Analysis of Distal Compound Muscle Action Potential Duration in Hereditary Transthyretin Amyloidosis with Polyneuropathy

Yumi Hoshino[1], Minori Kodaira[1]*, Nobuhiko Ohashi[1]
Hiroshi Morita[2] and Yoshiki Sekijima[1)3]

1) *Department of Medicine (Neurology and Rheumatology), Shinshu University School of Medicine*
2) *Center for Health, Safety and Environmental Management, Shinshu University*
3) *Institute for Biomedical Sciences, Shinshu University*

Background : Hereditary transthyretin (ATTRv) amyloidosis, a disorder accompanied by axonal polyneuropathy, is often misdiagnosed as chronic inflammatory demyelinating polyneuropathy (CIDP). Prolongation of distally evoked compound muscle action potential duration (DCMAPD), an electrophysiological parameter of heterogeneous conduction delay at the distal part of the motor nerve suggesting demyelinating neuropathies, is included in an index of diagnostic criteria for CIDP. However, DCMAPDs are strongly influenced by low-frequency filtering (LFF) settings, which differ across hospitals worldwide.

Aim : To analyze DCMAPD in patients with ATTRv amyloidosis with polyneuropathy (ATTRv-PN).

Methods : DCMAPs of the median, ulnar, tibial, and peroneal nerves were recorded under LFF settings of 2, 10, and 20 Hz in 50 patients with ATTRv-PN. The changes of DCMAPD accompanied with the changes of LFF settings were analyzed. The appropriateness of the cut-off values of the DCMAPD in the latest criteria for CIDP, which defined under various LFF settings, was also validated in ATTRv-PN patients.

Results : The DCMAPD was shorter with increasing LFF settings. Less than 10 % of patients with ATTRv-PN demonstrated prolonged DCMAPD of the ulnar, tibial, and peroneal nerves. In contrast, ten patients demonstrated prolonged DCMAPD in the median nerve under LFF settings of 2, 10, and/or 20 Hz. Nine of the ten cases were complicated with carpal tunnel syndrome (CTS).

Conclusion : Prolongation of DCMAPDs in the ulnar, tibial, and peroneal nerves is rare in ATTRv-PN patients. DCMAPD analysis of the median nerve in patients with ATTRv-PN requires caution, because they frequently develop CTS and those with CTS may demonstrate prolonged DCMAPD. *Shinshu Med J 72 : 87—94, 2024*

(Received for publication October 6, 2023 ; accepted in revised form December 20, 2023)

Key words : ATTRv amyloidosis, CIDP, low-frequency filtering, CMAP duration, carpal tunnel syndrome

I Introduction

Hereditary transthyretin (ATTRv) amyloidosis is a life-threatening and gain-of-toxic-function disease characterized by the extracellular deposition of amyloid fibrils composed of the transthyretin (TTR)[1]. Most patients with ATTRv amyloidosis demonstrate progressive axonal polyneuropathy[2]. Liver transplantation had been the only life-saving treatment. However, TTR tetramer stabilizers and nucleic acid therapeutics have been developed and have shown greater treatment effects[3)-6)]. In these situations, early diagnosis and treatment initiation are becoming more important.

However, early diagnosis of ATTRv amyloidosis may be difficult. Notably, several patients with ATTRv amyloidosis with polyneuropathy (ATTRv-PN) from non-endemic areas without a family history are misdiagnosed with other neuropathies, especially chronic inflammatory demyelinating polyneuropathy (CIDP)[7)8)].

* Corresponding author : Minori Kodaira
Department of Medicine (Neurology and Rheumatology)
Shinshu University School of Medicine, 3-1-1 Asahi,
Matsumoto, Nagano 390-8621, Japan
E-mail : mkodaira@shinshu-u.ac.jp

A nerve conduction study (NCS) is a useful electrophysiological tool for identifying the underlying pathophysiology in patients with neuropathy. However, patients with ATTRv-PN occasionally exhibit conduction delay, fulfilling the electrodiagnostic criteria of CIDP without a conduction block under severe axonal degeneration[9)10)].

