THORACOLUMBAR KYPHOSIS IN ACHONDROPLASIA: LITERATURE REVIEW

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Objective. To detect and summarize the existing data on the prevalence, diagnostic methods, prevention and treatment strategies of the thoracolumbar kyphotic deformities in patients with achondroplasia.

Material and Methods. Study design: literature review. The literature search was conducted in electronic databases: PubMed, CrossRef, Google Scholar and eLIBRARY.ru. The review includes articles in English and Russian languages. There were no limitations regarding the level of evidence and the year of publication.

Results. The selected 22 articles included 13 retrospective studies and 9 case reports. After dividing all studies into categories related to methods of diagnosis, prevention and treatment of thoracolumbar kyphosis, the following results were obtained: the definition of the border between the norm and pathology was reported only in two articles, and the prevalence rates could be counted based on the data of four articles. The identification of risk factors for the progression of deformity based on statistical analysis was presented in two articles. Three articles were devoted, among others, to the risk of neurological deficit. The description of brace treatment was found in four studies, and six retrospective studies and all selected case reports described surgical treatment.

Conclusion. There are very few publications devoted to the problem of thoracolumbar kyphosis in achondroplasia, despite possible poor prognosis of such deformities. Further research should be aimed at searching a consensus among experts in all issues concerning this type of deformity, from prevention to surgical correction of this pathology.

Key Words: achondroplasia, thoracolumbar kyphosis, spinal deformity, systematic review.

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Achondroplasia belongs to a group of rare hereditary diseases; for this reason, practitioners of all specialties do not often encounter this pathology in their practice. However, among skeletal dysplasias, the incidence rate of achondroplasia is one of the highest and amounts to one per 26,000 live births [1].

Since the late 19th century when the term "achondroplasia" was first used, and the main disease features were described, the genetic basis of this pathology and the pathogenesis mechanisms have been studied in detail. A mutation in the fibroblast growth factor receptor 3 (*FGFR3*) gene leads to disruption of endochondral ossification due to suppression of chondrocyte differentiation and impaired production and proliferation of cartilage matrix. The pathological process involves all long tubular bones, vertebrae, and some bones of the skull, which gives patients a characteristic appearance [2]. Body disproportion manifests by shortening of the limbs, mostly proximal parts, with the trunk length being relatively normal, and skull shape changes [3]. Usually, achondroplasia patients are characterized by an average health level in the population and a normal life expectancy. Life-threatening conditions are rarely diagnosed, but some otorhinolaryngological, neurological, and orthopedic complications in severe cases can lead to profound disability and death [4, 5]. In this regard, spinal disease in achondroplasia requires particular attention.

Thoracolumbar kyphotic deformity in two achondroplasia patients was first described in 1920 [6]. In most cases, these deformities are transient; the incidence rate is markedly reduced in children after one year of life [7]. But in some cases, the deformity progresses and requires conservative or surgical treatment [7, 8]. Unlike congenital kyphosis, kyphotic deformities in achondroplasia develop as a result of defective vertebral growth due to disruption of endochondral ossification [9]. Associated muscle hypotonia, joint hypermobility, large heavy head, and delayed motor development do not allow the child to keep the trunk upright and create an additional load on the dysplastic vertebrae, which promotes rapid development of wedge-shaped deformities of the vertebral bodies [7, 9, 10]. Small deformity is not associated with patient complaints, but as the deformity progresses, the risk of neurological deficits increases [11].

Because this pathology is quite rare, the literature has reported only a small number of cases of kyphotic deformity treatment in achondroplasia patients. Now, there are no generally accepted standards for the treatment and prevention of this deformity.

The aim of this study is to analyze and generalize the available data on the prevalence, diagnosis, prevention, and treatment of thoracolumbar kyphotic deformities in achondroplasia patients.

Material and Methods

The search for publications was performed in PubMed, CrossRef, Google Scholar, and eLIBRARY.ru electronic databases using the following keywords: achondroplasia, thoracolumbar kyphosis, spinal deformity, and treatment. The study included publications in Russian and English. There were no limitations on the level of evidence and the time of publication.

The study was divided into different categories:

1) criteria for diagnosis of thoracolumbar kyphosis in achondroplasia;
2) prevalence rate;
3) factors of deformity progression;
4) risk of neurological deficits;
5) preventive and conservative treatment options;
6) surgical treatment.

