



Orofacial function of persons having Noonan syndrome

Report from questionnaires

The survey comprises 16 questionnaires.

Estimated incidence: 40-100:100 000 live births.

Etiology: Autosomal dominant hereditary trait. The mutation is known in 75 % of cases.

General symptoms: Most children born with Noonan syndrome have some kind of cardiac defect. Short stature – adult individuals with Noonan syndrome are generally about 15 cm shorter than predicted height. Growth hormone production deficit. Late onset puberty is common, and in many boys the testicles remain undescended. Some individuals have delayed psycho-motor development and intellectual disabilities. There may be some increased tendency to bleed, but this is not severe.

Orofacial/odontological symptoms: Characteristic features are associated with the diagnosis. Feeding difficulties are common, particularly during the first years of life. Some children have a great deal of vomiting. Owing to eating and swallowing difficulties, some children with Noonan syndrome need to eat often, and require a special diet. This may lead to an increased risk of tooth decay. High palate, late teething and small jaws with crowded teeth have all been reported.

Orofacial/ odontological treatment:

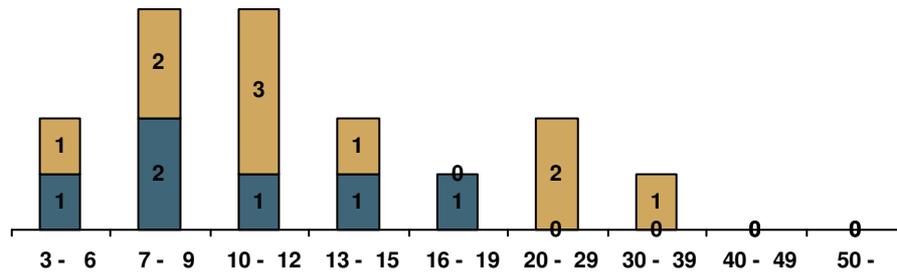
- Children with eating disorders often require extra dental care, including assistance with oral hygiene and fluoride treatments. However, the dental services should not advise on eating difficulties.
- Regular check-ups of dental and jaw development. Orthodontist should be consulted when needed.
- An increased tendency to bleed may result in complications when teeth are extracted.
- Oral motor training and stimulation may be relevant in cases of eating difficulties, speech impairment and drooling
- Feeding and swallowing difficulties are investigated and treated by a specialist team at the hospital or multidisciplinary treatment center.
- 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: 16

Ages: 3 - 38

Sex: M (6) F (10)

Additional diagnoses

Medical impairment	Yes	No	Missing data
Inborn heart defect	10	6	0
Other cardiovascular disease	0	16	0
Epilepsy	0	16	0
Asthma	2	14	0
Need of respiratory support	1	15	0
Allergy	6	10	0

Neuropsychiatric diagnosis	Yes	No	Missing data
ADHD/ADD	0	16	0
Autistic syndrome	1	15	0
Autistic traits	1	14	1
Asperger's syndrome	0	16	0
Tourette's syndrome	0	16	0

General disability	Yes	No	Missing data
Intellectual disability	2	11	3
Mobility dysfunction	4	8	4
Impaired vision	3	5	8
Impaired hearing	4	10	2
Difficulty in communicating	3	11	2

About dental care and oral health

Do you feel that you receive the dental care you need?

	Number
Yes, very much so	11
Yes, some what	3
No, not really	2
No, not at all	0
Missing data	0
Sum:	16

How many times per year do you normally seek dental care?

	Number
Three or more times per year	5
Twice per year	4
Once per year	5
Less than once per year	2
Missing data	0
Sum:	16

When were your teeth last X-rayed?

	Number
During the past two years	11
More than two years ago	0
Never had my teeth X-rayed	5
Missing data	0
Sum:	16

Do you look after your teeth in a good way?

	Number
Yes, very much so	6
Yes, some what	9
No, not really	0
No, not at all	0
Missing data	1
Sum:	16

Who brushes your teeth?

	Number
I always brush myself	8
Sometimes I brush myself	4
Someone else always helps me	4
Missing data	0
Sum:	16

How often are your teeth brushed?

	Number
Three or more times per day	0
Twice per day	11
Once per day	4
Not every day	1
Missing data	0
Sum:	16

About dental care and oral health

	Yes	No	Missing data
Does your mouth hurt?	3	13	0
Does your mouth feel dry?	1	12	3
Have you ever taken a serious hit to your permanent front teeth?	0	16	0
Do you feel that you have a divergent bite?	7	9	0
Have you had a brace?	6	10	0
Do you feel that you need orthodontics/a brace?	7	7	2

Do you grind or press your teeth at night?

