Post-Polio Syndrome: A Perspective from Three Countries

**CASE SCENARIO**

A 55-year-old woman with a history of polio at the age of 2 years presents with complaints of increasing fatigue and pain in her legs with moderate activity, including performing general household tasks and ambulating distances greater than a few city blocks. She reports that she had residual weakness in her legs that limited her participation in school athletics as a child and teenager, but no fatigue or pain. She has mild scoliosis but this is not accompanied by any symptoms and does not affect her activity or social participation. The symptoms of fatigue and pain began to emerge about 10 years ago and have steadily worsened to the point that she is having difficulty maintaining her previous level of function. Her parents told her that she did not require ventilatory support during the acute illness and never needed bracing during childhood. She describes her pain as primarily achy but sometimes more intense, sharp and radiating from the back. Rest usually helps to relieve pain and fatigue. She experiences the fatigue as both muscular and more generalized, gradually increasing with walking or other physical activity. These symptoms disturb her sleep intermittently during the week. She reports some mild depressive symptoms but does not believe that she suffers from depression. She is employed full-time as a paralegal, as she has been for many years. She used to be an avid golfer but has been unable to play the sport in recent years.

What additional evaluation, including history and physical examination, laboratory, or other studies, would you recommend for this patient? What do you think are the possible etiologies of her symptoms? How would you advise her concerning treatment of her condition including medications, lifestyle modifications, or exercise?

Jianan Li, MD, Responds

Over the last 20 years, post-polio syndrome (PPS) has become well-recognized as occurring in some patients with a history of polio. The case presented is a typical patient with PPS. China has the largest population of people infected by poliomyelitis in the world, but few PPS cases have been reported [1]. There may be several causes for the low incidence of PPS in China [2]: most polio patients are living in the countryside and have poor access to qualified doctors with sufficient knowledge regarding PPS. Most Chinese doctors have limited training in identification of PPS from a lack of PPS education in medical schools and postgraduate training [3]. PPS could be misdiagnosed as osteoarthritis, chronic pain syndrome (without a specific cause identified), depression, or deconditioning. Many Chinese polio patients are relatively young, so the contribution of aging to the disorder may not be evident yet.

Clinical Evaluation

Muscle strength assessment, electromyographic (EMG) studies and motion analysis may provide objective evidence of PPS pathology and help identify functional prognosis [4]. Motion analysis for a patient with weakness or endurance issues may help identify the possible beneficial effects of rehabilitation interventions such as orthoses and assistive devices.

Treatment

The recommended treatment for this patient may include energy-saving strategies for daily living, low-intensity exercise for maintenance of muscle efficiency and metabolism, as well as medications for pain, sleep disturbance, or depression. In addition, psychological treatment for depression might also be helpful.
Theoretically, traditional Chinese medicine is beneficial for adjustment of imbalanced neuromuscular function and psychological function through adjustment and balance of yin and yang (negative and positive energy). Some studies have demonstrated the effectiveness of acupuncture and Chinese exercise on polio survivors. However, there are no published randomized controlled studies providing clear evidence of the effectiveness of these Eastern medicine treatments for PPS. Cultural influences on clinical management, design of assistive devices, and environment modification as well as ergonomics in employment settings for PPS patients need to be considered in future studies.

Relevant Research Studies

The following data might be important to demonstrate a potential strategy for prevention of PPS in 20 to 30 years. There was a nationwide outbreak of polio in China from 1989 to 1990 affecting the Pi and Shuyang counties in Jiangsu Province. A total of 1012 cases were identified [5]. A survey showed that 42% had one limb affected, 47% had 2 limbs affected, and 11% had 3 or more limbs affected. This was the last major outbreak of poliomyelitis in the world. After 15 years of comprehensive rehabilitation services (provided since 1993), a cohort of 648 people (350 males and 298 females, age 20 years on average) were followed in 2008. The interventions included the following.

Surgery. Surgery on extremities was performed on 488 patients (830 operations); procedures included joint plasticity procedures and tendon lengthening, transferring, and release. Spinal surgeries were performed in 98 cases and spinal revision surgery was required in 6 cases.

