

Spinal Muscular Atrophy and Atelectasis: A Case Study

Tom Hall RN, RCP

Employed by Hill-Rom

Author Profile

Tom Hall, as Account Manager for Hill-Rom, has contributed to a variety of research endeavors and developed white papers, case studies and other supporting materials.

Case Study:

Spinal Muscular Atrophy and Atelectasis

Tom Hall, RN, RCP

The childhood spinal muscular atrophies (SMA) are a group of heritable neurodegenerative disorders, which vary greatly in terms of pathophysiology, severity, and prognosis.^{1,2} Severely afflicted individuals (Types I and II) usually die by the age of two or three. Those afflicted with milder forms of SMA (Type III) experience a slower disease progression and much longer life expectancy. In general, the SMA's are characterized by progressive weakness, abnormalities in spinal development, and deterioration in pulmonary function. In all forms of SMA, death is usually a result of respiratory failure secondary to bronchopneumonia.³ Prognosis is related closely to the level of pulmonary function.⁴

In Type III SMA patients, aggressive disease management is of paramount importance. With proper care, patients may live productive and satisfying lives. In addition to maintenance of joint mobility and muscle strength, treatment must focus on maximizing respiratory function. In SMA patients, pulmonary complications are related directly to the restrictions placed on thoracic movement and inspiratory ability by scoliosis, a universal feature of the disease. The intercostal and respiratory muscles are weakened, but the diaphragm is relatively spared, so a paradoxical breathing pattern and flaring of the lower portion of the rib cage are produced.⁵ The level of pulmonary dysfunction varies directly with the severity of the scoliosis.

The distorted thorax resulting from scoliosis affects normal tidal breathing as well as the ability of the patient to take a deep breath. Because of impaired inspiratory ability, patients with scoliosis may also suffer from atelectasis and associated pulmonary problems, including impaired gas exchange. Compromise in the ability to inhale maximally can interfere with the ability to generate an effective cough and thus diminish the ability to clear pulmonary secretions. Retained secretions provide a culture medium for bacterial pathogens. Consequent infections produce still more mucus, initiating a vicious cycle of mucus obstruction, recurrent bouts with pneumonia, bacterial colonization, and finally, the respiratory failure which accounts for most SMA deaths.

To prevent or manage such complications, rigorous bronchial hygiene, including effective airway clearance therapy, is an important component in the care of SMA patients.

The following is an account of a young SMA patient who was experiencing frequent, progressively worsening episodes of respiratory distress.

Case Summary

The patient is a nine-year old boy with SMA. In addition to progressive muscle weakening and wasting, this young man presented with severe scoliosis and other skeletal deformities, including pelvic distortion and a partially dislocated left hip. During surgical evaluation for possible transcutaneous rodding of the spine, a routine chest x-ray revealed marked atelectasis of the left lower lobe with a loss of volume and slight left shift of mediastinal structures. The etiology of the atelectasis was not clear, but differential diagnosis included secretion retention, ineffective cough, and restrictive lung disease secondary to skeletal deformity. Before developing atelectasis, the patient enjoyed relatively good health. He had suffered few upper respiratory infections (URIs) and required no hospitalizations for pulmonary exacerbations, although he had been hospitalized for orthopedic surgical procedures on his shoulder and hip. The patient exhibited mild to moderate paradoxical respiration and his rib cage showed distal flaring. Except during episodes of dyspnea, he used accessory muscles with minimal difficulty and recovered from oxygen desaturation quickly. His baseline heartrate was elevated, suggesting ventilation and perfusion mismatch.

During consultation with a pediatric pulmonologist, the patient's mother expressed concern that her son's airway clearance needs were not being met with traditional chest physiotherapy (CPT). Her anxieties were well founded; typically, patients with severe scoliosis cannot assume the positions required for effective drainage of secretions. To overcome these difficulties, the pulmonologist elected to try high-frequency chest wall oscillation (HFCWO) administered via The Vest™ Airway Clearance System ("The Vest™ system").

