Maintaining oral function in boys with Duchenne muscular dystrophy.

Stefan Axelsson, PhD, Specialist in Orthodontics, and Vesna Stoilkovic, Speech Language Therapist,
TAKO-centre, National Resource Centre for Oral Health in Rare Diagnoses, Lovisenberg Diaconal Hospital, Oslo, Norway

Background

Duchenne muscular dystrophy (DMD) is a serious X-linked neuromuscular disorder diagnosed in early childhood, affecting approximately 1 in every 3.500 live male births.

DMD results in progressive loss of muscle strength and is caused by a mutation in the gene that encodes for dystrophin. Because dystrophin is absent, the muscle cells are easily damaged. The progressive muscle weakness leads to serious medical problems, particularly affecting the heart and lungs.

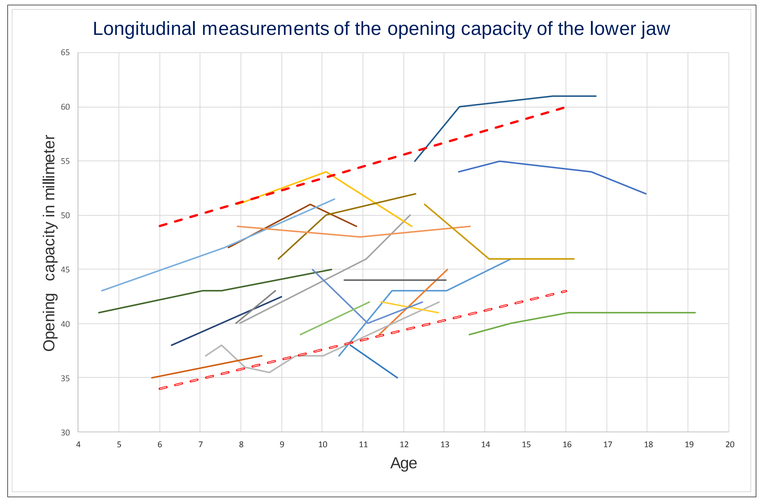
The muscular weakness may lead to contractures in the temporomandibular joints and limit the jaw opening capacity. The muscle weakness may also cause difficulties in chewing and swallowing.

Figure 1

Measuring the maximal mouth opening capacity with a metallic caliper between the incisal edges of upper and lower central incisors, and add the measurement of the vertical overbite to get the correct measurement.

Figure 2

Reduced maximal mouth opening capacity of the lower jaw (22 mm).



Methods

For several years the TAKO Centre has focused on maintaining oral function and jaw movement in boys with DMD through a systematic training program.

The boys and their parents/carers have been shown and thought simple training exercises aiming at avoiding contractures of the temporomandibular joint and increase strength and endurance of the masticatory muscles.

In addition, the boys have been advised to eat food items that give them chewing challenges, such as carrots cut in strips. Sugarfree chewing gum and chewing aids, such as the ChewyTube, have also been used in the program.

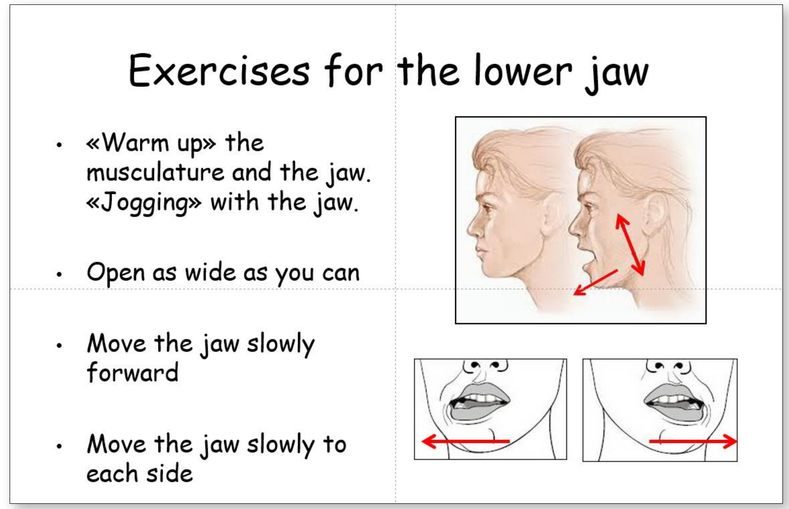
References

- van Bruggen et al. Reduced mandibular range of motion in Duchenne Muscular Dystrophy: predictive factors. J Oral Rehab. 2015;42(6):430-8.
- Ueki et al. Bite Force and Maxillofacial Morphology in Patients With Duchenne-Type Muscular Dystrophy. J Oral Maxillofac Surg. 2007;65:34-9.
- Scandinavian State Of The Art (SOTA) Document on Duchenne Muscular Dystrophy. Revised 2015

Results

As a result of systematic and daily training, the opening capacity of the lower jaw (mandible) has been maintained or even increased in many of the boys, as shown in the graph. The bold red dotted lines in the graph represents the variation of maximum opening capacity of the lower jaw in the general population.

Since the program started it has only been used it in children and adolescents, but it is believed that the program also will be useful in adults in a slightly altered and adjusted form.



Suggestions

We suggest that the systematic joint contracture prophylaxis for the temporomandibular joints and training exercises for the masticatory muscles should be included in a general physiotherapy program for boys with DMD from a young age and throughout adolescence and adulthood.