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The changing role for neurosurgeons and the treatment of spinal deformity

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Spinal deformity has classically and historically been studied by those in the discipline of orthopedic surgery. This may be attributable to the orthopedic interventionalists' experience with osseous fixation for long-bone and other skeletal fractures. Neurosurgeons have maintained a long-standing interest in complex cervical spinal disorders, and their interest in the larger field of complex spinal deformity has been expanding.

An understanding of spinal deformity disorders, biomechanics, bone biology, and metallurgy is necessary before clinical, teaching, and research activities can be undertaken within neurosurgery.

The authors describe basic and advanced concepts of spinal deformity management with cases to illustrate teaching points.

Key Words • **neurosurgery** • **spinal deformity** • **scoliosis** • **kyphosis**

Spinal deformity is a complex subspecialty that involves the management of disorders that affect the spinal alignment and balance. Neurosurgeons have traditionally treated various spinal disorders secondary to traumatic, congenital, neoplastic, and infectious causes involving the spinal column, cord, and nerves as well as a wide array of degenerative compression syndromes. As the ability to treat more complex neurosurgical spinal disorders has evolved, spinal instability has often required the use of modern reconstructive techniques to preserve neurological function, maintain or correct spinal alignment, and achieve structural stabilization. Alternatively, some complex neurological disorders are associated with spinal deformity, whereas certain neurological disorders are indeed caused by deformity of the spinal column (Fig. 1). It is such spinal deformity–related cases, as well as others with related disorders, that have generated interest in this discipline and recognition that a fundamental understanding of spinal deformity is essential for the practice of neurological surgery.

Types of Spinal Deformity

The practicing neurosurgeon should be familiar with the two broad classifications of spinal deformity: coronal-plane (scoliotic) and sagittal-plane (kyphotic and lordotic) abnormalities; these curvatures can be readily recognized on physical examination and confirmed on imaging (Fig. 2). Lordosis is also considered a deformity if present in abnormal locations such as the thoracic spine, a phenomenon that never occurs in the normal thoracic spine. Hyperlordosis of the cervical and lumbar spine can occur to compensate for thoracic kyphosis.

Because spinal deformity is a three-dimensional entity, the compound terms "kyphoscoliosis" and "lordoscoliosis" are often used to define further the character of these complex curves. Often more than one curve exists in cases in which the primary lesion (that is, structural curve) is the true pathological curve that is fixed and is defined by its larger size and rigidity; however, the secondary curves are often compensatory because of the primary curve.

Causes of Spinal Deformity

The origins of spinal deformity can be classified into two broad groups: idiopathic or acquired. These classifications also generally fall into two broad age categories of pediatric and adult, although notable exceptions exist. There are numerous causes of both adult and pediatric spinal deformities. Tables 1 to 3 provide relatively comprehensive lists of most causes/origins of spinal deformity.

Clinical Presentation and Decision Making

The practicing neurosurgeon must possess a fundamental ability to assess various spinal deformities. Additionally he/she must be able to recognize their clinical presentations

Abbreviations used in this paper: AP = anteroposterior; MR = magnetic resonance; PSO = pedicle subtraction osteotomy.

FIG. 1. Magnetic resonance image obtained in a 22-year-old patient presenting with progressive spastic quadriparesis due to severe basilar invagination and a large syrinx. He underwent a transoral approach to decompress the severe deformity and then occipitocervical fusion for stabilization.

and syndromes to establish appropriate patient care and surgical or nonsurgical treatment.

Idiopathic scoliosis typically develops in adolescent girls around the time of menarche and presents with a conspicuous chest-wall deformity. Most often the effects are cosmetic, whereas neurological function is normal and symptoms minimal. In cases of adolescent scoliosis, brace therapy is undertaken to delay or prevent the need for surgical intervention. Surgery, in fact, is often delayed as long as possible or until maximum possible skeletal maturity has occurred. Skeletal maturity is determined by the radiographic appearance of the iliac apophysis, more commonly known as the Risser sign, where skeletal maturity has been reached approximately at age 14 years in girls and age 18 years in boys.[1,24,29] Milder cases of scoliosis often will not progress significantly after onset of skeletal maturity and therefore will not require further treatment. If the deformity progresses to an advanced state, however, surgical correction becomes necessary; thus, the surgeon requires an understanding of the threshold and decision-making process for surgical indications. A general indication for surgery is the following: deformity exceeding 50° with continued and documented progression or intractable pain refractory to brace therapy. Severe curvature greater than 70° is also likely to be associated with cardiopulmonary or neurological symptoms even after establishment of skeletal maturity; these deficits are more difficult to correct in

TABLE 1

Classification and terminology of structural scoliosis

idiopathic	
	infantile
	juvenile
	adolescent
neuromuscular	
	neuropathic
	myopathic
congenital	
	failure of formation
	failure of segmentation
neurofibromatosis	
mesenchymal disorders	
	Marfan syndrome
	Ehlers–Danlos syndrome
	other
rheumatoid disease	
trauma	
	fracture
	surgical
	irradiation
extraspinal contractures	
	postempyema
	postthoracoplasty
osteochondrodystrophies	
	diastrophic dwarfism
	mucopolysaccharidosis
	spondyloepiphysial dysplasia
	multiple epiphysial dysplasia
infection of bone	
	acute
	chronic
metabolic disorders	
	rickets
	osteogenesis imperfecta
	homocystinuria
	others
related to lumbosacral joint	
	spondylolysis & spondylolisthesis
	congenital anomalies of lumbosacral region
tumors	
	vertebral column
	spinal cord

