Orofacial function of persons having
Silver-Russell syndrome
Report from observation charts

The survey comprises 37 observation charts.

Synonyms: Russell-Silver syndrome, Silver syndrome.

Estimated occurrence: Very rare.

Etiology: In approximately 50 % of individuals with Silver Russell syndrome the cause is unknown. The most common known cause is a defect on chromosome 11 (11p15) but in some individuals a change has been found in chromosome 17 and in others in chromosome 7.

General symptoms: Children with Silver-Russell syndrome are small at birth. These children do not gain weight or grow satisfactorily. The body often develops asymmetrically. The hands and feet tend to be small and the fifth finger, which grows slowly, eventually curves inward. Muscular weakness and delayed motor development are common. Learning disability is found in approximately 30% of these children. The average adult length of individuals with the syndrome who do not receive growth hormone treatment is approximately 140 cm (4 feet 7 inches) for women and approximately 150 cm (5 feet) for men.

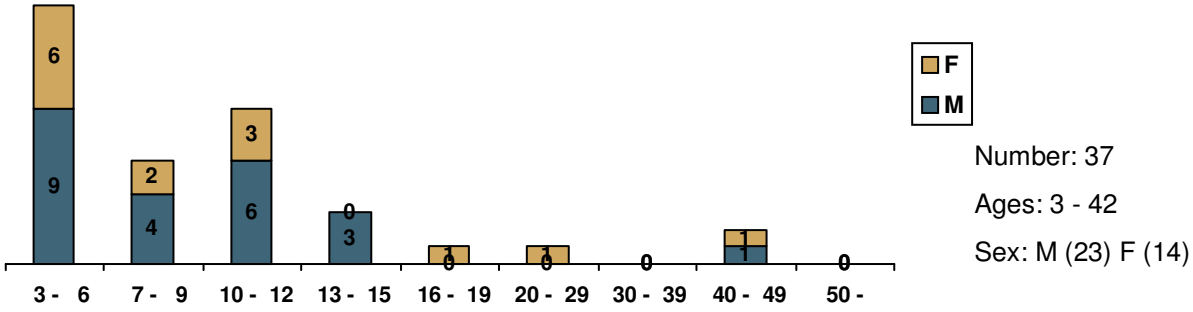
Orofacial/odontological symptoms: Children with Silver-Russell syndrome often have a special facial shape (triangular shaped face), the face being short and the mouth downward-turned. The head is large in relation to the body. Other characteristics include small jaws, a small, narrow chin, and a narrow, high palate. The occurrence of a large overbite is somewhat more common in these children than in a control group, as well as deep bite. The eruption of permanent teeth is often delayed one year. The teeth may be small and short. Enamel defects on the primary teeth and on the permanent frontal teeth is a frequent finding. Feeding impairment is common in children.

Orofacial/odontological treatment:

- Children with eating problems often require supplementary dental care, including help with their oral hygiene and fluoride treatment.
- Regular check-ups of dental and jaw development. Orthodontist should be consulted when needed.
- Feeding and swallowing difficulties are investigated and treated by a specialist team at the hospital or multidisciplinary treatment centre.
- A speech therapist may provide practical advice regarding feeding, as well as instruction for the stimulation of the mouth muscles.

Sources:
The rare disease database of the Swedish National Board of Health and Welfare.
The MHC database - The Mun-H-Center database on oral health and orofacial function in rare diseases.
The Documentation from the Ågrenska Center.

