



Orofacial function of persons having Huntington disease

Report from questionnaires

The survey comprises 15 questionnaires.

Synonym: Huntington chorea, Juvenile Huntington disease

ICD 10: G10

Estimated incidence: 6-12:100 000 inhabitants

Cause: Autosomal dominant inheritance. The disease is caused by a mutation in the gene on the short arm of chromosome 4. The gene controls the formation of a protein, huntingtin.

General symptoms: Huntington disease is a progressive neurological and neuropsychiatric disorder that entails motor function, cognitive and psychiatric symptoms. The first sign of the disease usually become noticeable between 30 and 50 years of age. An unusual juvenile form occurs which appears before the age of 20. The disease is characterised by difficulties with balance and random, jerky movements of arms and legs that increase in time. Complications from the respiratory organs are common.

Orofacial/odontological symptoms: Neurological symptoms often start with involuntary, scarcely noticeable movements of fingers, toes and tongue. There is a risk of bite damage to tongue and cheeks. Increased grimacing may occur. However, the facial expression is flattened as control of the muscles deteriorates. Oral motor difficulties affect the ability to clean the mouth. The impact on the oral motor function also affects the ability to speak, chew and swallow.

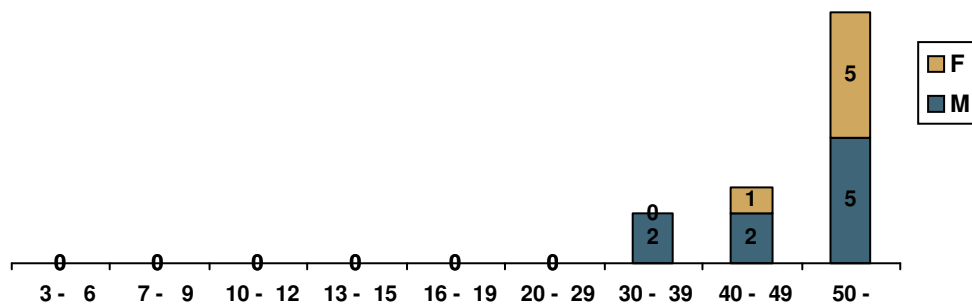
Orofacial/odontological treatment:

- Continuous intensified preventive oral care is very important
- People with Huntington disease may need reminding about times for treatment
- A soft plastic splint that protects against bite damage may be needed
- In the long term relatives and personal assistants need instruction on how to help with oral hygiene
- Dental treatment may be facilitated if the patient is helped to sit/lie in the dentist's chair as relaxed as possible – for example on Tumble pillows
- Avoid large amounts of water if there is a risk of aspiration.

Source

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.
Huntington disease – a practical guide on CD.

Age distribution



Number: 15

Ages: 31 - 83

Sex: M (9) F (6)

Additional diagnoses

Medical impairment	Yes	No	Missing data
Inborn heart defect	0	15	0
Other cardiovascular disease	1	14	0
Epilepsy	2	13	0
Asthma	1	14	0
Need of respiratory support	0	14	1
Allergy	5	10	0

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

General disability	Yes	No	Missing data
Intellectual disability	1	14	0
Mobility dysfunction	3	2	10
Impaired vision	7	4	4
Impaired hearing	2	12	1
Difficulty in communicating	3	3	9

About dental care and oral health

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

	Number
Yes, very much so	11
Yes, some what	3
No, not really	0
No, not at all	1
Missing data	0
Sum:	15

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

	Number
Three or more times per year	9
Twice per year	2
Once per year	2
Less than once per year	1
Missing data	1
Sum:	15

When were your teeth last X-rayed?

	Number
During the past two years	9
More than two years ago	3
Never had my teeth X-rayed	0
Missing data	3
Sum:	15

Do you look after your teeth in a good way?

	Number
Yes, very much so	5
Yes, some what	8
No, not really	1
No, not at all	1
Missing data	0
Sum:	15

Who brushes your teeth?