Rehabilitation. Training for strength, joint motion, gait, and activities of daily living; preoperative and postoperative therapy; and psychological interventions were provided. The number of orthoses prescribed for lower limbs was 2756 braces for 455 patients; 153 pairs of corrective shoes were provided. Spinal braces were provided for 232 patients. A total of 2193 crutches and 296 wheelchairs were provided.

As a result of the 15-year program, significant improvements in physical functioning have been demonstrated in all patients. The most important achievement is the recovery of ambulation capacity in 95.8% of patients. All patients with one-leg involvement achieved independent walking. A majority (90.9%) of the 296 patients with paraplegia and quadriplegia regained independent walking ability. Wheelchair dependence was seen in 27 patients with the most severe paralysis in 3 or 4 limbs in this group.

Dependence on orthoses for walking has been dramatically reduced over time. Among 455 patients with orthoses at entry into the study, 106 patients (23%) reduced their dependence on orthoses. For example, 65 patients with bilateral knee-ankle foot orthoses (KAFO) now only require a unilateral KAFO; 20 patients with a unilateral KAFO progressed to ankle foot orthoses (AFO); and 21 patients with an AFO progressed to walking without an orthosis. Of the cohort, 162 patients (36%) do not require an orthosis as they showed sufficient improvement in physical functioning; these included 71 patients with initial bilateral KAFOs, 70 patients with an initial unilateral KAFO, and 21 patients with an initial AFO. Only 187 (41%) are still requiring an orthosis, including 27 patients with bilateral KAFOs, 47 patients with a single KAFO, and 13 patients with an AFO. The authors conducted gait analysis for a selected group of 20 patients. The results indicated that 75% of patients who progressed from KAFO to AFO within 2 years significantly improved their gait and also reduced the oxygen cost of ambulation.

The question is whether the reduction in the use of orthoses will contribute to an increase in the incidence of PPS in 20 to 30 years. To help answer the question, the authors have commenced a long-term follow-up study of those polio patients. EMG and gait analysis will be conducted with selected patients. In addition, a community-based rehabilitation program has been organized for those patients. The authors postulate that well-designed activities and patient education will have the potential to reduce the incidence of PPS.

Further studies might be focused on the following questions: What is the pathology or etiology of PPS (ie, overuse or disuse of affected muscles, and how is it related to the degeneration of individual nerve terminals in the residual motor units)? Does biomechanical adaptation of affected limbs lead to a better alignment of lower limbs, resulting in better mechanical efficiency of muscles and lower oxygen consumption for walking and daily living activities? Is PPS predictable and preventable by careful monitoring of electrophysiology, gait and motion analysis, and symptoms?

REFERENCES


Gunnar Grimby, MD, PhD, FRCP (London), Responds

The patient represents a typical case of post-polio syndrome (PPS), probably from successive loss of muscle mass during later years. The patient also seems to overexert herself. There are no reasons to assume respiratory limitations are causing the reduced physical activity and would suggest the following further evaluations.
Clinical Evaluation

Exercise tolerance should be further analyzed with respect to type and amount of activities, habits of pauses and resting, and occurrence of pain and fatigue. The distribution of pain should be described using pain drawings [1]. Other reasons for pain and fatigue not directly related to late effects of polio should be considered and assessed using clinical examination and laboratory tests. Subjective experience of new muscle weakness and fatigue should be recorded with respect to specific muscle groups. For weak muscles, manual muscle tests could be performed but would not be relevant for 4-grade strength or higher. Entrapment syndromes should be excluded. Gait should be analyzed by clinical observation.

Muscle strength can be measured using stationary methods (isometric and/or isokinetic) or with handheld dynamosimeters for relevant muscle groups, such as quadriceps, hamstrings, dorsiflexors, planatarflexors, biceps, and triceps, and grip. Comparison should be made between sides and with reference values. To establish which muscle groups have been affected by polio, electromyography can be used. If possible, an estimate of motor unit size is recommended, preferably with macro-electromyography [2,3]. This will also provide information on the denervation/rennervation processes within a motor unit. The presence of very large motor units is a risk factor for further deterioration in muscle function. Even if respiratory function is not assumed to limit performance, vital capacity should be measured for baseline documentation. Exercise tolerance may be further analyzed using a bicycle ergometer. Even if maximal heart rate cannot be approached because of muscle weakness, the anaerobic threshold may be defined. Electrocardiogram and blood pressure should be recorded during the evaluation.

