RARE FORMS OF CLINICALLY SIGNIFICANT SCOLIOSIS

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Abstract: *This paper describes rare forms of scoliosis, their clinical manifestations, the causal factors behind their development, and treatment methods, focusing on preventive measures. All of this was described based on a literature review.*
Keywords: *scoliosis, groups, children, deformity, analysis, signs.*

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Annotatsiya. *Ushbu maqolada skolioz kasalligining kam uchraydigan shakillari, ularning klinik ko'rinishi, kelib chiqish sabablari va konservativ davolash usullari yoritilgan. Kasallikni kelib chiqishini oldini olish chora tadbirlariga urg'u berilgan. Maqola adabiyot malumotlariga asoslangan.*
Kalit so'zlar. *Skolioz, gurux, bolalar, deformatsiya, analiz, belgilar.*

РЕДКИЕ ФОРМЫ КЛИНИЧЕСКИ ЗНАЧИМОГО СКОЛИОЗА

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Аннотация: *в данной статье описываются редкие формы сколиоза, их клинические проявления, факторы лежащие в основе их развития и методы лечения. Кроме приведен акцент на профилактические меры. Все это описано на основе обзора литературы.*
Ключевые слова: *сколиоз, группы, дети, деформация, анализ, признаки.*

"Early-Onset Scoliosis" refers to scoliosis with early onset or early manifestation (EOM), a range of scoliotic deformities with different etiologies but rapidly progressing in early childhood, such as infantile and juvenile idiopathic scoliosis, congenital spinal anomalies, and a large group of associated scolioses requiring early surgical treatment of curvatures [Akbarnia B.A et al., 2005; 2010].

Grouped according to the early childhood manifestation, infantile IS from 1 to 3 years, and juvenile - from 5 to 8 years [James J.I-1954], such curvatures, in contrast to deformations in IS, pose significant risks to the health of the child, presenting many problems not only to vertebrologists, but also to doctors of other specialties [Campbell R.M; 2003, Canavese F., Dimeglio A., et al. 2007, Umarkhodzhaev F.R; 2021]. Considering that the prevalence of infantile and juvenile IS in the population of 6-7 year old schoolchildren with a scoliotic arc of 10° or more may be 0.05% for girls and 0.02% for boys [52], the share of SRM may account for about 1% of all IS registered in the USA, of which about 10% of those tolerant to conservative treatment will require early surgical treatment for scoliosis [7,54]. In contrast to IS, SRM affects boys more often than girls (3:2), and 75%-90% of deformities are represented by left-sided curvatures [36]. Congenital deformities of the

spine and congenital scoliosis (CS). According to most researchers, congenital curvatures are defined as axial curvatures: kyphosis, scoliosis, lordosis, and kyphoscoliosis, which are the inevitable result of anomalies and asymmetric growth of the vertebrae and/or ribs, which have an irreversible "global" effect not only on the spine and rib cage, but on virtually all visceral organs [19, 9, 13, 7, 33].

