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Predictive factors for masticatory performance in Duchenne muscular dystrophy

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Abstract

Patients with Duchenne muscular dystrophy (DMD) report masticatory and swallowing problems. Such problems may cause complications such as choking, and feeling of food sticking in the throat. We investigated whether masticatory performance in DMD is objectively impaired, and explored predictive factors for compromised mastication.

Twenty-three patients and 23 controls filled out two questionnaires about mandibular function, and underwent a clinical examination of the masticatory system and measurements of anterior bite force and masticatory performance. In the patients, moreover, quantitative ultrasound of the tongue and motor function measurement was performed. The patients were categorized into ambulatory stage (early or late), early non-ambulatory stage, or late non-ambulatory stage.

Masticatory performance, anterior bite force and occlusal contacts were all reduced in the patient group compared to the controls (all $p < 0.001$). Mastication abnormalities were present early in the disease process prior to a reduction of motor function measurement. The early non-ambulatory and late non-ambulatory stage groups showed less masticatory performance compared to the ambulatory stage group ($p < 0.028$ and $p < 0.010$, respectively). Multiple linear regression analysis revealed that stage of the disease was the strongest independent risk factor for the masticatory performance ($R^2 = 0.52$).

Anterior bite force, occlusal contacts and masticatory performance in DMD are severely reduced.

Keywords: Duchenne muscular dystrophy; Masticatory system; Masticatory performance; Bite force

1. Introduction

The most prevalent type of muscular dystrophy in children is Duchenne muscular dystrophy (DMD), which is characterized by a progressive loss of muscle cells and replacement by fat and connective tissue [1]. DMD is an X-linked recessive disorder or is a consequence of de novo gene mutation. It is caused by a defective gene for dystrophin, an essential component of the dystrophin–glycoprotein complex (DGC) maintaining the membrane integrity of muscle fibers [2]. DMD has an incidence of about 1:3500 live male births [3]. The onset of DMD symptoms occurs at around 2–6 years of age. Untreated, muscle strength deteriorates, and boys require the use of a wheelchair before their teens [4].

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Medical care for patients with DMD, which encompasses medical, surgical, and rehabilitation approaches, has improved the quality and duration of life. As a consequence of the increased duration of life, the non-fatal medical complications of the disease, among which mastication and swallowing difficulties, are now requiring more attention [5].

The subjective experience of mastication and feeding problems in DMD has previously been documented using questionnaires [6,7], yet not objectively using masticatory performance tests. Mastication problems may induce post swallow residue, which has been suggested to possibly contribute to a greater risk of food aspiration [8,9]. Video fluoroscopic swallow studies showed an increased pharyngeal post swallow residue of solid food in advanced stages of DMD [10].

In general, the main factors influencing masticatory performance are the number of occlusal contacts and bite force [11]. Furthermore tongue function is of importance because of its role in food-bolus mixing and positioning [12]. Since masticatory performance of DMD patients has so far not been assessed objectively, it is yet unclear which factors influence the masticatory performance in these patients.

The aim of this study is to compare the masticatory performance of patients with DMD and an age matched healthy control group, and to assess predictive factors for the masticatory performance.

2. Material and methods

The study was approved by the Committee on Research Involving Human Subjects of Arnhem and Nijmegen, the Netherlands (registration number 2009/331).

2.1. Participants

A case-control study design was used. Twenty-four patients with DMD from 6 to 38 years, and 24 age-matched male healthy controls were recruited between May 2010 and February 2012. Age limits were set above 5 years to ensure that the participants were capable of undergoing all physical examinations and of filling out questionnaires, whenever needed with the help of their caretakers.

The patients with DMD originate from the cohort of the swallowing study of van den Engel-Hoek et al., 2013 [11]. They were recruited by announcements of patient organizations. These patients had a genetically confirmed diagnosis of DMD, or a diagnosis of DMD as defined by the onset of symptoms and disease course of DMD. DMD patients who were entirely dependent on tube feeding were excluded.

The control group was recruited at a primary and a secondary school located in the western part of the Netherlands, and consisted of children and parents. Controls above 18 years were as well recruited at the College of Dental Sciences, Nijmegen, The Netherlands. Exclusion criteria for healthy controls were: a history of neuromuscular disease, temporomandibular disorder, orthodontic treatment and the morphologic dental malocclusions cross bite or tendency to cross bite.

Eligible participants and their parents were sent information letters with an invitation to participate in the study. After receiving informed consent from the adults or parents, and – if the subjects were older than 12 years – also from the participants, the participants underwent all examinations in the hospital during one day.

2.2. Protocol

The participants in both the patient and the control group, completed structured questionnaires, and underwent a clinical examination of the masticatory system, anterior maximum voluntary bite force (MVBF) and food mixing ability measurements. Additionally, the patients underwent quantitative muscle ultrasound imaging (QMUS) of the tongue [13], and an assessment of their general physical abilities.

2.2.1. Questionnaires

The patient group and the control group completed two questionnaires: Screen [14,15] and the Mandibular Functional Impairment Questionnaire (MFIQ) [16].

Screen contains questions related to: (1) quantitative and qualitative aspects of pain in the head, neck and shoulders such as the pain location(s) (which the patient could mark on a drawing of the head, neck and shoulders), the pain intensity as reported on a Visual Analogue Scale (VAS), and factors influencing pain; (2) symptoms of temporomandibular dysfunction such as joint noises, limited mouth opening and mastication difficulties; (3) oral habits, such as tooth-grinding and clenching; and (4) general health factors(e.g. medication, family history).

MFIQ was designed to measure impairments of the function of the masticatory system. MFIQ consists of 17 questions, which can be subdivided into two dimensions – functional capacity and feeding –, and is rated on a 5-point Likert scale ranging from '0' (no difficulty) to '4' (very difficult or impossible without help). The ratings are added to give a sum score (S, range 0–68). A higher score indicates more perceived mandibular function impairments and a MFIQ score of '0' indicates no impairment in mandibular functioning. The raw score of the MFIQ-scale (C, range 0–1) is obtained by dividing the sum of the items by four times the number of the items ($C = S/68$). The raw score can be converted to a qualitative level of impairment: no/low (I), moderate (II) and high (III). Previous studies have considered the MFIQ to be a reliable and valid tool [17].

2.2.2. Clinical examination of the masticatory system

All participants in this study were clinically examined following a validated procedure as described by