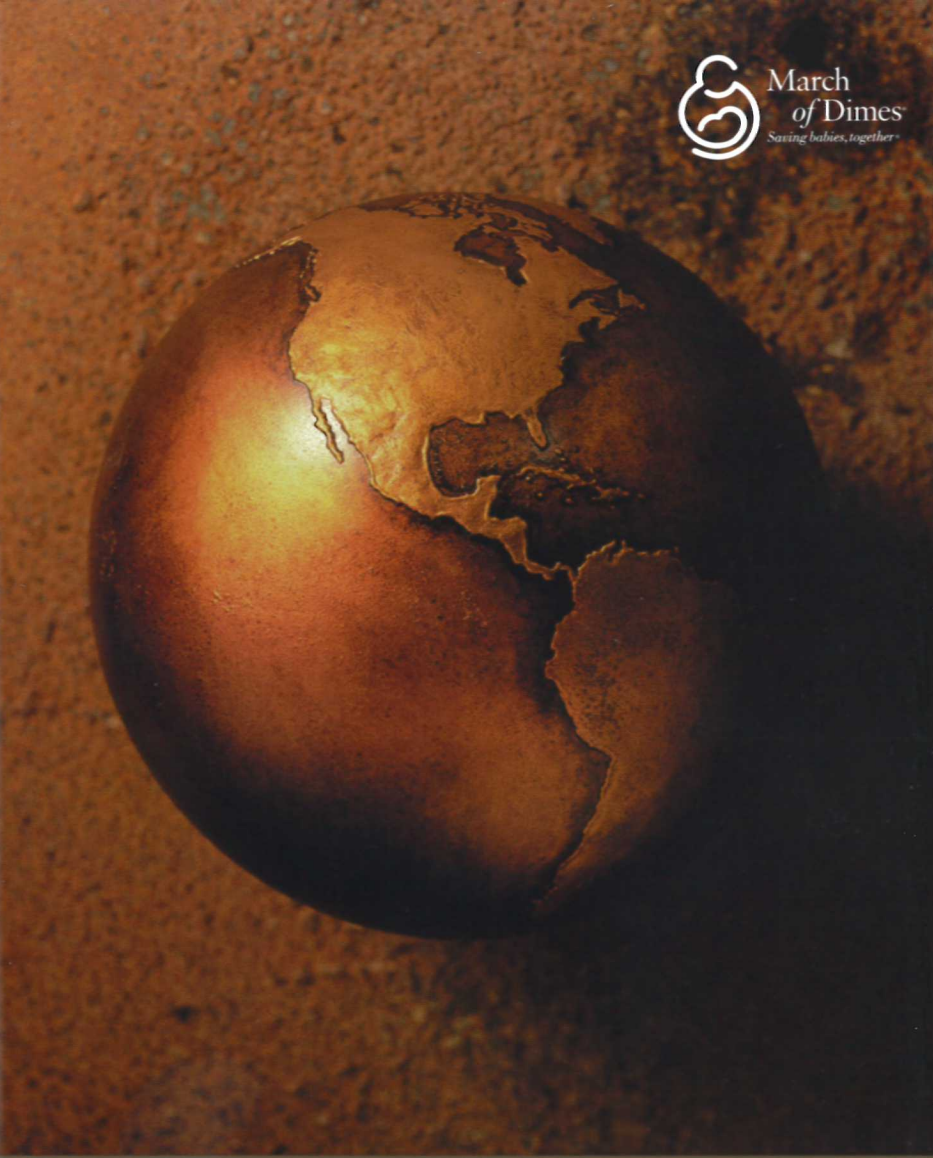

MARCH OF DIMES INTERNATIONAL CONFERENCE ON

Post-Polio Syndrome

Identifying Best Practices in Diagnosis & Care

For additional copies contact:
March of Dimes
1275 Mamaroneck Avenue
White Plains, NY 10605
USA

Or download from Global Programs at March of Dimes
website: www.modimes.org

STEERING COMMITTEE

Lewis P. Rowland, MD *(Chair)*, Department of Neurology, Columbia University College of Physicians and Surgeons, New York, NY
John R. Bach, MD, University of Medicine and Dentistry of New Jersey, New Jersey Medical School, Newark, NJ
Kristian Borg, MD, PhD, Head, Huddinge University Hospital, Stockholm, SWEDEN
Neil R. Cashman, MD, Center for Research in Neuro-Degenerative Diseases, University of Toronto, Ontario, CANADA
Marino C. Dalakas, MD, Neuromuscular Disease Section, National Institute of Neurological Disorders & Stroke, Bethesda, MD
Elizabeth Dean, PhD, PT, School of Rehabilitation Sciences, University of British Columbia, Vancouver, BC, CANADA
Lauro S. Halstead, MD, Post-Polio Program, National Rehabilitation Hospital, Washington, DC
Joan L. Headley, MS, International Polio Network, Saint Louis, MO
T. Jacob John, MD, Department of Clinical Virology, Christian Medical College, Vellore, Tamil Nadu, INDIA
Richard B. Johnston, Jr, Department of Pediatrics, National Jewish Medical and Research Center, Denver, CO
Rhoda Olkin, PhD, Department of Clinical Psychology, California School of Professional Psychology, Alameda, CA
Susan L. Perlman, MD, Department of Neurology, University of California at Los Angeles, Los Angeles, CA
Daria A. Trojan, MD, MSc, McGill University, Montreal Neurological Hospital, Quebec, CANADA
Barbara G. Vickrey MD, MPH, Department of Neurology, University of California at Los Angeles, Los Angeles, CA
William W. Wendling, Cleveland Municipal School District, Cleveland, OH

