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POSTURAL DEFECTS IN CHILDREN WITH CYSTIC FIBROSIS – PRELIMINARY REPORT

WADY POSTAWY U DZIECI Z MUKOWISCYDOZĄ – DONIESIENIE WSTĘPNE

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Abstract
Postural disturbances may cause the worsening of cardiopulmonary function in cystic fibrosis children and adolescents, as well as adversely affect their somatic and psychological development.

The aim: Evaluation of postural defects in the population of cystic fibrosis children and adolescents.

Material and methods: Physical orthopedic examination of 41 cystic fibrosis patients (20 girls, 21 boys), aged 2-17 years (mean 8.1). The patients were divided into age groups: I under 5 y.o – 8 patients, II 5-9 y.o. – 6 patients, III 9-13 y.o. – 11 patients, IV 13 y.o. and older – 16 patients. Postural disorders were divided into two groups. The first group included defects with a possible influence on the respiratory function of a cystic fibrosis patient: scoliosis, spine defects and chest deformities. The second group included lower extremities defects without influence on the function of the respiratory system.

Results: Postural as well as chest disorders were observed in 36 (87.8%) of the patients who were examined. Disorders with possible influence on respiratory system function, were observed in 13 (31.7%) of the 41 patients. Increased thoracic kyphosis (round back) was most frequent. In the group of patients aged 5 years and younger neither spinal nor chest abnormalities were observed. In the group of patients aged 5-9 years 1 (16.6%) case of congenital scoliosis and 1 (16.6%) case of plane back were found. In the group of children aged 9-13 years increased thoracic kyphosis was observed in 2 (18.1%) cases, plane back was observed in 2 (18.1%) children as well, while in 1 (9.0%) case increased lumbar lordosis was diagnosed. Barrell chest was observed in 2 (18.1%) cases. In the group of adolescents aged 13 years and more, 4 (25.0%) cases of scoliosis, 16 (100%) cases of increased thoracic kyphosis and 6 (37.5%) cases of barrel chest were observed. Because of the different methods of examination among posture studies of the healthy population, obtaining a control population is difficult.

Conclusions: The number of postural defects in children with cystic fibrosis increases with age, and is becoming a significant health problem. It seems that there is a need of a deeper study on a greater population.

Key words: cystic fibrosis, posture, poor posture, postural disturbances, scoliosis

Streszczenie
Wady postawy mogą prowadzić do pogorszenia funkcji układu oddechowego oraz krążenia u dzieci i młodzieży z mukowiscydozą, a także zaburzać przebieg rozwoju somatycznego i psychicznego.

Cel: Ocena wad postawy w populacji dzieci i młodzieży z mukowiscydozą.

Materiał i metody: Badanie przedmiotowe ortopedyczne 41 pacjentów z mukowiscydozą (20 dziewcząt, 21 chłopców) w wieku 2-17 lat (średnio 8.1). Pacjentów podzielono na następujące grupy wiekowe: I poniżej 5 r.ż. – 8 pacjentów, II 5-9 r.ż. – 6 pacjentów, III 9-13 r.ż. – 11 pacjentów, IV 13 r.ż. i starsi – 16 pacjentów.
INTRODUCTION

Human posture as well as postural defects are very difficult to define. Unfortunately, there is a lack of one precise definition of the problem. There are many definitions of posture and postural defects, including human morphological, physiological, dynamic and psychological status. Many authors agree that posture is “a motor habit shaped on a specified morphological and functional background; manifestation of physiological and psychological status of an individual; an indicator of the mechanical efficacy of the kinetic sense, as well as muscular balance and neuromuscular coordination” [1]. A defect of posture is any deviation from the typical posture among the population with the same distribution of age, gender, psychophysiological and demographic features. According to this definition there is a need of a reference population for the assessment of posture. Typical assessment of posture includes observation of the shoulders, back, chest, pelvis, lower extremities and feet. Observation may be extended with the use of various devices – a goniometer, cameras and computer assisted devices.

