



Orofacial function of persons having Möbius syndrome

Report from observation charts

The survey comprises 35 observation charts.

Synonyms: Möbius sequence, Moebius syndrome.

Codes:

ICD-10: Q87.0W

ORPHA: 570

Estimated occurrence: Very rare.

Etiology: Most cases are sporadic without a known etiology. The diagnosis is probably caused by an early fetal damage affecting the development of the cranial nerves.

General symptoms: The primary symptoms are congenital facial palsy (affecting the facial muscles) and abducens palsy (affecting the possibility of lateral eye movements). These palsies usually occur on both sides. Other cranial nerves may be affected, generally the hypoglossal nerve (controls tongue mobility), the glossopharyngeal/vagus nerves (important for swallowing and speech) and/or the trigeminal nerve (controls the muscles involved in chewing and the sensation of the face and mouth). Skeletal anomalies may occur. Some individuals with Möbius syndrome have a neuropsychiatric disorder and intellectual impairment.

Orofacial/odontological symptoms:

Facial palsy results in little or no facial expression. Many of these children have sucking, eating and speech difficulties owing to underdevelopment of the cranial nerves and craniofacial deformities including cleft palate and/or microglossia (underdeveloped tongue). There may be drooling problems. A post normal bite a small lower jaw and crowded teeth are all relatively common, as is the absence of some tooth buds. Studies have shown, that the saliva production may be reduced and some have a modified buffering capacity.

Orofacial/ odontological treatment:

- Early contact with dental services for intensified prophylactic care and oral hygiene information is essential.
- In cases of craniofacial deformities, a specialist team will be responsible for follow up and treatment.
- Regular check-ups of dental and jaw development. Orthodontist should be consulted when needed.
- Orofacial therapy and oral motor skill training should be considered.
- Speech, language and communication training are often justified.
- Feeding and swallowing difficulties are investigated and treated by a specialist team at the hospital or multidisciplinary treatment center.

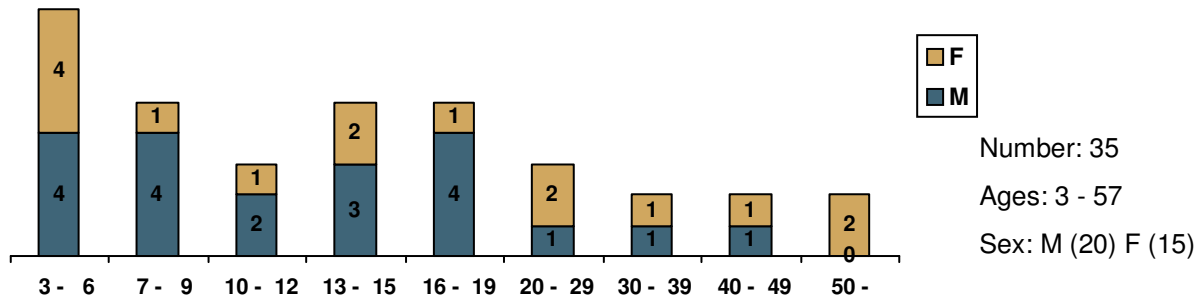
Sources

The rare disease database of the Swedish National Board of Health and Welfare.

The MHC database - The Mun-H-Center database on oral health and orofacial function in rare diseases.

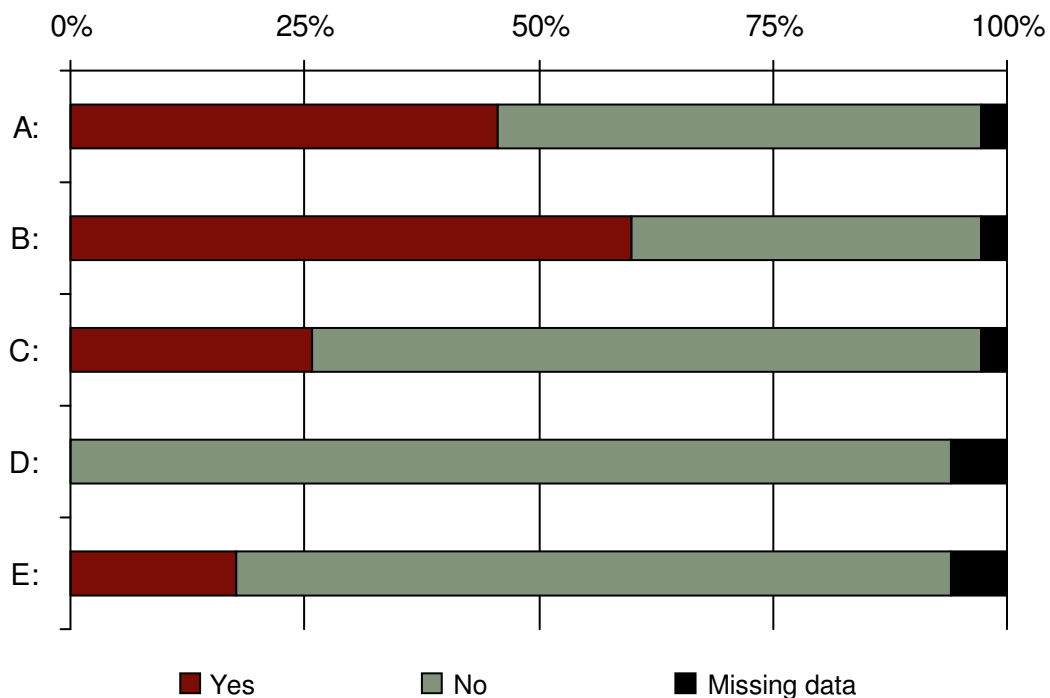
The Documentation from the Ågrenska Center.

