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ELECTROMYOGRAPHIC ANALYSIS IN SPINAL PROGRESSIVE MUSCULAR ATROPHY, JUVENILE UNILATERAL MUSCULAR ATROPHY, NEURAL PROGRESSIVE MUSCULAR ATROPHY AND MYELORADICULONEURITIS

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ABSTRACT

Recently, as diagnostic aids of neurological science, not only electromyography but also other techniques such as motor nerve conduction velocity, evoked potentials and strength-duration curve have been performed. In this paper, the first, the standard value of conduction velocities and the critical value of reduced conduction velocities in pathologic condition were discussed, because the problems of the values have not been determined with constancy yet. The second, applying four procedures to spinal progressive muscular atrophy, juvenile unilateral muscular atrophy, neural progressive muscular atrophy (Charcot-Marie-Tooth disease) and myeloradiculoneuritis, mutual interrelation of the measured values was studied and discussed the parallelism to clinical sigh and symptom with the disease. In combination of electromyography with subjective and qualitative expression and the other techniques with objective or numerical expression, the pathognomy of the cases could be assumed and, too, the course of the diseases in repeating the procedures frequently.

In clinical neurology electromyography with a unipolar and concentric needle electrode has been employed as a diagnostic aid. However, since in 1948 Hodes, Larrabee and German[1] pointed out clinical application of conduction velocity of the motor axons, many investigators have reported motor nerve conduction velocity in normal and neurological subjects. Among a few electrodiagnostics, Trojaborg[2] (1962) applied the technique of electromyography, motor nerve conduction velocity and strength-duration curve to polyneuropathy and non-traumatic mononeuropathy, and studied their correlation of the methods. Furthermore, Dawson and Scott[3] (1949) recorded nerve action potentials through the skin in man, which included evoked potentials measured at the wrist or ankle stimulating on the elbow or knee and sensory nerve conduction velocity from the finger or toe to the wrist or ankle, and there-

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after several reports have been presented to neurological science. In comparison with electromyography evaluated qualitatively, measurements of both motor nerve conduction velocity and evoked potentials may be characteristic of quantitative estimation of the peripheral nerve lesion. However, it seems to be a problem that the technique of measurement of the conduction velocity differs with laboratories and normal range of the conduction velocity has not been determined as a constancy. Purpose of this paper is to show some results of normal subjects and patients with several neurological diseases such as spinal progressive muscular atrophy, juvenile unilateral muscular atrophy, neural progressive muscular atrophy (Charcot-Marie-Tooth disease) and myeloradiculoneuritis in application of the electrodiagnostics as stated previously.

METHODS

1. Electromyography: A unipolar and concentric needle electrode was employed to small muscles of the hand and foot in all subjects.

2. Motor nerve conduction velocity: Needle electrode for stimulation and surface electrode for recording were used. In ulnar, median, peroneal and tibial nerves a proximal stimulation to the nerves was applied to the elbow or the vicinity of the knee joint and the distal stimulation to the wrist or ankle. The stimulus employed was 120 volts in maximum with 0.1 to 1.0 milliseconds. The recording was performed on the hypothenar eminence for ulnar nerve, the thenar eminece for median nerve, M. extensor digitorum brevis for peroneal nerve and M. flexor hallucis brevis for tibial nerve.

3. Evoked potentials: Needle electrodes for stimulation and recording were applied to the same points as the measurement of motor nerve conduction velocity and condition of stimulation was 120 volts with duration of 0.1 milliseconds in maximum.

During measurements of the conduction velocities room temperature was kept at between 21° and 25°C, and skin temperature on the subject remained between 30° and 35°C except for some instances with peripheral nerve lesion. Dividing the distance between the stimulating points by the difference of latencies in measurement, the conduction velocities were calculated.

