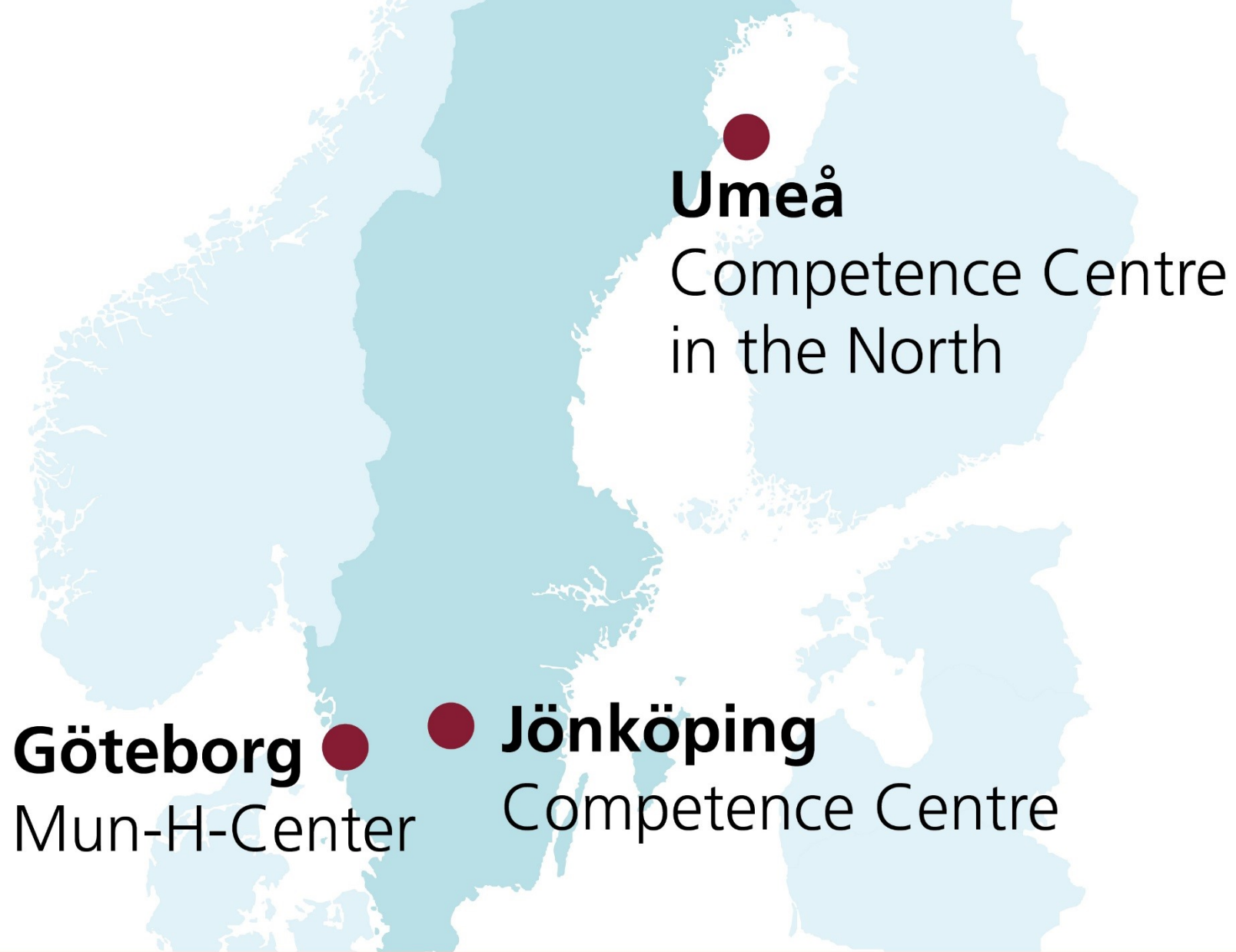


The Nordic Conference 2018
Rett Syndrome in Nordic Light

Rett syndrome and orofacial disturbances

Lotta Sjögren, Speech-Language Pathologist, PhD, Mun-H-Center





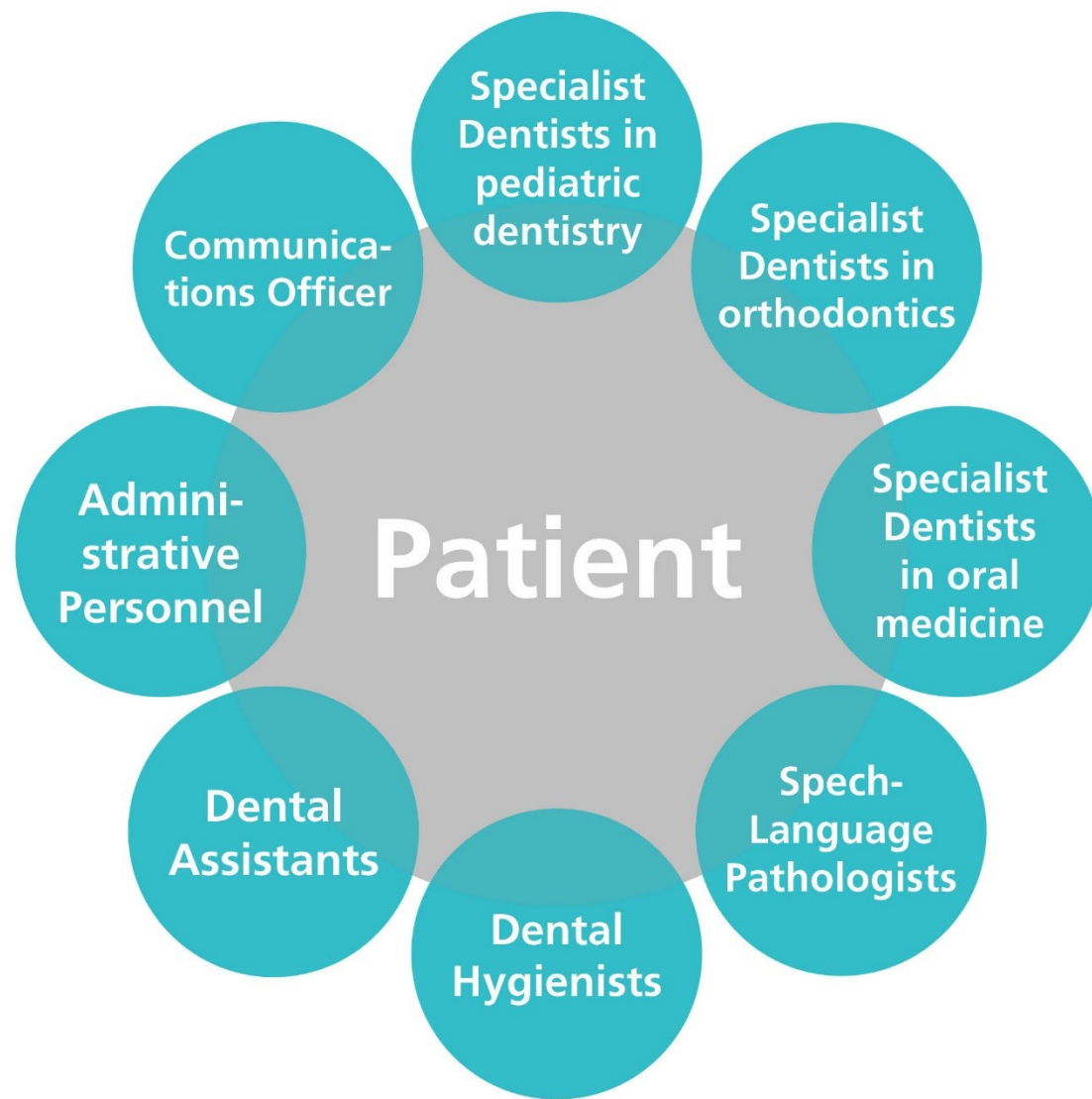
Our task

National

Gather, document and **spread** knowledge about oral health and orofacial funktion in rare medical and dental conditions.
Consultation and **advisory services** about treatment for people with confirmed or suspected rare disease.

Regional

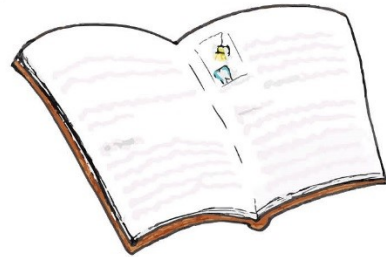
Consultation and even **certain treatment** in specialist dental care and speech-language pathology.



