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SAVE OUR SHOULDERS: A GUIDE FOR POLIO SURVIVORS

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Many polio survivors report new symptoms as they age. Some of the more common symptoms include loss of strength, fatigue, muscle twitching, cramping, and muscle or joint pain. These symptoms, combined with the muscle weakness caused by the polio virus, often lead to problems with activities like climbing stairs or getting up from a chair or sofa.

When muscle weakness and/or pain is present in one area of the body, people often compensate by putting more stress on another area of the body. For example, polio survivors who have weak leg muscles often rely on their arms to assist with mobility-related tasks, such as pushing off of the armrests of a chair when getting up or pulling up on the railing when going up a flight of stairs. This behavior can increase the risk for symptoms of shoulder overuse. These symptoms can include pain, swelling, weakness, and loss of motion or function in one or both the shoulders.

The purpose of this manual is to help polio survivors become more aware of the behaviors that can cause shoulder overuse and some of the treatment options that are available. While the information included in this manual is targeted to polio survivors, it will be useful to anyone with shoulder problems. Shoulder dysfunction is an orthopedic problem with similar symptoms in people with and without a history of polio. Treatment approaches are individualized and focus on minimizing or eliminating pain while maximizing function.

General Information About the Shoulder

The shoulder is made up of many muscles, bones, tendons, ligaments, and nerves that work together to help with movement of the arms. The shoulder joint is the only joint in the body that allows movement in a complete circle.

The bones that are part of the shoulder include the scapula, the clavicle, and the humerus.

The scapula is the bone that makes up the back of the shoulder. The clavicle is located in the front of the shoulder, and the humerus is the long bone in the upper arm that inserts into the shoulder socket. These bones all work together to make the shoulder a functional unit.

Many muscles support the shoulder by helping to keep the bones in proper position and aiding in moving the arms. The major muscles that help to keep proper bone position are known as the rotator cuff. These muscles are called a cuff because they give support to the shoulder joint.

A rotator cuff tear is the most common shoulder problem and is found in almost half of the people who seek help for a shoulder injury.

Facts About Shoulder Problems

- Shoulder problems are one of the most frequent complaints in the post-polio population.

- Shoulder issues are also common among older adults without a history of polio, occurring in about 30% of people who are 60 years and older.
- Shoulder problems can increase with age and changing levels of physical activity.
- People who are involved in sports are not the only ones to suffer from shoulder problems.
- Shoulder problems are common in those with long standing medical conditions, such as post-polio syndrome, arthritis, spinal cord injury, stroke (CVA), or multiple sclerosis.

Our research has shown that polio survivors who have weakness in their legs or who are overweight are at high risk for developing shoulder problems.

Risks for Developing Shoulder Problems

You may be at risk for developing shoulder problems if you:

- Had Polio
- Are over 60 years old
- Use your arms to compensate for weakness in other areas
- Use an assistive device such as a cane or walker
- Are overweight
- Had previous injury or trauma to your shoulder(s)
- Perform tasks that involve repetitive shoulder motion
- Perform activities that require frequent reaching overhead
- Feel pain, discomfort or muscle fatigue in your arms when doing your normal activities
- Use improper lifting techniques or poor body mechanics when performing tasks at work
- Do not do proper warm-up or stretching before physical activity

Common Shoulder Problems and Possible Causes

- Pain may result from:
 1. Fractures (broken bones) caused by a fall, bone disease such as osteoporosis, or medications.
 2. Impingement Syndrome (pain in the shoulder due to incorrect shoulder movement) caused by overuse, trauma, poor posture, weakness in some muscles of the shoulder, or bony changes.
 3. Tendonitis (swelling of the structure that joins muscle to bone) caused by overuse, trauma, or a disease process such as a stroke or polio.
- Weakness may result from:
 1. Disease processes such as polio, stroke, or nerve damage
 2. Inactivity

- Movement Difficulty may result from:
 1. Frozen Shoulder (unable to move arm because of severe pain which leads to stiffness in the joint) caused by an infection, injury, lack of movement in arm, or severe pain in shoulder.
 2. Bony changes caused by arthritis, fractures, calcium deposits, or birth defects
 3. Swelling caused by surgery, trauma, lack of motion, or a disease process such as a stroke.

Pain, weakness and movement difficulty frequently develop due to a cycle of symptoms. Individuals limit their use of a painful shoulder, which results in stiffness and loss of strength. This leads to increased pain and continues the cycle of pain, weakness, and loss of motion. An effective treatment program will focus on breaking this cycle.

