Typical and Atypical AIS. Pathogenesis

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Abstract. AIS hypothesis has the right to recognition, if it explains the transition of "healthy" vertebra column into status of "scoliotic" one. AIS is the most investigated disease in the history of orthopedics, but up the present time there is no clear explanation of some its phenomena: vertebra column mono-form deformation along with its poly etiology character, interrelation of its origin and development and child's growth process etc. The key for authors' view at AIS was scoliosis with non-standard (concave side) rotation. On the bases of its' multifunctional instrumental investigation results (Rtg, EMG, EEG, optical topography, hormonal and neuropeptides trials, thermo-vision methods and other) in comparison with typical AIS was worked out the new hypothesis, part of it is suggested for discussion. In the work under observation is the sequence of appearance of typical and atypical scoliosis symptomatology beginning from the preclinical stage.

Keywords. Adolescent idiopathic scoliosis (AIS), typical/atypical scoliosis, pathogenesis.

Introduction

Since the times of Hippocrates, who for the first time described scoliosis, the amount of information on the most mysterious of its kind - adolescent idiopathic scoliosis (AIS) – has become so great, that our first priority is not getting new information on the disease, but trying to find a place for the knowledge we already have in the greater AIS picture. Such a task can be compared with a puzzle game for children and adults, where to get a result – you have not only to find the key fragments of the picture, but also to remove items that are not relevant to the final picture. On the basis of this ideology that came to us over many decades of original research and of studying the works of other authors we have come to a number of conclusions, both conventional and original, that we bring to the discussion of our colleagues.

1. Background

The first and the most important fact is the connection of the AIS origin and its development with the child's growth process. This conclusion is so indisputable one, that can be hardly debated in the current literature.

The second, equally important conclusion, should be named the fact of two-component vertebra column character: the first of these components – the spinal cord, and the other – its bone-ligament-muscular sheath. This conclusion is as obvious as the previous one, and so practically is not debated. But at the same time, these components

together form the most complex in the body anatomical and physiological complex in which two organs, different in origin and staying at different hierarchical levels of the body, doomed to grow together, including the growth process.

The third conclusion is that the term "etiopathogenesis" cannot be applied to the AIS, as its obvious poly etiology cannot stay in the same linguistic conjunction with its own indisputable mono-form character. This goes due to the fact that for various AIS reasons (and because of general agreement - not debated) they all lead to this one single phenomenon, which is being independent of the initial cause, is triggering in any case mono-form AIS start and further development, [one C-curve, or C-shape and two C-curves, or S-form]. What is this phenomenon? There is no answer.

The following conclusion from the AIS reflection is that outside the frames of its study stays such a period of the formation of vertebra column 3D deformation, when a healthy (normal) spine is turning into a sick one (scoliotic). This period Dr. K.Bagnall called "the dark zone". And because of the lack of knowledge about this "zone" for a long time we just could not talk about effective prophylaxis of AIS and could only carry a hard fight against its progressive course. And in this struggle we were not always winners.

A "key" that generated a discussion of our proposed concept of the AIS pathogenesis was the revealing of its atypical kind, in which the main defining feature – "concave side rotation" – was noted at the beginning of the twentieth century. Half a century later, almost simultaneously in Russia (Dudin M., 1980, 1981) and in the USA (Armstrong G., 1981) was found that such a kind of pathological rotation takes place in the independent AIS form with the unexpected quality – a benign course.

According to the results of our study of such AIS kind has been revealed a number of clinical and radiographic features that give us opportunity confidently carry out differential diagnosis. These data are presented in Table 1.

Table 1. Summary table of clinical and radiological signs of scoliosis with **typical** and **atypical** pathological rotation of the vertebrae

