A clear understanding of the pathogenesis of idiopathic scoliosis is essential to appreciating its clinical behavior. Although epidemiological surveys have shown an enormous number of children with minor coronal-plane deformities of the spine, only a relatively small number show evidence of progressive idiopathic scoliosis, which means that other factors must be superimposed to make a deformity idiopathic and progressive. The environment of growth is clearly important, as with other progressive skeletal deformities. The prevalence rate of minor curves in boys is about half that for each age in girls, and the difference is progressively more obvious the greater the size of the curve. Boys are therefore in some way protected from idiopathic scoliosis, but their spines are going to be subjected to much the same neuromuscular, metabolic, and endocrine processes during growth as those of girls. On a commonsense basis it would seem unlikely that pointing the finger of suspicion, for instance, at the paravertebral musculature, brain, eyes, ears, spinal cord, nerve roots, muscles, collagen, and even platelets would not be a profitable line of research into the cause of idiopathic scoliosis. Notwithstanding, this path has largely been the focus of research activity in the etiology of idiopathic scoliosis over the past half century.

### Muscular Theories

Most hypotheses and speculations put forward about the cause of idiopathic scoliosis concern neuromuscular theories, although idiopathic scoliosis is defined as a spinal curvature in the absence of any associated musculoskeletal condition. Since Lerique and Le Coeur first demonstrated electromyographic asymmetry in the paraspinal muscles of patients with the condition,1 much work on the paraspinal musculature has been performed, including studies of muscle fiber type and ultrastructural differences.2–11 Although Zetterberg and colleagues showed that these abnormalities resembled the sort of changes encountered after endurance training, indicating a secondary or adaptive process (i.e., secondary to the presence of a spinal curvature) in their occurrence,12 Saartok et al went even further, stating that “a neuromuscular imbalance has not been shown to be an etiological factor for the idiopathic form of scoliosis.”13 The histological specimens were obtained during scoliosis surgery from a highly selected group of patients with larger curves, focusing on the difference between the right and left sides, when the geometric problem is instead a front-to-back buckling lordosis (vide infra).

Platelets contain actin and myosin, and because of their resemblance to those in skeletal muscle, these structures have also been examined in idiopathic scoliosis. Early studies reporting abnormal morphology and function of platelets in patients with idiopathic scoliosis14–16 were not confirmed in subsequent studies, which showed no difference between patients with idiopathic scoliosis and controls.17,18 Platelet aggregation abnormalities were shown to be more prevalent in those with larger scoliotic curves, again indicating a secondary effect.19,20 Calmodulin regulates the contractile properties of muscles and platelets through changes in calcium concentration, and higher platelet calmodulin levels have been demonstrated in patients with progressive curves,21 as have lower levels of melatonin, a calmodulin antagonist.22 It is unclear what these changes reflect other than the biochemistry of growth in general.

### Neurological Theories

Because scoliosis is associated with many neurological diseases, from those affecting the brain to those of peripheral nerves, several neurological abnormalities have been described in association with idiopathic scoliosis.

Electroencephalography, proprioception, and vibration sense have been examined in idiopathic scoliosis, as well as balance and electronystagmography.23–26 Recently, abnormal somatosensory function has been demonstrated in patients with adolescent idiopathic scoliosis,27,28 but its relationship to curve severity again suggests changes secondary to the presence of a deformity. Forty years ago it was thought that idiopathic scoliosis could be caused by a short spinal cord,29 and more recently Porter has shown that in patients with the condition the spinal canal is shorter than the spine itself.30–32 This theory has been fancifully called “uncoupled neuro-osseous growth,”32 and the concept of this difference in length has also been supported through screening with magnetic resonance imaging (MRI).33 Thirty years ago in Oxford, cadaver spines with idiopathic scoliosis were measured and it was shown that the spinal canal takes the shortest route down the spine.34
The Leeds Study Group looked at the same specimens in more detail and confirmed the findings that the posterolateral concave canal distance was the shortest spinal route. Since MRI scanning first became available, the Leeds Group has looked at patients with idiopathic spinal deformities. Whereas in Scheuermann’s thoracic hyperkyphosis the spinal cord hugs the back of the vertebral bodies at the apex of the curve, the spinal cord also takes the shortest route in the opposite deformity of idiopathic lordoscoliosis, being close to the back of the vertebral body/pedicle on the concave side of the curve (Fig. 3.1). Patients with idiopathic scoliosis do not have any known clinical neurological abnormality, nor does MRI scanning show any tethering effect in this condition, such as a low conus or a secondary Arnold–Chiari malformation. A spinal canal proportionately shorter than the rest of the axial skeleton in idiopathic scoliosis merely reflects what one would expect of the geometry of a lordoscoliosis.

