LETTERS TO THE EDITOR

ULTRASOUND VIEW OF A TRAUMATIC TWO-LEVEL MEDIAN NERVE LESION

After falling and landing on his outstretched hand in a motorbike accident, a 40-year-old man experienced a traumatic distal fracture of the radius and ulna on the left side; surgical treatment was successful.

Soon after the trauma, however, the patient complained of paresthesias in the first 3 fingers and palm of the left hand, severe impairment of hand movements, mild weakness of wrist flexion, and pain in his arm. Physical examination showed severe weakness of the abductor pollicis brevis (APB) (Medical Research Council Scale (MRC) 1/5] and the opponens pollicis (OP) (MRC 1/5), while the flexor digitorum profundus of the second and third digits (FDP), the flexor pollicis longus (FPL), and the flexor carpi radialis (FCR) were less weak (MRC 3/5). Although weakness of the proximal muscles could have been pain-related, the consistency of muscle contraction suggested neurogenic involvement. A sensory deficit in the median nerve territory (thenar area and fingers) was observed. Electrophysiological studies showed severe involvement of the APB and the OP (denervation signs and deficit of recruitment); there was only reduced neurogenic recruitment in the median-innervated forearm muscles [flexor digitorum profundus (FDP) of the second and third digits and FCR]. These findings, together with the clinical deficit, suggested a proximal median nerve lesion with greater involvement of distal median nerve branches.

We then scanned the median nerve along its course using a 5–18-MHz linear-array transducer (Esaote Lab 25 Gold, Genoa, Italy). At the point of fracture, in the third distal region of the forearm, the median nerve was focally swollen, with increased cross-sectional area (CSA 13 mm^2; Fig. 1A and B). Toward the middle of the forearm the median nerve had normal structure and echogenicity (CSA 7 mm^2; Fig. 1C and D). At the antecubital fossa, however, we observed a second focal swelling (CSA 18 mm^2, while contralateral CSA was 9 mm^2) of the median nerve, which was hypoechoic with two particularly swollen single fascicles (CSA 4 mm^2 and 2 mm^2, respectively; Fig. 1E and F).

As a result we hypothesized a double median nerve lesion. The distal lesion was a direct consequence of the bone fracture. The fracture may have caused proximal stretching of the median nerve (Fig. 1G) at the point where it enters the forearm between the two heads of the pronator teres muscle.

In our laboratory, of the 56 traumatic nerve lesions examined over a period of 13 months, we diagnosed 4 cases of nerve damage at two sites, based on ultrasound evaluation, performed not only at the point of the lesion but also along the whole length of the nerve. In the literature there is reference to the so-called "double crush

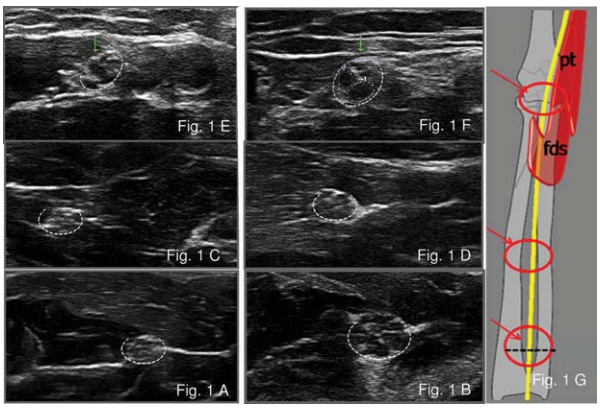

FIGURE 1. (A, B) The median nerve at the site of distal fracture. At the unaffected site **(A)**, the nerve was normal in structure, size, and echogenicity; at the affected site **(B)**, the median nerve was focally swollen, with increased cross-sectional area (CSA 13 mm^2). **(C, D)** Median nerve in the middle of the forearm. The nerve had normal structure and echogenicity (CSA 7 mm^2); there was no difference between the unaffected **(C)** and affected **(D)** sites. **(E, F)** The median nerve at the antecubital fossa. On the unaffected side **(E)**, the median nerve had normal structure and echogenicity (CSA 9 mm^2). At the affected site **(F)**, the CSA of the nerve was 18 mm^2 with two particularly swollen fascicles (CSA 4 mm^2 and 2 mm^2, respectively). **(G)** The median nerve along its course. We hypothesized a double median nerve lesion. The distal lesion was a direct consequence of the bone fracture, and the proximal lesion was the result of nerve stretch. Immediately distal to the antecubital fossa, the median nerve, at the point where it enters the forearm, runs between the two heads of the pronator teres (PT) muscle before passing deep to the flexor digitorum superficialis (FDS). In our case, the distal trauma may have caused stretching of the median nerve at this site. [Color figure can be viewed in the online issue, which is available at wileyonlinelibrary.com.]

syndrome" and "reversed double crush syndrome"[2] in relation to nerve compression, but the clinical picture, underlying mechanisms, and timing of the double crush syndrome are different from those hypothesized in a traumatic "double-level" nerve lesion. We believe that injury due to stretching could occur where the nerve is located at an anatomical–physiological angle or site of fixation.