The estimated prevalence of vertebral malformations in childhood, according to various sources, ranges from 0.5–1:1000 to 1:100 or more [4, 9]. Among axial spinal deformities, curvatures due to anomalies are detected in 2–11% of cases, 50% of which are detected in the first year of life. Among the latter, clinically significant cases account for 30%. By the age of three, severe forms of curvature already account for 50%, although no progression was noted in approximately 18% of cases [4]. Scoliotic deformities or SC, in the structure of congenital axial deformities of the spine, are detected much more often, up to 75% of cases, than "pure" congenital kyphosis [4, 37]. The most common spinal anomalies that provoke scoliosis are considered to be multiple underdevelopment of half a vertebra (44%). In second place is a segmentation defect (32%), and at the bottom of the list is a single hemivertebra (18%) [4]. Congenital deformities can be classified based on localization, the nature of the curvature, or the type of anomaly that led to the development of axial deformity. Depending on the type of vertebral body anomaly observed on radiographs, a two-dimensional classification of congenital scoliosis is used in clinical practice. Three types are distinguished: Type I - formation disorder; Type II – segmentation disorders, and type III – mixed, combining the two previous types of anomalies [4]. The greatest progression of VS occurs according to general rules, during two periods of rapid growth, during the first 3 years of life and in the adolescent growth spurt, therefore, a patient with an earlier onset and a longer history of the disease requires close attention from surgeons [15]. It should be noted that type I VS, due to anomalies of the lateral and posterolateral quadrants of the hemivertebral body, progresses by an average of 2.5° up to the age of 10 years, and then by 5° per year [33]. Posterior and lateral hemivertebrae have a slightly better prognosis, followed by butterfly and wedge-shaped vertebrae [35]. VS of the upper thoracic spine, due to underdevelopment of a hemivertebra, will progress by 1°-2.5° per year, causing a tilt of the shoulders, while a similar anomaly in the lower thoracic location causes a more rapid and greater progression of 2° to 3° annually [33]. Segmentation disorders or type II anomalies causing VS have a high risk of progression from 2° to 6.5° per year in the thoracic spine, between 6° and 9° in the thoracolumbar spine and about 5° per year in the lumbar region over the same period [33]. Anterior fusions show a slow progression of deformity of about 1% per year up to 10 years, while anterolateral ones had a worse prognosis [35]. Type III is the least common, includes type I and type II defects, and is considered the most aggressive malformation with a huge potential for rapid progression of scoliosis and kyphoscoliosis, 5° per year up to 10 years of age and 10° at older ages. In practice, such deformities present the greatest difficulties in treatment, either due to severe deformity and neurological impairment, or as deformities prone to significant progression [35, 33]. Scoliotic deformities associated with neurofibromatosis (NF) are one of the most common (1:3000-3500 newborns) genetically determined human diseases, affecting an equal number of men and women, a total of about 1,000,000 [44]. There are two clinical forms of this disease: NF type 1 (NF1) or peripheral, better known as Recklinghausen's disease, which accounts for up to 85% of all patients suffering from NF. Neurofibromatosis type 2, or central (NF2), is less common and is characterized by the development of tumors of the auditory or optic nerves and other intracranial and intravertebral neoplasms [12, 44]. Characteristic clinical symptoms of NF are slowly progressive lesions of the nervous system, soft tissues, mainly the skin, and abnormalities of the musculoskeletal system, among which severe spinal deformities are particularly prominent [4, 21, 6, 25]. Scoliotic deformities in NF are most often detected at the age of seven to sixteen years, 10%-60% of cases [11, 12, 6]. Scoliosis is more often detected in NF1, since scoliosis is often detected first and only after a detailed examination the diagnosis of NF is made [11]. All scoliotic deformities of the spine in neurofibromatosis are divided into dystrophic and non-dystrophic [11, 21]. Non-dystrophic curvatures are characterized by a benign course similar to idiopathic scoliosis, however, they can often transform into a dystrophic form [18]. Dystrophic curvatures are characterized by an acute-angled, rapidly progressing, on average by 5 degrees or more per year, short (4-6 vertebrae) kyphoscoliosis, usually with a predominance of hyperkyphosis, which in advanced cases can reach 180° or more [12, 21]. Classical manifestations of the dystrophic form of NF1 are: pronounced wedge-shaped deformation and rotation of the vertebrae, short arc, presence of kyphosis, sharpening of the "penciling" ribs, fusiform "spindling" deformation of the transverse processes, scalloping of the edges of the vertebral bodies, widening of the intervertebral foramina, meningocele and paravertebral tumors, spondyloretrolisthesis and spondyloptosis are common [21,34]. Surgical treatment of scoliotic and kyphotic curvatures of the spine in NF is associated with a large number of cases of pseudoarthrosis, progression of deformity and possible development of paraplegia [11, 12, 21]. The presence of degenerative bone changes in the spine, rigidity of the kyphotic

component of the deformity, osteoporosis, meningocele, neural tumors around the brain or nerve roots, anatomical anomalies, and the magnitude of the curvature, often reaching extreme angular values, place deformities associated with NF in the category of severe forms of scoliosis, with a more complex and dangerous tactic, often requiring vascularized transplantation. [12, 28, 52, 50].

Dystrophic spinal deformities associated with NF, compared with non-dystrophic ones, have a less favorable prognosis, and patients require earlier and more aggressive surgical intervention, starting at the age of 10 years [6]. According to American researchers, Marfan syndrome (MS) is detected in 1 in 10,000 surviving newborns [22]. The disease is inherited in an autosomal dominant manner, not linked to sex, with a 25% incidence of spontaneous mutation, and significant clinical variability ranging from mild dysplasia to life-threatening systemic disorders [40]. The hallmarks of Marfanoid habitus are well known. Manifesting in infancy, the disease is severe, and despite the best efforts of physicians in virtually every known specialty, the average life expectancy with SM does not exceed 43 years in men and 46 years in women [27]. Scoliotic deformities are detected in 58-63% of patients with SM. Of these, 2-2.5% are SRM, with kyphoscoliosis observed in approximately half of all SM cases [40, 45, 14].

