

UFMG Sydenham's Chorea Rating Scale (USCRS): Reliability and Consistency

Antônio Lúcio Teixeira, Jr., MD, PhD, Débora P. Maia, MD, and Francisco Cardoso, MD, PhD*

*Movement Disorders Clinic, Faculty of Medicine and Hospital das Clínicas, Federal University of Minas Gerais (UFMG),
Belo Horizonte, Brazil*

Abstract: Despite the renewed interest in Sydenham's chorea (SC) in recent years, there were no valid and reliable scales to rate the several signs and symptoms of patients with SC and related disorders. The Universidade Federal de Minas Gerais (UFMG) Sydenham's Chorea Rating Scale (USCRS) was designed to provide a detailed quantitative description of the performance of activities of daily living, behavioral abnormalities, and motor function of subjects with SC. The scale com-

prises 27 items and each one is scored from 0 (no symptom or sign) to 4 (severe disability or finding). Data from 84 subjects, aged 4.9 to 33.6 years, support the interrater reliability and internal consistency of the scale. The USCRS is a promising instrument for rating the clinical features of SC as well as their functional impact in children and adults. © 2005 Movement Disorder Society

Key words: rating scale; chorea; Sydenham's chorea

Sydenham's chorea (SC) is a delayed complication of group A β -hemolytic streptococcal pharyngitis and constitutes one of the major criteria of acute rheumatic fever.¹ The typical age at onset of SC is between 8 and 9 years old and there is a female preponderance.^{2,3} The presence of random and purposeless involuntary movements called chorea is the main feature of SC. Choreic movements in SC are generalized in most patients, but they determine variable degree of motor disability. Other motor symptoms, such as motor imperistence, hypometric saccades, dysarthria, and decreased muscle tone can be present.² Although the distinction between chorea and tics may be troublesome, the latter are reported frequently in SC.⁴ Neuropsychiatric symptoms including hyperactivity and obsessive-compulsive behavior are also noticeable in SC patients.^{5–7}

In 1998, Swedo and colleagues⁸ proposed that a subset of childhood tic disorders and behavioral abnormalities,

principally obsessive-compulsive disorder, could be causally related to streptococcal infection. The acronym PANDAS (pediatric autoimmune neuropsychiatric disorders after streptococcal infection) was then employed to name this condition.⁸ Considerable controversies followed the PANDAS hypothesis bringing renewed interest in SC, a disease that was restricted to underdeveloped countries.³ Despite advances in the understanding of SC, many questions remain unsolved, including the validity of the PANDAS construct.⁹

One limitation of the study of SC and related disorders is the lack of a validated rating scale. This study presents the development and initial testing of the first published scale of SC called the Universidade Federal de Minas Gerais (UFMG) Sydenham's Chorea Rating Scale (USCRS).

SUBJECTS AND METHODS

Subjects

The 84 subjects comprised children (64) and adults (20) (mean age \pm SD, 15.7 \pm 6.2 years; age range, 4.9–33.6 years; 28 males, 56 females) were enrolled in this study carried out from January 2003 to December 2003. All subjects had been diagnosed with SC and are currently followed at the Movement Disorders Clinic at UFMG, Belo Horizonte, Brazil. SC was diagnosed if the

*Correspondence to: Dr. Francisco Cardoso, Av. Pasteur 89/1107, 30150-290 Belo Horizonte, MG, Brazil.
E-mail: cardosofe@terra.com.br

Received 29 June 2004; Revised 24 August 2004; Accepted 22 September 2004

Published online 12 January 2005 in Wiley InterScience (www.interscience.wiley.com). DOI: 10.1002/mds.20377

patient had acute onset of chorea, met the modified Jones criteria for acute rheumatic fever,¹ and if other causes of chorea were excluded carefully.²

These patients were interviewed and examined by clinicians with extensive clinical experience with SC and trained in the use of the USCRS. For the interrater reliability assessment, 20 patients were each rated by three clinicians (A.T., D.M., and F.C.), with the clinicians independently evaluating the patients.

USCRS

After several months of pilot experience, the investigators formulated the USCRS that was designed to provide a detailed quantitative description of the performance of activities of daily living, behavioral abnormalities, and motor function of subjects with SC.