The Vest™ system, manufactured by Hill-Rom, consists of an air-pulse generator attached to an inflatable vest that covers the entire thorax. When the generator is turned on, the pressure in the vest increases and decreases between five and 25 times per second, thereby applying rapid oscillations to the patient's chest wall. This procedure is called high-frequency chest wall oscillation, or HFCWO. HFCWO has been shown to produce transient increases in airflow at low lung volumes, cough-like shear forces, alterations in the physical properties of mucus, and consequent increases in mucus mobilization. Because The Vest™ system operates according to a mechanism entirely different from traditional CPT, it provides effective airway clearance independent of postural drainage.

Method

The pulmonologist prescribed a regular schedule of The Vest™ system treatments to be delivered at a pressure setting of 5 and a frequency of 12 Hz. for sessions of 20 minutes administered twice daily. A registered respiratory therapist (RRT) was provided to train the mother, monitor the patient, and act as a liaison with the pulmonologist.

Results

On the first day of treatment, the RRT described a slightly built nine-year old male appearing pale and exhibiting circumoral cyanosis. His respiratory rate was 42 breaths per minute with a heart rate of 128 beats per minute. SaO₂ saturation was 92% on room air. Auscultation revealed diffuse ronchi over all fields and high-pitched expiratory wheezes bilaterally. The patient had a frequent but ineffective cough. The boy's mother reported that he was recovering from a URI and was on the fifth day of antibiotic therapy. He had been sent home from school that day because of respiratory distress.

To begin the initial treatment session with The Vest™ system, 2.5 mg of nebulized albuterol was administered via mask, followed by oraltracheal suction. A small volume of light yellow secretions was returned. The patient tolerated initial inflation and oscillation at the designated settings well. Following 2-3 minutes of oscillation, his cough rate increased, and orotracheal and pharyngeal suctioning was used to clear secretions. The treatment continued, alternating oscillation with frequent suctioning. Nebulized therapy was administered continuously during the 20-minute therapy session.

During therapy, the patient's respiratory and heart rates decreased gradually to 26-28 breaths per minute and 102 beats per minute. As therapy progressed, the secretions became increasingly tenacious and mucopurulent. SaO₂ increased from 92% to 96% after 8 minutes of therapy. Wheezing decreased, although, upon auscultation, ronchi persisted over all fields. The patient's color improved progressively, and he appeared to relax. The RRT stayed with the patient for an hour following HFCWO treatment to manage remaining secretions. During that time, the color of secretions changed from dark to pale yellow, and viscosity and volume diminished markedly. Heart rate, respirations, and SaO₂ stabilized respectively at 92 beats per minute, 26 breaths per minute, and 96%. The boy's mother volunteered that he occasionally experienced "blue spells" that she had attempted to treat with a combination of chest percussion, position changes, and suctioning. She was instructed to continue as ordered with routine nebulized therapy and to administer the second daily HFCWO treatment at bedtime.

During follow-up by telephone the next day, the mother reported to the RRT that the second treatment with The Vest™ system proceeded smoothly. Moderate volumes of secretions were mobilized and then cleared by suctioning.

The patient rested comfortably throughout the night and returned to school after a one day absence. The RRT passed this information on to the physician; schedules for both HFCWO and nebulized therapies were increased to t.i.d. for the next five days.

Five days after the initial treatment with The Vest™ system, the RRT visited the patient in his home. He found a bright, alert, cheerful young man lying on his bed studying spelling words. His color was good and he exhibited no signs of respiratory distress. During the interval, he had been able to tolerate a full six hour school day. For the first time in six months, he was able to wear rigid orthotics for spinal and cervical support for up to four hours. HFCWO therapy was going well, along with nebulizer treatments t.i.d., and his mother reported that he seemed to enjoy treatment sessions.

Ten days after HFCWO therapy had been initiated, the RRT made a second home follow-up visit. The patient appeared healthy and active, was continuing to use his rigid orthotics and, for the first time in a year, was able to travel to school in his wheelchair. His mother reported the appearance of "short, dark chunks" of material in his tracheal aspirate. The RRT believed those particles were probably mucous plugs being mobilized from peripheral areas of the lungs.

