



Orofacial function of persons having Turner syndrome

Report from observation charts

The survey comprises 62 observation charts.

Estimated occurrence: 1:2000 girls born.

Etiology: Girls with Turner syndrome are either missing one X chromosome (45,X) or part of it. Sometimes parts of the body have normal cells that contain two X chromosomes, while other cells are missing one X chromosome (mosaicism). There are also variants in which girls may have three X chromosomes, two of which are normal length and the third shorter. Why this chromosomal aberration occurs is not known today. Turner syndrome is not hereditary.

General symptoms: Short stature. Many of these girls have loss of ovarian function and they often do not enter puberty. Cardiac defects are common. Often there are frequent ear infections. Learning disability and behavioral problems can sometimes occur.

Orofacial/odontological symptoms: Characteristic facial features are associated with the diagnosis. In contrast to skeletal development, which tends to be delayed by more than two years, dental development tends to be early, and teeth may erupt earlier than normal. Teeth may have shape aberrations. The most common anomalies relate to the proportions between the root and the crown of the teeth, with relatively short roots on the incisors, canines and premolars. Teeth may be small, with thin enamel. There are reports of arched palate and a small lower jaw. Various malocclusions may occur, with a particular frequency of post normal bite and cross bite. Malocclusions appear early. Feeding problems are common during the first years of life. Some girls have difficulties with speech and language acquisition. Women who have not been treated with growth hormone can have a high pitched voice.

Orofacial/ odontological treatment:

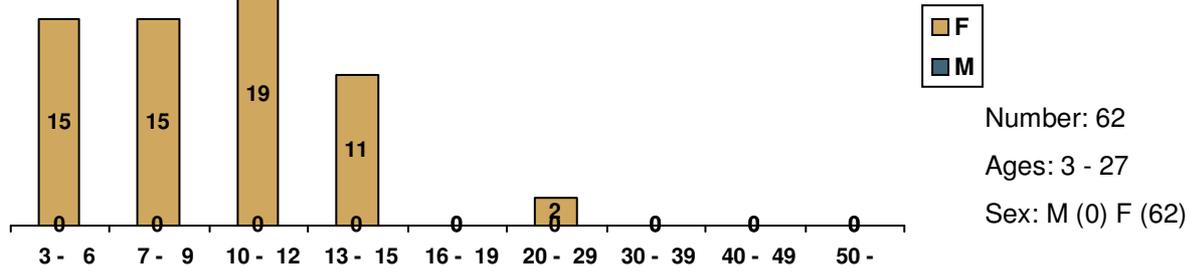
- Regular check-ups of dental and jaw development. Orthodontist should be consulted when needed.
- A specialist in pediatric dentistry or orthodontia may be a useful resource in this respect, and should also be consulted if treatment with growth hormone is being considered
- There are special intervention programs for children with Turner syndrome, with a focus on odontology. Tooth age determination is recommended as part of the diagnostic work-up. Anomalies beyond 2SD indicate that the child should be referred to a physician for further examination.
- Children with eating disorders often required extra dental care, including assistance with oral hygiene and fluoride treatments.
- Feeding and swallowing difficulties are investigated and treated by a specialist team at the hospital or multidisciplinary treatment centre.
- Speech and language difficulties should be diagnosed and treated by a speech therapist
- When treating medically compromised patients always contact their doctors for medical advice.

Source:

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

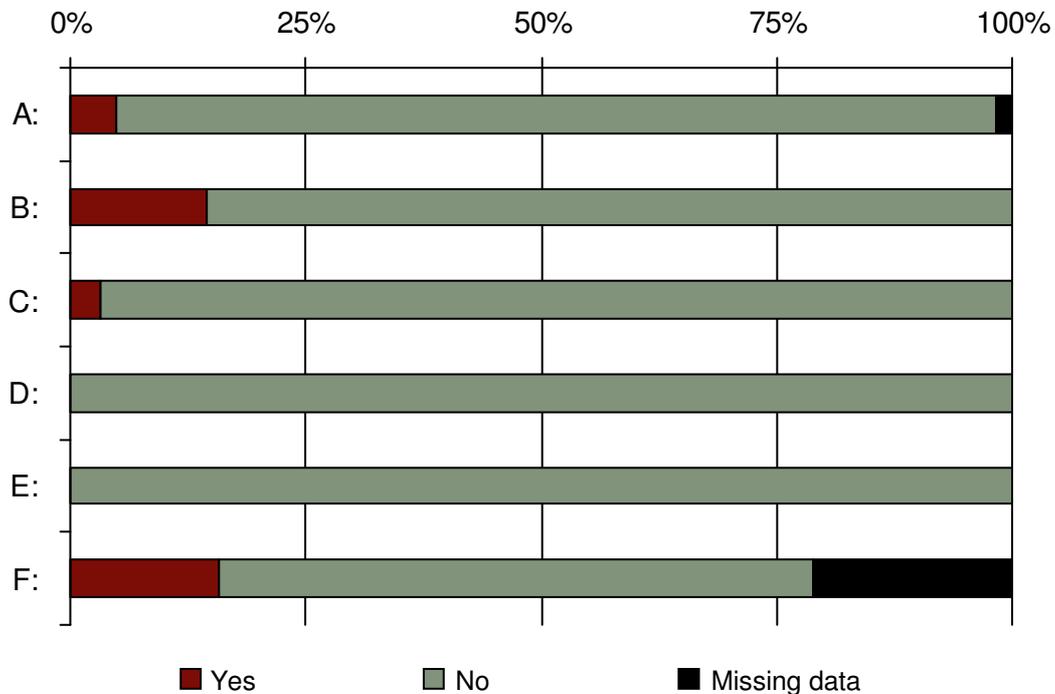
The Documentation from the Ågrenska Center. The intervention program for Turner syndrome, 2000, published by the Swedish Turner Academy.