ADVISER

Michael Katz, MD, Vice President for Research, March Of Dimes

MARCH OF DIMES STAFF

Christopher P. Howson, PhD, Director, Global Programs, *Editor*
Richard P. Leavitt, Director, Science Information, *Editor*
Ellen L. Fiore, Managing Editor, Science Information, *Associate Editor*
Amy L. Edelstein, Assistant to the Director of Global Programs
Joan Roe, Conference Coordinator
Michael J. Finnerty, Manager, Global Programs
Judy Jacobson, Director, Department and Chapter Operations, Volunteer and Chapter Support

ACKNOWLEDGMENTS

The committee is grateful to the many individuals who made substantive and productive contributions to this report. In particular, it thanks the conference participants whose background papers and discussion contributed importantly to the report: James Agre, MD, Howard Young Medical Center, WI; Anne Gawne, MD, Roosevelt Warm Springs Institute for Rehabilitation, GA; Charlotte Gollobin, MS, Nutrition Consultant, MD; Burk Jubelt, MD, SUNY Health Science Center, NY; Pesi H. Katrak, MD, Prince Henry Hospital, Sydney, Australia; Elizabeth Kowall, MS, OTR, Occupational Therapist, WI; Frederick Maynard, MD, Upper Peninsula Rehabilitation Medical Associates, MI; Susumo Sato, MD, National Institute of Neurological Disorders and Stroke, MD; Kimberly Sialtsis, Easter Seals/March of Dimes National Council of Canada, Ontario; Barbara C. Sonies, PhD, National Institutes of Health, MD; Andria Spindel, Easter Seals/March of Dimes National Council of Canada, Ontario; and Alexa Stuifbergen, PhD, RN, FAAN, University of Texas at Austin, TX.

The committee is also grateful for the important perspective, data and other information provided by the polio survivors, health care providers, and others who called in to the committee during its public session at the October 1999 meeting: Dr. Ulrich Alsentzer; Phyllis Bailey; Kathleen Blumberg; Mary Correia; Ellen Crim; Jenny Danielson; Katherine Decker; Dr. James Donovan; Paul Fick; Dr. Anne Gawne; Lucile Harford; Nickie Lancaster; Robbie Leonard; Dr. Paul Peach; Jim Powell; Frederick Price; and Dr. Robert Pyatt.

The committee thanks the following project staff of the March of Dimes: Dr. Christopher P. Howson, Richard P. Leavitt and Ellen L. Fiore for their substantive input into the content and design of the report and for keeping this project on schedule; and Joan Roe, Amy L. Edelstein, Michael J. Finnerty and Judy Jacobson for their logistical support of the committee. It also thanks Dr. Jennifer L. Howse, President of the March of Dimes, and Ms. Anna E. Roosevelt, Vice Chair of the March of Dimes National Board of Trustees and Executive Director of the Brain Research Foundation, Chicago, Ill, for their support and key contributions at the Warm Springs conference and to this report.

The committee is deeply appreciative of the excellent organizational efforts of the staff of the Roosevelt Warm Springs Institute for Rehabilitation: Frank Ruzycki and Carolyn Moreland.

Finally, the committee gratefully acknowledges the important support of Mrs. Lucile Harford, Englewood, Fla. This report is dedicated to her and to the many courageous polio survivors, worldwide.

Contents

PREFACE

On May 19-20, 2000, the March of Dimes Birth Defects Foundation (March of Dimes), in collaboration with the Roosevelt Warm Springs Institute for Rehabilitation, held an international conference on post-polio syndrome (PPS) in Warm Springs, GA. The purpose of the conference was to review current information on the syndrome's causes and promote information exchange on best practices regarding diagnosis, treatment and management of PPS. The two-day conference convened many of the world's leading experts on various aspects of PPS, including causes; diagnostic criteria; features of the disorder, including neurological, musculo-skeletal and respiratory problems; and treatments such as physiotherapy, occupational therapy, coping skills and nutrition. This report derives from that conference.

