

Musculoskeletal and neuromuscular interventions: a physical approach to cystic fibrosis

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Cystic fibrosis (CF) is an autosomal recessive genetic disease occurring in 1/3200 Caucasian births.¹ It affects the exocrine glands, causing multiple internal organ dysfunctions, particularly of the lungs, gastrointestinal tract, and pancreas.² In the last 45 years, dramatic advancements in the medical management of this disease have resulted in a remarkable improvement in the median age of survival. In 1959, infants with CF lived for only about 6 months—by 2003, the median age of survival was 33 years old.^{1,3,4} If the trend continues at the present rate, the median age of survival could be 50 years old by 2015, ensuring that most patients with CF would reach well into adulthood (Box 1). The bad news is that these children are now living long enough to develop secondary problems with their ‘external’ support systems: the musculoskeletal and neuromuscular systems. Although there is no direct impairment of either system according to the definition of the disease, secondary complications are becoming more frequently reported in the literature as the survival rate increases.^{5–8} The purpose of this paper is: (1) to explain how external or physical deficits can affect the function of internal structures and vice versa in the maturing child with CF; (2) to present screening procedures to identify secondary postural impairments of the spine, chest, and shoulders, and (3) to present possible interventions to minimize the impact of physical impairments on the function, morbidity, and mortality of patients with CF.

LITERATURE REVIEW: OPTIMAL TIME FOR MUSCULOSKELETAL INTERVENTIONS

Historical perspective

Children with CF were just beginning to reach skeletal maturity in significant numbers in the 1970s and 1980s, and researchers were already noting the development of secondary postural abnormalities such as thoracic kyphosis, musculoskeletal pain and arthritis.^{9–13} However, only Denton in 1981 suggested physical interventions to attempt to prevent the emerging musculoskeletal problems.¹⁰ It was not until the early 1990s that the majority of children with

Box 1 Cystic fibrosis: average survival age (Refs 1,3,4)

- 1959—6 months old
- 1979—15 years old
- 1991—23 years old
- 2003—33 years old
- 2015—50 years old?

CF were living long enough to reach skeletal maturity.⁴ Consequently, dysfunction related to the musculoskeletal and neuromuscular systems became a more pervasive problem at that time. These dysfunctions have been found to impair the patient’s ability to effectively support the function of their internal organs, particularly the lungs, thus contributing to the overall morbidity and mortality of the disease.¹⁴ Therefore, determining an optimal time to attempt medical and physical interventions to minimize or prevent these postural abnormalities would appear paramount in the rapidly maturing CF population.

The young child

No current studies reported postural abnormalities, decreased bone mineral density (BMD), or musculoskeletal pain in the child with CF who was less than 8 years old. The most recent study at the time of this writing documented the lack of a problem as old as 10 years old. Buntain *et al.* in 2004,¹⁵ investigated BMD differences across the lifespan in 153 individuals with CF compared with age matched controls and did not find any differences between children with CF and their peers in the youngest age groups, 5–10 years old even when adjusted for size and height.

The child nearing puberty

As the CF population neared puberty, numerous studies noted the emergence of musculoskeletal abnormalities. Unlike the studies of the younger children with CF, the results were mixed. Mortensen *et al.* in 2000,¹⁶ looked at children with CF aged 8–12 years and found no difference between children with CF and their matched age, weight, gender peers in terms of BMD. However, they did find a significant decreased level of vitamin D in the children with CF.¹⁶ On the other hand, in a similar age group of 9–12

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year-olds with CF, Fok *et al.*¹⁷ in 2002, found low BMDs and noted that it was most often associated with previous systemic corticosteroid use of over one month's duration. Sood *et al.* in 2001,¹⁸ added another twist, noting that the severity of the disease seemed to be a better indicator of early problems with BMD than age. Likewise, Hardin *et al.* in 2001,¹⁹ noted a decrease in BMD compared with age matched controls of children 10–12 years old. However, they noted that the apparent differences disappeared if you took the children's lean build and height into account. Hardin also posed the more perplexing question: is the decrease in BMD due to the disease process itself or is it a result of the medications and nutritional deficits? The latest article by Buntain *et al.* in 2004¹⁵ attempted to answer that question. They found a significant decrease in total body BMD even when matching for size in a slightly older grouping of 11–20 year-olds and determined a weak association of decreased BMD with nutritional status and severity of the disease: however, the study included pre- and post-pubescent children (11–20 year-olds) in a single grouping.

The adult

Unlike the mixed results during the pre-pubescent phase, by adulthood, the studies overwhelmingly note musculoskeletal limitations. Decreased BMD is now universally present in studies reported on adults with CF.^{14,15,19–22} Buntain's extensive lifespan study noted that their adolescent group had decreased BMD at 2/5 test sites but their adults had decreased BMD at 4/5 test sites even in spite of normal vitamin D levels.¹⁵ Specific spine deformities, especially a thoracic kyphosis, were more frequently noted, as were painful conditions and arthritis.^{5–7} In particular, Parasa's 1999 study noted that adults with CF have skeletal fracture rates that are twice as high as their peers (females > males) by age 16 years old, 62% of the adults had excessive kyphosis, and 94% of their adult patients reported back pain.