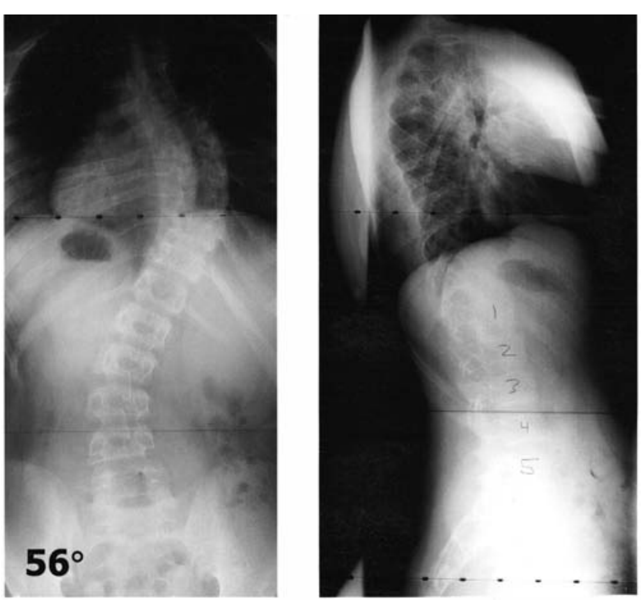

FIG. 2. Long-cassette 36 × 14–in AP *(left)* and lateral *(right)* radiographs obtained to evaluate spinal curvature and sagittal balance.

TABLE 2
Classification and terminology of nonstructural scoliosis

postural scoliosis
hysterical scoliosis
nerve root irritation
herniation of nucleus pulposus
tumor
inflammatory
related to leg length discrepancy
related to hip contracture

advanced stages.[2,28,30] In general, it is also important to understand that pediatric deformity is often distinctly different from that in adults because the spinal column in children is relatively flexible and mobile in contrast to the stiff and inflexible column present in adults.

The classification of adult deformity can include cases involving adolescent patients with scoliosis in whom the curvature has remained stable into adulthood when it then becomes symptomatic due to adult degenerative spinal disease. In such cases the thoracic and lumbar spine are typically involved and the characteristic rotatory deformity is present, consistent with adolescent deformity (Fig. 2). Alternatively the de novo adult lumbar degenerative scoliosis is not related to adolescent scoliosis, and in most of these cases the scoliotic deformity occurs in women older than 60 years of age and involves the midlumbar segments (Fig. 3).

TABLE 3
Classification and terminology of kyphosis

postural (roundback)
Scheurmann disease
neuromuscular
myelomeningocele
developmental (late paralytic)
congenital (present at birth)
congenital
defect of formation (hemivertebra)
defect of segmentation (Klippel–Feil syndrome)
traumatic
due to bone &/or ligament damage w/o cord injury
due to bone &/or ligament damage w/ cord injury
postop
postlaminectomy
after vertebral body removal
postirradiation
metabolic
osteoporosis
osteomalacia
osteogenesis imperfecta
other
skeletal dysplasias
acondroplasia
mucopolysaccharidosis
neurofibromatosis
other
collagen disease
Marie–Strupell disease
other
tumor
benign
malignant
inflammatory & infectious

