Justin had few friends, which concerned his mother. But when his second grade teacher asked if he had a medical problem or allergies, Justin's mother became worried. Recently his teacher found him frequently coughing, clearing his throat, and at times he seemed to be muttering words under his breath. Justin often blinked his eyes, which his mother had always called "his habit." But now it seemed he was disrupting the classroom, and the other children seemed to be avoiding him. Justin was unaware of any of his "habits," and he often seemed preoccupied with his own thoughts. This did not greatly affect his school performance. He was bright and a good reader, but it was true that sometimes he would miss what the teacher had said, or find himself continuing a project while the rest of the class had gone on to something else. In the evening, if he was reading a book, or watching television, he would be blinking away at rapid pace, until one of his parents would notice and call his name. Then he would look up and the blinking would stop. Justin's father nicknamed him "Blinky," which was cute, but now it was becoming a problem.

Annie was described by her mother as “immature” in her behaviors. She did not relate well to other children or to adults. She often seemed withdrawn with older children or adults, but with her sister or with younger children she would be inappropriately aggressive, taunting, and annoying. She required significant special needs help to keep up with academic work in the fifth grade. In the classroom she was very distracted, and over-active. She had a characteristic behavior of thrusting of her head, or turning of her neck, followed by a grunting noise, which sometimes would occur repetitively for several minutes. Other students couldn't help but notice these movements and sounds, and tended to avoid Annie. Although they did not understand why, there was no doubt that Annie was different. Annie's mother reported that Annie had a first cousin who was diagnosed with Tourette Syndrome.

Jonathan was brought urgently to the local emergency room after his mother spoke briefly on the phone with his pediatrician. That evening, after what was an uneventful dinner, Jonathan had suddenly started having rapid, violent jerking of his entire body. At least it seemed like his whole body was shaking, although his legs actually were moving in response to the extraordinary movements of his upper body and head. The primary concern was that Jonathan was having a seizure. But upon arrival at the hospital, it was clear that Jonathan was not distressed. He was awake, alert, and able to speak clearly without difficulty. Blood tests for blood sugar, calcium, magnesium, sodium, potassium, and other essential chemicals were all normal even as the movements continued. An emergency CT scan of his brain was normal. After a period of observation, and slight improvement, he was seen by a pediatric neurologist, who concluded that the jerking movements were most probably tics. Jonathan was treated with medication, with remarkable improvement over several days.
WHAT WE KNOW ABOUT TICS

Tics are involuntary quick muscle movements, most frequently involving the facial muscles, eye twitching, shoulder or upper body muscles. The movements are rapid jerks, which may occur singly, or in a flurry lasting a few seconds. Sometimes the tics may seem continuous if one flurry of movements follows another. Vocal tics may involve the throat, resulting in gutteral throat-clearing sounds. Sniffling or coughing may be a tic. When the muscles of speech are involved tics may manifest as sudden verbal outbursts. Usually these are not intelligible words, but sometimes distinct, out of context words may be articulated. Coprolalia is the medical term for tics consisting of sudden articulation of slang language, sometimes seen in severe tic disorders (Tourette syndrome).

Tics cannot be controlled by the child. They are involuntary, and originate in a part of the brain which regulates motor movement. A child cannot stop tics from occurring, although the tics may seem to worsen when a child is preoccupied with activities such as reading, or watching television. Upon calling to the child, the tics may stop briefly as the child becomes more "self-aware."

Benign tics often worsen with anxiety, stress, or "nervousness." Many people may experience "eye twitching" when they are "stressed-out", over-tired, or sleep-deprived. Stresses such as school, work, illness, or fever may cause an increase in tics.

When tics occur frequently, not just on occasion, the possibility of a tic disorder should be considered and medical consultation obtained. If the tic persists, even in the absence of any specific stressful situation, it is less likely to be a transient "nervous habit."

Mild "habit tics," or "nervous tics" are quite common, and almost everyone may experience eye blinking or facial twitching under stressful circumstances or when anxious. However, this would not be considered a tic disorder unless the tics were reoccurring regularly and varying in type and duration. True tic disorders are relatively uncommon, but the exact incidence in the population is not known. Some reports suggest that tics of any type may occur as frequently as one in two hundred.

