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Comparative assessment of limb function and conduction parameters in peripheral nerves in the course of two forms of amyotrophic lateral sclerosis

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Amyotrophic lateral sclerosis (ALS) is a progressive, incurable, neurodegenerative disease affecting the upper and lower motor neuron, which inevitably leads to the impaired fitness of patients and therefore deterioration of their quality of life.

The aim of the study was a comparative assessment of two forms of ALS in terms of limb function and electrophysiological parameters of peripheral nerves.

Material and methods. 20 persons participated in the study, where 10 suffered from bulbar-onset ALS and 10 had limb-onset ALS. Patients were examined clinically and electrophysiologically three times at three-month intervals. Rene Zazzo's card test and Mira Stambak's line-drawing test as well as the 10-metre walk test were used to assess limb function. The results of electrophysiological conduction in the area of nerves of upper and lower limbs were subjected to statistical analysis, as were the results of tests used to assess limb function.

Results. The comparative analysis of the obtained results demonstrated that patients with limb-onset ALS showed significant deterioration of conduction in proximal nerve sections compared to patients with bulbaronset ALS. Clinical reflection of this was impairment of limb function with a tendency for progression of these changes over time.

Conclusions. Progressive impairment of conduction in the area of proximal nerve sections and ventral roots of spinal nerves is reflected by the increasing disability of limbs in patients with ALS. In the course of the disease, patients with limb-onset ALS show both worse nerve conduction and limb function than patients with bulbar-onset

Key words: amyotrophic lateral sclerosis, neurodegenerative disease, limb function, conduction in proximal nerve sections

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Ocena porównawcza sprawności kończyn oraz parametrów przewodzenia w nerwach obwodowych w przebiegu dwóch postaci stwardnienia zanikowego bocznego

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Stwardnienie zanikowe boczne to postępująca, nieuleczalna choroba neurozwyrodnieniowa, dotycząca górnego i dolnego neuronu ruchowego, która w sposób nieunikniony prowadzi do upośledzenia sprawności chorych, a w związku z tym – pogorszenia ich jakości życia.

Celem pracy była ocena porównawcza dwóch postaci stwardnienia zanikowego bocznego pod względem sprawności kończyn oraz parametrów elektrofizjologicznych nerwów obwodowych.

Materiał i metody. W badaniu brało udział 20 chorych – 10 z postacią opuszkową i 10 z kończynową stwardnienia zanikowego bocznego. Chorych badano trzykrotnie klinicznie i elektrofizjologicznie w odstępach trzymiesięcznych. Do oceny sprawności kończyn wykorzystano test karty Rene Zazzo oraz test kreskowania Miry Stambak, oraz 10 metrowy test marszowy. Analizie statystycznej poddano wyniki przewodnictwa elektrofizjologicznego w zakresie nerwów kończyn górnych i dolnych oraz wyniki testów służących do oceny sprawności kończyn. Wyniki. Analiza porównawcza uzyskanych wyników badań wykazała,

że chorzy z postacią kończynową w porównaniu do chorych z postacią opuszkową, wykazywali istotne pogorszenie przewodnictwa w dosiebnych odcinkach nerwów, czego klinicznym odzwierciedleniem było upośledzenie sprawności kończyn, z tendencją do progresji tych zmian na przestrzeni czasu.

Wnioski. Postępujące upośledzenie przewodnictwa w zakresie dosiebnych odcinków nerwów oraz korzeni przednich rdzenia kręgowego znajduje odzwierciedlenie w narastaniu niesprawności kończyn u chorych na SLA. W przebiegu choroby chorzy z postacią kończynową SLA prezentują zarówno gorsze przewodnictwo nerwowe oraz gorszą sprawność kończyn niż chorzy z postacią opuszkową SLA.

Słowa kluczowe: stwardnienie zanikowe boczne, choroba neurozwyrodnieniowa, sprawność kończyn, przewodnictwo w dosiebnych odcinkach nerwów

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Amyotrophic lateral sclerosis (ALS) is a progressive, incurable disease of unknown cause and pathogenesis. In histopathology, it is defined as degeneration of both the upper and lower motor neuron, and it involves selective damage to nerve cells of the motor cortex, motor cells of nuclei of cranial nerves in medulla oblongata and motor neurons of anterior horns of the spinal cord [10,16,27].

