



NHS GG&C, Children's Specialist Services

Sydenham's chorea Chorea Information

Your child has been diagnosed with Sydenham's chorea. This leaflet aims to give you as much information as possible about this condition and to help you and your child cope with this condition.

What is Sydenham's chorea?

Sydenham's chorea is a disease affecting the brain. It was described by the English physician Dr Thomas Sydenham in the 17th century. 'Chorea' refers to a type of abnormal movements. Sydenham's chorea happens after a childhood infection because of a problem with the body's immune response to a common infection. The infection is caused by a type of bacteria called Streptococcus. In some children, the immune system reacts in an unusual way to this infection. It affects cells in parts of a child's brain called the basal ganglia. These are a collection of brain cells located deep within the brain and have a role in controlling movement and emotional responses. We do not know the reason why this happens but a risk of Sydenham's chorea can run in families.

Sydenham's chorea can develop up to six months after a child has had an infection. Not all children have a clear history of having had an infection by a bacteria called Streptococcus. Most people who might develop Sydenham's chorea do so before they reach eighteen years of age. It is more common in girls than boys. Sydenham's chorea is part of a wider group of inflammatory responses called rheumatic fever, that may involve other parts of the body including the skin, joints and heart.

What are the symptoms?

Your child may have some or all of the following symptoms:

- **Movement disorder:** often the child's behaviour changes before the onset of the abnormal movements. The movement disorder comes on over a period of hours to days with uncoordinated jerking movements in the arms, hands, legs, feet and face. These movements are called chorea. Sometimes the chorea only involves one side of the body.

- Behaviour changes, like changes in mood and behaviour, fatigue and restlessness.
- Physical symptoms may include loss of fine and gross motor skills, loss of muscle tone, motor and speech difficulties, abnormal patterns of walking, facial grimacing and headache. Some children go on to develop a different kind of movement problem - they may make movements or sounds that they cannot control (often described as “tics”). Other physical symptoms may include inflammation of the heart, and joint pain.
- Some children’s ability to learn may slow down.

How is it diagnosed?

There is no definite test for Sydenham’s chorea but tests may be important to rule out other conditions that might be suspected in some cases. Your General Practitioner (GP), a paediatrician or doctor at an A&E department may make the diagnosis after reviewing your child’s symptoms and a physical examination. They may take a blood sample or throat swab to look for signs of a streptococcal infection. Once Sydenham’s chorea has been suspected your child may be referred to a specialist paediatric doctor called a neurologist who will further assess them. The neurologist will ask questions and examine them, before confirming a clinical diagnosis of Sydenham’s chorea. Your child may need to go for an MRI scan to rule out other conditions but this is not always necessary. Depending on your child’s condition the neurologist may refer them on to other professionals. A list of these can be found in ‘who can help’.

What is the treatment?

Most doctors will try to treat any streptococcal infection by prescribing a course of penicillin followed by a lower dose of this drug to prevent a return of this bacteria. Your child may need to take penicillin until they are 18 years old - this is to prevent the development of inflammation of the heart. The neurologist may try to treat the movement disorder if it is causing problems for your child’s daily living. There has not been a lot of research which helps to decide which medication is best, but doctors often use a

medicine called sodium valproate to help reduce the chorea. Other medicines that can help include a course of steroids, and other drugs including haloperidol, pimozide, carbamazepine and clonidine.

How long will it last?

The movement disorder (chorea) almost always settles down either on its own or with the help of medication. This can take months or even longer and varies from patient to patient. Symptoms can vary over this time, for example the movement disorder is often more noticeable if a child is tired or unwell. The movement disorder can come back later in childhood (a relapse) but will settle down again over time. Doctors used to think that 1 in 5 children with Sydenham's chorea will have at least one relapse during childhood. The more up-to-date opinion is that relapses may affect more children than previously thought. It is very rare for patients to have relapses during adulthood, although it can happen during pregnancy (chorea gravidarum) and in association with the oral contraceptive pill. Some children find that the movements of chorea settle but a new kind of movement can develop. Children with chorea may develop tics or habits that may also come and go. Unfortunately, little is known about the long term effect on behaviour and mood in children who have had Sydenham's chorea. In the majority of cases the young person will make a full recovery after about two years, with no long lasting symptoms, and will not need on-going treatment or follow-up. However some children may experience behavioural and emotional changes that may start before and carry on after the movement disorder and may need ongoing support from a Child & Adolescent Mental Health Service. These changes may come and go and in some cases there are underlying changes in the brain which can continue into adult life. The symptoms that affect a child's mental health may need help and possibly treatment over a longer period of time than the chorea.

