Original Article

Patients with post-polio syndrome are more likely to have subclinical involvement as compared to polio survivors without new symptoms

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Abstract

Background: Post-polio syndrome (PPS) is a condition that affects polio survivors decades after recovery from an initial acute attack. It is a well-known entity that limbs thought to be nonaffected by polio survivors commonly demonstrate electromyography (EMG) evidence of prior polio. Although the diagnosis of PPS requires a remote history of acute paralytic polio, clinically unapparent damage caused by poliovirus can be associated with PPS later in life. **Objective:** To evaluate EMG abnormalities and late progressive symptoms in limbs thought to be nonaffected by polio survivors, in order to determine the prevalence of subclinical motor neuron involvement in those fulfilling criteria for PPS comparing to those without such symptoms. **Materials and Methods:** Clinical and EMG findings of 464 limbs in 116 polio survivors who had been admitted to our clinic were analyzed. Affection of the limbs by polio was classified based on the patient's self-report on remote weakness during the acute phase of poliomyelitis, muscle strength measured by manual muscle testing, and four-limb needle EMG. **Results:** Seventy-six of the patients (65.5%) met the criteria of PPS. Needle EMG studies revealed subclinical involvement in 122 out of 293 (42%) limbs with no history of remote weakness during the acute phase of poliomyelitis. Prevalence of subclinical involvement was found 47% in polio survivors who met the criteria of PPS compared to 33% in those without PPS ($P = 0.013$). Among the limbs that had developed new weakness in PPS patients, 33.5% had subclinical involvement. **Discussion and Conclusion:** Subclinical involvement is common in limbs thought to be nonaffected by polio survivors, and this is especially present in those fulfilling criteria for PPS. New muscle weakness may develop in apparently nonaffected, subclinically involved muscles. Thus we believe that four-limb EMG studies should be performed in all polio survivors, especially in those with the symptoms of PPS.

Key Words

Electromyography, poliomyelitis, post-polio syndrome

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Introduction

Acute paralytic poliomyelitis is an acute viral illness caused by the neurotropic poliovirus. Invasion of motor neurons by the virus causes dysfunction or cell death, which results in flaccid weakness and muscle atrophy, typically with an asymmetric distribution.[1] Earlier studies showed that once polio virus gained entrance to the central nervous system, there was usually widespread involvement with up to 95% of the anterior horn cells infected.[2] After the acute period, patients usually show either a full or partial recovery of strength, as a result of collateral reinnervation or "sprouting" from the surviving motor units. The degree of resultant paralysis or paresis is determined by the proportion of infected cells that survive.[1,3] Although paresis is an obvious clinical marker of previous

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paralytic polio infection, needle electromyography (EMG) has an important diagnostic role, either in documenting or excluding motor neuron involvement compatible with previous paralytic polio. In most cases with a history of paralytic polio, an EMG study detects different degrees of chronic neurogenic EMG findings in different muscles, usually with an asymmetric distribution. This is also true for the patients who had initial paralysis or weakness and experienced complete clinical recovery. It is well-known clinically that limbs thought to be nonaffected by polio survivors commonly demonstrate EMG evidence of involvement from the prior polio.[4-6]

Because of the success of poliovirus vaccine, acute poliomyelitis is extremely rare today. In recent years however, attention has been focused on the new musculoskeletal and neuromuscular symptoms reported several decades after the paralytic poliomyelitis infection; what is now known as post-polio syndrome (PPS).[7] Many polio survivors now seek assistance for this new symptomatology including new onset of weakness, fatigue, and pain. The diagnosis of PPS was made, if the patient has met the well-established clinical criteria.[8] Although no objective test is available that can reliably and specifically diagnose PPS, needle EMG is helpful to document the evidence of motor neuron involvement or to determine or exclude other neurologic disorders that might mimic the new symptoms of PPS.

Although the diagnosis of PPS traditionally has required a remote history of acute paralytic polio, significant, but clinically unapparent damage caused by poliovirus can be associated with PPS later in life.[9-11] This phenomenon has become more widely appreciated in recent years.

We have been systematically examining all patients with a history of paralytic poliomyelitis who admit to our post-polio clinic for the last 15 years. All patients answer a detailed questionnaire concerning their original illness and subsequent sequel and new neuromuscular and musculoskeletal complaints. Detailed physical examination including manual muscle testing of upper and lower limb muscle groups are performed in all of them. We routinely perform four-limb needle EMG studies to all of the polio patients at least once, to confirm lower motor neuron involvement consistent with previous poliomyelitis, to determine degree and extent of motor neuron loss, to identify or exclude other conditions that may cause the similar symptoms of PPS, and to detect concomitant nerve or muscle disorders. The diagnosis of PPS is made using the well-established criteria, based on the new neuromuscular and musculoskeletal complaints.[7] Analyses of this collected data allowed us to evaluate the prevalence of subclinical motor neuron involvement and late progressive symptoms in limbs that were considered to be nonaffected by patients with PPS.

