



Orofacial function of persons having Duchenne muscular dystrophy

Report from questionnaires



88 questionnaires

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.

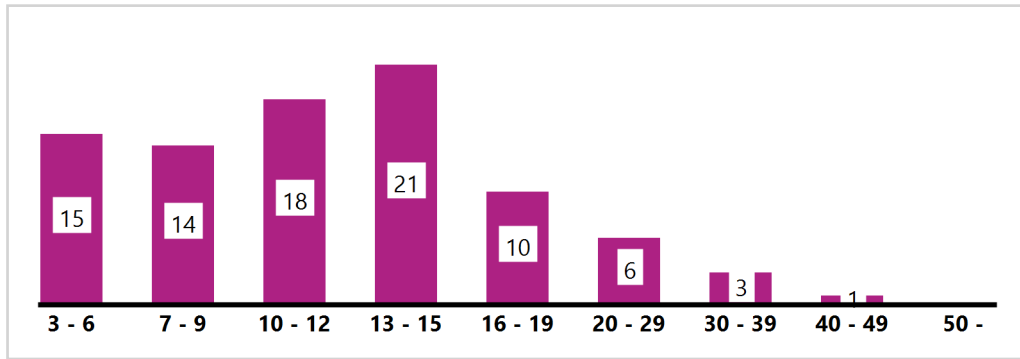
Sources The MHC database Rare diseases Dokumentation-Ågrenska

Age distribution

Number: 88

Ages: 4 - 48

♂ (88)
♀ (0)



Additional diagnoses

Medical impairment	Yes	No	Missing data
Inborn heart defect	1	84	3
Other cardiovascular disease	8	77	3
Epilepsy	0	87	1
Asthma	4	83	1
Need of respiratory support	12	75	1
Allergy	14	73	1

Neuropsychiatric diagnosis	Yes	No	Missing data
ADHD/ADD	16	68	4
Autism (Includes autism, Asperger syndrome and autistic traits)	13	37	38

General disability	Yes	No	Missing data
Intellectual disability	11	69	8
Motoric functional impairment	16	19	53
Visual impairment	13	70	5
Hearing loss	2	85	1
Communication difficulties	11	62	15

About dental care and oral health

Do you feel that you receive the dental care you need?	Number
Yes, very much so	55
Yes, somewhat	25
No, not really	5
No, not at all	1
Missing data	2
Total:	88

How many times per year do you normally seek dental care?	Number
Less than once per year	8
One time per year	28
Two times per year	19
Three or more times per year	32
Missing data	1
Total:	88

When were your teeth last X-rayed?	Number
During the past two years	61
More than two years ago	7
Never had my teeth X-rayed	15
Missing data	5
Total:	88

Do you look after your teeth in a good way?	Number
Yes, very much so	49
Yes, somewhat	33
No, not really	3
Missing data	3
Total:	88

Who brushes your teeth?	Number
I always brush myself	12
Someone else always helps me	50
Sometimes I brush myself	25
Missing data	1
Total:	88

How often are your teeth brushed?	Number
Not everyday	1
Once per day	14
Two times per day	68
Three or more times per day	4
Missing data	1
Total:	88

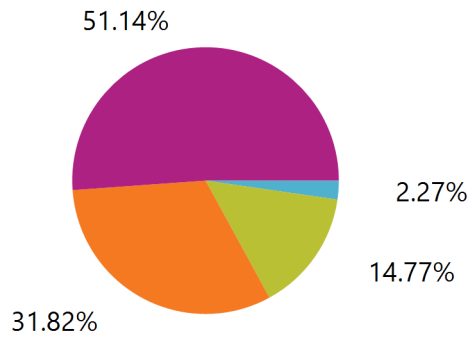
About dental care and oral health

	Yes	No	Missing data
Does your mouth hurt?	9	77	2
Does your mouth feel dry?	12	72	4
Have you ever taken a serious hit to your permanent front teeth?	14	71	3
Do you feel that you have a divergent bite?	16	64	8
Have you had a brace?	4	82	2
Do you feel that you need orthodontics/a brace?	13	70	5

Do you grind or press your teeth at night?	Number
Never	74
Once or twice per week	7
Every night	5
Missing data	2
Total:	88

Do you grind or press your teeth during the day?	Number
Never	81
Once or twice per week	4
Every night	1
Missing data	2
Total:	88

About eating



Do you have any problems with eating?

Response	Number
Yes, very much so	2
Yes, somewhat	13
No, not really	28
No, not at all	45
Missing data	0
Total:	88

	Yes	No	Missing data
Do you cough daily in connection with meals?	2	86	0
Do you gag daily in connection with meals?	0	87	1
Do you get acid reflux daily?	3	80	5
Do you throw up often (at least twice per week)?	0	87	1
Do you have a poor appetite?	12	74	2
Does it take a long time before you can swallow a mouthful?	3	41	44
Do you press your tongue forward when you swallow so that food ends up outside the mouth?	0	46	42
Do you find it difficult to chew, i.e. grind food using your molars?	14	72	2
Do you find it difficult to take food from the spoon using your lips?	1	84	3
Have you had problems with food and drink leaking out through the corners of your mouth?	1	45	42
Does food tend to remain in your mouth after meals?	10	75	3
Do you get nutrition in any other way than through your mouth?	4	84	0

About drooling

Do you drool?	Number
Never drool	83
Drool sometimes – not every day	3
Missing data	0
Total:	88

How much do you drool?	Number
Slight drooling, only on the lips	2
Moderate drooling, on lip and chin	1
Total:	3

Is your drooling a problem for you?	Number
Yes, somewhat	1
No, not really	1
No, not at all	1
Total:	3

Is your drooling a problem for your family or people around you?	Number
No, not really	1
No, not at all	2
Total:	3