



Orofacial function of persons having Apert syndrome Report from questionnaires

The survey comprises 17 Questionnaires.

Synonym: Acrocephalosyndactyly type 1

Estimated occurrence: 1:100 000 live births.

Etiology: Autosomal dominant genetic trait. Most often spontaneous mutation. Diagnosis based on symptoms.

General symptoms: Cranial and malformations, malformations of the hands and feet. Fusion of the cervical vertebrae is frequent. Children with Apert Syndrome often have delayed speech and language development, as well as learning disabilities. They also generally have vision and hearing problems. Severe acne is common while growing up.

Orofacial/odontological symptoms: Premature fusion of the bones at the base of the skull gives rise to craniofacial malformations. The head is broad and elongated, the midface underdeveloped, and the eyes bulge. The palate is often high and narrow, and some children are born with a cleft soft palate. Crowded teeth and an underbite are common. Constriction in the upper respiratory system may lead to breathing difficulties and sleep apnea (frequently stopping breathing when asleep). Craniofacial malformations lead to articulation difficulties and impair chewing ability.

Orofacial/odontological treatment:

In Sweden, treatment of craniofacial malformations is centralized, and carried out at Sahlgrenska University Hospital, Göteborg.

- Most individuals with Apert syndrome require both orofacial surgery and extensive orthodontia.
- Speech and language disabilities are dealt with by a speech therapist.
- Many of these individuals require supplementary prophylactic dental care.
- Snoring problems should be followed up by a physician.

Source

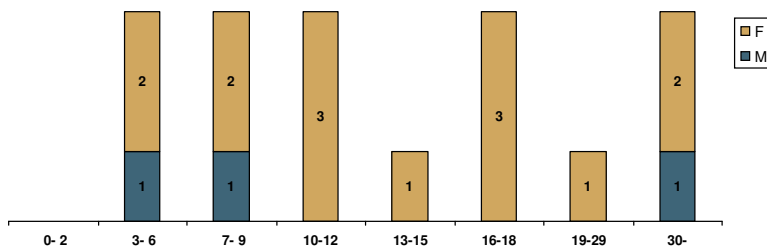
The rare disease database of the Swedish National Board of Health and Welfare.

The MHC database - The Mun-H-Center database of orofacial manifestations in rare diseases.

The Newsletter of the Ågrenska Center.