Emphasis was placed on clinical features that were likely to be relevant in SC and easily assessed by a brief physical examination. The scale comprises 27 items and each one is scored from 0 (no symptom or sign) to 4 (severe disability or finding). The behavioral assessment measures irritability, attention deficit, hyperactivity, obsessions, compulsions, and reduced verbal fluency. The functional assessment evaluates impairment and the level of independence of the patient to perform activities of daily living, such as handwriting, cutting food, dressing, walking, and hygiene. The motor function section of the USCRS assesses oculomotor function, motor persistence, dysarthria, chorea in the face and all limbs, bradykinesia, muscle tone, and gait. Tics were not assessed, as it is virtually impossible to distinguish them from chorea.

Some items of the USCRS such as ocular pursuit and leg agility were based on previously published scales, the Unified Huntington's Disease Rating Scale (UHDRS)¹⁰ and the Unified Parkinson's Disease Rating Scale (UPDRS),¹¹ respectively. Other items, such as muscle tone, were formulated by the authors based on their clinical experience. The USCRS is reprinted in the Appendix.

Statistical Analysis

The variables were processed in a database and statistical analysis carried out using the SAS software (SAS, Cary, IN). The interrater reliability was assessed using an intraclass correlation coefficient (ICC). The internal consistency of the USCRS was assessed by Cronbach's α and the Spearman correlation coefficient calculation. Factor structure was also analyzed using a principal components factor analysis.

TABLE 1. *Intraclass correlations for 3 raters and 20 subjects*

USCRS item	Intraclass correlation
Irritable behavior	0.99
Attention deficit	0.98
Hyperactivity	0.97
Obsessions	0.98
Compulsions	0.96
Verbal fluency	0.99
Dysarthria-s	0.99
Chorea-s	1.00
Handwriting	0.99
Cutting food	0.99
Hygiene	0.99
Dressing	1.00
Walking	0.99
Ocular pursuit	0.86
Dysarthria	1.00
Face chorea	0.98
Right arm chorea	0.98
Right leg chorea	0.98
Left arm chorea	0.97
Left leg chorea	0.99
Tongue protrusion	0.99
Right side finger tap	0.99
Left side finger tap	0.96
Right side leg agility	0.97
Left side leg agility	0.98
Muscle tone	0.96
Gait	0.96

USCRS, UFMG Sydenham's chorea rating scale.

RESULTS

The ICC values for individual items of the USCRS ranged from 0.86 to 1.0 (median 0.98) (Table 1). Cronbach's α value was 0.97. Table 2 presents the Spearman correlation coefficients between each item of the USCRS. There was a high degree of internal consistency in each component of the USCRS, except for some items of behavioral domain (obsessions, compulsions, attention deficit, and hyperactivity). Interestingly, when we carried out principal components analyses, two factors emerged (Table 3). The first factor comprised items of the motor function and performance of activities of daily living, whereas the second factor corresponded to behavioral items.

DISCUSSION

The basal ganglia are composed of several nuclei that are linked to one another and to the brain cortex by parallel segregated circuits, each subserving different functions. These corticostriatal pathways include a dorsolateral prefrontal circuit that mediates executive functions, an orbitofrontal circuit responsible for behavioral monitoring, an anterior cingulate circuit that mediates motivated behavior, and motor and oculomotor circuits.¹² The motor circuit facilitates voluntary move-

TABLE 2. Spearman's correlation coefficients between individual items of the USCRS