Results

Criteria for diagnosis of thoracolumbar kyphosis. Where is the normal–disease boundary? This question can baffle any clinician. In this regard, the diagnosis of thoracolumbar kyphosis is no exception. Investigation of the sagittal balance in patients without spinal pathology revealed that the spinal column shape (Cobb measurement) ranged from 3° of lordosis to 20° of kyphosis at the T10–T12 level and from 23° of lordosis to 13° of kyphosis at the T2–L2 level [12]. In a systematic review of the prevalence and development of kyphosis in achondroplasia, Engberts et al. [13] reported that none of the seven studies included in the review provided a definition of this deformity. The authors of only two articles included in the present review defined the lower limit of the norm for thoracolumbar deformities, based on studies devoted to assessing the sagittal balance [8, 14]. Khan et al. [8] defined kyphosis with a Cobb angle of more than 10° and the apex at the T11–L2 level as pathological. A range of 10 to 25° was associated with mild kyphosis, 26–50° with moderate kyphosis, and more than 50° with severe kyphosis. Margalit et al. [14] considered mild kyphosis with a Cobb angle ⩾20° and the apex at the T12–L1 level as a referent. According to the authors, defining thoracolumbar kyphosis as a Cobb angle of >0° would overestimate this condition in achondroplasia patients.

Borghuu et al. [15] defined deformities with a Cobb angle of less than 20° as success in spontaneous regression of the deformity, and Xu et al. [16] – during brace treatment. However, the authors did not explain why they chose this magnitude, and whether deformities below this magnitude may be considered normal.

Prevalence rate. In most of the analyzed papers, thoracolumbar kyphosis was included in the inclusion criteria; therefore, only four studies allow calculating the prevalence rate of deformity in the population of achondroplasia patients [7, 8, 11, 17]. Bethem et al. [17] examined 80 patients with various systemic diseases accompanied by impaired growth for spinal column pathology. Of 30 achondroplasia patients included in the study, 18 patients had thoracolumbar kyphosis of varying severity, which amounted to 60 %. Because detailed investigation of this deformity type was not the purpose of that study, the authors did not provide a detailed report on the age of examined patients and the magnitude of deformity. Comparable results are presented in a study by Kahanovitz et al. [11]. Of 36 patients who underwent lateral radiography of the thoracolumbar spine, kyphotic deformities were found in 18 (50 %) patients.

Given the transient nature of this pathology in achondroplasia, Kopits [7] investigated the prevalence rate of deformities depending on age. In patients under 1 year of age, kyphosis occurred in 94 % of cases. By the age of 10 years, the prevalence rate of kyphosis gradually decreased to 10 % and then again increased to 35 % by the age of 20–50 years. According to the author, an increasing prevalence rate of deformities in the older age group may indicate persistent kyphosis or be associated with an increase in the rate of consultation sought by the patients with this pathology. In that study, the average population prevalence rate of spinal deformities reached only 35 %.

Khan et al. [8] reported a higher prevalence rate. They found kyphotic deformities with a Cobb angle of more than 10° and the apex at the T11–L2 level in 79 % of patients. The mean age of examined patients was 18 years. Depending on the age group, the following results were observed: 0–2 years – 88.5 %, 3–12 years – 80.0 %, 13–19 years – 70.5 %, 20–40 years – 71.5 %, and over 40 years – 78.5 %. Therefore, there are no previously described significant differences in different age groups. Noteworthy, both of the above studies were conducted at the same medical university (Johns Hopkins University School of Medicine), 27 years apart.

Factors of deformity progression. In a study by Borkhuu et al. [15], the authors searched for a statistically significant relationship between deformity progression and various factors: gender of patients, a concomitant diagnosis of hydrocephalus, the presence of a ventriculoperitoneal shunt, previous foramen magnum decompression, delayed motor development, thoracolumbar kyphosis magnitude, thoracic kyphosis, lumbar lordosis, the percentage of apical wedging for height of the vertebra, and the percentage of apical vertebral translation. Two criteria were chosen to diagnose delayed motor development: an inability to sit without support by 6 months and an inability to walk independently by 15 months, which is comparable to the average population data. The patients were divided into two groups based on deformity progression. A statistically significant difference in the age at onset of walking ($p = 0.02$) was found. For example, the mean age at onset of walking for children with spontaneous regression of deformity was 17.9 months, while that for children with deformity progression was 22.7 months. An analysis of the radiographic parameters revealed a statistically significant difference for the magnitude of lumbar lordosis and the percentage of apical wedging for the height of the vertebra ($p < 0.05$). Deformity progression also depended on the magnitude of kyphosis ($p < 0.001$)

