



Orofacial function of persons having LCHAD

Report from questionnaires

The survey comprises 6 Questionnaires.

Synonym: Long Chain 3-Hydroxy-Akyl-CoA Dehydrogenase Deficiency

Estimated occurrence: Uncommon diagnosis. Thirteen known cases in Sweden (2004).

Etiology: Autosomal recessive inheritance with a defect in chromosome 2. Deficiency in an enzyme (LCHAD-enzyme) that oxidizes long chain fatty acids.

General symptoms: The disorder often causes hypoglycemia (low blood sugar), lethargy, hypotonia (low muscle tone) and affects the heart and liver. Many are susceptible to infection. Abnormalities in the retina may cause visual impairment and sensitivity to light. Brain damage and development delay may occur as a result of the child becoming seriously ill. Frequent high-carbohydrate meals are necessary and a fat-free diet is recommended. Nearly all those with LCHAD have a gastrosomy.

Orofacial/odontological symptoms: There are no known specific symptoms related to the teeth or bite in LCHAD. Eating difficulties and frequent vomiting are common. Many children who are enterally fed eat only small portions by mouth and lack appetite. When general development is delayed, even speech and oral motor development may be affected.

Orofacial/odontological treatment:

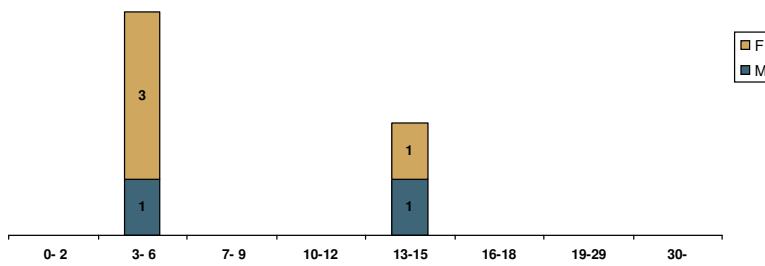
- Children with eating difficulties often need extra dental care, e.g. help with oral hygiene and fluoride treatment.
- Persons with heart defects may require prophylactic antibiotics prior to oral interventions where bleeding is to be expected.
- Eating and swallowing difficulties should be investigated and treated by a specialist team at the hospital (nutrition team or dysphagia team) or multidisciplinary treatment center.
- Orofacial therapy and oral motor skill training and stimulation is recommended for eating difficulties.
- Speech and language difficulties are treated by a speech-language pathologist.

Sources:

The MHC database - The Mun-H-Center database of orofacial manifestations associated with rare disorders.

The Newsletter of the Ågrenska Center.

