Physical Therapy for Children with Chronic Lung Disease

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Chronic lung disease is a major health problem among children. Estimates suggest that one child in six has a chronic respiratory condition. This article reviews three common chronic respiratory conditions occurring in childhood for which physical therapy is usually recommended. The cause, pathophysiology, and medical treatment are explained for asthma, respiratory complications of chronic neuromuscular disease, and cystic fibrosis. The rationale for physical therapy and the concepts of treatment for the three disorders are presented. Major clinical studies that have attempted to document the efficacy of physical therapy are discussed. Questions for future research are proposed.

Key Words: *Respiratory disease, Pediatrics, Physical therapy.*

Chronic lung disease is considered to be largely a disorder of adults who have encountered the ravages of tobacco, pollution, industrial exposure, disease, and time. A surprising fact is that about 18 percent of all children have some form of chronic respiratory dysfunction. Included in this percentage are children with nonserious conditions, such as hypertrophied tonsils and adenoids, but figures also show that an estimated 4 percent of all children have asthma, 15,000–20,000 have cystic fibrosis (CF), and many thousands with neuromuscular disorders have respiratory system involvement.[1] Physical therapy has been shown to be effective in the treatment of children with respiratory problems, and is used extensively in the comprehensive care of these patients. The purposes of this article are fourfold: 1) review three common causes of chronic lung disease in children—asthma, neuromuscular disease, and CF; 2) discuss physical therapy evaluation and treatment; 3) review pertinent literature; and 4) suggest directions for future research.

DISEASES

Asthma

Asthma, a reversible obstructive airways disease, has many varied definitions, but included in most are the following characteristics. Asthma results in widespread diminution of airway cross-section diameter due to the contraction of bronchial smooth muscle mediated by the parasympathetic division of the autonomic nervous system. Asthma occurs episodically, but may also be chronic and perennial. It may be stimulated by a multitude of factors including inhaled allergens, food, exercise, respiratory infection, environmental irritants, dry or cold air, and intense emotions. The major diagnostic feature of the disease is its reversibility by either therapeutic means or spontaneous occurrence.[2]

Estimates that asthma affects as many as 10 percent of the pediatric population may make this respiratory disease the most prevalent chronic disorder of these patients. Asthma mortality is rare, but its morbidity, noted by days lost from school and numbers of hospitalizations, is enormous.[3] Asthma in children is associated with allergic (atopic) individuals with a positive family history of asthma. Boys are afflicted at about twice the rate of girls. Symptoms or episodic attacks in children are often precipitated by respiratory tract infection, and may be intensified by severe emotional lability. The phenomenon of exercise-induced bronchospasm is present in a majority of children with asthma.[2]

The pathophysiology of asthma is airways obstruction caused by stimulation of the parasympathetic nervous system when specific mediator substances are released from mast cells within the airways. Parasympathetic stimulation causes bronchial smooth muscle contraction, edema of the bronchial epithelial lining, and secretion of mucous from goblet cells and mucous glands. These events cause airways obstruction resulting in an increased resistance to expiratory flow, an increase in hyperaeration, and a mismatching of ventilation and perfusion that causes arterial hypoxemia. Children with an exercise-induced component to their asthma respond similarly to strenuous exercise. After about six minutes of exercise during which the heart rate is maintained at about 170 bpm, and about five or six minutes after cessation of the exercise, a sudden drop in airway function occurs that is

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similar to an "attack" of asthma. This phenomenon has implications for the physical therapist when developing a rehabilitation program for an asthmatic child.

Medical treatment of chronic asthma includes use of pharmacologic agents, environmental control, and immunologic hyposensitization. Among the pharmacologic agents are sympathomimetics (taken orally or by aerosol), methylxanthines, corticosteroids, and sodium cromoglycate (taken by inhalation). These medications use differing mechanisms in an attempt to reduce bronchospasm. Environmental control of allergens is an important mode of therapy. The major factors are no dust, irritants, pets, or allergic foods. The active child who is likely to provoke exercise-induced bronchospasm must learn to use appropriate medications prior to strenuous exercise. Finally, many physicians use immunotherapy as a means of control. Once the allergen has been identified by testing, minute doses are periodically injected intramuscularly to stimulate the child's immunologic defenses that produce antibodies to the allergens. When sufficient antibodies are present, they will obviate the response of the child to the offensive allergen.

Chronic Neuromuscular Disease

Chronic neuromuscular disease in children may affect any part of the neuromuscular chain that is responsible for respiratory muscle contraction. Therefore, the respiratory complications of chronic neuromuscular disease in children are largely due to respiratory muscle weakness and dysfunction. The basic pathologic process will often determine the nature of the respiratory complications: mild and temporary or severe and progressive.

