SPINAL MUSCULAR ATROPHY: CLINICAL FEATURES AND TREATMENT OF SPINAL AND LIMB DEFORMITIES

Interstate Consensus Protocol

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Objective. To substantiate the protocol for the diagnosis and treatment of deformities of the spine and limbs in patients with spinal muscular atrophy basing on an assessment of the level of evidence of published data.

Material and Methods. Data on foreign protocols and their adaptation for use in Russia and CIS countries were analyzed and summarized. The main platform was the evidence-based systematization of studies reflecting modern approaches to the diagnosis and treatment (including surgery) of spinal and limb deformities in patients with spinal muscular atrophy. The formulated recommendations are based on literature data and the authors' own experience. Literature was searched in online databases of Medline, Embase, Web of Science, and Cochrane Library information platforms. Preference was given to studies that could be classified as evidence level 2+ and higher according to the ASMOK system. References are given in the order of their mention in the text. Search depth was 5 years. Methods used to assess the quality and strength of evidence were expert consensus and significance assessment in accordance with the rating scheme. Methods used to analyze evidence were reviews of published meta-analyzes and systematic reviews with evidence tables.

Results. Various aspects of clinical examination, respiratory support and postural control, conservative and surgical treatment of spinal and limb deformities, preoperative, intraoperative and postoperative management, and anesthetic risk assessment in patients with spinal muscular atrophy are highlighted.

Conclusion. Secondary orthopedic pathology in patients with spinal muscular atrophy causes not only severe violation of the musculoskeletal system functions (support, movement, and verticalization), but also pathological changes in the vital functions of internal organs and systems (respiratory, digestive, cardiovascular). A thorough analysis of the patient's condition (assessment of general somatic, neurological, and orthopedic statuses) based on the data of preoperative multidisciplinary examination allows assessing the risks of complications and developing individual program of surgical rehabilitation of the patient. Surgical correction of orthopedic pathology in spinal muscular atrophy improves the functional status of the patient, improves the quality of life and the level of self-care, and optimizes the function of external respiration.

Key Words: spinal muscular atrophy, spinal deformities, limb deformities, orthopedic syndrome, spine surgery, conservative treatment, surgical treatment.

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Spinal and limb deformities in patients with spinal muscular atrophy (SMA) should be analyzed on the basis of interdisciplinary assessment of the prognosis, management, and treatment of patients. SMA is a group of disorders requiring various approaches to patient care and involvement of a wide range of specialists (Fig. 1). The literature focuses on the fact that coordination of the interdisciplinary approach is usually performed by a neurologist or pediatric neurologist who is aware of the disease course and potential problems. This enables control of various aspects of the patient's condition, which lead to progression of the disease, and preventive care when possible [1–4].

The purpose of this study was to substantiate the protocol for diagnosis and treatment of spinal and limb deformities in SMA patients based on assessment of the evidence level of published data.

Design

Methods used for collection/selection of evidence included search in electronic databases. Preference was given to publications that might be classified as evidence level 2+ and higher according to the ASMOK system. References are given in the order of their mention in the text.

Description of the methods used to assess the quality and strength of evi-

dence: the evidence base for recommendations was publications included in the Medline, Embase, Web of Science, and Cochrane Library. The search depth was 5 years.

Methods used to assess the quality and strength of evidence:

– consensus of experts;

– assessment of significance in accordance with the rating scheme (the scheme is supplied).

Methods used to analyze evidence:

– reviews of published meta-analyzes;

– systematic reviews with tables of evidence.

Description of the methods used to analyze evidence: upon selection of publications as potential sources of evidence, the technique used in each study was analyzed separately to assess its validity. The result of this analysis affected the evidence level assigned to the publication, which in turn influenced the strength of the recommendations. To minimize potential errors, each study was evaluated independently. Any differences in assessments were discussed by all members of the working group. If consensus was not reached, an independent expert was involved.

Tables of evidence were filled in by the authors of the consensus (Tables 1, 2); all information was graded by the level of reliability/evidence (Table 1) depending on the quantity and quality of studies on a particular issue.

Methods used to assess the quality and strength of evidence:

– consensus of experts;

– assessment of the evidence level in accordance with the rating scheme.

Good Practice Points (GPPs): the recommended good practice is based on clinical experience of the authors of the developed recommendations.

Economic analysis: no cost analysis was performed, and no publications on pharmacoeconomics were analyzed.

Guidelines validation method: external expert evaluation; internal expert evaluation.

Description of the guidelines validation method. A draft of these guidelines was reviewed by independent experts who were first of all asked to comment on how understandable is the interpretation of the evidence underlying the guidelines. All comments of experts were carefully systematized and discussed by working group members (guidelines' authors). Each item was discussed separately.

Consultation and expert assessment. The draft guidelines were reviewed by independent experts who were first asked to comment on the intelligibility and accuracy of the interpretation of the evidence base underlying the recommendations.

Working group. For the final revision and quality control, the guidelines were re-analyzed by working group members who came to the conclusion that all expert comments and suggestions were taken into account, and the risk of systematic errors during development of the guidelines was minimized.

