



Orofacial function of persons having Duchenne muscular dystrophy

Report from observation charts



114 observation charts

Synonym

ICD-10 G71.0A

Estimated occurrence 30:1,000,000 inhabitants. Only affects boys.

General symptoms Symptoms normally present around the age of three. The child develops a waddling gait, has difficulty running, jumping and getting up off the floor. The thoracic girdle, the pelvic and the musculature of the back all become progressively weak. Upper arms and thighs are affected earlier than lower arms and calves. Curvature of the lumbar region develops. These boys generally stop being able to walk around the age of 10 to 12. Impaired cardiac function often develops. Muscular weakness increases over the years. Many individuals develop contractures and scoliosis. The respiratory musculature is weakened, and lung function reduced. Malignant hyperthermia has been reported and should be considered when treatment under general anaesthesia. There may be concentration problems and learning disability.

Oral symptoms Weakness of the main muscles used for chewing, the facial muscles and the tongue. Malocclusions develop successively owing to muscular weakness and the changed morphology of the tongue. The jaws increase in width, more in the lower jaw than in the upper, which often results in a posterior cross-bite. Later a lateral open bite may appear and later on a frontal open bite can be seen. Reduced muscular strength and endurance of the chewing muscles in combination with lack of occlusal (bite) contact may result in difficulties masticating food. In late stages of the disease, swallowing difficulties often develop. The mouth opening capacity can be reduced.

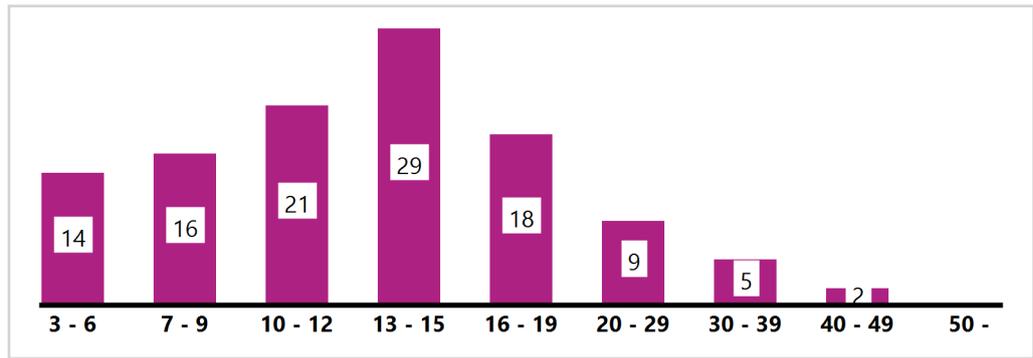
Oral treatment

- Problems in managing oral hygiene and eating difficulties justify extra preventive dental care. Co-operation with a dental hygienist and an occupational therapist is recommended.
- It is important to bear in mind in relation to dental treatment, that individuals with cardiac and respiratory difficulties have trouble sitting in a reclining position.
- Stretching of the jaw opening is required if the jaw opening capacity is reduced.
- When treating medically compromised patients always contact their doctors for medical advice (bleeding problems, heart diseases etc.).
- There is an increased risk of malignant hyperthermia during general anaesthesia.
- Eating and swallowing difficulties are investigated and treated by a specialist team at the hospital or multidisciplinary treatment centre.

