What is Idiopathic scoliosis?

80-90% of all scolioses are idiopathic, the rest are neuromuscular or congenital scolioses with manifest primary diseases responsible for the scoliotic pathogenesis. This condition affects girls four times as frequently as it does boys.

The term “idiopathic” means the condition develops "by itself." In the final analysis, the causes are not known (Greek: idios = self, pathos = suffering, disease).

Idiopathic scoliosis is a three-dimensional spinal deformity, the pathogenesis of which remains unknown to date. The three-dimensional quality of scoliosis refers fact that the changes it brings about occur in the three planes of the body, the frontal, sagittal, and transverse planes. Lateral deviation is observed in the frontal plane. An additional change is observed in the sagittal plane, which may alter the typical physiological lateral contours of the spinal column curvatures (kyphosis and lordosis). Scolioses of the thoracic spine frequently feature a flattening of the sagittal profile with the formation of a thoracic flat back. Scolioses in the transitional thoracolumbar spine region often feature a kyphosing (humpback) in the area. The deformation of the spinal column leads to structural changes in the mobile segments with changes in the vertebrae, vertebral joints, intervertebral discs and ligamentous apparatus, that begin suddenly and progress rapidly and which are caused by the altered static load on the segments.

What factors are considered to be causative for idiopathic scoliosis?

- **Genetic factors**
  
  Studies have revealed high interfamilial incidence patterns for scoliosis.

- **Growth acceleration**
  
  The spinal column may show some instability in periods of accelerated growth and is therefore, in combination with postural deficiencies, more susceptible to the formation of malpositions.

- **Posture**
  
  In comparative studies involving scoliosis patients and patients with normal spinal column findings, it has been demonstrated that the group of scoliosis patients also shows a much higher incidence of persons with a proprioceptive dysfunction. Proprioceptive functions include the sense of depth awareness, for example of muscle tonus. Disturbances in this function may lead to alterations in posture and thus to development of malpositions.

- **Mechanical factors**
  
  Biomechanical studies have demonstrated that a movement of the spinal column in the direction of one of the body’s axes causes a countermovement along one of the other axes. This phenomenon is called the “coupling effect.” This effect and the varying rotation of the vertebrae in a lordotic or kyphotic spinal column segment may, under the influence of an axial force (for example during a period of accelerated growth) result in a lateral deviation of the spinal column with rotation of the vertebrae.

How are idiopathic scolioses classified?

There is no internationally uniform classification of idiopathic scoliosis. In addition to many other group definitions, classification methods used are based on the age of first onset of the scoliosis or on the form and curvature type involved in the scoliosis.

The following classifications are differentiated on the basis of age at first diagnosis:

- **Infantile scoliosis**
  
  This type first occurs up to the age of 3, is rare, and typically involves scolioses of the thoracic spine.
Idiopathic scoliosis · Scoliosis · Deformities

- Juvenile scoliosis
  
  The juvenile type first occurs between the ages of 3 and 9 and features both thoracic and lumbar scolioses. Incidence in girls and boys is about equal.

- Adolescent scoliosis
  
  This type first occurs at the age of 10, is the most frequent form, and affects girls about four times as often as boys, often in the thoracic spine.

- Adult scoliosis types that first occur in adulthood
  
  With reference to the curvature type, scoliosis forms are differentiated by their occurrence in the cervical, thoracic or lumbar spine and in the transition sections occipitocervical, cervicothoracic, thoracolumbar and lumbosacral.

What are the most important aspects when considering the need for surgery?

A decision to perform surgical treatment on a scoliosis depends on a number of factors:

- Worsening of scoliotic curvature (progression)
- Existing unfavorable sagittal profile
- Avoidance of structural changes in compensatory curve
- Pain
- Avoidance of secondary complications (cardiovascular system and lungs)
- Factors such as patient age, Cobb angle, individual handicaps, and level of suffering also figure into the decision making process.

The following criteria should be considered when selecting a surgical technique:

- Flexibility of the major and secondary curves (rigidity)
- Sagittal profile
- Stable zone
- Determination of the terminal vertebra for instrumentation

What are the objectives of the surgical correction of scoliosis?

