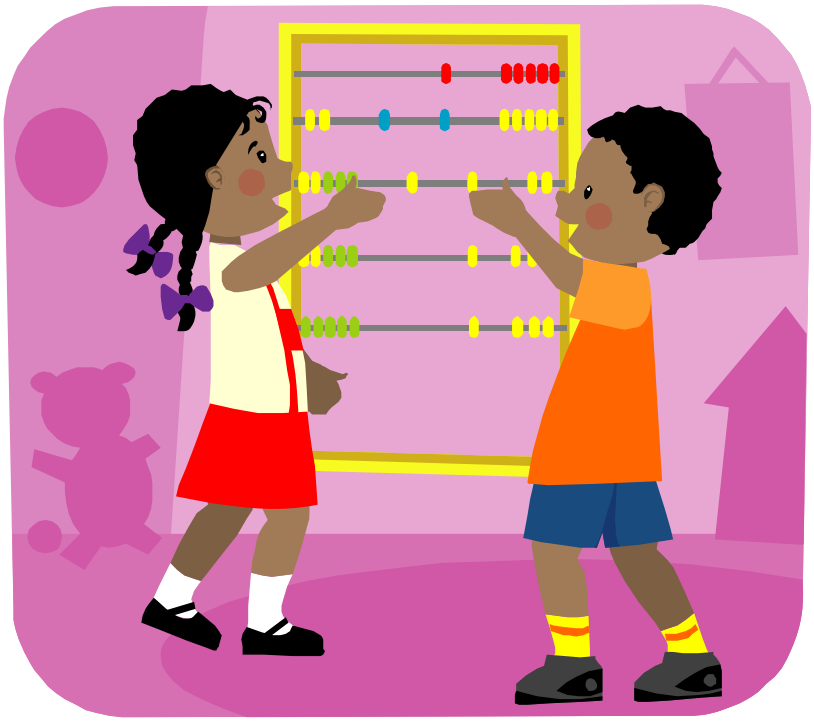
Sturge-Weber Syndrome in the Daycare Setting



The Sturge-Weber Foundation
www.sturge-weber.org

Sample Letter for Daycare Provider

Dear Caregiver,

Our child has been diagnosed with something called Sturge-Weber syndrome (SWS). Sturge-Weber syndrome is a highly unpredictable, progressive syndrome that can cause glaucoma, seizures and developmental disability, although not all patients have the same or all of these manifestations. 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.

If child is older and/or teasing is a concern:

We have worked very hard with (child's name) to encourage a healthy self-esteem, however, he/she does experience hurt feelings from stranger's 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.

If seizures are a concern:

There are many types of seizures, and children with SWS may present with more than one type. The seizure threshold may be lowered by cutting teeth, immunizations, illness, etc. To learn more about recognizing the signs of seizures, please see "Recognizing the Signs of Seizures" contained in this packet. It is important to record anything that may be a seizure on the enclosed "Seizure Observation" form and contact us immediately.

(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,

P.S. to learn more about SWS, please visit www.sturge-weber.org

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 _____

_____

The Sturge-Weber Foundation

Types of Seizures
During a seizure, the nerve cells leave their normal activities, and fire in massive, Synchronized bursts. After a few seconds or minutes, the brain regains control and the person returns to normal.

There are many different types of seizures, therefore, it is important to distinguish among the different types of seizures because they require different medications, and have different causes and outcomes. Some of the most common types of seizures are noted below. Each has a new name and an old name (in parenthesis).

Simple Partial Seizures (Focal Cortical Seizures)
Simple partial seizures result from epileptic activity which is localized in one part of the brain, usually the cortex or limbic system. Consciousness is not impaired, and children can talk and answer questions. They will remember what went on during the seizure. These seizures take different forms in different children:

Sensory: Some simple partial seizures consist of a sensory experience. The child might see lights, hear a buzzing sound or feel tingling or numbness in a part of the body. These Seizures are sometimes called Jacksonian sensory seizures.

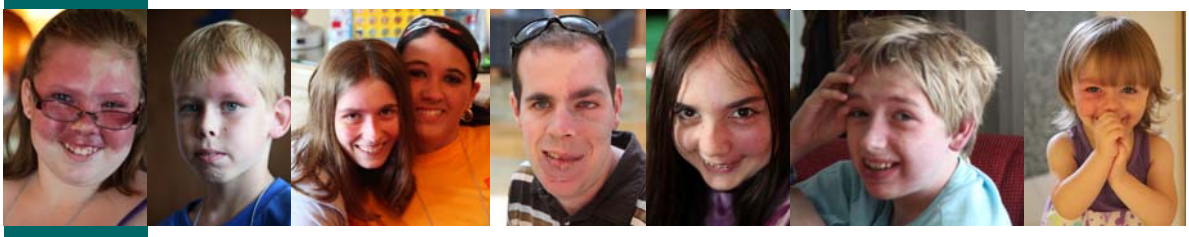
Motor: Other simple partial seizures consist of convulsive movements, which are generally clonic (jerking). Jerking typically begins in one area of the body: the face, arm, leg, or trunk. Jerking may then spread from that part to other parts of the body. These seizures are sometimes called Jacksonian motor seizures and the spread is called a Jacksonian march. It cannot be stopped.

