Dental and Swallowing Issues

Elizabeth Vroom, DDS
Lack of dystrophin

Muscle breakdown
Fibrosis
Infiltration Fatty Tissue

Leading to loss of function and deformities

Orofacial changes in DMD described since 1990’s by orthodontists but not picked up by the NMD community
Lack of dystrophin causes

Progressive weakness of masticatory and perioral muscle groups

Tongue enlarged and reduced motility (progressive)

Progressive oropharyngeal and hypolaryngeal weakness
Which causes

- Changes in dental arches
- Changes in occlusion
- Reduced contacts between the upper and lower (pre) molars
- Changes in bite force
- Difficulties oral cleaning
- Difficulties in speech
- Swallowing problems
- Skeletal changes
Reduced bite force and reduced contacts causes:

- Mastication difficulties including less fragmentation of food.
- Poor fragmentation of food in combination with weak pharyngeal clearance may increase pharyngeal post swallow residue.
- Which may cause in advanced stages swallowing difficulties like choking and the feeling of sticking food in the throat.
skeleton

dentition

function
Form follows function

- Thumb sucking
- Mouth breathing
- Swallowing
No Duchenne  Duchenne
L. Eckhardt and W. Harzer

facial structure and functional findings in patients with Duchenne

Study of dental and cranial development of 15 dmd patients compared with controls

American J Orthod Dentofacial Orthop 1996
(Aug;11092);185-90
Mean of sample

Progressive muscular dystrophy

Fig. 9. Superposition of cephalograms from 10-year-old patient with PMD and same age controls, along SN line at sella point.

M - masticatory muscles - weak
O - orbicularis oris - normal
T - tongue - weak and voluminous

⇒ change direction of morphology
Stavros Kiliaridis and Christos Katsaros

The effects of myotonic dystrophy and Duchenne muscular dystrophy on the orofacial muscles and dentofacial morphology

Department of Orthodontics, Faculty of Odontology, Goteborg University, GoÈteborg, Sweden and Department of Orthodontics, University of Saarland, Homburg/Saar, Germany

Acta Odontol Scand 1998
Orofacial changes

- transversal over-development of the dental arches
- sagittal shortening of the dental arches
- sagittal under-development of the cranial, maxillary and mandibular base

- reduction of overbite and overjet
- retrusion of incisors
- concave profile
- increase in bizygomatic width
Transversal expansion of skull and dental arches caused by

- Decreased activity of masticatory muscles starting 2 yrs earlier than the perioral muscles

- enlarged hypotonic tongue
Temporalis Muscle Hypertrophy and Reduced Skull Eccentricity in Duchenne Muscular Dystrophy

C. S. M. Straathof, MD, N. Doorenweerd, MSc, B. H. A. Wokke, MD, E. M. Dumas, PhD, J. C. van den Bergen, MD, M. A. van Buchem, MD, PhD, J. G. M. Hendriksen, PhD, J. J. G. M. Verschuuren, MD, PhD, and H. E. Kan, PhD

Journal of Child Neurology 29(10)
Transverse sections taken parallel to the anterior and posterior commissure (AC/PC) of a healthy 10-year-old boy (left) and a 10-year-old boy with Duchenne muscular dystrophy. The boy with Duchenne muscular dystrophy shows a rounder head and skull shape and bilateral temporal muscle hypertrophy (indicated by arrows).
Oral muscles are progressively affected in Duchenne muscular dystrophy: implications for dysphagia treatment.

van den Engel-Hoek L1, Erasmus CE, Hendriks JC, Geurts AC, Klein WM, Pillen S, Sie LT, de Swart BJ, de Groot IJ.
Tongue

- Thickness tongue increases with age/stage
- Tongue motility decreases
- Tongue pressure decreases
- Lies on the occlusal surfaces of the mandibular teeth as the jaws are relaxed
- Protrudes over the anterior teeth
3 superior longitudinal tongue muscle, 4 transverse tongue muscle.

**e** Ultrasound image from a 12-year-old boy with DMD (ENAS), transverse tongue muscle,

**f** Ultrasound image from an 18-year-old boy with DMD (LNAS), superior longitudinal tongue muscle, transverse tongue muscle. Note that the images of the 12-year-old boy are almost normal, except for the geniohyoid muscle.
Enlarged and weaker tongue causes

- Dental deformities (causing problems chewing)
- Problems to move the food through the mouth
- Problems with cleaning
- Problems with speech
- Problems with appearance
1 digastric muscles,
2 geniohyoid muscles.

