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FACING SURGERY WHEN BREATHING IS A PROBLEM

Scoliosis, Ventilation, & Surgery

Augusta Alba, MD, DNP-N, DPM&R, Goldwater Memorial Hospital, New York, New York

ow does scoliosis affect breathing? Consider a typical, moderate curve of the spine of an individual. We usually look at the outside of the body to determine what is happening to the inside of the body, to the lungs, and the heart. The twisting of the ribs backwards causes the lung to be similarly twisted into that shape and partially compressed. Distortion of the lung happens not only on the side of the convexity, but on the other side as well, the side of concavity.

When considering breathing, we are particularly interested in the cervicothoracic curve (apex at C7-T1), the thoracic curve (apex between T2-T11), and the thoracolumbar curve (apex at T12-L1). These are the ones that can affect cardiopulmonary function. Curves are classified by the number of degrees of lateral curvature. Curves in group III (31-50°) and group IV (51-70°) and beyond are the ones that cause more problems insofar as anesthesia and surgery are concerned.

There are several specific neuromuscular diseases which can cause curvatures including muscular dystrophies, spinal muscular atrophy, cerebral palsy, spinal cord injury, spina bifida, and arthrogryposis. Post-polio scoliosis is probably most similar to the scoliosis associated with spinal muscular atrophy. For the person with neuromuscular scoliosis, it is important to know specific management techniques when the curve is the result of muscle weakness.

The surgical treatment of scoliosis has already been discussed (*Polio Network News*, Vol. 11, Nos. 2 & 3). Non-surgical treatments include observation, bracing, custom seating, and electrical stimulation which for the most part has fallen out of favor, because it has not proved to prevent further progression of the curve. Postural exercises, exercises to maintain spinal flexibil-

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New Breathing Problems in Aging Polio Survivors

Respiratory Muscle Weakness

Ann Romaker, MD, Kansas City, Missouri

In my pulmonary practice over the past 15 years, I have seen individuals who have a history of polio and who have respiratory problems but are unaware of them. Only one required ventilatory support during acute polio, and one other had known bulbar polio. The others had isolated or extensive limb involvement, even quadriplegia. Most had involvement in just a single limb. When studied, all of them had measurable respiratory muscle weakness. Anyone with a prior history of a neuromuscular disease, such as polio, needs to be evaluated in depth before and around stressful, physiologic events such as surgery.

Function of the respiratory muscles ... The diaphragm is the major muscle of breathing and its descent increases the length of the thorax and increases lung capacity. The external intercostal muscles pull the ribs up and out. The scalene muscles raise the rib cage and the sternum (or breastbone). Another muscle, the sternocleidomastoid, elevates and expands the rib cage. Negative pressure is created within the chest, and the increase in the dimension of the chest increases the volume of the lungs. Air then enters the lungs through the nose, and is inspired.

Expiration is a much less active process. The abdominal muscles pull the ribs down and push the diaphragm up. The internal intercostal muscles pull the ribs down and in and squeeze the air out. If either the inspiratory or expiratory muscle groups, or both, are not functioning properly, some type of respiratory assistance may be needed.

Representative case ... An individual with involvement (that she was aware of) only in her legs and who wears leg braces, works full time as a nurse. After routine gynecologic surgery, she developed pneumonia

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ity, and other exercise routines are helpful and must be individualized. Physical therapists specializing in the treatment of scoliosis can provide this training.

For polio survivors with scoliosis, the main problem is a reduction of the biggest breath one can take. When the ability to breathe in deeply is impaired, the tidal volume (volume inspired with each breath) becomes smaller. Respiratory rate then becomes more rapid in order to maintain adequate ventilation. When a person is breathing more rapidly, the amount of air that actually reaches into the distal parts of the lungs where gas exchange occurs, the alveoli, is less. The work of breathing is increased because the flexibility of the chest wall is decreased due to the scoliosis. If the work is increased, the oxygen cost of breathing is also increased.

