LETTERS TO THE EDITOR

THE "SPLIT HAND SYNDROME"

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I am writing concerning the article by Kuwabara and coworkers that appeared in the July 1999 issue.[1] For the past 9 or 10 years, whenever we have performed nerve conduction studies (NCS) on patients with suspected amyotrophic lateral sclerosis (ALS) who have had hand weakness, we have routinely recorded compound muscle action potentials (CMAPs) from the ulnar nerve-innervated hypothenar muscles and first dorsal interosseous (FDI), as well as the median nerve-innervated thenar muscles (abductor pollicis brevis/opponens pollicis). Based on decreases in the CMAP amplitudes, we found, as Dr. Kuwabara and coworkers report, that the thenar muscles frequently are substantially more denervated than are the hypothenar muscles in ALS. However, the FDI is even more frequently severely denervated than the others, and sometimes it is significantly involved before the thenar muscles. We labeled this the "split hand," since the muscles on the lateral aspect of the hand preferentially were affected, compared to those on the medial aspect. We first reported this finding in 1992,[2] and then again in 1994.[3] The "split hand" has been discussed in two recently published books dealing with ALS.[4,5] However, we would point out certain caveats. First, not every ALS patient with hand-wasting shows such dissociation. In many, the wasting appears to be more or less equal between the lateral and medial hand muscles, especially when the process is advanced. Moreover, on rare occasions, a reverse "split hand" is seen, whereby the hypothenar muscles are the most severely affected. Second, this phenomenon is not limited to ALS. We have encountered it with Werdnig–Hoffmann disease, "benign" focal motor neuron disease, and remote poliomyelitis. However, it does seem to be specific for anterior horn cell disorders, as opposed to C8 radiculopathies. We consider the demonstration of a "split hand" during NCS on a patient referred to the EMG Laboratory with suspected ALS to be a very ominous finding. Almost never has a subsequent needle electrode examination demonstrated any alternative diagnosis.

CCC 0148-639X/00/010138-02

1. Kuwabara S, Mizobuchi K, Ogawara K, Hattori T. Dissociated hand muscle involvement in amyotrophic lateral sclerosis detected by motor unit number estimates. Muscle Nerve 1999; 22:870–873.
2. Wilbourn AJ. Dissociation of muscle wasting of ulnar-innervated hand muscles with motor neuron disease. Third International Symposium on ALS/MND; 1992 November; Solihull, UK.
3. Wilbourn AJ, Sweeney PJ. Dissociated wasting of medial and lateral hand muscles with motor neuron disease. Can J Neurol Sci 1994;21(Suppl 2):S9.
4. Wilbourn AJ: Electrodiagnostic evaluation of the patient with possible ALS. In: Belsh JM, Schiffman PL, editors. Amyotrophic lateral sclerosis. Armonk, NY: Futura Books; 1996. p 193.
5. Mitsumoto H, Chad PA, Pioro EP. Amyotrophic lateral sclerosis. Philadelphia: FA Davis; 1998. p 67.

RESPONSE TO CARBAMAZEPINE OF RECESSIVE-TYPE MYOTONIA CONGENITA

Myotonia congenita is characterized by myotonia with the "warm-up" phenomenon and generalized muscle hypertrophy; muscle strength deficits are either absent or, when present, are minimal and transient. Onset usually occurs in the first decade of life, and the clinical picture tends to be stable over time.[3,9] The disease can be inherited as an autosomal dominant or autosomal recessive trait[3,4,8–10]; the latter type is generally more severe. The motor impairment due to myotonia is usually mild, and medical therapy is not normally required, unless respiratory symptoms (e.g., stridor) or ocular disturbances (e.g., blurred vision, strabismus) occur or the myotonia is severe. The main problem when deciding upon a therapeutic course is choosing the safest and most effective drug.[7]