and the percentage of posterior translation of the apical vertebra (p = 0.01). A linear regression analysis revealed that only delayed motor development was associated with deformity progression (p = 0.03): patients with progressive kyphosis were 2 times more likely to have delayed motor development (in terms of the ability to sit and walk), relative risk (RR) 2.4; 95 % CI 1.10–5.25. Similar data were obtained in a study by Margalit et al. [14]. Clinical parameters such as the presence of a ventriculoperitoneal shunt, concomitant hydrocephalus, foramen magnum decompression, and gender of patients were not statistically significantly associated with progression of kyphotic deformity. As in the previous study, the only clinical parameter significantly associated with deformity progression was delayed motor development (p = 0.015); the same factor was the only significant one revealed by linear and multivariate regression analyses. In this study, criteria for motor skill development delay were as follows: an inability to sit without support and an inability to walk independently by 14 and 30 months, respectively, adapted to the population of achondroplasia patients. Differences in the percentage of posterior translation of the apical vertebra (retrolisthesis) (p = 0.001) and the percentage of apical vertebral wedging for height (p = 0.031) were statistically significant between patients with spontaneous regression of deformity and patients with persistent kyphosis one year after walking age.

Risk of neurological deficits. The gravest complication of thoracolumbar kyphosis progression is spinal cord compression. Kahanovitz et al. [11] divided patients into 5 groups depending on the severity of neurological complaints. Group 1 included patients without neurological deficits. They had kyphosis in 44 % of cases; the mean Cobb angle of deformity was 16°. Group 2 consisted of patients who presented with lumbar pain. In this group, kyphosis of 20° was diagnosed in 44 % of patients. Group 3 included patients diagnosed with herniated discs causing neurological deficits. Of two patients in this group, kyphosis was found in both and amounted to 62°, on average. A similar deformity magnitude was presented in Group 4 that included patients with complaints of neurogenic intermittent claudication. Of 8 patients, kyphosis was found in 3 (38 %). Group 5 consisted of patients with the most severe neurological symptoms, up to paraparesis, with pelvic organ dysfunction. Kyphotic deformity was found in 83 % of them, but the magnitude was only 31°. Therefore, this study shows that neurological symptoms are associated with kyphosis, but there is no dependence on the deformity magnitude, and, also, not all patients with kyphosis have neurological deficits.

Kopits [7] reports a direct relationship between kyphotic deformity and the presence and severity of spinal stenosis symptoms. The author also warns that kyphosis is not the only component of stenosis.

According to Tolo [18], the mean age in a group of patients without neurological deficit was less than that in a group of patients with neurological deficit, which was 8 and 22 years, respectively.

Prevention of progression and conservative treatment. Each parent of a child with achondroplasia should be informed about all possible risks of the development and progression of kyphotic deformity. One of the basic rules is to prevent the child from being in a sitting position without support. To ensure interaction of the child with the outside world; child seats with a 45° backrest angle should be used. In addition, parents should watch the position of the child held in arms and avoid the child from curling the lumbar spine [10].

The main conservative treatment for kyphotic deformity is the use of various braces. A modified Knight brace with three fixation points is described in a study by Kopits [7]. Seventy-six children used the brace for 29 months, on average. The mean deformity at the study onset was 56°; the age of children ranged from 8 to 26 months. The brace corrected deformity initially to 25–55 % of the original magnitude. According to the study, the brace corrected initial signs of deformity in 29 % of patients and stopped progression in 55 % of cases (residual deformity, 25–55°). In 10 (13 %) patients, deformity progressed and eventually reached 45–105°. Surgery was recommended for these patients. According to the author, treatment outcomes are affected by many factors: severity of deformity, concomitant muscle hypotonia, age of brace initiation, and compliance of the patient and his\her parents with the bracing program. The best option is to prescribe a brace between 8–18 months of age and to use it for at least 70 % of the time, particularly while the child is in the upright position. However, despite the timely prescription of a brace and adherence to the recommendations for the duration of its use, deformity progression was observed in some patients. On the other hand, there is no evidence that patients from the success group would not have spontaneous regression of deformity without using the brace.

