

## Sydenham's Chorea

### What is chorea?

Chorea is the name given to specific types of movement disorders characterized by twisting or jerky movements of the body or limbs.

Movement disorders are conditions that cause involuntary body movements. Parkinson's disease, cerebral palsy, and Lou Gehrig's disease (ALS) are all movement disorders.

### Sydenham's chorea

Sydenham's chorea (SC) is the most common form of acquired chorea. Unlike many movement disorders people are born with, it is caused by infection and said to be "acquired." Most frequently, it is caused by Group A beta-hemolytic streptococcal infection. It is part of the spectrum of disorders associated with rheumatic fever.

SC usually does not last. Most cases disappear on their own between 3 to 6 months after first appearing. Cases rarely last longer than one year.

Twice as many girls get SC than boys. Most patients are between 5 and 15 years old.

### Signs and symptoms of SC

With all movement disorders, abnormal signals from the brain cause patients to have trouble controlling the muscles of their bodies.

Children with SC usually have involuntary movements resembling fidgetiness or restlessness. They may also appear to have muscle weakness. These movements usually affect the whole body but may just affect one side. Or they may just affect the face, hands, and arms.

The movements can occur at rest or while active. Movements may increase with distracting activities such as counting or performing mental arithmetic.

Symptoms usually do not appear during sleep.

### Trying to hide the movements

Because the movements are relatively mild, children often try to hide them. They may incorporate an involuntary movement into one that appear to have a purpose. For example, an involuntary head movement may be turned into a flick of the hair. Sometimes children will sit on their hands to try to stop the movements.

Children may seem clumsy and may drop or spill things frequently. Sometimes children have repetitive "piano playing" type movements or they may be unable to hold things in their hands. SC may also cause bursts of uncontrolled speech.

### Longer term

About 3 in 10 children with SC will have a symptom months or years after the disorder has gone away the first time. Usually this happens about 2 years after the first signs of SC have cleared.

As they grow into adulthood, girls may have recurring symptoms if they take birth control pills or estrogen or if they get pregnant.

### Behaviour, feelings, and SC

Sometimes, children with SC also have or develop emotional issues. These might include depression, anxiety, personality changes, being

overly emotional, obsessive-compulsive disorder (OCD), and attention deficit/hyperactivity disorder (ADHD).

It is not known if these emotional effects are part of SC or a result of SC. Sometimes emotional outbursts happen just before involuntary movements begin.

If your child with SC is attending school, you may want to inform the school about your child's condition, including the possible emotional and behavioural issues.

## Diagnosing Sydenham's chorea

Diagnosis of SC can be difficult. There are many different types of movement disorders, many of which have different causes. Often at the beginning, their symptoms appear the same.

Even though SC is caused by a streptococcal infection, the infection may be gone from the body by the time movement symptoms begin.

A patient and family history is taken when diagnosing SC. This is because the doctors will want to find out if the child or other members of the family have had a current or recent infection as well as to distinguish from other types of hereditary chorea.

## Treatment

Sydenham's chorea is often not treated because the symptoms are so mild and the condition will

most likely go away on its own after a few months. More severe cases, where the movements interfere with function, may be treated with medications. These medications may include:

- Anticonvulsants, which can reduce the frequency and severity of movements
- Steroids and intravenous immune globulin (IVIG) may be used in severe and resistant cases. They help to get rid of antibodies which may cause symptoms to worsen.

Children with SC will also be given penicillin for 10 days to prevent rheumatic fever. If your child is having surgery, including dental surgery, she should have penicillin prophylaxis.

The doctor will also assess your child's heart. This is done because chorea is often associated with rheumatic fever. Patients who have Sydenham's chorea may have an affected heart.

## Key points

- Sydenham's chorea (SC) is an acquired movement disorder.
- SC is often mild and usually goes away on its own after a few months. Most cases of SC do not need to be treated with medication or therapy.