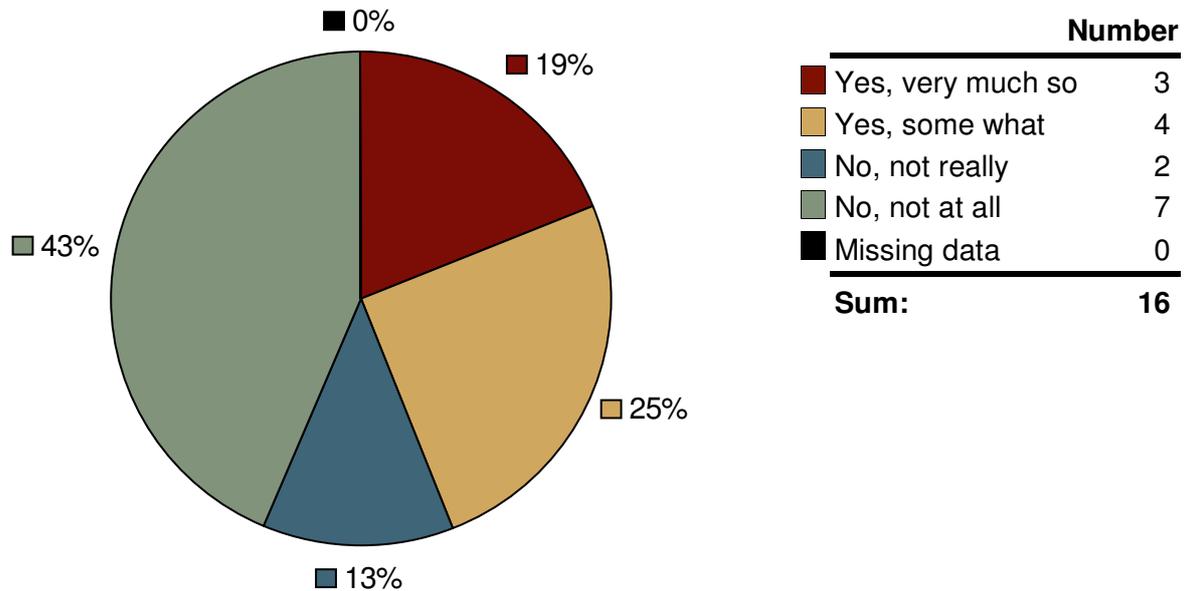
	Number
Never	8
Once or twice per week	5
Every night	2
Missing data	1
Sum:	16

Do you grind or press your teeth during the day?

	Number
Never	12
Once or twice per week	3
Every day	1
Missing data	0
Sum:	16

About eating

Do you have any problems with eating?



	Yes	No	Missing data
Do you cough daily in connection with meals?	2	14	0
Do you gag daily in connection with meals?	3	12	1
Do you get acid reflux daily?	0	16	0
Do you throw up often (at least twice per week)?	1	15	0
Do you have a poor appetite?	7	8	1
Does it take a long time before you can swallow a mouthful?	5	11	0
Do you press your tongue forward when you swallow so that food ends up outside the mouth?	0	16	0
Do you find it difficult to chew, i.e. grind food using your molars?	3	12	1
Do you find it difficult to take food from the spoon using your lips?	1	15	0
Have you had problems with food and drink leaking out through the corners of your mouth?	1	15	0
Does food tend to remain in your mouth after meals?	3	13	0
Do you get nutrition in any other way than through your mouth?	3	13	0

About drooling

Do you drool?

	Number
Never drool	15
Drool sometimes – not every day	0
Drool often – every day	1
Constant drooling	0
Missing data	0
Sum:	16

How much do you drool?

	Number
Slight drooling, only on the lips	0
Moderate drooling, on lip and chin	1
Profuse drooling, on clothes	0
Very profuse drooling, on hands/objects	0
Missing data	0
Sum:	1

Is your drooling a problem for you?

	Number
Yes, very much so	0
Yes, some what	1
No, not really	0
No, not at all	0
Missing data	0
Sum:	1

Is your drooling a problem for your family or people around you?

	Number
Yes, very much so	0
Yes, some what	0
No, not really	1
No, not at all	0
Missing data	0
Sum:	1