Orofacial function of persons having
Tuberous sclerosis

Report from questionnaires | 41 questionnaires

Synonym

ICD-10 Q85.1

Estimated occurance 10:100,000 live births.

General symptoms The anomalies may be found in one or more organs, and may be minor or major. Organs that may be affected include the brain, kidneys, heart, eyes, lungs, nails and skin. The most common symptoms are skin abnormalities, and the most discernible functional disabilities include epilepsy, mental retardation and autism or autistic traits. Hyperactivity is a serious problem in nearly half of all individuals with severe tubular sclerosis. Impulse control disabilities and temper tantrums are also very frequent symptoms.

Oral symptoms Enamel aberrations in the form of pitting are frequent. Gum fibromas may occur, particularly in the region of the front teeth. There is a risk of bone cyst formation in the jaw. Speech- and language development is often delayed, and some of these children never acquire speech. Eating difficulties and drooling are common.

Oral treatment

- Early contact with dental services for intensified prophylactic care and oral hygiene information is essential.
- When enamel pitting occurs, this seldom requires treatment but may increase the risk for caries.
- Because of the risk for bone cyst formation in the jaw, radiographic evaluation is recommended by the age of 6 to 7 years or earlier if asymmetry, asymptomatic swelling or delayed or abnormal tooth eruption sequence is evident.
- Orofacial therapy and oral motor skills training and stimulation in cases of difficulties with eating, speech or saliva leakage may be relevant.
- Speech, language and communication training are often required.
- Feeding and swallowing difficulties are investigated and treated by a specialist team at the hospital or multidisciplinary treatment centre.

Sources The MHC database Rare diseases Dokumentation-Ågrenska Teng et al. Dermatological and Dental Aspects of the 2012 International Tuberous Sclerosis Complex Consensus Statements. JAMA Dermatology 2014; 150:1095-1101.

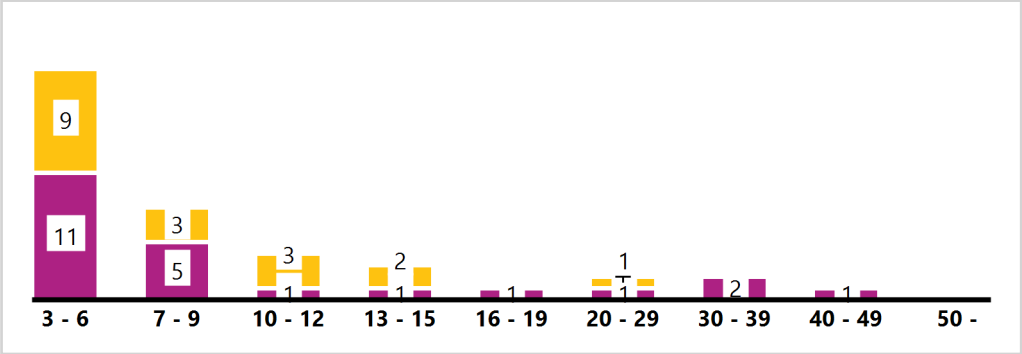
Age distribution

Number: 41

Ages: 3 - 46

♂ (23)

♀ (18)



Additional diagnoses

Medical impairment	Yes	No	Missing data
Inborn heart defect	6	33	2
Other cardiovascular disease	2	37	2
Epilepsy	32	7	2
Asthma	6	34	1
Need of respiratory support	0	39	2
Allergy	9	30	2

Neuropsychiatric diagnosis	Yes	No	Missing data
ADHD/ADD	4	33	4
Autism (Includes autism, Asperger syndrome and autistic traits)	26	9	6

General disability	Yes	No	Missing data
Intellectual disability	10	12	19
Motoric functional impairment	11	20	10
Visual impairment	6	28	7
Hearing loss	0	39	2
Communication difficulties	9	10	22

About dental care and oral health

Do you feel that you receive the dental care you need?	Number
Yes, very much so	19
Yes, somewhat	18
No, not really	2
No, not at all	1
Missing data	1
Total:	**41**

How many times per year do you normally seek dental care?	Number
Less than once per year	7
One time per year	12
Two times per year	11
Three or more times per year	11
Total:	**41**

When were your teeth last X-rayed?	Number
During the past two years	22
More than two years ago	3
Never had my teeth X-rayed	16
Total:	**41**

Do you look after your teeth in a good way?	Number
Yes, very much so	22
Yes, somewhat	17
No, not really	2
Total:	**41**

Who brushes your teeth?	Number
I always brush myself	4
Someone else always helps me	30
Sometimes I brush myself	5
Missing data	2
Total:	**41**

How often are your teeth brushed?	Number
Not everyday	1
Once per day	6
Two times per day	32
Three or more times per day	2
Total:	**41**

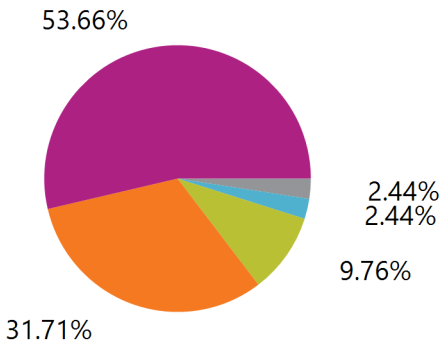
About dental care and oral health

	Yes	No	Missing data
Does your mouth hurt?	1	31	9
Does your mouth feel dry?	2	31	8
Have you ever taken a serious hit to your permanent front teeth?	10	30	1
Do you feel that you have a divergent bite?	6	32	3
Have you had a brace?	2	39	0
Do you feel that you need orthodontics/a brace?	1	31	9

Do you grind or press your teeth at night?	Number
Never	25
Once or twice per week	11
Every night	4
Missing data	1
Total:	**41**

Do you grind or press your teeth during the day?	Number
Never	28
Once or twice per week	11
Missing data	2
Total:	**41**

About eating



Do you have any problems with eating?	Number
Yes, very much so	1
Yes, somewhat	4
No, not really	13
No, not at all	22
	1
Missing data	0
Total:	**41**

	Yes	No	Missing data
Do you cough daily in connection with meals?	6	35	0
Do you gag daily in connection with meals?	0	40	1
Do you get acid reflux daily?	0	36	5
Do you throw up often (at least twice per week)?	0	40	1
Do you have a poor appetite?	4	34	3
Does it take a long time before you can swallow a mouthful?	1	28	12
Do you press your tongue forward when you swallow so that food ends up outside the mouth?	0	29	12
Do you find it difficult to chew, i.e. grind food using your molars?	3	37	1
Do you find it difficult to take food from the spoon using your lips?	2	38	1
Have you had problems with food and drink leaking out through the corners of your mouth?	0	29	12
Does food tend to remain in your mouth after meals?	1	40	0
Do you get nutrition in any other way than through your mouth?	0	41	0

About drooling

Do you drool?	Number
Never drool	27
Drool sometimes – not every day	8
Drool often – every day	6
Missing data	0
Total:	**41**

How much do you drool?	Number
Slight drooling, only on the lips	5
Moderate drooling, on lip and chin	7
Profuse drooling	2
Total:	**14**

Is your drooling a problem for you?	Number
Yes, somewhat	3
No, not really	8
No, not at all	3
Total:	**14**

Is your drooling a problem for your family or people around you?	Number
Yes, somewhat	4
No, not really	5
No, not at all	5
Total:	**14**