

Modern illness or a thing of the past?
Surveillance study of childhood/adolescent Sydenham's chorea in the UK and the Republic of Ireland

Abstract	Chorea is the term for abnormal movements described as rapid, irregular, aimless and involuntary. The most common cause of childhood chorea is Sydenham's chorea (SC), which is often accompanied by emotional and behavioural symptoms such as obsessions and compulsions. The condition is associated with prior Group A streptococcal infection and is a major diagnostic criterion for acute rheumatic fever. We plan to study the paediatric service related incidence of SC in 0-16 year olds in the UK and ROI over 2 years. We are asking paediatricians to report cases to us when they first see new cases of suspected or confirmed SC in their service. The notification questionnaire will ask about onset, antecedents, clinical presentation and initial investigations and management. Follow-up questionnaires at one year and two years after notification will enquire about ongoing symptoms, impairment in functioning, investigations, management and outcomes including course and recovery.
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Website	www.rcpch.ac.uk/bpsu/sydenhams
Background	SC is a potentially severe illness with widespread impact which can affect functioning at home and at school. At the peak of their illness, children may become entirely dependent on their families. ¹ Reports suggest that the disorder is often not recognised promptly by professionals, perhaps due to a widespread belief that it is no longer seen in developed countries, but little is known about the current pattern of the disorder. ² This study will help to fill this gap in scientific knowledge, and will be the first such study performed on this condition in the UK and ROI. It will raise awareness of the condition and report on current clinical practice in terms of how such cases are investigated and managed. Data on outcomes will also help clinicians give the most accurate information to families about what to expect.
Coverage	United Kingdom and Republic of Ireland
Duration	November 2018 - November 2020 (25 months surveillance), with a 12-months and 24-month follow-up questionnaire until November 2022.
Research Questions	<ol style="list-style-type: none">1. The paediatric service-related incidence of new presentations of SC in children and young people in the UK and ROI over two years2. The characteristics of children and young people who present with SC, including antecedents of onset of the disorder and the clinical features of their presentation,3. Current practice and variation regarding the diagnosis and management of SC4. The clinical outcomes of children and young people with SC at 12 and 24

- months following presentation to services. These outcomes will include the onset and course of associated physical and psychiatric symptoms/disorders, recovery and recurrence of chorea
5. The impact of SC on children and young people's functioning over a two year period, including on education and activities of daily living.

Case definition

Surveillance Case Definition

According to the Jones criteria for Acute Rheumatic Fever, Sydenham's chorea is defined as "purposeless, involuntary, nonstereotypical movements of the trunk or extremities, often associated with muscle weakness and emotional lability" (3). Sydenham's chorea is typically of acute or subacute onset, meaning that chorea reaches a peak within days or weeks rather than months. The Jones criteria include the differential diagnoses which must be excluded in order to confirm a diagnosis of Sydenham's chorea – these are listed on the notification form. Chorea is frequently a clinical diagnosis. It is important to note that laboratory confirmation of streptococcal infection provides supportive evidence of SC, but absence of such evidence does not preclude clinical confirmation. Cases may be either:

- o Suspected: cases presenting with chorea with acute or subacute onset, but where no diagnosis of SC has yet been made
- o Confirmed clinically: cases where a new diagnosis of SC has been made, with chorea presenting with acute or subacute onset, and lack of clinical or laboratory evidence of an alternative cause as defined by the Jones criteria

Reporting instructions

Please report children and young people aged 0-16 presenting for the first time to you during the reporting period with a first episode of suspected or confirmed Sydenham's chorea (SC) (i.e. with no prior diagnosis of SC)

Methods

- 1) A 25-month surveillance study of paediatric service related incidence of Sydenham's chorea in children and young people aged 0-16 years. Paediatricians reporting a case will be sent a notification questionnaire enquiring about presentation and initial management.
- 2) Follow-up questionnaires sent to the reporting paediatrician at 12-months and 24-months from reporting to capture data on further management, course and outcomes

Ethics approval

This research has been approved by the Confidentiality Advisory Group of the Health Research Association (18/CAG/0086), by the London Bloomsbury NHS Research Ethics Committee (18/LO/0760), and in Scotland by the Public Benefit and Privacy Panel (1819-0077)

Support group

The Sydenham's Chorea Association: <http://www.sydenhamschorea.org.uk/index.php>

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Key References

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