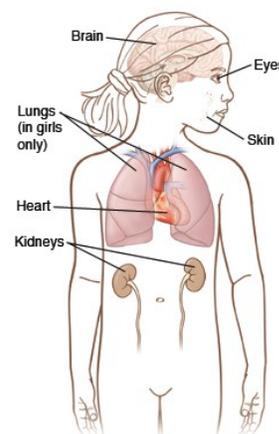


Factsheet: Tuberous Sclerosis

What is it?

Tuberous Sclerosis Complex (TSC) is a multi-system genetic condition caused by a mutation of the tumor suppressor gene. It leads to non-malignant growths in various organs of the body. These growths are sometimes referred to as tubers or lesions. TSC is caused by either an inherited gene from one parent (1/3 of cases) or the result of spontaneous mutation (2/3 of cases). Although tumors can grow anywhere, areas most commonly affected include the brain, eyes, heart, kidney, skin and lungs. Effects on the brain including seizures, developmental delay, intellectual disability and autism can all greatly impact development and behavior. However, many children are undiagnosed because of the unfamiliarity of TSC.

The growth of tumors resulting from TSC is not as severely unregulated as in cancer, but these tumors may still cause serious problems. Such tumors can block the flow of CSF causing headaches, dizziness and behavior changes, changes in blood flow of the heart causing arrhythmia, blockage of the retina that affect vision, as well as disruptions of kidney function. There is no single clinical feature that is absolutely specific to TSC. Instead, a diagnosis of TSC is based on careful physical examination in combination with imaging studies. A diagnosis usually includes at least 2 major TSC features.



What are the signs and symptoms of TSC?

Tumors can form on many surfaces and layers of the brain. Lesions can form in the fluid filled brain ventricles and even potentially block the flow of fluid in the brain causing increased pressure. Children experiencing headaches, nausea and vomiting, as well as changes appetite and mood should be assessed by a neurologist. Some children may also already have a shunt in place to drain increased fluid in the brain. Tumors can also affect the kidneys causing cysts and even possible bleeding, grow on cardiac tissue, lung tissue, as well as affect the retina of the eye. Most noticeable may be changes in skin growth and pigmentation. TSC patients may also experience seizures, developmental delays and a range of intellectual disabilities. Behavior problems can include aggression, self-harming behavior, specific phobias, sleep disturbances, and anxiety. There is also strong evidence of correlation for increased rates of ADHD and autism.

COGNITION	BEHAVIOR
Social-communication deficits	Autism, Asperger's
Receptive and expressive language deficits	ADHD and related disorders
Attention deficits	Aggression, outburst, temper tantrums
Executive deficits (planning, sequencing)	Negativity
Memory deficits	Depression
Motor abnormalities (fine, gross, movement)	Anxiety
	Sleep disorders
	Epilepsy related psychotic disorders

Tuberous Sclerosis Association (2015)

What is the treatment?

There is no cure for TSC but early diagnosis and intervention can greatly impact the person's development and quality of life. Initially, a medication regime to control seizures is helpful but when not effective, other treatment aimed at seizure control can be utilized. Surgery is sometimes used when tumors that are blocking function can be accessed and surgically removed. Drug treatment that can shrink tumors is also sometimes prescribed.

Suggested school accommodations

Children with TSC can present differently in the educational environment. Although some children function within the normal IQ range, others may have some degree of intellectual disability. Common factors that have been identified include language and communication barriers, attention deficits, behavior outburst, and social interaction disorders. Such barriers and their progression may be directly related to the severity and/or control of seizures.

- Neurocognitive assessment
- Behavior assessment
- Consider 504 or update IEP
- Maintain a quiet, calm environment
- Maintain structure, daily routines
- 1:1 teaching as necessary
- Preferential seating
- Repeat instructions
- Check for understanding
- Use step-by-step instructions
- Offer information in outline form
- Offer short bursts of information
- State the obvious when teaching
- Require organization, planning
- Limit “multi-tasking”
- Obsessive behaviors
- Need structure, routine
- Encourage peer groups
- Practice social skills through role-playing
- Note difficulty with fine and/or gross motor
- Adaptive or assistive technology

SHNIC school nurses information:

Specific health issues for individual health care plans

- Diagnosis including all medical characteristics
- Baseline cardiac assessment (including blood pressure related to kidney function), lung assessment and skin assessment
- Documentation of type of seizure, description of, typical length, characteristics, triggers, warning signs, how often seizures occur, and student’s behavior following a seizure
- Orders for hidden device like a shunt or vagus nerve stimulator, education about how to use/manage device
- Current medication list including PRN medication
- Seizure action plan, when to call 911
- Dose, route, time to administer emergency seizure medications
- Documentation/log of seizures
- Nutrition orders (Ex. Ketogenic diet)
- Safety precautions for ambulating, transitioning in hallways, wearing a helmet, etc.
- Return to class protocol

Resources & Manuals

Tuberous Sclerosis Association
<http://www.tuberous-sclerosis.org>

Tuberous Sclerosis Alliance
<http://www.tsalliance.org/pages.aspx?content=2>

Teacher’s Guide: Educating a child with Tuberous Sclerosis Complex
<http://www.tsalliance.org/documents/TeacherGuidetoTSC2014.pdf>

Behavior Issues and Tuberous Sclerosis Complex
www.tsalliance.org/documents/Behavioral%20Issues%20and%20TSC.pdf