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Clinical case

## Correction of Congenital Kyphoscoliosis on the Background of Failure of Vertebral Segmentation in the Thoracic Spine Associated with Diastematomyelia at the Level of the Thoracolumbar Spine

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### Abstract

*Congenital scoliosis is one of the most common deformities of the spine in children. Intraspinal anomalies are always accompanied with congenital scoliosis. Diastematomyelia is one of the most common intraspinal pathologies in congenital scoliosis. To date, there is no standard method for correcting congenital spinal deformity associated with diastematomyelia.*

*We present a clinical case of simultaneous correction of congenital scoliosis by an endocorrector with excision of diastematomyelia.*

*Simultaneous tactics of surgical treatment in this clinical case was the method of choice for congenital scoliosis associated with diastematomyelia, since the total length of the operation and the total volume of blood loss is significantly lower, and exeresis of the bone septum reduces the risk of neurological complications.*

*Keywords: congenital scoliosis, failure of vertebral segmentation, diastematomyelia, correction.*

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## Introduction

Congenital scoliosis is the most common spinal deformity in children and affects approximately 2% to 11% [1]. Congenital scoliosis is a failure of vertebral formation, segmentation or a combination of them, arising from abnormal development of the vertebrae in the period from four to six weeks of pregnancy [2]. Congenital scoliosis is a failure of vertebral formation, segmentation or a combination of them, arising from abnormal development of the vertebrae in the period from 4 to 6 weeks of pregnancy [2]. The prevalence of intraspinal abnormalities in congenital scoliosis ranges from 20 to 58% [3]. The most common anomalies in congenital scoliosis are syringomyelia, diastematomyelia and a fixed spinal cord [4,5]. Diastematomyelia (Split Cord Malformation — SCM) is one of the variants of spinal dysraphy, when the spinal cord is divided into two arms by a septum located in the sagittal area. S.R. Olivier first described this anomaly in 1837 [6].

## Description of the clinical case

Patient M, 17, was admitted to the Department of Orthopedics No. 6 of the National Scientific Center of Traumatology and Orthopedics named after academician N.D. Batpenov Center with a diagnosis of congenital kyphoscoliosis on the background of failure of vertebral segmentation (concretion of Th8-Th12 vertebrae) with a violation of the frontal balance of the trunk. Spinal cord anomaly (diastematomyelia).

She complained of the presence of spine deformity with nagging pain in the thoracolumbar spine. According to the legal representatives, spinal deformity from birth has progressed in dynamics. She has constantly undergone courses of conservative treatment.

At the moment, there are several types of surgical correction of congenital scoliosis associated with diastematomyelia. A number of authors are supposed about the traditional, two-stage method of treatment [7,8]. At the first stage, diastematomyelia is removed, and at the second stage, scoliosis is corrected. Other authors talk about the opportunity of a one-stage operation [9-11]. Some authors do not exclude the possibility of correction of scoliosis without resection of diastematomyelia, however, to date this method has not been sufficiently studied and the risks of complications are very high [12]. Therefore, the choice of surgical tactics remains relevant.

**The purpose** of this manuscript is to describe a clinical case of simultaneous surgery in a patient with congenital scoliosis on the background of failure of vertebral segmentation with diastematomyelia.

During the examination, the patient moves independently, there is an asymmetry of the shoulder blades, shoulder pads. Pronounced scoliotic deformity of the thoracolumbar spine is determined, the arch of the bulge is turned to the right. Rib hump on the right, muscle roller on the left. Palpation is painful in the thoracolumbar spine. The Adams test is positive. Movements in the thoracolumbar spine are limited. The frontal balance of the trunk is disturbed, sagittal balance is preserved (Figure1). There are no neurological disorders on the periphery of the lower extremities, the functions of the pelvic organs are not violated.