ALS occurs all over the world. Incidence is estimated to be 1-3 cases per 100,000, whereas morbidity rate varies between

6 and 8 cases per 100,000. An increase of incidence has been observed in recent years. Most often, persons aged between 50 and 65 fall ill, and the disease occurs before 40 or after 70 in only 10% of cases. There are also cases of patients aged 20 or even less. The disease affects men slightly more frequently (1.5:1), albeit women predominate in the case of bulbar-onset ALS [2,10]. In 90-95% of patients, the disease is of sporadic nature (SALS/sporadic amyotrophic lateral sclerosis) while the other 5-10% is accounted for by the familial form

(FALS/familial amyotrophic lateral sclerosis). The familial forms are usually inherited in line with the autosomal dominant inheritance pattern. The following clinical forms are distinguished: classic – symptoms relate to the upper and lower motor neuron; progressive muscular atrophy (PMA) – symptoms relate to the lower motor neuron; primary lateral sclerosis (PLS) – symptoms relate to the upper motor neuron; progressive bulbar palsy (PBP). In practice, a slightly different division is used, i.e. bulbar-onset ALS, spinal-onset ALS as well as flail arm and flail leg ALS. The course of these affects mainly the lower motor neuron [18,24,26]. The diagnosis is based on the clinical picture supplemented by an electrophysiological examination. Currently, the modified El Escorial criteria from 1998 are used for clinical classification of ALS [1,4,5,11,12,14,21].

ALS is characterised by the occurrence of a fairly long, preclinical phase, since symptoms appear when approximately 40% of motor neurons of anterior horns of the spinal cord have been lost. In the clinical phase, regardless of the nature of the disease's onset - be it more often occurring weakening of muscles of upper limbs than lower limbs (classic spinal-onset form), weakening of bulbar muscles or damage of the upper or lower motor neuron only - a linear decrease in motor function occurs over time [2,22,24,26]. Impairment of motor fitness has a major impact on the quality of life of those patients. They become dependent on the environment in the basic activities of their everyday life, i.e. washing, dressing up, eating or breathing. The course of the disease is progressive with varying duration. Patients with spinal-onset ALS live slightly longer than those suffering from bulbar-onset ALS. Patients usually live for about 5 years.

MATERIAL AND METHODS

The study was conducted in a group of 20 persons with clinically certain or probable diagnosis of amyotrophic lateral sclerosis, including 5 women and 15 men aged 43-85 (mean age: 65.5). Patients included in the study were those with their diagnosis established on the basis of the modified EI Escorial criteria from 1998 (also referred to as the Airlie House criteria) [7]. All subjects were informed on the scope of clinical and electrophysiological examination and provided oral consents to the proposed diagnostic procedure.

All patients underwent a full neurological examination with an assessment of limb function. On the basis of the clinical picture, patients were divided into two groups. Group A consisted of patients with limb-onset ALS, whereas group B comprised patients with bulbar-onset ALS. Medical history of none of the subjects included other conditions which would have impaired their fitness. Patients from group B did not report weakness in their limbs.

Patients were examined three times at three-month intervals. The first measurement (preliminary) was marked with letter A, the examination after the first three months was marked with letter B and the one after another three months – with letter C.

To assess upper limb function, two tests used to assess lateralisation of limbs in children and adolescents were used: *Rene Zazzo's* card test and *Mira Stambak's* line-drawing test [19]. The control group consisted of 20 healthy persons at the age of 20-86 (mean age 59,6). During all three measurements, both upper limbs were assessed in both tests, the result of the fitter limb was chosen and then it was compared against the control group. Interpretation of the above-mentioned tests was as follows: the lower the result on *Rene Zazzo's* card test, the fitter the upper limb and the higher the result on *Mira Stambak's* line-drawing test, the fitter the limb. To assess lower limb function, the 10-metre walk test was employed where the lower the result, the higher the lower limb fitness.