Age distribution



Overview

	Yes	No	Missing data	N
A: Incomprehensible speech/No speech	3	58	1	62
B: Eating and drinking difficulties ¹	9	53	0	62
C: Profuse drooling, on clothes ¹	2	60	0	62
D: Breathing difficulties ^{1 2}	0	19	0	19
E: Grinding every day ^{1 2}	0	19	0	19
F: Severe malocclusions ²	3	12	4	19



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

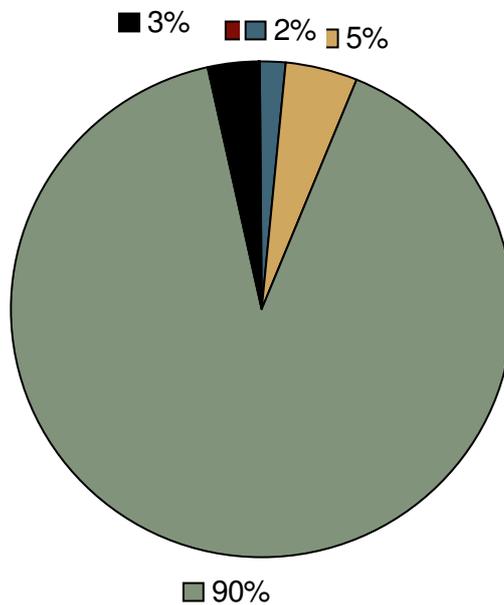
Oral health index (indices)¹

		0	1	2	3	4	5	6	Missing data	N
Calc	Calculus	15	2	0	0	0	0	0	2	19
GI	Gingivitis	10	1	3	0	1	2	0	2	19
Plaq	Coating	9	3	0	0	2	2	1	2	19
Toot	Tooth wear	13	1	0	0				5	19

- 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



	Number
None	0
Negative	1
Reluctant	3
Positive	56
Missing data	2
Sum:	62

Caries

	3-6 years	7-12 years	13-19 years	Adults
deft¹				
Examined	13	18		
Number of individuals with deft=0	10	13		
Mean	0,4	1,2		
Standard deviation	0,7	2,2		
Missing data	2	16		
DMFT²				
Examined		25	8	1
Number of individuals with DMFT=0		21	7	0
Standard deviation		0,7	1,7	0,0
Mean		0,3	0,6	1,0
Missing data		9	3	1

1: Number of carious or filled deciduous teeth

2: Number of carious or filled permanent teeth

Occlusal relationship

	Number
Neutral bite	43
Post normal	15
Pre normal	2
Missing data	2
<hr/>	
	Sum: 62

Maximum jaw opening

Children younger than 10 years

	Number
- 20	0
21 - 30	1
31 - 40	14
41 - 50	4
51 -	0
Missing data	11
<hr/>	
	Sum: 30

Children, 10 years or older, and adults

	Number
- 20	0
21 - 30	1
31 - 40	3
41 - 50	19
51 -	6
Missing data	3
<hr/>	
	Sum: 32

Profile¹

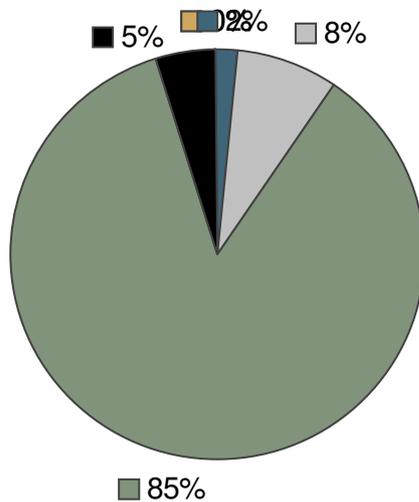
	Number
Normal	18
Convex	0
Concave	0
Missing data	1
<hr/>	
	Sum: 19

Mandibular plane¹

	Number
Normal	14
Increased	2
Reduced	2
Missing data	1
<hr/>	
	Sum: 19

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

Speech difficulty



	Number
■ No speech	0
■ Very incomprehensible	0
■ Incomprehensible speech	1
■ Slightly indistinct speech	5
■ No problems	53
■ Missing data	3
Sum: 62	

Clinical findings	Yes-answers			
	Total N=62 (%)	Boys/Men N=0 (%)	Girls/Women N=62 (%)	Missing data
Narrow palate	34 (57)	0 ()	34 (57)	2
High palate	28 (46)	0 ()	28 (46)	1
Spacing	20 (32)	0 ()	20 (32)	0
Open mouth at rest	14 (23)	0 ()	14 (23)	2
Frontal open bite	14 (24)	0 ()	14 (24)	3
M mentalis overactive	11 (18)	0 ()	11 (18)	0
Over crowding	7 (11)	0 ()	7 (11)	1
Deep bite with gingival contact	3 (5)	0 ()	3 (5)	5