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Original Research Article

The degree of damage in the peripheral facial nerve palsy in children depending on the cause: The role of Lyme neuroborreliosis

Elżbieta Młynarczyk [a,*], *Wacław Kopala* [b], *Jacek Morski* [a], *Aleksandra Melnyk* [a]

[a] *Department of Pediatric Neurology, Provincial Specialist Children's Hospital in Olsztyn, Poland*
[b] *Department of Pediatric Otolaryngology, Provincial Specialist Children's Hospital in Olsztyn, Poland*

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ABSTRACT

Introduction: Symptoms of peripheral facial nerve paralysis occur in patients of all age groups. Etiology of this disease is very diverse.
Aim: The aim of this work is to estimate facial nerve damage in relation to presumable cause, including cases in which Lyme neuroborreliosis has been confirmed by laboratory testing.
Material and methods: Thirty-three patients with symptoms of facial nerve paralysis were examined from January to September 2012 in the Department of Pediatric Neurology of the Provincial Specialist Children's Hospital in Olsztyn. Facial nerve function was classified on a three-stage scale created by the authors of this work. Full neurological examination was performed in all patients. Laboratory tests for Lyme-specific antibodies IgG and IgM by enzyme-linked immunosorbent assay has also been performed.
Results and discussion: In the study group, diagnosis of Lyme neuroborreliosis was confirmed in nine patients. Analysis shows that the stage of facial nerve damage in the group with Lyme disease was significantly heavier and that percentage of mild paralysis was significantly higher in the group of patients in whom Lyme disease has not been confirmed.
Conclusions: The role of neuroborreliosis among the infectious factors ought to be emphasized. It seems significant that in patients with confirmed Lyme disease the stage of facial nerve damage was considerably deeper and the recovery was slower.

* *Correspondence to*: Department of Pediatric Neurology, Provincial Specialist Children's Hospital in Olsztyn, Żołnierska 18 A, 10-561 Olsztyn, Poland. Tel.: +48 89 539 33 60; fax: +48 89 533 77 01.
E-mail addresses: neurodziec@wssd.olsztyn.pl (E. Młynarczyk), wkopala@onet.pl (W. Kopala).

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1. Introduction

Facial nerve, as one of the most important of the cranial nerves, has a very complex structure and course, which implies a diverse symptomatology of its damage. It consists of a motor root and the part described under the name of intermediate nerve, which contains sensory (afferent) and parasympathetic (secretory) fibers. Motor fibers that reach facial mimic muscles arise from the facial nerve nucleus located between the dorsal pons and medulla oblongata.[1–3] Symptoms of peripheral facial nerve palsy occur in patients of all age groups, beginning from the neonatal period. Etiology of palsies is very diverse.[4]

The most common unilateral facial nerve damage is defined as idiopathic peripheral facial paralysis – Bell's palsy, which represents 75% of facial nerve paralyzes. Currently, it seems that even these cases have a viral etiology and are often preceded by exposure to cold.[1,3,5,6]

Recently increasing attention is paid to *Borrelia burgdorferi* infection as a potential cause of the facial nerve paralysis. In case of *B. burgdorferi* infection there are probably two mechanisms responsible for the development of peripheral neuropathies. The first results from the direct interaction between the spirochete and the nerve cell, which leads to its damage, while the second is associated with stimulation of the immune response against this pathogen. Reports in the literature describe the T- and B-cell autoreactivity against endogenous neural structures, which in the presence of 41 kDa *B. burgdorferi* flagellin may elicit inflammatory mediators.[4,7] Inflammatory and angiopathic lesions, perivascular infiltrates, partially with thrombosis of the vasa nervorum are responsible for the axonal damage of peripheral nerves. Axonal neuropathies, in contrast to direct nerve damage, are less responsive to antibiotic therapy.[4,7,8]

Secondary causes include inter alia, congenital anomalies, such as Mobius syndrome, trauma, including iatrogenic trauma during surgical procedures, neoplasms, particularly cerebellopontine angle tumors or parotid tumors, and infections.[1,3,5]

2. Aim

The aim of this work was to assess the degree of facial nerve damage in relation to the presumable cause, with particular reference to cases of elevated antibody titers against *B. burgdorferi* in serum and cerebrospinal fluid (CSF) findings indicating neuroborreliosis.