Age distribution

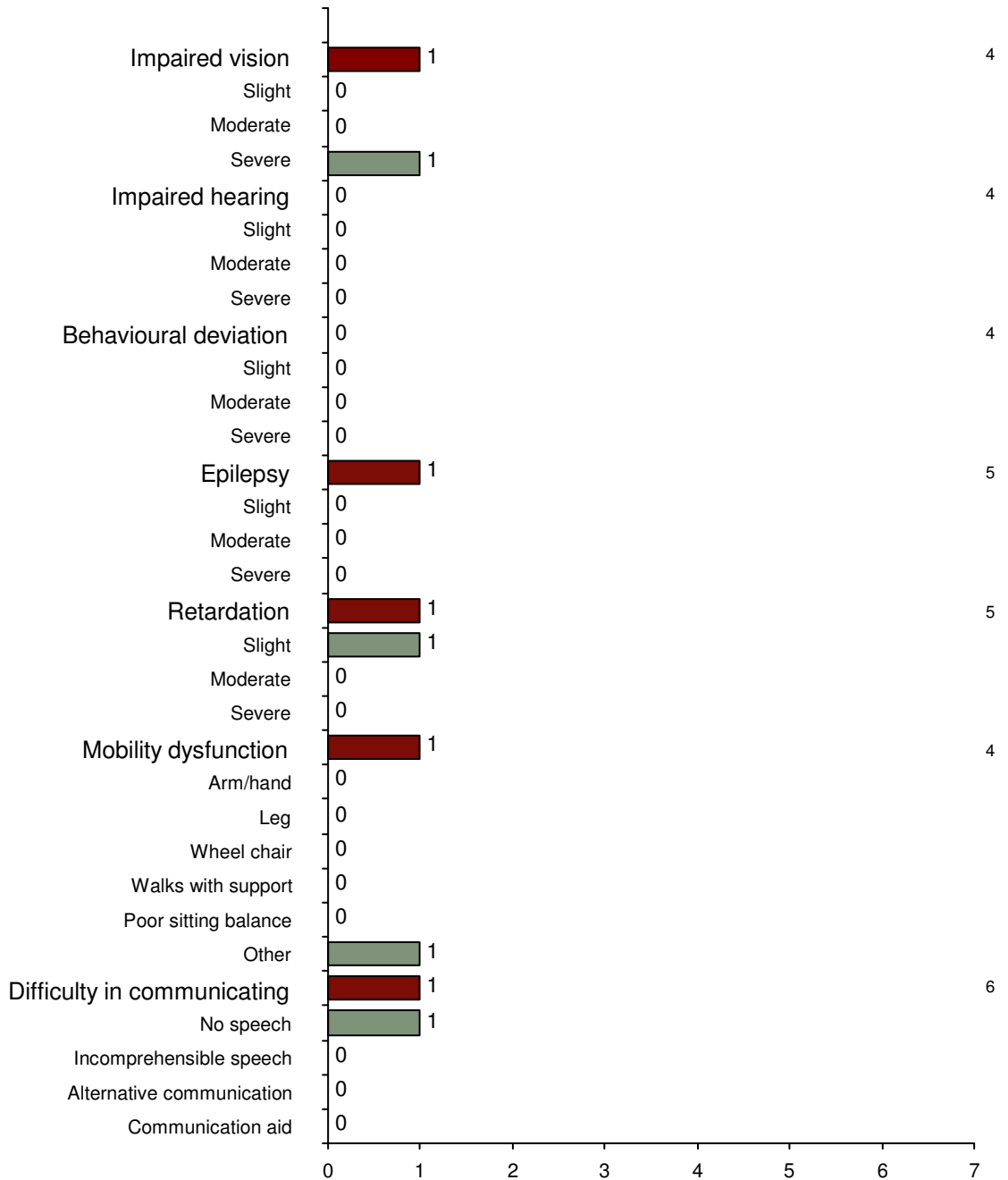


Number: 6

Ages: 3 -- 14 years

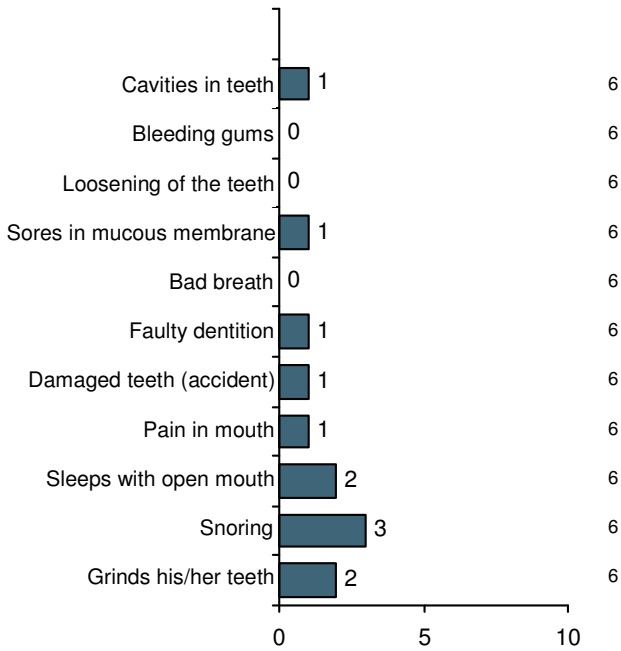
Sex: M (2) + F (4)

