



# Orofacial function of persons having Hereditary spastic paraplegia

## Report from observation charts

The survey comprises 24 observation charts.

**Synonyms:** Hereditary Spastic Paraplegia, Strumpell-Lorraine Syndrome

**Estimated occurrence:** Very rare.

**Etiology:** Genetic. Hereditary spastic paraplegia (HSP) consists of a number of subgroups with similar symptoms. A variety of inheritance patterns are represented. In persons with HSP, a gradual degeneration of motor neural structures takes place.

**General symptoms:** Spasticity and insidiously progressive muscle weakness that primarily affects gait pattern. Spasticity may lead to foot deformities. Arm and hand function are often unaffected. Unstable balance and ataxia (impaired control of voluntary movements) occur. Other symptoms that may be associated with HSP include speech difficulties, visual impairment, learning disability, epilepsy and urinary symptoms. Symptom onset occurs between 10 and 40 years of age.

**Orofacial/odontological symptoms:** Oral motor function may also be affected in HSP and produce symptoms such as eating difficulties, speech difficulties (dysarthria) and drooling. There are no known specific odontological symptoms associated with HSP.

**Orofacial/odontological treatment:**

- Speech and language impairments are to be diagnosed and treated by a speech-language pathologist
- Oral motor training and stimulation may be relevant in cases of eating difficulties, speech impairment and drooling

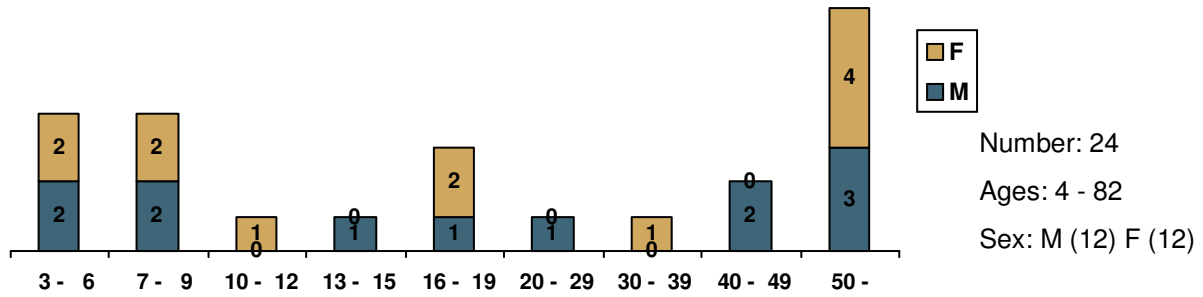
**Sources**

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

The MHC database - The Mun-H-Center database on oral health and orofacial function in rare diseases.

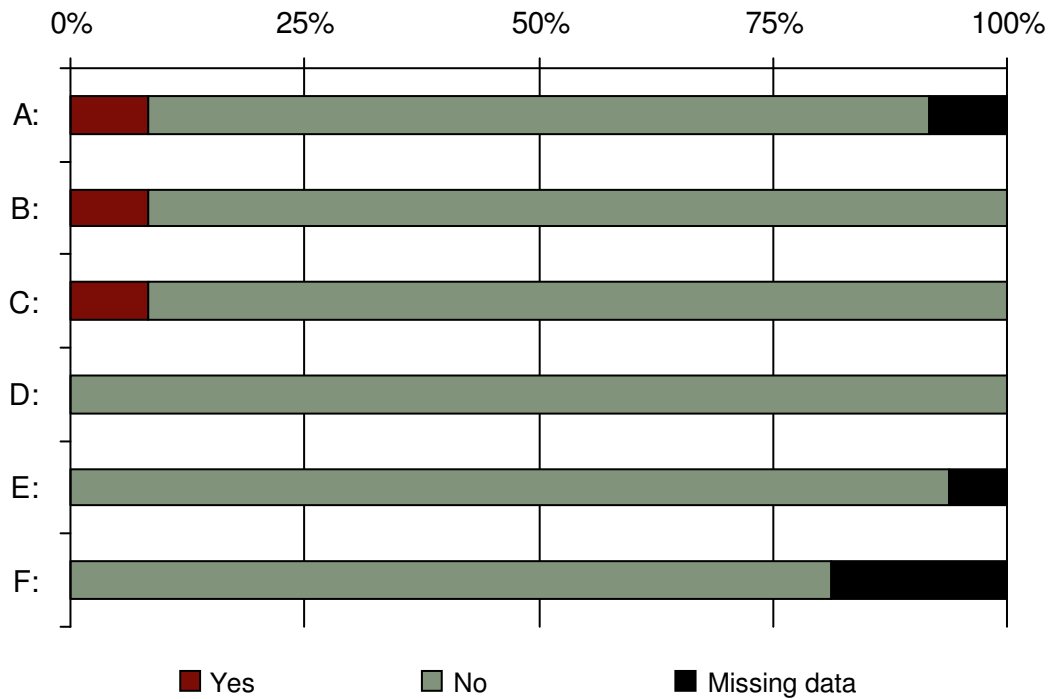
The Documentation from the Ågrenska Center.

