DOWNLOAD PDF MOVEMENT DISORDERS IN CHILDHOOD

Chapter 1 : Movement Disorders in Childhood Thalamic Tumors

Movement disorders can result from many types of brain injury, such as head trauma, infection, inflammation, metabolic disturbances, toxins or unintended side effects of medications. They can also be a symptom of other, underlying diseases or conditions, including genetic disorders. The brain.

Most of the thalamic tumors were low grade gliomas. Most movement disorders began shortly after surgical resection and only one developed symptoms months prior to surgery. The most common movement disorder was postural tremor, followed by ballism, athetosis, resting tremor, myoclonus and dystonia. Study neurologists were blinded to magnetic resonance imaging MRI data in order to analyze the extent of involvement of the thalamic nuclei. Out of the eight thalamic nuclei identified as affected in the MRI data of these patients, only the red nucleus was involved in all nine patients. Six of these patients also had substantia nigra involvement. This article finds that movement disorders associated with thalamic tumors are more commonly seen as a sequelae of surgical injury than as a presenting symptom of the tumor. Although the findings in this article focus on the occurence and outcome of movement disorders that develop after surgical intervention it is important to remember that brain neoplasms and other focal lesions need to be considered when otherwise healthy children develop the sub-acute onset of bilateral or unilateral tremor as well as other types of movement disorders [2]. While movement disorders associated with brain tumors are more commonly seen unilaterally, it is important to note that in this study, a subset of patients developed bilateral symptoms. Another important finding in this study is that the movement disorders did not improve significantly over time. This suggests a permanent injury from the tumor or surgical procedure, rather than post- operative inflammation, as the cause of the movement disorder associated with thalamic tumors. Whether a movement disorder develops before or after the diagnosis and treatment of a thalamic tumor, they represent significant morbidity for this population and can have a great impact in their quality of life. When addressing movement disorders associated with thalamic tumors, it is imperative to identify the correct phenomenology in order to implement appropriate pharmacological and non-pharmacological interventions that are appropriate for each individual patient [3]. Even with correct phenomenology identification, secondary movement disorders are difficult to treat regardless of the underlying cause of the involuntary movement. In these settings, it is common for multiple medications to be tried without success. As investigators suggested, the treatment of secondary movements disorders merits further research. Treatment can be extremely challenging and many times ineffective in this this vulnerable patient population. Disclosures The author s have declared that no competing interests exist. Clinical characteristics and long-term outcomes of movement disorders in childhood thalamic tumors. Pediatr Neurol Dec Two children with tremor. CMAJ Apr 7: Advances in management of movement disorders in children. Lancet Neurol Jun 7: Movement Disorders in Childhood Thalamic Tumors. Pediatric Neurology Briefs, 32, p. Pediatric Neurology Briefs, 32, 4.

Chapter 2 : Movement Disorders Clinic | Childrenâ€™s Pittsburgh

Movement Disorders in Childhood, Second Edition, provides the most up-to-date information on the diseases and disorders that affect motor control, an important area of specialization within child neurology.

