

# Prevalence of refractive errors in Möbius sequence

## Prevalência de erros refrativos na sequência de Möbius

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### ABSTRACT

**Purpose:** To assess the prevalence of refractive errors in Möbius sequence.

**Methods:** This study was carried out during the Annual Meeting of the Brazilian Möbius Society in November 2008. Forty-four patients diagnosed with the Möbius sequence were submitted to a comprehensive assessment, on the following specialties: ophthalmology, neurology, genetics, psychiatry, psychology and dentistry. Forty-three patients were cooperative and able to undertake the ophthalmological examination. Twenty-two (51.2%) were male and 21 (48.8%) were female. The average age was 8.3 years (from 2 to 17 years). The visual acuity was evaluated using a retro-illuminated logMAR chart in cooperative patients. All children were submitted to exams on ocular motility, cycloplegic refraction, and fundus examination.

**Results:** From the total of 85 eyes, using the spherical equivalent, the major of the eyes (57.6%) were emmetropics (>-0.50 D and <+2.00 D). The prevalence of astigmatism greater than or equal to 0.75 D was 40%.

**Conclusion:** The prevalence of refractive errors, by the spherical equivalent, was 42.4% in this studied group.

**Keywords:** Möbius syndrome; Refractive errors/epidemiology; Strabismus; Anisometropia; Astigmatism

### RESUMO

**Objetivo:** Avaliar a prevalência de erros refrativos em crianças portadoras da sequência de Möbius.

**Métodos:** Trabalho realizado durante o encontro anual da Associação Möbius do Brasil (AMoB) em novembro de 2008. Quarenta e quatro pacientes com diagnóstico de sequência de Möbius foram submetidos a avaliação multidisciplinar: oftalmológica, neurológica, genética, psiquiátrica, psicológica e odontológica. Quarenta e três pacientes colaboraram com exame oftalmológico. Vinte e dois (51,2%) eram do sexo masculino e 21 (48,8%) do sexo feminino. A idade média foi de 8,3 anos (2 a 17 anos). A medida da acuidade visual foi realizada com tabela logMAR retro-iluminada, nos pacientes que colaboravam. Todas as crianças foram submetidas a exame da motilidade ocular, refração sob cicloplegia e fundo de olho.

**Resultados:** Do total de 85 olhos estudados, usando o equivalente esférico, a maioria dos olhos (57,6%) são emétopes (>-0,50 D e <+2,00 D). A prevalência de astigmatismo maior que 0,75 D foi 40%.

**Conclusão:** A prevalência de erros refrativos, pelo equivalente esférico, no grupo estudado foi de 42,4%.

**Descritores:** Síndrome de Möbius; Erros de refração/epidemiologia; Estrabismo; Anisometropia; Astigmatismo

### INTRODUCTION

The Möbius syndrome was first reported in 1881<sup>(1)</sup>, when Von Graefe described one of his patients as having congenital facial diplegia<sup>(2)</sup>.

In 1888, Möbius traced a relationship between congenital facial diplegia and other malformations. He also described the classical signs of this syndrome: absence of abduction in both eyes, along with the deficiency of other cranial nerves (V, IX and XII especially)<sup>(3)</sup>.

The Möbius syndrome designation has recently been replaced by Möbius sequence. A sequence is thought to represent a pattern of multiple anomalies derived from a single structural defect or mechanical factor, usually due to multiple etiologies compared with the designation "syndrome" which implies a single cause<sup>(4,5)</sup>.

In a recent publication, Souza-Dias and Goldchmit have stated that the variation in ocular motility in Möbius sequence patients is, in reality, horizontal conjugate eye movement paralysis<sup>(6)</sup>. Its patho-

genesis is unclear, but it seems to be related to an embryonic insult from heterogeneous causes, which affects the developing cranial nerve nuclei<sup>(7,8)</sup>.

Many ophthalmic disorders in Möbius sequence patients have been described, but we only found two studies in the literature regarding refractive errors in such patients<sup>(9,10)</sup>. Uncorrected refractive errors can interfere in school performance, reduce employability and economic productivity, and generally impair quality of life<sup>(11)</sup>. Therefore, the main purpose of this study was to assess the prevalence of refractive errors in a group of patients with Möbius sequence.

### METHODS

This is a multidisciplinary and collaborative study, approved by the Ethics Committee of the Human Research of the Santa Casa de Misericórdia de São Paulo Hospital (400/08).

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Patients were referred from the following institutions: Department of Ophthalmology, Federal University of São Paulo - UNIFESP; Department of Ophthalmology, São Paulo University - USP; Department of Ophthalmology, Santa Casa Medical School; Disable Children Care Association and from Brazilian Möbius Society.

