TYPES OF EPILEPSY

- **PRIMARY GENERALIZED EPILEPSY (AKA IDIOPATHIC GENERALIZED EPILEPSY)** - an epilepsy syndrome of idiopathic or unknown cause. An idiopathic disease is a "primary" or "intrinsic" disorder that cannot be attributed to a known underlying condition. So, while other types of epilepsy may be caused by a brain tumor, stroke, or other neurological disorder, idiopathic epilepsy is a primary brain disorder of unknown cause. In fact, most idiopathic epilepsy syndromes are presumed to be due to a genetic cause, but in most cases the specific genetic defect is not known and a family history of epilepsy may not be present.
  - **CHILDHOOD ABSENCE EPILEPSY** – Absence seizures that occur between the ages of 5 and 9. When diagnosing CAE, typical absence seizures need to be differentiated from atypical absence seizures which can occur at an earlier age. An EEG of a child with CAE will show a typical pattern known as 3-Hz generalized spike and wave complexes. Many children with CAE have normal neurological examinations and intellectual abilities. However, some children may have developmental and intellectual impairments and may have other types of seizures including, but not limited to, tonic clonic seizures.
  - **JUVENILE MYOCLONIC EPILEPSY** characterized by quick little jerks of the arms, shoulders, or occasionally the legs. These usually occur in the early morning, soon after awakening. The myoclonic jerks sometimes are followed by a tonic-clonic seizure or tonic-clonic seizures can occur independently. Absence seizures also may occur, where the patient seems to 'blank out' for a short period of time that can last from seconds to several minutes. The seizures of JME may begin between late childhood and early adulthood, usually around the time of puberty.
  - **JUVENILE ABSENCE EPILEPSY** - Common type of epilepsy that typically begins on or after puberty, between the ages of 10 and 17. About one-third of patients with JAE have a family history of seizures. Children may experience a few absence seizures per day. The child may also experience tonic-clonic seizures upon awakening.
  - **MYOCLONIC ASTATIC EPILEPSY / DOOSE SYNDROME** - An epilepsy syndrome of early childhood. Most symptoms appear between ages one and five. Characterized by difficult to control generalized seizures. Generally resistant to medication, children may experience several seizures daily.
- **SYMPTOMATIC GENERALIZED EPILEPSY** - Majority of seizures are generalized, but partial onset seizures can also occur. The types of generalized seizure that occur include myoclonic, tonic, atonic, atypical absence, and generalized tonic-clonic. Virtually any type of partial onset seizure can also occur, depending on the underlying brain pathology. Usually (but not always) there is a known underlying brain disorder or injury, which is often severe. These syndromes may occur in the setting of certain neurological diseases or may be due to lack of oxygen at birth, trauma, infection, developmental malformations, chromosomal abnormalities or other causes.
  - **INFANITLE SPASMS/WEST'S SYNDROME**- a very uncommon form of epilepsy, begin between 3 and 12 months of age. The seizures, or spasms, consist of a sudden jerk followed by stiffening. With some spells, the arms are flung out as the body bends forward ("jackknife seizures"). Other spells have more subtle movements limited to the neck or other body parts. A brain disorder or brain injury, such as birth trauma with oxygen deprivation, precedes the seizures in 60% of these infants, but in the other 40% no cause can be determined, and development is normal prior to the onset of seizures.
  - **LENNOX-GASTAUT SYNDROME** – Characterized by mental deficiency that can range from slight to profound cognitive impairment, a distinct brain wave pattern and multiple seizure types. The peak age of onset is between the ages of 3 and 5 years. There is a slightly greater prevalence in boys than girls. It is estimated that between 20 and 60 percent of children with this syndrome have a prior history of infantile spasms. This syndrome is rare and occurs in about .3 in every 1,000 live births.
  - **PROGRESSIVE MYOCLONIC EPILEPSY** - rare and frequently result from hereditary metabolic disorders. They feature a combination of myoclonic and tonic-clonic seizures. Unsteadiness, muscle rigidity, and mental deterioration are often also present.

