Muscle Length and Joint Range of Motion in Children with Cystic Fibrosis Compared to Children Developing Typically

Allison Mandrusiak, Donnae Giraud, Julie MacDonald, Christine Wilson, Pauline Watter

ABSTRACT

Purpose: To explore range of motion in children with cystic fibrosis (CF) compared to children developing typically. With the increasing longevity of people with CF, musculoskeletal concerns are becoming more prevalent and should be identified and considered in management plans. Reduced range of motion (ROM) in older people with CF has been demonstrated, and the age at which these changes begin to occur must be explored.

Methods: Participants were 38 children with CF (aged 7–14 years) and 38 children developing typically, matched for age and gender. Muscle length and joint ROM measurements were compared, using analysis of variance, in the thoracolumbar region (extension, flexion, lateral flexion), upper limb (pectoralis minor, pectoralis major, shoulder horizontal abduction), and lower limb (hip internal and external rotation, hamstrings, gastrocnemius-soleus).

Results: Children with CF had significantly shorter pectoralis minor, pectoralis major, and gastrocnemius-soleus muscles and significantly greater range of shoulder horizontal abduction than matched controls.

Conclusion: Altered ROM was identified in children with CF, strengthening the rationale to support proactive physiotherapy management of the musculoskeletal system, aiming to optimize function across the increasing lifespan. Evaluation and management of ROM and its impact on activity and participation are important areas for future research and clinical practice.

Key Words: children, cystic fibrosis, ICF, joint range of motion, muscle length

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RÉSUMÉ

Objectif : Explorer l'amplitude de mouvement des enfants atteints de fibrose kystique (FK) comparativement à celle des enfants se développant normalement. Avec la longévité croissante des personnes atteintes de FK, les problèmes musculo-squelettiques deviennent plus fréquents et devraient être identifiés et pris en compte dans le plan de gestion de la maladie. Il a été démontré que les enfants atteints de FK qui sont plus âgés ont une amplitude de mouvement réduite, à l'âge auquel de tels changements commencent à survenir devrait être étudié.

Méthode : Les participants regroupaient 38 enfants atteints de FK (de 7 à 14 ans) et 38 enfants ayant un développement normal, jumelés en fonction de leur âge et de leur sexe. Les mesures de la longueur des muscles et de l'amplitude de mouvement des articulations ont été comparées, à l'aide d'une analyse de la variance, pour la région thoracolumbaire (extension, flexion, flexion latérale), membres supérieurs (petit pectoral, grand pectoral, abduction horizontale de l'épaule) et membres inférieurs (rotation interne et externe de la hanche, ischio-jambiers, gastrocnémiens soléaires).

Résultats : Les petits pectoraux, les grands pectoraux et les gastrocnémiens soléaires sont considérablement plus courts chez les enfants atteints de FK et ceux-ci affichaient une amplitude de mouvement beaucoup plus importante à l'abduction horizontale de l'épaule que le groupe de contrôle qui leur était associé.

Conclusion : Une amplitude de mouvement réduite a été observée chez les enfants atteints de FK, ce qui vient appuyer l'argument pour une gestion proactive du système musculo-squelettique par la physiothérapie dans le but d'optimiser les fonctions pour toute la durée de vie, laquelle est en croissance. L'évaluation et la gestion de l'amplitude des mouvements et ses effets sur l'activité et la participation sont des sujets importants pour la recherche future et la pratique clinique.

Mots clés : amplitude de mouvement des articulations, enfants, fibrose kystique, ICF, longueur musculaire

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INTRODUCTION

Range of motion (ROM) is dependent on joint range and muscle length, reflecting the properties of bony structure, muscle, and connective tissue.¹ The impact of altered range on function should be considered using the frame of reference provided by the International Classification of Functioning, Disability and Health (ICF).² A discussion of the multidirectional nature of the relationships between the domains of the ICF model in young people with cystic fibrosis (CF) has been presented by Mandrusiak et al.³ In the context of the current study, not only may impairment in range of motion at the body structure and function level lead to activity limitations and participation restrictions, but reduced participation in physical activity may in turn limit functional capacity, increasing impairments in body structures and functions. Thus, it is important to identify and address impairments in ROM to optimize overall functioning.

