



# NEUROGENIC SPINAL DEFORMITIES IN ADULTS: MODERN PROBLEMS AND APPROACHES TO TREATMENT

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**Objective.** To present the features of clinical manifestations, diagnostic aspects and approaches to the treatment of neurogenic deformities of the spine in adults based on the literature data.

**Material and Methods.** The literature review was performed using PubMed, Medline, Web of Science, Scopus, CrossRef, AOSpine, Clinical Key, eLibrary databases and references of key articles published in the period from 06.02.2017 till 04.11.2017.

**Results.** General trends in the treatment of adult patients with neurogenic deformities of the spine have been determined. Assessment of the risk from performing an intervention, taking into account possible complications and potential outcome, determines the approach to surgery in these patients. It is necessary to develop protocols of management with the definition of the main clinical symptoms, the rationale for the use of non-invasive, minimally invasive or other options for care. Surgical treatment of patients with spinal deformities associated with neurodegenerative diseases (Parkinson's disease) is accompanied by a high rate of complications and repeated interventions. Tactical algorithms for these patients should include the consistency of non-surgical and mini-invasive techniques and considering clinical manifestations of myopathy, myelo-, radiculopathy, which, in comparison with diagnostic tests, can determine the indications and volume of decompressive interventions, and the extent and levels of spinal fixation.

**Conclusion.** The complexity of pathogenetic mechanisms and ambiguous results of non-surgical and surgical treatment determine the need for multidisciplinary approach and the development of protocols for the management of adult patients with neurogenic deformities of the spine.

**Key Words:** neurogenic spinal deformities, neuromuscular adult scoliosis, postural deformities in Parkinson's disease, spinal deformities in CP in adults.

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Neurogenic scoliosis is more often perceived as and associated with childhood deformities of the spine. This is due, above all, to early and rapid progression of deformities and identification of indications for surgical treatment already in the childhood. In adults, spinal deformities caused by various disruptions of neuromuscular regulation occur both as a result of neurological diseases or spinal cord lesions and as outcomes of childhood neuromuscular pathology, congenital anomalies in the development of the spine, injuries and tumors of the brain and spinal cord. Such deformities are characterized by complex nature of their pathogenetic mechanisms (accretion of degenerative changes), polymorphism of clinical manifestations, changes in bone density, and concomitant diseases. There is no

defined tactics for management and treatment of such patients.

The aim of the study was to use literature data to demonstrate features of clinical manifestations, diagnostic aspects and approaches to treatment of neurogenic deformities of the spine in adults.

Study design: Literature review.

## Material and Methods

The literature review was performed using PubMed, Medline, Web of Science, Scopus, CrossRef, AOSpine, Clinical Key, and eLibrary databases. Search period: 06.02.2017 to 04.11.2017.

Search keywords: neurogenic deformities of the spine in adults, spinal deformities in Parkinson's disease, spine deformities in neuromuscular diseases of various etiologies in adults, treatment tactics

for spine deformities in Parkinson's disease, treatment tactics in neuromuscular diseases in adults.

The inclusion criteria were: publications with search keywords with a level of evidence of 1–5.

The exclusion criteria were: publications on neuromuscular diseases without spinal deformity, Parkinson's disease without spinal deformity, spine deformity in children (under 18 years), non-clinical studies, reports.

## Results

### *Types of neurogenic deformities of the spine in adults*

In adult patients, the deformities can be outcomes of neuromuscular childhood scoliosis, congenital developmental anomalies (myelomeningocele, MMC),

curvature of the spine due to cerebral palsy, variants of spinal muscular atrophy (SMA) of types 3 and 4, Friedreich's ataxia, post-traumatic (brain and spinal cord injury) and postoperative (after removal of spinal cord tumors) [3, 31, 52] or can be caused by primary neurological diseases.

Many people with type 3 SMA survive to adulthood and are able to walk up to the age of 30–40 years, although some lose their ability to walk already in their youth [100]; in type 4 SMA (adult form of the disease), the human body remains soft along its entire length and the disease is characterized by atrophy of the muscles of the arms, legs and tongue, which appears after 35 years of age [37]. The average life expectancy for patients with Duchenne muscular dystrophy ranges from adolescence to 20–30 years [34]. In Friedreich's ataxia, scoliotic deformity develops in 60–79 % of cases [58]. The prognosis for patients is unfavorable and the average life expectancy from the onset of disease progression is 15–20 years [28].