	Irritability	Attention deficit	Hyperactivity	Obsessions	Compulsions	Fluency	Dysarthria-s	Chorea-s	Writing	Cutting	Hygiene	Dressing	Walking
Attention deficit	0.501												
Hyperactivity	0.000	0.392											
Obsession	0.299	0.000	0.220										
Compulsion	0.006	0.258	0.044	0.373									
Fluency	0.300	0.175	0.251	0.000	0.069								
Dysarthria-s	0.006	0.111	0.021	0.003	0.531	0.690							
Chorea-s	0.009	0.663	0.713	0.976	-0.010	0.000							
Writing	0.574	0.310	0.241	0.083	0.928	0.581							
Cutting	0.000	0.004	0.027	0.452	0.285	0.000							
Hygiene	0.660	0.314	0.475	0.317	0.009	0.000	0.685						
Dressing	0.000	0.004	0.000	0.003	0.409	0.000	0.000						
Walking	0.537	0.307	0.436	0.174	0.525	0.485	0.745						
Pursuit	0.000	0.005	0.000	0.004	0.113	0.000	0.000						
Dysarthria	0.533	0.262	0.420	0.235	0.178	0.632	0.751	0.833	0.628				
Face chorea	0.000	0.016	0.000	0.031	0.104	0.000	0.000	0.824	0.000				
R arm chorea	0.475	0.229	0.295	0.307	0.165	0.599	0.620	0.726	0.749				
P	0.000	0.036	0.006	0.004	0.134	0.000	0.000	0.000	0.000				
R leg chorea	0.508	0.283	0.393	0.223	0.146	0.647	0.678	0.757	0.737				
P	0.000	0.009	0.000	0.041	0.186	0.000	0.000	0.000	0.000				
L arm chorea	0.532	0.263	0.424	0.206	0.174	0.638	0.770	0.794	0.623				
P	0.000	0.016	0.000	0.061	0.150	0.000	0.000	0.000	0.000				
L leg chorea	0.415	0.225	0.450	0.142	0.051	0.637	0.543	0.622	0.620				
P	0.000	0.040	0.030	0.196	0.642	0.000	0.000	0.000	0.000				
Protrusion	0.459	0.193	0.449	0.053	0.044	0.813	0.813	0.646	0.530				
R finger tap	0.000	0.079	0.000	0.635	0.694	0.000	0.000	0.000	0.000				
R finger tap	0.425	0.301	0.426	0.112	0.059	0.708	0.641	0.703	0.703				
L finger tap	0.000	0.005	0.000	0.311	0.596	0.000	0.000	0.000	0.000				
P	0.539	0.297	0.450	0.237	0.159	0.518	0.507	0.837	0.813				
P	0.000	0.006	0.000	0.078	0.148	0.000	0.000	0.000	0.000				
L arm chorea	0.483	0.260	0.449	0.193	0.177	0.577	0.558	0.816	0.724				
P	0.000	0.017	0.000	0.078	0.108	0.000	0.000	0.000	0.000				
L arm chorea	0.602	0.312	0.464	0.090	0.266	0.557	0.615	0.780	0.585				
P	0.000	0.004	0.000	0.413	0.015	0.000	0.000	0.000	0.000				
L leg chorea	0.563	0.274	0.427	0.066	0.245	0.588	0.659	0.742	0.510				
P	0.000	0.012	0.000	0.551	0.025	0.000	0.000	0.000	0.000				
Protrusion	0.470	0.344	0.449	0.171	0.107	0.464	0.543	0.612	0.549				
R finger tap	0.000	0.001	0.000	0.120	0.331	0.000	0.000	0.000	0.000				
L finger tap	0.512	0.316	0.456	0.184	0.175	0.593	0.535	0.780	0.677				
P	0.000	0.003	0.000	0.094	0.112	0.000	0.000	0.000	0.000				
L leg chorea	0.411	0.189	0.394	0.064	0.197	0.586	0.548	0.691	0.576				
P	0.564	0.085	0.444	0.561	0.072	0.000	0.000	0.000	0.000				
R leg agility	0.000	0.295	0.444	0.222	0.178	0.617	0.636	0.768	0.706				
P	0.000	0.006	0.000	0.042	0.105	0.000	0.000	0.000	0.000				
L leg agility	0.449	0.217	0.368	0.091	0.237	0.686	0.664	0.713	0.586				
P	0.000	0.048	0.001	0.411	0.030	0.000	0.000	0.000	0.000				
Muscle tone	0.506	0.243	0.357	-0.065	0.139	0.629	0.585	0.677	0.555				
P	0.000	0.026	0.001	0.558	0.206	0.000	0.000	0.000	0.000				
Gait	0.460	0.237	0.366	0.086	0.162	0.734	0.732	0.783	0.657				
P	0.000	0.030	0.001	0.435	0.141	0.000	0.000	0.000	0.000				

(continued)

TABLE 2. (Continued)