The advent of MRI scanning demonstrated a greater prevalence of a syrinx in the spinal cord than had previously been found, particularly for less common curve patterns such as a left thoracic curve, a stiff curve, a very painful curve, or a progressive curve in a boy (Fig. 3.2). These unusual curves do have a neuromuscular basis, and syrinx drainage or shunting usually leads to curve stabilization or improvement.

Experimental animal models of root or cord damage have produced nonidiopathic curves, usually instantly upon awakening of the animal from anesthesia. Interestingly, Langenskiold and Michaelsson produced scoliosis in rabbits by dividing the costotransverse ligament, but the resulting curves again turned out to be neuromuscular curves. De Salis and colleagues showed that the segmental artery to the spinal cord in rabbits runs just under the costotransverse ligament, and that damage to this segmental blood supply produced a neuromuscular type of deformity. In rabbits, the spinal cord depends upon a segmental feeding vessel at each level (Fig. 3.3). Not surprisingly, when the costotransverse ligament was divided in primates with an Adamkiewicz type of cord blood supply, the spine remained straight. Thermal coagulation of the facet joint capsules in rabbits also produces a spinal deformity from ischemic cord damage.
Connective-tissue Abnormalities

Because connective tissue disorders such as Marfan or Ehlers–Danlos syndrome are associated with an increased prevalence of spinal deformity collagen structure and metabolism have been extensively investigated both in the skin and in the intervertebral discs in idiopathic scoliosis.45–50 Again the findings were thought to be secondary to the presence of a spinal deformity, and indeed, recent research has excluded collagen abnormalities as potential genetic causes of idiopathic scoliosis.51,52

Genetic Theories

In the late 1960s and early 1970s, the familial nature of idiopathic scoliosis was clearly demonstrated in both Scotland53 and the United States.54 It was thought that idiopathic scoliosis might be inherited in a sex-linked dominant mode, but with variable expressivity and incomplete penetration. Genomic screening and chromosome studies have suggested chromosome 19 as a possible candidate for a genetic source of the disorder,55,56 but idiopathic scoliosis is so multifactorial that it is extremely unlikely that only a single gene is responsible for it.

Longitudinal studies of growth in relation to idiopathic scoliosis show that early reports of children with the condition having been taller and having advanced earlier in...
adolescent growth, while later experiencing growth retardation, were not strictly correct because they relied upon historical controls already shown to be unreliable.\(^57\) When compared with contemporaneous controls, these children showed no differences in comparison with straight-backed counterparts. However, children with bigger curves are significantly taller at each age, but do not grow faster, indicating that a genetically tall stature may be related to the progression potential of idiopathic scoliosis.\(^57\) In such families one would expect to find a high prevalence rate of idiopathic scoliosis, and the concept of a gene for idiopathic scoliosis therefore loses credibility when the familial nature of the disorder can be explained in part on the basis of stature. Moreover, the whole pattern of growth during adolescence is strongly familial,\(^58\) with, for instance, girls and their mothers having their menarches at similar chronological ages.

If idiopathic scoliosis is a matter of abnormal spinal shape that runs in families, then how that shape is achieved must also be genetically determined. Delmas\(^59\) and Stagnara et al\(^60\) both put forward the notion that children have a spinal physiognomy just as they have, for instance, a facial one, and suggested that lateral profile may be governed genetically just as are many other aspects of body shape.

Recently, the familial nature of sagittal spinal shape has been investigated in schools, using the Quantec surface-shape scanning technique, which can noninvasively register the lateral spinal profiles virtually the same. This is a very important genetic element in the pathogenesis of idiopathic scoliosis.

