mun-h FOLK-TAND-VÅRDEN

Orofacial function of persons having
Cri Du Chat syndrome

Report from observation charts | 34 observation charts

Synonym 5p deletion syndrome.

ICD-10 Q93.4

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

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.

Oral symptoms Characteristic facial features are associated with the diagnosis. Small lower jaw and narrow palate is common as well as 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.

Oral 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.).

Sources The MHC database Rare diseases Dokumentation-Ågrenska

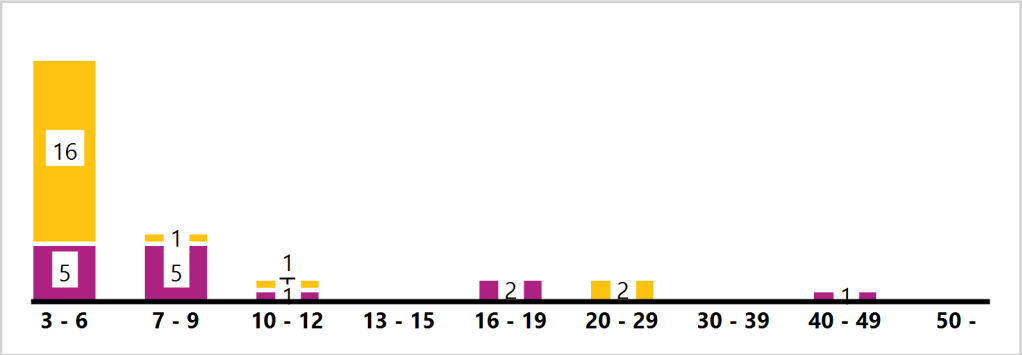
Age distribution

Count: 34

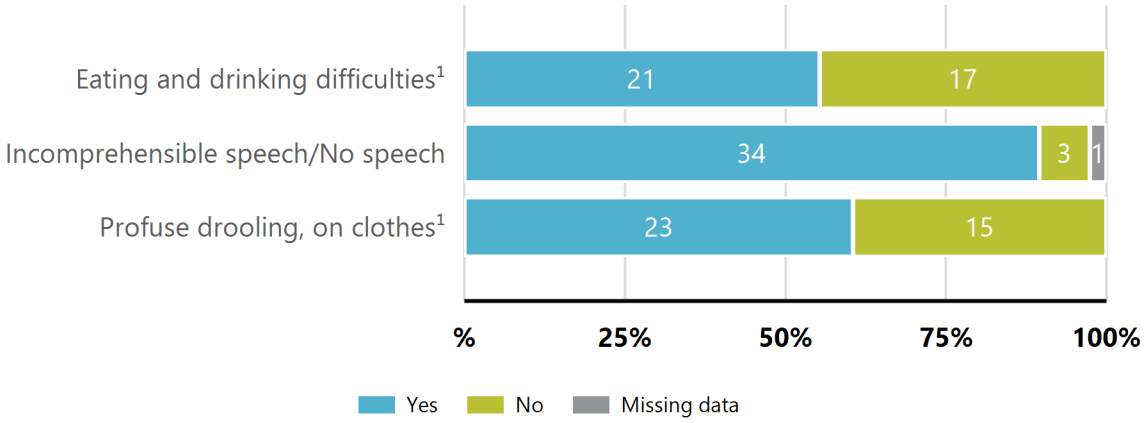
Ages: 3 - 43

♂ (14)

♀ (20)



