



Orofacial function of persons having Neurofibromatosis 1 Report from questionnaires

The survey comprises 51 Questionnaires.

Synonym: Recklinghausen's disease

Estimated occurrence: 1:3,000 inhabitants (Neurofibromatosis type 1)

Etiology: The locus of the gene that causes this syndrome is the long arm of chromosome 17. Autosomal dominant heredity. 50-60% of the occurrences are spontaneous mutations.

General symptoms: Café au lait spots and neurofibromas of the skin are characteristic. The latter are benign tumors that develop in the supportive tissue around the nerves. There may be up to several hundred of them. One-third of affected individuals develop plexiform neurofibromas, which are not nearly as restricted as neurofibromas of the skin. This disease, or more correctly disorder, may also impair the ocular and auditory nerves, the central nervous system, and skeletal development. Endocrine tumours occur. Children with this diagnosis should have annual physical examinations. Some developmental delay, learning difficulties and concentration problems may occur, as well as epilepsy. Scoliosis is occasionally found.

Orofacial/odontological symptoms: Neurofibromas may occur in the oral mucous membranes, but these are normally not treated unless they grow or become uncomfortable. Neurofibromas may also occur in the jawbone, where they may be found with the aid of general radiographs. If they are found, they should be checked at regular intervals. There may be enlarged papillae on the tongue. Early tooth eruption has been reported, as well as abnormal tooth positions. Eating and speech difficulties and drooling are found, as well as occasional sleep apnea (frequent suspension of breathing while asleep).

Orofacial/ odontological treatment:

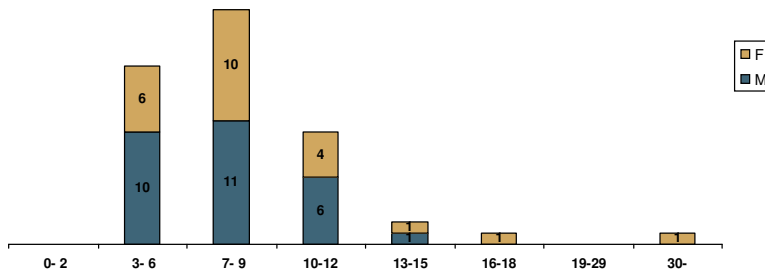
- Many of these children require extra preventive dental care.
- Training in oral motor skills and extra stimulation in cases of eating disorders, speech difficulties and drooling may be necessary.
- Speech, language and communication training are often justified.
- Snoring problems should be followed up by a physician.
- 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.

Sources:

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

The Newsletter of the Ågrenska Center.

