Multifocal motor neuropathy: long-term clinical and electrophysiological features of the disease

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Little attention has been paid abroad to the problem of the long-term course of multifocal motor neuropathy (MMN). In our country, catamnestic studies of MMN have not been conducted at all. However, the results of such an analysis are extremely important for understanding the course and prognosis of the disease.

Objective: *to analyse the clinical and neurophysiological data of patients with MMN with a disease duration of more than 5 years.*

Material and methods. *The study included 28 patients with MMN: 9 women (32%) and 19 men (68%); the median age at admission was 50 [44; 56] years; the median disease duration was 10 [8; 13] years. Medical documentation, medical history, complaints, neurological examination results (scored on the MRC and INCAT scales) and results of electroneuromyography (ENMG) of the long nerves of the hands were analysed.*

Results. *The median time between onset of the disease and diagnosis was 5.5 [2; 10] years. Paresis <3 points on the MRC scale was found in the extensor muscles of the hand and fingers (12/28; 43%), in the median (15/28; 53%) and ulnar (20/28; 71%) muscle groups of the hands, in the extensors (11/28; 39%) and flexors (9/28; 32%) of the feet. The median total score for the degree of disability on the INCAT scale was 3 [2; 3] for the hands and 1 [0; 2] for the legs. The comparative analysis of the severity of the neurological deficits on the MRC and INCAT scales at the onset of the disease and in the long-term catamnesis revealed no significant differences (p>0.05). An objective assessment of sensory disorders revealed no changes when testing tactile, pain and temperature sensitivity, while half of the cases (14/28; 50%) showed a disturbance of vibration sensitivity in the lower extremities. The ENMG examination was consistent with the electrophysiological criteria of the disease, one third of the patients showed significant secondary damage to the axons of the motor fibers of the hand nerves, and in half of the cases a slight impairment of the axons of the sensory fibers was registered.*

Conclusion. *MMN is a curable disease. Unfortunately, our retrospective analysis showed that in the Russian Federation there are problems with its diagnosis and quality care of this category of patients. Late diagnosis, delayed start of treatment and non-compliance with the schedule of pathogenetic therapy lead to persistent disability of patients.*

Keywords: *multifocal motor neuropathy; long-term catamnesis; retrospective analysis; electroneuromyography.*
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Multifocal motor neuropathy (MMN) is a rare chronic dysimmune neuropathy characterized by autoimmune attacks against the nodal zones of motor nerve fibers, resulting in conduction blocks (CB) and, as a consequence, muscle weakness [1]. Different authors estimate the prevalence of MMN to be one to two cases per 100,000 population per year, with a higher incidence among males, particularly those in the active age group. The primary clinical manifestation typically involves a slowly progressive, asymmetrical, upper (predominantly distal) flaccid paraparesis, less often tri- or tetraparesis. Sensory and conduction disorders, pyramidal symptoms are not typical [2]. In 2010, the European Federation of Neurological Societies/Peripheral Nerve Society (EFNS/PNS) formulated diagnostic criteria for MMN, which are still used today [3]. The only effective method of pathogenetic therapy is high-dose intravenous immunoglobulin (IVIg) [4].

Very little attention has been paid to the problem of the long-term course of MMN, with few studies published abroad dedicated to its analysis. These studies mainly assess the efficacy of various dosing regimens of intravenous or subcutaneous immunoglobulin, and most of them include a small number of patients. In our country, prospective cohort studies of MMN have not been carried out at all. Nevertheless, the results of such an analysis are crucial for understanding the course of the disease, its prognosis and impact on the patientXs quality of life, as well as the need for maintenance therapy in order to plan health care costs for the treatment of this patient population.

The study **goal** is to analyze the clinical and neurophysiological data of patients with MMN with a disease duration of more than 5 years.

Material and methods. The register of the Center for Diseases of the Peripheral Nervous System of the Scientific Center of Neurology (FSBRI SCN) includes records of 69 patients diagnosed with MMN. For this study, we selected 28 individuals (40%) with a disease duration of more than 5 years: 9 women (32%) and 19 men (68%); the median age at the time of inclusion in the study was 50 [44; 56] years; median disease duration was 10 [8; 13] years. A mixed retrospective and prospective cohort study was conducted.