Scoliosis in SM is similar to IS, but there are a number of fundamental differences. Almost always, early onset of scoliosis is recorded, a high prevalence of double thoracic and large triple curves [45], while 40% of patients have a kyphosis of more than 50 °. The above-mentioned features of the association of scoliosis with SM directly indicate the need to choose a more aggressive tactic for treating spinal deformities in such patients already in early childhood. When choosing a surgical tactic, it is necessary to note the disturbances of the respiratory function due to the "thoracic insufficiency" syndrome, which can be a consequence of the double action of not only the circular scoliotic deformity of the chest, but also an additional funnel-shaped or keel-shaped deformity in front [13]. Disturbances in the activity of the cardiovascular system in the complex of instrumental correction of scoliosis in SM are often fatal [14, 48]. Echocardiography, MRI to determine dilation of the root and ascending aorta, aortic or mitral valve prolapse, and beta-blockers to prevent complications associated with aortic dilation are indicated [14, 48]. Before and during surgery, it is necessary to involve a hematologist, conduct a thorough examination of the platelet count, hemostasis, and administer anticoagulation therapy [14]. Bone density in SM can also be reduced, which is aggravated in 63% to 92% of patients by thinning of the pedicles and arches of the vertebrae due to dural ectasia [14], which reduces the stability of instrumented fixation [14]. In order to maintain stability, it is recommended to use the maximum number of hooks, screws, tapes, and/or cement to better distribute the load. It is very important that the patient and his or her legal representatives be warned about the high rate of fatal complications, since 28.6% of patients die during treatment from cardiovascular complications not related to spinal surgery [43, 14]. In scoliosis surgery, complications related to deep infection account for approximately 10% of the complications, while dural ruptures and large blood loss volumes of 2400-5300 ml account for 8% [14, 26]. Scoliotic deformities associated with syringomyelia (SGM). Syringomyelia is classified as an organic disease of the central nervous system associated with neural tube developmental anomalies and anatomically characterized by glial proliferation and the formation of longitudinal cavities in the cervicothoracic spinal cord, brainstem, and internal capsule [2]. Most patients with CMS have a malformation of the cerebellar tonsils (Arnold-Chiari I malformation), which presumably disrupts cerebrospinal fluid dynamics and, as a result, the size of the central canal of the spinal cord increases, causing slow progression of myelopathy with an expansion of the central canal of the spinal cord of more than 1 mm over at least 2 vertebrae [32]. According to researchers, the prevalence of CMS varies significantly from 3.3 to 130 cases per 100,000 population [1]. The association of IS and CMS is considered the most common pathology, 11%-15% of all cases of IS [41, 16]. Approximately 2/3 of patients with CMS have curvatures of less than 20°, and only the remaining 4-5% have clinically significant curvatures of more than 20-25°. Of the latter cohort, patients with juvenile and infantile idiopathic scoliosis together account for more than half, 27% and 25%, respectively [30, 19, 17, 41].

In asymptomatic cases, approximately 14-27% of cases of SGM in scoliosis are detected incidentally [29, 41], whereas in the presence of neurological symptoms, SGM is detected in almost 100% of cases, and in the absence of apical lordosis or the presence of apical kyphosis in the scoliotic curvature, only in 66.6-80% of cases [39, 41]. When SM is associated with IS in the presence of neurological disorders, double curves are more common than single ones, and single lumbar curves are completely absent [29, 41]. It should be noted that with left-sided curvatures of the thoracic localization, an asymptomatic course of SM is more often observed, while in the presence of neurological symptoms, scoliosis is characterized by double arcs of the thoracolumbar localization [16]. Thus, the following main criteria for a high risk of the presence of SM in idiopathic scoliosis can be identified: neurological disorders [42], infantile and juvenile scoliosis [19, 17, 30], atypical deformity (left-sided arc), and abnormal sagittal profile of the spine [42, 39, 17], deformities with two arcs of

