

# Telecanthus as A Presenting Sign of a Fronto-Ethmoidal Mucocele in an Eleven Year-Old

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## Abstract

An eleven year-old girl with no prior history of trauma, sinusitis, or cystic fibrosis presented with an isolated sign of recent onset telecanthus, which was not present in photographs from two years prior. Orbital CT-scan showed a fronto-ethmoidal mucocele. The patient was referred to an ear, nose and throat surgeon, who performed an endoscopic drainage and marsupialization of the mucocele. Although mucoceles most commonly present in adults with proptosis, headache, diplopia, globe displacement, and/or epiphora, this case illustrates that it may also uncommonly present in the pediatric population as telecanthus.

**Keywords:** Pediatric mucocele; Telecanthus; Fronto-ethmoidal mucocele

## Introduction

Langebeck first described paranasal sinus mucoceles in 1820, labelling them as “hydatides.” It was not until the early 1900s that mucoceles were given their name by Rollet and popularized by Gerber, who published 178 cases. In 1955, Lambert attributed frontal or ethmoidal mucoceles as the most common nasal condition to produce proptosis [1].

Mucoceles are lined by epithelium containing a collection of mucus and reside within an air sinus. Although benign, mucoceles are capable of expansion by resorption and bony remodeling of the paranasal sinuses, which may cause a malignant appearance. The condition is most commonly unilateral (90%) and generally occurs in the fronto-ethmoidal region (70-98%) [2,5]. Mucoceles are most commonly caused by postoperative scarring; however, they can also be formed by chronic inflammatory processes such as benign tumors, sinusitis, and trauma [3]. Consequently, paranasal sinus mucoceles are rare in children but relatively common in adults.

In the literature, the etiologies of pediatric mucoceles have focused on impaired secretion processes such as cystic fibrosis, pathological pneumatization processes, atopy and trauma [3,4]. Nicollas et al. published a large series on pediatric paranasal mucoceles with ten cases of which six patients had underlying cystic fibrosis, two had facial trauma, and one had chronic sinusitis; in one case there was no identifiable contributing factor [5].

Mucoceles progress slowly, their associated clinical symptoms, such as nasal obstruction and headaches, occur as a result of expansion or inflammatory changes. When affecting the orbit, the most common symptom is proptosis [7]. Other orbital signs and symptoms include epiphora, diplopia, visual loss, raised intraocular pressure, and lateral and inferior globe displacement [8,9]. Nevertheless, to the best of our knowledge, there have been no reported cases of a patient presenting with a sole finding of telecanthus from a paranasal mucocele, especially in a pediatric patient with no identifiable risk factors.

## Case Report

An eleven year-old girl was referred with a presumed diagnosis of right dacryocystitis because her parents had noticed swelling of the

right medial canthal area for two and a half weeks, associated with occasional tearing. The patient denied fevers, chills, redness, pain or localized tenderness. The patient had no history of trauma, previous sinus infections or upper respiratory infections.

On physical exam, the patient’s vision was 20/20. Her extraocular movements were intact and visual fields were full to confrontation. Examining the medial canthus of the right eye, there was no induration, erythema or tenderness upon palpation; however, there was significant telecanthus (Figure 1). The distance from her mid nose to canthus and Hertel’s exophthalmometry were 22 and 16 millimeters and 16 and 15 millimeters with a base of 85 millimeters on the right and left, respectively.

The patient’s lacrimal system was probed and irrigated without evidence of obstruction. An orbital CT-scan was then performed which showed a large fronto-ethmoidal mucocele (Figure 2). A sweat test was negative. The patient was instructed to bring in old photographs, which lacked evidence of telecanthus two years earlier.

The patient was then referred to an ear, nose and throat surgeon,



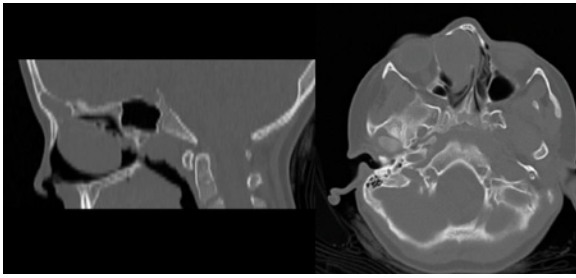
Figure 1: This is an eleven year old patient with significant telecanthus.

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**Figure 2:** This is an axial and sagittal view of a large right sided fronto-ethmoidal mucocele.

who performed an endoscopic drainage and marsupialization of the mucocele two months after her initial presentation. Six months later, the patient's telecanthus was imperceptible.

## Discussion

Paranasal sinus mucoceles, though uncommon in the pediatric population, should be considered in the differential of proptosis, telecanthus, epiphora, diplopia, and globe displacement. Many of these signs and symptoms are secondary to the progressive growth and bony remodeling into the orbital cavity, thus, it is very common for these patients to present to an ophthalmologist. Conversely, in our case, the orbital cavity was largely spared, as the mucocele grew into the nasal cavity producing only telecanthus. Moreover, a pediatric patient presenting with signs and symptoms suggestive of a mucocele should also be evaluated for inflammatory processes (such as polyposis and chronic sinusitis) and also, congenital, benign and malignant tumors (such as dermoids, meningoceles, gliomas, lymphangiomas, angiofibromas, Ewing's sarcoma, rhabdomyosarcoma, neuroblastoma, adenocarcinoma, and metastases) [3].

Within the work-up of patients with suspected mucoceles, CT-scan is the preferred technique as it provides the surgeon with necessary bony details for preoperative planning. On CT-scan, mucoceles appear as round homogenous masses with bony erosions. Alternatively, MRI provides useful information to rule out other pathological soft tissue masses but lacks the bony detail that the CT-scan provides [2]. Additionally, since cystic fibrosis is by far the most common association between mucoceles and pediatric patients, it is advisable to conduct a sweat test [3]. Notably, there have been many cases described by Hartley and Lund involving a pediatric patient with an idiopathic

mucocele such as in our case [6]. It is still unclear why mucoceles can occur in otherwise healthy children.

Surgical management of sinus mucoceles is almost universally performed through an endoscopy as opposed to an external approach allowing for a quicker recovery, minimal bleeding, and better cosmetic result. The objective of the surgery is adequate drainage with complete marsupialization of the mucocele. Complications of this approach are rare but include damage to intraorbital structures and penetration of the intracranial cavity resulting in cerebrospinal fluid leak. The reported recurrence rates for both children and adults are close to negligible. If they do occur, then they happen from two to eighteen years after endoscopic surgery [2].

In summary, paranasal sinus mucoceles are more common in adults than children. When they do occur in the pediatric population, an ophthalmologist should consider risk factors such as impaired secretion processes, pathological pneumatisation processes, atopy, trauma, and previous surgeries [4]. Of particular note, the slowly progressive growth of paranasal sinus mucoceles can cause bony remodeling and mass effects creating a variety of signs and symptoms, including an isolated finding of telecanthus.

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