Treatment

Detailed, individualized recommendations depend on the results of the extended history and laboratory tests that become available, but more general advice for PPS can be provided. The physician and the rest of the rehabilitation team should discuss and monitor physical activity patterns to find strategies for reducing fatigue and pain as well as developing individualized energy-saving strategies. Clinical and research experience has shown that pool exercise may reduce pain and improve performance and it can be generally recommended in this case [4]. For muscle groups with moderately reduced muscle strength, a submaximal resistance training program can be used with close monitoring of muscle strength changes to avoid overuse weakness [3]. If gait analysis and assessments of muscle function indicate that the use of a cane may be appropriate, the patient should be encouraged to try using one. However, it may not be accepted immediately. The need for other assistive technologies or home modifications should also be considered.

It is important for the physician to provide information on PPS and to discuss the etiology and prognosis of the disorder with the patient and family members. Finally, it is worth mentioning that treatment with intravenous immunoglobulin has given some promising results in Swedish studies for improving function and health-related quality of life [5] and may be considered in this case if available.

The patient should be seen by a physiatrist or other physician with competence in treating patients with late effects of polio when the results of testing are available, followed by rechecks at 3 or 6 months. A multiprofessional assessment and follow-up with a rehabilitation team is important. Further appointments can then be discussed.

REFERENCES

referral pattern? Is the pain in normal muscles; functionally intact, polio-affected muscles; or muscles that are severely atrophic? Further information is also needed on the aggravating and alleviating factors. Does the pain present during activity or later in the day? Do pain and fatigue occur simultaneously? Do symptoms progress as she continues certain activities? Can she alleviate symptoms by simply standing still, leaning forward, or sitting down? Do symptoms persist for several days after the activity? When the pain and fatigue disturb her sleep, does she have difficulty falling asleep or frequent awakening during the night? Is pain at night related to position? Are the episodes of sleep disturbance associated with muscle cramping and twitching? Is there any relationship between the amount of activity she has done during the day and the episodes of sleep disturbance? Is her generalized fatigue related to activity or quality of sleep? Does she wake up refreshed or tired? Medical history, current medications, family and social history, and a complete review of systems may also add pertinent information. Our patient reports some mild depressive symptoms. Further questioning or screening with a depression inventory will narrow the differential diagnosis.

The importance of the physical examination should not be underestimated in evaluating a polio survivor. Unlike many other neuromuscular diseases, the residual effects of polio are variable in each individual, and careful assessment is key to understanding body mechanics and function. Assessment of vital signs, heart, and lungs will indicate general wellness and cardiovascular fitness level. Specific spine examination, evaluating scoliosis, screening for lumbar radiculopathy, and special maneuvers may identify spine pathology. Evaluation of the extremities must include joint function and provocative maneuvers. Although limbs severely affected by polio may have discoloration and decreased temperature, peripheral pulses and skin sensation should be intact in the absence of peripheral vascular disease or neuropathy. A careful and complete manual muscle examination of all major muscle groups will provide insight into the patient’s unique body mechanics and areas at risk from agonist-antagonist muscle imbalances or increased/abnormal stress with weightbearing or movement. Clinical gait evaluation is an excellent tool to further understand biomechanics of functional mobility and compensations.

Additional laboratory or radiographic studies might be indicated based on information and findings obtained from the history and physical exam. There are no specific diagnostic studies for PPS, but studies may confirm other etiologies or conditions contributing to symptoms. Electrodiagnostic studies are not necessary for the diagnosis of PPS but may provide evidence of the effects of polio in cases that do not have a clear clinical history [2]. They can be used to screen for nerve entrapments or lumbar radiculopathy. Radiologic studies can reveal the degree of lumbar scoliosis and the extent of degeneration in the spine or other lower extremity joints. If neurogenic claudication remains in the differential, magnetic resonance imaging of the spine is indicated. Arterial Doppler studies and segmental pressures can assess the vascular system. Chronic muscle pain with increased weakness, especially in a limb-girdle distribution or with a temporal relationship to statin use, may prompt measurement of creatine phosphokinase and liver function levels. A patient with fatigue should have a complete blood count, blood chemistry, and thyroid panels to screen for anemia and endocrine/metabolic disease.

