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POLIO ABOVE THE NECK

WITH DR. SUSAN PERLMAN

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Dr. Susan Perlman, Director of the UCLA Post-Polio Clinic, spoke at the May 2, 1999 meeting of the Orange County Post-Polio Support Group. In addition to her responsibilities in the Post-Polio Clinic, Dr. Perlman also serves at the University of California-Los Angeles (UCLA) as Associate Clinical Professor of Neurology, and Director of the Neurogenetics Clinic.

Previously much attention has been given to post-polio problems involving the limbs -- a weak leg, fatiguing arms, etc. These manifestations of Post-Polio Syndrome (PPS) occur in people who had spinal poliomyelitis – the acute infection that affected anterior horn cells in the spinal cord segmentally.

People who had nonparalytic polio, by definition had polioencephalitis. They had involvement of those brain areas above the spinal cord, and could well have had poliovirus changes in the brainstem (bulbar polio). So breathing and swallowing problems may be present even in people who ostensibly had nonparalytic polio and also in others who may have no complaints about their legs or arms.

This report focuses on the problems of bulbar polio and other problems that are now manifested in people who have PPS. The cause of these problems is polio damage that occurred in the upper cervical spine and upward. Bulbar is defined as polio involvement of the motor nerves in the brainstem.

Early Studies:

Polio autopsy reports following the epidemics of the 1940s and 1950s showed signs of acute polio infection throughout the body; it was not just restricted to the spinal cord. There were changes found in upper motor neuron pathways and there were changes found in central brain structures that control alertness, central fatigue, and autonomic functions such as temperature regulation etc. There were also many changes seen in the brainstem itself.

Probably greater than 90% of a polio survivors motor neurons were somehow affected during the acute stage and had some damage, even if not paralytic. Autopsy studies have shown that during the acute attack as few as 3 or 4% of the motor neurons remained intact.

There are not many polio survivors seen now who had severe residual effects from the acute bulbar polio infection. This is probably because when there is a 50% loss of the motor neurons that control breathing or swallowing, a person is going to be in serious trouble. Many did not survive the acute infection. Those who survived bulbar polio truly are survivors, said Dr. Perlman.

Respiratory Problems:

There are some people who had only mild breathing weakness with the original polio, (chest wall weakness or a little diaphragmatic weakness) but their breathing centers in the brainstem were actually intact. People who had true bulbar polio had involvement that didn't trigger breathing – they needed to be assisted until those centers came back. So when looking at post-polio breathing problems, people who had primary muscular manifestations should be included along with those who had bulbar polio.

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In other parts of the body such as arms and legs, surviving motor neurons remodeled and adopted the injured or orphaned ones in order to take over function. However this was not as common in the brain and brainstem. Since the brainstem has less plasticity and flexibility, it was harder for remodeling to occur to the nerves of the pharyngeal muscles and to those of the upper part of the esophagus. For years, many survivors have been using compensatory strategies, such as swallowing on the other side of their throat. Those who had residual problems such as vocal cord paralysis may also have been able to compensate for this. But when the muscles on the “good” side of their throat begin to weaken, they become aware of a “new” problem in that area.

Muscles that control breathing and swallowing are the same groups of muscles that are involved with speech, to some extent. These groups of muscles are located in the throat. So people with PPS who are having problems with swallowing, breathing, or shortness of breath during activities of daily living, might also complain of speech problems. Although facial, jaw, and throat muscles are controlled in the brain stem, they have rarely been thought of in connection with polio.

Are people who had bulbar polio going to be having increasing bulbar dysfunction? The following criteria re used to determine increasing bulbar problems:

1. Severity of residual disability.
2. Residual bulbar or respiratory signs.
3. Later age at onset of acute polio (past age 10).
4. (Possibly) recent falls or injuries; or surgical procedures in the pharyngeal area; or weight gain, because it puts more pressure on the diaphragm and pharyngeal area.

Fortunately our bodies have several safety factors to assure that breathing muscles meet our oxygen and carbon dioxide demands. The primary muscle for breathing is the diaphragm and the secondary breathing muscles are the external intercostals (the muscles between the ribs). So if a person has a fatigable diaphragm that tires out by the end of the day, the intercostals will kick in and the person will begin breathing from the chest. There are also accessory respiratory muscles that help lift the chest from the shoulders.

With post-polio breathing problems, part of the problem is going to be muscular. There may be a diaphragm or secondary assistive muscles not kicking in as well as they should. There can also be central (brain) changes contributing to this, such as decreased respiratory drive, if those centers were affected originally. There may be changes in the chemoreceptors: perhaps the sensors are not sensing carbon dioxide (CO₂) buildup as sensitively as before.

Scoliosis can also cause restriction of breathing. A person who has a scoliotic spine cannot expand his chest as well so he underbreathes because of it. If scoliosis is getting worse as a post-polio symptom, it's going to make that aspect worse and will interfere with breathing.

When doctors are looking at respiratory problems of PPS patients, they should look at not only peripheral problems and muscular problems, but also central disregulation of breathing as well. All these factors are addressed at the UCLA sleep lab, directed by Dr. Frisca Yan-Go.

