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Late Effects of Poliomyelitis

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ABSTRACT. Cosgrove JL, Alexander MA, Kitts EL, Swan BE, Klein MJ, Bauer RE: Late effects of poliomyelitis. Arch Phys Med Rehabil 68:4–7, 1987.

• Muscular atrophy and decreased functional abilities are recognized as late complications of poliomyelitis. This study sought to more clearly define late-onset, postpolio muscular weakness—age of onset, symptoms, and severity—and to determine whether people might benefit from environmental modification, respiratory aids, and orthoses. A total of 183 postpolio patients were examined by a physician and completed a questionnaire on their ambulatory status and related musculoskeletal and respiratory symptoms. Of those participating in the study, 154 claimed to be experiencing late deterioration in strength. The syndrome included decreased endurance, more limited ambulation, and increased weakness in the previously affected limb/s. For those describing late-onset weakness, average ages were determined for the onset of polio (8.3 years), the onset of postpolio muscular weakness (42.3 years), and the latent period of stable functioning (34.8 years). Patients claimed to have experienced a new, lower level of strength for an average of 4.7 years. All 33 patients who had undergone muscle transfer surgery were experiencing late-onset weakness in that extremity. Fasciculations (51%) and long-bone fractures secondary to falls (21%) were common sleep disturbances occurred frequently (31%) even in those without prior bulbar involvement. Upper motor neuron signs were present in only one case. Bracing was rare (16%) and the braces used were usually old. The subjects did not report a steadily or rapidly progressive decline, but rather described a steplike decrement with long plateaus. The population described losses in strength that had significant bearing on functional status and general health. Many patients had not sought routine care in years, let alone special evaluation of neuromuscular function. Perhaps most alarming was the large number of sleep complaints. Clearly, many postpolio patients might benefit from closer medical supervision, with orthotic and pulmonary intervention being commonly required. On completion of this study, 31% of the patients had new orthoses prescribed and 18% were referred for further pulmonary evaluation.

KEY WORDS: Fasciculations; Fractures; Orthotic devices; Paralysis; Poliomyelitis

The syndrome of late muscular weakness with antecedent poliomyelitis was first documented by Charcot in 1875. Since that time many authors have commented upon this phenomenon.[1-4,8,11,14,16,18,22] Though epidemiologic data is not abundant, a prevalence rate of 22% has been estimated.[3]

Myriad neuromusculoskeletal complications have been associated with the late effects of poliomyelitis. Late-onset weakness usually has been described in muscles that were previously atrophied.[9,14,16,18] However, the contralateral, uninvolved limb has often been involved as well.[2,8,16] Weakness has been generally perceived as slowly progressive. However, it has been suggested that a subgroup of more active individuals may have a more rapidly progressive form of the syndrome.[4] Two deaths have been reported secondary to late motor neuron dysfunction.[16,18] Joint pain, joint instability, and long-bone fractures are also identified as late complications of polio.[1,2,8,11,16] Yet the relative frequency of these conditions is not known. Muscle fasciculations have been described,[1,2,9,11,15,16,22,23] but often no distinction is made between true fasciculations and contraction fasciculations.[11] Muscle cramps, myalgia and fasciculations, commonly associated with late muscular weakness, are often found in old polio patients without new weakness.[6,11] Upper motor neuron signs have been rare, having been present in only six of the 100 cases reported between 1969 and 1984.[1,2,6,11,14,16,18]

Respiratory insufficiency also has been implicated as a late complication of previous bulbar poliomyelitis.[7,9,12,20] Formal sleep studies have revealed the presence of central sleep apnea in patients with prior bulbar involvement who had been weaned from respiratory support.[7,11,18] An increase in the use of respiratory support of between 25%[8] and 80%[3] from the time of optimal neurologic functioning has been reported.

Despite the number of case reports and descriptions of late-onset, postpolio muscular weakness (LOPPMW) in the literature, no large series of patients has been examined systematically. The intent of this study is to describe the syndrome of LOPPMW more completely and the role of continuing medical intervention.

MATERIALS AND METHODS

The subjects were 183 volunteers with previous paralytic poliomyelitis who attended one of four conferences on the late effects of poliomyelitis. They were examined by one of the six authors. At that time a detailed questionnaire was completed regarding demographic data, initial presentation of polio, extent of current involvement, functional areas of impairment, use of orthotics, previous orthopedic procedures, frequency and severity of falling, fasciculations, initial bulbar involvement, respiratory difficulties, use of ventilatory support, and the quality of sleep.

Late-onset weakness and decreased endurance were ascertained through the questionnaire and the physician's history.

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In both the questionnaire and the examination an effort was made to separate true fasciculations from contraction fasciculations. True fasciculations were accepted as muscle twitching that occurs at rest, not under voluntary control. Bulbar involvement was defined as having difficulty swallowing or requiring respiratory assistance of a rocking bed, supplemental oxygen, iron lung, or any artificial means of ventilation. Sleep disturbances were determined by patient report. Prolonged apneic spells during sleep, loud snoring, excessive daytime sleepiness, early morning headache, and waking at night gasping for air were considered significant sleep disturbances.