Ataxia Ataxia is a degenerative disorder affecting the brain, brainstem or spinal cord. This can result in clumsiness, inaccuracy, instability, imbalance, tremor or a lack of coordination while performing voluntary movements. Movements are not smooth and may appear disjointed or jerky. Patients may fall down frequently due to an unsteady gait. Ataxia also can affect speech and movement of the eyes. If a metabolic disorder can be identified as the underlying cause, specific treatment may be available in select cases. The cornerstone of treatment for ataxia of parkinsonism or parkinsonism of any cause is the use of oral L-DOPA. Other medications used to treat ataxia associated with parkinsonism or parkinsonism of any cause include anticholinergics , dopamine agonists , amantadine , selegiline and entacapone. In children with ataxia, generally only anticholinergics are prescribed. Dystonia Dystonia is a neurological muscle disorder characterized by involuntary muscle spasms. Dystonia results from abnormal functioning of the basal ganglia , a deep part of the brain which helps control coordination of movement. These regions of the brain control the speed and fluidity of movement and prevent unwanted movements. Patients with dystonia may experience uncontrollable twisting, repetitive movements or abnormal postures and positions. These can affect any part of the body, including the arms, legs, trunk, eyelids and vocal cords. General dystonias involves the entire body. Depending on what part of the body is affected, the condition can be very disabling. There is a three-tiered approach to treating dystonia: These may be used alone or in combination. Botox injections help block the communication between the nerve and the muscle and may lessen abnormal movements and postures. Surgery is considered when other treatments have proven ineffective. The goal of surgery is to interrupt the pathways responsible for the abnormal movements at various levels of the nervous system. Some operations purposely damage small regions of the thalamus thalamotomy , globus pallidus pallidotomy or other deep centers in the brain. Recently, deep brain stimulation DBS has been tried with some success. Other surgeries include cutting nerves leading to the nerve roots deep in the neck close to the spinal cord anterior cervical rhizotomy or removing the nerves at the point they enter the contracting muscles selective peripheral denervation. Essential Tremor Essential tremor is an uncontrolled shaking or trembling, usually of one or both hands or arms, that worsens when basic movements are attempted. Essential tremor affects about five million people in the U. National Library of Medicine, essential tremors are found most commonly in adults over the age of It is caused by abnormalities in areas of the brain that control movement and is not tied to an underlying disease e. About 50 percent of patients have a family history of the condition. This condition usually does not result in serious complications, but it certainly can interfere with daily activities and cause distress. In some cases, physical therapy or changes in lifestyle may improve symptoms. About 50 to 75 percent of patients taking medications have a reduction of their tremor. Beta-blockers , anti-seizure medications , benzodiazepines and carbonic anhydrase inhibitors often are prescribed. Beta-blockers usually are prescribed for younger patients because they may cause memory loss and confusion in older patients. Botox injections help block the communication between the nerve and the muscle and may lessen tremor. If the tremor is so severe that is causes a disability, surgery may be recommended. Thalamotomy purposely destroys a portion of the area deep within the brain that receives sensory messages, and area known as the thalamus. About 75 percent of patients undergoing this procedure find relief on one side of their body. Surgery on both sides of the thalamus is rarely done due to the high risk of speech loss. Deep Brain Stimulation is another surgical option in severe cases of essential tremor that have not responded to medication. A hair-thin wire is implanted in the thalamus and connected to a neurostimulator implanted under the collarbone. The neurostimulator sends electrical impulses along the wire to the thalamus, interrupting signals that cause tremor. Onset most often occurs between ages 35 and 50, with the condition progressing without remission over 10 to 25 years. A juvenile form of the disease affects patients age 20 and younger, accounting for about 16 percent of all cases. Symptoms include jerking; uncontrollable movements of the limbs, trunk, and face; progressive loss of mental abilities; and the

development of psychiatric problems. Doctors may prescribe antipsychotics, antidepressants , tranquilizers , mood-stabilizers or botox injections. These are prescribed in the lowest effective dosage, as all of these medications may have side effects. Researchers have observed that the earlier in life the symptoms occur, the faster the disease often progresses. Because symptoms, onset and severity of MSA vary from person to person, differing ranges of symptoms were designated initially as three different diseases: Shy-Drager syndrome , striatonigral degeneration and olivopontocerebellar atrophy. All of these now are classified under MSA. Symptoms include stiffness or rigidity; freezing or slowed movements; instability; loss of balance; loss of coordination; a significant fall in blood pressure when standing, causing dizziness, lightheadedness, fainting or blurred vision orthostatic hypotension ; male impotence ; urinary difficulties ; constipation ; and speech and swallowing difficulties. Medication may be prescribed to treat some of the symptoms associated with this disease. Orthostatic hypotension can be improved by prescribing drugs that raise blood pressure. As MSA progresses, the benefits of medication lessen. In cases that have progressed and are more severe, a feeding tube may be needed when the patient cannot swallow food on his or her own. Myoclonus Myoclonus is a twitching or intermittent spasm of a muscle or group of muscles. Myoclonus is classified into several major types and many subcategories. The most common type is cortical myoclonus, which arises from an area of the brain known as the sensorimotor cortex. Jerky movements usually have a regular rhythm and may be limited to one muscle or muscle group focal or several different muscle groups multifocal. They may occur without an obvious cause or be a result of many diseases. Subcortical myoclonus usually affects many muscle groups generalized and may be the result of abnormally low levels of oxygen in the brain hypoxia or a metabolic process, such as kidney or liver failure. Spinal myoclonus usually is caused by a focal spinal lesion, such as multiple sclerosis , syringomyelia , trauma, ischemic myelopathy or an infection such as herpes zoster , Lyme disease , E. The jerking often lasts longer and is more variable than in cortical or subcortical myoclonus and continues during sleep. The most common type of peripheral myoclonus is hemifacial spasm , which may occur for no underlying reason or be caused by compression of the facial nerve. Movements persist during sleep and may last for only a few days or for as long as a few months. The exact type of myoclonus is delineated further by the parts of the body affected and by the underlying causes. Myoclonus is treated through prescribing medications that may help reduce symptoms. In some cases, effective results are achieved by combining multiple drugs. Some of the medications prescribed are barbiturates , phenytoin , primidone , sodium valproate and the tranquilizer clonazepam. All of these medications have potential side effects, so it is very important for patients to work closely with their doctor on medication management. These nerve cells die or become impaired, losing the ability to produce an important chemical called dopamine. Some common medications used are dopamine precursors, dopamine agonists and anticholinergics. Surgery is considered when medications have proven ineffective. Thalamotomy can help stop tremor by placing a small lesion in a specific nucleus of the thalamus. People with PSP experience a gradual loss of specific brain cells, causing slowing of movement and reduced control of walking, balance, swallowing, speech and eye movement. Often, there are personality and cognitive changes, causing emotional outbursts and a decrease in intellectual abilities. This disease more commonly affects people ages 40 to 60 and usually runs its full terminal course in six to 10 years. While the cause of PSP is unknown, researchers know that a brain protein called tau accumulates in abnormal clumps in certain brain cells in people with PSP, causing the cells to die. There appears to be a genetic predisposition. Unfortunately, there is no effective medication to treat PSP, but research is ongoing. Medications that may have a slight benefit are levodopa, amantadine and amitriptyline. Botox injections may be used to treat the blepharospasm involuntary eyelid closure that occurs in some people with PSP. Rett Syndrome Rett Syndrome is a progressive neurological disorder that causes debilitating symptoms, including reduced muscle tone , autistic-like behavior , repetitive hand movements, irregular breathing, decreased ability to express feelings, developmental delays in brain and head growth, gait abnormalities and seizures. Loss of muscle tone usually is the first symptom. According to the International Rett Syndrome Foundation , about one in every 10, to 23, infant girls is diagnosed with Rett, but the prevalence may be much higher due to undiagnosed cases. Rett can affect boys, but they account for a very small percentage of cases. Children with Rett appear to develop normally until six to 18 months of age, at

