VERTEBRAL DEFORMITY ASSOCIATED WITH THORACIC SPINE SEGMENTATION DISORDER IN NEWBORNS AND INFANTS DURING THEIR FIRST SIX MONTHS OF LIFE*

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Objective. To analyze clinical features, X-ray anatomy and dynamics of deformities of the spine and thorax in newborns and infants during their first six months of life with vertebral segmentation disorders.
Material and Methods. Twenty four children in neonatal period and 8 children in the first 5 months of their lives were examined. X-ray anatomy of malformation, dynamics of congenital spinal deformity, intervertebral disc and spinal canal status were studied. Thorax development pattern, and dynamics of congenital vertebral block and spinal deformity were followed in 15 children aged between 2–8 years.
Results. Most children with vertebral segmentation disorders were born with grade III–IV spinal deformity. An asymmetric congenital block was detected almost in 70 % of patients. This malformation enables rapid progression of scoliosis in more than 1/3 of patients. True bone block was observed in 3 % of cases; other patients of this age group had either fibrocartilaginous or combined block. Gradual maturation and ossification of this block is observed. Asymmetric growth of the spine with extended congenital vertebral block (five or more segments) results in formation of grade III–IV spinal deformity in most newborns.
Conclusion. Spinal deformity associated with vertebral segmentation disorder in infants of the first weeks and months of life offers a challenge of passing from passive observation of such patients to active orthopedic treatment at an early age.
Key Words: vertebral segmentation disorders in the thoracic spine, in newborns and infants during their first six months of life, congenital spinal deformity, asymmetric growth of the spine.

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This paper is dedicated to the memory of our teacher and one of the pioneers of surgical neonatology in the USSR, the Corresponding Member of the Academy of Medical Sciences, Prof. G.A. Bairov

Congenital segmentation disorder (also known as congenital block, which is less correct in pathogenetic terms) of the thoracic vertebrae is a malformation causing severe and usually rapidly progressing deformities that require orthopedic treatment during the entire growth period, considerably worsen the quality of life, and are virtually surgically incorrectable. This anomaly is frequently accompanied by severe malformations in the internal organs, extremities and spinal cord [1]; the clinical manifestations of these malformations become predominant and make it impossible to timely correct the spinal deformity. Most studies devoted to vertebral segmentation disorder focus on the age periods when the spinal deformities formed are almost incurable [1–4, 6–9, 11–14].

This article was aimed at analyzing the clinical features, the X-ray anatomy presentation and dynamics of spine and thorax deformity in newborns and infants during their first six months of life with segmentation disorder.

Material and Methods

The data of 32 children with segmentation disorder in the thoracic spine, including 24 newborns (1–30 days old) and 8 infants with age varied from 43 days to 5 months, were used in this study. All patients underwent X-ray of the spine in two projections. MRI and CT of the spine were performed on two and one of the patients, respectively. It was impossible to perform a more thorough examination because of the necessity to use anesthesia, while the overall condition of the patients was severe. All the children were personally examined by the authors of this article.

The following groups of indicators were analyzed during the study:

1) clinical indicators: the features of perinatal history, physical development of children and concomitant anomalies;

2) X-ray indicators: the initial value of the major deformity curve; presence of counter curves, the sagittal profile of the thoracic spine, and position of the pelvis; the features of the main malforma-

tion (type and length of vertebral and rib blocks, disc status and the size of the base of curves near the block and in the adjacent segments, and the width of the spinal canal at the level of the block) were examined more thoroughly.

The scoliotic angle was determined using the Cobb procedure; the degree of deformity was assessed according to V.D. Chaklin's classification [5]. The thorax status was additionally assessed according to the left and right hemithoracic volume, the presence and degree of pulmonary hypoplasia. The dynamics of the aforementioned indicators were followed in 15 children aged between 2–8 years.

Results

Most children (84.0%) were born full term; the body weight of 71.8% lied within the normal range. Concomitant anomalies were detected in 14 (43.7%) patients; the average number of these anomalies being 1.7 per patient (Table 1).

Three children died under the age of 1 month after surgeries performed for severe concomitant anomalies.

