Duchenne muscular dystrophy — a dental healthcare program

Prof. Maria Mielenk-Błaszczak, PhD, DDS; Borowska Małgorzata, PhD, DDS

Paedodontics Department, Medical University of Lublin, Poland.
*Corresponding author e-mail: maria.mielnik@am.lublin.pl


Introduction

Duchenne Muscular Dystrophy — DMD (dystrophia musculorum pseudohypertrophica) — is a disease that results from a mutation in a gene on the X chromosome, which prevents the production of dystrophin, a normal protein in muscle tissue. This gender-linked, recessive disorder affects males at a rate of one in 3,500 births; death usually occurs within 15 years of clinical onset.1 DMD is characterized by a progressive degeneration of particular groups of muscles.2

The disease begins at a young age in the form of impaired mobility. It is caused by symmetrical dystrophy of muscles, initially of the pelvic girdle and spine extensors. As the disease progresses, other symptoms develop, e.g., contractions in the knee, ankle and hip joints. These result from the asymmetric distribution of muscular atrophy. Walking becomes waddling and lumbar lordosis is pronounced.3,4

Weakening of the muscular force gradually affects other groups of muscles (shoulder girdle, forearms and wrists), which leads to immobility of the child commonly between the ages of 9 to 14 years. At that time, serious respiratory failure develops due to chest and spinal deformities. Tendon reflexes become weak and finally disappear. However, reflex sensitivity is not impaired.5,6

Around the oral cavity, muscular atrophy is observed, often masked by the overgrowth of connective and adipose tissue. The process results in the hypertrophy (real and false) of the orbicularis oris, masseter muscles, and muscles of the lips and tongue.5,6

Defects within the jaws are also quite typical. Cross-bite and open bite malocclusion are frequently observed, probably caused by the changes in cranial base shape, i.e., shortening of its posterior part and increase of the basilar angle. With age, the open bite and increase in the angle between the base of the maxilla and the base of the mandible become more pronounced. Flattening of the palate is also visible.

According to Eckhard and Harzer, the activity of the masseter muscles diminishes about two years earlier than the orbicularis oris muscles. This, along with an enlarged hypotonic tongue, results in transverse widening of the alveolar arch, especially in the mandibular region with an associated cross-bite.

With greater hypotonia of the orbicularis oris, patients develop the habit of mouth breathing, which worsens the defect.6

Patients with DMD typically survive to 15 or 20 years of age. Death results due to circulatory failure or respiratory tract infection, especially for patients whose treatment of immobilizing joint contractures and spinal deformities was neglected.4,6

Patients with DMD may react adversely to general anesthesia, leading them to develop a complex resembling malignant hyperpyrexia.7 Malignant hyperpyrexia is a dangerous complication of general anesthesia, which occurs in individuals with an underlying muscle disease. The essential clinical features of the syndrome are a drastic and sustained rise in
body temperature, metabolic acidosis, and widespread muscular rigidity. This is caused by a massive and sudden release of calcium into the myoplasm from the calcium-storing membranes in the muscle cell when exposed to general anesthetic agents. This is why oral rehabilitation under general anesthesia is contraindicated for persons with DMD.16

The aim of this paper is to describe the oral health and prophylactic regimens used to prevent caries when providing dental treatment for children with DMD.

Materials and methods

In the Department of Pedodontics, Medical University of Lublin, seven patients with Duchenne Muscular Dystrophy have been treated since 1998. These patients were 10- to 14-year-old boys in whom the disease had reached the stage in which it was impossible for them to move (in most cases since they were 7 to 10 years of age).7

Patients were examined for the presence of malocclusion and oral hygiene. To evaluate the patients’ periodontal disease, the community periodontal index of treatment needs (CPITN), and caries experience (DMFT) were assessed using the ball end of a periodontal probe, sharp explorer, and dental mirror. Radiographs were used for evaluation when available. The survey considered issues at the time of diagnosis, such as what concomitant diseases were present, the past dental treatment of the patients, and any caries preventive measures undertaken so far.

Results and discussion

In this study, DMD was diagnosed at different ages, ranging from 3 to 8 years of age. In four patients, their elder brothers had also been diagnosed with DMD and they had died at the ages of 14 to 18 years.

