



# Orofacial function of persons having Becker muscular dystrophy

## Report from observation charts

The survey comprises 11 observation charts.

**Estimated occurrence:** The total number of persons in Sweden with Becker muscular dystrophy is unknown; however, its incidence is estimated to be about 40 per 1 000 000 male inhabitants. The disease occurs nearly exclusively in boys.

**Etiology:** The disease is caused by a mutation in the dystrophin gene. The type of mutations occurring in Becker muscular dystrophy results in severe deficiencies of the protein dystrophin in the muscle fibers. Dystrophin is important for muscle fiber cohesion. The absence of dystrophin renders the muscle fibers more susceptible to degeneration. The dystrophy gene is located on the X chromosome. In about a third of the cases, the disease is caused by a new mutation.

**General symptoms:** The symptoms associated with Becker muscular dystrophy closely resemble those of Duchenne muscular dystrophy but are milder. Pronounced symptoms may however sometimes occur. Although muscle weakness is observed in a few cases during infancy, symptoms appear much later in many. Muscle weakness nearly always begins in the legs. About 20% of the cases will require some aid of a wheelchair; the age at which this occurs greatly varies. Muscle weakness also often progresses with age to the upper body and arms. As dystrophin is normally found in the skeletal musculature and heart muscle, as well as to a lesser degree in the central nervous system, symptoms from the heart and central nervous system may occur. Most have no symptoms from the respiratory tract. A tendency to slightly lower learning ability is occasionally observed.

### **Orofacial/ odontological symptoms:**

Muscle weakness in the masticatory and facial muscles, especially if occurring at an early age, may lead to malocclusions. Muscle strength and endurance in the masticatory musculature may gradually decrease. It then becomes more difficult to masticate food, which in turn may eventually lead to altered dietary habits. In cases of reduced muscle strength in the arms and hands, toothbrushing may be difficult.

### **Orofacial/ odontological treatment:**

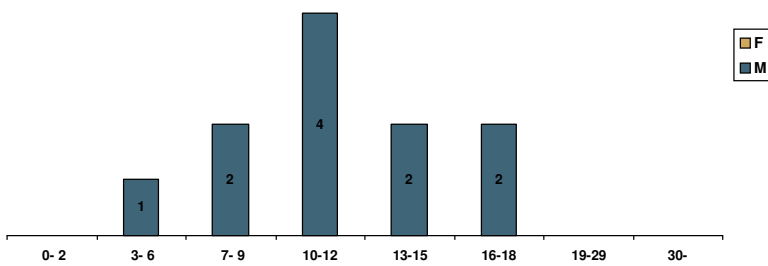
- Problems in managing toothbrushing and dental care due to tired and weak arm and jaw muscles justify extra prophylactic dental care.
- Orofacial assistive devices should be tried when needed; among these, an electric toothbrush may be of great help.
- Eating difficulties should be investigated and treated by a specialist team.

### **Sources**

The rare disease database of the Swedish National Board of Health and Welfare.

