

Support, Educate, Empower

Sturge-Weber Syndrome In the Elementary School Classroom



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The Sturge-Weber Foundation

Care. Connect. Collaborate

Dear Parents, Teachers and School Faculty,

When unforeseen challenges are presented to a teacher or the school faculty, such as needing to provide the staff and students with insight into a certain disease, it can be discouraging when the resources are not readily available. This packet is intended to offer educators the tools to implement Sturge-Weber syndrome education into the classroom.

What is appropriate for the students in a classroom is of course dependent first and foremost upon the child's level of openness. It is important to let the child lead at a comfortable pace. It is crucial to consider what struggles are going on with your student in particular; with Sturge-Weber syndrome, the personal challenges vary.

Also, I would suggest that you implement any discussion you do on Sturge-Weber syndrome or self-esteem and difference into the classrooms of students older and younger than their peer with Sturge-Weber syndrome. With a year more of maturity and sensitivity, the older students will serve as advocates to their younger friend. The students below will learn a valuable lesson in humanity, and will use their innocence to ask important and poignant questions regarding Sturge-Weber syndrome.

Please accept the enclosed material as the first step to supporting, educating and empowering your students. As the manifestations of Sturge-Weber syndrome may change over time, so may the needs of your student. I invite you to contact me, should you see the need for additional information and thank you for your dedication and concern.

Sincerely,

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**Back to School -
Advice, Tips and Suggestions From the School Nurse's Perspective
By Lynne Siegel, RN, who has been on "both sides of the fence".**

The school nurse is a key member of the team you will be working with during your child's school career.

Classroom teachers may change every year, but usually the school nurse will know your child for his whole school experience. So it is important to establish communication early and maintain it often.

Phone calls really aren't as effective as seeing the parent and child in person. You don't need an appointment to stop by at the beginning of the school year to visit your school nurse.

In my role as a school nurse I am able to discuss things like the medications and the appropriately filled out forms and properly labeled bottles at the beginning of the year with all of my parent "visitors", saving time for them and myself by not having to toss the paperwork back and forth between the doctor's office and the parent, etc. And I love getting to know the parents in a non-emergency way.

Ask for the written policies of your school district on these issues and you will save yourself tons of time and aggravation. Nurses sometimes work with an assistant called a "health room technician" although different states may have a different name for the same position. They are there EVERY day while some nurses have to visit several school health rooms every week. Find out from the school secretary what days the RN will be stationed in your child's school.

There are things called "pre-printed care plans" that the nurse keeps in each child's medical folder. They are general for all medical problems or seizure types. If you want the specifics added to your child's care plan (and they should be because every kid's needs are different), make sure you see it and check it over in person as often as you need to.

(For instance, standard protocol for seizures is usually to call 911, stay with the student and administer Diastat if there is a doctor's order.)

AND.....this is a biggy...make sure you put copies of as many tests, clinical notes and doctor's reports as possible in your child's record - make sure you are present when they get filed in the chart. Make copies for the teachers so everyone can be on the same page.

If you can, have your doctor write educational recommendations like "no contact sports" or "extra time for testing". These will support your demands for reasonable accommodations. The more documentation, especially neuro/psych/educational - the better prepared you will be to pursue school staff support.

Make sure the RN is present in the IEP meeting (or at least try to invite her in advance of the meeting, the school staff might not always remember). Sometimes the nurse is so busy or is absent, but she can still send a written assessment to the meeting. Her input is very important.

If she can make it to the meeting, make sure the medical accommodations are specific. For example, "due to the nature of the medications, the child will need a general pass to go to the bathroom as needed" or "to prevent dehydration, the child may bring a water bottle to class".

If you need to keep non-aspirin (Tylenol) in the health room to use as needed (spelled out as PRN in the records) for aches and pains, you will need a doctor's written orders even for over-the-counter preparations. By the way, you cannot just send medications on the bus to be delivered to the health room. Check the rules of each school district.

