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Combination of pectus carinatum and Scheuermann's disease in children: An empirical pattern or somite syndrome?

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Abstract

BACKGROUND: In recent years, the number of children with pectus carinatum tended to increase. The literature describes only a few cases of a combination of keeled chest deformity with a more serious pathology of the spine, i.e., Scheuermann's disease.

AIM: This study aimed to examine the frequency, clinical manifestations, and timely diagnosis of combined structural lesions of the thoracic spine in children with keeled chest deformity.

MATERIALS AND METHODS: This observational single-center cross-sectional study included patients aged 5–17 years with pectus carinatum. Categorical values were described by indicating absolute values and percentages in the sample, and quantitative indicators with normal distribution were described using arithmetic means and standard deviations and 95% confidence intervals. Quantitative indicators without normal distribution were described using the median and interquartile range.

RESULTS: Scheuermann's disease was detected in 11 (9.3%) of 118 children with pectus carinatum. "Pterygoid scapulae" was noted in 97 (82.2%) children with pectus carinatum, increased cervical lordosis in 93 (79.7%), and sloping, anteriorly adducted shoulders in 99 (83.9%), which significantly hampered the clinical assessment of the extent of thoracic kyphosis. At the time of examination, a rigid thoracic kyphosis was formed in a 16-year-old boy. In younger children (5–14 years old), Scheuermann's disease had no clinical manifestations and was detected only during screening X-ray examination, whereas in the older age group (15–16 years), 3 of 4 adolescents complained of back pain.

CONCLUSIONS: The frequency of Scheuermann's disease in children with pectus carinatum exceeds the average prevalence in the population. In patients with keeled chest pterygoid scapulae, excess cervical lordosis, and rounded shoulders are associated with the difficulty of the assessment of the magnitude of thoracic kyphosis. Asymptomatic progression of Scheuermann's disease is typical in younger children with pectus carinatum, and pain syndrome and the clinical picture of the disease develop only by the age of 15–16 years. Thus, all patients with pectus carinatum and posture disturbance should undergo a screening X-ray examination of the thoracic and lumbar spine to detect Scheuermann's disease and initiate treatment in time.

Keywords: pectus carinatum; thoracic wall; Scheuermann's disease; kyphosis; posture, children.

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ЭКО•ВЕКТОР

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Сочетание килевидной деформации грудной клетки и болезни Шейермана – Мау у детей: эмпирическая закономерность или сомитный синдром?

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Аннотация

Актуальность. В последние годы отмечена тенденция к резкому увеличению числа обращений детей с килевидной деформацией грудной клетки. В литературе описаны лишь единичные клинические наблюдения сочетания килевидной деформации грудной клетки с более серьезной патологией позвоночника — болезнью Шейермана – Мау.

Цель — исследование частоты клинических проявлений и способов своевременной диагностики структурных поражений грудного отдела позвоночника у детей с килевидной деформацией грудной клетки.

Материалы и методы. В обсервационное моноцентровое одномоментное срезовое исследование были включены пациенты 5–17 лет с килевидной деформацией грудной клетки. Категориальные значения описывали путем указания абсолютных значений и процентных долей в выборке; количественные показатели, соответствующие критериям нормального распределения, — при помощи средних арифметических величин и стандартных отклонений, границ 95 % доверительного интервала; не соответствующие критериям нормального распределения при помощи медианы и интерквартильного размаха.

Результаты. Из 118 детей с килевидной деформацией грудной клетки болезнь Шейермана – Мау была выявлена у 11 (9,3 %) пациентов. Клиническую оценку величины грудного кифоза значительно затрудняли отмеченные у 97 (82,2 %) детей с килевидной деформацией грудной клетки «крыловидные лопатки», у 93 (79,7 %) — усиление шейного лордоза и у 99 (83,9 %) покатые, приведенные кпереди плечи. Ригидный грудной кифоз на момент осмотра сформировался у одного юноши 16 лет. У детей младшей возрастной группы (5–14 лет) болезнь Шейермана – Мау не имела клинических проявлений и была выявлена только при скрининговом рентгенологическом исследовании, в то время как в старшей возрастной группе (15–16 лет) 3 из 4 подростков с симптомокомплексом «килевидная деформация грудной клетки + болезнь Шейермана – Мау» самостоятельно или при активном сборе анамнеза жаловались на боли в спине.