Age distribution



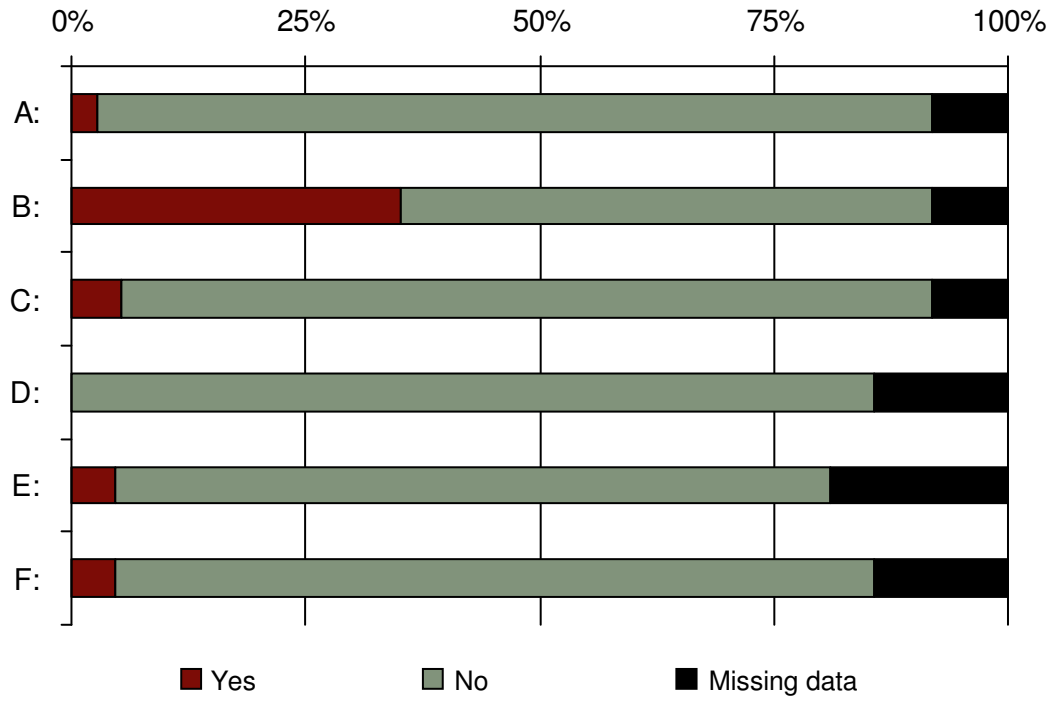
Number: 37

Ages: 3 - 42

Sex: M (23) F (14)

Overview

		Yes	No	Missing data	N
A:	Incomprehensible speech/No speech	1	33	3	37
B:	Eating and drinking difficulties[1]	13	21	3	37
C:	Profuse drooling, on clothes[1]	2	32	3	37
D:	Breathing difficulties[1] [2]	0	18	3	21
E:	Grinding every day[1] [2]	1	16	4	21
F:	Severe malocclusions[2]	1	17	3	21



Note that the diagram is based upon less than 100 individuals.

1: Compiled using questionnaire

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

Oral health

Oral health index (indices)[1]

		0	1	2	3	4	5	6	Missing data	N
Calc	Calculus	13	1	1	0	1	0	0	5	21
GI	Gingivitis	13	2	0	0	1	0	0	5	21
Plaq	Coating	12	1	1	0	2	0	0	5	21
Toot	Tooth wear	12	2	2	0				5	21

C 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.

GI 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.

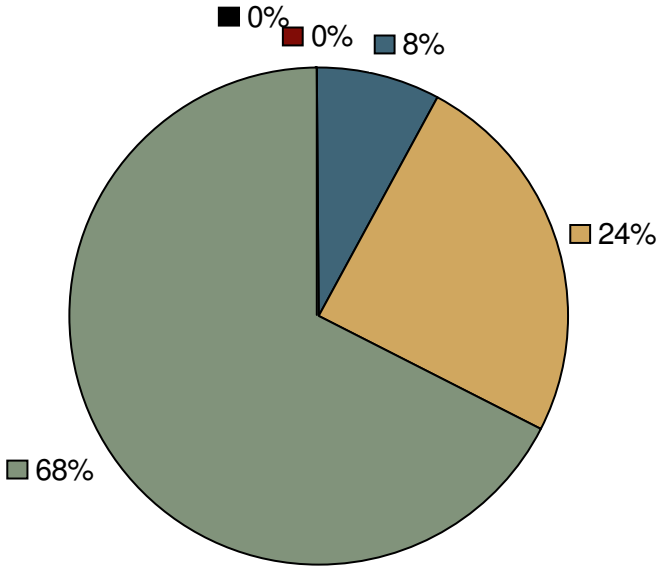
PI Plaque index is based on the presence of visible plaque on the buccal surface of 6 index teeth. 0 indicates that there is no plaque, 6 indicates plaque on all index teeth.