Prolongation of distally evoked compound muscle action potential duration (DCMAPD), an electrophysiological parameter of heterogeneous conduction delay at the distal part of the motor nerve suggesting demyelinating neuropathies, is included in an index of criteria for CIDP[11)-13)]. The cut-off value of DCMAPD in the European Federation of Neurological Societies/Peripheral Nerve Society (EFNS/PNS) 2010 diagnostic criteria for CIDP had been based on results obtained under only a low-frequency filtering (LFF) of 20 Hz[11)12)]. However, the DCMAPD recorded under lower LFF settings is more prolonged[14)15)]. Furthermore, LFF settings differ across hospitals worldwide. Accordingly, Mitsuma et al. proposed cut-off values of DCMAPD under LFF settings of 2, 5, 10, and 20 Hz to diagnose CIDP[15)]. They defined these cut-off values based on the results of NCSs in patients with typical CIDP and diabetic polyneuropathy, and healthy subjects. Recently, these cut-off values proposed by Mitsuma et al. have been recommended for the analysis of DCMAPD by the latest CIDP criteria, the revised European Academy of Neurology/Peripheral Nerve Society (EAN/PNS) 2021 guideline for CIDP[13)15)]. Validation of these cut-off values under each LFF setting in diseases occasionally misdiagnosed as CIDP is needed. We analyzed the effects of LFF settings on DCMAPD and validated whether the cut-off values of DCMAPD under each LFF setting are appropriate in patients with ATTRv-PN.

Ⅱ Methods

A Participants

Patients with disease-causing variants of *TTR* genes and TTR-derived amyloid deposition in biopsy specimens were included in this study. All patients showed symptoms of polyneuropathy, and those with other factors affecting peripheral nerve function, such as diabetes mellitus and alcoholism, were excluded. The clinical stage of patients with ATTRv amyloidosis was defined using the criteria of Countinho et al. criteria[16)] : Stage 0 : patients have some autonomic dysfunction without polyneuropathic symptoms or signs ; Stage I : polyneuropathy is localized to the lower limbs, and patients can walk without assistance ; Stage II : polyneuropathy involving the upper and lower limbs, and patients are disabled but can still walk with assistance ; Stage III : severe polyneuropathy and autonomic dysfunction confining patients to a wheelchair or bed.

B Electrophysiological examinations

All NCSs were performed using a MEB2312 (Nihon-Koden, Tokyo, Japan). Motor NCSs were performed using the belly-tendon method on the one-sided median, ulnar, tibial, and peroneal nerves[17)]. The distance from the cathode of the recording electrode to the stimulating electrode cathode for measuring distal latency was 7, 7, 10, and 8.5 cm for the median, ulnar, tibial, and peroneal nerves, respectively. CMAP amplitude, distal latency (DL), motor nerve conduction velocity (MCV), F-wave latency, and DCMAPD were measured in motor NCSs. CMAP amplitude was defined as the amplitude between the baseline and negative peak. DCMAPD was defined as the period from the onset of the initial negative phase to the return to the baseline of the last negative deflection of the CMAP at a sensitivity of 500 μV/div. High-frequency filtering (HFF) was set and fixed at 10 kHz throughout the motor NCSs. LFF was set at 2 Hz. The LFF setting was changed from 2 Hz to 10 Hz and 20 Hz only when the DCMAPDs were recorded to evaluate the effects of the LFF settings on the DCMAPD. A hum filter was not used. Sensory NCSs were conducted on the median, ulnar, and sural nerves using an antidromic method[17)]. In the median and ulnar nerves, the cathode of the stimulating electrode was placed on the wrist in the same position as the motor NCSs of each nerve. In the sural nerve, the stimulating electrode cathode was placed 14 cm proximal to the cathode of the recording electrode. Amplitude of sensory nerve action potential (SNAP) and sensory nerve conduction velocity (SCV) were measured in sensory NCSs. The SNAP amplitude was defined as the

amplitude between the initial positive and negative peaks. The HFF and LFF were fixed at 3 kHz and 20 Hz, respectively, during sensory NCSs. The skin temperature of the examined segment was ≥ 30 ℃ throughout the electrophysiological examination.

C Analyses

The effects of LFF settings on DCMAPD were evaluated using the Jonckheere–Terpstra test, which is consistent with previous studies[15]. The DCMAPD was analyzed according to the EAN/PNS 2021 criteria for CIDP. Correlations between the DCMAPD recorded under an LFF setting of 2 Hz and other electrophysiological parameters, including CMAP amplitude, DL, MCV, F-wave latency, SNAP, and SCV, corresponding to each nerve, were analyzed using Spearman's rank correlation coefficient. Correlations between DCMAPD in the lower limb nerves and sensory nerve function were analyzed using the results of the sensory NCS in the sural nerve. When a group with abnormally prolonged DCMAPD was detected, subgroup analyses between patients with and without prolonged DCMAPD were performed using the Mann–Whitney U test or Fisher's exact test. Statistical significance was set at $p < 0.05$. Statistical analysis was performed using the Bell Curve for Excel (Social Survey Research Information Co., Ltd., Japan).