	Number
I always brush myself	4
Sometimes I brush myself	1
Someone else always helps me	10
Missing data	0
Sum:	15

How often are your teeth brushed?

	Number
Three or more times per day	2
Twice per day	6
Once per day	5
Not every day	2
Missing data	0
Sum:	15

About dental care and oral health

	Yes	No	Missing data
Does your mouth hurt?	3	10	2
Does your mouth feel dry?	10	4	1
Have you ever taken a serious hit to your permanent front teeth?	5	9	1
Do you feel that you have a divergent bite?	2	12	1
Have you had a brace?	2	12	1
Do you feel that you need orthodontics/a brace?	0	15	0

Do you grind or press your teeth at night?

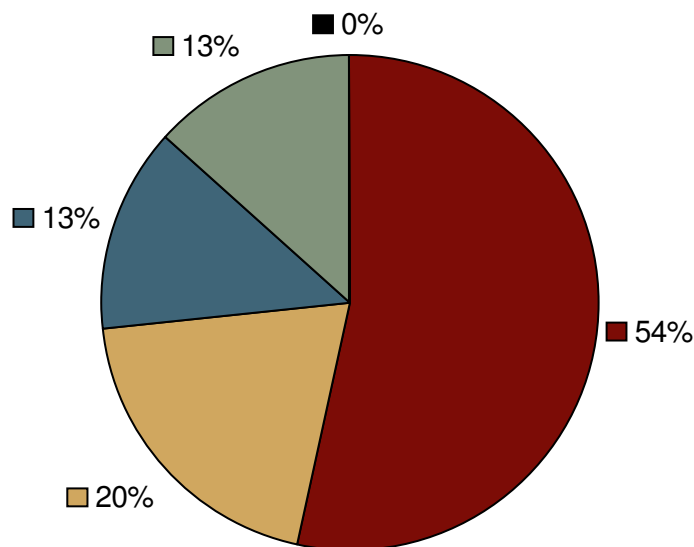
	Number
Never	10
Once or twice per week	2
Every night	2
Missing data	1
Sum:	15

Do you grind or press your teeth during the day?

	Number
Never	11
Once or twice per week	4
Every day	0
Missing data	0
Sum:	15

About eating

Do you have any problems with eating?



	Number
Yes, very much so	8
Yes, some what	3
No, not really	2
No, not at all	2
Missing data	0
Sum:	15

	Yes	No	Missing data
Do you cough daily in connection with meals?	9	5	1
Do you gag daily in connection with meals?	5	9	1
Do you get acid reflux daily?	3	11	1
Do you throw up often (at least twice per week)?	2	12	1
Do you have a poor appetite?	0	14	1
Does it take a long time before you can swallow a mouthful?	6	8	1
Do you press your tongue forward when you swallow so that food ends up outside the mouth?	3	10	2
Do you find it difficult to chew, i.e. grind food using your molars?	10	3	2
Do you find it difficult to take food from the spoon using your lips?	3	10	2
Have you had problems with food and drink leaking out through the corners of your mouth?	6	7	2
Does food tend to remain in your mouth after meals?	7	6	2
Do you get nutrition in any other way than through your mouth?	5	10	0

About drooling

Do you drool?

	Number
Never drool	6
Drool sometimes – not every day	6
Drool often – every day	3
Constant drooling	0
Missing data	0
Sum:	15

How much do you drool?

	Number
Slight drooling, only on the lips	4
Moderate drooling, on lip and chin	2
Profuse drooling, on clothes	1
Very profuse drooling, on hands/objects	1
Missing data	1
Sum:	9

Is your drooling a problem for you?

	Number
Yes, very much so	2
Yes, some what	0
No, not really	3
No, not at all	2
Missing data	2
Sum:	9

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

	Number
Yes, very much so	0
Yes, some what	2
No, not really	3
No, not at all	2
Missing data	2
Sum:	9