RESULTS

Of the 183 patients in the study group, 29 were experiencing no late effects of poliomyelitis. Of the 154 with late weakness there were 58 men and 96 women. The following data concerns those 154 patients, who were experiencing the syndrome of LOPPMW.

The initial onset of paralytic polio occurred between 2 months and 33 years of age (mean, 8.3 years). All 154 patients reported stable neurologic status during adulthood. Late-onset weakness began between years 18 and 74 (mean, 42.3 years). The latent period ranged from 15 to 72 years (mean, 34.8 years). The patients had noticed diminished function for 1 to 40 years (mean, 4.7 years). Most patients had difficulty giving an exact date for the late onset of weakness. Those with minor residual deficits and very active lifestyles could, however, pinpoint an exact month and year of the onset of their late symptoms. In no subgroup was a steady downhill progression seen. All 154 were experiencing late-onset weakness in muscles that were clinically affected by polio previously. Seventy patients (45%) noted weakness in muscles that previously they had considered normal. The single most common complaint was decreased endurance (153 of 154). Increased difficulty with stair climbing was almost universal (152 of 154).

Bracing was found only in 25 of 154 (16%). Most braces were old, with an average age of 15.3 years. Only 30 patients (19%) had begun to use a cane or crutches. Six patients (4%) had ceased to ambulate and were wheelchair bound. The use of braces or alteration in bracing was recommended for 56 of the 183 patients examined. Fifty of these 56 were complaining of late weakness. Also, 13 of the 154 patients were advised to begin using either a cane or crutches. Though scoliosis was present in 30 of 154 patients, only three were braced because of deformity. No changes in orthoses were made with these patients.

Seventy-eight patients had undergone orthopedic surgery for the treatment of polio-related disorders. Of those, 33 had muscle transfer surgery. All of those patients experienced late-onset weakness in that extremity.

Falling was a frequent complaint. Falls had caused long-bone fractures in 32 patients. Only eight of these 30 patients with fractures had begun to use ambulatory aids or bracing.

Though 76 patients claimed on the questionnaire to have had fasciculations, only 35 could be verified on physical examination. Three other patients denied having fasciculations, which were nevertheless noted on physical examination. In total there were 79 of the 154 patients who either complained of or who were observed to have fasciculations. All patients reported increased fasciculation after exhaustive exercise (usually stair climbing). Fasciculations were noted quite often in muscles that were not clinically weak. In fact, eight of the 29 patients without LOPPMW were noted on physical examination to have fasciculations.

In one patient, there was noted a positive Babinski sign. In all others, no upper motor neuron signs of Babinski, Hoffmann or clonus were found on examination. In that patient, the diagnosis of multiple sclerosis has not been ruled out.

Nineteen patients complained of bulbar involvement as a part of the initial presentation of poliomyelitis. Twenty-six patients complained of late occurrence of dysphagia and dysphonia. Only one patient was on supplemental oxygen or respiratory assistance (chest cuirass at night time only). None required positive pressure ventilatory support.

However, 47 of 154 patients with weakness and one of 29 patients without weakness complained of a sleep disturbance. Nine of the 19 patients with previous bulbar involvement had sleep complaints. Four patients complained of apneic spells, six complained of loud snoring, ten complained of gasping for air, and 27 complained of early morning headache. Interestingly, 19 of the patients with headache had sought the advice of a physician and had been diagnosed as having "allergies." None had experienced relief with decongestants or antihistamines. Sleep apnea had been diagnosed in the one patient mentioned above, utilizing the chest cuirass. All 19 patients with bulbar involvement initially were advised to undergo baseline pulmonary function tests. Those patients with reports of apnea or gasping were referred to a pulmonologist for further evaluation.

DISCUSSION

Remarkably consistent throughout previous literature and corroborated by this study is the time course of stable neurologic functioning. Between the third and fourth decade after onset of poliomyelitis is the average age of presentation for LOPPMW.[1,2,3,8,11,14,16,18,22] Our findings indicate a mean latent period of 34.8 years. Yet the wide range in years of stable functioning (15 to 72 years) indicates that the time course of the onset of the weakness can be extremely variable.

The progressive nature of this syndrome is difficult to assess. Often interpretation of the data is complicated by increased age and weight. The patients in this study had noted the presence of late-onset weakness, as evidenced by diminished ability to ambulate, for an average of almost five years. As in the previous literature, they described a slow, sporadic decline in strength, with plateaus of stability.[1,2,14,15,16,22] Dalakas and colleagues[4] were able to distinguish between two separate subgroups of LOPPMW (ie, progressive and nonprogressive), but we were unable to do so. Patients with more minor residual deficits, however, were much more exact in describing the onset of their late symptoms.