To 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: Oral health index (indices) was (were) introduced in the observations in 2008

Acceptance of dental examination



	Number
None	0
Negative	3
Reluctant	9
Positive	25
Missing data	0
Sum:	**37**

Caries

	3-6 years	7-12 years	13-19 years	Adults
deft[1]				
Examined	12	9		
Number of individuals with deft=0	10	8		
Mean	2,1	0,3		
Standard deviation	5,4	0,9		
Missing data	3	6		
DMFT[2]				
Examined		9	3	3
Number of individuals with DMFT=0		7	2	1
Standard deviation		1,0	0,9	4,0
Mean		0,4	0,7	3,3
Missing data		6	1	0

1: Number of carious or filled deciduous teeth

2: Number of carious or filled permanent teeth

Occlusal relationship

	Number
Neutral bite	23
Post normal	10
Pre normal	0
Missing data	4
	Sum: 37

Maximum jaw opening

Children younger than 10 years

	Number
- 20	0
21 - 30	4
31 - 40	9
41 - 50	0
51 -	0
Missing data	8
	Sum: 21

Children, 10 years or older, and adults

	Number
- 20	0
21 - 30	1
31 - 40	7
41 - 50	7
51 -	1
Missing data	0
	Sum: 16

Profile[1]

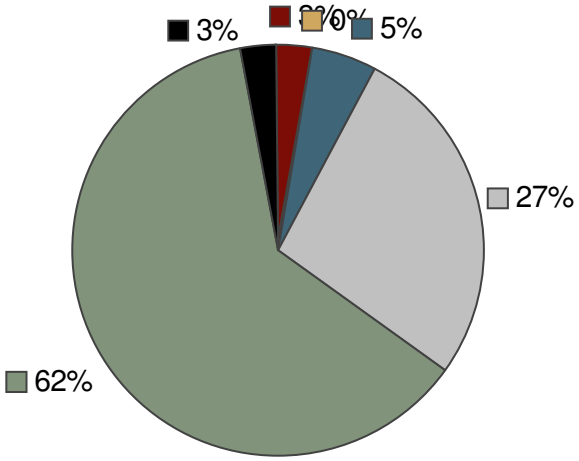
	Number
Normal	13
Convex	6
Concave	0
Missing data	2
	Sum: 21

Mandibular plane[1]

	Number
Normal	12
Increased	6
Reduced	0
Missing data	3
	Sum: 21

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

Speech difficulty



	Number
No speech	1
Very incomprehensible	0
Incomprehensible speech	2
Slightly indistinct speech	10
No problems	23
Missing data	1
Sum:	**37**

Clinical findings	Yes-answers Total N=37 (%)	Boys/Men N=23 (%)	Girls/Women N=14 (%)	Missing data
Open mouth at rest	**15** (41)	10 (43)	5 (36)	0
Over crowding	**12** (35)	9 (41)	3 (25)	3
High palate	**9** (25)	5 (23)	4 (29)	1
Impaired tongue motility	**7** (19)	7 (30)	0 ()	1
Low muscle tone in lips	**5** (14)	4 (18)	1 (7)	1
Narrow palate	**5** (14)	4 (18)	1 (7)	1
M mentalis overactive	**4** (11)	3 (14)	1 (7)	1
Reduced opening capacity	**4** (14)	3 (18)	1 (8)	8
Deep bite with gingival contact	**4** (12)	2 (10)	2 (15)	3
Facial asymmetry	**3** (8)	2 (9)	1 (7)	0
Short tongue frenulum	**3** (8)	3 (13)	0 ()	1