Clinical examination revealed a relative shortening of the body in all children. In patients with unilateral segmentation disorders, the thorax was asymmetric and had defects associated with rib aplasia or underdevelopment of the ribs; rigid scoliosis curve with the gibbus deformity and, less frequently, kyphosis of the thoracic segment were observed (Fig. 1).

Grade I-II respiratory failure of a restrictive type was detected in 13 (40.6%) patients.

The scoliotic angle varied from 3 to 69o with the prevalence of severe deformities: grade III and IV scoliosis was found in 16 (50.0%) and 2 (6.3%) children, respectively; grade I and II scoliosis was detected in 6 (18.7%) and 8 (25.0%) children, respectively. Grade II-IV deformities could be visually detected as early as in newborns, while grade I scoliosis was usually revealed incidentally when performing X-ray examination for concomitant malformations.

Table 1
Concomitant malformations in children with the disorder of thoracic vertebral segmentation

Malformation type	Patients, n (%)
Esophageal atresia	7 (21.9)
Anorectal malformations	3 (9.4)
Renal agenesis	3 (9.4)
Urinary tract malformations (except for renal agenesis)	2 (6.2)
Pulmonary hypoplasia	2 (6.2)
Micrognathia	2 (6.2)
Relaxation of the cupula of the diaphragm	1 (3.1)
Medullispinal hernia	1 (3.1)
Radial clubhand	1 (3.1)
Fallot's tetrad	1 (3.1)

Fig. 1
Appearance and radiograms of 6-year-old patient V. with congenital scoliosis associated with T3–T12 segmentation disorder: **a** – right-sided gibbus is shown with an oval; **b** – spina bifida of the bodies and arches in the upper eight thoracic vertebrae, non-segmented T3–T10 block on the left side represented by partially ossified cartilages, and bone block of ribs at the level where the non-segmented spinal column localizes are visualized

The congenital vertebral block in all patients localized within the T2–T12 region and comprised 3–11 segments: blocks of less than five segments were detected in 13 (40.6%) children; blocks of 5–10 segments were detected in 17 (53.1%) children; and blocks of more than 10 segments were found in 2 (6.3%) children. The deformity apex usually localized in the center of the block, while the primary curve angle clearly correlated with its length. Among 19 children with the block of more than 5 segments, the initial scoliotic deformity corresponded to grades III and IV in 10 and 2 children, respectively.

