

## **Educating a Child with Tuberous Sclerosis Complex**

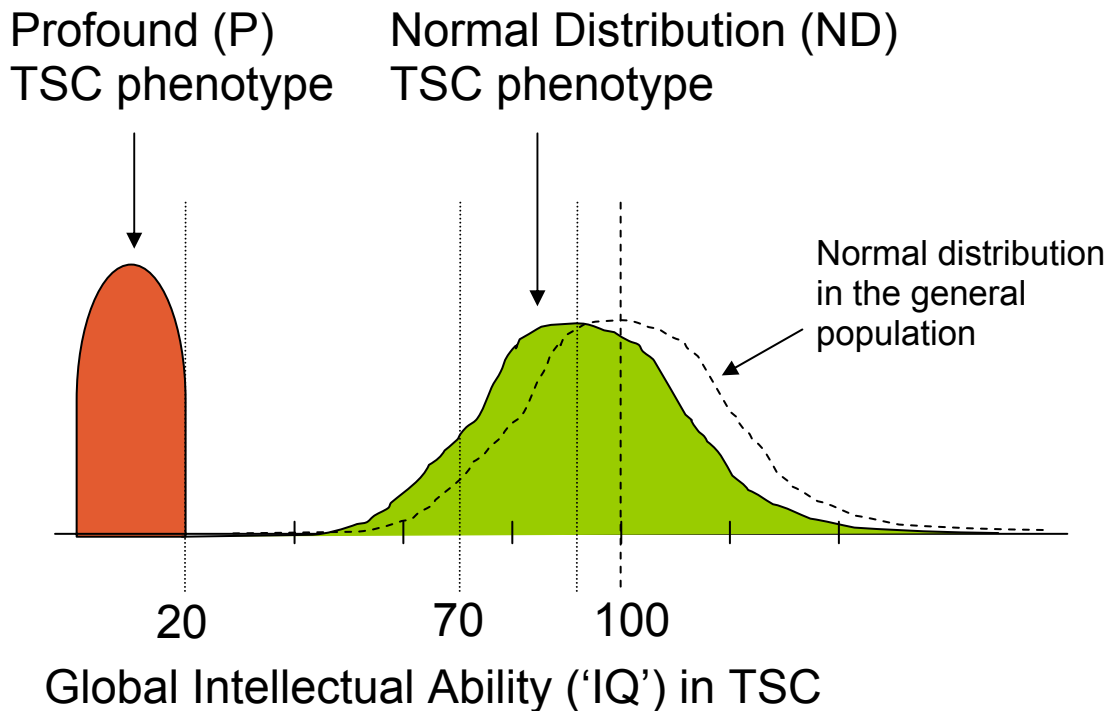


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## What is Tuberous Sclerosis Complex?

Tuberous sclerosis complex (TSC) is a genetic disease that affects one in every 6,000 live births. TSC causes tumors to develop in the skin, kidney, brain, heart, eyes, lungs, liver, oral cavity, and many other organs. The severity of an individual with TSC can vary from mild to severe, depending on where those tumors are and the manifestation caused by the tumors.

Below is a chart that shows the large range of abilities of students with TSC.



Prather & de Vries, JCN 2004, de Vries & Prather, NEJM, 2007

In the chart above, you can see that students with TSC can have a normal to gifted IQ, as well as an IQ in the severe developmental delay (mentally retarded) area. Many children with appropriate accommodations can learn and develop to be independent adults. The key is making sure the appropriate services and supports are provided. Because each individual with TSC has different educational needs it is important to understand each of these manifestations and their educational implications.

## How Does TSC Affect the Brain?

Let's start with the brain and the types of brain abnormalities that may be seen in students with TSC.

**Cortical tubers** – The cortical tuber is a disorganized area of the brain similar to a birthmark in the brain. Some individuals have numerous tubers, and others will not have any. Tubers and/or the brain area surrounding a tuber play a role in the development of seizures.