4. Strength-duration curve: The curve was plotted by voltages of stimulation which occurred minimal muscle contraction, providing the voltage of 0.1 to 80 milliseconds. Discontinuity of the curve and elevation over 2 in ratio of stimulating voltages with 0.1 and 80 milliseconds was determined to be an abnormal pattern, in which the former was classified as partially denervated pattern and the latter as denervated pattern. Examined muscles were the thenar and hypothenar eminences of the hand, and M. extensor digitorum brevis and M. flexor hallucis brevis of the foot, corresponding to those

in measurements of the conduction velocities.

MATERIALS

1. Control group: As this group healthy fifteen male subjects with age distribution of 16 to 49 year-old were examined, in each of whom motor nerve conduction velocity and evoked potentials of ulnar and median nerves in the upper extremity, and peroneal and tibial nerves in the lower extremity were measured.

2. Spinal progressive muscular atrophy: There were examined nine patients including three patients with Aran-Duchenne type and six patients with Vulpian-Bernhard type, in the latter of which two patients were done only in the upper extremity. The youngest patient was 35 year-old and the eldest 59 year-old. Their clinical courses since the onset were three to eleven years.

3. Juvenile unilateral muscular atrophy: This disease was reported first by Hirayama *et al.*[4] (1959) and Takagi *et al.*[5] (1959) and reveals muscular atrophy on the unilateral forearm and hand of young adult predominantly in male. In this group ten patients were examined on both the affected and non-affected sides of the arms. Age distribution was 17 to 41 year-old, and duration of the disease since the onset was two to twelve years.

4. Neural progressive muscular atrophy (Charcot-Marie-Tooth disease): Examined patients of this group consisted of eight cases, all of whom were male. Three of them were brother's cases and the others sporadic. All patients were 20 to 50 year-old in age with clinical course of five to thirty years. They showed muscular atrophy and motor weakness below lower one-third of the thigh with mild sensory impairment, and some cases revealed minimal symptom in the upper extremity.

5. Myeloradiculoneuritis: This group contained thirteen patients, five males and eight females. Clinical course of them was three months to five years and their age at the examination was 16 to 56 year-old. Clinical observation revealed slight impairment of motor activity with mild muscular atrophy, hyperactivity of deep tendon reflexes with appearance of pathologic reflex, autonomic symptom in the lower extremity and sensory involvement below the level of the lower thoracic cord.

RESULTS

1. Control group:

As a control study fifteen of normal subjects were examined for motor nerve conduction velocity and evoked potentials of ulnar and median nerves between the elbow and wrist and also peroneal and tibial nerves between the

knee and ankle. Table 1 demonstrates the conduction velocities over distal portion of the extremities. In measurement of motor nerve conduction velocity the calculated values are very similar to the data reported by other investigators, indicating a little difference in each other (Hodes *et al.*[1] (1948), Thoma *et al.*[6] (1959), Mayer[7] (1963)).

In measurement of evoked potentials the conduction velocity was slower than motor nerve conduction velocity in each nerve. The conduction of potentials means not only that of the sensory nerve fibers but that of the motor nerve fibers in part, and may be called a compound nerve conduction. As the values of the conduction velocities in normal subjects are not distinctly constant at different laboratories, it is a great problem how much reduction of normal conduction velocity is considered as a pathologic condition. However, as seen in Table 1, the peripheral nerves examined have 50 to 60 meters per second in the conduction velocities. Accordingly, in the conduction velocities of ulnar and median nerves measured between the elbow and wrist it will be suggested that the values below 45 meters per second of motor nerve conduction velocity and 40 meters per second of evoked potentials can be estimated as an abnormal reduction and pathologic, and in peroneal and tibial nerves 40 meters per second of both conduction velocities abnormal (Blom *et al.*[8] (1964), Yomura[9] (1964)). In the following discussion this standard at the abnormal reduction has been utilized throughout, and in a group of the reduced conduction velocity the reduction and no response on the procedure were included.