In all patients, the standard electrophysiological examination was conducted three times. Motor conduction was examined in both median nerves and ulnar nerves as well as in the right peroneal nerve and tibial nerve. Distal latency, conduction velocity

and the compound muscle action potential (CMAP) were assessed. The shortest F wave latency was also determined. Furthermore, sensory conduction was investigated in both median nerves and ulnar nerves as well as in the right sural nerve using the orthodromic method. Latency, conduction velocity and the sensory nerve action potential (SNAP) were assessed.

The results of tests assessing limb function and electrophysiological examinations were analysed in the entire investigated group as well as in patients with limb-onset and bulbaronset ALS.

The obtained results underwent statistical analysis where the arithmetic mean (X) and the standard deviation (SD) were calculated for the measurable characteristics. For characteristics consistent with the normal distribution, the Shapiro-Wilk test was employed. Furthermore, goodness of fit was assessed based on the coefficients of skewness and kurtosis and the standard errors as well as on the basis of visual assessment of its histogram and Q-Q plot. In comparisons between the groups, Student's t-test was used. In order to determine the equality of variance in samples, Levene's test was employed. For calculation purposes, the maximum permissible type I error was taken to be $\acute{a}=0.05$, and the statistical significance level – p<0.05.

RESULTS

The analysis of results during the preliminary measurement indicated that, relative to the laboratory's own standards, all subjects (100%) obtained normal results with regard to the distal latency of the right ulnar and the right peroneal motor nerve as well as with regard to the conduction velocity of the left median, the left ulnar, the right peroneal and the right tibial motor nerve. The vast majority of subjects also showed normal results with regard to the distal latency of the left ulnar (95%) and the right tibial (90%) motor nerve as well as with regard to the conduction velocity of the right median (95%) and the right ulnar (90%) motor nerve as well as in terms of the minimum F wave latency of the right (70%)/left (85%) median, the right (85%)/left (90%) ulnar, the right peroneal (95%) and the right tibial (85%) motor nerve.

Most often, abnormalities related to the value of the compound muscle action potential (CMAP) of the right (70%)/left (70%) median, the right (35%)/left (40%) ulnar, the right peroneal (85%) and the right tibial (45%) nerve (tab. 1).

The analysis of the results of sensory conduction in the area of both median/ulnar nerves or the right sural nerve did not reveal any significant abnormalities.

The assessment of test results (*Rene Zazzo's* card test, *Mira Stambak's* line-drawing test and the 10-metre walk test) showed that, relative to the laboratory's own standards, all subjects (100%) obtained negative results on the card test, the vast majority (80%) had a negative result on the walk test and the majority (60%) – on the line-drawing test (tab.2).

During preliminary measurement A, the analysis using Student's t-test for independent samples did not reveal any statistically significant differences between the groups in terms of the investigated variables (> 0.05). That meant that, during preliminary measurement A, persons with limb-onset ALS did not differ from persons with bulbar-onset ALS in terms of the individual parameters of electrophysiological assessment of the motor nerve. During the next examination after 3 months (B), the analysis showed statistically significant differences between the groups in terms of the minimum F wave latency of the right peroneal nerve: t(17) = 2.49; p < 0.05; d = 1.15 and the right tibial nerve: t(18) = 2.60; p < 0.05; d = 1.16. That meant that persons with limb-onset ALS were characterised by a higher value of the minimum F wave latency of the peroneal nerve and the tibial nerve than persons suffering from bulbar-onset ALS. No statistically significant differences during this measurement were demonstrated in terms of the other parameters of electrophysiological assessment of the examined motor nerves.

Table 1. Electrophysiological assessment of the motor nerve during measurement A, B and C in persons with limb-onset ALS (n=10) and persons with bulbar-onset ALS (n=10)

Tabela 1. Elektrofizjologiczna ocena nerwu ruchowego podczas pomiaru A, B i C u chorych z postacią kończynową SLA (n=10) i chorych z postacią opuszkową SLA (n=10)