Diffuse pathologic conditions of the CNS caused by disease, trauma, or perinatal anoxia may result in acute respiratory failure, chronic respiratory insufficiency, or complications associated with poor secretion clearance and decreased lung volumes. Children with severe problems in motor control or cerebral palsy often have perinatal anoxia as the etiologic condition. Altered reflex and neural control mechanisms may change the physiologic response to chemical and mechanical stimuli within the lung, brainstem, major vessels, blood, and cerebrospinal fluid. These altered mechanisms are often found in children with familial dysautonomia, sleep apnea, and obesity-hypoventilation syndrome. Spinal cord injury can result in numerous respiratory complications, which have been described elsewhere in this special issue. Lesions of the anterior horn cell, such as Werdnig-Hoffmann and Guillain-Barré syndromes, have common respiratory complications that often lead to the mortality associated with these disorders. Finally, primary degenerative muscle diseases (pseudohypertrophic muscular dystrophy being the classic example) exhibit a progressive decline in pulmonary function. This disease progression ultimately diminishes the child's ability to maintain adequate oxygenation and carbon dioxide levels in the arterial blood and death is often the result of respiratory failure following an acute pneumonia. Regardless of disease process, the respiratory complications can be described as altered and inefficient breathing, lack of thoracic and shoulder mobility, and inability to adequately clear mucous secretions.

Cystic Fibrosis

Cystic fibrosis is the most common lethal genetic disorder of the white race. First identified in 1936, CF occurs once in every 2,000 white children, and has an estimated carrier rate of 5 percent. Cystic fibrosis is an exocrine disorder characterized by the symptom triad of high sweat electrolytes, pancreatic enzyme deficiency, and chronic suppurative lung disease. Other body systems involved include the portal system and genitourinary system of the male. Because CF is a multisystem disorder, the appearance of symptoms is extremely variable and the disease is often misdiagnosed as asthma, allergy, chronic diarrhea, celiac disease, or chronic bronchitis. Cystic fibrosis is transmitted in a Mendelian recessive pattern. When both parents are carriers there exists for each pregnancy 1) a 25 percent chance of the offspring having the disease, 2) a 50 percent chance of it being a carrier, and 3) a 25 percent chance of it being free of the CF gene. There is currently neither an accepted heterozygote (carrier) test nor a prenatal homozygote (disease) test.[4]

The clinical course of the disorder, like its onset, is extremely variable. Although CF is still considered a fatal disease, the survival rates have continued to improve slowly for the past 25 years. Recent reports of large numbers of adults with CF have appeared.[5,6] It must be noted that CF is one of the few diseases among young people in which there seems to be a female disadvantage.[7]

Pulmonary involvement in CF, which results in the greatest mortality, begins when thick voluminous mucous secreted by goblet cells and mucous glands is retained within the smaller airways. These secretions provide a culture medium for bacterial pathogens, notably *Staphylococcus aureus* and *Pseudomonas aeruginosa*, with the resultant infection producing still more mucous. A vicious cycle of mucous obstruction and infection is begun. Continual reinfection leads to bronchiectasis and bronchiolectasis, irreversible processes causing destruction of bronchial structures. In addition to the destructive process, the goblet cell and mucous gland hyperplasia results in copious volumes of purulent secretions that cause the severe airways

obstruction common to CF. The progression of pulmonary disease and efficacy of treatment appear to play a major role in determining survival for children with CF. The pulmonary complications of CF include pneumothorax, hemoptysis, lobar or segmental atelectasis, pulmonary artery hypertension, and cor pulmonale. The pulmonary aspect of CF has been fully described previously.[4,8]

Medical treatment of the pulmonary symptoms is directed toward controlling the pulmonary infection. The keystone of medical care is the identification of bacterial pathogens and determination of their sensitivity to antimicrobials. Appropriate choice of medication is based upon the results of sputum culture. Antimicrobials may be administered orally or parenterally. Currently there is no effective oral preparation to combat *Pseudomonas aeruginosa* and intravenous or intramuscular methods must be used.[4]

Management of gastrointestinal symptoms, aggressive nutritional support, physical rehabilitation, and early psychosocial and emotional counseling are all made available through a national network of CF centers supported by the Cystic Fibrosis Foundation. Because of the improved life expectancy, career counseling, marital counseling, and family therapy have all become important issues in caring for individuals with CF.

PHYSICAL THERAPY EVALUATION AND TREATMENT

Physical therapy evaluation of children with chronic lung disease must include not only a comprehensive assessment of the respiratory status, but should also determine the child's general physical condition. The respiratory evaluation should examine the pattern of breathing, strength of the respiratory musculature, mobility of the thorax and shoulders, and ability to clear secretions. The nonrespiratory evaluation should determine overall strength, range of motion, endurance, and response to physical activity.

