What is “congenital scoliosis”?

Congenital scoliosis is a spinal deformity with lateral deviation and rotation of the spinal column, where congenital dysfunctions in embryonal vertebra development cause one or more malformed vertebrae. The vertebrae are incompletely formed, leading to asymmetrical spinal column growth. Congenitally anomalous vertebrae may occur in any part of the spine.
Normal spinal growth is disrupted by such formation defects, segmentation defects or combined forms of vertebral anomalies. As the spine continues to develop, this may lead to the developmentof scoliosis.
Congenital scolioses are rare, but they may require early surgery due to the severity of the spinal deformity involved.

How are the vertebra formed in embryonal development?

Weeks 4 to 8 of a pregnancy are called the embryonal period, during which the organs develop out of the three germ layers (organogenesis).
The three germ layers are:
The outer germ layer (ectoderm) out of which the spinal cord, nervous system, brain, skin, and hair develop.
The inner germ layer (entoderm) out of which the digestive tract, liver, pancreas, urinary bladder and urethra, thyroid gland, and respiratory tract develop.
The middle or 3rd germ layer (mesoderm) out of which the spinal column, ribs, and muscles develop.
The somites (primitive vertebrae) are formed by cell migration along the midline (paraaxial) of the mesoderm, where at first 42-44 somite pairs are formed next to the notochord and the neural tube. The individual somites form segmentation furrows that are visible in the embryo.
The somites continue to differentiate, whereby three developmental structures are formed from the somite wall: The dermatome forms the skin and subcutaneous tissues, the myotome forms the striated muscles of the arms, legs and torso and the sclerotome forms the spinal column and ribs. There are more primary (provertebral) segments than there are fully developed vertebrae. As embryonal development continues, 10 somites cease developing and the final number of 32-33 fully developed vertebrae is established.
Between the 7th and 10th week of pregnancy, the vertebrae are formed when the sclerotomes divide down the middle into front and back sections, whereupon each vertebra is formed by the unification of two adjacent half-somites (provertebral segments). The place where they are joined later becomes the intervertebral space. The intervertebral discs develop from the front somite half, which has fewer cells. This reorganization of the body’s axis is known as resegmentation.
If malformations occur during the fusion of the half-somites, this may result in the formation of anomalous vertebral bodies that manifest as formation defects or segmentation defects of the vertebral bodies.

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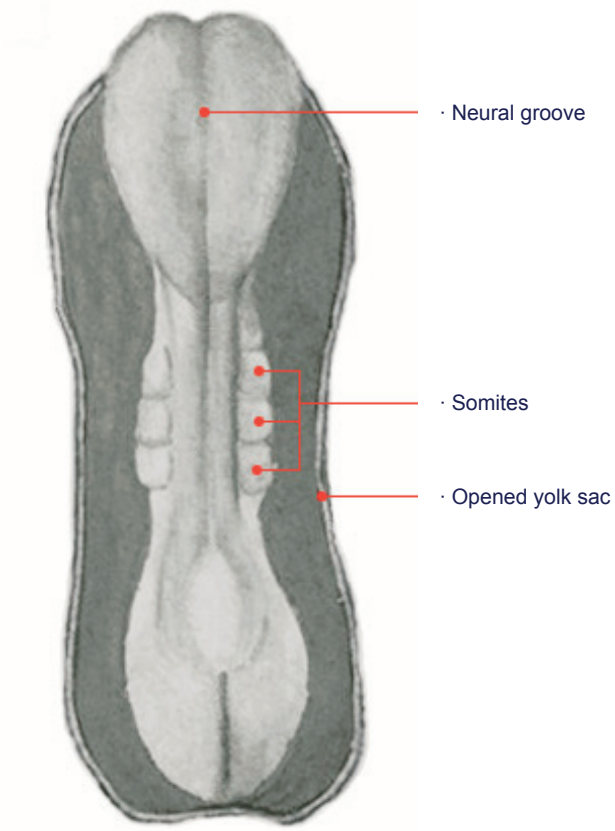
• Three formed somite pairs in a human embryo in about the 3rd to 4th week of pregnancy (from the rear, yolk sac opened)



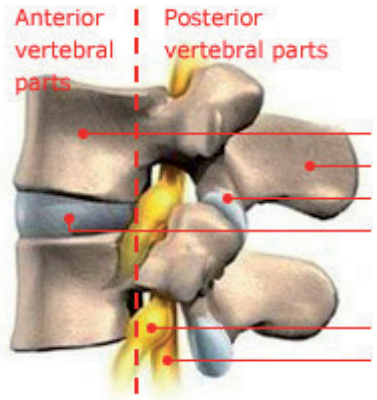
What forms of congenital vertebral anomalies are there?

There are three types: formation defects, segmentation defects and a combined form of these vertebral defects.

Segmentation defects

A segmentation defect is when the intervertebral disc space is formed incompletely or not at all and the growth joint of the vertebra is lacking.
If an entire intervertebral disc space is missing, this anomaly is called a block vertebra. If the vertebra is not segmented in certain areas only, this results in bar formation, which may be dorsal (back), lateral (on the side) or posterolateral (on the back and side).