TABLE 1. Conduction velocities in the forearm and lower leg in controls with age distribution of 16 to 49 year-old (meters per second)

Nerve	Number of individuals	Segment of nerve	Motor nerve conduction velocity		Evoked potentials	
			Mean	Range	Mean	Range
Ulnar	15	elbow to wrist	62.0±4.6	(72.5−52.2)	56.8±3.5	(70.5−50.6)
Median	15	elbow to wrist	62.7±4.6	(75.6−51.8)	56.5±5.4	(71.2−49.7)
Peroneal	15	knee to ankle	53.2±4.4	(65.6−41.9)	51.2±4.3	(63.3−45.9)
Tibial	15	knee to ankle	52.0±5.9	(68.7−43.9)	52.9±4.3	(65.2−43.9)

2. Spinal progressive muscular atrophy:

As seen in Table 2, clinical signs were more remarkable in the upper extremity than in the lower extremity, and all patients showed motor weakness, muscular atrophy, fasciculation, and reduced deep tendon reflexes in the extremities in a high percentage but had no appearance of pathologic reflexes and sensory loss. In the electromyographic finding at rest there was no any potential, and during voluntary effort severe reduction of interference voltage (severe loss of activating motor units) was more marked in the upper extremity. High amplitude NMU (Neuro-Muscular Unit) voltage over 4 mV was observed in six of nine patients in the upper extremity and six of seven

TABLE 2. Clinical signs, electromyography, motor nerve conduction velocity, evoked potentials and strength-duration curve in present series (see the text in detail)

Disease	Spinal progressive muscular atrophy		Juvenile unilateral muscular atrophy		Neural progressive muscular atrophy		Myeloradiculoneuritis
Site	upper extremity	lower extremity	affected side	non-affected side	upper extremity	lower extremity	lower extremity
Number of cases	9	7	10	10	7	8	13
Motor weakness slight	6	4	5	1	5	3	10
Motor weakness severe	3	0	5	0	0	5	1
Muscular atrophy slight	7	4	7	0	2	2	5
Muscular atrophy severe	2	0	3	0	0	6	0
Fasciculation	8	6	8	0	0	0	0
Tendon reflex hypoactivity	9	5	6	2	2	8	1
Tendon reflex hyperactivity	0	2	2	0	0	0	12
Pathologic reflex	0	0	0	0	0	0	6
Sensory loss slight	0	0	0	0	2	5	13
Sensory loss severe	0	0	0	0	0	0	0
Autonomic symptom	0	0	9	0	1	7	13
Fibrillation voltage	0	0	1	0	0	0	0
Fasciculation voltage	0	0	2	0	0	0	1
Reduced interference voltage slight	5	5	8	6	7	5	12
Reduced interference voltage severe	4	1	1	0	0	3	1
Low amplitude NMU voltage	1	1	0	0	2	7	7
High amplitude NMU voltage	6	6	10	9	1	4	3
Complex NMU voltage	1	0	3	0	0	1	4
Reduced motor nerve conduction velocity ulnar	2		0	0	2		
Reduced motor nerve conduction velocity median	1		1	0	1		
Reduced motor nerve conduction velocity peroneal		0				7	1
Reduced motor nerve conduction velocity tibial		0				6	7
Reduced evoked potentials ulnar	0		0	0	1		
Reduced evoked potentials median	0		0	0	1		
Reduced evoked potentials peroneal		1				8	6
Reduced evoked potentials tibial		1				5	8
Abnormal strength-duration curve ulnar	4		9	1/8	3		
Abnormal strength-duration curve median	4		8	4/8	5		
Abnormal strength-duration curve peroneal		4				6	7
Abnormal strength-duration curve tibial		—				8	3/7

patients in the lower extremity, but appearance of low amplitude NMU voltage and complex NMU voltage (polyphasic potential) were seen only in a few cases. Motor nerve conduction velocity was slowed in two of ulnar nerve and one of median nerve, and evoked potentials were abolished in one of the patients (Table 3). Strength-duration curve showed partially denervated pattern with discontinuity of the curve in four of nine patients of the upper extremity and four of seven patients of the lower extremity.

