



# Orofacial function of persons having Prader-Willi syndrome

## Report from questionnaires

The survey comprises 27 questionnaires.

**Estimated occurrence:** 6-8:100 000 live births.

**Etiology:** Chromosomal aberration on chromosome 15. The gene or genes that cause the syndrome have not yet been identified.

**General symptoms:** There are great variations between different individuals and different ages. Pronouncedly poor muscle tone/hypotonicity from birth. After the age of one or two, distinct eating disorder occur, with an insatiable appetite despite a low need of energy (approximately 50-60% of what is considered normal). Without a special diet, this results in extreme obesity and an increased risk of type II diabetes. Most of these individuals are mildly to moderately intellectually impaired and delayed motor development is common. There is an increased risk of behavior problems. Individuals with PWS often have short stature and underdeveloped sexual organs (hypogonadism).

**Orofacial/odontological symptoms:** Characteristic facial features are associated with the diagnosis. Muscle laxity in newborns leads to most PWS infants being unable to be breast-fed. Many require a special bottle or tube feeding. Delayed speech and language development and problems with oral motor skills are common. Many suffer from dry mouth. Narrow nasal and throat airways may result in snoring and an increased risk of sleep apnea (frequent suspension of breathing while asleep). Delayed teething, defective tooth enamel, an excessive number of teeth and small teeth have all been reported. A high, arched palate and malocclusion are common. Some individuals have tooth grinding. There is an increased risk of caries owing to the eating disorder and dry mouth.

### **Orofacial/ odontological treatment:**

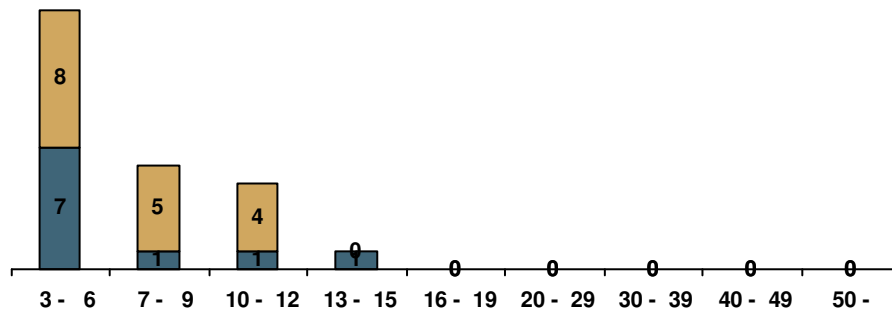
- Early contact with dental services for intensified prophylactic care and oral hygiene information is essential.
- Regular check-ups of dental and jaw development. Orthodontist should be consulted when needed.
- Tooth grinding should be followed up, and be managed with a splint when necessary.
- Training in oral motor skills may be relevant.
- Speech, language and communication training are often justified.
- Snoring problems should be followed up by a physician.

### **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: 27

Ages: 3 - 14

Sex: M (10) F (17)

## Additional diagnoses

Medical impairment	Yes	No	Missing data
Inborn heart defect	1	25	1
Other cardiovascular disease	0	26	1
Epilepsy	0	26	1
Asthma	4	22	1
Need of respiratory support	0	26	1
Allergy	6	20	1

Neuropsychiatric diagnosis	Yes	No	Missing data
ADHD/ADD	0	25	2
Autistic syndrome	0	23	4
Autistic traits	8	13	6
Asperger's syndrome	0	23	4
Tourette's syndrome	0	24	3

General disability	Yes	No	Missing data
Intellectual disability	10	7	10
Mobility dysfunction	16	2	9
Impaired vision	12	11	4
Impaired hearing	3	22	2
Difficulty in communicating	12	4	11

## About dental care and oral health

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

	Number
Yes, very much so	22
Yes, some what	5
No, not really	0
No, not at all	0
Missing data	0
<b>Sum:</b>	<b>27</b>

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

	Number
Three or more times per year	5
Twice per year	12
Once per year	5
Less than once per year	2
Missing data	3
<b>Sum:</b>	<b>27</b>

### When were your teeth last X-rayed?

	Number
During the past two years	12
More than two years ago	2
Never had my teeth X-rayed	13
Missing data	0
<b>Sum:</b>	<b>27</b>

