



Orofacial function of persons having Möbius syndrome

Report from questionnaires

The survey comprises 16 questionnaires.

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.

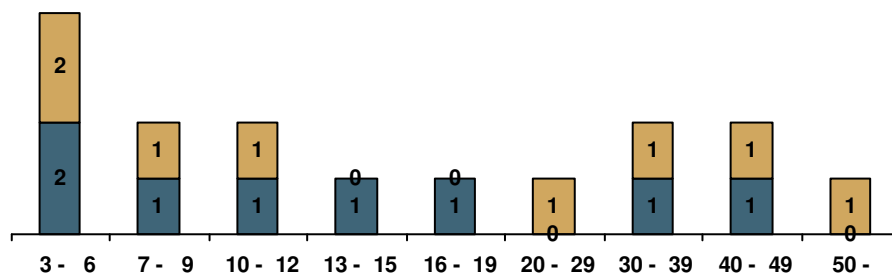
Sources

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



Number: 16

Ages: 3 - 57

Sex: M (8) F (8)

Additional diagnoses

Medical impairment	Yes	No	Missing data
Inborn heart defect	0	14	2
Other cardiovascular disease	0	14	2
Epilepsy	1	13	2
Asthma	2	13	1
Need of respiratory support	2	12	2
Allergy	2	12	2

Neuropsychiatric diagnosis	Yes	No	Missing data
ADHD/ADD	1	13	2
Autistic syndrome	0	14	2
Autistic traits	3	10	3
Asperger's syndrome	1	13	2
Tourette's syndrome	0	13	3

General disability	Yes	No	Missing data
Intellectual disability	0	12	4
Mobility dysfunction	3	7	6
Impaired vision	6	1	9
Impaired hearing	2	8	6
Difficulty in communicating	3	8	5

About dental care and oral health

Do you feel that you receive the dental care you need?

	Number
Yes, very much so	10
Yes, some what	5
No, not really	0
No, not at all	0
Missing data	1
Sum:	16

How many times per year do you normally seek dental care?

	Number
Three or more times per year	5
Twice per year	3
Once per year	4
Less than once per year	4
Missing data	0
Sum:	16

When were your teeth last X-rayed?

	Number
During the past two years	13
More than two years ago	1
Never had my teeth X-rayed	2
Missing data	0
Sum:	16

Do you look after your teeth in a good way?

	Number
Yes, very much so	7
Yes, some what	7
No, not really	0
No, not at all	0
Missing data	2
Sum:	16

Who brushes your teeth?

	Number
I always brush myself	9
Sometimes I brush myself	3
Someone else always helps me	4
Missing data	0
Sum:	16

How often are your teeth brushed?

	Number
Three or more times per day	2
Twice per day	12
Once per day	2
Not every day	0
Missing data	0
Sum:	16

About dental care and oral health

	Yes	No	Missing data
Does your mouth hurt?	0	15	1
Does your mouth feel dry?	4	8	4
Have you ever taken a serious hit to your permanent front teeth?	2	14	0
Do you feel that you have a divergent bite?	5	10	1
Have you had a brace?	3	13	0
Do you feel that you need orthodontics/a brace?	3	11	2

Do you grind or press your teeth at night?

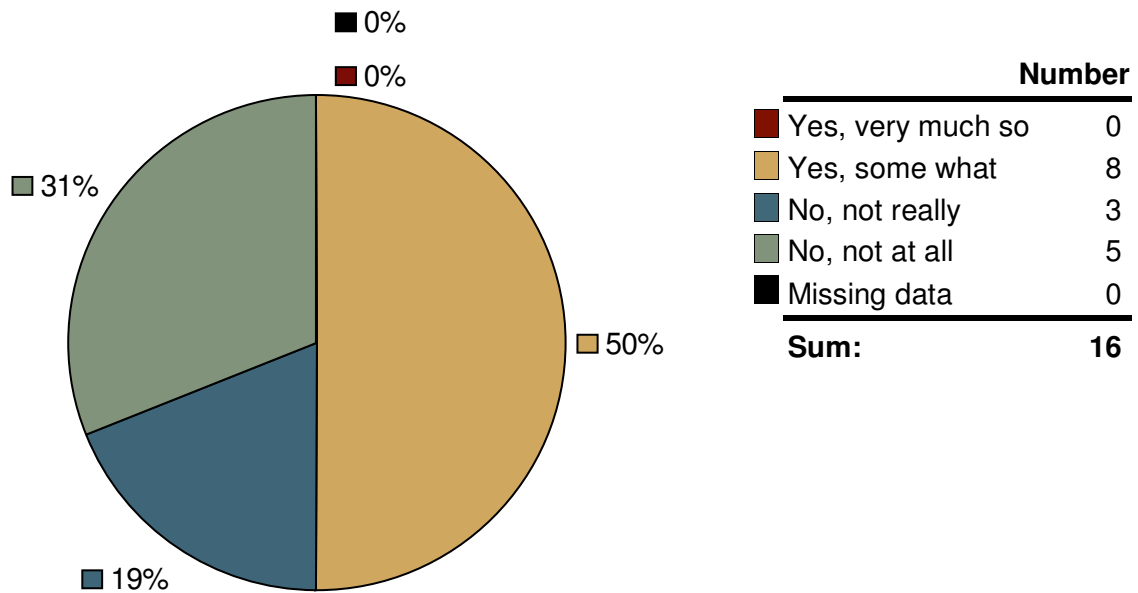
	Number
Never	7
Once or twice per week	3
Every night	3
Missing data	3
Sum:	16

Do you grind or press your teeth during the day?

	Number
Never	8
Once or twice per week	5
Every day	2
Missing data	1
Sum:	16

About eating

Do you have any problems with eating?



	Number
Yes, very much so	0
Yes, some what	8
No, not really	3
No, not at all	5
Missing data	0
Sum:	16

	Yes	No	Missing data
Do you cough daily in connection with meals?	1	15	0
Do you gag daily in connection with meals?	1	15	0
Do you get acid reflux daily?	1	14	1
Do you throw up often (at least twice per week)?	0	16	0
Do you have a poor appetite?	1	15	0
Does it take a long time before you can swallow a mouthful?	3	12	1
Do you press your tongue forward when you swallow so that food ends up outside the mouth?	0	11	5
Do you find it difficult to chew, i.e. grind food using your molars?	5	10	1
Do you find it difficult to take food from the spoon using your lips?	7	9	0
Have you had problems with food and drink leaking out through the corners of your mouth?	6	9	1
Does food tend to remain in your mouth after meals?	7	8	1
Do you get nutrition in any other way than through your mouth?	2	14	0

About drooling

Do you drool?

	Number
Never drool	6
Drool sometimes – not every day	6
Drool often – every day	3
Constant drooling	1
Missing data	0
Sum:	16

How much do you drool?

	Number
Slight drooling, only on the lips	3
Moderate drooling, on lip and chin	6
Profuse drooling, on clothes	1
Very profuse drooling, on hands/objects	0
Missing data	0
Sum:	10

Is your drooling a problem for you?

	Number
Yes, very much so	1
Yes, some what	1
No, not really	5
No, not at all	2
Missing data	1
Sum:	10

Is your drooling a problem for your family or people around you?

	Number
Yes, very much so	1
Yes, some what	1
No, not really	5
No, not at all	3
Missing data	0
Sum:	10