SELECTED LECTURES

ON SPINE SURGERY

EARLY ONSET SCOLIOSIS: THE FRENCH EXPERIENCE

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The problem of early onset scoliosis (EOS) is one of the vital problems in the spine surgery. Common conventional method for treatment of this pathology does not exist, which makes thing worse. One of the leading experts in the world, Jean Dubousset not only possesses a wealth of clinical experience, but also puts forward new bright ideas promoting the advancement of spine medicine. Original approaches to strategy and tactics in the treatment of EOS patients are developed in many countries and regions of the world. The lecture of Jean Dubousset is dedicated to the French experience.
Key Words: early onset scoliosis, surgical treatment, conservative treatment.

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Classification of early onset scoliosis

The concept of early onset scoliosis (EOS), popularized by Dickson et al. [1], is a good one for two reasons. On the one hand, it is a paradigm of spinal deformity that develops at a young age and has not been correctly defined in a child with a big potency of growth. On the other hand, it is not sufficiently adequate because it uses a single term (attractive and well-known) for conditions with different etiologies, which have very little in common, except the age of patients. We cannot compare congenital scoliosis due to hemivertebrae involving 1–2 segments, paralytic scoliosis that is associated with spinal muscular atrophy and involves the entire spinal column, and infantile idiopathic scoliosis involving the entire thoracic spine. Therefore, to continue using the EOS concept, we need to introduce etiological subgroups where each condition is described separately. There are two large groups of patients that should not be confused. Unfortunately, it is obvious that when we listen to reports or read papers evaluating treatment options and their applications, we do not distinguish between the groups, which, in my view, is unacceptable.

It is wrong to rely on the omnitude of this understanding of EOS because many surgeons take new treatment options as a panacea and gradually lose their skills of conservative treatment using a corrective brace and serial casting (a method that takes a lot of time and is poorly paid, ignored, and marginal). They do not realize that, by giving preference to surgery, they "put their finger into a gear", which initiates a cycle of repeated surgeries with an exponentially increasing risk of infection and other complications. In my opinion (and this is typical of France), we should classify all EOS cases in accordance with the patient's age and etiology of deformity.

Patient age. According to my experience, real EOS patients are children who have been diagnosed with the pathology and undergone treatment in the period between birth and 6 years of age (before starting school). In addition, a prepubertal group (from 6 to 9 years), a puberty onset group, and a postpubertal group may be distinguished. This distinction is necessary because each group has its own therapeutic strategy.

Etiology. This feature enables assigning each patient to one of five groups: group 1 – idiopathic scoliosis, group 2 – paralytic and neuromuscular scoliosis, group 3 – congenital scoliosis, group 4 – dystrophic scoliosis, and group 5 – iatrogenic scoliosis. It is important to remember that each group comprises numerous subdivisions (postpoliomyelitis syndrome differs from spinal muscular atrophy and congenital muscular dystrophy; postlaminectomy deformity differs from thoracogenic deformity; etc.). Each patient should be treated as a unique case. In this lecture, we will focus on the first age group of patients with real EOS (developing between birth and 6 years of age). However, the surgeon's thought process should encompass all age and etiologic groups.

Complete clinical and radiologic examination is a mandatory first step

After we have taken the general information about a patient's family history, pregnancy, and birth and after the classical orthopedic examination and measurements, a very thorough neurological examination, including a study of the cranial nerves

and abdominal reflexes, should be conducted. It is necessary to evaluate the condition of soft tissues and skin, joints mobility, etc. A radiologic study includes plain radiography, CT, and MRI. This is useful, or even mandatory, not only for diagnosis (e.g., a combination of syringomyelia and suspected idiopathic scoliosis) but also for treatment (detection of asymptomatic craniocervical instability in the setting of a congenital abnormality in a patient with chondrodystrophic scoliosis). Three-dimensional reconstruction, especially in a craniodorsal view is a very reliable prognostic indicator for identification of three types of idiopathic scoliosis: spontaneously regressive scoliosis, moderately progressive ("benign", according to Min Mehta) scoliosis that can be completely corrected using corrective braces or serial casting, and malignant progressive scoliosis that is resistant to conservative treatment (Fig. 1). This important information was presented at the SRS meeting in Illinois in 1980 but, unfortunately, was ignored like its publication in France [5] a few years later. This phase of a clinicoradiological study enables a more thorough identification of the deformity etiology, rather than just attaching a genetic label "Early Onset Scoliosis".

General considerations for solving problems in EOS patients

First of all, it is necessary to identify significant problems of the lungs and respiratory function associated, in turn, with a heart condition and subsequently with the thorax development. Thoracic insufficiency syndrome was described by Campbell. Now, we know that alveolar multiplication is completed at the age of 7–8 years. Therefore, thoracic insufficiency syndrome requires early diagnosis and treatment. In addition, spinal instability, from the craniocervical junction to the sacrum, should be detected as soon as possible. Otherwise, a minimal trauma can cause irreversible changes in the spinal cord, up to paralysis.