	Pursuit	Dysarthria	Face- chorea	R arm chorea	R leg chorea	L arm chorea	L leg chorea	Protrusion	R finger tap	L finger tap	R leg agility	L leg agility	Muscle tone
Attention deficit <i>P</i>													
Hyperactivity <i>P</i>													
Obsession <i>P</i>													
Compulsion <i>P</i>													
Fluency <i>P</i>													
Dysarthria-s <i>P</i>													
Chorea-s <i>P</i>													
Writing <i>P</i>													
Cutting <i>P</i>													
Hygiene <i>P</i>													
Dressing <i>P</i>													
Walking <i>P</i>													
Pursuit <i>P</i>													
Dysarthria <i>P</i>	0.663												
Face chorea <i>P</i>	0.000	0.625											
R arm chorea <i>P</i>	0.631	0.536	0.750										
R leg chorea <i>P</i>	0.560	0.569	0.796										
L arm chorea <i>P</i>	0.577	0.620	0.687		0.630								
L leg chorea <i>P</i>	0.457	0.558	0.745	0.904	0.679	0.000							
Protrusion <i>P</i>	0.527	0.605	0.643	0.632	0.599	0.630	0.520						
R finger tap <i>P</i>	0.559	0.562	0.681	0.827	0.806	0.659	0.642	0.606					
L finger tap <i>P</i>	0.538	0.518	0.679	0.567	0.618	0.768	0.762	0.000	0.688				
R leg agility <i>P</i>	0.511	0.564	0.717	0.739	0.814	0.599	0.701	0.529	0.779	0.626			
L leg agility <i>P</i>	0.548	0.618	0.707	0.568	0.630	0.744	0.801	0.545	0.000	0.808	0.807		
Muscle tone <i>P</i>	0.561	0.617	0.685	0.000	0.000	0.000	0.000	0.000	0.000	0.000	0.000	0.690	
Gait <i>P</i>	0.635	0.702	0.752	0.677	0.729	0.687	0.708	0.588	0.733	0.744	0.840	0.923	0.716
	0.000	0.000	0.000	0.000	0.000	0.000	0.000	0.000	0.000	0.000	0.000	0.000	0.000

The level of significance was set at $P < 0.05$. R, right; L, left.

ments and inhibit competing movements that might interfere with the desired movement.¹³

The basal ganglia have been implicated as a target of poststreptococcal autoimmune mechanism in SC.³ Indeed, structural or functional abnormalities within basal ganglia circuits may explain the clinical picture exhibited by SC patients. For instance, chorea may be explained by the motor circuit impairment whereas hypometric saccades may be explained by oculomotor circuit dysfunction. Some behavioral abnormalities that follow SC can be related to the orbitofrontal circuit syndrome¹² characterized by disinhibition and emotional lability. It can be hypothesized that different SC clinical phenotypes might reflect the topography of the immune-based dysfunction within corticostriatal circuits.¹⁴ Bearing this hypothesis in mind, we constructed an instrument called the USCRS to assess the several symptomatic dimensions of SC.

The high ICCs for all USCRS items indicate that these measures can be made with a high degree of accuracy by experienced clinicians. We also found a high degree of internal consistency of the USCRS expressed by the Cronbach's α value. However, on the Spearman correlation analysis, some behavioral items correlated poorly with other variables. As internal consistency represents the degree to which individual items are consistent with each other and, thus, are tapping the same construct, items with poor internal consistency can be dropped from the final scale form.¹⁵ We preferred to retain such items, because of their clinical relevance as proposed by several authors.⁵⁻⁷ The factor structure analysis of the USCRS also segregated all behavioral items from the motor/functional components. As SC is proposed to be an immune-mediated basal ganglia dysfunction, this may suggest that distinct basal ganglia circuits are affected differently in SC. Interestingly, analysis of the UHDRS also demonstrated that the behavioral component did not correlate well with the motor, cognitive, and functional components.¹⁰

Although the USCRS is not a diagnostic instrument, it would be interesting to evaluate patients considered to have PANDAS with it. As the fifth criterion for PANDAS refers to the presence of abnormal neurological findings described as choreiform movements,⁸ it would be of paramount importance to verify if some PANDAS patients might have in fact mild form of SC.⁹

In conclusion, the USCRS is a promising instrument for rating the clinical features of SC and their functional impact in children and adults. We believe that this scale might be a useful instrument to assess clinical changes in SC associated either with natural history or therapeutic intervention.

