Orofacial function of persons having
Neurofibromatosis 1

Report from observation charts | 99 observation charts

Synonym Recklinghausen´s disease.

ICD-10 Q85.0

Estimated occurance 30:100,000 inhabitants.

General symptoms Café au lait spots and neurofibromas of the skin are characteristic. The latter are benign tumors that develop in the supportive tissue around the nerves. There may be up to several hundred of them. One-third of affected individuals develop plexiform neurofibromas, which are not nearly as restricted as neurofibromas of the skin. This disease, or more correctly disorder, may also impair the ocular and auditory nerves, the central nervous system, and skeletal development. Endocrine tumours occur. Children with this diagnosis should have annual physical examinations. Some developmental delay, learning difficulties and concentration problems may occur, as well as epilepsy. Scoliosis is occasionally found.

Oral symptoms Neurofibromas may occur in the oral mucous membranes, but these are normally not treated unless they grow or become uncomfortable. Neurofibromas may also occur in the jawbone, where they may be found with the aid of general radiographs. If they are found, they should be checked at regular intervals. There may be enlarged papillae on the tongue. Early tooth eruption has been reported, as well as abnormal tooth positions. Eating and speech difficulties and drooling are found, as well as occasional sleep apnea (frequent suspension of breathing while asleep).

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.
- Oral motor training and stimulation may be relevant in cases of eating difficulties, speech impairment and drooling.
- Speech, language and communication training are often justified.
- Snoring problems should be followed up by a physician.

Sources The MHC database Rare diseases Dokumentation-Ågrenska

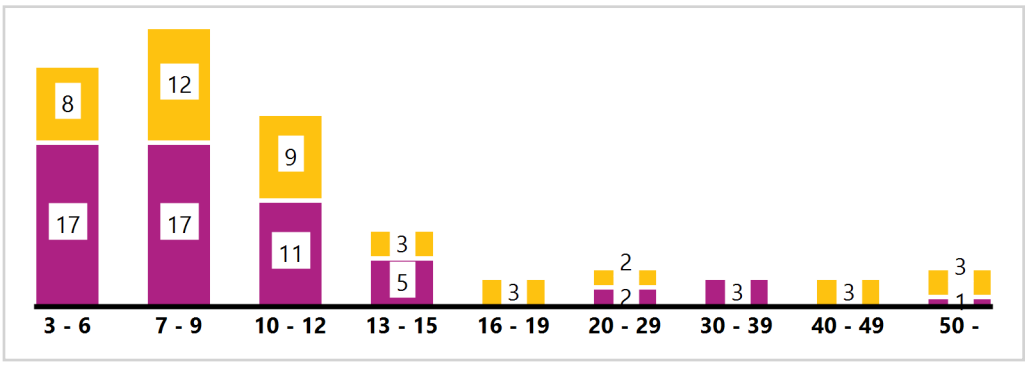
Age distribution

Count: 99

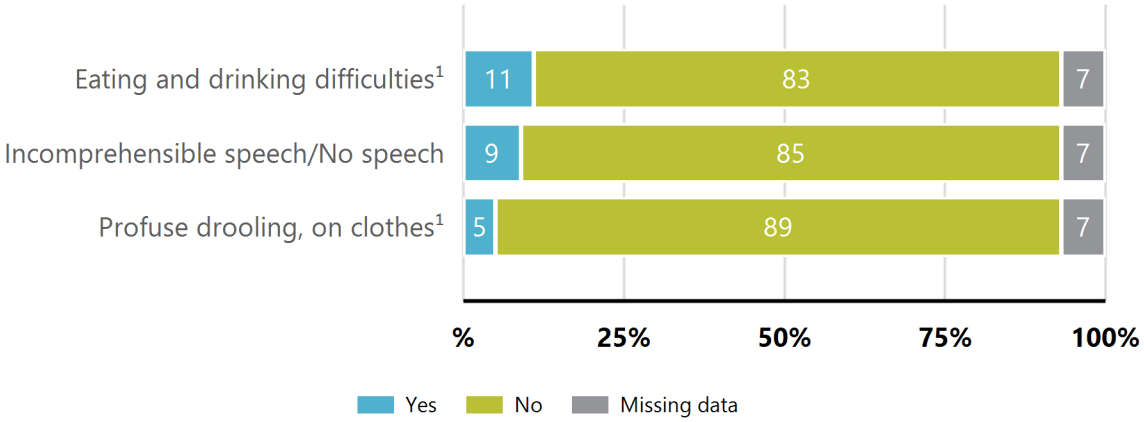
Ages: 3 - 75

♂ (56)

♀ (43)



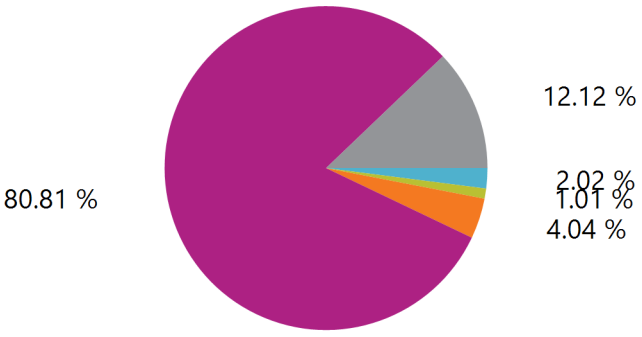
Summary



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	2
Negative	1
Reluctant	4
Positive	80
Missing data	12
Total:	99