Заключение. Частота встречаемости болезни Шейермана – Мау у детей с килеведной деформацией грудной клетки превышает среднюю распространенность в популяции. Более чем у 80 % пациентов с килевидной деформацией грудной клетки выявлено крыловидное выстояние лопаток, избыточный шейный лордоз и скругленная форма плеч, что затрудняет оценку истинной величины грудного кифоза. У детей с килевидной деформацией грудной клетки младше 14 лет болезнь Шейермана – Мау прогрессирует бессимптомно и клинически неотличима от постуральной круглой спины, однако к 15–16 годам развивается характерная клиническая картина заболевания. Таким образом, всем детям с килевидной деформацией грудной клетки и нарушениями осанки показана скрининговая рентгенография грудного и поясничного отделов позвоночника с целью своевременного выявления и начала лечения болезни Шейермана – Мау.

Ключевые слова: килевидная деформация грудной клетки; деформация грудной клетки; болезнь Шейермана – Мау; юношеский кифоз; дети.

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儿童胸廓龙骨畸形和Scheuermann病（休门氏病）的结合：是经验规律性还是体节综合征？

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简评

论证。近年来，患有胸廓龙骨畸形的儿童数量急剧增长。在文献中，只有个别的与胸廓龙骨畸形和Scheuermann病（休门氏病）结合有关的临床观察，要考虑到，休门氏病是一种更严重的脊柱病变。

目的是对患有胸廓龙骨畸形的儿童有胸廓结构性病变的发生率、最常见的临床表现并及时诊断方法的有效性进行评估。

材料和方法。5–17岁的胸廓龙骨畸形患者被纳入一项观察性、单中心、横向的研究中。分类值是用样本中的绝对值和百分比而描述的；符合正常分布标准的数量指标是用算术平均数和标准差、95%置信区间而描述的；不符合正常分布标准的数量指标是用中位数和四分位距而描述的。

结果。共有118名患有胸廓龙骨畸形的儿童参加了这项研究。检测到11名（9.3%）患有休门氏病的患者。97名（82.2%）儿童的“翼状肩胛”、93名（79.7%）儿童的颈椎过度前凸以及99名（83.9%）儿童的肩膀前倾使胸椎后凸的临床评估变得复杂。一名16岁的男孩在检查时已经有僵硬胸椎后凸。在年龄较小（5–14岁）的一组儿童中，Scheuermann病没有临床表现，只是通过筛查放射学检查而发现的。对较大年龄（15–16岁）组来说，在4名具有“胸廓龙骨畸形+Scheuermann病”症状复合体的青少年中，有3名单独或在积极询问病史时诉说背痛。

结论。胸廓龙骨畸形患儿有休门氏病的发病率高于人群的平均发病率。检测到，80%以上的胸廓龙骨畸形患儿具有“翼状肩胛”、颈椎过度前凸、肩膀前倾，这些因素使得胸椎后凸的临床评估变得困难。在14岁以下患有胸廓龙骨畸形的儿童中，Scheuermann病没有临床表现，临床上与姿势性驼背没有区别，但到15–16岁时，会出现该病的特征性临床表现。因此，对所有患有胸廓龙骨畸形和姿势障碍的儿童应进行胸椎和腰椎的筛查放射学检查，以便及时发现和治疗Scheuermann病。

关键词：胸廓龙骨畸形；胸廓畸形；Scheuermann病；青年性驼背；儿童。

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BACKGROUND

Over the past 20 years, pediatric thoracic surgeons and orthopedists have noted the progressive increase in the numbers of children and adolescents with complaints of pectus carinatum (PC), which is likely due to the increased public awareness regarding this problem and the development and spread of noninvasive treatment of PC [1]. PC accounts for up to half of all chest malformations; the ratio of the morbidity structure among boys and girls is 3:1–4:1 [1, 2]. PC manifests in adolescents during the period of active growth, but the etiology and pathogenesis of thoracic malformations remain unknown [2]. Studies on the rib cartilage tissue of children with PC have revealed molecular and cellular abnormalities indicating deterioration of cartilage metabolism [3]; however, reliable data on changes in its mechanical characteristics have not been obtained. In addition, relative elongation of the cartilage and relative shortening of the bone part of ribs were observed in children with PC [4]. Probably, the excessive suppleness and propensity for deformations of the thoracic skeleton are explained by a combination of histological and anatomomo-morphological changes.