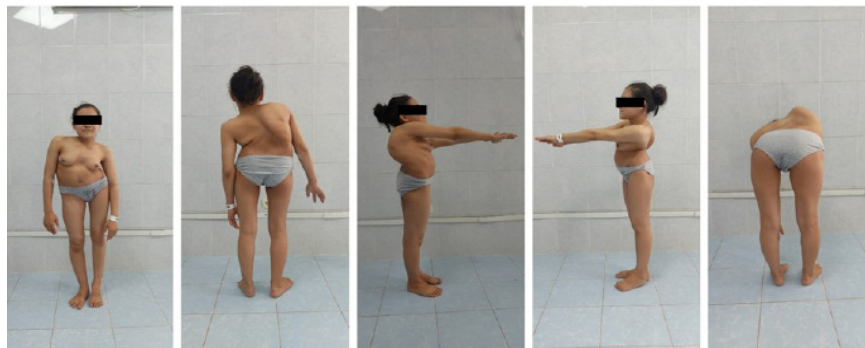


Figure 1 - View of patient A - in front, B - behind, C - right side, D - left side, E - The Adams test is positive

On the X-ray in the frontal projection, the Cobb angle was 110. Violation of frontal balance – 13 cm (Figure 2).

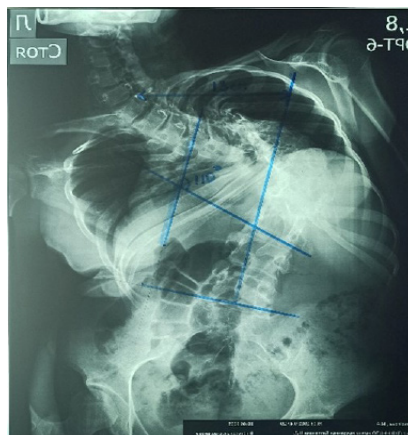


Figure 2 - X-ray in the frontal projection

The rigidity of the scoliotic arch is noted on x-rays with functional samples and on stretching (Figure 2).

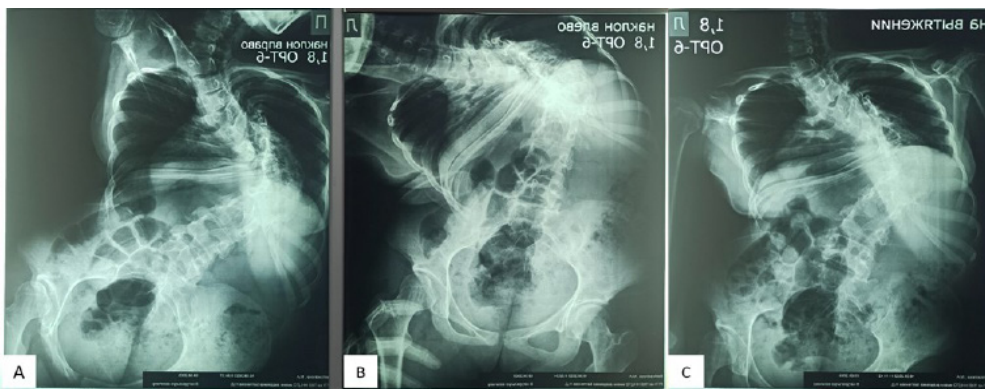


Figure 3 - X-rays with functional samples A - inclination to the right, B - inclination to the left, C - on stretching

Computed tomography (CT) visualizes Th12, L1 vertebrae (Figure 2). diastematomyelia in the form of a calcar at the level of Th11,



Figure 4 - CT in 3 projections A - axial, B - sagittal, C - frontal

Magnetic resonance imaging (MRI) visualizes the separation of the spinal cord into 2 arms at the Th11 level (Figure 5).