Inclusion criteria:
– age over 18 years;
– compliance with diagnostic criteria for MMN [3];
– signing an informed consent to participate in the study.
Exclusion criteria:
– age under 18 years;
– the presence of poorly controlled concomitant somatic diseases.

For all patients included in the study, we analyzed medical record, history data, concomitant disorders, duration and characteristics of the disease course, results of previous laboratory tests and instrumental investigations; complaints, results of physical and neurological examinations with assessment of motor, sensory functions, coordination and reflexes, including international scales – Medical Research Council Scale for Muscle Strength sum score (MR‘) [5] and Inflammatory Neuropathy Cause and Treatment (INCAT) [6].

All patients underwent nerve conduction studies (NCS) using a four-channel Keypoint Clinical System myograph (Medtronic, USA) according to the standard method (E. Stalberg et al. [7]) with a body surface temperature of at least 33 °C, which was controlled using a contactless electronic temperature sensor, and was measured before the investigation at the wrist joint level on both sides. Motor and sensory fibers of the median and ulnar nerves on both sides were assessed. Commonly accepted NCS parameters were analyzed. The values recommended by J. Kimura [8], with correction for the laboratory regulatory base of the FSBRI SCN, were used as reference.

Statistical data processing was carried out using Microsoft Office Excel 2016 and SPSS Statistics 23 software (IBM, USA). The main descriptive statistics used were percentage, mean and standard deviation, minimum and maximum, median and quartiles (Me [25th; 75th percentiles]). Qualitative data are described as frequencies and proportions.

The study was approved by the local ethics committee of the FSBRI "Scientific Center of Neurology" (protocol No. 10-4/21 of November 17, 2021).

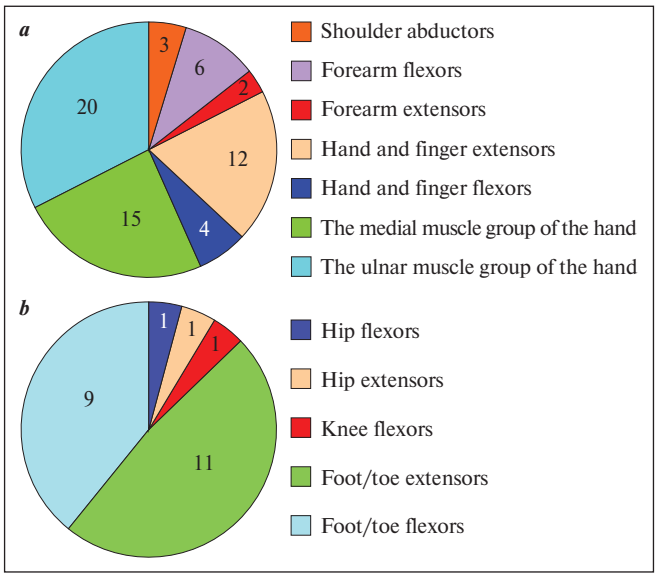
Table 1. *Severity of limb muscle paresis in patients with MMN at long-term follow-up, MRC scale, Me [25th; 75th percentile]*

Muscle group	on the right side	on the left side
Upper limbs:		
shoulder abductors	5 [5; 5]	5 [5; 5]
forearm flexors	5 [4; 5]	5 [4; 5]
forearm extensors	5 [4; 5]	5 [4.2; 5]
hand extensors	4 [4; 4.7]	4 [2.2; 5]
finger extensors	3 [2; 4]	3.5 [2; 5]
hand and finger flexors	5 [4; 5]	5 [4; 5]
the medial muscle group of the hand	3 [1.2; 4]	3 [2; 4.7]
the ulnar muscle group of the hand	3 [1.2; 4]	3 [2; 4.7]
Lower extremities:		
hip flexors	5 [5; 5]	5 [5; 5]
hip extensors	5 [5; 5]	5 [5; 5]
leg extensors	5 [5; 5]	5 [5; 5]
shin flexors	5 [5; 5]	5 [5; 5]
foot extensors	4 [2; 5]	4 [3; 5]
foot flexors	4 [3; 5]	4 [3; 5]