General disabilities

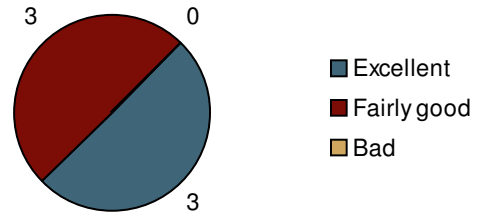


About dental health

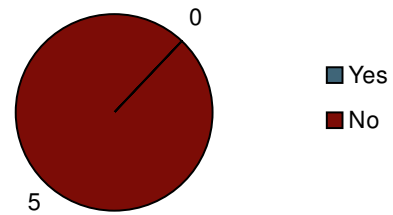
About dental health - problems



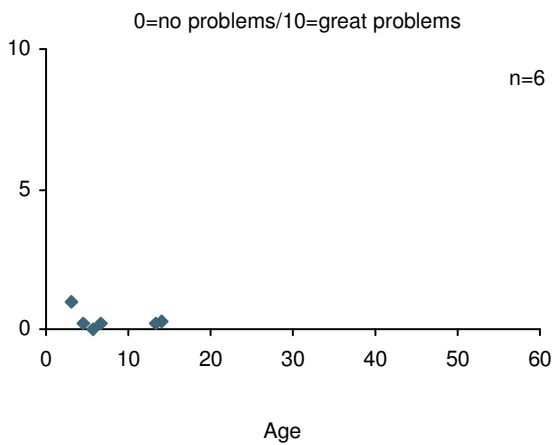
Oral health



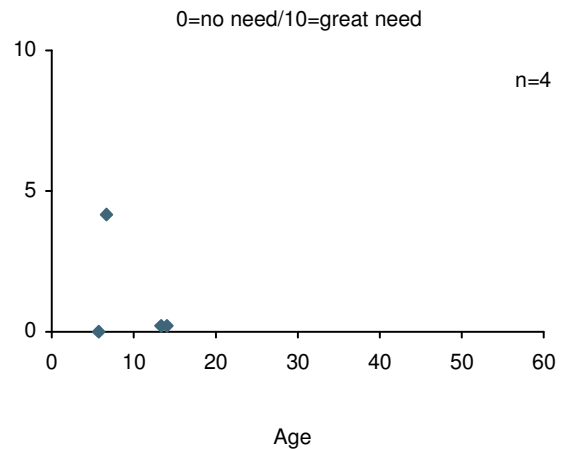
Has NN received orthodontic treatment?



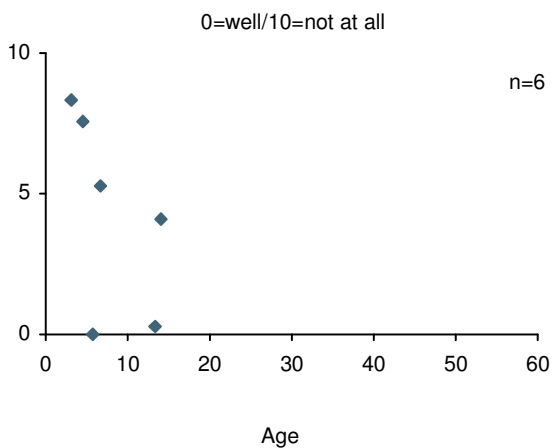
How does NN experience the dental care received?



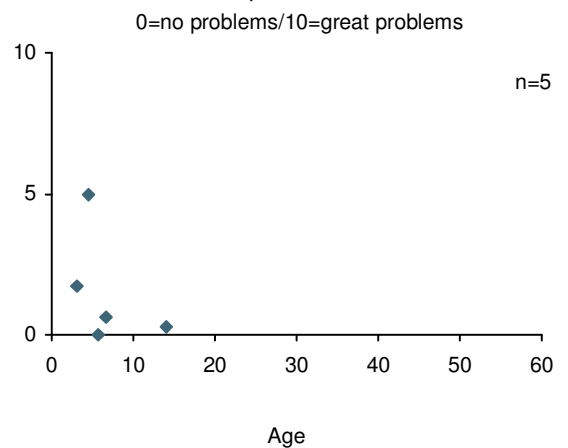
NN is considered to be in need of orthodontic treatment