3. Material and methods

In the period between the beginning of January and the end of September 2012 in the Department of Neurology of the Provincial Specialist Children's Hospital in Olsztyn, 33 patients with symptoms of peripheral facial nerve palsy were examined. Study group consisted of 10 boys (30.3%) and 23 girls (69.7%) aged 4–18 years. Median age was 8.5 years. Facial nerve function was classified on a scale created by the authors. In the study, visual determination of each separate function was performed: wrinkling the forehead, closing the eyelids, drooping of the corner of the mouth, whistling and grinning. Each of this functions was given a specific number of points (on a scale of 0–3), where 0 points – no damage of the motor function, 1 point – minimal damage that becomes visible by mimic movements with modest asymmetry of the face, 2 points – moderate damage characterized by dysfunction visible also at rest with distinct muscle weakness and facial muscle movements still present with great effort, and 3 points – severe damage to the function with great asymmetry of the face and lack of movements even with great effort.

Corneal reflex was also evaluated. The presence of corneal reflex was given 0 points, uncertain reflex was assessed as 1 point, and lack of reflex as 2 points. In such an examination, the maximum number of points indicating a severe damage was 17 points.

Dependent on the number of points obtained on examination, patients were divided into three groups:

- *1st degree of damage* (mild): this group included patients in whom facial nerve function on the scale described above ranged from 1 to 10 points,
- *2nd degree of damage* (moderate): range of points from 11 to 13,
- *3rd degree of damage* (severe): range of points from 14 to 17.

For all patients a thorough medical history was obtained, that included presumable etiology of facial nerve palsy, particularly tick bite within the preceding six months and potential rash at the site of the tick bite.

Laboratory tests for serum *B. burgdorferi*-specific IgG and IgM antibodies detected by immunoenzymatic method were performed, and in case of positive results, by Western Blot assay. Lumbar puncture was also performed to determine concentrations of *B. burgdorferi*-specific IgG and IgM antibodies in CSF in all patients with elevated serum antibody titers. Pleocytosis and elevated protein concentrations in the CSF were also analyzed.

4. Results

In the examined group of patients it was found that facial nerve paralysis occurs more frequently on the left side and in girls.

Patients, in whom no potential cause of facial nerve palsy was found, constituted a group of 15 subjects (45.5%). Other possible causes in the examined group of patients might have included: multiple sclerosis (2 patients), herpes (1 patient), temporal bone fracture (1 patient), acute otitis media (1 patient), history of chemotherapy for leukemia (2 patients), electrocution (1 patient), and Epstein–Barr infection (1 patient), making a total of 9 cases (27.3%). Elevated serum *B. burgdorferi*-specific IgG and IgM antibodies concentrations were found in the 9 patients (27.3%) (Fig. 1 and Table 1). After obtaining guardian consent, these children had lumbar puncture performed, which confirmed elevated IgM and IgG antibody titers in the CSF in 4 patients. Pleocytosis and elevated protein concentrations in the CSF were found in 2 cases (Table 2).

Table 1 – Clinical and laboratory characteristics of patients with peripheral facial nerve paralysis with positive serum IgM and IgG antibody titers against *B. burgdorferi*.