Results. ***Clinical and history characteristics.*** The median period from the onset of the disease to diagnosis in the patients included in the study was 5.5 [2; 10] years, and the delay in initiation of treatment was 6 [4; 10] years. The range of erroneous diagnosis included: carpal tunnel syndrome (12/28; 43%), chronic inflammatory demyelinating polyradiculoneuropathy (8/28; 28%), motor neuron disease (5/28; 18%), osteochondrosis (3/28; 11%)

At the time of inclusion in the study, 20 (71%) patients were receiving IVIg: the total duration of the therapy was 4 [1; 8] years, the interval between maintenance courses was 8 [5; 12] weeks. The median duration of the IVIg effect was within 4 [4; 6] weeks. Non-compliance with the intravenous immunoglobulin (IVIg) dosing regimen or non-adherence to an adequate dose was reported in every second patient; in every third case the efficacy of therapy was insufficient (after the administration of IVIg, the degree of paresis did not change or weakness regressed by no more than 1 point on the MRC scale).

Patients with a disease duration of more than 5 years persistently complained of asymmetric weakness in the proximal and distal arms (in 35 and 100% of cases, respectively), as well as in the proximal and distal legs (in 14 and 71% of cases, respectively). All patients complained of weight loss of paretic muscles, muscle twitching, and impaired fine motor skills. Every third patient (8/28; 28%) complained of impaired gait; the same num-



Number of patients with muscle paresis of the right upper (a) and lower (б) limbs (⩽3 points on the MRC scale)

Table 2. *Severity of neurological disorders in patients with MMN at the onset and long-term follow-up, total score, Me [25th; 75th percentile]*

Score	Assessment at the onset of the disease	Assessment during long-term follow-up	p
MRC	50 [46; 53]	53 [49; 56]	0.08
INCAT (hands)	3.3 [3; 4]	3 [2; 3]	0.1

ber of patients (8/28; 28%) complained of sensory disturbances in the limbs (numbness and paresthesia). None of the patients had pain.

The results of assessment of the main muscle groups of the limbs according to the MRC scale in the patients included in the study are presented in Table 1. There was greater involvement of the distal limb muscles with a slight predominance of weakness in the dominant arm (p>0.05).

In the MMN patients under long-term follow-up, the most common findings included paresis of ⩽3 points on the MRC scale in the extensors of the hand and fingers (12/28; 43%), in the medial (15/28; 53%) and ulnar (20/28; 71%) muscles of the hands, in extensors (11/28; 39%) and flexors (9/28; 32%) of the feet (Fig. 1). The median total score for the degree of dysfunction on the INCAT scale was 3 [2; 3] for the hands and 1 [0; 2] for the legs.

Table 3. *Analysis of the results of ENMG of the motor fibers of the peripheral nerves of the hands in patients with MMN during long-term follow-up*