In recent years, several research groups have independently noted the high incidence of combined anterior thoracic wall deformities, particularly PC with various changes in the posterior thoracic framework–postural or structural defects of the thoracic spine. According to N. Alaca et al. [5], 83.3% of children with PC showed a kyphotic posture. In a later publication, the same research group reported back pain in 13.3% of children with PC, and pathological kyphosis was detected in 35.6% of patients [6]. Thus, sagittal balance abnormalities in children with PC are not limited to postural abnormalities; a literature review revealed three papers describing the combination of structural anterior and posterior thoracic pathologies, keel or funnel chest deformity, and Scheuermann's disease (SD) disease [7–9]. However, all these publications focused on the description of individual clinical observations or small samples (less than 10 people) and did not imply the assessment of the prevalence of the phenomenon. According to K.A. Likhotay [7], signs of spinal osteochondropathy were detected in almost half (3 of 8) of the children included in a study on PC, which resulted in the skepticism regarding the postural nature of kyphosis in children with "bird's chest" and suspicion of more serious disorders of the thoracic spine in this patient group.

SD disease, an idiopathic degenerative–dystrophic lesion of vertebral body apophysis, is clinically manifested by the formation of pathological rigid kyphosis in the lower thoracic (less often in the mid-thoracic or upper lumbar) spine, and pain syndrome that first appears in adolescence and progresses during life [10]. Thus, SD disease is considered a specific case of osteochondrosis (juvenile osteochondrosis). The incidence of SD disease ranges from 0.4%–8.3%, depending on the choice of diagnostic criteria [11]. The use of the radiological criterion for this variant of osteochondropathy — a wedge-shaped deformity of three or more adjacent vertebrae located at the apex of kyphosis by more than 5° — has become generally accepted [11]. In addition, radiographs of patients with SD disease may reveal other changes, such as an increase in the thoracic kyphosis of more than 40° (the norm established by the Scoliosis Research Society), necrosis of apophysis and usurpation of the closing plates of the involved vertebrae, and impingement syndrome of spinal discs with the formation of Schmorl hernia [10-12].

This work aimed to investigate the frequency, clinical manifestations, and methods of timely diagnosis of structural lesions of the thoracic spine in children with ciliated thoracic deformity.

MATERIALS AND METHODS

The clinical and instrumental data of children and adolescents with PC who came for outpatient visits to an orthopedist and/or thoracic surgeon at the Filatov City Children's Clinical Hospital were considered in the formation of the observational monocenter single-sample study sample. The inclusion criteria were the consent of parents or other legal representatives for the adolescents' participation in the study and detection of PC in the child during the clinical examination. Patients with infantile cerebral palsy and other neurological disorders, children with chronic respiratory diseases (bronchial asthma, tuberculosis, etc.), and those with a history of surgical interventions on chest organs, heart or lung malformations, spinal, and rib or sternum fractures were excluded from the sample.

During the initial examination of children and adolescents, we registered the complaints that prompted them to consult a physician. The patients were specifically questioned about the presence of back pain at rest and during physical exertion, family history, life history (particular the concomitant diseases included in the connective tissue dysplasia (CTD) complex, namely, myopia and mitral valve prolapse). All patients were measured for height and body weight, and the presence of skin striae was noted. During orthopedic examination, we determined the posture type, noted "wing-like" shoulder blades, rounded shoulder shape, axial limb deformities, and pathological foot position (valgus deformity or flat feet) and assessed skin elasticity and joint hypermobility (as a result of subjective results, the latter two parameters were excluded in the work).

All children underwent direct and lateral Adams functional tests. After completing the clinical examination, patients with severe posture disorders underwent radiography of the thoracic and lumbar spine in standing position in straight and lateral projections. Thoracic kyphosis was measured, and the presence or absence of wedge-shaped deformity of three or more adjacent vertebrae, aseptic necrosis of apophysis and usurpation of the vertebral body closure plates, and Schmorl herniation

were noted on the radiographs. The wedge-shaped deformity of three adjacent vertebrae by more than 5° was considered as a radiological diagnostic criterion for SD disease. In addition to signs of osteochondropathy, the following pathological "incidental findings" were noted in the evaluation of radiographs: splitting of vertebral arches, spondylolysis, spondylolisthesis, etc. The clinical study was approved by the local ethical committee of the Medical Research and Education Center of the Lomonosov Moscow State University (protocol No. 10/20 of September 14, 2020). Each legal representative of a child who met the criteria for inclusion in the sample was given an information sheet summarizing the contents of the study. After reading the information sheet, the parents or other legal representatives can inquire the physician if they have additional questions and decide whether to sign a consent form allowing the use of data constituting medical secrecy as a part of the research work. After signing the consent, each patient was assigned an individual number for the creation of an anonymous information base. The sample size was not preliminarily calculated. SPSS Statistics 28 software package (SPSS Inc., USA) was used for statistical data processing. Categorical values were described by indicating the absolute values and percentages of the sample. Quantitative measures were evaluated for normality using the Kolmogorov–Smirnov criterion. Quantitative measures with a normal distribution were described using arithmetic mean (*M*), standard deviation (*SD*), and 95% confidence interval limits. Given the small size of the group of patients with combined SD disease and PC, the results were described using nonparametric statistical tools. A series of quantitative data was characterized using median and interquartile range (–*Me* [Q_1–Q_3]). Anthropometric measures of height and body mass index (BMI) were compared with the population averages for children 5–19 years old published on the World Health Organization (WHO, 2007) website [13].