Pattern of Breathing

By examining the child's breathing pattern, the therapist can answer two major questions: Is the movement of air sufficient to ventilate the major lung fields without wasting a large portion of that ventilation? Is an economical muscular pattern used for breathing?

Auscultation to determine air entry and measurement of minute volume (respiratory rate times tidal volume) provide an objective answer to the first question. If an area of lung served by a patient's bronchial tree is poorly ventilated, the therapist must use localized thoracic expansion exercises to encourage increased movement of air toward the underaerated portion of lung. When a child has a rapid respiratory rate, breaths are more shallow and a larger percentage of the minute volume ventilates only the anatomical dead space. In this inefficient pattern of breathing, decreased air movement to the distal portions of the lower lobes is common. The therapist may teach the child to slow the rate and increase the volume of each breath by counting or by using a spirogram, metronome, or clock. The therapist must take care to maintain a rate and volume within normal limits for the child, or the presumed benefits of an improved ratio of minute ventilation to dead space ventilation may be lost.[9] This problem of rapid, shallow breathing is common to many children with chronic respiratory problems, but is particularly troublesome for those with neuromuscular disease.

The most efficient muscular pattern for inspiration is use of the diaphragm with assistance from the external intercostal muscles. When the child is acutely ill, the use of accessory muscles of inspiration and expiration to augment ventilation is physiologically and mechanically appropriate.[10] When the acute illness abates, many children with asthma and CF continue to breathe with the energy-wasting accessory muscle pattern. The physical therapist must train the child to discontinue this inefficient type of breathing. Suggested training methods include use of relaxation techniques and, more recently, sensory feedback devices. No objective data exist, however, to support the notion that short-term changes achieved during treatment have a residual effect on the muscular pattern of breathing.

Respiratory Muscle Function

Evaluation of inspiratory muscle strength and endurance as well as development of a strengthening program (when appropriate) are important facets of physical therapy for children with chronic lung disease. Based upon an objective evaluation, the therapist may develop a training program for inspiratory muscles. Merrick and Axen recently described objective expressions of inspiratory muscle shortening, maximal velocity of shortening, and strength by using spirometry, flow rates, and maximal inspiratory pressures, respectively.[11] As with other skeletal muscles both strength training and endurance training must be considered for respiratory muscles depending on whether increased strength or endurance is the specific goal. Improved respiratory muscle function may be beneficial when an increase in ventilation is necessary during periods of increased physical activity or during acute exacerbations of the child's chronic respiratory illness.

One traditional method of inspiratory muscle strengthening has been placing weights upon the epigastric area. The patient works against the resist-

ance by contracting the diaphragm with a deep inspiration.[12] Other means of inspiratory muscle strengthening include breathing against artificially increased resistance, using incentive spirometry to improve volumes, and using a rapid, deep breathing maneuver called the maximal sustainable ventilatory capacity (MSVC).[13-15] Improving strength and endurance in the accessory muscles may augment inspiratory reserve for periods of ventilatory stress.

Musculoskeletal Mobility

Regardless of disease, the child whose thorax or shoulder girdle lacks full range of motion will have increased work to maintain adequate ventilation. Thoracic expansion should be measured in the transverse, anteroposterior, and vertical planes for inspiration and expiration. Range of motion and configuration should be noted for the thoracic part of the spine and the shoulder girdle including acromioclavicular, sternoclavicular, and glenohumeral joints. Active efforts to improve chest expansion may be done for localized areas or for the entire thorax. These active efforts may be augmented by use of incentive spirometers or, in the presence of inspiratory muscle weakness, by use of intermittent positive pressure devices. A child, whose thorax is normally more compliant than that of an adult, should respond well to these techniques. Improved range of motion in the shoulder joints will also help improve thoracic mobility. Specific treatment techniques have been discussed by others.[12,16]

Bronchial Hygiene

Secretion clearance, a major concern for physical therapists who work with children having acute respiratory illnesses, remains a great concern for children with chronic lung disease. Because of the small cross-sectional area of a child's airway, the predisposition exists for small amounts of mucous to occlude abruptly and completely the peripheral bronchioles and the larger, more proximal bronchi. When a large percentage of airways is occluded by secretions, chronic respiratory dysfunction can rapidly deteriorate into acute respiratory failure. A critical portion of the physical therapy assessment, therefore, is evaluation for secretions and determination of the child's ability to clear those secretions actively.