APPENDIX

THE UFMG SYDENHAM'S CHOREA RATING SCALE (USCRS)

A. Instructions

USCRS was designed to rate the overall severity of Sydenham's chorea across different symptomatic dimensions. It is important to reassure that this is not a diagnostic scale.

The assessor should first complete the sections I and II (items 1-13) before performing the motor assessment (section III, items 12-21). The first two sections are completed based on all available information obtained from the patient and/or relatives. The style of the interview is semistructured, using the description of the items and the content of the anchor points as a guide. The third section is completed during the physical examination.

B. Inventory

I-Behavior (Information obtained from the patient and/or relatives)

1. Irritable behavior (impatient, impulsive, inflexible, uncooperative)
 - 0 = absent
 - 1 = minimal (less than 25% of the time)
 - 2 = mild (between 25 and 50% of the time)
 - 3 = moderate (more than 50% of the time, but less than 75%)
 - 4 = severe (more than 75% of the time)
2. Attention deficit (difficulty sustaining attention in tasks or play activities)
 - 0 = absent
 - 1 = minimal (present, but without functional impairment)
 - 2 = mild (with functional impairment, but the patient can perform activities demanding attention without help)
 - 3 = moderate (the patient can perform activities with help)
 - 4 = severe (the patient is unable to perform any activity demanding attention even with help)
3. Hyperactivity (distinct from chorea)
 - 0 = absent
 - 1 = minimal (present, but without functional impairment)
 - 2 = mild (with functional impairment, but the patient can control him/herself without external warning)
 - 3 = moderate (the patient can control him/herself only with external warning)
 - 4 = severe (the patient can not be controlled even with external warning)
4. Obsessions (recurrent and/or persistent ideas, thoughts, or images)
 - 0 = absent
 - 1 = minimal (present, but without functional impairment)
 - 2 = mild (with functional impairment, but the patient can perform most of the activities of daily living)
 - 3 = moderate (the patient can not perform most of the activities of daily living)
 - 4 = severe (activities of daily living are impossible)
5. Compulsions (uncontrolled repetitive behaviors)
 - 0 = absent
 - 1 = minimal (present, but without functional impairment)
 - 2 = mild (with functional impairment, but the patient can perform most of the activities of daily living)
 - 3 = moderate (the patient can not perform most of the activities of daily living)
 - 4 = severe (activities of daily living are impossible)

TABLE 3. USCRS item loadings on two factors

USCRS items	Factor 1	Factor 2
Irritable behavior	0.49	0.70
Attention deficit	0.14	0.63
Hyperactivity	0.24	0.75
Obsessions	-0.02	0.61
Compulsions	-0.03	0.65
Verbal fluency	0.89	-0.14
Dysarthria-s	0.83	0.10
Chorea-s	0.81	0.46
Handwriting	0.81	0.23
Cutting food	0.91	0.13
Hygiene	0.91	0.04
Dressing	0.92	0.07
Walking	0.84	0.25
Ocular pursuit	0.76	0.07
Dysarthria	0.90	-0.07
Face chorea	0.86	0.20
Right arm chorea	0.77	0.32
Right leg chorea	0.84	0.22
Left arm chorea	0.75	0.39
Left leg chorea	0.79	0.32
Tongue protrusion	0.68	0.29
Right side finger tap	0.85	0.20
Left side finger tap	0.79	0.26
Right side leg agility	0.89	0.19
Left side leg agility	0.84	0.26
Muscle tone	0.86	0.14
Gait	0.94	0.10

USCRS, UFMG Sydenham's chorea rating scale; n = 84.

6. Verbal fluency

- 0 = normal
- 1 = minimally impaired (reduced spontaneous speech but without impairment of the communication)
- 2 = mildly impaired (reduced spontaneous speech with impairment of the communication)
- 3 = moderately impaired (spontaneous speech abolished, but able to speak when requested)
- 4 = severely impaired (no speech even when requested)

II-Activities of Daily Living (Information obtained from the patient or relatives)

7. Dysarthria

- 0 = absent
- 1 = minimal (present, but completely comprehensible, or speech easily understood)
- 2 = mild (less than 25% of the speech is incomprehensible, or some difficulty in understanding speech)
- 3 = moderate (25-50% of the speech is incomprehensible, or marked difficulty in understanding speech)
- 4 = severe (incomprehensible speech)

