Idiopathic Scoliosis

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Two important points should be remembered when dealing with patients with idiopathic scoliosis:

1. Stiff deformity, painful deformity, or abnormal abdominal reflexes are not suggestive of idiopathic scoliosis.
2. Maximum reduction of the deformity by surgery is not always the optimal for balance, harmony, and function.

16.1 Definition

Idiopathic scoliosis (IS) is a deformity of the axial skeleton that develops from head to pelvis (Videos 16.6 and 16.10); IS is in relation with the erect posture of the human and it is a three-dimensional deformity located in the sagittal, coronal, and horizontal (rotation) plane [1]; the torsion of the vertebral column is the key deformity. It is called idiopathic because until now no clear etiology has been established. Most advanced research converges to a genetic neuro-hormonal cause in relation to the erect posture in otherwise healthy individuals.

IS involves up to 3% of the population (>90% females), and it must be distinguished from a scoliotic attitude where torsion does not exist.

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16.2 Natural History

IS can be artificially classified according to age into infantile (0–3 years) (Chap. 13), juvenile (3–9 years), adolescent (9–end of growth) (Chap. 17) and adult (neglected case or de novo deformity) (Chaps. 48 and 49). The majority of IS cases show typical deformity progression (worsening of the torsion) during the adolescent growth spurt (Appendix M). The rate and severity of deformity progression during childhood and adolescence is extremely variable. Generally, the end of skeletal growth stabilizes the deformity even though the deformity tends to progress mainly at lumbar level during adulthood.

The three-dimensional architecture of the deformity [2] and the anatomical location of the deformity are responsible of the major related problems:

Thoracic spine: Apart from the cosmetic issue of the hump, the respiratory problems are predominant if significant coronal or sagittal deformity exists.

Thoraco-lumbar and lumbar spine: cosmetic issues (waist asymmetry) and balance problems are predominant; moreover, the rotatory disorders (degenerative) become predominant during adulthood and are responsible for pain (Chap. 45).

Physical Examination (1) Patients do not complain of pain nor stiffness. A painful and stiff scoliosis is from tumor, trauma, or infection until proven the contrary. (2) The patient is clinically checked in a standing posture (Fig. 16.1a): (a) to rule out lower limb discrepancy; (b) the plumb line dropped from C7 help to detect and

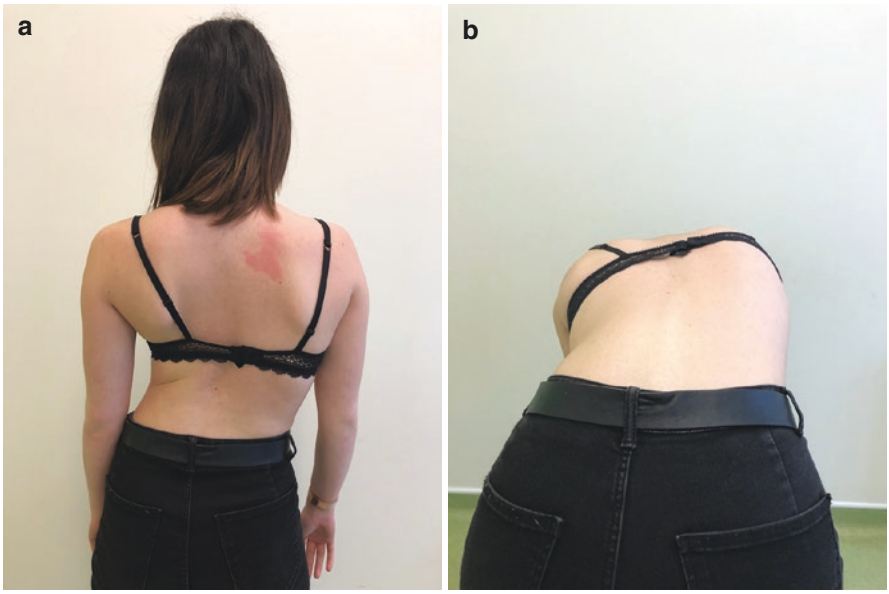

Fig. 16.1 Clinical view of a typical adolescent idiopathic patient. She has an apparent truncal shift and waistline asymmetry (**a**). Adam's forward bending test of the patient shows a right rib hump (gibbosity) which represents the axial rotation of vertebrae at the thoracic spine (**b**). (Courtesy of Prof. Jean Dubousset)

quantify right/left imbalance; (c) the Adam's forward bending test is used to detect, locate and measure the gibbosity (with the aid of the "scoliometer") (Fig. 16.1b); (d) to detect and record the severity of waist asymmetry (window sign); (e) additional signs: shoulder balance (elevated/symmetric/asymmetric), sagittal plane abnormality (kyphosis/lordosis; anterior hump. (3) Inspect the skin of the entire body to detect "café au lait" spots suggestive of Neurofibromatosis type-1 or McCune-Albright syndrome; search for angioma (midline or a tuft hair predicting spinal cord anomaly (Chap. 31). It is wise to check the elasticity of the skin and of the joints of the hand to rule out connective tissue diseases such as Ehlers Danlos, Marfan; if such diagnosis is suspected, echocardiography is more important than the radiographs of the spine in order to detect a latent aneurysm of the aorta! ***Complete neurological examination*** is mandatory: the examiner must check the cranial nerves, both upper and lower extremity osteo-tendinous reflexes, spasticity and clonus, Babinski, and extrapyramidal function (Videos 16.4 and 16.9); abdominal reflexes must always be texted as the absence of *cutaneous abdominal reflex* may lead to suspecting a spinal dysraphism (Chiari malformation, syringomyelia) (Chaps. 30 and 31).

