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Tremor and Other Hyperkinetic Movements

Case Reports

Functional Impact of Sydenham's Chorea: A Case Report

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Abstract

Background: Sydenham's chorea (SC) is the most common type of acquired chorea in childhood. In some cases, symptoms (most commonly described in terms of neurological signs) last up to 2 years, and many cases relapse. This report describes the clinical course in terms of functional abilities following diagnosis of SC.

Case report: Standardized assessments across the domains of activity and participation were administered following diagnosis, prior to and following treatment with haloperidol to measure treatment response and identify occupational therapy intervention needs. SC was observed to significantly reduce the child's participation and independence in activities of daily living. In this case, the standardized assessments administered highlighted difficulties with both motor and process skills. At 1 week after commencing haloperidol, both motor and process skills had improved. Clinically significant changes in self-care and mobility were noted with less improvement with handwriting. At 9 weeks, most symptoms and functional difficulties had resolved.

Discussion: Given the process difficulties detected in this case, and the possibility of enduring symptoms, the use of functional assessments is advocated in the routine management of SC. These findings illustrate the potential for motor and non-motor sequelae in acute childhood movement disorders and related functional disabling consequences.

Keywords: Sydenham's chorea, occupational therapy, rehabilitation, handwriting, functional ability, haloperidol

Citation: Gimeno H, Barry S, Lin JP, et al. Functional impact of Sydenham's chorea: a case report. Tremor Other Hyperkinet Mov 2013;3: http://tremorjournal.org/article/view/128

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Editor: Elan D. Louis, Columbia University, United States of America

Received: September 22, 2012 **Accepted:** February 4, 2013 **Published:** April 19, 2013

Funding: None.

Conflict of interest: The authors report no conflicts of interest.

Financial disclosures: None.

Introduction

Sydenham's chorea (SC) is an autoimmune disease that may be triggered by infection with group A beta-hemolytic streptococci in children.[1] Classical presentation of SC includes rapid, involuntary movements that are irregular, non-stereotyped and can affect all limbs, face, and trunk.[2] Other symptoms can precede the onset of chorea, such as obsessive–compulsive signs, attention difficulties, and emotional lability.[3] While patients with SC have been described to make a full recovery, some studies show up to 50% of patients continue to display movement problems years after the initial onset,[1,2,4] and it is estimated that up to 50% of cases can relapse.[1,4]

Cardoso and colleagues,[1] in their seminar on choreas, highlighted that most studies report on the presence or absence of involuntary movements. The involuntary movements associated with SC respond to medication (sodium valporate or haloperidol) in the majority of cases.[5–8] However, the clinical course and response to medication is variable, and the nature, severity, and duration of impact on daily life are poorly understood. Some authors suggest that as spontaneous remission is expected after a few months, occupational therapy and physiotherapy are not indicated.[9] Functional long-term issues and safe hospital discharge, while florid movements are still present, have however not been described. Although the SC rating scale[10] provides a scale to assess functional activities of daily living (ADLs), this is a simple measure of ability and does not provide sufficient detail to quantify functional impairment or need for intervention.

Given that the symptoms of SC can last up to 2 years,[1,2,4] the early identification of disabling sequelae is important to ensure implementation of therapeutic interventions for home and school life. The aim of this case report is to describe the functional disabilities arising from SC that may impact safe discharge and illustrate the importance of multidisciplinary involvement in the management of children with SC.

Case report

An 8-year-old girl, who was right handed and had previously met developmental milestones, presented at her local hospital with a sudden onset of involuntary movements 2 months prior to admission. Her speech, manual function, mobility, and most ADLs were affected. One month after being admitted to her local hospital, she was transferred to our tertiary hospital in central London, where this study took place. Her symptoms were similar to those described in other case reports.[8] Neurological examination at presentation revealed the features of a severe total body chorea including impersistence of the handgrip bilaterally known as "milk maid's sign." Truncal posture was only stable in the supine position. There was marked dysarthria with bilateral facial chorea; however, eye movements, deep tendon reflexes, Babinski plantar responses, and sensation were normal throughout the course of the illness. There was no alteration of consciousness at any time.

Functional concerns on admission, as described by the child's parents, related to requiring assistance with dressing and grooming activities. She required constant supervision when walking and going up and down stairs, as she bumped into objects and was at risk of falling. Concerns were also raised about her return to mainstream school because of difficulties with mobility, manual function and speech. It was observed that she did not use her left hand spontaneously as a supportive hand when completing activities (i.e., to support the paper with her non-dominant hand to avoid the paper moving when writing). Overall, the involuntary movements affected all aspects of daily living and the child's level of independence and participation.