RESULTS

For sample formation, the clinical status and instrumental findings of 118 children with PC who were presented to an orthopedist or thoracic surgeon at the N.F. Filatov City Children's Clinical Hospital were reviewed. Active questioning revealed 23 children who reported pain occurring with exertion or prolonged sitting. All children underwent a

Table 1. General characteristics of the sample, complaints, and state of the sagittal component of posture based on clinical examination results

Таблица 1. Общие характеристики выборки, жалобы и состояние сагиттального компонента осанки по результатам клинического осмотра

Indicator	Value
Gender, *n*	
Boys	104
Girls	14
Age, years, *M* ± *SD*	12.7 ± 2.7
Anthropometric indicators, *M* ± *SD*	
Height, cm	166.4 ± 13.7
Body mass index, kg/m^2	17.3 ± 2.1
Reason for treatment, *n*	
Unsatisfactory appearance of the chest	118 (100%)
Other complaints, *n*	
Posture disorder	81 (68.6%)
Back pain syndrome	23 (19.5%)
State of posture, *n*	
Normal	58 (49.2%)
Kyphotic	34 (28.8%)
Kypholordotic (poor posture)	20 (16.9%)
Lordotic	4 (3.4%)
"Straight back" (spina recta)	2 (1.7%)
Rigid thoracic kyphosis	1 (0.8%)
Other abnormalities of sagittal balance and muscle tone in the chest. neck, *n*	
Winged blades	97 (82.2%)
Increased cervical lordosis	93 (79.7%)
Sloped, fronted shoulders	99 (83.9%)

Table 2. X-ray parameters, signs of the spine osteochondropathy, and incidental findings in children and adolescents with pectus carinatum and Sheuermann–Mau disease

Таблица 2. Рентгенологические параметры, признаки болезни Шейермана – Мау и случайные находки у детей и подростков с сочетанием килевидной деформации грудной клетки и болезни Шейермана – Мау

Indicator	Value
Scoliosis, *n*	
Less than 10° (grade I scoliosis)	7 (63.6%)
10° to 25° (degree II scoliosis)	2 (18.2%)
Mean thoracic kyphosis, deg, *Me* [Q_1–Q_3]	52° [42–57]
Signs of Scheuermann's disease, *n*	
Cuneiform deformity of three adjacent vertebrae more than 5°	11 (100%)
Pathological kyphosis (over 40°)	9 (81.8%)
Aseptic necrosis of the apophysis	10 (90.9%)
Endplate usurpation of vertebral bodies	11 (100%)
Schmorrhoid hernias	2 (18.2%)
Other radiological findings, *n*	
Splitting of the L_V vertebral arch (*spina bifida occulta*)	2 (18.2%)
Spondylosis and Spondylolisthesis Grade I L_V-S_I	1 (9.1%)

complete orthopedic examination, and the results indicated pronounced posture disorders in 60 (50.8%) of them. Table 1 presents the general characteristics of the sample and condition of the sagittal component of the posture according to the results of clinical examination.

A total of 97 (82.2%) children with PC had "winged scapulae," 93 (79.7%) had increased cervical lordosis, and 99 (83.9%) had sloping, forward shoulders, which caused difficulty in the clinical assessment of thoracic kyphosis. In this regard, all children with poor posture underwent X-ray examination of the thoracic and lumbar spine in straight and lateral projections in standing position.

In addition, the formation of rigid thoracic kyphosis persisted during the Adams test of one 16-year-old patient. Thus, the remaining patients had no reliable clinical signs of SD disease at the time of examination.

According to the results of X-ray examination, 11 (9.3%) children with PC had SD disease. Table 2 shows the radiological parameters, signs of SD disease, and incidental findings in children and adolescents with a combination of PC and SD disease.

Given the need to identify clinical and anamnestic features and therefore the diagnostic criteria for patients with the combination of PC and SD disease, the general characteristics of this group of children were considered separately (Table 3).