BACKGROUND

PPS is a disorder of the nervous system that appears in many survivors of paralytic polio, usually 15 years or more after the original illness. Its main symptoms are new progressive muscle weakness, severe fatigue and pain in muscles and joints. Some patients develop potentially life-threatening difficulties with breathing and swallowing.

PPS first came to wide attention in the medical community during the 1980s—three decades after the worst polio epidemics in the United States, which struck more than 50,000 people in 1952 alone. All told, up to 250,000 U.S. polio survivors may have PPS.

While increasing numbers of post-polio patients have been showing up in doctors' offices and medical clinics throughout the United States and other countries of the world, few physicians and relevant allied health professionals have adequate knowledge of the syndrome or of best clinical practices for diagnosis and treatment. Three conferences on PPS have been held to date—in 1984, 1986 and 1994—but these focused largely on identifying research needs. There has, therefore, been a need for examination of current clinical practices, with the goal of

developing guidelines on how best to diagnose, treat and rehabilitate persons with PPS.

THE ROLE OF THE MARCH OF DIMES

The March of Dimes was founded in 1938 to combat epidemic polio in the United States, and its mission was realized by the early 1960s with development and widespread use of the Salk and Sabin vaccines. With polio rapidly dwindling as a public health threat in the United States, the March of Dimes changed the focus of its mission to improving the health of babies by preventing birth defects and infant mortality.

Although its mission had changed, the March of Dimes continued to be engaged in some polio-related issues. From 1986 to 1992, the Foundation was involved in a number of post-polio activities, including the publishing of public health information sheets on polio and post-polio syndrome. In 1999, in response to a growing number of contacts from polio survivors who reported that they were experiencing obstacles in the diagnosis and care of their fatigue, pain, weakness and other problems associated with PPS, the Foundation convened an international steering committee, chaired by Lewis P. Rowland, MD, Professor of Neurology at the Columbia University College of Physicians and Surgeons, to address an issue of critical importance to polio survivors: the need to improve knowledge among medical care providers and polio survivors of best practices in diagnosis and clinical management of PPS.

ACTIVITIES OF THE STEERING COMMITTEE

The steering committee, which included leading researchers in the fields of PPS, neurology, rehabilitation medicine, pulmonary medicine, virology, psychology and other disciplines, as well as individuals living with PPS, held its initial meeting in October 1999 in New York, to develop the agenda and identify speakers for the May conference. In addition, the committee provided an opportunity for individuals not physically present at the

meeting to call in with data, perspectives, and other information on PPS that they wished the committee to consider. Callers included polio survivors with PPS, and medical and allied health care providers, among others.

In reviewing the data presented in May 2000 at Warm Springs, the steering committee recognized that there was little in the way of new evidence on PPS diagnosis, treatment and rehabilitation published since the 1994 conference and that major gaps in knowledge persist. It concluded, therefore, that in addition to providing guidelines on how best to diagnose, treat and rehabilitate persons with PPS, the committee could best serve the PPS community by offering recommendations on research that might address some of the persistent gaps in knowledge. The committee has also recommended institutional mechanisms—i.e., the establishment of comprehensive post-polio centers—to concentrate clinical expertise and help stimulate collaborative research in PPS.

CONFERENCE REPORTS AND DISSEMINATION

Two reports were drafted by the steering committee. The first—this report for medical care providers—offers best practices in diagnosis and care. In addition to this Preface, the report contains chapters on pathogenesis, diagnosis, clinical management, recommendations for establishment of comprehensive post-polio centers and recommendations for research. Appendix A directs the interested reader to key summaries and research reports.

The second report is a patient brochure that provides similar information in layman's language. The brochure is designed to update patients and other interested lay readers on current best practices in the diagnosis and clinical management of PPS and to facilitate communication between patients and medical care providers.

As for report dissemination, the March of Dimes is working with the International Polio Network, a St. Louis-based organization that assists and coordinates information for post-polio support groups, to distribute the reports to polio survivors and their health care providers nationwide and abroad.

The March of Dimes is also collaborating with the Ontario March of Dimes, a member organization of the Easter Seals/March of Dimes National Council of Canada, in the distribution of the reports in Canada. The committee thanks both these organizations for their invaluable help and support. In addition, the March of Dimes will distribute the reports through professional organizations, schools, and agencies in both the public and private sectors associated with the care of polio survivors. To obtain copies, see the inside front cover.

Finally, and importantly, the American Academy of Neurology, in cooperation with the American Academy of Physical Medicine and Rehabilitation and several allied health care professional organizations, will develop independent joint practice guidelines for PPS in 2001. These practice guidelines will be informed by and coordinated with the recommendations of this report.

1 PATHOGENESIS

Post-polio syndrome (PPS) is a variable combination of new progressive muscle weakness and other symptoms in survivors of paralytic poliomyelitis, with onset usually at least 15 years after the acute illness.