**Treatment**

In this case, the symptoms of leg pain and fatigue are most likely polio-related and probably from PPS. The patient meets the criteria of previous polio, functional stability, and new muscle fatigability. Pain as reported by this patient is commonly associated with PPS, but not essential to meet the diagnostic criteria. If the pain is not simply overuse muscle pain, but from a musculoskeletal problem, the diagnosis of PPS should be delayed. If appropriate treatment of the musculoskeletal problem does not abate muscle fatigue, the diagnosis of PPS can be made.

The pathophysiology and treatment of PPS have not been clearly defined and proven. The peripheral disintegration model proposed by Weichers and Hubbell [3] describing gradual loss of axon sprouts has become the accepted explanation for weakening and fatigability of polio-affected muscles. The secondary symptoms of generalized fatigue and pain are less clearly defined with multiple potential etiologies. Initially, the symptoms of increased weakness and fatigue were attributed to overuse. Although many related musculoskeletal conditions are related to overuse or progressive degenerative changes, exercise or an active lifestyle has not been directly linked to increased loss of axon sprouts or development of PPS. Adaptations in functional activities, lifestyle modifications, and a controlled exercise program have proven to be beneficial in most cases [4]. As polio survivors have become aware of PPS, many have decreased activity to the point that deconditioning and disuse atrophy are common.

Whether a patient meets the exact criteria for PPS may not be important. Treatment of PPS is based on symptoms not pathophysiology. To date, the authors do not have a medication to stop the loss of axon sprouts or to slow the decrease in numbers of motor units with aging. Without a cure, interventions are based on management principles that may apply to all polio survivors with residual paralysis, not just those with PPS. Musculoskeletal problems and loss of biomechanical efficiency related to muscle imbalance, joint deformities, and asymmetric forces on joints, ligaments, and connective tissue are common. Pain may result from arthropathy, bursitis, tendonitis, peripheral nerve entrapments, radiculopathy, osteoporosis, and other conditions. These medical conditions are related to the sequelae of polio, but do not meet the criteria for diagnosis of PPS. They would be referred to as “other orthopedic conditions.” Although these musculoskeletal problems are not unique to polio survivors, they often go unrecognized by health professionals unfamiliar with sequelae of polio. Failure to recognize limitations of partially denervated muscles may result in inappropriate treatment.
Inexperienced health providers may be well-served by referring PPS patients to a post-polio specialist.

Recommendations for treatment must be individualized based on a detailed assessment by an experienced clinician. Ideally, pain can be controlled through avoidance of overuse and adaptations to decrease stress on weak muscles and unstable or degenerative joints. The adaptations may include therapies to optimize muscle balance or joint alignment, orthoses, or assistive devices. Pain medications should be used for a specific functional benefit, not to mask the signs of overuse. Fatigue can be managed with pacing of activities and regular rest periods. Sleep hygiene should be addressed and optimized. Regular mild to moderate exercise should incorporate stretching, gentle strengthening of muscles grade 3 or higher, and cardiovascular conditioning [5,6]. The patient must recognize signs of muscle overuse (cramping, twitching, crawling sensation, or pain) and learn to adjust exercise and activity accordingly.

A self-educated polio survivor may be ready to accept recommendations immediately, whereas others will need to process the information and deal with the psychological issues associated with new or recurrent disability before accepting any intervention. A psychologist or local support group is often helpful during the process of acceptance. Long-term decisions regarding employment and recreation are best addressed later in the treatment course after the basic recommendations have been implemented.

