



Orofacial function of persons having Silver-Russell syndrome

Report from questionnaires

The survey comprises 17 questionnaires.

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.

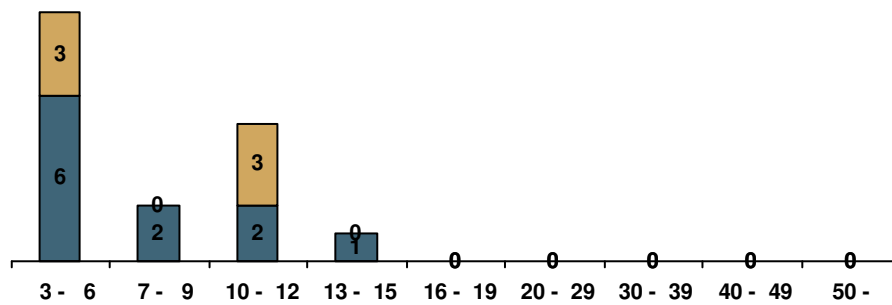
Sources:

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: 17

Ages: 3 - 14

Sex: M (11) F (6)

Additional diagnoses

Medical impairment	Yes	No	Missing data
Inborn heart defect	6	11	0
Other cardiovascular disease	1	16	0
Epilepsy	0	17	0
Asthma	7	10	0
Need of respiratory support	0	17	0
Allergy	4	13	0

Neuropsychiatric diagnosis	Yes	No	Missing data
ADHD/ADD	3	11	3
Autistic syndrome	2	15	0
Autistic traits	1	16	0
Asperger's syndrome	1	16	0
Tourette's syndrome	0	17	0

General disability	Yes	No	Missing data
Intellectual disability	5	12	0
Mobility dysfunction	7	9	1
Impaired vision	5	10	2
Impaired hearing	2	14	1
Difficulty in communicating	2	13	2

About dental care and oral health

Do you feel that you receive the dental care you need?

	Number
Yes, very much so	13
Yes, some what	2
No, not really	1
No, not at all	0
Missing data	1
Sum:	17

How many times per year do you normally seek dental care?

	Number
Three or more times per year	7
Twice per year	2
Once per year	5
Less than once per year	2
Missing data	1
Sum:	17

When were your teeth last X-rayed?

	Number
During the past two years	8
More than two years ago	0
Never had my teeth X-rayed	7
Missing data	2
Sum:	17

Do you look after your teeth in a good way?

	Number
Yes, very much so	5
Yes, some what	12
No, not really	0
No, not at all	0
Missing data	0
Sum:	17

Who brushes your teeth?

	Number
I always brush myself	2
Sometimes I brush myself	7
Someone else always helps me	8
Missing data	0
Sum:	17

How often are your teeth brushed?

	Number
Three or more times per day	1
Twice per day	14
Once per day	2
Not every day	0
Missing data	0
Sum:	17

About dental care and oral health

	Yes	No	Missing data
Does your mouth hurt?	0	17	0
Does your mouth feel dry?	0	14	3
Have you ever taken a serious hit to your permanent front teeth?	2	14	1
Do you feel that you have a divergent bite?	8	7	2
Have you had a brace?	0	16	1
Do you feel that you need orthodontics/a brace?	8	6	3

Do you grind or press your teeth at night?

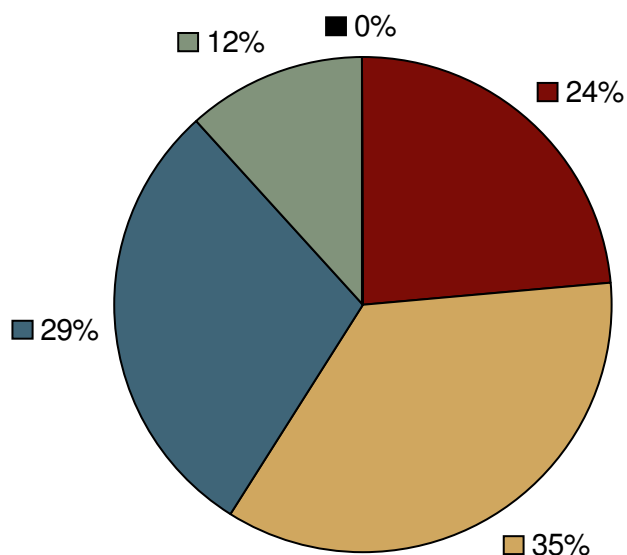
	Number
Never	10
Once or twice per week	2
Every night	4
Missing data	1
Sum:	17

Do you grind or press your teeth during the day?

	Number
Never	12
Once or twice per week	4
Every day	0
Missing data	1
Sum:	17

About eating

Do you have any problems with eating?



	Number
Yes, very much so	4
Yes, some what	6
No, not really	5
No, not at all	2
Missing data	0
Sum:	17

	Yes	No	Missing data
Do you cough daily in connection with meals?	2	15	0
Do you gag daily in connection with meals?	2	14	1
Do you get acid reflux daily?	1	14	2
Do you throw up often (at least twice per week)?	2	14	1
Do you have a poor appetite?	11	6	0
Does it take a long time before you can swallow a mouthful?	7	10	0
Do you press your tongue forward when you swallow so that food ends up outside the mouth?	0	16	1
Do you find it difficult to chew, i.e. grind food using your molars?	6	10	1
Do you find it difficult to take food from the spoon using your lips?	3	13	1
Have you had problems with food and drink leaking out through the corners of your mouth?	1	16	0
Does food tend to remain in your mouth after meals?	2	14	1
Do you get nutrition in any other way than through your mouth?	8	9	0



About drooling

Do you drool?

	Number
Never drool	17
Drool sometimes – not every day	0
Drool often – every day	0
Constant drooling	0
Missing data	0
Sum:	17

:

:

: