



Orofacial function of persons having Vacterl syndrome

Report from observation charts

The survey comprises 19 observation charts.

Synonyme: VATER

Estimated occurrence: 10-20:100 000 live births.

Etiology: Unknown. The malformations are thought to be caused by an interruption of the cell formation during early foetal life.

General symptoms: Low birth weight and premature birth are common. The term VACTERL is an acronym with each letter representing the first letter of the organ in which the anomaly may occur.

V	Vertebral (vertebrae, bones of the spinal column)
A	Anorectal (anus /rectum)
C	Cardiovascular (heart)
T	Tracheal (windpipe)
E	Esophageal (tube connecting the throat to the stomach))
R	Renal (kidney)
L	Limb (arms/ legs)

The diagnosis is made if anomalies occur in at least three of the above groups. Other anomalies may also occur, for example hydrocephalus and choanal atresia.

Orofacial/odontological symptoms: Feeding difficulties are common in children with breathing difficulties, heart defects and esophageal atresia. Some children therefore need to be tube fed during infancy or later. Gastroesophageal reflux (leakage of stomach fluids into the esophagus) and vomiting are relatively common and contribute to eating difficulties and increase the risk of caries.

Orofacial/odontological treatment:

- Children with eating difficulties are often in need of supplementary dental care e.g. help with oral hygiene and fluoride treatments
- Feeding and swallowing difficulties are investigated and treated by a specialist team at the hospital or multidisciplinary treatment centre.
- When treating medically compromised patients always contact their doctors for medical advice.

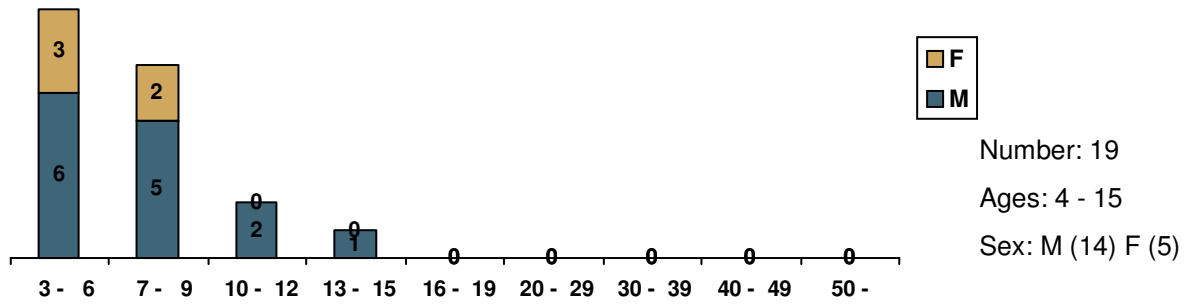
Sources

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.

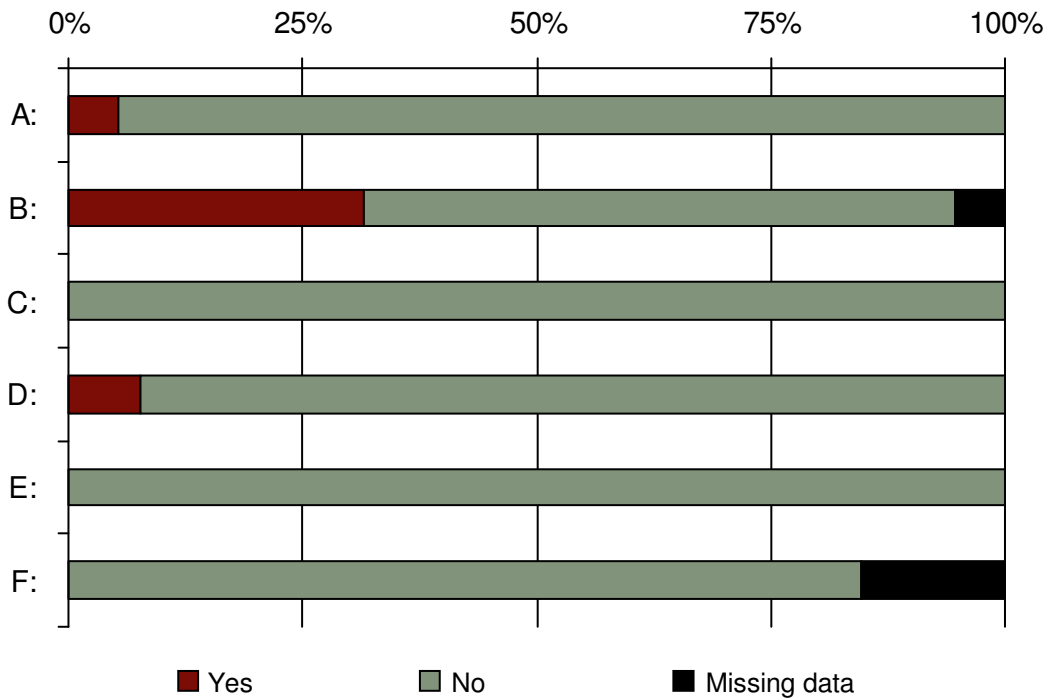
The Documentation from the Ågrenska Center.

