# LITERATURE REVIEW

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## SCOLIOSIS IN PATIENTS WITH ARTHROGRYPOSIS: A LITERATURE REVIEW

© ***O.E. Agranovich[1], I.A. Komolkin[2]***

[1] The Turner Scientific Research Institute for Children's Orthopedics, Saint Petersburg, Russia;
[2] Saint Petersburg State Pediatric Medical University, Saint Petersburg, Russia

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The reported incidence of scoliosis in patients with arthrogryposis varies greatly between 2,5% and 69%. Curves can progress quickly. The most typical localizations of scoliosis are the thoracic and the thoracolumbar parts of the spine. In some cases, thoracolumbar scoliosis is combined with contractures in the hip and pelvis oblique. Scoliosis in children with arthrogryposis, as opposed to limb abnormalities, is generally rarely diagnosed at birth. The diagnosis is often established at the age of 5 years and can be explained by a period of rapid spine growth. The poor prognostic signs that determine the rapid development of scoliosis include progression at an early age, paralytic deformity, and pelvis oblique. Treatment options include spine casting, bracing, expandable implant surgery, and spinal fusion. The treatment goal is to allow optimal growth and development of the chest and a well-balanced spine. This article analyzes the modern approach to treatment scoliosis in patients with arthrogryposis.

**Keywords:** arthrogryposis, scoliosis, bracing, surgical treatment.

## ЛЕЧЕНИЕ СКОЛИОЗА У БОЛЬНЫХ С АРТРОГРИПОЗОМ (ОБЗОР ЗАРУБЕЖНОЙ ЛИТЕРАТУРЫ)

© ***О.Е. Агранович[1], И.А. Комолкин[2]***

[1] ФГБУ «НИДОИ им. Г.И. Турнера» Минздрава России, Санкт-Петербург;
[2] ФГБОУ ВО «Санкт-Петербургский государственный педиатрический медицинский университет» Минздрава России, Санкт-Петербург

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Сколиоз у пациентов с артрогрипозом встречается в 2,5–69 % случаев и часто характеризуется быстро прогрессирующим течением, а также ригидностью. По локализации наиболее часто отмечаются деформации грудного и грудопоясничного отделов позвоночника, при этом возможно поражение одного либо обоих отделов. Часто сколиоз грудопоясничного отдела позвоночника сочетается с контрактурами в тазобедренных суставах и с перекосом таза. Сколиоз у детей с артрогрипозом, в отличие от аномалий конечностей, как правило, редко диагностируется при рождении, чаще в возрасте 5 лет, что может объясняться периодом быстрого роста позвоночника. Плохими прогностическими признаками, обусловливающими быстрое развитие сколиоза, служат: прогрессирование в раннем возрасте, паралитический характер деформации, а также наличие перекоса таза. Цель лечения деформаций позвоночника у больных с артрогрипозом — создание предпосылок для нормального роста и развития грудной клетки, а также восстановление баланса туловища и оси позвоночника. Основными методами лечения данной патологии при артрогрипозе являются: гипсовая коррекция, корсетирование, использование спинальных конструкций, позволяющих осуществлять коррекцию деформаций позвоночника и грудной клетки в процессе роста, а также выполнение спондилодеза.
В статье анализируются современные подходы к лечению сколиоза у пациентов с артрогрипозом.

**Ключевые слова:** артрогрипоз, сколиоз, корсетирование, оперативное лечение.

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## Introduction

In the international literature, the term "arthrogryposis" refers to a heterogeneous group of diseases (>300 nosological entities) representing congenital contractures of ≥2 large joints of non-adjacent areas. These diseases have different etiopathogenesis (sporadic conditions, gene and chromosomal abnormalities, syndromes, and teratogenic pathology) and are divided into three groups.

1. Amyoplasia, the classical form of arthrogryposis (the most common variant of arthrogryposis).

2. Distal forms of arthrogryposis characterized by a predominant lesion in the distal segments of the limbs (hand, foot) with/without maxillofacial anomalies.

3. Congenital joint contractures in different syndromes and diseases (Larsen syndrome, myopathy, diastrophic dysplasia, pseudotrisomy X, myopathy, etc.) [1-6].

In the Russian Federation, it is customary to isolate the third group of arthrogryposis mentioned above as an independent nosological entity, thus the term "arthrogryposis" refers only to amyoplasia and distal forms of arthrogryposis [7, 8].

Conducting an information search in the PubMed database using the terms "arthrogryposis, scoliosis" listed references related to clinical cases and methods of scoliosis treatment in patients with congenital joint contractures in different syndromes and diseases not related to arthrogryposis, and there are very few articles published on the difficulty in treating spinal deformities in patients with amyoplasia and distal forms of arthrogryposis.