**Subependymal nodules (SENs)** – SENs are small accumulations of cells that are located in the spaces in the brain that contain cerebrospinal fluid. They often accumulate calcium and are used to diagnose TSC through a CT scan.

**Subependymal giant cell tumor (SGCT)** – Another name for a SGCT is a subependymal giant cell astrocytoma (SEGA). It is a type of non-cancerous brain tumor. A SGCT/SEGA usually grows during the adolescent to young adult years. When these nodules grow they can cause changes in learning, moods, seizures, and behavior.

### *Epilepsy/Seizure Disorders*

Sixty to 90 percent of individuals with TSC have seizures. Many children with TSC are diagnosed with TSC in infancy after they begin having a type of seizure called *infantile spasms*. Older children and adults may develop multiple types of seizures including generalized, complex partial and other focal seizures.

### *Types of Seizures*

**Generalized Seizures** – Generalized seizures affect both sides of the brain. They cause loss of consciousness, either briefly or for a longer period of time:

***Generalized tonic clonic seizures (grand mal seizures)*** – They begin with stiffening of the arms and legs, followed by jerking of the arms, legs and face.

***Myoclonic seizures*** – These seizures are brief contractions of body muscles that are rapid and usually on both sides of the body. Sometimes they may involve one arm or a foot.

***Atonic seizures*** – These seizures can cause a person to fall without warning. They can also cause his/her head to drop, or have loss of posture without warning.

***Absence seizures (petit mal seizures)*** – These seizures are associated with staring that last only for seconds. There is no after-effect or warning when these seizures occur. Because of the subtleness, these seizures are very hard to identify and a teacher may think the child is simply day dreaming.

***Infantile Spasms*** – Onset of infantile spasms peaks between four and six months of age, but can occur anytime in the first two years. Infantile spasms are seizures that result in sudden jerks involving all or part of the body in a forward or backward motion. It can be as subtle as a slight bob of the head or a thrust of the chin, but over time the seizures usually become more pronounced and occur in clusters of many seizures.

**Partial Seizures** – Partial seizures affect one side or area of the brain. There are two types of partial seizures:

***Simple partial seizures*** – These seizures do not cause unconsciousness but do cause sudden jerking and sensory issues. They last approximately 90 seconds. There might be weakness or loss of sensation following this type of seizure.

***Complex partial seizures*** – There might be precursors to this seizure such as picking at clothes, wandering, lip smacking, and lack of environment awareness. They last one to two minutes, and the individual may have confusion, loss of memory, and/or sleepiness following the seizure.

### *Types of Treatment for Seizures*

#### **Medication**

It is important to understand the types of treatments for seizures and their side effects. Many side effects can cause issues in learning. Medication for childhood epilepsy may cause issues with cognitive development and

learning. These possible effects are mainly in the area of speed of processing and attention processes. There is also increasing evidence that various types of behavioral disturbance may occur from the use of anti-epileptic medication. When you are teaching a student who is on anti-epileptic medicines, you should ask the parent about possible side effects.

### **Vagus Nerve Stimulation (VNS)**

The VNS is surgically placed under the skin of a child's chest. This device provides short bursts of electrical energy that are directed into the brain through a large nerve (vagus nerve) in the neck.

### **Ketogenic Diet**

This diet is a very strict and should be monitored by a physician together with a nutritionist. This diet is very high in fats and low in carbohydrates. Because it is high in fats and low in carbohydrates, it makes the body burn fat for energy instead of glucose. When this diet is used, it usually helps two out of three children and can prevent seizures in some children.

### **Resective Surgery**

Resective surgery is used in individuals with seizures where there is a clear specific seizure focus in an area of the brain that can be safely removed without causing a significant loss of function. There have been good surgical outcomes for many children with TSC receiving this type of surgery. They may have improvement in behavioral and cognitive development after successful surgery that results in a reduction of seizures.

