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Childhood/adolescent Sydenham's chorea in the UK and Ireland: a BPSU/CAPSS surveillance study

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ABSTRACT

Objective To conduct the first prospective surveillance study of Sydenham's chorea (SC) in the UK and Ireland, and to describe the current paediatric and child psychiatric service-related incidence, presentation and management of SC in children and young people aged 0–16 years.

Design Surveillance study of first presentations of SC reported by paediatricians via the British Paediatric Surveillance Unit (BPSU) and all presentations of SC reported by child and adolescent psychiatrists through the Child and Adolescent Psychiatry Surveillance System (CAPSS).

Results Over 24 months from November 2018, 72 reports were made via BPSU, of which 43 met the surveillance case definition of being eligible cases of suspected or confirmed SC. This translates to an estimated paediatric service-related incidence rate of new SC cases of 0.16 per 100 000 children aged 0–16 per year in the UK. No reports were made via CAPSS over the 18-month reporting period, although over 75% of BPSU cases presented with emotional and/or behavioural symptoms. Almost all cases were prescribed courses of antibiotics of varying duration, and around a quarter of cases (22%) received immunomodulatory treatment.

Conclusions SC remains a rare condition in the UK and Ireland but has not disappeared. Our findings emphasise the impact that the condition can have on children's functioning and confirm that paediatricians and child psychiatrists should remain vigilant to its presenting features, which commonly include emotional and behavioural symptoms. There is a further need for development of consensus around identification, diagnosis and management across child health settings.

WHAT IS ALREADY KNOWN ON THIS TOPIC

⇒ Sydenham's chorea (SC) is considered a rare disease, but there have been no previous prospective surveillance studies in the UK and Ireland.

WHAT THIS STUDY ADDS

⇒ This study provides the first estimate of the service-related incidence of SC in the UK.
⇒ Although cases reported by paediatricians had high rates of emotional and behavioural difficulties, child and adolescent psychiatrists did not report any cases to this study, suggesting a lack of awareness or involvement.
⇒ The clinical management of SC appears variable in the UK.

HOW THIS STUDY MIGHT AFFECT RESEARCH, PRACTICE OR POLICY

⇒ This study highlights the importance of remaining vigilant to the presenting features of SC.
⇒ This study also describes high rates of emotional and behavioural symptoms, but very limited reports of onward referral to CAMHS, suggesting a need to review how such symptoms are recognised, assessed and managed in paediatric care pathways and consider potential 'triggers' for CAMHS involvement.
⇒ Clinical management was variable, and consensus is needed on optimum diagnostic and management approaches.

INTRODUCTION

Sydenham's chorea (SC) is a neuropsychiatric condition largely affecting children and adolescents,[1] associated with prior Group A Streptococcal infection. SC is a major criterion for rheumatic fever diagnosis[2] and occurs alone or alongside other features. SC is characterised by purposeless, involuntary, non-stereotypical movements of the trunk or extremities (chorea), often associated with muscle weakness and emotional lability. Symptoms range from mild to severe but can severely impact a child's ability to perform activities of daily living.[3] The chorea is often accompanied by emotional and behavioural symptoms, including anxiety, tics, obsessions and compulsions that may be severe and persistent.[4] Symptoms usually resolve within 2 years, but for 20% may become chronic.[5–7]

SC is currently considered a 'rare disease' but it is unclear how many children are affected by it, their clinical journey or their needs. Previously, only single-centre retrospective studies in paediatrics have taken place in Western Europe, but such methods have limitations in capturing cases and clinical detail.[8 9] Reports also suggest that SC is often not recognised promptly.[6 7 10] Rare conditions may have greater impact on families, who may become more isolated, lack information on prognosis and experience more diagnostic delays.[11–16] This research was therefore developed in partnership

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Table 1 Case definitions for BPSU study

Surveillance case definition	Children and young people aged 0–16 years presenting to the reporting clinician for the first time in the surveillance period with a first episode of suspected or confirmed Sydenham's chorea (SC).
Reporting instructions for paediatricians	Please report: ► Children and young people presenting to you for the first time in the surveillance period with a first episode of suspected or confirmed SC. ► According to the Jones criteria for acute rheumatic fever, SC is defined as 'purposeless, involuntary, non-stereotypical movements of the trunk or extremities, often associated with muscle weakness and emotional lability'. ► SC is typically of acute or subacute onset, meaning that the chorea reaches a peak within days or weeks rather than months.[2] ► The Jones criteria list the differential diagnoses which must be excluded in order to confirm a diagnosis of SC (detailed in full in the reporting card). ► Chorea is frequently a clinical diagnosis. It is important to note that laboratory confirmation of streptococcal infection provides supportive evidence of SC, but the absence of such laboratory evidence does not preclude clinical confirmation.
	Please report cases presenting to you for the first time during the surveillance period, who are new cases of suspected or confirmed SC (with no prior diagnosis before the current episode). These cases may be either: ► Suspected: cases presenting with chorea with acute/subacute onset, but where no diagnosis has yet been made. ► 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 above by the Jones criteria.

BPSU, British Paediatric Surveillance Unit.

with the family-led charity, the Sydenham's Chorea Association (http://www.sydenhamschorea.org.uk/).