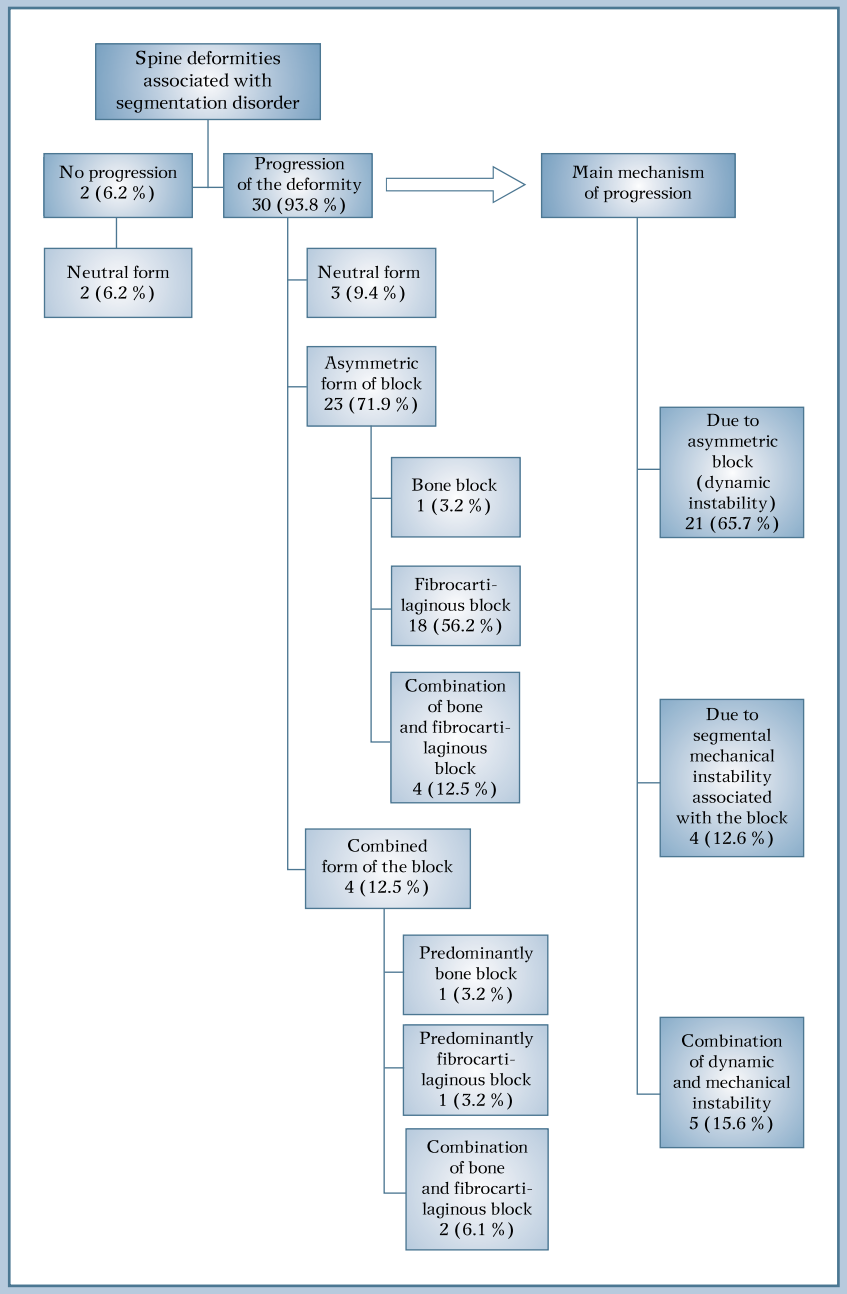

Fig. 2
Dependence of the aggravation in spinal deformity on the shape of vertebral block

An increase in the number of blocked segments (the so-called maturation of the block) during the dynamic follow-up study was observed in three cases; length of the block increased by two and three segments in one and two cases, respectively. This was accompanied by a 31–39% increase in deformity, which resulted in aggravation of the deformity four years later: from grade III to grade IV in two patients and from grade I to grade III in one patient.

In the group under study, the asymmetric forms of vertebral block significantly predominated over the neutral ones and were observed in 26 (81.3%) and 6 (18.8%) of cases, respectively. The right- and left side non-segmented blocks were detected in 14 (43.8%) and 12 (37.5%) of patients, respectively. The neutral shape of the complete vertebral block was diagnosed in one (3.1%) patient.

The Klippel–Feil syndrome was detected in seven (21.9%) children; however, its rate is underestimated, since many patients did not undergo X-ray examination of the cervical spine.

The sagittal profile changes were observed in all 32 patients. The flat-spine variant predominated in 24 (75.0%) of children; lordosis up to 21o due to block of the posterior column was revealed in one (3.1%) patient. The 3–33o kyphotic component associated with grade III–IV scoliosis was observed in seven cases (21.9%).

The counter curves that were formed independently of patients' verticalization and developed either antenatally or during the first few weeks of life were detected in 10 children, including counter curves in the cervicothoracic spine (C6–T5) in two patients and those in the thoracolumbar spine (T6–L5) in eight patients. It should be mentioned that no compensatory (secondary) pelvic distortion caused by the block of thoracic vertebrae was observed. The presence of compensatory pelvic distortion in three patients was attributed to the concomitant anomalies of formation and fusion of the thoracic vertebral bodies and the low boundary of the caudal end of the scoliosis curve.

The features of X-ray presentation of vertebral segmentation disorder. Three types of vertebral segmentation disorder were distinguished: bone, fibrocartilaginous, and mixed ones. It is of interest that the pure bone block in newborns was detected only in one (3.1 %) case, while being observed more frequently in infants. The fibrocartilaginous block (the asymmetric block with poor ossification) was detected in seven (21.9 %) patients; the mixed type with the regions of well-pronounced bone and fibrocartilaginous block was found in 24 (75.0 %) patients.