When posteroanterior (PA) X-ray films of idiopathic scoliosis are inspected, it can be observed that the direction of rotation of the spine is constant, with the posterior elements turning toward the curve concavity, and with this rotation being maximal at the curve apex. (Fig. 3.6; see also Fig. 2.4) The posterior elements of the spine are therefore running, as it were, the shorter, inside lane of the “running track,” as this appearance clearly indicates that the back of the spine must be shorter than the front. The PA X-ray view of the patient’s spine is, however, a PA view of everything except the structural curve, because from the neutral vertebra above down to the apex of the scoliotic curve, each vertebra is progressively more rotated out of the frontal plane before recovering from the apical vertebra to the lower neutral vertebra. If the apical vertebra is, for instance, rotated by 30 degrees, then to make a true anteroposterior (AP) film, either the patient or the X-ray beam has to be rotated by 30 degrees from the frontal plane, in which case the size of the deformity is maximized. Stagnara devised this AP view and termed it the plan d’élection view (see Figs. 2.9 and 2.10).\(^63\) If a true lateral X-ray film of the curve apex is to be made, the X-ray beam has to be rotated 90 degrees with reference to the AP plan d’élection view (Fig. 3.7). When this is done, the essential lordosis is visualized.

The Leeds Group studied articulated skeletons with idiopathic scoliosis at the Royal College of Surgeons of Edinburgh Museum, which helped to visualize the lordosis and the nature of the seemingly complex three-dimensional deformity in the disorder.\(^35,64\) Figure 3.8 shows one such specimen. The PA view shows a significant deformity, with
Fig. 3.5  A 14-year-old girl with a 45-degree spinal curve. In the erect position (A) the deformity is much less obvious than (B) on forward bending, in which the rib hump is maximized.

Fig. 3.6  PA X-ray film of a thoracic idiopathic scoliosis with the tips of the spinous processes marked with triangles and the middle of the vertebral bodies with dots. It can be seen that the distance down the back of the spine is shorter than the distance down the front, confirming that all structural scolioses are lordotic.

Fig. 3.7  A true lateral view of the apex of the curvature shown in Fig. 3.6, demonstrating the lordosis.
considerable rotation, and the PA X-ray film demonstrates almost 90 degrees of rotation, with the apical vertebra being seen in an almost lateral projection. The lateral view of the specimen would appear to show the presence of a kyphosis, but it can be seen at the curve apex that the spinous processes are pointing almost directly backward. The lateral X-ray film of the specimen now looks more like an AP view of the curve apex, again confirming a rotation of almost 90 degrees. A true lateral view of the apex thus unmasks the essential lordosis.

Going back to the clinical inspection of patients, it is possible to see the lordosis in idiopathic scoliosis if one knows where to look. Figure 3.9A shows a young man with a 30-degree right thoracic curve. His whole thoracic
kyphosis is flattened, and there is clearly a lordosis in the middle. Figure 3.9B shows a girl with a 70-degree curve. Again looking at the concavity at the curve apex, there is clearly a lordosis. Figure 3.9C shows an extreme degree of infantile progressive scoliosis. The structure bulging backward underneath the convex ribs is in fact the front of the spine, with the vertebral bodies, whereas looking toward the concave side of the curve apex clearly shows the lordosis.

Returning to the biomechanics of forward bending, the axis of spinal-column rotation normally passes in front of the thoracic kyphosis and behind the cervical and lumbar lordoses (Fig. 3.10). This confers protection to the thoracic spine against buckling, because this region of the spine is normally under tension. With the development of a thoracic lordosis, however (Fig. 3.11), the vertebral bodies move progressively further forward, toward and in front of
This axis of rotation, making them very vulnerable to buckling and explaining the increased rotational prominence seen on forward bending in idiopathic scoliosis (Fig. 3.9).65

If one compares the true lateral view of the apex of the idiopathic thoracic scoliotic curve with a lateral view of Scheuermann’s kyphosis, they would appear to be opposite directional deformities in the sagittal plane (Fig. 3.12). Thoracic hyperkyphosis is, however, progressively further behind the axis of spinal-column rotation, and therefore progresses solely in the sagittal plane (Fig. 3.13). However, it is well known that more than two-thirds of patients with Scheuermann’s idiopathic thoracic hyperkyphosis have an idiopathic scoliosis below this deformity, and this is where the lumbar hyperlordosis, which exists to balance the thoracic hyperkyphosis above, buckles to produce Scheuermann’s disease and idiopathic scoliosis in the same spine (Fig. 3.14).66

