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Severe scoliosis in neurodevelopmental disabilities: clinical signs and therapeutic proposals

A. FERRARI [1, 2], C. FERRARA [2], M. BALUGANI [2], S. SASSI [2]

[1]*Department of Neuroscience, University of Modena and Reggio Emilia, Modena, Italy*
[2]*Children Rehabilitation Unit, S. Maria Nuova Hospital Reggio Emilia, Italy*

Scoliosis is an important cause of disability in childhood, due to its incidence and limitation on ability. In neurodevelopmental disabilities, scoliosis worsens the already limited functional capacities of the child and can thwart abilities partially recovered through rehabilitation. In cerebral palsied children (CP), scoliosis is considered a consequence of damage to the central nervous system or a complication of a peripheral impairment, in particular, through obliquity of pelvis, of the unilateral dislocation of hip. In order to explain the scoliosis of central origin, primitive and pathological reflexes, reactions or automatisms must be taken into account, especially the asymmetrical ones. This contradicts the absence of scoliosis in hemiplegia. On the contrary, symmetrical patterns should act as protective factors. However, the greater incidence of scoliosis in tetraplegia than in dipegia does not confirm this idea. Scoliosis is secondary when it is linked to an unilateral hip dislocation for side of convexity, proximity, measure and timing sequence. In childhood neuromuscular diseases (NMD), scoliosis is the unavoidable consequence of muscle weakness. The only protective factor may be muscle stiffness in case of fibrotic degeneration. The main curve is reducible for longer periods, while the less reducible secondary one at cervical level can limit the correction possibilities, due to the need to keep the head aliened. In spina bifida (SB) children, lesional and supralesional scoliosis can be present. In the former, the malformative ones directly derive from the vertebrae involved in the myelomeningocele and the secondary ones develop from unilateral hip dislocation similar to CP, or from muscular imbalance (a typical feature of SB, especially for lower lumbar levels) or from primitive skeletal malformations of pelvic girdle or lower limbs. The acquired ones are caused by muscle weakness as in NMD. In the supralesional scoliosis, the curve is the consequence of an impairment in neurological structures especially of ponto-cerebellar carrefour or of an acquired tethered cord and may affect any segment of spinal column, also above the primitive lesion level. The scoliosis in neurodevelopmental disabilities can be treated conservatively with corsets and postural systems, with limited results, or through a surgical approach, often made difficult due to the multiple impairments present in the same patient.

KEY WORDS: Scoliosis - Cerebral palsy - Neuromuscular diseases - Spina bifida occulta.

Corresponding author: A. Ferrari, Children Rehabilitation Unit, S. Maria Nuova Hospital, Reggio Emilia, via Risorgimento 80, 42100 Reggio Emilia, Italy. E-mail: ferrari.adriano@asmn.re.it

Among the major causes of childhood disability, scoliosis, given its incidence and severity, is one of the most critical. Its natural history produces a great negative impact on the course of adult life. Scoliosis can occur in every child, at any age, but more often in those children affected by a motor pathology such as cerebral palsy (CP) (see Appendix), neuromuscular diseases (NMD) or spina bifida (SB). Scoliosis, by impairing postural control, severely compromises the organizing and interfacing abilities with the surrounding world, deprives social relations and reduces the quality of life.

Scoliosis in cerebral palsy

In the different clinical forms of CP,[1] scoliosis is a recurrent and severe complication that causes negative effects on thoracic structures and on the development of adaptive functions (see Appendix).[1] Scoliosis affects motor activities recovered by the child through rehabilitation and limits participation in social life.[2]

Epidemiology and etiopathogenesis

The onset of scoliosis is somehow related to the nature of the primary defect and to the natural history of CP, since its occurrence is significantly higher compared to reference populations (from 15% to 80% according to Koop).[3] It arises even before the tenth year of age and advances also during adulthood, becoming potentially surgical in one out of seven cases.[4] The incidence of scoliosis is directly correlated to the level of motor and cognitive impairment,[5] and therefore to the degree of the central lesion. Its occurrence increases proportionally to the topographic distribution of the palsy, affecting 30% of tetraplegic, 10% of diplegic and an insignificant percentage of hemiplegic forms.[3] This latter data, though apparently logic, introduces one of the contradictions of scoliosis of central origin, as originally coined by Stagnara.[6] In fact, if we consider asymmetry between right and left side in its possible expression, such as:

— difference in muscle tone and postural attitude (sustainment);

— difference in muscular excitability, force and endurance;

— difference in muscular length and passive stiffness;

— difference in length and trophism of the homologous limbs (heterometry);

— different joint range of motion;

— occlusive and chewing defects from alteration of temporo-mandibular joints;

— different repertory of motor patterns, combinations and sequences;

— different automatic reactions between the two hemisides (straightening);

— different balancing ability between the two hemisides (equilibrium);

— different ability in motor control on the two hemisides;

— different use of the homologous limbs (motor initiative);

— disturbs of lateralization (eye, hand, foot);

— attention defects (neglect);

— sensorial and sensitive deficits (visual, proprioceptive, etc.);

— perceptive and gnosic impairments.

As a factor predisposing scoliosis, we should see a greater incidence of scoliosis in the unilateral forms with respect to the bilateral ones. If, on the contrary, we consider symmetry inducing primitive/pathological patterns (see Appendix) as protective factors in keeping trunk *en bloc* (*i.e.*, propulsive reaction (see Appendix) or startle reaction (see Appendix), or the prevalence of straightening pattern (see Appendix) over rotational-derotational one, which is typical of "spastic" forms) [7] scoliosis should occur less often in tetraplegic than in diplegic forms. Nowadays, it is well known that beside central neurological factors (postural organization, antigravity competence (see Appendix), straightening ability, fixation (see Appendix) and space alignment (see Appendix), balancing (see Appendix), motor coordination, muscle and postural tone (see Appendix), etc.) and peripheral biomechanical factors (muscle strength, stiffness (see Appendix), bone geometry), there are several other factors triggering scoliosis, among which: genetically predisposing, metabolic (the structure of collagen) and hormonal factors (somatic growth). As already stated by Marbini *et al.*, many of these elements are also directly involved in the genesis of CP motor defects.[8]

In the "spastic" forms of CP, scolioses usually have an Italic "S" shape, but "C" or "double S" shaped deformities are also possible. In these forms, the most affected tract is the dorso-lumbar one.[4] In "dyskinetic" forms, scoliosis appears in athetosis and in dystonia, but only in tetraplegic, diplegic (reversed) and double hemiplegic forms. Scolioses are unusual in chorea and in ballism and almost completely absent in hemidystonia, despite the intensity of twisting spasms and the extreme asymmetry of the pattern they trigger.

Characteristic clinical aspects

Of great relevance is the distinction between primitive and secondary scoliosis (Table I). The former are not worsened by hip luxation; lumbar spine is, in fact, rectilinear and the kinematic pattern of lower

TABLE I.—*Clinical aspects of the main forms of scoliosis in CP.*

	Primitive scoliosis	Secondary scoliosis
Example	Scoliosis starting from head Mid-thoracic scoliosis	Compensatory scoliosis due to a pelvic obliquity
Common site	Cervico-thoracic	Lumbo-sacral
Causes	Multifactorial causes, in particular primitive and pathological reflexes, reactions or automatisms	Unilateral hip luxation provoked both by a valgus and anteverted femoral neck and by a dominant pattern in flexion-adduction at the lower limbs
Diagnostic criteria	S Italic, C, double S o bizzarre shape (rectilinear lumbar spine, hips in situ and pelvis aligned) Head, eyes, and mouth can be diverted in the same direction of the curvature Can occur plagiocephaly	Curve is concordant, coherent, consecutive and consequent to pelvis obliquity. Can occur windswept hips deformity with the same direction of convexity
Compensatory curves	Usually absent. Curves are often uncompensated. The sitting posture is unstable	Usually present compensatory cervical curves that easy a stable sitting posture
Complications	Early respiratory, postural, visual and relational complications	Late respiratory complications

limbs does not justify the onset of scoliosis, as in the case of medio-thoracic scolioses and in the ones starting from head. Secondary scolioses, on the contrary, are those actually determined by an unilateral hip luxation.