Optimal time for intervention

The current research clearly demonstrates significant musculoskeletal problems in the adult population with CF, and no discernible problems in the youngest age group, particularly under the age of 8 years old. The musculoskeletal problems begin to present themselves during the pre-pubescent years, beginning for some children as young as 8 years old, and are generally present by the end of puberty. Because it is easier to prevent than to correct a postural problem from both the musculoskeletal perspective and the motor planning/execution perspective, research appears to support preventative physical retraining interventions. At the 2004 North American Cystic Fibrosis

conference in St Louis, Missouri, Lannefors concurred, arguing that postural abnormalities are caused not by decreased BMD alone, but rather by the abnormal respiratory workload repetitively imposed upon a child's developing musculoskeletal frame and the neuromuscular support of that frame.²³ She made a strong argument that prevention of musculoskeletal deformities is easier than restoration, and that avoiding damage to the vertebral segments, especially wedging of the vertebrae, should be aggressively pursued. Taking all the clinical and research information currently available, the optimal time to attempt to minimize or prevent musculoskeletal deformities related to CF appears to be during the pre-pubescent years, around 8–12 years old.

In the second half of this paper, I will suggest physical therapy intervention strategies that attempt to reduce the known secondary musculoskeletal deformities. These strategies are based on normal anatomical relationships, normal developmental maturation of the musculoskeletal and neuromuscular systems, and common adaptations secondary to the presentation of a chronic childhood disease.^{24–26}

RELATIONSHIP BETWEEN RESPIRATION AND POSTURE

Understanding the relationship between the respiratory muscles and the postural muscles is critical in planning a potentially effective strategy to address the secondary musculoskeletal abnormalities related to CF. Fundamentally new research on the interaction of respiration and posture has been spearheaded by Paul Hodges in Australia. He has demonstrated that every muscle of the trunk is both a respiratory *and* a postural muscle, especially the diaphragm.^{27–33} He also demonstrated that if the physiologic support for breathing is compromised, the postural response of the trunk muscles will be reduced in order to focus on the immediate needs of respiration.²⁸ Thus, if the muscles that are needed for respiration and respiratory related activities such as coughing are over-utilized, they will directly affect the postural capabilities of those same muscles. This conflict can lead to the postural adaptation that may not be effective for optimal growth and maturation. In addition, Hodges *et al.* have recently demonstrated that painful conditions cause an ineffective neuromotor recruitment of the core trunk stabilizers, which are the same muscles that support respiration and posture.^{34–36} Parasa reported that 94% of adults with CF complain of pain.⁶ Consequently the presence of an ineffective motor recruitment strategy of the trunk muscles due to pain, may indeed predispose the individual with CF to the further development of significant musculoskeletal deformities.

SODA-POP MODEL OF POSTURAL CONTROL AND RESPIRATION

The dual function of the respiratory and postural muscles means that neither activity should be evaluated as an isolated response. External and internal forces that affect respiration will also affect postural responses and vice versa. For the maturing child with CF, this concept is critical when developing a programme to optimize the postural support muscles to help maintain the best spinal alignment possible for that child while simultaneously developing that child’s respiratory programme. The following model, *The Soda-pop Can Model of Postural Control and Respiration*, seeks to illustrate the relationship between respiration and posture.³⁷

Structurally weak, yet functionally strong

The aluminium shell of a can of soda-pop is thin, flimsy and inherently weak. Yet, when unopened, it is functionally strong. It is almost impossible to compress or deform the unopened can unless the exterior shell is punctured. However, when it is opened, it loses its ‘strength’ and is easily crushed. It is the internal pressure from the carbonation, not the aluminium exterior, which gives the can strength. The trunk of the body uses a similar concept, using its muscular contractions, to prevent its flimsy casing, the skeleton, from being ‘crushed’ by external forces such as gravity (Figure 1).

Pressure support

The trunk is composed of two chambers (thoracic and abdominal cavities) which are completely separated by the diaphragm, thus rendering each chamber capable of creating different internal pressures. The diaphragm becomes the primary pressure regulator of the two cavities. The chambers are functionally sealed at the top by the vocal folds and at the bottom by the pelvic floor muscles (Figure 2). The entire system is dynamically supported by



Figure 1 Clinical application of the concept of the ‘crushed soda-pop’ can. Melissa, age 3½ years, had a C5 complete spinal cord injury secondary to a traumatic birth experience. Notice that her skeletal frame (her ‘aluminium shell’) has collapsed upon itself. Melissa’s paralysis prevents her from generating adequate internal pressures in her thoracic and abdominal cavities that could counteract the force of gravity upon her ‘aluminium shell’, thus her skeletal shell gives way to the external pressure