Systematic evaluation of orthoses in postpolio patients had not been performed by previous investigators. Few of our patients used braces or walking aids and those that were used were quite old. Few had sought medical advice concerning ambulation, despite the universal impression of increased weakness (100%) and the high incidence of falls resulting in broken bones (21%). The explanation of this behavior appears

to lie deeper than simple denial of disability. More likely it is attributable to the medical, family, and social influences of their early rehabilitation experience. The patients reported that their ritual of exercise was religiously followed, with the goal being to rid themselves of braces. A return to braces, then, may be viewed as failure. Although initial resistance to bracing was almost unanimous, with the aid of counseling and through demonstration, prescriptions were written for 56 orthoses. Another 13 patients were advised to use walking aids.

Scoliosis, though present in 20% of our population, did not represent a major clinical problem. Only three of the patients were braced and none appreciated any progression of their curvatures. Furthermore, no new spinal orthoses were prescribed. The absence of progression in scoliotic curves is difficult to reconcile with more marked changes in the appendicular musculature. Perhaps the decrease in muscle strength tended to be symmetrical, thus causing no dramatic spinal changes. Since routine pulmonary function tests were not done, we cannot comment on the presence or absence of restrictive lung disease.

The long-term results of muscle transfer surgery have not been studied by previous authors. In our population, 21% had undergone muscle transfer surgery, all of whom were experiencing LOPPMW in the muscle group/s that had been transferred. It thus appears that muscle transfers are best considered as a temporizing measure.

Fasciculations in LOPPMW have been noted by many investigators.[1,2,6,9,11,15,16,22] We attempted to separate contraction fasciculations from true fasciculations, both in the questionnaire and during the examination. Only fasciculations at rest were counted as true fasciculations. Fasciculations were quite common, being observed in 38 patients and reported by 41 others. However, fasciculations frequently occurred in muscles that were not clinically weak. Furthermore, many patients reported having fasciculations "all my life." Though the significance of fasciculations remains unclear, they do not appear to portend imminent decline in muscle strength.

In only one patient was an upper motor neuron sign of Babinski, Hoffmann or clonus elicited. In other studies upper motor neuron signs have also been rare, having been present in only six of 100 cases reported.[1-4,8,11,14,16,18,22,23] Their presence, however, has led some authors to suggest that LOPPMW may represent a forme fruste of amyotrophic lateral sclerosis (ALS).[16,18] The clinical data presented above, specifically the rate of progression and the relatively limited areas of involvement, clearly indicate that ALS and LOPPMW represent different syndromes.

Bulbar involvement during the initial illness was not as prevalent in our population (19 of 154) as in other studies.[3,8] Furthermore, only one patient was requiring any type of ventilatory support (<1%), compared with 5% and 9% as noted by Halstead and Rossi[8] and Codd and colleagues,[3] respectively. Despite the apparent good respiratory health of this study population, 26 complained of dysphagia/dysphonia and 47 complained of sleep disturbances.

In contrast to the findings of Hamilton and coworkers,[9] none of the patients in our study complained of increased sensitivity to cold, decreased memory, or decreased capacity for intellectual work. The most common sleep complaint, early morning headache (27 of 47), is recognized as a symptom of hypercarbia and hypoxemia. Hypercarbia and hypoxemia due to central sleep apnea have been well-described as late complications of poliomyelitis.[7,12,20] Unlike previous cases cited in the literature, however, most of our patients denied having previous bulbar polio. One might argue that sleep complaints are quite common in the general population and that our results represent an unrelated epiphenomenon. However, the findings in 19 of 27 patients who had headache severe enough to seek medical care, none of whom experienced relief with medical management, is certainly suggestive of a central process.

CONCLUSION

Our examinations demonstrate that the late symptoms of poliomyelitis are not restricted to areas previously considered weakened by polio. Seventy of 154 patients experienced weakness in limbs that they did not feel were involved originally. Thirty-eight patients described sleep disturbances but denied ever having bulbar polio. The etiology of these symptoms remains unknown. Congenital predisposition,[5,14,17,18] recrudescence of a latent virus,[3,13,15,18] impaired cell-mediated immunity,[5] premature anterior horn cell death,[11,22] and failure of the neuromuscular junction[21] have been proposed as theories. It has also been found in postmortem examinations that subclinical diminution of the number of anterior horn cells can occur without apparent weakness.[19] Furthermore, electromyographic (EMG) results indicate that complete reinnervation can occur if more than 50% of the anterior horn cells remain, thus obscuring the extent of involvement of poliomyelitis.[10,11] Therefore, we suggest that individuals with a decreased number of viable anterior horn cells later undergo premature failure of the (remaining) motor units due to one of the mechanisms mentioned above. The result is late-onset weakness of the skeletal and bulbar musculature previously considered normal. The treating physician must therefore perform a complete physical examination, being alert for subtle evidence of weakness, and not limiting the examination to areas of obvious clinical atrophy.

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Index to Volume 67 to be Republished

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