



Orofacial function of persons having Spinal muscular atrophy

Report from questionnaires

The survey comprises 38 Questionnaires.

Synonyms: SMA I (Werdnig-Hoffmann disease, SMA II, SMA III (Kugelberg-Welander disease).

Estimated occurrence: Every year, 4-6 children in Sweden develop the most severe form of spinal muscular atrophy - SMA I. SMA II and III are somewhat less common. For an unknown reason, SMA is somewhat more common in boys than in girls.

Aetiology: Defect on the SMN1-gene on chromosome 5. SMA types I, II and III are inherited via autosomal recessive inheritance. Spinal muscular atrophy (SMA) is caused by destruction of the motor neurons in the diencephalon section of the brain, the medulla and the anterior horn cells of the spinal cord, resulting in muscular weakness and atrophy.

General symptoms: Muscular weakness and atrophy are most pronounced in the proximal musculature, including the chest, back and neck muscles. Intellectual development is not affected.

SMA I (Werdnig-Hoffmann disease): Symptoms from birth, or prior to the age of 6 months. Respiratory function is severely affected, and these children are at high risk from infections.

SMA II (known as the intermediate form): Symptoms present around the age of 6-18 months. Muscular weakness is often more pronounced in the legs than in the arms. Scoliosis (curvature of the spine) is common. Respiratory function may be affected.

SMA III (Kugelberg-Welander disease): Presents around the age of 2 years. Weak musculature in the trunk (proximal), back problems, and ambulatory difficulties are common and increase with age.

Sometimes, SMA 0 is used for the severe congenital type and SMA IV for the milder type with adult onset.

Orofacial/odontological symptoms: Infants with SMA I are unable, owing to their weak neck musculature, to lift their heads. Musculature weakness in the throat affects the ability to suck and to swallow. The musculature of the tongue is also weak, and fasciculations (slight trembling) in the tongue may occur. Adolescents and adults with SMA sometimes develop an impaired jaw opening capacity. Some have malocclusion

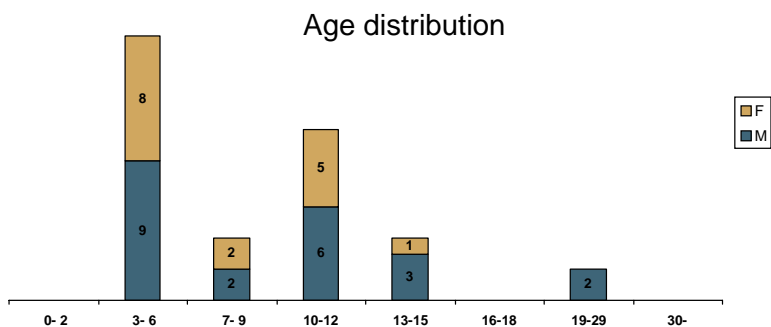
Orofacial/odontological treatment:

- Early contact with dental services for intensified prophylactic care and oral hygiene information is essential.
- Regular check-ups of dental and jaw development. Orthodontist should be consulted when needed.
- When the jaws do not open properly, the function of the jaw joint should be investigated, and appropriate treatment thereafter prescribed.
- Feeding and swallowing difficulties are investigated and treated by a specialist team at the hospital or multidisciplinary treatment center.

Sources

The rare diseases 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.