REFERENCES

M. Elizabeth Sandel, MD, Senior Editor, Commentary

World Polio Day on October 24 is a reminder that great strides have been made in eradicating poliomyelitis through vaccination (Salk, 1955; Sabin, 1962 [1]). The Global Polio Eradication Initiative launched in 1988 has certainly led to the sparing of millions of children and adults from death and disability. In 2008, there were 1652 confirmed cases of paralytic polio reported worldwide. At this time, 4 countries—Afghanistan, India, Nigeria, and Pakistan—are endemic for polio. The World Health Organization estimates there are 20 million people worldwide with disability from poliomyelitis [2]. According to surveys by the National Center for Health Statistics, there were about one million polio survivors in the United States in the 1990s, a subpopulation primarily of “baby boomers” who contracted the disease in the 1940s and 1950s [3].

In this discussion, a distinction is made between late effects of polio (mainly musculoskeletal) and the subcategory post-polio syndrome (PPS), a neurologic disorder. The prevalence of PPS is reported to range from 28% to 64%, and the manifestations of PPS are reported to be similar around the world [4]. The functional course, risk factors, and prognosis for PPS are unclear [5]. Fatigue is the most prominent symptom and may be due to central or peripheral causes. Patients report symptoms of both fatiguing muscles and generalized fatigue. The importance of evaluating neuromuscular effects on respiratory function must be emphasized, especially as these effects may be a contributor to fatigue through hypoventilation [6].

There are differing emphases in these commentaries with no major disagreements, and certainly a physiatric approach is used by each of the discussants. The evaluation of the patient with late effects of polio, including PPS, especially those in mid- to late-life, may be very difficult. PPS is indeed a diagnosis of exclusion as there is no biomarker for the disorder. Other diagnoses may emerge as the patient ages.

Dr. Li discusses the situation in China where the population may be less (or not yet) affected by PPS or perhaps not identified because of various factors. The opportunity certainly exists in China to study the progression in a recently infected cohort and readers must await further studies from that country. Dr. Grimby emphasizes a specific workup for neuromuscular aspects, including electrophysiologic studies to confirm the effects of the polio virus and for identification of what muscles are subclinically involved. Dr. Li also refers to the benefits of evaluations such as electromyography and motion analysis. Dr. Vandenakker focuses her discussion on the differential diagnosis using electrophysiologic, laboratory, and radiographic studies in some cases to make sure that no other musculoskeletal, neurologic, or metabolic/endocrinologic disorder is missed.

Pain management and rehabilitation are based on specific symptom-based assessments and an understanding of the patient’s level of activity and participation. Individualized prescription may include therapeutic exercise and lifestyle modifications, weight reduction programs, energy conservation, strengthening programs, adaptive devices, bracing, mobility aids, and ventilatory support, but treatment plans will vary widely. The clinician must be aware that 3 factors may contribute to PPS: aging, overuse, and disuse; the contributions of each will vary in an individual patient and are difficult to distinguish. In addition, the limitations of the manual muscle test are clear in this disorder because of the fatigability of muscles that have no apparent loss of strength.

Intravenous immunoglobulin is not approved by the United States Food and Drug Administration for this disor-
der. Use of intravenous immunoglobulin is more common in Scandinavian countries, where most of the research has been conducted. The cost may be prohibitive for widespread use. Other pharmacological interventions, acupuncture, or other alternative medicine approaches have not been proven to be effective. Readers must await additional studies on the biology of PPS, which remains poorly understood.

The case represents what physiatrists do best when pathophysiology is unclear and medical cure is elusive, yet treatment is not. Physiatrists integrate a comprehensive history of the condition, including associated symptoms, with a thorough physical examination, supplemented with diagnostic studies to employ a variety of strategies, with therapists and other providers, to reduce disability using an individualized approach. This patient, with the right rehabilitation treatment plan, may stay working and even get back to the golf course.

