

## THE IMPORTANCE OF POSTURING AND PHYSICAL THERAPY IN SPINAL MUSCULAR ATROPHY

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### Abstract

**Introduction:** Spinal amyotrophy is part of a group of autosomal recessive degenerative diseases that affect the motor neurons in the spinal cord, often also those in the brain stem, resulting in their death, followed by loss of muscle mass and muscle strength. The motor deficit does not symmetrically affect the left and right hemispheres. This problem determines in time changes in the thoracic cage, associated with respiratory deficiency, disorders of vertebral statics, muscle retractions in the upper and lower limbs, but also in the neck muscles. The clinical picture can be aggravated by the appearance of the redors that affect the large joints, but also the small ones and especially the temporomandibular joint that slows the child's feeding.

To prevent the appearance of these alterations, careful supervision of these patients is required and also to establish a physical therapy program, led and monitored by a multidisciplinary team.

**Materials and methods:** For the prevention or recovery of deformations, have been used several medical devices, such as pillows, sandbags, corsets, orthoses and other devices.

The physical therapy program includes manual orthopedic therapy, hydrotherapy, ergotherapy and robotic therapy.

**Result:** Early application of treatment maintains the joints functional, the muscles maintain their characteristics, breathing does not become a risk factor, all of which allow the establishment of medicinal treatment with Spinraza.

**Discussions and conclusions:** Implementing a recovery program in specialised centers, but also its continuation at home, may prevent or sometimes only delay the appearance of changes in the locomotor apparatus.

**Key words:** *Spinal amyotrophy, posturing, physiotherapy*

**Introduction:** Spinal muscular atrophy (SMA) is part of a group of autosomal recessive degenerative diseases that affect the motor alpha neurons in the spinal cord, the consequences being progressive muscular atrophy, weakness and paralysis [1].

Sometimes the neurons in the brain stem are affected, resulting in their death, followed by loss of muscle mass and muscle strength.

The onset age of this disorder determines various degrees of severity, early onset of the disease makes the clinical picture more severe. This shows muscular hypotonia, symmetrical and progressive motor deficiency at the left and right hemisphere level [2]. We have encountered cases of children with SMA type 1 to which the motor deficit does not manifest symmetrically.

Muscle weakness is proximal, affecting the muscles of the pelvic belt and the lower limbs more than the muscles of scapular belt and the upper limbs [3].

Children with spinal muscular atrophy face difficulties lifting the head off the supporting surface or maintaining the head upright when lifted into seated position, deficit that progresses and spreads to the distal extremity of the limbs.

At the thoracic level, it can be observed a weakness of the intercostal muscles and breathing adopting a paradoxical pattern.

The diaphragm is affected to a lesser extent, but the modifications mentioned determine the appearance of a specific form of the thoracic cage, namely "bell-shaped Thorax" [4].

Muscle weakness and the positions in which children with SMA are held, determines in time disorders of vertebral statics, deformation of the thoracic cage, associated with respiratory deficiency, muscle retractions in the upper and lower limbs, but also at the neck muscles level.

The clinical picture can be aggravated by the appearance of the stiffness, that affect the large joints, but also the small ones and especially the temporomandibular joint. Decreasing the amplitude of opening the mouth, generates problems related to the phonation and especially the patient's feeding.

To prevent the appearance of these alterations, careful supervision of these patients is required, as well to establish a treatment program, led and monitored by a multidisciplinary team [1], [4].

### Materials and methods

Spinal muscular atrophy (SMA) represents a condition that has generated interest and study programs at international level. Thus in 2004 an international conference led to the establishment of a committee of experts in spinal muscular atrophy to create a consensus statement on care standards [3].

In 2007 a report of the consensus declaration makes it possible for the guidelines that underpin the care guides and protocols to be adopted at the international level [2].

For the prevention or rehabilitation of deformations, have been used several medical devices, such as pillows, sandbags, corsets, orthoses and other devices. The physical therapy program includes manual orthopedic therapy, hydrotherapy, ergotherapy and robotic therapy.

### **Posturing**

Posturing represents one of the most important anakinetic techniques we use in the child's physical therapy program with spinal muscular atrophy.

„Posturing implies an attitude imposed on the whole body or at least on parts of it, for therapeutic or preventive purposes, to correct or avoid the installation of static deviations and vicious positions or to facilitate a physiological process” [5].

The duration of the posturing is variable, depending on the specific characteristics of each patient and the purpose it is made for. Patients with spinal muscular atrophy with a higher motor deficit, remain in certain positions for a longer period of time. These positions are not always the most accurate. The fact that they are placed in different positions on the bed for rest, in carts during the day, or they are held in an uncontrolled manner, makes these positions become true posturings.

Corrective postures are most commonly used in the physical therapy of SMA patients.