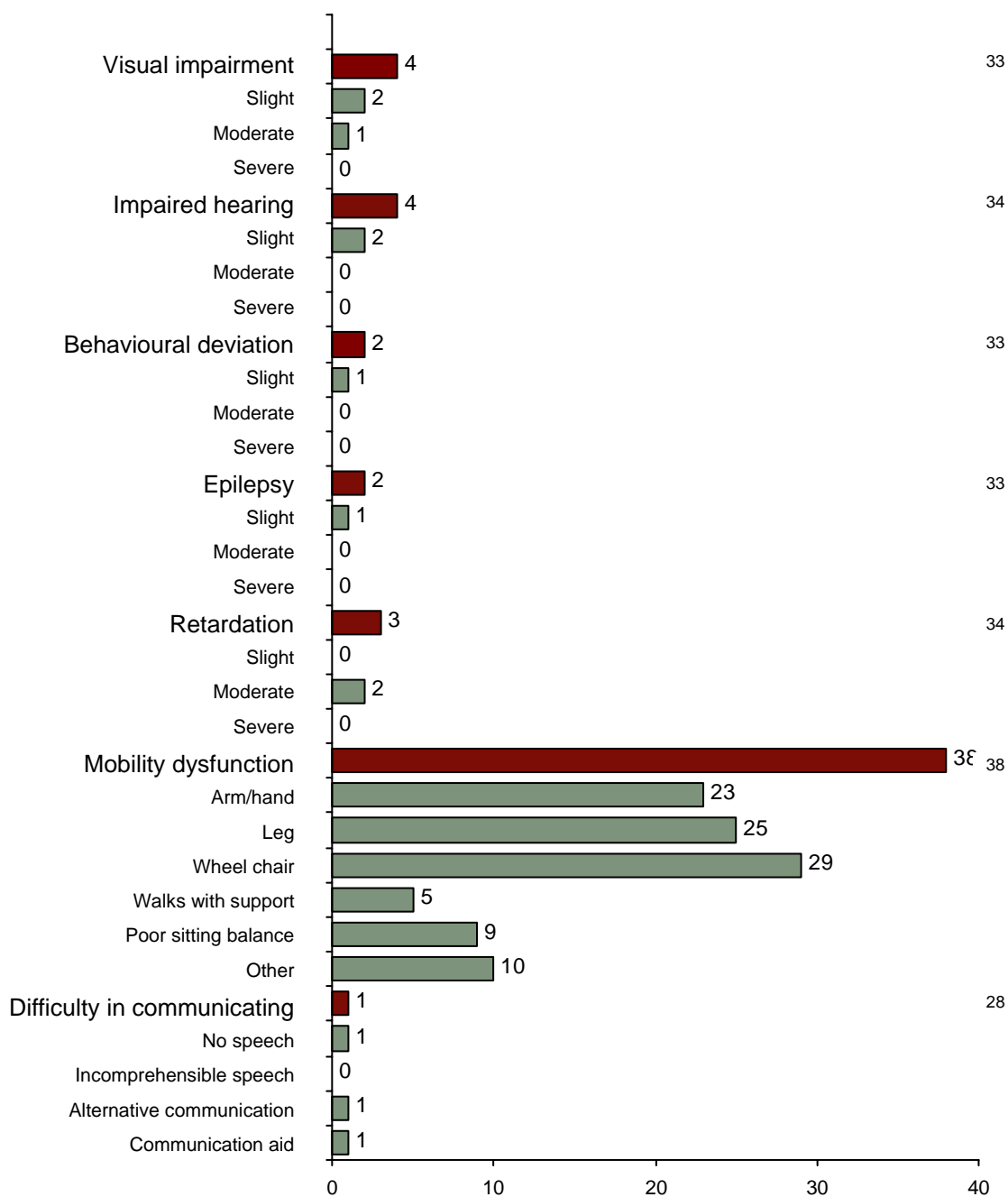


Number: 38

Ages: 3 -- 27 years

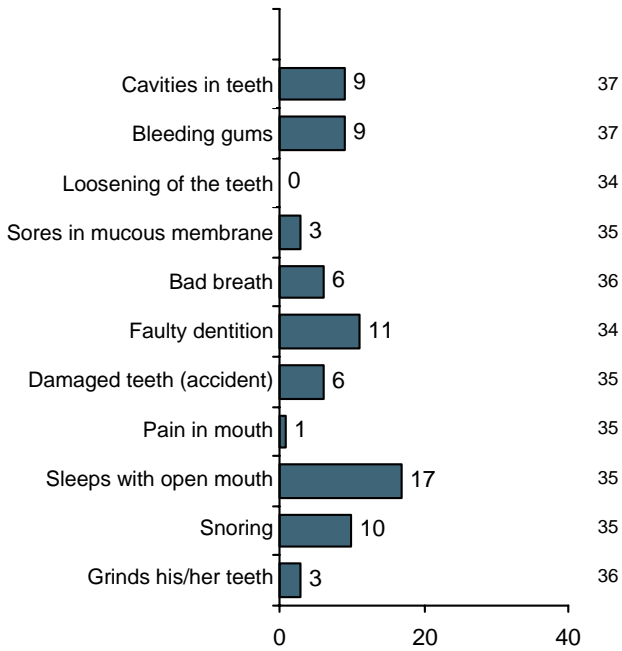
Sex: M (22) + F (16)