Primitive scoliosis

Among the patterns which are able to affect axial motricity (see Appendix) and, therefore, suspected to be a major cause in the onset of scoliosis (at least in primitive forms) there are asymmetrizing motor reflexes, reactions and automatisms. The most known is the asymmetric tonic neck reflex (swordsman position) (see Appendix), but also the Galant reflex (see Appendix), the Branco Lefevre (see Appendix) and the tonic neck reflex starting from head inclination (see Appendix) are also worth mentioning. Among symmetrizing patterns, therefore with a protective feature, the most relevant are the Juanico-Perez reflex (see Appendix), the labyrinthic tonic reflex (see Appendix), the symmetric tonic neck reflex (see Appendix) and the tonic lumbar reflex (see Appendix). A particular role is played by the spasms in extension-torsion (see Appendix), considered to be directly responsible for the onset of the so called "scoliosis starting from head". A synthetic description of these reflexes and the definition of some technical terms used in this paper are reported in appendix. From the short description of the reflexes that have effects on the spine, emerges that some are responsible for trunk inclination movements on the frontal plane, and/or flexor-extension on the sagittal plane, and/or rotation on the horizontal plane, but also that every reflex acting in a certain direction can be ideally nullified by contrasting one able to produce an opposite movement or postural attitude. Therefore complex movements in three planes, typical of central origin scolioses, are not "simply" referable to the dominance of this or that reflex. In substance there are many suspicions, but no definitive verdict. It should also be emphasized that, especially in dyskinetic forms, pathological reflexes are activated by emotional stimuli or voluntary movements, thus becoming a personalized way to interact with the body and with the physical and social context. Therefore, scoliosis becomes not an absolute defect, but the result of a primitive and sometimes patiently constructed strategy.

Among scolioses with central origin, scoliosis starting from head deserves a separate discussion. This term indicates the postural-motor pattern of CP children (generally spastic tetraplegic with dyskinetic components, mental retardation and epilepsy), who, usually in sitting position, but sometimes also in prone or supine ones, "obstinately" assume a scoliotic attitude, in some cases associated with complete trunk flexion and in others with its extension, with persistent deviation of the head towards one side. The direction of head inclination/rotation is always the same. Eyes look continuously to the same side and there

is often nystagmus. Given the continuous production of "grimaces", even the mouth can be diverted in the same direction as eyes and head. Gradually the skull becomes deformed by acting as a "first vertebra" (plagiocephaly). Scoliosis may reach any part of the spine, with "bizarre" deformities which are difficult to contain, especially in cervical and dorsal segments. Curves are often "uncompensated" and frequently destabilize postural balance, as they cause the trunk barycentre to be projected outside the support base. When we try to bring patient affected by scoliosis starting from the head back to a more correct posture (for example using postural aids or orthoses), he skillfully manages to get around the introduced restrictive device. If he cannot succeed, he struggles against it thus increasing muscular effort in order to regain the previous pathological posture. This scoliosis is clearly a "neurological" alteration (tonic neck reflexes?, labyrinthic reflexes?, lumbar reflex?, oculogyral reactions?) which over time can be overlapped by an "orthopedic" secondary deformity (kyphoscoliosis extended to the head often associated to a windswept deformity of the inferior limbs).

Secondary scoliosis

By secondary scoliosis, or compensatory scoliosis, we mean a scoliosis that follows an alteration of pelvis alignment on the frontal plan, most frequently a pelvic obliquity (lift) produced by unilateral hip luxation, a frequent event in CP, with or without pelvic twisting as an effect of windswept lower limbs.

Other deformities have the same nature of passive scoliosis, *i.e.*, the deformities produced on sagittal plane by pelvis rotation on its transverse axis as a result of functional compromise among the dominant flexion pattern of the upper body and the extension one which prevails in pelvis and in lower limbs, with consequent muscular contractures.[9] In case of pelvis anteversion due to prevalence of hip flexors, there will be lumbar hyperlordosis, whereas in the case of pelvis retroversion due to prevalence of hamstring shift, a large dorsal kyphosis will extend downward, thus transferring base from pelvic bone to sacrum.

Pathogenetic mechanisms of secondary hip luxation in CP and its possible influence on spine structure still represent an uncertain field in the clinic. It is generally accepted that deformity progression respects same stages in the following order: valgism (increase of inclination angle) and anteversion (increase of declination angle) of femoral neck, femoral head lateralization with progressive deformation to Basque beret, suffering of acetabular margin and consequent accentuation of the roof inclination and finally the gradual ascent of femoral head towards a posterolateral direction, up to subluxation and complete dislocation.[10] The prevailing flexion-adduction of the thigh (adductors, ileopsoas, medial hamstrings), universally indicated as the pathogenetic factor of the secondary hip dislocation, can extend its influence on pelvis, which goes up ipsilaterally to the compromised hip, and on spine, which consequently develops a lumbar-sacral scoliosis with opposite convexity. Hip luxation is opposite to scoliosis direction, whereas the direction of windswept hip deformity has the same direction of scoliosis convexity,[11] so that the femur of the luxated hip goes in the same direction of the curve concavity.[12] It follows that, if we do not take into consideration the problem of pain, bilateral luxation in CP is less severe than the unilateral one due to the absence of scoliosis like a secondary complication.

The recent literature shows that, without making a distinction between primary and secondary scoliosis, the relationship between scoliosis and hip dislocation may be more ambiguous. Associations between the direction of scoliosis and direction of pelvic obliquity, and between the direction of windswept hip deformity and the side of hip subluxation/dislocation are confirmed. A significant association is also seen between the direction of scoliosis and the direction of the windsweeping such that the convexity of the lateral spinal curve is more likely to be opposite to the direction of windsweeping.[12] In accordance with Loeters *et al.*,[13] on the contrary, evidence is weak for an association between the severity of CP, hip dislocation and pelvic obliquity and scoliosis. Also Senaran *et al.* [14] assert that "Unilateral hip dislocation causes a significant increase of pelvic obliquity but does not affect the rate of scoliosis curve progression". While Hodgkinson *et al.*[15] assert that "We were unable to find any relationship between the side of the pelvic obliquity and the side of the scoliosis convexity, the side of the hip excentration, or the deviation attitude, but the deviation attitude appeared to be a risk factor for pelvic obliquity, which itself was a risk factor for excentration, which was a risk factor for scoliosis". According to Koop [3] there is a clear relationship between windsweeping and convexity of scoliosis, which have opposite di-

rections, but there is no strong correlation between unilateral luxation and scoliosis. Lattre [5] and Lonstein [16] argue that there is a correlation between scoliosis and pelvic obliquity, which have opposite directions, but there is no correlation between scoliosis and windsweeping. Letts [11] claims that there is a correlation between windsweeping and ipsilateral scoliosis. Prevailing unilateral left luxation justifies a higher incidence of scoliosis convex to the right. Significantly more windswept deformities on the right, hips subluxed on the left and lateral lumbar/lower thoracic spinal curves convex to the left were observed.[12]

It seems likely that cases of secondary scolioses actually consequent to a single lateral hip dislocation and cases of concomitant scolioses are mixed differently in the samples studied by the authors. In order to distinguish them, the following criteria may be useful.