The objective is the three-dimensional restoration of the spinal column:

- In the frontal plane, distraction must be used to straighten the scoliosis from the lateral curve deviation into a plumbline-straight position.
- In the transverse plane, derotation is done to eliminate the rotational and torsional malposition.
- The profile of the spinal column must be restored in the sagittal plane.
- The instrumentation should ensure primary stability so that follow-up treatment with a brace is not required. This is feasible by means of dorsal instrumentation with pedicle screws and double-rod instrumentation. In ventral procedures, the necessity of follow-up brace therapy depends on the bone quality. If the bone quality is poor, brace therapy for two or three months post surgery may be required. If the bone is intact, ventral surgical procedures with cage support on the ventral side will usually not necessitate postoperative use of a back brace.
- The fusion length (i.e. of the spondylodesis) must be kept as short as possible to ensure as good residual mobility of the spinal column as is permitted by the individual situation and findings.
Idiopathic scoliosis · Scoliosis · Deformities

Three-dimensional correction can be achieved by either ventral or dorsal instrumentation.

a. **Dorsal instrumentation**

Dorsal instrumentation, which is currently generally used with pedicle screws in both the thoracic and lumbar spine, can achieve a favorable correction in the frontal plane once the joints have been properly mobilized. Influence on the sagittal profile is limited. For this reason, a ventral release operation is required in cases of prior pronounced thoracic lordosis. It is also difficult to influence the axial rotation plane by way of dorsal rod rotation, since rod rotation does not rotate the spinal column itself. What is involved here is rather a translation of the instrumented spinal column section in the transverse plane.

b. **Ventral instrumentation**

The ventral instrumentation approach goes back to Dwyer and Zielke. Its main advantage is that the intervertebral discs are removed, making it possible to effect direct segmental rotation between the vertebral bodies as well, which is not possible using dorsal instrumentation (see above). At the same time, it must be kept in mind that, in ventral instrumentation, the removal of the intervertebral discs shortens the anterior column, so that ventral instrumentation always has a kyphosing effect, which can be put to good use for thoracic scolioses, which are frequently accompanied by a thoracic lordosis. Therefore, in modern surgical procedures, thoracic scoliosis combined with apical lordosis result in the ideal indication for ventral instrumentation.

On the other hand, lumbar and thoracolumbar curves are also well-suited for ventral instrumentation. In these cases, however, the procedure must always be combined with ventral support between the vertebral bodies to avoid the kyphosing effect of ventral instrumentation (see above).

c. **Combined access**

In highly rigid scolioses and in cases with double curves in particular, a ventral release operation in the thoracic or lumbar spine is often required, that is then followed by dorsal instrumentation. After the ventral release procedure, the axial profile and rotation can be readily influenced because the intervertebral disc has been removed, making direct rotation possible, a process that cannot be done unless the disc has been removed.

In addition to correction in the frontal plane, an important objective of surgery must be the restoration of a very good sagittal profile. Another important object is favorable horizontal positioning of the vertebral body that has just been instrumented. This means the vertebral body last instrumented in the caudal direction should be positioned horizontally in relation to the sacrum.

The big advantage of dorsal instrumentation is that postoperative treatment can normally be done without a brace. Depending on bone quality, 2-3 months of brace support are required post surgery with ventral instrumentation.
Idiopathic scoliosis · Scoliosis · Deformities

What different forms occur and how can they be surgically corrected?

First of all, it is necessary to differentiate between thoracic and lumbar scolioses. Then there is the difference between single curve and double curve scolioses. The double curve scolioses usually occur in the thoracic and lumbar regions together. A large single curve scoliosis in the shape of a big C is less frequently seen in idiopathic scoliosis, but is frequent in neuromuscular scoliosis.

Thoracic curves
Thoracic scolioses are scolioses with the major curve in the thoracic spine and a compensatory minor curve in the lumbar spine. An additional thoracic minor curve has to be included in instrumentation, since otherwise a dysbalance may develop despite effective correction of the major curve.

Thoracic scolioses are characterized in particular by a change in the sagittal profile such that a tendency to form a hypokyphosis (flat back) up to a lordosis (sway back) is in evidence. This sagittal malposition is considered by Dickson, for example, to be a primary cause of scoliosis.

Thoracic scolioses can be corrected by surgical approaches with a ventral (front), dorsal (back) or combined access route to the spinal column.

Surgical methods using ventral access
Most thoracic scolioses of Lenke types 1A, 1B, 1C and 3C can usually be corrected quite effectively in three dimensions by a ventral access operation. The advantage of ventral access is that the anterior column is always shortened by the removal of the intervertebral discs, automatically translating a hypokyphotic-lordotic malposition in the sagittal profile into normal kyphosis.