Several evaluative procedures will aid in locating secretions. Auscultation of the lungs is a useful method for identifying airways occluded by mucous. Rhonchi, wheezes, and, occassionally, moist rales denote secretions. Decreased or absent breath sounds may indicate atelectasis that is often accompanied by mucous obstruction of an airway. Palpation of the thorax for tactile fremitus is an alternate means of finding secretions in larger airways. Chest radiographs are not helpful for identifying secretions except when there is radiographically visible evidence of atelectasis, pneumonia, or bronchiectasis. Older children, when questioned, can identify areas of their lungs in which they are congested.

The child's cough must be evaluated. Necessary components for an effective cough include a large inspiratory volume and a coordinated glottic (upper airway) closure closely followed by a sudden forceful contraction of the abdominal muscles. These three events will cause a marked increase in intrathoracic pressure that is released, as a cough, by a sudden opening of the glottis. The therapist must assess the child's inspiratory effort, ability to control the glottic musculature, abdominal muscle strength, and coordination of these factors to produce a cough that will raise sputum.

Treatment to clear secretions in children with chronic lung disease varies depending generally on the disease process and its severity and specifically on the amount and viscosity of the secretions being produced. A child with asthma will have intermittent production of secretions that is more prevalent following an exacerbation of the asthma or a respiratory infection. Secretions in the child with neuromuscular disease vary widely. A youngster with muscular dystrophy may have no secretion retention until very late in the progression of his disease, yet a child with cerebral palsy who has poor swallowing function and chronically aspirates food may have large amounts of secretion that need to be cleared. The youth with CF who has significant pulmonary involvement will need daily secretion clearance.

Modalities of treatment to clear sputum, that also must be taught to parents, include positioning for gravity-assisted bronchial drainage; manual techniques including percussion, vibration, and chest-wall shaking; and secretion evacuation including coughing, huffing, and, if necessary, endotracheal aspiration. These techniques have all been described elsewhere in this issue.

Physical Rehabilitation

In addition to the respiratory evaluation, the child's overall physical strength, exercise tolerance, and posture must be assessed. In a recent review of pulmonary rehabilitation, Lertzman and Cherniack did not examine pediatric rehabilitation.[17] Quantitative strength testing for major muscle groups can be performed with equipment routinely available in the physical therapy department. Evaluation of posture may take different forms, but a commonly used method includes a grid system as described by Kendall and Boynton.[18] Exercise tolerance and endurance may be determined either by sophisticated exercise testing or

by semiquantitative means. A useful and simple index of fitness is the heart rate achieved during a standard work load and the time necessary for heart rate to return to the preexercise resting level. A treadmill, bicycle ergometer, or timed distance walking can be used as the work load.[19] The child with exercise-induced asthma must receive before the exercise period medication to prevent bronchospasm. Also, the child with neuromuscular dysfunction should be evaluated, if appropriate, for coordination, ability to perform activities of daily living, developmental level of function, and oral motor function.

A physical rehabilitation program based upon the strength and exercise tolerance assessment should be developed for the child. Standard methods of muscle strengthening may include progressive resistive exercise, isometric exercise, and use of isokinetic devices. Training methods to improve exercise tolerance may include running on a treadmill, riding a bicycle ergometer, free-running, jogging, or swimming. In children with advanced lung disease, arterial blood gas determinations during exercise are necessary to document the child's ability to maintain adequate levels of carbon dioxide and oxygen.

REVIEW OF LITERATURE

Except for secretion removal techniques, there is a dearth of well-designed, controlled studies to support or refute our customarily performed treatments for children with chronic respiratory disorders. This lack of objective data is most apparent for breathing retraining (including altering breathing pattern) and increasing thoracic mobility. Clinicians commonly refer to studies performed using adult subjects with chronic lung disease and infer that the benefits of treatment will apply to children. However, the significant difference between adult and pediatric respiratory physiology, especially for mechanics of breathing, makes extrapolation between the two groups speculative at best.[20]

Pattern of Breathing

There have been no studies evaluating attempts to alter patterns of ventilation in children by changing the pattern of respiratory muscle use. Nor have investigators justified the claim that children will be spared physical work by changing their breathing pattern from that of shallow breathing with accessory muscle predominance to a pattern increasing tidal volume and decreasing respiratory rate. One author has suggested that because of the increased inspiratory work needed to achieve large lung volumes by overcoming elastic recoil of the lung and chest wall, a pattern of rapid, shallow respiration may be more efficient for the child in respiratory distress than the supposedly improved pattern of slow, deep breathing.[21] Another recent study of deep inspiratory and expiratory efforts in patients with asthma reported a large increase in airway resistance following deep inspiration and a similar, but weaker, response following deep expiration. Therefore, the use of unusually deep respiratory efforts in children with asthma must be cautiously initiated because of the possibility of increased airways resistance.[22]