Neurological examination was performed by a movement disorders specialist pediatric neurologist (J.P.L.). Magnetic resonance imaging (MRI) findings showed no brain abnormality. No medications were prescribed prior to admission to the tertiary hospital. Sodium valporate was introduced for 7 days following hospital transfer at an initial dose of 5 mg/kg/day and increased daily to 270 mg twice daily (20 mg/kg) with no effect and discontinued before baseline was obtained.

Functional data were obtained at three time points by occupational therapists (HG, SB, AG) to inform clinical management. Time point 1 (baseline) was obtained at 10 days after admission, prior to MRI for which the patient had been fasted (time point 1=no medication). Time point 2 took place 1 week following introduction of haloperidol (500 μg twice daily), and time point 3 was taken 9 weeks after haloperidol was started) (250 μg twice daily).

Standardized assessments across the domains of activity and participation were selected to enable monitoring of functional ability over time, to obtain a formal baseline for comparison following medical intervention, and to highlight areas of unmet need amenable to occupational therapy treatment. The information obtained also helped to inform intervention needs teaching strategies to overcome some of the difficulties, i.e., thinking about where the child was positioned in relation to the task items for skill development using cognitive strategies, which is described as a restorative approach to intervention. This was combined with an adaptive approach using simple aids such as non-slip mats (dycem®), adapting the environment (i.e., height of chair and table), and changing the task (i.e., sitting down to butter toast). These strategies were used in occupational therapy sessions and generally incorporated into the child's daily routine.

The Pediatric Evaluation of Disability Inventory (PEDI)[11] was administered. This parental questionnaire measures functional ability and caregiver assistance in the areas of self-care, mobility, and social function. Scaled scores were obtained in the areas of self-care and mobility across all three time points, but the social function domain was only completed at baseline as the patient showed no difficulties in this area. Activities assessed in the self-care areas in this domain include washing, dressing, eating with utensils, and managing toileting activities. The areas in the mobility section of this parental questionnaire include walking indoors and outdoors, going up and down stairs, and transfers. A difference of 11 points in scale scores indicates a clinically significant change.[12]

Handwriting legibility, speed, and self-generation were measured using the Detailed Assessment of Speed of Handwriting (DASH).[13] The DASH consists of four timed pencil–paper sub-tests. Standard scores have been validated for children aged 9 years and above, and as our patient was soon to be of that age the standard scores of the patient compared with a 9 year old at all three time points are presented. Standard scores are calculated based on population norms (mean=10, standard deviation [SD]=3).

Finally, the Assessment of Motor and Process Skills (AMPS)[14] was completed at all time points. The AMPS is a standardized measure of motor and process components of functional daily tasks. For this assessment, the child chooses two ADLs including personal and domestic activities (i.e., dressing or making a sandwich). The AMPS uses Rasch methodology with raw scored converted into logits. A change of 0.50 in logits is needed to be statistically significant. Motor and processing skills standard scores were also calculated (general population mean=100, SD=15).

The data obtained from the standardized assessments were analyzed between each time point to describe the nature and severity of disability and the changes over time.

On admission, the severity of her difficulties is reflected in baseline scores. Scores in the PEDI showed the self-care functional domain to be 65/100 and the caregiver domain to be 59/100, with the mobility domains scoring 68.7/100 and 70.5/100 for the self-care functional and caregiver domains, respectively. DASH scores were 2 SD below the mean for copying and 1 SD for alphabet writing and free handwriting. The AMPS demonstrated motor skills 4 SD below the mean and 1 SD below the mean for process skills. A marked improvement in functional ability including self-care, mobility and handwriting skills was evident 1 week after commencing haloperidol (500 μg twice a day) (see Table 1).

A clinically significant change on PEDI (>11 points)[12] was noted in the areas of self-care (functional and caregiver assistance) and in the mobility section (functional domain only) after 1 week on haloperidol and had full scores 9 weeks after starting medication. Despite a clear improvement in self-care and other ADLs, the DASH shows only a

Table 1. Functional Assessment Results

	PEDI				DASH				AMPS			
	Scaled Scores				Standard Scores (mean=10, SD=3)				Standard Scores (mean=100, SD=15)		Logit Scores	
	Functional Domain		Caregiver Domain		Copy Best	Alphabet Writing	Fast Copy	Free Writing	Motor Skills	Process Skills	Motor Skills	Process Skills
	SC	M	SC	M								
Baseline	65	68.7	59	70.5	4[1]	6[1]	3[1]	5[1]	<45[1]	81[1]	–0.64	0.25
1 week after medication	85.1	89.2	89.7	78.3	7	7	NT	NT	93	114	1.44[2]	1.15[2]
9 weeks after medication	100	100	100	100	11	9	8	6[1]	120	114	2.39[2]	1.15[2]