Scoliosis should be:

— concordant: in the opposite direction from the dislocated hip;

— coherent: proportionate to the pelvic obliquity degree;

— consecutive: without interposition of "neutral" vertebrae;

— consequent: following over time the pelvic obliquity appearance.

A lumbo-sacral scoliosis is surely a complication of hip dislocation, but we should remember that in CP the scoliosis which do not accompany dislocation are generally more severe, as if the hip resistance leads greater fragility to the spine.

Elements of process complexity

Other factors affecting the whole process should be considered: first of all, the kind of contractile activity. On one side, it seems logical to confer the main pathogenetic responsibility of dislocation to spasticity; nevertheless, it is difficult to explain why similar deformities can also appear in situations of complete muscle weakness, as for example in cases of spinal muscular atrophy (SMA) types I and II and of congenital myopathy. SB, on the other hand, shows unmistakably the pathogenetic importance of muscular imbalance, especially for levels L5 and L4, when preserved flexor and adductor muscles cannot be counterbalanced by paralyzed extensor and abductor ones.

However, the deforming force could also not be of a purely contractile nature, as progressive muscular dystrophies clearly evidence. Indeed, these are illnesses where muscular impairment is always considerably symmetrical, but the loss of force is often not symmetrical and muscular retractions are never symmetrical (as a rule).

Anyway, spasticity itself, or better spasms, could still be a cause of anterior dislocations of femoral head, instead of posterolateral one, through an excessive retroversion of femoral necks (batrachian posture, torsion dystonia, etc.) though not associated with scoliosis, even when unilateral.

Regardless of inherent strength, joints always come out the worse in the conflict muscle-bone-joint. We only need to consider the extreme rarity of muscular tears in all childhood neurodevelopmental disabilities compared to the great frequency of joint dislocations and torsion deformities of long bones.

Beside coxofemoral joint, also other joints may show important subversions: for example the sacroiliacs, especially in windswept deformities, or the sacral complex also involved in pelvic anteversion and retroversion. The responsibility of biarticular muscles in deformity onset cannot be clarified without considering the complex role of chain movements, otherwise there would be an aggravation of the defect to be corrected. Hamstrings cannot be blamed for dislocation if, for example, pelvic is strongly anteverted; on the contrary, we can not talk about responsibility of ileopsoas muscles if pelvic is retroverted. Furthermore, the adductors are not responsible if the pelvis is lifted on the opposite side, whereas spinofemoral muscles cannot support thigh abduction if femur is simultaneously extrarotated, and so on. The retraction of medium gluteus anterior fibers may accentuate windsweeping as well as the adductors on the opposite side, which win over joint strength and then significantly reduce their activity by effect of insertion approachment, produced by the rising of femoral head (false innocence). In the presence of windswept deformity the torsional components of scoliosis are intensified by knees flexion, supported in turn by hamstring contracture and sometimes accompanied by the failure of the patellar tendon (patellar lift off), more than inclination components which are induced by pelvic twisting.

Scoliosis in neuromuscular diseases

Scoliosis is frequently observed in many neuromuscular diseases, particularly those whose onset occurs during childhood, because the skeletal apparatus is still growing and therefore much more susceptible to deforming forces. For this reason, the illness is particularly significant for frequency, severity and rapidity of progression in Duchenne muscular dystrophy (DMD) and in type II spinal muscular atrophy (SMA).[17, 18]

Duchenne muscular dystrophy

In DMD, a scoliosis of progressive nature (dystrophic scoliosis) debuts, according to most authors,[19] one to three years after the natural loss of walking ability. The incidence varies significantly, from 48% according to Robin,[20] to 64% Dubowitz,[21] to 90% Siegel,[22] to 93% in a subsequent study by Robin.[23] These differences are due to discrepancies in enrolled populations, not especially homogeneous in terms of age.

In DMD, a progressive scoliosis, with the characteristics of the idiopathic form, can sometimes appear even before the loss of walking. This is defined as scoliosis in dystrophic child, rather than dystrophic scoliosis. This condition should be considered separately, both for the characteristics of the curve and for the therapeutic measures to be adopted, considering the negative repercussions that both scoliosis and corset can have on patient residual walking ability.

The typical curve has a wide C-shape. It affects the dorso-lumbar segment and is accompanied by a cervical compensation curve in order to maintain the head balanced and aligned. The lateral tilt movement of vertebral bodies is associated with a rotation on convex side, maximum in the lumbar region, for ligaments and anulus fibrosus tensioning.[24] The main curve produces spinal muscles retraction on the concave side, while on the convex one, superior trapezius and levator scapulae retraction may appear, as a result of their contraction in order to maintain the head vertical. The neutral top vertebra lies between T4 and T8. The neutral bottom vertebra is among the last lumbar ones and an extension of the curve down to the sacrum is also possible, with the appearance or accentuation of pelvic tilt. The apical vertebra affects the final two dorsal or the first two lumbar vertebrae. On the convex side, the intercostal space increases, accentuating the so-called posterior gibbus, while on the concave side, that space decreases, deforming the lateral part of chest as a wedge. On the convex side rib-vertebral corners are reduced because of soft tissue stretching, and increased on the concave side, caused by rib horizontalization. In the worst cases, in sitting position, the scapula can lean on the posterior iliac crest. On the concave side, the costal arch soon enters into conflict with the pelvis and ends up sinking into it, undermining the main brace support point. Curve reducibility is greater at dorso-lumbar level (main curve) than at cervical one (secondary curve). This should be taken into account when constructing the brace, because the correction degree depends on the reducibility of the secondary curve (cervical) and not of the main one (dorso-lumbar). The inclination on the sagittal plane (kyphosis, lordosis) can justify the rapidity of curve worsening, which is directly related to pelvic retroversion that the patient maintains in sitting position.[19] In fact, in NMD the presence of pelvic anteversion with accentuation of lumbar lordosis serves as a protective factor towards the development of scoliosis [25] and respiratory failure.

The deformity can rapidly develop, increasing by 2.1 degrees per month.[26] The deterioration continues, although more slowly, even after somatic growth has stopped. Therefore many authors consider it necessary to refer the patient to radiological anteroposterior rachis control, executed in sitting position, every six months,[19] although X-rays of the patient lying on his back, with half-bent knees supported by a pillow in order to reduce lumbar lordosis, with parallel femurs and patellae at the zenith, are more objective in investigating the real measure of skeletal deformities and not those produced by the postural attitude of the patient, still partially reducible by head traction.

