



Orofacial function of persons having Laurence-Moon-Bardet-Biedl syndrome

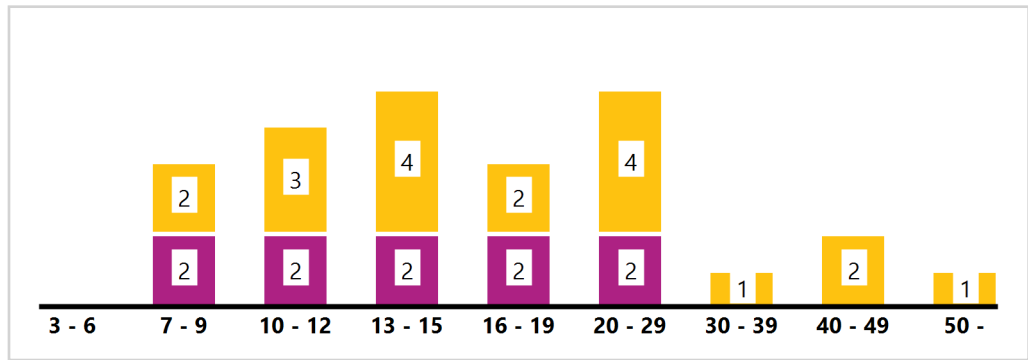
[Report from observation charts](#)

29 observation charts

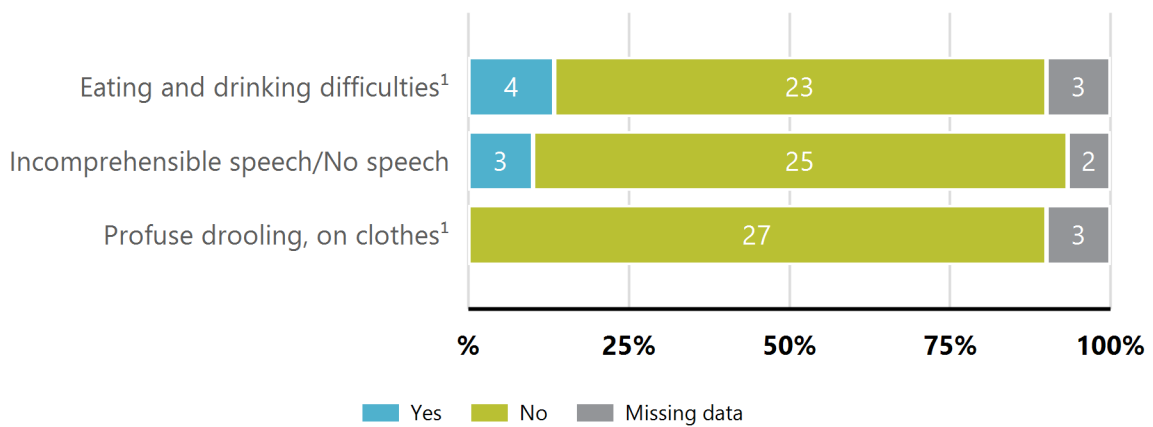
Synonym	Bardet-Biedl syndrome, Laurence-Moon syndrome
ICD-10	Q87.8
Estimated occurrence	Uncommon diagnosis. Thirteen known cases in Sweden (2004).
General symptoms	Symptoms may vary but the core symptoms are retinal changes, extra fingers and/or toes, obesity, underdevelopment of the male reproductive organs (hypogonitalism) and low concentrations of sexual hormones (hypogonadism), renal changes, intellectual disability and coordination difficulties.
Oral symptoms	Tooth anomalies may occur, such as lack of tooth buds for one or more teeth (hypodontia), small/thin teeth and short tooth roots. Snoring is common. Impaired oral motor function occurs and may cause speech and eating difficulties, as well as drooling.
Oral treatment	<ul style="list-style-type: none">• Early contact with dental services for intensified prophylactic care and oral hygiene information is essential.• Regular check-ups of dental and jaw development. Orthodontist should be consulted when needed.• Extra care is recommended in orthodontic treatment of patients with short tooth roots.• Snoring problems should be investigated by a physician in cases of suspected sleep apnea (frequent breathing pauses during sleep).• Oral motor training may be required in cases of eating difficulties, speech impairment and drooling.
Sources	The MHC database Dokumentation-Ågrenska

Age distribution

Count: 29
 Ages: 7 - 52
 ♂ (10)
 ♀ (19)