Motor nerve	Parameter	ALS form	Measur. A			Measur. B			Measur. C		
			n	Х	SD	n	х	SD	n	Х	SD
Left median nerve	Distal latency [ms]	limb-onset ALS	10	4.10	0.44	10	4.19	0.70	10	4.25	0.62
		bulbar-onset	10	4.06	0.39	10	4.26	0.72	10	3.87	0.46
	Amplitude [mV]	limb-onset ALS	10	3.26	2.15	10	3.26	1.60	10	3.30	1.93
		bulbar-onset	10	4.93	1.87	10	3.47	1.47	10	4.25	1.49
	Conduction	limb-onset ALS	10	53.54	2.60	10	53.16	1.51	10	53.05	1.63
	velocity [m/s]	bulbar-onset	10	53.58	2.26	10	54.78	3.37	10	54.78	3.52
	Min. F wave latency [ms]	limb-onset ALS	7	26.06	1.56	8	26.10	2.40	8	25.68	2.26
		bulbar-onset	10	25.25	2.00	8	25.94	1.42	8	25.10	1.56
Right median nerve	Distal latency [ms]	limb-onset ALS	10	4.35	0.65	10	4.23	0.74	10	4.29	0.62
		bulbar-onset	10	4.18	1.34	10	4.37	1.28	10	4.27	1.36
	Amplitude [mV]	limb-onset ALS	10	3.57	2.52	10	3.25	2.61	10	3.59	2.43
		bulbar-onset	10	4.80	2.59	10	3.99	1.91	10	4.22	2.51
	Conduction velocity [m/s]	limb-onset ALS	10	54.33	3.28	10	54.15	4.78	10	52.98	3.25
		bulbar-onset	10	52.59	2.99	10	53.11	3.55	10	54.89	4.84
	Min. F wave latency [ms]	limb-onset ALS	8	26.56	2.77	7	26.87	1.83	8	26.14	2.38
		bulbar-onset	9	27.09	4.56	8	25.04	2.09	9	24.21	1.58
Left ulnar nerve	Distal latency [ms]	limb-onset ALS	10	3.23	0.30	10	3.13	0.34	10	3.19	0.22
		bulbar-onset	10	3.15	0.41	10	3.15	0.30	10	3.06	0.23
	Amplitude [mV]	limb-onset ALS	10	5.22	3.44	10	4.70	3.37	10	4.81	3.23
		bulbar-onset	10	5.70	2.11	10	5.07	1.97	10	4.02	2.30
	Conduction velocity [m/s]	limb-onset ALS	10	57.7	4.01	10	55.48	3.87	10	58.31	8.24
		bulbar-onset	10	5788	4.26	10	55.92	3.39	10	57.57	3.01
	Min. F wave latency [ms]	limb-onset ALS	8	28.93	2.06	7	27.57	2.06	8	28.21	3.37
		bulbar-onset	10	26.9	2.91	9	25.54	2.91	9	26.26	1.66
Right ulnar nerve	Distal latency [ms]	limb-onset ALS	10	3.19	0.27	10	3.07	0.35	10	3.12	0.17
		bulbar-onset	10	3.00	0.40	10	2.99	0.52	10	3.10	0.19
	Amplitude [mV]	limb-onset ALS	10	5.21	3.68	10	4.91	3.62	10	4.44	3.39
		bulbar-onset	10	7.29	2.86	10	6.52	2.69	10	3.98	2.08
	Conduction velocity [m/s]	limb-onset ALS	10	57.10	4.22	10	57.83	4.02	10	55.09	3.26
		bulbar-onset	10	57.72	2.53	10	58.99	5.11	10	59.79	5.19
	Min. F wave latency [ms]	limb-onset ALS	7	27.19	2.31	7	27.24	2.61	7	27.41	2.09
		bulbar-onset	10	26.38	1.84	10	25.47	2.87	10	24.87	2.46
Right peroneal nerve	Distal latency [ms]	limb-onset ALS	10	4.21	0.38	10	4.08	0.53	10	4.15	0.42
		bulbar-onset	10	4.04	0.38	10	3.99	0.37	10	3.83	0.47
	Amplitude [mV]	limb-onset ALS	10	3.25	1.59	10	3.38	1.50	10	3.07	1.70
		bulbar-onset	10	3.80	2.53	10	3.01	1.72	10	2.69	1.07
	Conduction velocity [m/s]	limb-onset ALS	10	44.10	3.90	10	43.45	2.64	10	43.70	3.95
		bulbar-onset	10	46.14	3.39	10	44.01	3.02	10	44.77	3.23
	Min. F wave latency [ms]	limb-onset ALS	10	48.24	3.85	10	49.20	4.75	10	47.91	4.65
		bulbar-onset	10	46.00	7.89	9	43.98	4.33	10	44.85	3.30
Right tibial nerve	Distal latency [ms]	limb-onset ALS	10	3.51	0.21	10	3.54	0.28	10	3.52	0.25
		bulbar-onset	10	3.66	0.64	10	3.61	0.77	10	3.47	0.43
	Amplitude [mV]	limb-onset ALS	10	8.11	4.39	10	7.96	4.34	10	5.38	2.42
		bulbar-onset	10	5.47	3.73	10	5.77	3.55	10	4.57	2.15
	Conduction velocity [m/s]	limb-onset ALS	10	50.37	2.60	10	52.19	4.21	10	53.05	5.08
		bulbar-onset	10	51.24	2.52	10	54.36	2.83	10	53.30	4.12
	Min. F wave	limb-onset ALS	10	49.73	4.08	10	51.73	4.95	10	51.22	4.76
	latency [ms]	bulbar-onset	10	49.51	7.78	10	44.73	6.94	10	48.00	2.49