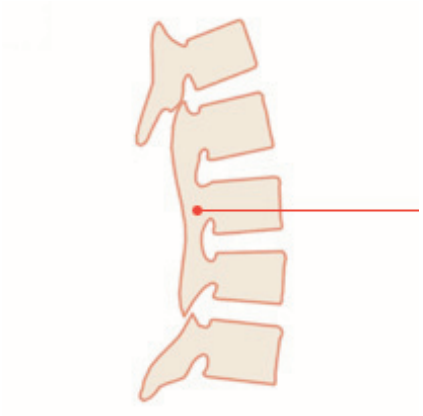
• Normally formed vertebral segment



The following types of segmentation defects are defined::

· Dorsal (posterior) segmentation defect

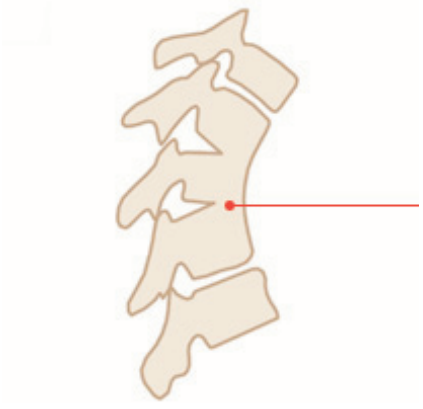
• Posterior segmentation defect, spinal column from the side



In a posterior segmentation defect, the vertebral arches and vertebral joints are not formed separated from one another, leading to the potential fusion of several vertebrae in their posterior sections. More pronounced growth of the anterior portions of the vertebrae may result in increased lordosis (hollow back).

· Ventral (anterior) segmentation defect

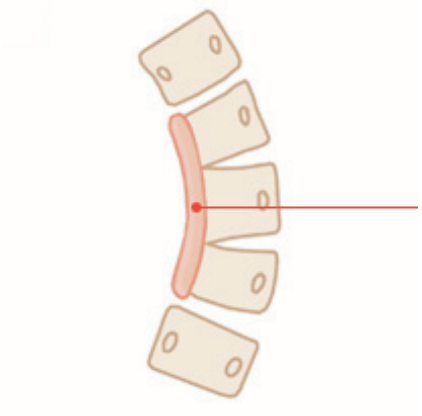
• Ventral segmentation defect, spinal column from the side



In a ventral (anterior) segmentation defect, the vertebral bodies of several vertebrae without the formation of intervertebral discs are fused. More pronounced growth of the posterior portions of the vertebrae results in development of a kyphosis (humpback).

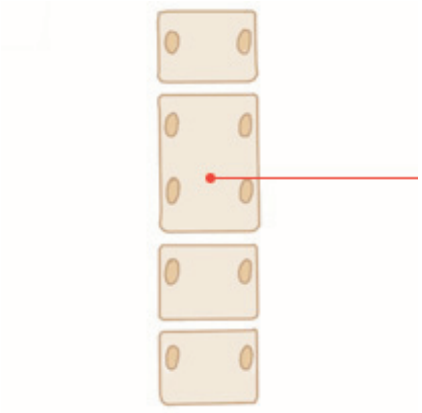
· Lateral (side) segmentation defect

• Lateral segmentation defect, spinal column from the front



Lateral segmentation defects are caused by an anomaly in the pedicles, resulting in a lateral fusing of several vertebrae with the formation of a lateral bony rod. Greater growth on the non-fused opposite side then results in the formation of scoliosis.

· Block vertebra formation



A complete segmentation defect in two vertebrae results in block vertebra formation, which will at most cause a reduction in length, but not a malposition of the spinal column.

Combined genetic malformations

Segmentation defects and formation defects may occur in combination with various vertebral anomalies or fused rib primordia (rib synostoses). Congenital scoliosis frequently occurs together with congenital anomalies of other organ systems (heart, gastrointestinal tract, kidneys, efferent urinary tract), which is why the organs should be carefully examined for malformations if anomalies are present in the vertebrae.

What are the possible courses of congenital scoliosis?

The spontaneous course of congenital scoliosis is variable and depends on the extent of the anomaly. Some anomalies, such as incarcerated or balanced hemivertebrae, have a favorable prognosis, unlike rapidly progressive forms such as hemivertebrae with contralateral bar formation. The formation of 2 unilateral hemivertebrae generally leads to a pronounced curvature requiring early intervention. A complete unilateral formation defect results in a hemivertebra, one of the commonest causes of congenital scoliosis. With the exception of a few incarcerated forms, a hemivertebra has a nearly normal growth potential, so that it forms a wedge-shaped deformity that grows worse over time. Both the dimension and the rigidity of the major curve increase with age. Minor countercurves develop in the originally healthy adjacent spinal column sections, which also rigidify with age, sometimes causing more problems than the wedge vertebrae themselves.

Simple hemivertebrae, according to McMaster and Ohtsuka, cause a progression of spinal column curvature of 1-3.5° per year. The course is least favorable if the wedge vertebrae are located in the lower thoracic spine and at the thoracolumbar transition. Hemivertebrae in combination with contralateral bar formation show a progression rate of 5-10° and more per year, clearly demonstrating the dramatic nature of this malformation. If the situation is unfavorable, the congenital scolioses may also be combined with a progressive kyphosis, which has a very unfavorable prognosis. In particular, this deformity may also result in myelopathy (spinal cord damage) with worsening neurological problems as the patient ages.

How are congenital scolioses treated?

Conservative therapy is not promising since the asymmetrical growth patterns cannot be influenced by brace therapy. The only possible therapeutic objective here would be to influence the developing minor countercurve. In view of the unfavorable spontaneous course, confirmed or expected progression represents an indication for surgery.

The surgical methods described to date are:
· In situ fusion
• Convex-side hemiepiphysiodesis
• Convex-side hemiarthrodesis
• Growth-guidance surgical methods
• Instrumentation with Zielke-Askani growing rod
• Vepter instrumentation

In addition to the surgical methods listed above, a further alternative is hemivertebral resection with fusion (removal of hemivertebra followed by surgical spondylodesis) of the affected spinal section.