



Orofacial function of persons having Noonan syndrome

Report from questionnaires

The survey comprises 26 Questionnaires.

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:

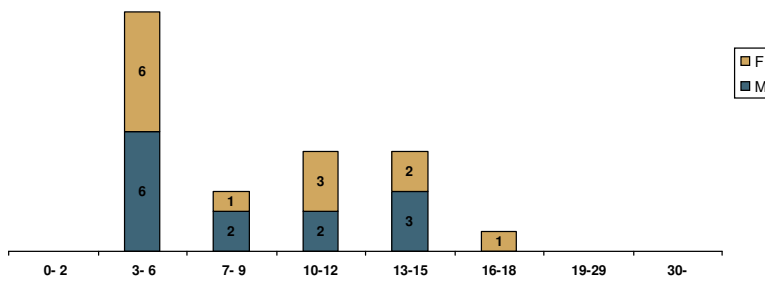
- Children with eating disorders often required extra dental care, including assistance with oral hygiene and fluoride treatments. However, the dental services should not advise on eating difficulties.
- An orthodontist should be consulted between the ages of 7 and 9 in order to identify dental aberrations 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.

Age distribution

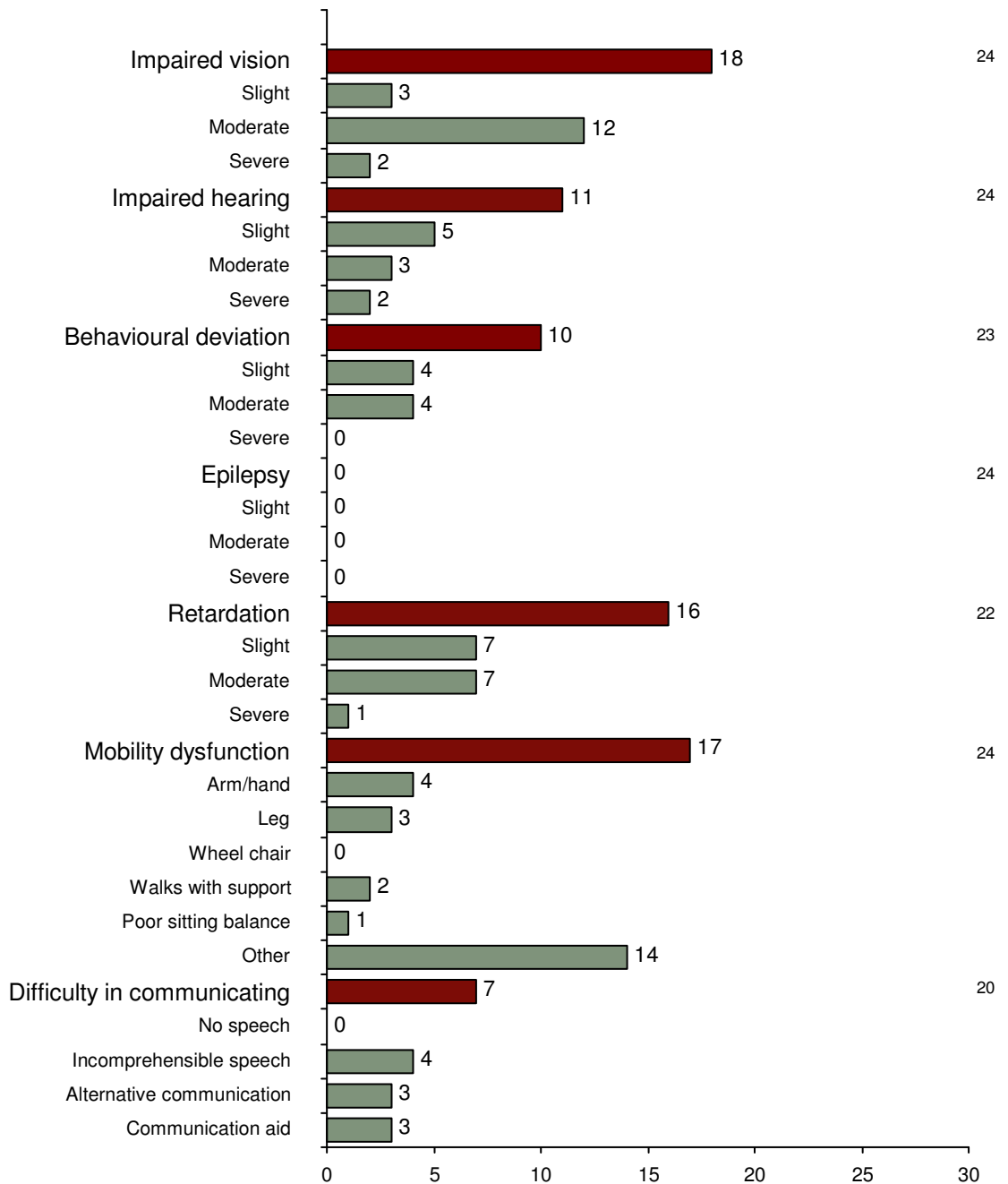


Number: 26

Ages: 3 -- 18 years

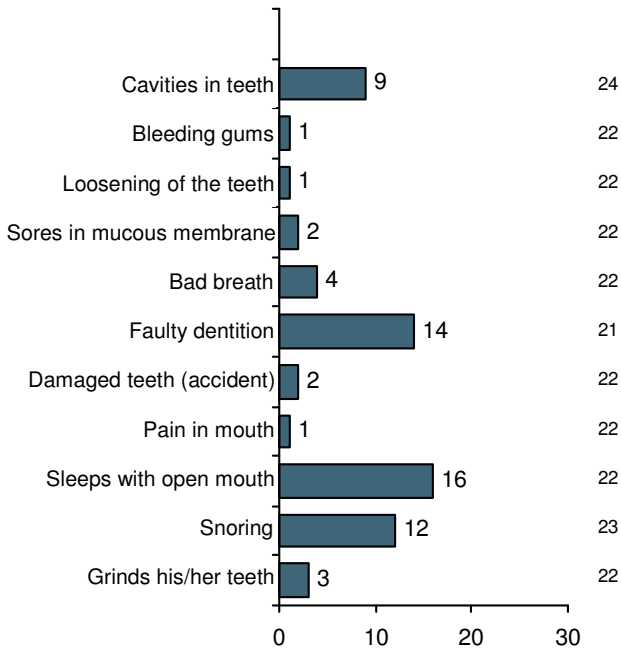
Sex: M (13) + F (13)

General disabilities

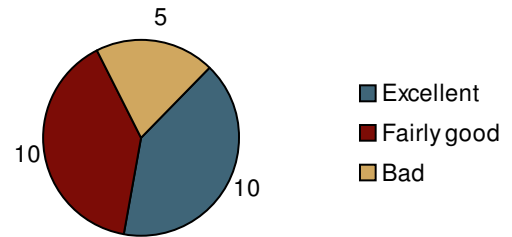


About dental health

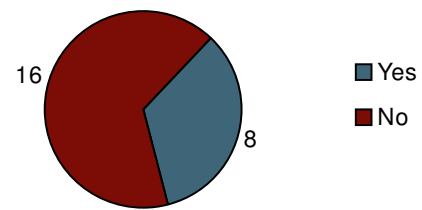
About dental health - problems



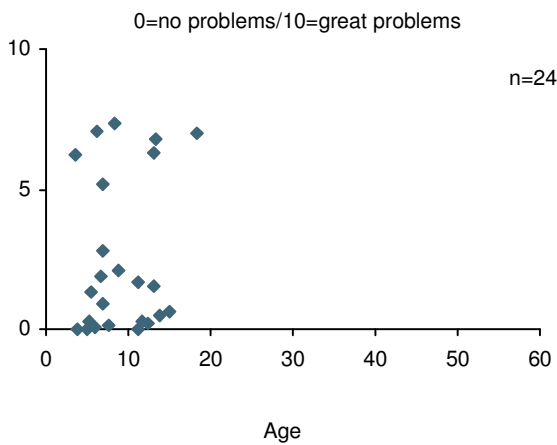
Oral health



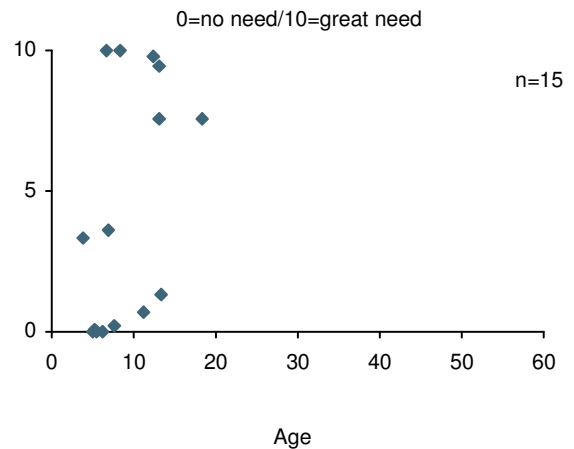
Has NN received orthodontic treatment?



