



Orofacial function of persons having Hereditary spastic paraplegia

Report from questionnaires

The survey comprises 16 questionnaires.

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

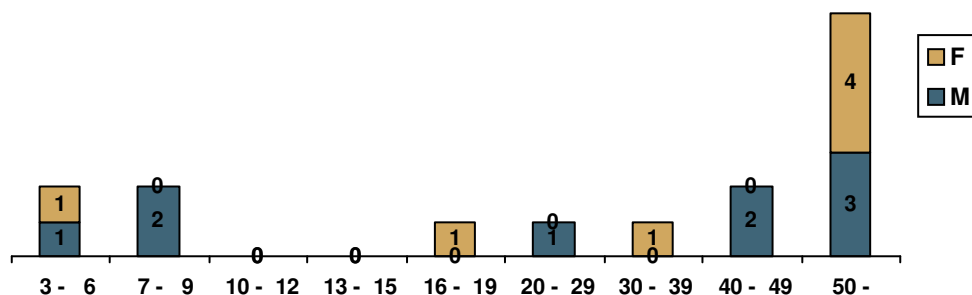
Sources

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



Number: 16

Ages: 6 - 82

Sex: M (9) F (7)

Additional diagnoses

Medical impairment	Yes	No	Missing data
Inborn heart defect	0	16	0
Other cardiovascular disease	0	16	0
Epilepsy	0	16	0
Asthma	0	16	0
Need of respiratory support	0	16	0
Allergy	1	15	0

Neuropsychiatric diagnosis	Yes	No	Missing data
ADHD/ADD	0	15	1
Autistic syndrome	0	15	1
Autistic traits	0	14	2
Asperger's syndrome	0	15	1
Tourette's syndrome	0	15	1

General disability	Yes	No	Missing data
Intellectual disability	0	14	2
Mobility dysfunction	1	2	13
Impaired vision	4	7	5
Impaired hearing	0	13	3
Difficulty in communicating	0	14	2

About dental care and oral health

Do you feel that you receive the dental care you need?

	Number
Yes, very much so	9
Yes, some what	5
No, not really	1
No, not at all	0
Missing data	1
Sum:	16

How many times per year do you normally seek dental care?

	Number
Three or more times per year	0
Twice per year	4
Once per year	7
Less than once per year	5
Missing data	0
Sum:	16

When were your teeth last X-rayed?

	Number
During the past two years	13
More than two years ago	2
Never had my teeth X-rayed	1
Missing data	0
Sum:	16

Do you look after your teeth in a good way?

	Number
Yes, very much so	8
Yes, some what	6
No, not really	1
No, not at all	0
Missing data	1
Sum:	16

Who brushes your teeth?

	Number
I always brush myself	10
Sometimes I brush myself	3
Someone else always helps me	2
Missing data	1
Sum:	16

How often are your teeth brushed?

	Number
Three or more times per day	1
Twice per day	12
Once per day	2
Not every day	0
Missing data	1
Sum:	16

About dental care and oral health

	Yes	No	Missing data
Does your mouth hurt?	0	16	0
Does your mouth feel dry?	2	14	0
Have you ever taken a serious hit to your permanent front teeth?	5	10	1
Do you feel that you have a divergent bite?	0	15	1
Have you had a brace?	2	14	0
Do you feel that you need orthodontics/a brace?	2	13	1

Do you grind or press your teeth at night?

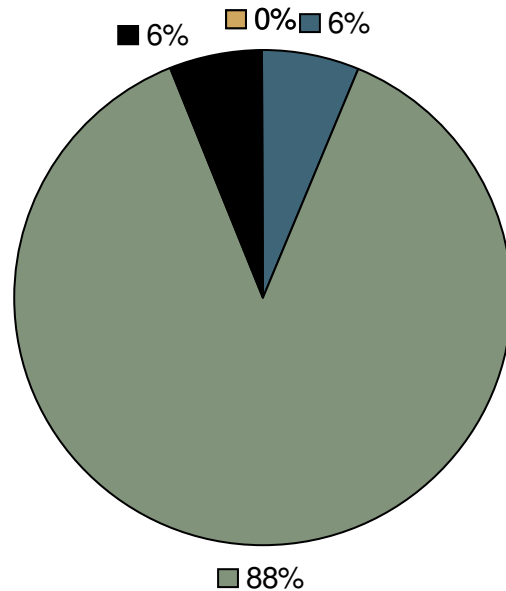
	Number
Never	13
Once or twice per week	1
Every night	2
Missing data	0
Sum:	16

Do you grind or press your teeth during the day?

	Number
Never	14
Once or twice per week	1
Every day	1
Missing data	0
Sum:	16

About eating

Do you have any problems with eating?



	Number
Yes, very much so	0
Yes, some what	0
No, not really	1
No, not at all	14
Missing data	1
Sum:	16

	Yes	No	Missing data
Do you cough daily in connection with meals?	0	15	1
Do you gag daily in connection with meals?	0	15	1
Do you get acid reflux daily?	1	14	1
Do you throw up often (at least twice per week)?	0	15	1
Do you have a poor appetite?	0	15	1
Does it take a long time before you can swallow a mouthful?	0	15	1
Do you press your tongue forward when you swallow so that food ends up outside the mouth?	0	15	1
Do you find it difficult to chew, i.e. grind food using your molars?	0	16	0
Do you find it difficult to take food from the spoon using your lips?	0	16	0
Have you had problems with food and drink leaking out through the corners of your mouth?	0	16	0
Does food tend to remain in your mouth after meals?	0	16	0
Do you get nutrition in any other way than through your mouth?	0	15	1



About drooling

Do you drool?

	Number
Never drool	14
Drool sometimes – not every day	1
Drool often – every day	0
Constant drooling	0
Missing data	1
Sum:	16

How much do you drool?

	Number
Slight drooling, only on the lips	1
Moderate drooling, on lip and chin	0
Profuse drooling, on clothes	0
Very profuse drooling, on hands/objects	0
Missing data	0
Sum:	1

Is your drooling a problem for you?

	Number
Yes, very much so	0
Yes, some what	0
No, not really	0
No, not at all	1
Missing data	0
Sum:	1

Is your drooling a problem for your family or people around you?

	Number
Yes, very much so	0
Yes, some what	0
No, not really	0
No, not at all	1
Missing data	0
Sum:	1