Before I was a school nurse, I did not know that the medical documentation would have so much impact when used to gain more in the way of services for my son. Had I only asked the neurologist at that time to refer my son for neuro-psych educational testing! We all thought everything was fine and the teachers were criticizing him for being "difficult". Testing revealed that he will never be able to keyboard with both hands and all fingers. The teachers thought he was copping out. I thought so too. Would you believe I finally figured all of this out AFTER I became a school nurse!

You will probably have to remind the school staff to review your child's medical records right before the IEP meeting. Do not settle for the IEP being put off. Document your requests in writing and your child will not be placed at the end of the list.

Of course when your child leaves high school it is like starting all over again, only colleges will not make modifications based on need and your request. They make them based on medical recommendations IN WRITING from an educational tester and a doctor. Each higher education institution has its own requirements; they are not part of the public school system that has known your child all along. And as your child gets older, there are legal issues of confidentiality that arise once a son or daughter is over 18. When my son started community college they wouldn't use all the stuff I had for him when he was 16 in high school.

When I was younger and my son was starting school, I just trusted that the school would "do what had to be done" after I signed the releases for the medical information to be sent from the doctor.

If I knew then what I know now, I would have had every test from every specialist from kindergarten to senior year in high school documented and included in the record.

I know how hard this is and how much time it takes, but if you get started early and keep adding doctors' letters to the medical record, the school will take a more serious interest in your child's educational needs.

One more thing, this time from the mom, not the school nurse. My kid liked going to the health room, to hide out or get what he would call "sweet pity" from the health room technician. So there was a lot of fabricating and exaggerating illnesses, bumps and bruises, in order to get out of doing things. The pediatrician and I thought he looked great neurologically and functionally. He was seizure free from age 6 through age 16 and no deficits were seen outwardly.

But guess what? There are deficits you cannot see, and they were not completely related to his vision problems. With avoidance behaviors, he could avoid the things that were much more difficult for him than any of us knew!

And he could avoid the teasing by charming the health room tech into letting him hang out during lunch or PE. Middle school seemed to be the worst as far as teasing. In retrospect, I should have pursued the problem more energetically. It really pays to be a squeaky wheel when it comes to your kid's health issues in the school.

Lynne Siegel, RN is the mom of an adult son with SWS. She is a Community Health Nurse II working in school health services of the Montgomery County, MD, Department of Health and Human Services.

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Sample letter to be sent to the Teacher & Staff

Dear Teachers and Staff,

Our son/daughter is starting kindergarten this fall. He/she has been diagnosed with a condition called Sturge-Weber syndrome (SWS). Sturge-Weber syndrome is a highly unpredictable, progressive syndrome that can cause glaucoma, seizures and developmental disability. Of the one million individuals with a Port Wine Stain, SWS affects 8-16 %. Those who develop a seizure history in infancy usually have a poorer prognosis. Based on seizure history and brain involvement, in a physical sense, Sturge-Weber syndrome can be disabling. In an emotional sense, it can be challenging, especially for those with a facial port wine stain.

We have worked very hard with (child's name) to encourage a healthy self-esteem, however, he/she does experience hurt feelings from strangers' remarks and comments. This is where we would like to ask for your help! Knowledge is power and if his/her schoolmates are aware of (child's name)'s story, we believe that the teasing can be kept to a minimum. We would appreciate any help you can give in the education of others about Sturge-Weber syndrome. We would also like to know of any teasing of (child's name) so that we can help him/her deal with it.

(child's name) has already gone through (list treatments/surgeries); there is no cure for Sturge-Weber syndrome. He/she currently taking (list medications and reason). Please feel free to contact us with any questions, suggestions or comments.

Thank you for your help and support,

Sample letter to be sent to the parents of classmates, or published in an open letter in the school newsletter. It is recommended that you go through the appropriate channels before sending this; approval may be needed from the school's principal. Please edit this letter as you see fit based on your child's manifestations and treatments.

To the parents of all students at (name of school),

You and your family are probably as excited as we are about the start of school. Our son/daughter, (child's name), will be attending (school name) this year in the (grade). He/she is eager for school to start, and looking forward to renewing friendships and making new friends. As his/her parents, we share his/her enthusiasm, but we also feel very anxious about a unique situation with (child's name), and that's why we are writing to all the parents with children at (school name).

