



# Orofacial function of persons having Noonan syndrome

## Report from observation charts

The survey comprises 37 observation charts.

**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:

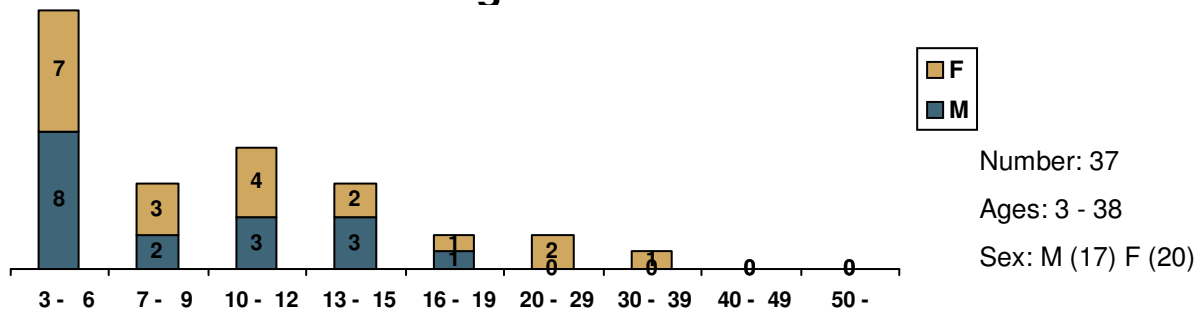
- Children with eating disorders often require extra dental care, including assistance with oral hygiene and fluoride treatments. However, the dental services should not advise on eating difficulties.
- Regular check-ups of dental and jaw development. Orthodontist should be consulted when needed.
- An 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.

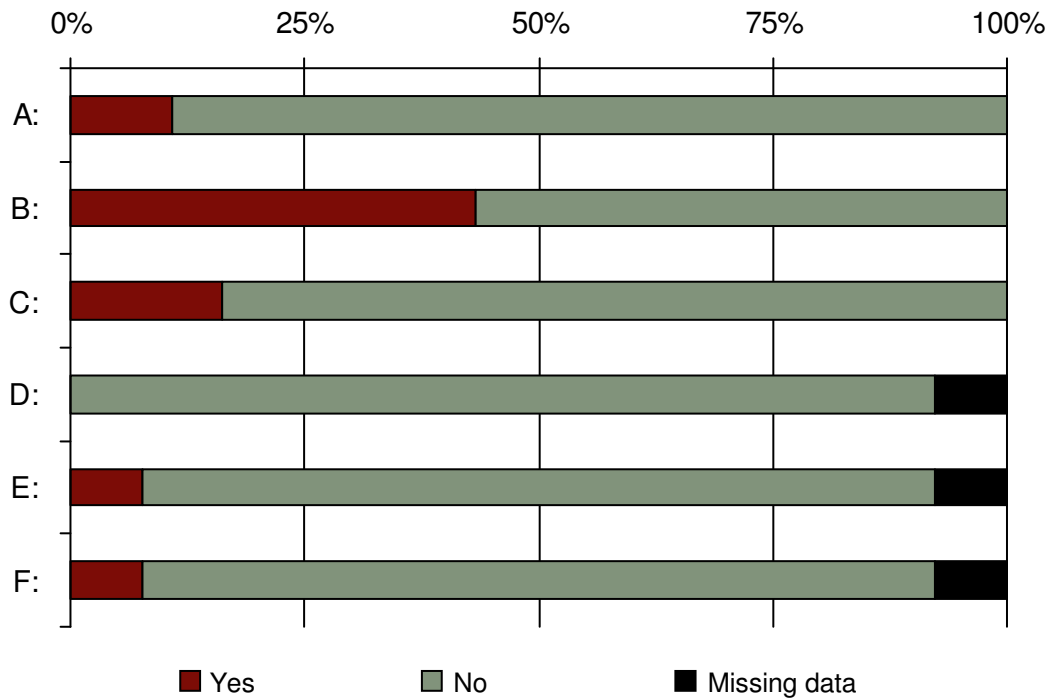
The Documentation from the Ågrenska Center.

### Age distribution



### Overview

	Yes	No	Missing data	N
A: Incomprehensible speech/No speech	4	33	0	37
B: Eating and drinking difficulties <sup>1</sup>	16	21	0	37
C: Profuse drooling, on clothes <sup>1</sup>	6	31	0	37
D: Breathing difficulties <sup>1 2</sup>	0	12	1	13
E: Grinding every day <sup>1 2</sup>	1	11	1	13
F: Severe malocclusions <sup>2</sup>	1	11	1	13