Age distribution

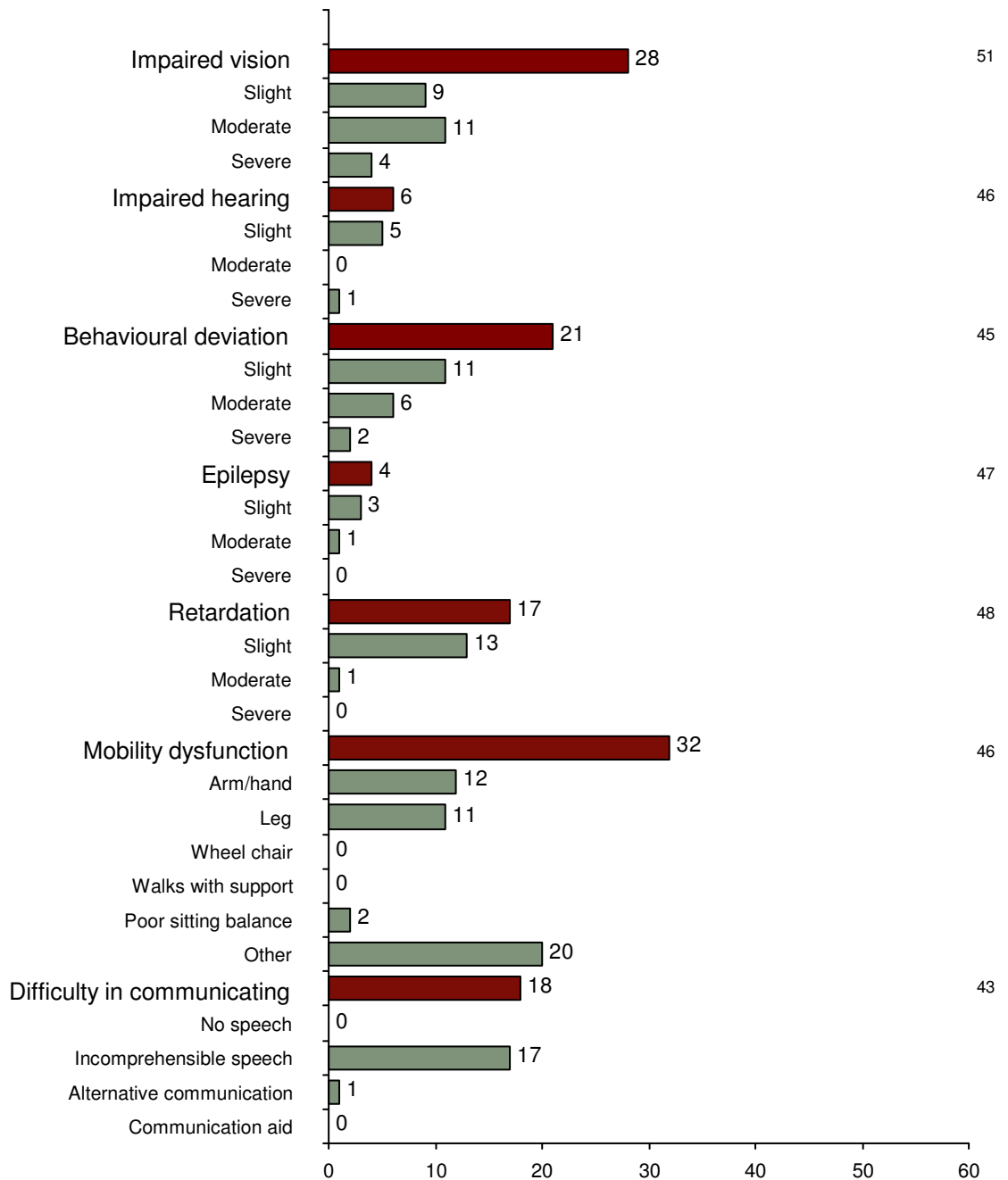


Number: 51

Ages: 3 -- 51 years

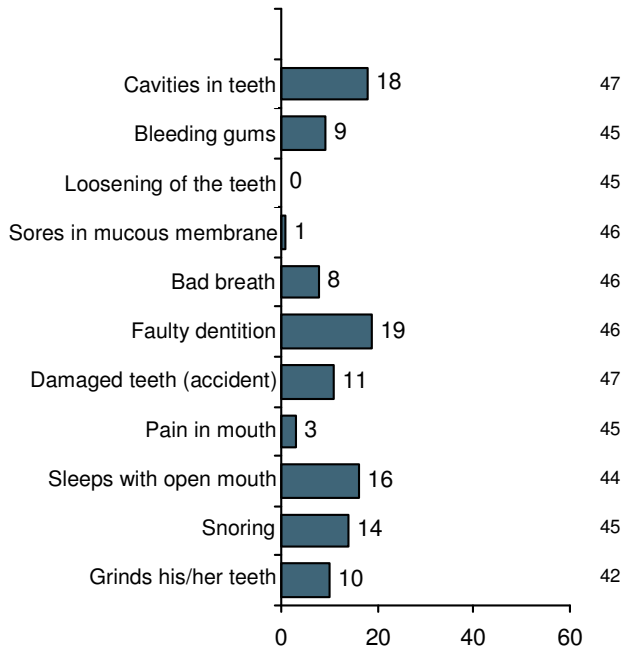
Sex: M (28) + F (23)

