ORIGINAL RESEARCH PAPER

Volume-8 | Issue-1 | January-2019 | PRINT ISSN No 2277 - 8179

INTERNATIONAL JOURNAL OF SCIENTIFIC RESEARCH

# MOBIUS SYNDROME WITH BILATERAL HORIZONTAL GAZE PALSY

Ophthalmology

**Dr. Vidya Lakshmanan** MS, FAICO , Sankara Eye Hospital, Varthur Main Road, Kundalahalli Gate, Bengaluru, Karnataka 560037.

**Dr. Kanika Gupta*** MS, DNB, FICO, Sankara Eye Hospital, Varthur Main Road, Kundalahalli Gate, Bengaluru, Karnataka 560037. *Corresponding Author

## ABSTRACT

Mobius is a rare syndrome with congenital bilateralnon progressive facial weakness with bilateral abduction deficit.Sixth and seventh nerve involvement is the minimum diagnostic criteria. It is commonly associated with primary or secondary brainstem atrophy. Convergence is preserved in these cases. Hereby we report the case presenting as bilateral horizontal gaze palsy with poor convergence and the use of prism glasses for the latter.

## KEYWORDS

## INTRODUCTION

Mobius syndrome is a type of Congenital cranial dysinnervation disorder characterised by congenital, non-progressive facial weakness and limited abduction of one or both eyes. It is typically attributed to agenesis of the abducens and facial cranial nuclei or nerves.

Here we are summarizing a case of a 6 year old girl (born prematurely with low birth weight) with complaints of watering both eyes(BE) since birth. Child had intellectual and speech delay. Father gave history of incomplete closure of both eyes since birth. On head to toe examination, she had flat occiput, mask like facies(figure 1), mongoloid slant, microstomia,misaligned supernumerary teeth (figure 2), high arched palate and short stubby extremities. Neurological examination revealed bilateral facial nerve weakness with lagophthalmos. Bells phenomenon was good(figure 3). Corneal sensations were normal.Visual acuity was difficult to assess because of poor intellect and speech difficulty.



**Fig 1.Mask like facies**



**Fig 2.Microstomia and supernumerary teeth**

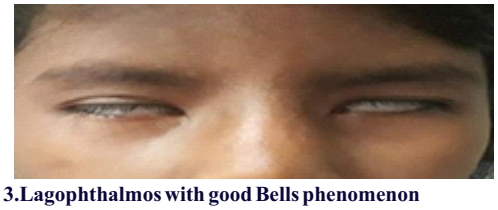


**Fig 3.Lagophthalmos with good Bells phenomenon**

Extraocular movements (EOM) revealed limitation of abduction and adduction of -4. Horizontal saccades and pursuits were absent. Vertical movements were normal in BE. Convergence was absent ( fig 4)



**Fig 4 . Bilateral horizontal movement deficit with preserved vertical movement**

Hirshberg's test revealed right eye hypertropia. Anterior segment was normal with normal pupillary reactions. Posterior segment was normal. Cycloplegic refraction showed myopic astigmatism of -2.0D in both eyes.

Child underwent MRI brain scanning which showed flattening of anterior aspect of left half of medulla and lower part of pons (fig 5)

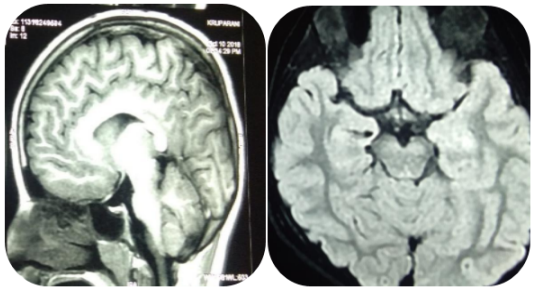


**Fig 5.Flattening of anterior aspect of left half of medulla and lower part of pons**

The child was diagnosed to have Mobius syndrome with Bilateral Horizontal gaze palsy.

## DISCUSSION

Paul Julius Mobius, a German neurologist, first described the clinical entity of bilateral combined palsies of the 6th and the 7th cranial nerves.

Abramson et al actually classified and graded the syndrome on the basis of the clinical findings of cranial nerve palsies and musculoskeletal anomalies by using the acronym CLUFT (*c*ranial nerve, *l*ower limb, *u*pper limb, *f*ace, and *t*horax)[1]

In another extensive review of 37 patients (largest series of Mobius patients ), authors observed a uniform clinical picture characterized by facial diplegia of the upper and lower facial muscles, bilateral eye abduction impairment, hypoglossia, craniofacial and limb

malformations, and long tract symptoms.[2]

According tostudies done by Miler et al[3], Cromnemberger et al[4] , Amaya et al[5], Santos et al[6].and Henderson[7], the most common strabismus in Moebius syndrome is esotropia (33.3%-77.8%) followed by orthotropia (11.1%-48.9%) and exotropia (3.6%-12.5%). Limitations of abduction (73.8%-100%) was more commonly seen than adduction (45.5%-95.8%).[3-7]

Rucker et al studied ocular motor findings of 40 subjects (23 months to 64 years; 24 females, 16 males) with congenital facial weakness.The most common ocular motor pattern was bilateral horizontal gaze palsy(BHGP) with intact vertical rangefrom pontine abducens nuclear defects, rather than abducens nerve involvement.[8]

Convergenceis uniquely known to remain intact in the presence of abducens nuclear lesions causing BHGP in contrast to our case where convergence was involved.

MRI findings in mobius syndrome from previous studies showed brainstem hypoplasia with flattening of fourth ventricle floor Involving facial colliculus and 6th nerve nucleus which was normal in our case.

Our case had unique presentation of bilateral facial diplegia with bilateral horizontal gaze palsy with hypertropia in primary gaze and convergence insufficiency with brainstem hypoplasia involving anterior half of pons and medulla which has not been reported before. In addition we emphasize the importance of correction of refractive error to prevent amblyopia and base in prism to aid in convergence.

## REFERENCES


1. Abramson DL, Cohen M, Mulliken JB. Mo¨bius syndrome: classi-fication and grading system. PlastReconstrSurg1998;102:961–967
2. Verzijl HT, van der Zwaag B, Cruysberg JR, Padberg GW. Mobiussyndrome redefined: a syndrome of rhombencephalicmaldevelop-ment. Neurology2003;61:327–333
3. Miller MT, Stromland K. The mobius sequence: a relook. J AAPOS. 1999 ;3(4):199-208.
4. Cronemberger MF, de Castro Moreira JB, Brunoni D, Mendonça TS, AlvarengaEH, Rizzo AM, Diogo SM. Ocular and clinical manifestations ofMobius' syndrome. J PediatrOphthalmolStrabismus.2001;38(3):156-62.
5. Amaya LG, Walker J, Taylor D. Möbius syndrome: a study and report of 18cases. Binocul Vis Q.1990;5:119-32.
6. Santos LPF, Ventura LMVO, Almeida HC, Miller M, Colier AC.Achadoso ftalmo lógicosem 28 criançasportadoras da seqüência de Möebius. Arq BrasOftalmol. 2004; 67(4) :591-5.
7. Henderson JL. The congenital facial diplegia syndrome: clinical features, pathologyand aetiology: a review of sixty-one cases. Brain. 1939;62(4):381-403.
8. Janet C. RuckerBryn D. WebbTamieshaFrempong, Harald Gaspar,Thomas P. Naidic and Ethylin Wang Jab .Characterization of ocular motor deficits incongenital facial weakness: Moebius andrelated syndromes|.Brain 2014: 137; 1068–1079