



Orofacial function of persons having Becker muscular dystrophy

Report from observation charts



20 observation charts

Synonym

ICD-10 G71.0B

Estimated occurrence 3-5:100,000 live births. Only affects boys.

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.

Oral 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.

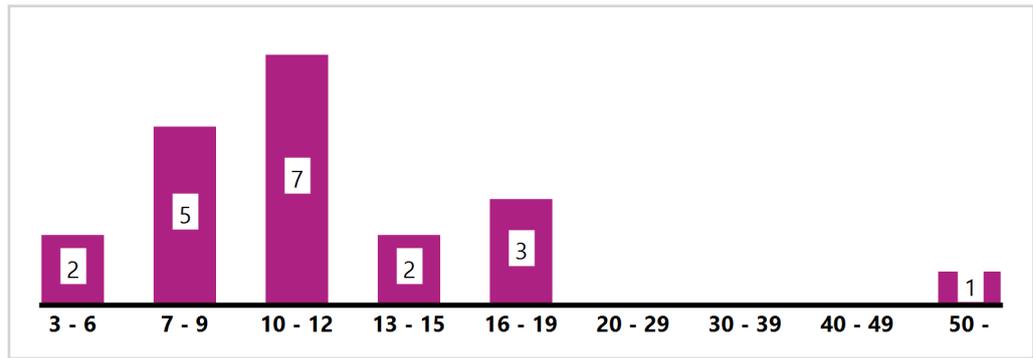
Oral treatment

- Problems in managing toothbrushing and dental care due to tired and weak arm and jaw muscles justify extra prophylactic dental care.
- An orthodontist should be consulted between the ages of 7 and 9 in order to identify dental aberrations or malocclusions and to plan any necessary orthodontic treatment.
- 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.

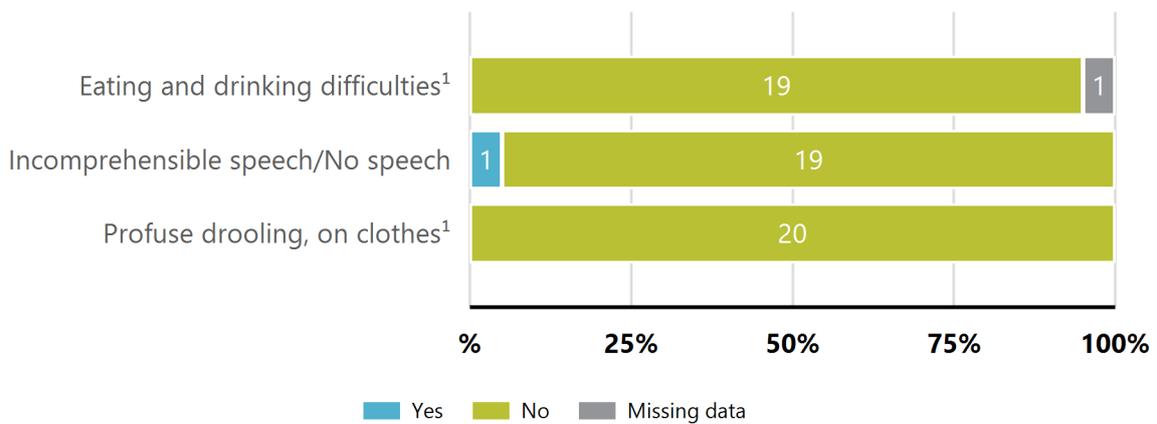
Sources The MHC database Rare diseases Dokumentation-Ågrenska

Age distribution

Count: 20
 Ages: 3 - 61
 ♂ (20)
 ♀ (0)



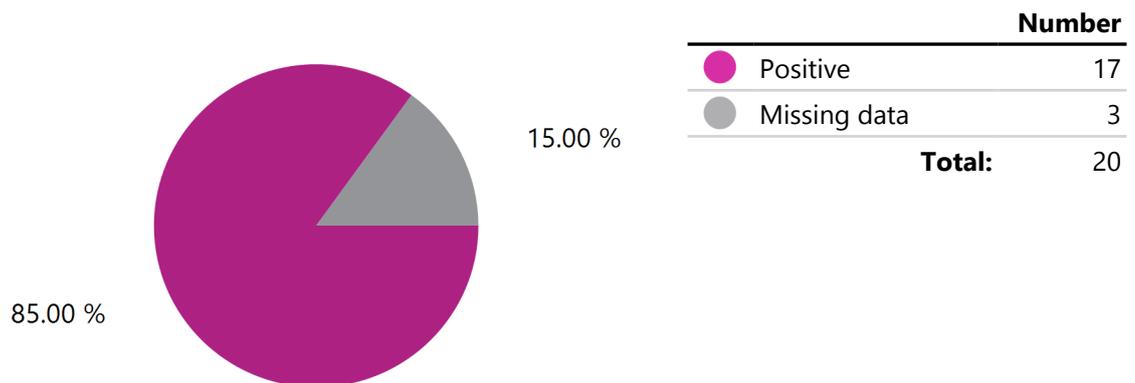
Summary



Certain caution should be observed when interpreting the chart because the number of individuals is less than 100.