What to Do if You Have a Shoulder Problem

It is important that you seek the care and advice of a medical professional qualified to diagnose and treat shoulder problems. Depending on the problem that you are having, you may be referred to a specialist.

Regardless of who evaluates your shoulder, information will be collected about your specific shoulder problem. If you are a polio survivor, it is important to seek the help of a healthcare provider who is informed about polio and its effects on your treatment and recovery.

In order to get the most helpful information, you may be asked questions such as:

- Tell me your medical and polio history, and medicine you are taking.
- Tell me about your shoulder problem.
- Have you ever had this problem before? – what did you do for it then... Did you have any tests done?
- Have you had therapy, surgery, or other treatments for the same problem?
- Does anything make your shoulder feel better? ... Worse?
- Does your shoulder bother you more at a particular time of the day? ... When?
- Does your shoulder problem wake you from sleep?
- Does the problem affect different areas of your body? ... Where?
- Do you need help with getting washed, dressed, or household chores?
- Do you use assistive devices such as a reacher or a long handled sponge because of shoulder problems?
- If you need to take medication for your shoulder problem, how often are you taking it? ... Does it help?

Shoulder problem treatment options

Many approaches can be used to treat shoulder problems. Some examples of treatment options are:

- Rest
- Cold Pack or Hot Pack
- Massage

- Ultrasound
 - Aquatic Therapy
 - Electric Stimulation
 - Anti-inflammatory medicines or cortisone injections
 - Pain Management at a pain clinic
 - Modifications (home/work; adaptive devices or equipment; lifestyle changes)
 - Surgery
1. The most common treatment for a shoulder problem begins with rest, ice, and heat, and the use of anti-inflammatory medicines. If these conservative treatment options do not help, the doctor usually recommends cortisone injections to the painful area. After injections, the physician will often refer you to physical therapy for evaluation and treatment.
 2. In physical therapy there are several treatment techniques that can be combined to relieve the pain, and increase the strength and use of the arm. Physical therapists may use exercise, stretching, ice, heat, ultrasound (a deep heat), massage, electric stimulation (electrical current used to decrease pain or increase movement), water therapy, or hands on techniques to help your shoulder problem.
 3. The final option recommended by the doctor is usually surgery. Most doctors will try non-invasive options before recommending surgery.
 4. The success of treatment lies with the patient and the health-care provider working together so that the greatest benefit of the treatment goals can be reached.

The Importance of Education and Exercise

Most treatment plans will include education and exercise.

Why Education?

- Learning about the problem you are having with your shoulder and how it is affecting you will make you more aware of things that can aggravate the problem.
- Understanding why a treatment plan has been chosen, and how to follow it will improve your ability to get the most benefit from the plan.
- Involving you in the treatment process is important so that further injury can be minimized and proper treatment of the current problem can be set up based on your needs.

Why Exercise?

- It is inexpensive.
- It is easy.
- It doesn't take much time.
- It helps to maintain or increase your flexibility, balance, coordination, and stability.

- It helps to maintain or increase your strength and endurance.
- It helps keep you independent with daily activities.
- It helps you control weight and body fat.
- It helps to reduce stress.
- It helps increase energy levels.

It is important to work with a health care provider who is educated in the effects of polio and exercise.

Before beginning your exercise program it is important to remember:

- Exercise is not always appropriate for every shoulder problem or every individual. In some cases, some exercises can actually do more harm than good.
- Some medical problems can influence which exercises are appropriate for you to perform.
- It is important to get clearance from your doctor BEFORE you begin any exercise program.

General Information on Exercise Programs

- The benefits of exercise can be achieved by doing as little as 30 minutes of moderate physical activity three to five times a week.
- Exercise can be a source of enjoyment for many individuals. Exercise can be performed in a group or alone

Before you begin an exercise program you should always get approval from your doctor to participate. This is especially important if you have any medical problems.

About Exercise Programs in General

- An exercise program consists of three key phases.
 1. Warm up
 2. Exercise
 3. Cool down
- The warm-up and cool down involve stretching, getting your blood flowing, and your heart pumping.
 1. The warm-up gets your body ready for exercise because it increases the blood flow, preparing the heart and body for exercise.
 2. Stretching prepares your muscles for exercise.
 3. Stretching involves moving your body part to the point of feeling a pull in the muscles. It is important not to stretch your muscle beyond the point of pain . If you feel pain, stop the stretching motion.
 4. Most stretches are held for 30 seconds for 3 repetitions.
 5. The cool down slows your body down from the exercise and prepares your body for rest. It slows your heart down

gradually, instead of rapidly ending the session after strenuous activity.