The combined height of 94 (79.7%) children with PC and 10 (90.9%) children with PC + SD disease was greater than the average for their age group (according to WHO, 2007) [13], and BMI was lower than the combined age median in 89 (75.4%) children with PC and 10 (90.9%) children with PC and SD disease [13]. The comparison of clinical and instrumental parameters between children with PC and children with a combination (PC + SD disease) was not feasible because the patients with a combined pathology were significantly older, which prevented the exclusion of the conditionality of differences between age-related changes and the stage of pathology development in the groups.

Critically, all seven children 9–14 years of age with a combination of PC and SD disease denied pain in the

Table 3. Characteristics of the children with pectus carinatum and Scheuermann's disease: sex and age, anthropometric characteristics, and complaints

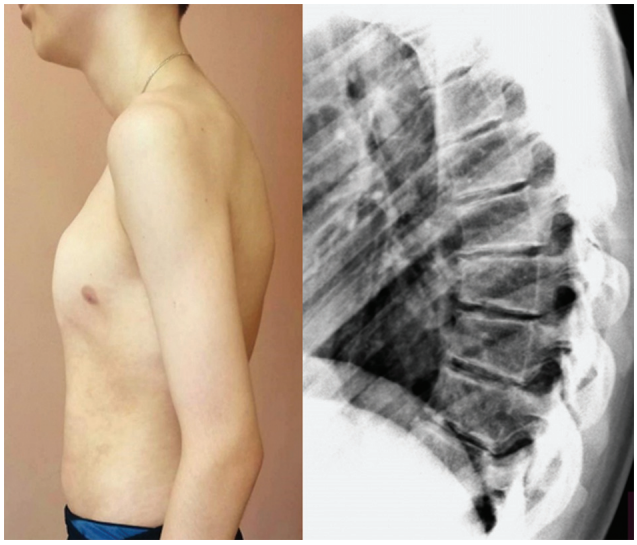
Таблица 3. Характеристики группы детей с сочетанием килевидной деформации грудной клетки и болезни Шейермана – Мау: половая и возрастная структура, антропометрические характеристики и жалобы при обращении к врачу

Parameter	Value
Gender, *n*	
Boys	10
Girls	1
Age, years, *Me* [Q_1–Q_3]	14 [13–15]
Anthropometric indicators, *Me* [Q_1–Q_3]	
Height, cm	176 [165.8–180.6]
Body mass index, kg/m^2	18.0 [17.1–19.2]
Complaints, *n*	
Unsatisfactory appearance of the chest	11 (100 %)
Posture Disorder	8 (72.7 %)
Back pain syndrome	3 (27.3%), adolescents 15–16 years old

Table 4. Family history, comorbidities related to the connective tissue dysplasia syndrome, and type of pectus carinatum in children with pectus carinatum and Scheuermann's disease

Таблица 4. Семейный анамнез, сопутствующие заболевания, относящиеся к синдрому дисплазии соединительной ткани, и форма килевидной деформации грудной клетки у детей в сочетании с болезнью Шейермана – Мау

Parameter	Number of patients, *n*
Family history	
Deformities of the thorax:	
keeled	1 (at my uncle's)
funnel shaped	1 (at my sister's)
Scoliosis of degree II or higher	1 (mother's)
Flat-valgus foot deformity	2 (mother and sister)
Anamnestic and instrumental signs of connective tissue dysplasia	
Mitral valve prolapse without hemodynamically significant regurgitation	4 (36.4%)
Mild to moderate myopia	3 (27.3%)
Clinical signs of connective tissue dysplasia	
Striae	2 (18.1%)
Flat-valgus foot deformity	4 (36.4%)
Type of keel-shaped chest deformity	
Symmetrical	2 (18.2%)
Asymmetric	9 (81.8%)

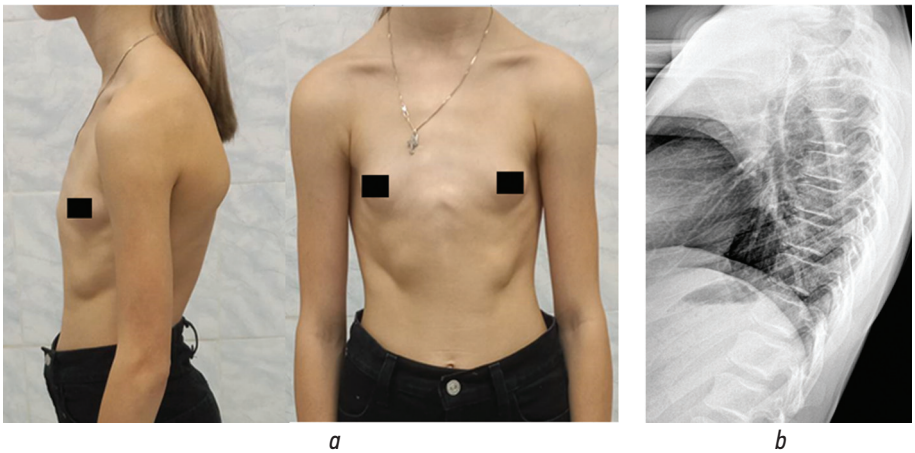


a *b*

Fig. 1. Appearance (*a*) and results of chest X-ray (*b*) of a 14-year-old adolescent with pectus carinatum and Scheuermann's disease
Рис. 1. Внешний вид (*a*) и результаты рентгенографии грудной клетки (*b*) подростка 14 лет с килевидной деформацией грудной клетки и болезнью Шейермана – Мау

