Postpolio syndrome: Clinical and epidemiological studies
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Chapter 1

Introduction and aims of the study

Polymyositis, an acute muscle disease, is a highly communicable disease resulting from infection by poliovirus. The disease affects either men or women, and often displays symptoms of muscular weakness, fatigue, and pain. The muscle weakness is usually symmetrical and usually affects the extremities. Early weakness has been recorded in about 10-15% of the patients with paralytic polymyositis. In the spinal cord, neurons in the anterior horns are specifically affected. Therefore, paralytic poliomyelitis has the specific features of anterior horn disease, including muscle flaccid paralysis and atrophy, anhidrosis, and fasciculations. Mortality in paralytic polymyositis is due to respiratory failure and is estimated to be between 5-10 percent.

Before vaccination, spinal polio infections were the main victims during the polio epidemic. In countries where vaccination is common, small children can occur among young children that do not react bacterially. Vaccinated children and adults are also typically affected. Paralytic poliomyelitis is an example of a disease that can occur in vaccinated individuals. Typing for virulence in vaccinated for the severe poliomyelitis can be a helpful measure in terms of the risk of paralytic paralytic poliomyelitis and the severity of the disease.

In the United States, the vaccination rate has been relatively high for the past 30 years. However, the vaccination rate has decreased in recent years. The decline in vaccination rates has been attributed to several factors, including the growing concern over the potential risks associated with vaccines, public health initiatives, and the increasing use of non-vaccination options. As a result, the incidence of poliomyelitis has decreased significantly, but the disease continues to be a concern worldwide. The increasing number of cases of poliomyelitis has prompted the World Health Organization to launch a global poliomyelitis eradication initiative.
2-postpolio syndrome
Poliomyelitis anterior acuta (polio) is a highly communicable disease resulting from infection by poliovirus. A large majority of infected patients are either asymptomatic or display mild systemic symptoms. Approximately 1% of the affected patients present with a variable degree of paralysis (paralytic poliomyelitis). The muscle weakness is usually asymmetric and mainly affects the extremities. Bulbar weakness has been recorded in about 10-15% of the patients with paralytic poliomyelitis. In the spinal cord, neurons in the anterior horns are specifically affected. Therefore, paralysis in polio has the specific features of anterior horn disease, including muscle flaccid paralysis and atrophy, areflexia and fasciculations. Mortality in paralytic poliomyelitis is due to respiratory failure and its rate is estimated to be between 5-10 percent.

Before vaccination against polio, children were the main victims during the polio outbreaks. In countries where vaccination is common, small outbreaks can occur among community groups that do not practice immunization. Unvaccinated children and adults are then equally affected.

There is considerable recovery of muscle strength after the acute stage in most cases and many patients recover without residual paresis. The neurons that had only partially been damaged can regain their function. There are two mechanisms to compensate for the permanent loss of neurons. (1) Distal branching of axons of intact motor neurons (collateral sprouting); (2) Compensatory hypertrophy of the non-denervated muscle fibres.

Motor functions of patients with residual paresis can be improved in the subsequent years by additional operations, such as muscle transpositions and by the use of orthotic devices and adaptive measures. Patients reach after some years a period of stability. However, decades after paralytic poliomyelitis, polio survivors may experience new neuromuscular complaints. These complaints include new or increasing muscle weakness, muscle atrophy, muscle cramps, muscle and joint pains, fasciculations, fatigue, diminished endurance and increase of disabilities. Anecdotal reports on so-called late onset polio sequelae have been published since the beginning of this century. However increased interest for the late sequelae arose in the United States since 1980, when large groups of polio survivors of the outbreaks in the 1940-1960 period started to report new neuromuscular complaints. Major differences in the prevalence of the late onset polio sequelae, varying between 25 to 85 percent, have been reported in different studies. These differences could be attributed to differences in the selection of the study populations, but there is controversy also about the clinical manifestations. Dalakas who was the first to draw attention on the deterioration of polio survivors claimed, that there are two distinctive groups of patients. The first is manifesting new progressive muscle weakness and atrophy while the second has new neuromuscular complaints other than new muscle weakness. For the patients with confirmed new progressive muscle weakness, he introduced the term "progressive postpoliomyelitis muscular atrophy". However, distinction between patients with or without progressive muscle weakness is not
as clear as suggested by Dalakas, because the reported decrease in muscle strength can not always be confirmed by muscle strength assessment\(^1\). Ongoing research is concerned with the occurrence, rate of progression and assessment of the new muscle weakness in postpolio patients. The term "postpolio syndrome" is now more widely used to classify a complex of new neuromuscular symptoms in polio survivors\(^5\).

Considerable research effort has been addressed toward finding diagnostic tools capable of distinguishing polio survivors with stable neuromuscular conditions from those with the postpolio syndrome\(^4,7,12\). However, no diagnostic tests have yet been found for the reliable diagnosis of the postpolio syndrome.

The initial search for pathogenetic mechanisms of the late polio sequelae and the new muscle weakness was in the field of neurology\(^8\). However, questions arose gradually about the subsequent disabilities and handicaps resulting from the new clinical complaints. Consequently, these aspects attracted the attention from workers in the field of rehabilitation medicine. Researchers in this discipline shed new light also on the possible etiology of the late complaints putting forward the concept of overuse of muscles, ligaments and joints\(^10\). Nevertheless, the pathogenesis remains unsolved. It is conceivable that several factors contribute to the development of the postpolio syndrome.

This thesis clearly parallels the lines of investigation which evolved during recent years. In the first part (chapters 2 and 3) of the thesis, emphasis is on the neurological aspects of late polio sequelae, including new muscle weakness and diagnostic methods, whereas the last part (chapter 4) deals with disabilities and handicaps.

The main objectives of this study have been:

- to evaluate the new neuromuscular complaints in polio survivors (chapters 2a, 2b, 2c and 4);
- to estimate the extent of progression of new muscle weakness in patients with postpolio syndrome (chapter 2a);
- to evaluate diagnostic tests which may distinguish between polio survivors with stable neuromuscular conditions and patients with postpolio syndrome (chapters 3a and 3b);
- to determine the level of impairments, disabilities and handicaps in a representative cohort of Dutch polio survivors (chapter 4).

A prospective clinical study on the presentation of late neuromuscular complaints and on the time-course of new muscle weakness development in polio survivors is presented in chapter 2a. Chapters 2b and 2c concentrate on dysphagia and sleep complaints in postpolio patients. Chapter 3 investigates the value of two diagnostic measures i.e., macro electromyography and computed tomography of the skeletal muscles, as diagnostic tools for postpolio syndrome. Chapter 4 focuses on the epidemiology of late polio sequelae in The Netherlands, namely on the prevalence of the new neuromuscular symptoms and the
present level of impairments, disabilities and handicaps in a representative sample of Dutch polio survivors of the last polio outbreak before vaccination against polio. Chapter 5 presents a general discussion of the results of this study, viewed against the background knowledge from the literature and the prospects for future research on late sequelae of polio.

A summary in English and Dutch concludes this thesis.

References