Surgical methods using dorsal access
A dorsal access operation should be considered if, in addition to the thoracic major curve, an additional rigid upper thoracic minor curve is also present. If this upper thoracic minor curve is not taken into consideration, decompensation may result since spontaneous correction of this upper thoracic minor curve will not be sufficient.

An operation with sole dorsal access is the method of choice under the following conditions:
• Good mobility of the major curve, easily determined by the bending test
• No pronounced thoracic hypokyphosis-lordosis.

Surgical methods using combined ventral and dorsal access
Combined methods are always used when the major curve is very rigid and the sagittal profile of the spinal column is unfavorable. This approach is also taken in the presence of a rigid major curve and an additional upper thoracic rigid minor curve. In these cases, the rigidity (stiffness) of the spinal column can be improved by a ventral release procedure. Dorsal instrumentation can then be carried out at the same time or in a second operation, resulting very good three-dimensional corrections. The sagittal profile of the spinal column is an important factor in the decision whether to do a ventral release operation with or without ventral instrumentation.

In the presence of a pronounced thoracic lordosis, a ventral release is always required to ensure sufficient rekyphosing of the thoracic spine.

In highly rigid scolioses, preoperative extension treatment with halo extension can be applied, where a specially attached system is used to exert permanent tensile force in the direction of the longitudinal axis of the spinal column so as to loosen the stiffened curve prior to surgery.

This additional procedure should be used for the treatment of very rigid scolioses with a Cobb angle exceeding 70°. In massively ankylosed scolioses, it is sometimes necessary to begin with a halo extension procedure lasting 2 weeks, followed by dorsal instrumentation. Very good corrective results can be achieved, even in cases of highly rigidified scolioses, with this rather complex and time-consuming procedure.
Thoracolumbar and lumbar curves
These curve types can be treated ideally via ventral surgical access. Dwyer and Zielke were mainly responsible for developing this surgical approach in the 1970s. Following the introduction of the CD instruments by Cotrel-Dubousset, the method initially had a bad reputation because, despite the very good corrective results obtained in the frontal plane in terms of straightening the spinal column, the sagittal column profile was often left even worse than before, frequently due to insufficient correction.

The changes in the ventral surgical approach suggested by Harms, in particular the use of structural ventral support materials (cages), have enabled surgeons to completely avoid the drawbacks of this approach, i.e. formation of a lumbar kyphosis.

Therefore today, for most thoracolumbar or lumbar curves, ventral lordosing-derotating spondylodesis with good ventral support between the vertebral bodies can be considered the therapy of choice.

In the presence of very rigid thoracolumbar and lumbar scolioses with a pronounced thoracolumbar kyphosis, a combined access approach may be necessary for these curve types as well. By employing ventral release and dorsal instrumentation with a double-rod system, very good correctional results can be obtained, even with such findings, and follow-up treatment with a brace is often avoidable. This technique is frequently used in older patients.

Double major curve
This curve type features double curves, normally a right convex thoracic and a left convex lumbar curve. The problem with this type is that the scoliosis is often not recognized until later in its development because the counter curves compensate each other effectively, so that clinical abnormalities manifest late. In the double major curve, the sagittal profile is usually not changed as unfavorably as in the thoracic or lumbar scolioses.

Earlier, surgical treatment of this curve type was approached with reservations. It has now been demonstrated that pronounced lumbar curves, which are hardly problematical in younger years, may over the course of their progression lead to early onset of degenerative changes in the pathologically stressed intervertebral discs, accompanied by pain. For this reason, surgery is also advisable when these double curves are present, and in particular double curves with a tendency to progress.

Both dorsal and ventral approaches can be used to treat double major curves. These double curves usually show good flexibility, making them suitable for dorsal instrumentation alone. The frontal and sagittal spinal column profile can be very effectively corrected with pedicle screws.

If bending tests carried out on a double curve show that one of the two curves cannot be sufficiently straightened, or if the sagittal profile is unfavorable, especially in the thoracic region, ventral instrumentation can also be used on both curves.

The ventral procedure then often makes it possible to shorten the fusion length towards the bottom. Not having to fuse one extra segment might be sufficient grounds to choose the ventral procedure with these findings, since each additional mobile segment in the lumbar spine can be of key importance to the overall function of the spinal column.