



Orofacial function of persons having Primary ciliary dyskinesia

Report from observation charts

The survey comprises 13 observation charts.

Synonyms: Immotile Cilia Syndrome, Kartagener Syndrome

Estimated occurrence: 4-6: 100 000 live births.

Etiology: The disorder is caused by mutations identified in genes on chromosomes 5, 9 and 17. The mutations cause a congenital defect in the function of the cilia. The disorder is inherited autosomal recessively. New mutations may possibly have another inheritance pattern.

General symptoms: Cilia are found in several bodily organs, e.g., respiratory tract (from the nose and sinuses to the trachea), in the sperm tail and in olfactory cells. Mucus clearance from the lungs may be impaired already at birth. Ear and eustachian (auditory) tube infections as well as wet cough that become chronic are common during the first years. Recurrent lung infections lead to impaired lung function and possibly poor oxygenation. Sinus infections, poor sense of smell and eye anomalies are common. Fertility is often reduced. Half of those with Kartagener syndrome have major organs on the opposite side to normal. In these cases the diagnosis is often discovered early.

Orofacial/odontological symptoms: Persons with Primary Ciliary Dyskinesia do not have more orofacial or odontological problems than do people in general.

Orofacial/odontological treatment:

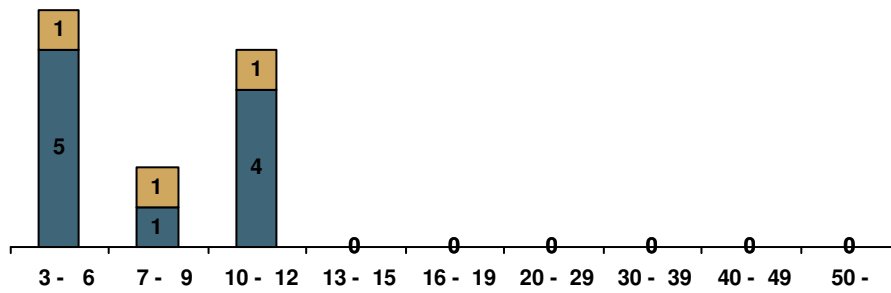
- As infections may be prolonged and severe, especially if lung function is affected, dental care should be clearly prevention oriented.
- Early contact should be made with dental services for assessment and treatment planning.
- Antibiotic therapy should be considered in oral infections.
- Dental visits should be planned taking into account the time of day that the patient feels best given medications and symptoms.

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.



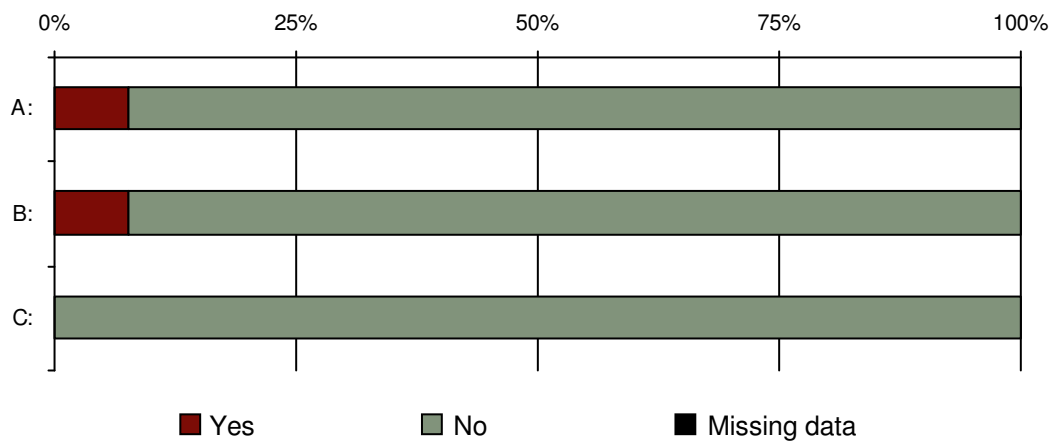
Age distribution



Number: 13
 Ages: 4 - 11
 Sex: M (10) F (3)