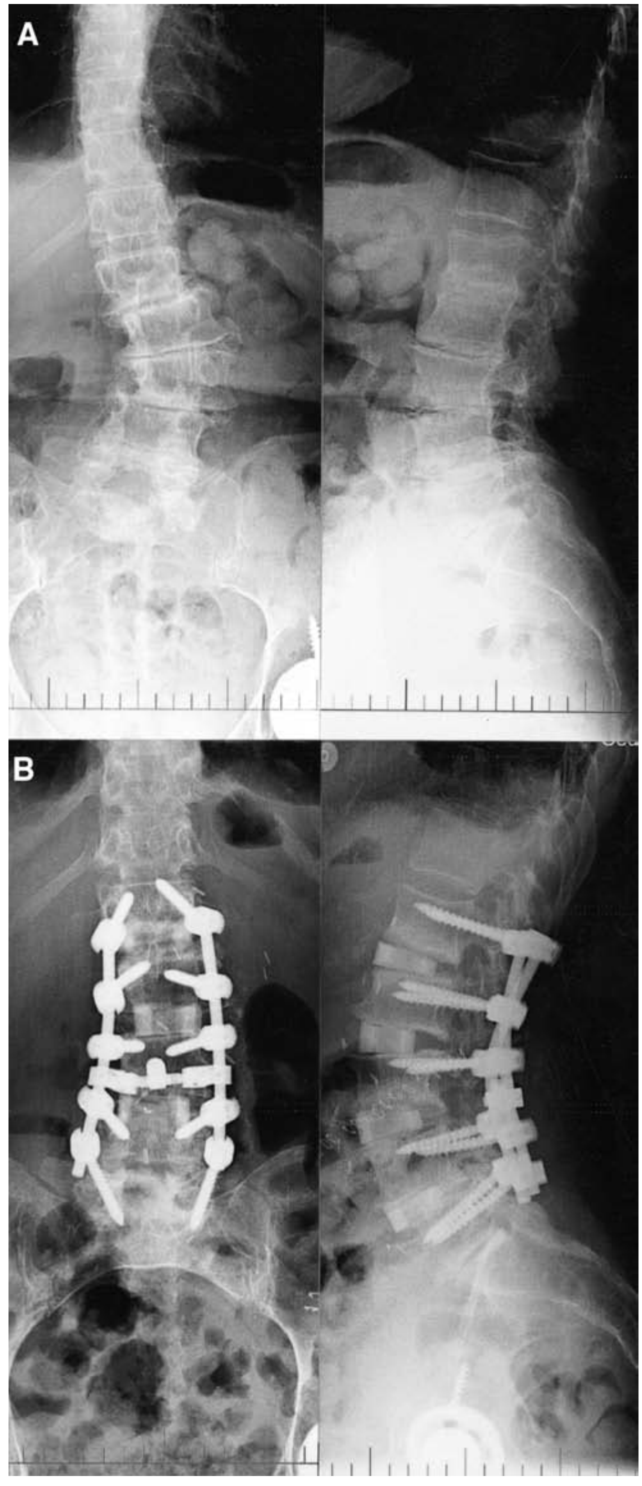

FIG. 3. Adult patient with degenerative lumbar scoliosis *(left panels)* who underwent a multilevel anterior lumbar interbody fusion and posterior instrumentation-assisted fusion *(right panels)*.

Kyphotic deformities are caused by forward sagittal-plane decompensation and can occur in the cervical, thoracic, or lumbar spine. The classic thoracic kyphotic deformity is Scheurmann kyphosis (Fig. 4), which occurs in

FIG. 4. Sagittal MR image revealing Scheuermann kyphosis with classic multilevel degenerative discs, multiple endplate irregularities, and wedging of multiple vertebral bodies *(arrows)*.

pediatric and adult patients and presents primarily with thoracic pain. The prototypical lumbar kyphotic deformity is the iatrogenic flat-back syndrome, usually occurring if the patient is incorrectly positioned on the operating table (in a nonlordotic position) due to instrumentation-induced loss of lordosis (Fig. 5).

Evaluation and Diagnosis of Spinal Deformity

The practicing neurosurgeon is familiar with clinical

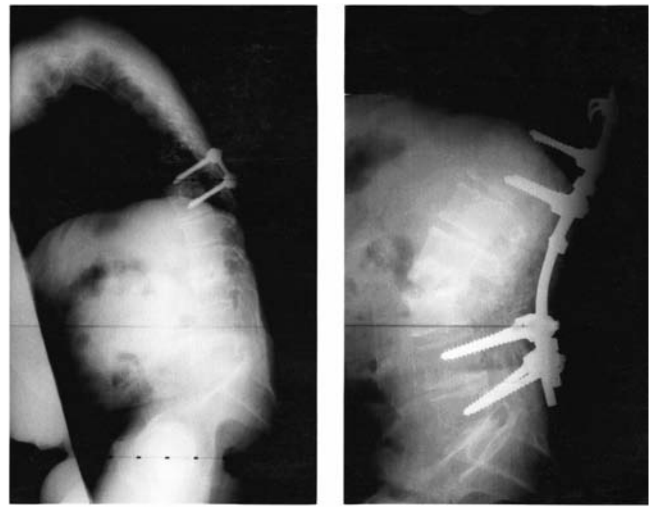

FIG. 5. Postoperative images of posterior lumbar L2–S1 spinal fusion *(left)* and a T11–12 fusion *(right)* causing severe flat-back syndrome with a typically stooped position. The patient was treated with an L-3 posterior PSO to correct the deformity.

evaluation of the spine to determine the presence of significant medical, musculoskeletal, or neurological abnormalities that may be associated with various spinal disorders; this evaluation may be expanded to include basic assessment of deformities.[19,25] Other clinical and radiographic/neuroimaging abnormalities should be examined in patients with curvatures to include body habitus and spinal column alignment that may exhibit a characteristic asymmetrical hump in the presence of significant scoliotic deformity (Fig. 6). Full-length (36 × 14–in) standing spinal radiography is performed to assess the overall spinal balance (Fig. 2). Cobb angles are measured on these radiographs by using a goniometer in both AP and lateral orientations (briefly, a perpendicular line is drawn from the endplates of the most angulated vertebrae involved in the pathological curve [Fig. 7]). Coronal and sagittal alignment is assessed using a plumb line in the AP and lateral projections to determine if decompensation is present (Fig. 8). Dynamic

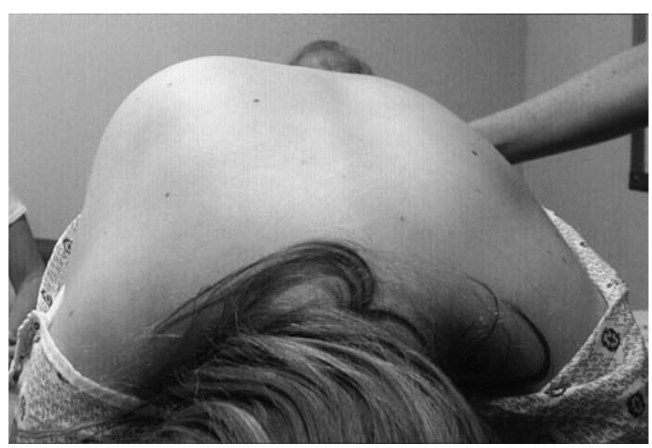

FIG. 6. Photograph of a patient with asymmetrical elevation of the chest wall (that is, rib hump) due to scoliosis causing spine rotation in rib-cage elevation.