The next task is assessment of normal and abnormal growth of the skeleton, especially the spine [4, 8], because this determines the shape of the spinal canal at all levels and, therefore, the possibility of spinal cord compression and the development of progressive deformities (scoliosis, lordosis, kyphosis). These deformities, in turn, can subsequently affect functions of internal organs, the locomotor function, and balance and cause appearance problems.

The third priority is a study of the nervous and muscular systems that enable maintaining an upright posture when sitting and standing as well as the locomotor function.

The result of all these studies should always be in the focus of surgeon's attention. Does spinal deformity change the shape of the spinal canal, increasing the risk of injury to the dura mater and its contents? In particular, the inner wall of the canal in kyphoscoliosis can be flat but can be step-like deformed. Do the vertebral bodies form a prominence on the canal wall, which can cause compression of the spinal cord? Is there any disc protrusion into the spinal canal? Are there any signs of immediate or potential instability? Are shapes of the arch roots and articular processes normal? Is the deformity mobile or rigid? Clinical examination is crucial, especially neurological examination and radiologic studies, including functional ones (traction, bending, and extensions with a roller). Upon initial examination, CT, MRI, and sometimes MSCT are recommended for clarifying the etiology of disease. For example, infantile scoliosis may be considered as idiopathic scoliosis, until MRI reveals syringomyelia, after which the diagnosis should be changed to neurogenic scoliosis.

The next issue is the effect of spinal deformity on vital organs. Are there abnormalities of the cardiopulmonary system (dysplasia, agenesis), thorax (synostosis or agenesis of ribs), muscles (aplasia of the thoracic or abdominal wall and diaphragm), and internal organs (kidneys or intestine)? What is the impact of all these diverse pathologies on the life and development of the child? All these aspects should be carefully considered and evaluated before making a decision about treatment.

The most important principles of treatment

Conservative treatment

The main requirements for this treatment in the growing child are as follows [4]:

– avoid thorax compression;
– correct or prevent vertebral collapse;
– provide spinal derotation, avoiding significant limitations to vital structures;
– provide conditions for normal life of the child (playing, walking, running) to minimize a negative psychological effect.

Serial casting. In many aspects, casting (Fig. 2) is the method of choice, being used immediately after detection of spinal deformity, in some cases soon after birth. Casting is performed under general anesthesia using a nasotracheal tube because it is necessary to apply head-pelvic traction (always very mild) and to prevent the situation when the child's teeth clamp the

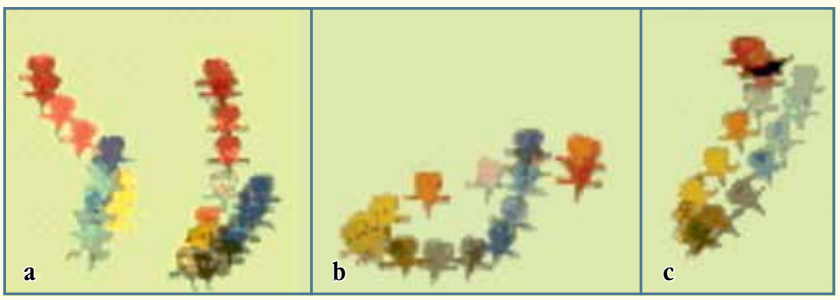

Fig. 1
Three types of infantile idiopathic scoliosis (3D-reconstruction, top view): **a** – spontaneous self-healing; **b** – moderate progression; **c** – malignant progression