### **Retention Intervention**

Most research states that retention is not a good intervention for children with disabilities. But, in cases where a young child receives resective surgery many of these children's cognitive abilities improve. By retaining these children, it gives them a chance to fill the gaps in their learning that were caused by their seizures and the anti-epileptic medication. Requiring that a child repeat a grade level and immerse them in the curriculum helps to fill those learning gaps. Passing them to the next grade level and trying to address their gaps in special education is a far less effective way to fill their

needs. So, in the case of a young child with TSC having resective surgery, **retention is a good intervention.**

## **Seizure First Aid**

- 1. Help the person lie on the floor, placing him/her on their side.**  
During a seizure, a person is unable to control his/her movements and posture. Lying on the floor minimizes the risk of physical injury. Also, an individual will often vomit during a seizure episode. Laying him/her on their side prevents choking. Make sure to move objects away that are potentially dangerous to the individual (sharp objects, glass, etc.).
- 2. Do not put anything in the person's mouth.**  
It is common belief that an individual may swallow their tongue during a seizure. Although it is possible that person may bite their tongue, this rarely, if ever, causes serious injury. On the other hand, the human jaw is very powerful, and people have bitten off spoons, sticks and other objects during seizures. In addition, placing anything in the person's mouth poses a greater danger of choking.
- 3. If the seizure activity lasts more than five minutes, call 911.**  
Most seizures are brief and self-limited, but they occasionally do last longer. If you have a rectal medication (i.e. Diastat) available, administer it as directed. Do not attempt to administer medications by mouth! A person is unable to swallow during a seizure and will most likely choke if something is placed in the mouth.
- 4. Check to see if the individual has a medical I.D. bracelet or necklace that says "epilepsy" or "seizure disorder."**  
Some individuals wear a medical I.D. bracelet or necklace to help other people know that they are being treated for seizures. Often, the I.D. will have a list of the medication(s) on the back of the I.D. so that health care professionals will know immediately what medications the person uses to treat the seizure disorder.
- 5. Try to remain calm.**  
Remember that most seizures are brief and self-limited, and the individual is rarely in danger. However, if at any time there is

concern for someone's safety, call 911. Speak softly and reassuringly to the person having the seizure and offer them help until the seizure ends. Stay with the person until he or she is completely awake and aware again.

**6. There are some special things to remember if a seizure occurs while the individual is in the water (in the bathtub or while swimming).**

You should support the person having a seizure in the water, and make sure their head is tilted so the face and head stay above the surface of the water. Help them to get out of the water as soon as possible. Once on dry land, make sure they are breathing. If they are not breathing begin rescue breathing at once and call 911. Anyone who's had a seizure while in the water should go to the emergency room for a checkup, even if he/she seems to be alright afterward. If a lot of water was swallowed, there could be damage to the heart and lungs.

**When should you call 911?**

- You have no way of knowing whether or not the person has epilepsy.
- The person having a seizure does not have epilepsy because it could be a sign of serious illness.
- The person having a seizure is pregnant, has diabetes, is injured in some way, or appears to be ill.
- The seizure goes on longer than five (5) minutes.
- Another seizure starts right after the first one ends.
- The person has trouble breathing, seems hurt, or is in pain.
- The person isn't getting back to normal the way he or she usually does following a seizure.

**You do not need to call 911 if:**

- The seizure ends after a couple of minutes;
- The individual wakes up and starts breathing normally again; and
- Is fully awake and aware after a short rest.

*"Seizure First Aid" was written by Elizabeth Thiele, Ph.D., M.D., Director of the Carol and James Herscot Center for TSC and Director of the Pediatric Epilepsy Program, Massachusetts General Hospital, Boston, Massachusetts, and an assistant professor at Harvard Medical School.*

## Parent and School Communication About Seizures

Children with TSC may go years without the occurrence of seizures. It is important to have ongoing communication with a student's parents if there is any type of change in a student's social interaction, behavior, learning, and energy level (tired). Some seizures can go unnoticed because they might be very subtle. They can be as unnoticeable as a student just staring, and might only last for seconds. If you notice any changes in the student's behavior contact the parents and talk with them about your concerns.