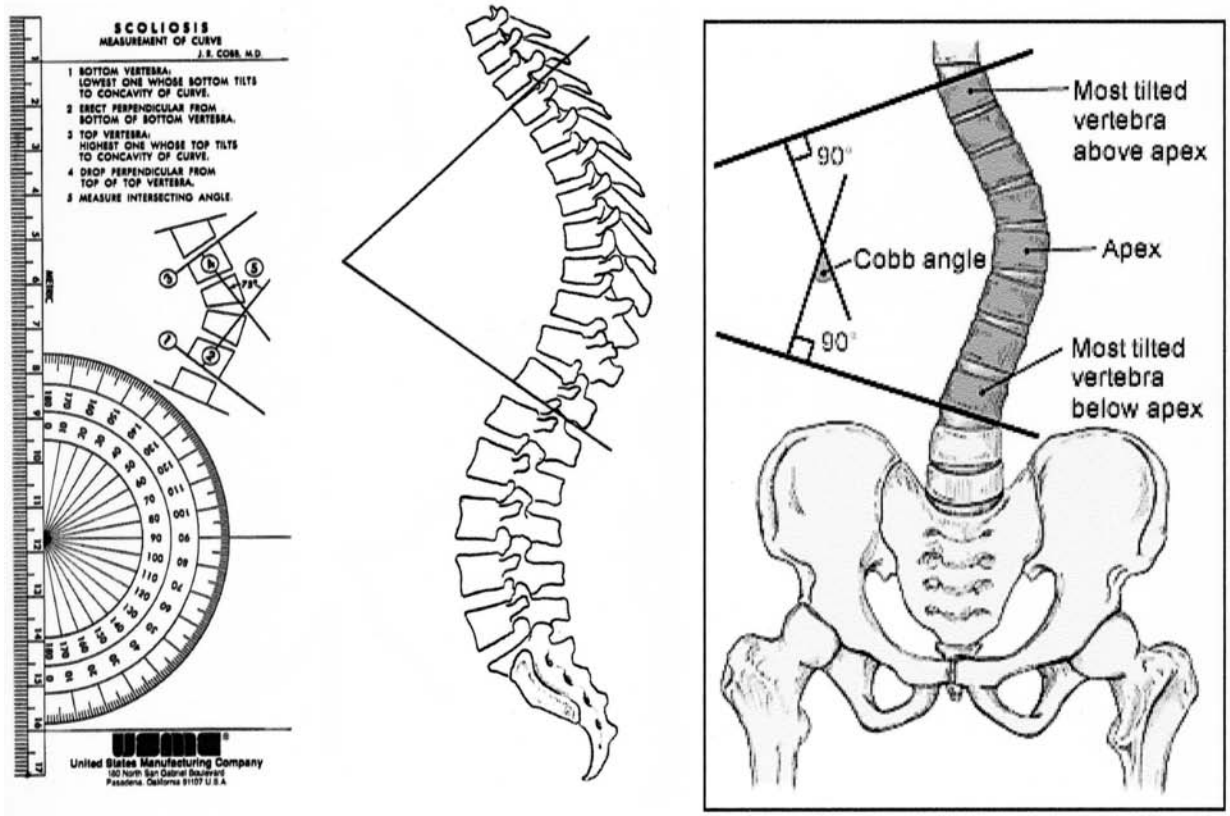

FIG. 7. Diagrams showing the use of a goniometer *(left)* for measurement of Cobb angles from the vertebral endplates at the distal ends of the curve and measured to determine the maximum curve angle, measurement of the sagittal-plane angle *(center)*, and measurement of the coronal-plane angle *(right)*.

lateral view flexion–extension radiography is performed to evaluate sagittal-plane instability (that is, spondylolisthesis), and AP lateral bending radiography is conducted to evaluate the location of a rigid structural curve of a deformity (Fig. 9). Additional data are acquired, as needed, using MR imaging or computerized tomography myelography for routine clinical evaluation. In general there are fewer indications for MR imaging and computerized tomography scanning in idiopathic scoliosis cases, although most patients with acquired deformity will need to undergo some form of advanced imaging.

Epidemiology and Classification of Spinal Deformity

The practicing neurosurgeon should also have a basic understanding of the epidemiology, natural history and current classifications of deformity to manage and communicate with patients as well as to confer with other physicians regarding spinal deformity. Standard terminology established in a glossary by the Scoliosis Research Society is available on their website (www.SRS.org) and serves as a common language for defining spinal deformity. Although some familial tendency for adolescent scoliosis is likely, there is no clear inheritance pattern nor is there an established cause of scoliosis.[32] A minimum curve of 10° must be present to establish a diagnosis of scoliosis (incidence ~ 3% of the population). Curves greater than 20° appear in 0.5% and those greater than 40° are present only in 0.1% of the population.

The anatomical classification of spinal deformity is based on its physical location (thoracic, thoracolumbar, or lumbar region);[7] other systems for defining treatment of adolescent scoliosis were published in 1983[15] (King classification) and 2001[16–18] (Lenke classification). The clinical presentation of a deformity is somewhat bimodal, with idiopathic deformity occurring in the adolescent population and adult deformity presenting after 50 years of age.