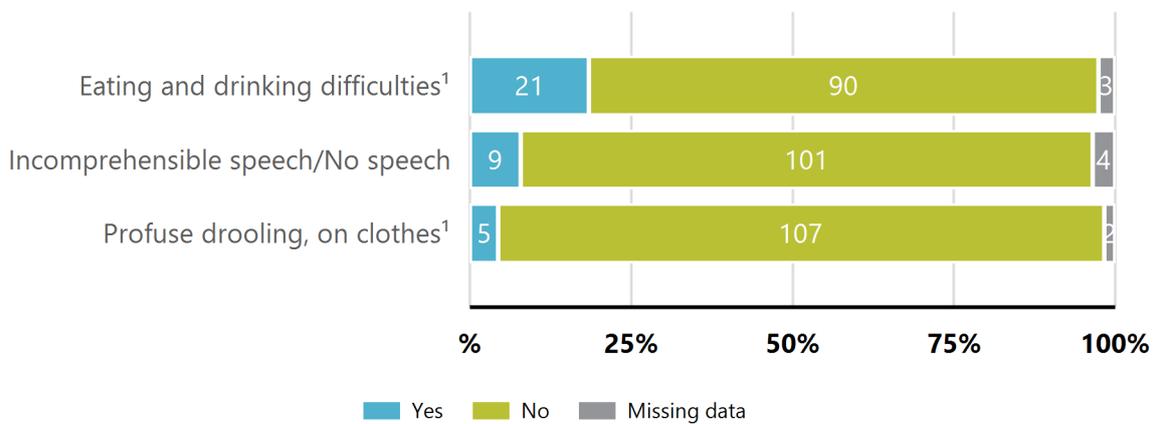
Sources The MHC database Rare diseases Dokumentation-Ågrenska

Age distribution

Count: 114
 Ages: 4 - 48
 ♂ (114)
 ♀ (0)

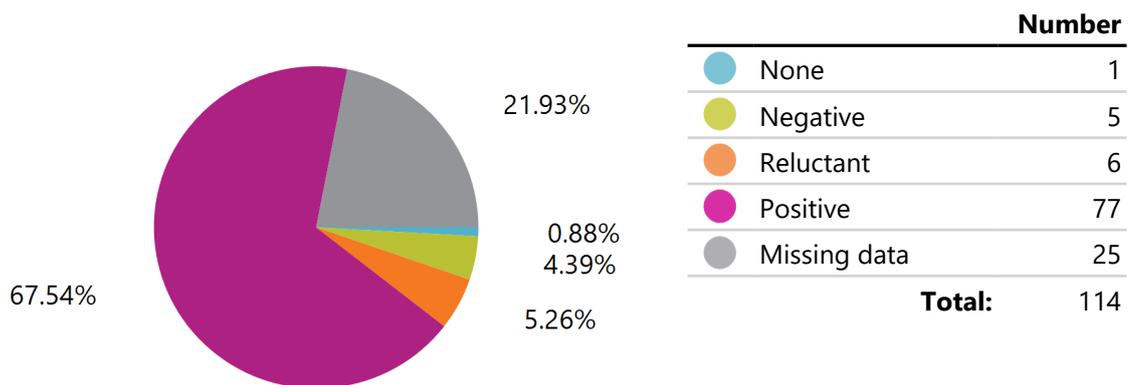


Summary



¹ Reported via questionnaire

Acceptance of dental examination



Oral health

Caries

		3-6 years	7-12 years	13-19 years	>19 years
deft	Examined	14	37		
Number of carious or filled deciduous teeth	Number of individuals with deft=0	13	33		
	Mean	0.6	0.7		
	Standard deviation	1.5	1.5		
	No data	7	20		
DMFT	Examined		37	47	16
Number of carious or filled permanent teeth	Number of individuals with DMFT=0		37	40	9
	Mean		0.0	0.9	3.0
	Standard deviation		0.0	2.1	4.8
	No data		20	20	4

Oral health index (indices)¹

	0	1	2	3	4	5	6	Missing data	Number
Calculus	49	9	5	1	0	0	0	50	76
Gingivitis	38	14	6	4	2	0	0	50	76
Plaque	24	10	8	8	7	4	3	50	76
Tooth wear	50	15	0	0	0	0	0	49	76

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	71
Overbite	14
Underbite	17
Missing data	12
Total:	114