In conclusion, ultrasound evaluation simplifies diagnosis of this kind of lesion, which is difficult to detect with electrophysiology alone. Treatment and rehabilitation are thus enhanced.

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TALIPES EQUINOVARUS AS LEADING SYMPTOM OF CONGENITAL MYOTONIC DYSTROPHY TYPE 2

In their study, Renard and colleagues described a 2.5-year-old patient with congenital bilateral talipes equinovarus (CTEV).[1] Both in the patient and his mother, CCTG expansions typical for myotonic dystrophy type 2 (DM2) were found in the *ZNF9* gene with 85 and 88 repeats, respectively. Mother and patient were otherwise clinically asymptomatic. The authors discussed a possible association of CTEV with DM2.

In 2008, we reported the first patient with possible congenital DM2.[2] In addition to reduced intrauterine movements and postnatal moderate muscular hypotonia, the patient also had congenital pes equinovarus. Our patient is now 5 years old and shows mild speech and motor retardation. Myotonic or other symptoms have not been noticed so far.

Our initial observation is in line with the findings of Renard and colleagues and suggests that a congenital variant of DM2 exists with CTEV as the leading early symptom. Many different neuromuscular diseases have been linked to congenital CTEV. The clinical findings in these two patients suggest that, in addition to myotonic dystrophy type 1,[3] DM2 could also be considered as a possible underlying disorder in patients with congenital CTEV, with or without other signs of an unclassified neuromuscular disease. Careful clinical and family analysis is recommended and, if appropriate, molecular testing could help to reach the right diagnosis.

Editor's note. Renard and colleagues were asked if they wished to reply to this letter. Because it agreed with their initial report, they felt that no reply was necessary.

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AWAJI ISLAND MODIFIED CRITERIA FOR ALS—INCREASED SENSITIVITY WITHOUT CHANGE IN SPECIFICITY: ARE THEY REALLY TWO SIDES OF THE SAME COIN?

We read with interest the article by Chen and colleagues on the use of the Awaji criteria for the electrodiagnosis (EDX) of amyotrophic lateral sclerosis (ALS) and its impact on sensitivity of diagnosis.[1] We agree that in many patients with suspected ALS the presence of fasciculation potentials, especially with widespread distribution on EDX studies, might suggest the diagnosis of motor neuron disease (MND). In our opinion, to substitute such findings for fibrillation potentials/positive sharp waves may prove to be premature. The strict Lambert criteria, which were subsequently modified in the El Escorial criteria, make certain such errors of oversensitivity could be avoided. The implications of their observations could be far reaching for both patients and physicians alike in clinical practice, and hence we offer the following comments.

Since the publication of the original Awaji criteria in 2008,[2] new data have been published that show a statistically non-significant occurrence of complex fasciculation potentials in both benign fasciculation syndrome and in ALS.[3] These observations suggest that even in the right clinical settings complex fasciculation potentials may not be specific for ALS.

Based on the proposed electrodiagnostic criteria the definitive ALS category in the Chen et al. study increased only by 1 case (2 definite before to 3 definite after EDX

examination) and not surprisingly, the authors reported an increase in the number of ALS cases classified as probable (24–37 EDX cases).[1] Further in their experience 2 definite ALS cases evolved into 7 definite cases by clinical diagnosis alone. It would be of interest to know the breakdown of the diagnostic class these 5 patients belonged to in the initial Awaji criteria. In Chen et al. report an ALS specialist evaluated 50% of the patients in the study group, resulting in a pretest probability bias toward correct diagnosis. This is probably different from routine clinical practice when neurologists are confronted with such cases. A larger cohort of patients with long-term follow-up could provide the statistical evidence to support such change in EDX practice.

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REPLY

We appreciate the interest of Drs. Govindarajan and Galvez-Jimenez in our work as well as their comments. In response, we first emphasize that the Awaji modifications apply to the electrodiagnostic criteria, which by themselves are never sufficient for the diagnosis of ALS. Second, the Awaji guidelines use the combination of fasciculation potentials with chronic neurogenic changes (defined as motor unit potentials of increased duration or amplitude, and/or reduced recruitment) as electrodiagnostic evidence of lower motor neuron dysfunction in the diagnosis of ALS. In our study, we did not use or propose using fasciculation potentials alone to diagnose motor neuron disease.