As the result of Sturge-Weber syndrome (child's name) has (a facial port wine stain, seizures and/or glaucoma). We would really appreciate it if you would take a few moments now to read this important letter and be sure to share this information with your family before school begins. While (child's name)'s seizures have been managed for (some time, one year, etc.), there is the possibility that a seizure can occur. In addition, he/she may have laser treatments for his/her port wine stain, which can result in "purple dots" on his/her face. These things can be visibly quite shocking, if you're not used to them.

Sturge-Weber syndrome is a poorly understood brain and skin disorder—currently there is no cure. You can't "catch" Sturge-Weber syndrome; it is not contagious. Sturge-Weber syndrome affects both sexes, and all ages. Sturge-Weber syndrome is a rare syndrome, of the one million individuals with a Port Wine Stain, SWS affects 8-16 %. The course of SWS is highly unpredictable; seizure activity and/or glaucoma may develop at any time.

Depending on the manifestations, Sturge-Weber syndrome can be medically disabling, as you can imagine, it can be very challenging emotionally. The emotional pain can be overcome with one's own inner resources and the support of others. As you know, beginning around age six children are becoming much more concerned about how others view them, how they may differ from others, and whether others might be making fun of them. Since children at this age have become so aware of individual differences, they unfortunately can sometimes be disposed to poke fun at those who don't fit their definitions of "normal." One of the most frequent problems of teasing described by children with Sturge-Weber syndrome is name calling due to their port wine stain. One of the reasons we have chosen (school name) for (child's name) is because of the school's firm commitment to teaching and upholding high standards of individual respect and dignity. We hope you feel the same way, and share our commitment, and that (child's name) will not endure ostracism or suffer any shameful or humiliating experiences. We really need your assistance and are asking you to discuss (child's name)'s situation with your child, and to point out and emphasize standards of behavior consistent with mutual respect.

Like all parents, we're eager to help make (child's name)'s school experience as positive and successful as possible. We hope you find this letter helpful in understanding her particular circumstance, and that it will help you talk with your child about it before school begins. We are aware that this will be a new and different situation for your child to grasp. Like any new behavior pattern, it may need reinforcement throughout the school year and we hope we can count on all the families at (school name) to understand this.

If we can answer any further questions, please don't hesitate to call us. In advance, thank you for your help and support.

Sincerely,

Self Esteem and Students *(Adapted from Teacher Talk, Indiana University)*

The school year can pose a particular problem for children who have Sturge-Weber syndrome. It is a time of life when school-age children and adolescents are especially sensitive about their appearance. An individual's school experience can have a profound effect on the socialization process. Since most children are unfamiliar with Sturge-Weber syndrome, many children are uncomfortable with interactions. Teachers can help the child with Sturge-Weber syndrome to cope and can help peers to be sensitive and understanding. How you feel about yourself is, in part, a reflection of how you think others view you. As children grow and develop a greater sense of who they are, it is important that acceptance and integration exist for them. There are various factors that shape and affect self-esteem:

* The respect, acceptance and concern of significant others. "Significant others" change from family members for the very young child, to teachers for the school-age child, to peers for the adolescent. Giving others the opportunity to ask questions and to voice their concerns develops empathy, understanding and ultimately, acceptance.

* A history of successes. By encouraging recognition of the positives, children can form a basis in reality for self-esteem. Children can be "steered" towards experiences that are likely to be successful, in order to enhance a positive feeling of self-worth.

* Values and aspirations. The more successes we have, the higher our aspirations and expectancies of what we can achieve. People who have overcome their physical limitations can provide positive role models. Encourage the child's expression of thoughts feelings and dreams.

* Problem solving. Teaching problem solving skills increases the likelihood that a child will feel a sense of control over life. Difficult situations can be viewed as learning experiences, rather than as a devaluation of self.

These ideas may be used as a starting point by parents and teachers inside the classroom and in the community. Most important is that the focus be creative and interactive. Remember that each child is an individual.