D Protocol approval and ethical concerns

The Shinshu University Certified Review Board of Clinical Research approved this prospective study (approval number : 4244) and written informed consent was obtained from each patient.

Ⅲ Results

A Background of patients with ATTRv-PN

Fifty patients with ATTRv-PN were included in this study. The clinical profiles of patients are shown in **Table 1**. Notably, 86 % of the patients had a family history of ATTRv amyloidosis, and 36 % were from an endemic area of ATTRv in Nagano Prefecture. Six patients from non-endemic areas had no apparent family history of ATTRv amyloidosis. Thirty-five patients had the *TTR* variant, Val30Met (p.Val50Met). Fifteen patients had non-Val30Met variants [two Ile107Val (p. Ile127Val), two Asp38Ala (p. Asp58Ala), two Ser50Ile (Ser70Ile), two Phe44Ser (p. Phe64Ser), one Ser50Arg (p. Ser70Arg), one Ser90Arg (p. Ser110Arg), one Thr60Ala (p. Thr80Ala), one Ala36Pro (p. Ala56Pro), one Glu42Gly (p. Glu62Gly), one Tyr114His (p. Tyr134His), and one Phe53Val (p. Phe73Val)]. Forty-five patients received disease-modifying therapies, such as liver transplantation, TTR tetramer stabilizers (tafamidis), and/or ribonucleic acid interference therapy (patisiran). The clinical stage of most patients was either stage I or II. Seventeen cases were complicated with carpal tunnel syndrome (CTS).

Table 1 Patients' clinical background

Age (y)	52.4 ± 14.5
Female/Male	16/34
Age at onset of neuropathy (y)	42.8 ± 15.3
Duration from onset of neuropathy (y)	10.4 ± 6.3
Duration from start of treatments (y)	6.7 ± 5.8
Presence of family history (%)	86
Patients from non-endemic area (%)	64
Clinical Stage Ⅰ (%)	42
Clinical Stage Ⅱ (%)	52
Clinical Stage Ⅲ (%)	6
Complication of carpal tunnel syndrome	17

Data are presented as mean ± SD. y : years
Treatments include liver transplantation, transthyretin tetramer stabilizers, or oligonucleotide therapeutics.

Table 2 The DCMAPDs of the median, ulnar, tibial, and peroneal nerves under each LFF setting

		LFF settings		
		2 Hz	10 Hz	20 Hz
Median	(ms)	6.9 (1.5)	6.4 (1.4)	5.8 (1.2)
Ulnar	(ms)	6.8 (1.2)	6.6 (1.3)	6.0 (1.1)
Tibial	(ms)	6.1 (1.9)	5.8 (1.3)	5.6 (1.4)
Peroneal	(ms)	6.9 (1.7)	6.6 (1.6)	6.3 (1.8)

Data are presented as mean (SD).
DCMAPD, distally evoked compound muscle action potential duration ; LFF, low-frequency filtering.

B Effects of LFF settings on DCMAPD in ATTRv-PN patients

CMAPs were not detected in the median, ulnar, tibial, and peroneal nerves in three, one, 11, and 16 patients, respectively. Therefore, the effects of LFF settings on the DCMAPD of the median, ulnar, tibial, and peroneal nerves were analyzed in 47, 49, 39, and 34 patients, respectively. The effects of the LFF settings on the DCMAPD are summarized in **Table 2**.

Table 3 Results of electrophysiological examinations.