There might also be times when you will need to help the parents and doctors monitor medications. You might have to collect data on how many seizures, what type, and when a student may be having seizures. Below is a chart to help with this process.

### Seizure Communication Form

Student's Name \_\_\_\_\_

Date	Time of day	Where did the Seizure occur?	What did the Seizure look like?	How long did it last?	How did the Student act after the Seizure	What interventions were provided?

Teacher Signature \_\_\_\_\_ Date \_\_\_\_\_

### Why a Neuropsychological Evaluation?

Children with TSC can have anything from a mild learning disability to autism. When you are looking at the disability categories under the Individuals with Disabilities Education Improvement Act of 2004 (IDEA) children can have any one of the disabilities identified under IDEA and some may have more than one disability. Because TSC is a genetic condition, it is important to have a good baseline of the student's abilities and understand how the student learns.

TSC experts recommend that all children diagnosed with TSC have a thorough neuropsychological evaluation at the time of diagnosis so early intervention can be implemented. A consensus on the initial neuropsychological evaluation and subsequent follow-up was published in 2005 and includes recommendations for an initial evaluation and follow-up neuropsychological testing at times of school transition and/or as needed based on the needs of the individual with TSC.

A neuropsychological evaluation is the best form of data to have in identifying a student's baseline and needs. A typical school evaluation is done to see if the student qualifies for Special Education to support progressing the in the general education curriculum. A neuropsychological evaluates specific cognitive abilities that are affected by the TSC lesions (cortical tubers, subependymal nodules, and subependymal giant cell astrocytomas) in the brain. This type of evaluation provides valuable information about a child's development in areas such as language, memory, attention, perception, coordination, and personality – basically “how the child learns.”

Many parents of children with TSC are already working with a neuropsychologist and have current neuropsychological evaluations. It is in your best interest as a teacher and school district to use this information when developing a child's IEP. The recommendations on the neuropsychological evaluation will support the IEP Team in developing a plan that best fits the needs of the student.

If you have had a child with TSC in your classroom in the past, don't assume you understand the needs of another child with TSC. Students with TSC can have mild to severe learning issues. There is nothing consistent about TSC except that it is inconsistent. TSC manifests differently in every individual with TSC.

### **Learning Disabilities**

When a student with TSC has brain involvement learning disabilities can occur. Students with TSC may be doing fine at the beginning of the school year and start struggling in the middle or end of the school year depending on their seizures, tumor growth, and the area of the brain where the tumor or seizure are located. Please note that you might have to develop a new IEP more than once during a school year depending on what is going on

medically with the student. Behaviors and learning issues are a red flag for changes in tumor growth and seizure activities in the brain. Below are some learning disabilities that can happen to children with TSC.

### *Dyslexia*

According to the U.S. National Institute of Health, dyslexia is a learning disability that can hinder a person's ability to read, write, spell, and sometimes speak. The area of the brain that controls the ability to translate images received from the eyes or ears into understandable language may be affected. The child's vision or hearing ability has nothing to do with their learning difficulties.

Warning signs of dyslexia:

- Trouble moving to the rhythm of music
- Cannot remember content of stories
- Does not understand left from right
- Trouble with visual spatial concepts
- Uncoordinated (skipping is difficult)
- When speaking, may use wrong word or reverse words
- When writing reverse letters, words, and/or numbers
- Cannot proof their own written work
- Does not understand time
- Does not understand seasons

It is important to do early identification and intervention for students suspected of having dyslexia to be successful in school. The response to intervention (RTI) process should be implemented as soon as a child is suspected of have dyslexia. Through this process a child can be identified early and placed on an IEP to avoid depression, behaviors, low self esteem, and losing interest in school.