which point symptoms start to appear. Rett leaves its victims profoundly disabled, requiring maximum assistance with all aspects of daily living. Unfortunately, there is no cure for Rett. Treatment for the disorder focuses on the management of symptoms and requires a supportive, multidisciplinary approach. The disorder progresses through four major stages, each with characteristic symptoms and medical implications. Medication may be needed for breathing irregularities and motor difficulties. Antiepileptic drugs may be used to control seizures. Occupational therapy , education and supportive services are geared towards helping individuals with Rett cope with daily challenges and maintain a quality of life. Although it is severely debilitating, individuals with Rett have lived to middle age, but rarely beyond ages 40 to Many of the medications used to treat this condition have potential side effects, so it is very important to work closely with your doctor on medication management. Spasticity Spasticity is increased muscle contractions causing stiffness or tightness of the muscles that may interfere with movement, speech and walking. Spasticity usually is caused by damage to the portion of the brain or spinal cord that controls voluntary movement.

Chapter 3 : Download ebook Movement Disorders in Childhood pdf

Pediatric movement disorders is a relatively new and growing field of child neurology. Whereas hypokinetic disorders such as Parkinson disease predominate in adults, children more commonly demonstrate hyperkinetic disorders such as tics, tremor, chorea, and dystonia.

Programs and Services Movement Disorders In Depth Movement disorders can result from many types of brain injury, such as head trauma, infection, inflammation, metabolic disturbances, toxins or unintended side effects of medications. They can also be a symptom of other, underlying diseases or conditions, including genetic disorders. All these structures are thought to act together in a network, so damage to just one might be enough to cause a movement disorder. Treatment of movement disorders often focuses on correcting the underlying cause. How well a child will do depends on the type of movement disorder, its cause, how well the movement disorder is treated and how soon treatment is started. For example, a child with primary dystonia can live a normal life if she is treated early, whereas dystonia that goes undiagnosed and untreated for many years can become permanently disabling. Other movement disorders, such as tic disorders, may eventually go away on their own or become much milder. If the movement disorder is part of a serious genetic or degenerative disorder, there may be no curative treatment. Types of movement disorders include: Chorea Chorea is a symptom of a neurologic injury, not a disease in and of itself. They can be slow and writhing athetosis or more forceful ballismus. If chorea affects the legs, children may frequently stumble and have difficulty walking but rarely fall. Chorea in children can be caused by a brain injury at birth, head trauma, infection, inflammation or as a toxic reaction to medications. In some cases, it can have a genetic cause. Specific types of chorea include: Syndenham chorea, also called rheumatic encephalitis, is a short-lived, autoimmune form of chorea that can occur as an after-effect of a group A strep infection. Juvenile Huntington disease is a very rare genetic form of chorea not seen commonly in children. Chorea from genetic causes, such as NKX2. Tremor Tremor is a rhythmic shaking or trembling of a limb. It sometimes occurs in children as a complication of an underlying illness or head injury, as a side effect of heavy medications or as a consequence of genetic disease. Tremor can occur at rest or during movement, can occur alone or be accompanied by other neurological symptoms such as weakness of the affected limb and difficulty in fine-tuning movements. Tremor also sometimes runs in families. The most common inherited form is known as essential tremor. Unlike most other movement disorders, children can sometimes consciously suppress tremors. Myoclonus Myoclonus is a movement disorder involving very quick, sudden, involuntary muscle jerks that the child cannot suppress. The jerks can be occasional or frequent, and may occur randomly or in a semi-rhythmic pattern. Myoclonus sometimes has triggers, like holding the body in a certain posture, being touched or startled; or symptoms may seem to appear for no reason. Myoclonus may occur after a severe brain injury, or can result from a metabolic disorder or a neurodegenerative disease. Occasionally, a child will have myoclonus alone without any accompanying problem. There is also a benign form of myoclonus that affects newborns during sleep and usually goes away by 6 months of age, and another benign form that begins at about 6 months of age and goes away within 6 to 18 months. Dystonia In children with dystonia, faulty brain signals cause groups of muscles to contract abnormally. Rather than contracting in a coordinated fashion, muscles may contract in opposition to each other: Dystonia is often triggered by specific actions, like writing or walking, but can also occur when the child is at rest. Symptoms often start in one part of the body, such as the hand, leg or mouth, and then spread to other body areas. Deep brain stimulation surgery early in the course of the dystonia, before fixed deformities develop, is an important treatment to consider for certain children; for others, surgically implanting a medication pump may be the best option. Primary dystonia is thought to be genetic. Children with primary dystonia do not have any accompanying neurologic disorder. Primary dystonia tends to progress and spread most rapidly in the first five years after onset, then begins to stabilize. Secondary dystonia, the most common form of dystonia in children, results from a brain injury during birth, from a stroke or trauma, or from an inherited degenerative syndrome or metabolic disorder. The severity and symptoms of secondary dystonia vary depending on its underlying cause. Secondary dystonia often occurs together with