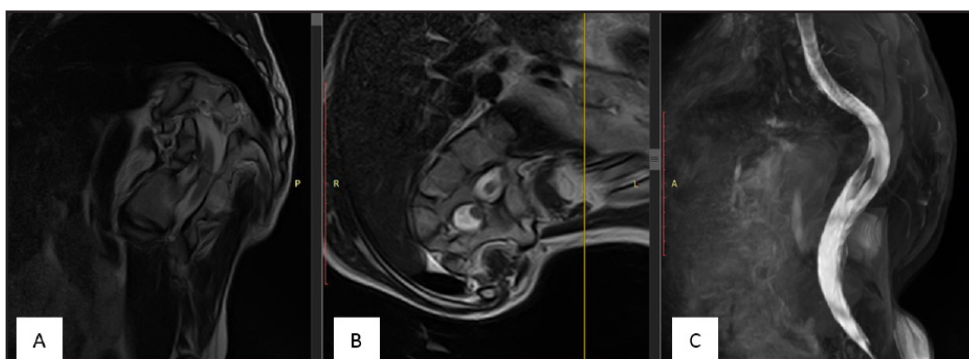


Figure 5 - MRI in 3 projections A - sagittal, B - axial, C - frontal

In our clinic, the patient underwent surgery in the following volumes: Laminectomy of Th11, Th12, L1 vertebrae, partial excision of diastematomyelia and decompression of the spinal cord. Pedicular subtraction osteotomy at the level of Th9, Th10 vertebrae with resection of 11, 12 ribs on the right. Correction of congenital deformity of the spine by the Medtronic spinal system. Posterior spondylodesis (Figure 6).

In the early postoperative period, the patient complained of moderate pain in the area of surgery. There were no neurological disorders on the periphery of the lower extremities. The function of the pelvic organs is

normal. On the 7th day after the operation, the patient underwent a control radiography. After surgery, the Cobb angle is 67°, the correction was 60.9%, the frontal balance was 9 cm (Figure 7).

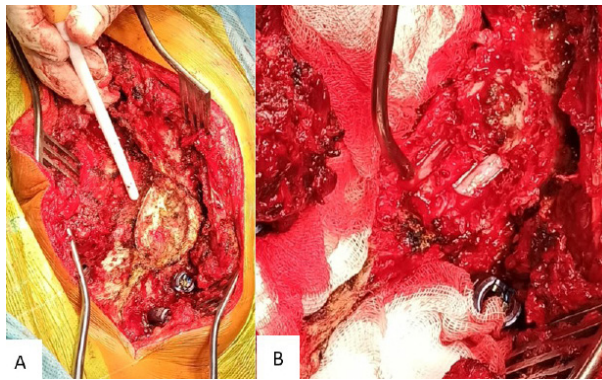


Figure 6 - Intraoperative photos. A – splitting of spinous processes of Th11, Th12, L1 vertebrae, B – separation of the spinal cord into 2 arms (indicated by arrows)

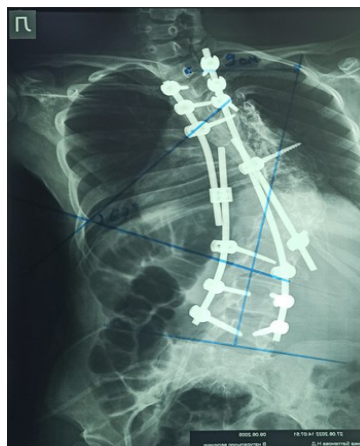


Figure 7 - X-ray in the frontal projection after surgery

The patient was released from hospital on the 10th day after the operation with good health (Figure 8).