In a study by Borkhuu et al. [15], a brace was prescribed to 14 (29 %) patients with progression of deformity; 10 patients had improvement due to its use. Because the study was aimed at assessing risks of deformity progression, the authors did not indicate the brace type, percentage of correction, duration of use, and other parameters necessary to evaluate the brace treatment.

Only 2 out of 60 patients were treated with a brace in a study by Margalit et al. [14]. For this purpose, nighttime braces with 3-point bending were used. One patient underwent brace treatment from 18 months to 5 years of age. For this period, the deformity decreased from 73 to 25°. Recall that the authors of this article defined the referent for kyphotic deformity (Cobb angle) below 20°. Another patient used a brace from 10 months to 4 years of age and achieved improvement at the end of treatment. The deformity at that time was 25°, with the initial magnitude being 40°. However, after discontinuation of bracing, the deformity recurred, and, the kyphosis magnitude was 68° by the age of 7 years. Subsequently, the patient underwent surgical correction of the deformity.

The most detailed study on brace treatment for thoracolumbar kyphosis in achondroplasia patients was published by Xu et al. [16]. An analysis was carried out in a group of 33 patients with a mean age of 2.3 years. All patients were prescribed a Boston brace. The recommended usage time was 22 h per day. The mean kyphosis at the study onset was 41.7° and decreased to 29.5° by the end of follow-up. The mean period of bracing was 32.2 months. Bracing was discontinued when there was no correction of the deformity for a year. Compliance assessed by a survey of patients' parents was more than 95 %. The bracing success was defined as a decrease in the deformity to <20° (Cobb angle), which was achieved in 18 (54.5 %) patients. Subsequent comparison of the radiographic and clinical data of patients from the success and failure groups revealed that the age of treatment initiation ($p = 0.04$), kyphosis magnitude ($p < 0.001$), percentage of apical vertebra wedging ($p = 0.01$), and pelvic tilt (PT) ($p = 0.03$) were significantly less in patients of the success group. Parameters such as the lumbar lordosis magnitude and pelvic incidence (PI) did not affect the treatment outcome. Out of 33 patients, retrolisthesis of the apical vertebra was found in 7 patients, which was also associated with higher risk of an unsatisfactory treatment outcome ($p = 0.03$). Concomitant scoliosis, which was present in 18 patients, was not associated with progression of kyphotic deformity and was represented equally in both groups. Plotting the receiver operating characteristic curve (ROC-curve) showed the optimal cutoff points for kyphotic deformity (40°), apical vertebral wedging (60.0 %), and PT (15°). For example, in 17 patients with initial deformity of less than 40°, the mean correction rate was 48.6%, which was significantly higher than that (17.9 %) in patients with initial kyphosis of more than 40°.

Surgical treatment. Kyphosis progression despite conservative treatment, clinical manifestations of neurological deficit or risk of its development, and a pronounced cosmetic defect are the main causes that determine the need for surgical correction of deformity. Numerous surgical techniques have been described; some of them currently have no practical value and are of purely historical interest. Among the papers included in this review, 6 retrospective studies [18–23] and 9 case reports [24–32] are devoted to surgical treatment. Several cases of surgical correction of deformity without a detailed description of the surgical technique are presented in three studies [14, 15, 17] where the authors report 59, 1, and 4 patients operated on, respectively.

Shikata et al. [19] reported 3 cases of surgery in 1971–1979 due to severe neurological deficits. The Cobb angle of kyphotic deformity was 28–32°. Radiographs showed the characteristic findings of achondroplasia: an altered vertebral shape, a decreased interpedicular distance, and hypoplasia of the pedicles and facet joints. At the time of surgical treatment, the patients' age ranged from 19 to 33 years. All patients underwent multilevel laminectomy. In only one case, laminectomy was combined with autologous bone fusion and Harrington instrumentation. By the end of follow-up, neurological deficit persisted or worsened in patients who underwent laminectomy without instrumented fixation; in one patient, deformity progressed to 57°. A positive result was achieved only in the case of using Harrington instrumentation, which was reflected in the absence of complaints after a 2-year follow-up period.

