Basic principles of idiopathic scoliosis initiation and progression

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Abstract
The etiology of idiopathic scoliosis is still a matter of debate as its common cause had not been found. But some basic principles in its initiation and progression do exist. The curvature of the spine is always accompanied by rotation of the vertebrae and rib cage as ribs are connected by intercostal muscles and cannot spread apart on the convex side when the spine bends. The rib cage rotates instead. The deformation becomes irreversible if bone resorption and remodeling change shape of the ribs and vertebrae or if ligaments are not firm enough. When the rib cage and vertebrae become structurally rotated, the vertebrae lose balanced support from the ribs from both sides. Shear forces from the ribs turn vertebrae even more and push vertebral bodies toward the convexity. Continuous progression of scoliosis starts and can be stopped only with a properly designed brace.

Keywords
Idiopathic scoliosis, Etiology, Thorax, Bone remodeling

Introduction
Scoliosis, the side curvature of the spine, was known from the time of Hippocrates. Many different hypotheses were postulated on its etiology, but no common cause had been found [1]. However, some basic principles governing its initiation and progression definitely exist. If we want to explain them, we should answer some questions. 1., Why is side curvature of the spine always accompanied by rotation of the rib cage and vertebrae; 2., Is structural scoliosis fixed by primary bone growth in growth plates of the vertebrae or by bone remodeling by which bones adapt to external loads; and 3., What drives the progression of scoliosis? Some alternative views to the prevailing explanations are discussed below.

Curvature and rotation
Human spine is a vertical column of 24 articulating segments, seven cervical, twelve thoracic and five lumbar vertebrae separated by intervertebral discs. Such a structure would be very unstable and prone to collapse without additional support. Ligaments and muscles provide some support but are not able to prevent rotation or buckling of the vertebral column. The role of the rib cage is essential for the stability of the human spine. Each of the thoracic vertebrae are supported by a pair of ribs, connected in front by the sternum and costal cartilage. Ribs articulate with vertebrae at two points, one on the vertebral body and the other on the transverse process. The symmetrical rib cage fixes vertebrae in the midline of the body and prevents rotation of the vertebrae with dual articulating surfaces on each side of the vertebrae (Figure 1).
Fig. 1: Ribs of a symmetric chest stabilize vertebrae and straighten the spine.

When somebody leans or curves to the side, the spinal column bends. The gaps between the ribs on the convex side should become larger in this case. However, this is possible only to a very limited degree. Ribs are connected by intercostal muscles and cannot spread apart. When the spine is curved to the side, a tension builds up in the thoracic wall. It can be released only with rotation and deformation of the rib cage which is possible due to flexibility of the ribs and especially the costal cartilage. The convex side of the rib cage could slip forwards or backwards, but slight rotation to the right already exists in the thoracic vertebrae of adolescents [2] and paravertebral muscles at the back prevent rotation forwards. They are known to be active and stronger on the convex side of the scoliotic thorax [1]. As a consequence, the rib cage slips backwards on the convex side and vertebrae rotate with the ribs. This is reversible as long as the ribs and ligaments are flexible enough to regain their original form upon straightening of body posture. But when bone structure is remodeled after bone resorption stimulated by frequent deformations, the rotated ribs and vertebrae become fixed in the new position. Structural scoliosis develops. If joint ligaments are not firm enough the deformation of the rib cage becomes easier irreversible. Scoliosis develops in patients with congenital laxity of connective tissue [3] and in children with idiopathic scoliosis joint hypermobility occurs more frequently than in healthy controls [4].

Rotation of the vertebrae is usually explained as a consequence of hipokyphotic or lordotic thoracic spine because of anterior spinal overgrowth. Vertebral bodies would rotate when a person bends forward. This theory does not explain why rotated vertebrae and ribs are found in all structural scoliosis cases. If bending of the thoracic spine is not possible without rotation of the rib cage, the coupling of both phenomena is inevitable.

**Bone growth and remodeling**

Most spinal deformities begin as a nonstructural scoliosis [5]. With time, the deformations are fixed by structural changes. Vertebrae can become wedge-shaped and this is usually regarded as the crucial step in the formation of structural scoliosis. As scoliosis develops most frequently in adolescents during rapid growth spurts, the role of primary bone growth is regarded as responsible
for the abnormalities. While vertebrae of an adult change shape only with bone remodeling, in a child they grow in length. Growth takes place with ossification of cartilage in growth plates under the articulating surfaces. In the twisted spine unevenly loaded cartilage is compressed on the concave side and stretched on the convex side. So the bone grows faster on the convex side of the vertebrae which become wedge shaped [6]. But wedge-shaped vertebrae are not always present, in many cases cartilaginous intervertebral discs are transformed more (Figure 2). The curvature usually progresses slowly also in adulthood. Linear rate of progression at about one Cobb degree per year had been demonstrated in progressive adult scoliosis [7]. Scoliosis can also appear anew in adulthood, when only bone remodeling can be blamed for it.