Patient	Age (years)	Sex	Neurological history	*B. burgdorferi* in serum IgG (AU/mL)	*B. burgdorferi* in serum IgM (AU/mL)
WR	8	F	No tick bite	42.8	6.1
KC	11	M	Tick bite, rash (+)	262.9	22.0
LH	7	F	No tick bite	5.0	58.5
EW	12	F	No tick bite	16.8	4.6
RJ	16	M	Tick bite, rash (–)	54.0	7.2
JM	10	M	Tick bite, rash (+)	60.0	104.1
GP	13	M	Tick bite, rash (+)	19.6	25.6
MM	6	F	Tick bite, rash (–)	23.2	2.0
EB	8	F	Tick bite, rash (–)	21.2	20.9

Note: Qualitative assessment of serum IgM antibodies against *B. burgdorferi*: positive result ≥22 AU/mL; borderline result between 18 AU/mL and 22 AU/mL; negative result <18 AU/mL. Qualitative assessment of serum IgG antibodies against *B. burgdorferi*: positive result ≥15 AU/mL; borderline result between 10 AU/mL and 15 AU/mL; negative result <10 AU/mL.

Table 2 – Clinical and laboratory characteristics of cerebrospinal fluid of patients with peripheral facial nerve paralysis with positive serum IgM and IgG antibody titers against *B. burgdorferi*.

Patient	Age (years)	Sex	CSF protein (mg/dL)	CSF pleocytosis (/μL)	*B. burgdorferi* in CSF IgG (AU/mL)	*B. burgdorferi* in CSF IgM (AU/mL)
WR	8	F	24.9	1	0.2	0
KC	11	M	18.7	2	6.2	19.80
LH	7	F	24.3	1	5.5	17.30
EW	12	F	34.6	1	0.2	7.50
RJ	16	M	32.4	2	2.3	0.01
JM	10	M	82.3	51	6.8	16.00
GP	13	M	63.6	60	35.4	84.90
MM	6	F	23.0	2	1.7	3.60
EB	8	F	26.2	1	0.2	0.22

Note: Qualitative assessment of cerebrospinal fluid IgG antibodies against *B. burgdorferi*: positive result ≥5.50 AU/mL; borderline result between 4.50 AU/mL and 5.50 AU/mL; negative result <4.50 AU/mL. Qualitative assessment of cerebrospinal fluid IgM antibodies against *B. burgdorferi*: positive result ≥3.50 AU/mL; borderline result between 2.50 AU/mL and 3.50 AU/mL; negative result <2.50 AU/mL.

The degree of facial nerve damage was assessed according to the scale presented above in the two groups. The first group consisted of patients seropositive for *B. burgdorferi*, the second group included rest of the patients.

Analysis of Figs. 2 and 3 indicates that the degree of facial nerve function damage in a confirmed Lyme disease group was significantly heavier, while proportion of mild paralysis was significantly higher in a group of patients with no serologic confirmation of Lyme disease.

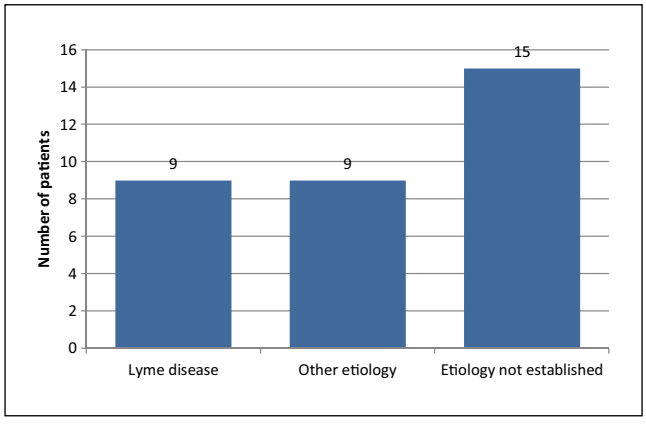

Fig. 1 – Etiology of facial nerve paralysis. First column includes patients with elevated serum antibody titers against *B. burgdorferi*.