Respiratory and pulmonary function (pulmonary function tests) must be assessed at rest and during effort. It is important to remember that *growth of the spine and growth of the lungs are linked together from birth to 7 years of age when alveolar multiplication stops.*

A precise evaluation of secondary sexual characteristics is needed though it requires a lot of tact; breast and pubic hair development in girls as well as knowledge about the menarche status: growth spurt starts with the first pubic hairs and stops with the full horizontal pubic hair development; the first menarche is the end of the ascending phase of the pubertal diagram (Appendix M). In boys the augmentation of testis size highlights the beginning of the pubertal growth spurt which ends with the appearance of the facial hair (beard).

Finally, it is wise to ***check the family history*** and to rule out if other family members have the disease because of the frequent genetic inheritance.

16.3 Imaging

Remember radiographs are only the shadows of the three-dimensional reality as they can show the spine in two planes only. The essential imaging is the full spine standing (anterior-posterior and lateral projections); the cervical spine up to the skull and the entire pelvis should be included (if possible with low dose radiation allowing a 3D computer reconstruction of the spine; EOS system). The full spine radiographs allow locating and measuring all the curves (Video 16.6) and identifying the apex of the deformity, the "junction" between curves and between spinal segments and the axial rotation of each vertebra. The Cobb angle with its perfect reproducibility is still the basic measurement; the evaluation of the apex and of the junctional levels must be done on both projections.

The flexibility of the curve can be assessed on traction or bending films or fulcrum bending films; the examiner must evaluate the Cobb angle (Video 16.6) and the rotation of the vertebrae. This allows differentiating between structural and compensatory curves; in particular, axial rotation does not disappear in structural curves while it disappears completely in compensatory curves.

A computed tomography (CT) scan and a bone scan have limited indications in patients with idiopathic scoliosis; CT is useful to measure the spinal penetration index.

Magnetic resonance imaging is highly recommended in the infantile group and every time a spinal dysraphism is suspected (left thoracic curve, painful scoliosis, abnormal neurological exam).

Three-dimensional reconstruction of the spine with vertebral vectors representation and view from the top help to evaluate in a glance the deformities and help to choose the levels to be treated [3]. Axial and intervertebral rotation can be measured by the Nash and Moe or the Perdriolle method. Bone maturation is assessed with the Risser sign, the hand and elbow maturation, and the vertebral listel ossification (Appendix M).

16.4 Treatment Options

At any age, treatment options can be observation, exercises, cast, brace, or surgery. Exercises alone are ineffective and the brace is often the treatment of choice (Chap. 18).

Surgery cannot be performed at any age to the negative effects of early arthrodesis on spine and thoracic cage growth and function; modern techniques allow the spine to grow even though autofusion and subsequent stiffness are frequent (Chap. 13).

For adolescents and adult patients, goals of treatment are Cobb angle correction, cosmetic improvement, balance and harmony of the spine, and the best possible function in relation to the three-dimensional mobility of the unfused segments above and below the fusion mass (major importance of selecting properly the limits of fusion) (Chap. 19). Surgery is indicated in case of deformity progression not responsive to brace treatment (>45°); however, current orientation is not to wait too much for worsening of the deformity but to get a reliable prognosis for progression and to treat by brace as soon as possible even with very low Cobb angle in order to prevent surgery (Video 16.6).

16.5 Potential Complications

Potential complications may occur following serial casting or surgical treatment.

Complications related to casting are pressure sores and brachial plexus injury; such complications can be prevented by good technical expertise and proper advice

to the patient and parents. Surgical complications can be (1) **neurological** (roots or cord): usually secondary to over-stretching of the cord or misused instrumentation; prevention: preoperative progressive awake traction, spinal cord monitoring (during surgery), wake-up test (during surgery); (2) **infection:** mostly early infection (<3 months) (Chap. 66); prevention: perioperative antibiotics; surgical setting. Treatment requires early reoperation in order to keep the hardware in place. Late infection (after fusion obtained) can also occur; the treatment consists of hardware removal and antibiotic treatment; (3) **nonunion or pseudarthrosis**: revision surgery (localized).

16.6 What Should Patient and Family Know?

Families and patients must be aware that (1) when brace (or cast) treatment is prescribed, compliance is mandatory, and patient and parents' collaboration is required; (2) a straight spine on post-operative radiographs is not the ultimate goal of surgical treatment: balance, harmony, and residual motion are the main goals.

Further Readings

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