Key recommendations. The grades (strength) of recommendations (Table 2) based on the appropriate levels of evidence are given in the References at the end of each citation.

Clinical Examination

Assessment of the SMA patient involves a physical examination with allowance for functional disorders of the musculoskeletal system. A set of assessment methods is determined by relevant aspects for each severity degree. Examinations of SMA patients should include various methods of assessing muscle strength and amplitude of joint movements, motor functional scales, and time tests to monitor functions and activities in everyday life [5–9] (Table 3).

Scoliosis is a typical orthopedic manifestation of SMA type 1, 2, and 3. The incidence rate is 60–90 % with onset in early childhood [2, 10]. The spinal curvature in children with reduced muscle tone continuously progresses throughout childhood. Also, most patients develop thoracic spine hyperkyphosis of varying severity. The spine should be examined as part of a routine clinical examination. If kyphoscoliosis is suspected, an examination in a standing or sitting position and in the Adams position (forward bending) is required. In the presence of spinal deformity, radiographs of the spine are performed in the frontal (anteroposterior) and lateral projections in the most possible vertical position of the patient achieved independently of assistive devices (i.e., in a sitting position in children who can sit on their own and in a standing position in SMA type 3) to quantify the degree of spinal deformity in both the frontal and sagittal planes.

In patients with SMA type 1 and 2 and scoliosis and with a Cobb angle of more than 20°, examination should be performed every 6 months until the end of growth (Risser IV or more) and then, after full skeleton development, annually.

Commentary. Recommended examinations should be performed by specialists regularly every 6 months or more often in the case of certain indications (new clinical signs of pathology and worsening of the condition). In this case, the frequency of examinations should be determined by a dedicated expert or a multidisciplinary team.

Regular monitoring enables detection of possible changes and timely definition of indications for corset therapy or surgery with allowance for the therapeutic and age corridor, as well as monitoring of their result [11, 12].

Methods of Physical Examination

Physical examination of SMA patients should be performed taking into account functional disorders of the musculoskeletal system:

– muscle strength tests;

– identification of limb contractures;

– analysis of the frontal and sagittal balance of the spine with assessment of C7 deviation from the central sacral vertical line (CSVL) and posterior sacral vertical line (PSVL);

– traction test for assessing spinal mobility;

– X-ray and clinical analysis of the pelvic position in the sagittal and frontal projections;

– control of the pain intensity (if present) using VAS;

– determination of BMI (body mass index).

Methods of Instrumental Examination

Radiographic examination should include all spinal departments (cervical, thoracic, and lumbar) with involvement of the pelvis and hip joints in the frontal and lateral projections in a sitting (in patients verticalized in the sitting position) or lying (in non-verticalized patients) position. X-ray telemetry of the spine is optimal. Radiography should be performed each year if the spinal curvature is less than 15–20°, and every 6 months if the deformity is more than 20°, until skeleton maturation (Risser I–III). An increase in the time interval between radiographies over 1 year may lead to missing progression of scoliosis. After skeleton maturation, the decision on radiography is made on the basis of clinical assessment of the patient's functional state.

Before surgery, CT of the cervical, thoracic, and lumbar spine is performed; MRI is carried out if signs of myelopathy are present.

Echocardiography is performed if indicated as well as when preparing for surgery.

Nocturnal pulse oximetry or cardiorespiratory monitoring is performed if indicated as well as when preparing for surgery.

ECG is performed as part of preoperative examination and identification of indications for cardiotropic therapy.

A pulmonary function test and osteodensitometry are performed according to similar indications as well as to identify indications for therapy.