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

1: Compiled using questionnaire

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

## Oral health

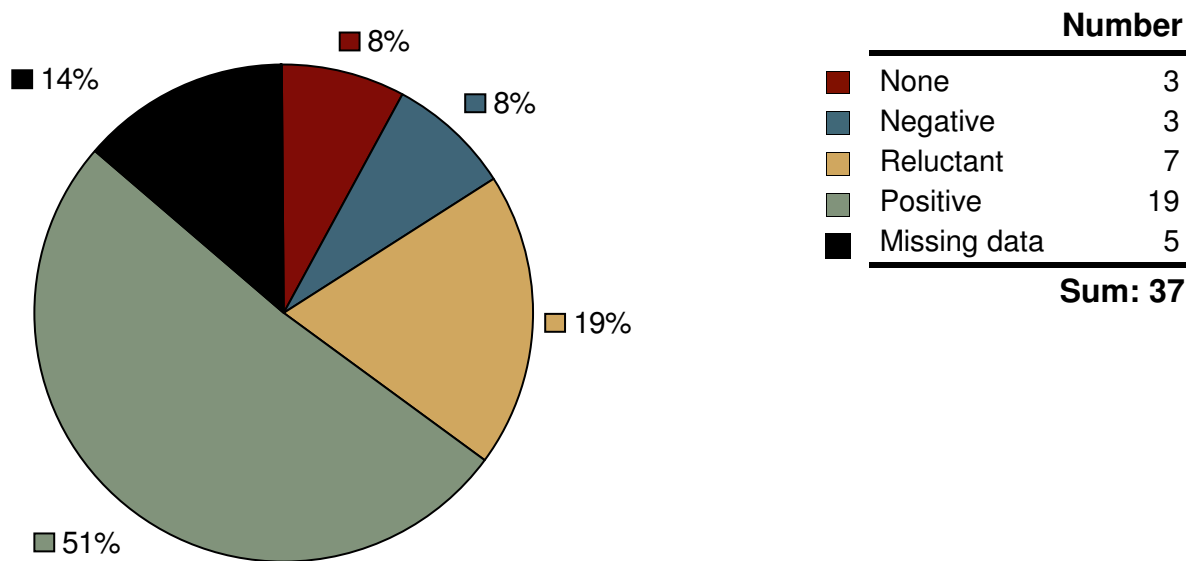
### Oral health index (indices)<sup>1</sup>

		0	1	2	3	4	5	6	Missing data	N
Calc	Calculus	9	0	0	0	0	0	0	4	13
GI	Gingivitis	8	1	1	0	0	0	0	3	13
Plaq	Coating	9	1	0	0	0	0	0	3	13
Toot	Tooth wear	7	2	0	0				4	13

- C 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.
- GI 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.
- PI 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.
- To 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.

<sup>1</sup>: Oral health index (indices) was (were) introduced in the observations in 2008

## Acceptance of dental examination



## Caries

	3-6 years	7-12 years	13-19 years	Adults
<b>deft<sup>1</sup></b>				
Examined		9	8	
Number of individuals with deft=0		6	6	
Mean		2,3	1,4	
Standard deviation		3,7	3,0	
Missing data		6	4	
<b>DMFT<sup>2</sup></b>				
Examined		10	7	3
Number of individuals with DMFT=0		9	4	0
Standard deviation		1,2	2,4	3,7
Mean		0,4	1,3	5,0
Missing data		2	0	0

1: Number of carious or filled deciduous teeth

2: Number of carious or filled permanent teeth

## Occlusal relationship

	<b>Number</b>
Neutral bite	23
Post normal	9
Pre normal	2
Missing data	3
<hr/>	
	<b>Sum: 37</b>

## Maximum jaw opening

Children younger than 10 years

	<b>Number</b>
- 20	0
21 - 30	0
31 - 40	7
41 - 50	3
51 -	0
Missing data	10
<hr/>	
	<b>Sum: 20</b>

Children, 10 years or older, and adults

	<b>Number</b>
- 20	0
21 - 30	0
31 - 40	5
41 - 50	9
51 -	0
Missing data	3
<hr/>	
	<b>Sum: 17</b>

## Profile<sup>1</sup>

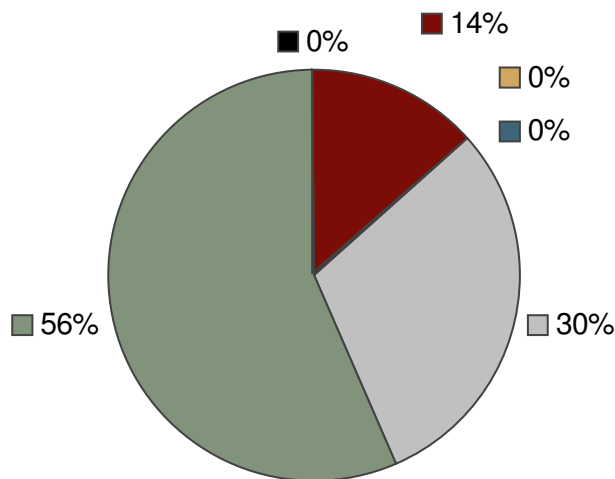
	<b>Number</b>
Normal	8
Convex	2
Concave	1
Missing data	2
<hr/>	
	<b>Sum: 13</b>

## Mandibular plane<sup>1</sup>

	<b>Number</b>
Normal	8
Increased	2
Reduced	0
Missing data	3
<hr/>	
	<b>Sum: 13</b>

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

## Speech difficulty



	<b>Number</b>
<span style="color: red;">■</span> No speech	5
<span style="color: gold;">■</span> Very incomprehensible	0
<span style="color: blue;">■</span> Incomprehensible speech	0
<span style="color: lightgray;">■</span> Slightly indistinct speech	11
<span style="color: green;">■</span> No problems	21
<span style="color: black;">■</span> Missing data	0
<b>Sum: 37</b>	

Clinical findings	Yes-answers			
	Total N=37 (%)	Boys/Men N=17 (%)	Girls/Women N=20 (%)	Missing data
Open mouth at rest	<b>14</b> (38)	5 (29)	9 (45)	0
Low muscle tone in lips	<b>11</b> (30)	3 (18)	8 (40)	0
High palate	<b>10</b> (30)	4 (27)	6 (33)	4
Frontal open bite	<b>9</b> (26)	4 (25)	5 (28)	3
Narrow palate	<b>7</b> (22)	3 (21)	4 (22)	5
Over crowding	<b>6</b> (18)	1 (7)	5 (28)	4
Impaired tongue motility	<b>5</b> (14)	0 (0)	5 (25)	1
M mentalis overactive	<b>4</b> (11)	1 (6)	3 (15)	0
Spacing	<b>3</b> (9)	2 (13)	1 (6)	3