Respiratory Muscle Function

Based on the results of several recent projects, the concept of specific strength and endurance training for the respiratory muscles appears to be justified for respiratory treatment programs. Merrick and Axen state that the diaphragm, like other skeletal muscle, can adapt to training because its fiber composition and oxidative capacity are modified in response to functional demands.[11] Because the diaphragm, and, presumably, other respiratory muscles, will modify themselves in response to training regimens, it is necessary to adapt a respiratory training program to the appropriate needs of the child. Leith and Bradley demonstrated that static contractions result in increased respiratory muscle strength and that deep breathing at high rates improves endurance but not strength.[23] Hence, a program that includes high resistance to respiratory effort combined with increased rate and depth of ventilation will (as has been demonstrated for patients with quadriplegia) improve respiratory muscle strength and endurance.[13]

A well-conceived and designed study of ventilatory muscle endurance training was reported by Keens and associates, in 1977, using patients with CF as subjects and physical therapists as controls.[15] The CF subjects were assigned to one of two groups—ventilatory muscle training or general physical activity. The former group engaged in a ventilatory training program using the MSVC as a training modality. The MSVC, a technique of fast and deep breathing similar to the breathing in the maximal voluntary ventilation test, is used for respiratory endurance training. The latter group was engaged daily in vigorous physical activity such as rowing and basketball. After a four-week period of daily MSVC training, the ventilatory training group had a statistically significant improvement in MSVC of 52 percent. After an identical MSVC training program, the control group showed an improvement in MSVC of 26 percent. The remarkable finding in this study was the 57 percent improvement in MSVC for the physical training group after four weeks of daily activity. The authors postulated that the normal ventilatory response to vigorous physical activity served as a training tool to increase endurance of the ventilatory muscles.[15]

Bronchial Hygiene

Of all modalities included in chest physical therapy, the most definitive studies have been performed in an effort to document the effects of bronchial hygiene. Most of these studies had as subjects patients with CF, but at least one author confined his investigation to children with asthma. The experimental efforts can be divided based upon dependent variables. A number of authors have examined the effects on pulmonary function of bronchial hygiene, and others have relied upon changes in sputum transport or removal.

Denton studied 23 patients with CF who were alternately treated with gravity drainage and cough, gravity drainage and placebo, and gravity drainage with mechanical percussion and vibration.[24] With gravity and cough and with gravity and placebo, 19 of 23 and 18 of 23 patients, respectively, produced no measurable sputum. With the addition of percussion and vibration, 22 of the 23 subjects produced sputum ranging in volume from 4.5 ml to 40.0 ml.

Lorin and Denning found an increased amount of sputum per cough and per treatment session with 20 minutes of gravity drainage as well as percussion and vibration when compared to a control test of five voluntary coughing attempts.[25] Their subjects with CF were treated in five gravity-assisted drainage positions.

Wong and colleagues examined the effects upon mucociliary tracheal transport rates of gravity drainage in subjects with CF.[26] They found that when patients were in a 25 degree Trendelenburg's position, there was a marked increase in mucociliary transport in those who had initially absent or abnormal transport rates.

Several groups of investigators evaluated the effects on pulmonary function of bronchial drainage, percussion, and vibration in patients with CF.[27-29] Motoyama, in 1973, and Feldman and associates, in 1976, found improvement in test results of both large and small airway function. This improvement was of the greatest magnitude at 45 minutes after treatment.

Levison and Godfrey found no change in four pulmonary function values after a treatment that consisted of only two drainage positions.[30] Schwenk and associates reported no change in pulmonary function following drainage and percussion in 10 patients with CF who had only mild pulmonary symptoms.[31] That these two studies found no improvement in pulmonary function is no surprise. Levison and Godfrey employed only two drainage positions—hardly to be considered a complete treatment. By studying mildly involved patients, Schwenk and associates did not have the most important indication for treatment—large amounts of secretion.

Huber and co-workers treated one of two groups of children having moderately severe bronchial asthma.[32] The treatment group, which received postural drainage with percussion, showed a 10.5 percent increase in mean forced expiratory volume in the first second (FEV_1) compared to a slight decrease in FEV_1 for the control group.

Two studies have compared the efficacy of mechanical devices and manual techniques for chest percussion and vibration.[33, 34] Children with CF were treated using gravity-assisted bronchial drainage and both manual and mechanical percussion and vibration. Both treatments produced improvement in expiratory flow rates, indicating improved airway status, but no difference could be determined between the two mechanisms for loosening secretion.

We can state with certainty that for children with CF who have disease of moderate severity or worse, and possibly for children with asthma, bronchial drainage with percussion and vibration is beneficial over a short period. Secretion removal is greatly enhanced and large and small airways obstruction, denoted by pulmonary function tests, is decreased.