What is the result of inadequate ventilation due to a scoliotic chest wall? One develops an increased resistance in the circulatory system of the lung which leads to hypertension of the pulmonary artery system — the system which sends blood to the lungs from the heart. If pressure in the pulmonary artery increases, the right side of the heart becomes enlarged and hypertrophied. The heart muscle becomes thicker in an effort to pump blood against a higher pressure. If the scoliosis is not corrected, the heart eventually dilates. This condition is known as cor pulmonale.

With a scoliotic curve there can be a displacement of the heart within the chest, with it sometimes appearing to be on the right side of the chest. The aorta, which is the major blood vessel of the body, may also be twisted to follow the scoliotic curve. It is rarely kinked. The ultimate outcome with inadequate ventilation is congestive heart failure.

What recommendations can be made when polio survivors with breathing problems are facing surgery? There is no special pre-op or post-op care if the vital capacity is 70 to 100% of normal, with a good ability to cough, and if expiratory peak flow, hemoglobin, and EKG are normal.

If curvature and neuromuscular weakness have decreased the vital capacity to 50 to 70% of normal, the expiratory flow is slightly reduced, and hemoglobin is slightly elevated but EKG is normal, pre-op management is regular. However, post-op management should include intermittent positive pressure breathing treatments on a regular basis with assisted coughing.

When vital capacity is further decreased to 30 to 50%, there are more serious problems affecting the heart and lungs. Expiratory flow is markedly reduced, and hemoglobin is elevated above normal. With the secondary complication of increased hemoglobin, the blood is more viscous which increases the chance of thrombo-

sis. The EKG reflects the increased size and hypertrophy of the right side of the heart. Pre-op management must include assisted ventilation on a regular basis and, if improvement occurs, then surgery can be considered.

If assisted ventilation helps pre-op, continue it post-op on an indefinite basis so that hemoglobin and carbon dioxide will not be elevated and oxygen will not be depressed.

In post-op management, artificial ventilation should continue until the individual has regained at least 75% of pre-op vital capacity (VC). Aerosol mists with assisted cough; frequent turning to prevent atelectasis (small mucus plugs in the lung); and a limit on post-op sedation, which reduces the ability to breathe deeply and to cough, are recommended.

Persons with scoliosis with 10 to 30% of predicted VC may have a greater degree of cor pulmonale, a further increase of hemoglobin and will need even more assisted ventilation. If the persons is helped by assisted ventilation, then proceed with surgery. In some cases with vital capacity in the 10 to 30% range and the 30 to 50% range, consider a pre-op tracheostomy.

The post-op treatment of peoplet with 10 to 30% of predicted VC is the same as that for persons with 30 to 50%.

Considerations Before Surgery

Oscar A. Schwartz, MD, FCCP, St. Louis, Missouri

Certain polio survivors may be at increased risk for surgical procedures, and the benefits must be weighed with the risks for determining candidacy. Prior to surgery, a good evaluation of the respiratory system is necessary. Polio survivors can develop breathing problems due to weakness of muscles originally (and obviously) involved with polio as well as those partially compromised but continuing to function. The effects of scoliosis also need to be considered, as Dr. Alba has discussed.

The evaluation should focus on the individual's reason for going into surgery as well as the complications that may result from anesthesia. Anesthetic complications can include those which are secondary to the general anesthetic, as well as those which may be complications of use of the anesthetic. The program that is developed should have the goals of supporting the respiratory system if it is impaired, or decreasing the amount of risk associated with the surgical procedure.

Patient-focused care involves a team. Team members are determined by the type of surgery whether it involves surgery of the spine or abdominal surgery such as gallbladder, colon surgery, or hysterctomy. The team is established so that the surgeon, anesthesiolo-

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gist, as well as the professional following the polio survivor, communicate with each other.

With healthy individuals, surgical procedures may have limited risk. In polio survivors, even minor surgical procedures that involve general anesthetics or nerve blocks that may paralyze the only functioning respiratory muscles may have disastrous effects.