With regard to the characteristics of fasciculation potentials seen in ALS and benign fasciculation syndrome, we had no information on the waveform morphology or the firing frequency of the fasciculation potentials recorded in our study; as a retrospective analysis we used tabulated EMG reports. We did not claim in our study that the fasciculation potentials seen in ALS are more complex or unstable, as proposed by the expert consensus made in Awaji.

Regarding the initial Awaji diagnoses for the 5 patients who showed clinical progression and who later were diagnosed with clinically definite ALS, 4 (cases 2, 8, 61, and 69) were initially diagnosed with probable ALS, whereas 1 (case 66) was initially diagnosed with possible ALS by either Airlie House or Awaji criteria (see the supplementary table accompanying the article). We did not discuss these results in our investigation, because (a) the sample size was small, and (b) it was not our objective to determine whether the certainty of the diagnosis was correlated or predictive of clinical progression, although that is certainly an important issue.

Since the publication of our study, several reports have emerged suggesting that the use of the Awaji guidelines can be useful in the early diagnosis of ALS.[1–3] We agree that a larger prospective study without selection bias would be helpful in determining the utility of the Awaji guidelines in routine clinical practice with the aim toward the earlier diagnosis of ALS.

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CAN WE ACCURATELY MEASURE THE ONSET LATENCY TO THE FIRST DORSAL INTEROSSEOUS?

I have read the short report by Takahashi and Robinson with interest.[1] First dorsal interosseous (FDI) and abductor digiti minimi (ADM) compound muscle action potentials (CMAP) from two healthy subjects were recorded using four bipolar montages (A–D) and four monopolar montages (E–F). The investigators proposed that the initial deflections of the belly–tendon "FDI" (montages C and D) and "ADM" (montage A) CMAPs might be affected not by the activation of FDI and ADM themselves, but by volume-conducted potentials from more proximal ulnar-innervated intrinsic muscles. These volume-conducted potentials should be incorporated in "belly–CMAP" (montages

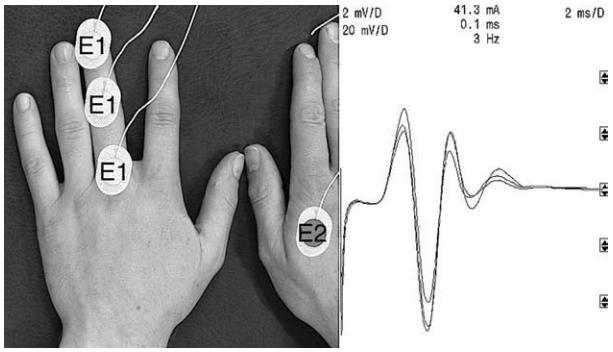

FIGURE 1. Three monopolar recordings (superimposed curves) with increasing distance between the ulnar stimulation site at the wrist and the active recording electrode E1 placed over digit 3 (E2 contralateral). Latencies to potential peaks do not increase with increasing distance.

A–D) but also in "tendon–CMAP" (montages F–H).

From my point of view, the "tendon–CMAP" is not a volume-conducted potential, but rather it is a far-field potential. More precisely, it is a stationary wave originating from myotendinous junctions from all ulnar-innervated muscles, including more proximal muscles than the FDI.[2–5] Proof of this is illustrated in Figure 1. When the distance between the stimulation site at the wrist and the recording electrode E1 placed over digit 3 (E2 contralateral) increases, latencies to potential peaks do not change, indicating that these potentials are not volume-conducted. Thus, potentials called "belly–CMAP" correspond to the summation of the FDI CMAP and myotendinous far-field potentials, whereas potentials called "tendon–CMAP" are mainly related to myotendinous far-field potentials. Consequently, the initial positive deflection in montage B (FDI index) indicates that positive far-field potentials, originating from myotendinous junctions, are steeper when recorded over the index than over the FDI belly.

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REPLY

We appreciate the interest and the comments by Dr. Wang regarding our short report on the origin of the response recorded from over the first dorsal interosseous (FDI) muscle.

We believe that Dr. Wang's letter and figure augment the information provided in our report. In our report, we used the term "volume-conducted potentials" to indicate that potentials originated not from directly under the electrode, but from a distance. Specifically, we consider volume conduction as "spread of current from a *potential* source through a conducting medium, such as body tissues."[1]

We do not disagree with Dr. Wang that these volume-conducted potentials could be, at least in part, far-field potentials from myotendinous junctions of proximal muscles, although we cannot be sure which muscles contribute and how much of the far-field potential is from the myotendinous junction vs. proximal muscle bellies. Because the differences between "near-field" and "far-field" are somewhat arbitrary,[1] we did not use that term in our report.

Our objective in the short report was to demonstrate the significant influence of the tendon electrode on the belly–tendon compound muscle action potential waveforms recorded from FDI. Dr. Wang's information provides additional information on possible origin of the potentials recorded at the tendon and reinforces the need to carefully interpret responses recorded in this region of the hand.

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