Mun-H-Center



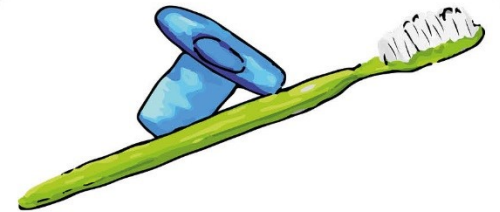
Specialist
dental care



Information
dissemination



Research and
development



Orofacial
assistive devices

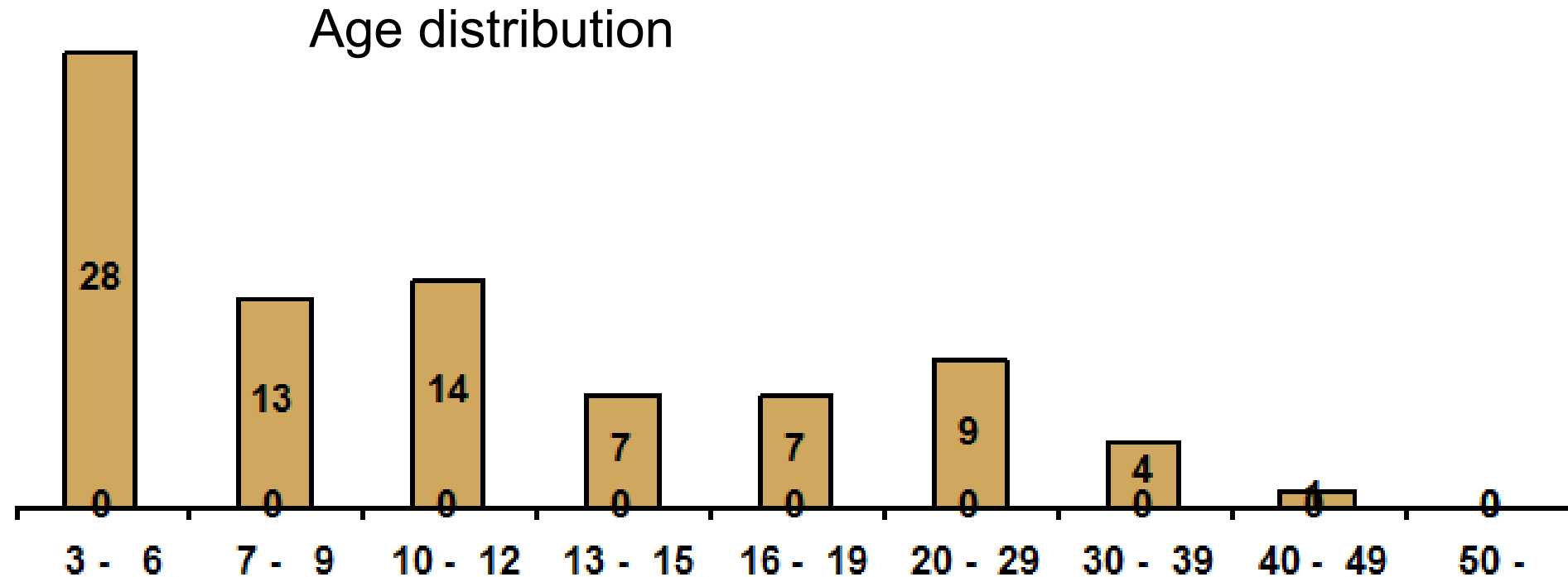
MHC database

A national database for documentation of orofacial manifestations in rare diseases.



Examined patients with Rett syndrome since 1996

N=83, 3-47 years



The MHC Orofacial Observation Chart

Dental examination

- Orofacial morphology, dental occlusion, oral health and oral hygiene

Oromotor examination

- Speech intelligibility, muscle tone and mobility of the facial, tongue and jaw muscles

The image shows a screenshot of a digital form titled "Observationsschema" (Observation Scheme) from the "mun-h" and "FOLKTANDVÅRDEN" (Public Dental Care) systems. The form is designed for recording patient information and clinical observations. It includes fields for patient name, address, date of birth, and personal identification number. There are sections for medical history, including previous diagnoses and other conditions. The form also contains checkboxes for various clinical findings, such as speech disorders (e.g., "Talsvårigheter", "Saknar tal"), facial and jaw issues (e.g., "Kraniofacial missbildning", "Nedsatt stabilitet i nacken"), and oral conditions (e.g., "Intraoralt", "Makroglossi", "Mikroglossi"). A "Noteringar" (Notes) section is provided at the bottom for additional observations. The form is numbered "1" in the bottom right corner.

The MHC Questionnaire

- Questions about medication, general disabilities, oral hygiene, dental care, eating and drinking ability and saliva control
- Answered by caregivers

VERS 2018/01

 **FOLK TAND VÅRDEN** **Frågeformulär**
Frågor om munhälsa och orofacial funktion

Frågorna är ställda till dig som patient, även om någon annan svarar i ditt ställe. Ibland kan de besvaras av föräldrar, vårdpersonal eller någon annan, men alla frågorna handlar om dig. En del frågor kan upplevas som svåra att svara på eller som om de inte alls passar in på dig. Det beror på att frågorna ska täcka in ett brett område och att samma frågor ställs till alla oavsett ålder. Försök att svara på alla frågor.

Dina personuppgifter

Namn: Personnummer:

Adress: Telefon:

Vårdnadshavarens namn (för barn under 18 år):

God mans namn (i förekommande fall):

Datum när frågeformuläret fylls i:

Vem har fyllt i formuläret? Jag själv Vårdnadshavare/
God man Kontaktperson/
vårdpersonal Annan

Samtycke

Jag är medveten om att deltagandet i kartläggningen är fullt frivilligt och att jag när som helst och utan närmare förklaring kan avbryta deltagandet.

Jag godkänner att jag får undersökas av tandläkare och logoped från Mun H Center och att insamlade data får journalföras.

Ort och datum:

Underskrift 1:

Namnförtydligande:

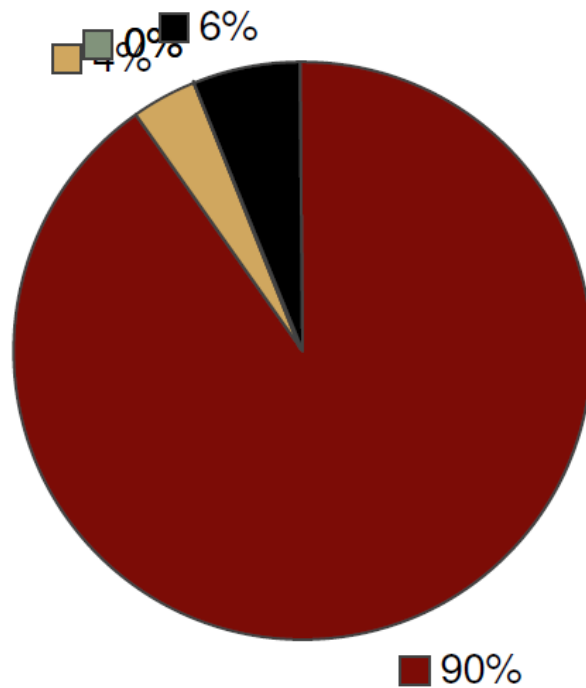
Underskrift 2:







Namnförtydligande:

