THORACIC INSUFFICIENCY SYNDROME IN INFANTILE CONGENITAL SCOLIOSIS*

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Review of modern literature on the problem of thoracic insufficiency syndrome in small children with severe congenital scoliosis associated with abnormal development of vertebrae and ribs is presented. Pathogenesis and clinical picture of this condition are described; the examination methods allowing one to objectify the thorax and spine status in a child in the process of natural development of pathology and its treatment are considered.

Key Words: congenital scoliosis, hemithorax, thoracic function, lung growth.


The problem of infantile congenital scoliosis diagnosed in the first days and months of life has always been regarded as a rather complex one in spinal surgery, since no efficient treatment methods have been known. This is particularly true for scoliosis associated with abnormal development of vertebrae and ribs (Fig. 1), when malignant progression of these abnormalities inevitably causes patient’s early disability and death. The treatment of these deformities is in most cases reduced to a complex of general spine reinforcement procedures, which often include the absolutely inefficient brace treatment (this type of therapy should be considered harmful, since it has a negative effect on body muscles, while failing to stop the progression of spine deformity). The attempts at surgical treatment of these deformities typically included various versions of epiphysiodesis limiting spinal growth.

The past 20 years have been characterized by the emergence and wide spread of the fundamentally new treatment procedure associated with the surgical control and correction of spine and thorax deformities before skeletal maturity is achieved. The emergence of this method dates back to 1987, when American orthopaedist Campbell [6, 7] used a chest wall prosthesis consisting of a silicon plate and vertically oriented Steinmann pins for the first time in a 6-month-old child with congenital floating thorax, scoliosis, and high risk of early death from respiratory insufficiency. The surgery made it possible both to abandon the use of oxygen respiration support (which used to be necessary earlier) and to observe that an increase in distance between the ribs limiting the chest wall deformity was accompanied by significant correction of spine deformity in the control radiographs. The vertical expandable prosthetic titanium rib (VEPTR) was designed for further treatment of this child; Steinmann pins have been replaced with this device since 1989. The expansion thoracoplasty surgery, which includes the use of VEPTR both as a chest wall prosthesis and as a distraction device to normalize the hemithorax shape and volume, was subsequently proposed by Campbell.
The concept of thoracic insufficiency syndrome (TIS), which explained the mechanisms of pulmonary lesions in patients with the thoracic cage reduced in size, stiff and deformed because of the block-age, deformity or absence of ribs, was simultaneously developed. The pathology of the syndrome was additionally interpreted in study by Boffa et al. [4], who presented the autopsy data obtained for a 36-year-old woman with untreated severe spinal deformity. She has died because of acute heart failure associated with severe lung lesions (lung volume corresponded to that of a 6-year-old child, while the number of alveolar cells was comparable to that in a 1-year-old child). Campbell et al. [6] emphasized that after early epiphysiodesis, children with this pathology of the chest wall can live and retain motion activity close to the normal one; however, respiratory insufficiency inevitably develops in late adolescence because of the abruptly increasing body weight, while pulmonary infection may result in death of patient as early as in the third decade of life.

The prevalence of latent respiratory insufficiency in young patients with thoracic deformities is unknown. Patients with restrictive lung disease can be clinically tolerant to them during a long period; however, many of the patients over 40 years of age require oxygen respiration support and the mortality rate among them increases dramatically.

The development of the lung tissue due to the increase in number of alveolar cells continues in children until the age of eight years [13]. Correction of the deformity of spine and ribs at this age can facilitate the normalization of growth of all the chest cage components, including spine and lungs. Lung growth is impossible in older patients.

Campbell et al. [6] have formulated the definition of the thoracic insufficiency syndrome: it is the inability of the thorax to maintain normal respiration and lung growth. Thus, this syndrome consists of two major components.

1. Thorax status that fails to ensure normal respiration. The failure to provide normal secondary respiration of the defective thorax is compensated for by increased breathing frequency (tachypnea) or reduced motion activity to sustain the normal level of arterial oxygenation. These mechanisms make it possible for a child with mild thoracic insufficiency syndrome to normally exist. The aggravation of the thorax deformity results in further limitation of its mobility and volume, while lung expansion becomes completely dependent upon diaphragm function. The situation is aggravated by a respiratory infection. Compensation failure occurs as a result; the thoracic insufficiency syndrome manifests itself first in the form of dyspnea and subsequently in inability of living without oxygen respiration or even ventilatory support.