As stated in Gibson et al.⁴ CF is one of the most common life-shortening genetic diseases affecting Caucasian populations and, less commonly, other ethnic groups. Because of the seriousness of the respiratory manifestations of CF, musculoskeletal issues have previously been somewhat overlooked.⁵ The musculoskeletal system is not directly implicated in this condition; because their lifespan is increasing, however, children with CF are increasingly susceptible to secondary effects.⁵⁻⁷ Research into musculoskeletal manifestations affecting children with CF has reported mainly on their reduced bone-mineral density and muscle strength. Although it has been suspected that children with CF have altered ROM compared with children developing typically,^{7,8} this has yet to be confirmed objectively. Such information may provide direction for the timing and nature of intervention.

Reduced ROM of the shoulder, chest, and trunk has been reported in older people with CF⁸ and is associated with the high prevalence of back pain in this population.^{7,9,10} Rose et al.⁸ explained that the habitual flexed posture caused by altered breathing mechanics is reinforced by chronic coughing and results in further tightening of muscles and restriction of movement of the shoulder, chest, and trunk. It is also suggested that the dyspnea associated with CF may make this population less active, leading to more time spent sitting in flexed postures that promote reduced extension of the thorax and tightening of muscles in this area.¹⁰ Importantly, reduced ROM of the chest may further impair respiratory function and may make effective airway-clearance therapy difficult.⁹ This suggests that musculoskeletal concerns for people with CF across all ages should be identified and managed, using a holistic approach.

It is not clear when the changes in ROM of the shoulder, chest, and trunk observed in adults with CF begin to occur, and thus the optimal age at which intervention to

maintain range is warranted is also unclear. The ROM in the lower limbs should also be explored, as this may affect activity and participation. In addition, differences in ROM in children with CF based on contextual factors, such as in-patient and outpatient status, are not reported in the literature. Findings of our study of children aged 7 to 14 years may guide further research into the type and timing of strategies to address ROM concerns for children with CF, with the overall goal of better understanding the multidirectional dynamic relationships within and between the domains of the ICF model.²

The main purpose of this study was to compare ROM in children with CF to ROM in children developing typically, using standard measures. It was hypothesized that there would be differences between the CF and typical groups, with the CF group displaying reduced ROM, and between in-patient and outpatient CF groups, with the in-patient group displaying reduced ROM.

METHODS

Children with CF ($n = 38$) were recruited as part of a separate study at the Royal Children's Hospital and Health Service District (RCH), which used a sample of consecutively admitted in-patients (hospitalized due to exacerbation of respiratory symptoms) and outpatients. Subsequently, children developing typically were recruited from local suburban schools and matched for age (± 6 months) and gender to children in the CF group. With 38 participants per group, we had a power of 0.85 to detect an effect of >0.70 , significant at the two-tailed alpha level of 0.05.¹¹ Recruitment was open to children aged 7 years, 0 months, to 13 years, 11 months. Children with CF were excluded if they had any additional diagnoses (e.g., bowel obstruction), unstable vital signs, or existing musculoskeletal injury. Children were excluded from the control group if they had any diagnosed medical condition or existing musculoskeletal injury. Ethical clearance was obtained from the Medical Research Ethics Committee of the University of Queensland and from the Ethics Committee of the RCH. Informed assent was obtained from all participants, and informed consent from their parents/guardians, prior to their participation.

