



Orofacial function of persons having Primary immunodeficiency

Report from observation charts



15 observation charts

Synonym

ICD-10

Estimated occurrence 12:100,000 live births.

General symptoms

There are over 200 known immunodeficiencies in Sweden, some of which are Common Variable Immunodeficiency (CVID), Hyper IgM syndrome, Severe Combined Immunodeficiency (SCID), Chronic Granulomatous Disease (CGD), Severe Congenital Neutropenia (Kostmann syndrome) and Hereditary Angiodema (HAE). The most common primary immunodeficiency diseases cause frequent and difficult to treat bacterial infections. Warning signs of typical symptoms for children and adults have been developed. Warning signs in children can be more than six inflammations of the ear per year, recurring pneumonia, more than two bouts of sinusitis or inflammations that do not heal as expected during antibiotic treatment. For adults, it is four or more respiratory tract infections needing antibiotic treatment over a three year period, or other recurring, difficult to treat bacterial infections. The severity of the primary immunodeficiencies varies greatly. Many need gamma globulin as a substitute for the deficiency caused by the disease. The most severe of the primary immunodeficiencies are life threatening, and the children have to have a bone marrow transplant early on.

Oral symptoms

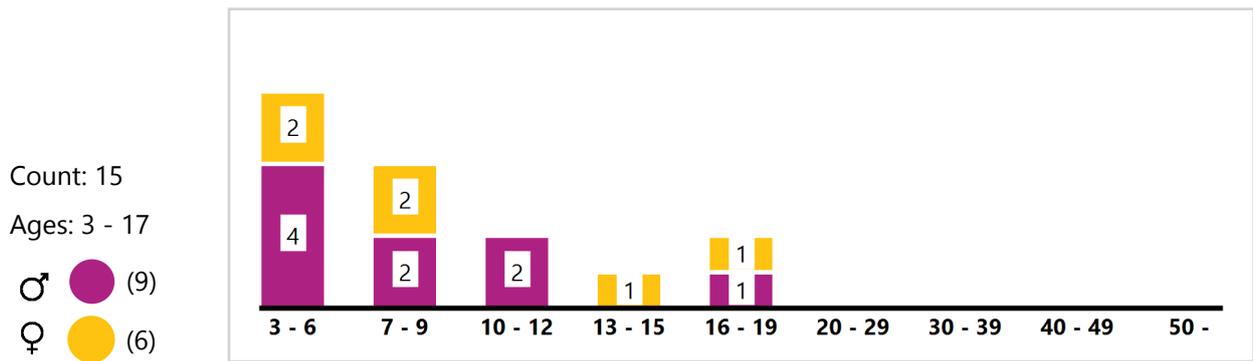
Long-lasting, pronounced fungal infections of the oral cavity or in the skin are possible symptoms. With Kostmann syndrome, the child experiences painful sores in the oral cavity early on, and after a tooth appears, there is often a chronic gum inflammation (gingivitis), and sometimes early tooth loss.

Oral treatment

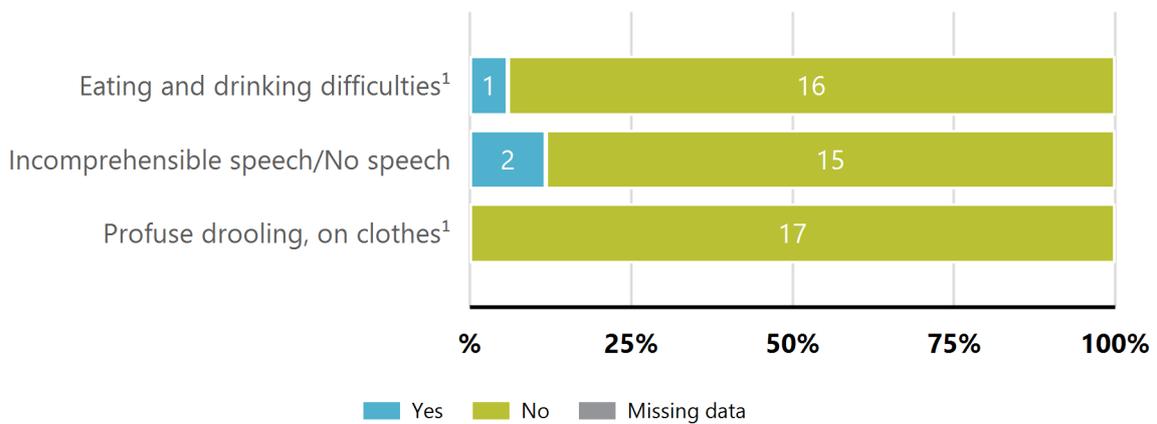
- It is important that individuals with primary immunodeficiency come in early contact with the dental services for extra preventive dental care and information about oral hygiene. Frequent infections, poor nutrition, and poorly mineralized enamel all increase the risk of caries.