n - number, X - mean, SD - standard deviation, Measur - measurement

The analysis of the results of electrophysiological examination of motor nerves during the last measurement (C) showed statistically significant differences in terms of the minimum F wave latency of the right ulnar nerve: t(15) = 2.23; p <

0.05; d =1.11 and the conduction velocity in the motor fibres of that nerve: t(18) = 2.42; p < 0.05; d = 1.08. That meant that persons with limb-onset ALS were characterised by a higher value of the minimum F wave latency of the right ulnar nerve

Table 2. Diagnosis of limb function during measurement A, B and C in persons with limb-onset ALS (n = 10) and persons with bulbar-onset ALS (n=10)

Tabela 2. Diagnoza sprawności kończyn podczas pomiaru A, B i C u chorych z postacią kończynową SLA (n=10) i chorych z postacią opuszkową SLA (n=10)

Parameter	ALS form	Measur. A			Measur. B			Measur. C		
		n	X	SD	n	х	SD	n	х	SD
Rene Zazzo's card test	limb-onset ALS	10	29.20	12.33	10	30.20	13.27	10	32.90	17.97
	bulbar-onset	10	20.20	3.33	10	24.10	5.24	10	24.20	6.86
Mira Stambak	limb-onset ALS	10	92.20	27.68	10	83.90	23.67	10	81.90	30.78
line-drawing test	bulbar-onset	10	130.40	27.42	10	120.60	29.64	10	113.20	21.34
10-metre	limb-onset ALS	10	16.50	3.84	10	4.45	4.45	10	21.00	4.47
walk test	bulbar-onset	10	11.90	2.69	10	12.80	3.05	10	14.40	2.46

n - number, X - mean, SD standard deviation, Measur - measurement

than persons suffering from bulbar-onset ALS. On the other hand, persons with bulbar-onset ALS achieved a higher conduction velocity value in the motor fibres of that nerve than persons suffering from limb-onset ALS. No statistically significant differences between the groups were demonstrated for the other variables.

The assessment of the results of the examined electrophysiological parameters of the investigated sensory nerves did not reveal any significant differences between the forms of ALS during any of the measurements either.

The results of tests used to assess limb function, i.e. *Rene Zazzo's* card test, *Mira Stambak's* line-drawing test and the 10-metre walk test, during the first examination showed statistically significant discrepancies between the investigated groups of patients with ALS.

Patients suffering from bulbar-onset ALS were characterised by a lower result on *Rene Zazzo's* card test $\{t(10.30) = 2.23; p < 0.05; d= 1.00\}$ and a higher result on *Mira Stambak's* line-drawing test [t(18) = 3.10; p < 0.01; d = 1.39], meaning that they showed better fitness in the area of upper limbs than patients with limb-onset ALS (fig. 1, 2).