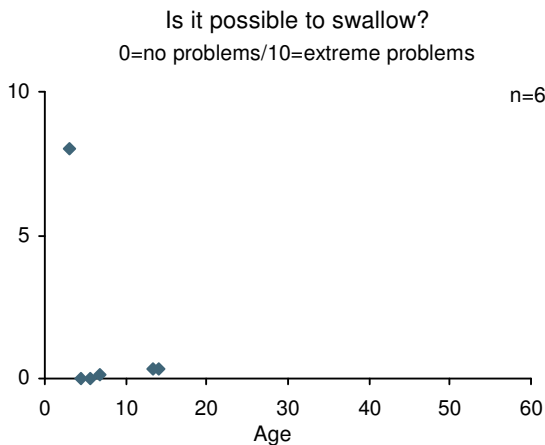
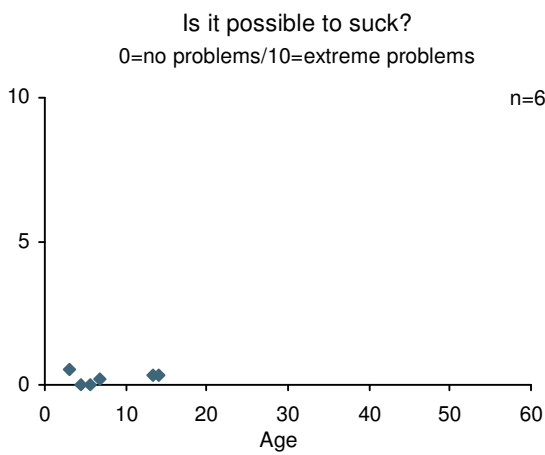
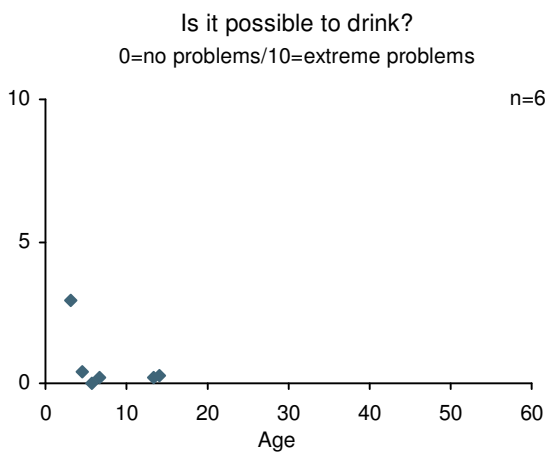
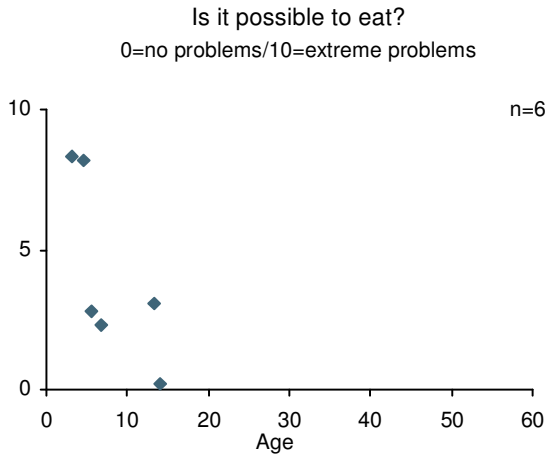
How does NN manage to brush his/her teeth?



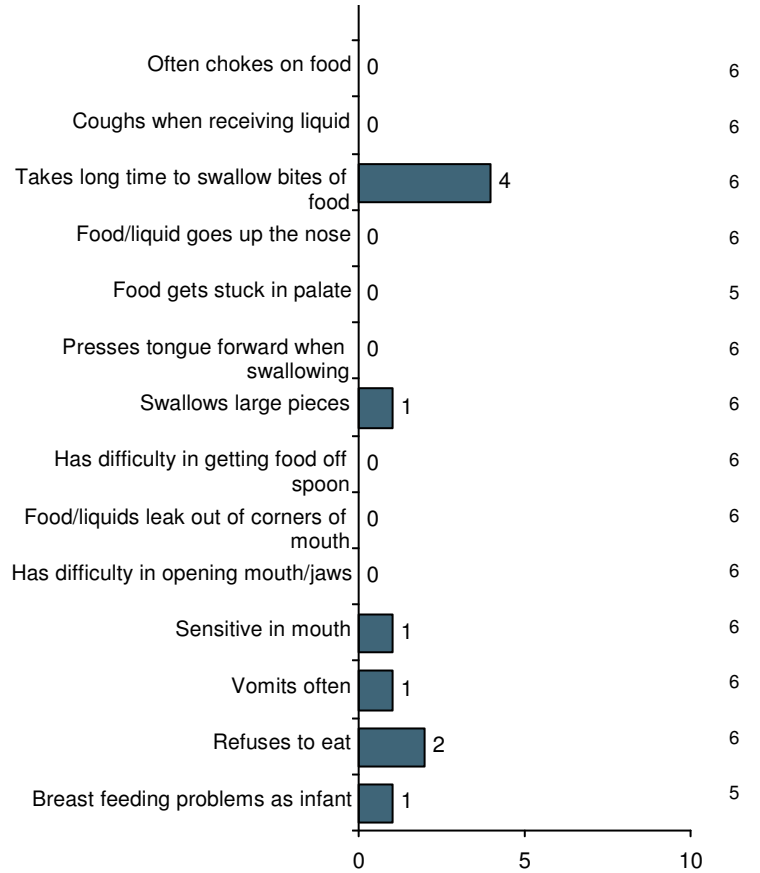
If help is needed with brushing teeth, how does it proceed?