The incidence of scoliosis (curvature more than 10° according to Cobb) in adults with cerebral palsy is 64 to 77 %. In this category of patients, pronounced deformities with sagittal imbalance occur in 7–10 % of cases [49, 54, 55, 74, 88].

A separate group of spinal deformities in adults includes those caused by disruption of neuromuscular regulation due to primary neurological diseases (Parkinson's disease, Alzheimer's disease, Lewy body disease, multiple systemic atrophy, progressive supranuclear palsy, Huntington's disease, subacute sclerosing panencephalitis, amyotrophic lateral sclerosis).

Parkinson's disease is a chronic progressive neurodegenerative disease. According to the literature [32, 59, 83], the incidence of the trunk deformities in this disease is 2 to 60 %. The central (trunk dystonia) and peripheral (myopathies, degenerative diseases of the spine) mechanisms are considered in the pathogenesis of postural deformities in Parkinson's disease [4, 59, 86].

#### *Features of neurogenic deformities of the spine in adults*

A particular feature of the outcomes of neuromuscular scoliosis is long, slightly-sloping arc of the thoracic and lumbar spine with a pronounced obliquity of the pelvis, presence of hyperkyphosis or hyperlordosis of the spine with corresponding violation of the trunk balance. Consequences of spine and thorax deformity include shortening of the trunk, decrease in the mobility of the ribs and diaphragm, significant displacement of the thorax organs. In patients with untreated paralytic scoliosis, the lower ribs of the concave side of deformity touch the wing of the ilium, which causes severe discomfort when sitting [8]. These patients have gross neurological manifestations of the underlying disease, they are limited in movement and even verticalization in the sitting position. Some diseases are accompanied by pronounced changes in the respiratory, cardiovascular, urinary systems. Patients are often emaciated, have problems with consumption and absorption of food, their paravertebral musculature is poorly developed, and they have osteopenia [2, 9].

Progression of congenital malformations of the spine in adulthood leads to multicomponent and multiplanar curvatures. For example, patients with MMC at the level of the thoracolumbar region are characterized by a pronounced kyphotic component of deformity, due to the anatomical features of the anomaly [38]. In addition to neurological disorders and muscle paralysis, the progression of the spinal deformity can be caused by hydrocephalus, disruption of liquor circulation and tethered spinal cord syndrome. Deformities in MMC are difficult to treat surgically due to changes in the vertebrae and ribs, shortening of the trunk, non-closure of the posterior elements of the spine, and compensatory lordosis in the thoracic spine [8].

The following types of postural deformities are distinguished in Parkinson's disease: antecollis (dropped head syndrome, 5–6 %) [14, 45], lateral flexion of the trunk (Pisa syndrome, 2–16.5 %) [22, 86, 99], camptocormia (bent spine

syndrome, 3–18 %) [10, 51] and scoliosis (9–33 %) [14, 17] (Fig.).

Manifestations of camptocormia and Pisa syndrome are alleviated in a horizontal position and aggravated under stress and fatigue. A feature that distinguishes Pisa syndrome from scoliosis in Parkinson's disease is the disappearance of deformity in the prone position. Three variants of the disease course are distinguished based on the rate of progression: rapidly progressing (acute), slowly progressing (subacute) and chronic (staged) [4, 59, 86].

In the opinion of some authors, the onset and progression of postural deformities [4, 36, 50] are associated with the duration and severity of the disease, age, duration of levodopa intake, concomitant dementia, whereas other authors do not believe they are correlated with these factors [10, 90].

Accretion of degenerative changes is typical for adult patients with neurogenic deformities of the spine. The curvature becomes rigid which may lead to secondary progression and cause pain and compression syndromes of osteochondrosis, secondary myelopathy and decompensation of somatic status to appear in the clinical presentation.