Similarly, the results of analysis of the 10-metre walk test revealed better fitness in the area of lower limbs in patients suffering from bulbar-onset ALS than in patients with limb-onset ALS, which was reflected by a lower result on that test in the former group $\{t(18) = 3.11; p < 0.01; d = 1.39\}$ (fig. 3).

During the second examination (B), still patients with bulbaronset ALS showed a higher result on *Mira Stambak's* line-drawing test $\{t(18) = 3.06; p < 0.01; d=1.37\}$ and a lower result on the 10-metre walk test $\{t(18) = 3.34; p < 0.01; d=1.49\}$ (fig. 2,3). No statistically significant differences between the subjects were demonstrated with respect to *Rene Zazzo's* card test (Fig. 1). The last measurement (C), similarly to the second one (B), revealed a higher result on *Mira Stambak's* line-drawing test in subjects with bulbar-onset ALS {t(18); p < 0.05;

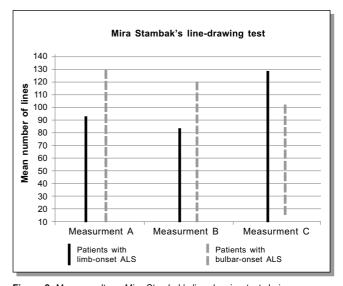


Figure 2. Mean result on *Mira Stambak's* line-drawing test during measurement A, B and C in persons with limb-onset and bulbar-onset ALS **Rycina 2.** Średni wynik testu kreskowania *Miry Stambak* podczas pomiaru A, B i C u chorych z kończynową i opuszkową postacią SLA

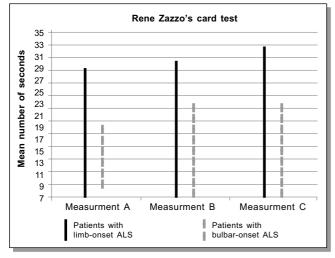


Figure 1. Mean result on *Rene Zazzo's* card test during measurement A, B and C in persons with limb-onset and bulbar-onset ALS **Rycina 1.** Średni wynik testu karty *Rene Zazzo* podczas pomiaru A, B i C u chorych z kończynową i opuszkową postacią SLA

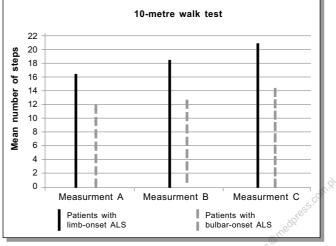


Figure 3. Mean result on the 10-metre walk test during measurement A, B and C in persons with limb-onset and bulbar-onset ALS Rycina 3. Średni wynik 10-metrowego testu marszowego podczas pomiaru A, B i C u chorych z kończynową i opuszkową postacią SLA

d = 1.18} and a lower result on the 10-metre walk test {t(18) = 4.09; p < 0.001; d = 1.83} than in persons suffering from limbonset ALS (Fig. 2,3). No differences with respect to Rene Zazzo's card test were demonstrated, meaning that both groups achieved similar results (fig. 1).

DISCUSSION

In patients with amyotrophic lateral sclerosis examined electrophysiologically, axonal motor neuropathy with impairment of conduction in proximal sections of motor nerves is most frequently diagnosed. Furthermore, in clinical terms, those patients show varying degrees of impairment of their physical fitness.

In the discussed group, during the preliminary examination, the compound muscle action potential (CMAP) was found to be significantly lowered in the majority of patients, regardless of whether they had bulbar-onset or limb-onset ALS. In addition, we found impaired conduction in proximal nerve sections. The results obtained by us were similar to those obtained by other authors [3,9,17,25].

Similarly to Liu XX. et al., we showed that a decrease in compound muscle action potential was reflected by worse physical fitness [20]

Multiple clinical reports mention the loss of neuromuscular function in the course of ALS [13,28]. Our research results during the preliminary measurement showed that almost all patients, both with bulbar-onset and limb-onset ALS, obtained negative results on the conducted tests.