This is a multidisciplinary study, approved by the Ethics Committee of the Human Research of the Santa Casa de Misericórdia de São Paulo Hospital (400/08).

**Table 1. Visual acuity (VA), cycloplegic refraction, type of ocular deviation (D), presence of anisometropia (Anis.) in patients with the Möbius sequence**

P #		VA		Cycloplegic refraction		D	Anis.
		OD	OS	OD	OS		
1	Nc	0.80	0.63	+2.50 -0.75 x 180°	+3.00 -2.50 180°	Ortho	
2	Wc	0.20	LP	+6.50 -2.50 x 180°	RD	Ortho	
3	Nc	0.80	0.80	+0.75	+0.75	ET	
4	Wc	1.00	1.00	+2.50 -1.50 x 15°	+2.75 -0.25 x 165°	ET	
5	Nc	Ni	Ni	+2.50 -1.50 x 180°	+3.00 -2.00 x 180°	Ortho	
6	Nc	0.40	0.40	-1.00	-1.00	ET	
7	Nc	Ni	Ni	+0.50	+0.75	Ortho	
8	Wc	0.80	0.63	+2.50	+6.00 -1.00 C x 90°	ET	x
9	Wc	0.80	0.80	+3.00 -1.00 x 180°	+2.75 -1.00 x 180°	Ortho	
10	Wc	0.25	0.40	+4.00	+6.00 -1.50 x 180°	XT	x
11	Nc	0.50	0.50	+1.50	+1.50	Ortho	
12	Nc	0.50	0.63	-2.00 x 90°	-2.00 x 90°	XT	
13	Wc	0.50	0.50	-2.25 -1.50 x 180°	-5.25 -5.25 x 180°	Ortho	x
14	Wc	Ni	Ni	-10.00 -2.00 x 30°	-7.00	ET	x
15	Nc	0.32	0.32	+4.00 -2.25 x 180°	+4.50 -2.50 x 180°	ET	
16	Nc	0.63	0.50	+1.00	-1.50	ET	x
17	Nc	Ni	Ni	+0.50	+0.50	Ortho	
18	Nc	0.50	0.32	+1.00	+1.00	ET	
19	Nc	0.20	0.25	+4.00 -2.50 x 160°	+3.75 -3.25 x 20°	ET	
20	Nc	0.50	0.50	+0.50	+0.50	Ortho	
21	Nc	0.40	0.50	+2.75 -0.50 x 90°	+1.25	Ortho	x
22	Wc	0.32	0.50	-7.50 -2.75 x 180°	-6.00 -0.75 x 180°	ET	x
23	Wc	0.80	0.63	+2.50 -2.00 x 120°	+2.50 -2.00 x 20°	ET	
24	Nc	Ni	Ni	+0.75 -1.00 x 180°	+1.50	ET	x
25	Nc	0.40	0.32	+1.00	+1.00	ET	
26	Wc	0.50	0.80	+1.50 -1.50 x 30°	+0.75 -1.00 x 100°	Ortho	
27	Wc	0.63	0.80	+1.50 -0.50 x 180°	+2.00 -0.75 x 180°	ET	
28	Nc	0.63	0.63	+0.75	+1.00	Ortho	
29	Nc	0.90	0.90	Plano	-0.50 x 180°	XT	
30	Nc	0.50	0.32	+1.50 -0.50 x 180°	+1.50	Ortho	
31	Wc	cf	0.63	+4.50 -5.25 x 175°	+4.75 -3.25 x 10°	Ortho	x
32	Wc	0.50	0.60	+3.00	+3.00	Ortho	
33	Nc	Ni	Ni	+4.50	+4.50	ET	
34	Nc	Ni	Ni	+1.75	+1.50	ET	
35	Nc	Ni	Ni	+0.75	+0.75 -0.50 x 90°	Ortho	
36	Nc	0.32	0.32	-2.00	-2.25	ET	
37	Wc	0.32	0.50	+4.00	+4.00	Ortho	
38	Nc	0.32	0.32	+1.50	+0.50	ET	
39	Nc	1.00	1.00	+1.75 -1.00 x 180°	+1.75 -1.00 x 180°	Ortho	
40	Nc	0.40	0.40	-0.50	+0.25 -0.75 x 90°	ET	
41	Nc	0.80	0.80	Plano	Plano	Ortho	
42	Nc	Ni	Ni	+1.50	+1.50	Ortho	
43	Wc	0.30	0.50	+1.50 -2.75 x 10°	+1.50 -1.50 x 170°	Ortho	

OD= right eye; OS= left eye; Nc= no correction; Wc= with correction; Ni= not informed; Cf= counting fingers; LP= light perception; RD= retinal detachment; Ortho= orthotropia; ET= esotropia; XT= exotropia.