RTI is a process in which the school system documents a child's response to scientific, research-based intervention using a tiered approach. In contrast to the discrepancy criterion model, RTI provides early intervention for students experiencing difficulty learning to read. RTI was authorized for use in December 2004 as part of the Individuals with Disabilities Education Act

(IDEA). This provides the student with early intervention and identification into special education.

### ***Dyscalculia***

When a child has TSC involvement in the language and visual processing centers of the brain dyscalculia can result. Dyscalculia is an inability to understand the meaning of numbers and/or inability to apply math principles to solve problems.

Warning signs of dyscalculia:

- Does not understand numbers and their quantities
- Does not understand addition
- Does not understand subtraction
- Does not understand multiplication
- Does not understand division
- Does not understand abstract concept (algebra)
- Has difficulty in making change and handling money
- Does not recognize patterns
- Does not understand days, weeks, months
- Does not understand time
- Has difficulty lining numbers up on a page
- Telling time

Because students with dyscalculia do not understand the basic concepts of numbers and quantities, they can not build on their knowledge to learn more complicated concepts.

There are strategies for educators to use to assist students with dyscalculia. These include:

- Assistive technology (calculator)
- Using music to teach math facts
- Use pictures for word problems
- Color code math problems (red subtraction-green addition)
- Manipulative
- Number chart
- Graph paper to keep numbers in line

Students with severe dyscalculia should be given every opportunity to learn math concepts. However, the IEP Team should determine when a child should have the curriculum modified to reflect what part of the general math curriculum should be taught. Many times a student needs to be taught basic math concepts; telling time, counting money, and understanding the days of the week are more important than learning higher math concepts. It is the IEP Team's job to assess what part of this curriculum will help the student reach his/her fullest potential. Sometimes that does not include learning algebra or geometry.

### ***Dysgraphia***

Dysgraphia is a learning disability that affects written expression and difficulty in processing the spoken language. Students with dysgraphia have visual-spatial difficulties and struggle organizing letters, number and words on a line or page. They can also have language processing difficulty. It may take a child with language processing 10 seconds or more to interpret what is being asked of them.

Warning signs of dysgraphia:

- Trouble writing or forming letter shapes
- Is not consistent with spacing between words and letters
- Tires of writing even short assignments
- Refuses or is reluctant to do writing assignments
- Does not like to draw or color
- Awkward pencil grip and unnecessary pressure on pencil point
- Cannot stay on the line when using scissors to cut
- Older students may struggle getting thoughts down on paper

Many educators try to get students with dysgraphia to slow down to help them write correctly, thinking this will give them time to concentrate on writing correctly. But slowing down can, in fact, cause them to get stuck on the details for writing and lose their train of thought.

There are strategies for educators to use to assist students with dyscalculia. These include:

- Use a computer for writing assignments
- Provide occupational therapy
- Use a tape recorder to tape ideas and organize thoughts
- Use a scribe
- Utilize a peer note taker
- Test orally
- Minimize writing assignments

### **Attention-Deficit/Hyperactivity Disorder (AD/HD)**

Attention-Deficit Hyperactivity Disorder (ADHD) is a common neurobehavioral disorder. Approximately 5 to 8 percent of school-age children have symptoms of ADHD that interfere with academic performance. ADHD may be even more common in child with TSC. There are as many as 25 to 50 percent of children with TSC who exhibit some signs of ADHD. ADHD often accompanies epilepsy and learning disabilities, and the symptoms of ADHD are usually present in children with autistic spectrum disorders. Perhaps one-third of the children with TSC plus epilepsy or TSC plus learning disability will have evidence of ADHD.