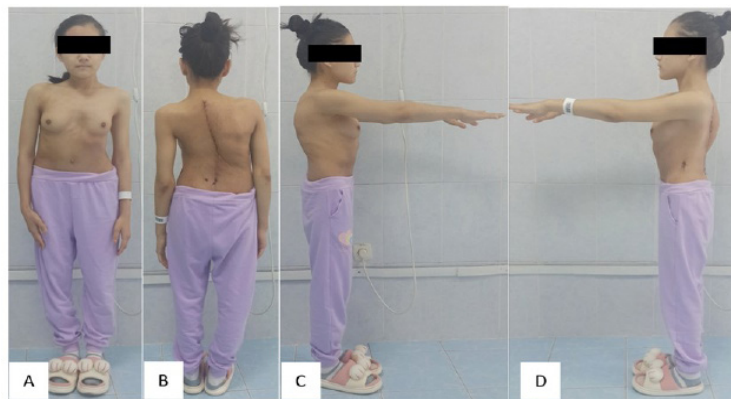


Figure 8 - View of patient after surgery A – in front, B – behind, C – right side, D – left side

The patient gave written informed consent to the publication of her clinical data.

### Discussion

Congenital deformities of the spine should be operated on as early as possible. There are several variants of surgical treatment of patients with congenital deformities associated with diastematomyelia. A number of authors follow to a two-stage tactic, during which the first stage is the removal of diastematomyelia, and the second stage is the correction of scoliosis after 3-6 months [7,8]. However, with this method, the total duration of the operation, the total amount of blood loss is much higher. Other authors are make choise one-stage correction, in which both diastema resection and deformation correction are executed in one

operation [9-11]. During this operation, the total time of the operation is shortened and the total amount of blood loss is decreased. The presence of modern equipment enables intraoperative monitoring in order to minimize possible complications. There are also studies that describe cases of correction of scoliosis without resection of the bone spur [12].

However, during our operation, there was a strong tension of the dura mater of the spinal cord in the process the

correction of deformation, which could lead to neurological complications with a preserved bone septum.

## Conclusions

Therefore, simultaneous tactics of surgical treatment may be the method of choice for congenital scoliosis associated with diastematomyelia, since the total

length of the operation and the total volume of blood loss is significantly lower, and exeresis of the bone septum reduces the risk of neurological complications.

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## Кеуде-бел омыртқалары деңгейінде диастематомиеліамен байланысты кеуде омыртқасының сегментациясының бұзылуы аясында туа біткен кифосколиозды түзету

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## Түйіндеме

Туа біткен сколиоз – бұл балалардағы омыртқаның кең таралған деформацияларының бірі. Интрапинальды патологиялар арқашан туа біткен сколиозбен бірге жүреді. Диастематомиеліа туа біткен сколиоз кезінде жиі кездесетін интрапинальды патологиялардың бірі. Бүгінгі күні диастематомиеліамен байланысты омыртқаның туа біткен деформациясын түзетудің стандартты әдісі жоқ.

Біз диастематомиеліаны жоя отырып, эндокорректормен туа біткен сколиозды бір сатылы түзетудің клиникалық жағдайын ұсынамыз.

Түйін сөздер: туа біткен сколиоз, сегментацияның бұзылуы, диастематомиеліа, түзету.

## Коррекция врожденного кифосколиоза на фоне нарушения сегментации в грудном отделе позвоночника, ассоциированного с диастематомиелией на уровне груднопоясничного отдела позвоночника

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### Резюме

Врожденный сколиоз представляет собой одну из распространенных деформаций позвоночника у детей. Интраспинальные аномалии всегда сопровождаются с врожденным сколиозом. Диастематомиелия одна из часто встречающихся интраспинальных патологий при врожденном сколиозе. На сегодняшний день нет стандартного метода коррекции врожденной деформации позвоночника, ассоциированной диастематомиелией.

Нами представлен клинический случай симультанной коррекции врожденного сколиоза эндокорректором с удалением диастематомиелии.

Одномоментная тактика хирургического лечения в данном клиническом случае был методом выбора при врожденном сколиозе, ассоциированном с диастематомиелией, поскольку общая продолжительность операции и общий объем кровопотери значительно ниже, а экзереэз костной перегородки снижает риск неврологических осложнений.

Ключевые слова: врожденный сколиоз, нарушение сегментации, диастематомиелия, коррекция.