Materials and Methods

We retrospectively evaluated clinical and electromyographic data of 152 patients with a history of paralytic poliomyelitis. Thirty-six patients who had concomitant neurological, rheumatologic, cardiovascular, or thyroid disorders that could cause symptoms of pain and weakness were excluded. Remaining 116 patients (74 women, 42 men, age between 19 and 65 years, with a mean age of 34.6 ± 14.8) were included in the analysis.

Medical reports of the patients concerning their original illness and subsequent sequel and new neuromuscular and musculoskeletal complaints, results of manual muscle testing of selected upper and lower limb muscle groups (knee extension, ankle dorsiflexion, ankle plantar flexion, elbow flexion, and shoulder abduction), and findings of needle EMG studies in selected proximal and distal muscles (quadriceps, tibialis anterior, gastrosoleus, biceps, and deltoid) in all four limbs were analyzed retrospectively.

Needle EMG investigations had been performed by an experienced electromyographer using concentric needle electrode. Analysis of spontaneous activity at rest had been assessed visually. Motor unit potential (MUP) analysis had been performed automatically on approximately 20 different MUPs with multi-MUP analysis program.[12] Results had been evaluated in relation to reference values described previously.[13] Quantitative analysis had not been made in the muscles in which chronic neurogenic changes were prominent by visual assessment. The interference pattern at strong voluntary contraction had been assessed visually. Confirmation of prior poliomyelitis involving motor neurons had been made, if EMG study had detected evidence of chronic denervation and ongoing reinnervation in the muscles under study. This typically includes the presence of high amplitude, long duration MUPs with reduced interference pattern.

Affection of the limbs by polio was first classified into "affected" and "nonaffected" based on the presence of remote weakness during the acute phase of poliomyelitis [Table 1]. History of remote weakness with variable recovery was defined by the patient's self-report. All "affected" limbs had decreased strength and/or atrophy and EMG evidence of prior poliomyelitis. "Nonaffected" limbs were further classified based on the muscle strength measured by manual muscle testing and needle EMG study [Table 1]. The "nonaffected" limbs with normal strength and a normal EMG were classified as "normal". The "nonaffected" limbs with normal/slightly decreased strength and an EMG evidence of prior polio were classified as "subclinically involved".

The diagnosis of PPS was made based on the well-established criteria, based on the new neuromuscular and musculoskeletal complaints.[7] Complaints of "new muscle weakness" in the limbs were defined by the patients' self-report.

Table 1: Classification of the limbs based on the patient's self-report on remote weakness during the acute phase of poliomyelitis, muscle strength, and needle EMG study

Measure	History of remote weakness with variable recovery		
	(+) Affected limbs	(–) Nonaffected limbs	
		Normal limbs	Subclinically involved limbs
Muscle strength	Decreased/atrophic	Normal	Normal/slightly decreased
Needle EMG	Prior polio	Normal	Prior polio

EMG = Electromyography

Statistical analyses were performed with the 15.0 Statistical Package for Social Sciences (SPSS). Frequency analyses were applied for all the data. Rates of subclinical involvement were calculated as the percentages of the limbs that were classified as "nonaffected". Prevalence of subclinical involvement in patients diagnosed as PPS with those without PPS was compared using simple cross tabulations and Pearson's chi-square test. *P* - value below 0.05 was considered to indicate statistical significance.

Results

Among the total 464 limbs investigated, 171 (37%) limbs were classified as "affected" and remaining 293 limbs (63%) were classified as "nonaffected" [Table 2]. Needle EMG studies revealed subclinical involvement in 122 out of 293 (42%) limbs with no history of remote weakness during the acute phase of poliomyelitis [Table 2]. Among these limbs, 216 (74%) had normal muscle strength, while muscle strength was slightly decreased in the remaining 77 (24%) limbs.

Seventy-six patients (65.5%) met the criteria of PPS[8] (47 women and 29 men, age between 27 and 52 years, with a mean age of 32.2 ± 12.2). The average duration until the onset of new PPS symptoms was 38.9 ± 15.2 years (19-46 years) and the average duration since onset of new symptoms was 13.7 ± 15.6 months (2-60 months). Among the patients with PPS, 118 out of 304 (39%) limbs were classified as "affected". In these patients, needle EMG studies revealed that 87 out of 186 "nonaffected" limbs (47%) had subclinical involvement [Table 3]. Among the non-PPS patients, subclinical involvement was found by needle EMG, in 35 out of 107 (33%) limbs that were classified as "nonaffected"[Table 3]. Percentage of the subclinically involved limbs was found to be statistically significantly higher in polio survivors diagnosed as PPS compared to those without PPS (χ^2 (2, N = 293) = 5.53, P = 0.013).