Sometimes a tic will seem to improve, or disappear altogether, only to be replaced by a different tic. It is not unusual for a parent to notice a particular tic, say eye blinking, during several weeks. Then for no obvious reason, the tic stops. But within days another tic, such as throat clearing begins.

Sometimes a child may consciously attempt to suppress the tic, and in fact the tic may improve. However, usually this is short lived, and some children notice that this may result in a worsening of their tic, as though it "rebounds" after being suppressed.

In many children, tics do improve with neurologic growth and maturity, and the tics may resolve completely at any time. This usually occurs over months or years. Other children may
continue to have tics on into adulthood. This is more likely if there is a history of tics in the family.

Tics have been categorized as simple or complex. While the most common are simple motor tics involving muscles of the face and eye twitching, complex motor tics may involve the whole body, hyperactive or aggressive behavior, and inappropriate touching. Simple vocal tics may begin with unusual sounds, such as snorting, grunting, barking, and throat clearing. More complex vocalizations may include distinct words in about half the children who have vocal tics. Coprolalia, unsociable or slang language, and echolalia, repeating words, are complex vocal tics but they are quite unusual. Since tics are sudden, uncontrollable events, they often occur in socially inappropriate circumstances, and may lead to misunderstanding and embarrassment. This may certainly create difficulties for a child in school whose teacher may misinterpret what is happening, and whose friends may be confused by what seems to be unacceptable behaviors.

**Tourette Syndrome**

Georges Gilles de la Tourette was a French neurologist who lived in the early 1800's. He is credited with the first reports of a patient with a neurologically based tic disorder.

Tourette syndrome is the medical diagnosis for severe tic disorders. Tics occur frequently, usually many times a day, and usually vary in their location and type. Both motor and vocal tics are present. The problem continues over many months, and it would be difficult to label tics as Tourette syndrome unless they persisted for at least a year. Tourette syndrome always begins in childhood, usually between age 2 and 13 years, whether or not it persists into adulthood. Boys are affected about four times as often as girls.

The basic chemical process is probably the same for mild infrequent tics as it is for severe cases. The main difference is the quantity, and to some degree the quality, of the tics. If there are vocal tics, or if slang language is occurring, the condition is more serious, and would probably be labeled Tourette syndrome. There is a range from mild infrequent muscle twitch tics (Benign Tic Disorder) to frequent, complex tics (Tourette syndrome).

**Diagnosing Tic Disorders**

Unfortunately, there is no specific test, x-ray, or blood test which can diagnose a tic disorder. The diagnosis is based on the symptoms over a period of time. For this reason it is extremely important that symptoms not be ignored. If you are worried, ask your doctor. An experienced pediatrician or pediatric neurologist can be very reassuring. But even the most experienced physician may not have the opportunity in the office to see the characteristic symptoms which a parent is concerned about. This is particularly true of conditions which cause intermittent symptoms, such as tics. It may be particularly helpful to video tape a child while the tics are occurring. Such a video could be immensely helpful to the physician in arriving at the correct diagnosis.
Sometimes an EEG (electroencephalogram) may be performed on a child with intermittent symptoms. This test measures the electrical function of the brain as it is occurring. It is most helpful in making sure the symptoms are not due to seizures. The EEG should be normal in tic disorders. Also, scans of the brain, such as MRI (magnetic resonance imaging) and CT (computerized tomography) are normal in tic disorders. No specific abnormalities are seen on blood testing, or in spinal fluid. The chemical basis of tics in the brain cannot be measured directly.

**Why Tics Occur**

Tics are caused by a chemical imbalance in the part of the brain which controls and regulates muscle movements. This is not the conscious part of the brain that tells the muscle to move, but a deep unconscious automatic nucleus in the brain that takes over after a muscle movement command is initiated. How remarkable it is that we are able to perform even the simplest movement so effectively. Think about how the brain accomplishes this. If I want to pick up a pencil on my desk, all I need to do is begin the idea that I will pick up the pencil. My brain does the rest automatically, with visual guidance. Do I need to think, "bend the right elbow 20 degrees, rotate the shoulder, extend the elbow, flex the thumb and forefinger on the pencil, flex the elbow...well, you get the point! My brain is doing a whole lot more than what I am consciously thinking. Involuntary movements occur when the brain takes matters into its own hands, so to speak, and attempts to regulate a movement in the absence of the conscious initiating message. This is kind of like a car going into gear even though you did not move the shift lever. Not good.