AMPS, Assessment of Motor and Process Skills; DASH, Detailed Assessment of Speed of Handwriting; M, Mobility; NT, Not Tested; PEDI, Pediatric Evaluation of Disability Inventory; SC, Self-Care; SD, Standard Deviation.
[1]>1 SD below population mean.
[2]Clinically significant change in logit score.

slight improvement in handwriting, suggesting ongoing impairments in the child's ability to record written work. At 9 weeks, handwriting returned to average scores (Figure 1) with the exception of the free handwriting sub-test. This requires the child to self-generate and organize thoughts and ideas and record them, rather than just copying a given text. A statistically significant improvement (>0.50 change in logits) was obtained in both motor and process skills at time points 2 and 3 using the AMPS. Discussion

This case study found that SC affected the child's functional abilities and level of dependence on parental assistance in daily tasks. Standardized assessments of functional abilities prior to the introduction of haloperidol indicated that motor and process component skills were impaired, and this was reflected in functional areas including mobility, hand use, self-care, and school-related tasks.

Impairment in process skills may include difficulties in problem solving, initiation of tasks, and planning, organizing, and adapting performance. A possible explanation for the functional disabilities measured in this case was the combination of both motor and process impairments (as measured by the AMPS). This adversely affected the child's ability to develop new problem-solving strategies to cope with her new movement difficulties. Once the involuntary movements were better controlled at time point 2 of assessment, there was a concomitant improvement in process skills (to within normal ranges). It is possible that the child was better able to attend to her environment and the cognitive demands of tasks when able to rely on improved postural control and voluntary movements. Preliminary functional data in childhood dystonia (occupational therapy complex motor disorders service data – unpublished results) shows involvement of both motor and process skills during functional activities in children with movement disorders.

Given that functional recovery is not well understood in SC and the potential risk for relapse in up to 50% of cases, an early functional assessment could help clinicians identify medium- to long-term health needs. The early identification of functional difficulties can help improve safe discharge, early return to child-appropriate activities and environments such as school and promote speed of recovery beyond impairment.

In this single case study, the effects of haloperidol in combination with intervention from occupational therapy taking both a restorative and adaptive approach to facilitate safe discharge showed a marked alleviation in carer burden and functional independence over time.

In children affected with SC, recovery times vary from a few months to 2 years. In the case described, the child presented with symptoms lasting several months, resulting in substantial carer burden in functional daily tasks with both motor and process impairments. Following medical intervention and ongoing occupational therapy input, improvements were rapidly evident in home and school independence, and the child was successfully discharged to home and school life.

Given that SC is a condition that affects the developing brain, deterioration in performance lasting months or years could have a detrimental effect on the child's ability to learn new tasks. A functional baseline evaluation using standardized assessment of motor and

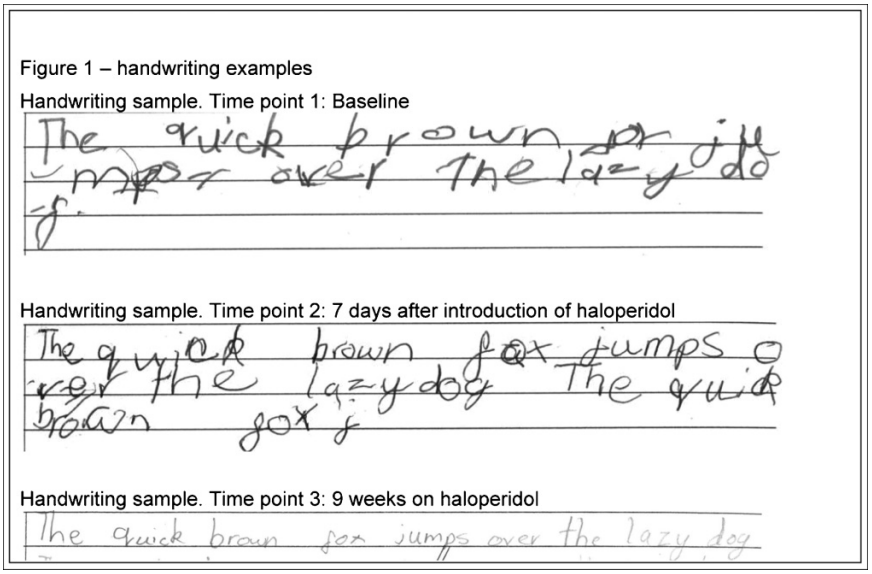

Figure 1. Handwriting Examples. Time point 1, baseline; Time point 2, 7 days after introduction of haloperidol; Time point 3, 9 weeks on haloperidol.

non-motor skills before pharmacology is initiated could provide important objective information. Greater understanding of the impact of SC on independence skills, safety, and carer burden and response to medical and rehabilitative interventions across these domains can support multidisciplinary interventions and targeting of resources to maximize functional gains.

Acknowledgements

We would like to thank the child and family that participated in this study.

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