IDIOPATHIC LOCALIZATION-RELATED EPILEPSIES (ILRE)

- **FRONTAL LOBE EPILEPSY** – recurring seizures beginning in the frontal lobe – the area of the brain located behind the forehead. Because the frontal lobe is responsible for planning and execution of movements and personality, frontal lobe epilepsy can have a dramatic affect on a patient's quality of life. Frontal lobe seizures are often very brief (<30 seconds) and tend to occur at night. They are typically simple or complex partial seizures and can quickly secondarily generalize. Because there are so many connections between the frontal and temporal lobes, it can be difficult to determine which section of the brain is being affected.
- **TEMPORAL LOBE EPILEPSY** - Recurring seizures beginning in the temporal lobe – the section of the brain located on the sides of the head behind the temples and cheekbones. Compared to other lobes in the brain, the temporal lobes seem to have a tendency to have seizures. The mesial portion (middle) of both temporal lobes are very important in epilepsy – they are frequently the source of seizures and can be prone to damage or scarring.
- **PARIETAL LOBE EPILEPSY** - Known as the "association cortex," the parietal lobe is responsible for connecting meaning to the brain's functions. It is here the brain creates a visual image, sounds are recognized as words, and the sense of touch is associated with a particular object. In other words, the parietal lobe is where perception becomes reality. Seizures beginning in the parietal lobe are rare.

- **OCCIPITAL LOBE EPILEPSY** - recurring seizures beginning in the section of the brain located in the back of the head primarily responsible for vision. When a seizure begins in the occipital lobe, flashing bright lights or other visual changes may be experienced off to the left side (if occurring in the right cortex), or the right side (if occurring in the left cortex).
- **NEOCORTAL EPILEPSY** - Characterized by seizures that originate from the brain's cortex, or outer layer. The seizures can be either focal or generalized. They may include strange sensations, visual hallucinations, emotional changes, muscle spasms, convulsions, and a variety of other symptoms, depending on where in the brain the seizures originates.
    - **IDIOPATHIC PARTIAL EPILEPSY** - An idiopathic disease is a disorder that cannot be attributed to a known underlying condition. most idiopathic epilepsy syndromes are presumed to be due to a genetic cause, but in most cases the specific genetic defect is not known and a family history of epilepsy may not be present.
        - **BENIGN RONANDIC EPILEPSY (AKA BENIGN EPILEPSY WITH CENTROTEMPORAL SPIKES)** - an epilepsy syndrome affecting children, characterized by twitching, numbness, or tingling of the child's face or tongue, and may interfere with speech and cause drooling. Seizures spread and become generalized seizures. In many cases, the seizures are infrequent and usually occur only at night. These seizures typically last no more than 2 minutes and the child remains fully conscious. BRE typically begins around ages 6-8, and is more likely to affect boys than girls. This syndrome represents about 15% of all epilepsies in children.