The progression of the deformity was determined by the shape of vertebral block. In case of neutral shape, the deformity did not increase, while spinal curvature usually was not very significant. In case of asymmetric or combined forms of the anomaly (69.0 %), the deformities aggravated, which was caused by the asymmetric growth of the vertebrae (the aggravation rate was 4–13° per year). If the vertebrae with the normally developed intervertebral discs (according to visual examination) were blocked at two levels, rapid progression of the deformity was detected at the level of the disc, which can be attributed to combination of the dynamic (growth asymmetry) and segmental instability (Fig. 2).

One of the objectives of this study was to examine the X-ray indicators of the intervertebral discs within the blocked area and in the adjacent area. An analysis of the X-ray data allowed us to distinguish the types of anomalies of the intervertebral discs: vertebral aplasia and hypoplasia.

Disc aplasia is accompanied by bone block of the intervertebral bodies over the entire cross-section area and is classified as a neutral type. If disc aplasia is extensive, spine deformities either do not occur or are insignificant and do not progress during the growth process (Fig. 3a).

Disc hypoplasia has three anatomical subtypes:

1) uniform hypoplasia of the disc over the entire surface area of the bodies of the upper and lower vertebrae (Fig. 3b); the X-ray presentation of the mild subtype of hypoplasia appears as a disc with a significant (most typically uniform) decrease in height and either a rudimentary or absent nucleus pulposus. This subtype is frequently the only sign of segmentation disorder that is not accompanied by bone or fibrocartilaginous vertebral block whose growth is not disturbed;

2) partial disc hypoplasia (Fig. 3c) always accompanies the asymmetric block of vertebral bodies; the major or minor portion of the disc localizes on the side opposite to the block and contains no nucleus pulposus; epiphyseal plates of the vertebrae around the defective disc continue functioning, which results in a deformity that inevitably progresses during growth;

3) alternating hypoplasia (Fig. 3d) is observed in the cases when the partial anomaly on one side is compensated for by a similar malformation on the other side; it is how one of the types of neutral block that is not accompanied by rapid progression of deformity is formed.

Almost all patients had a combination of the anatomic subtypes of disc hypoplasia; however, symmetric partial hypoplasia was the predominant subtype.

The X-ray parameters of the vertebral arches (width and height of their base) within the block area and in the boundary lower thoracic segments were studied in all 32 children in order to substantiate the possibility of using instrumental fixation of the spine. In patients aged younger than 1 month, the area of arch base at the block apex varied from 1 to 9 mm^2; from 1 to 9 mm^2 and from 3 to 6 mm^2 in patients younger than 3 and 5 months, respectively. The area of arch base of the adjacent vertebrae varied from 4 to 12 mm^2 and from 6 to 12 mm^2 in patients younger than 5 months. Thus, no age correlation with the area of the vertebral arch base in patients during the first 5 months of their lives was detected. Only the minimal and maximum size are given, while the significant discrepancy between the indicators is most likely to

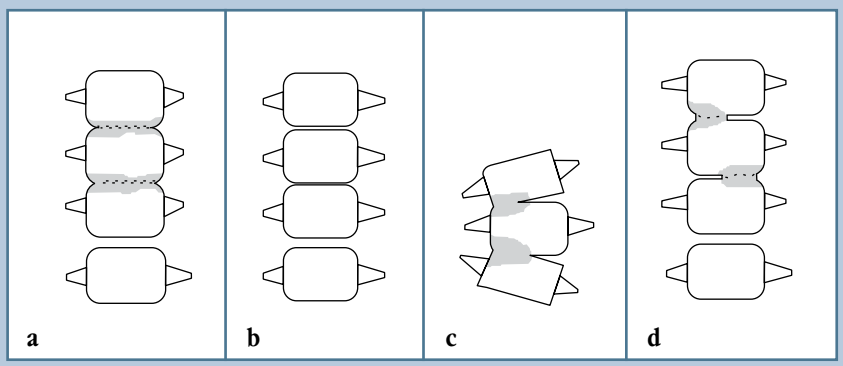

Fig. 3
Types of anomalies of the development of intervertebral discs: **a** – disc aplasia; **b** – symmetric hypoplasia of the discs; **c** – partial hypoplasia of the discs; **d** – alternating hypoplasia of the discs