Nerve	CMAP (mV)	DL (ms)	MCV (m/s)	F-wave latency (ms)	SNAP (μV)	SCV (m/s)
Median	7.3 (4.8)	4.1 (1.2)	50.8 (8.8)	29.1 (5.1)	10.8 (11.2)	53.7 (10.3)
Ulnar	6.4 (3.9)	3.1 (0.9)	55.7 (9.9)	28.9 (5.3)	10.2 (10.5)	51.5 (9.6)
Tibial	6.1 (6.3)	4.1 (0.9)	44.4 (7.1)	49.2 (5.8)		
Peroneal	1.5 (1.7)	4.5 (0.9)	43.0 (5.9)	50.7 (4.1)		
Sural					5.3 (6.5)	48.1 (5.6)

Data are presented as mean (SD).
CMAP, compound muscle action potential ; DL, distal latency ; MCV, motor nerve conduction velocity ; SCV, sensory nerve conduction velocity ; SNAP, sensory nerve action potential

Table 4 The proportion of patients showing prolonged DCMAPD according to the EAN/PNS 2021 criteria

Nerve	Number of patients	Number of patients showing prolongation of DCMAPD		
		2 Hz	10 Hz	20 Hz
Median	31 without CTS	0 (0 %)	1 (3.0 %)	0 (0 %)
Median	17 with CTS	8 (47.1 %)	7 (41.2 %)	6 (35.3 %)
Ulnar	49	2 (4.1 %)	2 (4.1 %)	2 (4.1 %)
Tibial	39	1 (2.6 %)	1 (2.6 %)	1 (2.6 %)
Peroneal	34	3 (8.8 %)	4 (11.8 %)	3 (8.8 %)

＜Cut-off value of DCMAPD[13]＞
Median ＞ 8.4 ms, Ulnar ＞ 9.6 ms, Tibial ＞ 9.2 ms, and Peroneal ＞ 8.8 ms under an LFF setting of 2 Hz.
Median ＞ 7.8 ms, Ulnar ＞ 8.5 ms, Tibial ＞ 8.2 ms, and Peroneal ＞ 8.3 ms under an LFF setting of 10 Hz.
Median ＞ 7.4 ms, Ulnar ＞ 7.8 ms, tibial ＞ 8.0 ms, and Peroneal ＞ 8.1 ms under an LFF setting of 20 Hz.
CTS, carpal tunnel syndrome ; DCMAPD, distally evoked compound muscle action potential duration ; LFF, low-frequency filtering.

As the LFF setting increased, the DCMAPD of the median, ulnar, and peroneal nerves became shorter ($p<0.001$, $p<0.001$, $p<0.05$, respectively). The effect of LFF on the DCMAPD of the tibial nerve was insignificant ($p=0.08$). However, DCMAPD tended to be shorter under higher LFF settings.

C Relationship between DCMAPD and other electrophysiological parameters

The results of the other electrophysiological examinations are summarized in **Table 3**. The SCV was negatively correlated with DCMAPD in the median nerve ($rs=-0.35$, $p<0.05$). MCV was also negatively correlated with DCMAPD in the ulnar nerve ($rs=-0.32$, $p<0.05$). No other parameters correlated with DCMAPD in any of the nerves.

D Prolonged DCMAPD in ATTRv-PN using CIDP electrodiagnostic criteria

Table 4 shows the number of patients with prolonged DCMAPD, according to the EAN/PNS 2021 diagnostic criteria for CIDP. Only a few patients (< 10 % of patients, except for those with DCMAPD of the peroneal nerve under the LFF setting of 10 Hz) showed prolonged DCMAPD of the ulnar, tibial, and peroneal nerves, fulfilling the EAN/PNS 2021 criteria **(Table 4)**. On the other hand, 10 patients showed prolonged DCMAPD of the median nerve under LFF settings of 2 Hz, 10 Hz, and/or 20 Hz. The clinical stages were Ⅰ and Ⅱ in two and eight patients, respectively. The ten patients with prolonged DCMAPD of the median nerve had slower MCV ($p<0.01$) and SCV ($p<0.05$) in the median nerve than those without prolonged DCMAPD of the median nerve. They also tended to show a longer DL ($p=0.07$) and smaller SNAP ($p=0.09$) in the median nerve than the latter. Patients with prolonged DCMAPD of the median nerve also tended to have longer DCMAPD of the ulnar nerve under an LFF setting of 2 Hz ($p<0.05$) and DCMAPD of the peroneal nerve under LFF settings of 2, 10, and 20 Hz ($p<0.05$, $p<0.005$, $p<0.05$, respectively). However, other electrophysiological