Strategies for teachers to use to help raise student esteem

- Use student names
- Have conversations with every student
- Provide multiple ways for students to be successful in your class
- Display student work
- Give each student a responsibility in the classroom
- Provide opportunities for student work to be judged by external audiences
- Take time to point out positive aspects of your students' work
- Never criticize a student's question
- Take time to help struggling students understand the material
- Try to get to know about the student's life outside of school
- Ask students about their other activities (ex. "How was the soccer game, Natalie?")
- Help students turn failure into positive learning experience
- Encourage students to take risks
- Provide opportunities for students to make their own decisions about certain aspects of your class
- Provide opportunities for students to work with each other
- Don't make assumptions about student behavior
- Allow students to suffer the consequences of their behavior-don't be overprotective
- Allow students to explore options in different situations
- Celebrate your student's achievements, no matter how small

Seizure Observation

Name of Person: _____

Date Seizure Occurred: _____ Time: _____

Duration seizure activity: _____ Duration recovery period: _____

1. Activity prior to seizure: _____

2. Describe what you did to assure health & safety: _____

3. Did the person recall any sensory experience (i.e. bad odor, tingling, feeling of fear, etc) before seizure? _____

4. Describe person's activity following seizure (confused, sleepy, dazed, etc.): _____

Check All That Apply:

Aura

- yelled
- action (describe)

Breathing

- noisy
- shallow
- stopped (how long)

Eyes

- pupils constricted dilated
- turned right turned left
- rolled up
- stared straight ahead

Movement

- jerked-(circle) whole body
- r arm, r leg, l arm, l leg
- limp
- jackknife
- purposeful movement

Color

- flushed
- pale
- bluish
- bluish around mouth

Mouth

- salivated
- chewed
- smacked lips
- cried
- talked/mumbled

Muscle Tone

- rigid-(circle) whole body
- r arm, r leg, l arm, l leg
- limp
- fell down

Sphincters

- urinated
- defecated

Mental State

- unchanged
- vacant
- unresponsive to commands
- unconscious

Describe any apparent injuries or information not listed above _____

A GUIDE FOR SCHOOLS

Sturge-Weber Syndrome (SWS) is a congenital, non-familial orphan disease of unknown cause, incidence, or prevalence. It is characterized by a facial birthmark and neurological abnormalities. Other symptoms associated with SWS can include eye and internal organ irregularities. Each case of SWS is unique and exhibits the distinctive findings to varying Degrees.

Due to the prominent facial birthmark, some children become embarrassed and experience a drop in self-esteem. Depression is not an unusual response to a curious public. Educators need to be aware of the psychological impact port wine stains and SWS play on a child's life. As an educator, you are on a daily basis reaching children in their most formative years. Because of documented studies linking impaired appearance with low self-esteem, you can be a positive influence on the child (with SWS) and his classmates.

60 percent of parents and 73 percent of teachers report a number of behaviors among SWS kids that result in poor academic performance. These behaviors may be related to cognitive impairments, attention problems, seizures, anticonvulsant medications and psychological factors.

Seizures typically occur within the first to two years of life but have been known to have a later onset. The seizures vary from child to child but children with SWS usually have focal seizures, infantile spasms, or partial seizures. They exhibit these seizures in a variety of ways: staring spells, twitches, jerks, loss of muscle tone, turning of heads to the right or left, etc.

Seizures are treated with anticonvulsant medications. Keep in mind a child with SWS may have to be excused from class to take medication. If a seizure does occur while the child is in class, try to make the child comfortable on his side and free from harmful objects. Call the school nurse and parents to notify them of this occurrence. After the seizure, it is important that you briefly discuss the event and answer any questions the students may have! Resume your normal routine as soon as possible. It would be prudent to point out to the students that seizures are only one aspect of the child with SWS. Remind classmates of the child's (with SWS) other special qualities.

Glaucoma is simply an increase in fluid in the eye causing pressure which left untreated can cause the eye to go blind. Glaucoma can occur anytime in a child with SWS.

Glaucoma is treated with eye drops and eye surgery. The child with SWS may wear glasses as a result of loss of vision due to the glaucoma.

Facial Birthmarks can be treated with a laser to remove the port wine stain beneath the skin. Some children prefer to wear make-up and others choose to do nothing. It is important that there is open communication when a child under goes laser treatments. It can take years of repeated laser treatments to remove or significantly lighten a child's birthmark.