TABLE 3. Motor nerve conduction velocity (MCV) and evoked potentials (EP) in present series (meters per second)

Nerve			Spinal progressive muscular atrophy				Juvenile unilateral muscular atrophy			
			Tested nerves	No response	Mean	Range	Tested nerves	No response	Mean	Range
	elbow to wrist									
Ulnar	MCV	affected side	9	0	56.2	(75.8–24.8)	10	0	64.8	(72.7–53.3)
		non-affected side					10	0	63.1	(74.6–45.2)
	EP	affected side	9	0	61.3	(70.2–49.6)	10	0	57.9	(64.7–45.8)
		non-affected side					10	0	58.3	(74.8–45.6)
Median	MCV	affected side	9	0	56.8	(64.5–32.9)	10	0	57.4	(73.3–42.9)
		non-affected side					10	0	64.4	(71.8–53.0)
	EP	affected side	9	0	66.4	(68.3–46.6)	10	0	58.2	(69.7–47.2)
		non-affected side					10	0	57.9	(64.9–49.5)
	knee to ankle									
Peroneal	MCV		7	0	57.3	(67.0–45.4)				
	EP		7	1	58.5	(68.5–50.5)				
Tibial	MCV		7	0	50.7	(64.9–41.3)				
	EP		7	1	57.7	(68.8–46.4)				

Nerve		Neural progressive muscular atrophy				Myeloradiculoneuritis			
		Tested nerves	No response	Mean	Range	Tested nerves	No response	Mean	Range
	elbow to wrist								
Ulnar	MCV	7	0	58.3	(75.2–39.5)				
	EP	7	1	58.9	(67.4–49.6)				
Median	MCV	7	0	62.2	(75.5–35.2)				
	EP	7	1	59.2	(68.5–53.3)				
	knee to ankle								
Peroneal	MCV	8	5	39.3	(53.2–28.1)	13	0	50.9	(63.8–35.3)
	EP	8	8	—	—	13	6	49.5	(56.7–40.0)
Tibial	MCV	8	3	33.8	(51.6–10.9)	13	0	36.9	(52.1–11.2)
	EP	8	5	42.5	(53.2–35.0)	13	8	57.1	(66.5–44.5)

3. Juvenile unilateral muscular atrophy:

On the affected side, muscular atrophy was slight in seven of ten patients and severe in the others, involving moderate motor weakness, and tremor-like fasciculation was observed in eight patients and autonomic symptom (coldness and "Kaltparese") was complained in nine patients (Table 2). On the non-affected side slight motor weakness and reduced deep tendon reflexes were seen only in a few patients. Of electromyogram at rest fibrillation voltage in one patient and fasciculation voltage in two observed only on the affected side. Reduction of interference voltage was revealed in nine patients of the affected side and in six of the healthy side. High amplitude NMU voltage to probably detect lesion of the anterior horn cells was observed in all cases of the affected side and in nine of the non-affected, but complex NMU voltage

was seen in three patients of the affected side. In the conduction velocities (Table 3), one patient showed slight reduction only in measurement of motor nerve conduction velocity of median nerve of the affected side. Strength-duration curve revealed slightly partially denervated pattern in seventeen of twenty atrophic small hand muscles and five of sixteen non-atrophic muscles.