1

Orofacial dysfunctions - speech

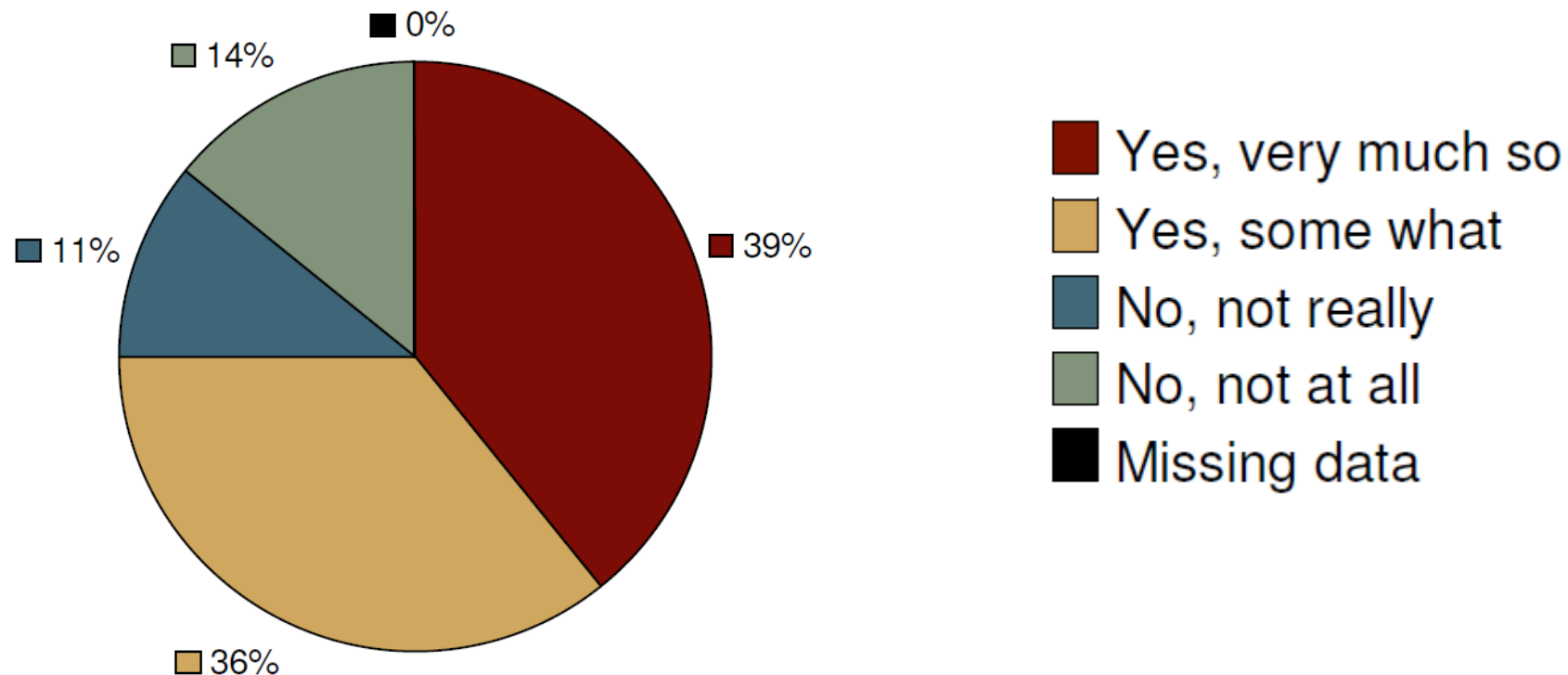
- No speech, 96%
- Very incomprehensible speech, 4%



	Number
 No speech	75
 Very incomprehensible	3
 Incomprehensible speech	0
 Slightly indistinct speech	0
 No problems	0
 Missing data	5
<hr/>	
	Sum: 83

Orofacial dysfunctions – eating and drinking

Do you have any problems with eating?



Orofacial dysfunctions – eating and drinking ability

- Food and drink is leaking out through the mouth, 66%
- Food remains in mouth after meals, 47%
- Takes long time to swallow food, 43%
- Coughing in connection with meals, 27%
- Gastrostomy feeding, 19%
 - 1996 - 2010: 10% (7/69)
 - 2011 - 2018: 65% (11/17)

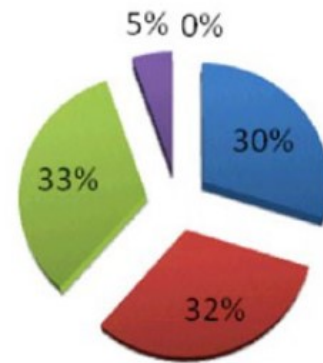
Dysphagia in Rett Syndrome: A Descriptive Study

Annals of Otology, Rhinology & Laryngology
2017, Vol. 126(9) 640–645
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DOI: 10.1177/0003489417723033
journals.sagepub.com/home/aor


Chiara Mezzedimi, MD¹, Walter Livi, MD, PhD¹,
Claudio De Felice, MD², and Serena Cocca, MD¹

- 65 females with Rett syndrome, 2-33 years with reported feeding problems
- Fiberoptic evaluation of swallowing (FEES)

