Postpolio syndrome: Clinical and epidemiological studies
Ivanyi, B.

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Summary

Postpolio Syndrome: clinical and epidemiological studies.

Poliomyelitis anterior acuta (polio) is a viral disease with an asymptomatic or mild course in most of the affected patients. In a minority of the patients the virus invades the nervous system. When this results in muscle weakness the term paralytic poliomyelitis is used. Paralytic poliomyelitis has a characteristic course. Acute onset with subsequent muscle weakness is followed by a two-year period of recovery of muscle strength. When maximal recovery is reached, some patients are still left with residual paresis. Usually a period of relative stability of several decades follows, whereafter some patients may experience new neuromuscular complaints. When other causes of progressive neuromuscular symptoms have been excluded, this complex of symptoms is called "postpolio syndrome". The complaints mainly concern new or increasing muscle weakness, decrease in muscle size, muscle cramps, muscle and joint pains, involuntary 'twitches' of muscles, fatigue, diminished stamina and increase of disabilities.

This thesis focusses on several aspects of the postpolio syndrome that are still a matter of debate. The main objectives of this study are discussed in chapter 1.

We investigated: (1) the clinical features of the postpolio syndrome, i.e., the occurrence of the new neuromuscular complaints and the extent of progression of new muscle weakness; (2) the diagnostic means to distinguish postpolio patients from polio survivors with stable neuromuscular conditions; (3) the prevalence of new neuromuscular complaints resulting in an impaired function in Dutch polio survivors.

In chapter 2 the clinical features of the postpolio syndrome are described.

In chapter 2a we present the results of a prospective clinical study on the presentation of late neuromuscular complaints and on the time-course of new muscle weakness development in postpolio patients.

In this study 43 self-referred postpolio patients and 13 polio survivors with stable neuromuscular conditions were included. All patients completed an extensive questionnaire investigating aspects of acute poliomyelitis, the stable period following recovery, and their present health conditions. Their functional levels were assessed using questions related to 17 daily life activities. The muscle strength of 26 muscles in all four limbs of each patient was assessed by manual muscle testing and was also measured isometrically using a handheld dynamometer over a mean follow-up period of 2.1 years. The mean interval between acute poliomyelitis and complaints of new muscle weakness was 33.7 years (range 18-46). Other neuromuscular complaints such as fatigue and pain usually preceded the complaints of new muscle weakness by a few years. Most frequent complaints were general fatigue, low back ache and muscle pain (97.7%, 86% and 79.1%, respectively). Decrease in the ability to walk, to stand, and to climb stairs were the most frequently reported restrictions in daily life activities (80%, 70%, and 67.5%, respectively).
respectively). New weakness in more than one limb was reported in 63% of the patients. New muscle weakness was also experienced in limbs (32%) of which the patients could not recall whether those had been involved during acute polio. During the follow up-period, we did not find a significant decrease in muscle strength in the symptomatic patients as compared to patients without new neuromuscular complaints, despite the patients’ complaints of increasing muscle weakness.

We argued that the detection of new muscle weakness of single muscles in postpolio patients may be hampered by stable muscle strength in the majority of the muscles and is perhaps also impaired by the lack of sensitivity of the measures of assessment.

In chapter 2b a study on dysphagia in postpolio patients is described.

New muscle weakness in postpolio patients may involve bulbar muscles resulting in dysphagia. In postpolio patients, prevalence, severity and progression of new swallowing problems are as yet not sufficiently documented. In the original cohort of the 43 postpolio patients described above, 8 patients (4 men and 4 women, mean age 45.4 years) complained of dysphagia and became subject of this study.

