



Orofacial function of persons having Crouzon syndrome

Report from observation charts



12 observation charts

Synonym

ICD-10 Q75.1

Estimated occurrence 4:100,000 live births.

General symptoms

The growth seams, or sutures, of the skull fuse prematurely, causing the head to take on an abnormal shape. The eyeballs appear to bulge owing to the shallowness of the eye sockets. The inadequate development of the skull sutures may result in excessive pressure on the brain. Symptoms of this pressure include headaches, vomiting, lack of appetite, and damage to the optic nerve. Hearing impairment may occur. The syndrome varies in severity from mild to very serious.

Oral symptoms

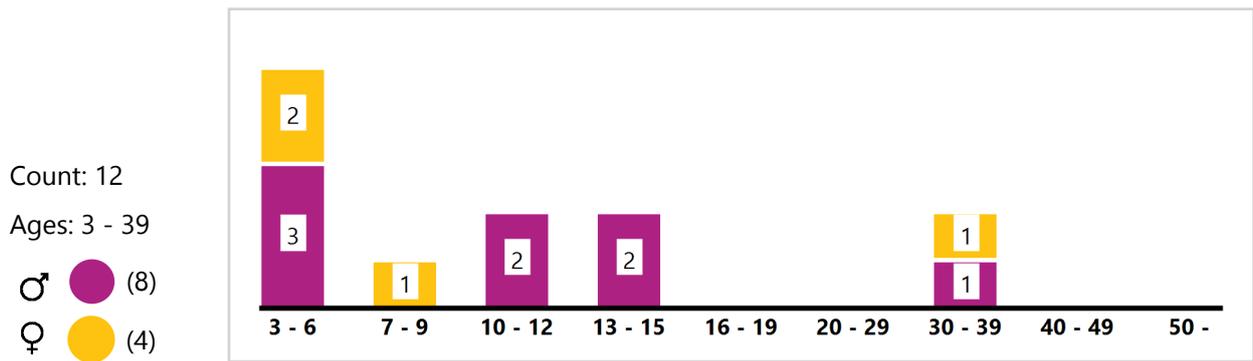
The midline third of the face and the upper jaw are underdeveloped, owing to the disturbance in the growth of the skull sutures. This may affect breathing, swallowing and speech. The palate tends to be high and narrow. Cleft lip and palate are not common, but do occur. The teeth may be crowded, particularly in the upper jaw. The bite may also be open, in the area of the front teeth. The reduced size of the lower jaw may lead to development of an underbite.

Oral treatment

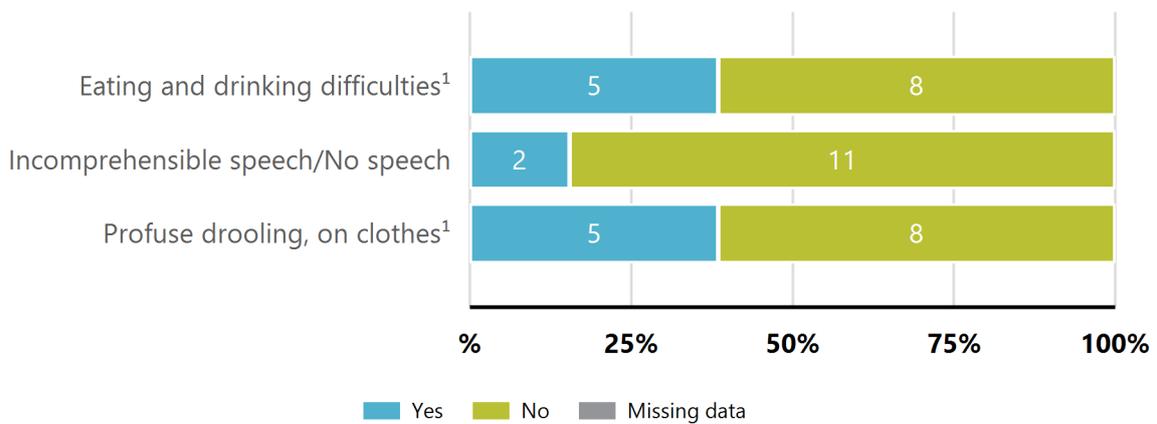
- In cases of craniofacial deformities, a specialist team will be needed for follow up and treatment.
- Many individuals with Crouzon syndrome require orthodontic treatment, often in combination with jaw surgery.
- Early contact with dental services for intensified prophylactic care and oral hygiene information is essential.