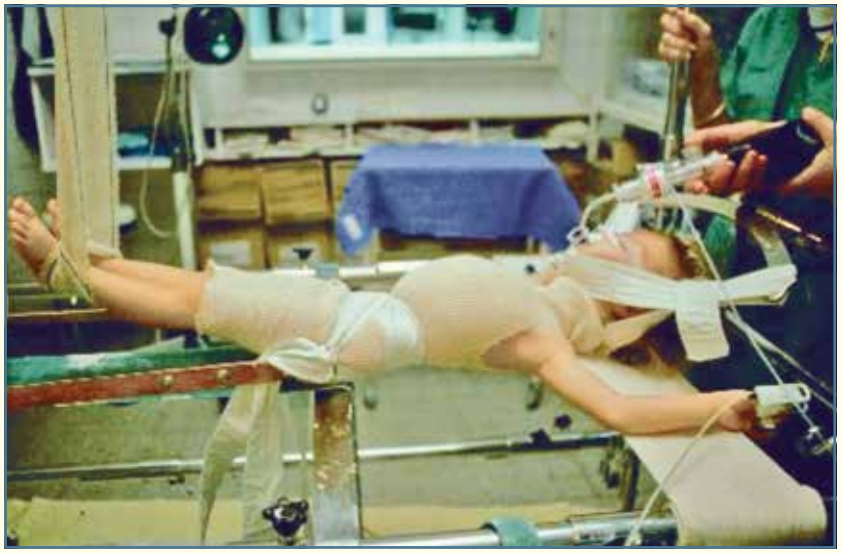

Fig. 2
Plaster cast molding. General anesthesia using transnasal intubation

intubation tube. Mild traction on a special frame makes the procedure easier to the patient, plaster technician, and surgeon. It also reduces the rate of respiratory and gastrointestinal complications. The child's body is very carefully (without any folds!) wrapped with 2–3 layers of cotton fabric. If the deformity is relatively flexible, an EDF (elongation, derotation, and flexion) technique is applied, with the use of linen straps for traction and hand molding of plaster. During the preparation, we place pieces of felt around the thorax, up to the infraclavicular area, which are removed when the cast dries, allowing room for thoracic excursion and preventing vertical orientation of the ribs. Pressure is applied to the ribs on the convex side of the deformity, below the apex, in a posterolateral direction. Windows are made in the cast to allow pressure and counter pressure, which provides a necessary derotation effect, with the vital capacity being minimally restricted.

If the deformity is large and rigid, immediate traction and derotation with linen straps do not work. In this case, correction with a Donaldson and Stagnara elongation cast should be used. The principle of this cast is based on growing distraction between the head and pelvic girdle by means of two turnbuckles fixed in both halves of the cast. A half turn of the turnbuckles, twice a day, enables 4–5 mm elongation per day. This technique is similar to that described previously, with allowance for the following clarifications:

– careful and precise hand molding of the pelvic girdle;

– the same manipulation with the occipital mandibular support;

– the use of a sufficient number of felt pads around the thorax, which are removed after the cast dries to free the skin from pressure and to provide the ribs with room for motion;

– minimal head-pelvic traction when the patient is on the cast frame.

For patients with very rigid deformities, we recommend halo traction applied to the calvarial bones, with the patient in a lying position with a load of 3–4 kg, overnight. A halo-ring is not attached to the cast to allow rotary motions and prevent degenerative changes in the movable joints of the cervical spine. This technique uses the viscoelastic properties of soft tissues of the spine: ligaments, joint capsules, tendons, and muscles. Traction is used in conjunction with positive pressure ventilation and is continued until monitored parameters reach stable values. The treatment is followed by localized surgery (spinal fusion), as in the case of congenital abnormalities; in the absence of indications for surgery, the cast is changed to a new one in accordance with the EDF principles. The cast is changed as needed (usually every 3 months), until treatment with a corrective brace starts. For the hygienic purpose, a cotton pad is changed every 15 days.

Bracing treatment. A corrective brace for a young child should meet the following requirements:

1) should be easy to put on with the help of one person (usually, the mother);

2) should not compress the thorax to avoid disrupting breathing;

3) to be preferably used after serial casting treatment;

4) to be adjustable, if possible, in height, width, and entirety of the body to match patient's growth for at least one year;

5) to exert an active, passive, or combined effect depending on the etiology of spinal deformity.

I prefer to use a Garchois corset (Fig. 3), with or without a chin support, for any type of EOS, especially in patients with poorly developed muscles. Monovalve or thoracic-lumbar-sacral-orthotic (TLSO) braces are difficult to use, and sometimes even harmful to the chest function. A Milwaukee brace is good for patients with normal muscles but requires involvement of an experienced physiotherapist. In patients with good muscles, an underarm three-dimensional carbon brace with derotational and kyphogenic effects is recommended in cases of pronounced lordoscoliosis. Any braces are individual; therefore, the technician's experience is very important.

A respiratory framework and physiotherapy are also an important part of the EOS treatment; therefore, it is advisable to engage a pediatric pulmonologist in the treatment.

Family involvement. The value of the family in the treatment of children with scoliosis is very great, so relatives should have a good understanding of the treatment goals as well as the fact that the period of active child growth is a difficult time in life of the whole family. They should be aware that surgical treatment may be required. Full transparency and mutual trust among all

participants of the treatment process the surgeon and members of his team (nurses, a physiotherapist, technician, secretary, and social worker), family, and patient – is a must.

Surgical treatment

Regardless of the scoliosis etiology, there are three major patient groups.

Patients with a localized pathology who need surgery in early childhood. Progressive congenital deformity (a hemivertebra associated with body imbalance, kyphosis, or lordosis) requires resection of the abnormal vertebra and spinal fusion at one or two levels, with or without instrumentation, which enables complete elimination of the deformity, without affecting spine growth. A similar situation is observed in dystrophic deformity (neurofibromatosis type 1 or chondrodystrophy) when a localized rotational dystrophic dislocation can lead to irreversible injury to the spinal cord. Localized dorsoventral fusion, with or without an endocorrector, resolves this acute problem. A similar approach is used in cases of inflammatory and neoplastic lesions (e.g., postlaminectomy deformity).

