



Orofacial function of persons having Marfan syndrome

[Report from observation charts](#)[49 observation charts](#)

Synonym

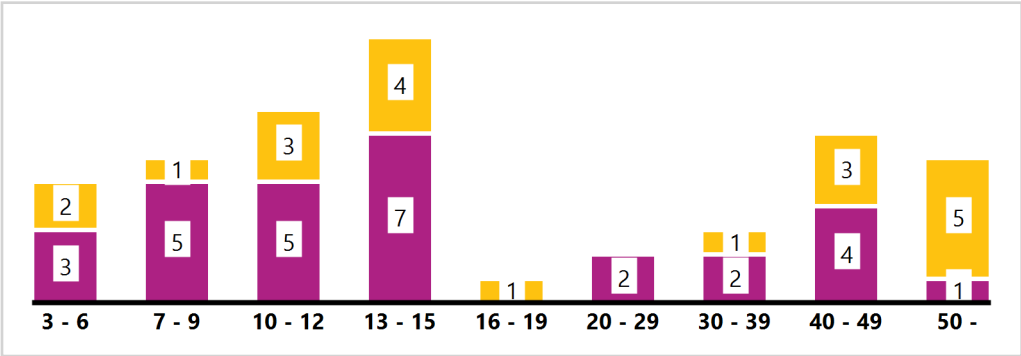
ICD-10 Q87.4**Estimated occurrence** 10-20:100,000 inhabitants.**General symptoms** Individuals with Marfan syndrome are often extremely tall. They may have problems associated with various body parts. Most common are muscular and skeletal problems, joint laxity, dilatation (widening) of aorta, mitral valve problems, vision impairment and some times pulmonary disorders.**Oral symptoms** Common symptoms include a high, sometimes narrow palate, and lack of space for teeth in the jaws. Snoring and an increased risk of sleep apnea may occur. The temporomandibular joint is often affected, and there may be instability of the joint-capsules. Tooth grinding implies extra pressure on the jaw, which may increase the joint problems. For the same reason, the patient may have difficulty opening his or her mouth wide and for a long time, as may be necessary, for example, at dental appointments.**Oral treatment**

- Extra prophylactic dental care may be relevant in order to prevent the need for dental treatment that would be very demanding for these patients because of their jaw problems. In some patients there is a risk of endocarditis if oral bacteria gain access to the blood stream. Improved oral and especially gingival health reduces this risk.
- Regular check-ups of dental and jaw development. Orthodontist should be consulted when needed.
- In cases of temporomandibular joints disorders, this should be investigated and appropriate treatment thereafter prescribed.
- Problems associated with snoring and sleep apnea should be followed up by a physician.
- When treating medically compromised patients always contact their doctors for medical advice.

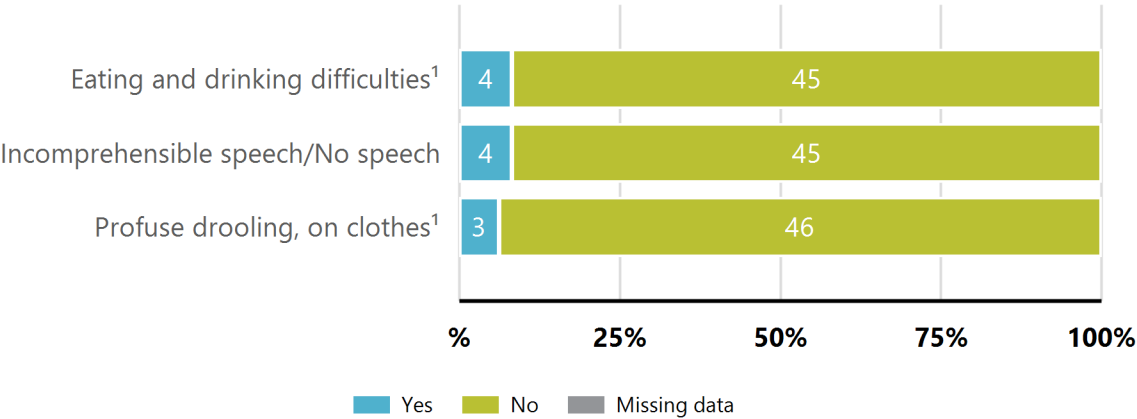
Sources The MHC database Rare diseases Dokumentation-Ågrenska