#### *Diagnostics*

The diagnosis of spinal deformities is based on clinical data, taking into account the etiology of the underlying disease. X-ray is used to determine the type and severity of deformity with an assessment of the position of the pelvis. Functional radiography (bending-test, maximum extension and flexion, lying and standing) is necessary to assess the rigidity (mobility) of a deformity.

The X-ray parameters of the postural deformities in Parkinson's disease are: for camptocormia, the thoracolumbar kyphosis of more than 45° in the standing position [10, 51], for antecollis, cervical kyphosis of more than 45° [14, 45], for Pisa syndrome, lateral deflexion of the spine of more than 10° [32] and 15° in the standing position [22], for scoliosis, structural lateral curvature more than 10° according to Cobb with vertebral rotation at the apex of deformity, which does not change in the prone position [32].

MRI and electromyography of muscles of the extremities and paravertebral muscles, *m. rectus abdominis*, *m. iliopsoas* with biopsy and pathohistological investigation are necessary for differential diagnosis of pathological changes in muscles, assessment of their condition, including planning for treatment methods (antispastic therapy for cerebral palsy or botulinum therapy for Parkinson's disease).

One of the stages of the examination is the evaluation of the severity of degenerative changes in the spine and their neurological manifestations to identify indications for decompression and stabilization manipulations or pain therapy.

Functional assessment of the state of the respiratory system in patients with neuromuscular diseases is necessary to determine indications for non-invasive ventilation, for planning preparatory procedures prior to a surgery, anesthesia during interventions and patient management in the postoperative period. Currently, there is no traditionally accepted and proven lower limit of the vital capacity of the lungs, which defines feasibility of spinal surgery in patients with neuromuscular diseases [62].

The diagnostic algorithm includes the assessment of the condition and function of the cardiovascular system (ECG, Holter monitoring, echocardiography).

Laboratory tests and genetic analysis are performed to verify the etiology of the disease.

It is necessary to evaluate bone mineral density (dual-energy X-ray osteodensitometry).

Assessment of the functional status of patients with cerebral palsy is carried out on the basis of the clinical form and Gross Motor Function Classification System (GMFCS), Communication Function Classification System (CFCS) and assessment of patient mobility using Functional Mobility Scale (FMS).

For a unified analysis of the condition of patients with Parkinson's disease the following scales are used: Hoehn and Yahr (H&Y) scale for the assessment of severity, Unified Parkinson's Disease Rating Scale (UPDRS) for assessment of the severity of the main symptoms, Schwab

et England scale for assessment of daily activity, PDQ-39 scale for assessment of the quality of patients life, CC scale for assessment of the severity of camptocormia and pain syndrome. Assessment of the effect of dopaminergic drugs is based on the patient's diary.

The Russian-language version of the SRS-22 questionnaire is recommended for self-assessment of functional status in patients with no cognitive impairment [5].

Multidisciplinary approach should be implemented at all stages of treatment: joint observation by physicians with different specialties (neurologist, spinal surgeon, rehabilitologist, clinical psychologist).

#### *Frontal and sagittal balance of the trunk*

Curvature of the spine in the frontal plane is an integral part of neuromuscular deformities. Many researchers have studied the correlation between the spinal deformity in the frontal plane and clinical manifestations of diseases.

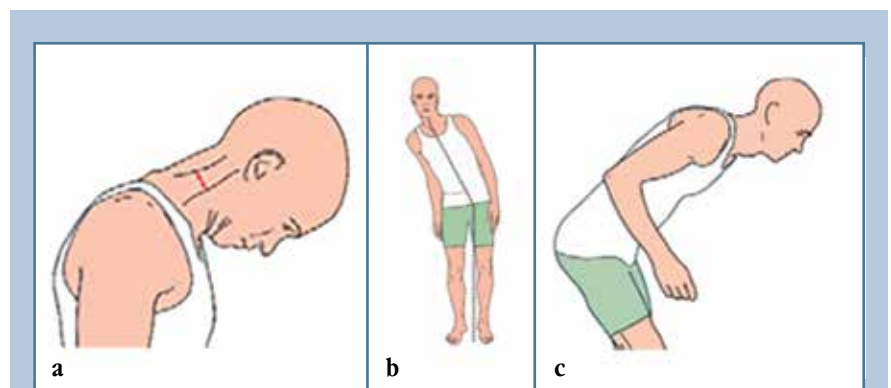
The frequency and type of scoliosis in cerebral palsy are correlated with the severity of neurological disorders and changes in motor functions (GMFCS). In adults, the curvature of more than 40° is more often diagnosed in quadriplegia (30 % of cases) than in diplegia (10 % of patients) [1, 96]. Scoliosis is detected more often in the spastic form of cerebral palsy than in the paralytic one [49, 55].