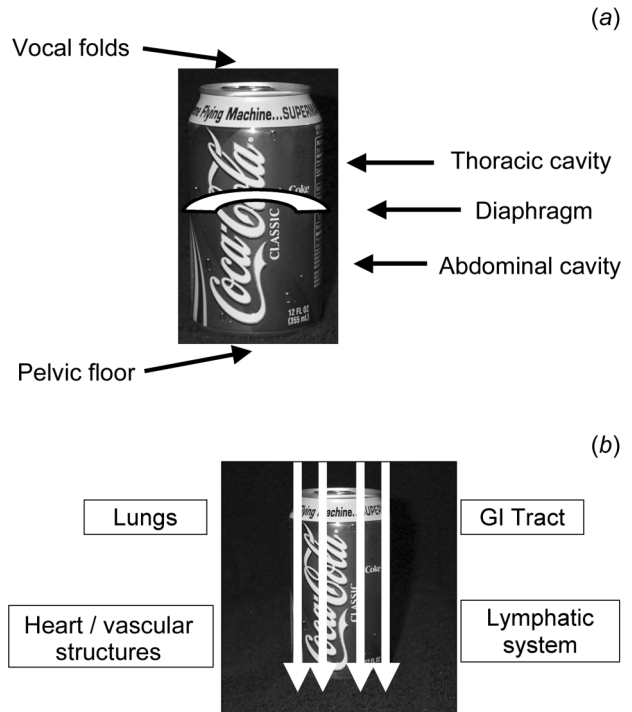


Figure 2 Soda-pop can model of postural control and respiration. (a) Shows the dual relationship of the trunk muscles for postural support and ventilation. Note that the control of the internal trunk pressures begins at the vocal folds and ends at the pelvic floor. Any breach in the system will result in an unintended release of pressure through the ‘weak’ spot. Clinically, this undesired release of internal pressure may be expressed by urinary stress incontinence, a poor balance reaction, poor breath support, weak cough, etc. (b) Illustration of the primary internal organ systems that either generate or use the internal pressures in the thoracic and abdominal cavities to improve their functional performance. For example, if the abdominal muscles are weak and cannot generate adequate abdominal pressure, the clinical result may be slow gastric emptying and/or low blood pressure. (GI, gastrointestinal)

muscles that generate, regulate and maintain internal pressures in both chambers, thus these muscles must simultaneously meet the respiratory and postural needs of that person. In other words, the trunk muscles act as one continuous functional unit, providing the core support for pressure regulation that allows the individual to multi-task, enabling ‘walking, talking, and chewing gum’ to occur simultaneously and effortlessly.³⁸⁻⁴⁵

Internal organs use of pressure

The functions of the internal organs are supported by these pressures as well, especially the lungs, heart, vascular structures, gastrointestinal system and lymphatic systems (see Figure 2). The loss of normal pressure support results in reduced haemodynamic movement of body fluids and slowed gastric motility. This is most dramatically seen in another diagnostic population: spinal cord injuries. Following a lower cervical spinal cord injury, the diaphragm

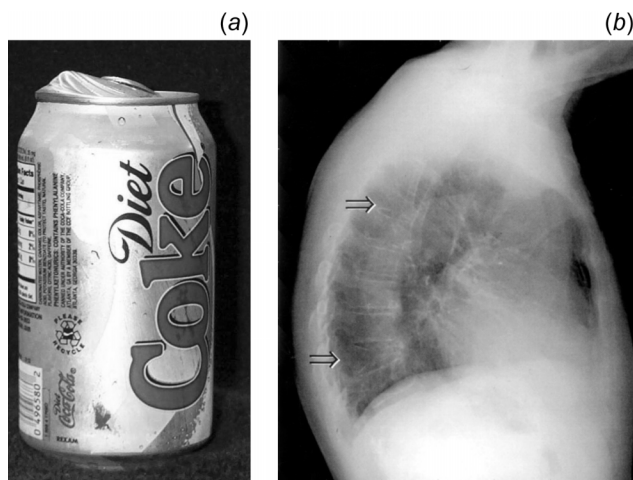


Figure 3 Pressure dysfunction related to cystic fibrosis (CF). Children with CF have the opposite problem than that of children with neurologic dysfunction. Due to their lung pathology, they exert excessive, repetitive, positive pressure on their skeleton secondary to coughing. This causes abnormal, prolonged outward pressures, like a soda-pop can that was frozen. (a) Note outward displacement of the top of the frozen can of soda-pop. (b) Clinically, this excessive pressure may be reflected as a thoracic kyphosis*, urinary stress incontinence, protracted scapulae, etc. [Ref. 14, Figure 1, with permission]

is still functioning, but the primary chestwall muscles (the intercostals) and abdominal muscles are paralysed. Without the support of all the trunk muscles the diaphragm's contractions do not result in adequate changes in pressures in the two trunk chambers. It becomes mechanically impossible for the patient with a cervical spinal cord injury to generate positive pressure in either cavity. Without any direct damage to the internal organs themselves, the patient suffers tremendous dysfunction; blood pressure becomes hypotensive, the cough becomes weak, inspiration becomes significantly impaired, and constipation develops.⁴⁶⁻⁴⁹

Pressure dysfunction related to CF

Patients with CF demonstrate the reverse problem. Due to their lung pathology, they exert repetitive positive pressure on the thin 'aluminium shell' of the skeleton secondary to coughing. This causes abnormal prolonged outward pressures in both chambers from the vocal folds to the pelvic floor. If the individual with CF cannot manage these excessive positive pressures with stronger and/or more effective muscular contractions, then the pressure will find the weakest point in the 'soda-pop can' to release some pressure much the way a soda pop can hyper-expands when frozen (Figure 3). Several types of cumulative pressure releases are noted in the literature for patients with CF especially as they reach skeletal maturity and adulthood (Box 2).^{5-8,13,17,20,23,50-55} For example, if the pelvic floor is the weak link in the pressure system, the outward pressure may be expressed as urinary stress

incontinence.^{28,29,56} In 2004 Langman reported that urinary stress incontinence was noted as occurring in 35-68% of women with CF, compared with 8-13% in the normal adult female population.⁵² Thus, the excessive internal pressures in the trunk associated with CF, can have severe mechanical repercussions anywhere where the 'soda-pop can' is weak, be it the pelvic floor, the abdomen, the spine, the chest or the vocal folds.

APPLICATION OF THEORETICAL CONCEPTS TO A SINGLE PATIENT CASE

The concepts of restoring optimal pressure support will be applied to a single patient case. 'Lauren' was chosen for this case not because she had severe CF symptoms and marked musculoskeletal issues, but rather because she represented a typical child with CF with emerging musculoskeletal issues within the 8-12-year-old range. Lauren's airway clearance issues, although a vital part of any programme for a child with CF, will not be covered in this paper.