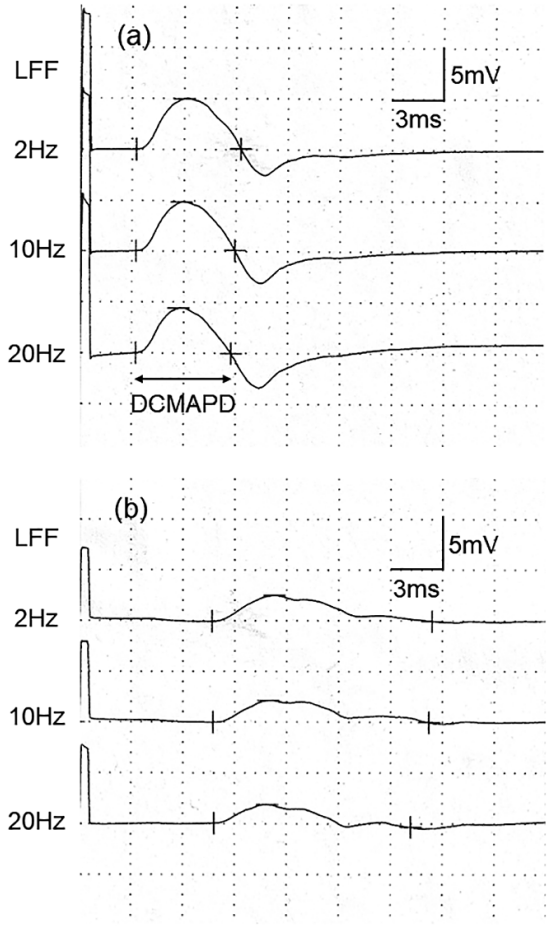

Fig. 1 Changes of DCMAPD in the median nerve following stimulation at the wrist under LFF settings of 2, 10, 20Hz

(a) CMAPs without prolongation of DCMAPD in patients with ATTRv-PN without CTS. (b) CMAPs with prolongation of DCMAPD in patients with ATTRv-PN with CTS. ATTRv-PN, hereditary transthyretin amyloidosis with polyneuropathy ; CMAP, compound muscle action potential ; CTS, carpal tunnel syndrome ; DCMAPD, distally evoked compound muscle action potential duration ; LFF, low-frequency filtering.

parameters of the ulnar, tibial, peroneal, and sural nerves did not differ between patients with and without prolonged DCMAPD of the median nerve. Of the 10 patients with prolonged DCMAPD of the median nerve, two had a surgical history of CTS and seven showed compatible clinical symptoms and signs of CTS. In contrast, eight of the remaining 37 patients without prolonged DCMAPD had a surgical history of CTS (n＝3) or clinical symptoms and signs of CTS (n＝5). Compared to ATTRv-PN patients without CTS, those with CTS tended to develop prolonged median nerve DCMAPD ($p<0.001$). **Fig. 1**. shows representative CMAP waveforms and changes of DCAMPD according to LFF settings in the median nerve from ATTRv-PN patients without and with CTS.

Ⅳ Discussion

This study confirmed the significant effects of LFF on DCMAPD in patients with ATTRv-PN. DCMAPD in patients with ATTRv-PN became shorter as the LFF setting increased. DCMAPD was determined to be within normal limits in most patients with ATTRv-PN according to the EAN/PNS 2021 criteria, which defines the cut-off values of DCMAPD under each LFF setting. However, more than one third of ATTRv-PN patients with CTS show prolonged DCMAPD of the median nerve, even when the cut-off values of the EAN/PNS 2021 criteria were used.

A Effects of LFF on DCMAPD in ATTRv-PN patients

The effects of LFF settings on DCMAPD in patients with ATTRv-PN were consistent with the results of previous studies in healthy participants and patients with diabetic neuropathy or CIDP[14)15)]. DCMAPDs of the median, ulnar, and peroneal nerves became shorter as the LFF setting increased. A similar tendency was observed in the tibial nerve. The effects of the LFF setting on tibial nerve DCMAPD were not statistically significant, which may be due to the difficulty in pre-

cisely measuring DCMAPD in small CMAPs because of contamination from potential spreads of other muscles[18] and electrical stimulating shock artifacts following high-intensity electrical stimulation to detect CMAPs under severe axonal degeneration.

DCMAPD did not correlate with other electrophysiological parameters of each nerve ; however, a weak negative correlation was observed between DCMAPD and SCV in the median nerve and between DCMAPD and MCV in the ulnar nerve. These results show that DCMAPD in patients with ATTRv-PN is less involved in the severity of damage in each nerve ; however, it is strongly affected by the LFF settings.

