, dilated NLD and intranasal nasal cystic mass [17].

The ideal management of intranasal NLDC causing nasal obstruction are best managed surgically[3] with endoscopic marsupialisation of the intranasal cyst and/or consequent NLD probing [16]. Endoscopic marsupialisation decompresses the system distally while probing the NLD allows proximal decompression. This re-establishes the continuity of the nasolacrimal drainage system and decompression of the mass effect of the cyst facilitates return to normal respiratory function [15].

## Conclusion

Congenital NLDC causing nasal obstruction is a rare diagnosis but it should be included in the differential diagnosis in infants with symptoms of upper airway obstruction, stertor, dysphonia or feeding difficulties. Surgical management is often the preferred treatment to relieve the airway obstruction and it results in immediate resolution of symptoms.

## Conflicts of Interest

None to declare.

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