Individuals who are diaphragm breathers may have response to inhaled anesthetic totally different from those who use the diaphragm as well as the intercostal muscles to breathe. Swallowing difficulties are of concern because of the potential risk of aspiration associated with the general anesthetic. Anesthesia can further compromise muscles in the head and neck area that may be involved with swallowing or breathing. Polio survivors should understand what the anesthesiologist will do and feel at ease with the procedure used.

After surgery, respiratory support may involve facilitation of cough. Support with a ventilator may be necessary, especially if the individual had a preexisting need for one. The post-operative period may be longer for polio survivors.

Preparing for Surgery

Kathleen Navarre, PhD, Essexville, Michigan

When I was six years I had bulbar polio as well as paralysis in my upper and lower body. In 1991 through the network, I connected with Oscar A. Schwartz, MD, in St. Louis. Since that time I have used LIFECARE's PLV-100 ventilator with a Respironics face mask for night-time ventilation. I am back to driving and teaching full time. I feel quite alive, and delighted to be so.

My experience with surgery, in 1993, was not a polio issue; it was a hysterectomy. I came to St. Louis to have the support of Dr. Schwartz and the team at St. Mary's Health Center which paid off for I was back in the classroom five weeks after the surgery.

My greatest fear as a polio survivor with breathing problems was the thought of abandoning myself to an anesthetic and letting something take over my breathing for me. I am a sensitizer (a person with internal locus of control), so to face this traumatic experience, I gathered all the details, even the gory ones. I suspect that most of us attending this conference want all of the information we can get.

Others come under the label of repressors (individuals who have an external locus of control). They say in essence to a physician, "Do whatever you need to do and wake me up when it is over." That is fine, too.

Having an internal or external locus of control or being a repressor or sensitizer is neither good nor bad. Recognize which you are and make it very clear to your physician.

When my sister and I walked into St. Mary's Health Center for my surgery, there was an iron lung in the hallway. Being a sensitizer, I said, "Oh, my goodness Mare, that's for me." Being a repressor, she said, "Oh no, that's always there." I responded, "There's about four inches of dust on it. Obviously it's been in a closet since 1954." I was iron lung phobic but I wanted to touch it, to feel it. After I did touch it and put my hands in it, and sort of kicked a tire or two, I felt better about it. Because it was Halloween, the nurses discussed putting a pumpkin at the head and that humorous idea made me feel more comfortable. (The iron lung was there for me in case I required it after surgery, but it was not needed.)

They tried to do a spinal for my surgery which did not work because of my scoliosis. It was impossible to provide the necessary regional anesthesia. I did receive a general anesthesia with appropriate intubation. My recovery was normal in terms of time.

Kathleen offers these suggestions for adjusting to using a face mask:

- ▲ Start with several 15-20 minute intervals on the mask during the day.
- Relax, watch TV, read, try to distract yourself to stay relaxed.
- ▲ Get into the rhythm of the vent's breathing by slowly letting it take over.
- ▲ During the first few nights, wake up, sit up, clear your head, and then start over again.
- ▲ At first the vent feels like a smothering enemy, but it will become a life-affirming friend. Give it a chance.

Arranging for Anesthesia

Judi Cox, Springfield, Illinois

For polio survivors, particularly ventilator users, anesthesia is not a pleasant undertaking. On two occasions I have needed to prepare for surgery with general anesthesia. With a knowledgeable pulmonologist as part of the physician, surgeon, and anesthesiology team at Memorial Medical Center in Springfield, Illinois, things went well.

I am a 56-year-old female who had all three types of polio at age 14 (1952). The initial attack resulted in quadriplegia and being in the iron lung for the first nine months. In the two years that followed, I used a cuirass, then a rocking bed, slowly progressing to inde-

pendent breathing. After months at Warm Springs, Georgia, I learned to walk independently.

In 1992 while going to work at a large regional hospital, I collapsed from respiratory failure. There had been months of extreme fatigue requiring return to a wheelchair and retirement from work. Since then I have used night-time ventilation with Aequitron's LP-6. With pacing, I have enjoyed retirement.