Human posture changes during a person's lifetime. Childhood and adolescence are the periods of most rapid change in human posture. Recent changes in lifestyle due to the development of civilization, nutrition and activity are connected with an increased number of postural defects in developed countries [1-3].

Scoliosis is not only a postural defect. It is a severe spinal disorder that occurs in children and adolescents [4]. The definition of scoliosis is: “side-to-side curvature of the spine over 10 degrees measured using Cobb method, combined with a rotation of the vertebrae” [4-5]. That means that scoliosis is not only a lateral curvature of the spine, but a three-plane deformation. A decrease of thoracic kyphosis is also characteristic.

Scoliosis is a disease usually classified as non-idiopathic or idiopathic. Non-idiopathic scoliosis is caused by malformation of the spinal column, or neuromuscular diseases, like cerebral palsy or myelomenigocele, or mesenchymal abnormalities, with insufficiency of passive stabilizers of the vertebral column, for example ligamentous laxity in the Marfan syndrome. Non-idiopathic scoliosis is less common.

Idiopathic scoliosis is more frequent. Diagnosis of idiopathic scoliosis is based on excluding non-idiopathic scoliosis. Idiopathic scoliosis is also classified with reference to the patient's age. Infantile idiopathic scoliosis occurs at the age of 0-3, and is rare – up to 1% of all idiopathic scoliosis cases. Juvenile idiopathic scoliosis occurs at the age of 4-10 and the prevalence is up to 10-15%. Both infantile and juvenile idiopathic scoliosis have poor prognosis of progression and often need operative treatment. Adolescent idiopathic scoliosis is the most frequent, comprising up to 90% cases of idiopathic scoliosis and occurs at the age of 11-18. The prognosis depends on age, gender and the scoliosis onset time (if later, then prognosis is better) [4].

Cystic fibrosis is a genetic, chronic, systemic disease that reduces life expectancy, and life quality as well. Chronic lung disease, malnutrition and reduced activity, caused by disease lead to postural disorders and scoliosis. Postural defects and scoliosis are believed to be more common in cystic fibrosis than in the healthy population. A British review of 316 patients affected with cystic fibrosis proved a higher level of scoliosis in cystic fibrosis patients. In this study there were 184 adults (age 17 years and over) and 132 children (age 0-6 years) from the Yorkshire Cystic Fibrosis unit. Their scoliosis rate was 20 times higher than in the healthy population aged 4-16 years [6].

Słowa kluczowe: Mukowiscydoza, postawa, nieprawidłowa postawa, wady postawy, skolioza

Postural defects in children with cystic fibrosis

Natomiast wady postawy podzielono na dwie grupy. Do pierwszej grupy zaliczono wady mające możliwy wpływ na funkcję układu oddechowego u pacjenta z mukowiscydozą: skoliozy, wady kręgosłupa i klatki piersiowej. Do drugiej grupy zaliczono wady kończyn dolnych, nie mające wpływu na funkcję układu oddechowego. Ze względu na fakt, że badania wad postawy u dzieci zdrowych prowadzone były przy użyciu różnych metod, trudno jest wyodrębnić grupę kontrolną.

Wyniki: Nieprawidłowości postawy i klatki piersiowej stwierdzono u 36 (87,8%) pacjentów, którzy zostali poddani badaniu. Wady, mogące mieć wpływ na funkcję układu oddechowego wystąpiły u 13 (31,7%) z 41 pacjentów. Zwiększona kifoza piersiowa (plecy okrągłe) występowała najczęściej. W grupie poniżej 5. r.ź. nie stwierdzono odchyleń od normy w zakresie kręgosłupa ani klatki piersiowej. W grupie 5-9 lat stwierdzono 1 (16,6%) przypadek skoliozy wrodzonej oraz 1 (16,6%) przypadek pleców płaskich. W grupie dzieci w wieku 9-13 lat zwiększona kifoza piersiowa została stwierdzona u 2 (18,1%) dzieci, plecy płaskie również u 2 (18,1%), w 1 (9,0%) przypadku rozpoznano zwiększona lordozę lędźwiową, klatka piersiowa beczkowata wystąpiła w 2 (18,1%) przypadkach. W grupie powyżej 13. r.ź. stwierdzono 4 (25%) przypadki skoliozy, 16 (100%) przypadków zwiększonej kifozy piersiowej oraz 6 (37,5%) przypadków klatki beczkowatej.