Age distribution

Count: 49
Ages: 4 - 66
♂ (29)
♀ (20)

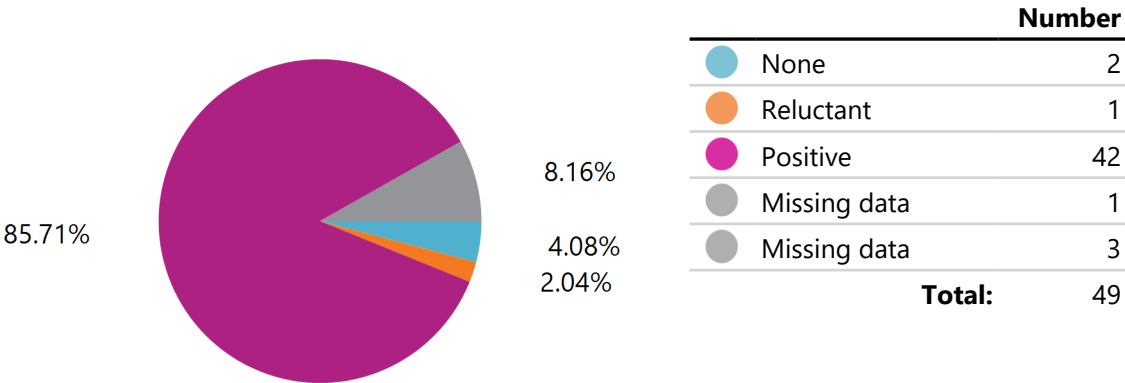


Summary



Certain caution should be observed when interpreting the chart because the number of individuals is less than 100.
¹ Reported via questionnaire

Acceptance of dental examination



Oral health

Caries

		3-6 years	7-12 years	13-19 years	>19 years
deft	Examined	5	14		
Number of carious or filled deciduous teeth	Number of individuals with deft=0	4	12		
	Mean	2.5	0.4		
	Standard deviation	3.5	0.7		
	No data	3	6		
DMFT	Examined		14	12	18
Number of carious or filled permanent teeth	Number of individuals with DMFT=0		13	6	3
	Mean		0.1	1.9	5.0
	Standard deviation		0.3	1.5	5.8
	No data		5	5	2

Oral health index (indices)¹

	0	1	2	3	4	5	6	Missing data	Number
Calculus	21	3	0	0	0	0	0	25	31
Gingivitis	23	3	0	0	0	0	0	23	31
Plaque	14	2	6	1	0	0	1	25	31
Tooth wear	16	10	1	0	0	0	0	22	31

CALCULUS

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.

GINGIVITIS

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.

PLAQUE

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.

