Orofacial function of persons having
Silver-Russell syndrome

Report from observation charts | 42 observation charts

Synonym Russell-Silver syndrome, Silver syndrome.

ICD-10 Q87.1G

Estimated occurance Very rare.

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 percent 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.

Oral symptoms Children with Silver-Russell syndrome often have a special facial shape (triangular shaped face), the face being short and the mouth downwardturned. 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.

Oral 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 MHC database Rare diseases Dokumentation-Ågrenska

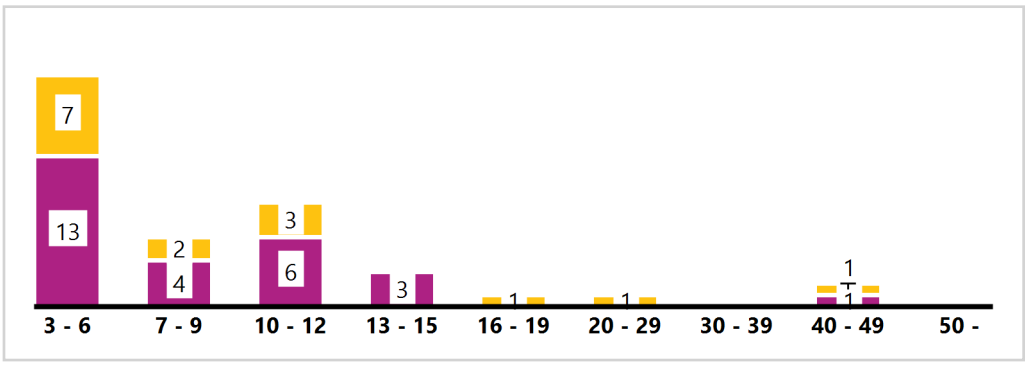
Age distribution

Count: 42

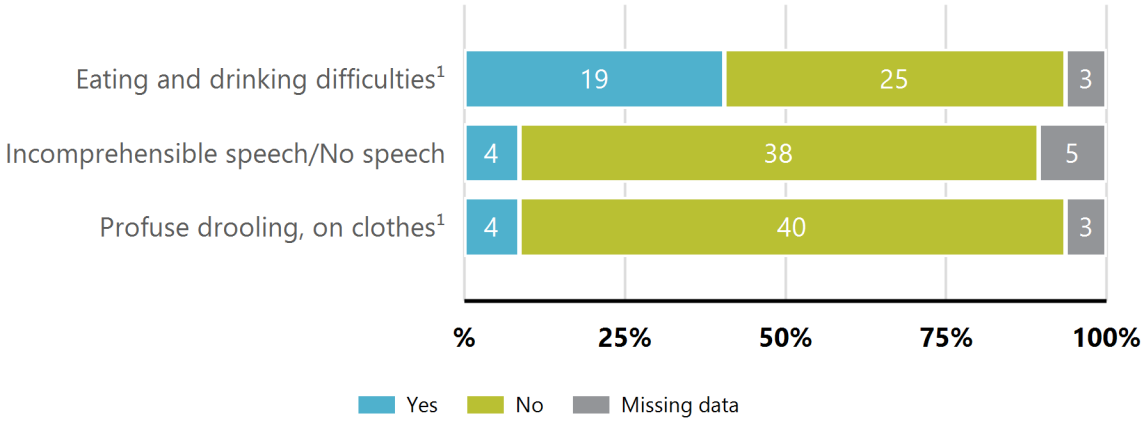
Ages: 3 - 42

♂ (27)

♀ (15)



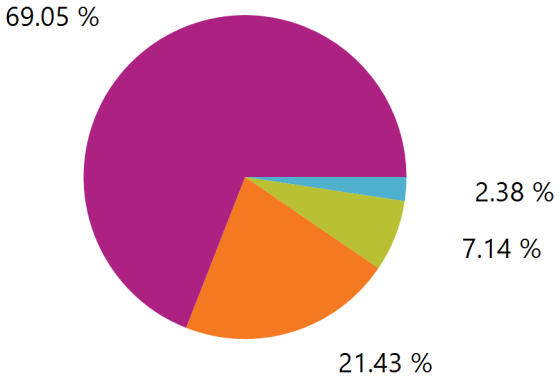
Summary



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
None	1
Negative	3
Reluctant	9
Positive	29
Total:	42

Oral health

Caries

		3-6 years	7-12 years	13-19 years	>19 years
deft	**Examined**	20	15		
Number of carious or filled deciduous teeth	**Number of individuals with deft=0**	18	14		
	Mean	2.1	0.3		
	Standard deviation	5.6	1.0		
	No data	8	6		
DMFT	**Examined**		15	4	3
Number of carious or filled permanent teeth	**Number of individuals with DMFT=0**		13	3	1
	Mean		0.4	0.7	3.3
	Standard deviation		1.0	1.2	4.9
	No data		6	1	0

Oral health index (indices)[1]