It is important to remember that some subjects with a confirmed DMD diagnosis do not develop scoliosis for a long time even after losing walking ability,[27] or develop a slow progressing scoliosis.[28] To justify this situation, the relationship that exists between muscle strength loss and development of muscular retractions, has to be considered in each patient. In most cases, loss of force precedes retraction development, but there are subjects who develop early retraction, causing premature loss of function, despite having a discreet residual muscular strength. Muscular retractions provide the rachis a

kind of passive protection against scoliosis, which is able to contain and compensate the destabilizing effects of muscle weakness. The inclusion in one or another form is easily assignable through clinical examination, comparing vertical performances with horizontal ones. Vertical performances (standing and walking) exceed horizontal abilities (sitting up, crawling, creeping, rolling) in the hyposthenic form because these require greater muscular strength. In the form with prevailing retractions, on the contrary, the horizontal performances exceed the vertical ones because they are less influenced by existing joints deformities (for example, flexor retractions of hip and knee and equinus feet are indifferent for crawling). Furthermore, inclusion in one or another form may be diagnosed by horizontal tomographic trunk scans to determine paravertebral muscle trophism, which is relatively preserved in the form with prevailing retractions. The natural history of disease confirms that hyposthenic patients can walk relatively longer, but show early scoliosis after loss of walking ability. Patients with prevailing retractions generally quickly lose walking ability, but they can also not develop scoliosis.

Applying head traction to a sitting patient, allows us to diagnose the following different clinical forms:

a) weakness without retractions: for a certain period an alignment of the spinal column is still possible because the "muscular" scoliotic attitude requires time to become a "structural" scoliotic deformity. During this transitional period, the use of brace or surgical stabilization of the spine is indicated;

b) weakness and retractions: despite limited head control, trunk balance, respiratory problems, etc., when head traction is applied, there is no trunk elongation, as if it were one rigid structure. The presence of severe limitations of elbow, knee and hip joint range of motion confirm the significant role of the retractile components. This second group of subjects can longer go without braces and surgical stabilization would be inappropriate.

The root cause of scoliosis in DMD patients is the loss of antigravitary trunk muscle strength, resulting in the progressive inability to resist gravitational stress. Additional factors will contribute to producing a drop to one side or another: firstly, pelvic asymmetry (generally an inclination with rotation), which can be caused by difference in lower limb length (*i.e.*, a structural equinus dominant on one side), or can be produced by an asymmetry of hip muscles retractions (mainly of superficial ones), or supported by the appearance of windswept in the lower limbs (retraction of tensor fasciae latae mainly on one side). Secondly, among the additional factors, there is sitting posture asymmetry. In order to respond to the dominant upper limb, the patient usually leans to the opposite side, which would explain the prevailing right convexity orientation of scoliosis. Finally the asymmetry in clinostatism produced by loss of ability to turn over in the bed should be considered. The skeletal age at the time of scoliosis onset is decisive in all types of scoliotic deformities, the earlier the onset relative to somatic growth, the worse the prognosis.

Spinal muscular atrophy

The frequency of scoliosis in SMA varies in literature, because of lack of sample homogeneity and different classification criteria: from 58% according to Hensinger [29] and 70% to Schwentker [30] typically reported in SMA II, to 95% according to Granata.[31] It is important to notice that the SMA I does not reach sitting position, while the SMA III exceeds adolescence before the hyposthenia becomes severe. Vertebral deviation in SMA II, that has an earlier onset than in DMD, involves a progressive functional damage represented by pelvic imbalance, inability to maintain a correct sitting position, reduction of the already precarious autonomy of upper limb, pain and bedsores. SMA II scoliosis has the same root causes as dystrophic scoliosis, but due to greater uniformity in hyposthenia distribution and low disease aggression, given equal degree of hyposthenia patients are able to stand and walk longer. In addition, SMA II C-shaped scoliosis has the same frequency of double curve scoliosis. It is interesting to note that while in DMD the head tends to stay straight even in extreme scoliosis, in SMA it tends to lean towards a shoulder due to cervical vertebrae deformity, that counts the skull like a last vertebra. It is not known why the head frequently shifts on the vertical of left shoulder.

Scoliosis in children with spina bifida

Lesional scoliosis (primitive, secondary, acquired)

In children affected by SB (myelomeningocele), the onset of a scoliosis directly related to the lesion can be attributed to different causes resulting from

primary malformations and variously combined among them:[32]

— vertebral malformations as hemivertebre (sometimes multiples), fusions, butterfly vertebrae or wedge vertebrae (triangular), accompanied or not by bifurcation, duplication, fusion or absence of ribs (primitive lesional scoliosis). Scoliosis may be congenital or occur after birth, progressing in relation to height growth. In the upper part of the spine, the presence of one or more hemivertebre, situated on the manifest Myelomeningocele cephalic side, generally produces a dorsal eccentric kyphoscoliosis with raised shoulder. In thoracic region, hemivertebrae may cause a short-range kyphoscoliosis, while at lumbar level more often there is an association between lordoscoliosis and pelvic tilt. Scoliosis may be accompanied by localized or generalized congenital kyphosis, mostly in the lumbar region (kyphoscoliosis). An isolated lordosis more or less rigid, instead, is due to pelvis anteversion caused by irreducible hip flexion, or developed to compensate a short-range congenital kyphosis. If there is a serious spinal malformation producing kyphosis, erector spine muscles along the sides of spinal column can no longer act as extensors but become abductors or even flexors.[33] The trunk inevitably "collapses" and the thorax sinks into the pelvis, accompanied by abdominal viscera compression and reduced respiratory capacity.[34] The skin above the kyphosis is particularly exposed to danger of decubitus ulcers owing to the absence of the subcutaneous tissue, poor vascularisation and general lack of sensibility;

— pelvic asymmetry caused by muscle-tendon retractions, from primitive skeletal malformations of pelvic girdle [sacral or coccyx agenesis, sacral dish asymmetry, caudal regression (see Appendix)] or of lower limbs, or by unilateral hip dislocation (secondary lesional scoliosis). The most frequent form is thoracolumbar lordoscoliosis, due to pelvic obliquity;

— insufficient support reaction (see Appendix) caused by primitive muscular weakness (acquired lesional scoliosis). It appears only in relation to higher lesional levels (above D12-L1) and follows the dystrophic scoliosis rules.

Primitive lesional scoliosis

In children with SB, congenital localized scoliosis and kyphosis are a relatively common deformity. They generally affect the thoracolumbar column region and cause orthopedic problems both at birth and at later ages. The vertebral bodies are abnormally wide and protrude at an acute angle.[35] This makes neurosurgical repair of myelomeningocele malformation extremely difficult and, as a consequence, increases the probability of bedsores. The congenital scoliosis and kyphosis show an acute curvature, whose fulcrum lies at the level of the malformed vertebra, usually hypoplastic. Typically the rachis recovers alignment above and below the main curve. In general the evolution of these deformities remains slight, but makes it difficult to adopt walking orthoses (pendulum balancing) and also penalizes sitting.[36]

In describing and studying the congenital lumbar kyphosis, which is commonly associated with thoracolumbar or high lumbar SB, Barson [37] related the fixed position of neural plate with development alterations of the underlying mesoderm somites, which give rise to each vertebrae. So, at a primitive lesion level, the vertebral ventral parts do not grow, while the dorsal ones do, therefore giving rise to a complex consisting of fused everted peduncles, creating the kyphosis.

Similarly, vertebrae underneath an asymmetric hemimyeloschisis (hemimyelocele) delay growth in one half of their structure and this part becomes the concavity of a scoliosis or a fixed primary kyphoscoliosis. The wedge apex is oriented towards the posterior longitudinal median line or sometimes towards the opposite side of the spinal column. Occasionally instead there can be a supernumerary vertebral body developed only in one half (accessorial hemivertebra).

Secondary lesional scoliosis

In secondary lesional scoliosis due to pelvic tilting, the curvature ray is less acute and the deformity is localized at the lumbosacral passage (transition between lumbar and sacral spine). In order to be defined secondary and compensatory, scoliosis must comply to the conditions set out above, in relation to CP, *i.e.*, be concordant, coherent, consecutive, simultaneous or subsequent.