No correlation has been found between the dominant side of the Parkinson's disease symptoms and the direction of curvature, but it has been established that postural deformities in the frontal plane are interrelated with the morphological changes of the spine and the apex of scoliosis in radiography [99]. Data on the dependence of the frontal imbalance on the severity and duration of the underlying disease, as well as on the patient's age, are contradictory [26, 89].

In modern spinal surgery, understanding the principles of sagittal balance is a prerequisite for achieving good outcomes in the treatment of various diseases of the spine, especially its deformities.

The assessment of the sagittal balance by standard method is impossible in patients with neuromuscular diseases accompanied by movement restrictions. The change in the magnitudes of lumbar lordosis and thoracic kyphosis, the patient's condition in the sitting position (forced positions of the limbs, head tilt, etc.) can be used as the indirect criteria.

The study of sagittal balance in patients with Parkinson's disease, conducted by Oh et al. [67], identified characteristic changes in the main indices. Bissolotti et al. [19, 20, 21] identified the correlation between the indices. Predisposing factors for disruption of the sagittal balance are gender, duration and



**Fig.** Types of postural deformities in Parkinson's disease: **a** – dropped head syndrome; **b** – Pisa syndrome; **c** – camptocormia (bent spine syndrome) [33]

severity of the disease (H&Y and UPDRS scales) [19, 67]. Additional factors affecting the sagittal balance are degenerative changes in the spine [21, 63]. Despite the ambiguous results, the researchers propose to continue studying the sagittal balance and believe that it necessary to take into account its parameters in any treatment options for patients with spinal deformities.

#### *Non-surgical methods of treatment*

In case of etiotropic treatment, the therapy is always based on the treatment of the underlying disease.

All variants of postural deformities in Parkinson's disease do not respond well to anti-Parkinson's therapy (levodopa, dopamine receptor agonists, amantadines, anticholinergic drugs) [4, 7, 25, 30, 32]. Correction of drug therapy in a form of exclusion or replacement of the main drug is appropriate in diagnostics of postural deformities [4, 7].

Physical therapy, corset therapy, including wearing a backpack (in Parkinson's disease) do not have sufficient evidence base [32]. Recommendations for physical therapy and corset therapy are the duration of the sessions, the patient's motivation, contact with the psychologist, and the lack of cognitive changes [29, 68].

Pain syndrome in the spine and paravertebral muscles of varying intensity occurs in 72–79 % of adult patients with cerebral palsy [12, 43]. Two surgical procedures have been developed to reduce the spasticity of paravertebral muscles and reduce the pain syndrome in cerebral palsy: selective dorsal lumbar rhizotomy (SDR) and implantation of the intrathecal baclofen pump (ITB) in patients with severe motor function limitations (GMFCS levels IV and V). Publications on the use of these techniques in adult patients indicate the relief of pain syndrome for up to 4 months [12, 39, 40, 49, 72]. There are no studies on the effect of these procedures on the magnitude of deformity in adults (Table 1).

Data on the effectiveness of injecting botulinum toxin into paravertebral muscles, *m. rectus abdominis*, *m. iliopsoas* in patients with Parkinson's disease are ambiguous [15, 56, 93]. A blind cross-sectional study by Bonanni et al. [22] showed

no improvement in the group of patients who received placebo, which indicates the effectiveness of the method. According to the results of Dupeyron et al. [33], the right choice of muscles for injection allows achieving a persistent positive outcome for the period of up to 1 year. An unsatisfactory result may be due to the uncertainty regarding the muscles for injections and adequate doses of drugs [4]. At present, there is no protocol for botulinum therapy based on the results of electromyography and identification of muscles for subsequent administration of the drug [59]. Therefore, this method requires further studies and development of practical recommendations.