Age distribution



Overview

	Yes	No	Missing data	N
A: Incomprehensible speech/No speech	16	18	1	35
B: Eating and drinking difficulties ¹	21	13	1	35
C: Profuse drooling, on clothes ¹	9	25	1	35
D: Breathing difficulties ^{1 2}	0	16	1	17
E: Grinding every day ^{1 2}	3	13	1	17



Note that the diagram is based upon less than 100 individuals.

1: Compiled using questionnaire

2: This variable was introduced in version 2 (2008) of the Observation chart.

Oral health

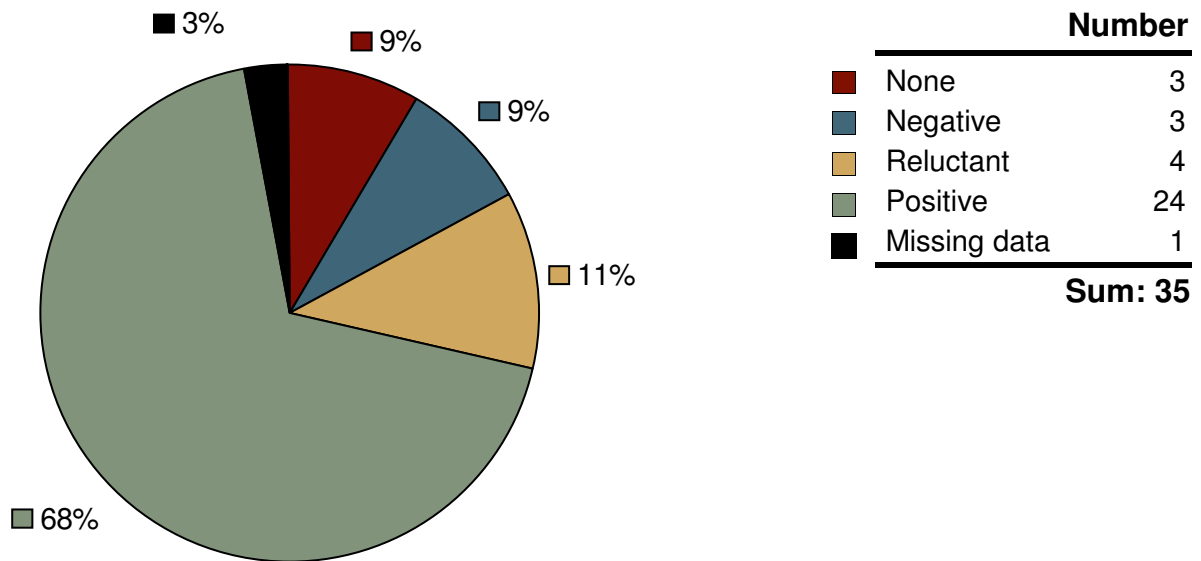
Oral health index (indices)¹

		0	1	2	3	4	5	6	Missing data	N
Calc	Calculus	12	2	0	0	0	0	0	3	17
GI	Gingivitis	6	2	1	2	2	0	0	4	17
Plaq	Coating	5	1	1	3	1	0	2	4	17
Toot	Tooth wear	8	4	1	0				4	17

- C Calculus index is based on the presence of visible calculus on the buccal surface of 6 index teeth. 0 indicates that there is no calculus at all, 6 indicates calculus on all index teeth.
- GI Gingivitis index is based on the presence of visible gingivitis on the buccal surface of 6 index teeth. 0 indicates that there is no bleeding, 6 indicates bleeding on all index teeth.
- PI Plaque index is based on the presence of visible plaque on the buccal surface of 6 index teeth. 0 indicates that there is no plaque, 6 indicates plaque on all index teeth.
- To Tooth wear index is a weighted summary of the degree of tooth wear on 6 different segments. Tooth wear is only evaluated in the permanent dentition, not in the primary teeth. The final index score is based on the degree of tooth wear found in most segments.
- 0: No tooth wear or minor wear of enamel in either of the segments
- 1: Marked tooth wear of the enamel, possibly exceeding into dentin
- 2: tooth wear in the dentine reaching up to 1/3 of the tooth crown
- 3: Tooth wear in the dentine reaching up to more than 1/3 of the tooth crown. If 3 is given in any segment then SI is 3.