2. Impossibility of normal lung growth caused by rigid abnormalities of the development of vertebrae and ribs, which are defined as severe forms of hemimetameric hypoplasia and multiple hemimetameric aplasias of the spine [1]. These deformities limit growth of the lungs, whose volume becomes inadequate for normal...
vital activity as early as in adolescence. Lethal abnormalities of the thorax shape and function are typically observed in patients with such conditions as asphyxiating thoracic dystrophy (Jeune syndrome) [17] or the total shortening of the thoracic spine and hemithorax in patients with spondylocostal dysplasia (Jarcho-Levin syndrome), which is accompanied by the 33 % mortality rate [15, 16].

With allowance for the pathological components of the syndrome, the strategy for its treatment should be focused on the recovery of the thoracic volume and function and retention of the achieved effect during the entire growth period.

Growth of the thorax is the complex process determined by elongation of the thoracic spine and symmetrical growth of the hemithoraces due to the growth and spatial orientation of ribs in accordance with child’s age. Thorax height is directly related to the growth of the thoracic spine, which normally grows at a rate of 1.4 cm/year during the period between birth and 5-year-old age; 0.6 cm/year during the period between 6 and 10 years; and 1.2 cm/year between 11 and 15 years [9]. Insufficient elongation of the thoracic spine caused by congenital scoliosis can be calculated by dividing its actual height by the one expected according to the child’s age. The specific relationship between the loss of the hemithorax volume and the shortening of the thoracic spine, as well as indirect negative effect on lung growth, remain unstudied. In any case, severe shortening of the thoracic spine abruptly reduces the thoracic volume and, correspondingly, lung volume. Thus, the thoracic spine in patients with Jarcho-Levin syndrome can be actually presented by a single block vertebra no higher than 1/4 of the normal height.

The thoracic width and depth play a very significant role. In a newborn infant, ribs have a horizontal orientation; the longitudinal growth of ribs directly increases the diameter of the thorax, whose shape in the horizontal plane approaches a square. The thoracic volume in children of this age is 6.7% of that in adults. By two years, the orientation of ribs in children changes: they become slanted downwards [14], while the thorax becomes oval in transverse section. The extremely abrupt inclination angle of the ribs flattens the thorax; its sagittal diameter decreases, so does its volume, by 5 and 10 years of life reaching 30 and 50 % of that in adults, respectively. During the final third of the period of skeletal growth (from 10 to 16–18 years), the thoracic volume increases most rapidly and eventually acquires the shape close to the rectangular one in transverse section.

Approximately 85 % of pulmonary alveolar cells are formed after birth [5]; the maximum increase in their number is observed in two-year-old children [13]. As mentioned earlier, this significant process is completed by the age of approximately 8 years; after it is attained, the lung volume increases only due to the hypertrophy of alveolar cells.

Three-dimensional deformity of the thorax associated with congenital scoliosis. The thorax consisting of the spine, ribs, and the sternum, can be regarded as the dynamic respiration chamber, which passively sustains primary respiration by providing a stable wall for the lungs expanding as the diaphragm goes down and ensures secondary respiration (vertical excursions of the ribs using secondary respiratory muscles). The symmetrical growth of the ribs and thoracic spine ensures normal lung growth. The thorax in healthy patients is characterized by the normal volume with adequate height, width, and depth of the chest cage and the ability to change this volume. The thoracic height is determined by the height of the thoracic spine, while width and depth are determined by the shape and position of ribs. The thorax needs to be stable in order to maintain the normal volume.

