Orofacial function of persons having Noonan syndrome

Report from observation charts

The survey comprises 26 observation charts.

Estimated incidence: 15-20 live births per year in Sweden.

Etiology: Autosomal dominant hereditary trait. Most frequently spontaneous mutation.

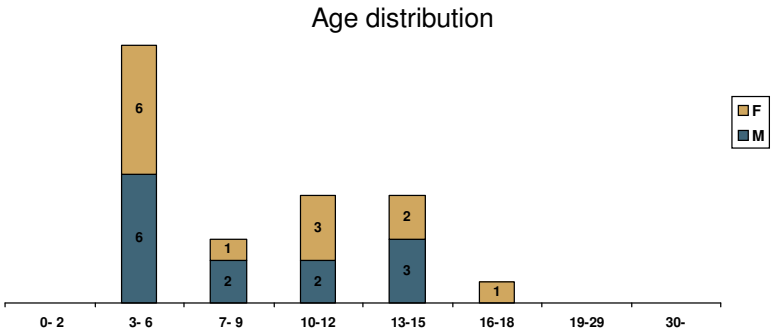
General symptoms: Most children born with Noonan syndrome have some kind of cardiac defect. Short stature -- Adult individuals with Noonan syndrome are generally about 15 cm shorter than predicted height. Growth hormone production deficit. Late onset puberty is common, and in many boys the testicles 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: A characteristic appearance with drooping eyelids, wide-set eyes, slanted eyes, an extra skin fold at the inner angle of the eyes, and short neck. 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 closely-spaced teeth have all been reported. However, it is difficult to establish exactly how much more common this is in children with Noonan than in others.

Orofacial/ odontological treatment:

- Children with eating disorders often required extra dental care, including assistance with oral hygiene and fluoride treatments. However, the dental services should not advise on eating difficulties.
- An orthodontist should be consulted between the ages of 7 and 9 in order to identify dental aberrations or malocclusions and to plan any necessary orthodontic treatment to correct bite problems.
- An increased tendency to bleed may result in complications when teeth are extracted.
- Individuals with cardiac defects may require prophylactic antibiotics when oral interventions associated with bleeding are undertaken.
- Training in mouth motor skills and extra stimulation in cases of eating disorders, speech difficulties and drooling may be necessary.
- Eating and swallowing problems will need to be investigated and treated by hospital specialist teams (either a nutrition team or a dysphagia team, or by other multidisciplinary treatment specialists.)

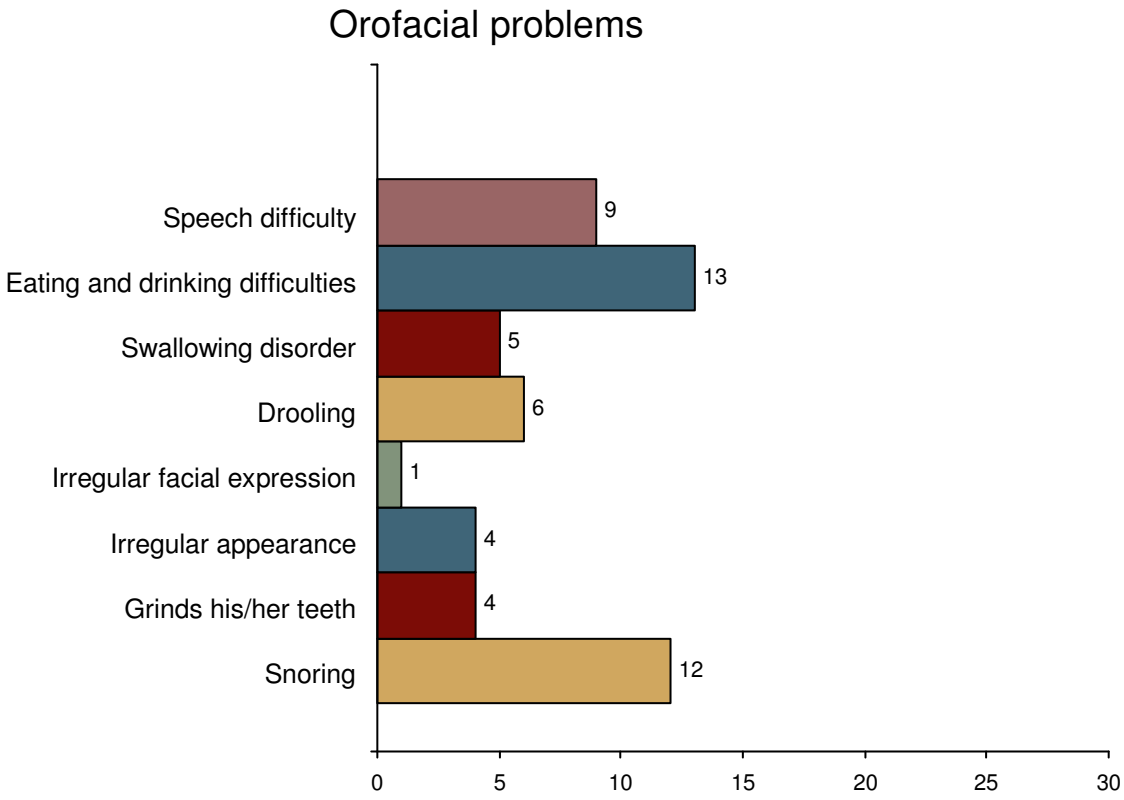
Source:
The rare disease database of the Swedish National Board of Health and Welfare.
The MHC database - The Mun-H-Center database of orofacial manifestations in rare diseases.
Noonan syndrom – folder from Smågruppscentrum.