- There are many different ways of exercising. There are aerobic, strengthening, range of motion, and flexibility exercises.
- For the best results, follow recommended precautions.

Polio Survivors Need to Pace Themselves

Stop exercising if you are tired or have increased pain and exercise slowly. One of the most important tips for polio survivors is to respect their body signals, and adapt their lifestyle to minimize their limitations. It is important for polio survivors to respect their fatigue and take frequent rest breaks when performing an exercise program.

Exercise Tips:

- Exercise slowly and take breaks between each exercise.
- Do not hold your breath while exercising (count out loud to encourage breathing.)
- Do not continue to exercise if you get sharp or new pain in the area you are exercising.
- If the exercise increases your pain, stop the exercise, or perform the exercise in a pain free range of motion.
- Do not push to perform the extreme of the motion or exercise.
- Stop exercising if you have muscle twitching, muscle cramping, or muscle fatigue that does not go away after two minutes rest.
- Polio survivors should not exercise to the point of fatigue.
- It is important to recognize that your fatigue may not appear immediately. It may show up at the end of the day or the following day. You should look at your recent activities and modify them if your fatigue increases or occurs at the end of the day or the next day.
- It is very important to **listen to your body** for signs of tiredness, pain, unusual muscle twitching, inability to perform a task that you can normally do, or the need for help with a task that you can usually do without help.

These are signs suggesting muscle fatigue or overuse, and continuing activities that cause these signs could be harmful.

- Most individuals who have a history of polio will have a system that works for them; some prefer to exercise in the morning because they have the most energy, others exercise at the end of their day because they can rest after the program, and still others may 'split' their exercise program into two sessions a day.
- Everyone will have his or her own way to do things, and when it comes to exercising, each person needs to find a system that works best for them.
- ***However, it is crucial to prevent fatigue because it may do irreversible harm to your muscles.***

Tips to Avoid Making Your Shoulder Problem Worse

There are many changes that you can make in your normal activities that may help decrease problems you may be having with your shoulder(s). Below are examples of simple things that can be done to reduce the stress placed on your shoulders.

REMEMBER: you are the one who knows your body best. Respect and learn to listen to your body's signals or limits.

- Rest or take a break from any sporting or leisure activity that aggravates your arm or causes shoulder pain.
- Limit prolonged activities by taking frequent rest breaks.
- Avoid excessive overhead activities with your painful arm.
- Avoid activities or movements that increase your pain.
- Take pain and anti-inflammatory medicine as prescribed by your doctor to control the pain and allow you to function.
- When you enter a room take a minute to look at the chairs and choose the highest one with armrests so that you do not have to put so much pressure on your shoulders when rising.
- Avoid regular stalls in public bathrooms, as most of the toilets are very low; handicap stalls have higher toilets and grab bars.
- Modify or seek help with work or home settings to avoid improper positioning and poor body mechanics.
- Keep items used most frequently in cabinets below your shoulder level.
- When possible sit in a chair with armrests to keep your arms supported and to lessen the amount of stress on your shoulders when you 'push up' to stand.
- When doing tasks at a table sit on a cushion to raise your body.
- When doing a sitting job, try to have everything you need at hand to avoid pushing yourself out of the chair more than necessary.
- Do not push a vacuum or lawn mower if you can get someone else to do it.
- Ask for help with heavy lifting tasks to prevent injuries.
- Use adaptive equipment to perform your activities of daily living, such as a reacher for getting items from a high shelf, or a long handled sponge to wash hard to reach body areas.
- Put your shirt, blouse, or jacket on your painful arm first.
- Do not sleep on your painful shoulder.
- Put a pillow between your arm and body while lying on your non-painful side.
- While lying on your back, put a pillow under your painful shoulder to support your arm.

Conclusion

Most people can easily identify what tasks are difficult for them to do or what activities cause a problem with their shoulders. For instance, many people have trouble climbing stairs or getting up from a chair. However, many do not realize just how much stress and strain is placed on the shoulders when doing these activities.

It is our hope that the information in this manual has helped you learn about your shoulders and has improved your awareness of some of the things that can increase the stress and strain placed on them. We know this information is important because keeping your shoulders as healthy as possible can help you maintain your

independence.