B Analysis of DCMAPD in ATTRv-PN using the latest criteria for CIDP

Patients with ATTRv-PN ideally should not show prolongation of DCMAPD, because primary pathophysiology of them is axonal damage. In our retrospective study using NCSs in the ulnar and tibial nerves[9], we observed that 45 % of the patients with ATTRv-PN demonstrated prolonged DCMAPD in the ulnar nerve recorded under an LFF setting of 2 Hz compared with the cut-off value of DCMAPD (≥6.7 ms in the ulnar nerve) of the EFNS/PNS 2010 diagnostic criteria for CIDP based on results of DCMAPD obtained under a LFF setting of 20Hz. Contrarily, we confirmed that only 2 % of the patients with ATTRv showed prolongation of DCMAPD in the ulnar nerve, fulfilling the cut-off value (>9.6 ms) under the LFF setting of 2 Hz proposed by Mitsuma et al.[15] in the study. However, the effects of LFF settings, except for 2 Hz, on DCMAPDs in the ulnar and tibial nerves and DCMAPDs in the median and peroneal nerves have not been investigated in patients with ATTRv-PN.

The results of DCMAPD in the ulnar, peroneal, and tibial nerves in the present study confirmed that the use of the cut-off values of DCMAPD under each LFF setting indicated in the revised EAN/PNS 2021 criteria for CIDP minimizes the misinterpretation of prolonged DCMAPD in patients with ATTRv-PN. Similarly, DCMAPD of the median nerve in ATTRv-PN without CTS did not prolong.

In contrast, this study found that ATTRv-PN patients with CTS tended to show prolonged DCMAPD of the median nerve, even when analyzed using the cut-off values of the EAN/PNS 2021 criteria. Patients with prolonged DCMAPD of the median nerve also tended to have longer DCMAPD of the ulnar and peroneal nerves ; however, other parameters of neurophysiological examinations, except for those of the median nerve, did not show any differences between patients with and without prolongation of DCMAPD in the median nerve. Furthermore, most of ATTRv-PN patients did not develop prolongation of DCMAPD in the ulnar, tibial, and peroneal nerves. These results also suggest that CTS may have a big influence on prolongation of DCMAPD in the median nerve of patients with ATTRv-PN. Mitsuma et al. reported that less than 5 % of patients with diabetic polyneuropathy without CTS showed prolongation of DCMAPD surpassing the cut-off values of the revised EAN/PNS 2021 criteria[15]. At present, the effects of CTS with or without axonal polyneuropathy on DCMAPD have not been established. Although pathological analysis of CTS in humans is scarce[19], focal demyelination and remyelination have been observed in a rat model of CTS using chronic nerve compression to the sciatic nerve[20]. In the experimental study using rats, heterogeneous demyelination and remyelination (i.e., region was located at the outer half of the nerve with normal finding at the center half) was also observed at early stage of chronic compression[20]. Investigation of DCMAPD in patients with CTS alone using the cut-off values of the revised EAN/PNS 2021 criteria is needed ; however, results of these pathological studies support a notion that CTS may induce prolongation of DCMAPD in the median nerve. Analysis of DCMAPD in the median nerve of patients with ATTRv-PN requires caution, because they frequently develop CTS[21].

C Limitations

Of the study participants, 36 % of the patients were from an endemic area for ATTRv amyloidosis, and they all had a family history of ATTRv amyloidosis. Furthermore, 86 % of participants, including patients from non-endemic areas, had family history of ATTRv amyloidosis. These patients are less likely to be misdiagnosed as having CIDP in clinical practice.

V Conclusion

Prolongation of DCMAPDs in the ulnar, tibial, and peroneal nerves is rare in patients with ATTRv-PN by using the cut-off values of DCMAPD under each LFF setting. Analysis of DCMAPD of the median nerve in patients with ATTRv-PN requires caution, because they frequently develop CTS and those with CTS may demonstrate prolongation of DCMAPD suggestive of demyelinating neuropathies.

Conflict of interest

The authors declare that they have no conflicts of interest.

Funding

None.