¹ Reported via questionnaire

Acceptance of dental examination



Oral health

Caries

| | | 3-6 years | 7-12 years | 13-19 years | >19 years |
|---|--|-----------|------------|-------------|-----------|
| deft | Examined | 2 | 12 | | |
| Number of carious or filled deciduous teeth | Number of individuals with deft=0 | 2 | 10 | | |
| | Mean | 0.0 | 0.2 | | |
| | Standard deviation | | 0.4 | | |
| | No data | 1 | 3 | | |
| DMFT | Examined | | 12 | 5 | 1 |
| Number of carious or filled permanent teeth | Number of individuals with DMFT=0 | | 12 | 3 | |
| | Mean | | 0.0 | 3.0 | 17.0 |
| | Standard deviation | | 0.0 | 4.4 | |
| | No data | | 3 | 2 | 0 |

Oral health index (indices)¹

| | 0 | 1 | 2 | 3 | 4 | 5 | 6 | Missing data | Number |
|-------------------|---|---|---|---|---|---|---|--------------|--------|
| Calculus | 9 | 0 | 0 | 0 | 0 | 0 | 0 | 11 | 9 |
| Gingivitis | 8 | 0 | 1 | 0 | 0 | 0 | 0 | 11 | 9 |
| Plaque | 5 | 2 | 0 | 2 | 0 | 0 | 0 | 11 | 9 |
| Tooth wear | 8 | 1 | 0 | 0 | 0 | 0 | 0 | 11 | 9 |

CALCULUS

Calculus index is based on the presence of visible calculus on the buccal surface of 6 index teeth. 0 indicates that there is no calculus at all, 6 indicates calculus on all index teeth.

GINGIVITIS

Gingivitis index is based on the presence of visible gingivitis on the buccal surface of 6 index teeth. 0 indicates that there is no bleeding, 6 indicates bleeding on all index teeth.

PLAQUE

Plaque index is based on the presence of visible plaque on the buccal surface of 6 index teeth. 0 indicates that there is no plaque, 6 indicates plaque on all index teeth.

TOOTH WEAR

Tooth wear index is a weighted summary of the degree of tooth wear on 6 different segments. Tooth wear is only evaluated in the permanent dentition, not in the primary teeth. The final index score is based on the degree of tooth wear found in most segments.

0: No tooth wear or minor wear of enamel in either of the segments.

1: Marked tooth wear of the enamel, possibly exceeding into dentin.

2: tooth wear in the dentine reaching up to 1/3 of the tooth crown.

3: Tooth wear in the dentine reaching up to more than 1/3 of the tooth crown. If 3 is given in any segment then SI is 3.

¹ This variable was introduced in version 2 (2008) of the Observation chart.

Occlusal relationship

| | Number |
|---------------------|---------------|
| Neutral bite | 18 |
| Overbite | 2 |
| Underbite | 0 |
| Missing data | 0 |
| Total: | 20 |

Maximum jaw opening

Children younger than 10 years

Children, 10 years or older, and adults

| | Number |
|---------------------|---------------|
| -20 | 0 |
| 21-30 | 0 |
| 31-40 | 4 |
| 41-50 | 2 |
| 51- | 0 |
| Missing data | 1 |
| Total: | 7 |

| | Number |
|---------------------|---------------|
| -20 | 0 |
| 21-30 | 0 |
| 31-40 | 0 |
| 41-50 | 7 |
| 51- | 6 |
| Missing data | 0 |
| Total: | 13 |

Profile¹

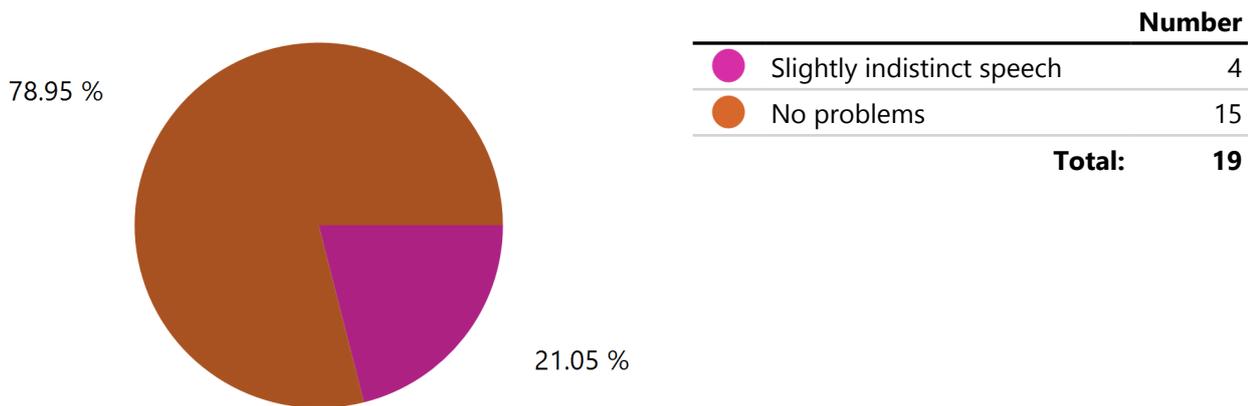
| | Number |
|---------------------|---------------|
| Normal | 9 |
| Convex | 0 |
| Concave | 0 |
| Missing data | 0 |
| Total: | 9 |

Mandibular plane¹

| | Number |
|---------------------|---------------|
| Normal | 8 |
| Increased | 1 |
| Reduced | 0 |
| Missing data | 0 |
| Total: | 9 |

¹ This variable was introduced in version 2 (2008) of the Observation chart.

Speech problems



Clinical findings

Number of yes-answers

| | Total = 20 (%) | ♂ = 20 (%) | ♀ = 0 (%) | Missing Data |
|--|----------------|------------|-----------|--------------|
| Open mouth at rest | 2 (10) | 2 (10) | #Error | 0 |
| Frontal open bite | 2 (10) | 2 (10) | #Error | 0 |
| Impaired tongue motility | 2 (10) | 2 (10) | #Error | 0 |
| Over crowding | 2 (10) | 2 (10) | #Error | 0 |
| Deep bite with gingival contact | 2 (10) | 2 (10) | #Error | 0 |
| Narrow palate | 1 (5) | 1 (5) | #Error | 1 |