■ NO DYSPHAGIA ■ MILD DYSPHAGIA ■ MODERATE DYSPHAGIA ■ SEVERE DYSPHAGIA



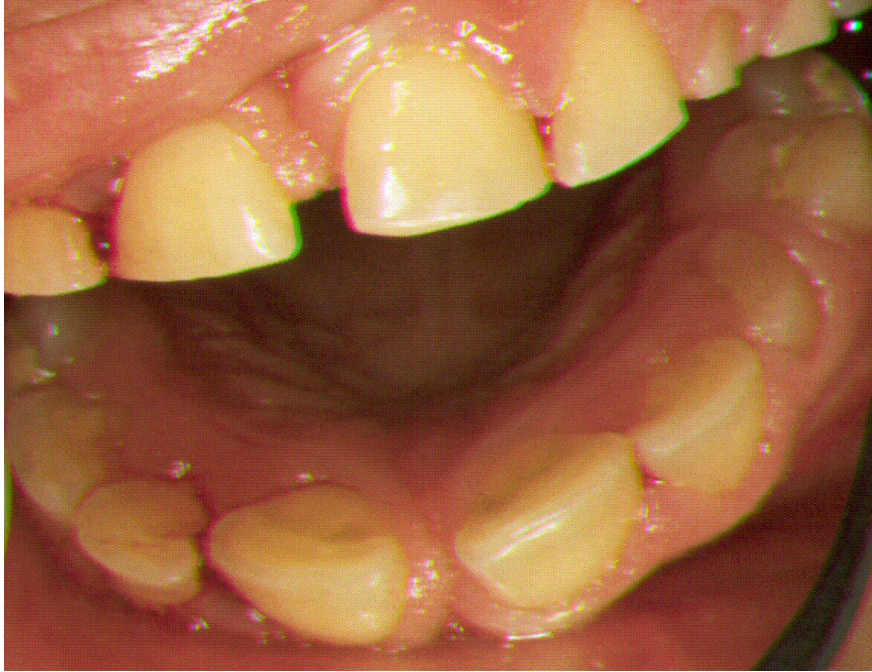
Mezzedini et al, 2017)

Orofacial dysfunctions - saliva control

Saliva leakage/drooling, N=75 (78%)

- Slight drooling, only on the lips, 6
- Moderate drooling, on lip and chin, 32
- Profuse drooling, on clothes, 24
- Very profuse drooling, on hands and objects, 13

Orofacial dysfunctions - tooth grinding

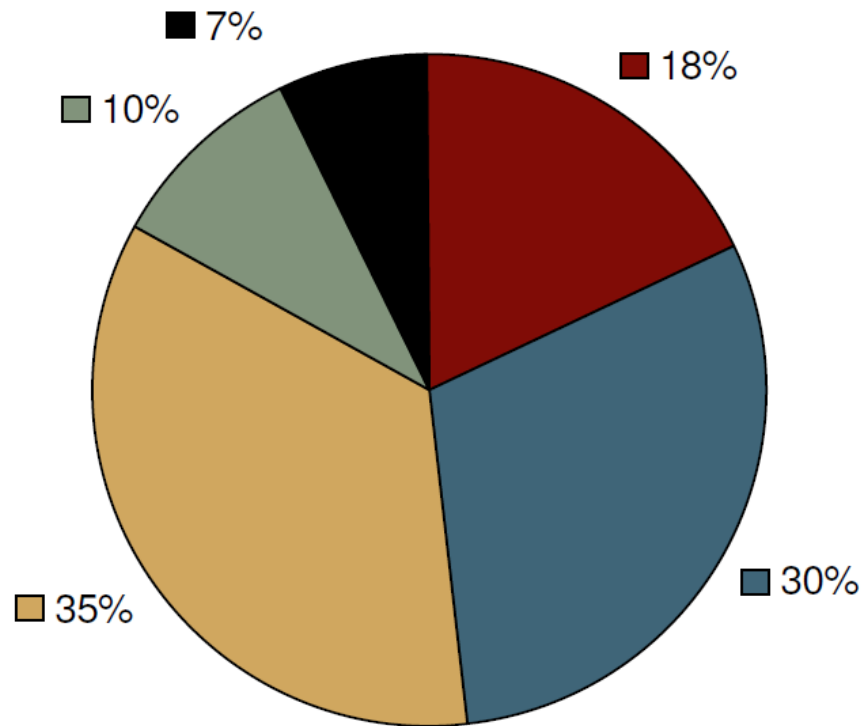


Tooth grinding at night
and/or during the day,
69/85 (81%)

Oral motor function (N=69)