The purpose of the physical therapy (PT) evaluation was to determine if Lauren's external support system, consisting of musculoskeletal alignment and neuromuscular control of that alignment, might be contributing to the recent decline in her functional and health status. The PT evaluation consisted of: (1) an initial screening for physical deficits; followed by (2) specific testing of the noted deficits; and finally by (3) designing a targeted intervention programme tailored to address Lauren's specific impairments (Figure 4). A summary of the findings, rather than the specific test results of Lauren's evaluation, will be presented in this paper in order to focus on the broader implications.

Box 2 Potential secondary complications resulting from repetitive excessive positive pressure in the trunk

- 1 Thoracic kyphosis and secondary mal-alignment of the shoulder girdle due to outward pressures on the thoracic spine and the prolonged use of trunk flexion posture for coughing
- 2 Chest pain secondary to excessive outward pressure on the internal chestwall
- 3 Eventual barrel shaping of the chest due to outward pressure and changes in the structure of the lungs from the disease
- 4 Low back pain secondary to outward pressure of the abdominal cavity on to the lumbar spine and muscle imbalance between the abdominal wall and deep spinal extensors
- 5 Urinary stress incontinence secondary to prolonged downward pressures on the pelvic floor, especially detrimental when performed repeatedly in a flexed trunk (coughing) posture.

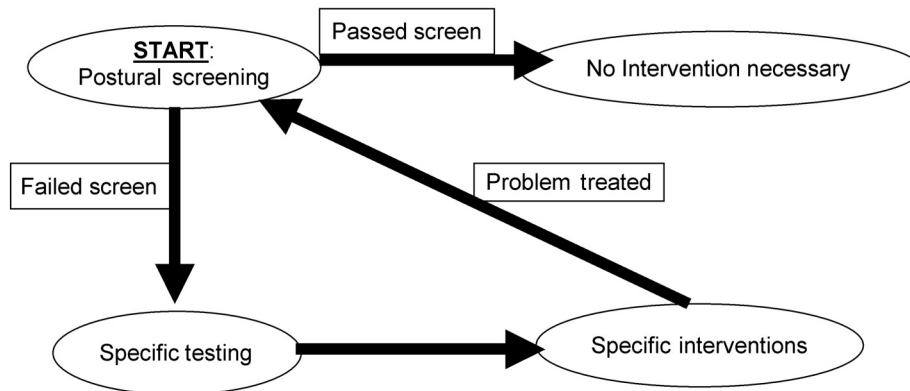


Figure 4 Physical therapy screening flow chart: postural assessment and interventions

Lauren, a 9½-year-old girl with CF, was referred to physical therapy by her pulmonologist. Lauren’s disease had been stable and mild until around age 6 or 7. It then progressed to a moderate level by age 9. She began to develop more pulmonary symptoms, including declining pulmonary function test results (PFTs) and she was having difficulty gaining weight. Lauren’s mother reported that functionally Lauren was showing limitations in endurance compared with her peers and she noted that Lauren was upset that her disease was affecting her ability to play with her friends.

Summary of Lauren’s initial physical therapy examination and evaluation

The musculoskeletal problems associated with CF tend to be more proximal than distal, thus the screening process focused on: (1) an overall postural assessment; followed by (2) a screening for spine, shoulder, scapulae and rib cage alignment impairments; and finally (3) a screening of the neuromotor control of those joints. Lauren’s musculoskeletal screening indicated impairments in all areas. Specific testing of each joint revealed limitations in spinal movement in all three planes of motion, restrictions in shoulder and scapular movement probably as a secondary result of the thoracic spine limitations, and tightness in some portions of her rib cage (Box 3). Lauren’s neuromuscular control of her trunk and shoulder joints also showed impairments that would make her movements less efficient and less supportive of needed internal pressures (Box 4).

Alignment (posture)

Lauren’s overall posture showed moderate deficits for a 9-year-old. Each individual component was only mildly impaired, but when combined, they presented moderate abnormalities in postural alignment for her age. Her

postural alignment is typical of the postural abnormalities that this author has noted with other young children with CF, most notably:

- 1 *The beginnings of exaggerated spinal curves:* Lauren showed increased lumbar lordosis, thoracic kyphosis, and cervical lordosis. Her primary trunk extension pattern was achieved with the lumbar spine, rather than a combination of the lumbar and thoracic spine. This sets up a pattern of ‘over-use’ of one spinal area (lumbar) which will predispose her to low back pain (Figure 5c–d).
- 2 *Forward head posture:* The sternocleidomastoid (SCM) muscle is a neck flexor and assists the larger trunk muscles as a trunk flexor. In CF, and with Lauren, the SCM often becomes perpendicular to the ground, rather than diagonally aligned, which has implications for potential future vocal fold dysfunction. Children with CF sometimes use their SCM muscle as a primary trunk flexor, rather than using the abdominals as the primary flexor, and thus they eventually pull the SCM forward toward the chest from over-use. This changes the alignment of the head relative to the shoulders and reduces the effectiveness of the SCM as an accessory muscle of ventilation (Figure 5c,d,f,g).
- 3 *Shoulder restrictions:* Her shoulders were forwardly rotated (protraction), elevated, internally rotated and the humerus was anteriorly tipped. If the thoracic spine is kyphotic, as it was for Lauren, the scapulae and entire shoulder complex is pushed laterally and anteriorly. This contributes to a significant loss of shoulder flexion and overall shoulder reach. In Lauren’s case, at only 9 years old, her shoulder flexion was limited to 140° instead of 180° and thus forced her to over-use her lumbar spine (lordosis) to try to increase her reach (Figure 5a–d,f,g).
- 4 *Scoliosis:* Lauren had a functional lumbar scoliosis. I have frequently noted a functional or positional scoliosis to