	0	1	2	3	4	5	6	Missing data	Number
Calculus	15	1	1	0	1	0	0	24	26
Gingivitis	15	2	1	0	1	0	0	23	26
Plaque	14	1	2	0	2	0	0	23	26
Tooth wear	12	2	4	0	0	0	0	24	26

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	27
Overbite	12
Underbite	0
Missing data	3
Total:	**42**

Maximum jaw opening

Children younger than 10 years

	Number
-20	0
21-30	5
31-40	9
41-50	0
51-	0
Missing data	12
Total:	**26**

Children, 10 years or older, and adults

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

Profile[1]

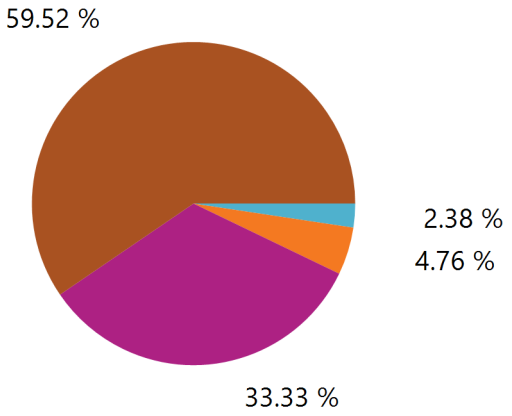
	Number
Normal	14
Convex	9
Concave	0
Missing data	3
Total:	**26**

Mandibular plane[1]

	Number
Normal	15
Increased	6
Reduced	1
Missing data	4
Total:	**26**

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

Speech problems



	Number
No speech	1
Difficult to understand	2
Slightly indistinct speech	14
No problems	25
Total:	**42**

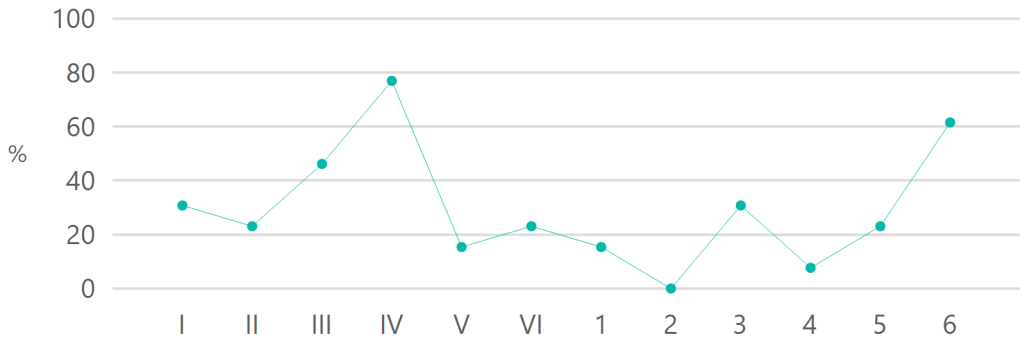
Clinical findings

Number of yes-answers

	Total = 42 (%)	♂ = 27 (%)	♀ = 15 (%)	Missing Data
Open mouth at rest	18 (43)	13 (48)	5 (33)	0
Over crowding	14 (33)	10 (37)	4 (27)	4
High palate	9 (21)	5 (19)	4 (27)	2
Impaired tongue motility	9 (21)	9 (33)	0 (0)	0
Low muscle tone in lips	6 (14)	5 (19)	1 (7)	2
Narrow palate	5 (12)	4 (15)	1 (7)	2
Reduced opening capacity	4 (10)	3 (11)	1 (7)	7
Deep bite with gingival contact	4 (10)	2 (7)	2 (13)	6
Facial asymmetry	3 (7)	2 (7)	1 (7)	1
Short tongue frenulum	3 (7)	3 (11)	0 (0)	1
Frontal open bite	2 (5)	1 (4)	1 (7)	6
Low muscle tone in tongue	2 (5)	1 (4)	1 (7)	0
Low muscle tone in masticatory muscles	1 (2)	1 (4)	0 (0)	13
Spacing	1 (2)	1 (4)	0 (0)	6

NOT-S

Total mean score: 3.54



Number: 13
Ages: 3 - 14

♂ (10)
♀ (3)

NOT-S interview

		Count	%
I	**Sensory function**	4	30.8%
II	**Breathing**	3	23.1%
III	**Habits**	6	46.2%
IV	**Chewing and swallowing**	10	76.9%
V	**Drooling**	2	15.4%
VI	**Dry mouth**	3	23.1%

NOT-S examination

		Count	%
1	**Face at rest**	2	15.4%
2	**Nose breathing**	0	0.0%
3	**Facial expression**	4	30.8%
4	**Masticatory muscle and jaw function**	1	7.7%
5	**Oral motor function**	3	23.1%
6	**Speech**	8	61.5%

The report is based on data from the MHC database - the Mun-H-Center database on oral health and orofacial function in rare diseases. Data was collected by dentists and speech-language pathologists using the Orofacial Observation Chart.