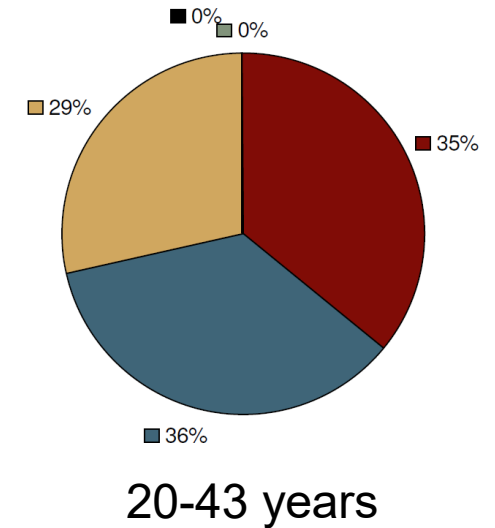
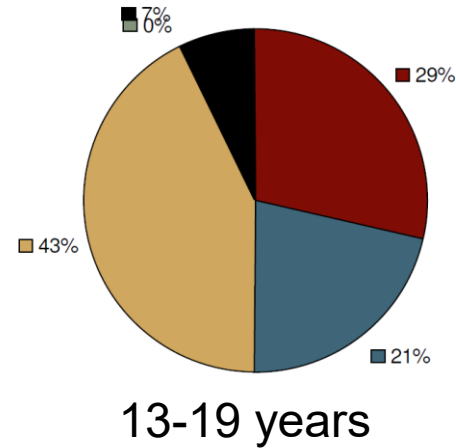
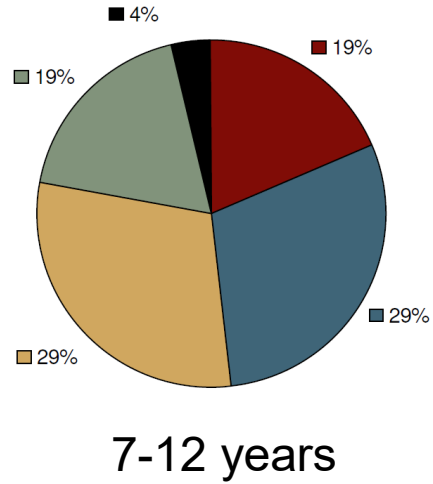
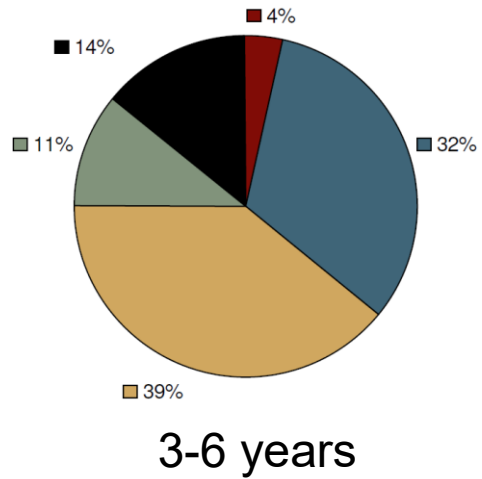
- Impaired tongue motility 81%
 - Delayed oral motor development
 - Oral dyspraxia
 - Involuntary tongue movements
- Open mouth at rest 58%
 - Low muscle tone/muscle weakness
 - Mouth breathing
- Low muscle tone in lips and/or tongue 54%
- Increased muscle tone in masticatory muscles 47%

Dental care – acceptance of dental examination



	Number
None	15
Negative	25
Reluctant	29
Positive	8
Missing data	6
Sum: 83	

Dental care – acceptance of dental examination



Oral health - caries free (N=64)

- 3-6 years 100% (20/20)
- 7-12 years 95% (20/21)
- 13-19 years 82% (9/11)
- > 19 years 67% (8/12)

Missing data: N=19

Occlusion (7-43 years)



Neutral bite

51%



Overjet

42%



Underbite

7%



Deep bite

15%



Open bite

23%

Over crowded teeth

31%

Orofacial/odontological symptoms in summery