Sources The MHC database Rare diseases Dokumentation-Ågrenska

Age distribution



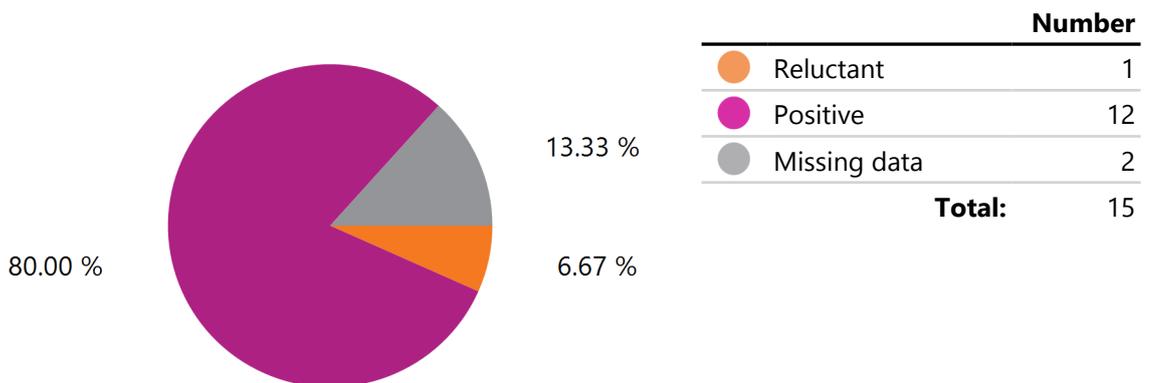
Summary



Certain caution should be observed when interpreting the chart because the number of individuals is less than 100.

¹ Reported via questionnaire

Acceptance of dental examination



Oral health

Caries

		3-6 years	7-12 years	13-19 years
deft	Examined	6	6	
Number of carious or filled deciduous teeth	Number of individuals with deft=0	6	4	
	Mean	0.0	2.3	
	Standard deviation	0.0	2.9	
	No data	0	2	
DMFT	Examined		6	3
Number of carious or filled permanent teeth	Number of individuals with DMFT=0		5	3
	Mean		0.2	0.0
	Standard deviation		0.4	0.0
	No data		0	0

Oral health index (indices)¹

	0	1	2	3	4	5	6	Missing data	Number
Calculus	9	0	0	0	0	0	0	6	9
Gingivitis	8	1	0	0	0	0	0	6	9
Plaque	7	1	1	0	0	0	0	6	9
Tooth wear	6	3	0	0	0	0	0	6	9

CALCULUS

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.

GINGIVITIS

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.

PLAQUE

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.

TOOTH WEAR

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

Occlusal relationship

	Number
Neutral bite	14
Overbite	0
Underbite	2
Missing data	-1
Total:	15

Maximum jaw opening

Children younger than 10 years

Children, 10 years or older, and adults

	Number
-20	0
21-30	0
31-40	4
41-50	5
51-	0
Missing data	1
Total:	10

	Number
-20	0
21-30	0
31-40	0
41-50	3
51-	2
Missing data	0
Total:	5

Profile¹

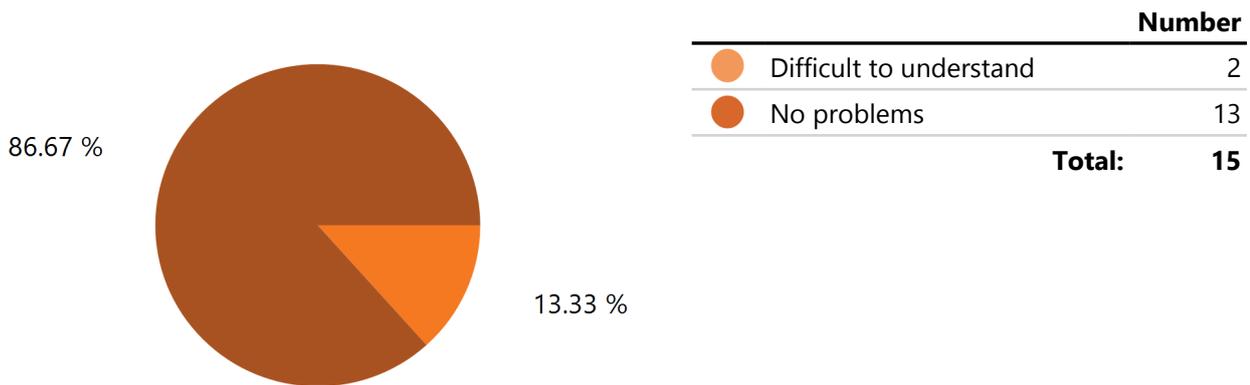
	Number
Normal	8
Convex	1
Concave	0
Missing data	0
Total:	9

Mandibular plane¹

	Number
Normal	9
Increased	0
Reduced	0
Missing data	0
Total:	9

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

Speech problems



Clinical findings

Number of yes-answers

	Total = 15 (%)	♂ = 9 (%)	♀ = 6 (%)	Missing Data
Open mouth at rest	2 (13)	2 (22)	0 (0)	1
High palate	2 (13)	2 (22)	0 (0)	0
Spacing	2 (13)	2 (22)	0 (0)	0
Low muscle tone in masticatory muscles	1 (7)	1 (11)	0 (0)	1
Narrow palate	1 (7)	1 (11)	0 (0)	0
Over crowding	1 (7)	0 (0)	1 (17)	0