The current literature review aims to standardize the worldwide experience in the treatment of scoliosis in patients with arthrogryposis.

## Incidence and clinical characteristics

Scoliosis incidence in patients with arthrogryposis is reported to range from 2.5% to 69% [1–3, 5, 9, 10–12].

Spinal deformities vary in intensity from mild to severe. The literature is extremely contradictory on gender distribution; some authors indicate the predominance in boys, whereas others point out the prevalence in girls [13].

J. Hall et al. examined 560 amyoplasia patients and identified five disease subtypes with different clinical manifestations:

1) with a symmetrical involvement of all four limbs;

2) with a severe symmetrical involvement of all four limbs;

3) with a symmetrical involvement of three limbs;

4) with an involvement of only the upper limbs;

5) with an involvement of only the lower limbs.

In addition to limb deformities, scoliosis was diagnosed in 11.3% of amyoplasia patients with a difference in the incidence of each subtype: subtype 1, 8.6%; subtype 2, 41.5%; subtype 3, 7.4%; subtype 4, 11.1%; and subtype 5, 7.1% [5].

In addition, cases of scoliosis in distal arthrogryposis were described: type 2A (Freeman–Sheldon syndrome), type 4, type 5A, and type 9 (Beals syndrome) [14-17].

Unlike limb abnormalities, scoliosis in children with arthrogryposis is rarely diagnosed at birth and often revealed at approximately 5 years of age, which is probably associated with a period of rapid spine growth. Poor prognostic signs indicating rapid progression of scoliosis are rapid progression observed at an early age, paralytic nature of the deformity, and the presence of a pelvic distortion [13]. B. Kowalchuk et al. recommend an early, thorough spine examination for arthrogryposis patients to detect scoliosis, in view of its high incidence in arthrogryposis [18].

There are three types of scoliosis with arthrogryposis: congenital, paralytic, and idiopathic. Spencer et al. [9] and D.S. Drummond, D.A. Mackenzie [12] reported that a significant number of patients have congenital anomalies of the spine. W. Yingsakmongkol and, S.J. Kumar examined 32 patients with arthrogryposis and scoliosis, but detected no congenital anomalies of the spine in any case [11]. J. Hall et al. observed the most significant spine deformities in subtype 2 of amyoplasia, including severe hyperextension in all parts of the spine (41.5%), wry neck (31.7%), congenital scoliosis (41.5%), and hypoplasia (aplasia) of the back and neck muscles [5]. J.F. Sarvark et al. reported that although congenital scoliosis has been diagnosed in patients with various syndromes, no congenital malformations of the spine have been detected in those with amyoplasia [19]. N.D. Fletcher et al. observed 6 patients

with congenital scoliosis of the cervical spine and arthrogryposis with asymmetric lesions of the upper limbs and detected segmentation disorders, vertebrae formation, and mixed anomalies. Furthermore, the Cobb deformity angle was >10°. Paralytic scoliosis was the most characteristic variant in amyoplasia, especially in patients with severe conditions who were unable to move independently [20].

The thoracic and thoracolumbar spine deformities are the most in scoliosis, with a possibility of either one or both parts being affected. Often, scoliosis of the thoracolumbar spine is combined with contractures in hip joints and pelvic distortion [21]. According to D.S. Drummond and D.A. Mackenzie, most patients with arthrogryposis and scoliosis have a C-shaped arch in neuromuscular scoliosis [12]. K.C. Soultanis et al. also noted extensive paralytic deformities in a number of patients [21]. N. Astur et al. found that the arch is usually localized at the $Th_5$-$L_2$ level [13].

## Treatment

The goal of treatment of scoliosis in arthrogryposis patients is to lay the groundwork for normal growth and development of the chest as well as to restore balance in the body and spinal axis.

The primary treatment methods are plaster correction; use of body jackets, spinal structures that enable to correct spine and thoracic deformities during growth; and spondylosyndesis.

The choice of treatment method primarily depends on the nature of progression and the age of the patient, and not on the severity of scoliosis. Even moderate-sized arches of deformity that are present at birth can remain stable until middle or late childhood. Most scoliosis is progressive and can be rapidly progressive, especially if detected in infancy.

The Mehta method of plaster correction can efficiently treat children aged <5 years with an arch size >30°, allowing a delay in surgical spine interventions. Plaster correction is performed under anesthesia every 8–16 weeks. Plaster treatment is started at the age of 12–18 months, and thermoplastic body jackets can be used for children aged <1 year. A body jacket is recommended in infants with an arch <30°. Unlike other pathologies that cause infantile scoliosis, the Mehta method does not provide a full recovery in arthrogryposis, but it slows scoliosis progression and promotes a symmetrical growth of the chest until the child reaches an age when they can receive more invasive treatments such as installation of spinal structures that corrects deformities of the spine and chest during growth. In children aged >5 years with moderate scoliosis, an alternative method of treatment is using body jackets to further delay surgical treatment [4, 9, 13].