General disabilities

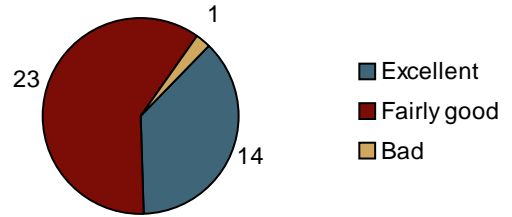


About dental health

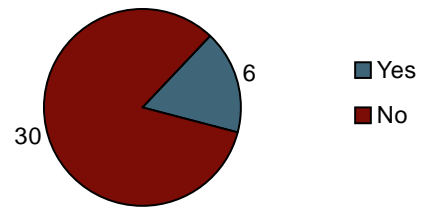
About dental health - problems



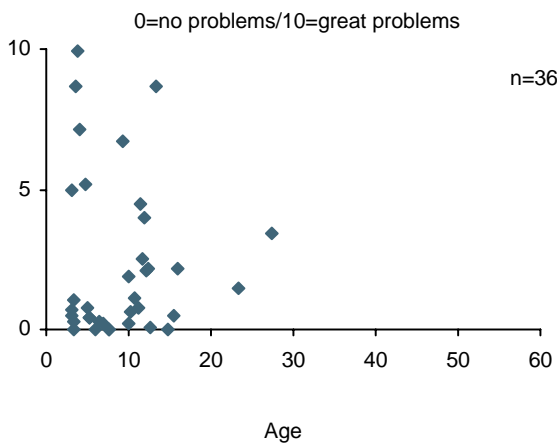
Oral health



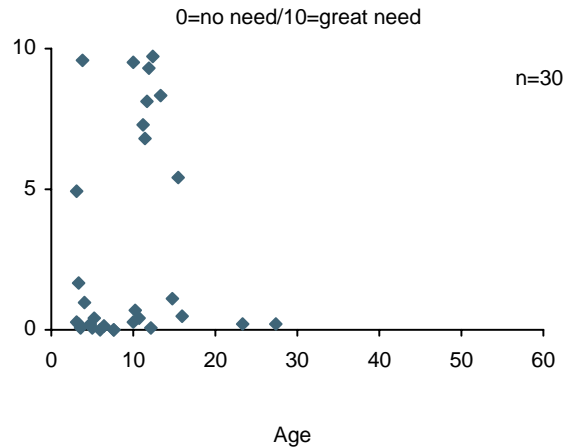
Has NN received orthodontic treatment?



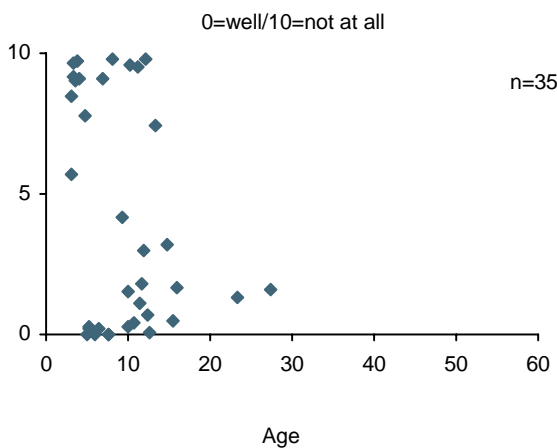
How does NN experience the dental care received?