Wniosek: Liczba wad postawy u dzieci i młodzieży z mukowiscydozą wzrasta wraz z wiekiem, stanowi więc znaczący problem zdrowotny. Wydaje się, iż konieczne jest przeprowadzenie pogłębionych badań na większym materiale.

Słowa kluczowe: mukowiscydoza, postawa, nieprawidłowa postawa, wady postawy, skolioza
Postural disturbances may cause worsening of the patients' cardiopulmonary function. A vicious cycle is the final result of the combination of poor posture and cystic fibrosis. In 2003 a review of postural disorders of adult patients with cystic fibrosis was published. The authors listed typical complaints of patients with cystic fibrosis. The list included back pain and postural deformities. The most common postural deformity was increased thoracic kyphosis. The authors noticed that postural defects and back pain had both a physiological and a psychological influence on the cystic fibrosis patient. Chronic pain was a burden which caused low mood, reduced self-confidence and decreased motivation. Stiffness and pain in the back and ribs inhibited ventilation and airway clearance resulting in a vicious cycle [7].

The aim of this paper was to evaluate postural disturbances in the population of cystic fibrosis children treated at the Warsaw Institute of Mother and Child.

MATERIAL AND METHODS

The investigation was carried out in 41 cystic fibrosis patients (20 girls, 21 boys), aged 2-17 years (mean 8.1) and was performed by an orthopedic surgeon. Patients were divided into age groups: I under 5 y.o. – 8 patients., II 5-9 y.o. – 6 patients, III 9-13 y.o. – 11 patients, IV 13 y.o. and older – 16 patients.

Clinical orthopedic examination included the spine, chest, pelvis, knees and feet. Examination of the spine included assessment of curvatures, range of motion, plumb line, observation during bending and scolimetry (a manual scolimeter was used for this test). Chest and pelvis observation from the front, back and both laterals, and range of motion of the hips were also performed. The standing view of both lower extremities was assessed. Knee alignment was measured (in degrees) with the use of a manual goniometer. The feet were examined in posterior view in standing position. No electronic devices, cameras or endoscopes were used during the examination, which was based only on orthopedic physical examination.

All the children were examined in July and August 2014 in the Warsaw Institute of Mother and Child. All of them were seen by one and the same orthopedic surgeon. They are all under the care of the Cystic Fibrosis Centre in Warsaw Institute of Mother and Child.

Postural disorders were divided into two groups. The first group included defects with possible influence on the respiratory function of the cystic fibrosis patient: scoliosis, spine defects and chest defects. The second group included lower extremities defects without influence on the respiratory system function.

Because of the different methods of examination among posture studies of the healthy population, obtaining a control population is difficult.

RESULTS

Some postural disorders were observed in 36 (87.8%), of the 41 patients who underwent clinical orthopedic examination.

Defects of this kind are important for the patient with cystic fibrosis, because a possible influence on the respiratory system occurred in 13 (31.7%) of the 41 patients. Those defects included 5 (12.2%) cases of scoliosis, and 8 (19.5%) barrel chest combined with increased thoracic kyphosis.