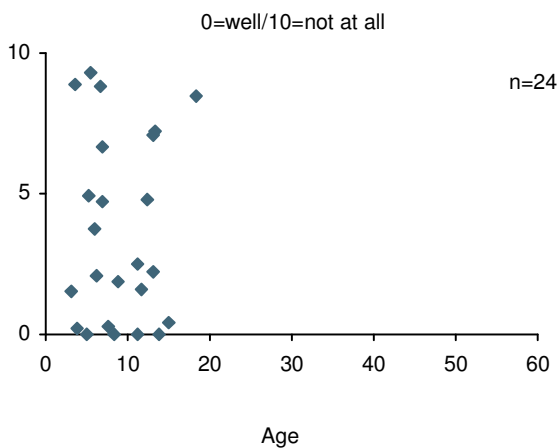
How does NN experience the dental care received?



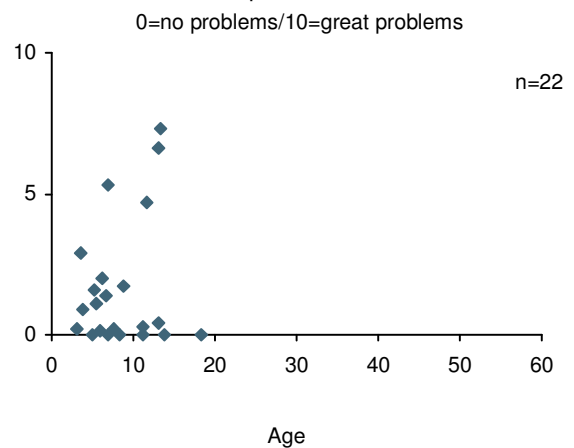
NN is considered to be in need of orthodontic treatment



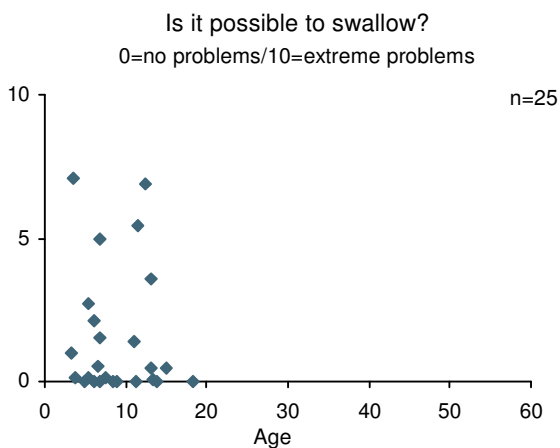
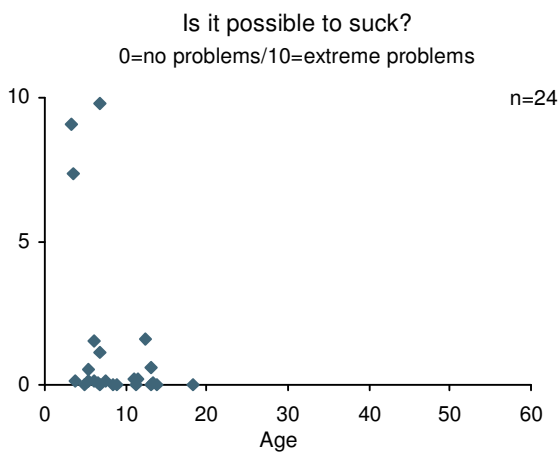
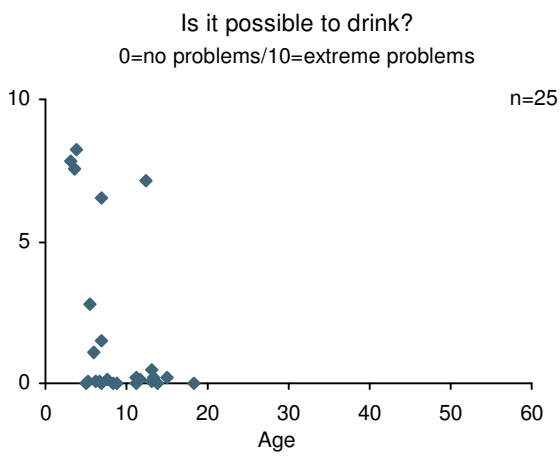
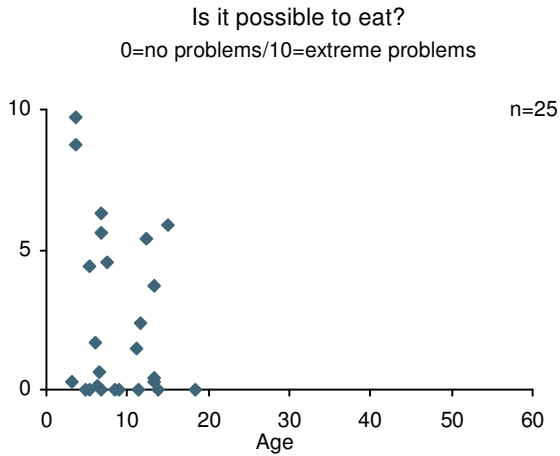
How does NN manage to brush his/her teeth?