Maximum jaw opening

Children younger than 10 years

	Number
-20	0
21-30	0
31-40	7
41-50	12
51-	2
Missing data	9
Total:	30

Children, 10 years or older, and adults

	Number
-20	3
21-30	5
31-40	13
41-50	36
51-	21
Missing data	6
Total:	84

Profile¹

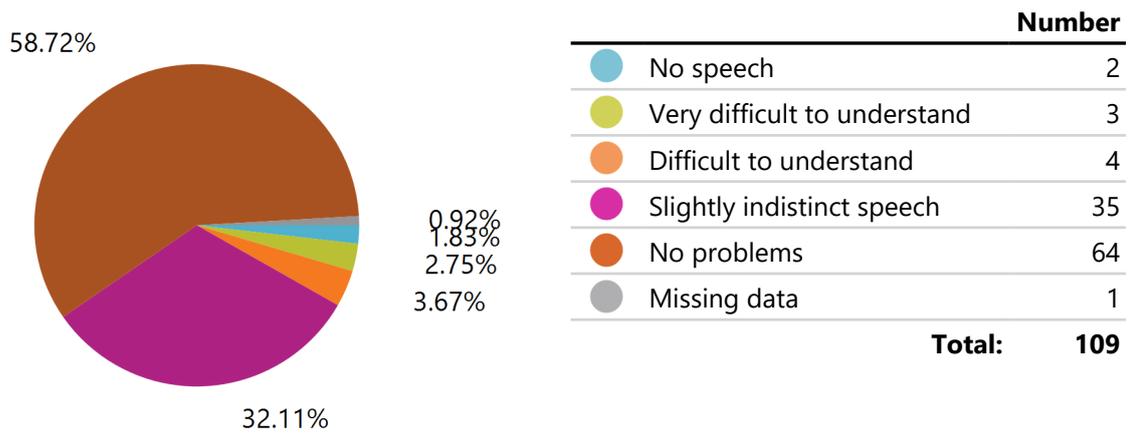
	Number
Normal	61
Convex	3
Concave	4
Missing data	6
Total:	74

Mandibular plane¹

	Number
Normal	60
Increased	5
Reduced	0
Missing data	11
Total:	76

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

Speech problems



Clinical findings

Number of yes-answers

	Total = 114 (%)	♂ = 114 (%)	♀ = 0 (%)	Missing Data
Frontal open bite	49 (43)	49 (43)	#Error	11
Reduced stability in neck	46 (40)	46 (40)	#Error	5
Low muscle tone in masticatory muscles	30 (26)	30 (26)	#Error	12
Open mouth at rest	27 (24)	27 (24)	#Error	2
Over crowding	26 (23)	26 (23)	#Error	8
Low muscle tone in lips	22 (19)	22 (19)	#Error	11
Low muscle tone in tongue	17 (15)	17 (15)	#Error	6
Reduced opening capacity	16 (14)	16 (14)	#Error	5
Impaired tongue motility	14 (12)	14 (12)	#Error	4
Spacing	9 (8)	9 (8)	#Error	9
High palate	5 (4)	5 (4)	#Error	13
Short tongue frenulum	5 (4)	5 (4)	#Error	6
Facial asymmetry	4 (4)	4 (4)	#Error	6
High muscle tone in lips	4 (4)	4 (4)	#Error	7
Narrow palate	3 (3)	3 (3)	#Error	13
High muscle tone in tongue	2 (2)	2 (2)	#Error	4

NOT-S

Total mean score:
2.58

Number: 69
Ages: 4 - 48

♂ (69)

♀ (0)



NOT-S interview

		Count	%
I	Sensory function	7	10.1%
II	Breathing	24	34.8%
III	Habits	21	30.4%
IV	Chewing and swallowing	24	34.8%
V	 Drooling	3	4.3%
VI	Dry mouth	6	8.7%

NOT-S examination

		Count	%
1	Face at rest	20	29.0%
2	Nose breathing	0	0.0%
3	Facial expression	20	29.0%
4	Masticatory muscle and jaw function	20	29.0%
5	Oral motor function	9	13.0%
6	Speech	24	34.8%

The report is based on data from the MHC database - the Mun-H-Center database on oral health and orofacial function in rare diseases. Data was collected by dentists and speech-language pathologists using the Orofacial Observation Chart.