

Cystic Fibrosis with Severe Atelectasis: A Case Study

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Case Study:

Cystic Fibrosis with Severe Atelectasis

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In the United States, cystic fibrosis (CF) is the most commonly inherited, progressively debilitating disease among children. Impaired mucociliary clearance as a consequence of defective ion and water transport and the effects of chronic airway infections lead to pathological secretion retention and incremental pulmonary damage. The patient's clinical course is defined by recurrent, increasingly severe bouts of pulmonary obstruction, inflammation, and infection. Among such patients, respiratory infection and compromised pulmonary function are the leading causes of morbidity and mortality.¹ Moreover, the bulk of medical expenses, chiefly hospital costs, are incurred as a result of such respiratory diseases.

Epidemiological data demonstrate a dramatic improvement in survival for patients with CF over the last few decades, and projections suggest that trend will continue. Currently, the average CF patient may expect to live 30.6 years.² Considerable progress has been made in identifying treatments that delay the progression of pulmonary disease in cystic fibrosis and that promote an acceptable quality of life for such patients. Typically, those patients who do well are those maintained on individualized therapeutic regimens which include airway clearance therapy, appropriate antibiotics, the use of effective mucolytics and bronchodilators, anti-inflammatory therapy, and well-monitored nutritional support.³

Airway clearance therapy is the most important prophylactic and therapeutic intervention in the management of patients with CF. The removal of secretions has been shown to relieve atelectasis and prevent or slow the proteolytic destruction of airways by discouraging bacterial colonization.⁴ Unfortunately, it is the component of treatment most likely to be neglected. Chest physiotherapy (CPT), the airway clearance modality prescribed most frequently, is fraught with intrinsic weaknesses.⁵ CPT is a complicated, labor-intensive, time- and energy-consuming technique that requires the services of a trained caregiver, often several times daily. Properly administered CPT requires positioning for postural drainage, chest wall percussion, coughing, suctioning when indicated, and, in patients who are physically able, the application of breathing exercises. The entire procedure must be repeated until all regions of the lungs have been percussed and drained. As a consequence of such difficulties, the quality and frequency of CPT treatments often is poor. Not surprisingly, non-compliance with CPT is a major problem reported by virtually every CF clinic.^{6,7} Thus, the airway clearance needs of many CF patients are not being met.

The following is an account of a ten-year-old girl with significant lung disease secondary to CF. For a period of about three years, her health had been deteriorating rapidly. Chief among her problems were severe respiratory exacerbations resulting in multiple hospitalizations. Moreover, she was becoming so listless, malnourished, and socially withdrawn that at one point her physician suggested hospice placement. Her bronchial hygiene therapy, which included CPT treatments administered four to six times daily, was ineffective. During a clinic visit for an episode of severe wheezing and respiratory distress, in an effort to find an intervention to halt her disease progression, her physician prescribed high-frequency chest wall oscillation via a device called 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 mechanism of action of The Vest™ system's is not reliant on gravity, The Vest™ system provides effective airway clearance independent of postural drainage.

Case Summary

The patient is a ten-year-old girl whose health history has been precarious since birth. Gestation and delivery were normal, but shortly after her birth, the 6 lb 3/4 oz. infant was found to have meconium ileus. The condition was treated by surgical decompression and ileostomy. (Take-down of ileostomy and reanastomosis occurred in August 1996.) Subsequently, the infant was placed on extracorporeal-membrane oxygenation (ECMO) for five days and on a respirator for two weeks. The parents report that the baby was

given supplemental O₂ for three months, during which time they were told she had *Psuedomonas pneumonia*. At age one month, a presumptive diagnosis of CF was made.

The child began a precipitous decline beginning with a hospitalization for pulmonary exacerbation and *Aspergillus pneumonia*. She had a port-a-cath placed at that time. A month later, she was rehospitalized for recurrent pneumonia and failure to thrive.

During the half-dozen hospitalizations that followed, episodes of pneumonia were attributed to pathogens including

Staphylococcus Aureus, *Pseudomonas Aeruginosa*, and *Aspergillus*. In addition to pulmonary exacerbation, admitting diagnoses included chronic sinusitis, persistent failure to thrive, and lower-left-lobe atelectasis. In June of 1996, she underwent gastrostomy and fundoplication and flexible endoscopic sinus surgery.