How does it affect life?

Sydenham's chorea should not stop your child living and enjoying life. At first chorea movements may make it difficult for your child to get out and about like they normally do. This part of the condition should pass quite quickly as the movement disorder reduces. Some children complain about being tired and this may impact on their ability to enjoy school, hobbies, friendships and family time. This tiredness can be a problem even when there is no heart problem and after movements seem to have settled. It can help to decide with your child which weekly activities are not enjoyable or completely necessary and may be increasing their tiredness. Reducing some of these activities may help to focus their energy and time on getting the best from their body physically and mentally. Your child will build up their strength gradually.

Feelings and behaviour

Some children will develop emotional or behavioural difficulties. These might include: depression, anxiety about separation from parents, personality changes, being overly emotional, obsessive-compulsive disorder (OCD), tic disorders and attention deficit/hyperactivity disorder (ADHD). Sometimes emotional outbursts happen just before involuntary movements begin. Some children are able to cover up symptoms in school but behave very differently at home. This is hard for parents to manage but it is a sign that the child feels safest at home. This may mean that a child can cope in school by letting problems out with the family. The Liaison Psychiatry Team can help with this.

How does this affect a family?

Changes in your child's mental health and their behaviour can be very difficult for parents, grandparents, external family and siblings to adjust to. Some parents talk about seeing their child changing before their eyes. Siblings might find help from local young carers group. Please ask your GP or staff at the hospital about young carers services.

How will Sydenham's chorea affect school?

You and your child might have lots of hospital appointments to attend initially. Together with the disease this can affect your child's attendance at school. Every child has the right to continue their education at times of illness and the school should help to achieve this. It is not helpful for children to be worrying about getting behind with work. It is helpful for parents or carers to link closely with school to minimise the work a child is missing and to help them to keep up. It is useful to pick up school work for your child to complete at home and sometimes teachers may visit your child to complete lessons at home. Starting back at school may be easier with a part-time timetable. You may want to ask the school to refer your child to the school's educational psychologist for additional support.

If your child is attending school, you may want to inform the school about the condition, including the possible emotional and behavioural issues. Your child may notice that their concentration and attention is affected and this can make learning in the classroom harder on some days.

Sydenham's chorea is not well known. Other pupils might be more likely to bully a child, if they don't understand the symptoms. It might be helpful to ask the school to give a lesson on Sydenham's chorea to help educate the class on the condition. Some children find the chorea movements difficult to manage in school and health professionals can help you to think about how you manage this more successfully. For example: sometimes a change of seating position in the class can mean that movements are less troubling for the child and those nearby; extra time in exams and tests may be needed.

It may be helpful for health professionals to visit your child's school to discuss Sydenham's chorea with staff and pupils, explain how it has affected your child and how they can support them.

What can be helpful?

- Getting advice on symptoms and sharing your knowledge with everyone who is involved in your child's care (eg grandparents, school teachers, close friends).
- Learning to live with the condition so that it has as little effect on life as possible.
- Speaking to other children, young people or families who have experience of Sydenham's chorea.
- Visiting the Sydenham's chorea web page or getting in touch with the network:
(www.sydenhamschorea.org.uk)
- Young Carers.
- Action for Sick Children Scotland - helpful to find out about sick children and young people's rights.



Competition winner picture by 11yr old JS

Who can help?

At the Royal Hospital for Sick Children, Yorkhill, you may visit several departments and might have lots of appointments with different professionals.