Age (years), height (metres), weight (kilograms), and body mass index (BMI) (kg/m^2) were recorded for each participant, as well as percentage of predicted forced expiratory volume in 1 second ($\text{FEV}_1\%$) for those with CF. Standard measures were used to assess ROM^{1,10,12} of the trunk and limbs using either a measuring tape (scored in centimetres) or a universal goniometer (scored in degrees). Each measure assessed range of either a single structure (e.g., pectoralis minor muscle) or multiple structures (e.g., thoracolumbar extension). For each measure, a practice trial was given to familiarize the child with the procedure, and the second trial was recorded. As part of the larger RCH study, children with

Table 1 Descriptive Details: Comparison of Means between Children with Cystic Fibrosis (CF) and Age- and Gender-Matched Controls

	CF (n = 38) mean (SD)	Matched controls (n = 38) mean (SD)	F (df = 1, 75)	p
Age (years)	10.2 (2.0)	10.3 (2.0)	0.002	0.97
Height (m)	1.41 (0.12)	1.42 (0.11)	0.20	0.65
Weight (kg)	33.97 (8.15)	35.28 (8.67)	0.46	0.50
BMI (kg/m ²)	16.9 (2.0)	17.2 (2.3)	0.57	0.45

BMI = body mass index (kg/m²)

CF in the in-patient group were assessed within 48 hours of admission; those in the outpatient group were assessed on the day of their CF clinic appointment. Children in the control group for the present study were assessed in a private room at their school.

Thoracolumbar extension and flexion were measured with a tape measure over the spinous processes of C7 to S1. Distance between the neutral and end position was recorded¹² and calculated as a percentage of the neutral position, as suggested by Rose et al.⁹ To measure thoracolumbar lateral flexion, the participant flexed to one side, with the pelvis stabilized and shoulders against a wall; the distance from the tip of the middle finger to the floor was measured at the start and end of the movement, and the difference was recorded.¹²

Range of horizontal shoulder abduction was assessed using a goniometer, with the participant sitting with the shoulder supported in 90° abduction and neutral rotation and the elbow flexed to 90°.¹² The participant's thorax was stabilized as the arm moved into horizontal abduction. Length of the pectoralis minor was assessed with the participant in supine with the arms by the sides. The shoulders were externally rotated with the forearms supinated and lumbar spine flat against the surface. The participant relaxed his or her shoulders, and the distance from the posterior border of the acromion to the support surface was measured using a measuring tape.¹ Length of the pectoralis major was assessed with the participant in supine with the hands clasped together behind the head, with cervical and lumbar spines in neutral; the participant relaxed his or her shoulders, and the distance between the olecranon of the humerus and the support surface was measured.¹

Hip-joint internal and external rotation were assessed using a goniometer, with the participant in supine with the hip joint in the anatomical position and the knee flexed to 90° over the edge of a plinth. The opposite hip and knee were flexed with the foot flat on the table; the hip was passively moved through internal and external rotation, with the distal end of the thigh stabilized and pelvic movement prevented.¹² Hamstring length was assessed using a goniometer via a passive straight leg raise; the knee joint was held in extension by the examiner, with the ankle joint relaxed and pelvis in the neutral

position.¹² The weight-bearing dorsiflexion test, as outlined by Bennell et al.,¹³ was used to assess length of the tendo-Achilles, a reflection of gastrocnemius-soleus muscle length.¹⁴ The participant placed the test foot perpendicular to a wall and moved the knee forward; the foot was progressively moved away from the wall until the maximum dorsiflexion range was reached without lifting the heel. The distance to the wall from the tip of the big toe was then measured with a tape measure.

Although this was not a reliability study per se, the two examiners used standard protocols^{1,10,12} and practised together to maximize accuracy with respect to identification of landmarks, measurement procedure, and readings. As a further effort to maintain consistency and avoid experimental bias, one examined the CF group and one the typical group. Importantly, most of the measures used in this study have established strong interrater reliability, as reported by Mayerson et al.,¹⁵ who investigated 22 joint positions with goniometry (yielding a coefficient of $r = 0.97$); reliability of the weight-bearing dorsiflexion test was reported by Bennell et al.¹³ at $r = 0.99$.