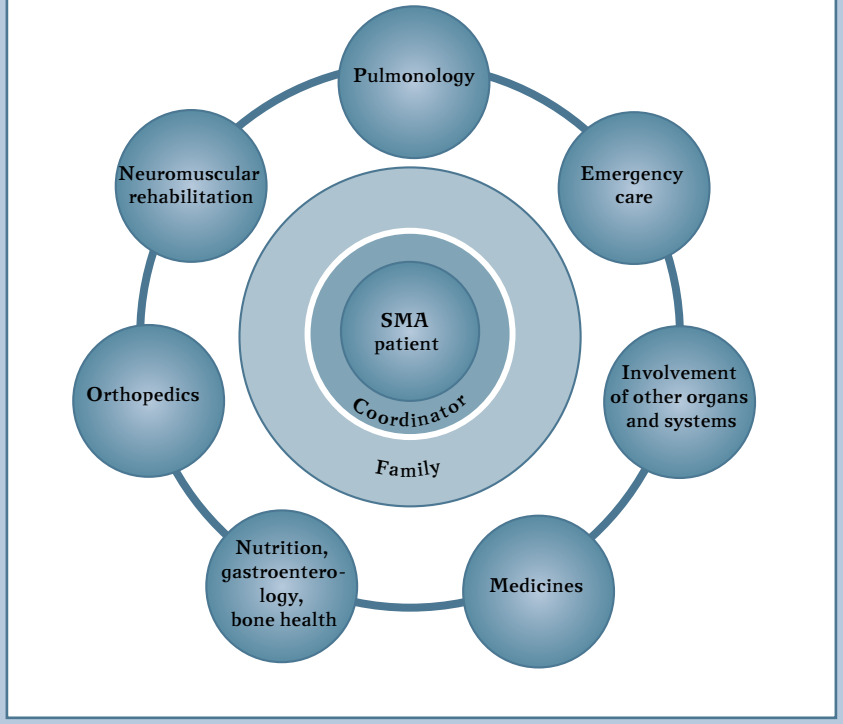

Fig. 1
Diagram of an interdisciplinary approach to assessing the status of patients with spinal muscular atrophy (SMA) [3]

Table 1
Levels of evidence

Level of evidence	Data type
1a	Meta-analysis of randomized controlled trials
1b	At least one randomized controlled trial
2a	At least one well-performed controlled non-randomized trial
2b	At least one well-performed quasi-experimental study
3	Well-performed non-experimental studies: comparative, correlation, or case control
4	Expert consensus opinion or clinical experience

Conservative Treatment

Rehabilitation of mobile patients (ambulatory stage)

The main tasks of rehabilitation of patients at the ambulatory stage are to restore, improve, or maintain mobility function, a sufficient range of motion in the limb joints, as well as improving balance and endurance.

Exercise/physical activity programs. Physical activity programs should include dynamic and static exercises for balance. Swimming, walking, cycling, yoga, hippotherapy, rowing, aerobics exercises, and general developmental workouts are recommended. The exercise program should be developed and supervised by a physical therapy specialist or occupational therapist with competencies in SMA. The optimal duration of aerobics exercises is at least 30 min.

Orthosis/prevention of contractures is aimed at maintaining activity and preventing the formation of contractures. Tactically, this is achieved via combina-

Table 2
Strength of recommendations

Grade	Level of evidence	Basis of recommendation
A	High	Large double-blind, placebo-controlled trials as well as data obtained from meta-analysis of several randomized controlled trials (evidence level I)
B	Moderate	Small randomized and controlled trials where statistics are based on a small number of patients (evidence levels I, II)
C	Low	Non-randomized clinical trials in a limited number of patients (evidence level IV or extrapolation of data from studies of levels II and III)
D	Very low	Consensus of an expert group on a particular problem (evidence level V; either unapproved or inconclusive trial of any level)

Levels of evidence and strength of recommendations are presented in references at the end of each citation.

tion of redressement and orthosis techniques. The minimum stretching frequency is 2–3 times a week, the optimal frequency is 3–5 times a week. AFO and KAFO orthoses of the lower limbs are mainly used. TLSO corsets are usually used to improve sitting position and are not used during walking because they can adversely affect mobility and limit effective compensatory abilities.

Positioning. Spinal orthoses are often used to support weakened muscle tone of the spine and to treat scoliosis of more than 20°, especially in children in a growth spurt [13, 14].

Commentary. There is currently no consensus regarding the type of orthosis that should be used in this category of patients. Both rigid and soft spinal thoracolumbar orthoses with an abdominal window are recommended.

Mobility devices and exercises. If endurance of patients is restricted, it is recommended to use lightweight manual wheelchairs or power wheelchairs to ensure mobility. For traveling independently over long distances, power wheelchairs or motorized scooters may be considered.

Rehabilitation of patients verticalized in a sitting position (early non-ambulatory stage)

The main objectives of rehabilitation in sitting patients (early non-ambulatory stage) are prevention of contractures and scoliosis, maintenance, restoration, and improvement of functions and mobility [3].

Prevention of contractures and control of range of motion. Stretching techniques include methods that can be achieved manually and using active auxiliary stretching, various verticalizers, orthoses, and staged gypsum casting techniques. Stretching procedures should be performed and/or supervised by a physical fitness specialist or occupational therapist. Parents and nurses should also be instructed about application of these methods in everyday life. The focus should be placed on the joints that are most at risk of contractures: hip, knee, ulnar, and carpal joints. The minimum frequency of exercise is 5–7 times a week.

Orthosis/positioning. Thoracolumbosacral orthoses (TLSO) are recommended for improving posture and function. Neck immobilization should be used to ensure transportation safety. Orthoses should be removed 60 min before bedtime. The minimum frequency of orthosis use is 5 times a week.

The use of various verticalizers is recommended for static axial load, prevention of contractures of the lower limbs, improvement of the spine and whole body posture, improvement of body functions, and preservation of bone density. The maintained vertical position of the patient should be no longer than 60 min, the minimum execution frequency is 3–5 times a week, and the optimal frequency is 5–7 times a week. To maintain posture and standing ability, knee immobilizers, KAFO, AFO, TLSO, and arm splinters are recommended.

Mobility devices and exercises. All patients who can be verticalized only in a sitting position should have power wheelchairs or a customized chair to maintain posture (like a turtle). In patients after two years of age, the use of power wheelchairs should be assessed [15]. If the patient has preserved the function of the upper limbs and sufficient muscle strength, lightweight manual wheelchairs or power wheelchairs may be recommended to stimulate independent movement.