The nerve	Nerve conduction studies variables	Parameters
n. medianus dex.	Number of patients with distal M-wave (dM-wave), n (%)	20 (71)
	dM-wave terminal latency, ms; Me [25th; 75th percentile] (N<3,5)	3.6 [3.4; 4]
	dM-wave amplitude, mV; Me [25th; 75th percentile] (N>5,0)	4.9 [1.4; 7.4]
	dM-wave duration, ms; Me [25th; 75th percentile]	6 [5.4; 6.4]
	CB at the level of the forearm, n (%); CB degree, %, Me [25th; 75th percentile]	11 (39) 48.7 [43; 72]
	CB at the level of the lower third of the upper arm, n (%); CB degree, %, Me [25th; 75th percentile]	2 (7) 60 [53; 65]
	CB in the proximal region (Erb's point), n (%); CB degree, %, Me [25th; 75th percentile]	5 (18) 75 [64; 89]
	Number of patients with M-response dispersion, n (%)	0
	CVm m/s; Me [25th; 75th percentile] (N>50) forearm elbow bend	52 [49; 54] 62 [50; 66]
n. ulnaris dex.	Number of patients with distal M-wave (dM-wave), n (%)	23 (82)
	dM-wave terminal latency, ms; Me [225th; 75th percentile] (N<3,0)	3 [2.6; 3.6]
	dM-wave amplitude, mV; Me [25th; 75th percentile] (N>6,0)	5 [1.3; 7]
	dM-wave duration, ms; Me [25th; 75th percentile]	5.8 [5.2; 6.3]
	CB at the level of the forearm, n (%); CB degree, %, Me [25th; 75th percentile]	13 (46) 50 [40; 63]
	CB at the level of the elbow joint, n (%)	0
	CB at the level of the shoulder, n (%)	0
	CB in the proximal region (Erb's point), n (%); CB degree, %, Me [25th; 75th percentile]	3 (11) 50 [50; 70]
	Number of patients with M-response dispersion, n (%)	0
	CVm m/s; Me [25th; 75th percentile]; (N>50) forearm elbow	56 [43; 65] 52 [41; 58]
n. medianus sin.	Number of patients with distal M-wave (dM-wave), n (%)	20 (71)
	dM-wave terminal latency, ms; Me [25th; 75th percentile] (N<3,5)	3.6 [3.3; 3.8]
	dM-wave amplitude, mV; Me [25th; 75th percentile] (N>5,0)	4.4 [1.4; 7.5]
	dM-wave duration, ms; Me [25th; 75th percentile]	6.2 [5.4; 6.9]
	CB at the level of the forearm, n (%); CB degree, %, Me [25th; 75th percentile]	10 (35) 71 [56; 78]
	CB at the level of the lower third of the upper arm, n (%)	0
	CB in the proximal region (Erb's point), n (%); CB degree, %, Me [25th; 7sth percentile]	5 (18) 75 [66; 91]
	Number of patients with M-response dispersion, n (%)	0
	CVm m/s; Me [25th; 75th percentile] (N>50) forearm elbow bend	52 [42; 54] 62 [56; 66]
n. ulnaris sin.	Number of patients with distal M-wave (dM-wave), n (%)	23 (82)
	dM-wave terminal latency, ms; Me [25th; 75th percentile] (N<3,0)	3 [2.6; 3.4]
	dM-wave amplitude, mV; Me [25th; 75th percentile] (N>6,0)	4.6 [2; 7.7]
	dM-wave duration, ms; Me [25th; 75th percentile]	5.7 [5.3; 6.6]
	CB at the level of the forearm, n (%); CB degree, %, Me [25th; 75th percentile]	11 (39) 55 [45; 61]
	CB at the level of the elbow joint, n (%)	0
	CB at the level of the shoulder, n (%); CB degree, %, Me [25th; 75th percentile]	3 (11) 60 [45; 74]
	CB in the proximal region (Erb's point), n (%); CB degree, %, Me [25th; 75th percentile]	5 (18) 83 [80; 93]
	Number of patients with M-response dispersion, n (%)	0
	CVm m/s; Me [25th; 75th percentile] (N>50) forearm elbow	55 [50; 64] 48 [39; 57]

Note. dM-wave, the distal motor response; CB, conduction block; CVm, the conduction velocity along of motor fibers; Here and in Table 4: n – number of patients; N – normal.

A comparative analysis of the severity of neurological deficit in MMN patients at the onset of the disease (retrospective assessment) and during long-term follow-up (assessment upon inclusion in the study) did not reveal clear trends; there were no significant differences in the total scores on the MRC and INCAT scales ($p>0.05$; Table 2).

Although a third of the patients had sensory complaints, objective evaluation of sensory disturbances revealed no changes in tactile, pain and temperature sensitivity. However, impaired vibration sensitivity in the lower extremities was noted in half of the cases (14/28; 50%).

Thus, the analysis of motor disorders in patients with advanced MMN from our sample revealed notably persistent and asymmetric paresis of the distal muscles of the arms in the vast majority (70%) of patients with a high level of disability (INCAT score: 3 points), involvement of the muscles of the lower extremities associated with gait impairment in every third patient, and the absence of a clear improvement on the therapy. Another noteworthy observation is objectively detectable impairment of vibration sensitivity in the legs in half of the cases among MMN patients under long-term follow-up.