TOOTH WEAR

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

Occlusal relationship

	Number
Neutral bite	32
Overbite	11
Underbite	5
Missing data	1
Total:	49

Maximum jaw opening

Children younger than 10 years

	Number
-20	0
21-30	0
31-40	4
41-50	5
51-	0
Missing data	2
Total:	11

Children, 10 years or older, and adults

	Number
-20	0
21-30	0
31-40	10
41-50	17
51-	7
Missing data	4
Total:	38

Profile¹

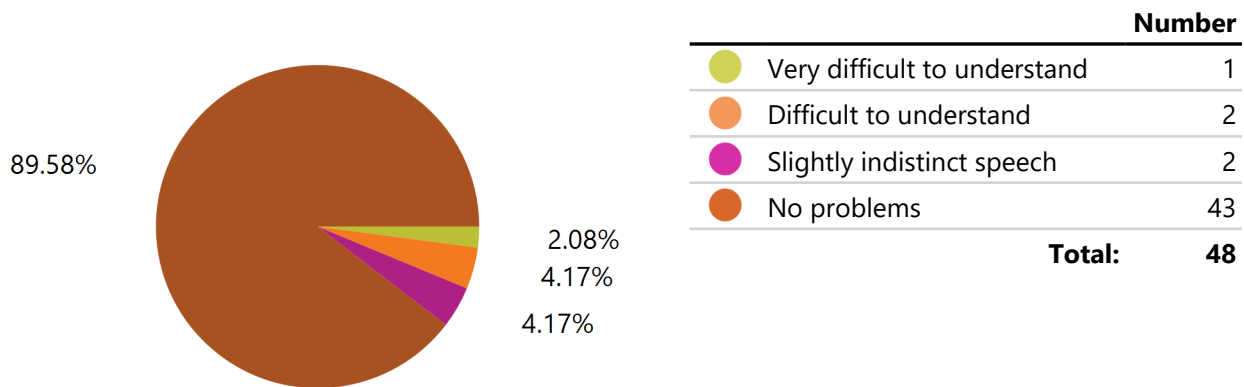
	Number
Normal	26
Convex	2
Concave	0
Missing data	1
Total:	29

Mandibular plane¹

	Number
Normal	19
Increased	4
Reduced	1
Missing data	7
Total:	31

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

Speech problems



Clinical findings

Number of yes-answers

	Total = 49 (%)	♂ = 29 (%)	♀ = 20 (%)	Missing Data
High palate	30 (61)	20 (69)	10 (50)	0
Narrow palate	19 (39)	11 (38)	8 (40)	0
Open mouth at rest	18 (37)	12 (41)	6 (30)	0
Over crowding	18 (37)	11 (38)	7 (35)	0
Low muscle tone in lips	7 (14)	6 (21)	1 (5)	0
Reduced opening capacity	7 (14)	4 (14)	3 (15)	0
Frontal open bite	4 (8)	4 (14)	0 (0)	0
Low muscle tone in masticatory muscles	4 (8)	3 (10)	1 (5)	1
Facial asymmetry	4 (8)	3 (10)	1 (5)	2
Short tongue frenulum	3 (6)	2 (7)	1 (5)	1
Impaired tongue motility	2 (4)	1 (3)	1 (5)	2
Deep bite with gingival contact	1 (2)	1 (3)	0 (0)	1

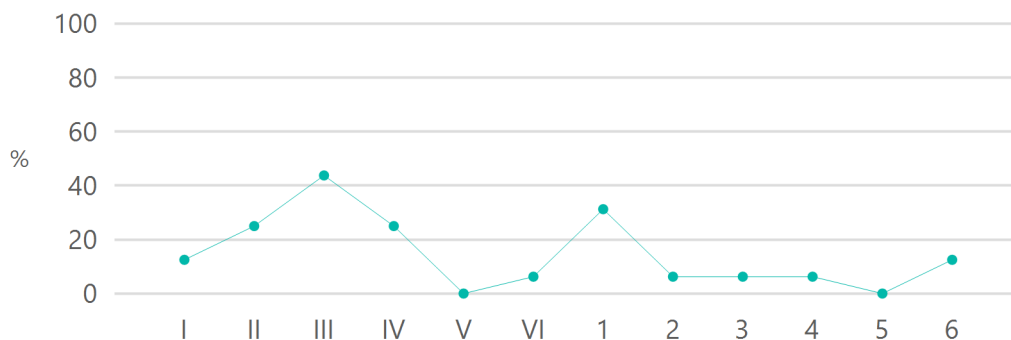
NOT-S

Total mean score:
1.75

Number: 16
Ages: 4 - 56

♂ (10)

♀ (6)



NOT-S interview

		Count	%
I	Sensory function	2	12.5%
II	Breathing	4	25.0%
III	Habits	7	43.8%
IV	Chewing and swallowing	4	25.0%
V	Drooling	0	0.0%
VI	Dry mouth	1	6.3%

NOT-S examination

		Count	%
1	Face at rest	5	31.3%
2	Nose breathing	1	6.3%
3	Facial expression	1	6.3%
4	Masticatory muscle and jaw function	1	6.3%
5	Oral motor function	0	0.0%
6	Speech	2	12.5%

The report is based on data from the MHC database - the Mun-H-Center database on oral health and orofacial function in rare diseases. Data was collected by dentists and speech-language pathologists using the Orofacial Observation Chart.