Apert syndrom – folder from Smågruppscentrum

Age distribution

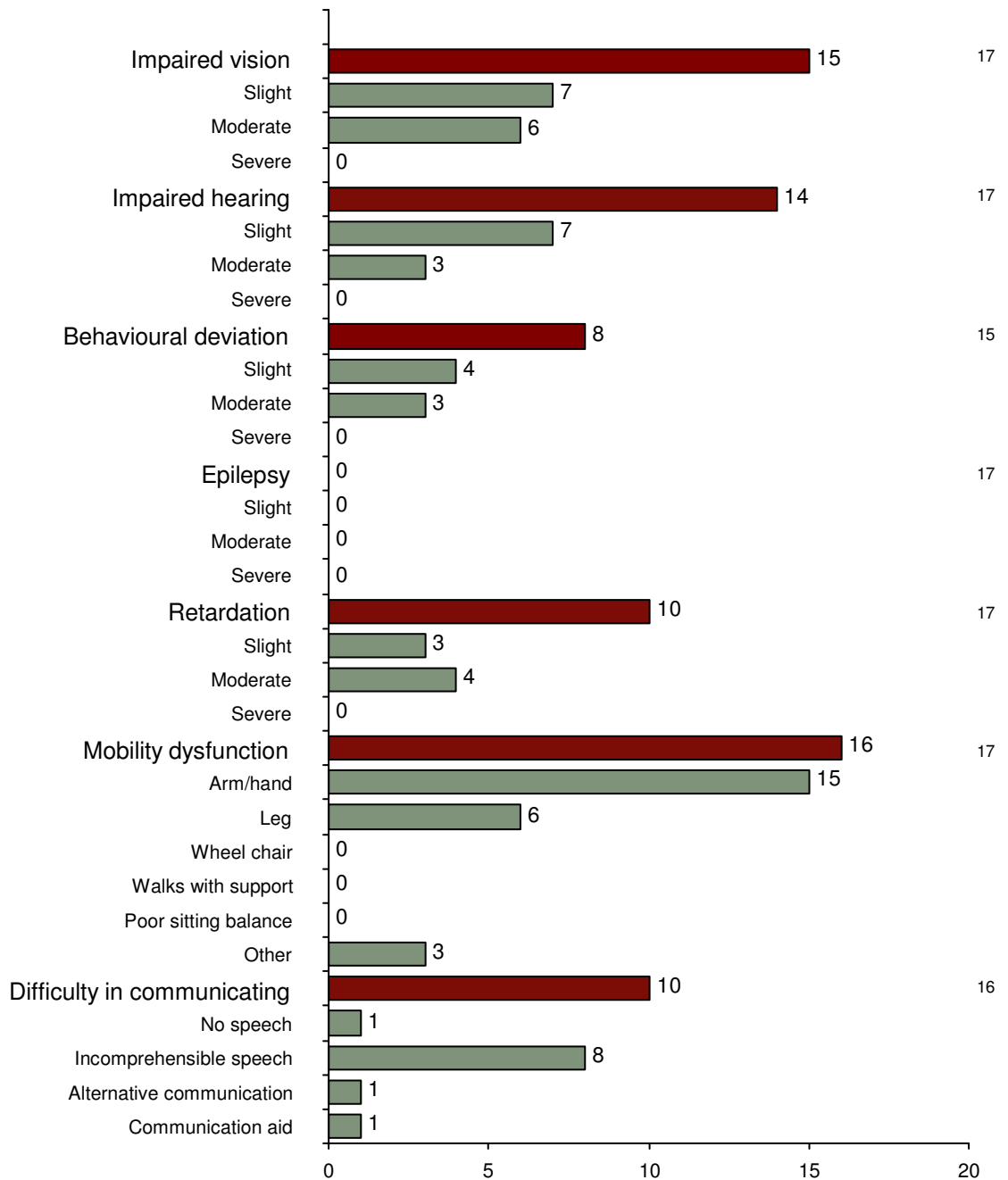


Number: 17

Ages: 4 -- 35 years

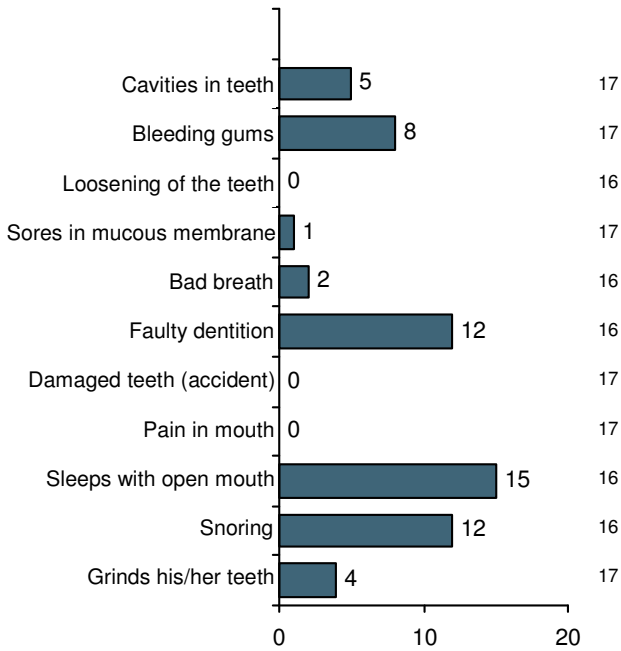
Sex: M (3) + F (14)