After a laser treatment, the treated area will have purple dots. The area may have to be bandaged or have some antibiotic ointment. It should not be exposed to the sun-light. Your sensitivity will carry over to the child's classmates during this time. With understanding, most children will react with a caring attitude towards the child under-going laser treatment. For further questions on any of these medical aspects of SWS, contact the parents or the Sturge-Weber Foundation.

"Names can never hurt me?" Children of all ages are sensitive to teasing and name-calling. Yet, at this age many children can virtually forget they have a birthmark in their eagerness to learn and participate. Unlike teenagers, young children have not quite become focused on appearance. As a teacher, you can support children with SWS by creating a classroom with only constructive comments, kind and thoughtful interaction, and one that does not tolerate insults.

Use any incidents of teasing to educate the "teaser" and comfort the "victim." Children with SWS need reassurance that the significant adults in their lives will support them, regardless of appearance.

Some children and parents opt to forego laser or treat later in life when the child can have input into the laser treatment. Another tool at their disposal is cosmetic cover make-up. There are various types of make-up. Some parents feel make-up is not for children. When a child is unhappy or embarrassed, any attempt to restore self-esteem should be viewed in a positive light. The decision to do laser and/or make-up should involve the child's input when possible. Some children with SWS have high self-esteem and feel special with their birthmark and do not want any alterations done to the birthmark. Again, listen to the child.

It is important to give children with SWS special, but not undue attention when they find themselves in new situations: the first few weeks of school, field trips, etc. As time passes, children with SWS usually relax and make friends but incidents may still occur where reassurance is needed.

Highlight the child's strong points as you do all children. Self-esteem is boosted for all of us by praise for a job well done or a skill mastered.

Flexibility with regard to your own rules is sometimes needed. Children with SWS want to be treated as you would any other child - so be judicious in your flexibility and special attention. As an educator, you have an instinct for the uniqueness in each child. Use that skill to determine if a child with SWS is shy, rambunctious, or outgoing. Then, tailor make your teaching and "teachable moments" for that particular child.

In cases of SWS, here are a few guidelines to remember:

- Give support to the child with SWS by your example and your teaching. If you avoid the touch of a child with SWS, you should not be surprised to see other pupils act accordingly.
- Do not ignore teasing or name-calling from other pupils. Intervene in situations where the child's self-esteem is being harmed. Integrate the child into group activities.
- Parents and teachers should be partners! Let parents know how the child is adjusting, the strengths and weaknesses you see in the child. If parents are unaware of laser treatments or make-up, inform them of these options.
- Regular eye and glaucoma exams should be done to assure adequate pressure control. Communicate closely with parents to keep abreast of other medical changes that may occur.
- Refer any student with SWS to a counselor or therapist if they seem troubled. Medical and physical challenges can cause psychological scarring which if caught early on can be healed.

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Epilepsy in the Classroom....The Teacher's Role

When a seizure actually occurs, it is a frightening experience, especially with a child in a classroom. The strangeness of the unusual behavior or the dramatic suddenness of a convulsion is upsetting, plus the teacher may be afraid for the child's welfare and/or may themselves feel vulnerable. Unless the occurrence of a seizure is handled properly, the fear generated may be translated into fear of the child who had the seizure. This progression can cause the child with the disorder to be shunned, teased, or both.

Communication: An understanding of epilepsy can be obtained through communication. As soon as possible following a seizure episode, an explanation by the teacher, school nurse or principal should take place properly explaining to other children what happened, answering questions, giving them a chance to discuss openly what they were feeling. All this can reduce the social impact on the child with the seizure disorder. Key Points to communicate to assist children understand epilepsy are:

- What happened to the child is called a seizure.
- It happened because just for a minute or two the child's brain did not work properly and sent mixed up messages to the rest of the body. Now that the seizure is over, the child's brain and body are working properly.
- Having seizures is part of a health condition called epilepsy, which some children have. It is not a disease and cannot be caught from other children.
- Children who have this condition take medicine to prevent seizures, but sometimes one happens anyway.

The teacher should emphasize that the child was not in any danger even though it looked as if he was, and the child does not have a mental illness.