The information provided is a basic guide for shoulder problems. If you have a shoulder problem, you should be evaluated by a healthcare professional that can work with you to develop a plan that best meets your needs.

The information contained within this article is for reference only and is not a substitute for professional medical advice. Before beginning any exercise program consult your physician.

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SOME RECENT THOUGHTS ABOUT POLIO AND POST POLIO SYNDROME

By Tim Zingale

Some might view what I am about to say as complaining or feeling sorry for myself. And it could be that, but I think it is more of a reflection of how I have seen my life. As I watched my children grow up, as I am watching my grandson grow up, I am elated at how they can run, jump, ride a bike, do all those things that I have never been able to do.

As I grow older, it is strange that I miss those things more now than ever. I don't know why, but I do. Maybe it is because as I am in my late 50's, I see my life winding down, maybe it is because I have been more reflective with our house hold being empty now.

I don't know, but I do wonder now what it feels like to run and feel the wind in your face and your hair being blown back by the wind. I wonder what it is like to jump and have both feet off the ground with nothing holding you down. I wonder what it is like to be free on a bicycle, to pedal fast and feel the wind, and the freedom of moving that fast down the street or sidewalk.

I wonder what it would be like to go down a flight of stairs, not hanging onto the rail, not taking one step at a time, but running down, or jumping down those final few steps.

I wonder what it would be like to walk with both legs being able to bend at the knee, as I wrote a long leg brace that locked at the knee when I was walking.

I remember when I was in 5th grade the teacher asked us to write about something we would do. some wrote about being a fireman, or policemen, or some other career, but I wrote about running and jumping, and she did not know what to make of my story.

People do not realize what it is like not being able to do those things that they take for granted.

I just wonder about these things now more than I ever have. I wonder, do you?

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The very, very best thing to help with fatigue . . . is having somebody do all the hard stuff so I can play on my computer.



Some people try to turn back their odometers. Not me. I want people to know why I look this way. I've traveled a long way and some of the roads weren't paved.

EMOTIONAL IMPACT

By Margaret E. Davis

Yesterday I was hit by a ball of anger that I have not experienced in a long, long time. Pure hard cold anger that has caused me to have a huge temper tantrum. This is the emotional impact that PPS has in my life at times. I had dealt with this anger on a large scale before I found out that I had PPS.

All I knew at the beginning was that I was losing life as I had known it and I didn't know why. When one of my doctors talked about putting me in the hospital entirely for depression, I went into a huge rage. I knew I was angry. Big time angry but I knew I had a problem. The problem was physical. No, I couldn't explain why I would vomit so hard and long that I would end up in the hospital. (I vomit when I get extremely fatigued.) I also have SLE (Lupus.) Doctors blamed my problems on that until my lab reports started showing that my Lupus was coming under control.

I couldn't explain why I fell several times a day. I couldn't explain why I was so exhausted by the time I got to work that I couldn't stand long enough to look up documents in the files for the boss. My supervisor started doing the stand up stuff for me if I needed to stand when I first got to work. I couldn't explain why I reached the point during each day when I could not hold a pen and there was no way I could put enough pressure to get copies through the multiply carbons required. Then, the fastest, most accurate key punch person in the office started making errors. Why in hell was my left hand no longer working properly on the computer?? Angry?? Yes!! This was in the late 80's. Thank God not all of my doctors turned their back on me.

But it was an old friend who brought a newspaper to my husband Robert and showed him a long article about PPS. This lady is about two or three years older than me. She was a neighbor when I had Polio. She remembered. She knew of the problems I was currently having. That was amazing. I read and reread that paper. I was reading about people who were having the same problems I was having. Doctors had treated them as the one doctor had treated me.

There was a phone number for more information. I called. The lady who answered spoke in low tones. She had trouble breathing. After a few questions she gave me a crash course in making the best of it and resting more. She frightened me. I had a lot of but, but, but, and she was reaching the point she couldn't talk much longer and with much irritation she explained that she could no longer wipe her own butt. She asked me if I wanted to be in the same condition!! Talk about getting my attention - she got it!

Within the same week, another friend brought me a smaller article from a magazine. It was small enough I knew my good doctor would read this all the way through. The article and I went to the doctor. He read every word twice! When I left his office, I had an appointment with a Neurologist who had just returned from a conference about PPS. And that was the beginning of a new way of life. I learned quickly. The

Neurologist used me to teach others. But, I did not talk with this doctor or any of my other doctors about the anger I was dealing with.