Emotional and Other: Simple partial seizures which arise in or near the temporal lobes often take the form of an “odd experience.” Children may see or hear things that are not there. They may feel an emotion: often fear, but sometimes sadness, anger or joy. They may smell a bad, smell or taste a bad taste. They may have a funny feeling in the pit of their stomach or a choking sensation. These seizures are sometimes called simple partial seizures of temporal lobe origin or temporal lobe auras.

Simple partial seizures usually last just a few seconds, although they may be longer. If they do not involve convulsive movements, they may not be obvious to the onlooker. In some children, simple partial seizures lead to complex partial seizures or to tonic-clonic convulsions.Simple partial seizures are not hysteria, acting out, or mental illness.

Complex Partial Seizures (Psychomotor or when arising from the temporal lobe, Temporal Lobe Seizures)
Complex partial seizures occur when epilepsy activity spreads to involve a major portion of the brain but does not become generalized. They often occur after a simple partial seizure particularly when it is of temporal lobe origin.

They may not involve convulsions, but consciousness is impaired. The child will no longer respond to questions after the seizure starts. Following the seizure, the child will have Incomplete memory of what went on though they often have memory of an aura at the onset of the event.



Complex partial seizures often begin with a blank look or stare and then may progress to chewing or uncoordinated activity. The child may appear unaware of the surroundings and may seem dazed. Some children sometimes perform meaningless bits of behavior, which appear random and clumsy (automatisms). They may pick at their clothes or try to take them off, walk about, pick up things, or mumble. They may appear afraid, and try to run and struggle.

Complex partial seizures usually last a few minutes (often 2-4), and they may be followed by a state of confusion that lasts even longer. Once the pattern of the seizures is established in a given child, it will usually be repeated with each subsequent seizure. Complex partial seizures sometimes resist anticonvulsant medication. In some children, they lead to tonic-clonic seizures. Complex partial seizures are not day dreaming, mental illness, bad behavior, drunkenness or drug abuse. Do not grab hold of the individual (unless there is a danger), shout, or expect to be heard.

Generalized Tonic-Clonic Seizures (Grand Mal)
Generalized seizures occur when epileptic activity occurs throughout the entire brain. The child is unconscious from the start, and will have a major convulsion with both a tonic (stiffening) and clonic (jerking) phase. After the seizure, the children are unconscious and then groggy for a while. They may want to sleep. There will be no memory of what went on during the seizure. The seizure begins with a fall, possibly accompanied by a sudden cry. The body stiffens (tonus), and then, after a while, begins to jerk (clonus). There may be shallow breathing or temporarily suspended breathing, with bluish skin or lips. (Note that childrendo not stop breathing long enough to hurt themselves.) There may be a loss of bladder or bowel control. Toward the end of the seizure, children may salivate profusely. Tonic-clonic seizures usually last 1 to 3 minutes, seldom longer. Tonic-clonic seizures are not a heart attack, a stroke, fainting, breath holding spells, or reflux from the stomach. Do not put anything in the person's mouth. Do not give liquids, try artificial respiration, or shout at the individual.

Absence Seizures (Petit Mal)
In absence seizures, epileptic activity also occurs throughout the entire brain. It is a milder type of activity, however, which causes unconsciousness without causing convulsions. After the seizure, there is no memory of what went on. An absence seizure consists of a period of unconsciousness with a blank stare, and begins and ends abruptly, without warning. There is no confusion after the seizure, and the child usually can resume full activity immediately. Seizures may be accompanied by chewing movements, rapid breathing, or rhythmic blinking.

Absence seizures are short, usually lasting 2-10 seconds. They are very mild, and may go unnoticed by parents and teachers. Absence seizures may recur frequently during the day, however, and the child may have difficulty learning if they are not recognized and treated. Absence attacks are not daydreaming, actions of deliberately ignoring adult instructions, or a lack of attention.

Atonic Seizures (Drop Attacks)
Although relatively uncommon, when they occur, they can be very hard to deal with. The seizures can occur without warning. The individual abruptly loses consciousness, collapses and falls to the floor. There is no convulsion, but the individuals may bang their head as they fall. Recovery occurs after a few seconds. When the person regains consciousness, he or she can stand and walk again. Atonic seizures are not indicative of clumsiness or a problem with walking, nor are they fainting.

Infantile Spasms
Infantile spasms are not common and normally occur only in the first year of life. The seizures consist of a cluster of sudden, quick movements that usually start at around 3-7 months of age. Generally, if the child is sitting up, the head may fall forward, the arms will flex forward and the body may flex at the waist. If the child is lying down, its knees will be drawn up, with arms and head flexed forward as if the body is reaching for support.
The seizures last one or two seconds and often repeat in a series of 5 to 50 or more. The seizures are more likely to occur when the child is drowsy, waking up or going to sleep. Infantile Spasms are not colic or normal movements for a baby.

Myoclonic Seizures
Myoclonic seizures occur in several different types of epilepsy. The seizures involve abrupt muscle jerks in part or all of the body. The seizures may involve a hand suddenly flinging out, or a shoulder shrug, a foot kicking, or the whole body may jerk. The events may occur individually, or in a series. Consciousness is not impaired. Myoclonic Seizures are not tics or “startle” responses.

Status Epilepticus
Any type of seizure that continues for a period of more than 10 minutes or repeats without the individual recovering is called status or status epilepticus. Immediate medical attention is necessary.