Avoiding Overprotection

Another major problem for children with epilepsy is the good intentions of adults to protect them from harm. People tend to limit or even eliminate a child's participation in normal childhood activities for fear that a seizure will occur. Vigorous physical activity is generally not associated with a greater number of seizures. In fact, studies suggest fewer seizures will occur when the child is active.

When the child is excluded from experiences due to a seizure disorder, he misses opportunities to develop social skills and self-confidence. The sense of being different, of being unable to join what others are doing, encourages dependence in the child and may keep him socially immature.

Whenever possible, the child should be encouraged to take part in all school activities. This will offer an opportunity to break the tendency to overprotect the child and instead will provide the child a healthy social and academic environment.



EPILEPSY AND LEARNING

The range of intelligence among children with epilepsy is the same as from the rest of the population. The majority of affected children will experience little or no learning difficulty. However, teacher surveys indicate that children with epilepsy have twice as many problems – such as lack of concentration, restlessness, and fidgeting – as their classmates. Some will have learning difficulties. These may stem from undiagnosed and subtle learning disabilities, from the psychological and behavioral problems that often accompany the condition, from mild or severe retardation, or from under or over medication.

Emphasis should be given that learning difficulties, when they exist, are not as much the result of the seizures (unless they are very frequent) as they are the result of the brain damage that causes both the seizures and the learning disabilities. Determining the underlying cause can be complicated by the fact that learning disorders may also be behavioral rather due primarily to the seizures or brain damage. This behavior may be the result of reactions by the child and by society to epilepsy. If this is the case, the solution may be very different from special educational endeavors, and could include psychological counseling for the child, family counseling or other procedures for modifying inappropriate individual and societal attitudes toward the condition.

Some children with epilepsy need special help at school but they do not need walls. They should integrate freely into the social and academic life of the school. Even if a child has a seizure, it is only a minute or two out of his school experience. At other times most kids with epilepsy are perfectly normal and healthy and there should be no difference in the way they are treated.

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FUNCTIONING IN THE ACADEMIC SETTING

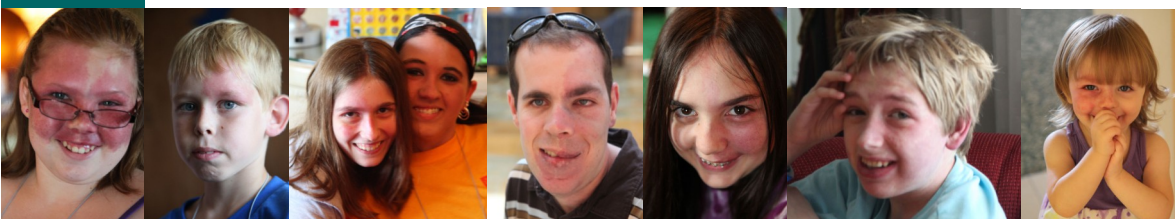
In a recent survey, 65% of parents of children with Sturge-Weber reported that their children were receiving special education services. 70% of the parents surveyed also reported that they were at least moderately concerned about school problems. Knowing that a student is receiving special education services tells us little about the underlying basis for that student's academic problems. This is particularly true for students with Sturge-Weber because the disorder is commonly associated with a number of other conditions that may impact learning. These conditions include impairments of intellectual functioning, seizures and Attention-Deficit/Hyperactivity Disorder. Each of these associated disorders and their implications for academic performance will be discussed in this section.

Mental Retardation and Learning Disabilities

Approximately 50 to 60% of individuals with Sturge-Weber syndrome also have mental retardation. The range of intellectual functioning in this group of individuals, however, appears to be quite wide. In one series of 32 young people with the disorder, assessed intelligence ranged from the moderately retarded to superior. There are a number of factors that appear to place individuals with Sturge-Weber at risk for lower levels of intellectual functioning, including a history of seizure disorder and higher frequency of seizures. Bilateral interictal EEG abnormalities and smaller hemispheric asymmetries in cortical metabolism have also been associated with lower levels of intellectual functioning.