I was very busy adjusting, adjusting and helping others do the same. Even so, days would come when I would struggle with the emotional impact of all of this. My response was cold, hot anger! I learned to tell my family that I was not angry with any of them. I was having trouble dealing with all the changes in my life.

Yesterday, my son, daughter-in-law and grandson came for a visit. They brought a blackberry cobbler that they had picked the berries on their property and son had baked the cobbler. We had a perfect visit. It was too hot, but I went outside to check on my flowers. I came inside to cool off once and went back out and did some transplanting. Yes, this kind of work is demanding but it is one of the things I love and can still do within limits. LIMITS -LIMITS-LIMITS.

The old anger hit me hard and unexpected. Robert thought he had done so mething. The dogs went to another room. I explained my emotions to Robert. Robert likes to fix this!! My butt hurt from sitting so long in my wheelchair. My back hurt. My shoulders hurt. I could deal with the physical pain.

The emotional impact – that's another thing. You can't rub your emotions. You can't put a heating pad on them. I was dealing with too much anger to go to bed where I needed to be. This was the first time this amount of anger has boiled within me in a long, long time. I didn't see it coming. I was shocked. I was disappointed in myself.

I had a visit with myself. OK, so it's been a long time. Even so, I still remember my other life. I wanted my other life back. There was no way to get it back. I like to get what I want. Daylight was about to peek upon me before I went to bed. But, me, myself and I had come to terms with each other.

I wish I could honestly say that I'm so happy and bubbly and life looks so good this day. That would be a lie and that would be pushing my emotions in the corner instead of dealing with them. I will not push my emotions into a back corner of my mind. I will face them. I will be fine. I've fought the anger before.

Being me, I don't expect to ever be thankful that I have PPS and SLE. Being me, I will survive and laugh and play and work and enjoy life. The rocks, bugs, flowers, family, sun, rain, snow and friends and the many, many wonders of life. I will enjoy them. I will be thankful for my wheel chair and all the other items that give me such a full life.

Why do I share this?? We have some new members. I want to tell you, hey, it's ok to be upset with the changes in your life. You will react in your own way. We are all different even though we have so much in common. I also want you to know that you can share your problems with the emotional impact just the same as you can share your physical pain and just the same as you can share your smiles.

All my love to each of you, be you new to PPS or be you an oldie.

Note: Margaret gave the Lincolnshire PPS newsletter permission to use this text from an e-mail she posted to the St. Johns University Polio E-mail List. To correspond with Margaret write to : medavis@copper.net

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RESEARCH I: WHAT IS BEING DONE? WHAT NEEDS TO BE DONE?

Who Is More Likely to Report PPS? Results of the 94/95 NHIS

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Background: Since the 1980s, researchers have been working to quantify the number of polio survivors who are affected by Post Polio Syndrome (PPS), and to identify what factors may put some survivors at risk for disease. Relying primarily on convenience samples of polio survivors and/or PPS patients, estimates of the prevalence of PPS within the survivor community have ranged from 20% to more than 70%. Risk factors that have been identified in various studies have included gender, the extent of the survivor's original polio infection, and the polio survivor's personality (or Type A behaviors in survivors). Common drawbacks of these studies are that they have not included data that are representative of all polio survivors, and have generally only studied one risk factor at a time.

Methods: In 1994 and 1995, the National Health Interview Survey (NHIS), a national health survey that samples a representative group of the U.S. population on an annual basis, identified and asked questions of a national sample of polio survivors. Information was collected about survivors' polio infection, current health status, experience with PPS, and personality traits. These data were used to determine the national prevalence of PPS, and to analyze what factors are more likely to be reported by survivors with PPS. A multivariate model, which included physiological, social, and personality variables, was constructed and tested using logistic regression to determine differences between survivors who reported a diagnosis of PPS and those who did not.

Major Findings: In this nationally representative sample, while nearly 25% of survivors reported that they believed they suffered from PPS, only 11% had been diagnosed with PPS by a physician. In multivariate analyses, the following characteristics were found to increase the odds that a survivor would report a medical diagnosis of PPS: having more years of education, having contracted polio as an adult, having more than five muscle groups affected by polio, and having required hospitalization for polio. An increasing score on the NHIS Type A-like personality traits scale was associated with decreasing odds of reporting a diagnosis of PPS. Patient gender was not associated with diagnosis of PPS; there were not enough cases of non-white survivors diagnosed with PPS in order to draw conclusions about the relationship between race and diagnosis of PPS.