Age distribution



Overview

	Yes	No	Missing data	N
A: Incomprehensible speech/No speech	1	18	0	19
B: Eating and drinking difficulties ¹	6	12	1	19
C: Profuse drooling, on clothes ¹	0	19	0	19
D: Breathing difficulties ^{1 2}	1	12	0	13
E: Grinding every day ^{1 2}	0	13	0	13
F: Severe malocclusions ²	0	11	2	13



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

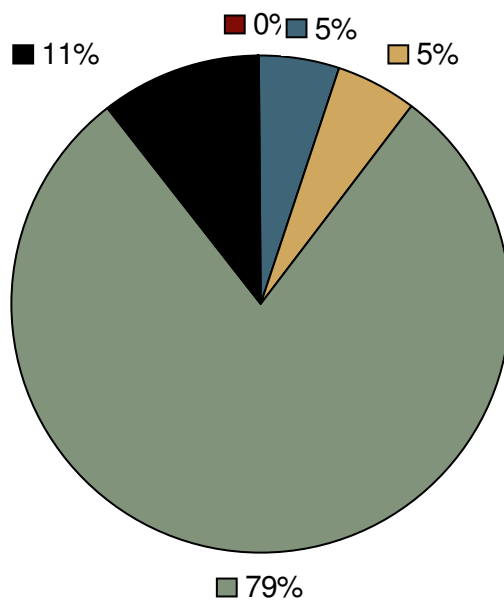
Oral health index (indices)¹

		0	1	2	3	4	5	6	Missing data	N
Calc	Calculus	10	1	0	0	0	1	1	0	13
GI	Gingivitis	11	0	0	1	1	0	0	0	13
Plaq	Coating	8	3	0	0	0	0	2	0	13
Toot	Tooth wear	12	0	0	0				1	13

- 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.

¹: Oral health index (indices) was (were) introduced in the observations in 2008

Acceptance of dental examination



	Number
None	0
Negative	1
Reluctant	1
Positive	15
Missing data	2
Sum:	19

Caries

	3-6 years	7-12 years	13-19 years	Adults
deft¹				
Examined	4	7		
Number of individuals with deft=0	4	5		
Mean	0,0	1,4		
Standard deviation	0,0	2,3		
Missing data	5	2		
DMFT²				
Examined		7	1	0
Number of individuals with DMFT=0		5	1	
Standard deviation		1,2	0,0	
Mean		0,7	0,0	
Missing data		2	0	

1: Number of carious or filled deciduous teeth

2: Number of carious or filled permanent teeth

Occlusal relationship

	Number
Neutral bite	15
Post normal	1
Pre normal	0
Missing data	3
Sum: 19	

Maximum jaw opening

Children younger than 10 years

	Number
- 20	0
21 - 30	0
31 - 40	7
41 - 50	6
51 -	0
Missing data	3
Sum: 16	

Children, 10 years or older, and adults

	Number
- 20	0
21 - 30	0
31 - 40	1
41 - 50	2
51 -	0
Missing data	0
Sum: 3	

Profile¹

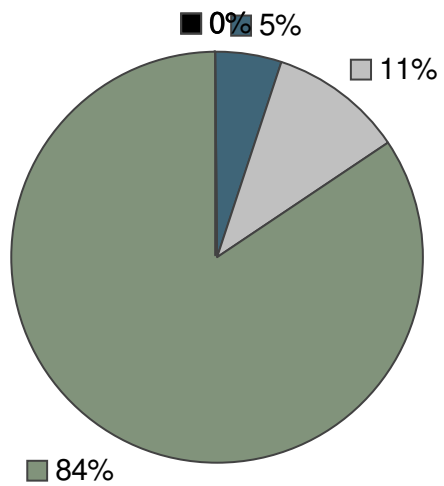
	Number
Normal	10
Convex	0
Concave	0
Missing data	3
Sum: 13	

Mandibular plane¹

	Number
Normal	11
Increased	0
Reduced	0
Missing data	2
Sum: 13	

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

Speech difficulty



	Number
No speech	0
Very incomprehensible	0
Incomprehensible speech	1
Slightly indistinct speech	2
No problems	16
Missing data	0
Sum: 19	

Clinical findings	Yes-answers			
	Total N=19 (%)	Boys/Men N=14 (%)	Girls/Women N=5 (%)	Missing data
High palate	5 (28)	5 (36)	0 ()	1
Over crowding	4 (21)	2 (14)	2 (40)	0
Frontal open bite	3 (16)	3 (21)	0 ()	0