This paper describes the first prospective surveillance study of SC in the UK and Ireland, with paediatricians via the British Paediatric Surveillance Unit (BPSU) and with child and adolescent psychiatrists through the Child and Adolescent Psychiatry Surveillance System (CAPSS).[17] The parallel surveillance method was chosen as children with SC may be seen by child psychiatrists due to the high prevalence of emotional and behavioural symptoms.

The study aimed to answer the following three main questions:

- What is the paediatric service-related incidence of first presentations of SC in children and young people aged 0–16 years in the UK and Ireland?
- What is the psychiatric service-related incidence of children and young people with SC and associated psychiatric symptoms in the UK and Ireland?
- What are the most common presenting features of SC, and what is the current clinical practice in terms of investigations, management and referral?

METHODS

BPSU surveillance

We conducted a prospective surveillance study of suspected or confirmed first presentations of SC in children and young people aged 0–16 years (case definition in table 1) with paediatricians in the UK and Ireland from November 2018 to November 2020 via the BPSU using their standard 'Orange card' anonymous notification methodology,[18] summarised in figure 1. SC is frequently a clinical diagnosis, based on the distinctive presentation.[1] Laboratory confirmation of streptococcal infection and echocardiographic findings provide supportive evidence of SC. However, the absence of these does not preclude clinical diagnosis, as SC onset may be delayed following infection with no persisting serological evidence.[1 10] Consequently, our case definition of confirmed cases included all clinically confirmed cases.

Questionnaires were piloted by paediatricians before finalising and included items on characteristics of case and service; clinical presentation and antecedents; investigations and management, referrals, service-related outcomes, impairment and impact on activities of daily living using the Universidade Federal de Minas Gerais (UFMG) SC rating scale[19]; impact on education (see online supplemental material).

CAPSS surveillance

CAPSS uses a very similar surveillance methodology to BPSU, with monthly e-cards sent to child and adolescent psychiatrists registered with the system.[20] The case definition (table 2) and questionnaire (online supplemental materials) differed between the CAPSS and BPSU, capturing service-related incidence of any presentations of SC or associated psychiatric symptoms to child and adolescent psychiatrists, not only new onset SC, as psychiatric symptoms may appear much later.[4] Surveillance through CAPSS took place from May 2019 to December 2020 (18 months).

Analysis

Our analysis of characteristics included all cases meeting the surveillance case definition were analysed, with sensitivity analysis in Stata V.17.0 performed using confirmed cases.[21] Descriptive analysis was performed presenting data rounded percentages for the full sample following statistical disclosure principles. Where possible, incidence was calculated using official national mid-year population estimates for children aged up to 16 for the appropriate geographies.[22]

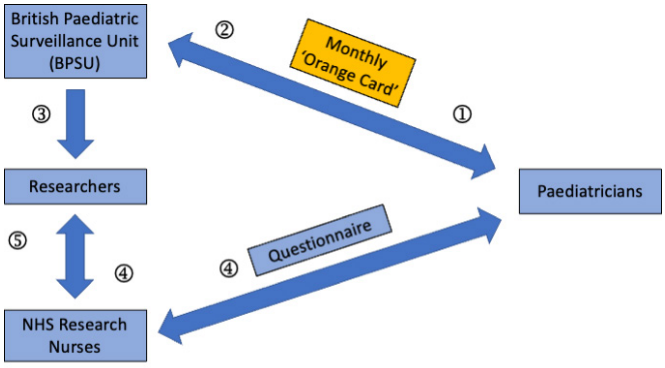

Figure 1 A summary of the BPSU 'orange card' surveillance study reporting and follow-up process. BPSU, British Paediatric Surveillance Unit.

Table 2 Case definition and reporting instructions for CAPSS study

Surveillance case definition	Children and young people aged 0–16 years that have or have had a suspected or confirmed diagnosis of Sydenham's chorea (eg, by paediatrician) and present to the reporting clinician for the first time within the current episode of care with psychiatric symptoms.
Reporting instructions to child and adolescent psychiatrists	Please report: ► Children and young people aged 16 years or under who have or have had a suspected or confirmed diagnosis of Sydenham's chorea (eg, by paediatrician) and present to you for the first time within the current episode of care (regardless of whether this is their first contact with CAMHS or not) with one or more psychiatric symptoms. ► Chorea is defined as a state of excessive, spontaneous movements, irregularly timed, non-repetitive, randomly distributed and abrupt in character. These movements may vary in severity from restlessness with mild intermittent exaggeration of gesture and expression, fidgeting movements of the hands, unstable dance-like gait to a continuous flow of disabling movements. ► Sydenham's chorea is frequently a clinical diagnosis. Laboratory confirmation of streptococcal infection (eg, by throat swab) provides supportive evidence of Sydenham's chorea but the absence of investigation does not preclude clinical confirmation. Cases may fall into any of these categories: – Suspected Sydenham's chorea: cases presenting with chorea but where no clear diagnosis has yet been made. – Confirmed Sydenham's chorea: where a diagnosis of Sydenham's chorea has been made (eg, by paediatrician) and alternative causes have been excluded.

CAPSS, Child and Adolescent Psychiatry Surveillance System.