Knowing the direction in which the spine tends to deform and how the musculoarticular tissue is affected, allows the establishment of a prevention program. It may include corrective and hypercorrective posturing, which is designed to preserve the normal amplitude and elasticity of the joints and soft tissues involved.

Hypercorrective posturing (applied preventively) must be made alternately and at equal amplitudes, and the time the posturing is maintained should be the same for both ways.

In the case of patients who collaborate, corrective positions can be:

- freely chosen, they could adopt and maintain them voluntarily;
- Free-aided postures, using sand bags, rolls, bands or other devices;
- Fixed, which are externally corrective postures, using devices or robots and have the role of stretching soft tissues and restoring the amplitude of movement.

1 Facilitation postures used for SMA patients have the role to facilitate a physiological process, disrupted by the disease, out of which we mention here:

- Bronchial drainage postures, which help relieve respiratory pathways from secretions;
- Slanted forward or upward postures to promote or prevent return circulation;
- Respiratory or cardiac facilitating postures [6].

### **Posturing for breathing assistance**

Breathing is the vital function that suffers due to the muscle strength loss, but also due to the deformation of the thoracic cage. These disorders cause the increase in the amount of secretions that stagnates in the lungs and the bronchial tree. In these conditions the breathing becomes superficial, and the gas exchanges at the alveoli level are inefficient.

In order to reduce these shortcomings, there are recommended slanted backward or horizontal postures, on lateral or ventral decubitus. These postures are used to perform assisted breathing with various devices or the AMBU balloon.

### **Posturing for thoracic cage assistance**

Shallow breathing, but also the positions in which these children are held, determines the deformation of the thoracic cage. Posturing is recommended to emulate normal development of the thorax. The ventral decubitus position is indicated in this situation.

### **Posturing for kyphoscoliosis assistance**

Kyphoscoliosis is a deformity that occurs consistently in SMA. Free posturing or using devices from physical rehabilitation rooms are used to prevent or delay the worsening of these deviations (photo 10, 11, 12). It generally uses all the decubitus positions, as well as others that favor the elongation of the spine.

### **Posturing for scapular belt and lower limbs assistance**

The scapular belt is affected by the muscle weakness and by the positions in which the patients are held. The following positions are specific to this condition: the internal rotation of the upper limb, elbow flexion, ulnar inclination and pronation of the forearm. Recommended for posturing are the positions opposing those in which these segments are found. The positions for the spine assistance are also used, as they strengthen the anterior torso muscles and determine the external rotation of the upper limb.

### **Posturing for hip extension assistance**

Prolonged maintenance of the hip flexion determines the shortening of the iliopsoas muscle and the reduction of the extension. The dorsal decubitus posture at the edge of the table with a bent limb and the other one hanging, represents a position that does not involve effort to maintain it.

#### **Posturing for knee extension assistance**

The flexion of the hip attracts knee flexion. In these conditions the hamstring muscles are shortened together with soft tissues that form the posterior part of the knee joint. For correction, different positions are used, free or assisted by devices or therapist, where pressure is made on the knee joint.

#### **Posturing for calf muscle and ankle assistance**

The use of orthoses is a solution to prevent the shortening of the triceps surae muscle. From the seated position, exerting pressure on the knee with the thigh at 90 °, it can elongate the soleus muscle.

#### **Posturing for adduction assistance**

The shortening of the tensor muscle of fasciae latae as a result of flexing and external rotation of the thigh, is prevented by the application of elastic bands, sandbags, orthoses, which maintain the knees close together.

#### **Posturing for seated position assistance with or without devices**

The seated position is recommended to patients when using corsets and other devices that favor an elongation of the spine. It is advisable to have head support for those who cannot maintain it voluntarily, the forearms to be properly supported, the angle at the knee and ankle level to be 90 °.

#### **Posturing for upright position assistance**

Orthostatism is a position where the SMA child can be postured when using orthosis for the pelvic girdle and lower limbs, torso orthosis and head support. This position can be maintained with verticalizers after the body has been fastened with straps so that the pressure exerted in the coxofemoral joint is reduced.

#### **Posturing during transportation/carrying the patients**

The child with SMA needs special attention. The child must be carried with the spine in an aligned position, having the body weight transferred to the shoulder of the carrier. Another option is carrying the child facing down. If the child has a corset, he/she can be held with the face or the back towards the carrier.

#### **Physical therapy**

In spinal muscular atrophy the application of physical therapy programs involves the prior fulfillment of a thorough respiratory,

neuromuscular and musculoskeletal assessment. This will be adapted to each type of spinal muscular atrophy: Type 1 (non-sitters), type 2 (Sitters), type 3 (patients with mobilization capabilities) [7].

The evaluation will include somatometric measurements, but also evaluation using recommended scales in SMA.