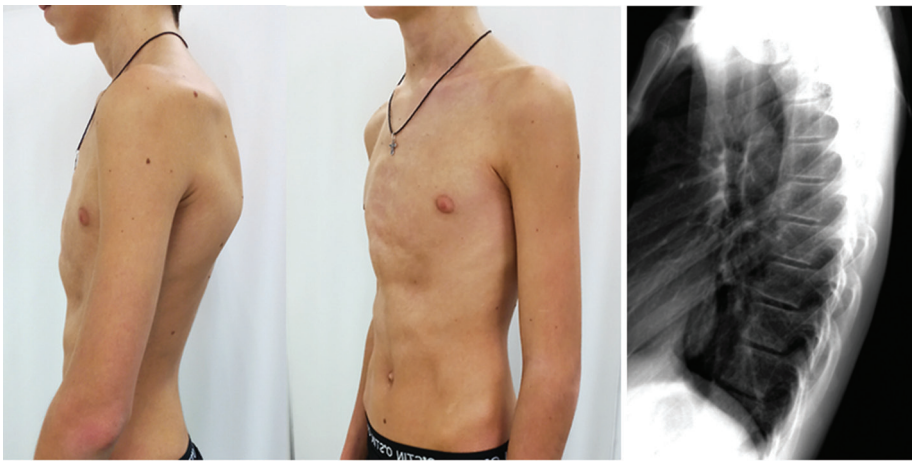
thoracic or lumbar spine, and in the older-age group (15–16 years), 3 out of 4 adolescents recognized pain syndrome on their own or during the active collection of their history.

During history taking and examination of children with a combination of PC and SD disease, a number of signs of nonspecific CTD were detected. Table 4 shows the data on the abnormalities detected and the form of PC in this group of patients.



a *b*

Fig. 2. Appearance (*a*) and radiographs (*b*) of a 12-year-old child with pectus carinatum, kyphotic posture, and pterygoid protrusion of the shoulder blades. Some signs of osteochondropathy do not meet the criteria of Scheuermann's disease (apophysial necrosis and usuration of the vertebral bodies' endplates without their wedge-shaped deformation) on lateral radiographs
Рис. 2. Внешний вид (*a*) и рентгенограммы (*b*) ребенка 12 лет с килевидной деформации грудной клетки, кифотической осанкой и крыловидным выстоянием лопаток. На рентгенограммах в боковой проекции признаки остехондропатии, не отвечающие критериям болезни Шейермана – Мау (некроз апофизов, узурация замыкательных пластинок тел позвонков без их клиновидной деформации)



a *b*

Fig. 3. Appearance (*a*) and radiographs (*b*) of a 14-year-old child with pectus carinatum, back pain, neck hyperlordosis, and rounded shoulders. No pathological findings were noted on the lateral radiographs
Рис. 3. Внешний вид (*a*) и рентгенограммы (*b*) ребенка 14 лет с килевидной деформации грудной клетки и жалобами на боли в спине, усилением шейного лордоза, крыловидным выстоянием лопаток и приведением кпереди плеч. На рентгенограммах в боковой проекции патологии не выявлено

For comparison, we presented the appearance and radiographs of a 12-year-old patient with PC, kyphotic posture, and wing-like protrusion of the scapulae. The radiographs in the lateral view show signs of osteochondropathy, which do not meet the criteria for SD disease (necrosis of apophysis and usuration of the closing plates of vertebral bodies without a wedge-shaped deformity; Fig.2), of a 14-year-old patient with PC, increased cervical lordosis, wing-like protrusion of the scapulae, and rounded shoulder shape. The child complained of back pain during prolonged walking with a backpack. X-Rays in the lateral view showed no abnormalities (Fig. 3).