In order to understand tics, it is necessary to know how the chemicals in the brain work. Think of the brake pedal in your car. If you want the car to stop you must keep your foot on the brake. But if you accidentally step on the accelerator also, the car might momentarily lurch forward a bit. This is how a tic occurs in the brain. Tics are involuntary quick muscle movements or twitches. They occur when neurotransmitters (chemicals, especially dopamine) in the brain act when they are not supposed to. Every time the "accelerator is pushed," a brief muscle movement occurs. This is controlled by a deep, unconscious part of the brain called the basal ganglia. This part of the brain helps to control and modulate both motor activities and sensory perceptions. Recent evidence suggests that rather than too much of the neurotransmitter (dopamine) being released, it may be that some neurons (brain cells) are very sensitive to even very small amounts of dopamine. In either case, the problem is due to an excess dopamine effect on the brain.

There is a lot we do not understand about brain chemistry, and we certainly do not understand exactly what causes tics. However, there are medications which can help to control tics, and the effect of these medications on the brain helps to understand the chemical cause of tics.
Tics and Seizures

Tics are not a type of seizure. Seizures are episodes in which brain cells "fire off" too much electrical activity, which may cause sudden uncontrollable muscle shaking or jerking activity. If the electrical activity of the brain is measured with an electroencephalogram (EEG), it will show abnormal amounts of electrical voltage in the brain. In tic disorders, the EEG is normal. Tics do not cause an excessive electrical discharge in the brain. The reason for this is that tics probably cause a normal electrical discharge in the brain which results in a muscle movement. Also, tics originate in a deep nucleus of the brain, which is not measured by a routine EEG with electrodes on the surface of the head. Tics are due to a chemical imbalance, rather than an electrical abnormality.

The Inheritance of Tics

While tics may develop in children without any history of other relatives or family members with similar symptoms, some studies have found a familial tendency for some cases of tic disorder. Tourette syndrome is thought to be inherited as a dominant trait, which means that it may or may not be transmitted from a parent to a child. The risk for each child would be 50-50, that is, each child would have a fifty percent risk of inheriting the trait. It may be inherited from either father or mother, but sons are somewhat more likely than daughters to inherit Tourette syndrome. The reason for this is not known. The severity of the condition in a child is not necessarily related to the parent's condition, and many children who inherit the trait may have a mild tic disorder which does not require treatment. Other children may have severe Tourette syndrome even though a parent had only a minimal tic.

What other movement problems should be considered?

Seizures, myoclonus, sleep myoclonus, shuttering attacks, paroxysmal choreo-athetosis, chorea.

There are many neurologic conditions which interfere with normal movements or cause involuntary movements. In general, tics can be distinguished from other movement disorders by visible characteristics of the movements. Tics do not affect consciousness, and patients with tics remain alert and awake during tics. There is no mental "spacey-ness" or confusion. After a flurry of tics, the child does not "snap back." The tics simply stop.

In contrast, seizures often involve a decrease in mental alertness, and usually cause unconsciousness. Often there is a "post-ictal" period following the seizure, in which the child is confused, lethargic, or even agitated.

Myoclonus is a rapid jerk-like movement of muscles, usually involving a specific part of the body, such as an arm or leg. It is not usually sustained, and does not usually occur daily. Myoclonus often occurs while falling asleep or waking up, and is experienced by almost everyone. Myoclonus may be confused with tics, but careful observations over a short period of time will distinguish the two conditions. While myoclonus may be a mild, benign movement, it
can be associated with a number of serious neurologic disorders, most of which are very uncommon. It is the job of a pediatric neurologist to consider these possibilities, and when appropriate, do the necessary testing to rule out serious conditions.

Shuttering attacks are infrequent benign movements which occur in young children, usually between about 4 months and 3 years of age. They do not persist and are entirely benign. The child may appear to be shivering, even though the child is not chilled. They usually go on for only a matter of weeks, or possibly months, and then the child will simply "grow out of it."

Chorea is the medical term for involuntary writhing movements of any part of the body, but extremities are most commonly involved. This movement is very different from tics, in that it tends to be slow and sustained for several seconds. However, paroxysmal (sudden) dyskinesia (abnormal movement) is a condition which may be difficult to distinguish from tics. In this condition, the movements begin without warning, although sometimes a sudden movement can precipitate them. Usually they last at least several minutes, and do not alter consciousness. This condition may be inherited. Sometimes it responds to treatment with medications used for seizures, although EEG's do not show any seizure activity.