Sources The MHC database Rare diseases Dokumentation-Ågrenska

Age distribution



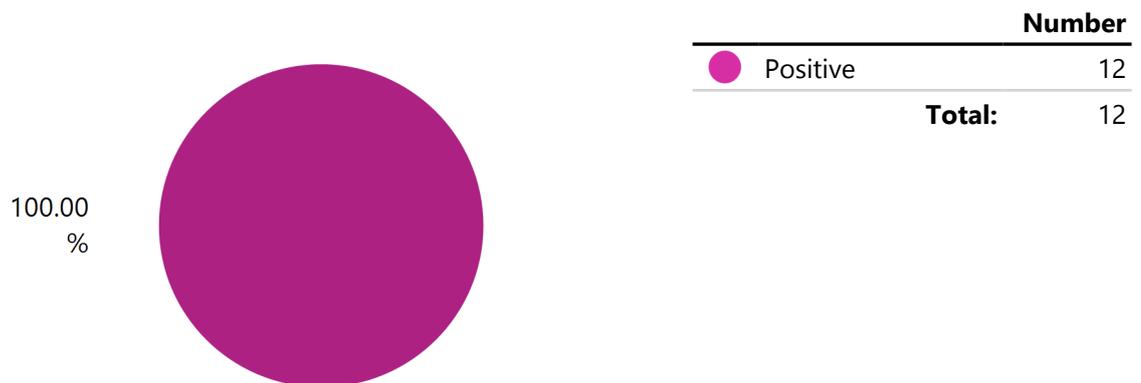
Summary



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	3		
Number of carious or filled deciduous teeth	Number of individuals with deft=0	4	2		
	Mean	0.4	1.0		
	Standard deviation	0.9	1.7		
	No data	0	0		
DMFT	Examined		3	2	2
Number of carious or filled permanent teeth	Number of individuals with DMFT=0		2	1	
	Mean		1.3	0.5	15.0
	Standard deviation		2.3	0.7	14.1
	No data		0	0	0

Oral health index (indices)¹

	0	1	2	3	4	5	6	Missing data	Number
Calculus	1	0	0	1	0	0	0	10	3
Gingivitis	1	0	0	0	0	0	1	10	3
Plaque	1	0	0	0	0	0	1	10	3
Tooth wear	3	0	0	0	0	0	0	9	3

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	0
Overbite	0
Underbite	13
Missing data	-1
Total:	12

Maximum jaw opening

Children younger than 10 years

Children, 10 years or older, and adults

	Number
-20	0
21-30	0
31-40	3
41-50	0
51-	0
Missing data	3
Total:	6

	Number
-20	0
21-30	1
31-40	3
41-50	1
51-	0
Missing data	1
Total:	6

Profile¹

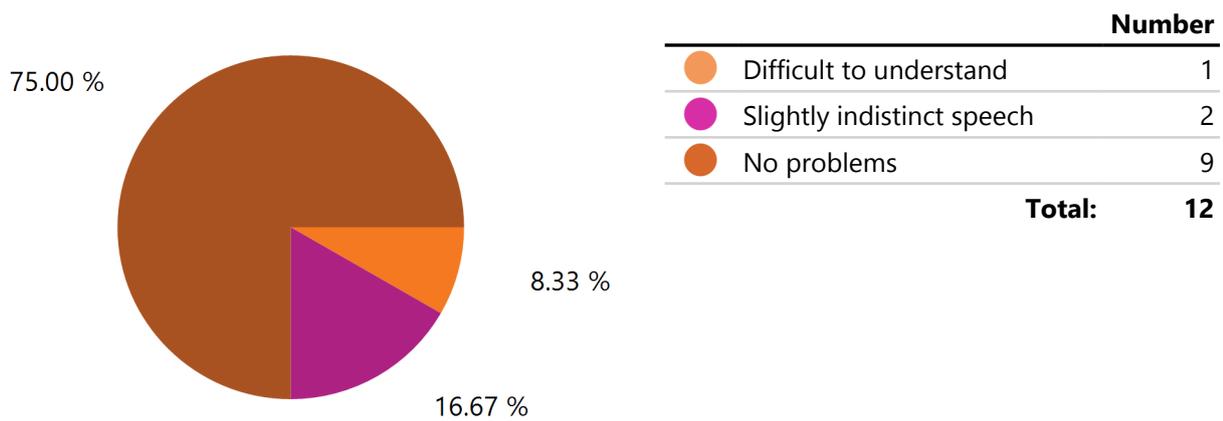
	Number
Normal	0
Convex	1
Concave	2
Missing data	0
Total:	3

Mandibular plane¹

	Number
Normal	2
Increased	0
Reduced	0
Missing data	1
Total:	3

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

Speech problems



Clinical findings

Number of yes-answers

	Total = 12 (%)	♂ = 8 (%)	♀ = 4 (%)	Missing Data
Open mouth at rest	9 (75)	6 (75)	3 (75)	0
Over crowding	7 (58)	5 (62)	2 (50)	0
High palate	6 (50)	4 (50)	2 (50)	0
Frontal open bite	5 (42)	3 (38)	2 (50)	1
Narrow palate	5 (42)	4 (50)	1 (25)	0
Reduced opening capacity	4 (33)	4 (50)	0 (0)	0
Facial asymmetry	3 (25)	2 (25)	1 (25)	0
High muscle tone in tongue	1 (8)	0 (0)	1 (25)	1