In some patients with congenital and degenerative deformities, epiphysiodesis (ventral in lordosis, dorsal in kyphosis, and dorsoventral on the convex side in scoliosis) gives satisfactory results. This is the consequence of a decision made with allowance for a three-dimensional effect of epiphysiodesis and the patient's age.

In all these cases, follow-up and control of the spine condition should be performed until the completion of patient growth. This principle should be followed even if some patients need to use a brace in the period of sustained growth, or if a massive surgical intervention is performed at the end of this period.

Patients with a pathology involving the entire spine and often the entire body. This includes cases of various pathological processes caused by lesions of the upper or lower motor neuron, most often spinal muscular atrophy, cerebral palsy, *spina bifida*, and damages to soft tissues (Ehlers-Danlos and Marfan syndromes, muscular dystrophy, and congenital myopathy).

Initial treatment is usually conservative. In this case, excellent results can be sometimes achieved if the patient and family strictly adhere to the regimen, and the surgeon's choice and strategy are adequate. In most cases, when patient growth is nearing completion, instrumental correction and spinal fusion are performed to stabilize the spine and discontinue external immobilization. Because of technical or psychological problems, these interventions are sometimes performed before completion of skeletal growth [10]. These patients will subsequently need ventral spinal fusion to prevent the development of the crankshaft phenomenon [2], even if this adverse effect in some of the patients with minimal residual torsion can be prevented by means of bilateral implantation of pedicle screws throughout the curve.

But sometimes, the severity of deformity and the general patient condition push the surgeon to start staged surgical treatment (fusionless treatment) to help the brace. This is done to delay the final fusion until the age of skeletal maturation in the case of conservative treatment failure in patients of 7–10 years of age. Several technique options have been developed, which have a common mechanical basis distraction forces applied to implants fixed to the spine or ribs with hooks, rings, or pedicle screws connected by shafts or rods of various sizes. Technique options, which are methods of applications of distraction or compression forces, may be divided into three categories.

1. Purely mechanical surgical procedure. Direct distraction can be performed intraoperatively using instruments that allow endocorrector elongation of 2, 5, or 8 mm. The achieved effect is fixed, and the wound is closed. The distraction maneuver is repeated every 4 or 6 months. Limitations of the technique include:

– the need for repeated anesthesia and surgery, which is associated with a risk of infection, even with minimal surgical approaches;

– inadequate distraction (not always controlled) associated with a risk of neurological complications;

– frequent hospitalizations increasing the cost of treatment.

In addition to the described complications, spontaneous bone blocks were observed during final fusion in almost 40 % of cases when this technique was used, which was difficult to relate to previous mini-surgeries. According to our observations, these spontaneous blocks are more pronounced in the case of dual rod correction that we used many years ago with pediatric Cotrel – Dubousset rods. I have no experience with VEPTR instrumentation developed by my friend Campbell.

2. An electrically powered system was developed in 1997, based on expanding prostheses to replace removed tumors. A preloaded spring is placed in a plastic tube. The spring decompresses after local heating of the plastic tube by electric current delivered through a transcutaneous wire. This technique has several advantages over that described previously: distraction is performed without anesthesia and hospitalization; the treatment process is controlled by the surgeon and patient; distraction is quite sudden. The technique was used for several years, which resulted in a reduced rate of infection, although rod fractures occurred. There appeared the possibility to delay final fusion and better control spinal deformity.

3. A magnetically controlled system, which was experimentally tested at our clinic in 2003, is the latest generation of compression-distraction spinal rods. Initially, a small magnet, which was connected to a threaded rod and activated by a large external magnet, was implanted. It enabled distraction or compression, depending on the direction of external magnet rotation. Further development of the system resulted in the use of the external magnet alone. The amount of planned correction ranged from 4 to 8 cm. Rod fragments fixed to the spine or ribs are linked via a domino-type connector. The rod may be curved according to the anatomy and is implanted through small incisions. The advantages of the technique are as follows:

– a minimal approach and subcutaneous implantation;

– progressive elongation of 1/10 to 1/2 mm per one revolution of the magnet;