Acquired lesional scoliosis

In acquired lesional scoliosis due to inadequate trunk support reaction, the curve takes on the paralytic

or neuromuscular scoliosis characteristics (C or Italic S shape). These are different from idiopathic scoliosis, because they appear with the same frequency in males and females, can be both right and left convex, and have maximum evolution in preadolescence.

Generally, in primitive lesional scoliosis (congenital, secondary and acquired), therapeutic measures (orthopedic corsets and functional surgery to rebalance the pelvis) are sufficient to slow the pejorative evolution and enable the more severe cases to reach the minimum surgery age with the spine still in good condition.

Supralesional scoliosis

The supralesional scoliosis pathogenesis is attributable to mechanical and/or neuropathic factors. Mechanical factors are due to a spinal cord stretch. The spinal cord is anchored at the level of the primitive surgical repair scar (tethered cord symptomatic).[38] As a result of the existing disproportion between spine growth and spinal cord stretching, the spinal cord first runs along the shortest course inside the vertebral canal, thus "cutting curves", then it progressively stretches and finally leads to an evolutionary spine deformation (scoliosis, hyperkyphosis, hyperlordosis). Neuropathic factors should not be attributed to a nerve root stretch on one side, therefore to the muscular excitability asymmetry, but rather to damage to spinal cord and to posture calibration structures of pontocerebellar "carrefour". In fact, as result of spinal cord stretching, the function of these structures, that organize egocentric and geocentric (see Appendix) body axis (see Appendix) alignment [cephalocaudal straightening reaction (see Appendix)], static and dynamic balance control,[10] collecting and processing visual labyrinthic and proprioceptive information, is altered. The lack of spatial coordinates for axial straightening exposes SB child to gravitational stress and other injuries, related to prevailing postures and to compensating mechanisms adopted during motor activities. Also perceptual disorders (posture change intolerance, fear reactions to emptiness and depth of surrounding space, etc.) can be considered as a result of body space alignment mechanism alteration. The pontocerebellar carrefour impairment may be linked to the upward extension of spinal cord tension caused by tethering, favored by cerebral malformations that are often congenitally associated with SB, such as Arnold Chiari malformation II, and by the action carried out from above by the hydrocephalus.[38]

Since spinal cord stretching is a phenomenon linked to length growth of the spine, supra lesional scoliosis onset is relatively late and coincides, more often, with spurts in growth rate.

The suspicion of a component that "feeds" neurogenic scoliosis arises when you have one or more of the following elements:

— occurrence in a previous normal innervation area;

— rapid pejorative evolution of scoliosis, independent of the skeletal age;

— lack of concordance with pelvic tilt or lower limb deformities, such as a right hip dislocation and a right convex scoliosis;

— non-balanced scoliosis;

— lack of consistency, *i.e.*, proportionality, between degree of scoliosis and degree of accompanying pelvic tilt;

— lack of consecutiveness between pelvic tilt and scoliosis, due to the presence of a number of neutral vertebrae, *i.e.*, of normal conformation, between one and another. This is the most important aspect to consider;

— "bizarre shape" scoliosis;

— lack of response to therapeutic measures (body brace, physiotherapy).

In these cases scoliosis is defined supralesional. In order to understand its nature, it is necessary to explore the presence of possible neurological complications, either isolated or variously associated with each other, such as:

— disturbances of long intersegmental spinal pathways caused by an ischemic vascular damage, primitive or secondary to myelomeningocele repair, which compromises postural tone and support reaction;

— spinal cord damage from symptomatic tethered cord (TC), with structural failure at less resistant areas (myelopathic scoliosis);

— spinal cord damage in vertebral canal caused by repeated micro-trauma due to eccentric spinal cord position, especially in thoracic segment (scoliotic myelopathy), with lesional level corresponding to the inside angle of greater scoliotic curvature. These micro-trauma could cause ischemia and myelomalacia;

— symptomatic hydrocephalus with worsening on Arnold Chiari II malformation;

— symptomatic Arnold-Chiari malformation (cerebellar wedging in large occipital foramen);
— symptomatic hydrosyringomyelia;
— diastematomyelia and symptomatic diplomyelia;
— symptomatic vertebral dislocation.

Since a supralesional scoliosis is present in more than half the patients with symptomatic TC, in case of anchored spinal cord stretching, the emergence of this developmental deformities can be considered an almost inevitable complication. To assess the importance of this sign, however, it is necessary to keep in mind that scoliosis in patients with SB can also be influenced by:

— discordant height growth (hypothalamus-pituitary axis disorders triggered by suprapituitary recess stimulation resulting from tri-or tetra-ventricular hydrocephalus);
— anomalous puberty development;
— bone metabolism disorders (nephropathy).

Supra lesional scoliosis evolves very quickly and is non-respondent to the usual physiotherapeutic, orthotic and conservative treatment.

It is therefore necessary to distinguish the so-called lesional scoliosis (primitive, secondary, acquired), which are generally not very evolutionary and have a compensatory arrangement above the primary deformation, from supralesional scoliosis due to spinal cord tethering or stretching or other neurological conditions. The latter generally worsens quickly and still constitutes a very complex problem. In fact, scoliosis is very serious from the start of its appearance and can become so severe as to lead to respiratory insufficiency, therefore requiring surgical correction. But an alignment of the scoliotic curve necessarily implies an additional stretching of the tethered spinal cord. All the traditional techniques for scoliosis correction, such as surgery for spinal stabilisation, remedial casts in preparation to the stabilisation, trans-skeletal traction (halo-traction) and body braces which make use of column lengthening, such as the dynamic self-correction, are very dangerous, as they may exasperate damage to spinal cord up to cerebellar tonsil wedge in foramen magnum. Correcting this scoliosis is anything but simple. Today, by early recognition of the presence of a tethered and stretched spinal cord, it is possible through a neurosurgical intervention of detethering, not only to avoid more serious or even fatal consequences, but also to block the direct causes of pejorative progression of scoliosis.[38]

Therapeutic proposals

The treatment aims are attenuation of scoliosis progression, pain reduction, maintaining stable sitting position [4] and head verticality, which are essential to facilitate vision, communication and upper limb functions.[39] By preserving postural control, feeding is favored, lung function is promoted, risk of gastroesophageal reflux and bronchial aspiration are minimized. These aspects are essential to increase social participation and quality of life.[40]

The physiatrist is responsible for differential diagnosis and prognosis. The therapeutic project and definition of treatment aims, shared with patient and his family, are based on these skills. Physiatrist should evaluate the possibilities offered by physiotherapy, protecting patients from useless and ineffective therapy (corrective gymnastics), designing orthoses and seating systems in terms of geometry as well as materials and constructive methods (positioning for plaster cast), detecting the need for surgery and estimating the amount of required correction (compensatory curve, pelvic conformation). In any case, the therapeutic project should consider collaboration among different professionals, patient's willingness and family consent, since an absolute solution to the problem is unattainable.

Physiotherapy

The validity of motor treatment, the so called therapeutic exercise, carried out also by non-professionals, continues to be widely accepted. However, no developmental scoliosis etiologic factors (altered axial motricity, reduced motor control, muscle weakness, vertebral malformations and secondary deformity) can be significantly improved through re-education.