Movement disorders represent an extremely complex part of the nervous system, which is not well understood. Neurologists are often in the dark about what causes abnormal movements, and there are very few treatments which work effectively. Fortunately, many involuntary movement problems which occur in childhood improve with growth and maturity of the brain. In our knowledge of the brain, we know the least about the chemical reactions, which are the basis of the brain's function as a control center for muscle movement.
HOW TICS ARE TREATED

Should tics be ignored? For the child, it seems easy to ignore the tic. For the parent, it is often impossible to ignore. However, it is very important not to focus on the tic, which may adversely affect a child's self esteem. The child cannot control the tic and should not be punished for it. One young third grader had a vocal tic which was quite active whenever he was working at the blackboard in his classroom. As his teacher considered his "noises" to be impudent behavior, he was sent to a "time-out chair" on several occasions. When the diagnosis of a tic disorder was finally made, the teacher was quite embarrassed and made every effort to support the child for having been inappropriately punished. Do not punish children for tics. Do inform their teacher that the tic activity is a medical condition. Do not assume that teachers and school officials know about tics.

Medications to Treat Tics

The chemical basis of tics allows tics to be treated by medications which block the "leak" of neurotransmitter, primarily, dopamine. Thus, dopamine blocking medications, such as haloperidol (Haldol®) are effective in treating tic disorders. Other medications which have been found to be effective include pimozide (Orap®), a selective dopamine blocker, and clonidine (Catapres®). Less frequently, clonazepam (Klonopin®) and fluphenazine (Prolixin®) have been tried.

None of these medications is a cure. They can be helpful in reducing the frequency of tics, but if the medications are stopped, the tics increase again. Usually, in this situation, restarting medication will control the tics once again. Sometimes after a child has been on medication for a while, the tics may increase again, or change to another type of tic. A child who was having, say, frequent eye blinking, may develop jerking movements of a shoulder when the eye blinking resolves. It is not unusual to need to increase the dose of medication after a period of weeks or months. It is best to start with a low dose of medication in order to avoid side effects, and to determine the least amount of medication necessary to control the tics. The dose can always be increased in subsequent steps. The medications used for treating tics can have significant side effects, and for this reason the decision to treat with medication should be based on the severity of the tics, and the adverse effect on a child's life. When the tics are relatively infrequent and there has been no apparent adverse effect on a child's self-esteem, social adjustment, or learning, then it would be best to avoid using medications. Medication must be taken daily, which in itself is a major change in a child's life-style.

Side effects of medications for tics

The most common adverse effect of medications for tics is drowsiness. This is true for clonidine, although clonidine generally is the least likely of tic medications to cause side effects. Because clonidine may be used to lower blood pressure in hypertensive patients, blood pressure should be checked. However, in reasonable doses, clonidine usually does not lower the blood pressure of an otherwise healthy child.
The best approach to using medication is to start with a low dose and gradually increase the dose provided side effects do not develop. The primary side effect is likely to be drowsiness. If a child is excessively sleepy and the tics have not improved, it is probably time to try another medication. Although haloperidol is generally the most effective medication, it also is the most likely to cause unwanted side effects. Therefore, it is reasonable to start with clonidine first, and pimozide second.

Haloperidol can be quite sedating as the dose is increased. In addition, because haloperidol is a potent dopamine blocker, side effects can include movement problems such as tardive dyskinesia, an ironic side effect when one is treating involuntary movements. Fortunately, abnormal movements such as tardive dyskinesia are rare side effects, and usually improve when the medication is discontinued. Haloperidol may potentially increase the risk of seizures in patients with a seizure disorder, and should be avoided in this population. Other reported side effects of haloperidol include depression, irritability, aggression, and confusion.

Pimozide can be used safely, but it currently is approved as a secondary medication, when other medications have failed or have not been tolerated. It may cause drowsiness, and rarely may adversely affect heart beat.
PROBLEMS ASSOCIATED WITH TIC DISORDERS

What other problems occur in children with tics?

Behavior, attention, adjustment, sleep, obsessive compulsive behaviors.

