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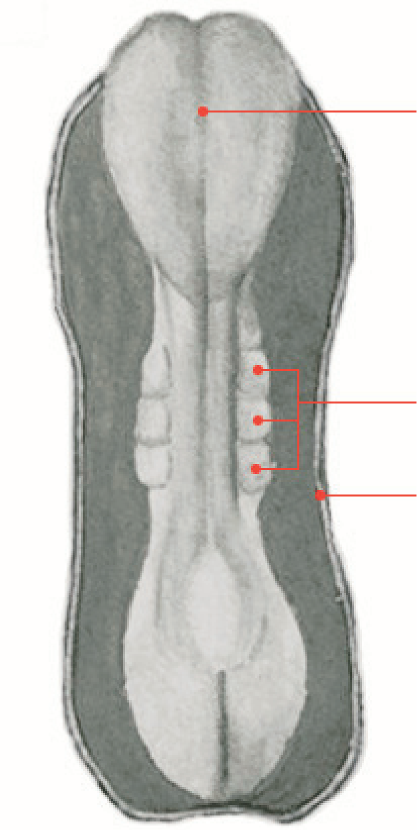
• Three formed somite pairs in a human embryo in about the 3rd to 4th week of pregnancy (from the rear, yolk sac opened)



What forms of congenital vertebral anomalies are there?

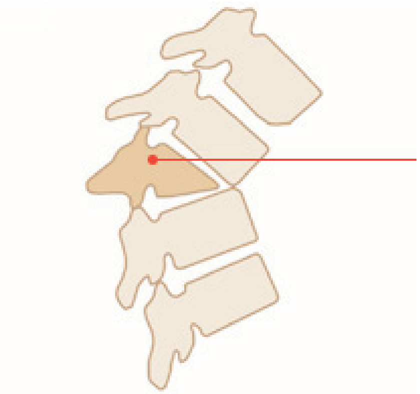
There are three types: formation defects, segmentation defects and a combined form of these vertebral defects.

Formation defect

In a formation defect, a vertebral body is incompletely formed.
This occurs in different forms:

• Anterior formation defect (wedge vertebra)

• Wedge vertebra (sagittal view)

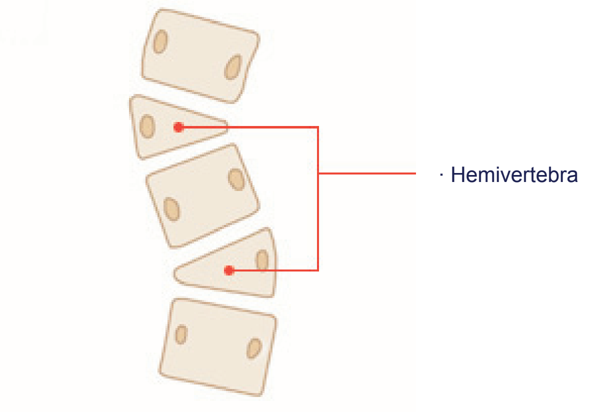


• Normal mid-thoracic spine, seen from the side

A wedge vertebra results from dysplastic (faulty) formation of a vertebral body on one side where the pedicles are still present.

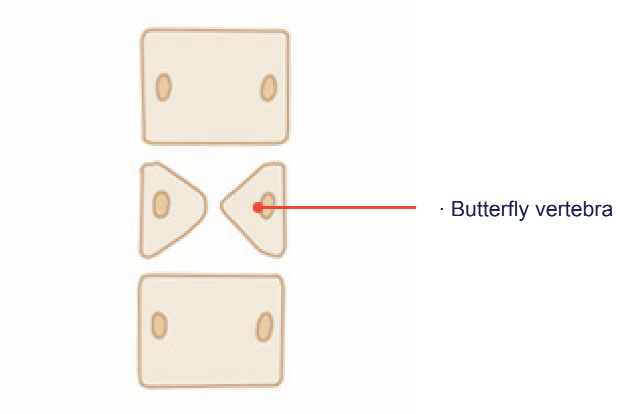
· Lateral formation defect (hemivertebra)

• Vertebral section with two hemivertebrae, compensated



Lateral formation defects result in hemivertebrae with dysplastic formation of one side of the vertebra. Hemivertebrae occur in various forms. On one side of the vertebra the pedicles and vertebral joints may not be formed at all, or the vertebral contour may show just a slight inclination.
Hemivertebrae may occur singly or there may be more than one and they can lead to a deformity of the spinal column in the frontal and sagittal profiles. In cases where two hemivertebrae are present and face in opposing directions above and below an intact formed vertebra, the two hemivertebrae may compensate the tendency of the spinal column to assume a pathological curvature (see illustration).

