Orofacial function of persons having **Neurofibromatosis 1**

Report from questionnaires | 65 questionnaires

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

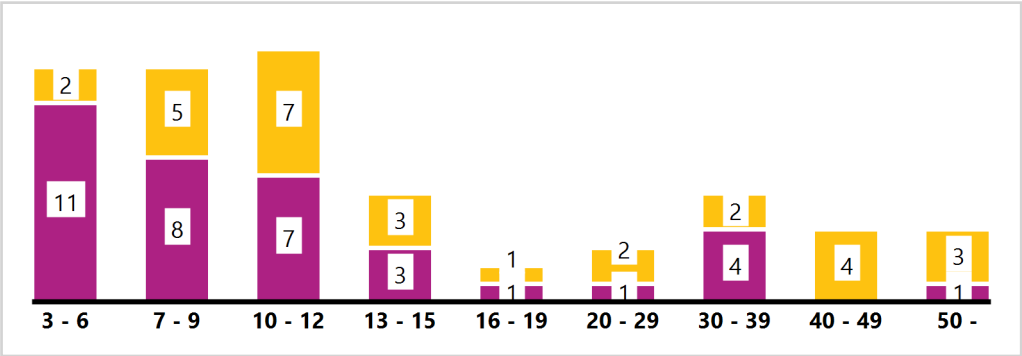
Age distribution

Number: 65

Ages: 4 - 75

♂ (36)

♀ (29)



Additional diagnoses

Medical impairment	Yes	No	Missing data
Inborn heart defect	1	61	3
Other cardiovascular disease	0	63	2
Epilepsy	3	58	4
Asthma	3	57	5
Need of respiratory support	2	61	2
Allergy	13	48	4

Neuropsychiatric diagnosis	Yes	No	Missing data
ADHD/ADD	10	47	8
Autism (Includes autism, Asperger syndrome and autistic traits)	5	47	13

General disability	Yes	No	Missing data
Intellectual disability	5	52	8
Motoric functional impairment	22	28	15
Visual impairment	12	31	22
Hearing loss	5	53	7
Communication difficulties	11	42	12

About dental care and oral health

Do you feel that you receive the dental care you need?	Number
Yes, very much so	38
Yes, somewhat	18
No, not really	3
No, not at all	1
Missing data	5
Total:	**65**

How many times per year do you normally seek dental care?	Number
Less than once per year	11
One time per year	28
Two times per year	8
Three or more times per year	15
Missing data	3
Total:	**65**

When were your teeth last X-rayed?	Number
During the past two years	40
More than two years ago	8
Never had my teeth X-rayed	9
Missing data	8
Total:	**65**

Do you look after your teeth in a good way?	Number
Yes, very much so	31
Yes, somewhat	29
No, not really	4
Missing data	1
Total:	**65**

Who brushes your teeth?	Number
I always brush myself	28
Someone else always helps me	13
Sometimes I brush myself	24
Total:	**65**

How often are your teeth brushed?	Number
Once per day	4
Two times per day	56
Three or more times per day	4
Missing data	1
Total:	**65**

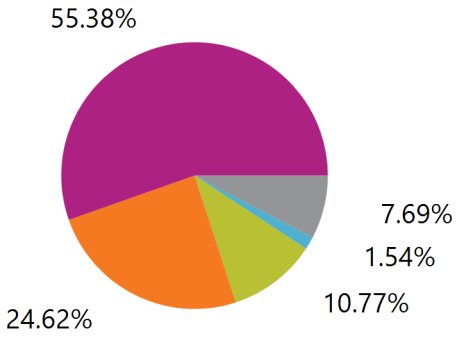
About dental care and oral health

	Yes	No	Missing data
Does your mouth hurt?	9	51	5
Does your mouth feel dry?	9	51	5
Have you ever taken a serious hit to your permanent front teeth?	5	57	3
Do you feel that you have a divergent bite?	9	51	5
Have you had a brace?	8	56	1
Do you feel that you need orthodontics/a brace?	16	40	9

Do you grind or press your teeth at night?	Number
Never	48
Once or twice per week	8
Every night	4
Missing data	5
Total:	**65**

Do you grind or press your teeth during the day?	Number
Never	53
Once or twice per week	6
Every night	3
Missing data	3
Total:	**65**

About eating



Do you have any problems with eating?	Number
Yes, very much so	1
Yes, somewhat	7
No, not really	16
No, not at all	36
	5
Missing data	0
Total:	**65**

	Yes	No	Missing data
Do you cough daily in connection with meals?	1	61	3
Do you gag daily in connection with meals?	0	60	5
Do you get acid reflux daily?	3	56	6
Do you throw up often (at least twice per week)?	1	62	2
Do you have a poor appetite?	5	57	3
Does it take a long time before you can swallow a mouthful?	4	44	17
Do you press your tongue forward when you swallow so that food ends up outside the mouth?	2	44	19
Do you find it difficult to chew, i.e. grind food using your molars?	5	53	7
Do you find it difficult to take food from the spoon using your lips?	3	57	5
Have you had problems with food and drink leaking out through the corners of your mouth?	2	43	20
Does food tend to remain in your mouth after meals?	1	57	7
Do you get nutrition in any other way than through your mouth?	0	63	2

About drooling

Do you drool?	Number
Never drool	51
Drool sometimes – not every day	5
Drool often – every day	3
Missing data	0
Total:	**65**

How much do you drool?	Number
Slight drooling, only on the lips	4
Moderate drooling, on lip and chin	4
Total:	**8**

Is your drooling a problem for you?	Number
	1
Yes, somewhat	2
No, not really	4
No, not at all	1
Total:	**8**

Is your drooling a problem for your family or people around you?	Number
Yes, somewhat	2
No, not really	2
No, not at all	4
Total:	**8**