Deep brain stimulation (DBS) as a method of treating postural deformities in Parkinson's disease shows results ranging from significant improvement [35, 42, 60, 76, 97, 98] to negligible one [6, 25, 69, 94, 98] or absence of improvement [4, 15, 79, 81, 92] in single observations. The method involves unilateral or bilateral implantation of electrodes into the deep cores of the brain (subthalamic nucleus STN or globus pallidus internus GPI) followed by one-stage or chronic electrostimulation. In the largest study by Umemura et al. [91] (8 patients with camptocormia, 10 patients with Pisa syndrome), the bilateral DBS STN proved to be ineffective in 6 cases. In patients with mild to moderate deformity, a positive outcome was observed already in the early postoperative period. In severe types of deformity, the improvement progressed gradually over a long period of time after the surgery. The use of unilateral (both ipsi- and contralateral) DBS STN for Pisa syndrome did not lead to a persistent clinical effect [59]. Sakas et al. [75] presented two cases of the use of DBS GPI in young patients with a short-term (for several days) and long-term (42–44 months) clinical effect.

The low efficiency of the procedure in some cases is caused by pronounced degenerative changes in the spine (spondylosis), especially at the level of the cervicothoracic junction [64, 84]. Therefore, the use of DBS is advisable in the early stages of the disease, before the develop-

ment of rigid deformities [91]. A meta-analysis published by Weaver et al. [95] demonstrated the need for randomized controlled trials to evaluate the effectiveness of DBS.

There are publications on the use of noninvasive technique of transspinal magnetic stimulation (TSMS) in treatment of patients with postural deformities in Parkinson's disease [13, 56]. Arii et al. [13] conducted a randomized, double-blind, cross-sectional, placebo-controlled trial of TSMS efficacy in 320 patients with camptocormia. A temporary positive effect was noted with the need for repeated courses to analyze the outcomes [56, 87].

Stereotactic destruction (ventrolateral thalamotomy, pallidotomy, subthalomotomy and their combination) is mainly used in postural instability and gait disorders without long-term effect due to low efficiency [11] (Table 2).

#### *Surgical treatment and its complications*

The publications devoted to the analysis of surgical treatment of adult patients with neuromuscular diseases are rare: they either refer to individual clinical cases or involve larger cohorts of patients, including children (Table 1). The greatest correction of deformity is achieved with using segmental transpedicular fixation involving the sacrum and pelvis [38, 57, 61]. Preoperative halo-traction with respiratory support are used as a preparatory stage [18, 46].

Discotomies [57, 73, 82] or vertebro-tomies are performed to create mobility in the spine in case of rigid, multi-plane deformities, and the type of surgery depends on the severity of curvature and mobility [24, 38, 44, 53, 85]. It is recommended to perform vertebro-tomy with corrective manipulation under the control of intraoperative neuromonitoring [41, 85].

The largest group of adults with scoliosis in cerebral palsy is analyzed in the study by Modi et al. [61]: 35 patients over 18 years of age (a total of 52). Segmented posterior instrumental fixation involving the pelvis allowed to achieve correction of scoliosis by 63 % with improvement in the functional status of patients. Suh et al. [85] achieved 53–66 % correction

Table 1

Outcomes of treatment of adult patients with neuromuscular diseases (according to the literature)