In 6 patients dysphagia was a newly reported symptom as they had no swallowing difficulties after recovery from acute polio. In 2 patients some swallowing difficulties remained after acute polio and now these patients reported deterioration years after a period of stability. The mean interval between acute polio and onset of new swallowing difficulties was 27.1 years (range 23-45 years). The symptoms were relatively mild, no one reported complications as aspiration pneumonia. Patients were followed for 12-36 months (mean 18 months). During the follow-up period all patients reported slight worsening of dysphagia. Videofluorography, a tool to evaluate oropharyngeal function, was performed at the start of the study in all patients and at the follow-up examination in 7 of them. Initial videofluorography showed signs of slight to moderate oropharyngeal dysfunction in 6 patients. The follow-up fluorography showed unaltered findings in 6 patients and slight deterioration in 1 patient.

The prevalence of new swallowing difficulties in postpolio syndrome patients in our cohort study was 18.6%. The swallowing difficulties and the videofluorographic abnormalities were milder than reported in earlier studies. We did not find a significant loss in oropharyngeal function on videofluorography on 1-3 years follow-up.

In chapter 2c a study on sleep complaints in postpolio patients is described.

Several studies point out the occurrence of sleep complaints in postpolio patients. Since sleep-related complaints are also frequent in the general population we argued that these symptoms might not be related to the postpolio syndrome.

Therefore, we have undertaken a study to determine the prevalence of sleep complaints and complaints of sleep-disordered breathing in our cohort of the 43 postpolio patients described above. The patients received a mailed questionnaire consisting of the validated Sleep Wake Experience List and a list of 18 questions related to sleep-disordered breathing. The patients were requested to select two age and gender matched healthy
controls from their neighborhood, family or friends who also filled out the questionnaire. The response rate was 88%. The frequency of sleep-related complaints such as tiredness on waking up and during the day, headache on waking up, daytime sleepiness and restless legs, was significantly higher in the postpolio patients group as compared to the controls. Estimated sleeping time was significantly longer in postpolio patients. However, there was no significant difference in the frequency of the complaints specifically related to sleep-disordered breathing between the postpolio patients and the controls. This study shows that sleep complaints are more frequent in postpolio patients than in the general population. These results confirm that sleep complaints constitute a part of the symptom complex of the postpolio syndrome although as yet there is no satisfactory explanation for this relationship.

Conventional electrophysiological and morphological techniques do not differentiate between postpolio patients and polio survivors with stable neuromuscular conditions. Chapter 3 concerns the value of two other diagnostic measures in postpolio syndrome. In chapter 3a the value of macro electromyography (EMG) as diagnostic tool for postpolio syndrome is investigated. In polio survivors the territory of the remaining motor units may be increased as a result of collateral sprouting, a mechanism that compensates for the loss of motor units in acute polio. A decrease in motor unit size in postpolio patients may indicate a decompensation of this compensatory mechanism causing new muscle weakness. Macro EMG provides information about the size of the motor unit territory and may therefore be a tool to diagnose postpolio syndrome. We investigated the motor unit area of 5 patients with postpolio syndrome, of 6 stable patients with prior poliomyelitis, and of 5 healthy volunteers. The motor unit territory was assessed by measuring amplitudes of motor unit potentials recorded by the macro EMG techniques. Patients were followed for 11 to 20 months (mean 15.6).

The median macro MUP amplitudes in both patient groups were markedly increased (p=0.02). There was no significant difference in the initial and the repeated amplitude values between the two patient groups. During the follow-up a decline in muscle strength was documented in 3 individual postpolio syndrome patients. These three patients had significantly increased initial median macro MUP amplitudes compared to stable polio patients (p=0.04), indicating a higher degree of reinnervation. On follow-up, two of these three patients showed a decrease in median macro MUP amplitudes. These findings suggest that in due course, breakdown of the oversized motor unit may play a role in the pathogenesis of the postpolio syndrome. Macro EMG follow-up examinations seem to be a promising diagnostic tool in postpolio syndrome. However, further research with larger patient and control groups is needed.