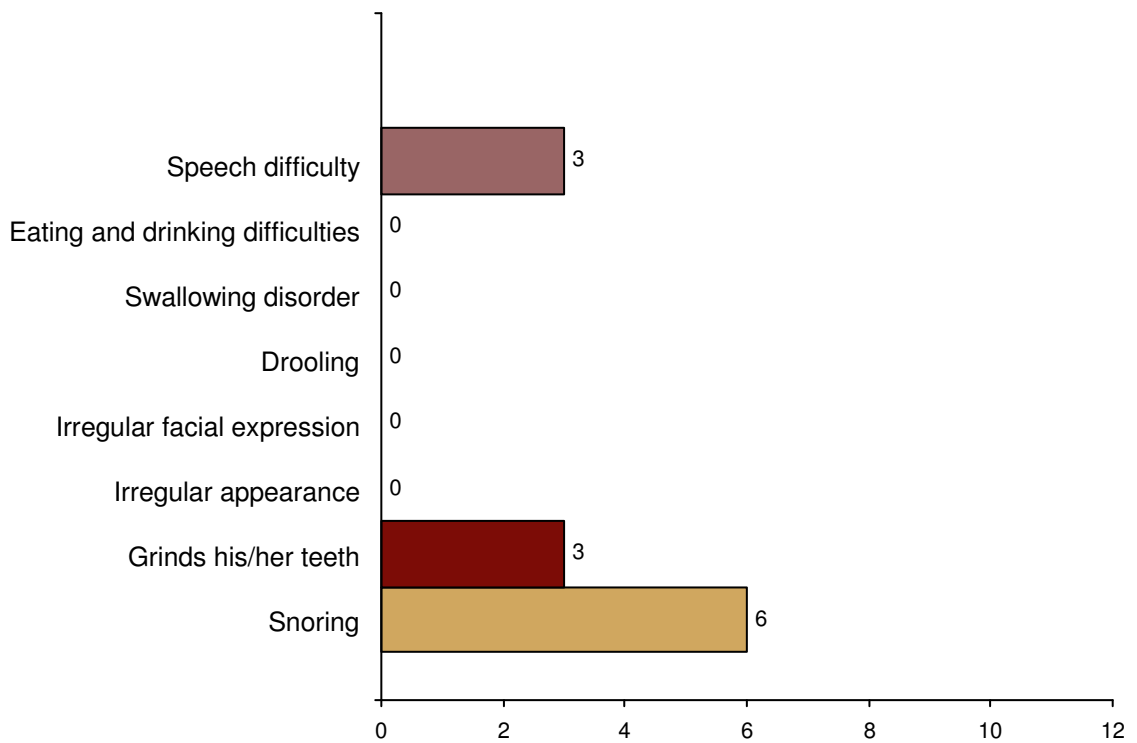
The MHC database - The Mun-H-Center database of orofacial manifestations in rare diseases.

Age distribution

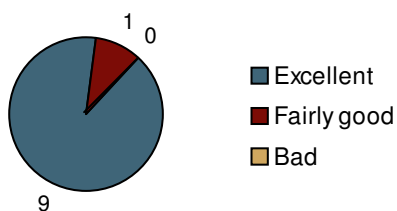


Number: 11  
 Ages: 3 -- 18 years  
 Sex: M (11) + F (0)

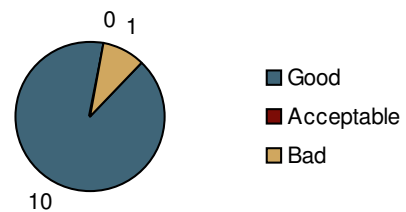
Orofacial problems



Oral health

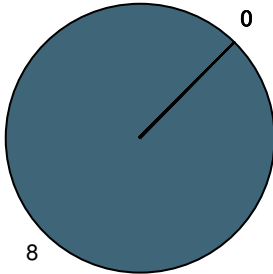


Oral hygiene



## Behaviour in the treatment situation

How calm and co-operative is the patient at time of examination?



- (8) ■ Examination is possible without problem
- (0) ■ Examination is possible without problems, some reaction is observed
- (0) ■ Examination can continue if adjusted to patient's reactions
- (0) ■ Reactions are considerable and examination is obviously affected
- (0) ■ Examination is practically impossible to complete
- (0) ■ Patient refuses examination

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0=/10=

Age

If there are treatment problems:

0=/10=

0=/10=

Age

Age



<b>Clinical findings</b>	<b>Total N=11</b>	<b>Boys/Men N=11</b>	<b>Girls/Women N=0</b>	<b>Not evaluated</b>
Cross bite	4	4	0	0
Speech difficulty	3	3	0	0
Post normal bite	2	2	0	0
Tongue seems large	2	2	0	0
Deep bite without gingival contact	1	1	0	0
Dental trauma	1	1	0	0
Edge to edge bite	1	1	0	0
Enamel hypoplasia	1	1	0	0
Frontal open bite	1	1	0	0
Gingivitis	1	1	0	0
Hypomineralisation	1	1	0	0
Narrow palate	1	1	0	0
Over crowding	1	1	0	0
Posturing forward	1	1	0	0
Reduced mobility in tongue	1	1	0	0
Retroclined lower incisors	1	1	0	0
Upper jaw seems small	1	1	0	0