        - **EARLY ONSET BENIGN CHILDHOOD OCCIPITAL EPILEPSY -** Consciousness is usually impaired or lost, either from the onset or the course of the fits, but in a few children, it may be preserved. Duration varies from a few minutes to hours (partial status epilepticus). Seizures are usually nocturnal, but semiology is similar in nocturnal or diurnal fits. Onset is between 1 and 12 years with a peak at 5 years. One third of children have a single seizure, the median total number of fits is two to three, and the prognosis is invariably excellent, with remission usually occurring within 1 year from onset. A few children may later develop rolandic or other benign partial seizures. The likelihood to have seizures after age 12 years is exceptional and rarer than that of febrile convulsions.
        - **LATE ONSET BENIGN CHILDHOOD OCCIPITAL EPILEPSY -** The age of onset 4-12. Both sexes are equally affected. The children with LOS had a higher incidence of seizures, shorter duration of seizures and more frequent diurnal onset (than those with EBOS); also, although not statistically significant, the LOS group had more frequent visual hallucinations and headaches.
- **REFLEX EPILEPSY** - seizures are triggered by specific stimuli in the environment. In the most common type of reflex epilepsy, absence, myoclonic or tonic-clonic seizures are triggered by flashing lights. This is called “photosensitive epilepsy”, which usually begins in childhood and is often outgrown by adulthood. Other environmental triggers in reflex epilepsy include sounds such as church bells, a certain type of music or song, or a person’s voice. For some people, activities such as arithmetic, reading, writing, and even thinking about specific topics can provoke seizures. These non-visual stimuli may trigger generalized or partial-onset seizures. Some patients with a reflex epilepsy can have spontaneous seizures that occur without exposure to their specific trigger.
- **STURGE-WEBER SYNDROME -** A neurocutaneous disorder (affecting the brain and skin) identifiable by the “port-wine stain” (known as an angioma) located on the forehead area around the eye. Seizures occur in more than 80% of children diagnosed with SWS, weakness on one side of the body (hemiparesis), developmental delays, and increased pressure in the eye (glaucoma). While not considered to be a genetic trait, SWS is a developmental abnormality of the blood vessels of the face and brain.
- **PANAYIOTOPOULOS SYNDROME/BENIGN OCCIPITAL EPILEPSY (AKA BENIGN FOCAL EPILEPSY WITH OCCIPITAL PAROXYSMS) –** A hereditary type of epilepsy that represents about 3 percent of all childhood epilepsy cases. There is a somewhat higher incidence in girls than in boys. This type of epilepsy can be grouped into two categories, depending on the age of the child when seizures begin. Panayiotopoulos type begins between 15 months and 17 years. Seizures are infrequent and typically occur at night, shortly after the child falls asleep. Episodes usually last less than 10 minutes and may include vomiting and gazing toward one side, and frequently evolve to rhythmic muscle contractions on one or both sides of the body. Triggers may include turning off lights, going from lighted areas to dark ones, or from dark areas to light ones. Gastaut type begins between the ages of 3 and 16 years and has a peak onset between ages 7 and 9. Children may experience visual hallucinations with the seizure. Headaches are common before, during, or after the seizures.
- **MITOCHONDRIAL DISORDERS** - Mitochondria are the energy factories of the cell. Abnormalities in mitochondrial DNA or genes produce metabolic disorders that affect different parts of the body, including muscle and brain. Mitochondrial disorders can be