Box 3 Types of tests used in Lauren's musculoskeletal examination

1 *Screening Tests*

- Overview of postural alignment
- Mobility and alignment of
 - Spine
 - Rib cage
 - Shoulders/scapulae

2 *Specific Impairment Tests**

- Spine mobility tests (particularly of thoracic spine, but could include entire spine)
 - Joint end feels
 - Posterior–anterior mobility (flexion/extension mobility)
 - Lateral mobility in sitting (axial rotation mobility)
 - Functional tests
 - 'Superman' test in prone (functional use of spinal extension)
 - Measuring lateral side bending in standing (functional use of lateral side bending mobility)
- Rib cage mobility tests
 - Joint end feels
 - Grouped rib segments (upper ribs 1–3, mid ribs 4–7, lower ribs 8–10)
 - Individual rib segments
 - Functional tests
 - Lateral side bending measurements (functional opening of rib cage in conjunction with thoracic side bending)
 - Measuring chest wall excursion measurements for both quiet breathing (tidal volume) and deep breathing (vital capacity)
- Shoulders/scapulae mobility tests
 - Joint end feels
 - Glenohumeral joint
 - All scapular and clavicular joints
 - Functional tests
 - Observing and measuring range of movement of shoulder and scapulae, especially: shoulder flexion, external rotation, scapular retraction and scapular depression

3 *Designing 'targeted' interventions to meet the specific deficits noted during examination*

*Other tests may be warranted for any particular patient—this list is not inclusive.

the right (concave right lumbar) in children with CF and other chronic childhood conditions, but as of yet, I do not know why this occurs (Figure 5b).

- 5 *Chestwall shape and mobility*: The limitation in the spine and the change in normal neuromuscular breathing strategies, probably contributed to the delayed maturation of her chest shape. Her upper anterior

chest is flat and narrow and her lateral expansion was limited. She did not have any sternal abnormalities or lower rib flares (Figure 5a,c,f,g).

Muscle imbalances

In addition to the postural alignment deficits, Lauren showed muscle imbalances throughout the trunk and upper extremities, with significant potential to develop painful over-use syndromes of her lumbar and cervical spine as she matures. Due to the limitations in the mobility of her thoracic spine and shoulders, she is over-using her lumbar spine and cervical spine to extend her spine and maximize her upper extremity reach. Over time, this spinal muscle imbalance will likely cause increased skeletal mal-alignment leading to potentially painful conditions and contribute to the forces that create arthritis. Another muscle imbalance was noted in Lauren's abdominal muscles. She had a rectus diastasis (split abdominal muscles in trunk midline) and delayed activation of her abdominal external oblique abdominal muscles and the deep transverse abdominus

Box 4 Types of tests used in Lauren's neuromuscular examination

1 *Screening muscle recruitment patterns*: screening for motor planning or motor control deficits

- Breathing pattern strategies
- Arm movement strategies
- Trunk movement strategies

2 *Specific Testing*

- Impairment tests: muscle weakness, muscle imbalances, delayed activation, poor sequencing, etc.
- Functional tests: in what activities does the motor control break down?