During home visits on days 20 and 30, the patient continued to demonstrate both clinical stabilization and impressive quality of life gains. Although particulate matter continued to appear in suctioned material, the patient no longer required "deep suctioning" and his previously fitful sleep pattern was now uninterrupted and restful. Scattered ronchi were resolved quickly with oropharyngeal suctioning. Daily therapy included nebulized treatments t.i.d. and HFCWO b.i.d. Wheezing and rales have ceased, and his SaO₂ has remained stable at 96-97% on room air.

Both parents stated independently that they have been deeply gratified by dramatic improvements in their child's social life. Since his stamina and sleep patterns have improved, he has been able to utilize his supportive orthotics without difficulty, and has shown a new zest for life. School absences are rare, and he has become a more active student. For the first time, he was invited to the home of a schoolmate for an after-school play session, and enjoyed an afternoon of activities, which included an assisted bike ride. The boys made plans for future play dates.

On day 28, during an automobile trip for a surgical consultation, the patient was able to be "up" for six hours, sitting by himself in the back of a van in a secured wheelchair. His parents reported that this was the first time in years that his mother did not need to sit in the back with him to be available for frequent suctioning. The entire family enjoyed the trip thoroughly. Both parents have expressed their belief that HFCWO has had a positive impact on their son's health. Because they no longer had to watch their child struggle for breath or spend anxious nights administering

suctioning, the parents reported feeling both a sense of relief and reduced stress.

The patient underwent orthopedic surgery for insertion of a Harrington Rod to reduce scoliosis progression, repair of a partial hip dislocation, and ligament lengthening. Although he experienced some of the post-operative complications anticipated in high-risk individuals with neuromuscular disease, the patient responded well to therapy and recovered uneventfully.⁶

Since the initiation of The Vest™ therapy, the patient has been seen in clinic every two to three weeks. Although he has suffered three URIs, none required emergency room visits or hospitalizations. Following the second URI, ipatropium bromide (Atrovent™) 0.5 ml was administered until his airway reactivity was stabilized. During the third URI, a chest x-ray was taken to rule out pneumonia. No evidence of pneumonia was present. On the contrary, the chest film revealed complete resolution of the persistent atelectasis in the left lower lobe and improved overall aeration in lung fields.

During the fall/winter semester, the young man missed eleven days of school, the fewest ever during that season. Subsequently, he has enjoyed his best ever overall school attendance record. Perhaps as a result of the support system provided by Hill-Rom, compliance with both HFCWO therapy and nebulized treatments has been excellent. The patient's viral URIs have been markedly milder and less debilitating than previous infections. After more than a year of The Vest™ therapy, and six months after orthopedic surgery, this bright, optimistic young man is attending school in a mainstreamed setting, excelling academically, and socializing with his peers. The chronic pulmonary problems, which prompted substitution of HFCWO for CPT, have been resolved completely, and the quality of his life has improved significantly.

Discussion

A literature search reveals very little documentation on the clinical course and natural history of Type III spinal muscular atrophy. Although the disease is generally regarded as a progressive condition, experienced clinicians have made the observation that, at least in some SMA patients, functional deterioration is a result of secondary effects, including scoliosis and contractures and not of continued loss of motor neurons.⁷ A recent study using reproducible strength measurement techniques suggests that although SMA patients do not gain strength at normal rates, neither do they lose it.⁸ Thus, SMA patients may suffer from a defect in motor unit maturation rather than progressive deterioration.