DISCUSSION

To date, the etiopathogenetic association of PC with spinal disorders and sagittal posture abnormalities is unclear. In accordance with the national concepts, PC is included in the symptom complex of genetically determined CTS, including posture disorders and spinal deformity as particular manifestations [14]. In addition, the simultaneous formation of a number of local orthopedic disorders at the corresponding levels of the spine (thoracic, lower cervical, and upper lumbar spine) in patients with PC suggests the possible mutual mediation of these pathologies.

A biomechanical relationship exists between the thoracic wall frame and thoracic spine [7-9, 15-17]. E.E. Berg described two cases of pronounced kyphosis in patients with concomitant fractures of the sternum and thoracic spine with minimal displacement; based on the results, Berg concluded the importance of the sternum in maintaining the stability of the thoracic spine and proposed that the sternum be considered as the “fourth column” of the spine [16]. J.S. Butler et al. [17] described the formation of pronounced thoracic kyphosis in patients with myeloma disease and pathological fractures of the spine in the presence of concomitant fractures of the sternum. Experiments on cadaveric material revealed a 25.8% increase in the sagittal mobility of the thoracic spine after sternal osteotomy and rib and sternum release (largely due to an increase in the extension amplitude) [18]. Nagasao T. et al. [15] described changes in the progression dynamics of thoracic spine scoliosis after surgical correction of asymmetric funnel chest deformity according to Nass. E. Fotiadis et al. [19] observed a statistically significant sternal shortening in patients with SD disease (16.7 cm compared with 17.8 cm in healthy children from the control group). By contrast, the height of adolescents with osteochondrodysplasia was higher (166.8 cm compared with 163.5 cm in the control group).

During the analysis of the literature on the combination of thoracic deformities and SD disease, we found three works devoted to the description of individual clinical observations or small samples [7–9]. K.A. Likhotai [7] demonstrated signs of thoracic spine osteochondropathy in 3 out of 8 children included in the study with PC. The combination of PC and SD disease was also described in a young man aged 18 years old; the patient underwent a one-stage surgical correction of both pathologies [8]. P.A. Sugrue et al. [9] reported the dramatic progression of SD disease with the formation of a 105° rigid kyphosis in a 14-year-old adolescent after Nass surgery for a funnel-shaped thoracic deformity. The authors attributed their observation to the interference in the structure of the rib–chest complex and violation of its biomechanical relationship with the spinal column; however, the primary radiographs presented in the work showed distinct signs of osteochondrodysplasia, which at that time did not meet the criteria of the classical form of SD disease, namely, necroses of apophysis, usuration of the closing plates,

and wedge-shaped deformation of individual vertebrae [9]. SD disease and PC are characterized by a set of similar clinical signs, such as manifestation during the adolescent growth spurt in tall, asthenic adolescents and characteristic postural abnormalities (rounded shoulder shape due to increased pectoral muscle tone and cervical lordosis) [5]; however, the etiopathogenetic link between these conditions remains unclear: perhaps, both processes are independent manifestations of CTD, but mutual biomechanical mediation of these states cannot be excluded. The leading role of the hereditary factor (CTD) is supported by the detection of other manifestations of dysplasia syndrome (flat feet, myopia, and striae) in patients. The possible significance of the biomechanical factor is evidenced by literature data on changes in the dynamics of scoliosis or thoracic kyphosis in patients with sternal fractures [16, 17] or after thoracoplasty [9, 15].

Decreases in back muscle strength (18.8 kg compared with 32.7 kg in healthy adolescents) and general asthenia, that is, a decrease in BMI (17.5 kg/m^2 compared with 20.7 kg/m^2 in the control group), are considered as possible causes of pathological kyphosis in children with PC [6].

Despite the gaps in the understanding of the etiopathogenesis of the simultaneous formation of PC and SD disease in adolescents, the present study allowed us to draw important conclusions from a clinical point of view. The authors identified a higher incidence of SD disease among adolescents with PC than in the population (9.3% versus 0.4%–8.3% according to the literature) [11]. Moreover, no clinical criteria that were identified allowed the verification or showed a high degree of probability to suspect SD disease in a child with PC. Moreover, asymptomatic osteochondropathy of the thoracic spine with evident structural changes in the vertebrae, including their wedge-shaped deformity, was observed in 100% of children aged 9–14 years. In the older-age group (15–16 years), 3 out of 4 (75%) adolescents with a combination of SD disease and PC reported pain in the thoracic or lumbar spine.