3 *Designing targeted interventions around impairments*

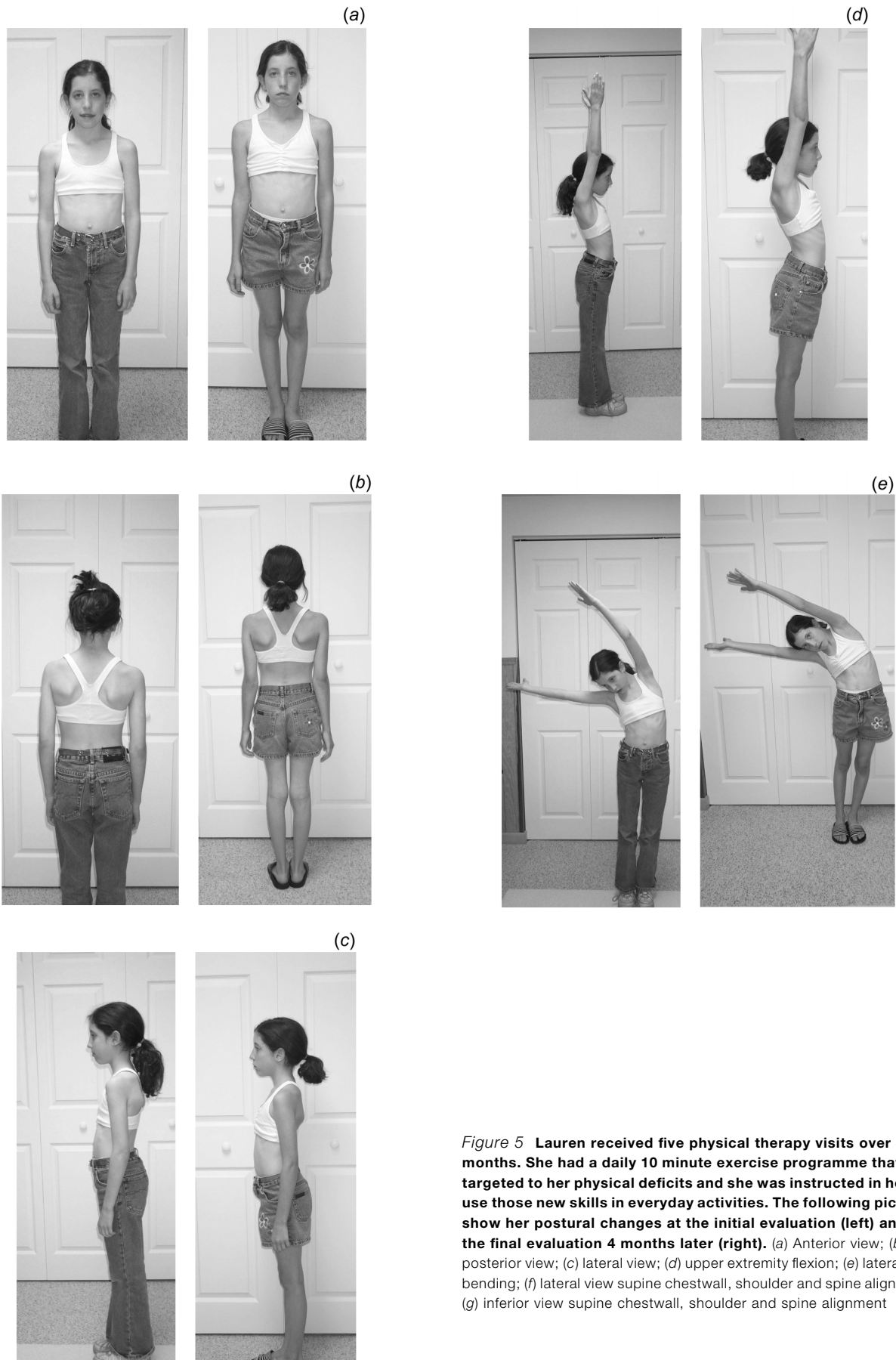


Figure 5 Lauren received five physical therapy visits over 4 months. She had a daily 10 minute exercise programme that was targeted to her physical deficits and she was instructed in how to use those new skills in everyday activities. The following pictures show her postural changes at the initial evaluation (left) and at the final evaluation 4 months later (right). (a) Anterior view; (b) posterior view; (c) lateral view; (d) upper extremity flexion; (e) lateral side bending; (f) lateral view supine chestwall, shoulder and spine alignment; (g) inferior view supine chestwall, shoulder and spine alignment

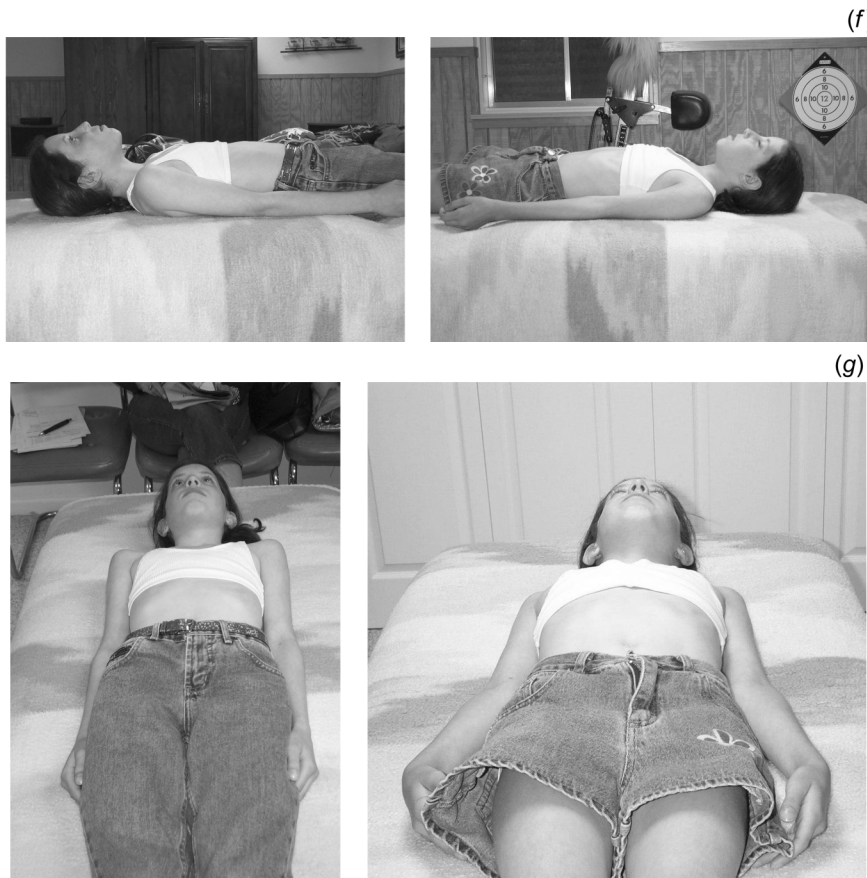


Figure 5 (continued)

muscle. This predisposes her to inadequate abdominal pressure management for lumbar spine which can result in low back pain and slow lower gastrointestinal emptying. It also causes her to lose some of the expiratory forces she needs for effective cough.

Neuromuscular control (motor strategies)

Lauren did not have weakness due to a neuromuscular ‘disease’. Her trunk muscle strengths and weakness were a result of her unique need to use these muscles to support multiple body systems: to meet the higher ventilation demands associated with her CF as well as normal developmental postural demands of childhood activities. Thus, Lauren’s problem was neuromuscular sequencing, not true weakness. Once her musculoskeletal alignment was changed through specific PT interventions, Lauren’s muscles could be retrained to (1) support that alignment and (2) support the dual function of the increased ventilation demands due to CF and the postural needs of physical activities.

