



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

## Report from observation charts

The survey comprises 14 observation charts.

**Synonyms:** Bardet-Biedl syndrome, Laurence-Moon syndrome, LMBBS

**Occurrence:** 8–55: 1,000,000 inhabitants. These figures are uncertain and the prevalence differs in different populations. It is estimated that 1-4 children with LMBBS are born annually in Sweden. The disorder affects both sexes.

**Etiology:** Unknown. Genetically determined. Changes have been found in six chromosomes with different mutations in different families. LMBBS is an autosomal recessive disorder.

**General symptoms:** Symptoms may vary but the core symptoms are retinal changes, extra fingers and/or toes, obesity, underdevelopment of the male reproductive organs (hypogonadism) and low concentrations of sexual hormones (hypogonadism), renal changes, mental retardation and coordination difficulties.

**Orofacial/odontological symptoms:** Tooth anomalies may occur, such as lack of anlage 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.

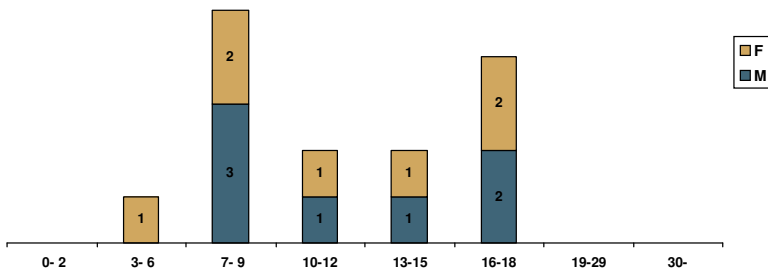
### Orofacial/odontological treatment

- Problems in managing tooth brushing justify extra preventive dental care and brushing with an electric toothbrush may be helpful.
- An orthodontist should be consulted when the child is between the ages of 7 and 9 in order to determine whether there are any dental or bite anomalies and whether corrective treatment is necessary.
- 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 rare disease database of the Swedish National Board of Health and Welfare.  
MHC-basen - Mun-H-Center's database of orofacial manifestations in rare disorders.  
Ågrenska's Newsletter (Swedish)

Age distribution

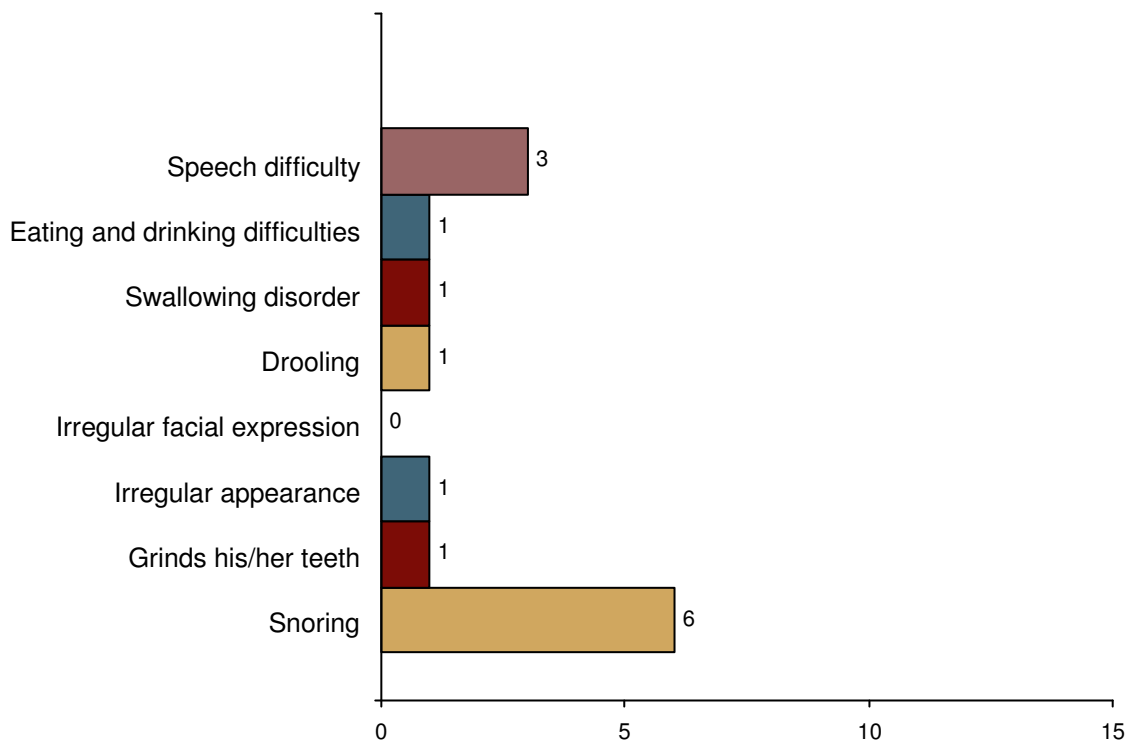


Number: 14

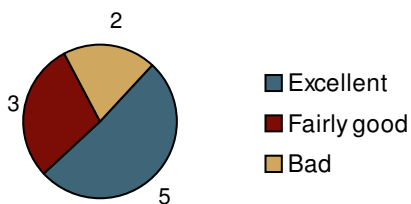
Ages: 5 -- 18 years

Sex: M (7) + F (7)

