Orofacial function of persons having
Charcot-Marie Tooth disease

Report from observation charts | 10 observation charts

Synonym	Hereditary motor sensory neuropathy (HMSN), Peroneal muscular atrophy (PMS)
ICD-10	G60.0
Estimated occurance	30:100,000 inhabitants.
General symptoms	CMT is a peripheral nerve disease (polyneuropathy) that affects the muscles entailing a slowly progressing muscle weakness in legs, feet, hands and arms. Foot drop, extremely high arch, hammer toes, scoliosis and poor balance are common. Respiratory problems may occur. The loss of nerve function often causes a pricking and a burning sensation in hands and feet. CMT belongs to the group of neuromuscular diseases.
Oral symptoms	CMT has no specific affect on teeth and bite. Nor are the muscles in the mouth and face generally affected since they are controlled by cranial nerves. These nerves are not usually affected by the disease. If children are extremely tired or if breathing is affected, speech may be somewhat unclear. Some medicines can cause dryness in the mouth.
Oral treatment	• In the event of dryness in the mouth intensified preventive dental care is recommended.
Sources	The MHC database Rare diseases Dokumentation-Ågrenska

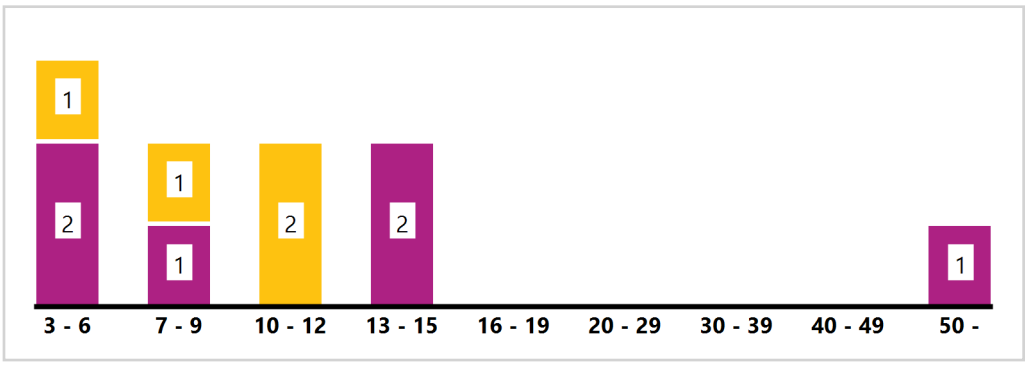
Age distribution

Count: 10

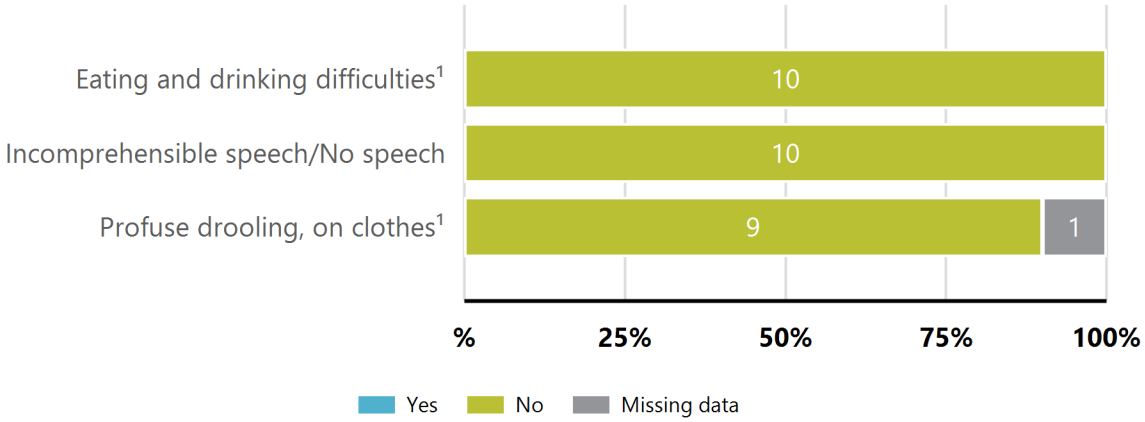
Ages: 4 - 53

♂ (6)

♀ (4)



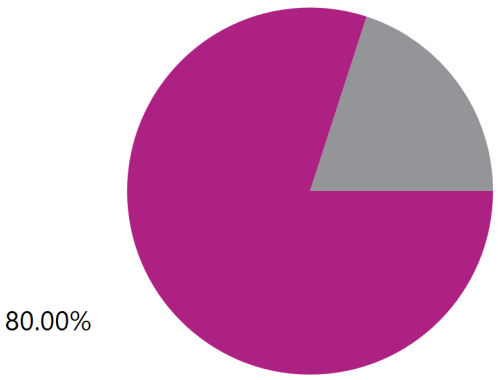
Summary



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



	Number
Positive	8
Missing data	2
Total:	10

Oral health

Caries

		3-6 years	7-12 years	13-19 years	>19 years
deft	**Examined**	3	4		
Number of carious or filled deciduous teeth	**Number of individuals with deft=0**	3	4		
	Mean	0.0	0.0		
	Standard deviation	0.0	0.0		
	No data	0	1		
DMFT	**Examined**		4	2	1
Number of carious or filled permanent teeth	**Number of individuals with DMFT=0**		3	2	
	Mean		0.3	0.0	8.0
	Standard deviation		0.5	0.0	
	No data		0	0	0