5. Discussion

Children who were included in the study were residents of the north-eastern Poland. This region is considered as a Lyme disease endemic area. Seasonal incidence of Lyme disease is also characteristic. The vast majority of facial nerve palsy with elevated serum antibodies against *B. burgdorferi* observed in the study population was reported in the summer–autumn period, which is confirmed by observations of other authors.[9,10] Objective clinical assessment of the degree of facial nerve function damage, from a discrete paresis to complete paralysis, may be presented in a 6-point House–Brackmann (HB) scale.[11]

In this work, a scale created by the authors was adopted, which includes also corneal reflex and lack of patients meeting criteria of 1st and 5th HB stage of facial nerve damage in the study group. HB scale is a descriptive scale, which prevents scoring of individual symptoms of facial nerve palsy. The extent of diagnostic tests performed results from the assumed etiology of facial nerve paralysis. First, infectious causes should be excluded by performance of cultures and tests for serum *B. burgdorferi* antibodies.

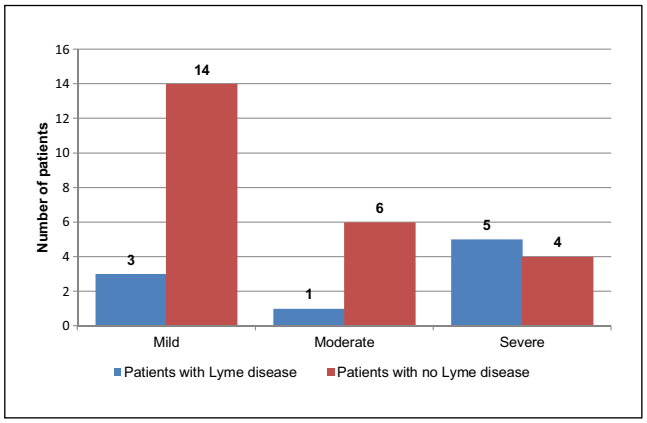

Fig. 2 – Number of patients in relation to the degree of facial nerve damage and presence of serum *B. burgdorferi*-specific antibodies.

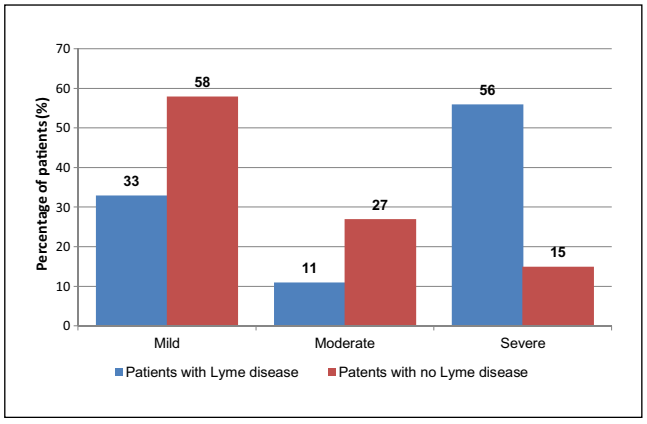

Fig. 3 – Percentage of the degree of facial nerve damage in groups by serum antibody titers.

In the analyzed material, elevated *B. burgdorferi*-specific IgM antibody titers were found in nine children, which constitutes 27.3% of the study group. Dąbrowska et al.[9] in their work have reported a lower proportion (18.9%) of patients with peripheral facial nerve paralysis, in whom elevated serum *B. burgdorferi* antibody titers were found. Similar to our results is the one presented by Dudzińska and Stettner[10] (22%). In the case of elevated serum antibodies, in accordance with recommendations of the European Federation of Neurological Societies (EFNS), with patients consent, lumbar puncture is performed in order to exclude neuroborreliosis. According to EFNS guidelines, for definite Lyme neuroborreliosis three of the following criteria should be met: (1) clinical symptoms, (2) CSF pleocytosis, and (3) *B. burgdorferi*-specific antibodies produced intrathecally. Possible diagnosis can be made when two of the following criteria are fulfilled. In the analyzed group, in two cases definite and in another two cases possible Lyme neuroborreliosis was diagnosed. Our patients with Lyme neuroborreliosis constituted 12.1% of the study group, which is more than twice the proportion presented by Dąbrowska et al.[9] (approximately 5.8%).

