



# Orofacial function of persons having Sturge Weber syndrome

## Report from observation charts

The survey comprises 12 observation charts.

**Synonym:** Encephalotrigeminal Angiomatosis

**Estimated occurrence:** 2-3:1000 000 births.

**Etiology:** Unknown

**General symptoms:** The syndrome is divided in three subgroups: Sturge Weber type I includes facial and brain angiomas, epilepsy and glaucoma; Sturge Weber type II includes only facial angiomas and glaucoma may occur; Sturge Weber type III includes only brain angiomas. Facial angiomas, a port wine colored birthmark, may be seen on the forehead, upper eyelid and on one side (or sometimes both sides) of the face. The color is light pink to dark purple and is caused by an overabundance of capillaries around the trigeminal nerve. Brain angiomas are vascular malformations of the brain surface which can cause learning disabilities, behavioral abnormality, hemiparesis/ weakness and headaches/ migraines.

**Orofacial / odontological symptoms:** Facial angiomas can cause facial asymmetry and in some persons the front teeth of the upper jaw may be offset to the side that does *not* have the skin lesion. Certain medications for epilepsy can give dry mouth as a side effect, increasing the risk of cavities (tooth decay). Children with Sturge Weber syndrome and neurological impairment may have delayed speech and language development.

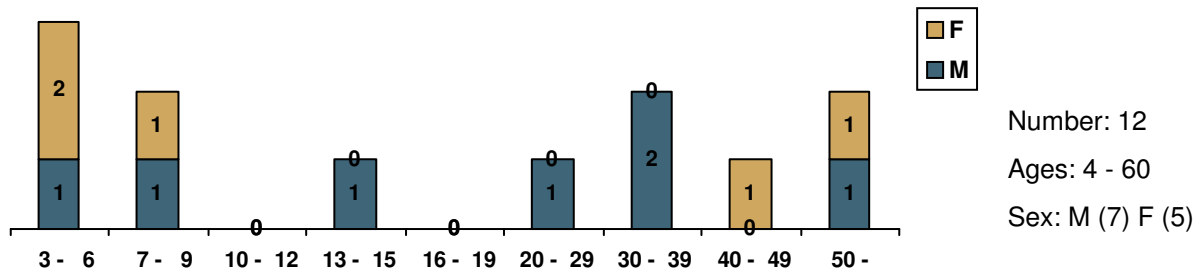
### Orofacial/odontological treatment:

- 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.
- Speech and language difficulties should be treated by a speech therapist.

### Source:

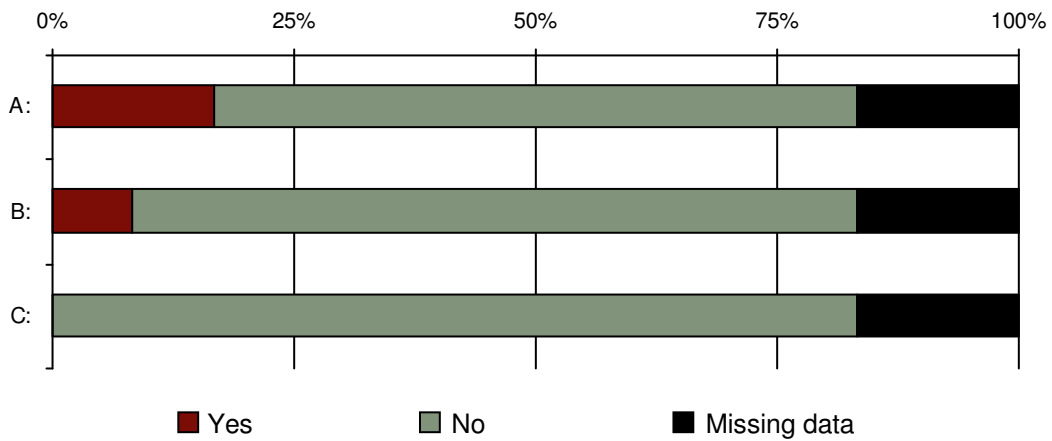
The rare disease database of the Swedish National Board of Health and Welfare.  
The MHC database - The Mun-H-Center database on oral health and orofacial function in rare diseases.  
The Documentation from the Ågrenska Center.

