

# Bilateral Isolated Congenital Microblepharon: A Rare Entity and Review of Literature

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## Abstract

Microblepharon is an extremely rare form of congenital anomaly of the eyelids defined as vertical shortening of anterior lamellae of the eyelids. Fuch's had defined the 'height' of upper lid as the distance from the margin of the upper lid to the centre of the brow when the eyes are closed, and 'length' as the same measurement when the lid is stretched maximally. The ratio of lid length to height normally exceeds 1.5 and any value less than this is taken as microblepharon. Microblepharon is usually associated with other congenital anomalies, as described in many literatures. Isolated microblepharon is extremely rare and exhaustive literature search revealed only one such previously reported case. We report a case of bilateral isolated congenital microblepharon, and the first of its kind which required no surgical intervention.

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**Keywords:** Microblepharon, Eyelid shortening, Isolated Microblepharon

## Introduction

Microblepharon is a rare congenital anomaly characterised by vertical shortening of anterior lamellae of the eyelids.<sup>1</sup> The condition is usually associated with other ocular as well as systemic abnormalities. Isolated microblepharon is an extremely rare situation and after an extensive literature search through databases of Pubmed, ScienceDirect, Google Scholar and Scopus we were able to find only one case prior to this. Herein, we report a case of isolated bilateral congenital microblepharon.

## Case Report

A 17-year-old female presented to the ophthalmology outpatient department with complaints of inability to close her eyelids since birth. (Figure 1) There was no history of redness, photophobia, or watering or use of glasses. She also had no systemic illnesses, any developmental delay, or any positive family history. On general examination, she was alert conscious with stable vitals and there was no gross facial dysmorphism.

On ophthalmological examination, uncorrected visual acuity in both of her eyes was 6/6 on the Snellen's chart with accurate projection of rays. The intraocular pressures were 13 mm Hg and 12 mm Hg in the right and left eye, respectively. On examination of the eyelids, bilateral upper and lower eyelid vertical shortening along with mild ectropion was noted. The upper eyelid creases were absent. On attempted closure of the eyelids, there was a lagophthalmos of 2 mm in both eyes. The Bell's phenomenon was good, and no part of the cornea was exposed at the area of lagophthalmos. The lid height or the distance from the eyebrow to the upper eyelid margin on attempted downgaze was measured to be 25 mm and the maximal lid length when measured after pulling the upper eyelid down with the help of cilia was measured to be 30 mm in both eyes. The vertical height coefficient was therefore 1.2. The rest of the ocular structures were within normal limits.

The patient was thoroughly evaluated for any syndromic associations; however, none was found. She was counselled regarding her eyelid condition and keeping in mind the likelihood of developing scars after any form of surgery, and the mild nature of the presenting condition, she was advised not to undergo any intervention and has been on follow-up ever since.

## Discussion

Microblepharon is a rare entity which was first mentioned by Cornaz in 1848 in his discussion of congenital anomalies of lids; later its clinical characteristics were described in detail



**Figure 1:** (1a). Clinical photograph of the patient showing bilateral short upper eyelids with mild ectropion.

**Figure 1:** (1b). Clinical photograph of the patient showing lagophthalmos of 2mm on attempted closure of eyelids.

by Fuchs in 1885. It was Fuchs who defined the ‘height’ of the upper lid as the distance from the margin of the upper lid to the centre of the brow when the eyes are closed, and the ‘length’ as the same measurement when the lid is stretched maximally.<sup>2</sup> This condition has been described to be associated with other congenital eyelid anomalies such as ectropion, absence of puncta, and tetrastichiasis.<sup>3,4,5</sup> The usual symptoms are irritation, foreign body sensation and minor disturbances of cornea; mainly attributed to the lagophthalmos. In severe lagophthalmos there can be extensive exposure of the cornea which can lead to keratitis, corneal ulcer and may require major surgical intervention to preserve vision.<sup>6</sup> Table 1 compares the present case with the previously documented case of isolated microblepharon. (Table 1)

Table 1: Showing comparison between the cases of isolated microblepharon		
Features	Meel et al <sup>6</sup>	Present case
Age at presentation	27 days of birth	17 year
Gender	Male	Female
Chief complaints	Inability to close both eyes since birth, redness, photophobia, discharge, and discolouration of both eyes	Inability to close both eyes completely since birth
Local examination findings	Ectropion and microblepharon of bilateral upper and lower eyelids, severe lagophthalmos, and infective keratitis of both corneas	Mild ectropion and microblepharon of both upper and lower eyelids, Mild lagophthalmos of 2 mm and good bell's phenomenon in both eyes
Systemic anomalies	None	None
Intervention	Control of keratitis followed by bilateral upper eyelid skin grafting with tarsorrhaphy.	None
Further Plan	To open up the tarsorrhaphy after correcting the iatrogenic entropion over 6-12 months	Regular follow up

The complete absence of eyelid is called ‘ablepharon’, which is also quite infrequent with limited reports in the literature. Microblepharon involving only one eye has also been reported.<sup>7</sup>

Microblepharon has been found to be associated with many complex congenital disorders and defects. Friderici et al. described a case of microblepharon in a stillborn and Gallenga et al. has documented associated anomalies of limbs, genitals, face, and skull.<sup>8,9</sup> Waardenburg et al has reported associated ocular abnormalities in cases of microblepharon, namely lid colobomas, microphthalmos. He has also described microblepharon with oral ocular clefts

and in an unusual case of twin monster.<sup>10</sup> There are reports in infants with ectropion and bilateral aphakic microphthalmos and in children with “ablepharon macrostomia” syndrome.<sup>11,12</sup> Merriem et al has also described microblepharon with multiple congenital anomalies in a child of a cocaine addict mother.<sup>13</sup> It has also been attributed to chromosomal anomalies like trisomy 21 and trisomy 12p.<sup>14</sup>

The route to understanding microblepharon demands knowledge of the normal embryological development of the eyelids. The lower lids develop from the maxillary processes and the upper lids from the frontonasal prominence.<sup>15</sup> By day 45, both upper and lower lid folds are appreciable, they develop from the proliferating ectoderm, grow towards each other and fuse with each other at around week 9, and they separate again at around 6 months. The mesenchyme found in the lid folds is a neural crest derivative and its deficiency can give rise to this condition, microblepharon. On the other hand, the cranial neural crest cells contribute to formation of other structures of the eye as well as to tissues of face, lips, palate and bones of orbit, maxilla, and palate. Any damage to these cells may explain the associated ocular and facial deformities.<sup>16</sup>

The treatment of microblepharon depends mainly on its severity. If lagophthalmos is mild, and the cornea is well protected by adequate Bell’s reflex, it may require no treatment at all like in our case, or can be managed on lubricating eye drops and ointment. However if corneal exposure is significant leading to visual disturbances, early surgical interventions are needed. The surgical procedures mainly involve eyelid height augmentation with free, full thickness skin grafts, pedicle flaps or inflatable plastic reservoirs in mild cases.

In our case, since the patient has a milder form of isolated microblepharon with adequate Bell’s, with no other associated anomalies, thus she required no ophthalmic intervention.

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