History of Surgical Treatment for Spinal Deformity

The treatment of spinal deformity originated in ancient times,[9,21] dating to Hippocrates,[14] when traumatic spinal injuries were treated with a "rack system" to straighten the spine, indicating a long-term recognition of spinal deformity (Fig. 10).

One of the earliest surgical interventions involving an implant to stabilize the spinal column was described in 1891 by Hadra,[10] who used wire fixation to correct tuberculous deformity. In 1924 Hibbs[13] established the effectiveness of in situ osseous fusion to stabilize spinal deformity. According to most experts the modern era of spinal instrumentation was initiated with the development of hook/rod technology introduced by Harrington[12] in 1962. In 1976 Luque[20] improved on the latter by introducing the

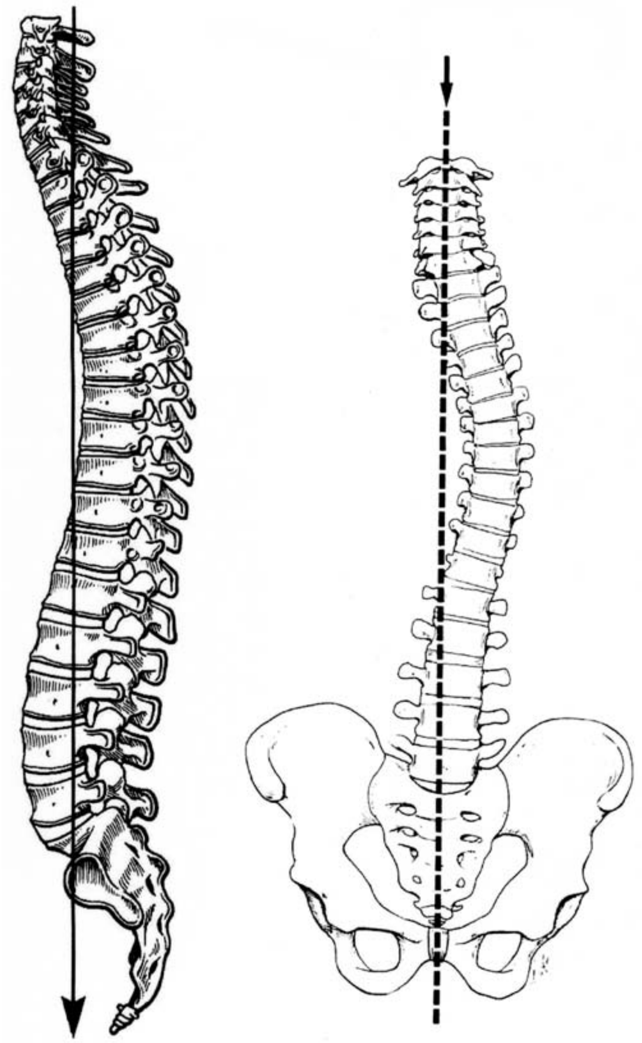

FIG. 8. Method for establishing a sagittal plumb line (from center of the C7–T1 disc and the anterior S-2 border; *left*) and coronal plumb line (C1–sacrum; *right*) along the long axis of the spinal column.

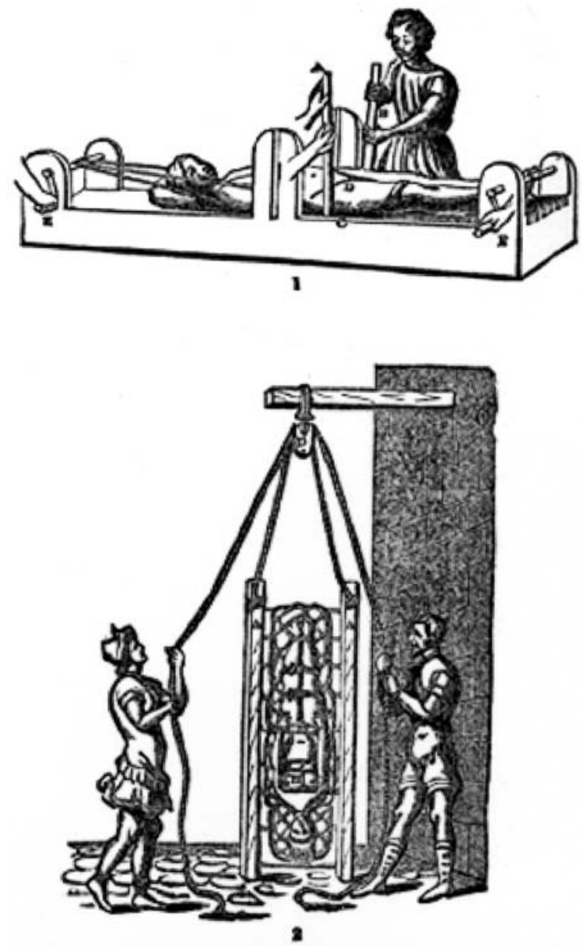

FIG. 10. Illustrations from the time of Hippocrates. The attempt to correct spinal deformity is depicted.