Plane	**Kind of scoliosis**	
	AIS with typical pathological vertebrae rotation (convex side rotation)	**AIS with atypical pathological vertebrae rotation (concave side rotation)**
Frontal plane	Frontal vertebra column curvature, more often: the right-sided in thoracic region and left-sided – in the lumbar one. On the anteroposterior radiograph – a clear picture of the frontal vertebra column curvature.	Frontal vertebra column curvature, more often: the left-sided in thoracic region and right-sided – in the lumbar one. On the anteroposterior radiograph – a clear picture of the frontal vertebra column curvature.
Sagittal plane	Lordosis of physiological thoracic kyphosis and increased physiological lordosis in the lumbar spine ("flat back"). On the lateral radiograph – **vertebral column is vertical.**	Intensification (with signs of rigidity) of a physiological thoracic kyphosis and smoothing of physiological lordosis in the lumbar spine ("sway back"). On the lateral radiograph – **vertebral column is bow-shaped**.
Horizontal plane	In clinical picture asymmetrically enlarged paravertebral muscular-roller is detected on the **convex side** of the vertebra column front-side	The clinical picture of asymmetric enlarged para-vertebral muscular-roller is detected on the **concave side** of the vertebra column front-side

	curvature. On the anteroposterior radiograph – all signs of the rotation of bodies vertebras in the **convex side** of the frontal curve, the leading one from them – asymmetric increase in the transverse dimension of X-ray projection of the base root of the vertebral arch on the side of **convexity**.	curvature. On the anteroposterior radiograph – all signs of the rotation of bodies' vertebras in the **concave side** of the frontal curve, the leading one from them – an asymmetric increase in the transverse dimension of X-ray projection of the base root of the vertebral arch on the side of the **concavity**.
X-ray anatomy of vertebrae's bodies (wedge-shaping, wedging)	The wedge base is on the **convex** side of vertebra column frontal curve.	The wedge base is on the **concave** side of vertebra column frontal curve.
Character of the course	**Progressive course character is possible**	**Absolutely benign course and there is even a possibility of self-correction.**
Frontal plane	Frontal vertebra column curvature, more often: the right-sided in thoracic region and left-sided – in the lumbar one. On the anteroposterior radiograph – a clear picture of the frontal vertebra column curvature.	Frontal vertebra column curvature, more often: the left-sided in thoracic region and right-sided – in the lumbar one. On the anteroposterior radiograph – a clear picture of the frontal vertebra column curvature.

Analysis of the sequence of symptoms observed in the transition of a healthy spine to a scoliotic one, showed the following logical stages:

Typical AIS

1-st – development of a "flat back";

2-nd – body torsion (shoulder and pelvic girdle front axles parallelism disorder);

3-d – the first signs of pathological convex side rotation of vertebrae in the zone of apex scoliotic curve (positive Adams test);

4-th – further development of the AIS (progressive or non progressive).

Atypical AIS

1-st – the development of kyphosis ("sway back");

2-nd – the first signs of vertebrae pathological concave side rotation in the zone of concave scoliotic curve (Adams test isn't always positive);

3-d – the level <15º for frontal and <15º for the horizontal component are their maxima, after which they won't increase. It's possible (without treatment) if the vertebra column deformation increases only in the sagittal plane.

Listed staging in clinical symptoms development is fully confirmed by the instrumental diagnosis (Rtg, EMG, Optical Topography, RND, Hormonal and Neuropeptide research et al.).

Mathematical modeling and the results published in the IRSSD papers in 1998, gave evidence that both variants of AIS are to be the defensive reaction on the two variants of conjugacy disorders of the dynamical processes of the longitudinal growth of the spinal cord and its bone sheath. In a typical AIS is compensated the relative excess length of the bone vertebra column, and in atypical - its shortage. But all this can be only in a **growing child**!

Meanwhile, the maintenance and provision of this conjugation is under the control of, in the first place, the nervous and endocrine systems. In addition, different embryonic origin of the spinal cord and its bone sheath suggest "personal" programs for their development. It follows that in a complex system of regulation of vertebral

complex longitudinal growth there are enough weak links, the failure of which leads to the necessity of compensatory responses. In the case of AIS, we can observe the effect of Ch. Sherrington (Nobel Prize 1932) - the multitude of initiation factors leads to a final reaction (response).

The scope of this publication does not allow us to consider all the results on the topic we have obtained through the research phases, so will be reflected the main one: with a relatively intense or insufficient bone growth of the spine, in particular because of the peculiarities of the osteotropic hormonal profile (materials IRSSD, 1998), are developed "discomfort" conditions for the spinal cord. In the first case it is "stretched" and in the second – "constrained".

1. A typical AIS **develops only in the case of spine cord extension,** if there is no necessary correction of hormonal regulation of bone vertebra column growth inhibition. Such a correction – is the first normal physiological response to the situation. If it is not sufficient, the excess of the longitudinal growth of bone vertebra column increases, as increases the stretching of the spinal cord.