¹: Oral health index (indices) was (were) introduced in the observations in 2008

Acceptance of dental examination



Caries

	3-6 years	7-12 years	13-19 years	Adults
deft¹				
Examined	6	6		
Number of individuals with deft=0	5	6		
Mean	1,3	0,0		
Standard deviation	3,0	0,0		
Missing data	2	2		
DMFT²				
Examined		7	10	8
Number of individuals with DMFT=0		7	2	0
Standard deviation		0,0	4,4	6,1
Mean		0,0	4,3	13,0
Missing data		1	0	1

1: Number of carious or filled deciduous teeth

2: Number of carious or filled permanent teeth

Occlusal relationship

	Number
Neutral bite	24
Post normal	9
Pre normal	1
Missing data	1
Sum: 35	

Maximum jaw opening

Children younger than 10 years

	Number
- 20	0
21 - 30	1
31 - 40	7
41 - 50	2
51 -	0
Missing data	3
Sum: 13	

Children, 10 years or older, and adults

	Number
- 20	1
21 - 30	4
31 - 40	6
41 - 50	7
51 -	1
Missing data	3
Sum: 22	

Profile¹

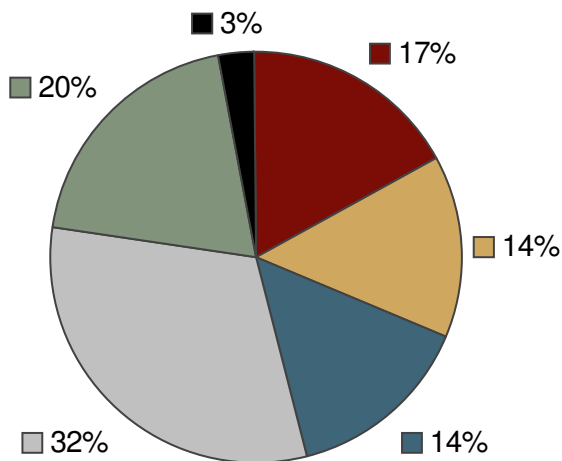
	Number
Normal	11
Convex	1
Concave	0
Missing data	5
Sum: 17	

Mandibular plane¹

	Number
Normal	11
Increased	1
Reduced	0
Missing data	5
Sum: 17	

1: This variable was introduced in version 2 (2008) of the Observation chart.

Speech difficulty



	Number
No speech	6
Very incomprehensible	5
Incomprehensible speech	5
Slightly indistinct speech	11
No problems	7
Missing data	1
Sum: 35	

Clinical findings	Yes-answers			
	Total N=35 (%)	Boys/Men N=20 (%)	Girls/Women N=15 (%)	Missing data
Facial palsy	34 (97)	20 (100)	14 (93)	0
Impaired tongue motility	25 (71)	14 (70)	11 (73)	0
Low muscle tone in lips	25 (74)	15 (79)	10 (67)	1
Open mouth at rest	20 (59)	12 (63)	8 (53)	1
Low muscle tone in tongue	17 (49)	9 (45)	8 (53)	0
Asymmetric tongue	16 (57)	7 (41)	9 (82)	7
Over crowding	12 (35)	8 (42)	4 (27)	1
Facial asymmetry	12 (36)	7 (35)	5 (38)	2
Reduced opening capacity	11 (34)	6 (32)	5 (38)	3
M mentalis overactive	10 (29)	7 (37)	3 (20)	1
High palate	10 (29)	6 (32)	4 (27)	1
Narrow palate	8 (23)	7 (35)	1 (7)	0
Cleft lip and palate	6 (18)	5 (26)	1 (7)	2
Low muscle tone in masticatory muscles	5 (15)	3 (16)	2 (14)	2
Cranio-facial abnormality	5 (14)	4 (20)	1 (7)	0
Microglossia	4 (12)	3 (16)	1 (7)	2
Short tongue frenulum	3 (10)	3 (17)	0 (0)	4
Gingival hyperplasia	3 (9)	3 (16)	0 (0)	1
Reduced stability in neck	3 (9)	2 (10)	1 (7)	1
High muscle tone in masticatory muscles	3 (9)	1 (5)	2 (14)	2