Department/Service	Their Support Can Include:
<p>Fraser of Allander Neurosciences Unit</p> <p>Paediatric specialist doctors called neurologists, nurses, speech and language therapists, occupational therapists, physiotherapists, neuro-psychiatrists and neuro-psychologists can offer your child different levels of help with the neurological symptoms, chorea movements, sleep difficulties, moods and concentration problems or impulsivity.</p>	<ul style="list-style-type: none"> • Appointments with the various team members for assessment. • Treatment - medication reviews, advice on managing symptoms. • Referral to other professionals.
<p>Neuropsychologist</p> <p>Sydenham's chorea can affect the brain in a way that means children have cognitive difficulties e.g. with learning, memory or mood. This could make things more difficult at school, or with day to day life. Often these problems will improve over time. If you are concerned that Sydenham's has affected your child in these areas, then please discuss this with your consultant and he/she will consider a referral to the neuropsychology team for an assessment.</p>	<ul style="list-style-type: none"> • The neuropsychology team will meet with you to discuss your concerns. • They will carry out an assessment with your child, to uncover any areas of difficulty and also highlight your child's strengths. This assessment normally takes around 2 hours, and involves asking your child to complete a series of tasks; children normally find these tasks quite enjoyable. • The team will get in touch with your child's school and help plan the best ways of supporting your child's learning at this time. • They will also talk to the other professionals involved in your child's care and may make suggestions regarding additional supports that may be useful.

<p>Cardiology</p> <p>Not all cases of Sydenham's chorea will need this service. However, if your child does have cardiac (heart) symptoms paediatric specialist doctors called cardiologists and nurses will help. If there are problems, these usually involve inflammation of the heart valves. If this happens your child will have appointments to monitor the heart's function. They may need medication. Some young people may need a heart valve replacement in later life. It is important to remember that anxiety about the heart can make a child feel heart symptoms with no heart disease present</p>	<ul style="list-style-type: none"> • Appointments with a cardiologist and a nurse with repeat tests • Treatment - your child may need medication and/or surgery
<p>Rheumatology Department</p> <p>Paediatric specialist doctors called rheumatologists, occupational therapists, physiotherapists and nurses may help with joint problems. They may offer physiotherapy sessions and prescribe pain medications if necessary.</p>	<ul style="list-style-type: none"> • Appointments with a rheumatologist, occupational therapist, physiotherapist or nurse. • Treatment - medication, exercise sessions, physical aids and supports.
<p>Paediatric Liaison Psychiatry Team/Neuropsychiatrist</p> <p>Paediatric specialist doctors called psychiatrists, clinical psychologist and nurse therapists who will help with mood difficulties, behaviour, concentration, sleep difficulties, slowed cognition (learning ability), restlessness and fine motor skills (movement of fingers).</p>	<ul style="list-style-type: none"> • Appointments with different members of this team. • Treatment - psychosocial therapies and sometimes medication. • Cognitive testing.

Community based services may be involved from the beginning and will be important if your child has more long term problems:

**Community Paediatrician/
School Nurse**

They may have a role to help coordinate your child's care and link with your child's school and family.

General Practitioner (GP)

Your family doctor will receive all letters and information from health professionals about your child's care. They are responsible for your child's overall care needs. They will look after their physical symptoms and prescribed medication, help with other physical health problems, coordinate health care, liaise with other professionals, and refer to other professionals.

- Appointments when needed.
- Repeat prescriptions when needed.
- Other treatments when needed.

Educational Psychologist

They may help if your child's symptoms affect their progress at school.

**Physiotherapists/Occupational
Therapists**

They can help children with the muscle and joint pains, loss of fine and gross motor skills, loss of muscle tone, motor difficulties and abnormal pattern of walking.

- Appointments - for assessment and advice.
- Treatment - exercise class, homework and splints.

Speech & Language Therapists

They can assess your child's ability to chew and swallow and advise on movements affecting the mouth.

Notes:

Please use this space to write down any questions or useful information for you and your child

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