



## Post-Polio Sequelae: Preventing Ultimate Burnout

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It's 8:00 p.m. and only one light is burning at corporate headquarters. Mr. Gray, MBA, the 55-year-old CEO, is awakened by the phone. He lifts his head from the desk to answer and hears his wife asking when he's coming home. Feeling as if he weighs a ton, his muscles burning, Mr. Gray wheels himself to the car and, with barely enough strength to pull his chair in behind him, drives home. He greets his kids, rolls into the bedroom and throws himself on the bed. It's the third night this week he has gone to bed without dinner and slept in his clothes.

The alarm rings at 5 a.m. A clean suit and a three cups of coffee later, Mr. Gray is on the road again. Driving to work he hears the same faint "sizzling" sound he hears every morning, like electricity arcing between two frayed wires. But on this, as on every other morning, he ignores the "sizzling," ignores his increasing fatigue and muscle weakness and pain. He has a corporation to run. There's no time to "give in" to his body. But he does wonder: "What is that "sizzling" sound?"

Well, Mr. Gray, that *metaphoric* "sizzling" is the sound of your nerves burning out. Nearly 80% of North America's one million+ polio survivors hear the same sound as they too, experience Post-Polio Sequelae (PPS), the unexpected and often disabling symptoms - overwhelming fatigue, muscle weakness, muscle and joint pain, sleep disorders, heightened sensitivity to anesthesia, cold intolerance and difficulty swallowing and breathing - that occur decades after the acute polio infection when polio survivors' reduced number of remaining, poliovirus damaged neurons "brownout" or fail and die due to overuse. Unfortunately, most polio survivors are just like Mr. Gray: hard working, time-conscious, perfectionistic super achievers who, as a way of life, push themselves beyond their physical limits. Polio survivors ignore the "sizzling" sound, refuse to give up control, refuse to "give in" to new fatigue and weakness and pain even though their bodies are begging them to stop.

### POST-POLIO PAST

Why won't polio survivors listen to their bodies and slow down? When you understand what it was like to have had polio, you can appreciate why polio survivors feel that "giving in" to PPS is the same as giving up their lives. "During the epidemics," says PPS researcher Dr. Nancy Frick, a polio survivor herself, "polio was America's summer terror. The adorable March of Dimes poster children, in heavy metal braces and leaning precariously on their crutches, were everywhere saying, 'Give money to find the polio vaccine. Don't let your child become a cripple like *me!*'"

When the polio vaccine arrived in 1954, the poster kids were needed no more. Their braces, crutches and wheelchairs were evidence of a horror that Americans wanted to forget. "So polio was eagerly forgotten by everyone," Frick says, "including those disabled by it. To be accepted back into society, polio survivors had to look 'normal' again. And since buildings were totally inaccessible, even paraplegic polio survivors had to be able to walk if they wanted to go to school or get a job."

So polio survivors were ripped away from their families for months or years and admitted to rehabilitation hospitals where they underwent multiple surgeries and draconian physical therapy. Many were verbally abused, slapped or even beaten with rubber truncheons by therapists or family members to "motivate" them to walk. "Is it any wonder that polio survivors discarded their braces and crutches when they came home from the hospital?" asks Frick. "I have a theory that polio survivors would have done almost anything to look 'normal' and stop the abuse."

In the 1995 International Post-Polio Survey Nancy Frick and I put her theory to the test. She surveyed more than 1,100 polio survivors, asking about the experiences surrounding their polio, including hospitalization, surgeries and emotional and physical abuse by family members, peers and medical professionals. She also measured their Type A behavior, sensitivity to criticism and failure and asked whether they had been evaluated or treated for PPS.