Authors	Underlying disease	Number of patients, n (age)	Treatment method	Outcome	Complications
Koop et al. [50]	CP	1 (21 years)	Selective dosral lumbar rhizotomy	Pain syndrome relief	Scoliosis progression of 10° over the course of two years
Reynolds et al. [73]	CP (spastic diplegia)	21 (>18 years)	Selective dosral lumbar rhizotomy	Reduction of pain syndrome (for the period of 4 months)	None
Fuerderer et al. [39]	Myelomeningocele; kyphosis	1 (20 years)	Transpedicular screw fixation + vertebrotoomy at the apex of the kyphosis	23 % kyphosis correction	No data
Sink et al. [83]	CP	41 (9–36 years)	Luque –Galveston construction; front release: 21; anterior spondylodesis: 12	Correction of the deformity was achieved in 19 patients	Proximal junctional kyphosis: 11 (32 %); migration of distal support: 11
Mertz et al. [58]	Neuromuscular scoliosis	1 (45 years)	Anterior release of T7–L2; transpedicular screw fixation of T4–L4	Correction of the curvature in the thoracic spine: 43%, in the lumbar: 46 %	None
Modi et al. [62]	CP	A total of 52 (aged 8 to 38 years): older than 18 years – 35	Transpedicular screw fixation – 10	Correction of scoliosis: 62,9 %, improvement of functional status	32 % (n = 52) lethal outcome: 2; neurological deficit: 1
Suh et al. [86]	CP – 7; spinal muscular atrophy - 1	8 (older than 18 years)	Transpedicular screw fixation + VCR of 4–5 vertebrae on the apex of the deformity, neuromonitoring	Correction of spastic forms: 53.6 %, paralytic: 66.2 %	Migration of 3 screws after the corrective maneuver
Piazzolla et al. [72]	Neuromuscular scoliosis	A total of 24: from 20 to 30 years: 6; older than 30 years: 1	CDI	Correction of scoliosis: 57.2 %, correction of hyperlordosis and hyperkyphosis	8.3 % (n = 24), lethal outcome: 1; deep infection: 1
Bao et al. [18]	Neuromuscular scoliosis; respiratory dysfunction	21 (older than 18 years) Neuromuscular scoliosis: 7	Halo-gravity traction + respiratory preparation (non-invasive ventilation)	Correction of scoliosis by 23.6°, improvement of respiratory indices	None

of the deformity in treatment of 8 adult patients with cerebral palsy using transpedicular fixation and VCR vertebrotoomy of 4–5 vertebrae. Piazzolla et al. [71] achieved correction of scoliosis by 57 % with correction of sagittal balance using CDI in 7 adult patients with neuromuscular scoliosis (Table 1).

It is impossible to assess the rate of complications of surgical interventions in adult patients with neuromuscular diseases based on the available publications. In 2017, Cognetti et al. [27] published the SRS report on complications and mortality after surgical interventions in patients with neuromuscular diseases over the period of 12 years (2004–

2015). The average age of patients with complications was  $15.0 \pm 7.2$  years, the average rate of complications was 6.3 %. Compared to 2004, there has been a 10 % decrease in complications, with a result of 4.4 % in 2015, with an almost identical mortality rate (0.34 %). Main types of complications: infectious ( $3.2 \pm 0.3$  %) including superficial inflammation

(0.55–1.0 %) and deep infection (2.0–2.7 %), neurological (0.5 %) and respiratory (0.08 %) [27].

Spinal surgery in Parkinson's disease is characterized not merely by technical complexity of the intervention, but rather by high rate of complications and repeated interventions [16, 30, 78]. This is due to many factors: decrease in mineral density of bone tissue, muscle condition (atrophy, fibrosis, adipose degeneration), age of patients, clinical manifestations of the disease (tremor, gait disorders, dementia). These factors define the indications for an extended instrumental fixation from the upper thoracic spine to the sacrum and pelvic bones [23, 48, 78] with mandatory additional fusion with auto- or allograft [69].

The analysis of the results of surgical treatment of different magnitude and levels of fixation revealed that repeated interventions were performed in 33.3–86.0 % of cases [16, 23, 48, 77, 80]. The main reasons for this are: instability of instrumentation [48, 77], progression of the deformity [77, 81], and infectious complications [48, 77]. Clinical and radiologic outcomes of treatment are defined by the authors both as satisfactory [23] and as unsatisfactory [63, 77] (Table 2).

A particular problem is manifestation of degenerative diseases of the spine: neurological disorders and pain syndromes. Accretion of pathogenetic changes and symptoms of diseases creates difficulties in choosing the tactics of treatment, especially when determining indications for surgical intervention.