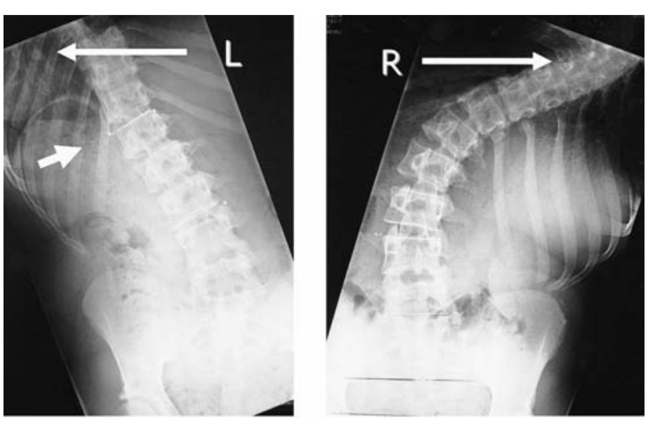

FIG. 9. Idiopathic scoliosis evaluated on AP lateral radiographs with the patient bending to the left *(left)* and right *(right)*, demonstrating a structural curve *(short arrow)* to distinguish it from flexible, nonstructural curves *(long arrows)*.

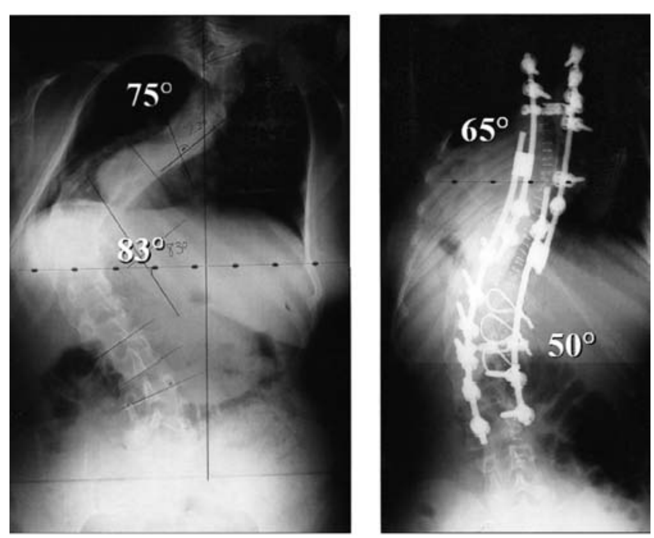

FIG. 11. Preoperative *(left)* and postoperative *(right)* radiographs obtained in a 14-year-old patient with severe idiopathic scoliosis who underwent anterior release and posterior multisegmental screw fixation.

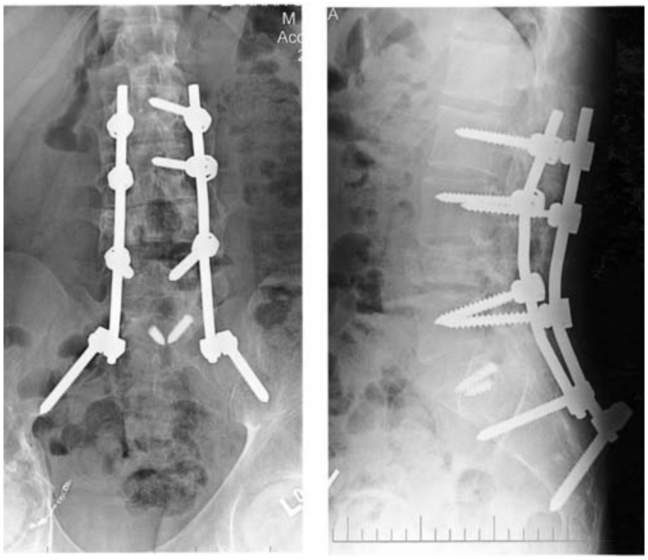

FIG. 12. Anteroposterior *(left)* and lateral *(right)* radiographs demonstrating pelvic fixation utilized in a long-segment correction of posttraumatic kyphosis in a patient treated with L-4 PSO.

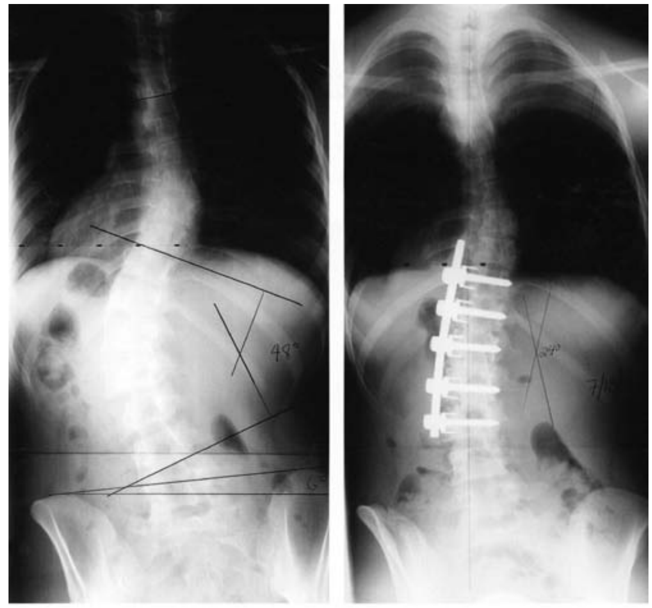

FIG. 13. Preoperative *(left)* and postoperative *(right)* radiographs acquired in a 15-year-old patient with thoracolumbar scoliosis who underwent short-segment anterior segmental deformity correction.