One mother of two children with DMD was found through a genetic profile test to be a carrier of the disease; this diagnosis was clinically confirmed by neurologological examination; the mother had symptoms such as fatigue and problems with walking up stairs.14 Apart from the basic disease, the boys had either mild or severe mental impairment, infantile cerebral palsy or epilepsy.

In the course of the dental examination, open bite was seen in two subjects and was accompanied by upper incisor retraction, mandible shape changes (increased angle), and enlarged, hypotonic tongues in most of the boys (Figure 1). The open bite malocclusion was associated with mouth breathing and a large, hypotonic tongue.

In all the patients, delayed eruption of the permanent dentition was observed, most frequently related to the premolars. A radiographic examination revealed the presence of all permanent tooth buds (Figure 2).

The mean DMF (Decayed, Missing, Filled) in the group was 6 at the time treatment was introduced. Oral hygiene status was unsatisfactory. Gingival inflammation, heavy plaque accumulation and calculus deposits were observed around the lower anterior teeth. The remaining primary teeth had extensive caries.

Five patients (71.5%) had a maximum community periodontal index (CPI) of 2. Two others had slight gingivitis (CPI-1). None of the patients had a healthy periodontium. High DMFT and CPI values could be related to difficulties with daily oral hygiene and malocclusion, accompanied by mouth breathing and lack of regular professional dental check-ups according to data from the patients’ dental histories.

One patient (PL, Figure 3) had large erosions of the enamel of the maxillary incisors and hypoplastic changes taking the form of white spots on the enamel surface of the mandibular teeth. These may have been due to chemotherapy the patient had received in 1997 for granulomatosis of the lymph nodes (six cycles
ABVD for Hodgkin’s lymphoma in st. III B (yp. NS).

Preventive dental treatment in our department for patients with DMD includes a regular prophylaxis, use of topical fluoride, instruction on oral hygiene and home care, and restoration of carious and hypoplastic defects.

Dental and prophylactic treatment should be introduced to children with DMD at the earliest age possible. Dental treatment often is difficult because these young patients may be uncooperative and require the use of general anesthesia. However, it is not recommended for children with DMD to have general anesthesia as it may cause the patients to develop malignant hyperpyrexia — a life-threatening complication.

In the group of 10- to 14-year-old children suffering from Duchenne’s muscular dystrophy, the mean DMFT was 6, which was higher than the value obtained in the Polish national epidemiological study of a group of healthy 12-year-olds, where it was 4.23. In the group of Polish 12-year-olds, the DMFT values were closer to the data obtained in Romania (4.1) and Greece (3.7) and higher when compared to Norwegian children (2.4). Similarly, the boys with DMD had periodontal disease as none had a healthy periodontium compared to 25% in the national study. A high percentage (71.5%) of the children with DMD had calculus on their teeth compared to 21% in the national study.

According to Symons et al., dental treatment and preventive measures should be implemented early in patients with DMD; they reported a satisfactory ability to maintain the teeth and periodontium in good health. In a group of 11- to 22-year-old patients with DMD, the mean DMFT was 2.9, which was significantly lower in comparison to the data obtained in Latvia (8.1), Germany (8.25), and Denmark (10.4).

There is no standard protocol for the prevention of both caries and periodontal diseases in patients with muscular dystrophy. Patients and their parents should be strongly encouraged to see the dentist regularly and it is important for them to understand the need for daily oral hygiene.

Dental healthcare program

For children with DMD, dental care 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.

The first stage of dental care for the child with DMD should involve a conversation with the parents (caregivers) concerning the following:

- proper diet
- oral hygiene (from 3 years of age oral hygiene agents should include fluoride)
- fluoride prophylaxis
- pit and fissure sealants and fluoride varnishes
- regular dental check-ups

From the age of 1, children should be gradually taught how to brush their teeth with a proper toothbrush, initially without toothpaste. Toothpaste should be introduced into oral hygiene after the child has learned to rinse his mouth (about the third year of the child’s life). In the same period, topical fluoride prophylaxis should be introduced, including fluoride varnish.

Conclusion

Dental and prophylactic treatment should be introduced at an early age in children with DMD. Lack of cooperation often makes dental treatment difficult in these young patients with DMD. While a general anesthetic is usually administered in such difficult patients, it is not recommended for children with DMD as it may cause the patient to develop malignant hyperpyrexia — a life-threatening complication.

References