RESULTS

Ascertainment

Over 24 months, 72 reports were made via BPSU, of which 43 met case definition (n=12 suspected, n=31 confirmed cases). The remainder were ineligible (n=8 not SC or not new, n=6 duplicates, n=15 unable to confirm). Returned questionnaire response rate was 79% (57/72).

No reports were made via CAPSS over the 18-month reporting period.

Incidence

Based on a mid-year UK population estimate (including all four nations) of 13 468 262 children aged 0–16 years, the cases (suspected and confirmed) reported to our study would result in a paediatric service-related incidence rate of new SC cases of 0.16 per 100 000 children per year (95% CI 0.11 to 0.25). Using only confirmed cases to generate an estimate, the incidence rate would decrease to 0.12 per 100 000 per year (95% CI 0.07 to 0.19). Applying an alternative assumption, that 84.3% of the cases lost to follow-up would meet the case definition (using the rate of ineligible or duplicate cases of 15.7% among returned questionnaires), produces an incidence estimate of 0.21 per 100 000 per year (95% CI 0.15 to 0.31). There were no reported cases from the ROI, hence no incidence estimate was calculated.

Characteristics

Mean age of presenting cases was 9.4 years (range 4–16), with 68% being female. Of these cases, 87% were of white ethnic background, slightly exceeding the UK Census estimates (81.7%).[22] About 74% of cases reported were from England; few cases were from Wales (n=<5), Scotland (n<5) or Northern Ireland (n<10). Of these, 55.6% of cases were reported by consultant general paediatricians and 30.2% by consultant paediatric neurologists.

Presenting features

Categories of clinical features at presentation are reported in figure 2. All cases had chorea, as dictated by our case definition, which was moderately severe at presentation in 72% of cases (n=31), with the remaining either severe (14%), or minimal or mild (7%). The most common presenting symptoms (besides chorea) were gait disturbance and loss of fine motor skills, both of which were reported in 91% of cases (n=39).

Overall, at least one neuropsychiatric, emotional or behavioural feature was reported in 86% (n=37) of cases. Emotional lability was seen in 77% of all cases, some form of anxiety was present in 51%, tics in 37% and inattention/attention deficit in 35%. Other criteria for rheumatic fever (besides SC itself) such as erythema marginatum, subcutaneous nodules, carditis and polyarthritis appeared rare as presenting features in this sample (see figure 2). A sensitivity analysis of presenting features in confirmed cases only found no statistically significant differences in presentation (p<0.05 threshold).

Clinicians were asked to rate the child's functioning at presentation.[18] The mean score on the UFMG rating scale functional impairment domains was 2.0 (0 is 'no impairment' and 4 is 'severe impairment'); 51% of children had severe impairment due to chorea on at least one of the six domains (hygiene, handwriting, dressing, speech, walking and handling utensils).

Investigations and results

In all reported cases, anti-streptolysin titre (ASOT) was performed. ASOT measures levels of anti-streptolysin antibodies, indicating previous streptococcal infection. Nineteen per cent of reported cases had an elevated ASOT (200 IU/mL or above) and the mean titre was 600 IU/mL. The majority of patients had an ECG (95%), MRI head (86%), echocardiogram (84%) and a throat swab for microscopy, culture and sensitivity (77%). Other tests reported as performed are presented in online supplemental table 1.

Antecedents

About 54% (n=24) of cases reported sore throat in the 6 months prior to the onset of chorea and 21% were diagnosed with acute

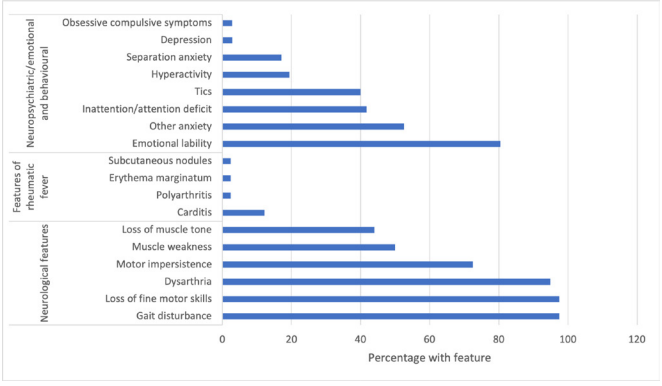

Figure 2 Presenting features of children with suspected or confirmed Sydenham's chorea (SC). *Where responses were blank or not known they were excluded from the denominator.

rheumatic fever (ARF) according to paediatrician report. There was only one case with a family history of SC and no reports of family histories of rheumatic fever or other movement disorders.

Management

About 77% of patients (n=33) were admitted to hospital during the clinical episode, with a median stay of 5 days. All but one were prescribed antibiotics, but dose and duration were not specified by most respondents. The most commonly prescribed course was 10 days of penicillin V followed by long-term prophylaxis (23%, n=10), but antibiotic regimes varied. Other treatments used included symptomatic treatment of chorea with anticonvulsants and neuroleptics (53%, n=23), most commonly sodium valproate, haloperidol or carbamazepine. About 28% of patients received immunomodulation, with 16% prescribed steroids, and 12% receiving intravenous immunoglobulins. A total of 51% of children were referred to occupational therapy or physiotherapy, and 14% to clinical psychology or neuropsychology. Fewer than five patients were referred to CAMHS.