The results obtained are consistent with those described in the literature; complaints of back pain in SD disease can occur at 8–12 years of age and in adulthood, and a clear radiological image of osteochondropathy can be obtained in 12–13-year-old patients regardless of the pain syndrome [11, 12]. Adolescents who consulted a physician at a later age and did not receive treatment in the early stages of the disease exhibited a more pronounced and rigid deformity of the spine during the initial examination [11]. We observed such a deformity in one adolescent 16-year-old boy. In the initial stages of disease development, rigid thoracic kyphosis may be absent; in this case, the clinical manifestation of SD disease is identical to that of kyphotic posture (so-called postural roundness of the back, PCS), and differential diagnosis of these conditions is possible only through the evaluation of radiological images [12]. As conservative therapy aimed at slowing down the destruction of the apophysis is effective only in the early stages of the disease, the timely diagnosis of SD disease is critical to improve the treatment prognosis [11]. In our study sample, the younger children (9–14 years old) who were presented to a physician because of poor chest appearance, clinical signs of SD disease (pain syndrome in the spine and rigid kyphosis) possibly manifested later, which hindered the timely initiation of treatment and preventive measures and led to a poorer clinical outcome.

LIMITATIONS

The present study focused on the combination of PC and SD disease to determine its true incidence among children with PC, identify clinical manifestations, and shape approaches to the diagnosis of structural spinal pathology against the background of sagittal imbalance, which is typical among the vast majority of children with PC. The study of radiographs of children with PC revealed cases of spinal osteochondropathy in children without structural kyphosis. Thus, the incidence and manifestation of spinal osteochondropathy in children with various forms of PC against the background of certain spinal sagittal balance abnormalities require further study.

CONCLUSION

The incidence of SD disease in children with ciliated thoracic deformity exceeds the average prevalence in the population (9.3% versus 0%–8.3% according to various literature sources). More than 80% of patients with PC showed wing-like protrusion of the scapulae, excessive cervical lordosis, and rounded shoulders, which caused difficulty in the assessment of the true value of thoracic kyphosis. In children with PC under 14 years of age, SD disease progresses asymptomatically and is clinically indistinguishable from the postural round back. However, by 15–16 years of age, complaints of pain syndrome appear, and a characteristic clinical image of the disease develops. Thus, screening radiographs of the thoracic and lumbar spine is indicated for all children with PC and postural abnormalities to detect and initiate the treatment of SD disease in a timely manner.

ADDITIONAL INFORMATION

Author contribution. Thereby, all authors made a substantial contribution to the conception of the study, acquisition, analysis, interpretation of data for the work, drafting and revising the article, final approval of the version to be published and agree to be accountable for all aspects of the study. The contributions of each author: E.A. Vorobyeva — clinical examination and treatment of patients with pectus carinatum, analyzing the data, writing the text of the article; A.Yu. Razumovskiy — clinical examination and treatment of children with pectus carinatum, organization of a multidisciplinary medical team working, editing the text of the article; V.E. Dubrov

— development of the research design, writing and reviewing the text of the article, organizing interaction with the local ethics committee and preparing materials for its meeting; D.Yu. Vybornov — clinical examination and treatment of children with pectus carinatum, organization of a multidisciplinary medical team working, editing the text of the article; I.V. Krestyashin, Z.B. Mitupov, E.L. Vakhova — clinical examination and treatment of patients with pectus carinatum, writing and reviewing the text of the article.

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Consent for publication. Written consent was obtained from the patient for publication of relevant medical information and all of accompanying images within the manuscript.

ДОПОЛНИТЕЛЬНАЯ ИНФОРМАЦИЯ

Вклад авторов. Все авторы внесли существенный вклад в разработку концепции, проведение исследования и подготовку статьи, прочли и одобрили финальную версию перед публикацией. Вклад каждого автора: Е.А. Воробьева — обследование и лечение пациентов с килевидной деформацией грудной клетки, статистическая обработка и анализ данных, написание текста статьи; А.Ю. Разумовский, Д.Ю. Выборнов — обследование и лечение детей с килевидной деформацией грудной клетки, организация работы мультидисциплинарной бригады для оказания помощи таким пациентам, редактирование текста статьи; В.Э. Дубров — разработка дизайна исследования, написание и редактирование текста статьи, организация взаимодействия с локальным этическим комитетом и подготовка материалов для его заседания; И.В. Крестьяшин, З.Б. Митупов, Е.Л. Вахова — обследование и лечение пациентов с килевидной деформацией грудной клетки, написание и редактирование текста статьи.

Конфликт интересов. Авторы декларируют отсутствие явных и потенциальных конфликтов интересов, связанных с проведенным исследованием и публикацией настоящей статьи.

Источник финансирования. Авторы заявляют об отсутствии внешнего финансирования при проведении исследования и подготовке публикации.

Информированное согласие на публикацию. Авторы получили письменное согласие законных представителей пациента на публикацию медицинских данных и фотографий.

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