Tolo [18] reported the results of surgical treatment of kyphotic deformity in 17 patients aged 3 to 39 years; of these, 10 patients presented with neurological complaints. The mean deformity at the time of surgery was 85°. Eleven patients underwent deformity correction through two approaches without laminectomy, with five of these patients undergoing the procedure as a one-stage operation. Six patients were operated on with laminectomy or after previous laminectomy (5 – through two approaches and 1 – through an anterior approach alone). Two patients had previous attempts of instrumented fixation using devices located within the canal (the author did not indicate which ones); both patients developed evoked potential changes without subsequent neurological deficit. Four patients underwent spinous process wire fixation; three patients developed somatosensory evoked potential changes intraoperatively, which required removal of the instrumentation. One patient achieved a positive result with spinous process wire fixation and Luque instrumentation. Fixation with Cotrel – Dubousset instrumentation was used in only one patient, in which case a positive result was also achieved. According to the author, the mean deformity correction was 20°, and the neurological status improved.

Ain et al. [20] defined the indications for surgical correction of deformity with kyphosis of more than 60° with progression of more than 10° per year. In that study, the indications for surgery and the surgical technique using a combined approach were similar to those for congenital kyphotic deformities. The mean deformity was 71.5°. To avoid neurological deficits, possible correction was calculated from a lateral radiograph in the hyperextension position and did not exceed intraoperatively the calculated value. In total, 4 children, 4 to 8 years old, were operated on. No patient had preoperative neurological deficit. Surgery consisted of two stages. First, apical disc excision with placement of morcellized bone graft into the disc space was performed through an anterior approach; then, 4 mm screws were inserted in the bodies of the end vertebrae of the curve, the deformity was corrected using a posterior rod, and anterior fusion with a rib or fibula bone graft was performed. Then, the patient was placed in the prone position, and facetectomy, decortication of the posterior vertebral structures, and posterior fusion with an iliac crest bone graft (taken from one side) were performed through a posterior approach without instrumented fixation. Decompression of the spinal canal contents was not performed. After the first stage of surgery, the patient was treated with a cast that was changed at least once until the second stage of surgery. After 4 months, posterior fusion was additionally performed (with an iliac crest bone graft

from the opposite side) to enhance fixation, decrease subsidence of the anterior column bone block, and reduce the time spent in the cast. The final correction was 26.4 %. No patient had any complications within the follow-up period.

Another study by Ain et al. [21] reported 12 cases of surgical treatment. The mean deformity was 64°. Three patients underwent surgery because of deformity progression; the others, in addition, had neurological deficits of varying severity. Four patients had previous laminectomy, and three patients without neurological deficit symptoms underwent a two-stage intervention as described in the previous study. In nine cases, laminectomy was required. In one case, anterior release and posterior instrumented fixation were performed. One patient underwent placement of instrumentation through anterior and posterior approaches. Seven patients underwent surgery through a posterior approach with placement of transpedicular instrumentation. The final deformity correction was 50 %. The following complications were reported: two patients had three iliac rod fractures due to trauma; one patient had a CSF leak that resolved without complications; one patient developed proximal junctional kyphosis (PJK) cranial to the fusion area. Based on the study results, the authors identified 2 basic principles for surgical correction of kyphosis in children with achondroplasia: 1) laminectomy should be followed by posterior spinal fusion, regardless of the initial curve magnitude; 2) patients under 4 years of age with progressive curves of more than 50° require surgical correction of the deformity. Additional indications for an anterior approach include: 1) pedicle size is insufficient to accommodate screw instrumentation; 2) the need for corpectomy to relieve anterior impingement; 3) hyperextension radiographs over a bolster show failure to correct the deformity to less than 50°.

Qi et al. [22] reported 4 cases of surgical deformity correction in patients aged 15–60 years with pronounced neurological deficits. In all the cases, a single-stage surgical intervention was performed through a posterior approach with concomitant decompression of the nerve structures and transpedicular instrumented fixation. If the apex vertebra was markedly hypoplastic, its spondylectomy and reconstruction of the anterior column using a titanium mesh cage were performed. The mean deformity correction was 43.6 %, with the mean initial kyphosis being 96.3°. All patients improved their neurological status after surgery, and, according to a survey, were satisfied with the correction outcome. The authors reported the following complications: one patient had partial laceration of the right T12 nerve root; another patient had a CSF leak that was repaired intraoperatively. These complications did not affect the neurological status in the postoperative period. In one case, intraoperative loss of fixation was observed during correction, which required replacement of pedicle screws. One of the complications was that, due to kyphotic deformity correction and sagittal balance changes, the patient was not able to reach his buttocks and experienced difficulties during hygiene procedures. The authors recommended paying attention to this aspect when planning correction, especially in elderly patients with limited mobility in the lumbar spine and fixed flexion contractures of the hip joints.