Numerous authors highlight the occurrence of serological window. It is the immunological phenomenon present in many infections that consist of a time delay in the appearance of specific antibodies against the infectious agent and associated symptoms. Thus, it may be assumed that the number of patients with Lyme neuroborreliosis is actually higher in the study group than it was initially found.[9,10,12]

6. Conclusions

1. The degree of motor function damage of the facial nerve in the course of peripheral palsy is dependent on the cause and in cases resulting from *B. burgdorferi* infection it might be more severe and require other treatment than that in Bell's palsy.
2. All patients with peripheral facial nerve paralysis should have serum *B. burgdorferi*-specific IgG and IgM antibodies determined.
3. Despite increasingly more precise diagnostic methods, in many cases the cause of facial nerve palsy remains unknown.

Conflict of interest

None declared.

REFERENCES

1. Bradley W, Daroff R, Fenichel G. *Neurologia w praktyce klinicznej. [Neurology in Clinical Practice]*. Lublin: Czelej; 2006:60–91.
2. FitzGerald MJ, Gruener G, Mtui E. *Neuroanatomia. [Neuroanatomy]*. Wrocław: Elsevier Urban & Partner; 2007: 128–144.
3. Rowland LP. *Neurologia Merritta. [Neurology]*. Wrocław: Elsevier Urban & Partner; 2005:101–132.
4. Oweaki M, Kozubski W. Obraz kliniczny neuroboreliozy [Clinical spectrum of neuroboreliosis]. *Wiad Lek*. 2007; 60(3/4):167–170.
5. Okurowska-Zawada B, Kułak W, Olchowik B, et al. Porażenie nerwu twarzowego w praktyce lekarza pediatry [Facial nerve palsy in pediatric practice]. *Klin Pediatr*. 2010;18(1): 14–19.
6. Szapłyko W, Chmielik J, Michałowicz R. Obwodowe porażenie nerwu twarzowego u dzieci – problemy diagnostyczne i lecznicze [Peripheral facial palsy in children – diagnostic and therapeutic problems]. *Klin Pediatr*. 1994;2(3):43–47.
7. Zajkowska JM, Hermanowska-Szpakowicz T, Grygorczyk S, et al. Neuroborelioza [Neuroborreliosis]. *Pol Przegl Neurol*. 2006;2(2):13–21.
8. Zajkowska JM, Pancewicz SA, Grygorczuk S, et al. Neuroborelioza – wybrane aspekty patogenezy, diagnostyki i leczenia [Neuroborreliosis – selected aspects of pathogenesis, diagnosis and treatment]. *Pol Merk Lek*. 2008;24(143):453–457.
9. Dąbrowska J, Sendrowski K, Olchowik B, et al. Neuroborelioza jako częsta przyczyna porażenia nerwu twarzowego u dzieci. Analiza retrospektywna z lat

2004–2010 [Neuroborreliosis as a frequent cause of peripheral facial nerve palsy in children. Retrospective analysis of the years 2004–2010]. *Neurol Dziec.* 2011;20(41):55–63.
10. Dudzińska M, Stettner D. Manifestacje kliniczne neuroboreliozy w materiale Oddziału Neurologii Dziecięcej w Chorzowie [Clinical manifestations of neuroborreliosis in the material of the Department of Child Neurology in Chorzów]. *Neurol Dziec.* 2011;20(40):27–33.
11. Kuczkowski J, Narożny W, Brzozowski W, et al. Zaburzenie czynności nerwu twarzowego w ostrym zapaleniu ucha środkowego [Facial nerve disorders in acute otits media]. *Ann Acad Med Gedan.* 2010;40(3):55–63.
12. Waespe N, Steffen I, Heininger U. Etiology of aseptic meningitis, peripheral facial nerve palsy and a combination of both in children. *Pediatr Infect Dis J.* 2010; 29(5):453–456.