In chapter 3b the value of computed tomography (CT) of the skeletal muscles in postpolio patients is investigated. This technique enables us to map the muscle areas in
which replacement by fat has taken place. Patients whose complaints of new muscle weakness involved one or both lower limbs were selected from the original cohort. The CT scan evaluation included 24 muscles at the lumbar region, pelvic girdle and lower limbs. Muscle strength of the lower limbs was assessed by manual muscle testing. Thirty-two postpolio patients aged 33 to 59 years (mean 47.2 years) complained of new muscle weakness and comprised the symptomatic patients group. Thirteen polio survivors with stable neuromuscular conditions aged 35 to 60 years (mean 42.3 years) comprised the asymptomatic patients group.

Significantly more CT scan abnormalities were found in the symptomatic patient group, whereas no significant difference in muscle strength between the symptomatic and asymptomatic patients occurred. However, we found no specific CT scan features to discriminate between symptomatic and asymptomatic patients. Thus, CT scanning of the skeletal musculature has proven not to be a diagnostic tool for postpolio syndrome. However, this investigative tool appeared to be valuable in the assessment of the neuromuscular status in individual patients. CT scan abnormalities were found in 33.8% of the compound muscles with normal strength on manual muscle testing. Therefore, it provides information about the skeletal muscles in addition to manual muscle testing and may contribute to seeking an explanation for the experience of new muscle weakness in an apparently 'healthy' extremity.

In chapter 4 a study on epidemiology of late polio sequelae in the Netherlands is described.

New neuromuscular complaints among polio survivors have been reported by others with a frequency of 25-85% but most of these data were derived from biased studies. Besides, there are only few data on the disabilities and handicaps resulting from the new complaints. Population-based surveys of late onset polio sequelae in Dutch polio survivors have not yet been performed.

In 1995 we mailed a questionnaire to a sample (350 subjects) of 1784 registered cases of paralytic poliomyelitis from the 1956 polio outbreak in the Netherlands. The questionnaire consisted of questions about neuromuscular complaints, disabilities and handicaps during the stable period following recovery from acute polio and at present.

The response rate was 74% (233 subjects, mean age 44 years, range 39-77). The frequency of all neuromuscular complaints was significantly higher than during the stable period following recovery of polio (range p-values 0.001 to 0.004). Fifty-six percent of the cases reported an increase in functional restrictions such as running, walking and climbing stairs, and there was an increased need for adaptive measures and devices. Fifty-eight percent of the cases reported an increase in muscle weakness.

Risk factors for increased muscle weakness were the neuromuscular complaints during the stable period and the current age.

From this study we conclude that nearly 60% of a more or less representative sample of
Dutch survivors of the 1956 polio outbreak experience increasing neuromuscular complaints, including muscle weakness. New handicaps are present, as indicated by increased need for new adaptive measures and devices and by occupational disability.

Chapter 5 presents a general discussion of the results of this study, their implications for the present state of knowledge about postpolio syndrome and the prospects for future research. Many questions about the postpolio syndrome still remain unresolved. However, we are presently facing a group of patients with increasing disabilities and handicaps who are looking for medical help. Given the high frequency of postpolio syndrome in polio survivors, more research is needed to unravel its pathogenesis and to find a cure. In the meantime, it is of utmost importance to recognize the symptoms of postpolio and to offer those suffering from this disease recommendations tailored to their needs, which often implicates a drastic change in life style.
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In chapter 4, a study on the epidemiology of postpolio syndrome was described. New neurological symptoms, some at a frequency of 2% per year, were studied. There are only few data on the incidence after the acute course of the disease. Population-based surveys have not yet been performed in the UK.

In 1995, we studied a group of individuals with postpolio paralysis, paralytic poliomyelitis cases in 1950, and compared their symptoms with those of patients who have not yet been affected. The outcome was comparable to the initial frequency of all neurological symptoms reported in other studies. The cases reported an increased number of mild symptoms, and the duration of the symptoms increased with the onset. From this study, we concluded...