"As in our two previous post-polio surveys," says Frick, "polio survivors were 21% more Type A and 15% more sensitive to criticism and failure as compared to nondisabled controls." A more upsetting finding was that polio survivors reported 34% more emotional abuse and 94% more physical abuse than did nondisabled control subjects. "All of that 'extra' abuse was related to polio survivors looking disabled," Frick explains. And those who were abused were at least 15% more Type A and sensitive to criticism than polio survivors who were not abused. "It is no mystery that polio survivors today refuse assistive devices that make them look more disabled," says Frick. "Using crutches or a wheelchair feels like painting a bullseye on your chest with the words, 'Abuse me, I'm disabled!'"

Frick's findings may also explain why polio survivors are so reticent about even being evaluated for PPS. "About 78% of polio survivors said they were not treated with concern by medical staff when they had polio," she says. "Those who were not treated well became very Type A and very sensitive to criticism and failure as adults." Our research suggests that Type A behavior developed as a protection against criticism and failure. We found that polio survivors who were the most Type A today are most likely to refuse treatment or even evaluation for their PPS.

Frick concludes, "Type A polio survivors refusal to treat their PPS and use assistive devices as a protection from the kind of abuse they experienced as children, as is polio survivors becoming Type A super-achievers. As our two previous surveys show, more polio survivors marry and go to college than do nondisabled Americans. Polio survivors work more hours of overtime and take fewer sick days than any other group. They became the leaders of their communities and the chief executives of the world's largest corporations. They become Mr. Gray. *That's* why their nerves have started to "sizzle."

## **POST-POLIO PRESENT**

Since we know that PPS are not caused by the return of the poliovirus that's been hiding in the body for decades, nor are the result of some new disease, the simplest explanation is that the reduced number of remaining, poliovirus-damaged neurons are mad as hell and aren't going to take it anymore.

**New Muscle Weakness and Pain.** When polio struck, large numbers of neurons in the brain stem were damaged by the poliovirus. For those who had paralysis, the poliovirus damaged 90% of the motor neurons in the spinal cord that run the muscles, and nearly half of those neurons died. The surviving but damaged neurons are less able to manufacture acetylcholine, the chemical that nerves release to make muscles contract. But, in spite of severe damage, motor neurons were still able to send out sprouts -- like extra telephone wires -- to turn on muscle fibers orphaned when their motor neurons were killed by the poliovirus. This sprouting allowed many people who were totally paralyzed during the initial polio attack to actually walk out of the hospital months later.

But since polio-damaged, over-sprouted motor neurons have been doing as much as 16 times the amount of work they had done before the poliovirus infection over 50-plus years, even polio survivors with minimal loss of function originally have been heading for a fall. Sprouted motor neurons can no longer make muscles contract as forcefully as they once did, or for as long as they once could, because their internal metabolic "factory" is failing after years of overworking. Muscles become weaker and hurt as they try to do more work than they are able to do." And because weakened muscles can no longer support the joints, polio survivors' shoulders and elbows ache and their knees

start to bend backward. As metabolically-damaged neurons fail, standing, walking, pushing a wheelchair -- sometimes even swallowing and breathing -- become more difficult or more painful.

This metabolic failure theory of PPS was demonstrated in 1997 by A.J. McComas of Canada's McMaster University. McComas used EMG to count the number of remaining motor neurons in polio survivors not being treated for PPS and then followed them for two years. McComas found that a limb weakened by polio had only 40% of the normal number of motor neurons. Even limbs that were thought *not* to have been affected by polio have only 60% of the motor neurons they should. Two years later, McComas found that 78% of polio survivors reported a decrease in muscle strength and had lost an average of 14% of their motor neurons, nearly twice the rate of loss expected in healthy subjects ten years older. Most alarming was the finding that the two survivors who reported the greatest decrease in strength had each lost 50% of their motor neurons over the two years. While these findings are frightening, they are also a guide to a rational treatment for PPS: *"Our findings make clear that polio survivors should not be treated using electrical stimulation that causes muscle contraction," warns McComas, "nor should they engage in fatiguing exercise or activities that further stress metabolically damaged neurons that are already overworking.*

**Brain Fatigue.** While new muscle weakness and pain are reported by more than 75% of polio survivors, the most common PPS symptom is fatigue. During the past 25 years, my team at The Post-Polio Institute has found that brain fatigue -- when polio survivors can't concentrate and have trouble staying awake as the day goes on -- is associated with an inability to focus attention and process information quickly plus a marked reduction in the brain-activating hormone ACTH. Magnetic resonance imaging revealed damage to the brain-stem neurons responsible for activating the brain, damage also found during autopsies of polio survivors done in the 1940s.