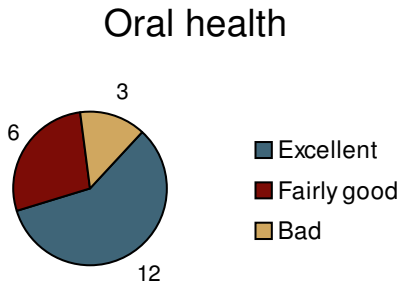


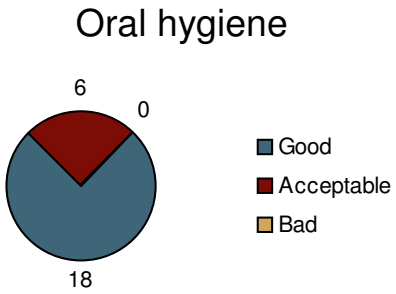
Number: 26

Ages: 3 -- 18 years

Sex: M (13) + F (13)

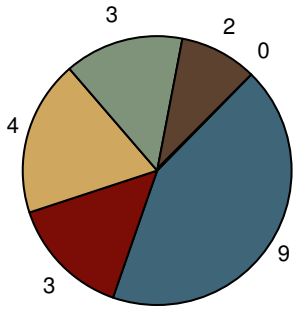






Behaviour in the treatment situation

How calm and co-operative is the patient at time of examination?



(9) Examination is possible w ithout problem

(3) Examination is possible w ithout problems, some reaction is observed

(4) Examination can continue if adjusted to patient's reactions

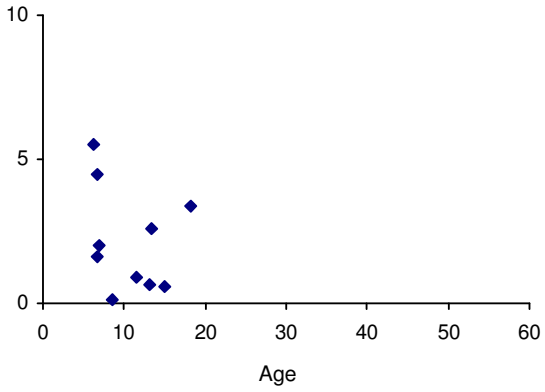
(3) Reactions are considerable and examination is obviously affected

(2) Examination is practically impossible to complete

(0) Patient refuses examination

How does the patient cope with treatment in general?

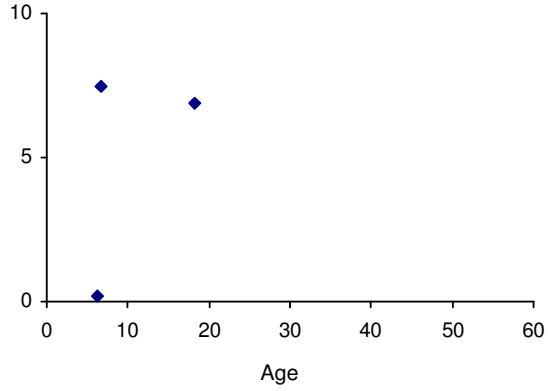
0=no problems/10=great problems



If there are treatment problems:

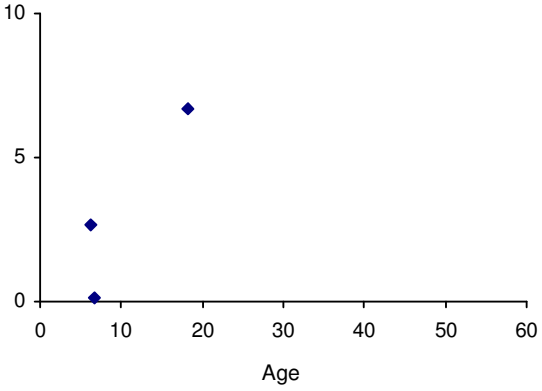
To what degree are the problems, if any, due to NN's handicap?

0=not at all/10=to a very high degree



To what degree are the problems, if any, due to NN's fear?

0=not at all/10=to a very high degree



Clinical findings	Total N=26	Boys/Men N=13	Girls/Women N=13	Not evaluated
Open mouth at rest	10	4	6	0
Hypomineralisation	9	4	5	3
Speech difficulty	9	2	7	0
Frontal open bite	8	4	4	0
Mask-like expression	8	2	6	0
Post normal bite	7	2	5	2
Drooling	6	3	3	1
Low muscle tone in lower lip	6	2	4	0
Low muscle tone in upper lip	6	2	4	0
High palate	5	3	2	2
Horizontal over-bite 6 mm or more	5	1	4	3
Hypodontia	4	1	3	12
M mentalis is overactive	4	1	3	0
Narrow palate	4	2	2	3
Other oral habits	4	2	2	0
Over crowding	4	1	3	2
Abrasion - significant	3	1	2	3
Gingivitis	3	1	2	2
Grinds his/her teeth	3	1	2	3
Molar contact only	3	2	1	2
Pre normal bite	3	1	2	2
Reduced mobility in tongue	3	0	3	0
Spacing	3	2	1	1
Cross bite	2	0	2	2
Edge to edge bite	2	1	1	3
Frontal inversion	2	1	1	2
Hyper-sensitive in oral cavity	2	1	1	2
Lower jaw seems small	2	0	2	2
Mouth breathing	2	1	1	10
Posturing forward	2	0	2	3
Reduced mobility in soft palate	2	0	2	7
Retroclined lower incisors	2	2	0	2
Short tongue frenulum	2	1	1	1
Tongue between front teeth	2	1	1	0
Tooth anomaly	2	1	1	4