In literature, indications for surgical treatment remain debatable and depend on the extent of the deformity. Thus, T. Cobb recommends surgery if the extent of deformity is >30°. According to Greggi et al. the indications for surgical treatment include a deformity arch of more than 40°, the presence of significant hyperkyphosis or hyperlordosis, and pelvic distortion [2]. Most authors agree that surgical treatment is recommended for a deformity arch >50° in patients with arthrogryposis and scoliosis [3, 9, 19].

Only a few articles in the available literature describe surgical treatment of scoliosis in patients with arthrogryposis, and the clinical material presented is scarce.

The first publication devoted to this topic was written in 1974 by R.M. Siebold who operated 5 arthrogryposis patients with scoliosis and found that spondylosyndesis helps to maintain the achieved deformity correction and prevent further progression, which was confirmed by other researchers in their work. In these 5 patients, most deformities were progressive and became rigid at an early age, and the progression of pelvic distortion correlated with the extent of spine curvature. Deformity progressed even in those patients who wore body jackets for 7 years. The author was not able to significantly correct rigid deformities, but surgical treatment stabilized the further progression of scoliosis. Siebold recommends conservative treatment with a body jacket when scoliosis is detected at an early age, but the treatment outcomes are unpredictable [22].

Y.H. Daher et al. studied the treatment results of 16 patients with arthrogryposis and scoliosis. Scoliosis was diagnosed at different times from birth to the age of 15 years in the study group. Two children died – one at 6 months from bronchopneumonia and the other died from cardiac arrest during surgery at the age of 16 years. Eight patients previously received only orthotics; 4 of them had deformity progression. Six children underwent spinal surgery

(4 had previously received treatment with body jackets) due to posterior spondylosyndesis without instrumental fixation (1 case), posterior spondylosyndesis with instrumental fixation (4 cases), and instrumental fixation without spondylosyndesis (1 case). Spine deformities in arthrogryposis often require surgical correction, especially in case of lordosis of the thoracic spine [23].

W. Yingsakmongkol et al. analyzed the long-term results of arthrogryposis treatment in 46 patients and found that thoracolumbar arch was predominant among them. The average extent of the deformity before surgery was 78.5°. After treatment, 3 patients who could not walk before were able to move independently. The authors recommend body jacket treatment in cases where the extent of deformity is <30° and the patient can move independently. Evaluation of surgical correction methods in the treatment of scoliosis showed the best results with a combination of anterior and posterior spondylosyndesis. According to W. Yingsakmongkol et al., spondylosyndesis with the use of modern instrumental fixation can delay scoliosis progression in order to achieve significant correction and satisfactory treatment results. Spinal implants and extension technologies without spondylosyndesis should be used at an early age to prevent deformity progression [11].

Greggi et al. analyzed the treatment of 6 children with arthrogryposis and scoliosis from 1987 to 2008. The average age of the patients at the time of surgery was 13.1 years (8–18 years).; all patients had rigid deformity that was rapidly progressing and severe by adolescence. One thoracolumbar arch was noted in 4 cases, and the main arch in the thoracolumbar region was associated with the compensatory arch in the upper thoracic or lower lumbar region in 2 cases. The extent of the main arch prior to surgery was 85° (42°–111°), and that of the compensatory arch was <30° (17° in 1 case and 28° in the other). Hyperkyphosis (73° and 74°) and lumbar hyperlordosis (103° and 82°) were each diagnosed in 2 patients. The Harrington–Luque system was used in 2 cases, the Luque system in 1 case, hooks and screws in 1 case, and spinal anchors and pedicle screws in 2 cases. Long-term results were studied for 1 to 11 years (average 4.2 ± 2.7 years).Local spondylosyndesis was achieved, preventing further deformity progression in 5 cases [2].

D. Ferrari et al. published a clinical case of spinal deformity treatment in 1 patient with Freeman–Sheldon syndrome with severe kyphoscoliosis and chest deformity. Scoliosis progressed rapidly despite conservative body jacket treatment, requiring a 2-step surgical intervention [14].

X.S. Wang et al. reported the experience of treating 6 children with contracture arachnodactylia and scoliosis, which was severe, rigid, and difficult to correct. All patients were diagnosed with kyphoscoliosis—thoracic in 2 and thoracolumbar in 4. The mean extent of scoliosis according to Cobb was 88.6° (85°–117°), while that of kyphosis was 93.6° (75°–123°). All patients underwent surgical treatment, including posterior instrumental fixation with pedicle screws and laminar hooks, and vertebrotomy was performed in 4 patients. Following surgical treatment, the mean extent of scoliosis and kyphosis according to Cobb was 37.6° (35°–52°) and 38.6° (28°–54°), respectively, and the degree of correction was maintained at 62.2 and 68.7%, respectively, over the long term [15].