In scoliosis vertebrae are not the only bones that change its form. Ribs are even more markedly transformed. Ribs on the convex side are pushed posteriorly and thoracic cage is narrowed by more strongly curved ribs which form a rib hump. Ribs on the concave side are pushed laterally and anteriorly. There are also other changes of the vertebrae besides wedging. Vertebral body is distorted toward the convex side, spinous process deviated to the concave side, vertebral canal is narrower on the convex side and lamina thinner. All these changes can be attributed to bone remodeling which is very active process during growth. Without it, the growing bones would be too heavy [8]. Bone remodeling slows down with adulthood, but never completely ceases.

The transformation of vertebrae and ribs in the process of bone remodeling is regulated by several hormones. One of them is melatonin, secreted by the pineal gland at night. In chickens and rats with destroyed pineal gland scoliosis developed, but administration of the melatonin prevented that [9]. It was suggested that lack of melatonin could be the cause of idiopathic scoliosis in humans also, but such a shortage was usually not detected in scoliotic patients [10]. Melatonin receptors can be impaired [11]. Melatonin suppresses bone remodeling by inhibition of bone resorption [12]. When there is a shortage of melatonin, bone remodeling accelerates.

Ishida et al. [13] found an increased amount of bone resorption marker in the majority of patients with adolescent idiopathic scoliosis, while the bone formation marker was at a normal level. Thus, in them bones degrade faster than regenerate. This decreases the strength of the bone and can lead to osteoporosis, known to cause scoliosis in the elderly.

Goldberg & Dowling [14] found statistically significant correlation between scoliosis configuration and handedness in 254 girls with idiopathic scoliosis. The curve pattern matched handedness in 82 percent. Of 228 right-handed children, 197 had a right convex curve pattern; of 26 left-handed children, 12 had a left convex pattern. Scoliosis without an obvious cause occurs only in humans. The same is true also of handedness: lateralization has not evolved to a similar degree in any other vertebrate. The influence of handedness on the curvature may be mediated through posture. When a child sits and writes with her right hand, she often bends to the left. Bones are loaded asymmetrically and scoliosis develops in children with rapid bone resorption and remodeling.
Fig. 2: Antero-posterior x-ray image of a scoliotic spine. If the curvature of very severe scoliosis resulted from uneven bone growth, vertebrae would be wedge-shaped. But here only intervertebral discs, which are made of cartilage, are transformed. They are stretched like bellows of an accordion.

**Progression**

Progression of idiopathic scoliosis is usually explained as uneven growth of vertebrae in length because of asymmetrical loading which brings more uneven load to the vertebrae and more asymmetrical growth in a vicious circle. But scoliosis can progress very rapidly in some cases, from 25 degrees Cobb angle to 45 degrees in just six months. Such a rapid progression of the curvature can not be attributed only to additional wedging of the vertebrae. Intervertebral discs may be deformed much more rapidly under constant pressure. The role of the ribs is underestimated again. The
importance of equal support of the spine through the ribs from both sides had been proved with experiments. Resection of posterior ends of ribs on one side induced progressive scoliosis in young animals. The spine curved to the side where heads and necks of the ribs had been removed [15].

When rotation of the rib cage and vertebrae happens, vertebrae lose balanced support from the ribs. Ribs on the concave side push only vertebral bodies toward the convex side and ribs on the convex side direct all their force to the vertebral processes. Shear forces cause additional turning of the vertebrae and bending of the spine toward the convex side. With gradual bone remodeling under constant pressure typical changes in the shape of the ribs and vertebrae evolve. Vertebral bodies are pushed closer to the rib arc on the convex side, narrowing the chest (Figure 3).

![Diagram showing forces transmitted by the ribs turning vertebrae and bending the spine sideways.](image)

**Fig. 3:** In a rotated chest forces transmitted by the ribs turn vertebrae and bend the spine sideways.

The importance of rib symmetry for infantile scoliosis resolution or progression was found by Mehta [16]. The difference between left and right rib-vertebra angle at the apex of the curve enabled him to distinguish between resolving and progressive infantile scoliosis. If the difference was less than 20° the spontaneous resolution of scoliosis appeared in 80 percent of cases, if greater than 20°, 80 percent of scoliosis cases progressed.

**Conclusions**

Side curvature of the spine is always accompanied by rotation of the rib cage because ribs are connected by intercostal muscles and cannot spread apart. The chest behaves like a flexible cylinder. When the spine bends, it rotates on its axis.

Bone remodeling is probably more important than primary bone growth in fixing the structural scoliosis. Both are active during growth spurts, but only remodeling proceeds into adulthood when scoliosis may progress further or starts anew. Children with progressive scoliosis may be genetically predisposed for more rapid bone resorption which is regulated by several hormones and their receptors.
When rib cage rotates, the shear forces transmitted by the ribs to the vertebrae turn vertebrae further and ribs on the concave side push vertebral bodies toward convexity, bending the spine. We can prevent the continuous deterioration with a brace, but braces should act against the rotating forces. If only side push is used to straighten the spine, rotation will increase and additional bending of the spine will follow.

**Competing interests**

The author declares that he has no competing interests.

**References**