curvature and asymmetry of superficial abdominal reflexes [41]. However, one of the shortcomings of all studies of scoliosis associated with SM is the uncertain diagnostic value of clinical symptoms. This is why routine MRI remains the most accurate in patients with IS, where scoliosis has been presented as a symptom in the presence of a neurologically asymptomatic neuroaxial abnormality. Not all detected neuroaxial abnormalities associated with scoliosis require neurosurgical intervention, and correction of the deformity is usually successful [31], but there is a risk of developing neurological disorders if the abnormality was not identified in the preoperative period [37, 38, 51]. Scoliotic deformities associated with Ehlers-Danlos syndrome. Ehlers-Danlos syndrome (EDS), also known as Chernogubov-Ehlers-Danlos syndrome, belongs to a group of hereditary mesenchymal dysplasias – heterogeneous conditions of connective tissue characterized by varying degrees of laxity, hyperextensibility of the skin, all ligaments and membranes, vascular fragility, which clinically manifests as joint laxity and dislocations, musculoskeletal pain, hypotension, hemorrhages, striae, ruptures of membranes, organs and vessels, hernias, scoliosis and kyphoscoliosis [10,38]. Due to the dual type of inheritance, it is difficult to accurately estimate the overall incidence of the disease; however, in the world population, it occurs with a frequency of 1:5000. Based on clinical and genetic characteristics, scoliosis is divided into six types, distinguished by varying degrees of joint instability, hyperextensibility and difficulty healing skin, and vascular fragility. Of greatest interest to vertebrologists is kyphoscoliotic type VI scoliosis, associated with a deficiency of lysyl hydroxylase, a collagen-modifying enzyme [24]. This rare pathology occurs in 1 in 100,000 live births and is inherited in 25% of cases.

In 50% of cases, the proband may be an asymptomatic carrier of the gene, while in 25% of cases, the proband may be affected but not a carrier. In addition, symptoms of the disease associated with the mutation have been identified [52]. Scoliosis in scoliosis is diagnosed at birth or within the first year of life. Progression of the deformity always leads to the need for surgical treatment [47,54], without which scoliosis leads to the development of respiratory disorders and severe kyphoscoliosis. Hypotonia can progress, leading to impaired motor development with the loss of the ability to move in the second and third decades of life. Although life expectancy may be normal, there is a risk of rupture of medium-sized arteries and the development of respiratory failure if kyphoscoliosis becomes severe [47,54]. According to some researchers, double-arc curvatures are more common, most of which worsen sharply at the beginning of the adolescent growth spurt, which necessitates consideration of surgical intervention at an average age of 11 years and 9 months with an average scoliosis magnitude of 88°-109° [24]. It should be noted that scoliosis can also be present in patients with any type of SED, and with its hypermobile type III, up to 50% of scoliosis cases are recorded [46, 24]. Due to the rarity of the pathology, there are only a limited number of small study groups describing the results of surgical treatment of spinal deformity in this disease [49,20, 24,31]. According to the majority of surgeons, spinal curvatures associated with SED almost always manifest themselves in the form of severe scoliotic deformities, the difficulties in treating which are due to the magnitude and rigidity of the deformity, despite the general hypermobility, early age, complexity and prevalence of surgical intervention, high risk of sagittal imbalance, hyperkyphosis and complications, and the risk of exacerbation of symptoms of the underlying disease. The average degree of curvature correction in SED is 30%-58%, which roughly corresponds to the mobility of the curvature [24], and is often (40% of cases) accompanied by revision interventions due to decompensation or failure of the structure, with wound edge divergence and hematomas requiring secondary suturing. The risk of potentially fatal vascular complications associated with massive blood loss during surgical treatment of patients with SED is quite high, which is equally confirmed not only by vertebrologists [49], but also by general surgeons [20]. Surgical treatment requires: hypotensive anesthesia, exclusion of blunt tissue separation, the use of electrocoagulation, delimitation of the discectomy zone and ligation of segmental vessels away from the aorta and vascular trunks. Vascular fragility resulting from distraction can lead to spinal cord infarction with the development of irreversible neurological complications. Therefore, it is recommended to limit correction solely to the magnitude of the functional component of the deformity, confirmed by preoperative radiography with stretching [49]. Thus, at the end of the literature review, it can be noted that this problem is relevant not only in traumatology, but also in medicine in general.

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