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inherited or sporadic. When inherited, the abnormal genes always come from the mother, since all mitochondria are of maternal origin. Two mitochondrial disorders can be associated with epileptic seizures.

- **MELAS:** mitochondrial encephalopathy, lactic acidosis (meaning too much lactic acid in the blood), and stroke-like episodes. MELAS can lead to stroke-like episodes at a young age (usually before 40), seizures, dementia, headaches, vomiting, unsteadiness, and ill effects from exercise. Persons with MELAS can have both generalized (including myoclonic and tonic-clonic) and partial seizures.
- **MERRF:** myoclonic epilepsy with ragged red muscle fibers). MERRF is one of the progressive myoclonic epilepsies. It can also be associated with hearing loss, unsteadiness, dementia, and ill effects from exercise. In addition to myoclonic seizures, patients with MERRF often have generalized tonic-clonic seizures. There are other mitochondrial disorders that do not fit clearly into the MELAS or MERRF syndromes but which can cause epilepsy and additional neurological problems.

- **LANDAU-KLEFFNER SYNDROME** - A rare childhood disease in which children experience a variety of seizure types, and gradually lose the understanding of language and eventually speech production (aphasia). Symptoms usually begin between age 3 and 8 and are more common in boys than girls.
- **LENNOX-GASTAUT SYNDROME -** The peak age of onset is between the ages of 3 and 5 years. There is a slightly greater prevalence in boys than girls. It is estimated that between 20 and 60 percent of children with this syndrome have a prior history of infantile spasms. Brain abnormalities are common with this syndrome, so physicians will perform brain imaging tests. The physician will also check for signs of prior seizures, mental retardation, hearing and visual impairment, and tuberous sclerosis which are sometimes present in children with Lennox-Gastaut.
- **RASMUSSEN SYNDROME (AKA RASMUSSEN'S ENCEPHALITIS)** - an autoimmune process that causes one hemisphere of the brain to become inflamed and deteriorate. Inflammation may stop without treatment, but the damage is irreversible. Rasmussen's syndrome is associated with intractable unilateral seizures, progressive hemiparesis or weakness on one side and intellectual dysfunction. Seizures are often the first symptom to appear.
- **HYPOTHALAMIC HAMARTHOMA** - Small tumors in the base of the brain that affect the hypothalamus can cause a syndrome consisting of abnormally early puberty, partial seizures with laughing as a frequent feature, and increased irritability and aggression between the seizures. The partial seizures may be simple or complex and there may be secondary generalized tonic-clonic seizures. Affected individuals are often short and have mild abnormalities in their physical features (dysmorphisms).
- **DRAVET SYNDROME (AKA SEVERE MYOCONIC EPILEPSY OF INFANCY or SMEI) -** a severe form of epilepsy. It appears during the first year of life with frequent febrile seizures – fever-related seizures that, by definition, are rare beyond age 5. Later, other types of seizures typically arise, including myoclonus (involuntary muscle spasms). Status epilepticus – a state of continuous seizure requiring emergency medical care – also may occur. Children with Dravet syndrome typically experience poor development of language and motor skills, hyperactivity, and difficulty relating to others. In 30 to 80 percent of cases, Dravet syndrome is caused by defects in a gene required for the proper function of brain cells. Borderline SMEI (SMEB) and another type of infant-onset epilepsy called generalized epilepsy with febrile seizures plus (GEFS+) are caused by defects in the same gene. In GEFS+, febrile seizures may persist beyond age 5.
    - **GENERALIZED EPILEPSY WITH FEBRILE SEIZURE PLUS (GEFS+) -** An unusual epilepsy syndrome. It describes families who have several members from different generations with epileptic seizures. The epileptic seizures nearly always start after a family member has had febrile convulsions. Febrile convulsions are seizures associated with a high temperature. Usually febrile convulsions stop after the age of six years. In GEFS+ families, children may go on to have febrile seizures well beyond this age. They may also develop other seizure types not associated with a high temperature. Very rarely, there may be a family member with a very severe type of epilepsy called Dravet syndrome.
- **RING CHROMOSOME 20 SYNDROME -** A ring chromosome is formed by the fusion of two arms of a straight chromosome during pre-natal (before birth) development. Why the formation of the ring causes epilepsy and other symptoms of the syndrome is not well understood. The problems associated with this syndrome can occur from birth to 17 years old. Seizures usually are the first and major clinical symptom of this syndrome, and usually do not respond to medications. They are often complex partial, usually with altered consciousness, staring, oral automatisms, unspecified automatic behavior, focal motor symptoms and/or head turning. Epilepsy is not present in all individuals with RC20, however, it is the most consistent clinical feature. Unlike other chromosomal disorders with epilepsy, malformed organs or appearance and other congenital malformations are rarely seen.