For example, Lauren’s neuromuscular recruitment (motor planning) of the trunk/respiratory muscles at rest was normal, but with minimal physical stress, such as

increased resistance and or increased endurance requirements, she quickly moved to recruitment of her accessory muscles as her primary respiratory support. The muscles of the trunk always play a role in both respiratory and postural needs of the person. If Lauren’s respiratory response from her central nervous system directs her to focus on ventilation at the expense of postural control, it will leave her trunk with inadequate ability to generate and regulate positive pressure adequately to meet the needs of the increased postural demand of that activity. Functionally, Lauren demonstrated this conflict by: (1) over-recruitment of her accessory muscles rather than her diaphragm for ventilation; and (2) over-recruitment of her lumbar extensors rather than abdominals, diaphragm and internal intercostals for postural control, leaving her lumbar spine without adequate positive pressure stabilization. Over time, this neuromuscular plan will predispose her to (1) repetitive stress injury and pain in the lumbar spine and (2) poor coordination between postural and respiratory needs of the trunk, which, as her mother has already reported, will be reflected in poorer physical endurance. In other words, ‘breathing’ won and ‘postural control’ lost, resulting in decreased physical participation. The research on CF clearly supports the need for lifelong physical activities in order to

maximize long-term health, higher level of BMD and optimal postures. Thus, resolution of the conflict between postural control and respiratory demands appears critical to maximize the potential health outcomes for Lauren or other patients with CF.¹⁵

Prognosis

I felt that Lauren had a very good prognosis for reversing her current musculoskeletal impairments and improving her motor control for postural support and ventilation needs. I anticipated that her individual mild musculoskeletal impairments would respond well to specific PT interventions. I expected the bigger outcome of the PT intervention to be improved overall alignment, improved postural support, and improved breathing mechanics. I was hopeful that the ‘sum of these changes’ would be greater than the individual improvements. The long-term focus of the PT intervention was to improve the musculoskeletal alignment and motor control of these new alignments in order to improve her overall physical performance and participation, as well as improve her overall health status. Lauren had two other strong factors influencing my favourable prognosis: (1) she was very motivated to be a ‘normal kid’; and (2) she had a supportive family who would follow through with home exercises and activities that would maximize the integration of new motor strategies into Lauren’s everyday life activities.

Treatment

The control of movement (postural control) results from the sum total of the interaction of all the body systems, not from a single system. Therefore the interventions had to be linked to multiple body systems. In this case, the intent was to specifically link the musculoskeletal, the neuromuscular

and the cardiopulmonary systems in order to optimize the function of all three systems. Lauren’s physical therapy programme was targeted to meet the specific musculo-skeletal and motor planning deficits that were identified on her initial evaluation. In general her programme consisted of mobilization of tight joints followed by specific neuromuscular retraining of the muscles that should be supporting that joint. Once this was accomplished, Lauren was taught how to use the new alignment and control in everyday tasks. Because her physical system only showed minor individual impairments, she quickly moved from mobilization interventions to neuromuscular retraining and strengthening of the muscles in that new position (shortened range). Lauren then took these new skills directly to her preferred physical activities fairly independently. If she had not been capable of transferring these specific skills to function, she would have required additional PT visits. Lauren needed only five PT visits over 4 months. Other children with CF may have needed more visits to accomplish the same goals. The major component of the programme was her daily 10 minute home programme and the functional carry over of new motor skills into her everyday activities.

For a measure of overall success, Lauren was asked to identify a motor activity that she would really like to (1) do better, (2) do longer, or (3) do at all. Lauren said that was easy: she wanted to be able to cycle ³/₄ mile to the local store with her friends, drink a flavoured ice drink, and cycle home without needing to call her mother for a ride due to fatigue. At this time, Lauren said she often needed to call her mother for a ride home. It is my belief that when patients set their own goals, they will be more motivated to do the specific stretching, strengthening and neuromuscular retraining activities at home that move them toward that goal.

Outcomes

Following the 4-month PT interval, Lauren was re-tested and re-photographed. It is impossible to say that Lauren’s improvements were due exclusively to her PT programme. They are more likely due to a combination of medical, physical, emotional and physical maturation changes, and/or the disease process itself. Lauren was re-evaluated for (1) changes in specific impairments, (2) carryover of new alignments and motor strategies into functional activities, and (3) the effect of these changes on her quality of life. Specific changes in impairment measurements will not be addressed in this paper in order to focus on the broader outcomes (Figure 5, Box 5).

These changes in impairments coincided with significant changes in her functional status:

- Lauren reported that she was running with ‘more energy’. Her mother noted that it was particularly

Box 5 Lauren’s musculoskeletal and neuromuscular changes following 5 PT visits in 4 months

1	Overall postural alignment in upright and supine was dramatically improved
2	Thoracic spine movements improved in all three planes of motion
3	Chest wall mobility and shape improved
4	Scapular mobility and resting position improved
5	Glenohumeral (shoulder) limitations were not significantly changed and would require further physical therapy interventions to make necessary improvements
6	Core stabilization of the trunk for postural control and simultaneous support of demanding ventilation manoeuvres such as chronic cough, improved significantly
7	Pulmonary function tests increased for the first time in the past year
8	Gained weight for the first time in months
9	No respiratory exacerbations during this 4-month period.

evident after she began to increase her calorific intake. Which was the 'chicken' and which was the 'egg'? Did Lauren gain weight and that then supported the energy needs of increased physical activity or did Lauren eat more because her improved respiratory mechanics reduced her ventilatory workload and made eating more desirable which then led to increased physical activity?

- Lauren reported that she could 'keep up with her peers now'.
- Lauren met her physical objective: she cycled to/from the store (1½ miles total) with her friends and consumed the flavoured ice drink without fatigue. In fact, Lauren told me quite confidently: 'that she could have done it again if she wanted to'.