It's fair to say that in the form of the vertebra column there are natural reserves to solve the emerging conflict. They are laid in the physiological curves - thoracic kyphosis and lumbar lordosis. The decrease of the first and increase of the second provide a certain reserve for maintaining a stable spinal canal length under the intensive growth of the column consisting of vertebrae bodies. Therefore, the change of vertebra column physiological curves we refer to the next (second) phase of a healthy compensation, opposing the spinal cord extension. And if the number of the above mentioned answers to the spinal cord stretching is sufficient, then the conflict situation is resolved.

But since the physiological curves' reserve is limited, the preservation of inconsistency of the spinal cord longitudinal growth to its vertically oriented bone sheath length, leads, from the point of view of theoretical mechanics, to the "launch" of the next and the only possible mechanism – twisting of a long part around the short one. Here, take on a leading role transverse-spinal (rotators) muscles and their task is to "take away the surplus" in the column of vertebrae bodies due to its twisting around the spinal cord and thereby reducing its tension.

And this, absolutely non-specific twisting, starting under the laws of biomechanics of the spine in the lumbar and lower-thoracic regions, by itself leads to disruption of the shoulder girdle position, which loses its strictly frontal orientation. As a result, there occurs as a functional difference in size of upper extremities, so as a number of other abnormalities in posture and the perception of the horizon's line. And this abnormal position, let us call it sub-pathological, needs for its compensation, which is not associated with the primary conflict between the spinal cord and bone vertebra column.

Thus, it turns out, that the result of a compensatory response to the growth non-conjugacy in the vertebral complex makes it necessary to have a second kind of compensation – compensation of torsion of the same complex. In the process begins the activation of transverse-spinal (rotators) muscles of the cranial area. At this level, for complete restoration of the shoulder girdle frontal position, they de-rotate vertebrae in an amount equal to the volume of the previous twisting. It is important to note that "the de-rotators" became the same transverse-spinal muscles, but from the contralalateral side, and by their reduce they "return" upper thoracic vertebrae in the middle position.

It's necessary to emphasize the fact of the vertebrae's saved rotational displacement between the vertebra column region with primary, or "caudal torsion" and the region of "compensatory de-torsion". As a result there is the formation of a primary frontal deviation (curve) of the column of vertebral bodies, which possesses all the features of 3D deformation. It has already lost the vertical stability and was acquired condition for the implementation of the **Hueter-Volkmann** law, according to which non-loaded for a long time growth zones are functioning more intensively than the loaded ones. As a result it is provided an irregular (non symmetrical) growth of the vertebral bodies and the generation of the wedge-shaping.

Here we are approaching the dramatic finale – the generated wedging leads to the frontal curve increase, which breaks more and more the vertical stability, which leads in-turn to creation of more favorable conditions for the new asymmetric growth of the vertebral bodies. Thereby appears so called "vicious circle" mentioned by J.Stokes (1996) – i.e. comes into existence the structural three-plane deformation – **scoliosis**!

To complete explanation of our point of view, we consider it possible to express our opinion on the AIS statistical peculiarity – its greater frequency in girls.

The factual basis for these arguments is confirmed by well-known in pediatrics Scammon's diagram (1930), according to which in children from early puberty period (10 years of age) up to its' end (16-18 years) the linear growth of nervous tissue (spinal cord! *author*.) practically does not occur. At the same time, the curve of the longitudinal growth of the body is directed steadily upwards (pubertal growth stimulus). Complementary to this are the data of J.Brock (1954), which specify one important feature – among the girls in this period the process of normal growth of the torso relatively to the length of the whole body is less intense than the same among boys. In other words, in girls of this age group vertebra column **does not have to grow rapidly concurrently maintaining a high rate of elongation of the lower extremities.**

We believe that the restraint character of the girl's vertebra column longitudinal growth (performing tissue) have to correlate with the same restraint longitudinal growth of the spinal cord (directive structures). But at puberty intensive growth of the lower extremities in girls, the leading factor for which is stimulating osteotropic hormonal profile general for the whole body, is increasing the role of spinal and central regulatory structures. Their goal is the selective control of osteogenesis process within only one segment – the bone vertebra column.

This naturally raises the tension in the regulatory apparatus, and it, in turn, increases the probability of disorders in security of the same type of conjugacy. Thus, the more complex the system is arranged, the more possible are the causes for its "failure" and it makes regulation of the growth vertebral complex elements conjugacy less secure and with more than in boys probability of its "failure".