B 12-year-old boy with DMD ENAS
C Ultrasound image from an 18-year-old boy with DMD LNAS
Dystrophic changes in masticatory muscles related chewing problems and malocclusions in Duchenne muscular dystrophy

Radboud University Medical Center, Nijmegen, The Netherlands

Neuromuscular Disorders 2016
Oral muscles are progressively affected in Duchenne muscular dystrophy: implications for dysphagia treatment

Lenie van den Engel-Hoek • Corrie E. Erasmus • Jan C. M. Hendriks • Alexander C. H. Geurts • Willemijn M. Klein • Sigrid Pillen • Lilian T. Sie • Bert J. M. de Swart • Imelda J. M. de Groot

• Oral muscles related to swallowing were progressively affected, starting in the AS with the geniohyoid muscle.

• Tongue (pseudo) hypertrophy was found in 70% of patients in the ENAS and LNAS.

• Oral phase problems and post-swallow residue were observed, mostly in the LNAS with solid food.

• sEMG and tongue pressure data of swallowing solid food revealed the lowest sEMG amplitude, the longest duration and lowest tongue pressure in the LNAS.
What you are looking at:

• 3 consistencies are swallowed. Starting with 5 ml thin liquid (with contrast).
• Thin liquid is swallowed and transported to the esophagus.
• After swallowing some residue is present in the valleculae and above the esophageal sphincter, due to weakness.
• A second bolus (pudding) is swallowed (time 0.09) and after swallow more pharyngeal residue is present, because pudding asks for more strengths. Several swallows are needed to clear the pharynx.
• At time 0.18 pureed potato is given and the first swallow can be observed.
• After 3 swallows there is still a lot of residue. Often this has to been cleaned with water.
• This pharyngeal residue can be a risk for aspiration (= material entering the airway)
Recommendations

• In case of swallowing problems in DMD, based on the disturbed mechanisms of swallowing, it is suggested to
  • (1) adjust meals in terms of less solid food,
  • (2) drink water after meals to clear the oropharyngeal area.
Reduced mandibular range of motion in Duchenne Muscular Dystrophy: predictive factors.

van Bruggen HW, Van Den Engel-Hoek L, Steenks MH, Bronkhorst EM, Creugers NH, de Groot IJ, Kalaykova SI.

• Mandibular movements in DMD are significantly reduced and become more hampered with loss of motor function, including the sitting position, arm function, and neck and head control.

• We suggest that measurement of the aMMO* becomes a part of routine care of patients with DMD.

* active maximum mouth opening
Fighting Against Disuse of the Masticatory System in Duchenne Muscular Dystrophy: A Pilot Study Using Chewing Gum

H. Willemijn van Bruggen, DDS, MSc1,2, Lenie van den Engel-Hoek, PhD3, Michel H. Steenks, DDS, PhD2, Andries van der Bilt, PhD2, Ewald M. Bronkhorst, PhD4, Nico H. J. Creugers, DDS, PhD1, Imelda J. M. de Groot, MD, PhD3, and Stanimira I. Kalaykova, DDS, PhD1

Mastication training in Duchenne muscular dystrophy using chewing gum for 4 weeks.

Mastication training by using sugar-free chewing gum in Duchenne muscular dystrophy patients improved their masticatory performance.

Since bite force did not improve, the working mechanism of the improvement in chewing may relate to changes of the neuromuscular function and coordination, resulting in improvement of skills in performing mastication.
Dental health

- Dental hygiene is poor in DMD
- More caries
- High DMF score (Decayed, Missing, Filled)
- Gingival inflammation
- Heavy plaque accumulation and calculus deposits (especially around the lower anterior teeth)
- Mouth breathing
- Very often unhealthy periodontium.
- Delayed eruption of permanent dentition

* Prof. Maria Mielnik-Baszczak, PhD, DDS;* Borowska Malgorzata, PhD, DDS
For children with DMD, dental and prophylactic treatment should begin early.

The pediatrician who diagnoses a child with DMD should be obliged to inform the parents about the necessity of regular dental check-ups.

Proper diet is important.

Oral hygiene (from 3 years of age oral hygiene agents should include fluoride)

Fluoride prophylaxis

Pit and fissure sealants and fluoride

Regular dental check-ups

Avoid treatment under general anesthesia

Prof. Maria Mielnik-Baszczak, PhD, DDS;* Borowska Malgorzata, PhD, DDS
Dental healthcare
(older boys)

• Dental hygiene more difficult when handfunction decreases and other people (often not trained) have to take over
• Large tongue
• Less ‘natural cleaning’ bij tongue and checks
• Think twice before removing wisdom teeth (every patient needs a personal benefit risk)
• Risk of aspiration
Recommendations

• Rinse after every meal
• Avoid mouth breathing and open mouth (tongue ‘in’)
• Don’t start orthodontic treatments without a thorough understanding of all aspects caused by the disease
• Consider chewing gum
Recommendations

In case of swallowing problems in DMD, based on the disturbed mechanisms of swallowing, it is suggested to:

(1) adjust meals in terms of less solid food
(2) drink water after meals to clear the oropharyngeal area.
Thanks

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