Orofacial function of persons having
Osteogenesis imperfecta

Report from observation charts | 28 observation charts

Synonym

ICD-10 Q78.0

Estimated occurance 5:100,000 live births.

General symptoms Type I-IV, from minor to severe symptoms. Moderate to severe bone fragility may cause multiple fractures, back pain and scoliosis. Other symptoms that may occur include discoloration of the sclera, hearing impairment, loose joints, instability of the cervical spine, cardiovascular problems, hyperthermia – "sweating", bleeding diathesis, varying degrees of shortness of stature, etc.

Oral symptoms The most common tooth development disorder is Dentinogenesis Imperfecta (DI). DI is characterized by:

- Tooth discoloration (light blue to dark brown with a transparent glaze).
- Dentin is softer than normal.
- Enamel "splinters" from the soft dentin.
- The soft dentin causes the teeth to wear down rapidly, especially the primary teeth.

Aplasia of one or more permanent teeth is common (some permanent teeth are missing). X-rays sometimes reveal elongated pulp chambers. The upper jaw is usually small causing malocclusion with underbite.

Oral treatment

- Early collaboration with specialists in child dentistry and orthodontics.
- Steel crowns on six-year-molars, in combination with long-term temporary dental filling therapy, reduce the risk for extensive tooth wear on the crossbite side.
- Plan early corrective surgical and prosthetic treatment to improve functional and esthetic occlusion.
- Increased risk associated with anesthesia due to instability of the cervical spine.

Sources The MHC database Rare diseases Dokumentation-Ågrenska

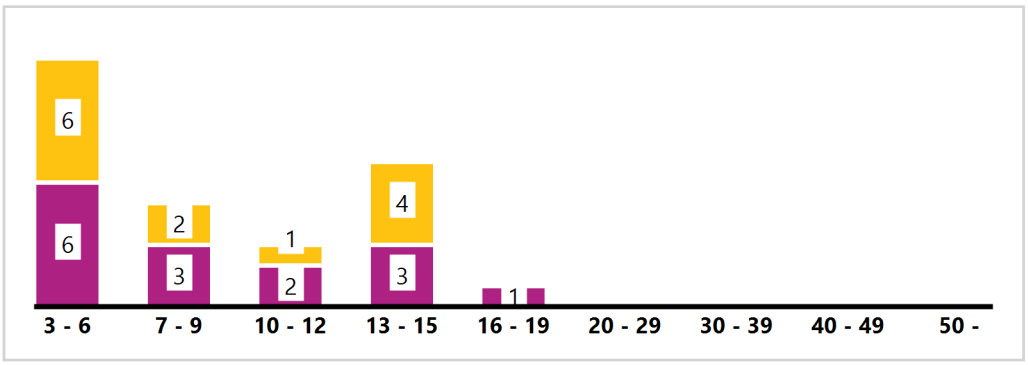
Age distribution

Count: 28

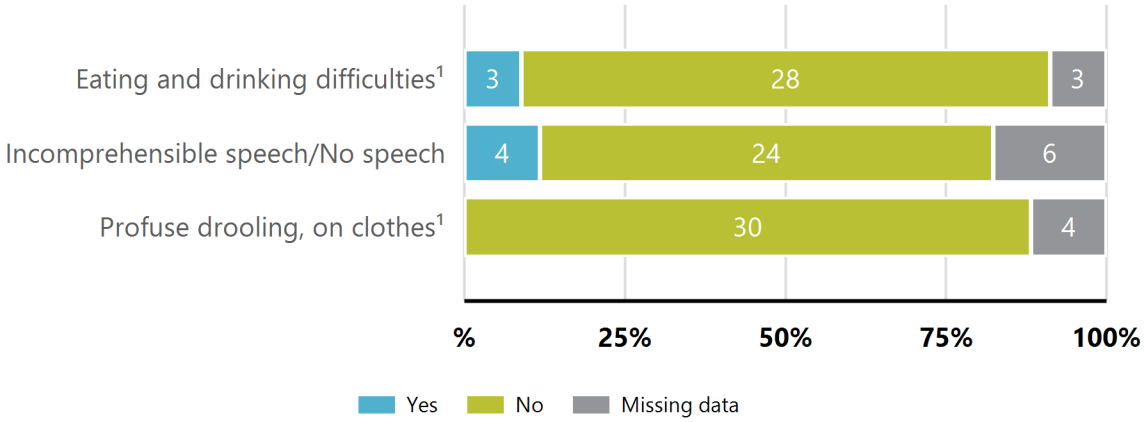
Ages: 3 - 18

♂ (15)