· Central formation defect



A central vertebral body defect results in the formation of a butterfly vertebra. The existing parts of the vertebra may show growth joints or they may be fused with the vertebral bodies of the adjacent vertebral segment. Butterfly vertebrae of normal height do not necessarily result in a deviation of the spinal column axis.

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.

Resection of a hemivertebra was first described by ROYL in 1928, but the method did not establish itself until much later. In 1979, LEATHERMAN was the first to report good results with a number of hemivertebral resections in two-session (dorsal, then ventral) operations. Correction after resection of the hemivertebra was achieved with a plaster or plastic brace worn over a period of about 6 months. Alternatively, the gap resulting from resection of the hemivertebra was closed using compression instrumentation (Harrington) hooks.

The disadvantages of combined wedge vertebra resection acc. to LEATHERMAN are as follows:

· Necessity of a second ventral surgical session with repeated anesthesia
· Correction in brace with long-term immobilization – difficult to realize in the thoracic region in particular
· Compression with hook instrumentation requires stable dorsal bony structures, making its use highly questionable and conditional in small children.

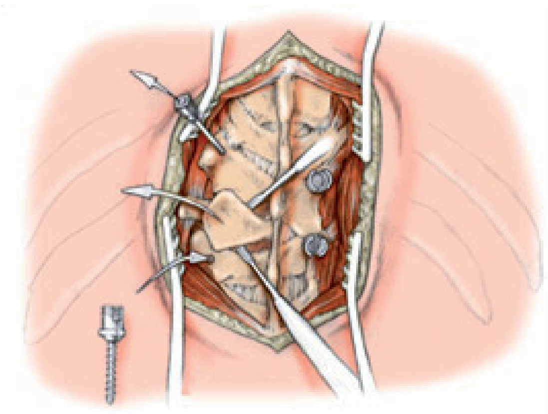
We developed a wedge vertebra resection technique designed to avoid the pitfalls of this method. Our method requires dorsal access only. It was carried out for the first time in 1991 and is characterized by two basic principles:

· Complete resection of the hemivertebra via dorsal access only
· Correction and stabilization by means of short-length and semirigid transpedicular instrumentation.

Surgical technique:

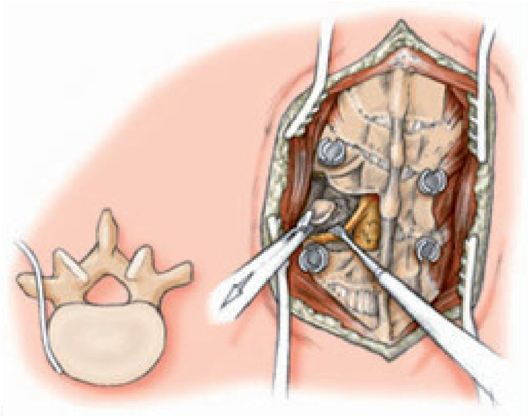
In dorsal access (from behind) the hemivertebra and adjacent vertebra are exposed including the small intervertebral joints. Preparation of the costal facet joints is also necessary in the thoracic spine. The entry points for the pedicle screws in the adjacent vertebrae are first marked with needles. The position and direction of the needles are then checked via the image converter (fig. 1).

fig. 1



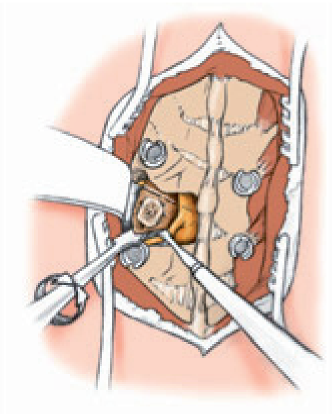
The pedicle screws are inserted following drilling and threading. The dorsal portions of the hemivertebra are resected (removed) including the lamina, joint facets, transverse process, dorsal portions of the pedicles. The dural sac and nerve roots above and below the pedicle of the hemivertebra are exposed (fig. 2).

fig. 2



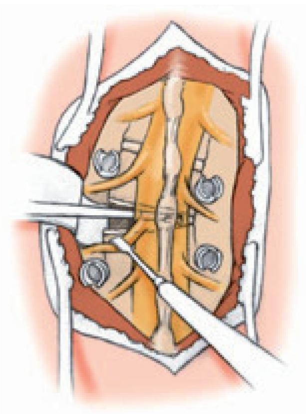
Following resection of the pedicles, the lateral portion of the hemivertebra is exposed: in the lumbar spine this requires resection of the transverse process, and in the thoracic spine the extra caput costae (posterior ends of the ribs) must be resected to expose the lateral wall of the vertebral body properly. Preparation of the lateral wall and anterior wall of the vertebral body is blunt, and can be done either below or outside the periosteum. It is important to intoduce a blunt spatula at this point to protect the vessels lying ventrally in front of the hemivertebra (fig. 3).