A 5-year-old boy was referred to us because of stiffness, difficulty in starting any motor activity, and frequent falls. From the age of 15 months, the patient had presented difficulty in walking and clumsiness. The family history was

noncontributory and, in particular, negative for myotonic phenomena. Examination of the child showed all the classic features of recessive myotonia congenita: active myotonia with the warm-up phenomenon, percussion myotonia of various skeletal muscles, and generalized muscle hypertrophy without associated weakness. Needle electromyography (EMG) revealed myotonic discharges but no other abnormality. We diagnosed myotonia congenita, very likely of recessive form. Both parents had a normal EMG, but this does not exclude the possibility that they are carriers of the gene.[5] The clinical picture remained stable until the child was 10 years old, when myotonia worsened, causing frequent falls and prompting a request for pharmacological therapy.

The drugs most commonly used in the treatment of myotonia are the type I antiarrhythmic agents such as phenytoin, procainamide, quinine, and mexiletine.[7] Of these, mexiletine seems to be relatively safe but is not commonly used in children; the others may have a variety of side effects. In the belief that any drug that inactivates sodium channels will block or reduce myotonic phenomena,[2,6,7] we started the patient on a course of carbamazepine (CBZ) treatment (daily dosage of 20 mg/kg administered in three doses). Before treatment, it took the boy 15 s to climb five steps after a 15-min rest. After 1 month of therapy, it took the patient 7 s to climb five steps under the same conditions. After 6 months of therapy, the results obtained were stable and the patient was without any side effects. The plasma level of CBZ was 4.9 mg/mL (normal range, 4–12 mg/mL). Currently, the patient can run more easily, and he describes his movements when walking and when getting up from the floor as "more fluid" than before. His falls are less frequent, as he is able to quickly recover his equilibrium when he loses his balance. The degree of myotonic involvement has improved from moderate to mild according to Becker's rating scale.[1]

Even though evidence of improvement in a single case is not a sufficient basis on which to reach definite conclusions, the use of CBZ in severe recessive myotonia congenita might be considered, with mexiletine as a possible second choice should CBZ prove ineffective.

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1. Becker PE, Lenz W, Vogel F, Wendt GG. Myotonia congenita and syndromes associated with myotonia. Stuttgart: Thieme; 1977.
2. Cannon SC. From mutation to myotonia in sodium channel disorders. Neuromusc Disord 1997;7:241–249.
3. Cannon SC. Ion-channel defects and aberrant excitability in myotonia and periodic paralysis. Trends Neurosci 1996;19: 3–10.
4. Koch MC, Steinmeyer K, Lorenz C, Ricker K, Wolf F, Otto M, Zoll B, Lehmann-Horn F, Grzeschik KH, Jentsch TJ. The skeletal muscle chloride channel in dominant and recessive human myotonia. Science 1992;257:797–799.
5. Koty PP, Pegoraro E, Hobson G, Marks HG, Turel A, Flagler D, Cadaldini M, Angelini C, Hoffman EP. Myotonia and the muscle chloride channel: dominant mutations show variable penetrance and founder effect. Neurology 1996;47:963–968.
6. Kwiecinski H, Ryniewicz B, Ostrzycki A. Treatment of myotonia with antiarrhythmic drugs. Acta Neurol Scand 1992;86: 371–375.
7. Moxley III RT. Myotonic disorders in childhood: diagnosis and treatment. J Child Neurol 1997;12:116–129.
8. Pusch M, Steinmeyer K, Koch MC, Jentsch T. Mutations in dominant human myotonia congenita drastically alter the voltage dependence of the CIC-1 chloride channel. Neuron 1995;15:1455–1463.
9. Zellweger H, Pavone L, Biondi A, Cimino V, Gullotta F, Hart M, Ionasescu V, Mollica F, Schieken R. Autosomal recessive generalized myotonia. Muscle Nerve 1980;3:176–180.
10. Zhang J, George Jr, Griggs RC, Fouad GT, Roberts J, Kwiecinski H, Connolly AM, Ptacek LJ. Mutations in the human skeletal muscle chloride channel gene (CLCN1) associated with dominant and recessive myotonia congenita. Neurology 1996; 47:993–998.