spasticity, and 5 to 15 percent of children with cerebral palsy also have dystonia. They are fairly common in children and usually can be treated by a general pediatrician. Muscle tics can involve any body part and may vary in severityâ€”from very mild and hardly noticeable to very disruptive, frequent and severe. If tics persist despite treatment, it may be beneficial for the child to see a specialist in movement disorders. About 10 to 15 percent of children will see their tics progress and become potentially disabling, but most tic disorders resolve or become minimal by the time a child reaches her 20s. If a child has both vocal and motor tics that last longer than a year, they are considered to have Tourette syndrome. Many children with Tourette syndrome also have attention deficit hyperactivity disorder ADHD , obsessive-compulsive disorder OCD or learning disorders, which can be evaluated and treated by a child neurologist or developmental pediatrician. Parkinsonism A child is considered to have parkinsonism if she has at least two symptoms shared by adults with Parkinson disease: By itself, parkinsonism is the least common movement disorder in children. Most often, the symptoms occur as a side effect of medications. Parkinsonism can also result from genetic conditions causing brain degeneration, such as Wilson disease, juvenile Huntington disease and lysosomal disorders, and from various forms of brain injury. As with adult Parkinson disease, treatment usually includes a trial of dopamine supplementation levodopa , with side effects carefully monitored. Ataxia Ataxia is a failure of motor coordination caused by injury or dysfunction of the cerebellum. Its symptoms are clumsiness, poor balance, irregularity of movements and inability to perform fine-tuned or smooth movements. Ataxia can have different patterns. Some cases begin abruptly, as a result of a stroke or brain bleed, inflammation or infection, and typically resolve as these causes are treated. Other types of ataxia repeatedly come and go, and are caused by epilepsy, genetic mutations, metabolic disorders or atypical types of migraine. There are also progressive forms of ataxia caused by various inherited genetic conditions known as the spinocerebellar ataxias. A condition known as functional neurological symptom disorder, sometimes called conversion disorder, can cause any of the movement disorders described above, even when no neurologic cause or condition can be identified. Functional disorders also can occur in children with other neurological diagnoses and make an existing movement disorder worse. Even though no physical cause can be discovered, these disorders can be just as disabling as the types of movement disorders described above. The good news is that most children recover from this type of movement disorder with retraining by physical therapists and continued supportive care from a neurologist. In some cases, children may benefit from joined care with a psychologist or psychiatrist. News and World Report for the fifth year in a row!

Chapter 4 : Movement Disorders In Depth | Boston Children's Hospital

Childhood movement disorders, taken together, are common. It is difficult to estimate the number of children with movement disorders as statistics about the frequency of these disorders in children is limited.