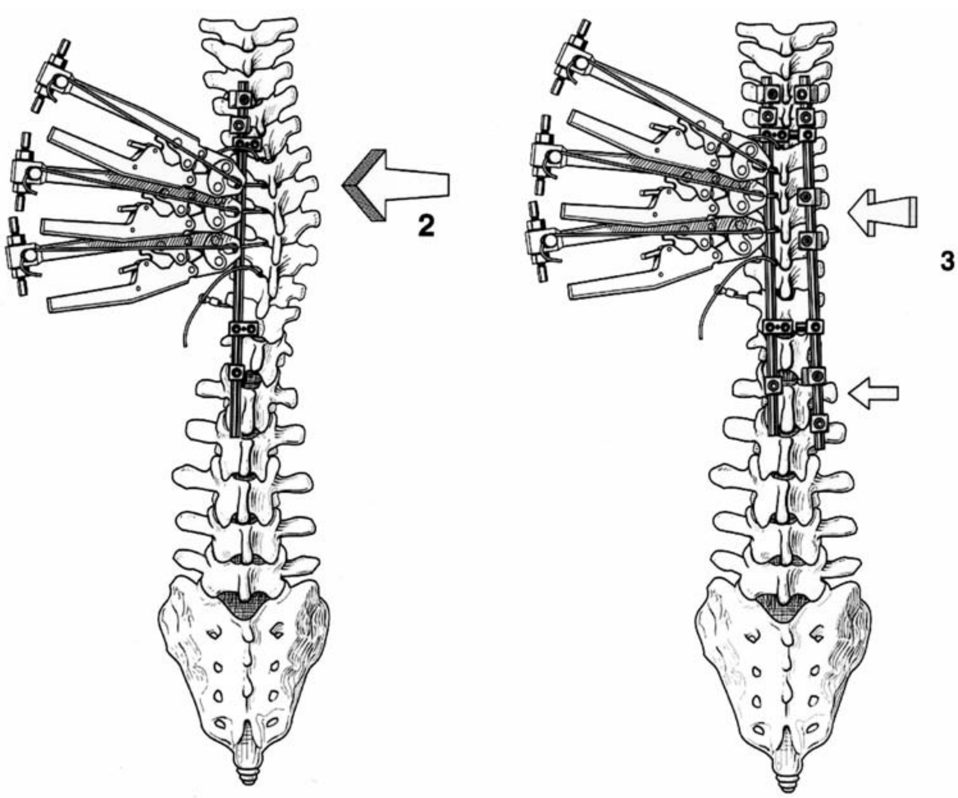

FIG. 14. Illustrations of three-point bending techniques used for correction of spinal curvatures.

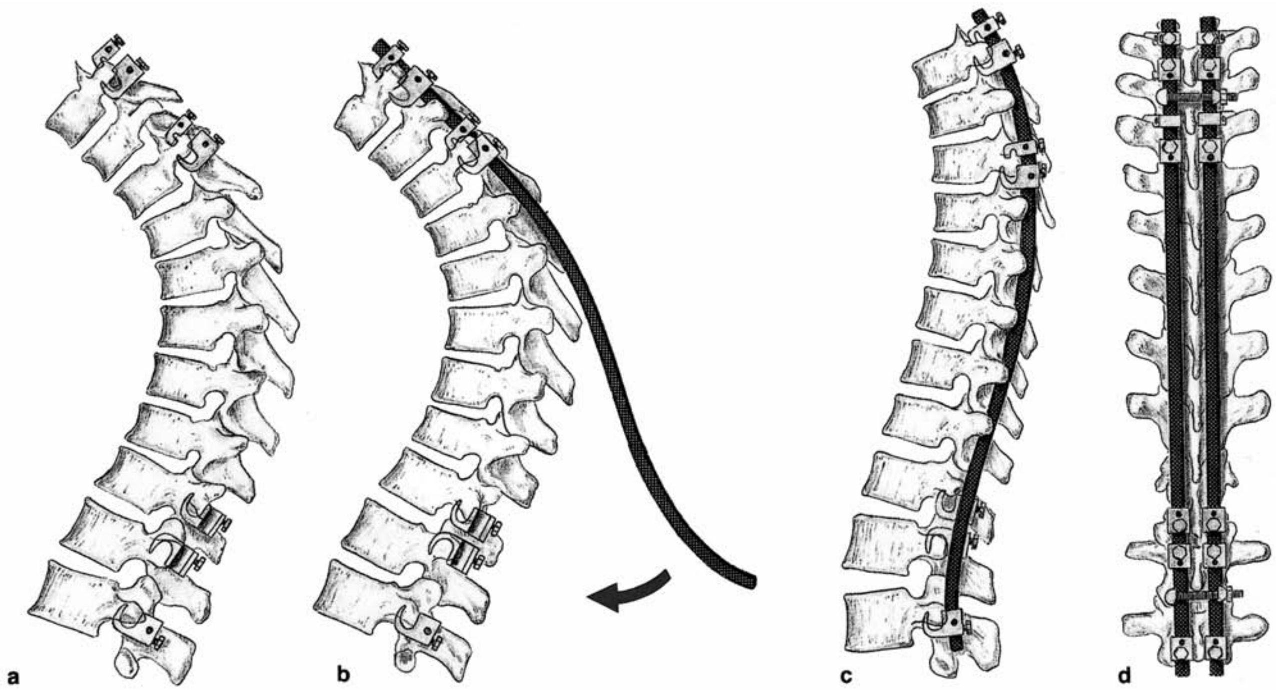

FIG. 15. Illustrations of cantilever techniques to achieve correction of kyphotic deformities.

concept of segmental fixation involving placement of sublaminar wires and rods. Providing an alternative approach for the correction of spinal deformity, anterior instrumentation was first described by Dwyer, et al.,[8] in 1969 and the technique was subsequently updated in 1976 by Zielke, et al.[33] The contemporary use of hook–screw/rod segmental fixation for deformity correction was described by Cotrel, et al.,[6] in 1988; this procedure laid the groundwork for the multitude of technologies that have evolved since.