Acute paralytic poliomyelitis results from poliovirus invasion of brainstem and spinal cord motor neurons, and is primarily a disease of the motor unit (defined as a motor neuron and all the muscle fibers that it innervates). More diffuse nervous system involvement with encephalitis can also occur in the acute attack. Motor neuron death causes denervation of muscle fibers with resultant weakness. Recovery of muscular force after acute polio occurs by recovery of some neurons and sprouting from remaining motor axons, and by muscle fiber hypertrophy in innervated muscle. Axonal sprouting can produce reinnervation of locally denervated muscle fibers, restoring the ability to produce muscle fiber contraction. The resulting motor units can be up to 8 times normal size.

The etiology of PPS is still unclear. The most widely accepted hypothesis, proposed by Wiechers and Hubbell,

attributes PPS to a distal degeneration of enlarged post-polio motor units. The surviving motor neurons innervate many more muscle fibers than normal, and may be unable to sustain this greatly increased metabolic demand indefinitely. Terminal axonal sprouts may degenerate, producing denervation of muscle fibers. Some of these denervated muscle fibers may become reinnervated by neighboring axons, causing a continuous "remodeling" process, but some may become permanently denervated, and thus produce permanent increased weakness. This distal motor unit degeneration can also produce neuromuscular junction transmission defects, which may be a cause of muscular fatigue in PPS.

Possible contributing factors to the development of PPS may be the normal aging process, and overuse or disuse of muscles. The aging process produces a progressive motor neuron loss in polio survivors, which can contribute to the onset of PPS.

2 DIAGNOSIS

CRITERIA FOR POST-POLIO SYNDROME

1. Prior paralytic poliomyelitis with evidence of motor neuron loss, as confirmed by history of the acute paralytic illness, signs of residual weakness and atrophy of muscles on neurologic examination, and signs of denervation on electromyography (EMG).
2. A period of partial or complete functional recovery after acute paralytic poliomyelitis, followed by an interval (usually 15 years or more) of stable neurologic function.
3. Gradual or sudden onset of progressive and persistent new muscle weakness or abnormal muscle fatigability (decreased endurance), with or without generalized fatigue, muscle atrophy, or muscle and joint pain. (Sudden onset may follow a period of inactivity, or trauma or surgery.) Less commonly, symptoms attributed to PPS include new problems with breathing or swallowing.
4. Symptoms persist for at least a year.
5. Exclusion of other neurologic, medical and orthopedic

problems as causes of symptoms.

CLASSIFICATION OF PPS SUB-TYPES

The diagnostic criteria include diverse signs and symptoms. Sub-types of PPS have been proposed, but conference members found insufficient basis for specific classification.

EVALUATION OF THE PATIENT

If the above criteria for PPS are met, there is usually little diagnostic difficulty.

If the new symptoms are less specific, there may be diagnostic uncertainty. Less specific symptoms include generalized fatigue, myalgia, aches and pains, exercise intolerance and cold intolerance. These symptoms are subjective and therefore difficult to evaluate in patients with or without prior paralytic poliomyelitis. For these patients, evaluation should include a general medical workup for coincident hypothyroidism, polymyalgia rheumatica or other systemic diseases. For patients with unequivocal exercise intolerance, mitochondrial or metabolic myopathy may be considered. Because symptoms are subjective, psychological problems are possible. For those with muscle pain and fatigue, fibromyalgia is a popular but controversial diagnosis. For patients with prominent fatigue as the main manifestation, sleep-disordered breathing with consequent daytime somnolence may be considered, and spirometry and sleep studies may be in order. Fatigue can be a consequence of serious disability from muscle weakness and atrophy, but it is also often a manifestation of depression.

PPS patients may be more susceptible than others to neurological problems originating in the spine when there is scoliosis or paravertebral muscle weakness. Among the neurological disorders that may be confused with PPS are adult spinal muscular atrophy, amyotrophic lateral sclerosis, cauda equina syndrome, cervical stenosis, chronic inflammatory demyelinating polyneuropathy, diabetic neuropathy, entrapment neuropathy, heavy

metal toxicity, inflammatory myopathy, multifocal motor conduction block, multiple sclerosis, myasthenia gravis, Parkinson disease, peripheral neuropathy, radiculopathy, spinal cord tumor and, notably, spinal stenosis. If a neurologist is available, consultation is in order.

Diagnostic problems arise if there are unusual late symptoms such as dysphagia or respiratory insufficiency, but these symptoms appear in some patients deemed to have PPS, especially if breathing and swallowing were affected in the acute earlier attack.

Some clinicians with experience in PPS have identified patients who lack a clear history of paralytic poliomyelitis, have little residual atrophy, but have progressive new weakness and evidence of diffuse denervation on EMG. This syndrome is uncommon and must be differentiated from spinal muscular atrophy.