Observation of the spine showed that 5 (12.2%) of the 41 patients met clinical criteria of scoliosis (scolometry >7 degrees). All those patients were examined with whole-spine x-ray in standing position for Cobb angle measurement. All the 4 (9.7%) cases of idiopathic scoliosis (without structural, neuromuscular or mesenchymal abnormalities of the spine) were in the group of 16 adolescents aged 13-17 years. One (2.4%) patient had congenital (non-idiopathic) scoliosis with Th1 and Th3 hemivertebrae. Idiopathic scoliosis was observed in 3 (15.0%) of the 20 girls and 1 (4.8%) of the 21 boys. Other spine abnormalities included 8 (19.5%) of the 41 patients with increased thoracic kyphosis. Plane back with decreased kyphosis was observed in 3 of the 41 patients (7.3%), increased lumbar lordosis was observed in only one patient.

Chest abnormalities were observed in 9 of the 41 patients (21.9%). Barrel chest was observed in 8 (19.5%) patients, hallowed chest (pectus excavatum) in 1 (2.3%) patient.

Pelvis asymmetry was combined with 5 (100%) cases of scoliosis. There was no isolated pelvis abnormality. No knee alignment abnormality was observed.

Flexible flatfoot was observed in 11 (26.8%) of the 41 cases. 6 (14.6%) of them were in patients under 5 years old, and were qualified as physiological. 3 (7.3%) older patients with flatfoot had symptoms of general laxity with a positive Marshall test. No cases of clubfoot, rigid congenital flatfoot (congenital vertical talus) nor any other foot disturbances were observed.

In group I – 8 patients under 5 years old, there were no important postural defects found. In 6 (75%) cases physiological flexible flatfoot was diagnosed.

In group II – 6 patients aged 5-9 years; 2 (33.3%) of the 6 patients had important postural defects. One (16.6%) case of congenital scoliosis with Th1-Th3 hemivertebrae and 1 (16.6%) case of plane back was found. 3 (50.0%) patients had flexible flatfoot combined with general laxity (Marshall test positive). Pelvis asymmetry was diagnosed in one (16.6%) patient with congenital scoliosis.

In group III – 11 patients aged 9-13 years; 8 (72.7%) of the 11 patients had important postural defects. 2 (18.1%) cases of barrel chest, 1 (9.0%) case of hallowed chest, 2 (18.1%) cases of increased thoracic kyphosis, 2 (18.1%) cases of plane back and 1 (9.0%) case of increased lumbar lordosis were found. In this group, none of the patients with barrel chest had spine abnormalities, but all of them were over 10 years old. There was no barrel chest deformity in patients younger than 10 years.

In group IV – all (100%) of the 16 children aged 13 years and more had important postural disorders. In the group aged 13 and older increased thoracic kyphosis was observed in all of the 16 (100%) patients, in 10 (62.5%) cases increased thoracic kyphosis was combined with other disorders. Barrel chest was observed in 6 (37.5%) of the 16 patients. In all those cases, barrel chest was combined with increased thoracic kyphosis. 4 (25.0%) of the 16
patients met the clinical criteria of scoliosis (scoliometry >7 degrees). All those cases were idiopathic scoliosis (without structural, neuromuscular or mesenchymal abnormalities of the spine) combined with increased thoracic kyphosis (kyphoscoliosis). Pelvis asymmetry was found in all the patients with scoliosis.

A summary of spine postural disturbances is shown in table 1. An example of a cystic fibrosis patient (a 16 year-old girl) with thoraco-lumbar scoliosis is presented in figures 1-4. Cobb angle of the thoracic curve is 40 degrees, lumbar 60 degrees. The patient was qualified for operative treatment of scoliosis.

**DISCUSSION**

The problem of postural disturbances in the cystic fibrosis population is becoming more and more remarkable. Postural disturbances are not an urgent life-saving problem like pulmonary, alimentary or liver dysfunctions for doctors handling cystic fibrosis. But they are really important for the patient, and are connected with longer life expectancy. Taking into consideration the possibility of lung transplantation, a postural formation with an intact spine and chest is becoming more and more important.