This means that in healthy children an allometry skeletal growth process is influenced by their sexual characteristics. There is no doubt that the latter are associated with the endocrine system, which itself operates in accordance with prevailing gender formed in the evolution of the human genetic code.

2. Atypical AIS **develops only in the case of the spinal cord "tightness"**, if does not take place the necessary correction of hormonal regulation of bone vertebra column growth activity. As in typical AIS – this is the first normal physiological response to the situation. If it is implemented, the lack of longitudinal bone growth of the spine increases, as increases and the "tightness" for the spinal cord and so occurs the need in other protective mechanisms and reactions.

In essence, such a sequence is identical to that, which occurs in typical AIS, but with the opposite sign, which emphasizes the unity of the conditions for the origin of the two types of three-plane vertebra column deformation – typical and atypical, which we regard as contradictory, clinically relevant, options for maintaining the physiological mechanisms balance between processes of the longitudinal growth of spinal cord and its bone sheath.

So, if the osseous formation process activation does not occur, primarily will be manifested at the height of the vertebral bodies, which is an important factor in normal vertebra column shaping. As a result, it is obligatory the increase of physiological thoracic kyphosis increase and reduce of lumbar lordosis. In clinical picture the doctor observes the "sway back or hunched back", which we regard as the sagittal component of possible in future 3D deformation.

If osseous forming process for several reasons remains insufficient and under the conditions, that vertebra column kyphosis process has a good reserve, then under the depletion of the latter, as well as in case of typical AIS, in accordance with the same laws of theoretical mechanics, mechanical compensation should come from the same "twisting" of the long part (but in this case – it is the spinal cord) around the short one (bone vertebral column).

However, opposite to the formation of typical AIS, in the formation of atypical scoliosis rotational component the major role is played by the long back muscles. We tend to think that these muscles with their unilateral reduction lead to atypical vertebral rotation. This is evidenced by the fact that is observed in normal torso bend over – to the rotation of the vertebral bodies in the direction of the formed frontal curve concavity.

This character of pathological vertebral rotation leads to the artificial elongation of the spinal canal, which is the final element of the compensation options of anatomical and functional disproportion character of spinal cord and bone vertebra column. Fortunately, the peculiarities of the anatomical elements of the latter bears such features, that under the realization of the described above compensatory mechanism the column of vertebral bodies becomes a rigid "core" with a minimum frontal mobility. This concrete fact stays on the way of the further vertebral complex frontal curvature, and as was noted above – this type of AIS is not progressing.

And the last. We would like to point out that the development of both typical and atypical AIS stop, if the "growth" conflict between the spinal cord and its bone sheath at any stage of development of 3D deformation is eliminated. This creates a wide range of cases with different clinical course variants – from rapidly progressive (compensatory mechanisms fail completely), progressive (compensatory mechanisms only partially achieve the objective) and to non-progressive types (compensatory mechanisms cope with their task).

2. Conclusions

In conclusion, let us formulate and state the following provisions:

- AIS pathogenesis (typical and atypical) – is a chain of consecutive, clinically relevant, compensatory responses (reactions) to nonconjugacy of spinal cord and bone vertebra column longitudinal growth;

- nonconjugated longitudinal growth of spinal cord bone and vertebra column has many causes, which can be conditionally divided into hormonal, spinal and central, that fully explains the spine 3D deformation poly etiology;
- regardless of the causes of nonconjugated longitudinal growth of spinal cord and bone vertebra column, the character of compensatory reactions is a stereotyped one, which determines the final clinical AIS mono form character;
- the most "intense" period for the directive systems of the organism (nervous and endocrine systems) – pre-puberty and puberty age final maturation periods and concretely with these periods is most often connected AIS beginning and development;
- excessive longitudinal growth of bone vertebra column (relatively to the spinal cord), especially in the pre-pubertal and pubertal periods, creates the conditions for the occurrence of typical AIS, and in the cases of its insufficiency - for atypical AIS;
- regulation of skeletal growth in girls at puberty period has its own peculiarities, which increase the risk of nonconjugated longitudinal spinal cord and bone vertebra column development;
  the described AIS pathogenesis gives us an opportunity to see fundamentally new directions of its prevention and treatment.

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