Physical and Breathing Exercises

Physical reconditioning to improve cardiovascular endurance and work capacity is another aspect of physical therapy for children with chronic lung disease.

Because swimming appears to provoke fewer symptoms of exercise-induced asthma, several investigators have conducted controlled studies of swimming as physical rehabilitation for children with asthma.

Sly and colleagues had a group of children with documented asthma engage in a 13-week program of two-hour swimming sessions three times each week.[35] Pulmonary function, personality traits, and numbers of days with wheezing were measured for the swimming group and for a similar group of control patients before and during the 13-week period. No change was observed for either group for pulmonary function or personality traits but the swimming group had a decrease in mean days wheezing from 31.3 before to 5.7 during the program. The number of wheezing days for the control group increased during the control period.

Fitch and associates had 46 children with asthma participate in a five-month swimming program and had a control group of 10.[36] Documented were a semiquantitative asthma score, physical and work capacity at heart rate of 170, a quantitative drug score, FEV_1, and response to an exercise challenge. Children in the treatment group had a statistically significant improvement for all values except FEV_1, and the parents spontaneously reported improved posture in these children. The bronchospastic response to a standardized exercise challenge remained unchanged.

There have been several efforts to evaluate the effects upon lung function of pulmonary rehabilitation programs for children with Duchenne muscular dystrophy (MD), and a recent study examined the benefits to children with cerebral palsy of a similar pulmonary program. Although the results of each study appeared promising, factors inherent in the experimental designs, including small numbers of subjects and lack of statistical analysis, prohibit generalizations regarding the usefulness to improve pulmonary function of exercises in children with chronic neuromuscular disabilities.

Hobermann, in 1955, studied seven subjects with MD whose vital capacities were below 75 percent of the predicted value.[37] The children, aged 7 to 14 years, took part for four months in an intensive rehabilitation program that included breathing exercises. Improved vital capacity was reported following the program, but no data were presented.

Houser and Johnson studied seven pairs of subjects with MD matched for age and functional level.[38] Forced vital capacity (FVC), forced expiratory flow between 25 percent and 75 percent of vital capacity (FEF_{25-75}), maximal voluntary ventilation (MVV), and peak expiratory flow rate (PEFR) were measured before and after a 12-week breathing program. The program included intermittent positive pressure breathing (IPPB) for six minutes on five days each week and deep breathing exercises, coughing assistance, and forced expiratory flow games used three days each week. At the end of the program no statistically significant differences between the treatment and control groups were found for pulmonary function.

Adams and Chandler found increased vital capacities in three boys with MD enrolled in a 30-minute program of swimming, water games, and IPPB in addition to routine physical rehabilitation activities.[39] The authors noted that when the swimming and IPPB regimen were discontinued, three subjects had losses in vital capacity. No statistical analysis of the data was performed.

Siegel, in 1975, instructed 10 ambulatory subjects with MD in techniques of diaphragmatic breathing, bronchial drainage, and IPPB.[40] Following three months of treatment, pulmonary functions were repeated and showed improvement in all of seven tests, except FEF_{25-75}, in all subjects except one. The subject who did not improve showed a marked decrease in general muscle strength during the testing period. As with two of the three studies of children with MD described above, no statistical analysis of data was performed.

Rothman studied breathing exercises in 10 children with spastic cerebral palsy.[41] Five subjects in the experimental group were matched as closely as possible for type of cerebral palsy, severity, age, IQ, and height with five other subjects who comprised the control group. The exercise program included eight activities whose sequence was varied during the eight weeks of the training project. The FVC and FEV_1 were measured at the beginning and end of the eight-week exercise schedule. The results showed a mean increase in FVC of 0.46 liter (31%) for the experimental group over their initial values. The control group had no change in FVC. The FEV_1 was within normal limits for both groups and did not change with the exercise program.

Major studies of physical reconditioning for children with CF have not been reported. Several authors have examined the responses of children with CF to vigorous exercise. Larson and Souhrada studied the pulmonary responses to exercise in 12 children with CF of mild or moderate severity.[42] They concluded that a majority of subjects had either no change or improvement in tests of airway function and a slight improvement in oxygen saturation. These changes were sustained for up to 30 minutes after the cessation of exercise.

Cropp and associates studied peak work rate, peak heart rate, minute ventilation, end-tidal CO_2, and oxygen saturation during incremental exercise for 17 normal children and 20 children with CF.[43] Children with mild or moderate CF had no significant differences from the normal child in any of the above values. Subjects with severe CF had significant diminution of all values. The authors concluded that limitation of physical activity was unnecessary for children with mild or moderate CF, but in those with severe involvement mild exertion could elicit significant arterial oxygen desaturation and carbon dioxide retention.