What's more, our study published in 1998 describes the measurement of polio survivors' brain waves and the hormone prolactin. Prolactin increases in the blood when there is too little of the most important brain-activating neurochemical, dopamine. Our finding that polio survivors with the most fatigue had the highest prolactin levels and the slowest brain waves suggests that they do not make enough dopamine to fully activate their brains. A dopamine shortage would explain symptoms of post-polio brain fatigue. This conclusion is supported by our finding that bromocriptine -- a drug that substitutes for dopamine in the brain -- reduces the symptoms of post-polio brain fatigue in survivors who do not respond to the conservative treatments of choice for PPS: reducing physical and emotional stress, using assistive devices, conserving energy, resting and pacing activities.

## **POST-POLIO FUTURE**

With the coming of PPS, the future might look bleak for polio survivors. Both the March of Dimes Birth Defects Foundation and the U.S. Congress have refused to set aside funds for PPS research. And two recent drug studies -- one testing insulin-like growth factor-1 (IGF-1) and another studying pyridostigmine (Mestinon) -- have failed to show any "significant impact on quality of life, fatigue or muscle strength." Even our study found that bromocriptine reduced symptoms only in polio survivors who were the most severely disabled by fatigue.

But the future is not bleak. Research during the past decade has shown that the conservative treatments of choice for fatigue are very effective in managing PPS. Polio survivors need to follow the "The Golden Rule for PPS:" *If something you do causes pain, fatigue or weakness, don't do it (or do less of it)!"*

"The Golden Rule" does *not* mean that polio survivors should sit home and become couch potatoes. "The Golden Rule" *does* mean that polio survivors should stop exhausting themselves, especially with exercise. Many survivors are prescribed -- or actually ask for -- the kind of physical therapy they had right after polio: exercising to the point of exhaustion. But several studies show that pumping iron will not increase the strength of muscles that are becoming weaker, and actually cause an irreversible

loss of strength and possibly the *death* of motor neurons. No laps in the pool, free weights, exercise bikes, or treadmills for polio survivors. "Feeling the burn" means nerves are burning out. Stretching, however, helps decrease pain and increase range of motion. And gentle, non-fatiguing exercise can be useful for some polio survivors to help maintain muscle strength and tone after getting a new brace.

Several studies show that changing survivors' physically and emotionally stressful Type A lifestyle is the best way to reduce weakness, fatigue and pain. One study shows that polio survivors who pace activity -- that is work and then rest for an equal amount of time -- can do 240% *more* work than if they push straight through the task. Another study shows that survivors use *three times less* energy walking if they use a short-leg brace on a weakened leg. And the follow-up study of The Post-Polio Institute's program of behavior modification, physical and occupational therapy shows that polio survivors who comply with treatment -- pace activities, conserve energy, take two 15-minute rest breaks a day and use assistive devices (a brace, cane, crutches or wheelchair) -- have up to 22% *less* pain, weakness and fatigue 16 months after therapy ends.