General disabilities

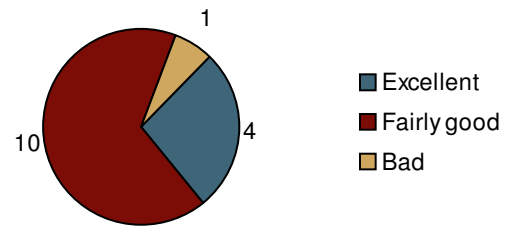


About dental health

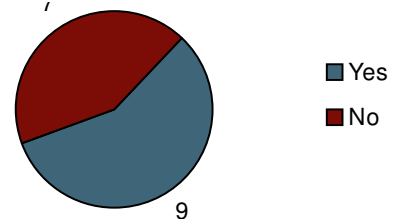
About dental health - problems



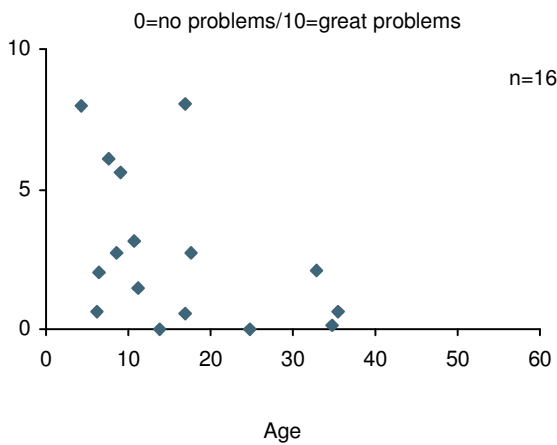
Oral health



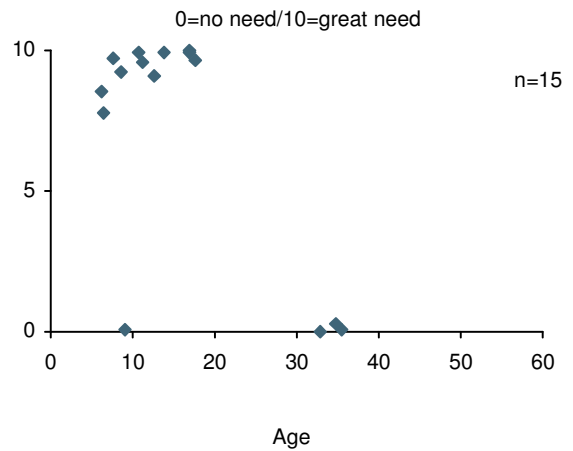
Has NN received orthodontic treatment?



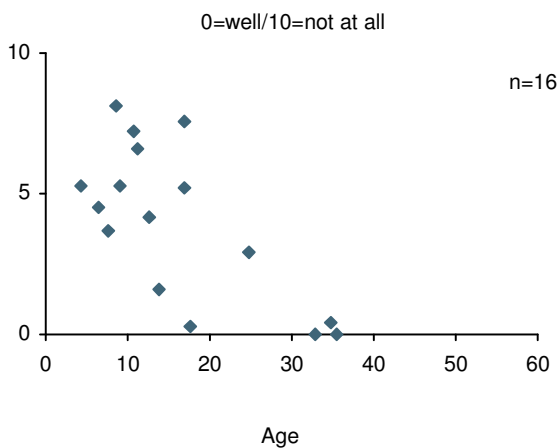
How does NN experience the dental care received?



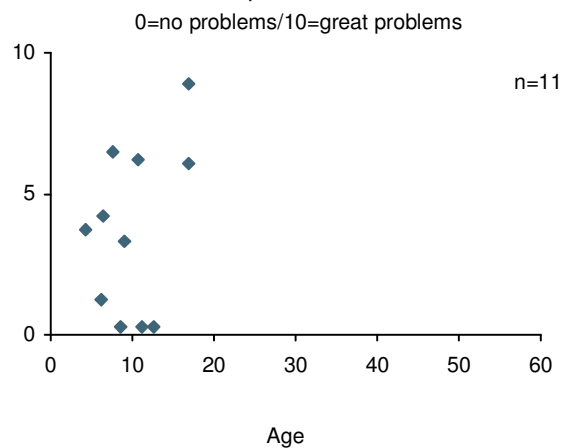
NN is considered to be in need of orthodontic treatment