DISCUSSION

Our estimates of incidence of 0.16 per 100 000 per year (or 0.21 per 100 000 corrected for reports which could not be followed up) are the first prospective estimates of incidence of SC for children in the UK, and align with Crealey *et al*'s earlier retrospective case note study from the Republic of Ireland, which estimated an incidence rate of 0.23 per 100 000.[10] We report similar age and gender profiles of children to other epidemiological studies from other settings.[23 24] The very low incidence of family history of SC identified in this study is comparable with recent Italian and Turkish findings.[9 25]

In the vast majority of cases, chorea was associated with moderate or severe functional impairment. 'Mild' cases may not be identified or recognised by families or professionals, reducing specialist referrals. The most common presenting features reported (besides chorea) were gait disturbance, loss of fine motor skills and dysarthria. Our reported rates of carditis (12%) were much lower than the rates of 'around 60%' found in an older Australian study and 81% presented by Orsini *et al* in 2022.[9 26] Despite the association of SC and rheumatic heart disease, 16% were reported as having no echocardiogram.

Neuropsychiatric, emotional and behavioural symptoms were common in line with the systematic review by Punukollu *et al*.[4] However, most children were not referred to CAMHS and no SC cases were reported via CAPSS over an 18-month period. This may be because co-occurring emotional and behavioural problems are not recognised or prioritised during the acute presentation of chorea, due to low confidence among paediatricians in assessing and making appropriate referrals, or because symptoms may be managed within paediatric services rather than referred onto CAMHS.

Alternatively, the absence of case reports from CAMHS consultants may be due to children being seen in CAMHS but not seen by the consultant child and adolescent psychiatrists, who are reporting into CAPSS, or to clinicians in CAMHS not making the link between a child's emotional or behavioural symptoms and a history of SC.

Strengths and limitations

The main strength of this study is the use of established prospective active surveillance methodology through the BPSU and CAPSS, meaning that clinicians are requested to return a response monthly even if no cases have been seen. The BPSU had an 88% response rate for surveillance cards in 2020.[27] In this study, another strength was also the high response rate: 80% of all questionnaires sent out were returned.

There are a number of weaknesses to consider in the methodology. During our study period, CAPSS had a lower response rate for surveillance cards (an average of 60% over the study period), meaning that cases may have been seen but not notified. Furthermore, while the BPSU and CAPSS achieve high coverage of consultants, there is a delay in updating contact lists which may mean not all new consultants or retiring consultants are included in or removed from databases. Furthermore, where cases are seen by other team members, they will not be reported unless specifically notified to the consultant. It is also possible that non-response to both the surveillance cards and the questionnaires may be associated with characteristics such as a particularly heavy workload. However, without data from non-responders or cases that might be seen elsewhere, we are unable to adjust our incidence estimates to reflect these effects.

Finally, after extensive consultation, and as SC is a clinical diagnosis, consultants were given the option to report cases as suspected or confirmed. We therefore report incidence using both our surveillance definition of confirmed or suspected SC; and our analytic definition of confirmed SC only, but present the remaining data on presentation and management using the surveillance definition.

Implications for research and practice

This study contains a number of implications for research and practice. First, it confirms the need for clinicians to remain vigilant to the presenting features of SC. Although a rare condition, it has not disappeared, and the impact on children's functioning can be severe.

Under-reporting of SC may also be linked to a question that has arisen in discussions with paediatricians and child psychiatrists, as well as through involvement with patient groups. This is the distinction between SC and paediatric autoimmune neuropsychiatric syndrome (PANS)/paediatric autoimmune neuropsychiatric disorder associated with *Streptococcus* (PANDAS). The distinctive feature of SC is the presence of clinically significant chorea resulting in functional impairment at disease onset.[4 28] However, there are overlaps in presenting features. While SC has been thought of as 'a thing of the past', PANS and PANDAS have an increasingly high profile. Ensuring that SC is recognised is important clinically as it requires long-term penicillin prophylaxis to prevent further attacks of ARF, sequelae of rheumatic heart disease and recurrent SC. Consensus development is needed in this area to combine expertise and evidence and provide greater clarity for clinicians and families.

Similarly, the findings do suggest variability in clinical management reported in our study, and in particular the use of antibiotics and immunomodulation, implying that harmonisation of management is merited. However, while there is evidence for the effectiveness of steroids, further well-conducted trials are needed to determine best practice.[24] There are challenges in conducting trials in rare disease research, but there are opportunities to address these through greater international collaboration, use of electronic medical records and adaptation of standard trial designs.[29]

In spite of the high proportion of psychiatric comorbidities among the children and young people reported there was minimal onward referral to CAMHS services and no reporting of cases from child and adolescent psychiatrists via CAPSS, this finding warrants further exploration. Assessment and

management in SC should take account of the high level of emotional and behavioural symptoms among children with SC; for example, including appropriate validated assessment tools for these symptoms and management strategies within guidance and care pathways.[30]

Finally, the surveillance we report here focusses on first presentations of SC in children. There has been very limited study of the course of SC over longer periods of time, and into adulthood, although older data suggest that recurrence of chorea may occur in up to 20 to 40% of cases.[7 31]

CONCLUSIONS

Our estimates of incidence suggest that SC remains a rare condition in the UK and Ireland but has not disappeared. Indeed, our findings also emphasise the impact that the condition can have on children's functioning and confirm that paediatricians and child psychiatrists should remain vigilant to its presenting features. The findings also highlight the need for further research and development of consensus around diagnosis and management.