The progressive weakness in PPS is sometimes accompanied by wasting and even fasciculations. This combination may be considered PPS if there is no progression or very slow progression, but even patients with old poliomyelitis can later have motor neuron disease. In such cases, therefore, neurological consultation is mandatory. Helpful examinations include EMG, CSF examination, brain and cord MRI, as well as magnetic stimulation of the motor cortex and magnetic resonance spectroscopy for evidence of upper motor neuron dysfunction.

If new weakness is focal, the differential diagnosis includes radiculopathy, and appropriate imaging is helpful.

For difficult diagnostic problems, it is reasonable to consult a neurologist or other specialist with expertise with PPS. The development of regional PPS centers, recommended by the committee in Chapter 4, would facilitate such consultation.

3 MANAGEMENT

Best practices in care for people with PPS are still evolving. Many symptoms of PPS seem to result from motor unit degeneration, or motor neuron attrition or dysfunction, combined with normal aging. Because both of these

aspects of PPS change over time, treatment regimens are adjusted to stages of the condition. Although there is no specific treatment for PPS, most patients benefit from a management program.

An interdisciplinary approach to management, with a team of physicians and allied health care personnel, is appropriate for many PPS patients because they often have a wide variety of problems.

The team may include a primary care physician, physiatrist, neurologist, and physical and occupational therapists. Consultants may include a pulmonologist, speech pathologist, psychiatrist or psychologist, orthopedist, rheumatologist, dietician, orthotist, nurse and respiratory therapist. Management should be symptom specific.

Management should also address conditions, such as osteoporosis or pressure palsies, that may occur more commonly in people with previous paralytic poliomyelitis than in the general population.

MANAGEMENT OF WEAKNESS

Managing weakness in PPS includes various strategies: judicious exercise (see below), avoidance of muscle overuse (exertion to the point of muscle pain and fatigue), weight reduction, bracing weakened muscles, and assistive devices such as crutches, manual wheelchairs, electric wheelchairs, and motorized scooters.

Orthoses and assistive devices can also be useful in managing pain, joint deformities, and gait difficulties.

Appropriate programs can be arranged in facilities for rehabilitation medicine.

MANAGEMENT OF FATIGUE

Excessive fatigue is an elusive symptom of diverse causes. Because it is heterogeneous in origin, subjective in nature, and influenced by psychological factors, there have been many different recommendations for management. None of these has been formally tested in controlled trials.

Among the recommendations for control of excessive

fatigue in PPS are energy conservation techniques, lifestyle changes, pacing of activities, regular rest periods during the day and improvement of sleep by relaxation techniques and medications. These include antihistamines, valerian, melatonin, amitriptyline, L-tryptophan and gabapentin. Noninvasive ventilation at night may alleviate fatigue in patients with underventilation. However, none of these interventions has been evaluated by rigorous clinical trial.

Some recommendations emerge from common sense and general rehabilitation experience. For instance, energy conservation techniques include discontinuing unnecessary energy-consuming activities, such as making a bed. Seating and work station arrangements should be optimized. Individuals can take advantage of special parking privileges, sit instead of stand, move items or supplies to make them more accessible (such as having the washer and drier on the first floor rather than in the basement), and use an electric scooter for longer distances.

Lifestyle changes include changing to a more sedentary job, or working fewer hours, and discontinuing selected activities. Regular naps or rest periods during the day, especially in the early afternoon, can help in managing general fatigue, and should be encouraged for patients with significant fatigue.

Pacing (regular rest periods during activity) is useful in managing focal (not general) muscle fatigue in PPS. Some experts recommend use of a formal rating scale by which patients can monitor their exertion (see an example in Appendix B) to help avoid fatigue.

EXERCISE AND MUSCLE OVERUSE

Almost everyone can benefit from some form of exercise. To be safe and effective, exercise programs for PPS must be cautiously customized to each person's needs, residual strength of individual muscles, and symptom patterns. For many individuals, the level of exercise may be nothing more strenuous than gentle stretching or various types of yoga. For others, it may be more vigorous, even including some forms of aerobic training.

There is no proof from prospective clinical studies that "muscle overuse" increases or creates persistent muscle weakness. Many physiatrists teach patients to avoid muscle overuse, manifest as aching upon exertion. Patients are advised that this is a warning sign, and that they should not exert themselves to this point. This may require reduction in activities, pacing, and education about energy conservation techniques. Problems with mobility and pain are addressed to be certain that the patient is safe and comfortable. After the patient has learned to monitor and manage weakness and fatigue, an individualized exercise program is started. However, additional exercise should be completely avoided in patients who are too weak and fatigued, and are already spending most of their energy simply performing activities of daily living.

When starting an exercise program, patients should be monitored to ensure that exercises are done correctly, and that there are no adverse effects. After patients understand the exercise program, and can monitor themselves, less frequent follow-up is necessary.