Because of my history, anesthesia was considered a risk. I researched and prepared as much data as possible about my status and discussed it thoroughly with the surgeon and anesthesia team members. We discussed oxygen sensitivity, cold sensitivity, uncomfortable positioning, recent symptoms of concern, and a list of questions, e.g., how long will the procedure last, etc.

The team felt I might go to ICU still intubated and could possibly remain on the ventilator for 24 hours post-op. I shared my concerns and my hope to get my mask and vent back as soon as possible. Because of preplanning, they removed the tube when I was fully awake in the recovery room and in eight hours post-op I was back with my own equipment. Using the LP-6 quite a bit over the next two days prevented any adverse effects of the anesthesia. Also, using minimal pain medication eliminated further problems. Nebulizer treatments and aid in coughing over the three to four days post-operatively helped with the mucus.

As part of the team, I prepared folders (typed and easy to read) for each person on the surgical team that included the following:

- ▲ A complete current medical history and brief detail regarding systems and possible complications, e.g., cardiovascular system: see attached echocardiogram 3/7/93, including extra data such as copies of relevant records of arterial blood gases, sleep tests, etc., for baseline comparison.
- ▲ A page describing allergies and how they are manifested, e.g., morphine causes a rash, and documenting other precautions such as no muscle relaxants and minimal pain medication.
- ▲ A page regarding the home mechanical ventilator and mask listing types, settings, sizes, facts, etc. (It may be necessary for the hospital to check with the legal department clearing the use of your own equipment.)
- ▲ A list of specific questions for each team member including ways I could assist myself and information about others involved in my care.
- ▲ Copies of pertinent literature, keeping it brief and highlighting data. (*Preparing for Surgical Interventions*, in "Pulmonary Dysfunction and Sleep Disordered Breathing as Post-Polio Sequelae: Evaluation and Management," written by John R. Bach, MD, and Augusta S. Alba, MD, published in *Orthopedics*, Dec. 1991, Vol. 14, No. 12 is excellent.)

Physicians will appreciate your efforts and provide the best care possible because of your thorough knowl-

edge. You will have peace of mind as a responsible contributor to your care as well as to the team. Trust them and, when possible, find humor together in the uniqueness of your situation.

Dealing with Breast Cancer Surgery

Ellen Fay Peak, Birmingham, Alabama

When I was diagnosed with cancer in my right breast on March 2, 1995, my first thought was not "Am I going to die?" but "Am I going to be able to type and feed myself?" On April 3rd the surgeon, whom I ultimately chose, first examined me and said that it would have to come out. I responded, "No, it does not have to come out." He listened closely as I explained the history of my arm and shoulder limitations and watched carefully as I, naked from the waist up, demonstrated my very narrow range of arm and hand use, a range so narrow that I must have full-time physical assistance. There I was in a doctor's office trying to prove that my body had enough to do in dealing with the late effects of polio. Cancer did not scare me. It was the treatment of cancer that tore me up.

What was I doing between March 2nd and April 3rd? On the same day that I learned that I had breast cancer from a routine mammogram, I had a sonar exam of the mass in my right breast, consultations with two surgeons, and a needle-core biopsy. Although the surgeons concurred with the idea of immediate surgery followed by radiation, I decided to do nothing until I had more information. I obtained some of the most useful from a friend who had undergone a modified radical mastectomy. Upon hearing my diagnosis, she came immediately to show me the results of her surgery, pointing out where muscles had been removed from her chestwall and down her underarm. She examined my breast and shoulder, drawing her fingers down my skin where the path of a surgery like hers would be. From that discussion, we determined that a simple mastectomy was as much as I could have and still use my arm. Anytime I needed her to, she would come and strip down and let me ponder my decision. She served as both my model and my patient advocate, putting forth ideas and intervening as needed.

On March 13, to try to determine if the cancer had spread beyond my breast, I had a bone scan, additional x-rays of my hips, and a chest x-ray, all of which appeared to be normal. Having pretty much decided that I would have no surgery that required general anesthesia, I then investigated radiation therapy. Because of shoulder tightness, my arm could not be positioned for radiation.