Although physiotherapy does not act directly on the scoliosis, it serves however to counteract the negative effects produced by constant corset use, to limit the onset of muscle retractions and to counteract intervertebral tightening (especially in CP and NMD), to maintain chest mobility and ventilation capacity (in particular in DMD and SMA). In SB, head traction maneuvers, due to the dangers linked to tethered cord and to malformation of Arnold Chiari II, must obviously be avoided.

Orthosis

A corset is recommended in scoliosis with Cobb angles exceeding 20-25° and in patients with difficulties in maintaining the sitting position.

Static univalve thoraco-lumbar-sacral orthosis (TLSO) is the most used. It is made in polypropylene or polyethylene, using a plaster-of-Paris mould performed on supine position, and worn during the day. Brace fastening is placed at the rear and made of velcro straps. This model is especially used in CP and SB, not in NMD because it penalizes respiratory function. A study on the effect on breathing by postural rigid braces, conducted on children with SMA (limited to 8 patients), reports that tidal volume and respiratory system compliance are significantly lower when wearing orthoses rather than not.[41]

For these patients, some authors [42] suggest the Gauchois brace. It lacks a thoracic support but has preclavicular and prehumeral ones, which permit greater costal breathing than in other braces. Gauchois brace also has a head support, which penalizes head movements but favors greater column alignment. Instead, diaphragmatic breathing is limited because of the absence of an abdominal opening. Considering that in advanced NMD breathing depends on diaphragm activity, in Italy the static-balanced corset, wrapped on the chest and opened on the abdomen, is preferred.[43] The pelvic part can be ileum-sacral, sacro-ischiatic or ischio-femoral, depending on the projection of the barycentre of trunk and on the degree of pelvis anteversion. A thoracic, pectoral or sternal chest support, and at least one axillary arch, are always present. In order to confer tubular resistance to the corset structure, a rigid metal closure is placed on the front.

Instead in CP and in SB, it is possible to indicate semi-rigid corsets, like Soft Boston Orthosis**,** to get the same respiratory advantages, but at the expense of the correction effectiveness.[44]

Lumbar corsets, such as soft boston orthosis or CLB (Bolognese lumbar corset) can also be used in SB in case of lumbar scoliosis not accompanied by serious pelvic deformity.

Although the plaster cast method seems to be the most common technique of brace construction, today the latest standard is CAD/CAM technology which produces corsets on a positive model realized by measurements made by an optical system on patient and subsequently elaborated through appropriate software.[45]

The degree of correction made by corset depends on mutual scoliotic curves and head/pelvis balancing. Skin tolerance, orthoses comfort and breathing must always be taken into account.[46, 47]

The corset use, if well tolerated, should be continued as long as possible over time, especially in patients who are not candidates for surgery.[48] In fact, the primary purpose of the conservative treatment of both corsets and seating adaptations, such as an adjustable head support, offset lateral chest rests, shoulder harnesses and straps and waist strap which are all fitted on the wheelchair, is to regain sitting position.

Instead, it has been shown that the use of the corset does not arrest the progression of scoliosis. In NMD patients with braces, the curve progression varies from 1° to 8° per year. For Cambridge,[49] braces do not slow down scoliosis progression because this is a direct consequence of muscle weakness. Other authors suggest using them only when surgery is refused or there are unsuitable conditions that make surgical treatment unfeasible.[50, 51]

In DMD patients, the most common objections to corset use concern respiratory function and neuromotor development.

Although braces limit costal mobility and reduce lung volumes as reported by several authors (Tangsrud *et al.*,[41] Noble-Jamieson *et al.*,[52] Ishida *et al.*[53]), there are no studies comparing respiratory function of patients who used orthoses, compared to patients who did not. Hence, when considering also respiratory function, it is not clear if it is the better to delay scoliosis progression with brace (thus worsening pulmonary volumes) or to favor breath avoiding brace. In any case, without the use of braces, scoliosis progression itself will nevertheless lead to secondary respiratory deterioration in a short time. In fact, performance assessment with and without brace are executed at the same time, generally in the morning. It would be more correct to compare morning respiratory function with the evening one, after a whole day of not wearing the brace. For some patients, muscular fatigue in maintaining sitting position could penalize breathing more than the brace itself, making its use advisable.

Considering the neuromotor development, it is important to remember that a corset that safeguards every function does not exist and it is necessary to

define from the start which one to promote: the corset cast in standing position will facilitate walking, whereas the one cast in sitting position will favor manipulation, etc.

In these children, walking does not stop scoliosis, which can cause, when it worsens, the loss of locomotion. Hence a conservative treatment will no longer be helpful. So the choice to delay walking loss for a few months, limiting the possibility to act on scoliosis, entails penalizing other functions, such as postural control and manipulation.

When the patient clinical condition worsens, seating adaptations, as listed above, should be added to the corset. When the brace is no longer feasible and tolerable, the half-reclining position in special wheelchair, with anatomically adjustable back or with padded seating shells, remains the only possible solution. Seating systems work on patients who are not able to autonomously change position due to severe hyposthenia, as in NMD, flaccid CP and high spinal cord lesion, instead they do not work on patients with an ante pulsed trunk, caused by a hyperlordosis or as compensation for head erector muscles retraction.

Surgical treatment

According to McCarthy,[54] children with progressive curves from 40° up to 50° degrees and unresponsive to bracing, are candidates for spinal fusion. Usually, in order to avoid that the arthrodesis could interfere with the bone formation, surgery is carried out once the vertebral formation is completed, typically between 15 and 17 years of age, or earlier if there are therapeutic reasons. In NMD, vertebral stabilization should be performed before respiratory function reduction can make a prolonged anesthesia problematic, or if the scoliosis worsens by more than 1-2° per month. Every 10° of scoliosis correspond to a 4% loss in forced vital capacity (FVC).[55] In any case, breathing continues to worsen during adolescence and does not improve after vertebral stabilization.

Different surgical techniques have been developed for scoliosis treatment: Loque, Harrington, Isola techniques or segmental spinal fusion. They rely on metal instrumentation (screws, bolts, laminas), which is able to guarantee a solid fixation of curves. For this reason post-operative plaster, used in the past, is now outdated and the verticalization of patient is possible from the first day after surgery. Arthrodesis while introducing a constraint to the mobility of the fixed spine segment, does not restrict the respiratory function that instead is always penalized with the use of corsets.

Although posterior spinal fusion with multisegmental fixation is the most common technique, others, such as an anterior release and/or fusion or combined procedures, are also considered. The study of Auerbach *et al.*,[4] demonstrates that an all-posterior spinal fusion provides an excellent correction in case of smaller and more flexible curves associated with severe pelvic obliquity; whereas an anterior release with posterior spinal fusion should be preferred in larger and less flexible curves. For those patients with significant pelvic obliquity or at risk of developing pelvic obliquity, the procedure of instrumentation should be extended up to the pelvic level, especially for non-walking children. In these cases, the spine correction with Unit Rod should be preferred. This is a rigid spinal instrumentation with long iliac anchors able to provide superior control over pelvic obliquity compared to custom rods, but it results less effective in proximal fixation, with a tendency towards junctional kyphosis. Furthermore Unit Rod also prevents crankshaft phenomenon (see Appendix) and produces a well-balanced spine with relatively uniform coronal and sagittal profiles.[56]

Surgical access and instrumentation choice should not only consider curve length and degree, but also spinal stiffness, rachis structure and degree of correction.

Light instrumentation, such as Loque, is suggested for NMD and flaccid CP scoliosis, because in these cases the only contrasting force is represented by trunk weight. Instead, in spastic CP, frequent and prolonged spasms work against instrumentation, creating mechanical stress up to the breaking point. In these cases heavy instrumentation, such as Cotrel-Dubousset,[57] should be preferred.