Commentary. For rehabilitation of patients verticalized in a sitting position, water procedures, aerobic training, concentric and eccentric exercises, and general training with and without resistance are recommended.

Manual techniques. Manual techniques involving changes in the body position, percussion massage, and vibration are important for patients, especially with SMA type 2, as they facilitate drainage of the respiratory tract. This is necessary for prevention of complications, especially in the perioperative period.

Table 3 (the ending is on page 84)
Rehabilitation evaluation and procedures recommended for patients with spinal muscular atrophy (SMA) [3]

Patients	Evaluation	Medical intervention	Care recommendations
Non-verticalized (bed) patients: late non-ambulatory stage	Postural monitoring; scoliosis; hip lability; ability to sit; chest deformity	Postural control and orthosis. Daily use of devices for sitting, supporting, and maintaining posture, chest braces and neck braces to support the head. Static chest fixation for respiratory support with an abdominal window. Surgical correction of spinal deformities	For effective use of orthoses, they should be removed 60 min before bedtime. The duration of training for effective stretching and increasing the range of motion depends on specific needs of the patient, condition of the joints, and goals of rehabilitation
	Contractures (goniometry, ROM)	Stretching. Daily use of orthoses for the upper limbs for stretching and preserving function and range of motion. Static orthoses. To maintain posture and stretching, it is recommended to use knee braces and arm splints. AFO, KAFO, and HKAFO orthoses can be used to stretch and support posture, TLSO – to support posture. Standing support. WHFO orthoses are recommended for control of the hand position. As an option, surgical correction of limb deformities can be performed	The minimum frequency of exercises for stretching and increasing range of motion is 3–5 times a week. The minimum frequency of effective orthosis use is 5 times a week
	Muscle weakness (movement against gravity); functional scales (CHOPINTEND); motor development (HINE)	Improving functions and mobility. Use of devices for sitting and mobility. Mobile shoulder supports are used to support the upper limb function	Toys with switches, light rattles, bath equipment, adapted beds, auxiliary devices for the upper limbs and lifts (elevators), environmental monitoring and eye movement recording devices for computers and communications, strollers with a hinged roof and power chairs, flat-lying/inclined adapted seats
Patients verticalized in a sitting position (sitting): early non-ambulatory stage	Postural control; foot and chest deformities; scoliosis and pelvic obliquity; hip lability	Positioning and orthosis. Chest fixation is recommended to maintain posture and improve functions. Neck fixation is often used to support the head for safety and transportation. As an option, surgical correction of spinal deformity can be performed	Orthoses should be removed 60 min before bedtime. The minimum frequency of orthosis use is 5 times a week
	Contractures (ROM, goniometry)	Stretching. Orthoses are used for the upper and lower limbs to improve function and ROM. Regular exercises to preserve the range of motion of the joints that are most at risk of contractures: hip, knee, ankle, elbow, and carpal joints. To maintain posture and ability to stand, it is recommended to use KAFO and AFO orthoses. RGO and KAFO can be used in patients capable of walking. TLSO and arm splints are used to maintain posture. As an option, surgical correction of limb deformities can be performed	The minimum frequency of stretching and ROM exercises is 5–7 times a week. During stretching or joint exercises, joint alignment should be ensured. Supported standing position should last up to 60 min; the minimum frequency is 3–5 times a week; the optimal frequency is 5–7 times a week

Table 3 (the beginning is on page 83)

Patients verticalized in a sitting position (sitting): early non-ambulatory stage	Functional scales (HFMSE, RULM, MFM); muscle weakness (strength tests)	Improving function and mobility. Use of devices for sitting and mobility. Use of devices for movement learning and mobility in patients capable of independent walking. Shoulders devices to support upper limb function	Exercises can effectively affect function, strength, ROM, endurance, ADL, participation, and balance
Patients capable of independent walking: ambulatory stage	Mobility; time tests; endurance assessment (6MWT); falls; functional scales (HFMSE, RULM); muscle weakness (strength tests)	Improving functional activity and mobility	Swimming, hippotherapy, and sports in wheelchairs. Sitting patients should be provided with power wheelchairs, individual posture support devices, and seating devices. Verticalizers should be provided for patients with weak strength of the shoulder girdle muscles. Lightweight manual wheelchairs are ideal for stimulating independent movement of patients with satisfactory strength of the shoulder girdle muscles. Exercises for aerobics and general developmental exercises for walking SMA patients. Options include swimming, walking, cycling, yoga, hippotherapy, rowing, elliptical/cross trainers. The exercise program should be designed and supervised by a physical therapy specialist or occupational therapist familiar with SMA. The optimal duration of aerobic exercises is at least 30 min
	Contractures (ROM, goniometry)	Stretching	The minimum frequency is 2–3 times a week; the optimal frequency is 3–5 times a week. Flexibility is maintained through active stretching and the use of orthoses in accordance with the specific needs
	Postural control; scoliosis; hip joint lability	Positioning and orthosis	Balance exercises. TSLO orthoses are used to improve posture in a sitting position. KAFO orthoses are used to maintain the function and range of motion in the lower limb joints