♀ (13)



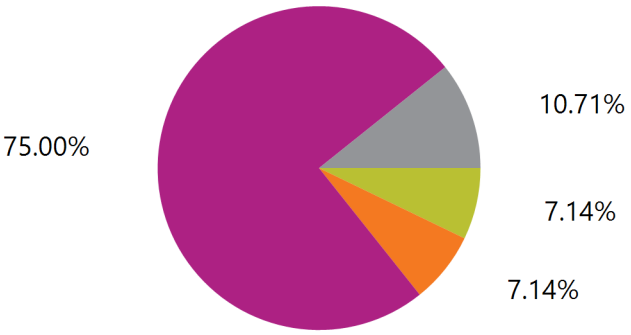
Summary



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



	Number
Negative	2
Reluctant	2
Positive	21
Missing data	3
Total:	28

Oral health

Caries

		3-6 years	7-12 years	13-19 years
deft	**Examined**	12	8	
Number of carious or filled deciduous teeth	**Number of individuals with deft=0**	11	7	
	Mean	0.3	0.3	
	Standard deviation	0.6	0.8	
	No data	9	2	
DMFT	**Examined**		8	8
Number of carious or filled permanent teeth	**Number of individuals with DMFT=0**		7	4
	Mean		0.3	1.8
	Standard deviation		0.8	2.2
	No data		1	0

Oral health index (indices)[1]

	0	1	2	3	4	5	6	Missing data	Number
Calculus	7	0	0	0	0	0	0	21	14
Gingivitis	7	0	0	0	0	0	0	21	14
Plaque	5	1	0	0	1	0	0	21	14
Tooth wear	7	0	0	0	0	0	0	21	14

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	19
Overbite	3
Underbite	5
Missing data	1
Total:	**28**

Maximum jaw opening

Children younger than 10 years

	Number
-20	0
21-30	1
31-40	2
41-50	5
51-	1
Missing data	8
Total:	**17**

Children, 10 years or older, and adults

	Number
-20	0
21-30	0
31-40	1
41-50	8
51-	1
Missing data	1
Total:	**11**

Profile[1]

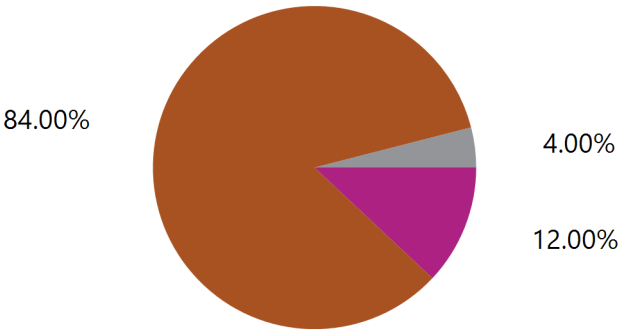
	Number
Normal	8
Convex	0
Concave	1
Missing data	3
Total:	**12**

Mandibular plane[1]

	Number
Normal	8
Increased	3
Reduced	0
Missing data	3
Total:	**14**

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

Speech problems



	Number
Slightly indistinct speech	3
No problems	21
Missing data	1
Total:	**25**

Clinical findings

Number of yes-answers

	Total = 28 (%)	♂ = 15 (%)	♀ = 13 (%)	Missing Data
Frontal open bite	5 (18)	2 (13)	3 (23)	0
Spacing	4 (14)	2 (13)	2 (15)	1
Over crowding	3 (11)	2 (13)	1 (8)	1
Open mouth at rest	2 (7)	2 (13)	0 (0)	0
Impaired tongue motility	1 (4)	1 (7)	0 (0)	1
Reduced opening capacity	1 (4)	0 (0)	1 (8)	1
Short tongue frenulum	1 (4)	1 (7)	0 (0)	1