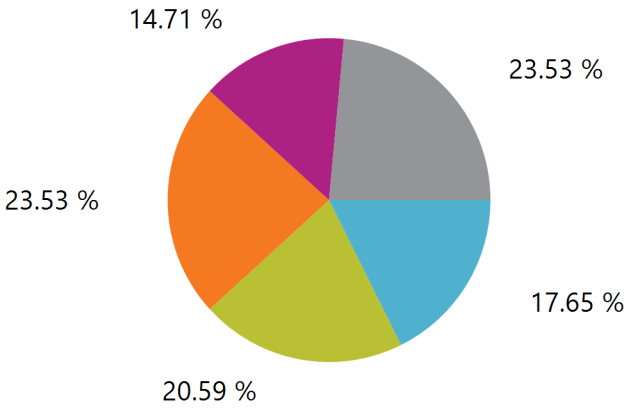
Summary



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



	Number
None	6
Negative	7
Reluctant	8
Positive	5
Missing data	8
Total:	34

Oral health

Caries

		3-6 years	7-12 years	13-19 years	>19 years
deft	**Examined**	21	8		
Number of carious or filled deciduous teeth	**Number of individuals with deft=0**	20	8		
	Mean	0.7	0.0		
	Standard deviation	2.3	0.0		
	No data	9	4		
DMFT	**Examined**		8	2	3
Number of carious or filled permanent teeth	**Number of individuals with DMFT=0**		7		
	Mean		0.3	4.0	10.3
	Standard deviation		0.8	1.4	3.5
	No data		2	0	0

Oral health index (indices)[1]

	0	1	2	3	4	5	6	Missing data	Number
Calculus	0	0	0	1	0	0	0	33	7
Gingivitis	0	0	0	1	0	0	0	33	7
Plaque	0	0	0	0	1	0	0	33	7
Tooth wear	1	0	0	0	0	0	0	33	7

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	15
Overbite	14
Underbite	1
Missing data	4
Total:	**34**

Maximum jaw opening

Children younger than 10 years

	Number
-20	0
21-30	0
31-40	1
41-50	1
51-	0
Missing data	25
Total:	**27**

Children, 10 years or older, and adults

	Number
-20	0
21-30	0
31-40	1
41-50	0
51-	1
Missing data	5
Total:	**7**

Profile[1]

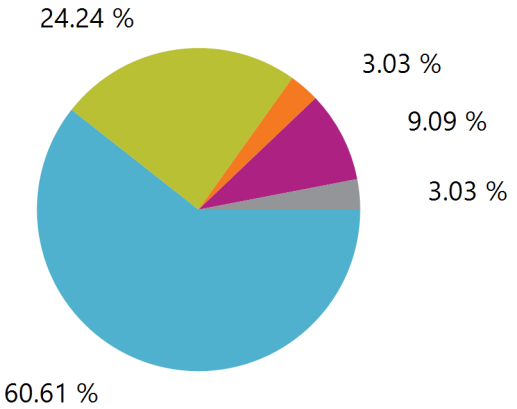
	Number
Normal	3
Convex	4
Concave	0
Missing data	0
Total:	**7**

Mandibular plane[1]

	Number
Normal	4
Increased	2
Reduced	0
Missing data	1
Total:	**7**

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

Speech problems



	Number
No speech	20
Very difficult to understand	8
Difficult to understand	1
Slightly indistinct speech	3
Missing data	1
Total:	**33**

Clinical findings

Number of yes-answers

	Total = 34 (%)	♂ = 14 (%)	♀ = 20 (%)	Missing Data
	29 (85)	12 (86)	17 (85)	2
Open mouth at rest	26 (76)	11 (79)	15 (75)	2
Impaired tongue motility	20 (59)	9 (64)	11 (55)	3
Frontal open bite	12 (35)	6 (43)	6 (30)	7
Narrow palate	8 (24)	3 (21)	5 (25)	8
Low muscle tone in tongue	6 (18)	5 (36)	1 (5)	4
High palate	5 (15)	3 (21)	2 (10)	8
Reduced stability in neck	4 (12)	0 (0)	4 (20)	3
Low muscle tone in masticatory muscles	3 (9)	1 (7)	2 (10)	11
Facial asymmetry	3 (9)	1 (7)	2 (10)	3
Spacing	2 (6)	1 (7)	1 (5)	9
Over crowding	1 (3)	1 (7)	0 (0)	8
Deep bite with gingival contact	1 (3)	0 (0)	1 (5)	8
	1 (3)	0 (0)	1 (5)	4