Congenital Scoliosis

12

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12.1 Definition

The term *congenital scoliosis* includes deformities of the spine in the coronal plane caused by malformations occurring within the early embryonic period. The formation of the somites (precursors of the spinal segments) starts between 3 and 5 weeks of gestation while segmentation occurs between 6 and 8 weeks of gestation. Failures of formation result in hemivertebrae which may be fully segmented with two adjacent disc spaces, semi-segmented, or non-segmented (incarcerated) (Fig. 12.1).

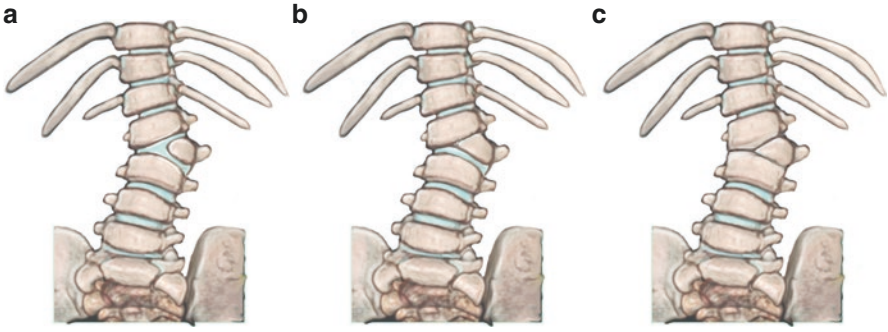

Fig. 12.1 Failures of formation: fully segmented (**a**), semi-segmented (**b**), non-segmented (incarcerated) (**c**)

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Segmentation defects include bar formations spanning one or more segments that may be located lateral, anterior, or posterior. Delay or cessation of growth within the bar usually results in asymmetric growth with progressive scoliosis, kyphosis, or lordosis (Fig. 12.2). Mixed deformities are frequent, often combined with abnormalities of the rib cage. Rib deformities, especially rib synostosis, may increase the scoliotic deformity of the thoracic spine (Fig. 12.3).

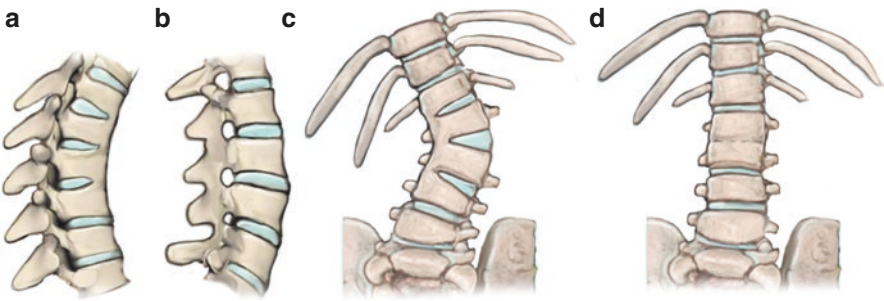

Fig. 12.2 Failures of segmentation: anterior bar (**a**), posterior bar (**b**), lateral bar (**c**), bloc vertebra (**d**)

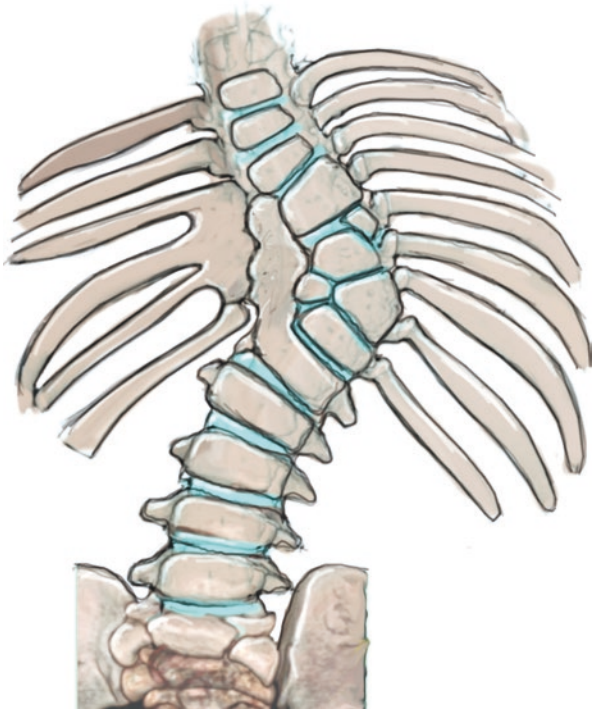

Fig. 12.3 Mixed deformity with hemivertebra, bar formations, and rib synostosis

12.2 Natural History

The natural history of congenital scoliosis depends on the type and the location of the congenital abnormality. Scoliosis progression may be expected during the growth spurt. The worst prognosis is usually seen in hemivertebrae with contralateral congenital bar (up to 14° of progression/year), followed by unilateral congenital bar, fully segmented hemivertebrae (1° to 5° of progression/year), and by non-segmented hemivertebrae which may remain stable [1].