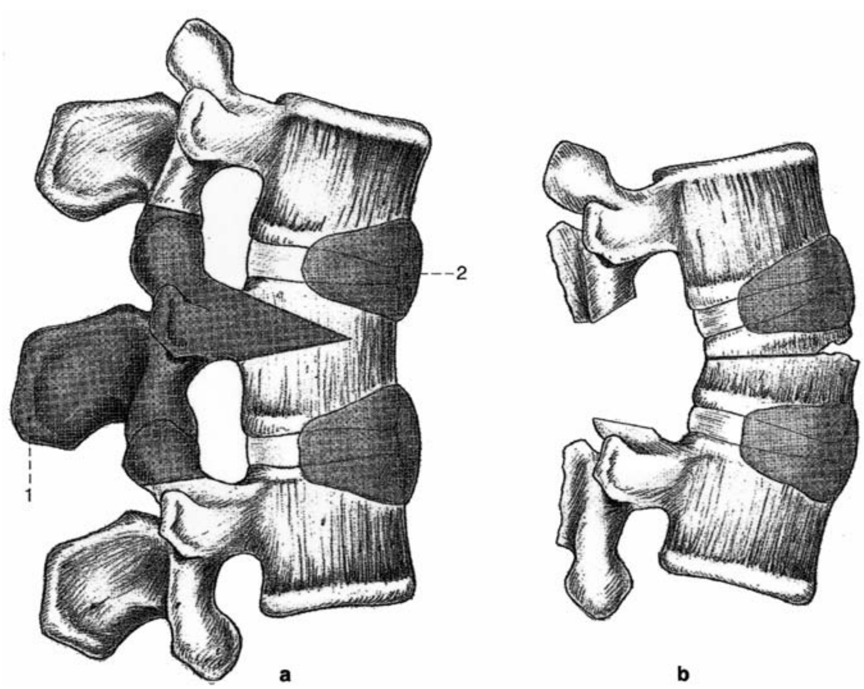

FIG. 16. Illustrations of the PSO procedure in which osseous resection of the facets and pedicles allows restoration of lordosis at a single vertebral level.

Contemporary Surgical Treatment of Spinal Deformity

Current surgical techniques for correction of spinal deformity have evolved significantly during the past decade, including numerous innovative devices for anterior and posterior correction procedures with segmental screw fixation.

In posterior spinal instrumentation procedures we currently use multisegmental screw fixation techniques[3,11,23,26,27] that can be performed at all spinal levels (Fig. 11). Pelvic fixation is similarly associated with improving outcomes after long-segment fixation across the lumbosacral joint (Fig. 12). Alternatively, in certain cases of adolescent scoliosis, correction can be achieved only via an anterior approach, which avoids posterior paraspinal musculature injury and reduces the number of segments fused because transvertebral screw/rod fixation is performed (Fig. 13). Posterior correction of the deformity is accomplished using the principles of three-point fixation and bending; in this, proximal and distal instrumentation is placed, and the deformed segment is reduced to the midline (Fig. 14). Posterior sagittal-plane correction requires a cantilever technique[27,31] for correction of kyphotic deformities (Fig. 15), necessitating multiple-level (that is, Smith-Petersen) facet joint osteotomy mobilization; additionally some will require an anterior release and multilevel discectomies, which are now performed in pediatric cases[22] with thoracic endoscopy. Patients with lumbar flat-back syndromes (that is, kyphosis) can undergo posterior PSO and placement of instrumentation by using a cantilever technique to reduce deformity (Figs. 5 and 16). An alternative strategy is single-level correction and a posterior PSO.[22,26] The latter requires a wedge-shaped resection of the posterior elements, including the pedicles, and an eggshell-type resection of the vertebral body allows a closing osteotomy to achieve up to 35° of correction.[4,5]

Neurological Manifestations of Spinal Deformity

Patients with spinal deformity can experience a wide variety of symptoms ranging from an absence of any symptomatology in the adolescent with idiopathic scoliosis to radiculopathy in the adult with degenerative scoliosis and more severe neurological involvement that includes myelopathy and syringomyelia (Fig. 1). The cause and effect of these entities remains unclear—some patients exhibit a deformity causing a syrinx, whereas in others scoliosis progression is slowed by treatment of the syrinx. The true relationship between spinal deformity and syringomyelia remains unclear, and further study is required.

Conclusions

The roles of neurosurgery and spinal deformity continue to evolve in relation to the discipline of spinal surgery. Orthopedic surgeons have traditionally been involved in the care of spinal deformity, making major contributions to our understanding of biomechanical factors; however, neurosurgeons have been involved in the care of a subset of patients with spinal deformity making similar and parallel advances in the understanding of spinal disorders. The combined efforts of the two disciplines in recent years have resulted in a synergy of innovations and patient care that will push the frontiers of spinal surgery to levels beyond imagination.

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