4. Neural progressive muscular atrophy (Charcot-Marie-Tooth disease):

Of this group, there was almost well-developed form in all cases, except for one case (Table 2). In the lower extremity such clinical signs as motor weakness, muscular atrophy, reduced deep tendon reflexes, sensory disturbance and autonomic symptom ("Kaltparese") were severer in degree than in the upper extremity. Electromyographic finding of this group showed slightly reduced interference voltage during voluntary effort in both extremities, providing increasingly reduced pattern in the lower extremity. And low amplitude NMU voltage was observed in two of seven patients in the upper extremity and seven of eight patients in the lower. High amplitude NMU voltage was seen in one of seven patients in the upper extremity and four of eight patients in the lower, whose region was restricted to lower one-third of the thigh observed just as the muscular atrophy. In measurements of conduction velocities in the upper extremity, two of ulnar nerve and one of median nerve revealed slight reduction in motor nerve conduction velocity, and evoked potentials showed disappearance in each of both nerves. In the lower extremity measurement of motor nerve conduction velocity showed no response in five of eight patients and slight reduction in two of eight patients on peroneal nerve, and no response in three, slight reduction in one and severe reduction in two of eight patients on tibial nerve. Furthermore, evoked potentials of the lower extremity had disappearance in all cases for measurement of peroneal nerve, while on tibial nerve there were disappearance in five of eight patients and slightly reduced conduction velocity in two patients (Table 3). Strength-duration curve showed partially denervated pattern in three of seven small hand muscles innervated by ulnar nerve and in five of seven muscles done by median nerve with denervated pattern in one patient. The curve in the lower extremity revealed more affected pattern, indicating denervated pattern in six of eight patients on small foot muscles innervated by peroneal nerve and in all cases on small muscles by tibial nerve.

5. Myeloradiculoneuritis:

Of clinical signs of this group with involvement of the lower extremity (Table 2), slight motor weakness was observed in ten of thirteen patients and sever in one, while slight muscular atrophy was noticed only in five patients. Twelve patients had hyperactivity of deep tendon reflexes in the lower extremity, except for hypoactivity in one patient, and six patients showed appearance of pathologic reflex. But sensory loss and autonomic symptom

were found in all patients. Electromyographic finding showed appearance of fasciculation voltage at rest in one case. Interference voltage during voluntary effort revealed slight reduction in twelve of thirteen patients and severe in one. The appearance of low amplitude NMU voltage in seven, high amplitude NMU voltage in three and complex NMU voltage in four were observed. In measurement of motor nerve conduction velocity, slight reduction of one patient on peroneal nerve was seen and tibial nerve showed sight reduction in five and severe reduction in two of fifteen cases. Evoked potentials could not be elicited in six peronal and eight tibial nerves. In plotting of strength-duration curve were there partially denervated pattern of four and denervated pattern of three of thirteen patients in peroneal nerve and the curve of the muscles innervated by tibial nerve was partially denervated pattern in one and denervated pattern in two of seven patients examined.

DISCUSSION

In neurological examination, measurements of nerve conduction velocities, especially motor nerve conduction velocity has been performed since the first report for clinical use of the technique by Hodes, Larrabee and German[1] in 1948, while electromyography had already become one of diagnostic aids in this field. But the measured values of motor nerve conduction velocity for normal subjects varied with reports by many authors, as the cause of which some factors are considered; the technique of measurement, i.e., choice of electrode on stimulation and recording (surface or needle electrode), characteristics of stimulator, individual difference in measurements of nerve length, and latencies between electrical artefact and onset of action potential and body temperature etc. Of the technique of measurement it may be suggested that the needle stimulating electrode is more available because of clinical necessity of smoothness and quickness in measurements of not only one nerve but also several nerves in the extremities and lesser stimulating current to the nerve. The body temperature has some influence on motor nerve conduction velocity (Henriksen[10] 1956) but English Schools have mentioned its little effect on clinical evaluation of the conduction velocity and not much attended to it.

Some of the factors on error of measurements have been well studied but some have not (Gassel[11] 1964). However, the values of normal subjects, as shown in Table 1, resemble the previous reports with the least difference (Hodes *et al.*[1] 1948, Mavor *et al.*[12] 1962, Mayer[7] 1963, Stensrud[13] 1963, Downie[14] 1964). Accordingly, it is necessary to consider the standard of nerve conduction velocity as pathologic condition as well as discussion on distinct constancy of the conduction velocity in normal subjects and the factors influenced to the measurements. In this paper the motor nerve conduction velocity of ulnar and median nerves measured between the elbow and wrist has been considered to

be pathologic in reduction below 45 meters per second, and of peroneal and tibial nerves between the knee and ankle be abnormal in reduction below 40 meters per second (Blom *et al.*[8] 1964, Yomura[9] 1964). In the conduction velocity of evoked potentials for this series, the reduction below 40 meters per second was adopted as the pathologic condition.

