



Orofacial function of persons having Crouzon syndrome

Report from observation charts

The survey comprises 11 observation charts.

Estimated occurrence: 4:100 000 live births.

Etiology: Gene mutations can be found on chromosome 10 (FGFR2) or on chromosome 4 (FGFR3).

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.

Orofacial/odontological 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.

Orofacial/ odontological 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.

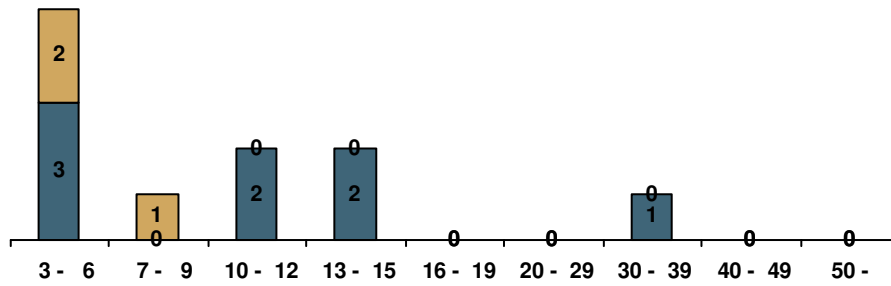
Sources

The 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.

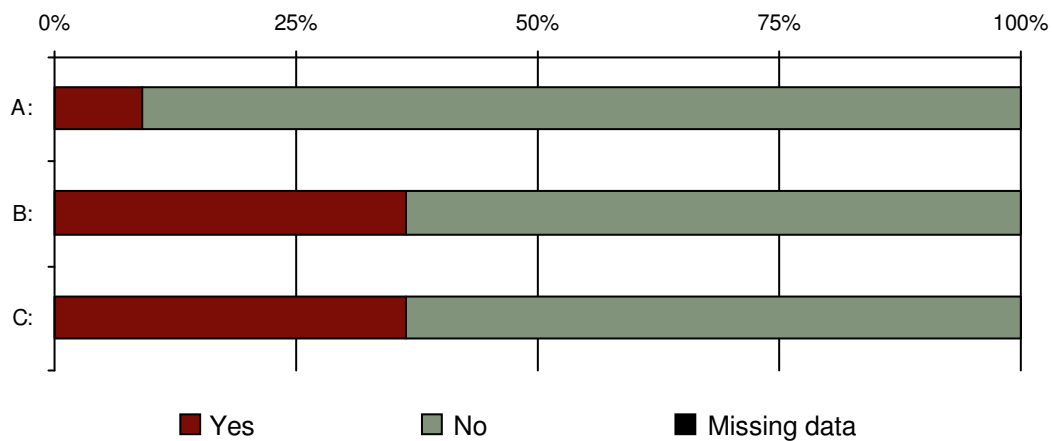
Age distribution



Number: 11
 Ages: 3 - 39
 Sex: M (8) F (3)

Overview

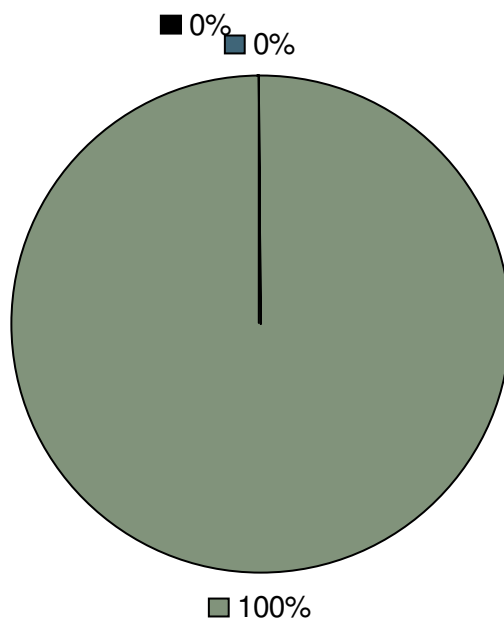
	Yes	No	Missing data	N
A: Incomprehensible speech/No speech	1	10	0	11
B: Eating and drinking difficulties ¹	4	7	0	11
C: Profuse drooling, on clothes ¹	4	7	0	11



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

¹: Compiled using questionnaire

Acceptance of dental examination



	Number
None	0
Negative	0
Reluctant	0
Positive	11
Missing data	0
Sum:	11

Caries

	3-6 years	7-12 years	13-19 years	Adults
deft¹				
Examined	5	3		
Number of individuals with deft=0	4	2		
Mean	0,4	1,0		
Standard deviation	0,8	1,4		
Missing data	0	0		
DMFT²				
Examined		3	2	1
Number of individuals with DMFT=0		2	1	0
Standard deviation		1,9	0,5	0,0
Mean		1,3	0,5	25,0
Missing data		0	0	0

1: Number of carious or filled deciduous teeth

2: Number of carious or filled permanent teeth

Occlusal relationship

	Number
Neutral bite	0
Post normal	0
Pre normal	11
Missing data	0
Sum: 11	

Maximum jaw opening

Children younger
than 10 years

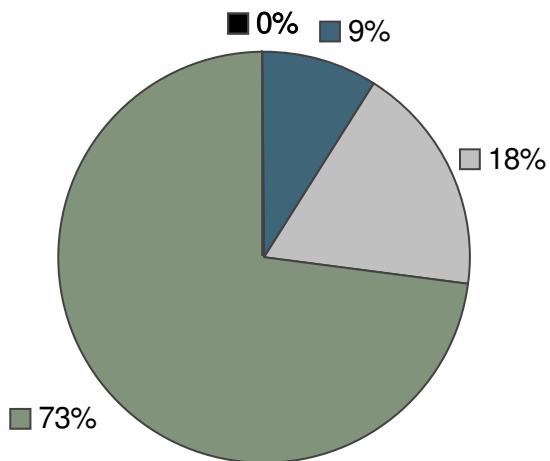
	Number
- 20	0
21 - 30	0
31 - 40	3
41 - 50	0
51 -	0
Missing data	3
Sum: 6	

Children, 10 years or
older, and adults

	Number
- 20	0
21 - 30	1
31 - 40	3
41 - 50	1
51 -	0
Missing data	0
Sum: 5	

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	2
No problems	8
Missing data	0
Sum: 11	

Clinical findings	Yes-answers			
	Total N=11 (%)	Boys/Men N=8 (%)	Girls/Women N=3 (%)	Missing data
Cranio-facial abnormality	11 (100)	8 (100)	3 (100)	0
Open mouth at rest	9 (82)	6 (75)	3 (100)	0
Over crowding	6 (55)	5 (63)	1 (33)	0
High palate	5 (45)	4 (50)	1 (33)	0
Frontal open bite	5 (45)	3 (38)	2 (67)	0
Narrow palate	4 (36)	4 (50)	0 ()	0
Reduced opening capacity	4 (36)	4 (50)	0 ()	0