In SB, given the anatomical malformations accompanying the posterior portion of the vertebrae involved in myelomeningocele, the skin anomalies and the high risk of infection, the anterior access (transpleural peridiaphragmatic retroperitoneal *via*) should be preferred, with ablation of several intervertebral disks in order to reduce the distance between head and pelvis and increase spine mobility. The anterior access can obtain a satisfactory vertebral fusion in lower thoracic-lumbar region.[58] If col-

umn stability is insufficient, after several weeks an instrumented arthrodesis with posterior access can be performed, although it is delicate and complex, in order to achieve pelvis-spine fixation.[59]

Another essential aspect to consider is the degree of correction, which is related to the defect nature. In spastic scoliosis, an excessive correction increases stress against instrumentations. In NMD scoliosis, an increase in correction degree could improve respiratory function, but on the other hand, if excessive, it risks compromising head control and, as a consequence, sitting position. In SB, surgery, also for minimum degree corrections, is not free from spinal cord stretching risk, that could result fatal.

Surgery results in short and long term are not always well defined.

In CP, with respect to surgical intervention, retrospective studies show that patients and parents are usually satisfied with long-term health benefits.[3] When dissatisfaction is reported, it usually refers to a deficient correction of the deformity, or to a recurrent deformity that needs further surgery, or to other major complications not solved.

In NMD, the effect of surgical correction on respiratory function is quite controversial: Kurz finds a slower decline of vital capacity [55] and another study [26] does not demonstrate any gain in respect to its natural deterioration. Instead Rideau and Galasko reported periods of 24 and 36 months respectively of VC stability after surgical treatment.[60, 61] A mortality reduction in DMD operated patients was also reported.[61] This result, however, has been contradicted by different authors who have not found a longer life expectancy or slowed reduction rate of respiratory function in DMD operated patients.[62-66] Due to the lack of randomized studies, it is difficult to determine if an increase in life expectancy is linked to the advent of non-invasive positive pressure ventilation, or to scoliosis surgical correction or to other types of treatment.[67]

Although the effect of surgical correction of scoliosis on respiratory function has not been fully defined yet, other advantages are listed in literature: greater physical comfort,[68] ability to more easily maintain sitting position,[49, 60, 62, 63] improved self image [68] and elimination of brace.[52]

The precarious respiratory condition of neuromuscular patients determines a high incidence (16%) of anesthetic and surgical complications,[62, 67] postoperative infections, bleeding, loss of gained correction, need for subsequent operations.[67] FVC of less than 35% is associated with a 50% complication risk.[62] Better prognosis are associated with VC values of more than 50% or with FVC values of more than 40%.[26, 68]

Up to now, no scientific evidence on the efficacy of surgical treatment has been produced.[67] The last Cochrane review concludes by reconfirming the following concept: "It is vital that the clinician assesses the patient from the cardio-respiratory point of view, to avoid surgical treatment in case of cardiomyopathy; it is also important that the patient and his family are informed about the potential complications and dubious efficacy of treatment".[69]

Because of the great risks, SB patients rarely undergo surgical correction, for example in cases of de-tethering procedure failure or of rapidly worsening scoliosis, despite the use of corset and seating systems.

Unfortunately, there is no evidence of potential benefit of surgery in comparing functional skills in patients with and without surgery. Despite the great number of studies which compare different surgical techniques, highlighting significant Cobb angle decreases, there is a lack of data concerning worsening or improving of functional abilities.[59] A case series by Schoenmakers [70] shows a postsurgical functional skill decline for at least six months, associated with a slight improvement after 18 months, concerning in particular self-care. However the walking abilities worsen in spite of early ambulation training and the surgical complication rate was 80%. These findings are essential in order to inform and prepare patients and parents for consequences of surgery.

Conclusions

Severe scoliosis in neurodevelopmental disabilities require great clinical and managerial skills from each team member: physiatrist, orthopedic surgeon, physiotherapist and orthotist.

The chances of limiting deformity and maintaining the quality of life of the patient and his family are related to the accuracy of periodic evaluations and to the professional efficiency of the team.

Mere physiotherapy is insufficient to stop vertebral deformity evolution; it should be combined with the use of orthosis and devices and with the correction of the associated conditions, above all hip dislocation.

Vertebral fixation is often the only possible solution: this awareness should be achieved as soon as possible, before respiratory insufficiency worsens the already critical clinical conditions of the patient.

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APPENDIX 1

(1) **Cerebral palsy** (CP): the term describes a group of disorders of the development of movement and posture, causing activity limitation, that are attributed to non progressive disturbances that occurred in the developing fetal or infant brain. Motor disorders of cerebral palsy are often accompanied by disturbances of sensation, cognition, communication, perception and/or behavior, and/or seizure disorder.[71]

(2) **Adaptive function**: function adapted to actor, suitable to context and appropriate to aim. The adaptive functions have cognitive value, that is they are targeted to specific aim or result. They acquire, for example, value of "praxia" in a Piaget sense, value of "gnosia" or percept that is having meaning (perceptive classification and semantic classification) values of "concepts", "abstractions" and "strategies" (of planning) in order "to resolve problems".[72]

(3) The term **primitive** indicates that the considered pattern belongs to normal repertory in a general sense, but is often observed outside the temporal range in which it should act. The term pathological indicates an observed pattern that does not belongs to a normal repertory. It is obvious that also primitive patterns that last indefinitely are expressions of a pathological organization of motor functions similar to those produced by CP or mental retardation.

(4) **Propulsive reaction**: this reaction belongs to a group also known as positive support reactions. A newborn baby is put in prone position on a crib with symmetrical lower limbs, abducted and extrarotated thighs and plantar surfaces held against examiner hands. The infant performs an active extension movement and moves the trunk forward (dragging reflex), while upper limbs on the side of trunk are moved forward and backward. The pattern is bilateral, diffused throughout all the body, symmetrical and synchronous. A healthy newborn baby already spontaneously performs these movements similar to "crawling-swimming", which would be reinforced by this maneuver.

The advancement can be with "abdomen on the ground", while four limbs, or only the upper ones "grip" the ground, or during every movement the abdomen is lifted off the ground. Some children, who demonstrate lower limb weakness or awkwardness, learn to drag trunk with upper limb force. The Milani propulsive reaction would be the expulsion mechanism during delivery: not only when the fetus pushes his feet against uterine wall in order to begin the expulsive contractions, but also to extend himself by assuming an aligned posture that more appropriately favors the passage through delivery canal.

(5) By the word **startle** we describe a reaction of alarm, surprise, fright, or panic that consists of a generalized jerky, sudden, involuntary, stereotypical motion produced by an unexpected stimulus (kinaesthesis, acoustic, visual, tactile, vestibular, thermal, aching). This reaction includes motor, autonomic, emotional elements. The motor expression, when complete, includes head extension, open mouth, anguished expression, upper limb abduction (arms opened widely) with contracted but slightly opened hands, trunk extension with chest in forced inspiration, abducted-extended thighs with semi-extended knees and varus supinated feet. The pathological startle reaction is an excessive or too frequent response, that is characterized by low-threshold and resistance to habituation. Startle reaction characterizes pathological conditions including CP. It can also be evoked by non-threatening, not sudden, weak, stimulus (ineffective stimulus under normal conditions) of various kinds (acoustic, tactile, visual, etc.). In its presence, anticipation capacity for self-induced stimuli is absent.