Pulmonary function appears to be the most important prognostic indicator in spinal muscular atrophy.⁹ A significant inverse linear relationship has been demonstrated between the severity of scoliosis and the percentage of

predicted vital capacity and peak flow. For many SMA patients, surgical modification of scoliosis offers important clinical and quality of life advantages.¹⁰ In patients undergoing surgical spinal stabilization with significant average curve correction, the decline in lung function seen preoperatively is not only reversed, but also markedly improved.¹¹

Prevention and early treatment of respiratory infections are the major management issues in type III SMA. Appropriate antibiotics should be given at the first sign of infection, and all measures taken to properly manage secretions and pulmonary insufficiency. In the case of the young man described in this report, those measures were not sufficient to stabilize him. His condition had deteriorated dangerously, and it was doubtful that he could survive surgery to correct his scoliosis. Following initiation of The Vest™ therapy, improvements in his respiratory status made him a much better surgical candidate.

Because this was not a formal study with controlled variables, it is impossible to assess with precision how much HFCWO contributed to the immediate clinical improvement and subsequent functional gains experienced by the young man described in this report. However, prior to beginning HFCWO airway clearance therapy, the patient's respiratory function and physical stamina were notably poor and both he and his family suffered significant deficits in their quality of life. The Vest™ system was the significant variable introduced to his treatment regimen, and the qualitative gains in his respiratory and physical status are unequivocal. Concomitantly, the patient's social functioning, sleep patterns, and energy level also improved. The family reported a reduction in levels of stress and anxiety. Our experience in this case suggests that further study is indicated concerning the role HFCWO might play in the prophylactic and maintenance respiratory care of SMA III patients. When the respiratory care and orthopedic problems of such patients are well managed, many may expect to live relatively full and independent lives.

References

- ¹ Russman BS, Melchreit R, Drennan JC. Spinal muscular atrophy: the natural course of the disease. *Muscle Nerve* 1983; 6(3): 179-181.
- ² Munsat, TL. The spinal muscular atrophies. *Current Neurology*, chapter 3, Vol. 14, 1994, (Mosby-Year Book, Inc.), pp. 55-71.
- ³ Chernick V, Boat TF, eds. *Kendig's Disorders of the Respiratory Tract in Children*, Sixth Edition, 1998. WB Saunders Co., Philadelphia, pp. 806-807.
- ⁴ Samaha FJ, Buncher CR, Russman BS, White ML, Iannaccone ST, et.al. Pulmonary function in spinal muscular atrophy. *J Child Neurol* 1994; 9(3): 326-329.
- ⁵ Munsat, op. cite, (n. 2).
- ⁶ Post-operatively, the patient developed atelectasis and Serratia species pneumonia; he was treated with IV antibiotics, traditional chest physiotherapy, and intermittent positive pressure breathing. Because there was initial concern that the surgical site and recently placed orthopedic hardware might be endangered by HFCWO, the medical team considered alternate modalities for secretion clearance; they elected to reinstate, under close supervision, a modified regimen

of The Vest™ Airway Clearance System therapy. With the posterior portion of the lumbar support orthotic in place and using a pressure setting of 2 and frequency of 20 Hz, the patient tolerated, with intermittent rest periods, a full 15-minute therapy session. Over the next five days, HFCWO therapy was ordered three times daily; infiltrates and atelectasis resolved and the patient was able to maintain SaO₂>95% without supplemental O₂. Two days later, he was discharged with orders to continue nebulized bronchodilator and The Vest therapy at home.

⁷ Munsat, op. cite, (n. 2).

⁸ Russman BS, Iannacone ST, Buncher CR, et. al. Spinal muscular atrophy: new thoughts on the pathogenesis and classification schema. *J Child Neurol* 1992; 7: 347-353.

⁹ Phillips, DP, Roye DP Jr, Farcy JP, et al. Surgical treatment of scoliosis in the spinal muscular atrophy population. *Spine* 1990; 15: 942-945.

¹⁰ Granata C, Cervellati S, Ballestrazzi A, et al. Spine surgery in spinal muscular atrophy: long-term results. *Neuromuscul Disord.* 1993; 3 (3): 207-215.

¹¹ Robinson D, Galasko CS, Delany C, Williamson JB, Barrie JL. Scoliosis and lung function in spinal muscular atrophy. *Eur Spine J* 1995; 4 (5): 268-273.