Tourette syndrome and other tic disorders Wilson disease Demographics The incidence rates and demographics vary for different types of movement disorders. The disorder can affect males and females and can begin at any age, although it may become worse as a person gets older. Causes and symptoms Causes Movement is produced and coordinated by several interacting brain centers, including the motor cortex, the cerebellum, and a group of structures in the inner portions of the brain called the basal ganglia. Sensory information provides critical input on the current position and velocity of body parts, and spinal nerve cells neurons help prevent opposing muscle groups from contracting at the same time. To understand how movement disorders occur, it is helpful to consider a normal voluntary movement, such as reaching to touch a nearby object with the right index finger. To accomplish the desired movement, the arm must be lifted and extended. The hand must be held out to align with the forearm, and the forefinger must be extended while the other fingers remain flexed. Movement of the right arm is begun by the left motor cortex, which generates a large volley of signals to the involved muscles. These electrical signals pass along upper motor neurons through the midbrain to the spinal cord. Within the spinal cord, they connect to lower motor neurons, which convey the signals out of the spinal cord to the surface of the muscles involved. Electrical stimulation of the muscles causes contraction, and the force of contraction pulling on the skeleton causes movement of the arm, hand, and fingers. Damage to or death of any of the neurons along this path causes weakness or paralysis of the affected muscles. One important refinement to it comes from considering the role of opposing, or antagonistic, muscle pairs. Contraction of the biceps muscle, located on the top of the upper arm, pulls on the forearm to flex the elbow and bend the arm. Contraction of the triceps, located on the opposite side, extends the elbow and straightens the arm. Within the spine, these muscles are normally wired so that willed voluntary contraction of one is automatically accompanied by blocking of the other. In other words, the command to contract the biceps provokes another command within the spine to prevent contraction of the triceps. In this way, these antagonist muscles are kept from resisting one another. Spinal cord or brain injury can damage this control system and cause involuntary simultaneous contraction and spasticity, an increase in resistance to movement during motion. In addition to sight, the most important source of information comes from the "position sense" provided by the many sensory neurons located within the limbs proprioception. Proprioception is what allows a person to touch a finger to his or her nose, even with eyes closed. The balance organs in the ears provide important information about posture. Both postural and proprioceptive information are processed by a structure at the rear of the brain called the cerebellum. The cerebellum sends out electrical signals to modify movements as they progress, "sculpting" the barrage of voluntary commands into a tightly controlled, constantly evolving pattern. Cerebellar disorders cause inability to control the force, fine positioning, and speed of movements ataxia. Disorders of the cerebellum may also impair the ability to judge distance so that a person under- or over-reaches the target dysmetria. Tremor during voluntary movements can also result from cerebellar damage. The basal ganglia send output messages to the motor cortex, helping to initiate movements, regulate repetitive or patterned movements, and control muscle tone. Circuits within the basal ganglia are complex. Within this structure, some groups of cells begin the action of other basal ganglia components and some groups of cells block the action. These complicated feedback circuits are not entirely understood. Disruptions of these circuits are known to cause several distinct movement disorders. A portion of the basal ganglia called the substantia nigra sends electrical signals that block output from another structure called the subthalamic nucleus. The subthalamic nucleus sends signals to the globus pallidus, which in turn blocks the thalamic nuclei. Finally, the thalamic nuclei send signals to the motor cortex. The substantia nigra, then, begins movement and the globus pallidus blocks it. This complicated circuit can be disrupted at several points. Disruptions in other portions of the basal ganglia are thought to cause tics , tremors, dystonia, and a variety of other movement disorders, although the exact mechanisms are not well understood. Some diseases

that cause sustained muscle contraction limited to a particular muscle group focal dystonia are inherited, but others are caused by trauma. Symptoms Abnormal movements are broadly classified as either hyperkinetic too much movement and hypokinetic too little movement. Dystoniaâ€”sustained muscle contractions, often causing twisting or repetitive movements and abnormal postures. Dystonia may be limited to one area focal or may affect the whole body general. Dystonia may be painful as well as incapacitating. Tremorâ€”uncontrollable involuntary shaking of a body part. Tremor may occur only when muscles are relaxed or only during an action or while holding an active posture. Ticsâ€”involuntary, rapid, non-rhythmic movement or sound. Tics can be controlled briefly. Myoclonusâ€”a sudden, brief, jerky, shock-like involuntary muscle contraction. Myoclonic jerks may occur singly or repetitively. Unlike tics, myoclonus cannot be controlled even briefly. Spasticityâ€”an abnormal increase in muscle tone. It may be associated with involuntary muscle spasms , sustained muscle contractions, and exaggerated deep tendon reflexes that make movement difficult or uncontrollable. Choreaâ€”rapid, non-rhythmic, uncontrolled jerky movements, most often in the arms and legs. Chorea also may affect the hands, feet, trunk, neck, and face. Choreoathetosis is a syndrome of continuous random movements that usually occur at rest and may appear to be fidgety, dancing, or writhing. Ballismâ€”like chorea, but the movements are much larger, more explosive and involve more of the arm or leg. This condition, also called ballismus, can occur on both sides of the body or on one side only hemiballismus. Akathisiaâ€”restlessness and a desire to move to relieve uncomfortable sensations. Sensations may include a feeling of crawling, itching , stretching, or creeping, usually in the legs. Athetosisâ€”slow, writhing, continuous, uncontrollable movement of the arms and legs. Bradykinesiaâ€”extreme slowness and stiffness of movement. Freezingâ€”inability to begin a movement or involuntary stopping of a movement before it is completed. Rigidityâ€”an increase in muscle tension when an arm or leg is moved by an outside force. Postural instabilityâ€”loss of the ability to maintain upright posture caused by slow or absent righting reflexes. Diagnosis Diagnosis of movement disorders requires a careful medical history and a thorough physical and neurological examination. The medical history helps the physician evaluate the presence of other conditions or disorders that might contribute to or cause the disorder. Records of previous diagnoses, surgeries, and treatments are reviewed. Genetic testing is available for some forms of movement disorders. Routine blood and urine analyses are performed. Brain imaging studies are usually performed. A lumbar puncture spinal tap may be necessary. Video recording of the abnormal movement is often used to analyze movement patterns and track progress of the disorder and its treatment. Other tests may include x rays of the spine and hips or diagnostic blocks with local anesthetics to provide information on the effectiveness of potential treatments. To aid diagnosis, a multi-disciplinary team may be consulted so the proper treatment can be planned. Occupational and physical therapy evaluations may be helpful to determine upper and lower extremity movement patterns and passive range of motion. In both tests, the examiner uses a computer, monitor, amplifier, loudspeaker, stimulator, and high-tech filters to see and hear how the muscles and nerves are responding during the test. In the nerve conduction study, small electrodes are placed on the skin over the muscles to be examined. A stimulator delivers a very small electrical current that does not cause damage to the body through the electrodes, causing the nerves to fire. In the electromyogram, a very thin, sterilized needle is inserted into various muscles. The needle is attached by wires to a recording machine. The patient is asked to relax and contract the muscles being examined. The electrical signals produced by the nerves and muscles during these tests are measured and recorded by a computer and displayed as electrical waves on the monitor. The test results are interpreted by a specially trained physician. An EEG electroencephalogram may be performed to detect seizures, analyze general brain functioning, and measure brain activity associated with movement or sensation. This test measures the electrical signals from the brain. Surface electrodes attached to the scalp measure voltages in the brain. The electrical activity can be measured while the child is resting or, in some cases, when the child is moving. An evoked potentials study may be part of the EEG test. Evoked potentials record the response of the brain to a sensory, visual, or auditory stimulus. Treatment Treatment of a movement disorder begins with a proper diagnostic evaluation. Treatment options include physical and occupational therapies, medications, surgery, or a combination of these treatments. Clinicians should work with the child and parents or caregivers to develop an individual treatment plan. Specific treatment goals will