For assessment of disability in the course of ALS, Couratier et al. and Ohashi et al. employed many scales, i.e. the ALS Functional Rating Scale, the ALS Severity Scale, the Appel Scale, the Norris Scale [6,23]. However, the authors of this paper made use of three other tests, i.e. Rene Zazzo's card test, Mira Stambak's line-drawing test and the 10-metre walk test. The results obtained were consistent with those of the aforementioned authors in spite of differences in the tests used.

In addition to the clinical tests, we used electrophysiological examinations to diagnose and monitor the progress of the disease as well. A comparison of the results of electrophysiological examinations of patients with limb-onset or bulbar-onset ALS did not show any statistically significant differences during measurement A. Nevertheless, differences were visible with regard to the minimum F wave latencies in the next two measurements, i.e. during measurement B of lower limb nerves and during measurement C of upper limb nerves. Patients suffering from limb-onset ALS showed higher F wave latency values than patients with bulbar-onset ALS, i.e. the former ones had worse conduction in the area of proximal nerve sections or ventral roots of spinal nerves. The analysis of the results of tests assessing physical fitness revealed that patients with limb-onset ALS showed considerably worse fitness than patients with bulbar-onset ALS as early as during the first measurement. Patients suffering from bulbar-onset ALS achieved better results on all three tests during measurement A as they did during measurements B and C on Mira Stambak's line-drawing test and the 10-metre walk test

In their paper, Fang et al. compared results of the minimum F wave latencies of the examined nerves in patients suffering from ALS and having symptoms of pyramidal tract syndrome and in patients suffering from ALS and not having any symptoms of pyramidal tract syndrome, showing worse conduction in the former group [8]. The authors of this paper made a similar comparison involving two groups of patients.

In their paper, Kulkantrakorn K. et al. described bulbar-onset ALS as the one associated with the worst prognosis [17]. In contrast, having analysed the results of their own research, the authors of this paper concluded that the deterioration with regard to electrophysiological conduction and limb function over six months affected much more the patients with limb-onset amyotrophic lateral sclerosis, meaning that they had a worse level of fitness than those suffering from bulbar-onset ALS.

Previous reports included mainly comparisons of patients with damage to the lower motor neuron only and the upper motor neuron only, where the patients with more quickly progressing deterioration of physical efficiency were the ones with the affected lower motor neuron according to six-month observations of the authors of this paper [15]. Nevertheless, the results of that research can be compared with the authors' own results as some similarities can be found.

CONCLUSIONS

- 1. Progressive impairment of conduction in the area of proximal nerve sections and ventral roots of spinal nerves is reflected by the increasing disability of limbs in patients with ALS
- In the course of the disease, patients with limb-onset ALS show both worse nerve conduction and limb function than patients with bulbar-onset ALS.