The ability to change its volume (thoracic function) depends on stability of the diaphragm attached to it at the level of the T12 vertebra, and active respiratory excursions of the ribs ensured by intercostal muscles and symmetry of the right and left hemithoraces. Congenital abnormalities changing the thoracic volume and function may cause the development of thoracic insufficiency syndrome.
The analysis of spine deformity in patients with congenital scoliosis in terms of three-dimensionality of the thorax gives grounds to regard the spinal column as a deformed dorsal support of the dynamic three-dimensional structure. The primary deformity of the thorax results from congenital rib fusion, which directly limits lung growth, or the absence of ribs and, correspondingly, thoracic instability and respiratory impairment. Curvature, rotation, and shortening of the thoracic spine may result in secondary rib deformity and abnormality of both volume and function of the thorax. Severe distortion of the thoracic shape due to vertebral rotation causes restrictive lung disease associated with external factors and the reduction of the hemithorax volume during the formation of the rib hump and limitation of rib mobility. This condition results in unilateral loss of the secondary breathing mechanism. If rotation is accompanied by thoracic hyperlordosis in this case, the ribs are “furled” around the spine like a sail around a mast. The hump acquires the leptokurtic shape; the pleural cavity becomes virtually slit-like; lung volume and rib mobility are severely disturbed. It is difficult to assess this three-dimensional asymmetry of the thorax from the conventional radiographs; the image of the deformed thorax in horizontal plane obtained using the more informative CT scanning is figuratively referred to in English-language literature as windswept thorax (Fig. 2).

By analogy with the abnormalities of vertebral formation and segmentation, unsegmented vertebrae accompanied by rib fusion should be classified as the unilateral anomaly of thoracic segmentation (Fig. 3), while hemivertebrae together with the absence of ribs should be classified as unilateral anomaly of thoracic formation (Fig. 4). In turn, the anatomical portions of the thorax corresponding to the rib block zone can be regarded as the zone of segmental hemithoracic hypoplasia; the hemithorax corresponding to the concave side of the scoliotic deformity and contralateral hemithorax are known as concave and convex, respectively.

The term “jumbled spine” can be used to describe a severe scoliotic deformity [12]; however, Campbell et al. [6] believe that the concept “jumbled thorax” more accurately characterizes the three-dimensional deformity.

Any spinal or rib malformation, which is accompanied by a decrease in the thoracic volume during the first years of life, has a negative effect on the lung size that is anatomically restricted by the thorax by the time the growth is finished. The
lower vital capacity of the lungs in patients with congenital scoliosis as compared to that in patients with idiopathic scoliosis of the same degree can be attributed to the presence of thoracic abnormalities.

Congenital scoliosis can be rapidly aggravated, in particular in the presence of a unilateral unsegmented bar; the coexistence of the rib fusion accelerates this process. The methods for surgical treatment conventionally used in these patients (anterior epiphysiodesis and/or dorsal fusion at the convex side of the curvature, removal of a hemivertebra or osteotomy of the spine) do not solve the problem of blocked ribs and its circumstances. Almost no data are known about the natural progression of thoracic deformities in patients with these malformations, although our own experience attests to severe disability and almost complete incurability of patients as early as they reach 12–15 years of age (Fig. 5). The question regarding the effect of spine surgeries on the progression of thoracic deformity and rib growth remains poorly studied. According to the data obtained by Emans et al. [11], fusion performed for at least five functional spinal units in children aged below 5 years subsequently results in a 30–79 % decrease in pulmonary function as compared to the age norm. The inverse relationship between the age at which the surgery was performed and the degree of the subsequent abnormalities has been detected in this case. The scoliosis caused by the rib block without spinal abnormalities has been described [8]. Campbell et al. believe that in cases of extensive rib fusion, the concave hemithorax serves as a powerful lateral tether to spinal growth, acting with great mechanical advantage to further unbalance the growth of a spine that is already deformed by asymmetric vertebral growth [6]. Correction of spine deformity has a certain effect but does not improve the situation with the blocked ribs.

Examination methods. Examination of a patient with thorax deformity is based on the history data (development of respiratory symptoms), physical examination, plain radiography, CT scanning, studying the pulmonary function, and laboratory tests.
When collecting the history data, special attention is paid to patient’s fatigability, frequency of respiratory infections, and need for oxygen support. Deformity is typically determined by chest palpation; a blocked rib region is usually shortened both longitudinally and transversely as compared to the opposite side. The semi-circumference of the chest is measured at the nipple line; the change in this value with respiration shows the degree of the active thoracic function of secondary respiration. The degree of unilateral loss of secondary breathing can be determined clinically with the thumb excursion test (Fig. 6). Areas of flail chest and their paradoxical excursions correspond to the zone where the ribs are absent.

