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## Postpolio Syndrome

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Postpolio syndrome (PPS) refers to a constellation of neuromuscular and orthopedic symptoms and signs that have been noted to occur in patients with remote antecedent poliomyelitis. It has been increasingly recognized that individuals recovering from acute poliomyelitis develop new symptoms, most commonly weakness, fatigue, and pain that develops decades after initial disease in the region previously affected. Associated symptoms may include dysphagia, respiratory insufficiency, new muscular atrophy, dysarthria, muscle cramps, fasciculations, sleep abnormalities, and cold intolerance.

Although the concepts of PPS was first described in the late 1800s, it was not until nearly 100 years later that the concept of PPS was more widely recognized and defined. This was due largely to the polio epidemic of the 1940s and 1950s that left many survivors in the world. The virtual epidemic of PPS that occurred among these polio survivors in the 1980s and 1990s has served as a catalyst to attract medical attention to this syndrome.

**Key Words:** Postpolio syndrome

(postpolio syndrome, PPS)

PPS가 (fasciculation), (cold intolerance)

1. PPS 28.5~64% 2.

PPS가 3.

15 가

1800 가

PPS

1940 1950 가 1.

1980 1990 1875 Raymond Charco가

PPS가 PPS

가 1.

PPS 100 가 1980 PPS

가 PPS 4-5

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(late-onset postpoliomyelitis progressive muscular atrophy),<sup>6</sup>

(late poliomyelitis muscular atrophy),<sup>7</sup>  
 (late postpoliomyelitis muscular atrophy),<sup>8</sup>  
 (progressive postpolio atrophy),<sup>9</sup>  
 (progressive postpolio muscular atrophy),<sup>10</sup>  
 (postpoliomyelitis progressive muscular atrophy),  
 (postpolio syndrome)<sup>11</sup>

가 PPS

2.  
 PPS 가 가 가  
 가 가  
 PPS가 PPS가

PPS  
 PPS 가 가 가  
 10 22  
 (discharge)  
 24 가 가  
 8  
 PPS  
 ter가 가 25 jit-(blocking)

(gliosis), (endomysium),<sup>13</sup>  
 oligoclonal band,<sup>14</sup>  
 M band inter-  
 leukin 2 interleukin 2 가,<sup>15</sup> GM1

26 가  
 jitte가 가 가  
 가 가 PPS  
 27  
 jitte가 가

PPS (neurofilament) 가 PPS, PPS가  
 가 PPS  
 17 PPS (genomic  
 sequences)가 PPS가

28 (synapse)가  
 29 PPS  
 (angulated) 가  
 13  
 PPS PPS

PPS PPS가 PPS가  
 PPS가 PPS가  
 5 가  
 PPS (attrition)  
 19

PPS PPS  
 PPS  
 (globus pallidus), (peri-aqueductal gray), (locus ceruleus),  
 30

nervation)가 PPS macro 가  
 21 PPS 가  
 22  
 23 60

70~96% PPS  
 32 PPS  
 31 PPS  
 (disuse) (overuse) 가  
 33 34

19 PPS  
 35

3. (Table 1) 50% 가 .

PPS 89% .

가 가 PPS (wall) PPS가 PPS가

PPS 40% PPS가 11,34,39,41

PPS 73% PPS 가 가 PPS가

PPS 가 가 가 PPS가 42

PPS 가 10~20% (bulbar) 가 PPS

가 가 가 PPS가 11

(bursa), 가 PPS 10 PPS가 1.9%

PPS 80% PPS가 (scoliosis) PPS 가

50% PPS 39

PPS 가

가 . , ,

**Table 1.** Clinical features of postpolio syndrome

Clinical feature	Percent of patients	
	Halstead and Rossi <sup>36</sup> (n=132)	Jubelt and Agre <sup>37</sup> (n=100)
Fatigue	89	86
Muscle Pain	71	73
Joint pain	71	73
Weakness of previously affected muscle	69	88
Weakness of previously unaffected muscle	50	59
Cold intolerance	29	53
Atrophy	28	52
Respiratory insufficiency	NA	36
Dysphagia	NA	36