Statistical analyses were performed using SPSS software version 14.0 (SPSS Inc., Chicago, IL). The distribution of scores was found to be normal. This comparative-design study examined differences between means of children in the CF group and those in the control group, and between children with CF in the in-patient and outpatient groups, using analysis of variance (ANOVA) with the threshold for significance set at $p \leq 0.05$.

RESULTS

Thirty-eight typically developing children were matched for age and gender to a data set of 38 children with CF (in-patient $n = 17$; outpatient $n = 21$). Mean scores for descriptive measures of all children are presented in Table 1. No significant differences were found between the groups for these measures (p range: 0.45–0.97).

Children with CF in the in-patient group were significantly older than children in the outpatient group (mean age (years) 11.0 ± 1.9 vs. 9.6 ± 1.9 , $p = 0.03$) and had significantly lower FEV₁% (mean = 58.9 ± 16.0 vs.

Table 2 Range of Motion: Comparison of Means between Children with Cystic Fibrosis (CF) and Matched Controls (and 95% Confidence Intervals of the Difference)

	CF (n = 38) mean (SD)	Matched controls (n = 38) mean (SD)	F (df = 1,75)	p	95% CI of the difference
Th/L Flx (%)	16.93 (5.4)	17.98 (6.55)	0.58	0.45	
Th/L Ext (%)	12.95 (6.80)	13.85 (5.23)	0.42	0.52	
Th/L LF (%)	17.73 (2.60)	18.87 (2.30)	3.49	0.13	
Pec Minor (cm)	5.71 (2.20)	2.58 (0.90)	65.93	<0.001	2.29 to 3.96
Pec Major (cm)	4.74 (3.42)	0.97 (0.49)	47.88	<0.001	2.63 to 4.89
Shoulder Ab (°)	54.37 (12.52)	45.18 (7.57)	14.81	<0.001	4.40 to 13.86
Hip IR (°)	24.82 (7.52)	22.21 (5.43)	22.33	0.04	0.17 to 6.36
Hip ER (°)	24.34 (6.76)	22.32 (4.78)	3.00	0.09	
SLR (°)	55.97 (20.04)	59.53 (14.08)	0.80	0.37	
WBDF (cm)	6.25 (2.62)	8.72 (2.47)	17.97	<0.001	-3.64 to -1.31

Th/L = thoracolumbar; Flx = flexion; Ext = extension; LF = lateral flexion; Pec minor = pectoralis minor; Ab = abduction; IR = internal rotation; ER = external rotation; SLR = straight leg raise; WBDF = weight-bearing dorsiflexion

78.9 ± 13.7 , $p < 0.001$). There was no difference in height, weight, or BMI, and all comparisons of ROM between in-patients and outpatients were non-significant ($F(1,37)$ range: 0–1.5, p range: 0.23–0.99), so the scores for in-patient and outpatient children with CF have been considered as one group.

For ROM measures, the mean scores, differences between means, and 95% confidence intervals (CIs) of the difference for the CF group and control group are presented in Table 2. Significant differences between the groups were found for pectoralis minor, pectoralis major, and gastrocnemius-soleus muscles; shoulder horizontal abduction; and hip internal rotation (p range: <0.001–0.04). The directions of these differences are shown in the table. Comparisons of ROM for all thoracolumbar movements, hip external rotation, and hamstrings were non-significant (p range: 0.07–0.69). For bilateral measures, only the right side is presented, as there was no significant difference between sides.

DISCUSSION

The main findings of this study were that significant differences in ROM were identified between children with CF and children developing typically, but not between in-patient and outpatient children with CF. Consistent with our hypotheses, children with CF displayed significantly reduced range for some muscles (pectoralis minor, pectoralis major, and gastrocnemius-soleus). However, in contrast to our hypotheses, children with CF displayed significantly greater range of shoulder horizontal abduction, and no significant differences for other ranges (thoracolumbar, hip rotation, and hamstrings).