In electrodiagnostics of spinal progressive muscular atrophy, remarkable electromyographic findings, indicating a neurogenic atrophy pattern with poor interference voltage and high amplitude NMU voltage, were observed in parallel to neurological signs of the disease. Motor nerve conduction velocity and evoked potentials in the extremities showed almost normal value with a slight reduction in small percentage of the patients as well as having revealed a partially denervated pattern of strength-duration curve in a few muscles. This tendency of slight reduction of the conduction velocity in progressive muscular atrophy has been previously reported, in this group including amyotrophic lateral sclerosis by Henriksen[10] (1956) and in amyotrophic lateral sclerosis by Lambert *et al.*[15] (1957) and Blom *et al.*[8] (1964). Normal conduction velocity of the examined nerves in these diseases may be reasonable to pathological findings of the disease and give a considerable aid of clinical diagnosis.

Juvenile unilateral muscular atrophy at first presented by Hirayama *et al.*[4] (1959) and Takagi *et al.*[5] (1959) may have a clinical entity of predominant affection in young adult and muscular atrophy in lower one-half of the forearm and the hand unilaterally. At present time pathognomy of the disease is not clear. In electromyographic finding of this disease were there observed a neurogenic atrophy pattern with slightly poor interference voltage and high amplitude NMU voltage in the both sides of most cases. Motor nerve conduction velocity and evoked potentials of the upper extremities were entirely within normal range except for one patient with slight reduction (42.9 meters per second) in median nerve of the affected side, but strength-duration curve showed a slight partially denervated pattern in high percentage of hand small muscles examined in the affected side and in small percentage of the muscles in the non-affected side. The findings of these electrodiagnostics have, to some extent, similarity to those of amytrophic lateral sclerosis and spinal progressive muscular atrophy, and may give some suggestions to locate a main lesion in the anterior horn cell. But, unilaterally restricted muscular atrophy and slower progression than the other of progressive muscular atrophy do not forgive an easy conclusion to the pathognomy of the disease. However, it is realized that the electrodiagnostics may sometimes throw a light to an assumption of pathologic lesion in the disease.

Patients with neural progressive muscular atrophy (Charcot-Marie-Tooth disease) showed findings of electrodiagnostics predicted from clinical course and pathognomy of the disease, which manifests more remarkable signs in the lower extremity. Electromyographic findings in this group revealed a neuro-

genic atrophy pattern with reduced interference voltage, high amplitude NMU voltage, involving low amplitude NMU voltage to suggest degeneration of muscle fibers in the atrophied muscles. Conduction velocities in the upper extremity had slight reduction in motor nerve conduction velocity of a few nerves, but in the lower extremity the reduction was more remaarkable. No response in measurement of motor nerve conduction velocity was likely to suggest degeneration of the peripheral nerve and muscle, giving some suggestions of condition and pathognomy of the disease when having utilized a recording method of muscle action potentials with a needle electrode applied by Dyck *et al.*[16] (1963), who observed reduced motor nerve conduction velocity in family member of Chacot-Marie-Tooth disease without any clinical sign and symptom. Conduction velocity of evoked potentials had almost the same tendency as that of motor nerve conduction velocity in the extremity. And strength-duration curve revealed abnormal pattern in about one-half of the examined hand muscles and in high percentage of the foot muscles in eight patients. The findings of these electrodiagnostics show no clear relationship to the clinical signs but are considerably helpful to obtain an information of course and situation of the disease. In diagnosis of myeloradiculoneuritis, determination of the lesion necessitates electrodiagnostics which could be recognized validly as a diagnostic tool. Electromyographic findings of the disease showed a neurogenic atrophy pattern with reduced interference voltage, high amplitude NMU voltage and complex NMU voltage, which are considered to present patho-physiologic character of the lesion involved the lower motor neuron. In measurements of the conduction velocities in present series, reduction of motor nerve conduction velocity was more remarkable in tibial nerve but, on the other hand, the conduction velocity of evoked potentials revealed no response or reduction in about one-half of both nerves examined, while strength duration curve had abnormal pattern in about half extensor digitorum brevis muscles and flexor hallucis brevis muscles. These findings in myeloradiculoneuritis do not clarify and explain directly the clinical feature but procedure of the examination is helpful in diagnosis to decide the lesion at level below the spinal cord. This fact may suggest that even if a patient with such clinical symptom as mentioned previously has been diagnosed as myelitis, he has the lesion of not only the spinal cord but also the peripheral nerve, and the diagnosis of myeloradiculoneuritis becomes adequate.