NA ; data not available







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## REFERENCES

- Jubelt B, Cashman NR. Neurological manifestations of the postpolio syndrome. *Crit Rev Neurobiol* 1987;3:199-220.
- Fisher DA. Poliomyelitis: late respiratory complications and management. *Orthopedics* 1985;88:891-894.
- Wiechers DO, Hubbell SL. Late changes in the motor unit after acute poliomyelitis. *Muscle Nerve* 1981;4:524-528.
- Dalakas MC. Post-polio syndrome 12 years later: how it all started. *Ann N Y Acad Sci* 1995;753:11-18.
- Dalakas MC. Pathogenetic mechanisms of post-polio syndrome: morphological, electrophysiological, virological, and immunological correlations. *Ann N Y Acad Sci* 1995; 753:167-185.
- Kurent JE, Brooks BR, Madden DL, et al. CSF viral antibodies: evaluation in amyotrophic lateral sclerosis and late-onset post-poliomyelitis progressive muscular atrophy. *Arch Neurol* 1979;36:269-273.
- Dalakas MC, Sever JL, Madden DL, et al. Late postpoliomyelitis muscular atrophy: clinical, virologic, and immunologic studies. *Rev Infect Dis* 1984;6(suppl 2): S562-S567.
- Dalakas MC, Sever JL, Fletcher M, et al. Neuromuscular symptoms in patients with old poliomyelitis: clinical, virological, and immunological studies. In: Halstead LS, Wiechers DO. *Late effects of poliomyelitis*. ed. Miami: Symposia Foundation, 1985;73-90.
- Block HS, Wilbourn AJ. Progressive post-polio atrophy: the EMG findings. *Neurology* 1986;36(suppl 1): 137.
- Dalakas MC, Elder G, Hallett M, et al. A long-term follow-up study of patients with post-poliomyelitis neuromuscular symptoms. *N Engl J Med* 1986;314:959-963.
- Dalakas MC. The post-polio syndrome as an evolved clinical entity: definition and clinical description. *Ann N Y Acad Sci* 1995;753:68-80.
- Pezeshkpour GH, Dalakas MC. Long-term changes in the spinal cords of patients with old poliomyelitis: signs of continuous disease activity. *Arch Neurol* 1988;45:505-508.
- Dalakas MC. Morphologic changes in the muscles of patients with postpoliomyelitis neuromuscular symptoms. *Neurology* 1988;38:99-104.
- Dalakas M. Oligoclonal bands in the cerebrospinal fluid of post-poliomyelitis muscular atrophy. *Ann Neurol* 1990;28: 196-197.
- Sharief MK, Hentges R, Ciardi M. Intrathecal immune response in patients with the post-polio syndrome. *N Engl J Med* 1991;325:749-755.
- Illa I, Leon-Monzon M, Agboatwalla M, et al. Antiganglioside antibodies in patients with acute polio and post-polio syndrome. *Ann N Y Acad Sci* 1995;753:374-377.
- Drory VE, Shapira A, Korczyn AD, et al. Antineurofilament antibodies in postpolio syndrome. *Neurology* 1998;51:1193-1195.
- Julien J, Leparac-Goffart I, Lina B, et al. Postpolio syndrome: poliovirus persistence is involved in the pathogenesis. *J Neurol* 1999;246:472-476.
- Tomlinson BE, Irving D. The numbers of limb motor neurons in the human lumbosacral cord throughout life. *J Neurol Sci* 1977;34:213-219.
- Kayser-Gatchalian MC. Late muscular atrophy after poliomyelitis. *Eur Neurol* 1973;10:371-380.
- McComas AJ, Upton AR, Sica RE. Motoneuron disease and ageing. *Lancet* 1973;2:1477-1480.
- Stalberg E, Grimby G. Dynamic electromyography and muscle biopsy changes in a 4-year follow-up: study of patients with a history of polio. *Muscle Nerve* 1995; 18:699-707.
- Nordgren B, Falck B, Stalberg E, et al. Postpolio muscular dysfunction: relationships between muscle energy metabolism, subjective symptoms, magnetic resonance imaging, electromyography, and muscle strength. *Muscle Nerve* 1997;20:1341-1351.
- Borg K, Borg J, Edstrom L, et al. Effects of excessive use of remaining muscle fibers in prior polio and LV lesion. *Muscle Nerve* 1988;11:1219-1230.
- Einarsson G, Grimby G, Stalberg E. Electromyographic and morphological functional compensation in late poliomyelitis. *Muscle Nerve* 1990;13:165-171.
- Wiechers DO. Acute and latent effect of poliomyelitis on the motor unit as revealed by electromyography. *Orthopedics* 1985;8:870-872.
- Maselli RA, Cashman NR, Wollman RL, et al. Neuromuscular transmission as a function of motor unit size in patients with prior poliomyelitis. *Muscle Nerve* 1992;15:648-655.
- Cashman NR, Trojan DA. Correlation of electrophysiology with pathology, pathogenesis, and anticholinesterase therapy in post-polio syndrome. *Ann N Y Acad Sci* 1995; 753:18-150.
- Herrera AA, Grinnell AD. Effects of changes in motor