Based on the results of a multicenter cross-sectional study, Nakane et al. [65] concluded that surgical interventions on the spine in the case of camptocormia lead to the progression of torsion dystonia. Some researchers indicate an increase in instability of fixation after osteotomies [23].

According to some authors [23], correction of sagittal balance is a key aspect in the surgical treatment of spine deformities in patients with Parkinson's disease. Other researchers [48] indicate that surgery for spinal deformities in Parkinson's disease does not require correction of the sagittal balance and that good

clinical outcomes with satisfaction from treatment can be achieved with different variants of interventions.

Given high rate of postoperative complications and repeated interventions, indications for stabilization should be considered only after failure of other minimally invasive methods of treatment [23, 70, 78, 92]. In this case, indications for surgery are defined by both neurological manifestations and limited mobility of patients and problems with self-care, dependence on others [23, 47, 66].

## Discussion

At present, there is no consensus on the treatment of patients with neurogenic deformities of the spine [23, 77, 92].

The treatment of adult patients with outcomes of neuromuscular diseases is not regulated and is represented only by single cases or small samples when analyzing the results of interventions in children.

Diagnostic criteria, severity scales, and algorithms of drug therapy are developed and applied in postural deformities in neurodegenerative diseases (Parkinson's disease), but only with reference to the underlying disease.

The protocol of management of patients with Parkinson's disease approved in 2005 defines the diagnostic, pharmacological, rehabilitation and social aspects of patients, but does not regulate the tactics of management in cases of postural deformities and indications for surgical treatment [7].

The analysis of published data make it possible to define general trends in the treatment of patients with neurogenic deformities of the spine: protocols of drug therapy of the underlying disease with correction according to indications are well established; the effectiveness of antispastic therapy is debatable; the results of stimulation of deep structures of the brain are ambiguous; the effectiveness of rehabilitation programs has no evidence base; the indications and volumes of surgical interventions are not defined; the rate of complications and repeated interventions in spinal surgery is high. Therefore, the tactics of treat-

ment and management of this category of patients have not been defined to date.

Despite the similarity of pathogenetic mechanisms, scoliosis caused by childhood neuromuscular diseases (outcomes of deformities) and curvatures due to primary neurological diseases (mainly neurodegenerative nature) have significant differences.

Outcomes of neuromuscular and congenital scoliosis in case of their progression are characterized by extensive, rigid, multiplanar curvatures that affect all parts of the spine in adulthood. It is caused by a long-term progressive course of the disease, and sometimes by the outcome of the underlying disease with pronounced muscular tissue damage, sub- or decompensation of the respiratory and cardiovascular systems, osteoporosis, and accretion of degenerative changes. Correction of such deformities requires extensive fixation, multilevel vertebro-tomy, preserved muscle and subcutaneous fat for tissue healing after osteosynthesis, compensated state of vital functions. In some cases, such interventions are impossible. The risk assessment for interventions, which accounts for possible complications and potential outcome, defines the approach to surgery in these patients. The management algorithms should include identification of the main clinical symptoms and potential outcomes of non-invasive, minimally invasive or other options for patient care.

Such algorithms have not been developed yet for this category of patients.

The therapeutic algorithms presented by some authors for patients with Parkinson's disease either define only the general directions of treatment [59] or are focused on the diagnosis of the main pathogenetic processes [56] and do not reflect the clinical aspect and do not define the stages of surgery.

The treatment algorithm by Upadhyaya et al. [92] is the most interesting one and has the highest practical value. In their analysis of the effectiveness of different methods of treating patients with spinal deformities in Parkinson's disease, the authors compare the diagnosis, pathogenetic and clinical aspects and directions of both conservative and

Table 2

Outcomes of treatment of adult patients with spinal deformities in Parkinson's disease (according to the literature)