(6) **Straightening pattern/reaction** includes automatic movements that develop starting from the first year of life under the guide of vestibular, visual, tactile data. This reaction serves to maintain or to reestablish alignment of head, trunk and limbs in egocentric space. It is possible to distinguish an axial straightening and a rotational-derotational straightening. In spastic syndromes, axial straightening proceeds in cefalocaudal direction and precedes, in an evolutionary sense, the rotational-derotational straightening. This latter can be compromised, bestowing an *en bloc* character to trunk motility, due to the difficulty in turning either left or right from any starting position. In dyscinetich syndromes, rotational-derotational straightening prevails over the axial one that proceeds in caudo-cephalic direction rather than cefalo-caudal sense. It allows for the possibility to turn right and left, sometimes also excessively beyond normal, but the difficulty for these patients is still to completely extend trunk and to maintain a correctly aligned head. This ideally represents the last link of the chain, especially in sitting and standing positions.

(7) **Support reaction**: expresses the ability to overcome the effect of the force of gravity applied to body mass (weight).

(8) **Fixation**: indicates stability that exists between body axis and extremities: firm extremities and mobile body axis, distal or centripetal fixation (from limbs towards trunk), mobile extremities and firm body axis: proximal or centrifuge fixation (from trunk towards limbs). Whereas in spastic syndromes at the debut of standing with the use of support (parallel bars, walkers), a distal fixation is clearly recognizable, which progressively transforms to a proximal fixation in order to allow gait with mobile supports (walkers, canes, crutches, etc.), in dyskinetic syndromes fixation remains variable. It can be disto-proximal (patient embraces himself while the body axis continues to move unstably), then completely distal, then completely proximal, etc, with consequently serious postural instability.

(9) **Spatial reference systems**: egocentric system: has as a reference point the body, especially its longitudinal axis (ideotropic vector). Allocentric or esocentric system: has as reference point the external space around the subject. Geocentric system: has as a references the vertical line, that is the direction of the force of gravity, and the horizon line, that is tangent to the terrestrial plane.

(10) **Balance reaction**: postural reaction that tends to prevent the projection of the barycentre from falling outside the support base. This is carried out through movement of the body or a part of it in the same direction but opposite to the deforming force.

(11) **Muscle tone**: number of motor units active in a resting muscle, which is detectable through repeated passive asynchronous lengthening of the same muscle. Postural tone: isometric contraction of antigravitary muscles which are responsible for maintaining a specific posture.

(12) **Stiffness**: degree of resistance or muscular fibers tension as a result of lengthening. Elastic coefficient: the opposing resistance by a muscle to stretching (neural and non-neural components).

(13) **Axial motricity**: a set of single movements that can actively modify spine posture.

(14) In order to evoke an **asymmetric tonic neck reflex**, the child is placed in the supine position. By passively rotating the head, the limbs on the rotated side spontaneously extend and on the other side the limbs flex towards the nape. The trunk laterally tilts with concavity on the nape side (swordsman position). This reflex is especially active in dyskinetic syndromes, where it interferes with the acquisition of derotational straightening reactions.

(15) The **Galant reflex** or spine tonic reflex, or trunk inclination reflex: with the child in the prone position or held in ventral suspension, unilateral stimulation of paravertebral region, proceeding from shoulder blade angle to lumbosacral zone, determines trunk inclination on the stimulated side, with homologous concavity, and its extension with pelvic lift. Upper limb on the stimulated side is abducted and moves away from trunk, lower limb abducts and flexes.

(16) **Branco-Lefevre reflex**: also this is a trunk inclination

reflex with ipsilateral concavity, but it is evoked by a pressure stimulation applied between the iliac crest and the 12° rib. Contrary to the Galant reflex, trunk inclination is associated with an extension of the ipsilateral lower limb and a flexion of contralateral one.

(17) **Tonic reflex** starting from head inclination is evocable in the vertical posture by tilting the head towards one side. This movement provokes an increase in extension tone on the inclined side (as if this side had to support body weight), and a lessening on the opposite side. The limbs on the head tilted side extend, while the contralateral ones flex. The trunk is laterally tilted with a homologous concavity of the head. The adopted attitude is exactly the opposite of what we described in the asymmetric tonic neck reflex.

(18) **Juanico-Perez reflex**: the child is held in a prone ventral suspension. Simultaneously stimulating the paravertebral regions starting at the top and moving down we obtain a head extension, upper limb abduction, pelvic lift with accentuation of lumbar lordosis and lower limb flexion at knees.

(19) **Labyrinthic tonic reflex** can be either in extension or in flexion. In the extension type with a child in the supine position, passive head flexion provokes an active extension of head and both shoulders, contraction of the trunk and hip extensors and mouth opening. Lower limbs are extended at hip and knee. In the flexion type with a child in the prone position, passive extension of the head provokes a head flexion, with flexion and abduction of upper and lower limbs and flexion of trunk (total flexion pattern).

(20) According to the action of **symmetrical tonic neck reflex**, in the prone position the downward and forward flexion of the head provokes upper limb flexion and lower limb extension. In a quadrupedic position, extending the head upwards and backwards provokes lower limb flexion and upper limb extension. In the supine position, flexing the head provokes the upper limbs to flex and the lower limbs to extend at the hip level, whereas head extension induces the upper limbs to extend and the lower ones to flex .

(21) **Shimamoto and Nakajma lumbar tonic reflexes** are provoked by position changes of upper body in respect to the pelvis. As an example, trunk rotation towards the right facilitates upper limb flexion and the homolateral lower limb extension and upper limb extension and contralateral lower limb flexion. While tonic neck reflexes demonstrate a marked effect on upper limbs, tonic lumbar reflexes manifest its effects on the lower ones.

(22) Spasms in **extension-torsion**: are involuntary and unexpected movements that are triggered from endogenous or external stimuli, sometimes also very common everyday sounds (a cough, a telephone, a motorcycle or a dog barking). Spasms begin rapidly and have a short duration, but devastating effects on postural control, which is immediately compromised without any possibility of restoration. Spasms are accompanied by manifestations of anguish (wide open eyes, respiration arrest, grimaces, startle reactions, etc). The dominant movement is overall extension, nearly always accompanied by elements of lateral inclination, rotation and twisting along body axis. Spasms can proceed in various directions: they can progress from feet towards head or vice versa, go from nape towards lower extremities, from shoulders and hips towards hands and feet, from pelvis towards upper and lower extremities or vice versa. In any case hips are abruptly extended compromising posture. Any attempt to block the spasm at its start, only intensifies and diffuses it. The general rule lets the spasm express itself through an extension-torsion of trunk relative to thighs in order to favor a quick return to initial position through an accentuation of hips and knee flexion.

(23) **Crankshaft phenomenon**: may occur in very young patients who undergo a posterior only spinal fusion for scoliosis. Although the back of the spine is fused, the vertebral bodies continue to grow causing, as a consequence, the develope of a severe and rigid hyperlordosis.

(24) **Caudal regression syndrome**, also referred to as caudal dysplasia and sacral agenesis syndrome, is a rare congenital malformation characterized by varying degrees of developmental failure early in gestation. It refers to a heterogenous constellation of congenital caudal anomalies affecting the lumbar, sacral and coccygeal vertebrae, and corresponding segments of the spinal cord, the hindgut, the urogenital system and the lower limbs. Although skin abnormalities are absent, the spine and the pelvis are not joined, for the absence of sacrum.

(25) **Body axis**: term that indicates a real or virtual image of the support structure (spine).