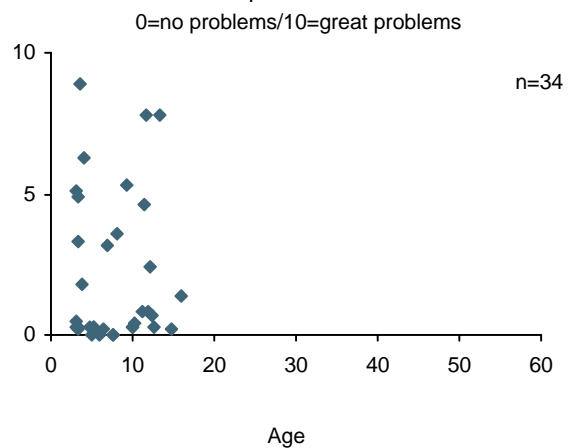
NN is considered to be in need of orthodontic treatment



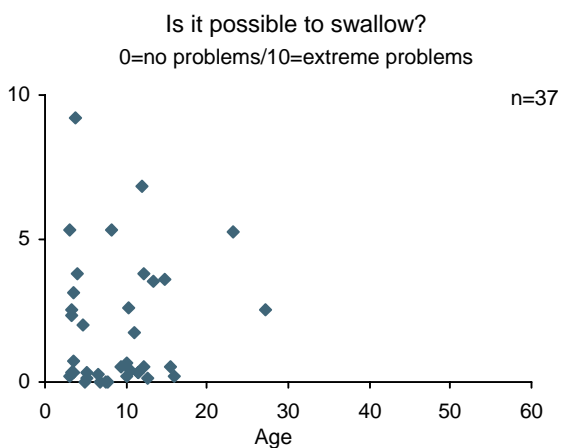
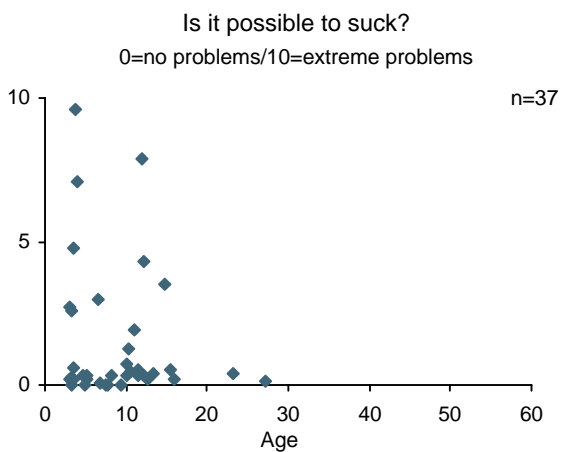
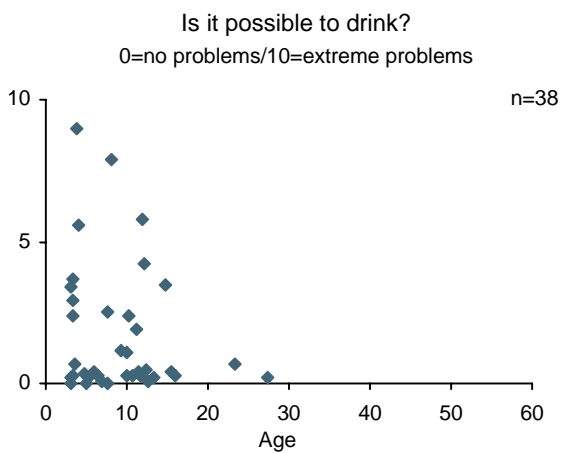
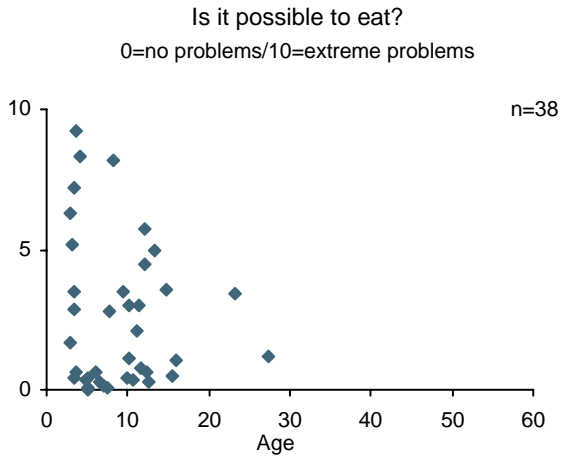
How does NN manage to brush his/her teeth?



