

Low compound muscle action potential amplitude with good prognosis in Guillain-Barré syndrome: a case report

Wei Chih HSU MD, Jiann Horng YEH MD, Hou Chang CHIU MD, Li Hsia TSENG RN

Department of Neurology, Shin Kong Wu Ho-Su Memorial Hospital, Taipei, Taiwan

Abstract

A 74-year-old woman presented with acute progressive quadriplegia and became ventilator-dependent on the third day after onset. The amplitudes of compound muscle action potential (CMAP) were less 10% of normal in all motor nerves. In spite of existence of all poor prognostic factors (low CMAP amplitude, rapid progression, need for ventilatory support, and older age), she recovered dramatically after plasmapheresis. The electrophysiological studies showed proportional improvement in the serial follow-ups. The rapid recovery in clinical and electrophysiological features suggest that distal conduction block rather than axonal degeneration is the more likely cause of low CMAP amplitude. This case support that sequential electrodiagnostic studies are useful in evaluating the prognosis of GBS. When there is low amplitude CMAP in the initial study, sequential study will be able to differentiate the distal conduction block with good prognosis from axonal degeneration with poor prognosis.

Key words: Guillain-Barré syndrome, compound muscle action potential, prognosis

INTRODUCTION

The clinical features and time course for recovery in the Guillain-Barré syndrome (GBS) are known to vary. The typical GBS is characterized by progressively ascending flaccid paresis with depression or loss of tendon reflexes. However, in some, a fulminant course with rapid progression, slow recovery, and severe disability occurs. Low compound muscle action potential (CMAP) amplitude, aging, rapid progression, and ventilator dependence are the major predictors of poor outcome.^{10,11} We report a case of GBS with all the above poor predictors, but rapid recovery in clinical and electrophysiological aspects after plasmapheresis.

CASE REPORT

A 73-year-old lady suffered from acute weakness of both legs on 27th May 1996. The weakness progressively involved the muscles of upper limbs on the next day. Three days after onset, she was admitted to our hospital in bedbound state. There was no preceding history of upper respiratory airway or gastrointestinal infection.

On assessment, she was conscious. Blood pressure was 177/104 mmHg. She was slightly pyrexial (37.6°C) and had regular respiration (18/min). Cranial nerve examination revealed

mild facial diplegia and dysphagia. There was generalized weakness of four limbs with predominantly proximal involvement. We evaluated the muscle strength and disability by Medical Research Council (MRC)-sumscore¹ and Hughes functional grade.² The MRC-sumscore summed the MRC scores of the muscles for arm abduction, forearm flexion, wrist extension, leg flexion, knee extension, and foot dorsiflexion in the either side. The Hughes functional grade system was defined as 0 for healthy, 1 for minor signs or symptoms, 2 for able to walk 5 meters without a walker or equivalent support, 3 for able to walk 5 meters with a walker or support, 4 for bed or chair bound, 5 for required assisted ventilation, and 6 for dead. In our patient, the MRC-sumscore was 9 and functional grade was 4 initially. No objective sensory loss could be identified, despite complaints of distal numbness. The tendon reflexes and Babinski sign were all absent. The electrophysiological tests performed immediately after admission disclosed prolonged distal motor latencies, low CMAP amplitudes (less than 10% of the normal), and relatively preserved sensory function. Sudden onset of apnea and decreased consciousness developed several hours later. After mechanical ventilation, consciousness recovered dramatically. Double filtration

plasmapheresis was started on the next day with plasmacure (Kuraray, Osaka, Japan) as a plasma separator and Evaflux 4A as a plasma fractionator. Episodic perspiration and unstable vital signs with hypotension (72/41mmHg) were noted during plasmapheresis. The patient was extubated after five sessions of plasmapheresis. The MRC sumscore and functional grade improved significantly during hospitalization (Fig.1). Electrophysiological studies on the 20th and the 42th day showed that CMAP amplitude increased markedly in contrast to the change in the conduction velocities (Table 1). However, no significant change was noted in the sensory nerve studies. She was discharged on the 30th day with nearly total recovery except generalised areflexia.

DISCUSSION

The clinical course and prognosis in the GBS are widely variable.³⁻⁵ Less than 5% of patients with GBS are characterized by rapid progression to severe paralysis and respirator dependence within 2 to 5 days of the onset with very poor and delayed recovery.^{6,7} Brown and Feasby reported that very low CMAP amplitude in the early course of the disease correlated well with the frequency of axonal denervation in the

muscles and unfavorable clinical recovery.⁸ In a prospective study done by Winer et al, the major features which associated with persistent disability were the time taken to become bedbound within four days, requirement for ventilation, age greater than 40 years, and small or absent CMAP of abductor pollicis brevis.⁹ McKhann et al proposed four parameters of poorer outcome: low CMAP amplitude less than 20% of normal, older age, time from onset to maximal deficits within 7 days, and need for ventilatory support.¹⁰ Among these considerations, the most powerful predictor was a low distal CMAP amplitude.^{10,11}

Our patient had all the poor prognostic factors (old age, rapid progression to bedbound status within 4 days, ventilator dependence, and low CMAP amplitude), but recovered dramatically both clinically and in her electrophysiological parameters after plasmapheresis.