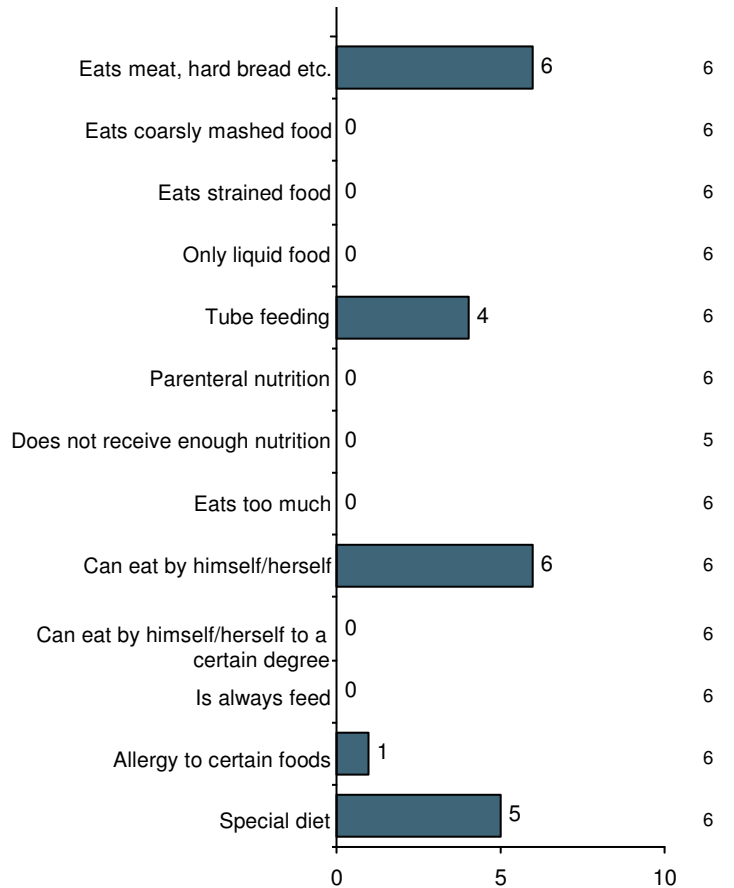
About eating habits



About eating habits - problems

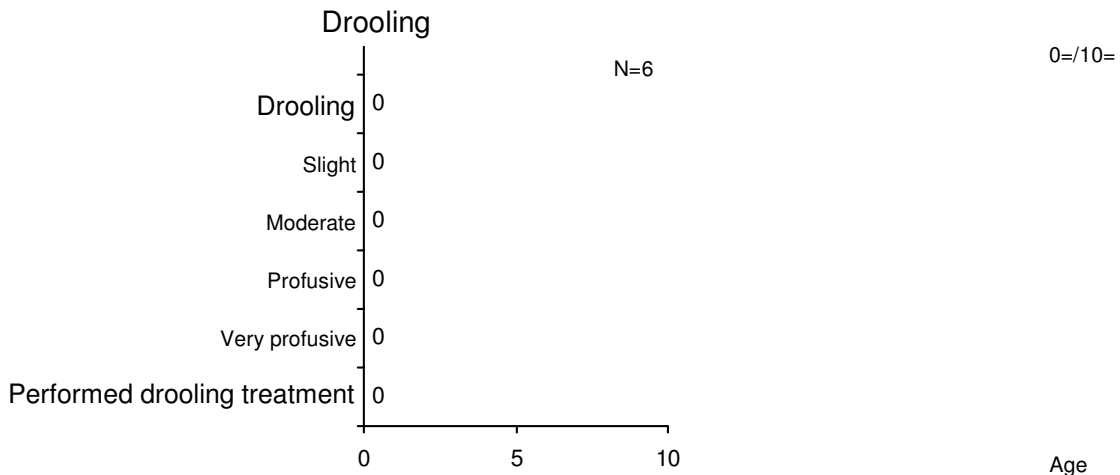


Food habits





About drooling



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Age

Age