Neither intelligence nor mental retardation has been easy to define. The history of psychology has been filled with debates about the definition of intelligence. One solution to the quandary has been to define intelligence as what intelligence tests measure. Whatever one considers intelligence to be, performance on intelligence tests is one of the strongest predictors of school performance. Intelligence tests assess many different cognitive abilities, such as language ability, memory, problem-solving and abstract thinking, and all of these abilities are essential for the acquisition of academic skills.

The most widely used definition of mental retardation is that proposed by the American Association on Mental Deficiency. A diagnosis of mental retardation is made when three criteria are met. First, the individual must perform two standard deviations below the mean for his or her age group on a measure of intelligence, producing an IQ of less than 70. Second, the individual's adaptive behavior must be significantly below age-expected levels. Adaptive behavior is defined as the demonstration of expected levels of personal independence and social skill. Finally, the cognitive and behavioral impairments must be apparent before the age of 18. The first criterion has the most direct implications for academic performance. An IQ of less than 70, in most cases, is an indication that the individual demonstrates significant deficiencies in all or most of the cognitive abilities assessed by the test of intelligence. Because those cognitive abilities are important for the development of reading, writing and math skills, the deficiencies present a major handicap for the student. Even within the range of mental retardation, IQ are is with academic progress.



Those individuals who are assessed to fall within the mild range of mental retardation may develop some functional academic skills while those who fall within the severe or profound range will very likely make little or no substantive academic progress. In all cases of mental retardation, a good school program is important for facilitating the development of adaptive skills.

Developmental problems can be limited to one or a limited number of cognitive abilities, rather than the global involvement that is characteristic of mental retardation. These more circumscribed cognitive impairment typically result in specific learning disabilities. For example, some students only exhibit deficiencies in some aspect of language functioning and this limitation results in reading and spelling disabilities. At the present time, there is no information available on the incidence of specific learning disabilities in individuals with SWS. Because individuals with epilepsy and many other neurological conditions are at increased risk for specific cognitive problems and learning disabilities it seems quite likely that those individuals with Sturge-Weber who have higher levels of intellectual functioning are at increased risk as well.

School districts offer cost-free assessments for young children with developmental problems and students with poor academic performance. Private evaluations can sometimes be more helpful than the psychoeducational evaluations conducted by the school. A neuropsychological assessment, for example, will typically provide a more in depth assessment of the specific cognitive processes than underlie learning. The more detailed characterization of the student's cognitive profile can guide the design of an educational plan that utilizes cognitive strengths to help the student compensate for cognitive weaknesses.

The Impact of Seizures and their Treatment on Learning

Frequent daytime seizures can interfere with a student's ability to take full advantage of his or her educational program. Learning clearly does not take place during a seizure but disrupted learning may be less obvious when the brain continues to process information inefficiently for a period of time afterwards, even though the student appears to have returned to baseline. Students who are having frequent seizures may spend a major part of their school day in a mental state that does not allow them to comprehend or remember material at their normal level. Fortunately, most of the young people who have seizure disorders secondary to SWS experience relatively good seizure control. For those who do not, care should be taken not to present new material or assess learning for several hours after a major seizure.

Anticonvulsant medications can improve learning by controlling seizures but, at the same time, they may produce side effects that impede learning. The cognitive side effects of anticonvulsants have not been easy to study because it is difficult to separate the effects of medication from many other factors, such as seizure type and frequency, that can also affect cognition. Most studies have focused on adults but the results of a smaller number of studies with children suggest that the effects are similar in both age groups. The older anticonvulsants have been fairly well studied and all of them have been shown to have the potential of causing reduced alertness, psychomotor slowing and impairments in memory. The likelihood of side effects is increased when dosages result in blood concentrations that exceed standard therapeutic levels and when medications are combined. When cognitive side effects do occur, they are usually mild. The most pronounced side effects are observed with phenobarbital, while carbamazepine, phenytoin and valproate have been found to produce less marked effects.