There are three types of ADHD:

1. Hyperactivity-Impulsivity ADHD
2. Inattentive ADHD
3. Combined ADHD

Warning signs of ADHD:

#### *Hyperactivity*

- A student who has endless energy and cannot seem to slow down
- A student who is jumpy, fidgety, squirmy, and noisy
- The student does not think before doing something (impulsive) both in action and speech
- The student is very disruptive
- The student cannot wait in turn taking
- The student shows emotions with lack of control

### *Inattentive*

- A student who can't stay focused on anything new
- A student who can't complete chores or homework
- A student who is very disorganized
- A student who forgets necessary items easily
- A student who can't do more than one step directions
- A student who can be hyper-focused on activity he/she enjoys (for example, playing video games or reading a book)
- A student who will stare off into space

### *Combined*

- A student who has endless energy and cannot seem to slow down
- A student who is jumpy, fidgety, squirmy, and noisy
- A student who does not think before doing something (impulsive) both in action and speech
- A student who is very disruptive
- A student who cannot wait in turn taking
- A student who shows emotions with lack of control
- A student who can't stay focused on anything new (task or idea)
- A student who can't complete chores or homework
- A student who is very disorganized
- A student who forgets necessary items easily
- A student who can't do more than one-step directions
- A student who can be hyper-focused on activities he/she enjoys (for example, playing video games or reading a book)

Strategies for educators to use to assist students with ADHD include:

- Seat the student in the location with the least distractions in the classroom (not by door or window)
- Post the classroom daily schedule and assignments in easy viewing
- Let the student know when activities are going to change 15 minutes, 10 minutes, 5 minutes before the end of activity

- Set up daily strategies within the classroom to help organize the students (backpack is always checked for books and homework assignments before leaving the classroom)
- Pair student up with others who are well-organized
- When giving instructions, also provide a visual example of the instructions to ensure understanding
- Break the instructions down into steps
- Provide priority lists for large assignments
- Provide extended time and small groups for test taking
- Let the student use small manipulative (chenille sticks) to help focus when listening to instruction
- Utilize a communication booklet between teachers and parents daily, weekly, or monthly depending on the age of the student

### **Autism Spectrum Disorder**

TSC is considered one of the leading genetic causes of autism spectrum disorder (ASD). Over the years, it has become recognized that between one-fourth and one-half of all children with TSC develop ASD. The rate of ASD in the general population is substantially lower (around 0.5 or 0.6 percent of the total population), so there is clearly a very substantial increase in the rate of ASD in children with TSC. Likewise, the rate of TSC in children diagnosed with ASD is around 1 percent. Although this is a relatively low rate, it is still clearly much higher than the rate of TSC in the general population (approximately 1 in 6,000 individuals).

Warning signs of ASD:

- An impairment in the ability to interact socially with people; often demonstrating a lack of eye contact and disinterest in physical contact such as hugging or hand-holding;
- An impairment in the ability to communicate using speech and/or gestures; and
- A tendency to have narrow patterns of interests and activities coupled with repetitive and obsessive behaviors, and a lack of pretend or imaginative play; often children with ASD find it necessary to have rigid and structured routines.

There are a wide range of variants and degrees of demonstrated behavior such that autism is often defined as “autistic spectrum disorder” or ASD.

Some children have clear signs of ASD in two of the main areas required for diagnosis, but have less obvious features in the third. In these instances, individuals are said to have an atypical form of autism. When the intellectual abilities are normal, early language development is not significantly delayed and speech is well developed, then individuals may meet criteria for another variant called Asperger syndrome. A third variant, termed pervasive developmental disorder (not otherwise specified) or PDD-NOS, describes individuals who have difficulties in all three areas but fail to meet full criteria in any of the areas, so are not diagnosed with ASD.

Teaching strategies vary based on the individual's age and ability. The focus of the treatment is often targeted at strengthening skills in individual areas of difficulty. Special education provisions and accommodations are incorporated in a child's individual education plan (IEP). This often includes the individual working with a multidisciplinary team of clinical professionals who provide several different services, including speech and language therapists, developmental and child psychologists, and pediatricians.