vary from one person to the next. Treatment should be provided by a movement disorders specialist or specially trained pediatric neurologist and a multi-disciplinary team of specialists that may include a physiatrist, physical therapist, occupational therapist, gait and movement specialists, social worker, and surgical specialists as applicable, such as a pediatric orthopedic surgeon or pediatric neurosurgeon. For example, some people with multiple sclerosis who experience significant leg weakness find that spasticity makes their legs more rigid, helping them to stand, transfer to a chair or bed, or walk. Physical and occupational therapies Physical therapy includes stretching exercises, muscle group strengthening exercises, and range of motion exercises to prevent muscles from shortening contracture , preserve flexibility and range of motion, and reduce the severity of symptoms. Exercises should be practiced daily, as recommended by the physical therapist. Prolonged stretching can lengthen muscles, and strengthening exercises can restore the proper strength to affected muscles. Aquatic therapy also may be recommended, since there is less stress on the body when in the water.

Chapter 5 : Disorders of Childhood: Stereotypic Movement Disorder

As they note, pediatric movement disorders usually represent a small chapter or section in larger adult movement disorder textbooks. The authors' ease and skill in handling these pediatric disorders is evident throughout the book.

Children and adolescents with this disorder may wave their hands, rock back and forth, twiddle their thumbs, twirl objects, or kick or contract muscles in their legs. More severe repetitive behaviors that children might engage in include banging their heads against hard surfaces and slapping or punching themselves, both behaviors which frequently lead to bruises, cuts, bleeding, infection and more serious injuries. These behaviors are experienced as irresistible; children cannot stop themselves from engaging in them for too long. Also, the behaviors serve no apparent function other than allowing children to experience the sheer physical sensation of performing the movement. Children are not trying to dry their hands, or get the attention of a friend, for instance, when they wave their hands about. The odd, repetitive movements must not be caused by another more appropriate diagnosis, such as Obsessive-Compulsive Disorder , or by a pervasive developmental disorder such as Autism. Stereotypic Movement Disorder is not very common in the general population of children and adolescents. It is a fairly common occurrence within the population of Mentally Retarded population, however. To a lesser extent, the behavior is also more common in populations of sensory disabled children, such as blind children. Head-banging is the most commonly acted out self-injurious behavior. Diagnosis of Stereotyped Movement Disorder The compulsive, repetitive and nonfunctional movement behaviors characteristic of Stereotyped Movement Disorder are not at all subtle, and point quickly to a small group of diagnoses only. Historical information helps clinicians to understand how the movement symptoms first developed and how they may have changed over time. Current observational and interview data helps the clinician determine frequency whether the behaviors are constant or intermittent , triggers do they occur during particular events or situations , and consequences how the behaviors influence parents, family members and peers. When practical, interviews and observations should be conducted in both home and school settings to determine whether the child behaves differently in one context versus another. Interviews may utilize a structured format such as the Child Behavior Checklist so that important questions are not missed. The PIC measures the developmental, emotional, personality, interpersonal social , and cognitive mental status of children and adolescents. The test yields measures of Hyperactivity, Conduct Problems, and Social Skills Deficits in children between the ages of 5 kindergarten and 19 senior year of high school years. As is the case with most mental illnesses, it is a good idea that a complete medical exam be conducted on children who are suspected of having Stereotypic Movement Disorder so that underlying medical causes of such behavior can be either identified and treated, or ruled out. There are multiple diagnoses characterized by self-injurious behavior, and Stereotyped Movement Disorder needs to be differentiated from these conditions. For instance, individuals with Borderline Personality Disordered will sometimes cut or burn themselves. In this example, Borderline cutting and burning behavior is intentionally self-inflicted as a punishment or as a means of feeling an intense sensation making it different in quality from the self-injurious behavior characteristic of Stereotyped Movement Disorder. Additionally, Borderline Personality is a syndrome in its own right with a characteristic set of symptoms which also must be present before that diagnosis can apply. Both conditions have as their primary symptom an irresistible compulsion to act out seemingly nonfunctional and meaningless behaviors. The conditions are differentiated on the basis of one being a motor disorder, involving body movements, while the other is often verbal in nature. There is such a thing as complex motor tics, of course, and it can be difficult to correctly know when stereotyped behaviors should be thought of as motor tics rather than a movement disorder.