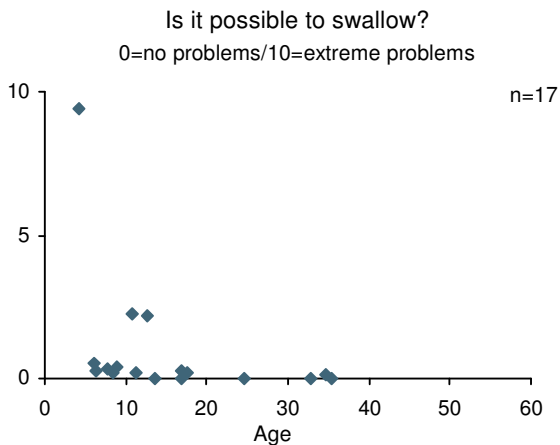
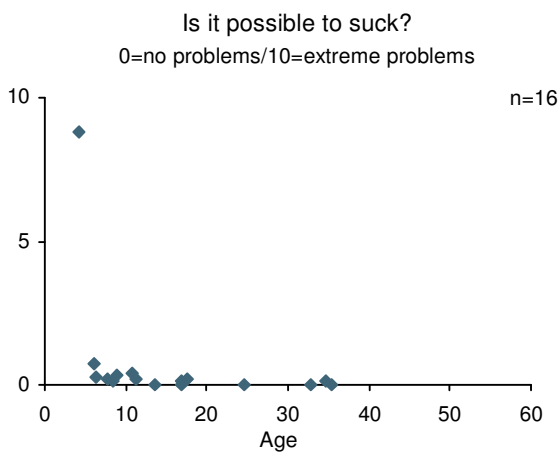
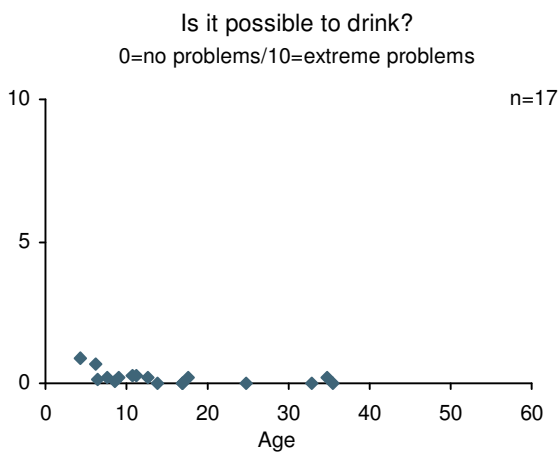
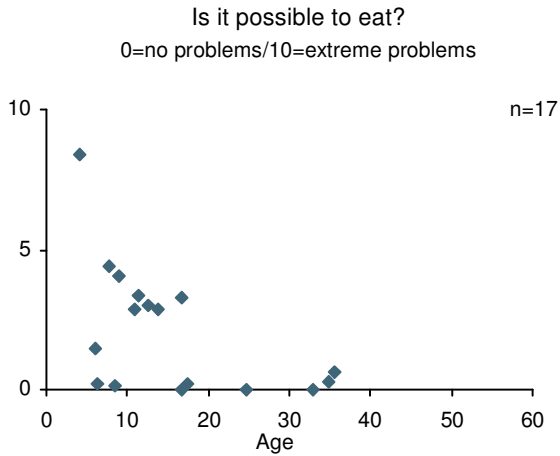
How does NN manage to brush his/her teeth?



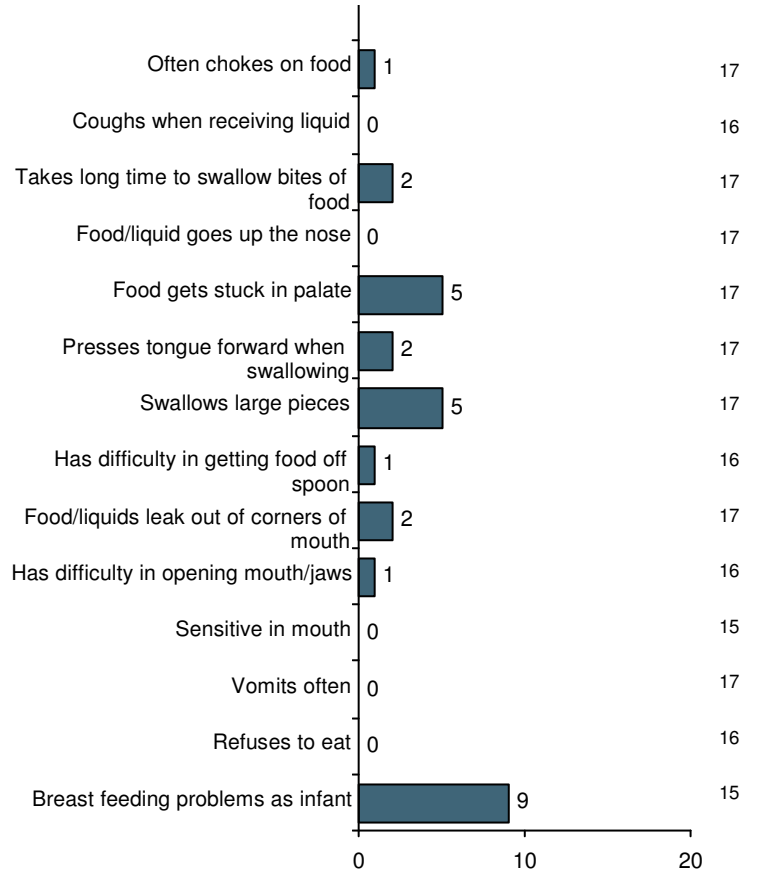
If help is needed with brushing teeth, how does it proceed?