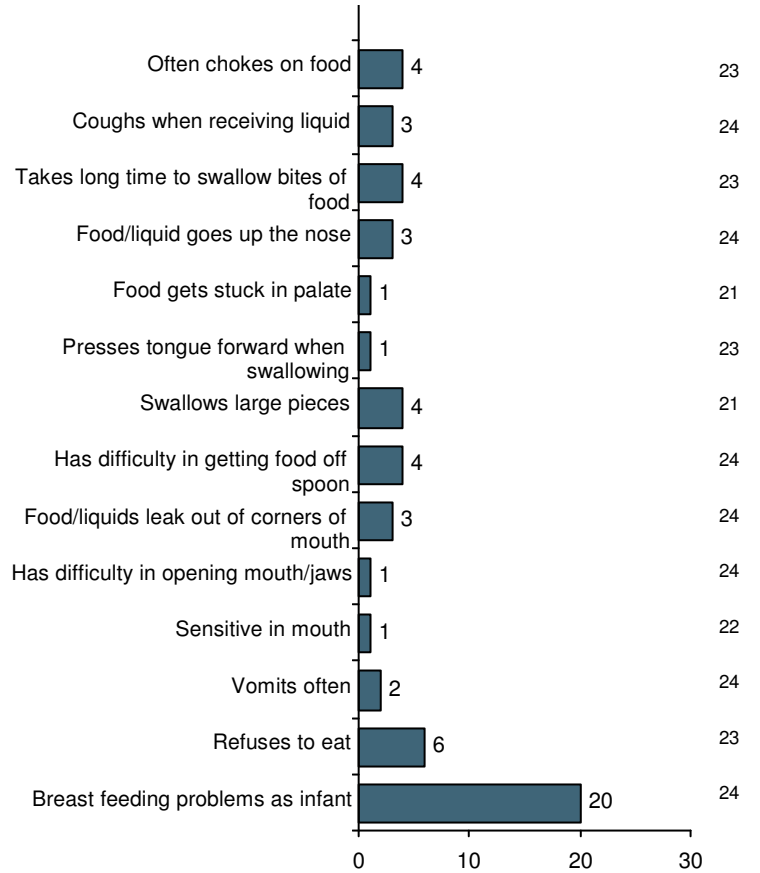
If help is needed with brushing teeth, how does it proceed?