Overview

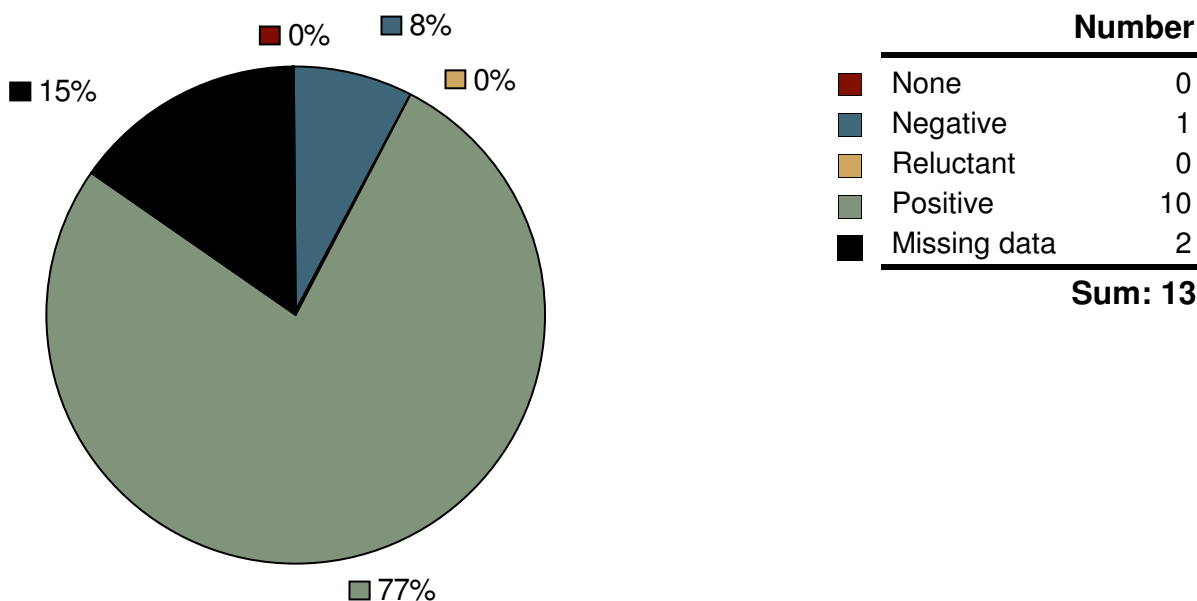
	Yes	No	Missing data	N
A: Incomprehensible speech/No speech	1	12	0	13
B: Eating and drinking difficulties ¹	1	12	0	13
C: Profuse drooling, on clothes ¹	0	13	0	13



Note that the diagram is based upon less than 100 individuals.

¹: Compiled using questionnaire

Acceptance of dental examination



Caries

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

1: Number of carious or filled deciduous teeth
 2: Number of carious or filled permanent teeth

Occlusal relationship

	Number
Neutral bite	10
Post normal	1
Pre normal	1
Missing data	1
Sum: 13	

Maximum jaw opening

Children younger
than 10 years

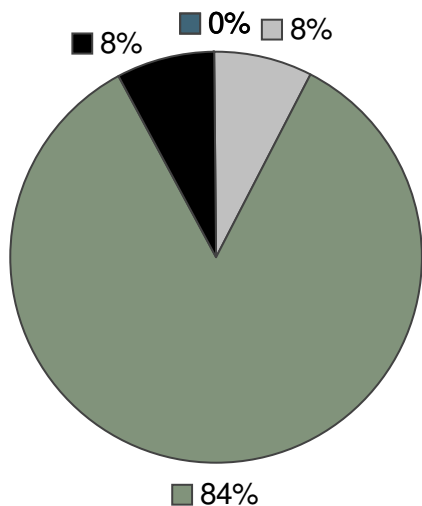
	Number
- 20	0
21 - 30	0
31 - 40	0
41 - 50	1
51 -	0
Missing data	7
Sum: 8	

Children, 10 years or
older, and adults

	Number
- 20	0
21 - 30	0
31 - 40	0
41 - 50	1
51 -	0
Missing data	4
Sum: 5	

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

Speech difficulty



	Number
No speech	0
Very incomprehensible	0
Incomprehensible speech	0
Slightly indistinct speech	1
No problems	11
Missing data	1
Sum: 13	

Clinical findings

	Yes-answers			
	Total N=13 (%)	Boys/Men N=10 (%)	Girls/Women N=3 (%)	Missing data
Open mouth at rest	7 (54)	5 (50)	2 (67)	0