The unaffected vertebrae adjacent to the congenital deformity are subject to asymmetric loading; in particular, asymmetric loading during growth can ultimately cause wedge-shaped deformity of the vertebral body. Over time, the stiffness of the main curve tends to increase. At the same time, secondary curves develop to promote trunk balance. These curves are flexible in the beginning but become structural with time.

12.3 Physical Examination

Physical examination includes documentation of

- Any deformity in the coronal plane.
- Pelvic/shoulder/head obliquity.
- Trunk balance/plumbline.
- Rib hump/lumbar prominence.
- Trunk height/ relation thoracic spine length - lumbar spine length.
- Any sagittal deformity.
- Flexibility of the curves/compensation mechanisms.

Neurological examination (Video 12.4) is mandatory although neurological compromise is rare; the examiner has to pay attention to pigment disorders, hairy patches, and other signs of dysraphism (Chap. 31). Associated malformations (cardiac, urogenital, gastro-intestinal; VACTERL syndrome) must be ruled out.

Special attention should be paid to congenital dislocation of the spine (rare malformation) secondary to a developmental failure of the spine and the spinal cord at a single spinal level (Chap. 24). The goal of treatment is to stabilize the spine in order to avoid secondary neurological damage (early cast immobilization and an early instrumented decompression-stabilization with circumferential fusion).

12.4 Imaging

Radiographs of the whole spine, anterior-posterior and lateral projection, in a standing position show the deformity secondary to the congenital malformation as well as compensatory curves (Video 12.6); radiographs are also needed to assess trunk balance and during follow-up. Bending films are obtained for preoperative planning, particularly to evaluate the flexibility of the secondary (compensatory) curves.

A computed tomography (CT) scan with three-dimensional reconstructions is important to evaluate the morphology and the location of the congenital hemivertebra as well as of the adjacent vertebrae and to provide information on bar formation or ribs synostosis. Three-dimensional reconstruction is helpful for surgical planning especially in mismatched abnormalities of the anterior and posterior structures; the size and diameter of the pedicles are measured to select proper implants.

Magnetic resonance imaging (MRI) is mandatory to evaluate the spinal cord and other soft tissue structures; malformations of the spinal cord must be detected or excluded.

The deformity is then classified according to the type and degree of the malformation.

12.5 Treatment Options

The goal of treatment is to achieve a straight spine with a physiological sagittal profile, with limited loss of motion, and limited growth deficit. To achieve these goals, early diagnosis and early surgical intervention in young children are needed due to the natural history of the disease: aging usually results in fixed, structural deformities (secondary curves), necessitating more aggressive surgical treatment usually resulting in longer instrumented fusions. If there is any doubt about further progression, especially in semi-segmented or incarcerated deformities, or hemivertebrae in the upper thoracic spine, frequent radiographic follow-up is mandatory.

In case of hemivertebra, resection of the hemivertebrae is the mainstay of treatment. Posterior resection with short instrumentation is the standard treatment option (Video 12.3). Surgery should be performed early before the adjacent vertebrae get deformed, and before secondary curves become structural.

Pedicle screws are inserted in the adjacent vertebrae (Videos 12.3 and 12.6). The hemivertebra and the adjacent discs are exposed and resected from the convex side. A cage may be inserted to ensure lordosis in the lumbar spine. In young children, the spine is highly flexible; thus, correction requires little force and is achieved by compression on the convexity via the instrumentation. With the early and complete correction of the focal deformity, the development of secondary changes can be avoided. Thus, the primarily healthy segments are allowed to grow physiologically: with short fusion, the overall growth deficit is minimal (Fig. 12.4) [2].

In the thoracic and lumbar spine, a posterior approach is usually sufficient. An additional anterior approach is required in the cervical spine, and it can also be performed at the lumbosacral junction in order to facilitate complete resection of the hemivertebra.

In patients with synostosis around the hemivertebra or congenital bar, especially older children or adults, an additional osteotomy at the concave side or a complete apical vertebrectomy is usually required; this procedure involves a complete disconnection of the spine [3].