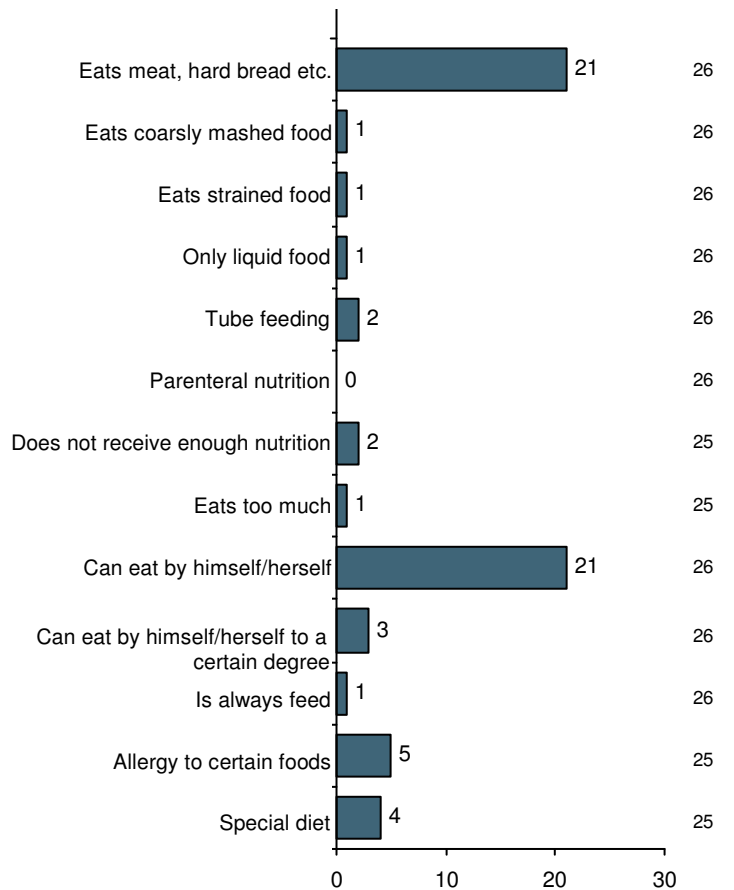
About eating habits



About eating habits - problems



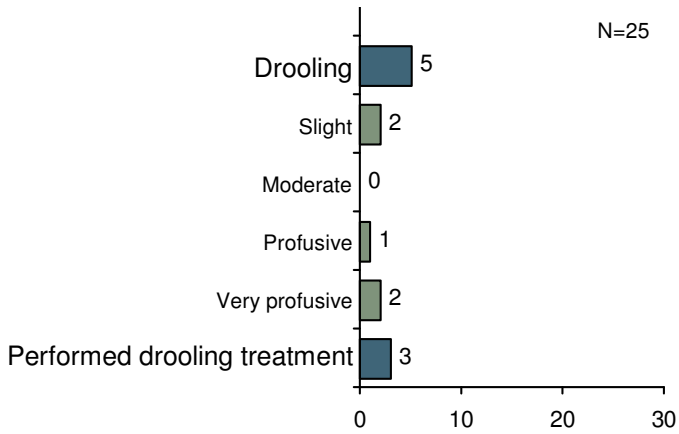
Food habits



About drooling

Drooling

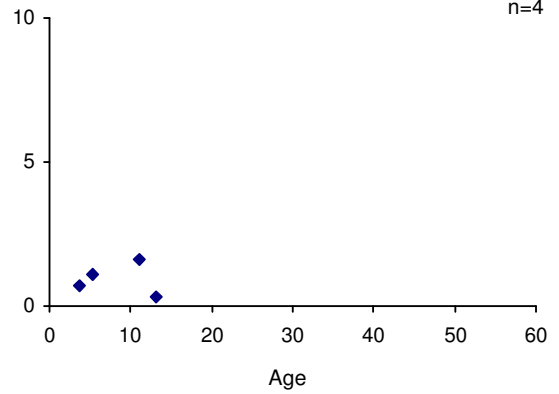
N=25



How does NN experience his/her drooling?

0=no problem/10=serious problem

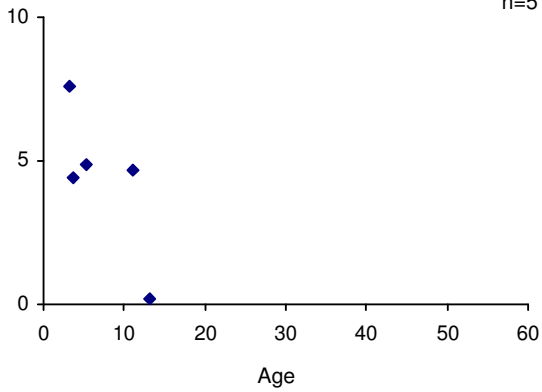
n=4



How does parent/attendant experience the drooling?

0=no problem/10=serious problem

n=5



How do people around NN experience the drooling?

0=no problem/10=serious problem

n=5