In diagnosis of lower motor neuron diseases, electromyography has developed as one of valuable diagnostics and even recently such evaluation of this procedure has been emphasized (Buchthal[17] 1962, Hamphrey *et al.*[18] 1962). The electromyographic findings can tell the pathological change of the lower motor neuron, if careful recording and estimation are performed. Measurement of motor nerve conduction velocity can show the disordered site along the peripheral nerve including sometimes intramuscular fiber by means of a

numerical expression as the conduction velocity of the fastest motor nerve. Combination of two techniques, elctromyography and motor nerve conduction velocity, becomes more effective to clarify the lesion in the peripheral motor nerve. Also, determination of residual latency (propagation time from end-plate to muscle fiber through intramuscular fiber) may indicate even the lesion beneath the end-plate and offer a basis to diagnosis of the distal lesion of the peripheral nerve. Of both techniques electromyography is probably more sensitive to uncover the peripheral lesion, even if one considers some problems on determination of the extent of the lesion and subjective expression of the examiner. The measurement of motor nerve conduction velocity is a method characteristic of showing the lesion in objective fashion. Conduction velocity of evoked potentials does not reveal the velocity of sensory nerve directly but compound velocity of that and motor nerve in part, while evoked potentials are smaller in amplitude than 50 microvolts and more difficult in measurement than other techniques. However, in the present series it was suggested that evoked potentials may appear mainly from the sensory nerve and at times showed distinctly an attitude of the sensory nerve in the peripheral nerve lesion. Thus, in a case with sensory disturbance this technique is available to decide the lesion as well as measurement of motor nerve conduction velocity, if the technique will be expanded to measurement of sensory nerve conduction velocity at distal portion of the peripheral nerve.

Strength-duration curve shows electrical excitability of the peripheral nerve in distal portion around the end-plate and muscle itself. Accordingly, in the present series the examination was applied to all of four diseases, especially more useful in neural progressive muscular atrophy and myelo-radiculoneuritis involved the peripheral nerve, and sometimes in progressive muscular atrophy the curve revealed rather the reduced excitability of the muscle fiber than of the peripheral nerve. Consequently, each of electro-diagnostics employed here for clinical diagnosis has different characteristics and limitation of application to the diagnosis of diseases, respectively. Of them, electromyography has a broad application to lower motor neuron diseases for revealing electrical evidence due to the lesion, but does not show an objectively quantitative expression of the lesion in parallel to clinical signs. The measurements of motor nerve conduction velocity and evoked potentials give objectively an available datum to the diagnosis in the peripheral nerve lesion which neural progressive muscular atrophy and myeloradiculoneuritis in this series have, but on the contrary, amyotrophic lateral sclerosis, spinal progressive muscular atrophy and juvenile unilateral muscular atrophy presumably involved the lesion in the anterior horn cell show little change in the conduction velocities. Strength-duration curve is more available in the former group than in the latter group with little lesion of the peripheral nerve.

SUMMARY

Measurements of motor nerve conduction velocity and evoked potentials in fifteen of normal subjects were performed between the elbow and wrist in ulnar and median nerves and between the knee and ankle in peroneal and tibial nerves. Values of the conduction velocities measured have well corresponded to those by many investigators and standard value of pathological reduction in the conduction velocities was discussed.