These changes had a direct effect on her quality of life. Lauren stated that she was happy that she could participate more readily with her friends and that she was no longer tired on a typical day. She said that her exercises have definitely helped: 'I have better coughing, better mobility (chest and trunk). It's just easier to move everything'. Lauren's mother noted similar changes in her daughter's status. She said that Lauren was: 'noticeably stronger, participated in more activities with her friends, and obviously felt better about herself'.

DISCUSSION

Lauren's outcomes should reflect typical outcomes for most children with CF. Of course, the results would vary according to the severity of the child's disease, the skill of the PT, the availability of services, the carryover by the family into the home environment and the motivation of the child. There is no 'one size fits all' programme that can be effectively dispensed to all kids with CF. It needs to be carefully designed around the deficits found in a comprehensive PT evaluation. In Lauren's case, the minimal investment in time and money were worth the outcomes according to Lauren's family, the PT and Lauren's physician. Lauren would benefit from further PT interventions to address her continuing glenohumeral joint limitations.

I did not do any 'treadmill' or other contrived endurance training programmes to achieve these results. It is my belief that the typical child, who enjoys socializing with their peers, wants to participate in recreational or sporting activities. Therefore if you can ease their respiratory and postural workload through specific, targeted interventions, this should lead to an increase in desire and participation of peer-related activities, like Lauren's cycling. When these children are sick, they may need to be motivated to move through contrived activities such as treadmill training or stationary bike riding, but the preferred method to achieve endurance is by helping the child to participate in life, not in exercise.

Box 6 Potential long-term benefits of musculoskeletal and neuromuscular programmes for children with cystic fibrosis (CF)

- *Improved mechanical support*
 - Optimal postural development
 - Optimal development of postural control/breathing
 - Decreased overall work of breathing (less fatigue, greater endurance, more participation)
 - Prevention or reduction in the adult onset of stress urinary incontinence
 - Maintenance of higher bone mineral density through adulthood because the improved mechanical support promoted increased engagement in physical activity
- *Improved breathing mechanics*
 - Improved ventilation volumes and efficiency of inhalation and exhalation manoeuvres
 - Improved mechanical support for airway clearance
- *Prevention of painful joint conditions due to mal-alignment or misuse*
 - Possibly reduced incidence of CF adult onset arthritis
 - Preventing loss of overall mobility and participation due to pain
- *Development of fewer pulmonary complications*
 - More postural support for breathing
 - More efficient breath support
- *Functional outcomes*
 - Decreased morbidity
 - Less hospitalizations
 - Less missed days of school or work
- *Quality of life issues*
 - Increased participation in life
 - Improved sense of well-being.

Potential long term benefits from a PT programme

A PT programme that focuses on optimizing the maturing musculoskeletal and neuromuscular systems of a child with CF could play a significant role in an improved long-term outcome of the child's physical and medical status (Box 6). Improving mechanical support for movement and breathing should lead to: (1) a reduction, or prevention of, secondary painful joint conditions; (2) fewer pulmonary complications that result from poor mechanics; (3) the prevention or reduction in the adult onset of stress urinary incontinence due to poor mechanical pressure support; and (4) maintenance of higher BMD through to adulthood because improvement in the child's postural support encouraged increased levels of physical activity. The ultimate goal of the intervention would be to positively influence the long term functional outcomes: reducing morbidity, reducing hospitalizations, reducing the number of missed days of school or work, and improving the overall quality of life and sense of well being for children growing up with CF.

SUGGESTIONS FOR FUTURE RESEARCH

Treatment suggestions in the current literature focused primarily on medical, nutritional or general physical activities, but not on specific musculoskeletal and/or

neuromuscular treatments. In the short term, I see a need to publish studies with short-term outcomes and/or case reports on specific musculoskeletal and neuromuscular interventions. In the long term, I see a need to establish longitudinal studies that assess the adult outcomes and efficacies of PT interventions that are begun in childhood. Another consideration for research is to look at the timing of the development of physical deficits. Although the literature points us towards initiating PT interventions around 8–12 years old when physical deficits are just emerging, researchers should consider whether the onset of physical impairments is inevitable or just probable. Could these deficits be delayed or even prevented if a PT programme was started earlier? If the infant or toddler with CF was monitored by PT for the development of effective postural control strategies, optimal postural alignment, adequate muscle strength and muscle balance, and effective coordination of their multi-system needs (especially musculoskeletal, neuromuscular and cardiopulmonary systems), could the emergence of postural deficits be prevented altogether?

SUMMARY

Children with CF are living longer than ever before, and thus issues pertaining to quality of life rather than just longevity of life need to be addressed by the entire healthcare team. This article addressed the issues pertaining to the external support of the dysfunctional internal organs: the secondary musculoskeletal (postural) and neuromuscular control deficits that occur to the maturing child with CF. The research pointed towards starting PT interventions for these deficits during the pre-pubescent phase when postural deficits were just emerging, but a suggestion was also made to explore whether these deficits can be even more effectively monitored and treated at an earlier age. The dual relationship between the muscles used to meet the increased respiratory demands of CF and the normal postural demands of physical activities was described through a model based on a soda-pop can and pressure support.

A pre-pubescent child with a typical progression of CF was presented as a case report to illustrate how a PT programme that was focused on postural deficits could be implemented and what type of outcomes might be possible. The child made significant changes within a relatively short time frame of 4 months, proposing that the musculoskeletal and neuromuscular systems may play a significant role in the medical and physical long-term outcomes of CF. For that reason, the physical as well as medical needs of the patient should be incorporated into a comprehensive multi-system approach to the disease across the lifespan.

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