Authors	Treatment method	Number of patients, n (age)	Outcomes of treatment	Complications
Bonanni et al. [22]	Botulinus toxin in the paravertebral muscles	9: main group - 5, control group - 4; group change	control group: no effect; the main group: varying degrees and duration of the effect	None
Tassorelli et al. [87]	Botulinus toxin into <i>m. iliopsoas</i> , into <i>m. rectus abdominis</i>	41: main group: 20; control group: 21	The main group: pain reduction and improvement of clinical manifestations for a longer period of time than in the control group	None
Sako et al. [77]	Stimulation of the subthalamic nucleus	6 (53–71 years)	The decrease in camptocormia by $78 \pm 9.1$ %; reduction in symptoms of Parkinson's disease (17 months)	None
Umemura et al. [92]	Stimulation of the subthalamic nucleus	18: camptocormia: 8 (59–79 years), Pisa syndrome: 10	The decrease in camptocormia in 5 patients (12 months); reduction in symptoms of Parkinson's disease in 7 patients (12 months)	None
Yamada et al. [99]	Stimulation of the subthalamic nucleus	Camptocormia: 17	Decrease in deformity (assessment based on a photo in standing position)	None
Arii et al. [13]	Transspinal magnetic stimulation	37: main group: 19; control group: 18	Reduction in the angle of the kyphosis while standing: main group: $10.9^\circ$ ; control: $0.8^\circ$ , short-term	None
Babat et al. [16]	Transpedicular screw fixation from T4 to the sacrum: 1; transpedicular screw fixation of 1–2 levels (L): 5; laminectomy: 2; decompression of 3 levels (L): 3; ACDF: 1; fixation of the cervical spine (lateral masses): 2	14 (51–79 years)	Reoperations – 12 (86 %)	Proximal junctional failure; spondylolisthesis; infection; scoliosis; restenosis; kyphosis; instability of the instrumentation
Koller et al. [49]	Transpedicular screw fixation from T to the sacrum: 15; transpedicular screw fixation of 3 levels: 18; anterior fusion: 15; PSO: 1; SPO: 7	23 (57–76 years), of them, 10 repeated surgeries	Correction of the sagittal balance: 25%; satisfaction with the clinical outcome: 78 %; partial satisfaction: 22 %	Medical complications: 7 (30.4 %); surgical: 12 (52.2 %); require correction of the sagittal balance: 33 %
Bourghli et al. [23]	Transpedicular screw fixation from T2 to the sacrum: 6 primary, 6 repetitive, SPO: 6	12 ( $68.0 \pm 6.2$ years)	Good clinical, radiologic outcome with satisfaction with the outcome of treatment; re-operations: 6 (50 %)	Instability of the instrumentation: 3; proximal junctional failure: 2; epidural hematoma: 1
Moon et al. [64]	Lumbar fusion: 1 level: 14; 2 levels: 5; 3 levels: 1	20 (48–81 years)	VAS before surgery 53.9; VAS after surgery 55.2; fusion: 15, without fusion: 5; poor clinical outcome: 14; increased dyskinesia: 4	No data

surgical treatment. In their opinion, non-surgical or minimally invasive treatment methods are preferable in this category of patients. Diagnosis of clinical manifestations of myopathy, myelopathy and radiculopathy determines, in comparison with diagnostic tests, the indications and volume of decompression interventions, the extent and levels of fixation of the spine. Adapted, extended and refined algorithm can become the basis of the protocol for managing patients with neurogenic deformities of the spine.

### Conclusion

The analysis of neurogenic scoliosis in adults which arises as a result of congenital disease or newly emerging deformity due to acquired disease

(most often Parkinson's disease or muscular dystonia) reveals high rate of postoperative complications and repeated interventions. It should be emphasized that for this disease the incidence of complications of non-surgical treatment is significantly lower than the incidence of surgical complications (up to 80 %). There are indications that surgical correction of the deformity is impossible in case of pronounced muscular atrophy. The investigation of the spinal cord specimens from patients with neurogenic scoliosis often indicates the development of myelopathy without signs of pronounced compression in MRI, apparently due to postural changes and gross deformities, which requires additional study. Indications for

correction of neuromuscular deformity in adults can be the following: failure of drug therapy and neurosurgical treatment, drastic decrease in the quality of life as a result of the deformity, the absence of pronounced dorsal muscle atrophy.

The multicomponent pathogenetic mechanisms, the ambiguity of the results of both non-surgical and surgical treatment call for a multidisciplinary approach and the development of protocols for the management of adult patients with neurogenic deformities of the spine.

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