Oral Care Programme

DM1 – Myotonic Dystrophy type 1

Mun-H-Center, a Swedish orofacial resource center for rare diseases and a specialist dental clinic, have many years experience from clinical treatment of patients with myotonic dystrophy type 1 (DM1). In this pamphlet we summarise the most important aspects of meeting and treating this group of patients.

Background

DM1 is an autosomal dominant neuromuscular disease, which means that one of the parents also has the disease. DM1 is caused by a mutation in the DMPK gene on chromosome 19.

The disease can be divided into four groups according to the age of onset: (1) congenital, (2) childhood onset, (3) classic and (4) mild forms. In general, the earlier the onset of the disease, the more severe are the symptoms. Muscles are affected to varying degrees, but other organs may also be affected. Common symptoms are muscle weakness, myotonia (rigidity in muscles after activation), cardiac symptoms (pacemakers are common), cataracts, gastro-intestinal problems, endocrine symptoms, fatigue, breathing difficulties and impaired sight and hearing. Learning difficulties and behavioural disorders are common among those with an onset before ten, but onset during adulthood can also affect cognitive functions.

The disease is slowly progressive. Weak muscles in and around the mouth cause impaired facial expression and an open mouth at rest. Muscle weakness can affect facial growth, which can cause malocclusion. A high arched palate and open bite are common. Temporomandibular disorders may occur.

There is an increased sensitivity to many anaesthetics.

Dental Care for People with DM1

Risk Factors for Oral Health

Most people with DM1 need regular and enhanced preventive dental care. There is an increased risk of developing oral diseases, such as caries and periodontitis. Reasons include food remaining in the oral cavity, dryness in the mouth, frequent intake of simple carbohydrates when short of energy, difficulties in carrying out effective oral hygiene due to motor limitations and tiredness.

Advice for Relatives and Assistants

Young people and adults with DM1 may also need assistance from relatives and assistants in carrying out oral care. To do this in the best way, they need practical guidance from the dental care and access to oral care equipment.

References

- Ahlborg B, Carlsson A, Kroksmark A-K, Lundälv E, Persson M, Zellmer M. Hjälpmedelsutprovning vid mun- och tandvård. Mun-H-Center förlag 2011.
- Antonarakis GS, Herzog G, Kiliaridis S. Vertical relapse after orthodontic and orthognathic surgical treatment in a patient with myotonic dystrophy. Eur J Paediatr Dent. 2019 Mar;20(1):53-58. doi: 10.23804/ejpd.2019.20.01.11.
- Engvall M, Birkhed D. Oral sugar clearance and other caries-related factors in patients with myotonic dystrophy. Acta Odontol Scand. 1997 Apr;55(2):111-5.
- Engvall M, Sjögreen L, Kjellberg H, Robertsson A, Sundell S, Kiliaridis S. Oral health in children and adolescents with myotonic dystrophy. Eur J Oral Sci 2007; 115: 192–197.
- Lindvall B (red). Dystrophia Myotonica (DM1), Skandinaviskt konsensusprogram, 2010 version 3, 2010-01-07.
- Mårtensson Å, Ekström A-B, Engvall M, Sjögreen L. Oral hygiene aspects in a study of children and young adults with the congenital and childhood forms of myotonic dystrophy type 1. Clinical and Experimental Dental Research. 4 AUG 2016 | DOI: 10.1002/cre2.36
- Sjögreen L, Tulinius M, Kiliaridis S, Lohmander A. The effect of lip strengthening exercises in children and adolescents with myotonic dystrophy type 1. Int J of Pediat Otorhinolaryngol 2010;74:1126-1134.

Links

- Socialstyrelsens kunskapsdatabas om ovanliga diagnoser: http://www.social- styrelsen.se/ovanligadiagnoser/dystrofi amyotonika
- Ågrenskas dokumentation, Dystrofia myotonika 2017 och Dystro a myotonika, vuxenperspektivet 2018: https:// www.agrenska.se/vi-erbjuder/ informationsmaterial/ dokumentationer/

If you want more guidance about dental treatment and oral care for those with myotonic dystrophy, or if you have any comments on the content of the oral care programme, please contact us at mun-h-center@vgregion.se/en, or call +49 10-441 79 80.

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Recommended oral care programme when treating patients with DM1 – Myotonic Dystrophy type 1

Adapt the dental care environment

- Use supportive cushions (Lasal dental cushions for example) to provide stability, security and better comfort in the dentist's chair.

Appointment procedures

- Consider using images in the invitation if the patient has cognitive difficulties.
- Arrange plenty of time for the appointment.
- Check with patients whether they want to have an extra reminder for the appointment just beforehand.
- Arrange the appointment to suit the patient's routines/ daily rhythm. Remember the patient may be tired; avoid any other tiring activities on the same day.

Adapt the dental treatment

- Work in short sessions if the patient:
  - needs to change position
  - has problems with breathing or phlegm production
  - is generally affected and tired
  - finds it difficult to keep the mouth open for a long time.
- Equipment that should be available to make dental treatment easier: bite support, mouth angle expander, oral wipes.
- If the patient finds it difficult to take part in dental care:
  - create a calm environment
  - offer an induction programme
  - use individually designed images for support.

Improved prophylaxis

- Annual dental examination (more often if necessary).
- Regular support treatment by dental hygienist (individual).
- Regular examination of jaw joints and range of jaw movement.

Orthodontics

- Orthodontics may be needed to reduce severe crowding, but also to reduce moderate crowding to make good oral hygiene easier.
- Work in short sessions.
- Improved prophylaxis during treatment.
- Life-long retention is often required, which should be followed.
- Orthognathic surgery is often required to reduce severe open bite but there is a major risk for relapse.

Oral motor training

- Muscle training with an oral screen should be considered from the age of five in the case of weak lip muscles.
- Muscle training with a Chewy tube should be considered from the age of three in the case of weak masseter muscles.
- Stretching with Therabite or Jaw trainer should be considered for young people and adults with impaired opening ability.
- Cooperation with a speech-language pathologist is recommended.

In cases with dysphagia

- Treat the patient in a semi-sitting position with the head tilted to one side.
- Consider drying the mouth with wipes if the patient cannot spit or rinse the mouth.

Sedation

- Always consult the attending doctor before giving sedation.
- Contraindications include muscle drugs.
- Anaesthesia:
  - It is important to inform about the diagnosis before a planned operation.
  - Increased sensitivity to many anaesthetics.
  - Post-operative monitoring.

Special dental care support

- If necessary, assist the patient with the application for dental care support.

Accompanying carers and assistants

- If necessary, give practical advice about oral care to accompanying carers and assistants.
- Give instructions on adapted working positions and useful equipment for oral care in the home.
- Give motivation and feedback on the carer's/ assistant's efforts. One way may be to make a running documentation of the patient's oral health using photos.

Self-care for young people and adults

- Let patients show how they manage to carry out their oral hygiene. If necessary, correct them gradually.
- Ask them how oral care works out at home. Are adaptations in the bathroom needed, such as a height adjustable washbasin? Contact an occupational therapist in the rehabilitation unit.
- Instruct the patient about adapted positions and good tools for oral care in the home.
- Individually adapted image support and a timetable programme can be used to help remember oral care routines.

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