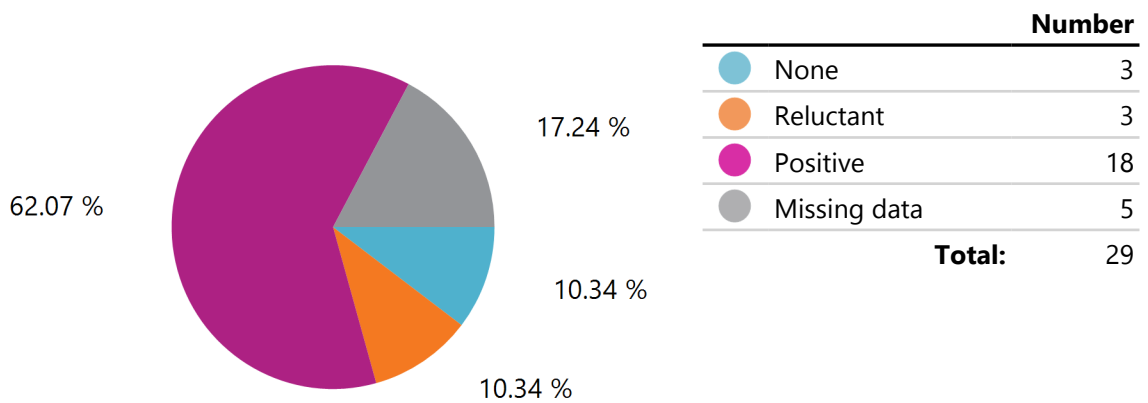
Summary



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

		7-12 years	13-19 years	>19 years
deft	Examined	9		
Number of carious or filled deciduous teeth	Number of individuals with deft=0	8		
	Mean	0.3		
	Standard deviation	0.6		
	No data	6		
DMFT	Examined	9	10	10
Number of carious or filled permanent teeth	Number of individuals with DMFT=0	8	6	3
	Mean	0.4	4.6	18.2
	Standard deviation	0.9	3.0	36.2
	No data	4	5	1

Oral health index (indices)¹

	0	1	2	3	4	5	6	Missing data	Number
Calculus	11	3	1	0	0	0	0	14	19
Gingivitis	8	2	2	1	0	2	0	14	19
Plaque	8	1	2	1	1	1	1	14	19
Tooth wear	11	3	1	0	0	0	0	14	19

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	15
Overbite	8
Underbite	2
Missing data	4
Total:	29

Maximum jaw opening

Children younger than 10 years

Children, 10 years or older, and adults

	Number
-20	0
21-30	0
31-40	0
41-50	1
51-	2
Missing data	1
Total:	4

	Number
-20	0
21-30	0
31-40	5
41-50	7
51-	7
Missing data	6
Total:	25

Profile¹

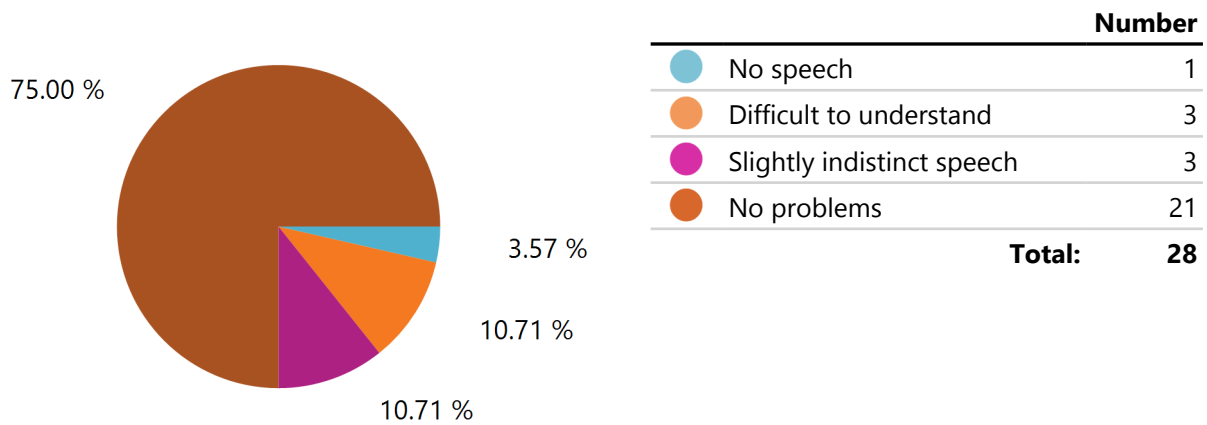
	Number
Normal	13
Convex	3
Concave	1
Missing data	2
Total:	19

Mandibular plane¹

	Number
Normal	15
Increased	1
Reduced	0
Missing data	3
Total:	19

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

Speech problems



Clinical findings

Number of yes-answers

	Total = 29 (%)	♂ = 10 (%)	♀ = 19 (%)	Missing Data
Open mouth at rest	14 (48)	6 (60)	8 (42)	0
High palate	9 (31)	1 (10)	8 (42)	3
Frontal open bite	7 (24)	3 (30)	4 (21)	3
Over crowding	5 (17)	2 (20)	3 (16)	4
Narrow palate	4 (14)	0 (0)	4 (21)	3
Spacing	4 (14)	3 (30)	1 (5)	5
Low muscle tone in lips	3 (10)	2 (20)	1 (5)	6
Impaired tongue motility	3 (10)	2 (20)	1 (5)	2
Reduced opening capacity	3 (10)	0 (0)	3 (16)	4
Deep bite with gingival contact	1 (3)	0 (0)	1 (5)	8
Short tongue frenulum	1 (3)	0 (0)	1 (5)	3