ROM – range of motion; CHOPINTEND – Children Hospital of Philadelphia Infant Test of Neuromuscular Disorders; HINE – Hammersmith Infant Neurological Examination; AFO – ankle foot orthosis; KAFO – knee ankle foot orthosis; HKAFO – hip knee ankle foot orthosis; TLSO – thoracolumbosacral orthosis; WHFO – wrist hand finger orthosis; HFMSE – Hammersmith Function Motor Scale Expanded; RULM – Revised Upper Limb Module; MFM – motor function measure; 6MWT – 6 minute walk test; ADL – activities of daily living.

Rehabilitation of non-verticalized patients (late non-ambulatory stage)

Primary goals of rehabilitation of non-verticalized patients (late non-ambulatory stage) include maintaining the possibility of verticalization with control of the body position and control of orthopedic disorders and prevention of their progression.

Prevention of contractures and control of range of motion/orthosis. Retraction of the muscles of the axial skeleton and limbs includes the use of orthoses, splints, active or passive positioning in verticalizers and in the supine position, and, less often, staged gypsum casting techniques. Upon verticalization, the use of TLSO corsets, KAFO splints for the lower limbs, or individual HKAFO systems for verticalization is recommended. Fixation of the neck with a Shants collar is often used in an upright position to minimize the risk of asphyxia and to control the head. Orthoses for the upper (WHFO) and lower (KAFO, HKAFO) limbs are used to stimulate functional activity, control, and/or expand the range of motion. For the effective use of

orthoses, they should be removed 60 min before bedtime.

Positioning. Systems for seating and orthostatic support should include all aspects of maintaining a normal sitting position: rollers, molded pillows, and supports. Individual and molded wheelchairs as well as individual sleeping systems are recommended. For mobility and transportation, wheelchairs and hardware wheelchairs with a falling/tilting back, adapted seats, and neck braces to support the head may be daily used. TLSO corsets with an abdominal window are recommended for respiratory support.

Physical exercises. To improve various functions of the patient, the use of auxiliary and adaptive equipment is recommended. To improve communication, devices monitoring eye movements may be used. Some bed patients can safely take water procedures with appropriate support for the head and neck and constant monitoring.

Percussion massage of the chest, along with passive excursion by an Ambu bag, is an important part of treatment. This technique is especially important during respiratory infection and the perioperative period to prevent respiratory failure and improve lung ventilation. Manual methods include percussion, vibration, and repositioning, which promotes postural drainage.

Surgical Treatment

Until the last decade, due to a low survival rate of patients with late non-ambulatory SMA, surgical correction of spinal and limb deformities has rarely been discussed as a possible treatment option in these patients. Patients with stable performance status, primarily respiratory one, were rare exception [1, 16]. The only options to treat the deformity were individual rigid orthopedic corsets and orthoses to support the sitting position of the trunk, provided that they do not impair pulmonary function (Fig. 2) [3].

The starting point for observing the course of spinal deformity is the Cobb angle value in a sitting position [16]. If corsets are used, X-ray of the spine is performed in a direct projection while sitting in a corset and without a brace.

Orthosis is a palliative method; it does not stop progression of the spinal deformity [14, 19], does not facilitate the use of technical rehabilitation equipment, and does not improve the quality of life [19–24]. Evaluation of the results of surgical correction of spinal curvature in SMA patients led to changes in the rehabilitation protocol in this nosological group [17, 18].

Spinal and chest deformities. Kyphoscoliosis is the most typical variant of spinal deformity in SMA. The decision on spinal surgery is based on key indications for surgical correction: a Cobb angle in the frontal plane of ⩾50°; hyperkyphosis of ⩾50° or deviation of ⩾20° from the upper boundary of the age profile in the sagittal plane with deviation from PSVL; an annual progression rate of ⩾10°. Other factors should also be considered: impaired respiratory function, chest deformity, and a twisted pelvis with imbalance of the body. External respiration function should be evaluated in advance to determine surgical risk and postoperative respiratory support.

However, there are a number of limitations to surgical treatment: BMI <12, marked osteopenia (Z criterion <-3 SD), and decompensation of respiratory and cardiac functions. These factors require active treatment by dedicated specialists with condition monitoring. The main focus in these situations in rehabilitation should be on postural control (braces and personalized wheelchairs), pain control, and nutritional support.

There was intra-rater agreement, but with a low level of evidence, that surgical treatment of spinal deformity should be delayed until patients reach 4 years of age under orthosis support [3].

In the case of an immature skeleton (Risser I–III) in patients aged 8 to 10 years, preference is given to dynamic systems for correcting and controlling scoliosis during growth [1, 19, 21, 25–28]. Currently, magnetically-controlled growing rods are more actively used as an alternative to traditional extendable rods requiring staged surgical extensions. However, the rate of complications requiring revision interventions amounts to 45 % [29–32].