I sought advice by phone and letter from doctors who knew me well. I discussed each development with my

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long-time internist who encouraged me to explore every avenue of information before making any decision. I had several informative conversations with the Medical Director of Post-Polio Services at Roosevelt Warm Springs (Georgia) Institute for Rehabilitation. His having my early polio records and those from his own evaluations of me through the years proved invaluable as we talked about the possible effects of general anesthesia and the types of breast surgery.

I pulled out Anaesthesia: Considerations for Polio Survivors, an article I had saved from the Fall 1991 issue of *Polio Network News*, and copies of both Dr. Susan Love's *Breast Book* and *Post-Polio Syndrome* by Halstead and Grimby. Because I was in the middle of planning the program for a state-wide forum sponsored by the Birmingham Post-Polio Support Group, I took the opportunity to discuss my situation with three of the major speakers. Each responded to my questions, providing me with an unexpected source of polio expertise.

Finally, I was in my surgeon's office arguing the fine points of tissue removal so as to preserve the little use I had of my right shoulder and arm: which lymph nodes I was willing for him to remove and what type of mastectomy I was willing for him to perform. I left there with a tentative surgery date, April 18th, to remove both breasts (the left breast looked threatening) and appointments with three more specialists to consult:

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On April 12th, I wrote the following letter to my surgeon, sending a copy to everyone else involved:

When I recently spoke with the Director of International Polio Network about my impending surgery and my agitation about my functioning afterwards, she pointed out that she knew of no situation like mine. Thus, I feel compelled to continue to provide my physicians with as much information as I can about my polio state. As an upside-down polio quadriplegic since 1936, with post-polio syndrome since the late '70s, I am functioning at the outer edge of possibility. Once my assistant, Kay, dresses me in the morning, I wash my face, brush my teeth, and feed myself, all with my right arm helping my left. For exercise throughout the day, Kay helps me to my feet and I walk around inside my house with either her help or a walker. The walker is merely a balancing tool. I cannot put any weight on it. I spend most of my day at my computer, my right arm supported in a sling attached to my office chair, typing with the eraser end of a pencil between my fingers (my left hand controls the mouse). Otherwise, I read and converse on the phone, activities that require the use of both my right and left arms.

I further say that, aside from its pain, cancer holds no more threat to me than polio. I am not in awe of it. I view it simply as one more possible obstacle to my functioning. Although in the past I have overcome many such obstacles, I now have so little margin of function left that I am doing everything within my power to protect it. First, I have chosen my surgeon carefully, indeed, with more care than I chose my internist, in those carefree days of the mid-'70s. Although that choice was kind of dumb luck, it has turned out to be one of my very best. I have discussed my situation with the individual I hope to have as my anesthesiologist. I have been evaluated by the physical therapist who will work with me after surgery. I have had the benefit of my internist's expertise, interest, and counsel at every step along the way.

As to the actual surgery: I understand that no paralyzing or other such compromising drugs will be used during the anesthesiological process. I also understand that you will remove both of my breasts and the first level only of the lymph nodes under my right arm. I would be very pleased if you decided to remove no lymph nodes!

Beyond the surgery: I plan to have no additional therapy beyond the drug Tamoxifen, or its like; plan to partake of inhalation therapy; and interact with the physical therapist to regain the function I now have, not expecting to be rehabilitated out of my wheelchair.

Finally, but perhaps most important, here are some facts to know about me that are a result of my particular bout with polio. Others have their own strangenesses.

Because of my lack of muscle mass, I am unable to generate body heat if I am chilled. Warm to you may be chilling to me. When I am chilled, I feel great fatigue; my veins contract. Things that have chilled me in the past are: air conditioning not adjusted for me; cold tables; refrigerated saline to humidify oxygen; refrigerated IV fluids; iced drinks, including water, etc.

The left side of my throat is weaker than the right; thus I am susceptible to choking on liquids.