REFERENCES
Attendees: Dr. William DeMayo (John P. Murtha Neuroscience and Pain Institute, Johnstown, PA), Joan Headley (Post-Polio Health International, St. Louis, MO), Barbara Duryea (John P. Murtha Neuroscience and Pain Institute, Johnstown, PA), Wendi Nagle (John P. Murtha Neuroscience and Pain Institute, Johnstown, PA), Jan Goodard (John P. Murtha Neuroscience and Pain Institute, Johnstown, PA), Dr. Fred Maynard (Marquette, MI), Dr. Carlos Vallbona (Baylor College of Medicine, Houston, TX), Dr. Walter Stolov (University of Washington Medical Center, Seattle, WA), Dr. Khema Sharma (University of Miami School of Medicine, Miami, FL), Cynthia Henley, PT (University of Miami, Miami, FL), Kathryn Wollam, PT (University of Miami, Miami, FL)

Agenda

I. Presentation/Discussion

Post-Polio Syndrome: A Perspective from Three Countries
Carol Vandenakker, MD
University of California – Davis Medical Center
Sacramento, CA

Article "Post-Polio Syndrome: A Perspective from Three Countries" and Discussion Points were sent to the Directors prior to the call.

II. Next Call

Topic – Functioning and Ageing with late-onset sequelae of poliomyelitis
Presenter – Dr. Frans Nollet, MD, PhD
Date – May 18, 2010 – 6:00 p.m. ET
(Third Tuesday of each month)

Disclaimer: The following are unofficial notes which have not been read by or approved by the speaker.

- The case scenario was vague and did not give much information.
- The editor's intent was to discuss PPS without too many specifics.
- Editor's idea beyond that was to get different perspectives from different places in the world.
- Dr. Vandenakker indicated a full assessment of the patient would be needed.
• Dr. Li's response was most interesting…from a country where there is still a lot of active polio and younger survivors.
• Some patients who immigrated here from other countries are dependent on family members to take care of them. Not the same personality as the U.S. survivors.
• Some younger survivors' walking problems are treatable, but some have developed deformities and have to be seen by a surgeon.
• Some are not amenable to bracing. They are not unhappy in their role of having the family do things for them and, if you cannot "fix" them, they are not interested in treatments.
• There is a culture issue.
• A few patients who walk with crutches do not want braces even though they are developing upper extremity problems.
• Often the immigrants have not had rehab.
• Dr. Li's questions at the end of his response were great. Is it inevitable or is it a secondary condition with aging or biomechanical issue?
• Dr. Grimby's response addressed using EMG and exercise tolerance testing. Do clinics do formalized exercise testing?
  o One clinic does a six-minute walk test to look at the quality of the gait. Another has the patient walk up and down the hall; checking pulse and respiration rate before and after the walk. Patient is usually able to walk more than they thought they could.
• Regarding the issue of pain – if patient had a definite history of polio and specific weakness cannot be determined, what else could be done to determine if this woman's vague complaints are due to overuse or something else?
  o Need to get a pattern of pain. The case scenario doesn't give an adequate pattern of the pain – when it comes or how it is alleviated. Is it joint pain from imbalance or deformity; is it hip or knee pain; is it more spinal?
  o Having the patient journal their pain will give physician a better pattern.
  o Do history and musculoskeletal exam.
  o If the clinical history of the acute onset is typical of polio it would not be necessary to do an EMG.
  o If not sure, an EMG should be done.
• Some patients being braced think that the brace will be heavy. However, they appear to expend a lot less energy walking with the brace than without. How can the patient be convinced the brace is helping them?
  o Have the orthotist and physical therapist together and make adjustments when the brace is put on. Use mirrors to let the patient see how they are walking. This often gives positive feedback. Also have the family present.
  o Education and training of the patient.
• Interesting article – but group would like to see more done. It validates that by curing the patient's mechanical problems, many of their pain and fatigue issues improve. Their function improves.
• More people listen, analyze and ask questions and then accept bracing.
• Pain and fatigue are not big problems with younger patients. They only go into a brace when they start to have pain.
• Part of the disabilities seen in later years are in patients who threw away their braces.
• Dr. Li raised that question and it is a good one. This has been seen in the US population. It's about how easily they can return to the brace they gave up and how well they do.
• They feel more disabled using a brace even if they are walking straighter.
• Those from other countries who never had a brace may be helped by being introduced to an American who has been using a brace. Those who are in support groups with older American survivors are more accepting of bracing.
• A patient who needs a fair amount of bracing can be helped by introducing a little at a time.