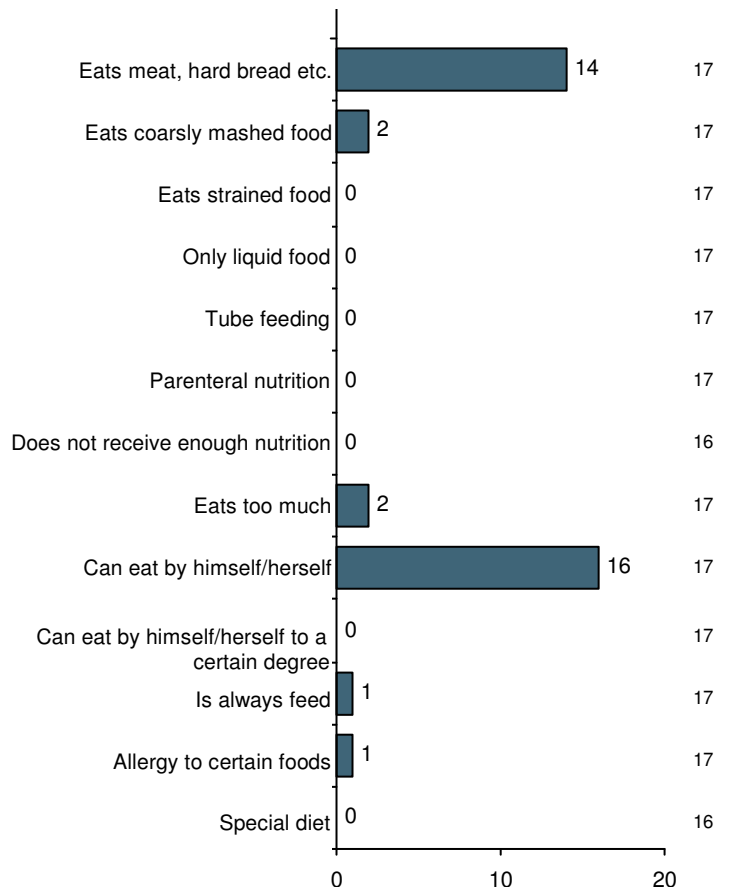
About eating habits



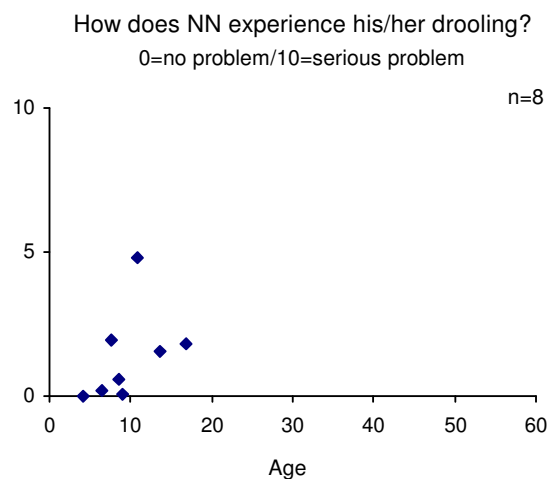
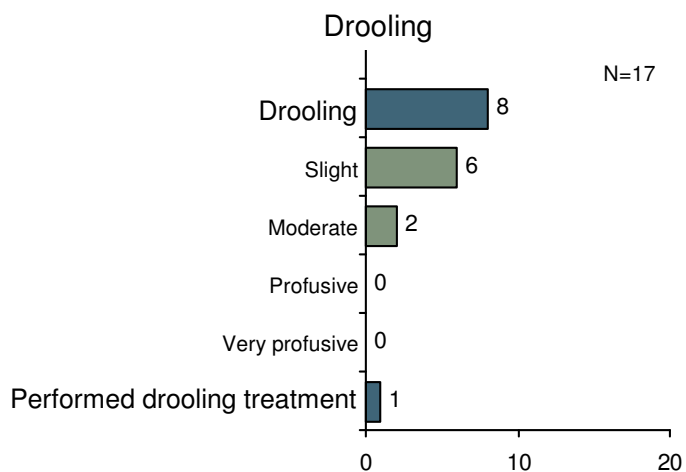
About eating habits - problems



Food habits

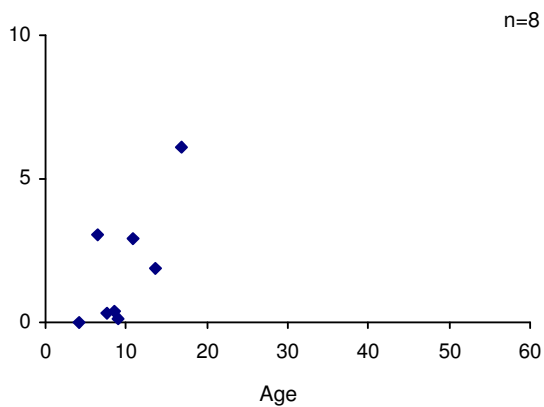


About drooling



How does parent/attendant experience the drooling?

0=no problem/10=serious problem



How do people around NN experience the drooling?

0=no problem/10=serious problem