Like the extremely elderly, I am very reactive to the effects of medicine; thus, I have always worked under the plan of starting with the lowest possible dose of any new medicine, and sometimes I wind up taking less than that.

These are my thoughts for the moment; now I can only hope for the success of all.

After-surgery note: I wish I had added to the list above two items. Because of my lack of muscle mass, my limbs offer no resistance to such things as blood-pressure cuffs, blood-work tourniquets, and manual positionings. My worst pain and a soreness that is still with me arose from the unnatural (to me) manipulation of my right shoulder on the operating table while I was under anesthesia.

Because of my enervated shoulder muscles, even the closure of incisions near my underarms became a major problem. The slightest roughness/lumpishness in that spot created a kind of a speed bump to my already-limited arm motion. I have recently had plastic surgery (under a local) to smooth out my right underarm.

I came through all of the surgery with little, if any, loss of function. Perhaps my arms and shoulders tire more quickly, but I am pleased that they serve me as well as they do. As for the cancer, the pathology report indicates that it was Stage One with negative lymph nodes, which means that it was caught early enough not to require follow-up radiation or chemotherapy. And because it contained no estrogen receptors, I am not even taking Tamoxifen. \square

Related resources:

Anesthetic Implications of Poliomyelitis by Joseph D. Tobias, MD; *The American Journal of Anesthesiology,* March/April 1995, pp. 102-104. New Neuromuscular Blocking Drugs by Jennifer M. Hunter, MB; *The New England Journal of Medicine*, Vol. 332, No. 25, pp. 1691-1699.

New Breathing Problems in Aging Polio Survivors: Respiratory Muscles Weakness continued from page 1

and had problems maintaining an adequate oxygen level. I was called in because the gynecologist and internist did not understand why the chest x-ray would not clear and why her oxygen level was so low. The first thing I ordered was a pulmonary function test to determine how strong her respiratory muscles actually were.

Pulmonary function tests ... In my opinion, the most accurate is a specific test of muscle strength. Some call it maximum inspiratory pressure. We call it negative inspiratory force. Expiratory strength can be measured by blowing as hard as possible against a resistance. We measure how much force is generated when someone tries to take a breath against an occluded mouthpiece. How hard one struggles to take a breath reveals how strong the respiratory muscles are. These special tests are helpful, easy to do, and they can be done at the bed-side. However, not all hospitals are properly equipped.

Standard respiratory testing involves having a clip placed over the nose while blowing as hard as possible. This measures actual lung volume and the ability to push air out in one second. Results of standard respiratory testing can give an indication of respiratory muscle weakness.

If any of the breathing muscles are weak, lung capacity and ability to expel air are reduced proportionally. It is important to note that with repeated effort most people have a learning curve and will get better results over the first three or four tries. On the contrary, someone with muscle weakness, who is asked to work harder and harder, will get worse with each try. A knowledgeable pulmonologist looking at that pattern will recommend tests to measure respiratory muscle strength.

Representative case ... The individual referred to earlier did have a significant decrease in respiratory muscle strength. The normal amount of pressure generated to breathe against an occlusion is about minus 60 to 70 centimeters of water pressure. She was generating about minus 20. As a matter of fact, no individual with prior neuromuscular disease who has been tested in my practice has done better than minus 26.

Stressors on the breathing system ... There may be no consequences of respiratory muscle weakness at all for someone in the minus 30 to minus 40 category until the breathing system is stressed, and surgery is often the stressor that highlights the fact that one has weak breathing muscles.

Infections also can highlight weak breathing muscles. If some find it difficult to cough, which is another function of the respiratory muscles, and have severe pneumonia, they may have difficulty clearing secretions, trouble maintaining oxygenation, and problems recovering from respiratory infection. Individuals with difficulty coughing due to weak muscles also may have a tendency toward bronchitis.

Medications also may unmask respiratory muscle weakness. Some post-polio individuals who believe they do not have respiratory weakness may take sleeping pills and then cannot cough, or take a deep breath. Others can react the same way to pain medications.