Implications for Future Research and Practice: While physiological features of survivors' original polio infections are associated with risk of PPS, there are other, non-physiological characteristics that warrant further research to ascertain their relationships with PPS. Race/ethnicity should be explored in future research models as well, in order to understand how the prevalence of and experience of PPS may differ between racial/ethnic groups. Finally, the theory that all polio survivors share a common, Type A personality that contributes to the development of PPS should be examined in larger samples of polio survivors in order to ascertain whether this relationship exists and in what contexts.

RESEARCH I: WHAT IS BEING DONE? WHAT NEEDS TO BE DONE?

A Mouse Model of Post-Polio Syndrome

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This project involves the development and characterization of a mouse model for Post-Polio Syndrome (PPS). An animal model may allow a determination of the cause(s) and therapeutic interventions for PPS. Patients with PPS develop new late weakness many years after their acute poliomyelitis. A similar picture is seen in this mouse model.

Mice were inoculated intracerebrally with 1011.0 TCD₅₀ of the attenuated W -2 strain of type 2 human poliovirus (PV2/W -2). Surviving paralyzed mice were examined at 4, 8 and 12 months for the development of new weakness. Strength and new weakness were determined by using the MRC manual muscle testing scale. Mice developing new weakness, stable paretic mice and normal controls were euthanized at 12, 24 and 48 months post-infection for pathologic and virologic changes. Mice were perfused with formaldehyde for pathologic studies. Other mice were euthanized and spinal cords/brains removed for RT-PCR for viral RNA.

In the initial cohort of 65 inoculated mice, 49% (32 mice) died during the acute infection (1-15 days after inoculation), 9% (6 mice) survived without paralysis and 42% (27 mice) survived with paralysis. Of the 27 mice with residual paralysis, 10 died from anesthesia during EMG testing. At one year, of the 17 surviving paretic mice, 4 had flaccid paralysis without recover (25%), 7 were stable (41%) and 6 (35%) developed new late weakness. In second cohort of 78 mice, there were 36 survivors. Of those, 20 had residual weakness from the acute infection. At one year post-infection 3 had flaccid paralysis without recovery (15%), 7 were stable (35%) and 10 (50%) developed late weakness.

Electromyographic (EMG) testing at 4 and 12 months revealed chronic denervation as is seen in human poliomyelitis. Qualitative pathologic studies revealed neuronal degenerative changes without inflammation. Virologic studies (RT-PCR) at about 2 years after inoculation detected viral genes in only a small number of mice, 2 of 15.

Mice infected with human poliovirus PV2/W -2 develop acute paralytic disease. About one-third of surviving paretic mice developed new late weakness at one-year post infection. EMG findings were consistent with old poliomyelitis as is seen in humans. Pathologic studies reveal that motor neuron degeneration continued after recovery from the acute disease and no inflammation was seen. Rarely was any evidence of viral infection found in these late poliomyelitis survivors. This appears to be an excellent model to determine the cause and treatment of PPS.

Studies are presently underway to determine if growth factors, specifically glial cell line-derived neurotrophic factor (GDNF) and insulin-like growth factor (IGF-1) will stop the late weakness. GDNF helps maintain the cell body while IGF -1 prevents terminal sprout dropout. Studies are underway to transport the genes of these growth factors to the motor nerve cells by insertion into a vector (adeno-associated virus) and injecting intramuscularly.

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RESEARCH I: WHAT IS BEING DONE? WHAT NEEDS TO BE DONE?

Update on Modafinil Study

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Post-Polio Syndrome (PPS) Fatigue: The New Challenge 50 Years After the Salk Vaccine:

Post-Polio Syndrome (PPS) is the term used to describe the reemergence of symptoms decades after recovery from acute poliomyelitis. Symptoms include increasing muscle weakness, pain, and atrophy, fatigue, breathing and swallowing difficulties, sleep disorders, and cold intolerance. After started, symptoms progress overtime and lead to gradual functional loss. Usually, but not always, PPS symptoms begin 15 or more years of stable function that follows recovery from paralytic polio.

