



Orofacial function of persons having Cri Du Chat syndrome

Report from observation charts

The survey comprises 33 observation charts.

Synonyms: 5p deletion syndrome

Estimated incidence: 2:100 000 live births. More common in girls.

Etiology: Deletion of chromosomal material on chromosome 5.

General symptoms: Newborns have a weak, high-pitched cry attributable to a small larynx. Mental retardation, delayed motor skill development and muscle laxity may be present to varying extents. Sensitivity to infection is common. One-third of these children have congenital heart defects.

Orofacial/odontological symptoms: Characteristic facial features are associated with the diagnosis. Bite abnormalities commonly occur, most often an open bite in the region of the front teeth and an overbite. Oral motor function is commonly affected and the majority has sucking difficulties in infancy, eating disorders and drooling problems. Learning disability and reduced oral motor skills lead to communication problems.

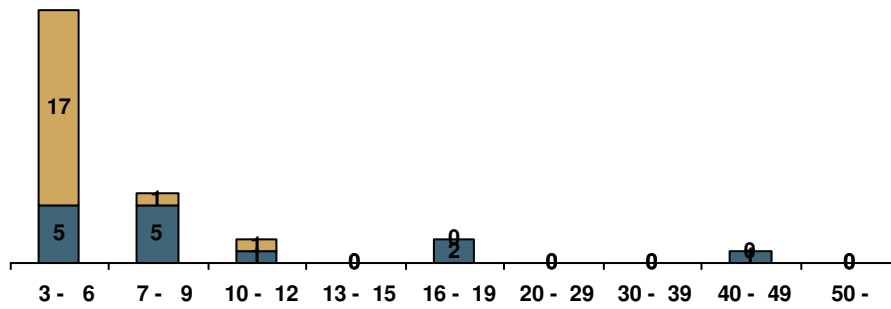
Orofacial/ odontological treatment:

- Early contact with dental services for intensified prophylactic care and oral hygiene information is essential.
- Regular check-ups of dental and jaw development. Orthodontist should be consulted when needed.
- Training in oral motor skills in cases of eating disorders, speech difficulties and drooling may be relevant.
- Speech, language and communication training are often required.
- When treating medically compromised patients always contact their doctors for medical advice (bleeding problems, heart diseases etc.).

Source

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.

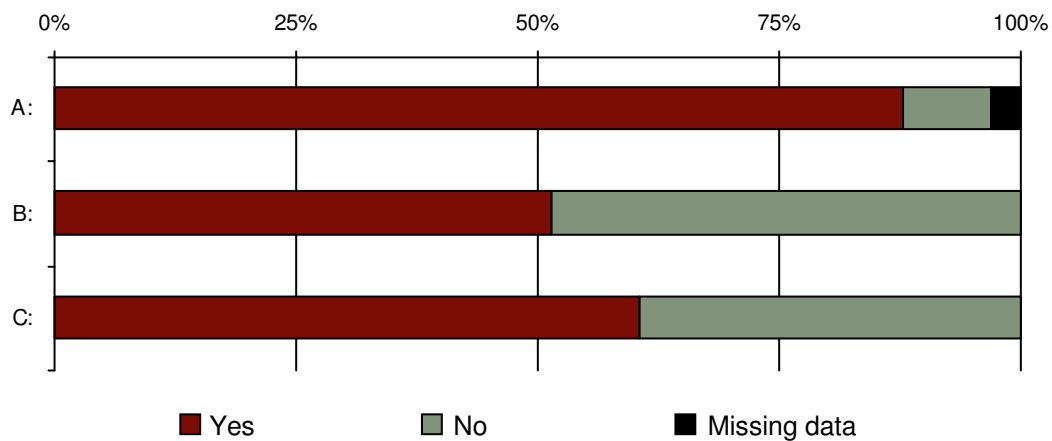
Age distribution



Number: 33
 Ages: 3 - 43
 Sex: M (14) F (19)

