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Post-polio syndrome: Pathophysiological hypotheses, diagnosis criteria, medication therapeutics

Syndrome post-poliomyélitique : hypothèses physiopathologiques, critères diagnostiques, traitements médicamenteux

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Abstract

Post-polio syndrome (PPS) refers to a clinical disorder affecting polio survivors with sequelae years after the initial polio attack. These patients report new musculoskeletal symptoms, loss of muscular strength or endurance. PPS patients are tired, in pain and experience new and unusual muscular deficits, on healthy muscles as well as deficient muscles initially affected by the *Poliovirus*. Once a clinical diagnosis is established, the therapeutic options can be discussed. Some pathophysiological mechanisms have been validated by research studies on PPS (inflammatory process in cerebrospinal fluid [CSF] and cytokines of the immune system). Several studies have been conducted to validate medications (pyridostigmine, immunoglobulin, coenzyme Q10) or physical exercises protocols. This article focuses on the relevance and efficacy that can be expected from these therapeutics. Very few studies reported some improvements. Medications combined to individual and supervised exercise training programs are promising therapeutic strategies for PPS patients care management.

Keywords: Post-polio syndrome; Therapeutic uses; Diagnostic techniques; Diagnosis; Pathophysiology

Résumé

Le syndrome post-poliomyélitique (SPP) est un diagnostic clinique. Il touche les personnes avec des séquelles de poliomyélite qui se plaignent de nouveaux troubles musculosquelettiques, d'une perte de force ou d'endurance musculaire. Un patient avec un SPP est fatigué, douloureux et perçoit de nouvelles déficiences musculaires inhabituelles, que ce soit sur des muscles sains ou déficients à la suite de l'infection par le *Poliovirus*. Une fois le diagnostic clinique établi, les alternatives thérapeutiques peuvent être discutées. Certains mécanismes physiopathologiques ont été validés par des travaux de recherche (dysrégulation inflammatoire dans le liquide céphalorachidien et cytokines). Un certain nombre de travaux ont été menés pour valider des procédures thérapeutiques médicamenteuses ou des protocoles d'exercices physiques. Cet article fait le point sur l'intérêt et l'efficacité attendue de ces thérapeutiques. Peu d'études thérapeutiques ont conclu à des effets positifs. Les traitements médicamenteux

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combinés à des exercices physiques individualisés et supervisés apparaissent comme des stratégies thérapeutiques prometteuses pour la prise en charge de personnes atteintes d'un SPP.

Mots clés : Syndrome post-poliomyélitique ; Étiologies ; Pathogénie ; Traitements médicamenteux

1. English version

1.1. Introduction

Acute anterior poliomyelitis or Heine-Medin disease corresponds to an infectious attack of the motor neurons by the *Poliovirus* (single-stranded RNA *Enterovirus* belonging to the *Picornavirus* group).

Before vaccines became available, this virus affected more than 600 000 children per year. The last great epidemics (pandemics) occurred in the 1950s. Poliomyelitis is an infectious disease of the neurological system and is the one causing the most motor disabilities in the world: 55,000 patients with polio sequelae in France, 700,000 in Europe, 20 millions in the world [20]. Years after these great epidemics and at least 15 years after the initial polio attack, new symptoms are reported in polio survivors. Several different studies and sharing the longitudinal follow-up of their cohorts led to the discovery of a nosological entity: post-polio syndrome (PPS) [16,18]. Diagnostic criteria were established. No additional exam is capable nowadays of identifying with certainty this syndrome. According to the studies, PPS prevalence is estimated between 20 and 60% [25,31,29], which excludes the psychological consequences related to the past history of this viral infection [33]. Once the diagnosis is established, a therapeutic needs to be defined and adapted to PPS. This work is a review of the literature on useful therapeutic procedures and their expected efficacy to treat patients with PPS.

1.2. Criteria for the diagnosis of post-polio syndrome

Several authors suggested criteria for PPS diagnosis. We mainly find the clinical trio including fatigue, pain and deterioration of the motor deficits. These non-specific symptoms must be detailed and we will focus here on Halstead et al. [16] criteria validated by a college of international experts:

- a prior episode of paralytic polio confirmed by history, physical exam, and typical findings on EMG;
- a period of partial or complete neurological recovery (that could have lasted several years) followed by;
- an extended interval of neurological and functional stability that lasted over 15 years;
- quick or progressive loss of endurance and/or muscular strength with or without muscular atrophy in previously unaffected and healthy muscles, associated to a general muscle and joint fatigue and cold intolerance;
- these symptoms are specific by their unusual and lengthy characteristics (progressing for more than 1 year). Other more rare symptoms can be noted (sleep disorders, respiratory disorders, dysphagia, dysarthria, fasciculations, joint deformities).

PPS is an exclusion diagnosis (Table 1): it is essential to exclude all other potential medical or surgical causes that could be responsible for these non-specific new symptoms before validating the PPS diagnosis. The existence of this nosological entity has been the object of discussions and controversies: natural or pathological evolution? We will refer here to the study of Munin et al. [23] who followed polio patients over a 3-year period. Every 6 months, they evaluated the muscular strength in the quadriceps in isometric and isokinetic settings without finding any significant strength decrease (controlled study). Thus, the natural history of poliomyelitis does not necessarily progress towards an aggravation or extension of the motor deficits. Then, how do we explain the pathophysiology of PPS?

1.3. Pathophysiology of PPS

PPS is related to the death or structural and functional dysfunction of the enlarged surviving motor units [3,5,8,10,11]. Humans are the only hosts for the *Poliovirus*. This virus specifically targets the motoneurons of the anterior horn cells of the spinal cord and the brainstem. Its multiplication is responsible for an apoptosis of the affected motoneuron. Secondly, the adjacent motoneurons re-innervate orphaned muscle fibers that have been denervated by acute polio infection creating giant motor units. After the initial infection, a partial or complete recovery occurs. Understanding the pathophysiological history of this infectious disease is essential to better understand the onset of new symptoms and deficits in

Table 1
Non-exhaustive listing of differential diagnoses for PPS.

Hypothyroidism
Other endocrinal disorders
Respiratory disorders
Sleep apnea syndrome
Cardiac insufficiency
Rheumatoid polyarthritis
Other joint affections
Hematological affections (anemia)
Neoplasia
Adult spinal muscular atrophy
Amyotrophic lateral sclerosis
Cauda equina syndrome
Cervical spondylotic arthritis
Lumbar spinal canal stenosis
Multiple sclerosis
Myasthenia
Radiculopathy