Patients diagnosed as PPS reported new weakness in 114 out of 304 limbs [Table 4]. Among the limbs that had developed new weakness, 38 (33.5%) were those with no history of remote weakness, but an EMG evidence of prior polio (subclinically involved limbs). Of these limbs, 28 (74%) had normal muscle strength, while muscle strength was slightly decreased in the remaining 10 (26%).

Discussion

The results of this study further support previous studies which found high prevalence of subclinical involvement in patients with a history of paralytic poliomyelitis.[4,9,14,15] The most important finding was that the prevalence of subclinical involvement was even higher in the patients diagnosed as PPS. Another important finding was that polio survivors might report new muscle weakness in the subclinically involved muscles with apparently normal strength.

Subclinical motor neuronal loss in seemingly nonaffected muscles in polio survivors is widely known for many decades. Previous EMG studies showed that, 21-40%, 5% of the muscles regarded as nonaffected by polio survivors had subclinical involvement on EMG examination.[9,14,15]

Table 2: Distribution of the limbs classified based on the remote weakness, muscle strength, and needle EMG study

Limbs	History of remote weakness with variable recovery			
	(+) Affected limbs	(–) Nonaffected limbs		
		Normal limbs	Subclinically involved limbs	Total
N	171	171	122	293
%	37%	58%	42%	63%

EMG = Electromyography

Table 3: Associations between the presence of PPS and subclinical involvement

Limbs			Nonaffected limbs			*P*-value
			Normal (%)	Subclinical (%)	Total (%)	
PPS	Yes	***N***	99	87	186	
		%	53	47	100	0.013
	No	***N***	72	35	107	
		%	67	33	100	
Total		***N***	171	122	293	
		%	58	42	100	

PPS = Post-polio syndrome

Table 4: Distribution of the limbs reported as having developed new weakness in PPS patients (*N* = 114)

Limbs	Affected limbs (%)	Subclinically involved limbs			Total (%)
		Normal strength (%)	Slightly decreased strength (%)	Total (%)	
N	76	28	10	38	114
%	66.5	74	26	33.5	100

PPS = Post-polio syndrome

Similar to our study, affection of a limb during the acute stage of poliomyelitis has been determined by the patient's self-report, which makes it highly subjective and difficult to verify. As the acute illness occurred decades earlier during infancy or childhood, memory recall and documentation can be patchy. It is even possible for patients to have subtle weakness which they are unaware of, as a limb to them may seem normal through their growth and development yet demonstrate clinical effects. In this case, extensive needle EMG studies play important role in detecting the presence of subclinically involved muscles. Prevalence of subclinical involvement (42%) among our patients with prior paralytic poliomyelitis was found to be higher than those found in the previous studies. A possible explanation for this might be that we performed more extensive EMG studies including all the four limbs. More advanced EMG methods like macro EMG would probably reveal even larger percentage of subclinical involvement in seemingly nonaffected limbs.

In our study, manual muscle testing did not detect significant weakness in the subclinically involved muscles. Among the limbs with subclinical involvement, 74% had normal muscle strength, while muscle strength was slightly decreased in the remaining 26%. Such a finding is inconsistent with a previous study that has reported manual muscle testing as the best clinical determinant of subclinical polio in nonaffected limbs.[15] This study concluded

that when an EMG study is not available or practical, physicians should use a thorough manual muscle testing to evaluate weakness and atrophy. However, the findings of the current study suggest that manual muscle testing may not be adequate for evaluation of subclinical involvement. Evaluation of muscle strength by dynamometer or by isokinetic devices might have opportunity to assess slight decreases in the muscle strength.

In the current study, we found even higher prevalence of subclinical involvement in polio survivors diagnosed as PPS (47%), as compared to those without PPS (33%). This finding supports the previous reports that the patients who had more severe and widespread involvement initially and whose original losses were largely regained during the recovery period are most at risk or most likely develop PPS.[7,16,17] These risk indicators are related to the number of motor neurons that were lost and the subsequent degree of adaptation resulting in increased motor unit size.[16,17]