Table 2
Average size of the base of vertebral arches in the examined patients (n = 32)

Area measured	Width, mm		Height, mm		Area, mm^2	
	right side	left side	right side	left side	right side	left side
Block apex (T4–T6)	2.2	2.2	2.3	2.4	5.4	5.5
Segments adjacent to the block (T9–L1)	2.7	2.7	3.2	3.2	8.6	8.7

be caused by the features of malformation anatomy. The mean size of the base of vertebral arches is listed in Table 2.

Since the segmentation disorder is frequently (up to 75.0% among patients older than 7 years) combined with spinal dysraphia, including diastematomyelia [1], the width of the spinal canal was studied using the radiograms.

In order to give definition to the term "normal width of the spine canal", the interpedicular distance at the level of the T6 vertebra was measured in two control groups consisting of 10 children without malformations of the axial skeleton. Group 1 was composed of full-term newborns; group 2 consisted of children aged 1.5–3 months. The frontal width of the spinal canal in group 1 fluctuated between 10 and 12 mm; that in group 2 was 11–13 mm [1].

In children with disorder of thoracic vertebral segmentation, the width of the spinal canal varied from 9 to 18 mm, being 10–13 mm in 22 (68.8%) patients. In two cases (children aged 26 days and 1 month 12 days), the interpedicular distance was 15 and 18 mm, respectively. This fact, along with the signs of hypotrophy of the lower limb, was an indication for MRI: diastematomyelia was detected in one child, while spinal cord lipoma was revealed in the other one. In addition, diastematomyelia was diagnosed two years later in two children with the normal width of the spinal canal during the neonatal period. These data should be taken into account when planning a spinal deformity correction surgery; the necessity of MRI or CT examination should be always substantiated despite the demand for an additional anesthesia.

The rib and thoracic malformations aggravate the clinical presentation of spine segmentation disorder [10]. The structure of anomaly types included blocks and aplasia (agenesis) of the ribs; most patients had a combination of different types of rib anomalies. Right side rib block was predominant: the average number of ribs blocked in the right and left hemithoraces was 5.5 and 5, respectively. Right- and left side agenesis of a part of ribs was observed in four and five patients, respectively.

One of the authors of this article, S.O. Ryabykh, has proposed to use the method for calculating the hemithoracic volume in order to determine and compare the volume of the right and left hemithoraces: the volume was calculated as the product of height (H), width (R), and the anteroposterior dimension (G). Furthermore, the thorax asymmetry index (ThAI) (the ratio between the volumes of the hemithoraces on the convex (a) side and the concave (b) side) was also proposed:

$$ThAI = \frac{Ha \times Ra \times Ga}{Hb \times Rb \times Gb}.$$

In children with segmentation disorder, the ThAI varied from 0.45 to 0.85 at N = 0.9–1.0.

The follow-up study of the natural course of the deformity in 15 children aged between 2–8 years has demonstrated that it progressed in 13 (86.6 %) of cases, including moderate aggravation up to 2° per year in 4 (12.5 %) patients and rapid aggravation with annual increment from 4.4 to 13° in 11 (34.4%) patients (Fig. 4).

The dynamics of indicators of progression of spine and thorax deformity are shown in Table 3.

An increase in block length was observed in three children. The ThAI remained stable as children grew in 5 (33.3 %) of cases; decreased in 5 (33.3 %) of cases; and increased in 5 (33.3 %) of cases.

Vertebral segmentation disorder is a relatively rare developmental malformation. However, these deformities are characterized by III and IV grade severity during the first few weeks of life in most cases (56.0%), which can be attributed to asymmetric intrauterine growth and asymmetric block of five and more vertebrae. In some patients the deformities are accompanied by the formation of counter curves already after they are born.