Table 4. *Analysis of the results of an ENMG examination of the sensory fibers of the peripheral nerves of the hands in patients with MMN in the long-term follow-up*

The nerve	Nerve conduction studies variables	Parameters
n. medianus dex.	Number of patients with registered SNAP, n. (%)	28 (100)
	The latency of SNAP, ms; Me [25th; 75th percentile] (N<3,0)	2.5 [2.2; 2.7]
	The amplitude of SNAP, mkV; Me [25th; 75th percentile] (N>15,0)	22.8 [14; 27]
	SNCV, m/s; Me [25th; 75th percentile] (N>50)	64 [57; 68]
n. medianus sin.	Number of patients with registered SNAP, n. (%)	28 (100)
	The latency of SNAP, ms; Me [25th; 75th percentile] (N<3,0)	2.5 [2.3; 2.7]
	The amplitude of SNAP, mkV; Me [25th; 75th percentile] (N>15,0)	30 [16; 37]
	SNCV, m/s; Me [25th; 75th percentile] (N>50)	61 [56; 67]
n. ulnaris dex.	Number of patients with registered SNAP, n. (%)	28 (100)
	The latency of SNAP, ms; Me [25th; 75th percentile] (N<3,0)	2.1 [2; 2.4]
	The amplitude of SNAP, mkV; Me [25th; 75th percentile] (N>15,0)	15.7 [10; 28]
	SNCV, m/s; Me [25th; 75th percentile] (N>50)	59 [56; 64]
n. ulnaris sin.	Number of patients with registered SNAP, n. (%)	28 (100)
	The latency of SNAP, ms; Me [25th; 75th percentile] (N<3,0)	2 [1.9; 2.3]
	The amplitude of SNAP, mkV; Me [25th; 75th percentile] (N>15,0)	18 [12; 28]
	SNCV, m/s; Me [25th; 75th percentile] (N>50)	58 [55; 65]

Note. SNCV, sensory nerve conduction velocity

Neurophysiological characteristics. We were unable to register the M-response from the abductor pollicis brevis (innervation by the median nerve) in 29% of patients included in the study and the abductor digiti minimi (innervation by the ulnar nerve) in 18%. These figures indirectly suggest the percentage of patients with severe damage to the axons of the motor fibers of the nerves under study and their inexcitability. The median amplitude of distal motor responses from the hand muscles was at the lower limit of normal (Table 3).

A comprehensive assessment of the data obtained showed that patients with MMN caused by nodopathy of motor fibers, under long-term follow-up, did not develop secondary demyelination: the latency and duration of the dM response, as well as the SPM values in the distal parts were within normal limits, the form of the motor response remained intact (no M-response dispersion). All patients with detected dM response retained motor CBs, which were most frequently recorded on the forearm (in 39% of cases for motor fibers of the right median nerve, 35% for the left one, 46% for the right ulnar nerve, 39% for the left one) and less frequently (up to 20%) in the proximal sections (Erb's point). None of the patients with MMN, despite the pathogenetic therapy carried out in the long-term follow-up, demonstrated normalization of the parameters of the study of motor fibers of the hand nerves (see Table 3).

Despite the fact that the average NCS parameters of the study of sensory fibers of the hand nerves in patients with MMN in the late stages of the disease were within normal values, a decrease in the amplitude of the action potential of the sensory fibers (AFP) of the median nerves <15 ?V was noted in 36% of cases (10/ 28), ulnar nerves P in half of the patients (14/28; 50%; Table 4).

Thus, in the long-term follow-up of MMN, the majority of patients remain in compliance with the electrodiagnostic criteria for the disease EFNS/PNS 2010; approximately a third of patients develop significant secondary damage to the axons of the motor fibers of the nerves of the hands; secondary demyelination is not typical, and in half of the cases mild involvement of the axons of sensory fibers is recorded.