- Severe oral motor impairment resulting in eating problems, saliva leakage and difficulties with oral hygiene.
- Most girls with Rett syndrome do not learn to speak.
- The majority have severe tooth grinding, resulting in tooth wear, and many also have involuntary tongue movements.
- Overbite and an open bite in the area of the front teeth are more common than in the general population.

Orofacial/odontological treatment

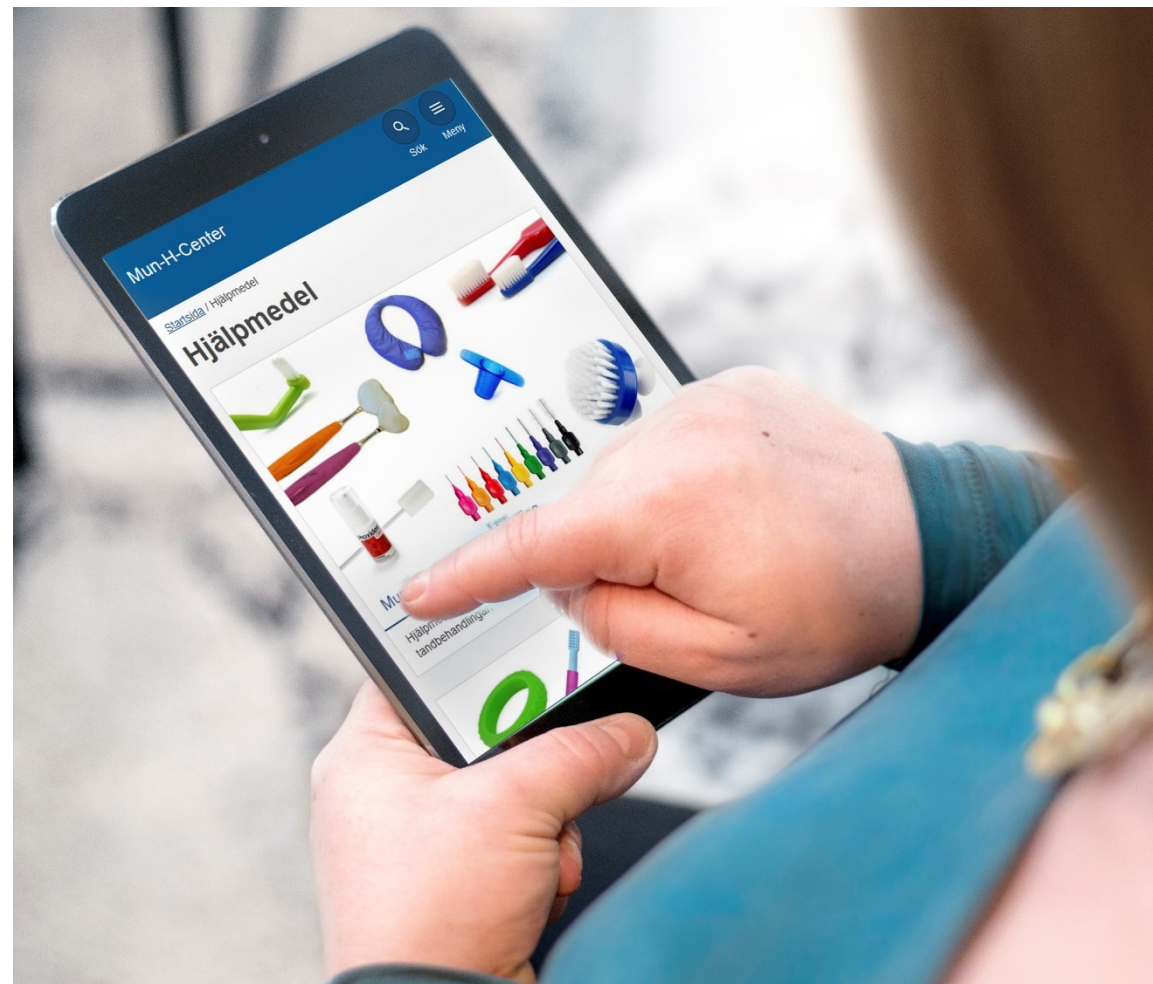
- Early contact with dental services for intensified prophylactic care and oral hygiene information is essential.
- Tooth grinding should be followed up, and be managed with a splint when necessary.
- Regular check-ups of dental and jaw development. Orthodontist should be consulted when needed.
- Feeding and swallowing difficulties are investigated and treated by a specialist team at the hospital or multidisciplinary treatment center.
- Communication skills training is frequently essential.
- Oral motor training and stimulation may be relevant.