Our study found that children with CF had reduced length of the pectoralis minor and major muscles, complementing the findings of Rose et al.,⁹ who explained the relationship between habitual shoulder protraction

and flexed posture and back pain in people with CF. Our findings suggest that physiotherapists should aim to maintain chest mobility and optimize respiratory function and airway clearance across the lifespan. Current approaches may include stretching of the pectoral muscles and postural education, even for this younger group, as part of holistic management. Employing the framework of the ICF model, which suggests that changes in one domain may be associated with changes in other domains, attention should also be directed toward maintaining physical activity and participation, in an attempt to promote optimal status within the body structures and functions domain.

Similarly, children with CF exhibited reduced range in the gastrocnemius-soleus muscle group, and although it is yet to be determined whether this difference is clinically meaningful, this is an interesting area for further consideration. These muscles are prone to tightness,¹⁶ and shortening of these muscles may be associated with a disadvantaged lever arm and reduced movement efficiency. Adequate range of ankle dorsiflexion is necessary for normal functional activities such as running¹³ and walking. According to Nixon et al.,¹⁷ young people with CF have reduced levels of physical activity, particularly of vigorous intensity. Although physical activity in this population is particularly influenced by respiratory function, it seems appropriate to identify and address impairments at the body structure and function level, such as dorsiflexion range, in order to facilitate activity and participation across the lifespan.

Interestingly, our study revealed increased range of shoulder horizontal abduction in children with CF. It is possible that this is a subconscious strategy to “open” the upper chest wall to achieve increased chest volumes via accessory muscle recruitment. Massery⁷ documented the secondary malalignment of the shoulder girdle in this population as a result of altered respiratory mechanics

and posture, and suggested that this contributes to loss of shoulder flexion range. In relation to our study, it may be that compensatory increases in length of the shoulder horizontal adductor muscles develop to preserve functional range of abduction in the presence of reduced length of the pectoral muscles. However, a study by Rose et al.⁹ found significantly reduced horizontal abduction range in a group of older people with CF (mean age 23.4 years) compared with age- and gender-matched controls; it may be that with increasing age, shoulder horizontal abduction also becomes restricted, further affecting the already restricted mobility of the chest and shoulders, as described by Rose et al.⁹ Although this highlights important areas for future research, an alternative explanation is the inherently low interrater reliability established for goniometry of shoulder horizontal abduction ($r = 0.30$).¹⁸

Although in our study of children aged between 7 and 14 years there were no significant differences in thoracolumbar ROM between children with CF and age- and gender-matched controls, the older group studied by Rose et al.⁹ showed significantly reduced flexion and extension range compared with matched controls. This suggests that these changes may occur with increased age, highlighting the need to monitor musculoskeletal concerns and incorporate proactive approaches to their management throughout the life stages.

It remains important to acknowledge that ROM changes were already apparent in some thoracolumbar and lower leg muscles in children with CF aged between 7 and 14 years; this finding suggests a need to address these issues early within the overall management of this population. However, there is limited evidence to support the efficacy of stretching in some populations,¹⁹ suggesting directions for future studies.

Comparison of in-patient and outpatient children with CF revealed no significant differences in ROM, indicating that the differences found between children with CF and children developing typically are not associated with an acute exacerbation of respiratory symptoms or with the differences in age or FEV₁% noted between the in-patient and outpatient groups. It is likely that the observed differences between children with CF and children developing typically have developed slowly in relation to subtle changes in body structure and function. These findings suggest that preventive as well as treatment plans may be indicated during both chronic and acute stages of CF to address these changes.