Oral health index (indices)[1]

	0	1	2	3	4	5	6	Missing data	Number
Calculus	8	1	0	1	0	0	0	0	10
Gingivitis	4	4	0	1	0	0	1	0	10
Plaque	4	2	1	2	0	0	1	0	10
Tooth wear	9	1	0	0	0	0	0	0	10

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	8
Overbite	1
Underbite	1
Missing data	0
Total:	**10**

Maximum jaw opening

Children younger than 10 years

	Number
-20	0
21-30	0
31-40	2
41-50	2
51-	1
Missing data	0
Total:	**5**

Children, 10 years or older, and adults

	Number
-20	0
21-30	0
31-40	0
41-50	2
51-	3
Missing data	0
Total:	**5**

Profile[1]

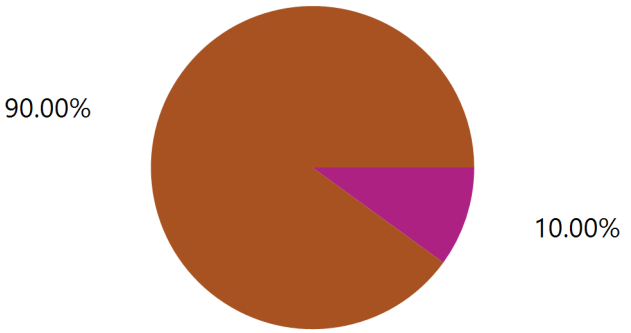
	Number
Normal	10
Convex	0
Concave	0
Missing data	0
Total:	**10**

Mandibular plane[1]

	Number
Normal	9
Increased	1
Reduced	0
Missing data	0
Total:	**10**

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

Speech problems



	Number
Slightly indistinct speech	1
No problems	9
Total:	**10**

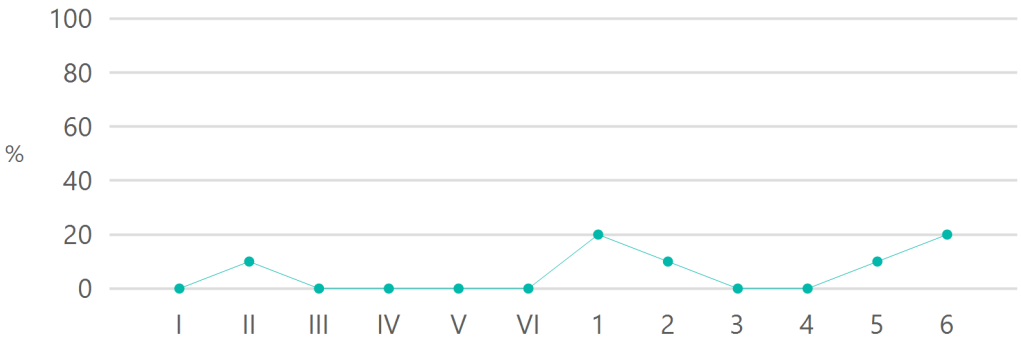
Clinical findings

Number of yes-answers

	Total = 10 (%)	♂ = 6 (%)	♀ = 4 (%)	Missing Data
Frontal open bite	3 (30)	3 (50)	0 (0)	0
Open mouth at rest	1 (10)	1 (17)	0 (0)	0
Over crowding	1 (10)	1 (17)	0 (0)	0
Reduced stability in neck	1 (10)	1 (17)	0 (0)	0
Spacing	1 (10)	1 (17)	0 (0)	0
Deep bite with gingival contact	1 (10)	1 (17)	0 (0)	0

NOT-S

Total mean score: 0.70



Number: 10
Ages: 4 - 53

♂ (6)
♀ (4)

NOT-S interview

		Count	%
I	**Sensory function**	0	0.0%
II	**Breathing**	1	10.0%
III	**Habits**	0	0.0%
IV	**Chewing and swallowing**	0	0.0%
V	**Drooling**	0	0.0%
VI	**Dry mouth**	0	0.0%

NOT-S examination

		Count	%
1	**Face at rest**	2	20.0%
2	**Nose breathing**	1	10.0%
3	**Facial expression**	0	0.0%
4	**Masticatory muscle and jaw function**	0	0.0%
5	**Oral motor function**	1	10.0%
6	**Speech**	2	20.0%

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.