



Orofacial function of persons having Spielmeyer-Vogt disease

Report from questionnaires



18 questionnaires

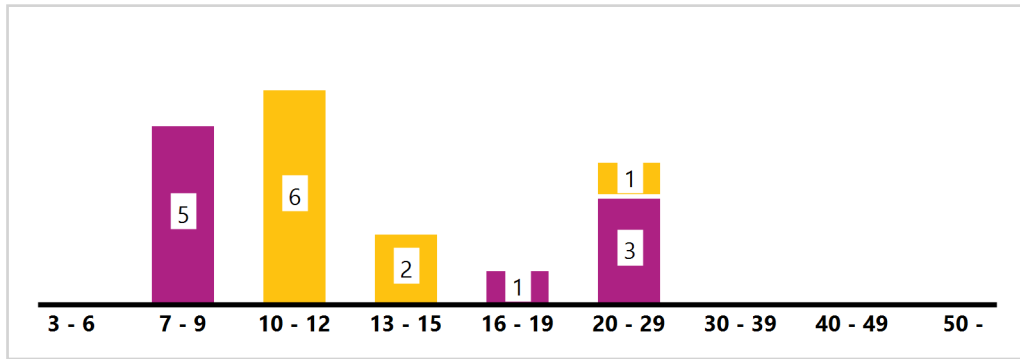
Synonym	JNCL, Spielmeyer-Vogt disease, Juvenile Batten disease, CLN3
ICD-10	E75.4
Estimated occurrence	5:1,000,000 inhabitants
General symptoms	As time goes by, the disease affects vision, motor skills, speech, language and cognition. Vision is impaired before school age, and by their early teens, most sufferers can't see. Motor skills change, and movement patterns become more stiff, with poor balance and bent-knee walking. Many develop epilepsy and neuropsychiatric symptoms such as compulsiveness and concentration difficulties. Aggression, fear, nightmares, anxiety, depression and sleep disorders may also occur.
Oral symptoms	Reduced oral motor skills are part of the disease progression, which leads to difficulties eating and speaking. Speech is often fast, stuttering and inarticulate. Alternative communication may become appropriate.
Oral treatment	<ul style="list-style-type: none"> • It is important that individuals with Spielmeyer-Vogts disease come in early contact with the dental services for extra preventive dental care and information about oral hygiene. • • Feeding and swallowing difficulties are investigated and treated by a specialist team at the hospital or multidisciplinary treatment centre. • Speech, language and communication training are often required. • Training in oral motor skills and extra stimulation in cases of eating disorders, speech difficulties and impaired saliva control may be necessary.
Sources	The MHC database Rare diseases Dokumentation-Ågrenska

Age distribution

Number: 18

Ages: 8 - 28

♂ (9)
♀ (9)



Additional diagnoses

Medical impairment	Yes	No	Missing data
Inborn heart defect	0	18	0
Other cardiovascular disease	0	18	0
Epilepsy	13	5	0
Asthma	0	17	1
Need of respiratory support	0	18	0
Allergy	1	17	0

Neuropsychiatric diagnosis	Yes	No	Missing data
ADHD/ADD	0	18	0
Autism (Includes autism, Asperger syndrome and autistic traits)	4	10	4

General disability	Yes	No	Missing data
Intellectual disability	4	6	8
Motoric functional impairment	8	2	8
Visual impairment	2	0	16
Hearing loss	0	17	1
Communication difficulties	4	3	11

About dental care and oral health

Do you feel that you receive the dental care you need?	Number
Yes, very much so	11
Yes, somewhat	4
No, not really	2
Missing data	1
Total:	18

How many times per year do you normally seek dental care?	Number
Less than once per year	4
One time per year	6
Two times per year	2
Three or more times per year	6
Total:	18

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	2
Missing data	1
Total:	18

Do you look after your teeth in a good way?	Number
Yes, very much so	9
Yes, somewhat	8
No, not at all	1
Total:	18

Who brushes your teeth?	Number
I always brush myself	3
Someone else always helps me	11
Sometimes I brush myself	4
Total:	18

How often are your teeth brushed?	Number
Once per day	3
Two times per day	15
Total:	18

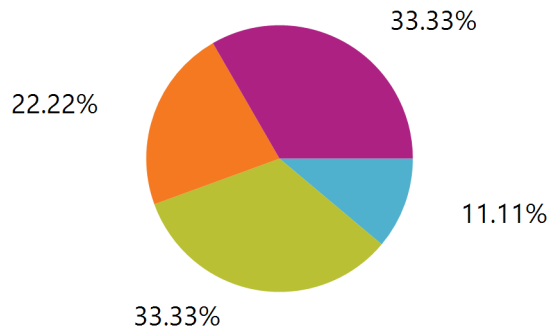
About dental care and oral health

	Yes	No	Missing data
Does your mouth hurt?	2	15	1
Does your mouth feel dry?	2	14	2
Have you ever taken a serious hit to your permanent front teeth?	5	13	0
Do you feel that you have a divergent bite?	2	15	1
Have you had a brace?	0	18	0
Do you feel that you need orthodontics/a brace?	2	13	3

Do you grind or press your teeth at night?	Number
Never	14
Once or twice per week	2
Every night	1
Missing data	1
Total:	18

Do you grind or press your teeth during the day?	Number
Never	15
Once or twice per week	1
Every night	1
Missing data	1
Total:	18

About eating



Do you have any problems with eating?

Do you have any problems with eating?	Number
Yes, very much so	2
Yes, somewhat	6
No, not really	4
No, not at all	6
Missing data	0
Total:	18

	Yes	No	Missing data
Do you cough daily in connection with meals?	2	16	0
Do you gag daily in connection with meals?	0	18	0
Do you get acid reflux daily?	0	17	1
Do you throw up often (at least twice per week)?	0	17	1
Do you have a poor appetite?	0	18	0
Does it take a long time before you can swallow a mouthful?	1	9	8
Do you press your tongue forward when you swallow so that food ends up outside the mouth?	1	9	8
Do you find it difficult to chew, i.e. grind food using your molars?	6	12	0
Do you find it difficult to take food from the spoon using your lips?	3	14	1
Have you had problems with food and drink leaking out through the corners of your mouth?	2	8	8
Does food tend to remain in your mouth after meals?	6	12	0
Do you get nutrition in any other way than through your mouth?	2	16	0

About drooling

Do you drool?	Number
Never drool	9
Drool sometimes – not every day	4
Drool often – every day	5
Missing data	0
Total:	18

How much do you drool?	Number
Moderate drooling, on lip and chin	4
Profuse drooling	5
Total:	9

Is your drooling a problem for you?	Number
Yes, somewhat	6
No, not really	3
Total:	9

Is your drooling a problem for your family or people around you?	Number
Yes, somewhat	5
No, not at all	4
Total:	9