General disabilities

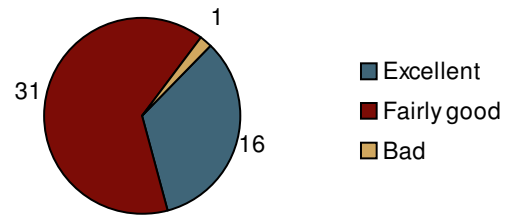


About dental health

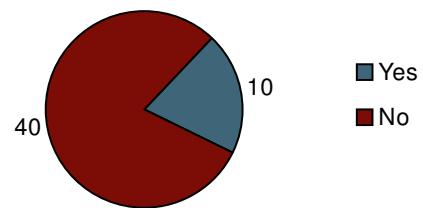
About dental health - problems



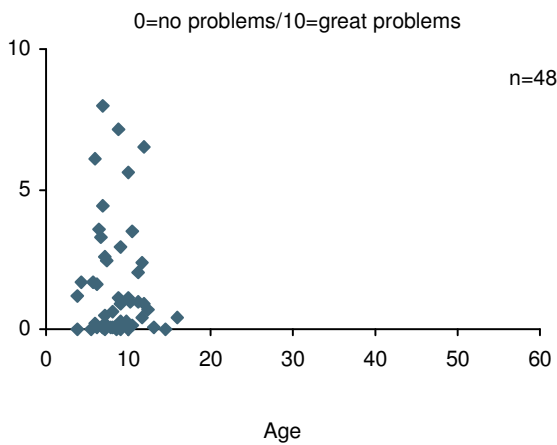
Oral health



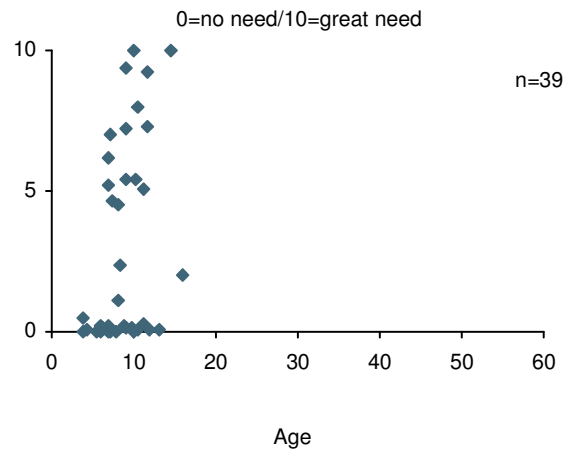
Has NN received orthodontic treatment?



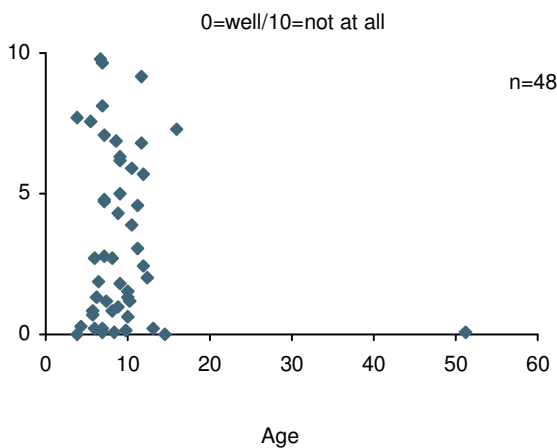
How does NN experience the dental care received?



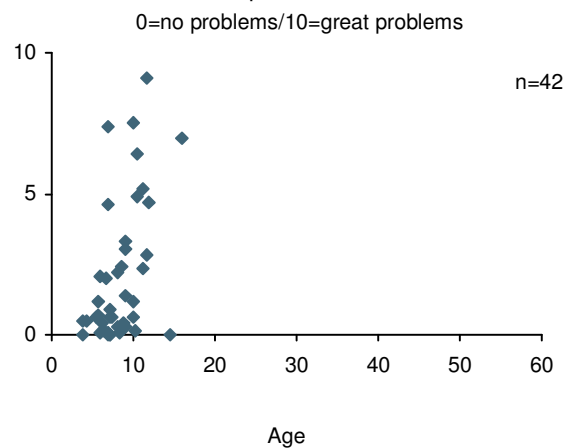
NN is considered to be in need of orthodontic treatment