However, polio survivors who refuse or quit therapy have 21% *more* fatigue and 76% *more* weakness 16 months after leaving The Post-Polio Institute. And there's the rub. It's only the refusal to listen to that "sizzling" sound that causes PPS to continue or progress. "The treatments for PPS may sound simple but they are not easy," says Frick. "Polio survivors have to deal not only with their bodies giving out on them again, but also with long-buried emotional pain." And it is those painful experiences of the past and memories of abuse, the fear of giving up "protective" Type A behavior and terror of looking disabled, that stop polio survivors from treating their PPS. So the most difficult aspect of treating PPS is not deciding whether a short or long-leg brace will be most helpful. The challenge is helping polio survivors face the pain of their abusive pasts and accept *appearing* more disabled -- by slowing down, asking for help and using new adaptive equipment -- so they won't *become* more disabled. That's why behavior modification and psychotherapy are required to help survivors accept their "second bout" with polio, change their super-achieving lifestyles and deal with the fear of looking disabled.

As Mr. Gray would tell you, this whole PPS business stinks. But isn't it far better to manage PPS now than to be forced to give up everything you do because you are too weak and fatigued to function? I know, you'll slow down and take care of yourself "when you're ready." You'll use a wheelchair "when there's no other choice." Well, you don't drive your car until it's out of gas. Why drive your body until it's out of neurons? Isn't it time to listen to the "sizzling" sound? Isn't it time to take care of yourself? To paraphrase one famous polio survivor who refused to take care of *himself*, "You have nothing to fear but fear itself."

### Red Flags for Polio Survivors [Mis]Diagnosis

**Diagnosis:** Many polio survivors have been incorrectly told they cannot have PPS because they didn't have paralytic polio. The National Institute of Health has just studied pairs of identical twins, one who had paralytic polio and one who had non-paralytic polio. Seventy-one percent of those who had paralytic polio have PPS. The surprise is that 42% of those who had "non-paralytic" polio also have PPS. The lesson: Don't let a doctor tell you that new symptoms all in your head just because you didn't have paralytic polio.

**Anesthesia:** Polio survivors are anesthetized very easily because the part of the brain that keeps you awake was damaged by the poliovirus. They also stay anesthetized longer and can have significant complications from anesthesia. Even nerve blocks with local anesthetics can cause problems. All polio survivors should have lung function tests before having a general anesthetic. "Polio survivors

with a lung capacity below 70% may need a respirator or respiratory therapy after surgery," says Dr. Augusta Alba. Your complete polio history and any new problems with breathing, coughing or swallowing should be brought to the attention of your surgeon or dentist -- and especially your anesthesiologist -- long before you go under the knife. Also, polio survivors should never have same-day surgery or procedures like colonoscopy that require general anesthetic.

**Pain:** Two studies have shown that polio survivors are twice as sensitive to pain as everyone else and need more pain medication for a longer time after surgery or an injury. However, survivors have had to develop an extremely high pain tolerance to deal with their increased pain sensitivity and are not likely to become addicted to pain medications.

**Sleep:** The majority of polio survivors have trouble sleeping due to pain, anxiety or sleep disorders, such as sleep-disordered breathing or generalized random myoclonus (GRM). GRM is when muscles in different parts of the body twitch and jump throughout the night and "wake up" the brain even if you don't wake up. Amazingly, most polio survivors aren't aware that they stop breathing or twitch. If you awaken with a headache, with your heart pounding, short of breath or don't feel rested during the day, call a local sleep disorders center for a sleep study.

**Trauma:** Our study found that eighteen percent of Post-Polio Institute patients reported that their PPS begin after trauma, including medical illnesses, surgeries, fractures, falls and especially auto accidents. Regardless of the type of trauma or location of the injury, the most common new symptom was muscle weakness (55%), followed by pain (34%) and fatigue (11%). Twenty-six percent of patients had new symptom in the injured area plus one other nearby location. For example, 40% of those who injured one leg developed weakness or pain in the other leg. (This is a common problem for polio survivors, who compensate for injury to one part of the body by overusing another part whose nerves were also damaged by the poliovirus.) The good news is that 86% of patients regardless of the cause of trauma or severity of their injuries had significant reductions in pain, fatigue and muscle weakness after complying with therapies known to be effective in treating PPS: reducing physical and emotional stress, using appropriate assistive devices, energy conservation, adequate rest and the pacing of activities.