For children aged 8 to 12 years, there is no single inter-rater agreement on a tactical approach. Correction techniques (dynamic systems or multi-support fixation) depended on the deformity value, growth rate of the spine and progression, maturity of the axial skeleton, and clinical data, in particular in terms of BMI, respiratory status, and pain [1, 19, 21, 25–32] (Fig. 2).

Deformity correction using polysegmental devices is indicated for patients older than 12 years of age and/or with Risser >IV. Pelvic fixation is recommended for patients with a non-ambulatory stage (SMA type 1, 2) or a twisted pelvis. This option is debatable in patients with an ambulatory stage (SMA type 3); the decision is based on the presence of a twisted pelvis relative to the cranial end plate of L5 [33].

Commentary. In cases of severe spinal deformities in patients with low BMI and hypotrophy, monolateral dual rod fixation through a minimally invasive approach may be considered as an alternative technique (prognostically for vital indications).

Intraoperative neuromonitoring (IONM) of the conducting structures of the spinal cord is recommended in patients with neuromuscular diseases regardless of the motor status to reduce the risk of traction radiculopathy and sensorimotor disorders [34–36].

Commentary. In the absence of published studies on the use of an intrathecal approach for injection of recently approved drugs (nusinersen) in SMA patients after spinal surgery, the decision on administration is made individually by a multidisciplinary team involving a spinal surgeon. The issue of choosing a dynamic system and the age of final instrumentation and fusion is also decided individually.

Chest deformity, thoracic insufficiency syndrome. Chest muscle hypotrophy in children with SMA leads to the formation of bell-shaped chest hypotrophy with thoracic insufficiency syndrome [19, 22, 23, 37–39]. Often this syndrome exacerbates spinal deformity and the rate of

its progression [19]. The most effective method for controlling chest hypotrophy is gymnastics with an Ambu bag [3, 4]. A retrospective study of children with thoracic insufficiency syndrome associated with scoliosis who were treated by dynamic systems (growing rods, VEPTR) showed a low efficiency in the treatment of rib deformities and enlargement of the chest volume; therefore, these methods are not recommended in this syndrome [37].

Hip joint instability. This syndrome is common in SMA patients [1, 19, 24, 40]. Earlier studies (2003–2012) did not recommend surgery. These studies demonstrated that recurrent subluxations or dislocations occurred during postoperative treatment despite the fact that this pathology was initially rarely accompanied by pain [1, 19, 24, 40]. In later studies, surgical treatment for unilateral and bilateral instability of the hip joint is recommended only in patients with severe pain [3].

Commentary. An indication for surgical correction of unilateral and bilateral instability of the hip joint in SMA patients is severe pain and, more rarely, the presence of functionally unfavorable contractures that prevent verticalization of the patient in a sitting position.

Limb joint contractures. Most reports describing the surgical approach to treatment of patients with pathology of the limbs in SMA are based on the personal clinical experience of specialists. In SMA patients, joint contractures develop due to imbalance of antagonist–agonist muscles, prolonged static position, and reduced range of motion [19, 41, 42]. Functionally and symptomatically, contractures can lead to pain and limited verticalization in patients with neuromuscular diseases in general [42–46] and SMA in particular [1, 15, 41, 47–53]. The need for surgical treatment of limb contractures should be considered if they facilitate the development of pain and a decrease in the patient's functional capabilities.

Surgical correction is aimed at eliminating contractures of the joints (hip, knee, ankle), improving the patient's positioning, and preserving the possibility of patient's verticalization and mobility, which is an important component of social rehabilitation. Also, it should be remembered that orthopedic correction of lower limb pathology in a complex treatment and rehabilitation process may reduce the risk of spinal deformities or reduce a progression rate of the existing deformity. If the foot is deformed, a separate relative indication is the inability to use standard shoes [3, 15].

Commentary. An important aspect is the understanding of the purpose of surgery, its outcome, and its risks by patients and their official representatives. Methods of surgical correction of orthopedic pathology, which are used in the treatment of patients with neuromuscular diseases, are as follows:

– various types of osteosynthesis for single-stage intraoperative correction; the use of internal (plates with angular stability, threaded wires, screws) osteosynthesis is recommended, which ensures primary stability of bone fragments and early axial load and reduces the duration of additional external immobilization;

– corrective surgery on limb bones (detorsion-varus osteotomy of the proximal femur, acetabuloplasty to eliminate hip dislocation, corrective foot osteotomy, femoral supracondylar extension osteotomy/resection for simultaneous extension of the leg, etc.);

– tendon-muscle repair/releases (lengthening of the knee flexor tendons and fascia lata, tenomyotomy of the hip joint flexors, achillotomy, transplantation of the tibial muscles, etc.).

In the postoperative period, comprehensive rehabilitation is required, including conservative treatment and orthosis of the limbs [48, 53].