Sun et al. [23] also preferred deformity correction and nerve structure decompression through a posterior approach. The study included 6 patients aged 12 to 36 years with a mean deformity magnitude of 53.3°. Correction was 75.0 %. Four patients who initially presented with neurological complaints improved their neurological status.

We found 9 papers [24–32] describing 10 clinical cases of surgical treatment of kyphotic deformities in achondroplasia patients. For ease of perception, the description of these papers is given in the Table.

Discussion

The review demonstrates various approaches to the diagnosis and treatment of thoracolumbar kyphosis in achondroplasia.

The first thing we have come across reviewing the literature is that most researchers have not defined parameters at which the deformity requires surveillance and treatment and is not a variant of the normal condition. Despite the apparent formality of the issue, a clear distinction between the normal condition and disease is of practical concern. Assessment of deformity progression and, consequently, decision-making regarding a treatment approach, among other things, depend on the deformity magnitude [15]. The most objective way to determine the magnitude of any spinal deformity, regardless of its etiology, is to measure the kyphosis angle. The most common procedure is a measurement of deformity angles using the Cobb method [33]. Errors at the stage of diagnosis often lead to the prescription of incorrect treatment and an unfavorable outcome, which can be defined as deformity progression with risk of neurological deficits.

Among the clinical parameters influencing deformity progression, only the motor development parameters are statistically significant: the ability to sit without support and walk independently by a certain age [14, 15]. Children with achondroplasia, during their development, demonstrate motor stereotypes not typical of healthy children [34]; their basic motor skills develop later than in the population, on average, which should be considered upon assessing the motor development [14, 34].

In cases without spontaneous regression of deformity and with increased risk of its progression, orthopedists usually recommend bracing, which is often reasonable and leads to a positive outcome [7, 15, 16]. As in any other treatment, the physician prescribing a brace should assess potential outcomes as objectively as possible based on clinical and radiographic parameters; however, the authors of only one study tried to define the optimal indications for using a brace based on statistical analysis [16]. There is no doubt that assessment of clinical and radiographic parameters affecting the outcome of bracing enables sound prescription of treatment in each spe-

cific case and, in the case of unfavorable prognostic signs, selection of patients for surgical treatment without waiting for progression of deformity.

The choice of a surgical technique depends on many factors. These include the magnitude of deformity, severity of dysplastic changes in the spinal column, patient age, and general somatic status. The experience and qualifications of the surgeon play an important role. Over the past decades, spinal surgery has made a breakthrough in its development, which could not but affect the treatment of thoracolumbar kyphosis in patients with achondroplasia. As mentioned above, those types of operations that were performed in the second half of the last century [18, 19] have receded into the past. At the present, the most common procedure is correction through one or two approaches with placement of transpedicular instrumentation and spinal fusion using bone allo- or autografts. If necessary, interventions are combined with decompression of the nerve structures [21, 22, 24–29, 31, 32].

Conclusion

A small number of papers have addressed the issue of thoracolumbar kyphosis in achondroplasia, despite the fact that progressive kyphosis significantly reduces the quality of life and can cause early disability in achondroplasia patients. Further research should be aimed at achieving a consensus on all issues related to this pathology, ranging from prevention to surgical treatment.

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Table
Summary table of cases of surgical treatment for kyphotic deformities in patients with achondroplasia (literature data)