MANAGEMENT OF PAIN

Pain in post-polio patients can be due to diverse causes, including joint and soft tissue abnormalities, muscle abnormalities, and pressure injuries of nerves in the arms and legs, including crutch injuries to the brachial plexus and carpal tunnel syndrome. Treatment depends upon the cause of pain.

Overuse is thought to be a common cause of pain in post-polio patients. The location of pain depends upon the patterns of motion and occurs more frequently in the legs and low back in patients who are still walking. Pain affects the arms more often in patients who use wheelchairs or crutches.

Some patients describe "post-polio muscle pain" as an aching sensation similar to that experienced at the time of acute polio, usually occurring late in the day. Patients may also experience muscle pain with activity, or muscle cramps. Management includes lifestyle changes, reduction of activity, pacing, stretching, and use

of ice or moist heat and assistive devices. Fibromyalgia is a controversial diagnosis, whose treatment can include amitryptiline, cyclobenzaprine, fluoxetine, aerobic exercise and other measures. Analgesics can be given but benzodiazepines and opiates should be avoided.

Joint and soft tissue pain can be due to osteoarthritis, tendonitis, bursitis, ligamentous strain secondary to joint deformities and previous surgical joint fusions (arthrodeses). Osteoarthritis of the hand and wrist occurs commonly in post-polio patients, more frequently in patients over age 50, and is usually associated with leg weakness, locomotor disability, and high usage of assistive devices.

Many of the causes of pain are treatable with modification of extremity use, physical therapy, strengthening (when possible), orthoses to control joint deformities and difficulties with previous joint fusions, assistive devices, nonsteroidal anti-inflammatory drugs, acetaminophen and, rarely, steroid injection, or surgery. These decisions warrant the attention of a specialist.

Superimposed neurological disorders occur commonly in post-polio patients, and can be a cause of pain. Use of assistive devices is a major risk factor for carpal tunnel syndrome. Spinal stenosis is another cause of back and leg pain. Treatments for carpal tunnel syndrome include splinting, use of pads on canes or crutches to place the wrist in a more neutral position and increase the weight-bearing surface of the hand, steroid injection and carpal tunnel release. For patients with low back pain, lumbosacral orthoses, shoe lifts, back or pelvic supports, non-steroidal anti-inflammatory drugs and physical therapy are options, before surgery is contemplated. Spinal stenosis can be treated with exercise, use of a cane or other walking device, transcutaneous electrical nerve stimulation (TENS), lumbosacral orthoses, epidural steroids and, in some cases, surgery.

MANAGEMENT OF RESPIRATORY PROBLEMS

Respiratory problems in PPS occur mainly in those who required ventilation at the time of acute poliomyelitis, but may also occur in those who did not recognize earlier respiratory involvement.

Respiratory muscle weakness is the main cause of respiratory insufficiency in post-polio patients. However, other contributing or causal factors include central hypoventilation due to previous damage from bulbar polio, scoliosis, kyphosis, sleep-disordered breathing, obesity, other lung diseases, smoking and heart disease.

Any contributing factors to respiratory problems that can be reversed should be eliminated or treated. All patients with respiratory dysfunction should receive pneumococcal vaccine and yearly influenza vaccinations. Ventilatory assistance may help those with hypoventilation or sleep-disordered breathing. Non-invasive methods are preferred because of better tolerance and lower complication rates. Supplemental oxygen should usually be used only for acutely ill hypoxemic patients with intrinsic lung disease, after alveolar ventilation and airway secretions have been optimally managed with respiratory muscle aids.

Sleep-disordered breathing, common in post-polio patients, should be identified and treated. Untreated sleep apnea can eventually result in cardiopulmonary failure.

Glossopharyngeal breathing, sometimes called "frog breathing," is a method of projecting boluses of air into the lungs by using the tongue and pharyngeal muscles, and can be taught to ventilator-dependent patients to allow periods of ventilator-free breathing.

MANAGEMENT OF DYSPHAGIA

Symptoms of difficulties in swallowing (dysphagia) include complaints that food fails to pass down the throat or esophagus, coughing or choking during eating and taking longer to eat meals. Dysphagia can occur without a history of previous bulbar polio.

The most common cause of dysphagia is weakness of pharyngeal or laryngeal muscles, but other disorders, such as structural lesions not related to polio, may cause or contribute to the problem. Management can include changing or restricting the diet to "safe" substances, such as purees and thickened liquids (usually when the need is determined by videofluoroscopy); use of special

breathing techniques; use of special swallowing techniques (such as turning the head to one side while swallowing); and avoiding eating when fatigued, or taking larger meals earlier in the day and smaller meals later.

MANAGEMENT OF PSYCHOSOCIAL DIFFICULTIES

PPS patients may have great difficulty in adjusting to this second, unexpected disability. In addition, because polio is now considered to be a conquered and largely forgotten disease, patients may have difficulties due to general lack of knowledge among physicians about the disease and its late consequences.