This study was carried out during the Annual Meeting of the Brazilian Möbius Society on November 2008, in the Department of Ophthalmology of Santa Casa de Misericórdia de São Paulo Hospital. Forty-four patients diagnosed with Möbius sequence were examined by a multidisciplinary group: ophthalmology, neurology, genetics, psychiatry, psychology and dentistry

A cross-sectional transversal study was performed on 43 cooperative patients to undertake the ophthalmic exam. Of these 43 patients, 22 (51.2 %) were male and 21 (48.8 %) were female. The average age was 8.3 years (2 to 17 years).

The visual acuity was measured each eye separately. The patients were tested with the retro-illuminated logMAR chart.

Measurement of ocular deviation was determined by cover test or a Krimsky test for uncooperative patients.

Retinoscopy was performed with loose trial lenses and cycloplegia was obtained using 1 drop of tropicamide 1% (Alcon), 1 drop of cyclopentolate 1% (Allergan), after anesthesia induced by proxymetacaine chloride 0.5% (Alcon). The refractometric examination was performed 40 minutes after the instillation of the last eye drops.

Prevalence of myopia spherical equivalent of -0.50 diopters or less and of hyperopia spherical equivalent of +2.00 diopters or greater were calculated. Emmetropia was considered greater than -0.50 spherical equivalent diopters or less than +2.00 spherical equivalent diopters. The prevalence of astigmatism was assessed at two levels:  $\geq 0.75$  to  $< 2.0$  cylinder diopters, and  $\geq 2.0$  cylinder diopters<sup>(12,13)</sup>.

The fundus exam was performed by an indirect ophthalmoscopy, soon after the refraction exam.

## RESULTS

Table 1 shows the data of the 43 patients who participated in the exam, with their respective results of visual acuity, cycloplegic retinoscopy of both eyes, and type of ocular deviation. The last column in table 1 shows the patients who had anisometropia. Out of the 43 patients, 9 (20.9%) were anisometropic.

Myopia was defined as spherical equivalent (SE) refractive error of at least -0.50 D in 13 (15.3%) eyes. The average myopia was -3.69 D (from -0.50 D to -11.0 D). Hyperopia as +2.00 D or more in 23 (27.1%) eyes. The average hyperopia was +3.47 D (from +2.00 D to +6.37 D). Forty-nine (57.6%) eyes were emmetropics ( $> -0.50$  D and  $< +2.00$  D), average +0.95 D (from -0.25 D to +1.87 D). The prevalence of astigmatism greater than or equal to 0.75 to less than 2 cylinder diopters was 17 (20%) and greater than or equal to 2 diopters was 17 (20%).

The fundus exam showed that 3 patients had an increased cup-to-disc ratio in both eyes (OU).

## DISCUSSION

Refractive error can place a substantial burden on the individual. School-age children constitute a particularly vulnerable group, because uncorrected refractive error may have a dramatic impact on learning capability and education potential<sup>(13)</sup>. Refractive error as a cause of blindness has been recognized only recently with the increasing use of presenting visual acuity for defining blindness<sup>(14)</sup>.

Reviewing the literature it was found only two studies on refractive errors related to Möbius sequence. Some authors<sup>(10)</sup>, in a group of 28 patients, the most frequent refractive error was astigmatism, considering hyperopic astigmatism, myopic astigmatism and mixed astigmatism, diagnosed in 33 eyes (58.9%) out of a total of 56, followed by hyperopia in 33.9% eyes. Other authors<sup>(9)</sup> found compound

hyperopic astigmatism to be the main refractive error (40.6%) in a series of 16 patients. Mean spherical equivalent was  $+1.90 \pm 2.49$  (median= $+2.00$ ). In our study, using spherical equivalent, most of the patients (57.6%) were emmetropic ( $> -0.50$  D and  $< +2.00$  D). The prevalence of myopia (at least -0.50 D) was 15.3% eyes and hyperopia as +2.00 D or more in 27.1% eyes. The prevalence of astigmatism greater than or equal to 0.75 D was 40%.

Uncorrected refractive error is recognized as the major cause of avoidable visual impairment in worldwide population, regardless age, sex or ethnicity. For children, there may be severe consequences such as delayed neuropsychomotor development, learning disabilities and special needs education in more severe cases. In a long term, it may result in a burden for the country and society<sup>(11)</sup>.

Furthermore, optical correction for refractive error is the most cost-effective intervention in eye health care<sup>(11)</sup>, due to easy detection, diagnosis and the correction with spectacles.

Especially for those patients in this study and considering their many individual limitations, detection and appropriated optical correction could improve their visual acuity and their daily activities and school performance, improving their quality of life.

## CONCLUSION

We conclude that the prevalence of refractive errors, by the spherical equivalent, was 42.4% in this studied group.

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