There are two possible explanations for the low CMAP amplitude or inexcitable motor nerves in GBS, a distal conduction block^{4,13,14} or an axonal degeneration^{9,12} in the motor axons. Cros and Triggs reported that recovery might occur within weeks with restoration of CMAP amplitude¹³ and that distal conduction block might be the pathophysiology in such inexcitable

TABLE 1: Results of nerve conduction studies

Nerve stimulated	Motor		
	DL (ms)	CV (m/s)	Ampl (mV)
Day 4			
Median	7.3*/6.7#	31.0/39.7	0.4/1.0
Ulnar	4.1/3.3	39.7/46.9	0.2/0.3
Peroneal	7.7/7.6	42.2/38.2	0.3/0.9
Tibial	8.2/10.1	33.7/39.7	0.2/0.2
Day 20			
Median	7.2/6.4	41.5/51.8	3.1/2.9
Ulnar	4.5/4.3	54.9/45.5	4.0/4.1
Peroneal	7.2/6.2	36.7/29.4	1.8/1.8
Tibial	6.8/6.5	34.7/34.6	4.9/1.9
Six Months			
Median	4.9/4.7	46.1/51.4	6.7/5.7
Ulnar	3.1/2.9	55.0/50.0	9.2/6.9
Peroneal	4.6/4.7	44.0/47.2	2.6/2.6
Tibial	4.5/4.3	43.2/43.0	9.1/4.9

DL = distal latency, CV = conduction velocity, Ampl = amplitude, * right, # left

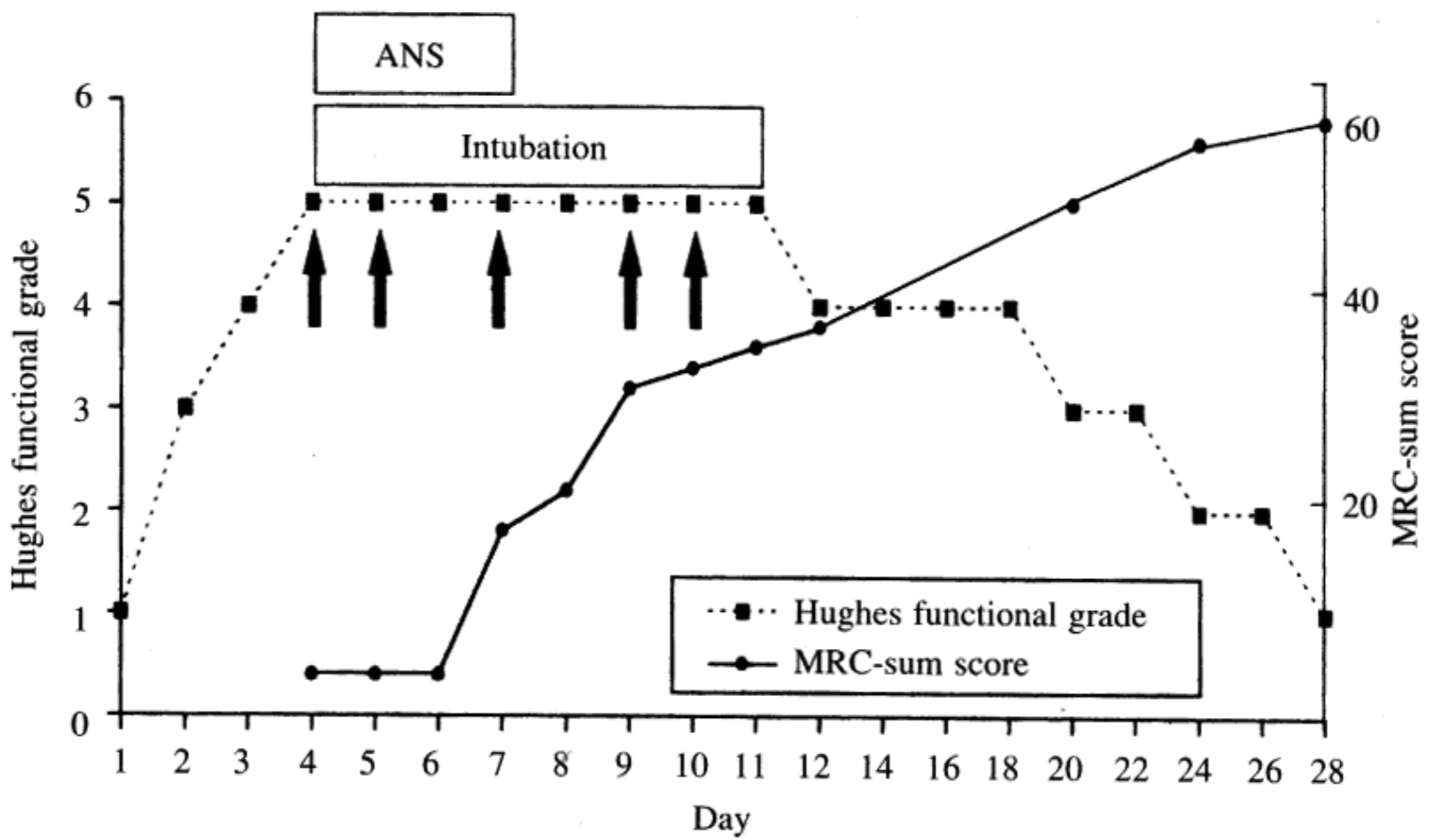


FIG. 1: Clinical course in a patient with Guillain-Barré syndrome
ANS = autonomic system dysfunction, arrow indicates plasmapheresis

or small CMAP studies. Distal conduction block is likely to be underlying pathological mechanism in our patient because of her rapid recovery. On the other hand, progressive loss of excitability and conduction indicates that the nerve fibers are undergoing axonal degeneration.⁷ This is correlated with poor outcome.¹⁵

This case supports the fact that sequential electrodiagnostic studies are useful in evaluating the prognosis of GBS.⁴ When there is low amplitude CMAP in the initial study, sequential studies will be able to differentiate between distal conduction block with good prognosis from axonal degeneration with poor prognosis.

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