On October 22, 1997, the patient was hospitalized for the eighth time since February 1996. The admission chest x-ray showed extensive bronchiectasis and persistent complete atelectasis in the lower left lobe. Despite aggressive therapy including IV antibiotics and supplemental oxygen, improvement was slight. The patient's pulmonary function remained poor, with an FEV₁ of 34% of predicted. After 17 days, the patient was discharged on supplemental oxygen and home IV therapy. Her prognosis was guarded. At that time, because her disease progression was rapid and intractable, the mother was asked to consider hospice care.

During the course of this young lady's illness, her family tried diligently to administer CPT. However, both parents worked outside of the home, and their daughter required airway clearance treatments several times daily. The child's grandmother, who cared for her during the workday, made efforts to deliver treatment, but was physically unable to administer effective chest percussion. Effective delivery of CPT was further complicated by 1) placement of a gastrostomy tube, which often caused reflux during the head-down position required for postural drainage⁸ and 2) chronic sinusitis, which caused severe head pain during certain postural drainage positions.

In August 1997, seeking an airway clearance method that would mobilize the patient's secretions more effectively than CPT, her physician prescribed The Vest™ system. At that time, the patient's weight had fallen from the 50th percentile of six months earlier to the 15th percentile, in part because her increased work of breathing was drawing heavily on her calorie stores. During a two-week family lakeside vacation, the child's mother reported that her daughter had remained inside the camper, listless, anorexic, and too weak to socialize with other children.

Unfortunately, the patient's insurance provider denied authorization for The Vest™ system, deeming the device "not medically necessary." For several months, an extensive series of resubmissions, denials, and appeals ensued. Because other therapeutic interventions were ineffective and the patient's health was declining rapidly, her physician contacted a case manager at Advanced Respiratory in an effort to obtain The Vest™ system. In March 1998, following hospitalization for a severe exacerbation, Advanced Respiratory dispensed a The Vest™ system to the patient without insurance authorization.

Method

The patient received The Vest™ system on March 17, 1998, and was trained by a healthcare professional within 24 hours. Treatments were administered four times daily for 20 minute intervals at a pressure setting of 5.5 and a frequency of 12 Hz. The patient was evaluated by her pediatrician at least every other day until March 23, and then followed up at 30, 60, and

180 days. The patient tolerated treatments with ease and adapted to the treatment regime without difficulty. At the same time, increased nutritional support was prescribed for the patient.⁹

Results

On March 2, 1998, a routine chest x-ray demonstrated chronic parenchymal changes throughout the right lung and evidence of atelectasis in the left-lower lobe with hyperaeration of the left-upper lobe. The patient's pulmonary status showed no improvement when compared to lung function determinations recorded during her severe exacerbation in October.

On March 16, 1998, the patient was seen in the clinic with wheezing and respiratory distress.

On March 18, following three Vest™ system treatments, improved breath sounds were noted.

On March 20-23, the patient's physician reported continued improvement in breath sounds in the atelectatic region of her lungs.

On April 12, the patient's physician noted a "marked improvement after implementation of her Vest™ system therapy." Her recovery from the exacerbation of early March had been rapid.

On April 20, 1998, during a clinic visit for a brief febrile episode, a chest x-ray revealed no new infiltrates superimposed on the chronic fibrosis in the right upper and lower lobes or in the consolidated left-lower lobe.

At 30 and 60 day follow-up interviews, it was noted that the patient continued to do well, although the atelectasis in her lower-left lobe remained persistent. Her cough was productive and her secretions were clearing well.

On September 15, 1998, 180 days after The Vest™ system therapy had been initiated, the patient's mother reported continued significant improvement. A check of The Vest™ system generator's "hour meter" monitor indicated the patient had complied precisely with her prescribed therapy protocol. After six months of treatment, there had been no serious pulmonary exacerbations and no hospitalizations since March. Fewer outpatient visits were required, and antibiotic therapy was reduced. Moreover, the patient's quality of life had improved dramatically. She enjoyed an active summer which included socializing with friends, participating in sleep overs, and enjoying water sports. Her energy level and sense of well being were improved significantly.

In October 1998, following a determination that progressive damage to the lower-left lobe was irreversible, the patient underwent a partial lobectomy for excision of the diseased portion. Recovery was rapid and uneventful. Post-operative chest x-rays demonstrated that both lungs were clear.