This study also showed that not only the affected limbs, but also the limbs with subclinical involvement may develop new muscle weakness and neuromuscular symptoms later in life. We found that among the patients diagnosed with PPS, 35.5% reported new muscle weakness in the limbs with no history of remote weakness. More strikingly, they reported new muscle weakness in 33.5% of the limbs with subclinical polio. These findings suggest that the muscles affected by polio virus are vulnerable for later functional impairment, regardless of whether or not they achieved complete recovery after the acute stage of poliomyelitis. Although the diagnosis of PPS traditionally has required a remote history of acute paralytic polio, it is now beginning to be appreciated that the syndrome may be diagnosed in individuals who had no clear history of paralytic disease.[9-11] Halstead and Silver described four cases of PPS in individuals not previously recognized to have had paralytic polio. The EMG studies revealed widespread changes compatible with polio virus involvement of anterior horn cells supplying muscles in all four limbs. Based on the history, physical examination, and EMG, and in the absence of other conditions that would cause their symptoms, the cases were diagnosed as PPS. The authors explained this phenomenon, by sufficient polio virus invasion of the central nervous system to put the nonparalytic patients at risk for PPS later in life. They assumed that, with just the right amount of neuronal loss distributed in various muscles and limbs, one could undergo fairly extensive anterior horn cell damage with no clinical paralysis.[11] Considering the fact that PPS is basically a disorder associated with motor neuron and muscle overuse, it has been suggested that the critical amount of initial anterior horn cell injury may vary from individual-to-individual depending on the intensity and frequency of muscular activity over many years.[11]

There are some limitations and weaknesses of this study. Absence of an objective clinical parameter to define whether the limb was affected by polio and complaints of new weakness is the major limitation. Another weakness of the study is that, given the referral nature of their population and the retrospective design, the generalizability of the results is limited.

In summary, needle EMG studies, when utilized appropriately are the most reasonable standard of determining the muscles affected by polio virus infection. Detection of the presence of subclinically involved muscles would be important in determining management strategies, because new muscle weakness may develop in apparently nonaffected, subclinically involved muscles. Thus we believe that extensive needle EMG testing using a standard sampling technique that evaluates one or more muscles innervated by each myotome in all four limbs should be performed at least once in all polio survivors. The findings of this study also support the suggestion that the emphasis on a history of paralytic polio as part of the PPS criteria can be misleading and confusing, thus the diagnostic criteria for PPS should be modified.[11]

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Conflicts of interest
There are no conflicts of interest.

References

1. Bodian D. Emerging concept of poliomyelitis infection. Science 1955;122:105-8.
2. Bodian D. Histopathological basis of clinical findings in poliomyelitis. Am J Med 1949;6:563-78.
3. Bodian D. The virus, the nerve cell, and paralysis; a study of experimental poliomyelitis in 190 the spinal cord. Bull Johns Hopkins Hosp 1948;83:1-107.
4. Agre JC, Rodriquez AA, Tafel JA. Late effects of polio: Critical review of the literature on neuromuscular function. Arch Phys Med Rehabil 1991;72:923-31.
5. Sharrard WJ. The distribution of the permanent paralysis in the lower limb in poliomyelitis; a clinical and pathological study. J Bone Joint Surg Br 1955;37-B:540-58.
6. Stalberg E, Grimby G. Dynamic electromyography and muscle biopsy changes in a 4 year follow- up: Study of patients with a history of polio. Muscle Nerve 1995;18:699-707.
7. Halstead LS, Rossi CD. Post-polio syndrome: Clinical experience with 132 consecutive outpatients. Birth Defects Orig Artic Ser 1987;23:13-26.
8. March of Dimes. March of Dimes International Conference on Post Polio Syndrome. Identifying Best Practices in Diagnosis and Care. White Plains, NY March of Dimes; 2000.
9. Gandevia SC, Allen GM, Middleton J. Post-polio syndrome: Assessments, pathophysiology and progression. Disabil Rehabil 2000;22:38-42.
10. Bruno RL. Paralytic vs. "nonparalytic" polio: Distinction without a difference? Am J Phys Med Rehabil 2000;79:4-12.
11. Halstead LS, Silver JK. Nonparalytic polio and postpolio syndrome. Am J Phys Med Rehabil 2000;79:13-8.
12. Stalberg E, Nandedkar SD, Sanders DB, Falck B. Quantitative motor unit potential analysis. J Clin Neurophysiol 1996;13: 401-22.
13. Bischoff C, Stalberg E, Falck B, Eeg-Olofsson KE. Reference values of motor unit action potentials obtained with multi-MUAP analysis. Muscle Nerve 1994;17:842-51.
14. Gawne AC, Pham BT, Halstead LS. Electrodiagnostic findings in 108 consecutive patients referred to a post-polio clinic. The value of routine electrodiagnostic studies. Ann N Y Acad Sci 1995;753:383-5.
15. Mayo-Ford C. Clinical predictors of electromyographic findings of remote polio in unaffected limbs of adults with a history of acute paralytic poliomyelitis. Post-Polio Health 2006;22:1-2.
16. Klingman J, Chui H, Corgiat M, Perry J. Functional recovery. A major risk factor for the development of postpoliomyelitis muscular atrophy. Arch Neurol 1988;45:645-7.
17. Trojan DA, Cashman NR, Shapiro S, Tansey CM, Esdaile JM. Predictive factors for post-poliomyelitis syndrome. Arch Phys Med Rehabil 1994;75:770-7.