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- **ANGELMAN SYNDROME** - A rare condition which used to be called the "happy puppet" syndrome because the children behave as though they are a "puppet-on-a-string". Children have learning difficulties and speech delay that are usually severe. They also have jerking movements, tongue-thrusting, a characteristic (typical) facial appearance, and a happy mood with sudden bursts of laughter and epilepsy. Many children also have a fascination for water, and usually running water. The condition is due to an abnormality on chromosome 15. The condition is usually diagnosed between four and 10 years of age, but sometimes earlier.
- **AICARDI SYNDROME** - A very rare syndrome which only occurs in girls. Affected girls have developmental delay and seizures. They also have structural brain abnormalities, including a partially or completely absent corpus callosum (the structure that links the two halves of the brain together). Girls with Aicardi syndrome have very typical eye abnormalities, which often help to make the diagnosis. The abnormalities on the retina at the back of the eye are called choroidal lacunae. They appear as distinctive, round footprint shaped yellow-white lesions (marks). As well as an absent corpus callosum, some babies may have brain cysts or other brain abnormalities. Other features of Aicardi syndrome include cleft lip and palate, asymmetry of the face (lopsided, uneven), microcephaly (a small head), abnormally formed bones in the backbone and scoliosis (curvature of the back). Aicardi syndrome does not appear to run in families. It is thought to be caused by a spontaneous mutation (random change) in the X chromosome (gene) which happens at conception. The precise genetic abnormality is still unknown.
- **UNVERRICHT-LUNDBORG SYNDROME (AKA BALTIC MYOCLONUS)** - This condition is very rare. Most children with this syndrome begin to have epilepsy between the age of six and 16 years. Half will have tonic-clonic seizures which often start in the night, during sleep, and the other half will have myoclonic seizures (jerks). Something as simple as starting to move or a simple daily task may cause a sudden jerk which might then cause the child to fall down. These movements are called 'action myoclonus', because the myoclonus is most noticeable when the child tries to do something. Following a period of malfunction, these nerve cells will eventually die. The motor problems are referred to as ataxia. The condition is inherited genetically.
- **RETT SYNDROME -** This syndrome happens in about one in every 10,000-12,000 girls. It rarely affects boys, but when it does, they are always affected far more severely than girls. Rett syndrome is caused by a genetic abnormality. Girls with Rett syndrome may show normal development for the first six or more months of their life. Then between six and 30 months of age (often between six and 18 months of age), their development slows down and may even go backwards. They become less interested in play, lose the ability to speak and may become irritable and scream for no obvious reason. They stop using their hands purposefully and they may, instead, begin to move their hands in a repetitive way. This often takes the form of hand-wringing or hand-washing movements. Epilepsy eventually happens in about nine out of 10 children. It usually begins after the age of two years, but may begin in the first year of life. The seizures may be of various types. When boys are affected, the epileptic seizures nearly always start within the first few weeks or months of life, and are often extremely difficult to treat.
- **OHTAHARA SYNDROME (AKA EARLY INFANTILE EPILEPTIC ENCEPHALOPATHY WITH SUPPRESION BURSTS)** – A very rare epilepsy syndrome. Seizures start before three months of age. Most babies have an underlying structural brain abnormality. This may be genetic in origin (passed on through the genes), or happen because of brain damage before or around the time of birth.
- **MIGRATING PARTIAL EPILEPSY IN INFANCY** - Most children start having seizures in the first few weeks of life and all will start having seizures by six months of age. The seizures may only happen now and then in the first few weeks after the epilepsy starts. However, very rapidly, the seizures increase in frequency to the point where they may happen many times every day. Seizures are usually shorter in younger children, lasting 30 seconds to one minute. But in older children the seizures may last many minutes. No cause has been found to explain this type of epilepsy although doctors think the cause is likely to be in the child's genes. Most babies make very little developmental progress or can even lose the progress they have made. They will be dependent for all their care and have problems with feeding.
- **GLUCOSE TRANSPORTER 1 (glut1) DEFICIENCY SYNDROME** - A genetic disorder that impairs brain metabolism. Most children start having seizures in the first few months of life, although some children can develop seizures later, or even have other neurological symptoms. Children can have a variety of symptoms which can be present all the time, or can come and go. Children with paroxysmal symptoms are often worse when they haven't eaten much, or before a meal. Seizures, of any type, are often the first feature of brain dysfunction. In this syndrome, abnormal paroxysmal eye movements may be noticed before the seizures start. Babies will often have slow head growth and developmental delay. They may go on to develop spasticity (stiffness of limbs) and abnormal posturing of limbs and/or trunk (dystonia).

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