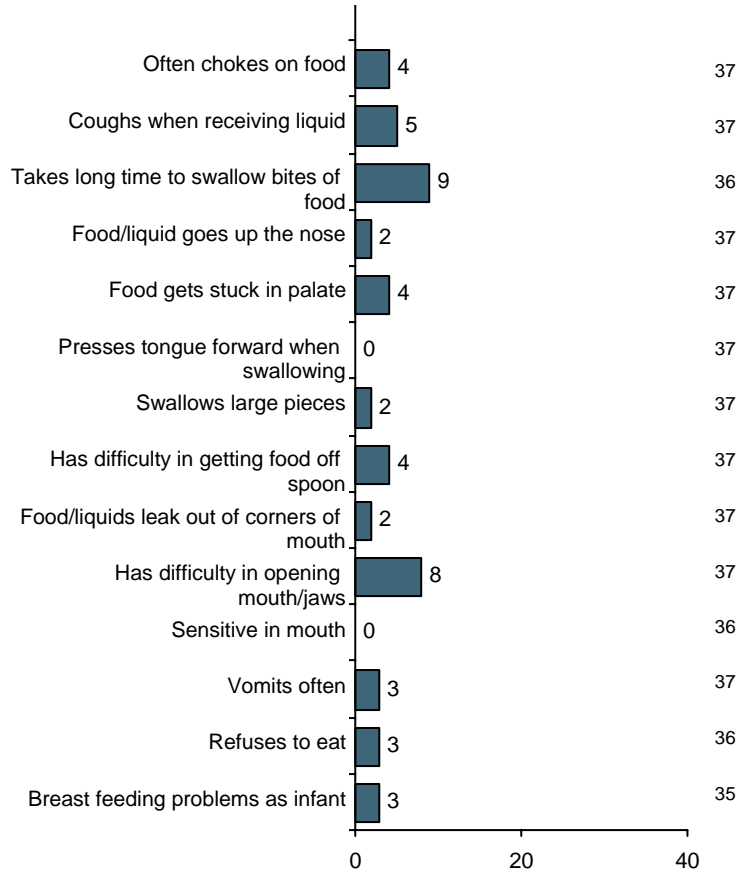
If help is needed with brushing teeth, how does it proceed?



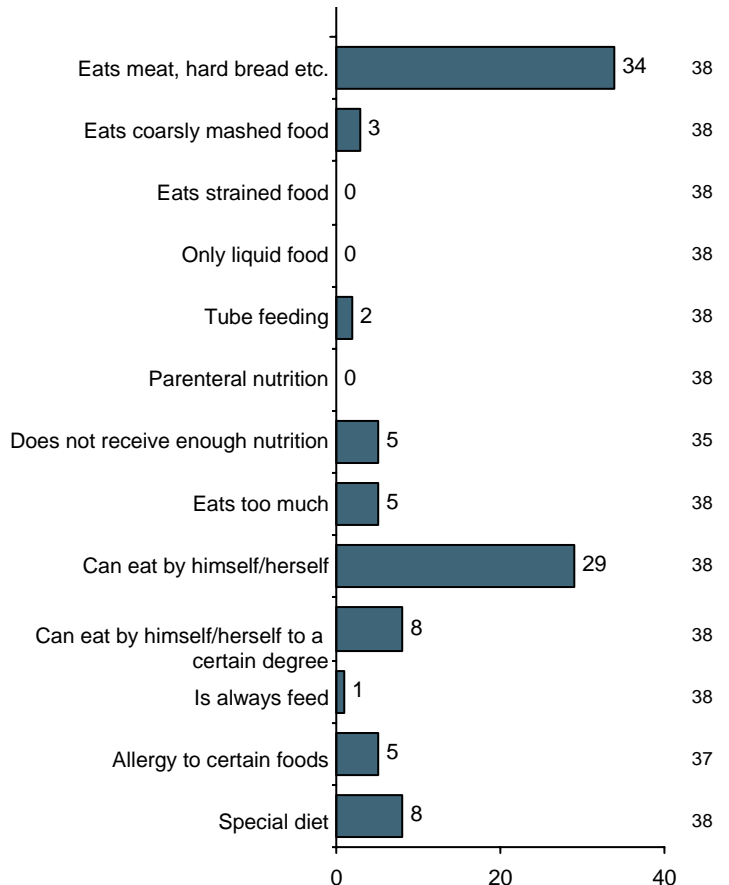
About eating habits



About eating habits - problems



Food habits



About drooling