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Contributors The study was designed with input from a Steering Committee who also advised on development of the questionnaires and study protocol. The steering committee included all authors (TN-D, MJSM, ML, OM, NM, ELW, AS, BK and TF). TN-D and OM oversaw the surveillance studies at BPSU and CAPSS, respectively. TN-D and ELW collated and presented the data which was discussed with all authors. ELW and TN-D led on manuscript drafting and editing and all authors contributed to the revision of all versions and approval of the final manuscript. TND is guarantor for the study, accepts full responsibility for the finished work and/or the conduct of the study, had access to the data, and controlled the decision to publish.

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Competing interests MJSM is Honorary President of the Sydenham's Chorea Association.

Patient consent for publication Not applicable.

Ethics approval Ethical approval was provided by the London Bloomsbury Research Ethics Committee (18/LO/0760). Approvals were obtained from the Confidentiality Advisory Group Ref: 18/CAG/0086 and the Public Benefit Patient Panel Ref: 1819-0077.

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BPSU No. D D/ N N/ Y Y

Case notification form Questionnaire – Strictly Confidential

Version 3

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

The first page of the case notification form will be stored separately from the rest of the questionnaire and personal identifying information for the case will be used only for linkage of records.

Reporting Instructions:

Please report any child or young person that you have contact with for the first time who presents with a first episode of **suspected** or **confirmed Sydenham's chorea.**

Please report any case even if you believe the case may have been reported from elsewhere.

Case Definition:

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)

Sydenham's chorea is a poststreptococcal autoimmune movement disorder, of which chorea is the hallmark. It is also one of the major manifestations of rheumatic fever**. 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"** (Gewitz et al., 2015). 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 below.

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:

- Suspected: cases presenting with chorea with acute or subacute onset, but where no diagnosis of SC has yet been made
- 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 below by the Jones criteria

Differential diagnoses of SC according to the Jones criteria

Drug intoxication	Lyme disease
Wilson disease	Hormonal e.g. hyperthyroidism
Tic disorder	Metabolic
Choreoathethoid cerebral palsy	Antiphospholipid antibody syndrome
Encephalitis	Autoimmune: e.g. Systemic lupus erythematosus
Familial chorea (including Huntington disease)	Vasculitis
Intracranial tumour	Sarcoidosis

BPSU No. D D / M M / Y Y

Section A: Reporter Details

1.1 Date of completion of questionnaire: D D / M M / Y Y Y Y

1.2 Consultant responsible for case: _____

1.3 Name of Person completing form (if not the above) _____

1.3 Name of clinic and Trust/Provider: _____

1.4 Telephone number: _____ Email: _____

Section B: Case Details

2.1 NHS/CHI No: ☐☐☐ ☐☐☐ ☐☐☐☐

2.2 Hospital No: ☐☐☐☐☐☐☐☐☐☐

2.3 First half of postcode only: ☐☐☐☐ If ROI - Town of Birth: _

2.4 Sex: M ☐ F ☐

2.5 Age of case: _ _ _ years _ _ _ months

2.6 Ethnicity*: ☐☐ If any "Other" background:, please specify: _

**Please choose the correct ethnicity code from Appendix A below*

Appendix A: Coding for Ethnic Group (ONS 2011)-

		Ethnicity Code	
A	**White**	**1**	Any White background
B	**Mixed**	**2**	White and Black Caribbean
		3	White and Black African
		4	White and Asian
		5	Any Other Mixed background, *please write in* ***section B***
C	**Asian or Asian British**	**6**	Indian
		7	Pakistani
		8	Bangladeshi
		9	Any Other Asian background, *please write in* ***section B***
D	**Black or British Black**	**10**	Caribbean
		11	African
		12	Any Other African background, *please write in* ***section B***
E	**Chinese or other ethnic group**	**13**	Chinese
		14	Any Other, *please write in* ***section B***
F	**Unknown**	**15**	Ethnicity not known

Wooding EL, *et al. Arch Dis Child* 2023;0:1–6. doi: 10.1136/archdischild-2023-325399

BPSU No. D D / M M / Y Y

Section C: Eligibility of case

3.1 Does the case meet the following criteria for this study?

	Yes	No
1. In **this** clinical presentation, is there a **history of acute or subacute chorea**, as defined above?	☐	☐
2. Does the case have currently have a **suspected or confirmed** diagnosis of Sydenham's chorea? (see case definition above).	☐	☐
If yes, please indicate whether suspected or confirmed by ticking one of the boxes below:		
• Suspected Sydenham's chorea	☐	
• Clinically confirmed diagnosis of Sydenham's chorea, where other causes of the presentation have been excluded (please see Jones criteria in case definition)	☐	
3. Is this a first episode of suspected or confirmed Sydenham's chorea?	☐	☐