Overview

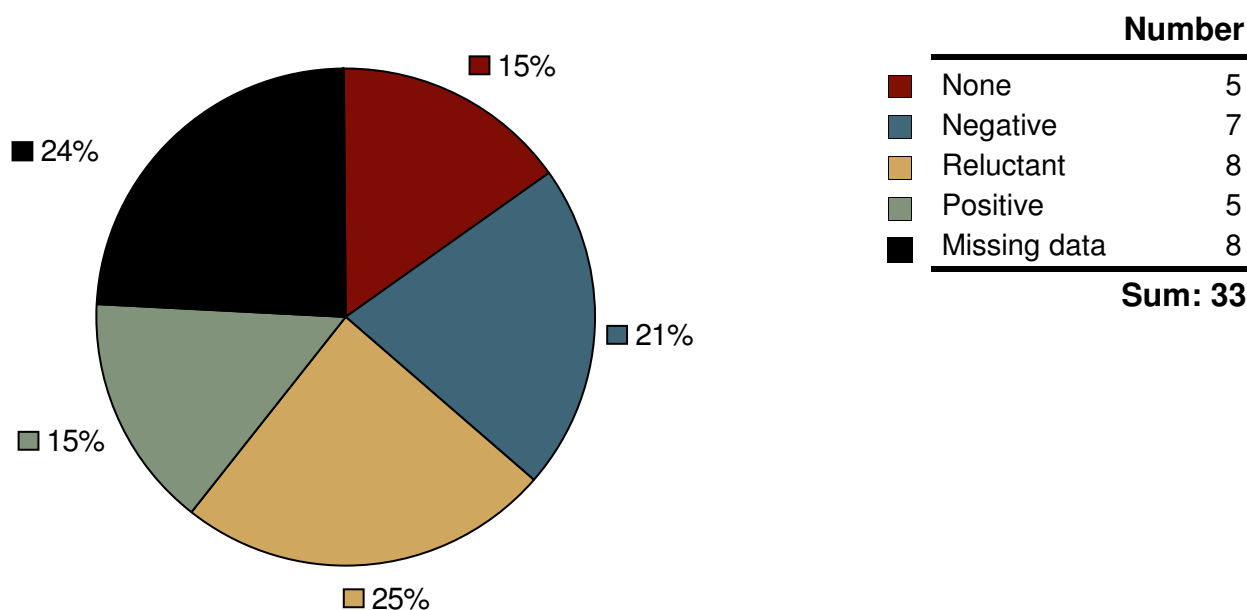
	Yes	No	Missing data	N
A: Incomprehensible speech/No speech	29	3	1	33
B: Eating and drinking difficulties ¹	17	16	0	33
C: Profuse drooling, on clothes ¹	20	13	0	33



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

¹: Compiled using questionnaire

Acceptance of dental examination



Caries

	3-6 years	7-12 years	13-19 years	Adults
deft¹				
Examined	13	4		
Number of individuals with deft=0	12	4		
Mean	0,6	0,0		
Standard deviation	2,1	0,0		
Missing data	9	4		
DMFT²				
Examined		6	2	1
Number of individuals with DMFT=0		5	0	0
Standard deviation		0,7	1,0	0,0
Mean		0,3	4,0	10,0
Missing data		2	0	0

1: Number of carious or filled deciduous teeth

2: Number of carious or filled permanent teeth

Occlusal relationship

	Number
Neutral bite	12
Post normal	13
Pre normal	1
Missing data	7
<hr/>	
	Sum: 33

Maximum jaw opening

Children younger
than 10 years

	Number
- 20	0
21 - 30	0
31 - 40	1
41 - 50	1
51 -	0
Missing data	26
<hr/>	
	Sum: 28

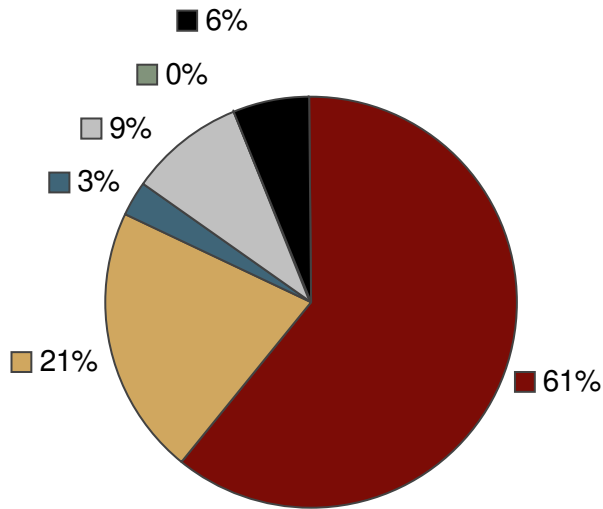
Children, 10 years or
older, and adults

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

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



Speech difficulty



	Number
No speech	20
Very incomprehensible	7
Incomprehensible speech	1
Slightly indistinct speech	3
No problems	0
Missing data	2
Sum: 33	

Clinical findings	Yes-answers			
	Total N=33 (%)	Boys/Men N=14 (%)	Girls/Women N=19 (%)	Missing data
Low muscle tone in lips	27 (87)	12 (100)	15 (79)	2
Open mouth at rest	26 (84)	11 (92)	15 (79)	2
Impaired tongue motility	19 (63)	9 (69)	10 (59)	3
Frontal open bite	12 (44)	6 (55)	6 (38)	6
Narrow palate	7 (27)	3 (25)	4 (29)	7
Low muscle tone in tongue	6 (20)	5 (38)	1 (6)	3
High palate	4 (15)	3 (25)	1 (7)	7
Reduced stability in neck	3 (10)	0 (0)	3 (17)	3
Facial asymmetry	3 (10)	1 (8)	2 (11)	2
Low muscle tone in masticatory muscles	3 (13)	1 (9)	2 (17)	10