The newer anticonvulsants, at this point, have been less well studied. The data that do exist suggest that, in general, their use is less likely to be associated with adverse cognitive consequences. Of the new anticonvulsants, topiramate has the greatest potential for causing a decline in cognitive functioning. Patients have reported trouble generating thoughts and producing speech and one retrospective study revealed cognitive declines in patients who received topiramate as a second medication. The changes were most dramatic in verbal intelligence, verbal fluency and verbal learning. Improvements were observed in other patients when topiramate was withdrawn. Although some patients may experience these undesirable side effects, many other patients tolerate topiramate very well and it appears that the likelihood of cognitive side effects is reduced if the medication is introduced gradually.

The data from studies of the other newer anticonvulsants do not indicate that cognitive side effects should be a major concern. One small study has suggested that zonisamide can have a negative effect on verbal learning but this side effect appears to diminish over time. Gabapentin has been shown to have some potential for negatively affecting cognition but the effects in studies have been mild and very limited in scope. Studies of lamotrigine, oxcarbazepine, and tiagabine, have not provided any evidence that these medications produce any deleterious effects on cognition.

The chances are greater that an anticonvulsant will produce unacceptable changes in mood than in learning. If it appears, however, that either cognition or mood has changed following either the introduction of an anticonvulsant or an increase in dosage, consideration should be given to the possibility that the medication is responsible. Usually the only way to know for certain that the medication is to blame is to give the child or adolescent a trial off of the medication. In some cases, the only anticonvulsant that has ever provided good seizure control is found to be responsible for a child's irritability and parents are left with the horrible dilemma of deciding which is worse--seizures or a disagreeable child.

Attention Deficit/Hyperactivity Disorder

In the survey mentioned earlier, 22% of the parents of children with SWS reported that their children had been diagnosed with Attention-Deficit/Hyperactivity Disorder (ADHD). This incidence is much higher than the prevalence of 3 to 5% that is commonly reported for the general population of school-aged children. The diagnosis of ADHD is made when a child, compared to other children of the same mental age, exhibits excessive levels of inattentiveness, impulsivity and hyperactivity. A subgroup of these children is predominately inattentive although most children with the disorder demonstrate significant levels of all three behaviors.

Some young people with ADHD develop social and conduct problems but they almost all experience some level of academic problem. They are often described as underachievers. Within the classroom setting, there are numerous complaints. Teachers report that they do not listen, and they have trouble finishing their work. Because these students are impulsive, they often fail to read or listen to instructions before beginning their work. Poor grades, particularly in math, are often simply due to carelessness. They often fail to complete their homework because they do not get the correct assignment and materials home. When they do, parents have to struggle with them to get it completed and, even if the work is completed, they may not turn it in. Their impulsivity and hyperactivity may also disrupt the classroom.

The only treatments for ADHD with demonstrated effectiveness are behavioral and pharmacological. Behavioral interventions involve increasing structure at home and in school. For example, students with ADHD should be seated close to the teacher. Attention and performance should be frequently monitored. The child should be asked to repeat instructions to ensure that he or she has heard them. Longer assignments or tasks at home should be broken down into shorter, more manageable segments. The child should be helped to check their work for careless errors. At school, a homework assignment sheet, checked by the teacher for accuracy, is recommended to ensure that the child's parents are always aware of their homework responsibilities. In both home and school settings, clearly communicated behavioral expectations and consistent limit setting are important as well.

Approximately 70% of children with ADHD will have a positive response to stimulant medications such as methylphenidate or D,L-amphetamine. The stimulants are effective in improving attention and impulsive control and reducing motoric overactivity. The stimulants can improve school performance although learning disabilities are not responsive to the medication and require a separate intervention. The most commonly reported side effects are appetite suppression and insomnia. Some children may also develop irritability. Tricyclic antidepressants, such as imipramine, have been used to treat children with ADHD who have either not responded to treatment with stimulant medication or are unable to take them because of undesirable side effects. The tricyclic antidepressants have been shown to decrease impulsivity and hyperactivity but, unfortunately, do not appear to improve attention.

Sturge-Weber Foundation Fact Sheets are intended to provide basic information about SWS, KT and/or PWS. They are not intended to, nor do they, constitute medical or other advice. Readers are warned not to take any action with regard to medical treatment without first consulting a physician. The SWF does not promote or recommend any treatment, therapy, institution or health care plan.