Section D: Onset and severity of the chorea

4.1 When was the onset of this episode of chorea?

__/__/____

4.2 How would you rate the severity of the chorea at its worst so far? (Please tick one)

Minimal (action chorea, or intermittent rest chorea) ☐

Mild (continuous rest chorea, but without functional impairment) ☐

Moderate (continuous rest chorea with partial functional impairment) ☐

Severe (continuous rest chorea with complete functional impairment) ☐

BPSU No. D D/ N N/ Y Y

Section E: Presentation/Clinical features

5.1 Have the following features been present during the current episode? Please tick :

	Yes	No	Don't know
Muscle weakness	☐	☐	☐
Loss of muscle tone	☐	☐	☐
Gait disturbance	☐	☐	☐
Dysarthria	☐	☐	☐
Loss of fine motor skills	☐	☐	☐
Motor impersistence (the inability to maintain postures or positions without repeated prompts)	☐	☐	☐
Tics	☐	☐	☐
Carditis	☐	☐	☐
Polyarthritis	☐	☐	☐
Erythema Marginatum	☐	☐	☐
Subcutaneous nodules	☐	☐	☐
Separation anxiety	☐	☐	☐
Other anxiety	☐	☐	☐
Obsessive-compulsive symptoms	☐	☐	☐
Emotional lability	☐	☐	☐
Depression	☐	☐	☐
Hyperactivity	☐	☐	☐
Inattention/attention deficit	☐	☐	☐

Other features, please specify_____

BPSU No. D D/ M M/ Y Y

Section F: Antecedents

6.1 Does the case have a history of pharyngitis/sore throat in the 6 months prior to the start of this episode of chorea?

Yes ☐ No ☐ Not known ☐

6.2 Has the case been diagnosed with Acute Rheumatic Fever? :

Yes ☐ No ☐ Not known ☐

6.3 Is there any family history of the following? Please tick whichever apply:

Sydenhams' chorea or St Vitus Dance ☐ Other movement disorder ☐ Rheumatic fever ☐

6.4 Is the case currently taking the oral contraceptive pill?

Yes ☐ No ☐ If yes, for how long have they been taking it? _____months _____years

6.5 Are there any other suspected triggers for the onset of chorea?

Yes ☐ No ☐ If yes, please specify_____

Section G: Investigations

7.1 During this episode of chorea, has there been evidence of prior streptococcal infection? (as confirmed by throat swab, serum anti-basal ganglia antibodies level or antistreptolysin O titre?)

Yes ☐ No ☐ Not known ☐

7.2 Have any of the following investigations been performed so far during this episode of chorea?

	Yes	No	Don't know	Result if known
Serum anti-basal ganglia antibodies level (ABGA)	☐	☐	☐	Positive ☐ Negative ☐
Throat swab for streptococcus	☐	☐	☐	Positive ☐ Negative ☐
Antistreptolysin O titre (ASOT)	☐	☐	☐	_____U/ml
Echocardiogram	☐	☐	☐	Result not required
MRI head	☐	☐	☐	Result not required
ECG	☐	☐	☐	Result not required
Other – please specify and give results if relevant				

BPSU No. D D/ M M/ Y Y

Section H: Initial management

8.1 During the current clinical episode, has the case been admitted as an inpatient at any point?

Yes ☐ No ☐

If yes, for how long in total were they or have they been an inpatient?____days _____weeks

8.2 Referrals: In the course of this episode, has the case been referred to any of the following specialties or services?:

	Yes	No	Not Known
Paediatric neurology	☐	☐	☐
Paediatric cardiology	☐	☐	☐
Paediatric rheumatology	☐	☐	☐
Clinical psychology or neuropsychology	☐	☐	☐
Physiotherapy and/or occupational therapy	☐	☐	☐
Educational psychology	☐	☐	☐
Child and adolescent psychiatry/CAMHS	☐	☐	☐

Other? Please specify:

8.3 Has initial management included any of the following?

	Yes	No	Not Known
Course of antibiotics	☐	☐	☐
If yes, please give antibiotic, and duration_____			
Pharmacotherapy for symptomatic treatment of chorea (e.g. valproate, haloperidol)	☐	☐	☐
If yes, please give drug name_____			
Immunotherapy	☐	☐	☐
If yes, please give drug name _____			
Psychological or psychiatric interventions or pharmacotherapy	☐	☐	☐
If yes, please give details if known _____			
Physiotherapy and/or occupational therapy	☐	☐	☐
If yes, please give details if known _____			
Review by special educational needs coordinator or educational psychologist	☐	☐	☐
If yes, please give details if known _____			

BPSU No. D D/ N N/ Y Y

Section I: Impact on functioning

9.1 At the time the case first presented to you, how would you rate the impact of their symptoms on the following activities of daily living?

Please tick one from each row

Domain	No impairment	Minimal impairment	Mild impairment	Moderate impairment	Severe impairment
Hygiene (e.g. washing, using toilet)	(normal)	(difficulty with hygiene tasks, but no help needed)	(help needed on less than half of occasions)	(help needed on half or more of occasions)	(completely dependent)
Dressing	(normal)	(difficulty with dressing, but no help needed)	(help needed on less than half of occasions)	(help needed on half or more of occasions)	(completely dependent)
Speech (dysarthria)	(absent)	(impairment present, but completely comprehensible, or speech easily understood)	(less than 25% of the speech is incomprehensible, or some difficulty in understanding speech)	(25–50% of the speech is incomprehensible, or marked difficulty in understanding speech)	(incomprehensible speech)
Handwriting	(normal)	(compromised handwriting, but all words are legible)	(not all words are legible)	(the majority of words are not legible)	(impossible handwriting)
Walking	(normal)	(walks with difficulty, but does not run into objects)	(walks with difficulty, running into objects)	(walks only with assistance)	(chorea paralytica; cannot walk at all, even with assistance)
Cutting food & handling utensils	(normal)	(difficulty with these tasks, but no help needed)	(occasional help needed, e.g., cutting meat)	(frequent help needed)	(needs to be fed)