Lannefors⁸ suggests that a proactive approach to potential changes in ROM is easier than remediating actual problems in later life. Further, Massery⁷ suggests that the optimal time to prevent or minimize musculoskeletal concerns related to CF appears to be during the prepubescent period (8–12 years of age), and findings from the present study support this view, as these impairments are already present by this time. It is recom-

mended that simple exercises to maintain muscle length and joint mobility be included in the daily physiotherapy programme from an early age.^{7,8,20} This programme is part of a holistic management approach that includes desirable levels of daily habitual activity in conjunction with attention to factors such as diet and airway-clearance techniques. A single-patient case study⁷ of a 9.5-year-old girl showed that identification and targeted intervention of postural impairments had beneficial outcomes not only at the body structure and function level but in terms of activity, participation, and quality of life. The child in this case was chosen to “represent a typical child with CF with emerging musculoskeletal issues within the 8- to 12-year-old range”^{7(p.58)}; the author therefore believes that the outcomes may reflect typical outcomes for children with CF.⁷ Results from our study provide objective support for the need for this approach. Although larger studies are indicated to examine the effectiveness of such interventions, it seems appropriate to suggest that when included in the management plan, they may reduce the risk of musculoskeletal manifestations with increasing age and any potential impact these changes may have.

It may be important for physiotherapists to identify and address ROM concerns for children with CF, as this may optimize their participation in physical activity, and vice versa. Massery⁷ proposes that strategies to improve postural development and thus enhance respiratory mechanics may encourage increased levels of physical activity. This is an especially important goal, based on the established knowledge that better fitness has far-reaching benefits for persons with CF, including increased survival rates²¹ and improved quality of life.²² Results of the current study provide important directions for physiotherapists working with children with CF, strengthening the evidence base for including ROM in the assessment of this population with the aim of optimizing their longer-term outcomes.

LIMITATIONS

Although the measures selected for this study are those commonly used in physiotherapy practice and are readily available in resources such as textbooks, in some cases other measures may be more appropriate, such as the popliteal angle method in place of the traditional straight leg raise test as a measure of hamstring length. The limited interrater reliability of the shoulder horizontal abduction measure is acknowledged, and it is important to establish inter- and intrarater reliability in future studies.

The age range selected may be considered narrow; however, it was chosen to reflect a time after a transition period of rapid maturation.²³ It would be useful to explore the same measures in children younger than 7 years and in adolescents approaching adulthood, to

provide a broader view of the changes in ROM across these stages.

It is difficult to align best evidence (e.g., emerging evidence on the limited efficacy of stretching)¹⁸ with current practice (whereby maintaining flexibility is considered to be desirable). Future studies should clarify the best approach for intervention for specific issues such as ROM across all domains of the ICF model.²

CONCLUSION

This study of children with CF between 7 and 14 years of age identified a number of significant differences in ROM compared with that of typically developing children. Reduced length of pectoralis minor, pectoralis major, and gastrocnemius-soleus muscles was found for children with CF. We found no differences between inpatients and outpatients, which suggests a gradual onset of this impairment for children in this age group; the identified changes in ROM do not appear to be associated with acute exacerbation of pulmonary symptoms. Further research is warranted to investigate the complex relationship between ROM and other parameters outlined by the ICF model² and explored theoretically by Mandrusiak et al.³ and to assess the efficacy of interventions targeting different domains of the ICF for people with CF across the lifespan.

KEY MESSAGES

What Is Already Known on This Subject

It is agreed that in adolescents and adults with CF, respiratory function is compromised by musculoskeletal changes, particularly with respect to upper limb and thoracic musculature. However, it is unclear when these changes become apparent and how they may best be addressed.

What This Study Adds

This study adds to the evidence by showing that a group of children with CF, as young as 7 years, already display differences in range of motion in the upper limb (reduced pectoralis minor and major length, increased shoulder horizontal abduction range) and in the lower limb (reduced weight-bearing dorsiflexion range). Further, no differences were seen between in-patient and outpatient groups. This study points to the usefulness of the multidirectional nature of the ICF model to aid our understanding of the links between and within domains, but the findings do not indicate whether intervention should be at the impairment or the participation level, indicating a need for further study.

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