N. Astur et al. analyzed the treatment results of 10 children (mean age, 5 years; average observation period, 4.2 years) with arthrogryposis and scoliosis using the VEPTR system. This technique enabled to reduce the arch from 67° to 43° (37% correction) and kyphosis from 65° to 48° (29% correction). Postoperatively, scoliotic and kyphotic deformities amounted on average of 55° (17% correction) and 62° (8%), respectively, over time. During the observation period, spine growth at $Th_1$-$S_1$ increased by an average of 4.2 cm (1 cm per year). Of the 62 surgeries performed in these patients, 6 complications were observed in 4 patients (inflammation in 3, rib fracture in 2, and implant fracture in 1 case). Local kyphosis developed in the long-term proximal to the fixation level in 6 patients with an extent of ≥45°. Extended bilateral pelvic/rib structures were used in 8 of 10 patients. Having analyzed the obtained data, N. Astur et al. recognized the effectiveness of the VEPTR system for correcting scoliosis and kyphosis as well as ensuring chest growth in arthrogryposis patients, but a technique for treating local kyphosis proximal to the system fixation level remains elusive [13]. Smith [13] reported that the use of bilateral structures to connect iliac bones with 2–4 ribs in 39% of cases led to a "crouch" gait in patients who were able to move independently. Considering this, N. Astur et al. (2014) believe that

short-term use of this technique is possible in patients with an identical motor status [13].

Constructions to correct spine and chest deformities during the growth cycle are indicated in children with arthrogryposis and scoliosis. The effective "growth" of an implant is about 5 years, which makes it possible to use implants in patients aged >5 years, and spondylosyndesis can be performed when children reach the age of 10 years [24, 25]. Two systems of sliding spinal implants generally are used: the VEPTR and the vertebral system using hooks and pedicle screws. The VEPTR system is indicated in cases of the "folded umbrella" chest deformity allowing restoration of the shape of the chest [13, 25, 26].

In patients with arthrogryposis who suffer from idiopathic scoliosis with thoracic arch, the choice of treatment method is similar to that in idiopathic scoliosis; less severe forms of scoliosis are common in these patients. With paralytic scoliosis, extended fixations with capture of the sacrum are required. Spondylosyndesis with sacrum is required in patients with pelvic distortion. In case of, Halo-femoral (halo-tibial) traction is indicated as well as anteroposterior release with spondylosyndesis with use of pedicle screws, vertebral body resection, and vertebratomy in rigid forms of scoliosis. In severely rigid forms of scoliosis, fixation of the sacrum can be effective [2, 3, 8, 9, 12].

When choosing a therapeutic approach for patients with scoliosis and arthrogryposis, it is necessary to assess the condition of the hip joint (presence or absence of contractures), as correction of the position of the spine/pelvis segment should enable the patient to sit after the surgery. If a child has extensive hip joint contractures and scoliosis, the contractures must be eliminated prior to spine surgery.

The results of surgical treatment of scoliosis with arthrogryposis are not always unambiguous. As a rule, the loss of the achieved correction with age is low in patients with idiopathic scoliosis. However, results for rigid forms of scoliosis with arthrogryposis are significantly worse than those in idiopathic scoliosis [9].

## Conclusion

Analysis of foreign literature revealed an insufficient representation of scoliosis treatment in patients with arthrogryposis. Available literature were mainly devoted to single clinical observations and contain general recommendations for patient management. There are currently no data on the clinical course of scoliosis in patients with amyoplasia and distal forms of arthrogryposis, hence this topic needs further study.

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*Information about the authors*

**Olga E. Agranovich** — MD, PhD, professor, head of the department of arthrogryposis. The Turner Scientific and Research Institute for Children's Orthopedics. E-mail: olga_agranovich@yahoo.com.

**Igor A. Komolkin** — MD, PhD, assistant of the chair pediatric surgery Saint Petersburg State Pediatric Medical University. E-mail: igor_komolkin@mail.ru.

**Ольга Евгеньевна Агранович** — д-р мед. наук, руководитель отделения артрогрипоза ФГБУ «НИДОИ им. Г.И. Турнера» Минздрава России.
E-mail: olga_agranovich@yahoo.com.

**Игорь Александрович Комолкин** — канд. мед. наук, ассистент кафедры детской хирургии ФГБОУ ВО «Санкт-Петербургский государственный педиатрический медицинский университет» Минздрава России. E-mail: igor_komolkin@mail.ru.