Individuals with PPS seem not to have higher levels of depression than the general population. However, when depression occurs, its recognition and evaluation can be complicated by post-polio symptoms, such as fatigue, that overlap with those of depression. Also, depression compounds fatigue, pain, and lifestyle management, and so is an important primary disorder that should be routinely considered in PPS patients.

Of likely special relevance to PPS patients is the general fact that reduction in activity is a risk factor for fatigue. A reported increase in fatigue after reduced activity, therefore, suggests reassessment for depression.

Treatment of psychosocial difficulties related to PPS can be accomplished with an interdisciplinary approach that includes a post-polio mutual support group, psychologist, social worker and psychiatrist, as necessary.

DRUG THERAPY

As in other aspects of PPS management, there has been little formal evaluation of drug therapy. Although certain medications have been reported to be helpful for primary symptoms of PPS, rigorous studies are required. These should include randomized, placebo-controlled, double-blinded studies designed with adequate power and disease-specific, responsive outcome measures. Adequate power requires multicenter trials. This purpose may best be served by establishment of comprehensive post-polio

centers, as recommended below.

Various medications have common or rare adverse effects that mimic new or worsening PPS symptoms such as weakness or fatigue. Such symptoms should therefore be assessed with attention to medications that a patient is taking, by prescription or otherwise.

NUTRITION

Except for weight control, no specific nutritional intervention is recommended.

Any limitation of physical activity is likely to lead to excessive weight gain. This can cause or aggravate features of PPS in at least two ways. It can invite fatigue and thereby further reduce physical activity, and so result in muscle atrophy from disuse. It also can lead to damage of skeletal structures, especially in a context of muscle weakness or easy fatigability. Diminished weight-bearing exercise is also a major risk factor for low bone density, with consequent limb fractures and compression of vertebrae. As in the general population, weight control by polio survivors is difficult, but of special importance that may call for professional nutritional consultation.

4 RECOMMENDATIONS FOR ESTABLISHMENT OF COMPREHENSIVE POST-POLIO CENTERS

The committee recommends creation of regional centers with comprehensive expertise in all aspects of PPS. It is desirable that each center be a physically unified facility. Where that is not possible, a "virtual" center should be created, comprising closely coordinated activities and active outreach among a network of appropriate other health care facilities and professionals within a region.

Chief components of each center should be services to patients and their families, teaching, research, and cooperation with other centers.

SERVICES

Centers are likely to be organized and led by a neurologist or a specialist in rehabilitation medicine. In some locales, an orthopedist may take the lead. All necessary specialties should be represented. Consultation should be available for orthopedics, respiratory medicine, psychiatry and pain management. Allied health professionals would provide expertise in physical and occupational therapy, orthotics, psychology, speech pathology, nursing, respiratory therapy, nutrition, social work, swallowing and assisted technologies (wheelchair and seating clinic). Diagnostic facilities include EMG; pulmonary function testing; sleep and swallowing studies; a blood laboratory; and radiologic capabilities for magnetic resonance imaging, computed tomography and X ray. Patients should have ready access to educational materials and instruction. Buildings and facilities should be fully accessible to the disabled.

TEACHING

Centers could be responsible for training physicians and other health care workers in the management of PPS, could educate patients as well as other physicians and health care workers, and could develop support groups to address the concerns of their patients.

Curricula and training in relevant medical specialties, including but not limited to neurology, orthopedics, physiatry and geriatrics, should be targeted for inclusion of specific information on PPS. This requires active participation by relevant professional societies.

RESEARCH

Centers should be actively engaged in research, publishing their results.

COOPERATION

Cooperation among the centers could be facilitated by a central administration that would make public the

number, location and activities of the several centers, including research in progress.

5 RECOMMENDATIONS FOR RESEARCH

In reviewing the evidence for this report, the committee recognized that there remain significant gaps in knowledge and understanding of the epidemiology, pathogenesis, diagnosis and optimal management of PPS, all of which need intensified research. The following recommendations for research are designed to help fill these gaps.

DIAGNOSTIC CRITERIA AND NOMENCLATURE

Because there is no biomarker for PPS, diagnostic criteria and nomenclature must be standardized, with a validation study to ensure inter-rater consistency, as a next step toward defining PPS and its clinical subgroups, and as a prerequisite for further research. It is necessary to make measurements and develop normative data, specific to polio survivors, of progressive weakness, fatigability, quality of life, and similar objective and subjective aspects of PPS. Definition of a post-polio muscular atrophy (PPMA) subgroup requires study. For study purposes, there need to be established consistent ways to separate neurological from musculoskeletal disabilities.