Recent Findings:-

Dr. Perlman cited a report (“Epidemiology of the Post-Polio Syndrome” by J Ramlow, et al. American Journal of Epidemiology, October 1, 1992) that involved 77 subjects with non-paralytic polio and 474 subjects with paralytic polio. A change in breathing was observed in 10% of the nonparalytic subjects and in 12% of the paralytic ones. Swallowing problems were noted in 6% of the nonparalytic subjects in 7% of the paralytic ones.

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In the larger ongoing studies at polio centers such as Mayo Clinic, Rancho Los Amigos, and centers in Canada, possibly as many as 40% of the people with PPS are having new respiratory complaints. (About 80% of PPS patients complain about fatigue.)

Another study of 74 polio survivors who were having shortness of breath found two good measuring devices for patients with increasing respiratory problems:

1. Forced expiration (a pulmonary function test) can be a very helpful monitor. This test shows how hard it is for a person with PPS to breathe out and it requires the use of some of the intercostals muscles and the abdominal muscles. Dr. Perlman recommends this test every year or so for her patients who have significant breathing complaints.

2. The other helpful thing is to monitor CO₂ levels in the blood. The question is not how much oxygen is a person inhaling, but how much CO₂ is a person retaining? Is a patient not breathing fast enough? Is the person fatiguing so that he cannot ventilate? It is the amount of ventilation a person gets that clears the CO₂. So if you are ventilating less efficiently, your CO₂ levels are going to go up slowly.

For patients at risk, these researchers felt that anything that was in danger of happening could be detected by measuring maximum expiratory pressure and carbon dioxide levels on a regular basis.

Swedish research on cardio-respiratory parameters in PPS patients found a significant incidence of deconditioning. This goes back to the old thought that everybody should be doing some exercise. Dr. Perlman says doctors are no longer saying people with PPS should do no exercise: some exercise, conditioning or aerobic exercise, is important. Survivors can improve heart function, circulation, and breathing to some extent by doing something that increases the heart rate. These researchers suggested increasing the heart rate to 70% of maximum by using a pool or other equipment. They felt these were tolerable levels for their PPS patients.

Researchers in Toronto looked at 3 areas of muscles (respiratory, diaphragm, chest wall), bulbar symptoms, the control rate, and scoliosis. They found that the control panel in the brainstem was the least important of the group. These people were having problems because of the diaphragm, chest wall fatigue, or due to progressing scoliosis.

A few of Dr. Perlman's patients have experienced increased breathing problems at higher altitudes. Adjustments can be made so those patients can travel at higher altitudes and not feel short of breath all the time.

Swallowing:

The motor neurons that control swallowing are located in the brainstem. In order to have an effective swallow, groups of muscles are used to insure that the food is chewed, forms into a bolus, and goes down properly. The swallowing center coordinates other activities related to swallowing: chewing, licking, gagging, coughing, sneezing, vomiting, belching, and breathing to some extent (when a person is swallowing, he doesn't breathe). It is hard to eat or swallow at the same time as breathing, because the two groups of muscles and nerves are competing against each other.

There are at least a dozen places in the body where a post-polio patient who had some pharyngeal problems or brainstem related swallowing problems could begin to have trouble now with swallowing – either in the steps or in the sequence of swallowing. For example, people who have weakness in the jaw muscle as a complication of PPS will find it hard to chew when fatigued. Or people who have a weak soft palette will find that food is slipping into their throat before it is fully chewed and food or drink – drink especially – may be coming out of their nose.

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Swallowing problems have been identified in recent studies. However it is rare to see a PPS patient who is having constant choking. Most people can think about it and use their conscious mind to control what is going on, thereby preventing swallowing problems from occurring. Any part of the gastro-intestinal tract – from the mouth all the way to the bottom – could be slowed, weakened, or not working properly due to PPS. But don't assume that every symptom a polio survivor gets is due to PPS.

Although common pathways are used for breathing and swallowing, not everyone who has swallowing problems has breathing problems and not everyone who has breathing problems has swallowing problems. Bulbar muscles can slowly dysfunction and there can be silent swallowing problems. In a 1991 swallowing study by Dalakas he concluded "...in bulbar neurons there is a slowly progressive deterioration similar to that in the muscles of the limbs."

Blood Pressure and/or Variable Heart Rate:

Can high or low blood pressure be a result of polio and PPS? Can variable pulse rate be made worse, not because of heart disease, but because of post-polio symptoms affecting the area?

The average internist may be hard pressed to believe it possible for polio to be related to current blood pressure or pulse rate problems. However, vaso-motor centers that control blood pressure and pulse rate are located in the medulla (in the lower brainstem) and also in the autonomic area of the brain. Since polio damage has been seen in the brain and in the brainstem, this is an area that needs further study.

As we understand what was involved in acute polio and the amount of brain that was involved, recovered motor neurons that might have looked quite complete on the surface could actually be functioning on very shaky ground. "The majority of motor neurons, in whatever region the poliovirus got to, were probably affected in some way or another", said Dr. Perlman.

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Audio Tape:

A recording of this complete talk is available for \$5.00 + \$2.00 postage and handling. Contact:

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