8. Chorea

- 0 = absent
- 1 = minimal (present, but without functional impairment)
- 2 = mild (with impairment of some motor or functional activities)
- 3 = moderate (with impairment of many motor or functional activities)
- 4 = severe (completely disabled)

9. Handwriting

- 0 = normal
- 1 = minimally impaired (compromised handwriting, but all words are legible)

- 2 = mildly impaired (not all words are legible)
- 3 = moderately impaired (the majority of words are not legible)
- 4 = severely impaired (impossible handwriting)

10. Cutting food and handling utensils

- 0 = normal
- 1 = minimally impaired (difficulty with these tasks, but no help needed)
- 2 = mildly impaired (occasional help needed, e.g., cutting meat)
- 3 = moderately impaired (frequent help needed)
- 4 = severely impaired (needs to be fed)

11. Hygiene

- 0 = normal
- 1 = minimally impaired (difficulty with hygiene tasks, but no help needed)
- 2 = mildly impaired (occasional help needed)
- 3 = moderately impaired (frequent help needed)
- 4 = severely impaired (completely dependent)

12. Dressing

- 0 = normal
- 1 = minimally impaired (difficulty with dressing, but no help needed)
- 2 = mildly impaired (occasional help needed)
- 3 = moderately impaired (frequent help needed)
- 4 = severely impaired (completely dependent)

13. Walking

- 0 = normal
- 1 = minimally impaired (walks with difficulty, but does not run into objects)
- 2 = mildly impaired (walks with difficulty, running into objects)
- 3 = moderately impaired (walks only with assistance)
- 4 = severely impaired (chorea paralytica; cannot walk at all, even with assistance)

III-Motor Assessment

14. Ocular pursuit (horizontal and vertical)

- 0 = normal or complete
- 1 = minimally impaired (jerky movement)
- 2 = mildly impaired (interrupted pursuits, but preserved full range)
- 3 = moderately impaired (incomplete range)
- 4 = severely impaired (cannot pursue)

15. Dysarthria

- 0 = absent
- 1 = minimal (present but completely comprehensible, or speech easily understood)
- 2 = mild (less than 25% of the speech is incomprehensible, or some difficulty in understanding speech)
- 3 = moderate (25-50% of the speech is incomprehensible, or marked difficulty in understanding speech)
- 4 = severe (more than 50% of speech is incomprehensible)

16. Chorea (Test face and each limb)

- 0 = absent
- 1 = minimal (action chorea, or intermittent rest chorea)
- 2 = mild (continuous rest chorea, but without functional impairment)
- 3 = moderate (continuous rest chorea with partial functional impairment)
- 4 = severe (continuous rest chorea with complete functional impairment)

17. Tongue protrusion

- 0 = can hold tongue protruded for more than 1 minute
- 1 = can hold tongue protruded for more than 30 seconds
- 2 = can hold tongue protruded for more than 10 seconds
- 3 = can hold tongue protruded for less than 10 seconds
- 4 = cannot protrude tongue

18. Finger taps (Patient taps thumb with index finger in rapid succession with widest amplitude possible. Test right and left)

- 0 = normal (>15/5 sec)
- 1 = minimally impaired (mild slowing and/or reduction in amplitude, 11–14/5 sec)
- 2 = mildly impaired (occasional arrests in movement, 5–10/5 sec)
- 3 = moderately impaired (frequent hesitation in initiating movements or arrests in ongoing movements, <5/5 sec)
- 4 = severely impaired (cannot perform the task)

19. Leg agility (Patients taps heel on ground in rapid succession with widest amplitude, picking up entire leg)

- 0 = normal
- 1 = minimally impaired (mild slowing and/or reduction in amplitude)
- 2 = mildly impaired (occasional arrests in movement)
- 3 = moderately impaired (frequent hesitation in initiating movements or arrests in ongoing movements)
- 4 = severely impaired (cannot perform the task)

20. Muscle tone (Test each limb. The value is the sum of the muscle tone for each limb divided by 4)

- 0 = normal
- 1 = minimal decrease (not apparent when the contralateral limb is simultaneously moved)
- 2 = mild decrease (apparent even when the contralateral limb is simultaneously moved, but without functional impairment)
- 3 = moderate decrease (apparent even when the contralateral limb is simultaneously moved and with functional impairment)
- 4 = severe decrease (loss of postural tone)