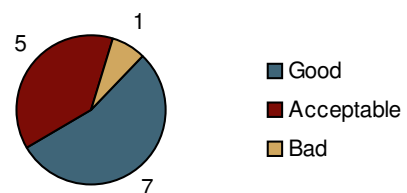
Orofacial problems



Oral health

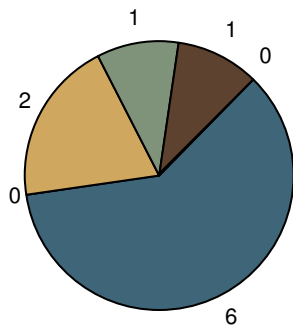


Oral hygiene



## Behaviour in the treatment situation

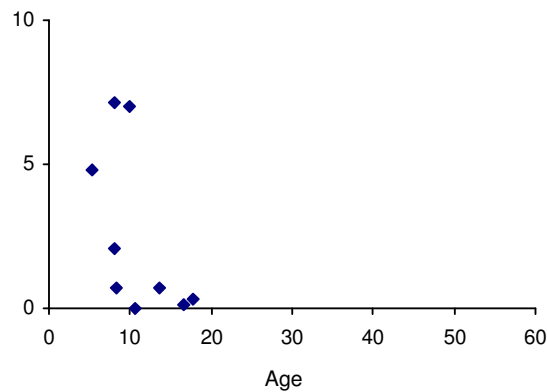
How calm and co-operative is the patient at time of examination?



- (6) Examination is possible without problem
- (0) Examination is possible without problems, some reaction is observed
- (2) Examination can continue if adjusted to patient's reactions
- (1) Reactions are considerable and examination is obviously affected
- (1) Examination is practically impossible to complete
- (0) Patient refuses examination

How does the patient cope with treatment in general?

0=no problems/10=great problems

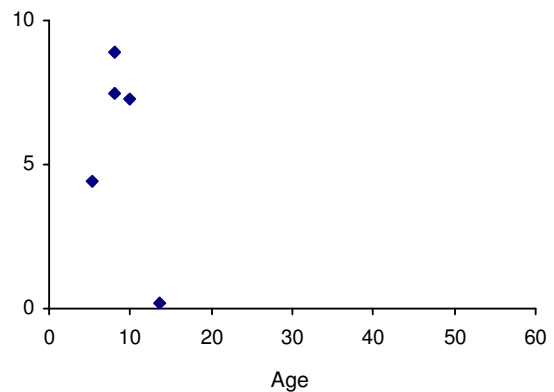
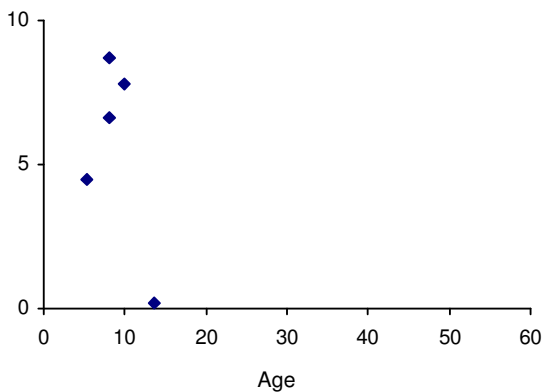


If there are treatment problems:

To what degree are the problems, if any, due to NN's handicap? To what degree are the problems, if any, due to NN's fear?

0=not at all/10=to a very high degree

0=not at all/10=to a very high degree



<b>Clinical findings</b>	<b>Total N=14</b>	<b>Boys/Men N=7</b>	<b>Girls/Women N=7</b>	<b>Not evaluated</b>
Tooth anomaly	10	6	4	1
Hypomineralisation	6	4	2	1
Open mouth at rest	5	3	2	0
Post normal bite	5	2	3	2
Frontal open bite	4	2	2	2
Mouth breathing	4	2	2	1
Narrow palate	4	0	4	1
Cross bite	3	1	2	3
Deep bite without gingival contact	3	2	1	2
Enamel hypoplasia	3	3	0	1
Gingival hyperplasia	3	1	2	0
High palate	3	0	3	1
Horizontal over-bite 6 mm or more	3	1	2	3
Lower jaw seems small	3	0	3	3
Molar contact only	3	1	2	3
Speech difficulty	3	1	2	0
Dental trauma	2	2	0	0
Frontal inversion	2	2	0	3
Gingivitis	2	1	1	0
Hypodontia	2	2	0	5
Low muscle tone in lower lip	2	0	2	0
Low muscle tone in upper lip	2	0	2	0
Over crowding	2	1	1	3
Proclined upper incisors	2	1	1	3
Retroclined upper incisors	2	1	1	3
Spacing	2	2	0	3
Upper jaw seems small	2	0	2	3
Corner of mouth lowered	1	0	1	0
Deep bite with gingival contact	1	0	1	2
Drooling	1	0	1	1
Dry mouth	1	1	0	1
Edge to edge bite	1	1	0	3
Jaw cleft	1	1	0	3
Lateral open bite	1	1	0	3
M mentalis is overactive	1	0	1	0
Mask-like expression	1	0	1	0
Misshaped occlusal plane	1	1	0	3
Posturing forward	1	1	0	3
Proclined lower incisors	1	1	0	3
Reduced mobility in tongue	1	1	0	0
Retroclined lower incisors	1	1	0	3
Scissor bite	1	1	0	3