### Age distribution



### Overview

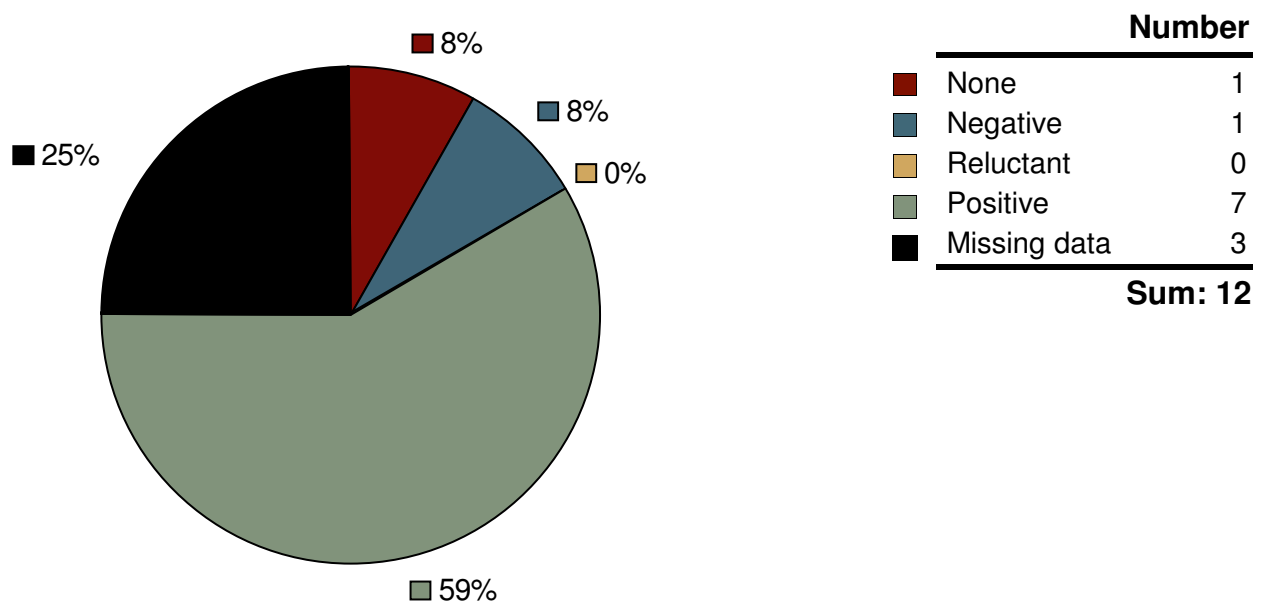
	Yes	No	Missing data	N
A: Incomprehensible speech/No speech	2	8	2	12
B: Eating and drinking difficulties <sup>1</sup>	1	9	2	12
C: Profuse drooling, on clothes <sup>1</sup>	0	10	2	12



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

<sup>1</sup>: Compiled using questionnaire

## Acceptance of dental examination



## Caries

	3-6 years	7-12 years	13-19 years	Adults
<b>deft<sup>1</sup></b>				
Examined	1	1		
Number of individuals with deft=0	1	1		
Mean	0,0	0,0		
Standard deviation	0,0	0,0		
Missing data	2	1		
<b>DMFT<sup>2</sup></b>				
Examined		1	1	5
Number of individuals with DMFT=0		1	1	1
Standard deviation		0,0	0,0	3,9
Mean		0,0	0,0	5,6
Missing data		1	0	1

1: Number of carious or filled deciduous teeth

2: Number of carious or filled permanent teeth

## Occlusal relationship

	<b>Number</b>
Neutral bite	5
Post normal	4
Pre normal	1
Missing data	2
<b>Sum: 12</b>	

## Maximum jaw opening

Children younger  
than 10 years

	<b>Number</b>
- 20	0
21 - 30	0
31 - 40	0
41 - 50	2
51 -	0
Missing data	3
<b>Sum: 5</b>	

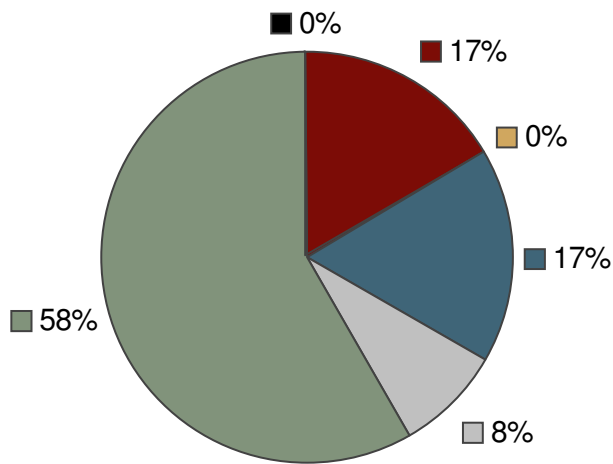
Children, 10 years or  
older, and adults

	<b>Number</b>
- 20	0
21 - 30	0
31 - 40	1
41 - 50	5
51 -	1
Missing data	0
<b>Sum: 7</b>	

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

### Speech difficulty



	<b>Number</b>
No speech	2
Very incomprehensible	0
Incomprehensible speech	2
Slightly indistinct speech	1
No problems	7
Missing data	0
<b>Sum: 12</b>	

Clinical findings	Yes-answers			
	Total N=12 (%)	Boys/Men N=7 (%)	Girls/Women N=5 (%)	Missing data
Facial asymmetry	8 (67)	5 (71)	3 (60)	0
Frontal open bite	5 (45)	3 (50)	2 (40)	1
Mucous membrane changes	4 (57)	4 (80)	0 ( )	5
Hyperplastic alveolar ridge	3 (50)	2 (50)	1 (50)	6
Gingival hyperplasia	3 (38)	2 (33)	1 (50)	4