On January 30, 1999, in a follow-up phone interview, the patient's mother reported that the young lady remains healthy, her school attendance is good, and she is riding her bicycle regularly. An improved appetite has increased her weight to a current sixty-five pounds. She continues to receive Vest™ system treatments two to four times daily.

Discussion

Chronic illnesses are now the predominant form of disease in the United States. Therapies are ameliorative rather than curative. Chronicity creates recurrent and growing costs, physical suffering, and diminished participation in the good things life has to offer. To contain costs, delay disease progression, and facilitate an acceptable quality of life for chronically ill patients, it is imperative that the treatment options most likely to support these outcomes be identified and implemented.

It is widely accepted in the literature that, in chronic respiratory disease, the frequency and severity of pulmonary exacerbations is chiefly a consequence of neglected or ineffective bronchial hygiene. In CF patients, where the critical importance of routine secretion clearance is well understood, patients whose airway clearance needs are met experience slower disease progression, fewer, less severe respiratory complications, and fewer hospitalizations.¹⁰ Many enjoy longer, healthier lives.

Because this was not a controlled study, it is difficult to assess quantitatively the contribution of HFCWO to the rapid improvement and subsequent good health experienced by the patient described in this report. Certainly, increased nutritional supplementation and vigilant medical management contributed to the deceleration of this child's disease progression. Nevertheless, the dramatic clinical, physical, and quality of life gains she experienced following the substitution of The Vest™ system therapy for traditional CPT airway clearance treatment suggest several areas for further study: 1) In the case of this patient, implementation of The Vest™ system therapy coincided with a marked decrease in disease progression, hospitalizations, and use of auxiliary medical care services. What role might HFCWO play in effecting clinical improvement and reduced medical costs in CF and other categories of chronic disease patients with airway clearance needs? 2) Non-adherence to therapeutic regimens is the single most significant factor in the failure of airway clearance treatment. How might The Vest™ system therapy be self-administered, technique-independent, and impact compliance behavior? 3) How might the simplicity of The Vest™ system therapy in contrast to the demands of other airway clearance modalities translate into quality of life gains?

In the management of chronic illness, the goal of medical treatment must be to extend both the length and quality of a patient's life, reduce morbidity and suffering, and control medical costs. All these criteria appeared to be met by the implementation of The Vest™ system therapy for this patient.

References

- 1 Cystic Fibrosis Foundation, Patient Registry 1997 Annual Data Report; Bethesda, Maryland, September 1998.
- 2 Ibid 19.
- 3 Marshall BC, Samuelson WM. Basic therapies in cystic fibrosis. Does standard therapy work? *Clin Chest Med* 1998; 19 (3): 487-504, vi.
- 4 Williams MT. Chest Physiotherapy and cystic fibrosis: why is the most effective form of treatment still unclear? *Chest* 1994; 106:1872-1882.
- 5 Eid N, Buceit J, Neuling M, Phelps H. Chest physiotherapy in review. *Respir Care* 1991; 36(4): 270-282.
- 6 Coates AL. Chest physiotherapy in cystic fibrosis: spare the hand and spoil the cough? *J Pediatr* 1997; 131: 506-508.
- 7 Passero M, Remor B, Salomon J. Patient reported compliance with cystic fibrosis therapy. *Clin Pediatr* 1981; 20: 264-266.
- 8 Button BM, Heine RG, Catto-Smith AG, Phelan PD. Postural drainage in cystic fibrosis: is there a link with gastro-oesophageal reflux? *J Pediatr Child Health* 1998; 34(4): 330-4. In this study, the investigators conclude that episodes of gastro-oesophageal reflux increase significantly during the postural drainage component of CPT. Gastroesophageal reflux may be an important cofactor in the progression of pulmonary disease.
- 9 In March, the patient's physician prescribed Scandi-shakes to replace Propeptide. Scandi-shakes offer 600 cal/8oz in contrast to the 230 cal/8oz provided by Propeptide. Four months after beginning this regimen, the patient had not only stopped losing weight, but had gained 2 pounds.
- 10 Saxauer WP, Fiel SB. New treatment modalities for cystic fibrosis. *Curr Opin Pulm Med* 1995; 1 (6): 457-464.