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New Onset Chorea in a Previously Healthy 7-Year-Old

A 7-year-old right-handed previously healthy boy presented with progressive clumsiness, abnormal movements of the right arm, and gait and speech abnormalities. Two weeks before his presentation, his father noted involuntary, nonrhythmic, nonsuppressible movements of his right arm that progressively developed into constant, writhing movements that did not occur during sleep. The movements had not changed in character but had progressively increased in intensity. He had decreasing dexterity at school and home and occasionally slurred words at the ends of sentences. He had no recent illnesses and no recent history of sore throat.

On admission, cranial nerve examination was notable for the presence of a darting tongue: when asked to protrude his tongue midline, the patient made fast, repetitive, nonvoluntary, retracting motions (**Video**; available at www.jpeds.com).[1] On right upper extremity examination, a unilateral milkmaid's sign was observed: when asked to grip the examiner's fingers, the left arm (**Figure**, A, unaffected side) demonstrated smooth constant grip, whereas the right arm (**Figure**, B, affected side) demonstrated an irregular pattern of flexion and extension and was unable to sustain consistent grip pressure (**Video**).[1] Cardiovascular examination revealed a soft, II/VI, systolic-ejection murmur at the left lower sternal border.

Antistreptolysin O titer and anti-DNASE B were elevated. His electrocardiogram showed a prolonged PR interval for age, and echocardiogram showed trivial mitral valve regurgitation. The patient was diagnosed with acute rheumatic fever (ARF) without carditis. Monthly intramuscular penicillin G benzathine was started and will continue until age 21 years. He received a 2-week steroid burst followed by a taper. He had full resolution of symptoms within 8 weeks.

The differential diagnosis for chorea is broad and can be divided into acquired and hereditary etiologies.

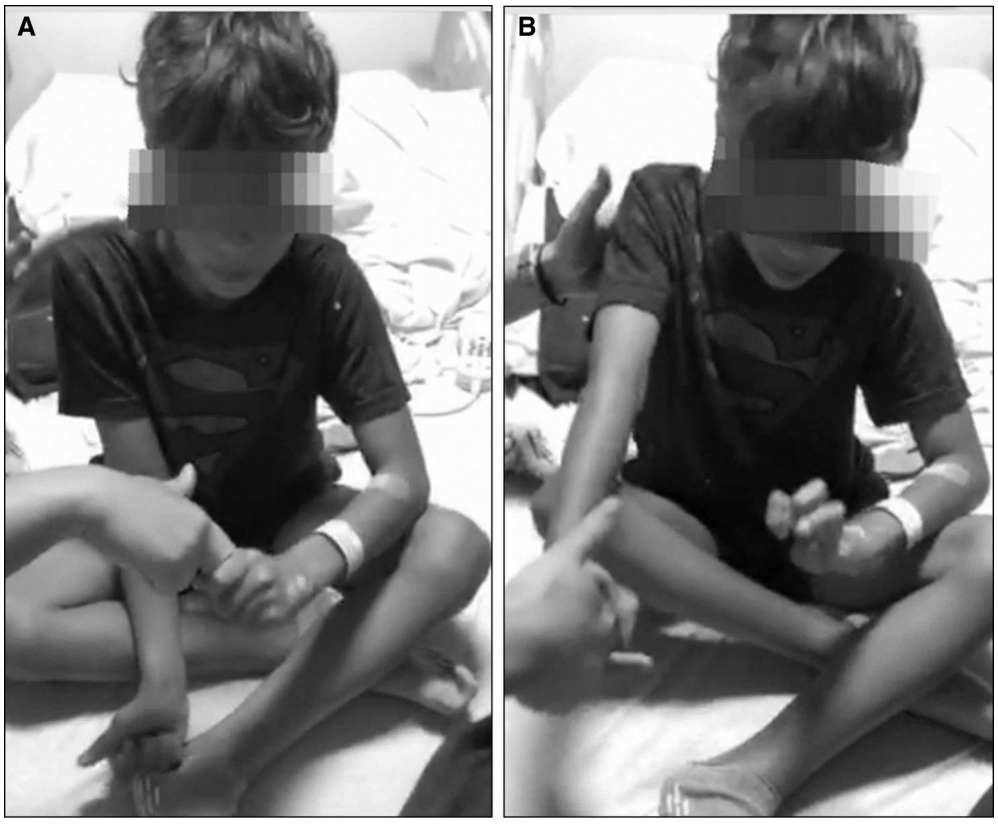

Figure. Milkmaid's sign: **A,** Unaffected arm. **B,** Affected arm. Still images are representative samples of the **Video**.

J Pediatr 2020;223:216-7.
0022-3476/$ - see front matter.
https://doi.org/10.1016/j.jpeds.2020.04.053

The most common cause of chorea in children is ARF, an autoimmune sequelae of Group A beta-hemolytic *Streptococcus* (GABHS) pharyngitis.[2] The diagnosis of ARF is based on the revised Jones Criteria.[3]

Sydenham chorea is the only major Jones criteria that can define a case of ARF on its own.[4] The pathophysiology of Sydenham chorea is only partially understood but is generally believed to be secondary to the binding of anti-N-acetyl-β-D-glucosamine antibodies with lysoganglioside and tubulin that are present in neurons of the basal ganglia.[5] Clinically, Sydenham chorea is characterized by neurologic motor symptoms with jerking movements of the arms and legs, as well as truncal restlessness. The chorea is most commonly bilateral; however, in 20%-30% of patients it can be unilateral. A classic feature is motor impersistence, which is the inability to sustain voluntary actions. This is most clearly evidenced by the darting tongue and milkmaid sign shown during our patient's examination. Dysarthria is also common, as well as emotional lability.[6] Sydenham chorea presents typically within 4-8 weeks after initial GABHS infection, but can appear up to 8 months later,[7] which makes the diagnosis more challenging as inflammatory markers may be normal and it may not be possible to document laboratory evidence of preceding GABHS infection .

The treatment of Sydenham chorea (and ARF) is supportive. Some studies have shown a hastened recovery of Sydenham chorea with the use of systemic steroids.[8] Recurrence of ARF and rheumatic heart disease can be prevented with the use of prophylactic penicillin.[9] ■

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Tense Bullae on the Hands

A 5-year-old girl presented to the dermatology clinic for evaluation of pruritic papules and blisters on the hands and forearms for three months. She was seen by her pediatrician, diagnosed with dyshidrotic eczema, and prescribed topical triamcinolone with no improvement. She reported itching everywhere, but her parents noted she most frequently scratched her hands and forearms. The itching was worse at night. No close contacts experienced pruritus. She had no history of inflammatory dermatoses or other medical problems.

Physical examination revealed tense erythematous bullae with discrete dermal-appearing papules and small nodules along the ventral aspect of the fingers, and excoriated erythematous papules on the flexor wrists (**Figure**). There was no other mucocutaneous involvement. The child was well-appearing with normal vital signs. Mineral oil preparation of scraping from the wrist papules revealed scabies mites. The child was diagnosed with bullous scabies and prescribed permethrin, with resolution of symptoms.

J Pediatr 2020;223:217-8.

https://doi.org/10.1016/j.jpeds.2020.04.050