Section J: You and your service

10.1 Please indicate your current role: Consultant in Community Paediatrics Consultant in General Paediatrics Consultant Paediatric Neurologist Consultant Paediatric Cardiologist Other – please specify:_____

10.2 Please indicate the service in which you saw this patient: Paediatric inpatient service Paediatric outpatient service – community paediatrics Paediatric outpatient service – cardiology Paediatric outpatient service – neurology Other – please specify_____

BPSU No. D D / N N / Y Y

Thank you for taking the time to complete the questionnaire

Please return the completed form by secure nhs.net mail to :

_____

If you have any questions about the study please do not hesitate to contact the investigators by email or telephone :

Telephone: _____ Email: _____

Wooding EL, *et al. Arch Dis Child* 2023;0:1–6. doi: 10.1136/archdischild-2023-325399

CAPSS CHILD & ADOLESCENT PSYCHIATRY SURVEILLANCE SYSTEM CCQI

Case notification form - Strictly Confidential

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

The first page of the case notification form will be stored separately from the rest of the questionnaire and personal identifying information for the case will be used only for linkage of records.

Reporting Instructions:

Please report any child or young person aged 0-16 years that you have contact with **for the first time within the current episode of care** for psychiatric symptoms and who **has or has had Sydenham's chorea**.

Please report any case even if you believe the case may have been reported from elsewhere.

Case Definition:

Please report:

Children and young people aged 0-16 years who **have or have had a suspected or confirmed diagnosis of Sydenham's chorea** (e.g., by paediatrician) and **present to you for the first time within the current episode of care** (regardless of whether this is their first contact with CAMHS or not) with one or more psychiatric symptoms.

Sydenham's chorea is a post-streptococcal autoimmune movement disorder defined as purposeless, involuntary, non-stereotypical movements of the trunk or extremities, often associated with muscle weakness and emotional lability. It **i**s frequently a clinical diagnosis (absence of laboratory confirmation of streptococcal infection does not preclude clinical confirmation). Cases may be either:

- Suspected: cases presenting with chorea but no diagnosis of Sydenham's chorea has yet been made.
- Confirmed: cases where a diagnosis of Sydenham's chorea has been made and alternative causes have been excluded.

Wooding EL, *et al. Arch Dis Child* 2023;0:1–6. doi: 10.1136/archdischild-2023-325399

Section A: Reporter details

1.1 Date of completion of questionnaire: D D / M M / Y Y Y Y

1.2 Consultant responsible for case: _____

1.3 Name of person completing form (if not the above): _____

1.4 Name of clinic and Trust/Provider: _____

1.5 Telephone number: _____ Email: _____

Section B: Case details

2.1 NHS/CHI No: ☐☐☐ ☐☐☐ ☐☐☐☐

2.2 Hospital No: ☐☐☐☐☐☐☐☐☐☐

2.3 Postcode: ☐☐☐☐ Town of Birth (if ROI) _____

2.4 Sex: M ☐ F ☐ Age: _ _ _ years _ _ _ months

2.5 Ethnicity*: ☐☐ Specify if any "Other" background _____

**Please choose the correct ethnicity code from Appendix A below*

Appendix A: Coding for Ethnic Group (ONS 2001)

		Ethnicity Code	
A	**White**	**1**	Any White background
B	**Mixed**	**2**	White and Black Caribbean
		3	White and Black African
		4	White and Asian
		5	Any Other Mixed background, *please write in* ***section B***
C	**Asian or Asian British/Irish**	**6**	Indian
		7	Pakistani
		8	Bangladeshi
		9	Any Other Asian background, *please write in* ***section B***
D	**Black or British/Irish Black**	**10**	Caribbean
		11	African
		12	Any Other African background, *please write in* ***section B***
E	**Chinese or other ethnic group**	**13**	Chinese
		14	Any Other, *please write in* ***section B***
F	**Unknown**	**15**	Ethnicity not known

Wooding EL, *et al. Arch Dis Child* 2023;0:1–6. doi: 10.1136/archdischild-2023-325399

Section C: Eligibility of case

3.1 Does this case meet the following criteria for this study?	Yes	No
1. Does this patient currently have a confirmed diagnosis of Sydenham's chorea?	☐	☐
2. Does this patient currently have a suspected diagnosis of Sydenham's chorea?	☐	☐
3. Does this patient have a past history of an episode with a confirmed diagnosis of Sydenham's chorea?	☐	☐

Section D: Clinical setting

4.1 Please indicate the setting in which you first saw this patient within the current episode of care (tick all that apply):

- ☐ CAMHS outpatient clinic or outreach service
- ☐ CAMHS inpatient or day patient unit
- ☐ Paediatric or general hospital setting
- ☐ Other (please specify) _____

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Section E: Psychiatric presentation

5.1 Please record the main presenting symptoms and any psychiatric diagnoses applicable during this episode of care (e.g., depressive symptoms, anxiety, emotional lability, obsessions and compulsions, motor or vocal tics, hyperactivity, inattention, fatigue, etc.)