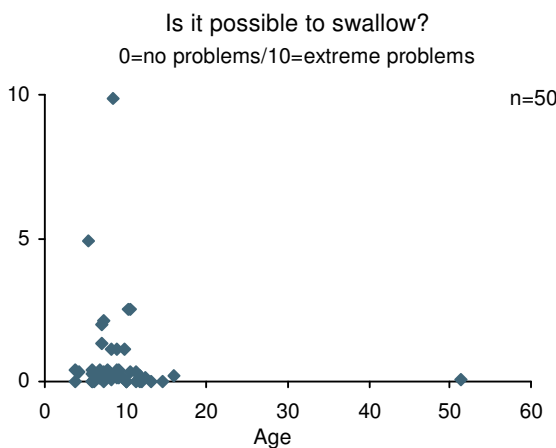
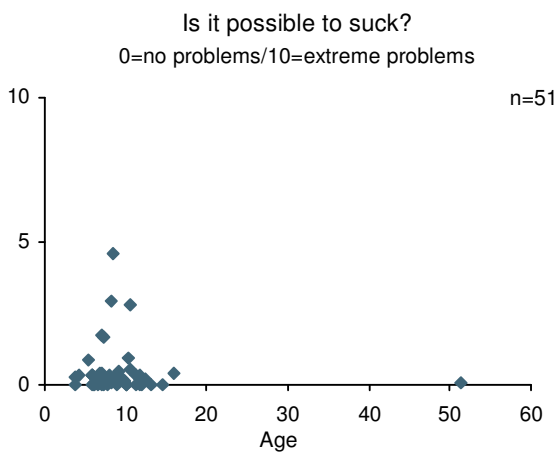
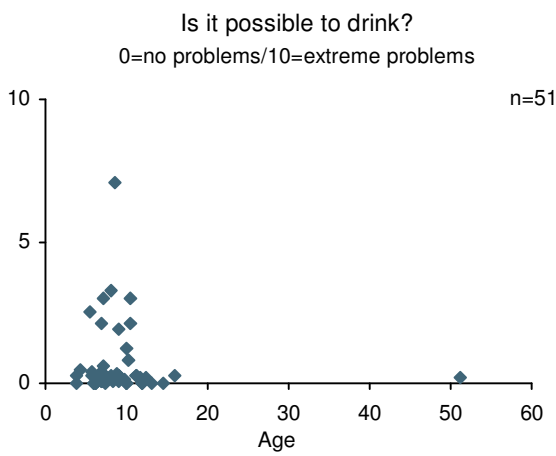
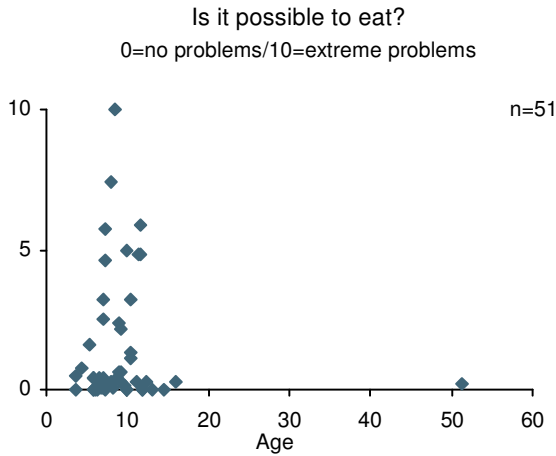
How does NN manage to brush his/her teeth?