fig. 3



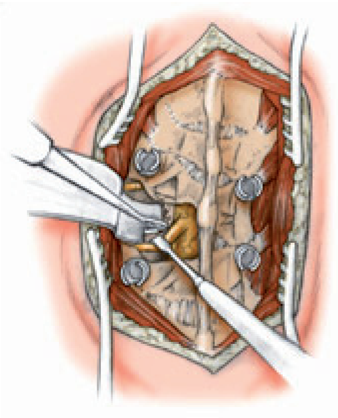
While protecting the dural sac (surrounding the spinal cord) and nerve roots, the intervertebral discs adjacent to the hemivertebra are then incised and removed. The remainder of the pedicle, together with the hemivertebra, is then mobilized and removed (fig. 4).

fig. 4



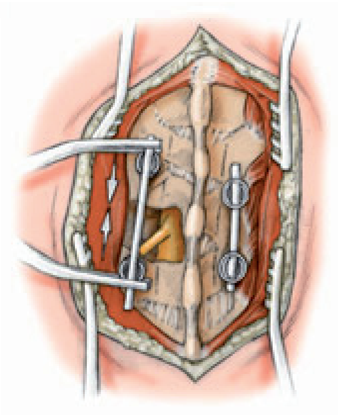
Then the intervertebral disc on the concave side connecting the two vertebral bodies on the cranial and caudal side of the hemivertebra is removed. The upper plates of the adjacent vertebra are then freshened (fig. 5).

fig. 5

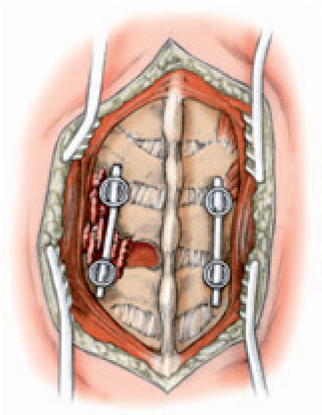


Now the longitudinal supports are inserted on the convex side with compression via the convexity, completely closing the gap resulting from the resection of the hemivertebra. The freshened endplates of the adjacent vertebral bodies move close together. The bone material from the hemivertebra is packed in to facilitate rapid bony fusion. Dynamic stabilization is also carried out in the concavity (figs. 6 and 7).

figs. 6



figs. 7



In the presence of an additional pronounced kyphosis, a titanium cage can be inserted as a ventral support into the intervertebral disc space. This cage then acts as a support or pivotal point for dorsal compression to achieve segmental lordosis. This also prevents the spinal cord from being shortened excessively.
For single hemivertebrae with no other anomalies, it usually suffices to fuse only the two vertebrae adjacent to the hemivertebra. For more pronounced structural changes in the adjacent vertebrae or a greater degree of kyphosis, additional segments can be temporarily involved in the instrumentation. In cases of contralateral bar formation and rib synostoses, the bar is severed and the concave-side capiti costae are resected. The instrumentation must be done over the entire length of the bar.

Follow-up treatment:

As a rule, patients can leave their beds on the first day after the operation. Depending on the stability of the instrumentation and the length of the fusion, a brace (2-shell orthotic device or Stagnara brace) is fitted and worn for about 12 weeks.

Assessment of the results

Correction of the major curve:

The preoperative segmental angle of the major curve averaged 37.6° (16-66°). This was corrected postoperatively to 8.7° (-1° to 29°) and amounted to 6.2° (-5° to 30°) at the last follow-up examination. This translates into an average correction of 31.4° or 84% (fig. 8).
The overall preoperative angle of the major curve averaged 45.9° (16° to 109°), corrected postoperatively to 11.9° (-1° to 45°) and 9.9° (-5° to 55°) at the last follow-up examination. This corresponds to an average correction of 36° or 78%.

Correction of the minor curves:

The correction of the minor curve is highly satisfactory. For the contralateral minor curve following cranially, a spontaneous correction of 80° was achieved in most cases, as was a spontaneous correction of 75° for the minor curve following caudally.

Sagittal plane correction:

The sagittal profile was also preserved or normalized in most cases.

Complications:

There were no cases of neurological complications. An implant rupture is a possibility since this is a dynamic instrumentation within an overall dynamic system. In a small number of patients, a scoliosis developed again in the course of further growth, requiring surgical correction once more.

Summary:

The overall results were highly satisfactory, and the number of cases requiring follow-up surgery can also be viewed as absolutely acceptable. It is important to remember that the patients were very young. The average age of our patients was 3.5 years (15 months to 6 years).
This clearly documents the considerable growth potential of the spinal column at the time of surgery. Reliable predictions of the effects on fusion within the growing skeleton are never possible. On the whole, however, we were amazed at how small the influence of growth on the fused segment turned out to be.