According to the Autism Society of America, treatment approaches include:

- Applied Behavioral Analysis (ABA) and Discrete Trial Training
- Treatment of Autistic and Related Communication Handicapped Children (TEACCH)
- Picture Exchange Communication System (PECS)

***ABA and Discrete Trial Training*** is often used interchangeably. These methods include intense repetitive, structured tasks in which good behavior is rewarded and undesirable behavior is ignored. It is time-intensive and focuses on changing current behaviors and does not prepare individuals to respond in new situations. Some individuals with TSC who have ASD have significantly benefited from ABA programs.

***TEACCH*** (Treatment of Autistic and Related Communication Handicapped Children) was developed at the University of North Carolina. TEACCH focuses on adapting the environment to the individual with ASD instead of trying to make the individual adapt to the environment. This is achieved through high structure, organizational charts and schedules. While many favor this approach, some feel it is too structured and makes the individual too dependent on charts and other organizational tools.

**PECS** (Picture Exchange Communication System) is used to encourage communication. By using pictures, an individual can point to or hand an object to someone to demonstrate what he or she wants.

Options vary and the treatment program needs to be tailored to the individual's age and ability. Treatment is targeted at fostering skills in the three main areas of difficulty: social and communication skills and the development of imaginative play. In addition, treatment aims to ensure that repetitive or obsessive behaviors do not become too marked or prominent and do not interfere with family life. Lastly, the treatment aims to help parents foster their child's development and support him/her during the early, often very demanding, years.

## **Mental Health Issues**

Individuals with TSC almost always exhibit at least one mental health complication, and some display multiple mental health issues. The seriousness of these issues varies from one individual with TSC to another.

### ***Anxiety***

Individuals with TSC experience a higher rate of anxiety than others. This anxiety can manifest itself into forms of excessive worrying, sporadic behavior, and unexplained panic attacks. Students who have gone to school without problems through elementary school can suddenly develop school phobia in middle school. The effects of this disorder can be devastating and seem to be more challenging as the student gets older.

### ***Aggressiveness***

Students who are severely affected by TSC often have outbursts and behaviors that cause self-injury. The successes to treating these behaviors are variable because they are used by trial and error. As the student gets older, these behaviors seem to decrease but are replaced with depression or anxiety. Medication can also cause aggressive behaviors in students. Many children with TSC act differently when starting new medications or changes in the dosage of medication. Medication issues are the first thing to rule out when trying to do a functional behavior assessment (FBA) on a student with TSC.

## ***Depression***

Individuals with TSC frequently have depression and coexisting mental health issues like anxiety. Many times, because a student may have coexisting mental health conditions, he/she is misdiagnosed. The depression can mask the anxiety or the anxiety can mask the depression. Other mental health issues such as aggression may be the result of the child being depressed.

## ***Obsessive Compulsive Disorder***

Many children with TSC will develop Obsessive Compulsive Disorder (OCD). OCD is an anxiety disorder that causes unwanted thoughts and/or repetitive behaviors. Repetitive behaviors such as hand washing, counting, checking, or cleaning, are done to control the unwanted thoughts in an effort to make them go away. This type of behavior is sometimes called “rituals.” These rituals provide temporary relief but cause extreme anxiety when they can not be preformed.

Because of the extreme negative influence, TSC-related behavioral issues can have on students with TSC, early identification and behavioral interventions are recommended to decrease the impact on learning. Setting up an IEP with a positive behavior intervention plan will be beneficial in educational success. Communicating and working with the student’s family, mental health professional and medical professional will also help impact educational success.

## **Other Health Impairments**

### ***Kidney***

Renal (kidney) angiomyolipomas (AMLs) are non-cancerous tumors and are the most common type of kidney tumors in TSC. AMLs occur in 70 to 80 percent of older children with TSC. These tumors begin to grow in childhood in many individuals with TSC, but usually grow very slowly and may not be problematic until young adulthood. When AMLs become larger than 4cm they can cause increase in blood pressure, cause back pain, and blood in the urine.

Some individuals with TSC will also have polycystic kidney disease (PKD). PKD occurs when kidneys have multiple cysts. These cysts grow and multiply over time, also causing the mass of the kidney to increase. Ultimately the diseased kidney shuts down, causing end-stage kidney disease for which dialysis and transplantation are the only forms of treatment.