Discussion. Studies conducted abroad demonstrate a favorable disease course on regular IVIg therapy. R.M. Van den Berg-Vos et al. [9] conducted a long-term follow-up study in 11 patients with MMN who received IVIg maintenance therapy. During the observation, the frequency of administration and dose of the drug were determined for each patient individually; the observation period ranged from 4 to 8 years. Against the background of IVIg, muscle strength increased, and disability of the arm decreased significantly at the last follow-up examination compared with the pre-treatment examination. An NCS study in patients on IVIg showed that motor CB regressed on average in six nerve segments, though the emergence of CB in other segments was observed during the follow-up. Similar findings were reported by other researchers [10, 11].

J.M. Leger et al. [12] retrospectively analyzed data from 40 patients with MMN: 22 of them had not previously received treatment, 18 were on maintenance IVIg. Between 1995 and 2003, all patients included in the study received regular therapy. The total score on MRC increased significantly in 14 of 22 treatment-naive patients ($p<0.0001$). At the end of the follow-up period (mean 2.2±2.0 years), only 8 of 40 patients (22%) had significant remission, while 25 (68%) patients were dependent on intermittent IVIg infusions. The number of CB decreased or remained unchanged in 12 treatment-naive patients.

J. Nemoto et al. [13] retrospectively evaluated data from eight patients with MMN between 2005 and 2020. IVIg was effective in four patients. Two patients did not require therapy due to minimal severity of symptoms with a stable clinical course.

In our sample, we obtained different results. Having retrospectively analyzed data on the development and course of the disease in 28 patients with MMN with a median disease duration of 10 [8; 13] years, we noted that the severity of neurological deficit, assessed using the MRC and INCAT scales, at the onset and during the follow-up of the disease did not differ significantly ($p>0.05$); most patients in the late stages of the disease have severe (>3 points on the MRC scale) paresis of the distal muscles of the arms, leading to impaired fine motor skills (INCAT score of 3 points). This observation necessitated clarification. Upon analyzing the timing of diagnosis and initiation of therapy, we noted that the delay in diagnosis in the patients included in the study was 5.5 [2.0; 10.0] years, and the start of pathogenetic therapy was delayed by 6 [4; 10] years. In addition, a third of patients did not receive pathogenetic therapy at the time of inclusion in the study, and every second patient did not adhere to the IVIg regimen. Thus, our study demonstrated a number of issues, on the one hand, related to the untimely diagnosis of MMN in our country, on the other, the low quality of care for this patient population, and indirectly sheds light on the disease course in the absence of adequate IVIg.

Interesting data emerged from a thorough a comprehensive analysis of the results of neurophysiological findings of the peripheral nerves most affected by MMNCthe long nerves of the upper limbs. We showed that in the patients with MMN included in this study in the late stages of the disease, in a third of cases, significant secondary damage to the axons of the motor fibers of the nerves of the hands was observed, while secondary demyelination of the motor fibers did not develop, and mild involvement of sensory fiber axons of the nerves of the hands was noted in half of the cases. In addition, the study of the nerves of the hands (in the case of recording a dM response from the muscles of the hand with an amplitude of >1 mV) showed that motor CB persisted in locations atypical for compression throughout the disease course. None of the MMN patients in our study exhibited normalization of parameters in the study of motor fibers of the hand nerves. The results of NCS in patients with MMN should be interpreted taking into account that pathogenesis-based therapy in the study patients was started with a significant delay, and in the majority of cases there was a violation of the therapy regimen characterized by prolonged intervals between infusions. In addition, recording of neurophysiological signs of gross damage to the motor fiber axons may explain inadequate response to IVIg observed in one-third of patients.

Our findings regarding the involvement of sensory fibers in patients with late-stage MMN are consistent with reports from other studies, necessitating further analysis and understanding [14, 15].

Conclusion. MMN is a manageable condition where maintaining a high level of patient independence and self-care is achievable, which was consistently demonstrated in the previous studies. Unfortunately, our retrospective analysis has revealed diagnostic and quality care challenges for MMN patients in Russia. It has been shown that late diagnosis, delayed initiation of pathogenesis-based therapy and non-compliance with the infusion schedule inevitably leads to disability in patients with MMN. The authors express hope that the draft clinical guidelines for MMN will be approved by the Ministry of Health of the Russian Federation, and this will improve the situation in our country.

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