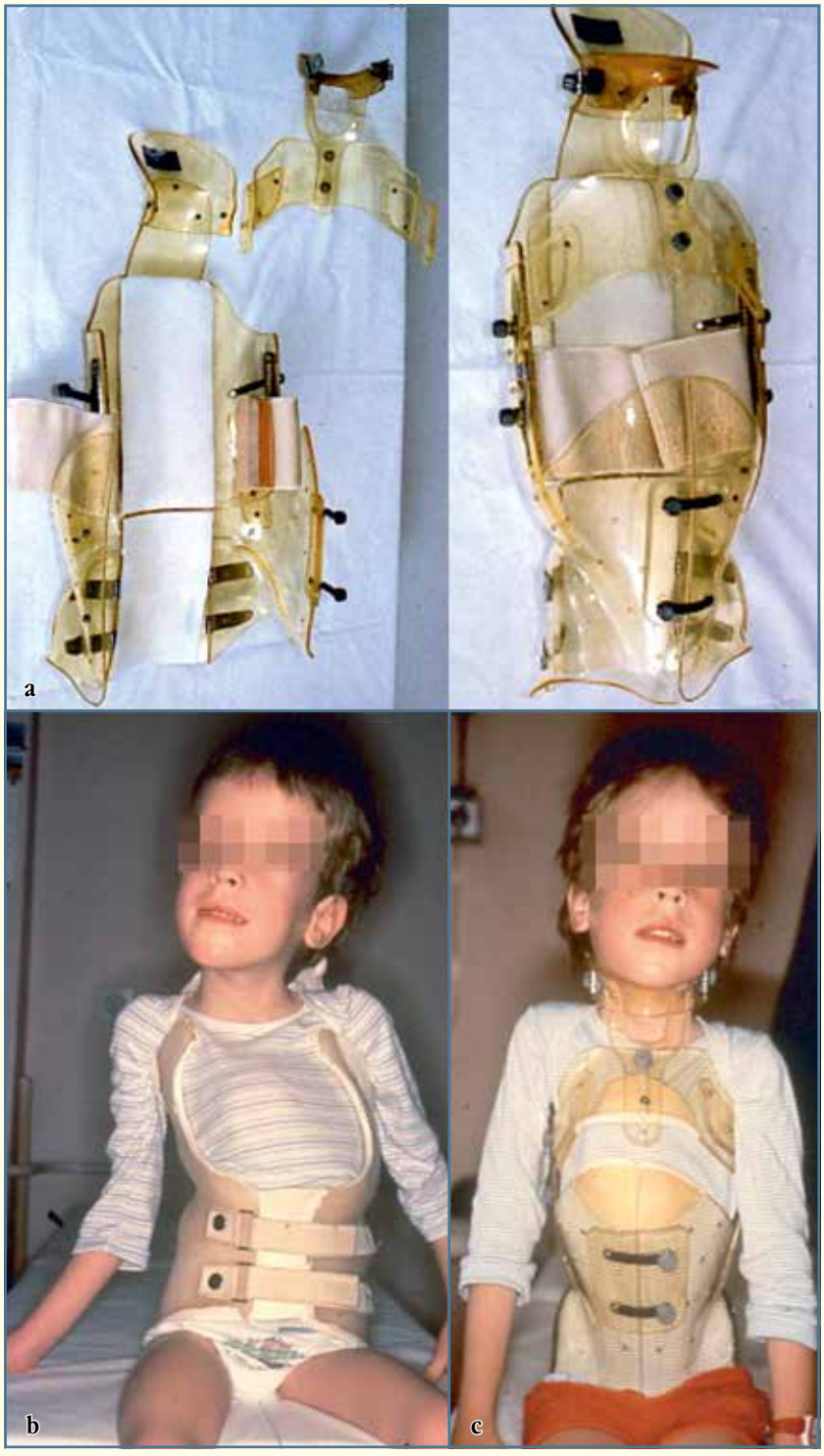

Fig. 3
Comparison between a Garchois brace and a "poor" brace: the Garchois brace is easy to use, can be adapted to the growing child, and has a high degree of rigidity and ability to support the position of the head; a pelvic portion opens like a book due to two rear hinges, can be adjusted in all directions as the child growth, and does not restrict breathing: **a** – appearance of the Garchois brace; **b** – a child in the "poor" brace; **c** – a child in the Garchois brace

– a painless procedure;
– no anesthesia;
– elongation is performed by parents at home.

The preliminary results of technique application are very promising, but it is too early to talk about its widespread use. It is used only at a few centers to explore in detail its capabilities. Unfortunately, production of this instrumentation was discontinued.

Also, the discussed group should include some congenital abnormalities or syndromes involving the entire spine in the pathological process (e.g., Jarcho – Levin syndrome). These deformities are so pronounced, and respiratory disorders are so severe that Campbell has described them as thoracic insufficiency syndrome. For example, when a segmentation defect involves the entire hemithorax, or when rib blocks are combined with long asymmetrical vertebral blocks, hemivertebrae, etc., surgical treatment is required because other methods are ineffective in these situations. The VEPTR instrumentation can be used as a rib distractor, but in some cases, it is excessively massive for a small child. That is why I prefer pediatric Cotrel – Dubousset rods, which are fixed to the spine or ribs, or hybrid constructs (spine and ribs). The main drawback is repeated surgeries every 6 months. They increase the risk of infection, while the distraction rate is insufficiently physiologic. Furthermore, we noted that a pre-existing kyphotic deformity is a relative contraindication for the technique and is a source of mechanical complications (displacements of a cranial anchorage regardless of its type – rings, hooks, pedicle screws, and tapes).