Anesthesiological Risks

Management of SMA patients by an intensivist. Assessment of syndromic status and perioperative risks. Various surgical interventions (surgical treatment of limb joint contractures, spinal surgery, acute surgical pathology and trauma, muscle tissue biopsy, gastrostomy, etc.) should be performed after detailed planning and analysis of the risks of possible complications.

During and after general anesthesia, SMA patients are at increased risk of muscle weakness, local and generalized muscle spasms, rhabdomyolysis, cardiovascular and respiratory disorders, malignant hyperthermia and hypothermia, and hyperkalemia. Also, these patients are at increased risk of early and long-term postoperative respiratory complications: acute respiratory failure, nosocomial and healthcare-associated infections, obstruction of the upper respiratory tract, difficult evacuation of airway secretions, hypoventilation, and lung atelectasis in the setting of a baseline high risk of prolonged forced respiratory care, late tracheal extubation, or even tracheotomy [54–57].

Physical examination methods. Apart from monitoring of the described conditions, control of changes in the neurological status of SMA patients is also important in the perioperative period.

Commentary. Identification of the SMA type is important for assessing and predicting anesthetic and surgical risks; therefore, preoperative analysis of the patient's condition should include assessment of the neurological status and disease progression degree [57].

Instrumental diagnostics. Preoperative assessment of the respiratory function is recommended with assessment of the risk of respiratory complications (evacuation of airway secretions, hypoventilation, aspiration, obstructive and central apnea). Underestimation of these conditions may lead to severe complications (infections, prolonged mechanical ventilation, tracheotomy) and even death.

Commentary. Assessment of the respiratory function should include taking of a detailed history and physical examination, assessment of the respiratory function and effectiveness of cough, sleep-associated respiratory disorders, and chest X-ray [57, 58].

Assessment of the respiratory function includes measurement of the vital capacity and daily pulse oximetry (SpO_2). SpO_2 of less than 95 % in atmospheric air is defined as a clini-

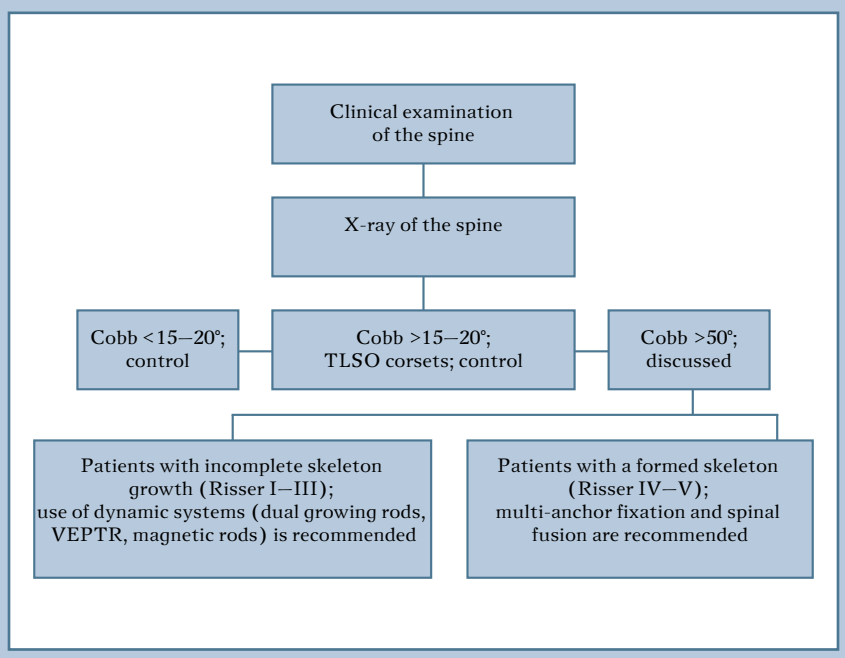

Fig. 2
Tactical diagram of treatment options in spinal deformity in patients with spinal muscular atrophy [3]

cally significant pathological value that requires additional assessment of the carbon dioxide partial stress. Respiratory failure is the most common cause of death [61].

Instrumental diagnostics in SMA patients should include:

– ECG monitoring, echocardiography; MRI and Holter monitoring of ECG and blood pressure (BP) before anesthesia or sedation are considered as additional options for analysis of cardiac dysfunction (various conduction blockades, myocardial hypertrophy, arrhythmias) for implementation of cardiotropic therapy [57, 58];

– daily monitoring of respiratory function and saturation [59];

– supervising cardiologist possessing necessary competencies in cardiac aspects of neuromuscular diseases [57, 58];

– intraoperative monitoring of motor evoked potentials, consistent with the Harvard standards, for control of neuromuscular conduction [60].

Preoperative Management

All patients admitted for surgical treatment should undergo preoperative preparation.