_____

_____

5.2 When have the presenting symptoms been first identified?

a) Onset clearly preceding the first episode of Sydenham's chorea ☐

and current presentation is an exacerbation of these pre-existing psychiatric symptoms ☐

b) Onset during the first episode of Sydenham's chorea ☐

c) Onset clearly following the first episode of Sydenham's chorea ☐

5.3 In addition to the presenting problems, has the patient had any other psychiatric symptoms or diagnoses (either ongoing or resolved)?

Yes ☐ and the onset of these symptoms clearly preceded the first episode of Sydenham's chorea ☐

No other psychiatric symptoms or diagnoses ☐

Section F: Neurological presentation

6.1 If this patient has been diagnosed with confirmed Sydenham's chorea please indicate:

1. How long since the first episode of Sydenham's chorea, if known? __weeks __months__years

	Yes	No	Don't know
2. Was the original diagnosis of Sydenham's chorea laboratory confirmed?	☐	☐	☐
3. Does the patient currently experience chorea?	☐	☐	☐
4. Has the patient had any relapses of chorea since the first episode of Sydenham's chorea? *If yes:*	☐	☐	☐

a) How many relapses have there been in total, if known? _____

b) How long has it been since the last relapse, if known? (leave blank if currently experiencing first relapse or no relapses) _____months_____years

Section G: Service involvement

7.1 Has the patient had contact with any of the following specialties/services since first episode of Sydenham's chorea?

	Current	Planned	Historical	Don't know
Clinical psychology or neuropsychology	☐	☐	☐	☐
General/community paediatrics	☐	☐	☐	☐
Paediatric neurology	☐	☐	☐	☐
Paediatric cardiology	☐	☐	☐	☐
Paediatric rheumatology	☐	☐	☐	☐
Physiotherapy and/or occupational therapy	☐	☐	☐	☐
Educational psychology	☐	☐	☐	☐

Other (please specify): _____

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Section H: Clinical and educational management

8.1 Has the management of the patient's neuro-psychiatric symptoms included any of the following?

	Current	Planned	Ever	Not known
Psychotropic medication (including psychotropic drugs for chorea e.g. haloperidol, valproate)	☐	☐	☐	☐
Please give details (e.g. drug, dose and duration), if known				
Course of antibiotics	☐	☐	☐	☐
Please give details (e.g. drug, dose and duration), if known				
Psychological therapy	☐	☐	☐	☐
Please give details (e.g. type of intervention, duration), if known				
Immunotherapy	☐	☐	☐	☐
Please give details e.g. drug, dose and duration, if known				
Physiotherapy and/or occupational therapy	☐	☐	☐	☐
Please give details (e.g. type of intervention, duration), if known				

Other interventions (please give details):

8.2 Educational support

	Yes	No	Not known
1. Is the patient currently in receipt of additional support at school or college? *If yes:*	☐	☐	☐
a) Is the additional support for needs arising from their Sydenham's chorea?	☐	☐	☐
b) Does the patient have a formal statement of special additional needs or education, health and care plan (if in England, Wales or NI), an additional support needs co-ordinated support plan (if in Scotland), or an education plan (in ROI)?	☐	☐	☐

Section I: Functional impact

9.1 At the time you first saw the patient within the current episode of care, how would you rate the impact of their symptoms on their ability to perform activities of daily living?

Overall functional impact	Psychiatric symptoms	Neurological symptoms
Normal (clinical feature absent)	☐	☐
Minimal impairment (clinical feature present but without functional impairment, no help needed to perform tasks)	☐	☐
Mild impairment (with functional impairment, but the patient can perform most of the activities of daily living, occasional help needed)	☐	☐
Moderate impairment (the patient cannot perform most of the activities of daily living, frequent help needed)	☐	☐
Severe impairment (activities of daily living are impossible, completely dependent)	☐	☐

Section J: Request to take part in a qualitative interview

10.1 Clinicians' general experience of case management

	Yes	No
We wish to interview a sample of clinicians about their **general experience** of managing cases of Sydenham's chorea, using a semi-structured telephone interview that will take approximately 30 minutes. We will NOT be discussing individual cases. Would you be willing to be contacted regarding taking part in such an interview? (This does not constitute any obligation to take part).	☐	☐

Thank you for taking the time to complete the Questionnaire

Please return this form using secure nhs.net mail to:

If you have any questions about the study please do not hesitate to contact the investigators by email or telephone:

Telephone: _____ Email: _____

Supplementary Table 1. List of other investigations requested by clinicians

Autoimmune investigations	• Anti-nuclear Antibody testing (ANA) • Anticardioliptin antibodies (IgG) • Anti DNAse • N-methyl-D-aspartate receptor (NMDAR) antibodies • Anti-mog • Anti-aquaporin antibodies
Infection screening	• Screening for Epstein Barr Virus (including assessing for acute and prior infection) • Lyme disease serology • *M. pneumoniae* serology • Lumbar puncture as part of screening for Encephalitis
Metabolic	• Copper and caeruloplasmin levels, for consideration of Wilson's disease as a differential diagnosis