Among the PPS symptoms, fatigue is the earliest and most disabling. Because of fatigue, many polio survivors are forced to leave work or lose the ability to live independently. The fatigue of PPS is not only debilitating, it affects the vast majority of polio survivor population. Estimates show that 79% to 89% of patients with PPS suffer from fatigue. A 1985 survey of 676 polio survivors showed that 91% of the sample experienced new or increased fatigue, with 41% claiming an interference of fatigue in performing work and 25% in self-care activities. A questionnaire completed by 276 Norwegian subjects with PPS showed that the prevalence of fatigue in PPS patients is significantly higher than in matched controls.

The cause of fatigue in PPS patients is only vaguely understood. It is thought that multiple domains of subjects' function are involved. In PPS patients, at least in part, fatigue is related to the gradual loss of individual nerve cells that make contact with other nerve cells (within the central nervous system), or with muscle fibers (within the peripheral nervous system). This results in subsequent loss of nerve transmission to these circuits. During the original polio infection, the poliovirus destroys nerve cells in the brain and spinal cord, particularly but not only, motor neurons. Among other things, this can result in loss of muscle function, including weakness or paralysis. However, to compensate for this loss, surviving neurons sprout out extra branches that are able to reestablish synapses (contacts), especially with orphaned muscle fibers that have lost their original nerve supply. Because of this process of reinnervation the individual is able to regain function. This is easily noticeable within the muscle system: body muscles are able to work again, sometimes as well as before. Ironically, these repaired circuits appear to wear down with aging. Some researchers have suggested that PPS develops because these extra sprouts cannot "hold" forever, but instead get weaker over time due to "over-use". Eventually, the sprouts degenerate, and function mediated by the neural contacts they have secured for years decline or disappears. This explains why recovered muscles gradually weaken and loose bulk when PPS settles in. An important lesson from the facts outlined above is that the fatigue faced by PPS patients is complex and involves multiple domains of function (emotional, intellectual, social, etc.), not simply the physical (muscular) dimension.

Unfortunately, except for supportive care, effective pharmacological therapies for the fatigue of PPS remain elusive. This problem remains as one of the most difficult new challenges faced by survivors of the last epidemics 50 years after the Salk vaccine.

Attempts at symptomatic management of other PPS symptoms have not met with much success either. In the past, several groups of researchers worked independently while investigating the origin of PPS and ways to reduce the burden of

the incapacitating fatigue. About half a dozen clinical trials directed to reduce fatigue, the most common and disabling problem faced by patient, were done but arrived at negative results.

In year 2001 the PPS Program was funded. The PPS Program is sponsored by the Uniformed Services University (USUHS) and administered under the auspices of the Henry M. Jackson Foundation (HJF). The mission of the PPS Program is to advance knowledge on the cause and treatment of post-polio syndrome. To achieve this goal several independent studies are taking place, some in the form of clinical trials intended to test the effect of medications to reduce the symptom burden in PPS patients.

Currently, the PPS Program is enrolling volunteers to participate in a clinical trial on PPS fatigue. This study will test if a medication called modafinil (Provigil) can help reduce the fatigue of patients with post-polio syndrome. This research is being done because, despite intense work, there still is no effective treatment for PPS fatigue, the most debilitating problem in persons with PPS. The nature of PPS fatigue is poorly understood but a central element is likely. This is supported by the damage caused by the poliovirus to neurons in supra spinal areas of the central nervous system, particularly the basal ganglia and reticular formation. Drugs reducing fatigue in neurological conditions usually act by facilitating central catechol-aminergic tone. The centrally-acting α -adrenergic agonist modafinil may help lower fatigue in PPS subjects. Modafinil has been used successfully to reduce fatigue in patients with other neurological disorders, including multiple sclerosis. Other studies in our program are directed to investigate different aspects of PPS. A second study (also actively enrolling) is looking at alterations in the brain and spinal cord of polio survivors that might help explain the development of PPS and the origin of PPS symptoms. This one is not a treatment trial. Instead, we are employing electrophysiology techniques and magnetic resonance to map possible residual abnormalities in the central nervous system induced by the polio virus during the original infection.

A third study is coming up soon. This one will examine if cognitive problems that are common in survivors with PPS, by measuring the brain ability to concentrate, sustain attention, register and memorize information, etc., with the use of traditional neuro-psychological tests. These studies aim to advance knowledge in several different areas of PPS and hopefully, help us design and test therapeutic interventions that can be safely used to reduce disability in polio survivors. For more information, please contact our research nurse coordinator, Ms. Kay Kelley, at 301-295-0231.

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