First of all, assessment of the expiratory reserve volume is necessary. This indicator is analyzed using X-ray or CT of the chest organs, spirometry, assessment of the peak cough rate, nocturnal pulse oximetry, and cardiorespiratory monitoring. Also, consultation of a pulmonologist trained in respiratory support is needed. If the expiratory reserve volume is limited, perioperative non-invasive ventilation of the lungs and manual and instrumental methods of stimulating and facilitating coughing (Ambu bag, cough assist machine) are necessary. This is more important for preparation of patients of the late non-ambulatory stage [57, 62, 63]. Assessment of the patient's cardiac activity should be performed at least 2 months before surgical treatment. In patients with impaired cardiac function, prescription and efficacy control of cardiotropic therapy are necessary before surgery. Administration of cardioprotective drugs can be started since the age of 10 years. Consultation with a nutritionist and optimization of the nutritional status are necessary [63]. Assessment of the bone mineral density is also needed. If osteopenia is verified, X-ray absorption densitometry (dual-energy X-ray absorptiometry (DEXA)) and consultation with an endocrinologist or osteologist are required. If osteopenia is detected, correction is necessary.

Premedication should exclude the use of drugs that can cause respiratory depression and hypoventilation of the lungs [57]; tracheal intubation should be performed according to the protocol for difficult airway management [57]. In cases of prolonged steroid therapy, the introduction of a stress dose of steroids should be considered during surgery [64, 65]. The central venous approach should be performed under ultrasound monitoring [66, 67]. Body temperature should be maintained at a normal level [68].

General Principles of Intraoperative Management of Patients with Neuromuscular Diseases

The management protocol is described in the Federal Clinical Guidelines "Perioperative management of patients with neuromuscular diseases" of the All-Russian Public Organization "Federation of Anesthesiologists and Resuscitators" [55].

Commentary. Anticholinesterase drugs are not recommended because of an unpredictable response to neostigmine. If possible, inhalation anesthesia should be avoided. Premedication with drugs that depress the respiratory center (opioids, benzodiazepines) is not recommended. Minimization of blood loss and surgery duration plays an important role.

Postoperative Management of Patients with Neuromuscular Diseases

In the postoperative period, all patients, regardless of the surgery amount and duration of anesthesia, should be observed in a critical and intensive care department [63].

Commentary. The capabilities of a critical and intensive care department enable comprehensive cardiovascular and respiratory monitoring, multicomponent therapy including non-invasive ventilation, and the use of devices facilitating coughing or aspiration of airway secretions.

Adequate analgesia is an essential component of treatment in the postoperative period. Doses should be adjusted to minimize inhibition of the respiratory centers. Preference should be given to multimodal anesthesia methods, with epidural anesthesia being a basic component.

Commentary. Timely prevention and relief of pain prevents secondary hypoventilation of the lungs due to muscle stiffness after thoracic surgery and surgical interventions on the upper abdominal cavity or spine. The dose of administered opioids should provide adequate analgesia, but not inhibit the cough reflex and spontaneous breathing.

In the postoperative period, disconnection from the respirator and tracheal extubation should be performed according to the protocols used in critical and intensive care departments for critical patients with mandatory CO_2 control. Also, there may be the need for prolonged mechanical ventilation after surgery and, in some cases, the likelihood of tracheostomy. Extubation and switch to non-invasive ventilation of the lungs (NIVL) should always be considered as an intermediate period before returning to the preoperative complex of respiratory support.

Stagnation of bronchopulmonary secretions, hypoventilation, and atelectasis can cause hypoxemia after surgery, so additional oxygenation should be performed carefully. First of all, it is necessary to exclude its typical causes; if they are present, initiate directed therapy. The use of oxygen is recommended only under NIVL and control of the acid-base state [1, 57, 69–72].

Prokinetics and gastric decompression through a thin nasogastric tube should be used to prevent intestinal motility disorders in the postoperative period. Nutritional support of the patient should also be considered.

Timely prescription of H2 blockers is required to prevent regurgitation and aspiration.

Commentary. Tracheal extubation should be performed under full control over bronchial secretion and upon achieving normal or borderline SpO_2 values in atmospheric air or in the presence of NIVL. At high risk of respiratory complications, combination of non-invasive pulmonary ventilation and cough stimulation is recommended.

Conclusion

In SMA patients, spinal and lower limb deformities are a frequent manifestation of the natural course of the underlying disease. Autochthonous progressive scoliosis is often combined with chest and limb deformities, contractures, and joint dislocations. Developing secondary orthopedic pathology causes not only gross dysfunctions of the musculoskeletal system (support, movement, and verticalization) but also pathological changes in the vital functions of internal organs and systems (respiratory, digestive, cardiovascular). For this reason, thorough analysis of the patient's condition is necessary (assessment of somatic, neurological, and orthopedic status). This is achieved via detailed preoperative multidisciplinary examination to asses the risks of complications and develop the treatment protocol and recommendations individually for each patient.

However, serious complications in SMA patients often develop in the peri- and postoperative periods; therefore, for prevention and treatment of dysfunctions in these patients, a system based on cardiorespiratory support techniques is required, recommended protocols of anesthesia and intensive care should be used, and adequate postoperative management and condition control should be performed.

Surgical correction of orthopedic pathology in SMA improves the performance status of the patient, improves the quality of life and self-care level, and optimizes the external respiration function.

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