Orofacial/odontological treatment

Early contact with dental services for intensified prophylactic care and oral hygiene information is essential.

- Dental care of excellence
- Knowledge about the diagnosis
- Cooperation with the medical team
- Aids and appliances for oral care



Aids and appliances for oral and dental care



Oral bite support



Double-action toothbrush

Aids and appliances for oral and dental care



TUMLE dental cushions

The management of tooth grinding

Tooth grinding should be followed up, and be managed with a splint when necessary. A relaxing facial massage is often appreciated.



Orthodontic follow-up and treatment

Regular check-ups of dental and jaw development.
Orthodontist should be consulted when needed.



- 16 years old
- Overjet
- Crowded teeth

Orthodontic treatment



Conscious
sedation

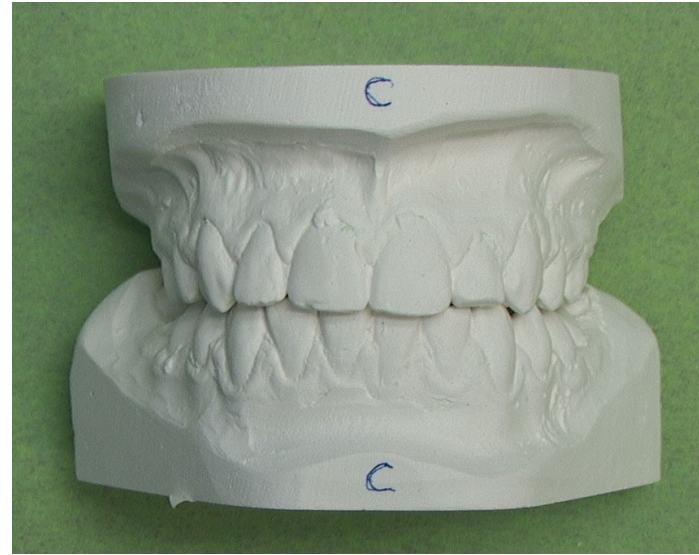


Extraction of
two teeth



Fixed appliance:
first in the upper jaw,
followed by appliance in
the lower jaw

Long-term retention to maintain the stability of the corrections obtained during treatment



She no longer bites her lips when having epileptic seizures

The management of dysphagia

Feeding and swallowing difficulties are investigated and treated by a **specialist team** at the hospital or multidisciplinary treatment center.

When there is an increased risk for:

- aspiration of food and liquids to the airways
- choking
- malnutrition

What can be done to facilitate eating and drinking?