Oral health

Caries

		3-6 years	7-12 years	13-19 years	>19 years
deft	**Examined**	25	49		
Number of carious or filled deciduous teeth	**Number of individuals with deft=0**	22	35		
	Mean	0.4	1.3		
	Standard deviation	1.1	1.8		
	No data	5	19		
DMFT	**Examined**		49	11	14
Number of carious or filled permanent teeth	**Number of individuals with DMFT=0**		42	9	3
	Mean		0.3	0.5	9.5
	Standard deviation		0.8	0.8	7.0
	No data		10	5	2

Oral health index (indices)[1]

	0	1	2	3	4	5	6	Missing data	Number
Calculus	29	8	0	0	0	0	0	62	51
Gingivitis	19	9	1	5	2	1	0	62	51
Plaque	22	6	4	2	2	1	1	61	51
Tooth wear	30	6	1	0	0	0	0	62	51

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	58
Overbite	12
Underbite	21
Missing data	8
Total:	**99**

Maximum jaw opening

Children younger than 10 years

	Number
-20	0
21-30	1
31-40	10
41-50	26
51-	6
Missing data	11
Total:	**54**

Children, 10 years or older, and adults

	Number
-20	0
21-30	0
31-40	7
41-50	18
51-	14
Missing data	6
Total:	**45**

Profile[1]

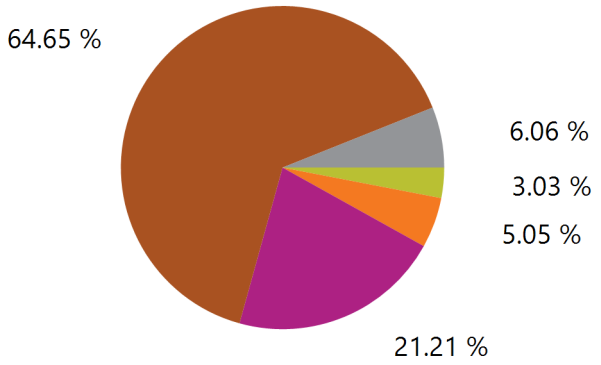
	Number
Normal	29
Convex	6
Concave	3
Missing data	13
Total:	**51**

Mandibular plane[1]

	Number
Normal	29
Increased	6
Reduced	1
Missing data	15
Total:	**51**

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

Speech problems



	Number
Very difficult to understand	3
Difficult to understand	5
Slightly indistinct speech	21
No problems	64
Missing data	6
Total:	**99**

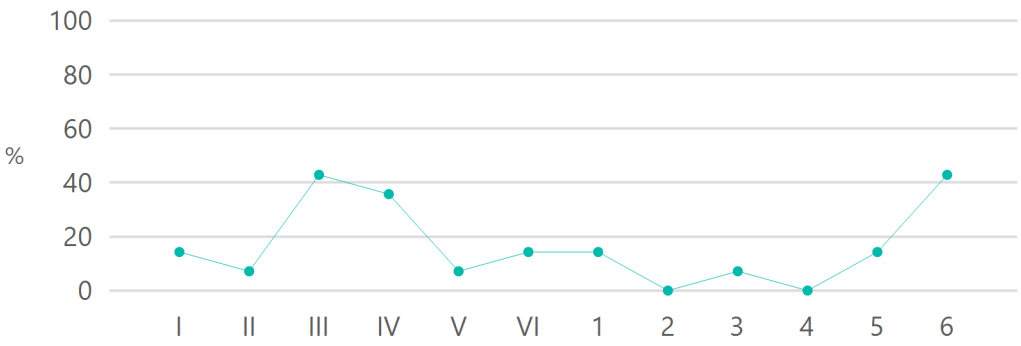
Clinical findings

Number of yes-answers

	Total = 99 (%)	♂ = 56 (%)	♀ = 43 (%)	Missing Data
Open mouth at rest	16 (16)	8 (14)	8 (19)	8
Facial asymmetry	16 (16)	10 (18)	6 (14)	7
Over crowding	14 (14)	9 (16)	5 (12)	11
Spacing	13 (13)	7 (12)	6 (14)	12
Frontal open bite	11 (11)	5 (9)	6 (14)	10
Low muscle tone in lips	10 (10)	7 (12)	3 (7)	9
Impaired tongue motility	10 (10)	7 (12)	3 (7)	10
High palate	9 (9)	6 (11)	3 (7)	11
Narrow palate	6 (6)	5 (9)	1 (2)	10
Low muscle tone in masticatory muscles	3 (3)	3 (5)	0 (0)	12
Deep bite with gingival contact	3 (3)	3 (5)	0 (0)	11
Low muscle tone in tongue	2 (2)	1 (2)	1 (2)	9
Reduced opening capacity	1 (1)	1 (2)	0 (0)	7
Short tongue frenulum	1 (1)	1 (2)	0 (0)	10

NOT-S

Total mean score: 2.00



Number: 14
Ages: 6 - 14

♂ (9)
♀ (5)

NOT-S interview

		Count	%
I	**Sensory function**	2	14.3%
II	**Breathing**	1	7.1%
III	**Habits**	6	42.9%
IV	**Chewing and swallowing**	5	35.7%
V	**Drooling**	1	7.1%
VI	**Dry mouth**	2	14.3%

NOT-S examination

		Count	%
1	**Face at rest**	2	14.3%
2	**Nose breathing**	0	0.0%
3	**Facial expression**	1	7.1%
4	**Masticatory muscle and jaw function**	0	0.0%
5	**Oral motor function**	2	14.3%
6	**Speech**	6	42.9%

The report is based on data from the MHC database - the Mun-H-Center database on oral health and orofacial function in rare diseases. Data was collected by dentists and speech-language pathologists using the Orofacial Observation Chart.