Thoracic lordosis, whose development reduces the sagittal diameter of the thorax, can be determined in profile radiographs. It is better to use CT scans and three-dimensional CT reconstructions to study vertebral rotation and thorax deformity.

The height of the thoracic spine corresponding to the thoracic height is measured using X-ray films and is compared to the normal indicators in order to determine the possible future shortening of the thorax [9]. The scoliotic

Fig. 6
Thumb excursion test [6]: physician’s palms embrace the lower half of the thorax; forefingers are located more ventrally from the anterior axillary line; forefinger tips are placed to the right and left at equal distances from the spine; the patient takes a deep breath; the distance by which each thumb has laterally moved is measured (+0 – no shift; +1 – shift by < 0.5 cm; +2 – shift by 0.5–1.0 cm; +3 – shift > 1.0 cm); the higher the shift degree, the more clinically pronounced the secondary breathing mechanism in the corresponding hemithorax is (+3 at the right side and +1 at the left side)

Fig. 7
Procedure for determining the space available for the lungs: hemithoracic height is the distance between the midpoint of the most cranial rib and the midpoint of the cupula of diaphragm (line A); the ratio expressed as a percentage is obtained dividing the height of the concave hemithorax by the height of convex hemithorax; in another version, the points of ribs (equally-spaced by the width of the corresponding vertebral body X from the spine) with the more caudal location as compared to the diaphragm (line B); deterioration of the index of total lung capacity in consecutive radiographs attests to the suppression of longitudinal growth of the concave hemithorax with progression of spine deformity [6]
curve angle to a certain extent correlates with the deformity of the concave hemithorax: higher scoliotic Cobb angle means that the height of the hemithorax is reduced to a more significant extent. The quantitative assessment of the degree of reduction of the hemithorax on the concave side of the deformity is characterized by the ratio between the heights of the concave and convex hemithoraces (expressed as a percentage), which is known as “the ratio between the spaces available for the lung” (Fig. 7).

The value of CT scanning consists in the ability to supplement the plain radiography with a large number of objective quantitative indicators to better understand the three-dimensionality of thoracic deformity and to assess its space–time dynamics, including that in patients who receive surgical treatment.

The degree of loss of hemithoracic symmetry can be characterized on the basis of the CT data by calculating the coefficient of dorsal hemithoracic symmetry (Fig. 8a), which makes it possible to early detect the “windswept thorax”. The other methods for CT diagnostics of thoracic asymmetry include measuring the angle of thoracic rotation formed by the sagittal plane of the spine connecting the midpoints of the anterior vertebral surface and the sternum (Fig. 8b) and the angle of spine rotation (the modified Aaron-Dahlborn procedure) [3] (Fig. 8c).

Relying upon the fact that hemithorax conventionally is a truncated pyramidal cone, S.O. Ryabykh [2] proposed to determine its volume by calculating the product of its height, width, and the anteroposterior dimension. This method can be used if the CT and/or MRI data for the thoracic spine are available (under the condition

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**Fig. 8**

Characteristics of the degree of symmetry loss [6]: a – dorsal hemithoracic symmetry ratio (a line is drawn through the tops of heads of ribs articulating with the vertebral bodies; the distances between the spine and the inner borders of the hemithoraces are measured, and the larger one is divided by the smaller one); in the normal condition, the ratio is equal to 1, but it increases with the emergence of a hump, indicating the onset of development of the thorax deformity; b – thoracic rotation (angle θ) is measured between the line dividing the chest cage into two hemithoraces, and the midsagittal plane of the vertebral body; the higher rotation, the smaller thoracic symmetry is; c – spinal rotation (angle θ) is measured between the sagittal plane of the apical vertebra (the perpendicular to the line connecting the heads of ribs) and the line perpendicular to the lower edge of the scan.
that the scan of the complete thorax is obtained. The thoracic height is measured by drawing a vertical line from the midpoint of the first rib to the point on the lowest rib located at a distance equal to the width of the lowest thoracic vertebra from the spine. If the lowest rib is rudimentary and is shorter than the vertebral width, the lower boundary point for the vertical axis will localize outside the rib on its conventional extension line. Hemithoracic width is measured by the value of horizontal line connecting the spine edge with the outer rib edge in the widest point of the corresponding hemithorax. The anteroposterior dimension is measured along the line connecting the anterior edge of the sternum and the spinous process in the widest hemithoracic portion (Fig. 9). The conditional character of this stereometric calculation (under actual conditions, the shape of hemithoraces is more sophisticated than that of truncated cones) smoothens the determination of the thoracic asymmetry index (ThAI) represented by the ratio between the hemithoracic volumes for the concave (a) and convex (b) sides:

