



Orofacial function of persons having Primary immunodeficiency

Report from questionnaires



9 questionnaires

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

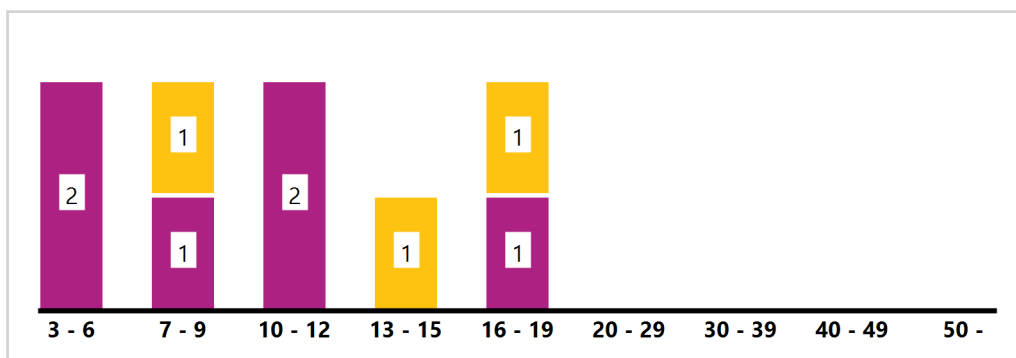
Age distribution

Number: 9

Ages: 6 - 16

♂ (6)

♀ (3)



Additional diagnoses

Medical impairment	Yes	No	Missing data
Inborn heart defect	1	8	0
Other cardiovascular disease	0	9	0
Epilepsy	0	9	0
Asthma	2	7	0
Need of respiratory support	0	9	0
Allergy	3	6	0

Neuropsychiatric diagnosis	Yes	No	Missing data
ADHD/ADD	0	9	0
Autism (Includes autism, Asperger syndrome and autistic traits)	0	9	0

General disability	Yes	No	Missing data
Intellectual disability	0	9	0
Motoric functional impairment	0	8	1
Visual impairment	3	5	1
Hearing loss	0	8	1
Communication difficulties	0	8	1

About dental care and oral health

Do you feel that you receive the dental care you need?	Number
Yes, very much so	8
Yes, somewhat	1
Total:	9

How many times per year do you normally seek dental care?	Number
Less than once per year	1
One time per year	4
Two times per year	2
Three or more times per year	2
Total:	9

When were your teeth last X-rayed?	Number
During the past two years	8
Never had my teeth X-rayed	1
Total:	9

Do you look after your teeth in a good way?	Number
Yes, very much so	7
Yes, somewhat	2
Total:	9

Who brushes your teeth?	Number
I always brush myself	4
Someone else always helps me	1
Sometimes I brush myself	4
Total:	9

How often are your teeth brushed?	Number
Two times per day	9
Total:	9

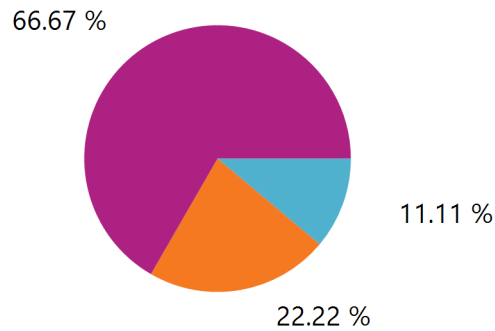
About dental care and oral health

	Yes	No	Missing data
Does your mouth hurt?	3	6	0
Does your mouth feel dry?	2	7	0
Have you ever taken a serious hit to your permanent front teeth?	1	8	0
Do you feel that you have a divergent bite?	1	8	0
Have you had a brace?	1	7	1
Do you feel that you need orthodontics/a brace?	1	8	0

Do you grind or press your teeth at night?	Number
Never	6
Once or twice per week	1
Every night	2
Total:	9

Do you grind or press your teeth during the day?	Number
Never	7
Once or twice per week	1
Every night	1
Total:	9

About eating



Do you have any problems with eating?

Response	Number
Yes, very much so	1
No, not really	2
No, not at all	6
Missing data	0
Total:	9

	Yes	No	Missing data
Do you cough daily in connection with meals?	1	8	0
Do you gag daily in connection with meals?	0	9	0
Do you get acid reflux daily?	2	7	0
Do you throw up often (at least twice per week)?	0	9	0
Do you have a poor appetite?	2	7	0
Does it take a long time before you can swallow a mouthful?	2	7	0
Do you press your tongue forward when you swallow so that food ends up outside the mouth?	0	8	1
Do you find it difficult to chew, i.e. grind food using your molars?	0	8	1
Do you find it difficult to take food from the spoon using your lips?	0	9	0
Have you had problems with food and drink leaking out through the corners of your mouth?	1	8	0
Does food tend to remain in your mouth after meals?	0	9	0
Do you get nutrition in any other way than through your mouth?	0	9	0

About drooling

Do you drool?	Number
Never drool	7
Drool often – every day	1
Missing data	0
Total:	9

How much do you drool?	Number
Moderate drooling, on lip and chin	1
Total:	1

Is your drooling a problem for you?	Number
Yes, somewhat	1
Total:	1

Is your drooling a problem for your family or people around you?	Number
No, not really	1
Total:	1