

Massery M. Referred by the Cystic Fibrosis Clinic's PT: Treatment of Posture and Pain. *Pediatric Pulmonology, Supplement.* 2008;31:112-114.

Dramatic improvements in the diagnosis and management of children with cystic fibrosis (CF) in the USA has resulted in the mean age of survival moving from infancy in 1959, to young adulthood by 1991, and to the mid 30's, by 2007. [1] The latest data from the United Kingdom indicates that for children born in this century, a projected mean survival age of >50 years should be easily reached, even without the development of new therapies. [2] With the success of survival have come unintended consequences. Children with CF are living long enough to develop secondary impairments in their musculoskeletal system and their neuromotor postural control strategies. [3, 4] These physical impairments can lead to pain and which has been shown to have an adverse effect on the quality of the patient's survival. [5]

Adverse musculoskeletal conditions have been identified in the literature as far back as the 70's and 80's [6-10]. Yet, few studies then or now have proposed interventions other than medications. The purpose of this presentation is 1) to make the case for on-going physical therapy screenings for all patients with CF in USA clinics in order to identify and refer patients to PT with developing musculoskeletal and/or neuromotor control impairments such as postural abnormalities, spinal deformities, shoulder limitations, low back pain, other painful conditions, impaired breathing mechanics, and urinary incontinence, to name a few., and 2) to make the case that with an ever-growing adult CF population, adult therapists and doctors need to be educated in common physical ailments in order to provide a timely diagnosis and treatment of these impairments across the entire lifespan.

In the last few years, the research has overwhelmingly reported decreased bone mineral density as young as 3 to 5 year old patients with CF, creating a skeleton that is potentially more vulnerable to abnormal stresses during maturation. [11-13] Armed with historical information and current research, the CF medical community should recognize the need for regular screening patients for spinal or other postural abnormalities as young as pre-school age in order to minimize or potentially prevent secondary musculoskeletal and painful impairments.

TREATMENT: In a single case report, this author reported the results of screening and treating secondary physical impairments in a 9 year old girl. [14] The child's health and quality of life had begun to deteriorate in the prior year. She was referred to PT from her pediatric pulmonologist to determine if her physical restrictions may have been contributing to her overall decline. Skeletal alignment, mobility and control of the child's upright posture, spinal curves, shoulder function, and rib cage function were found to be impaired. Postural deficits included an excessive lumbar lordosis, excessive thoracic kyphosis,), tight rib cage, forward head, loss of shoulder range of motion and reach, excessive scapular protraction, excessive humeral head internal rotation, excessive anterior pelvic tilt, knee recurvatum, impaired postural control strategies (poor core

strategies) and impaired breathing mechanics. Based on these findings, a physical therapy program was specifically designed and implemented. The child was seen for five PT sessions (one 2 hour evaluation and four 1 hour treatments) over a 4 month period. Each PT session was videotaped for carryover into a 15-20 minute daily home exercise program. Total monetary cost of PT program was \$625 USD.

RESULTS: It is impossible to state that the child's positive health and participation results were due exclusively to physical therapy. It is more likely that they occurred as a combination of medical changes, physical changes, emotional and physical maturation changes, and/or the disease process itself. Neither the child nor her mother reported any adverse reactions to the PT program.

Results of the 4 month PT program

1. Improved postural alignment in both upright and recumbent positions (see photos below).
2. Highest pulmonary function test (PFT) results in a year.
3. No pulmonary exacerbations during the 4 month PT program.
4. A weight gain of 3 pounds in 4 months in spite of the fact that she had been losing weight over the past year.
5. An increase in participation in peer related physical activities with self reports of "more energy and endurance". Her biking endurance doubled from $\frac{3}{4}$ miles to 1- $\frac{1}{2}$ miles before fatigue stopped her participation.
6. Improvement in quality of life as reported by the patient and her mother: less fatigue noted during daily activities, increased ease of performing airway clearance maneuvers, increased functional strength, and increased sense of well being. "It's just easier to move everything!" was the child's summary comment about her participation in the PT program.

PHOTOS:



Pre-treatment (left) posture; Post-treatment (right) following 4 months of PT interventions.

IMPRESSION: In this single case, the positive outcomes for this child's health, posture, participation and quality of life changes, along with the minimal cost and time associated with the PT intervention, makes an argument for utilizing physical therapy services for regular screening and treatment of secondary musculoskeletal and neuromotor postural control impairments associated with CF. Although this was a pediatric case, the increasing number of adult survivors and the increasing reports of postural and pain in adults suggest that all patients with CF may benefit from regular PT screenings. Educating adult CF providers about common physical impairments would help to improve the continuum of care from pediatric to adult settings.

Secondary physical problems associated with CF have been described in the literature for over 30 years. With a growing adult CF population, this author believes that it is time to go beyond describing the problem; it is time to systematically treat these physical impairments in the USA. Reducing postural abnormalities and painful conditions will go a long way to improving the quality as well as the quantity of the years of survival with CF. Research is needed to determine the optimal frequency of PT screenings to achieve both a time and financial cost-effectiveness for the patient, the therapist and the insurer. To date, no such data exists.

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