Chapter 6 : Movement disorders - Wikipedia

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In these situations, they can be part of normal infantile development. Stereotypies can also be secondary when they occur in the setting of a neurodevelopmental disorder such as autism spectrum disorder. A definitive cause of stereotypies has not been determined. Both a psychogenic and neuro-biologic basis of this movement disorder have been proposed. Dystonia Dystonia can be primary where dystonia is the only or predominant symptom or secondary where dystonia is due to another identified cause. Primary dystonia is typically caused by a single gene mutation. An example of this is DYT-1 dystonia which is a generalized dystonia that can present in childhood. In this disease, dystonia is the only symptom. In DYT-1 dystonia, children typically present with dystonia in a limb that then spreads and can involve their entire body. Secondary dystonia has varied causes. Dystonia can be a symptom of cerebral palsy. An example of how this occurs is in the setting of hypoxic-ischemic brain injury. The brain does not get enough oxygen and blood during delivery. This causes an injury to the basal ganglia which are the primary structures of the brain that control movement. Sometimes this injury can be seen on brain imaging with an MRI. Dystonia has many other causes. These include neurodegenerative diseases, brain infections, structural brain lesions, and medications. Dopamine-blocking medications can cause a specific type of dystonia called an acute dystonic reaction. In this case, dystonia presents abruptly after the medication is administered. Tremor There are many causes of tremor. On the least severe end of the spectrum, tremor can be physiologic and occur in otherwise healthy children. When tremor is more notable than the average healthy child, it is called an enhanced physiologic tremor. These tremors are exacerbated by stress and anxiety. Tremor can be hereditary as in the case with essential tremor. Children with essential tremor typically have an action tremor involving the hands. In some children, the head, voice, trunk, or legs can also be involved. Tremor can be caused by strokes or brain lesion which most commonly cause a unilateral tremor. Tremor can be caused by metabolic derangements such as low calcium, endocrine disorders such as hyperthyroidism, neurodegenerative diseases, medications, or toxins. Chorea Chorea can be seen in normal, healthy infants until they are about 6 month of age. After this age, chorea is typically pathologic. Chorea is rarely a primary disorder. Secondary chorea has numerous causes. In these cases, chorea is accompanied by additional neurologic symptoms and non-neurologic symptoms. Chorea can be acquired in childhood. On average months after the infection, children develop chorea. This is thought to happen when antibodies that formed to fight off the strep infection incorrectly attack and injure the basal ganglia. Chorea can be caused by metabolic derangements such as hypoglycemia or hyperglycemia low or high sugar , hyponatremia or hypernatremia low or high sodium , hypocalcemia low calcium and hyperthyroidism. Chorea can be seen after heart surgery when children have cardiopulmonary bypass post-pump chorea. Additional causes of chorea include infections, systemic lupus erythematosus, toxins, and neurodegenerative disorders. Chorea can be chronic as seen in a subtype of cerebral palsy. In this setting, chorea often co-exists with dystonia due to injury to the basal ganglia. Myoclonus Myoclonus can arise from multiple areas in the nervous system including the cortex, basal ganglia, brainstem, and spinal cord. In fact, myoclonus can be normal, as with sleep myoclonus. Most people have experienced this phenomenon where the body jerks as they are falling asleep, sometimes waking the person up while they are dozing off. There are numerous causes of pathologic myoclonus. Epileptic myoclonus must be differentiated from non-epileptic myoclonus. Sometimes this requires an electroencephalogram EEG to differentiate. Once myoclonus is confirmed as a movement disorder, the location and the appearance of the myoclonus can be helpful in determining the cause. Myoclonus can be seen in numerous inherited genetic diseases. It can also be acquired by exposure to toxins, infections, metabolic derangement, and hypoxic events. Parkinsonism Primary parkinsonism is typically caused be a single gene mutation. Secondary parkinsonism has a wide variety of causes. It can present as a symptom of a larger neurodegenerative disorder as in mitochondrial diseases. It can be caused by a brain