These data demonstrate that the malformation is predominantly represented

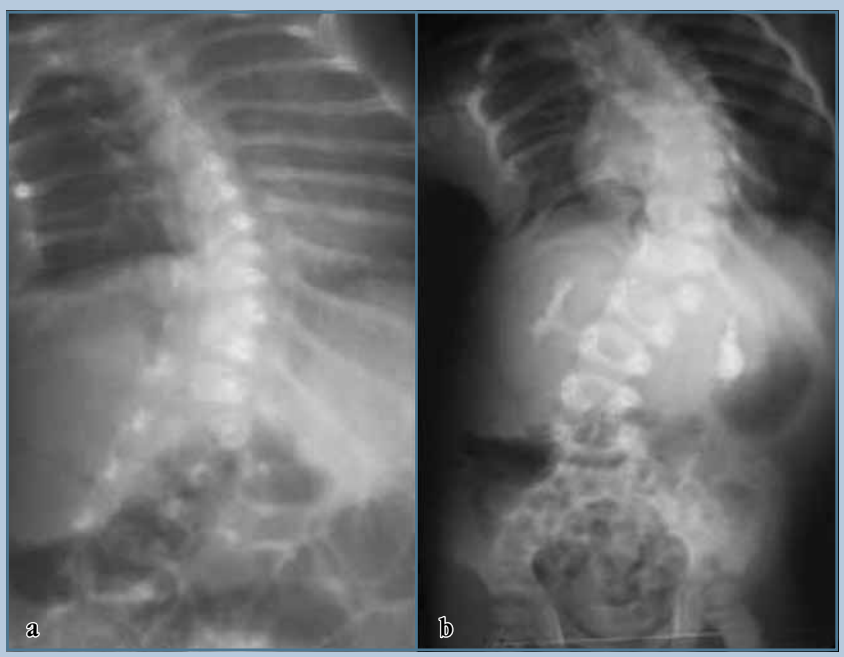

Fig. 4
Radiograms of patient M. with deformity progression associated with segmentation disorder in the thoracic spine: **a** – 1 month of age: disorder of segmentation at T3–T11 and formation of scoliotic curve (40°); **b** – 3 years of age: deformity progression – 44°; scoliotic deformity 78°

Table 3
Dynamics of the indicators of spine and thoracic deformity in the patients followed up

Patients	Follow up period, years	Deformity angle, deg.			Number of segments per block, n		Thorax asymmetry index		
		initial	final	gain, %	initial	in the follow up period	initial	in the follow up period	gain, %
1st	2.0	21	23	+4.3	5	5	0.90	0.70	+22.3
2nd	6.0	38	62	+39.0	9	12 (+3)	0.80	0.80	0
3rd	4.0	11	33	+44.0	7	7	0.90	0.60	+33.3
4th	2.0	28	32	+12.5	8	8	0.60	0.70	-14.3
5th	2.5	33	58	+43.0	9	9	0.70	0.70	0
6th	6.0	30	66	+54.0	7	8 (+1)	0.85	0.85	0
7th	8.0	14	14	0	4	4	0.72	0.72	0
8th	5.0	9	35	+74.0	5	8 (+3)	0.70	0.67	+4.3
9th	3.5	69	71	+2.8	9	9	0.90	0.90	0
10th	2.0	22	23	+4.3	9	9	0.65	0.69	-5.8
11th	2.5	29	29	0	10	10	0.72	0.82	-2.2
12th	3.5	36	36	0	4	4	0.87	0.80	+8.1
13th	4.5	37	57	+35.0	10	10	0.54	0.70	-22.9
14th	3.0	40	74	+85.0	8	8	0.52	0.70	-25.7
15th	4.0	44	64	+31.0	3	5 (+2)	0.60	0.43	+8.4

by the cartilaginous or fibrocartilaginous types of block in most children at birth. The true bone block develops much later. The fact that children of early age have no true bone block gives grounds for expecting a good outcome of the correction therapy during the early age. However, the lack of proper knowledge among pediatricians, pediatric surgeons and orthopedists, as well as detection of concomitant malformations requiring a long-term surgical treatment (sometimes comprising several stages), results in the fact that patients are referred to a pediatric vertebrologist very late.

Conclusion

The knowledge of the features of thoracic spine segmentation disorder in newborns and infants during their first six months of life offers a challenge of passing from passive observation of such patients to active orthopedic treatment at an early age.

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