The physical therapy program goals are:

- Preservation of vital functions: respiratory and cardiac functions;
- Maintaining the muscles characteristics;
- Maintaining joint amplitude;
- Avoid the deformations of the spine and thoracic cage;
- Preserving autonomy.

The Type 1 spinal muscular atrophy begins in the first trimester of life, which means these children need support from the first moves they make. Type 3 may have the age of onset at 18 months to 3years [7]. This means that the physical therapy program is addressed to patients who are small, sensitive and not always very collaborative.

#### **Respiratory therapy**

The respiratory therapy comprises techniques and methods that have as a result the assistance of the thoracic cage, the disobstruction of the respiratory paths and the realization of gas exchanges at the alveolar level.

The thoracic cage is regarded as a whole consisting of an osteo-articular structure, which under the action of the musculature mobilizes the soft tissues it contains. Breathing can be influenced by direct action from the outside to the inside or from the inside out.

From the outside mobilizations of the costovertebral, costosternal and intervertebral joints are made in order to maintain or increase the amplitude of their movement.

The mechanically assisted therapy it is done with action from the inside out, using breathing in-breathing out devices like Cough Assist, BIPAP or Vital Cough.

A technique of breathing therapy is the AMBU therapy, that can be applied separately or combined with the exhale assistance performed by the therapist or by the patient's relatives. To achieve this technique the patient can be positioned on the dorsal, lateral or ventral decubitus [8].

The increase of the diameter of the thoracic cage can be made by blocking the thoracic expansion one way and allowing the movement to the other ways. This is obtained during the voluntary

controlled breathing, through playing or using the AMBU device.

#### **Treatment of joint stiffness**

In order to prevent or delay the joint stiffness, it is necessary that the positions in which the SMA patient is maintained to be changed often and the joints to be mobilized on the whole amplitude of movement.

When joint limitations have occurred, treatment involves reduced amplitude mobilization and light pressures at the end of the movement. Posturing can be assisted by various devices such as rolls, sandbags, elastic straps or belts, as well as customized orthoses that create tension on periarticular and muscular tissues. Beforehand thermotherapy can be applied, which causes a local vasodilatation and stimulates the stretching of shortened tissues. Electrotherapy procedures could be used on older patients.

#### **Treatment of muscles retractions**

Treatment of muscle retractions is performed by wearing day and night orthoses, passive and active stretching on affected muscles and different physical exercises to stimulate voluntary contraction. For elongation of the shortened muscles there are recommended the serial positions, free or fastened in orthoses, simple or mobile. It is advisable to stimulate the antagonistic musculature to reduce the imbalance between agonists and antagonists.

#### **Stimulation of muscles contractions**

In the case of SMA the striated musculature impairment is subordinated to the neurological problem. This means that the muscle fiber is healthy and it requires the persistent stimulation of the isometric and isotonic contraction. No gravity exercises are recommended for Force 2 musculature, with the therapist exerting resistance or using toys approved by the child, at different times of movement. For the muscles that manage to mobilize different segments against gravity, this capacity should be speculated and the patients should be brought as often as possible in the situation to detach from the supporting surface of those specific segments.

#### **Treatment of vertebral deviations**

Treatment of vertebral deviations requires wearing of corsets or other devices that support the spine in a correct position. Preventively, asymmetrical positions that generates structural deformations of the bone system and shortening of soft tissues, must be avoided. Maximum amplitude mobilization in all motion plans and also pullups that stretch both joints and muscles must be performed. Posturing on various devices

help maintain the spine mobility within physiological limits. In the case of deviations that exceeded an angle of 60-70 °, surgical correction it is recommended, to avoid shortening of soft tissues from concavity and damaging of the internal organs.

#### **Parental counseling**

Parents of children with spinal muscular atrophy need to know how this disease evolves. They must participate in the rehabilitation program to acquire the knowledge and abilities of a therapist. They must ensure the continuity of the physical treatment at home. They must be able to intervene when a bronchic drainage is required, a discharge of secretions or when using noninvasive ventilation devices.

**Result:** Early implementation of posturing and physical therapy help maintaining the joints functional, muscles preserve their characteristics, breathing does not become a risk factor. If the alterations mentioned have already occurred, the correct application of the posturing and physical therapy program, in a consistent and progressive manner, allows for the rehabilitation to a variable extent of the motion amplitude and muscle elasticity. In the case of patients who have acquired gait, physical therapy helps them preserve their independence.

#### **Discussions and conclusions**

Implementing a rehabilitation program in specialised centers, but also its continuation at home, may prevent or sometimes only delay the appearance of changes in the locomotor system. The introduction of medicinal treatment programs brings a new hope for life for patients with spinal muscular atrophy. These two forms of treatment should complement each other to ensure the continuation with the injection program.

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