DIRECTIONS FOR FUTURE STUDY

Exciting possibilities exist for future study of the effects of physical therapy on children with chronic lung disease. To develop a list of priorities for research is difficult, but a suggested group of questions to be examined appears below.

Will efforts to expand localized areas of the thorax result in improved ventilation to the adjacent lung? This question has been studied inconclusively for the adult population, but the very compliant chest wall of the child may increase the likelihood of improved ventilation accompanying localized expansion of the chest.

Children with respiratory distress are routinely taught to decrease accessory muscle use and to breathe more deeply at a slower rate. Are these two objectives beneficial? If the energy cost of breathing decreases when the child performs as taught, is the decreased energy expenditure transient, or can it be prolonged?

Will the attainable increase in respiratory muscle strength and endurance improve either exercise tolerance or the ventilatory response to increased activity or disease? Must the child continue the exercise program in order to maintain improved respiratory muscle function?

Bronchial hygiene is the most carefully studied of all physical therapy modalities for children with chronic lung disease. However, no study has attempted to document changes occurring over periods greater than several minutes or hours. With the vast amount of time and effort spent providing bronchial hygiene, we must examine the long-term effect of this treatment on disease progression, especially for CF.

Physical rehabilitation and breathing exercises appear to be beneficial for children with chronic lung disease. The pulmonary effects of physical reconditioning and breathing rehabilitation have yet to be conclusively demonstrated for children with chronic neuromuscular disease or CF.

SUMMARY

This article has provided a brief description of three major groups of chronic lung disease in children. Asthma represented a reversible respiratory disease process; neuromuscular disease represented a nonpulmonary disease causing respiratory complications; and CF represented a progressive, irreversible pulmonary disorder. Physical therapy for the child with chronic lung disease was described. Studies of the efficacy of physical therapy were reviewed. The author has suggested several topics for future study in the hope that therapists with knowledge and interest in clinical investigation will be motivated to seek an answer to one or more of the questions raised.

REFERENCES

1. Prevalence of Selected Chronic Respiratory Conditions, United States, 1970. National Health Survey, National Center for Health Statistics, US Department of Health, Education and Welfare, publication number (HRA) 74-1511, 1973
2. Godfrey S: Childhood asthma. In Clark TJH, Godfrey S (eds): Asthma. Philadelphia, PA, W.B. Saunders Co, 1977, pp 324–331
3. Gordis L: Bronchial asthma. In Gordis L (ed): Epidemiology of Chronic Lung Disease in Children. Baltimore, MD, Johns Hopkins University Press, 1973, pp 4–52
4. Wood RE, Boat TF, Doershuk CF: Cystic fibrosis. Am Rev Respir Dis 113:833–878, 1976
5. Shwachman H, Kowalski M, Khaw KT: Cystic fibrosis: A new outlook. Medicine 56:129–149, 1977
6. diSant'Agnese PA, Davis PB: Cystic fibrosis in adults. Am J Med 66:121–132, 1979
7. Report of the Patient Registry. Rockville, MD, Cystic Fibrosis Foundation, Volume 1978
8. Tecklin JS, Holsclaw DS: Cystic fibrosis and the role of the physical therapist in its management. Phys Ther 53:386–393, 1973
9. Watts N: Improvement of breathing patterns. Phys Ther 48: 563–576, 1968
10. Campbell EJM, Agostini E, Davis JN: The Respiratory Muscles: Mechanics and Neural Control. London, England, Lloyd-Luke Ltd, 1970
11. Merrick J, Axen K: Inspiratory muscle function following abdominal weight exercises in healthy subjects. Phys Ther 61:651–656, 1981
12. Adkins HV: Improvement of breathing ability in children with respiratory muscle paralysis. Phys Ther 48:577–581, 1968
13. Gross D, Riley E, Grassino A, et al: Influence of resistive training on respiratory muscle strength and endurance in quadriplegia, abstracted. Am Rev Respir Dis 117:343 (April pt 2), 1978
14. Martin RJ, Rogers RM, Gray BA: The physiologic basis for the use of mechanical aids to lung expansion. Am Rev Respir Dis 122(Suppl):105–107, 1980
15. Keens TG, Krastins IRB, Wannamaker EM, et al: Ventilatory muscle endurance training in normal subjects and patients with cystic fibrosis. Am Rev Respir Dis 116:853–860, 1977
16. Warren A: Mobilization of the chest wall. Phys Ther 48:582–585, 1968
17. Lertzman MM, Cherniack RM: Rehabilitation of patients with chronic obstructive pulmonary disease: State of the art. Am Rev Respir Dis 114:1145–1165, 1976
18. Kendall HO, Boynton DA: Posture and Pain. Baltimore, MD, Williams & Wilkins Co, 1952, pp 5–14
19. Lunsford BR: Clinical indicators of endurance. Phys Ther 58: 704–709, 1978
20. Hogg JC: Age as a factor in respiratory disease. In Kendig EM, Chernick V (eds): Disorders of the Respiratory Tract in Children. Philadelphia, PA, W.B. Saunders Co, 1977, pp 177–187
21. Pride NB: Physiology. In Clark TJH, Godfrey S (eds): Asthma. Philadelphia, PA, W.B. Saunders Co, 1977, p 45
22. Gayrard P, Orehek J, Grimaud C, et al: Bronchoconstrictor effect of a deep inspiration in patients with asthma. Am Rev Respir Dis 111:433–439, 1975
23. Leith DL, Bradley M: Ventilatory muscle strength and endurance training. J Appl Physiol 41:508–516, 1976
24. Denton R: Bronchial secretions in cystic fibrosis: The effects of treatment with mechanical percussion vibration. Am Rev Respir Dis 86:41–48, 1962
25. Lorin MI, Denning CR: Evaluation of postural drainage by measurement of sputum volume and consistency. Am J Phys Med 50:215–219, 1971
26. Wong JW, Keens TG, Wannamaker EM, et al: Effects of gravity in tracheal transport rates in normal subjects and in patients with cystic fibrosis. Pediatrics 60:146–152, 1977
27. Motoyama EK: Lower airway obstruction. In Mangos JA, Talamo RC (eds): Fundamental Problems of Cystic Fibrosis and Related Diseases. New York, NY, Stratton Intercontinental Medical Book Corp, 1973, pp 335–343
28. Feldman J, Traver GA, Taussig LM: Maximal expiratory flows after postural drainage. Am Rev Respir Dis 119:239–245, 1976
29. Tecklin JS, Holsclaw DS: Evaluation of bronchial drainage in patients with cystic fibrosis. Phys Ther 55:1081–1084, 1975
30. Levison H, Godfrey S: Pulmonary aspects of cystic fibrosis. In Mangos JA, Talamo RC (eds): Cystic Fibrosis: Projections into the Future. New York, NY, Stratton Intercontinental Medical Book Corp, 1976, pp 3–24
31. Schwenk WF, Desmond KJ, Beaudry PH: Effect of postural drainage in cystic fibrosis children with mild pulmonary in-