infection encephalitis or an autoimmune inflammatory process autoimmune encephalitis. Parkinsonism can also be an adverse reaction to a medication. This can be seen with a class of medications called dopamine-blocking medications. For example, in an otherwise healthy 5-yold with eye blinking and sniffing, a tic disorder can be diagnosed without laboratory evaluation. When additional diagnostic testing is indicated, it may include the following: In some cases, the movement disorder can be observed during the clinic visits. If the movement disorder is intermittent, the physician must rely on the description of the movement from the parents; this can be challenging. Parents and children can also attempt to mimic the abnormal movement during the visit. If the abnormal movement in questions is intermittent, home videos are very helpful in identifying the phenomenology. In these cases, a short video from a cell phone may be all that is needed to arrive at the correct diagnosis. Movement disorders may need to be distinguished from other conditions which lead to the appearance of abnormal movements. The most common distinction that needs to be made is between a movement disorder and a seizure. This can often be done by simple observation but in more challenging cases, may require addition diagnostic testing to make this important distinction. The standard for diagnosis of a seizure disorder is an electroencephalogram EEG. In some cases, the identification of the type of movement disorder is all that is needed to come to a diagnosis. For example, a primary tic disorder may be diagnosed on history, examination, and observation alone. In other cases, additional diagnostic testing may be needed. Additional testing may include blood and urine testing, imaging studies, and testing of the cerebrospinal fluid. Diagnostic testing starts with attempting to identify the most common causes of the movement disorder as well as treatable forms of the movement disorder. Increasingly, more extensive genetic testing has enhanced the ability to diagnose movement disorders as well as lead to the discovery on new genetic movement disorders diagnoses. Some movement disorders are considered benign and do not require treatment. Other movement disorders are disruptive to a child, impact their quality of life, and necessitate treatment. Rarely, a pediatric movement disorder can be life-threatening and requires emergent treatment in a hospital. With some movement disorders, the goal of treatment is to lessen the severity of the involuntary movement. In other cases, the goal of treatment is to eliminate the abnormal involuntary movement. Many movement disorders can be difficult to eliminate even with our current best treatments. The benefit of the medication also must be weighed against the risk of potential side effects. For example, a slight decrease in tics might not be worth the sedation caused by a medication used to reduce tics. In cases of acquired movement disorders, it is important to treat the underlying cause. Treatment of the underlying cause may eliminate the movement disorder and obviate the need to treat the movement disorder itself. Tics Regardless of treatment, one-third of children will outgrow tics in adolescence or early adulthood, one-third will have improvement of tics in adolescence or early adulthood, and one- third will have tics that continue to be disruptive in adulthood. The decision to treat tics is based on whether the tics are bothersome or disruptive in day to day life. Often counseling the family to show that the tics are not the sign of an underlying serious neurologic condition provides enough reassurance that medications are not pursued. Providing information to teachers, classmates, and other family members can also reduce the stress caused by tics therefore minimizing the need for medication. In older children, tics may be embarrassing or the children may be teased about their tics. There are medications that can be used to treat tics. These include alpha agonists e. Medications decrease tics on average by one-third to one- half at best. Medication choices can be based on whether the child has tics alone or tics in combination with comorbid conditions. For example, if a child has tics and ADHD, the alpha agonists may be chosen as this medication can be helpful for both conditions. In some children with tics, the associated psychiatric symptoms are more disruptive than the tics themselves. For example, adequate treatment of ADHD may decrease stress at school and at home, secondarily decreasing tics. An alternative to oral medication is a cognitive-based behavioral therapy training program called the comprehensive behavioral intervention for tics. A key component of this behavioral approach is habit reversal therapy. This process teaches tic awareness and the development of a competing response which is an action that replaces the current tic. The child learns to stop the urge to have their tic with no movement at all.

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Even with correct phenomenology identification, secondary movement disorders are difficult to treat regardless of the underlying cause of the involuntary movement. In these settings, it is common for multiple medications to be tried without success.

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The International Parkinson and Movement Disorder Society (MDS) is a professional society of over 7, clinicians, scientists and other healthcare professionals dedicated to improving the care of patients with movement disorders through education and research.

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Experts in the field, Drs. Singer, Mink, Gilbert, and Jankovic, fill the gap in the market by offering the only comprehensive text devoted solely to the diagnoses and treatment of all pediatric movement disorders.