We also have experience with a Luque technique [6], especially in the most complex cases, such as myelomeningocele with severe scoliosis and pelvic obliquity or acute thoracolumbar kyphosis requiring kyphectomy before the age of 4 years, along with stabilization of the spine. This technique, if avoid aggressive manipulations during spine skeletization when passing wire loops, does not lead to the development of spontaneous blocks and preserves a vertebral growth potential. The technique is also used in spinal muscular atrophy and similar pathology; it does not require postoperative immobilization due to strong fixation of rods to the pelvic bones.

Therefore, the described techniques help overcome significant problems in patients with a poor prognosis, although it has been noted that the aforementioned constructs do not provide control of the deformity in the horizontal plane at the end of the growth period. Therefore, a pronounced rib hump is often seen in patients who have passed all stages of the treatment.

To avoid some of these complications, new technologies, e.g. magnetically controlled rods (under clinical trials), acting without additional interventions, anesthesia, and pain have been developed. They can be used at home, with the distraction rate being close to the parameters of patient's physiological growth.

Patients with gross deformity of a part of the vertebral column, e.g. the entire thoracic spine. This applies to cases of the so-called infantile and early onset juvenile scoliosis. Conservative treatment with successive use of serial casting, corrective braces, and again casting is optimal for this group of patients. In some cases, the patient reaches the end of the growth period with a normal spine in terms of shape, function, and mobility. On the other hand, some patients undergo surgery (correction and fusion) as soon as they reach the peak of growth in adolescence. Because these children are very active and have normal muscles, which allow them to run and jump, i.e. to act their age, there may be temptation to start staged treatment with growing rods to avoid external immobilization in childhood. Although corset therapy maintains normal growth of the spine, it is associated with physical limitations and appearance problems.

In the world, there are a large number of endocorrectors that are implanted using ventral and dorsal approaches. Ventral corrective braces were used as temporary epiphysiodesis in patients only before adolescence. Our experience demonstrates that the indications for this technique are quite limited. The dorsal technique of rod implantation on the concave side of deformity using a domino-type connector capable of extending is the simplest option. The technique enables correction and prevention of vertebral collapse, but is less suitable for control of the sagittal contour, and does not affect the deformity in the horizontal plane. Therefore, cosmetic surgery for a pronounced rib hump associated with the risk of respiratory function impairment is often performed at the end of puberty.

In addition, we present a method that is very simple, less aggressive, and less expensive compared to magnetic rods or VEPTR instrumentation, which was developed by Dr. Lofti Miladi, one of my students at the St. Vincent de Paul Hospital in Paris, in 2005. It does not require a meticulous technique. A single titanium rod, called a H3S2 construct, is implanted on the concave side of the curve through minimal incisions. Two supralaminar hooks and one pedicle hook are implanted as a cranial anchorage (H3). Two monoaxial pedicle screws (S2) form a caudal anchorage at the base of the curve. The spine is exposed extraperiosteally. The rod is bent according to a normal sagittal contour of the thoracic and lumbar spine and implanted transmuscularly in a caudocranial direction. The basic idea is to align the rod along the vertical axis of the patient's body. Anchorage implantation points are determined based on a spondylogram performed under longitudinal traction. Preservation of surrounding soft tissues and bones reduce the risk of fibrosis on the concave side and a spontaneous bone block. Preoperative traction is used if the deformity exceeds 50°. Axial traction and spinal cord monitoring are used intraoperatively. After surgery, no external immobilization is applied.

According to the Dr. Miladi's recommendations, the rod is elongated at its lower end every 8–12 months. If before surgery, the Cobb angle in the axial traction position exceeds 70°, ventral discectomy and fusion precede surgical correction. The first results (23 patients with a mean follow-up period of 3.5 years) are optimistic: Cobb angle correction of 57 % has been achieved. The rate of complications is 22 % (4 rod fractures, 2 infections, no neurological complications). Preoperative correction of severe rigid scoliosis, the absence of a connector, which is a weak point of the growing construct, and elongation due to a rod fragment distal to the anchorage site explain the low rate of complications. It should be noted that the described method is applicable for early onset scoliosis of any etiology.

To extend the discussion on various technical approaches, a Shilla technique, developed and popularized by McCarthy and McCullough [7], should be mentioned. The technique seems to be very promising because circumferential apical fusion is performed within a limited site, and longitudinal growth of the spine can continue, at least theoretically, above and below the limits. The results look quite good, but I have no experience of this technique.