Electromyography, motor nerve conduction velocity, evoked potentials and strength-duration curve were employed to the patients with spinal progressive muscular atrophy, juvenile unilateral muscular atrophy, neural progressive muscular atrophy (Charcot-Marie-Tooth disease) and myeloradiculoneuritis. In the preceding two diseases the conduction velocities were almost within normal range, while electromyography showed the findings predicted the lesion of the anterior horn cell and strength-duration curve sometimes revealed lowered excitability of the muscle fiber. In the latter two diseases all of four procedures showed the findings to be predicted in pathognomy of the diseases. These findings are helpful to reach a correct diagnosis including differential diagnosis, but in consideration of easiness and characteristics with the electrodiagnostics electromyography should be performed at first in subsidiary examination of neurology, and then the others.

REFERENCES

1) Hodes, R., Larrabee, M. G. and German, W., The human electromyogram in response to nerve stimulation and the conduction velocity of motor axons, *Arch. Neurol. Psychiat.*, **60**, 340, 1948.
2) Trojaborg, W., Motor nerve conduction, electromyography and strength-duration curve, *Danish Med. Bull.*, **9**, 23, 1962.
3) Dawson, C. D. and Scott, J. W., The recording of nerve action potentials through the skin in man, *J. Neurol. Neurosurg. Psychiat.*, **12**, 259, 1949.
4) Hirayama, K., Toyokura, Y. and Tsubaki, T., Studies on motor neuron disease, *Psychiat. Neurol. Jap.*, **61**, 1861, 1959 (in Japanese).
5) Takagi, S. and Okabe, Y., Juvenile unilateral muscular atrophy, *Psychiat. Neurol. Jap.*, **61**, 2170, 1959 (in Japanese).
6) Thoma, P. K., Sears, T. A. and Gilliatt, R. W., The range of conduction velocity in normal motor fibres to the small muscles of the hand and foot, *J. Neurol. Neurosurg. Psychiat.*, **22**, 175, 1959.
7) Mayer, R. F., Nerve conduction studies in man, *Neurology*, **13**, 1021, 1963.
8) Blom, S., Hagbarth, K. E. and Lundberg, P. O., Motor conduction velocities in amyotrophic lateral sclerosis, polyradiculoneuritis and Charcot-Marie-Tooth's disease, *Acta Neurol. Scand.*, **40**, 6, 1964.
9) Yomura, Y. Measurement of motor nerve conduction velocity, *Naika*, **14**, 867, 1964 (in Japanese).
10) Henriksen, J. D., Conduction velocity of motor nerves in normal subjects and patients with neuromuscular disorders, Thesis. University of Minnesota, Minneapolis, 1956.
11) Gassel, M. M., Sources of error in motor nerve conduction studies, *Neurology*, **14**, 825,

1964.
12) Mavor, H. and Libman, I., Motor nerve conduction velocity measurement as a diagnostic tool, *Neurology*, **12**, 733, 1962.
13) Stensrud, P. A., Conduction velocity in human peripheral motor nerves, *Acta Neurol. Scand.*, **39**, 323, 1963.
14) Downie, A. W., Studies in nerve conduction. Chapter 22 in "Disorders of Voluntary Muscle" edited by J. N. Walter, pp 511-535, 1964.
15) Lambert, E. H. and Mulder, D. W., Electromyographic studies in amyotrophic lateral sclerosis, *Proc. Staff Meet. Mayo Clin.*, **32**, 441, 1957.
16) Dyck, P. J., Lambert, E. H. and Mulder, D. W., Charcot-Marie-Tooth disease: nerve conduction and clinical studies of a large kinship, *Neurology*, **13**, 1. 1963.
17) Buchthal, F., The electromyogram. Its value in the diagnosis of neuromuscular disorders, *World Neurol.*, **3**, 16, 1962.
18) Humphrey, J. G. and Shy, G. M.. Diagnostic electromyography, *Arch. Neurol.*, **6**, 339, 1962.