### ***Heart***

Cardiac (heart) rhabdomyomas (non-cancerous tumors) usually form in the heart of infants with TSC and are at their largest size at birth. The incidence of these tumors in TSC has been reported to vary from 47 to 67 percent. A vast majority of cardiac tumors shrink and essentially disappear, but some individuals with TSC will have long-term heart rhythm problems that will need to be monitored throughout their lives. This can interfere with physical education classes, playground activities, and interaction with peers.

### ***Eye***

Benign tumors and depigmented patches can occur inside the eyes of individuals with TSC. Most of the time, these tumors and patches do not cause any visual loss or problem. But, a small a percentage of individuals do experience visual difficulties, so accommodations should be addressed on the student's IEP.

### ***Lung***

Lung involvement can sometimes occur in teenagers and is far more common in girls than in boys. Some young women who have lung involvement due to TSC may have lymphangiomyomatosis (LAM).

The first symptoms of lung involvement in an individual with TSC may be shortness of breath after mild exercise, cough, or spontaneous pneumothorax (a collection of air or gas in the chest causing the lung to collapse). Progression of such lung involvement to pulmonary failure can sometimes occur, and some individuals may require a lung transplant. Recent studies have shown that around 40 percent of women with TSC have LAM.

## *Teeth*

Oral involvement in TSC can include gum fibromas (tumors) and dental pits. The tumors appear as overgrowth of the gums and can be quite extensive. Dental pits can be observed in both primary and adult teeth; the incidence in individuals who are 11 years old and older is 100 percent, while it is 76 percent in individuals younger than 11. The pits are seen in both the front and back surfaces of the teeth, which are areas that do not normally develop cavities.

## *Skin*

Skin issues resulting from TSC include the following:

- **Hypomelanotic macules** – flat areas of skin that appear lighter than the surrounding skin. They can be any size or shape or may look like an ash-leaf. The skin cells in this area of the skin contain less pigment, so the area appears lighter than the surrounding skin.
- **Shagreen patch** – a patch of skin that is similar in color to surrounding skin but may be tough and dimpled like an orange peel. The shagreen patch is usually found on the lower back and nape of the neck but they may also be seen on other parts of the body.
- **Periungual or subungual fibromas** – small fibrous growths that appear around the fingernails or toenails. These growths can interfere with writing and small motor activities. Sometimes when they are on the toenails, they can cause so much pain that walking can be difficult.
- **Facial angiofibromas** – benign tumors of the face that often appear across the cheek and nose and on the chin. They are initially small reddish spots or bumps that may increase in size with age. Facial angiofibromas are rarely present at birth, but often appear as the child reaches 4 or 5 years of age or older. Adolescent students will be affected the most with angiofibromas because of peer pressure. Self esteem, depression, and anxiety can be side effects from this particular symptom.

- **Forehead plaque** – flesh-colored plaques that are often soft or compressible or doughy to hard lesions.

### **Support from the TS Alliance**

The TS Alliance offers a free, private online discussion group/listserv for professionals from a variety of fields in TSC research, treatment and education, which is not publicly accessible. Members are individually approved for subscription by the TS Alliance. Educators are welcome to join TS Alliance staff members and others who share information via this forum.

The online discussion group allows you to ask questions about TSC for doctors, researcher, and other educators who have experience dealing with TSC.

To join the Professional Online Discussion Group:

1. Go to [www.tsalliance.org](http://www.tsalliance.org)
2. At the top of the page, click on the button that says “Online Communities”
3. Once you choose “Online Communities,” a menu will drop down – then click on “Discussion Groups”
4. Look for the “TSC Research, Treatment, & Education Professionals” group
5. Then just follow the directions to join the group

If you would like to talk with someone at the TS Alliance, please contact:

Dena Hook  
Director of Advocacy & Education  
1-800-255-6872  
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