When applying techniques of staged surgical treatment for early onset scoliosis, I have always used external fixation with a light brace to protect fixation points from multiple repetitive stresses inherently associated with the physical activity of a small child. At the same time, I am satisfied with the results of H3S2 application without postoperative immobilization of preadolescent children who are even more active than small children.

The final problem for the third category of patients is the time to remove a temporary metal construct (a source of many artifacts during radiography in adult life) and to carry out the final fusion after endocorrector removal. In most cases, this surgery is necessary for the best results in the future.

With respect to all these operations, regardless of the deformity type, patient's age, and follow-up period, the primary surgeon's concerns are respiratory support and physiotherapy with or without a respiratory apparatus.

Indications for treatment

These indications are based on the treatment outcomes in many patients whom we have observed throughout our professional life.

Basic concepts

Except special cases, we should always begin with conservative treatment. We should remember that any surgery at any body site damages tissues, while cicatrization leads to the formation of scars. Fibrous tissue, which forms a scar, increases in the case

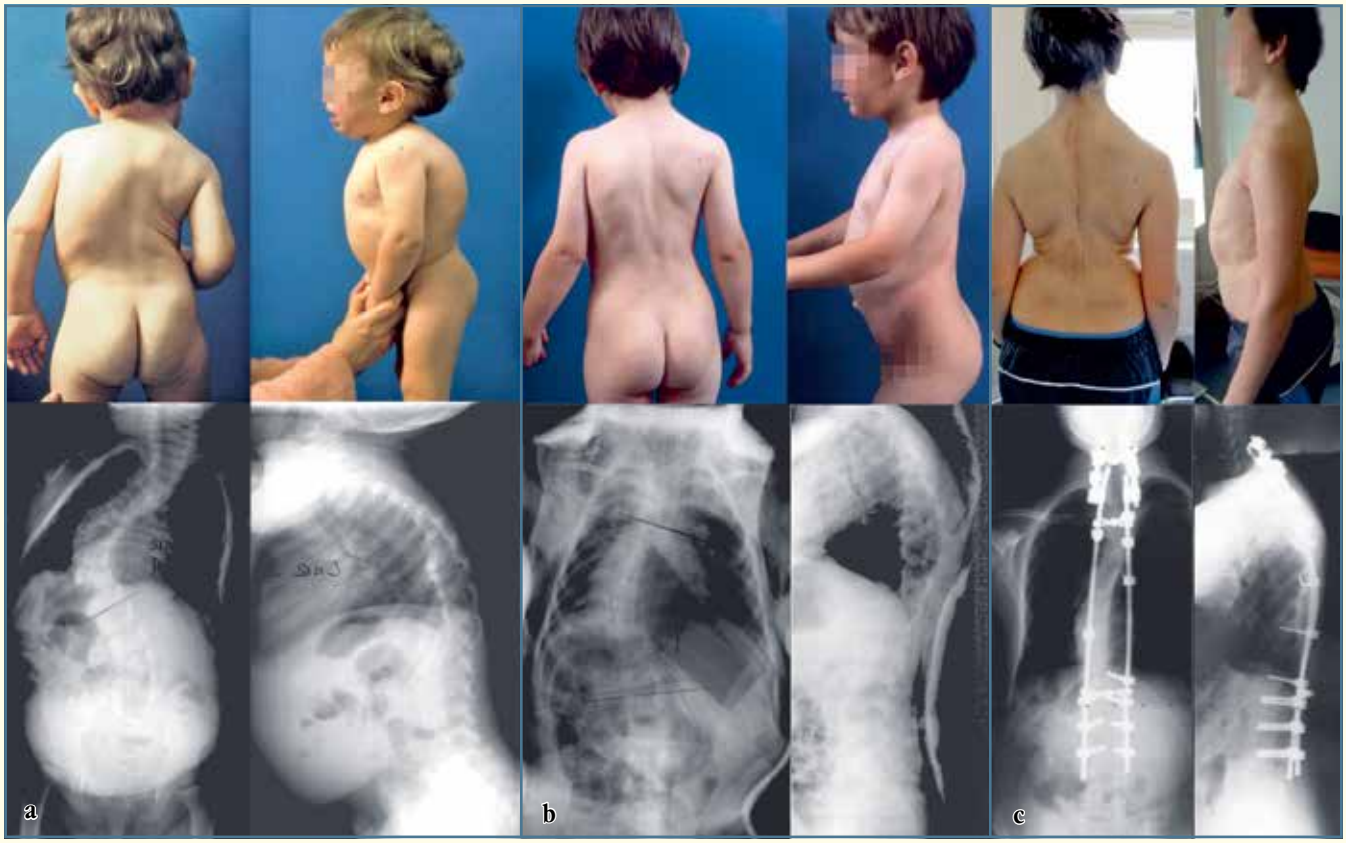

Fig. 4
Radiographs and photos of a child treated for malignant progressive idiopathic scoliosis (Cobb angle of 80°): **a** – age of 2 years (before treatment); **b** – age of 4 years (continuing treatment with a plaster cast); **c** – age of 16 years (after spinal fusion surgery)

of wound infection, especially if surgery is accompanied by implantation of a foreign body. It should be noted that re-operations at the same body site increase the amount of scar tissues and, therefore, the risk of infectious complications.