Tics are purely motor events, involving muscles and movement. They do not affect intelligence or cause progressive decline in cognitive function. However, as many as 35% of children diagnosed with Tourette syndrome also have problems with attention deficit and hyperactivity (ADD, ADHD). Most studies show a frequency of ADD of about 10% in the general school population, which means that children with Tourette syndrome are about three times more likely to have ADD. This may interfere with learning and lead to school failure and behavior disorders. Children with attention problems have difficulty concentrating, organizing their work, and completing tasks. Many children with Tourette syndrome will require a specialized individual education plan (IEP) in school to help deal with learning disabilities. The medical diagnosis of Tourette syndrome may help to provide a clear basis to justify providing educational services. There does not appear to be a common genetic basis of ADD and Tourette syndrome.

Children with Tourette syndrome may have sleep related difficulties, including difficulty falling asleep, awakening at night, sleepwalking, and night terrors. These problems are also seen in children with ADD and other developmental disorders, and may be related to associated underlying neuro-developmental problems, rather a direct result of the tic disorder. These problems may improve as the tic disorder is treated, but may require evaluation by a specialist.

Some children with Tourette syndrome have been found to have Obsessive-Compulsive Disorder (OCD), which may result in purposeless repetitive behaviors, "needing things to be only a certain way," and following pointless rituals in everyday activities. Tics themselves may seem "compulsive" to the child, who may feel a sense of tension building until the tics "release." As many as 50% of patients with Tourette syndrome may manifest obsessive behaviors, and up to 28% of patients were diagnosed with OCD in a study of school children with Tourette syndrome. Some researchers have suggested that both Tourette syndrome and OCD may have a common genetic basis.

Do medications for ADD cause tics?

Probably only in children who were likely to develop tics anyway.

While stimulant medications such as methylphenidate (Ritalin®) and dexamphetamine (Dexadrine®) may worsen tics in a child with a tic disorder, these medications probably do not cause a tic disorder. In a study of children treated with stimulant medications for ADHD, only 1-2% developed tics, and almost all of those resolved when the medication was stopped.
Many children with tics or Tourette syndrome may have symptoms of attention deficit before tics develop. When they are treated with stimulant medication for ADD, tics may appear, but it is likely that those tics would have developed eventually in that child. A family history of a tic disorder raises the likelihood that stimulant medications will cause tics. Many children are treated with stimulant medications for ADD and the vast majority of them never develop tics. Whether or not stimulant medication should be used in a child with tics should be decided on an individual case basis. However, in this situation, clonidine may be a useful medication as it has beneficial effects on attention and behavior as well as tics.

**Why are child psychiatrists sometimes involved?**

Some children with Tourette syndrome may manifest behavioral problems which are best managed psychologically. Child psychiatrists, psychologists, social workers may be experienced with Tourette syndrome, and may be a vital part of a professional team to treat all aspects of a child's function. Psychotherapy or family therapy can significantly improve a family's ability to cope with Tourette syndrome.

**What happens in the future? What is the prognosis?**

Many children with tics will improve as they grow and mature neurologically. Often tics will improve during adolescence, a time when significant developmental maturity occurs in the brain. The incidence of Tourette syndrome is much lower in adults than in children. Sometimes the tics simply "burn out" and resolve. Almost always this is a gradual process which occurs over a period of years. When there is a clear family history of tics, the risk is greater for tics to continue into adulthood, and possibly lifelong. Adults may be more adept at consciously suppressing tics, but it may create some social problems. Tourette syndrome does not affect normal lifespan or general health.

**Are there support groups for parents?**

Although every family and every case of Tourette syndrome is different, family support groups can provide a opportunity for discussion, information, and emotional support. Parents of children with significant tics and Tourette syndrome are constantly coping with the symptoms and its effects on their child's learning, behavior and development. Everyday presents new questions, new problems and dilemmas which demand attention and solution.

Many local support groups exist throughout the country. Your physician may be able to refer you, or else contact the Tourette Syndrome Association, which maintains an extensive list of related services.
Where can I get more information about tics and Tourette syndrome?

Tourette Syndrome Association
42-40 Bell Boulevard
Bayside, NY 11361
(716) 224-2999

The Tourette Syndrome Association can provide further information and valuable support to families of children with Tourette syndrome.

In addition, the Tourette Syndrome Association is a major supporter of research related to Tourette syndrome. Recent research studies have focused on the genetics and inheritance of Tourette syndrome, and the brain chemistry that causes the condition.