Tattersall and Walsh described postural disturbances as an element of a vicious cycle, when bad posture causes the worsening of the pulmonary condition [7].

Scoliosis has an influence on the pulmonary system even in the healthy population. Koumbourlis [8] listed the most important facts about scoliosis and cardiopulmonary function. In cases with moderate scoliosis, described as Cobb angle under 25 degrees, there are no respiratory complications.
In cases over 25 degrees, there is pressure in pulmonary arteries. In most advanced cases of scoliosis the author claims that patients may also develop evidence of lower airway obstruction. In cases over 70 degrees a significant decrease of lung volume may occur, while over 120 degrees there is a risk of chronic respiratory failure. Chronic airway inflammation, secondary to the poor clearance of secretions is also the result of severe scoliosis. Koumbourlis' study is based on the healthy population, however pulmonary complications may be more severe in cystic fibrosis.

Postural disorders are frequent even in the population of healthy children. A number of studies have been performed in Poland. It was believed that abnormal posture occurs in about 30-35% of healthy children. In our study the percentage was higher and reached 87%. The literature from Poland reveals quite divergent results of postural studies. Kaźmierczak reported data of 580 school children from the city of Bydgoszcz. Flatfoot was found in 39%, increased kyphosis in 36.9% [9]. Hanger et al reported poor posture in 64.5% of the children in the Mława district [10]. In our opinion the divergence in Polish studies lies in the methodology. Different methods of examination among healthy population postural studies were used, so studies can not be compared, or used as control population. In our opinion, there is a need of a great population study.
among children and the adolescent population in Poland with a unified examination protocol.

In Portugal the prevalence of scoliosis in the healthy population aged 10-16 was 4.2% [11]. In the Czech Republic, poor posture was diagnosed in 38.3% of 3520 school children [12].

No Polish studies about posture in cystic fibrosis have been found. According to international data, poor posture among healthy people is less common than in the cystic fibrosis population [7]. Typical and most common postural disorders in the population of adolescents with cystic fibrosis is increased thoracic kyphosis. In our study the prevalence of increased thoracic kyphosis in children over 13 years old was 100%. Okuro et al. studied increased thoracic kyphosis among forty-two patients with cystic fibrosis, and the prevalence was 61.9%. There was no difference in pulmonary function between the population with and without increased kyphosis in this study. [13].

Scoliosis among patients with cystic fibrosis is a serious problem, especially for those who expect lung transplantation. Kumar et al. have published a review of 316 cystic fibrosis patients from Yorkshire. The authors proved a higher level of scoliosis in cystic fibrosis patients than in the healthy population. There were 184 adults (age 17 years and over) and 132 children (age 0-6 years) from the Yorkshire Cystic Fibrosis unit. The scoliosis rate was 20 times higher than in the healthy population aged 4-16 years. The control group were 15,793 school children at a similar age and similar sex distribution from the same geographic area. In the population above, the prevalence was 9.8%, which is higher than that of the general population. Infantile scoliosis was described in this paper to be non-progressive [6].

Tattersall et al. published a paper about posture in cystic fibrosis. Unfortunately the population included in the study was adult, however the results are interesting even for a pediatrician. Poor posture is typical in adults with cystic fibrosis. Back pain is also typical in that population [7]. In our study, based on children, there was no problem of back pain, even in adolescents over 13 years old. Tattersall et al. claim that increased thoracic kyphosis is the most common postural abnormality in cystic fibrosis. This is typical for our population over 13 years. The authors write about psychological problems of cystic fibrosis and poor posture, which has an influence on the patients’ general condition and life quality [7].

CONCLUSION

The number of postural defects in children with cystic fibrosis increases with age, and is becoming a significant health problem.

There is a need of a trial about the postural development of children with cystic fibrosis in Poland, as well as one of the healthy population in Poland.

REFERENCES


Conflicts of interest/Konflikt interesu
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