EPIDEMIOLOGY AND HEALTH SERVICES RESEARCH

The impact of PPS on individuals and society is largely unmeasured. This fact hinders assessment of needs for resource allocation. To address this problem, the committee recommends the following actions:

- Analyze in depth and publish analysis of the Polio Survivor Data Set from the 1994-5 National Household Interview Survey (data made available by the National Center for Health Statistics).
- Study the total costs of PPS to the national economy.
- Conduct a population-based study of the quality of care and quality of life among polio survivors generally and

PPS patients in particular.

- Assess unmet health care needs of polio survivors, with emphasis on patients' perceptions of those needs.
- Evaluate PPS informational needs of practicing health care professionals.
- Evaluate international disparities in reported prevalence of PPS—for example, the very low rates reported in India, Italy and France. Such cross-cultural studies are needed to ascertain the true prevalence and costs of PPS globally.

PATHOPHYSIOLOGY

The committee recommends creation of a tissue bank for autopsy specimens of PPS spinal cord and other tissues. Volunteer patients could be educated about the value of their medical histories and findings on examination, and about the importance of information from postmortem examination. Instruction for the handling of tissues could be provided by a central administration. International collaboration in tissue banking is highly desirable.

Transgenic mice have been created that express the human cellular poliovirus receptor and thus are susceptible to paralytic poliovirus infection. Long-surviving animals should be explored as a possible model of PPS.

The possible roles of poliovirus persistence or of an immune component in the etiology of PPS are of low priority for further investigation because studies to date have been inconclusive or negative.

MANAGEMENT

Evaluating Interventions

PPS-specific, responsive, reliable and valid measures should be developed to assess disease progression and effectiveness of interventions. When validated, these should be widely disseminated and considered a standard of care.

There is need for rigorous, multicenter appropriately controlled assessment of the efficacy of interventions

now in use for PPS patients—i.e., of individualized, interdisciplinary rehabilitation programs, and of the individual components of rehabilitation programs, such as exercise, pacing, energy conservation, orthoses and use of assistive devices. These studies should include data on cost-effectiveness of specific interventions, as a basis for gaining approval by third-party payers.

Effective treatment is an important goal. Evaluations should include multicenter, doubleblind, controlled trials. Agents that might be evaluated include riluzole, neurotrophic factors, stem cells, modafinil, pyridostigmine and other agents.

Psychosocial Issues

The relationship between psychological disturbances and PPS is neither well-understood nor appreciated. Depression, sleep disturbances and diminished cognitive functions have been reported by PPS patients and deserve careful examination. The committee recommends that the following be undertaken in a sufficient sample of PPS patients:

- A rigorous evaluation of the role of depression and its relationship to fatigue.
- A rigorous evaluation of the role of sleep disturbances as a factor in fatigue.
- A systematic assessment of cognitive functions such as attention and memory.

Appendix A

SUGGESTED READING

Bach JR. Management of post-polio respiratory sequelae. *Annuals of the New York Academy of Sciences* 1995; 753:96-102.

Dalakas MC. Why drugs fail in postpolio syndrome. *Neurology* 1999;53:1166-1167.

Dean E. Clinical decision making in the management of the late sequelae of poliomyelitis. *Physical Therapy* 1991;71:752-761.

Halstead LS (ed.). *Managing Post-Polio: A Guide to Living Well with Post-Polio Syndrome.* Washington, DC: NRH Press, 1998.

Halstead LS, Silver JK. Nonparalytic polio and postpolio syndrome. *American Journal of Physical Medicine and Rehabilitation* 2000;79(1):13-18.

Jubelt B, Agre JC. Characteristics and management of postpolio syndrome. *Journal of the American Medical Association* 2000; 284:412-414.

Maynard FM, Headley JL (eds.). *Handbook on the Late Effects of Poliomyelitis.* St. Louis, MO: Gazette International Networking Institute, 1999.

Post Polio Information Package for Health Care Professionals. Toronto: Ontario March of Dimes, 2000.

Trojan DA, Cashman NR. *Current Trends in Post-Poliomyelitis Syndrome.* New York, NY: Milestone Medical Communications, 1996.

Appendix B

BORG RATING OF PERCEIVED EXERTION

To avoid excessive muscular and general fatigue, patients can monitor their perception of fatigue by using the Borg Rating of Perceived Exertion (RPE). This scale is a useful measure of effort, and therefore an aid in limiting exercise in PPS and other conditions. To prevent excessive fatigue, injury and overuse syndromes, PPS patients should end their activities at an RPE level of 14 or lower. (There are other scales for assessment of fatigue that may be equally useful in management of PPS patients.)

06
07 VERY, VERY EASY
08
09 VERY EASY
10
11 FAIRLY EASY
12
13 SOMEWHAT DIFFICULT
14
15 DIFFICULT
16
17 VERY DIFFICULT
18
19 VERY, VERY DIFFICULT
20

31-1522-01 2/01