That is why we should prefer conservative treatment of early onset scoliosis as long as possible and make a decision on surgery only when indications are evaluated as urgent. If the amount of surgery is limited, and the number of interventions is less than two, there are no restrictions at any patient's age. On the other hand, if surgery requires an approach to the spine over a large area (e.g., the entire thoracic spine), we should remember that if complications develop at the beginning of surgical treatment, we will have to perform staged surgeries before the age when the final intervention can be carried out. Therefore, I follow the principle of semi-delayed surgery using minimal approaches, such as with the H3S2 construct. In these conditions, new highly promising techniques, such as magnetically controlled rods, can demonstrate their efficacy.

Indications for treatment of main etiological forms of deformity

Idiopathic scoliosis. My preference is conservative treatment in possible cases. There are very rare situations where a disturbance of growth by fusion is preferred or acceptable if growth can be preserved without surgery. Only in very rare cases of malignant progression after repeated failures of corset therapy (including serial casts), we should consider surgery using a minimally invasive technique. In reality, this is a variant of delayed early surgery, as in the case of H3S2. We should try to restrict the surgical site only to a limited part of the vertebral column. Staged surgical techniques, when applied, should affect only a structurally changed part of the spine, without involvement of mobile segments, so as not to disrupt the capability of the vertebral column to compensate for balance impairments. In cases of malignant progression, when only three apical vertebrae are maximally rotated, the Shilla technique can be useful. At the same time, we call for caution because staged surgical treatment is associated with numerous complications. We believe that the best results can be obtained using serial casts and corrective braces before the age of puberty when the final spinal fusion, with or without anterior or posterior instrumentation, can be carried out (Fig. 4).

Congenital deformities. In cases of congenital malformations, it is easy to understand the difference among patients with different pathological situations of the same etiology. Localized surgery, such as resection of a thoracolumbar, lumbar, or

cervicothoracic hemivertebra followed by short fusion, solves the problem quickly and completely. The treatment is indicated as soon as deformity progression is verified. An asymmetric defect of segmentation that affects the entire thoracic spine and is accompanied (or not accompanied) by multiple synostoses of ribs on the concave side of the deformity can not be treated with early fusion on the convex side of the curve because this results in a short, small, and rigid thorax. As experience has demonstrated, consequences for the respiratory function in this situation are catastrophic because of limited growth of the thoracic spine, which creates extremely unfavorable conditions for the thorax development. In these cases, it is necessary to use distraction devices that are minimally invasive during implantation and require the least number of reoperations. Distraction should be started before the age of 8 years so as not to interfere with normalization of the thorax volume during the growth process. The whole process is optimized by an additional treatment – positive pressure lung ventilation, prescribed as soon as possible.

Mutual adaptation and combination of these two basic concepts of congenital scoliosis treatment depend on the individual anatomy, residual potential of growth, balance, and associated abnormalities.

Neuromuscular pathology. From my point of view, it would be a mistake to say that conservative treatment is ineffective in deformities of this etiology. We have data to refute this point of view; this becomes even more important with the use of a Garchois brace as a preventive measure in patients with pathology, such as spinal muscular atrophy, accompanied by a sharp decrease in the muscle tone. The brace is used as soon as the patient is able to sit. If large deformity has already developed, active treatment begins with a plaster cast and then is continued with the Garchois brace, with or without chin support, until the age when the final fusion can be performed. In some cases of infantile cerebral palsy, a light brace providing head support is used in the postoperative period. If preventive treatment is started before development of gross spinal deformity, and if the patient and family adhere to all doctor's recommendations, these measures may be sufficient until puberty when the final fusion is performed (or not performed). Otherwise, multi-staged surgery should be used. It is necessary to realize that many patients with this disease are not tolerant to this treatment when the patient's condition may require treatment every minute, which explains a high failure rate.

Treatment of patients with dystrophic pathology (neurofibromatosis type I or chondrodystrophy) is also long-term and difficult due to the fact that the apical dystrophic zone often requires early ventrodorsal localized fusion, and other spinal segments can be corrected by plaster casts and/or corrective braces before the age of final fusion implementation. Patients with soft tissue dystrophies are usually treated as patients with neuromuscular disorders. Syndromic pathology often involves a whole bunch of pathological changes and requires a successive use of various treatment options.

Conclusion

EOS patients represent a specific group with common basic treatment philosophy. A technique and goals of treatment are determined individually for each type of etiology, for each patient, with allowance for prognostic factors.

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