volvement, abstracted. Cystic Fibrosis Club Abstracts, 1977, p 78
32. Huber AL, Eggleston PA, Morgan J: Effect of chest physiotherapy on asthmatic children, abstracted. J Allergy Clin Immunol 53:2, 1974
33. Hartsell M, Traver G, Taussig LM: Comparison of manual percussion and vibration vs mechanical vibration alone on maximal expiratory flows, abstracted. Cystic Fibrosis Club Abstracts, 1978, p 49
34. Maxwell M, Redmond A: Comparative trial of manual and mechanical percussion techniques with gravity-assisted bronchial drainage in patients with cystic fibrosis. Arch Dis Child 54:542–544, 1979
35. Sly RM, Harper RT, Rosselot I: The effect of physical conditioning upon asthmatic children. Ann Allergy 30:86–94, 1972
36. Fitch KD, Morton AR, Blanksby BA: Effects of swimming training on children with asthma. Arch Dis Child 51:190–194, 1976
37. Hobermann M: Physical medicine and rehabilitation: Its value and limitations in progressive muscular dystrophy. Am J Phys Med 34:109–115, 1955
38. Houser CR, Johnson DM: Breathing exercises for children with pseudohypertrophic muscular dystrophy. Phys Ther 51: 751–759, 1971
39. Adams MA, Chandler LS: Effects of physical therapy program on vital capacity of patients with muscular dystrophy. Phys Ther 54:494–496, 1974
40. Siegel IM: Pulmonary problems in Duchenne muscular dystrophy: Diagnosis, prophylaxis, and treatment. Phys Ther 55: 160–162, 1975
41. Rothman JG: Effects of respiratory exercises on the vital capacity and forced expiratory volume in children with cerebral palsy. Phys Ther 58:421–425, 1978
42. Larson GL, Souhrada JF: The effect of exercise on pulmonary function in patients with cystic fibrosis, abstracted. Cystic Fibrosis Club Abstracts, 1979, p 5
43. Cropp GJA, Pullano TP, Cerny FJ, et al: Adaptation to exercise in cystic fibrosis, abstracted. Cystic Fibrosis Club Abstracts, 1979, p 32