Publication	Number of patients, n (age, years)	Kyphosis	Kyphosis before/after, deg. (correction, %)	Complaints	Surgery	Complications	Follow-up period, years	Results, conclusions, comments
Yilar et al. [24]	1 (4)	T12–L4	107.5/11.3 (89.5)	Cosmetic defect, gait disturbance	Single-stage. Posterior approach, TPF, T3–L4 growing instrumentation	No	–	Due to severe deformity, extended fixation was chosen
Miyazaki et al. [25]	1 (61)	L1–L3	105.0/32.0 (70.0)	Back and leg pains. Radiculopathy. NIC	Single-stage. 1 – anterior approach: XLIF at L3–L4 and L4–L5, anterior release, L1–L3 discectomy, partial L2 body resection; 2 – posterior instrumented fixation of T10–L5 (4-rod construct), laminectomy of T10–T11, T11–T12, T12–L1, L1–L2, and L2–L3, apical VCR (L2 body resection, expandable cage at L1–L3). Verticalization on day 3, TLSO brace for 6 months after surgery	No	2	No complaints. The first reported case of this surgery in an adult patient with achondroplasia
Ahmed et al. [26]	1 (5)	–	–	Back pain, stiffness in the lower extremities	Two-stage surgery: 1st – L1–L2 vertebrectomy; 2nd – 1 week later, posterior instrumented fixation, deformity correction, posterior fusion with bone allograft	No	4	No complaints
Popa et al. [27]	1 (38)	T10–L2	34.0/0.0 (100.0)	–	Two interventions: posterior instrumented fixation of L2–L5; L4–L5 TLIF; L2–L3 and L3–L4 XLIF; after 4 years, re-operation due to sagittal imbalance: L1 PSO, L1–L2 TLIF, T9–T12 Smith-Petersen osteotomies, extension of posterior fixation cranially to T8	No	–	–
Usikov et al. [28]	1 (44)	–	109.0/35.0 (68.0)	Pain, weakness in the lower extremities (s>d), numbness of the left leg, increased pain when walking. POD	Single-stage. Posterior approach: instrumented fixation 8 TPF of T11–L3 bilaterally, T12–L2 extended laminectomy, resection of the wedge-shaped L1 and accessory hemivertebra; deformity correction with a transpedicular system with external supports; anterior corporodesis of T11–L2 with autologous bone mesh	Lower distal paraparesis in the early postoperative period, which resolved in 3 weeks	1	On discharge, walking independently. No complaints at the end of follow-up

Kolesov et al. [29]	2 (5; 47)	–	1 – 95.0/25.0 (74.0); 2 – 51.0/20.0 (61.0)	1 – progression of kyphosis; 2 – lower flaccid paraparesis, POD	1 – in two stages. T11–L4 TPF, Smith-Petersen osteotomies of T11–L3, rods were not used. Anterior approach: T12–L1 and L1–L2 discectomy, anterior fusion. After 10 days, rod placement, kyphotic deformity correction. Activation on day 5. Leningrad-type brace for 8 weeks; 2 – one-stage. Posterior approach: L1–L2 laminectomy, decompression at the stenosis level, T11–L4 TPF. Anterior approach: L1–L2 body resection, anterolateral decompression, anterior fusion	1 – no; 2 – CSF leak (resolved spontaneously). Necrosis of postoperative wound edges in the back, secondary sutures	1 –3.5; 2 – 2	1 – no complaints; 2 – cane support, POD normalization
Auregan et al. [30]	1 (18)	–	180.0/180.0 (0.0)	Cosmetic defect, NIC	Single-stage. *In situ* fusion. Decompression through a posterior approach,: left-sided five-level hemilaminectomy, discectomy and partial decancellation of three apical vertebrae, spinal cord translation 2 cm anteriorly, anterolateral spinal fusion by cortical bone allograft, posterior spinal fusion by bone allograft, no deformity correction, no instrumented fixation. The spinal cord is uncovered by bone for some length. Verticalized on day 3. Cast for 3 months, TLSO for 6 months	No	3	Lumbago in the lower extremities on accidental contact with the skin above the spinal cord where it is uncovered by bone. No kyphosis progression. Disappearance of neurological claudication
Liao et al. [31]	1 (13)	T12–L2	55.0/22.0 (60.0)	Weakness of the back muscles, numbness of the lower extremities, NIC	One-stage. 1 – anterior approach: through two approaches, L1 partial resection, discectomy, anterior fusion of T12–L2 by allo- and autologous bone grafts; 2 – posterior approach: T12–L4 laminectomy with articular process preservation, T10–L4 TPF. Verticalized on day 2	No	1	The youngest patient at that time who had 2 approaches simultaneously
Aralak et al. [32]	1 (13)	T10–L2	97.0/32.0 (67.0)	Weakness of the lower extremity muscles during prolonged load	One-stage. Posterior approach, two-level PSO of T12 and L1, T4–L4 TPF. Bracing for 9 months	No	2	Authors opposed to anterior fusion due to risk of disturbed anterior vertebral growth

PSO – posterior subtraction osteotomy, VCR – vertebral column resection, TLIF – transforaminal lumbar interbody fusion, XLIF – minimally invasive lateral fusion, TLSO – thoracolumbar-sacral orthosis, TPF – transpedicular fixation, POD – pelvic organ dysfunction, NIC – neurogenic intermittent claudication.

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