## Do you look after your teeth in a good way?

	Number
Yes, very much so	17
Yes, some what	10
No, not really	0
No, not at all	0
Missing data	0
<b>Sum:</b>	<b>27</b>

## Who brushes your teeth?

	Number
I always brush myself	1
Sometimes I brush myself	10
Someone else always helps me	16
Missing data	0
<b>Sum:</b>	<b>27</b>

## How often are your teeth brushed?

	Number
Three or more times per day	1
Twice per day	24
Once per day	2
Not every day	0
Missing data	0
<b>Sum:</b>	<b>27</b>

## About dental care and oral health

	Yes	No	Missing data
Does your mouth hurt?	3	22	2
Does your mouth feel dry?	9	13	5
Have you ever taken a serious hit to your permanent front teeth?	2	24	1
Do you feel that you have a divergent bite?	3	22	2
Have you had a brace?	0	27	0
Do you feel that you need orthodontics/a brace?	3	20	4

## Do you grind or press your teeth at night?

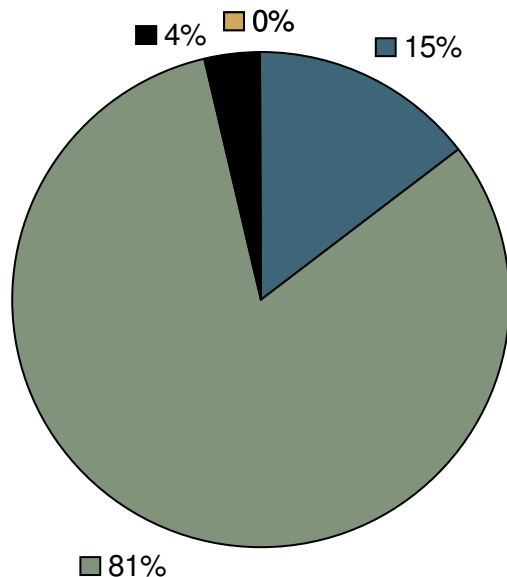
	Number
Never	18
Once or twice per week	6
Every night	1
Missing data	2
<b>Sum:</b>	<b>27</b>

## Do you grind or press your teeth during the day?

	Number
Never	20
Once or twice per week	5
Every day	1
Missing data	1
<b>Sum:</b>	<b>27</b>

## About eating

### Do you have any problems with eating?



	Number
Yes, very much so	0
Yes, some what	0
No, not really	4
No, not at all	22
Missing data	1
<b>Sum:</b>	<b>27</b>

	Yes	No	Missing data
Do you cough daily in connection with meals?	1	25	1
Do you gag daily in connection with meals?	0	23	4
Do you get acid reflux daily?	0	26	1
Do you throw up often (at least twice per week)?	0	26	1
Do you have a poor appetite?	0	26	1
Does it take a long time before you can swallow a mouthful?	1	25	1
Do you press your tongue forward when you swallow so that food ends up outside the mouth?	1	23	3
Do you find it difficult to chew, i.e. grind food using your molars?	2	24	1
Do you find it difficult to take food from the spoon using your lips?	0	25	2
Have you had problems with food and drink leaking out through the corners of your mouth?	1	25	1
Does food tend to remain in your mouth after meals?	3	21	3
Do you get nutrition in any other way than through your mouth?	0	26	1



## About drooling

### Do you drool?

	Number
Never drool	23
Drool sometimes – not every day	2
Drool often – every day	0
Constant drooling	0
Missing data	2
<b>Sum:</b>	<b>27</b>

### How much do you drool?

	Number
Slight drooling, only on the lips	1
Moderate drooling, on lip and chin	1
Profuse drooling, on clothes	0
Very profuse drooling, on hands/objects	0
Missing data	0
<b>Sum:</b>	<b>2</b>

### Is your drooling a problem for you?

	Number
Yes, very much so	0
Yes, some what	1
No, not really	0
No, not at all	1
Missing data	0
<b>Sum:</b>	<b>2</b>

### 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	1
No, not at all	1
Missing data	0
<b>Sum:</b>	<b>2</b>