- Feeding technique and positioning
- Food texture
- Feeding equipment
- Oral sensory-motor stimulation

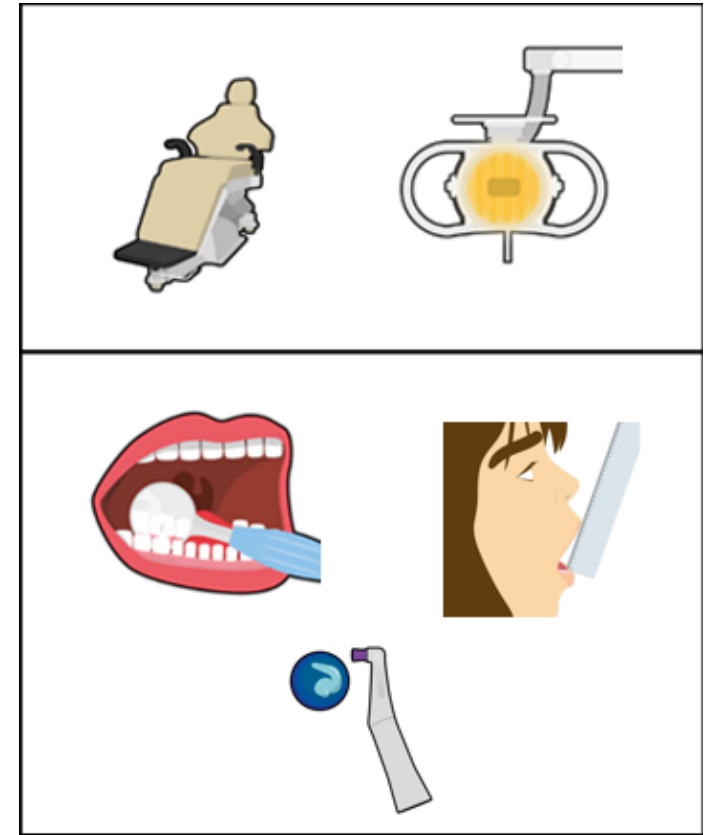
Feeding equipment to facilitate eating and drinking



Communication skills training

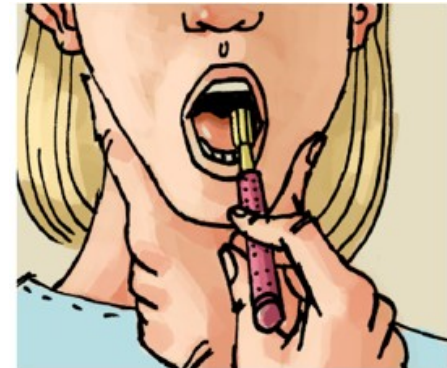
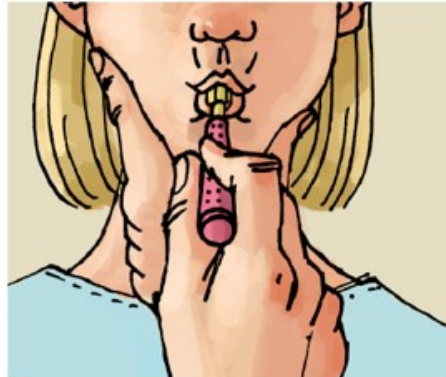


Image support in dentistry



www.bildstod.se

Oral motor training and stimulation



Chewing exercises



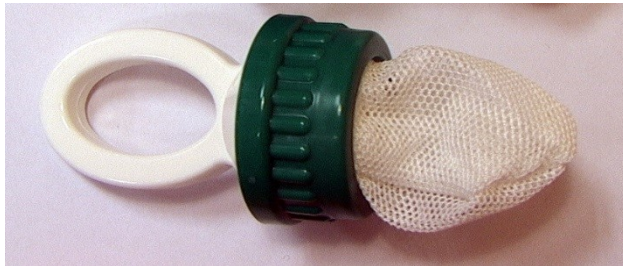
Chewy tube



ARK's Brick Bracelet
Textured Chew / Fidget



Wave bangles



Safe feeder,

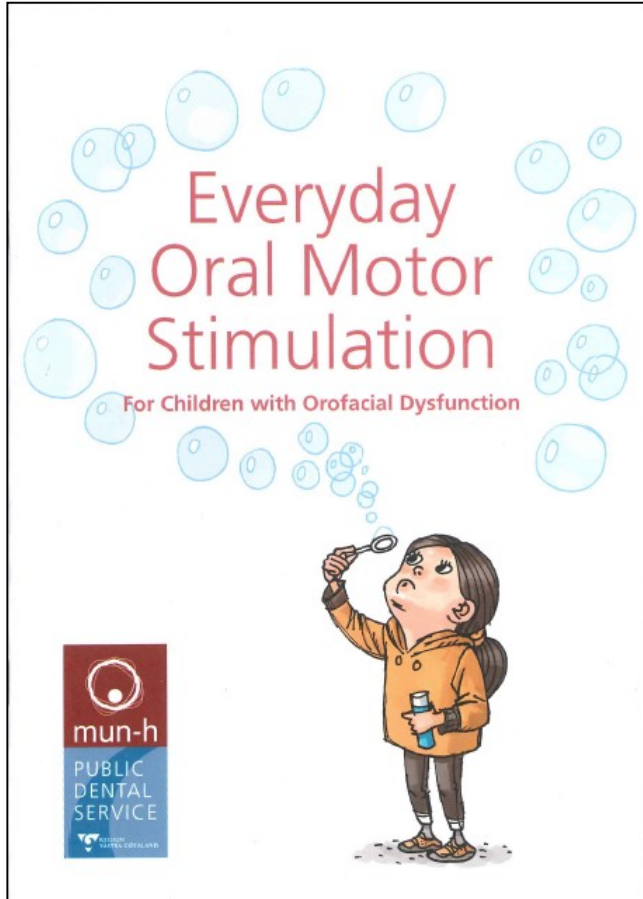


buDDy tube



Chews

Everyday Oral Motor Stimulation



Support for oral care in the MHC-app

Rare diseases



Advices about oral care for children



Information about familiarization to dental care