### Age distribution



### Overview

	Yes	No	Missing data	N
A: Incomprehensible speech/No speech	2	20	2	24
B: Eating and drinking difficulties <sup>1</sup>	2	22	0	24
C: Profuse drooling, on clothes <sup>1</sup>	2	22	0	24
D: Breathing difficulties <sup>1 2</sup>	0	16	0	16
E: Grinding every day <sup>1 2</sup>	0	15	1	16
F: Severe malocclusions <sup>2</sup>	0	13	3	16



Note that the diagram is based upon less than 100 individuals.

1: Compiled using questionnaire

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

## Oral health

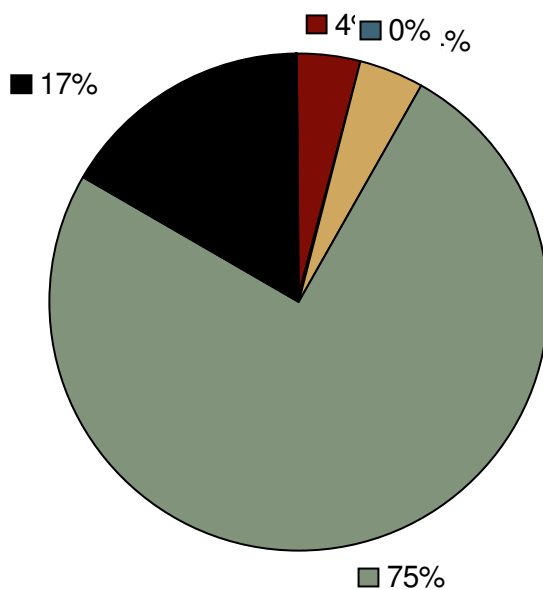
### Oral health index (indices)<sup>1</sup>

		0	1	2	3	4	5	6	Missing data	N
Calc	Calculus	12	3	0	0	0	0	0	1	16
GI	Gingivitis	11	3	0	1	0	0	0	1	16
Plaq	Coating	10	2	2	1	0	0	0	1	16
Toot	Tooth wear	1	8	3	2				2	16

- C 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.
- GI 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.
- PI 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.
- To 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.

<sup>1</sup>: Oral health index (indices) was (were) introduced in the observations in 2008

## Acceptance of dental examination



	Number
None	1
Negative	0
Reluctant	1
Positive	18
Missing data	4
<b>Sum:</b>	<b>24</b>

## Caries

	3-6 years	7-12 years	13-19 years	Adults
<b>deft<sup>1</sup></b>				
Examined		0	4	
Number of individuals with deft=0			4	
Mean			0,0	
Standard deviation			0,0	
Missing data			1	
<b>DMFT<sup>2</sup></b>				
Examined		5	2	9
Number of individuals with DMFT=0		5	1	0
Standard deviation		0,0	1,5	2,2
Mean		0,0	1,5	4,2
Missing data		0	2	2

1: Number of carious or filled deciduous teeth

2: Number of carious or filled permanent teeth

## Occlusal relationship

	<b>Number</b>
Neutral bite	16
Post normal	3
Pre normal	1
Missing data	4
<b>Sum: 24</b>	

## Maximum jaw opening

Children younger  
than 10 years

	<b>Number</b>
- 20	0
21 - 30	0
31 - 40	1
41 - 50	3
51 -	0
Missing data	4
<b>Sum: 8</b>	

Children, 10 years or older, and  
adults

	<b>Number</b>
- 20	0
21 - 30	0
31 - 40	1
41 - 50	5
51 -	9
Missing data	1
<b>Sum: 16</b>	

## Profile<sup>1</sup>

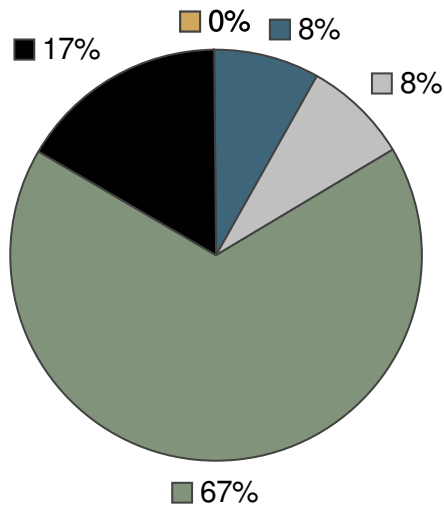
	<b>Number</b>
Normal	11
Convex	0
Concave	0
Missing data	5
<b>Sum: 16</b>	

## Mandibular plane<sup>1</sup>

	<b>Number</b>
Normal	8
Increased	1
Reduced	0
Missing data	7
<b>Sum: 16</b>	

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

### Speech difficulty



	<b>Number</b>
■ No speech	0
■ Very incomprehensible	0
■ Incomprehensible speech	2
■ Slightly indistinct speech	2
■ No problems	16
■ Missing data	4
<b>Sum: 24</b>	

Clinical findings	Yes-answers			
	Total N=24 (%)	Boys/Men N=12 (%)	Girls/Women N=12 (%)	Missing data
Over crowding	4 (20)	2 (20)	2 (20)	4
Frontal open bite	3 (16)	1 (10)	2 (22)	5