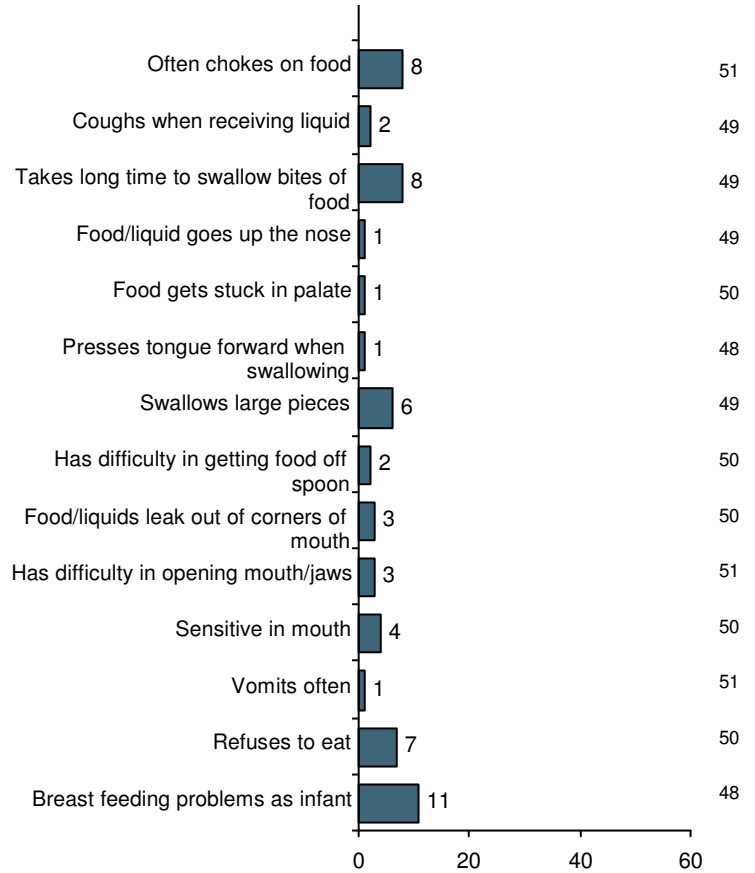
If help is needed with brushing teeth, how does it proceed?



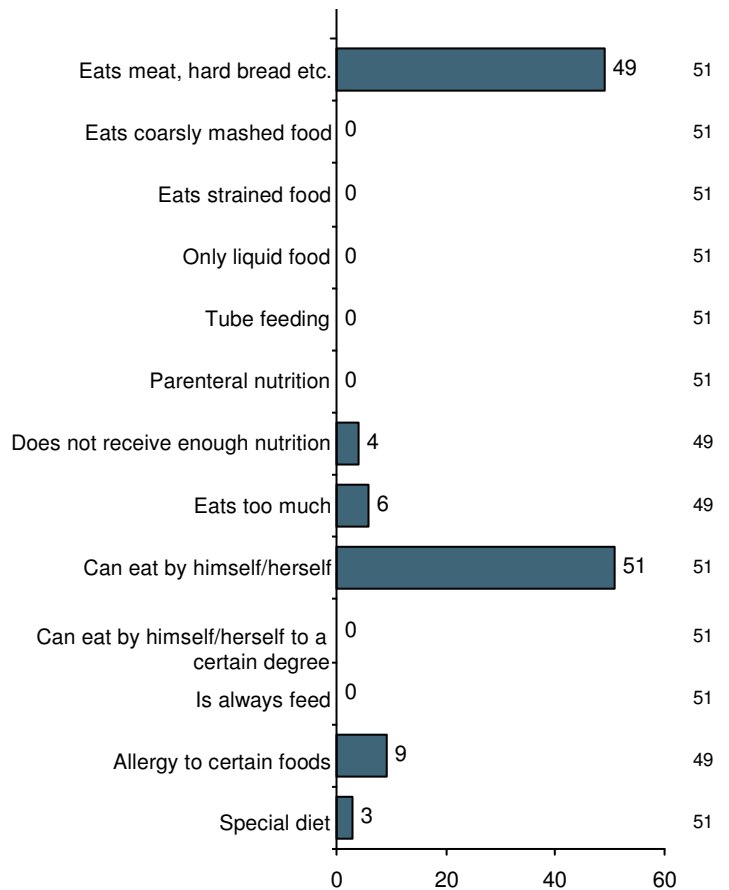
About eating habits



About eating habits - problems



Food habits



About drooling