- unit size on transmitter release at the frog neuromuscular junction. *J Neurosci* 1985;5:1896-1900.
30. Bruno RL, Cohen JM, Galaki T, et al. The neuroanatomy of post-polio fatigue. *Arch Med Rehabil* 1994;75:498-504.
  31. Bruno RL, Creange SJ, Frick NM. Parallels between post-polio fatigue and chronic fatigue syndrome: a common pathophysiology? *Am J Med* 1998;105:66S-73S.
  32. Bruno RL, Frick NM, Cohen J. Polioencephalitis, stress, and the etiology of post-polio sequelae. *Orthopedics* 1991;14:1269-1276.
  33. Agre JC, Rodriguez AA. Neuromuscular function in polio survivors. *Orthopedics* 1991;14:1343-1347.
  34. Agre JC, Rodriguez AA, Sperling KB. Symptoms and clinical impressions of patients seen in a postpolio clinic. *Arch Phys Med Rehabil* 1989;70:367-360.
  35. Shields RW. Postpolio syndrome. In: Brown WF, Bolton CF, Aminoff MJ. *Neuromuscular function and disease*. 1st ed. New York:W.B. Saunders. 2002;1295-1306.
  36. Halstead LS, Rossi CD: Post-polio syndrome: clinical experience with 132 consecutive outpatients. *Birth Defects* 1987;23:13-26.
  37. Jubelt B, Agre JC: Characteristics and management of postpolio syndrome. *JAMA* 2000;412-414.
  38. Halstead LS, Rossi CD: New problems in old polio patients: results of a survey of polio survivors. *Orthopedics* 1985;8:845-850.
  39. Jubelt B, Drucker J: Post-polio syndrome: an update. *Semin Neurol* 1993;13:283-290.
  40. Willen C, Grimby G: Pain, physical activity, and disability in individuals with late effects of polio. *Arch Phys Med Rehabil* 1998;79:915-919.
  41. Ravits J, Hallett M, Baker M, et al. Clinical and electromyographic studies of postpoliomyelitis muscular atrophy. *Muscle Nerve* 1990;13:667-674.
  42. Ivanyi B, Nollet F, Rodekop WK, et al. Late onset polio sequelae: disabilities and handicaps in a population-based cohort of the 1956 poliomyelitis outbreak in the Netherlands. *Arch Phys Med Rehabil* 1999;80:687-690.
  43. Windebank AJ: Differential diagnosis and prognosis. In: Halstead LS, Grimby G. *Post-polio syndrome*. ed. Philadelphia: Hanley and Belfus. 1995;69-88.
  44. Thorsteinsson G: Management of postpolio syndrome. *Mayo Clin Proc* 1997;627-638.
  45. Waring WP, McLaurin TM: Correlation of creatine kinase and gait measurement in the postpolio population. *Arch Phys Med Rehabil* 1993;73:37-39.
  46. Palmucci L, Bertolotto A, Doriguzzi C, et al. Motor neuron disease following poliomyelitis: Bioptic study of five cases. *Eur Neurol* 1980;19:414-418.
  47. Cashman NR, Maselli R, Wollmann RL, et al. Late denervation in patients with antecedent paralytic poliomyelitis. *N Engl J Med* 1987;317:7-12.
  48. Daube JR, Windebank AJ, Litchy WJ: Electrophysiologic changes in neuromuscular function over five years in polio survivors. *Ann N Y Acad Sci* 1995;753:120-128.
  49. Trojan DA, Gendron D, Cashman NR: Electrophysiology and electrodiagnosis of the post-polio motor unit. *Orthopedics* 1991;14:1353-1361.
  50. Boonyapisit K, Shapiro BE, Trojan DA, Cashman NR. Poliomyelitis and postpoliomyelitis syndrome. In: Katirji B, Kaminski HJ, Preston DE, Ruff RL, Shapiro BE. *Neuromuscular disorders in clinical practice*. 1st ed. Boston: Butterworth Heinemann. 2002;405-416.
  51. Steljes DG, Kryger MH, Kirk BW, et al. Sleep in postpolio syndrome. *Chest* 1990;98:133-140.
  52. Sonies BC. Dysphagia and post-polio syndrome: past, present, and future. *Semin Neurol* 1996;16:365-370.
  53. Swingler RJ, Fraser H, Warlow CP. Motor neuron disease and polio in Scotland. *J Neurol Neurosurg Psychiatr* 1992;55:1116-1120.
  54. Bach JR, Alba AS. Pulmonary dysfunction and sleep disordered breathing as post-polio sequelae: evaluation and management. *Orthopedics* 1991;14:1329-1337.
  55. Trojan DA, Collet JP, Shapiro S, et al. A multicenter, randomized, double-blinded trial of pyridostigmine in postpolio syndrome. *Neurology* 1999;53:1225-1233.
  56. Dinsmore, S, Drambosia J, Dalakas MC. A double-blind, placebo-controlled trial of high-dose prednisone for the treatment of post-polio syndrome. *Ann N Y Acad Sci* 1995;753:303-313.
  57. Stein DP, Dambrosia JM, Dalakas MC. A double-blind, placebo-controlled trial of amantadine for the treatment of fatigue in patients with the post-polio syndrome. *Ann N Y Acad Sci* 1995;753:296-302.