REFERENCES

- 1. Al-Chalabi A, Hardiman O, Kiernan MC, et al.: Amyotrophic lateral sclerosis: moving towards a new classification system. Lancet Neurol, 2016; 15(11)⁻ 1182-1194
- 2. Andrews J.: Amyotrophic lateral sclerosis: clinical management and research update. Curr Neurol Neurosci Rep, 2009; 9(1): 59-68.
- 3. Argyrion AA, Polychronopoulos P, Talelli P, et al.: F wave study in amyotrophic lateral sclerosis; assessment of balance between upper and lower motor neuron involvement. Clin Neurophysiol, 2006; 117(6):
- 4. Bresch S, Delmont E, Soriani MH, et al.: Electrodiagnosis criteria for early diagnosis of bulbar-onset ALS: a comparison of El Escorial, revised El Escorial and Awaji algorithm. Rev Neurol, 2014; 170(2):
- 5. Bromberg MB.: Electrodiagnostic studies in clinical trials for motor neuron disease. *J Clin Neurophysiol*, 1998; 15(2): 117-128. Couratier P, Torny F, Lacoste M.: Functional rating scales for amyotro-
- phic lateral sclerosis. Rev Neurol (Paris), 2006; 162(4): 502-507
- Dengler R.: El Escorial or Awaji Criteria in ALS diagnosis, what should we take? *Clin Neurophysiol*, 2012; 123(2): 217-218.
- 8. Fang J, Cui LY, Liu MS, et al.: F Wave Study in Amyotrophic Lateral Sclerosis: Assessment of Segmental Motoneuronal Dysfunction. Chin Med J (Engl), 2015; 128(13): 1738-1742.
- Felice KJ.: Nerve conduction velocities of single thenar motor axons based on the automated analysis of F wave in amyotrophic lateral sclerosis. Muscle Nerve, 1998; 21(6): 756-761.
- 10. Francis K. Bach JR. Delisa JA.: Evaluation and rehabilitation of patients with adult motor neuron disease. Arch Phys Med Rehabil, 1999;
- 11. Huynh W, Simon N, Grosskreutz J, et al.: Assessment of the upper motor neuron in amyotrophic lateral sclerosis. Clinical Neurophysiology, 2016; 127(7): 2643-2660.
- 12. Inghilleri M, Iacovelli E.: Clinical neurophysiology in ALS. Arch Ital Biol, 2011: 149(1): 57-63
- 13. Jensen L, Djurtoft JB, Bech RD, et al.: Influence of Resistance Training on Neuromuscular Function and Physical Capacity in ALS Patients. J. Neurodegener Dis, 2017; 2017: 1436519.
- 14. Joyce NC, Carter GT.: Electrodiagnosis in persons with amyotrophic lateral sclerosis. *PMR*, 2013; 5(Suppl 5): 89-95.
 15. Kent-Braun JA, Walker CH, Weiner MW, et al.: Functional significance
- of upper and lower motor neuron impairment in amyotrophic lateral sclerosis. Muscle Nerve, 1998; 21(6): 762-768.
- 16. Kiernan MC, Vucic S, Cheah BC, et al.: Amyotrophic lateral sclerosis. Lancet, 2011; 377(9769): 942-955.
- 17. Kulkantrakorn K, Suksasunee D.: Clinical, electrodiagnostic and outcome correlation in ALS patients in Thailand. J Clin Neurosci, 2017; 43: 165-169.
- 18. Lee CN.: Reviewing evidences on the management of patients with motor neuron disease. Hong Kong Med J. 2012: 18(1): 48-55
- 19. Lemonnier E.: Psychopathology in children with dyspraxia. Arch Pediatr, 2010; 17(8): 1243-1248.
- 20. Liu XX, Zhang J, Zheng JY, et al.: Stratifying disease stages with different progression rates determined by electrophysiological test in patients with amyotrophic lateral sclerosis. Muscle Nerve, 2009; 39(3): 304-309.
- 21. Ludolph A, Drory V, Hardiman O, et al.: A revision of the El Escorial Criteria-2015. Amyotroph Lateral Scler Frontotemporal Degener, 2015; 16(5-6): 291-292
- 22. Murray L, Butow PN, White K, et al.: Advance care planning in motor neuron disease: A qualitative study of caregiver perspectives. Palliat Med. 2016; 30(5): 471-478.
- 23. Ohashi Y, Tashiro K, Itoyama Y, et al.: Study of functional rating scale for amyotrophic lateral sclerosis: revised ALS FRS Japanese version. No To Shinkei, 2001; 53(4):346-355.

- 24. Rocha JA, Reis C, Simőes F, et al.: Diagnostic investigation and multidisciplinary management in motor neuron disease. J. Neurol, 2005;
- 252(12): 1435-1447.25. Theys PA, Peeters E, Robberecht W.: Evolution of motor and sensory deficits in amyotrophic lateral sclerosis estimated by neurophysiological techniques. *J.Neurol*, 1999; 246(6): 438-442. 26. Turner MR, Hardiman O, Brooks BR, et al.: Controversies and priorities
- in amyotrophic lateral sclerosis. The Lancet, 2013; 12(3): 310-322.
- Wood-Allum C, Shaw PJ.: Motor neurone disease: a practical update on diagnosis and management. *Clin Med (Lond)*, 2010; 10(3): 252-258.
 van Dijk JP, Schelhaas HJ, van Schaik IN, et al.: Monitoring disease progression using high-density motor unit number estimation in amyotrophic lateral sclerosis. *Muscle Nerve*, 2010; 42(2): 239-244.

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