Postpolio syndrome: Clinical and epidemiological studies
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Chapter 5

General discussion

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Postpolio syndrome

Pathogenesis
The pathogenesis of postpolio syndrome has as yet not been elucidated. Although this was not the objective of this thesis, we have found some support for two of the currently proposed theories. It is hypothesized that new muscle weakness in postpolio patients is due to the failure of an ongoing process of remodeling the size of the motor units. Reinnervation is needed to compensate for the loss of motor neurons caused by the acute polio infection. Motor neurons gradually become oversized due to reinnervation. These large motor neurons eventually cannot meet the metabolic demands of the reinnervated muscle fibers and result in the breakdown of terminal nerve branches. It has been suggested, that new muscle weakness may develop when reinnervation finally fails and denervation takes over, resulting in decreasing motor unit size. The results of our macro EMG study support this theory as they show greatly enlarged motor units in polio survivors and a decrease in the motor unit size over time in patients with increasing muscle weakness. Nevertheless, studies with larger numbers of patients are needed before final conclusions can be made. The question still remains what precisely causes this late decompensation. Another theory focuses on the chronic overuse in polio survivors that may cause the postpolio syndrome. This theory is supported by our epidemiological data. The presence of neuromuscular complaints during the stable period after recovery from polio is a risk indicator of the late complaints of increased muscle weakness. These early neuromuscular complaints may be indicative of early overuse, causing the later decompensation. However, the conclusions on pathogenesis, based on these epidemiological data are merely speculative. Further research on the effects of physical activity on the neuromuscular system compromised by a previous polio infection is warranted. Development of animal models affected by polio may open avenues to study the pathophysiology of compensatory mechanisms following acute polio and the subsequent decompensation.

Diagnostic measures
In the early studies on postpolio patients, EMG and muscle biopsy findings were thought to be specific for the postpolio syndrome. However, this opinion was abandoned when polio survivors with stable neuromuscular conditions were investigated. Thus, conventional electrophysiological examination and morphology of the skeletal muscles failed to differentiate between postpolio patients and polio survivors with stable neuromuscular conditions. Hence, these tests are not specific for the postpolio syndrome but are indicative of previous polio infection and of the compensatory mechanisms that occur during the recovery period from polio. In this thesis we have examined the value of macro EMG and computed tomography of
the skeletal muscles as diagnostic means for the postpolio syndrome. One can conclude from the results, that macro EMG could be a diagnostic tool for postpolio patients, though further studies with larger patient numbers are still needed. When designing future studies, one has to consider that different muscles in each patient can show different macro EMG findings, depending on the extent of the damage during the acute polio, following compensatory mechanisms and the superimposed changes due to late deterioration. Macro EMG technique causes some discomfort to the patients, therefore only a limited number of muscles can be examined in each patient. Recently a non-invasive multi-electrode surface EMG technique was described to obtain information about the motor unit size allowing more extensive examination of each patient. From the results of the computed tomography study we conclude that computed tomography is not a satisfactory diagnostic tool for the postpolio syndrome. However, this investigative tool appeared to be valuable in the clinical setting for the assessment of the neuromuscular status in individual patients. Computed tomography may show fatty changes in currently symptomatic muscles, which show normal strength and had been considered as unaffected during the acute polio period.

New muscle weakness
The presence of progressive muscle weakness is considered essential for the diagnosis of the postpolio syndrome. Nevertheless, new muscle weakness is a difficult concept in this context. Complaints about progressive muscle weakness in postpolio patients can not always be confirmed by muscle strength assessment. We must realize, that the new muscle weakness is superimposed onto residual weakness that has been present since the acute polio. Therefore, unlike in healthy persons, the sole presence of paresis is not sufficient to support the diagnosis postpolio syndrome. Progression of the muscle weakness must be confirmed by serial muscle strength assessments. In case there is slow progression of the new muscle weakness, a long term follow-up is needed. The distribution of muscle weakness differs in every patient and therefore it is difficult to assess the rate of the overall progression of muscle weakness in a group of postpolio patients. In the published studies, the duration of the follow-up and the methods to calculate the overall muscle strength in a group of postpolio patients show great differences amounting to estimates of the rate of progression of the new muscle weakness of about 1% per year and to 20% reduction over a 4-5 year period. Some authors still question the progressive nature of the muscle weakness in the postpolio syndrome. To resolve the present controversy, we advocate future studies involving larger patient numbers and a longer follow-up period than in our present study which had an average follow-up of only 2.1 years. However, detection of new muscle weakness in single muscles will be hampered by stable muscle strength in the majority of the muscles. Therefore, we conclude that there is not enough ground to claim at present that the diagnosis postpolio syndrome must be confirmed by new muscle weakness on serial
muscle strength measurements. In the clinical setting, postpolio syndrome can only be referred to as a complex of new neuromuscular complaints and symptoms in survivors of acute polio. The use of the term postpoliomyelitis muscular atrophy suggesting a clinical picture distinctive of postpolio syndrome is unfounded.

**Epidemiology**

The epidemiological results given in this thesis are the first data on the late onset polio sequelae in the Netherlands. We show that postpolio syndrome is a frequent cause of increasing disabilities and handicaps in polio survivors. We therefore advocate more research into preventive and therapeutic measures for postpolio syndrome and more attention within the field of rehabilitation medicine for polio survivors.

**References**

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The progression of muscle weakness in postpolio syndrome is characterized by muscle weakness that increases over time. In the majority of affected patients, the progression of muscle weakness is gradual, with a rate of about 1% per year. However, in some patients, the rate of progression can be more rapid. To quantify the rate of progression, we calculate the overall muscle strength in a group of postpolio patients using a standardized muscle strength test. The rate of progression is then estimated by a statistical model. The rate of progression is influenced by various factors, including age, gender, and the severity of the initial polio infection. Despite the variability in rates of progression, the overall trend is for muscle weakness to increase over time.