21. Gait (Evaluate the gait of the patient while walking at least 10 m over a line distant 50 cm from the wall)

- 0 = normal
- 1 = minimally impaired (walks with difficulty, but does not run into objects)
- 2 = mildly impaired (walks with difficulty, running into with objects)
- 3 = moderately impaired (walks only with assistance)
- 4 = severely impaired (chorea paralytica; cannot walk at all, even with assistance)

Acknowledgments: This work was supported by the Conselho Nacional de Desenvolvimento Científico e Tecnológico (CNPq), Brazil.

REFERENCES

1. Special Writing Group of the Committee on Rheumatic Fever, Endocarditis, and Kawasaki Disease of the Council on Cardiovascular Disease in the Young of the American Heart Association. Guidelines for the diagnosis of rheumatic fever: Jones criteria, 1992 update. *JAMA* 1992;268:2069–2073.
2. Cardoso F, Silva CE, Mota CC. Sydenham's chorea in fifty consecutive patients with rheumatic fever. *Mov Disord* 1997;12:701–703.
3. Cardoso F. Infectious and transmissible movement disorders. In: Jankovic J, Tolosa E, editors. *Parkinson's disease and movement disorders*. Philadelphia: Lippincott-Williams & Wilkins; 2002. p 584–595.
4. Mercadante MT, Campos MC, Marques-Dias MJ, Miguel EC, Leckman J. Vocal tics in Sydenham's chorea. *J Am Acad Child Adolesc Psychiatry* 1997;36:305–306.
5. Swedo SE, Leonard HL, Schapiro MB, Casey BJ, Mannheim GB, Lenane MC, Rettew DC. Sydenham's chorea: physical and psychological symptoms of St. Vitus dance. *Pediatrics* 1993;91:706–713.
6. Asbahr FR, Negrão AB, Gentil V, Zanetta DM, Paz JA, Marques-Dias MJ, Kiss MH. Obsessive-compulsive and related symptoms in children and adolescents with rheumatic fever with and without chorea: a prospective 6-month study. *Am J Psychiatry* 1998;155:1122–1124.
7. Mercadante MT, Busatto GF, Lombroso PJ, Prado L, Rosário-Campos MC, Valle R, Marques-Dias MJ, Kiss MH, Leckman JF, Miguel EC. The psychiatric symptoms of rheumatic fever. *Am J Psychiatry* 2000;157:2036–2038.
8. Swedo SE, Leonard HL, Garvey MA, Mittleman BB, Allen AJ, Perlmutter S, Lougee L, Drow S, Zamkoff J, Dubbert BK. Pediatric autoimmune neuropsychiatric disorders associated with streptococcal infections: clinical description of the first 50 cases. *Am J Psychiatry* 1998;154:110–112.
9. Kurlan R. The PANDAS hypothesis: losing its bite? *Mov Disord* 2004;19:371–374.
10. Huntington Study Group. Unified Huntington's Disease Rating Scale: reliability and consistency. *Mov Disord* 1996;11:136–142.
11. Fahn S, Elton RL, and members of the UPDRS Development Committee. Unified Parkinson's Disease Rating Scale. In: Fahn S, Marsden CD, Goldstein M, editors. *Recent developments in Parkinson's disease, II*. Florham Park: MacMillan Healthcare Information; 1987. p 293–304.
12. Tekin S, Cummings JL. Frontal-subcortical neuronal circuits and clinical neuropsychiatry: an update. *J Psychosom Res* 2002;53:647–654.
13. Mink JW. The basal ganglia and involuntary movements: impaired inhibition of competing motor patterns. *Arch Neurol* 2003;60:1365–1368.
14. Mercadante MT, Hounie AG, Diniz JB, Miguel EC, Lombroso PJ. The basal ganglia and immune-based neuropsychiatric disorders. *Psychiatr Ann* 2001;31:534–540.
15. Myers K, Winters NC. Ten-year review of rating-scales. I: Overview of scale functioning, psychometric properties, and selection. *J Am Acad Child Adolesc Psychiatry* 2002;41:114–122.