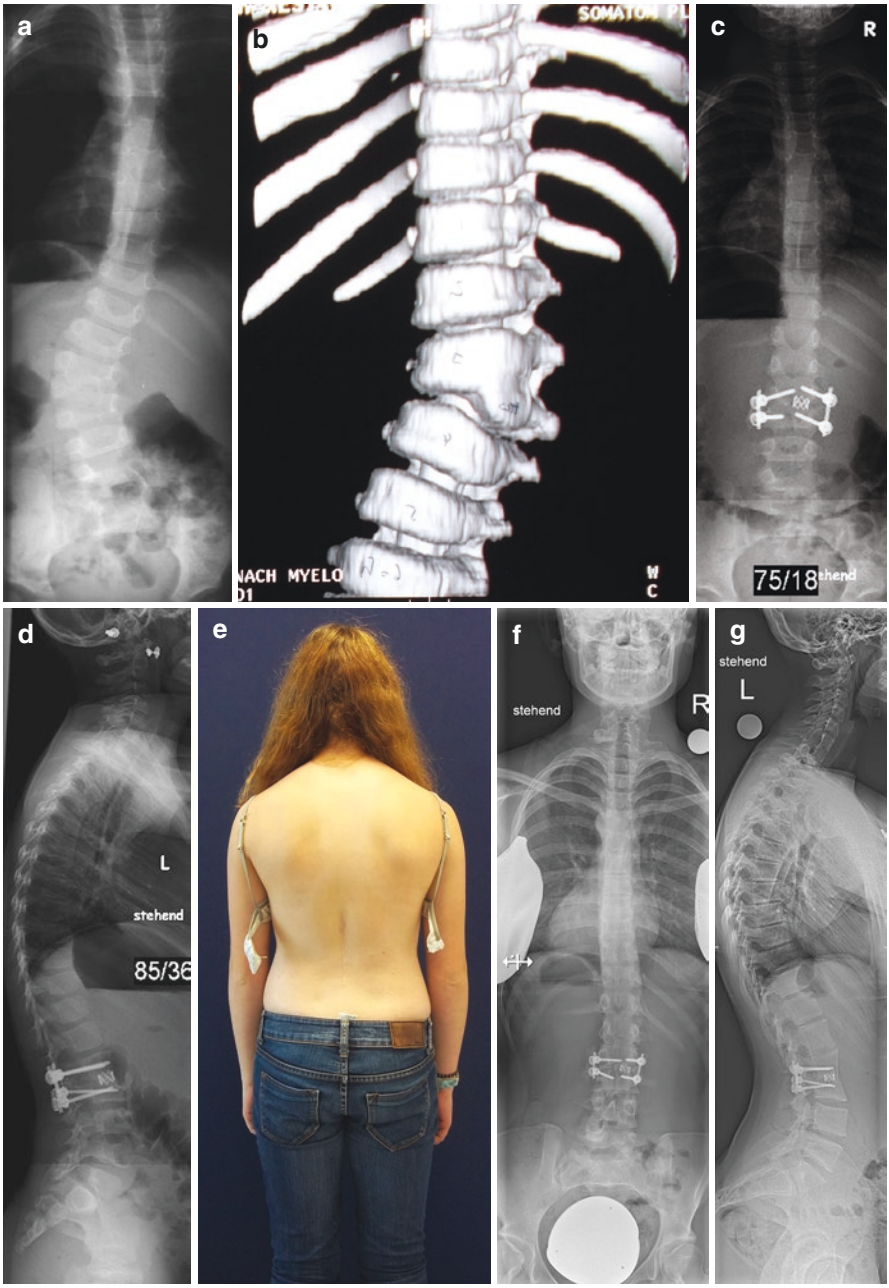

Fig. 12.4 A 4-year-old girl with semi-segmented hemivertebra L3a (**a**, **b**). Postoperative (**c**, **d**) at the age of 12 (**e**) and 17 (**f**, **g**) years

Multi-segmental deformities with a congenital bar in young children are often rapidly progressive. To avoid a long instrumented fusion potentially resulting in a short trunk, a combination of apical correction and "guided-growth" instrumentation of the adjacent segments may be useful.

12.6 Expected Outcomes

In patients with single hemivertebra, early resection with short fusion allows for complete and stable correction, and physiological growth. Complex deformity with congenital bar and rib synostosis may result in longer instrumented fusions and residual rib cage deformities. Depending on the primary growth plate abnormality, the trunk height may be short. Delayed treatment requires correction of the concomitant compensatory curves (usually stiff) resulting in longer fusion with the functional deficit.

12.7 Potential Complications

Neurologic compromise is rare. Progressive scoliosis may result in severe and very severe deformity; trunk imbalance, short trunk, pulmonary restriction, and loss of function are also possible.

12.8 What Patient and Family Should Know?

Patients must undergo regular follow-up until the end of growth. In the case of deformity progression, or new deformity, further surgery may be needed.

Further Readings

1. McMaster MJ, Ohtsuka K. The natural history of congenital scoliosis: a study of two hundred and fifty-one patients. J Bone Joint Surg Am. 1982;64:1128–47.
2. Ruf M, Harms J. Posterior hemivertebra resection with transpedicular instrumentation—early correction in children aged 1 to 16 years. Spine. 2003;28:2132–8.
3. Ruf M, Jensen R, Letko L, Harms J. Hemivertebra resection and osteotomies in congenital spine deformity. Spine. 2009;34(17):1791–9.