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\text{ThAI} = \frac{(Ha \times Ra \times Ga)}{(Hb \times Rb \times Gb)},
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where H is the height; R is the width; and G is the anteroposterior dimension.

The reduced sagittal diameter of the thorax showing the loss of the thoracic volume and symmetry is assessed using the spinal penetration index proposed by Dubousset et al. [10]. After analyzing the CT scans, the authors have formulated the concept of spine deformity as an endothoracic deformity resulting from protrusion (penetration) of the vertebral bodies into the thorax and forming the so-called endothoracic vertebral hump. The spinal penetration index is determined as follows: a line tangent to the posterior surface of vertebral arches on the concave and convex sides of the deformity is drawn for each transversal CT cross-section. The area limited by the rib contour and by the tangent line is normally formed by the sum of areas of the transversal cross-section of the spine with the adjacent paravertebral tissues and the thoracic cavity. The spinal penetration index is determined as the ratio between the area of the spine protruding toward the thorax and the total area of the cross-section. The index calculated for each consecutive cross-section for the entire height of the thorax determines the portion of the thoracic volume occupied by vertebrae. In the normal condition, this index is 8–10%; however, in patients with lordoscoliosis it may increase to 15, 20, and even 50% (Fig. 10).

The effectiveness of spine deformity correction can be assessed using the so-called interpedicular ratio: the terminal vertebrae of the scoliotic arch are selected and the midpoints of the medial surfaces of the shadows of arch pedicles are marked on them as points; straight lines are drawn between the corresponding points of the
upper and lower vertebrae in order to form the theoretical interpedicular line; the most laterally deviated pedicle is marked, and the distance from this point to the contralateral interpedicular line (B) is measured in mm; the distance between both interpedicular lines (A) is measured; the resulting index is the ratio between B and A and characterizes the lateral shift of the apical vertebra towards the theoretical interpedicular distance at this level (Fig. 11). Progression of scoliotic deformity increases this index, while deformity correction due to treatment reduces it. When the spine is virtually straight, the index is equal to 1. In the case under consideration, deformity correction reduced the index from 2.8 to 1.3 [7].

The conventional tests are used to assess the pulmonary function. The decrease in vital capacity (VC) of the lungs is a result of a number of factors: the primary loss of pulmonary tissue due to fibrosis (after recurrent infections or a congenital disease), the effect of respiratory tract lesions caused by asthma, secondary changes because of the reduction of thoracic volume and function, patient’s ability to adequately respond to the examination (which is usually absent in children younger than 5 years). The VC index is typically compared to the normal age index based on the data of patient’s height in the standing position. If child’s height is significantly reduced due to scoliotic spine deformity, the VC index may turn out to be erroneously high. The more reliable
index is calculated with allowance for arm span. The thoracic volume can be also determined using CT scanning; however, there can be some errors as well, even if high-speed spiral tomography machine is used, since the lower edge of the lungs constantly shifts with breathing.

The other methods for studying the pulmonary function may also yield erroneous results. Pulse oximetry is relatively poorly sensitive to small changes in blood oxygen level. The investigation of gases in arterial blood can be inaccurate because of hyperventilation in a crying child.

The full-fledged clinical and radial examinations together with the data of the pulmonary function tests allow one to obtain the comprehensive picture of the pathological process, to assess its dynamics, and to select the adequate treatment strategy.

We believe that the development of the thoracic insufficiency syndrome is possible in patients not only with congenital scoliosis, but also with the deformities of different etiologies; this fact should be borne in mind by spinal surgeons who deal with managing this pathology in patients.

References


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