References

1) Sekijima Y : Transthyretin (ATTR) amyloidosis : clinical spectrum, molecular pathogenesis and disease-modifying treatments. J Neurol Neurosurg Psychiatry 86 : 1036-1043, 2015
2) Ikeda S, Nakazato M, Ando Y, Sobue G : Familial transthyretin-type amyloid polyneuropathy in Japan : clinical and genetic heterogeneity. Neurology 58 : 1001-1007, 2002
3) Coelho T, Maia LF, Martins da Silva A, et al : Tafamidis for transthyretin familial amyloid polyneuropathy : a randomized, controlled trial. Neurology 79 : 785-792, 2012
4) Berk JL, Suhr OB, Obici L, et al : Repurposing diflunisal for familial amyloid polyneuropathy : a randomized clinical trial. JAMA 310 : 2658-2667, 2013
5) Benson MD, Waddington-Cruz M, Berk JL, et al : Inotersen treatment for patients with hereditary transthyretin amyloidosis. N Engl J Med 379 : 22-31, 2018
6) Adams D, Gonzalez-Duarte A, O'Riordan WD, et al : Patisiran, an RNAi therapeutic, for hereditary transthyretin amyloidosis. N Engl J Med 379 : 11-21, 2018
7) Planté-Bordeneuve V, Ferreira A, Lalu T, et al : Diagnostic pitfalls in sporadic transthyretin familial amyloid polyneuropathy (TTR-FAP). Neurology 69 : 693-698, 2007
8) Koike H, Hashimoto R, Tomita M, et al : Diagnosis of sporadic transthyretin Val30Met familial amyloid polyneuropathy : a practical analysis. Amyloid 18 : 53-62, 2011
9) Ohashi N, Kodaira M, Morita H, Sekijima Y : Electrophysiological demyelinating features in hereditary ATTR amyloidosis. Amyloid 26 : 15-23, 2019
10) Lozeron P, Mariani LL, Dodet P, et al : Transthyretin amyloid polyneuropathies mimicking a demyelinating polyneuropathy. Neurology 91 : e143-e152, 2018
11) Isose S, Kuwabara S, Kokubun N, et al : Utility of the distal compound muscle action potential duration for diagnosis of demyelinating neuropathies. J Peripher Nerv Syst 14 : 151-158, 2009
12) Joint Task Force of the EFNS and the PNS : European Federation of Neurological Societies/Peripheral Nerve Society Guideline on management of paraproteinemic demyelinating neuropathies. Report of a Joint Task Force of the European Federation of Neurological Societies and the Peripheral Nerve Society-first revision. J Peripher Nerv Syst 15 : 1-9, 2010
13) Van den Bergh PYK, van Doorn PA, Hadden RDM, et al : European Academy of Neurology/Peripheral Nerve Society guideline on diagnosis and treatment of chronic inflammatory demyelinating polyradiculoneuropathy : Report of a Joint Task Force-second revision. Eur J Neurol 28 : 3556-3583, 2021
14) Rajabally YA, Lagarde J, Cassereau J, Viala K, Fournier E, Nicolas G : A European multicentre reappraisal of distal compound muscle action potential duration in chronic inflammatory demyelinating polyneuropathy. Eur J Neurol 19 : 638-642, 2012
15) Mitsuma S, Van den Bergh P, Rajabally YA, et al : Effects of low frequency filtering on distal compound muscle

action potential duration for diagnosis of CIDP : a Japanese–European multicenter prospective study. Clin Neurophysiol 126 : 1805–1810, 2015

16) Coutinho P, da Silva AM, Lima JL, et al : Forty years of experience with type 1 amyloid neuropathy : review of 483 cases. In : Glenner GG, e Costa PP, de Freitas AF (eds), Amyloid and Amyloidosis, pp 88–98, Excerpta Medica, Amsterdam, 1980

17) Kimura J : Electrodiagnosis in diseases of nerve and muscle principles and practice. 4th ed, pp 74–179. Oxford University Press, New York, 2013

18) Sonoo M : Far-field potentials in the compound muscle action potential. Muscle Nerve 61 : 271–279, 2020

19) Thomas P, Fullerton P : Nerve fibre size in the carpal tunnel syndrome. J Neurol Neurosurg Psychiatry 26 : 520–527, 1963

20) Gupta R, Rowshan K, Chao T, Mozaffar T, Steward O : Chronic nerve compression induces local demyelination and remyelination in a rat model of carpal tunnel syndrome. Exp Neurol 187 : 500–508, 2004

21) Tojo K, Tsuchiya-Suzuki A, Sekijima Y, Morita H, Sumita N, Ikeda S : Upper limb neuropathy such as carpal tunnel syndrome as an initial manifestation of ATTR Val30Met familial amyloid polyneuropathy. Amyloid 17 : 32–35, 2010

（2023. 10. 6 received ; 2023. 12. 20 accepted）