The distribution of thoracic kyphosis is probably Gaussian, with patients at the flat end of the spectrum in danger of developing idiopathic scoliosis and those at the round end of the spectrum in danger of developing Scheuermann’s disease. The nature of the distribution would be confirmed by idiopathic scoliosis and Scheuermann’s disease having similar familial relationships and community prevalence rates.67

Considering the spine as the engineer’s beam or column, it can be confirmed that the column is subject to only two primary modes of failure: angular collapse (kyphosis) and beam buckling (lordoscoliosis) (Fig. 3.15). Furthermore, engineers have established laws of the behavior of flexible columns, the critical load being decreased by: (1) increased curvature; (2) increased length; and (3) increased intrinsic load.65 The greater the curve becomes, the more likely it will progress, as studies of the natural history of idiopathic scoliosis have clearly shown68 (the further the Leaning Tower of Pisa leans, the more it will be likely to fall down). Girls with idiopathic scoliosis are significantly taller than age-matched counterparts even when their spinal deformity has not been “uncoiled.”69,70 The concept of an increased intrinsic load refers to a situation in which the spinal column is weakened, and here one can bring in some of the other parts of the classification of spinal deformities. Thus, for instance, neuromuscular scoliosis occurs because the neuromuscular support to the spine is inadequate (Fig. 3.16A), whereas in neurofibromatosis or osteogenesis imperfecta, the more dystrophic the bone the greater the prevalence of structural scoliosis and the earlier its onset (Fig. 3.16B). With Marfan syndrome or Ehlers–Danlos syndrome the spine fails at the soft-tissue level (Fig. 3.16C).

The differences between scoliosis and kyphosis would appear to be very obvious, particularly with the established clinical conditions of, for example, 60 degrees of thoracic scoliosis and 60 degrees of thoracic hyperkyphosis, but the changes are much more subtle than that. The upper and lower thoracic vertebrae are either straight or are parts of the cervical or lumbar lordoses, leaving about eight real thoracic vertebrae. A figure of ~24 degrees would be reasonable for the thoracic kyphosis in early adolescence, and each of the eight vertebrae would therefore be kyphotically wedged by something of the order of 3 degrees. It is necessary to lose only a little more than 3 degrees of kyphosis to create lordosis and the danger of buckling into a lordoscoliosis (Fig. 3.17).65 Because these changes are so subtle, it should not be any wonder that school screening programs have demonstrated that 2.2% of girls aged 12 to 14 years have idiopathic scoliosis (a lateral curvature in excess of 10 degrees with rotation).71

Both Willner and Johnson in Sweden72 and the Leeds Group61 have shown that the thoracic kyphosis changes considerably during growth. It is at a minimum at about the age of 10 years before going up to its maximum of 30 to 40 degrees or so at the age of 15 years. Girls grow fastest between the ages of 10 or 11 years, when the thoracic kyphosis is at its minimum, and if they overgrow (a feature of the development of spinal deformities), they will therefore...
Fig. 3.11  (A) A lateral view of a growing spine with a biomechanically unstable lordosis. (B) The banister rail outside the operating theater in St. James’s University Hospital, Leeds. The banister rail makes a lordosis at each floor level, causing the black plastic handrail to buckle.

Fig. 3.12  (A) True lateral X-ray view of an idiopathic thoracic curve. (B) Lateral X-ray view of Scheuermann’s idiopathic thoracic hyperkyphosis. These are opposite deformities in the sagittal plane.
Fig. 3.13  (A) The center of gravity of the body lies just in front of the lumbar spine, and with hyperkyphosis the thoracic spine is therefore progressively behind the axis of spinal column rotation. (B) Consequently, the deformity progresses solely in the sagittal plane, with no buckling potential.

Fig. 3.14  (A) Lateral X-ray film of a boy with Scheuermann’s disease. (B) PA X-ray film showing that the compensatory lumbar hyperlordosis has buckled to produce idiopathic scoliosis below the area of Scheuermann’s disease.
Fig. 3.15  A column or beam can fail in only two ways: angular collapse (kyphosis) or beam buckling (lordoscoliosis).

Fig. 3.16  (A) Scoliosis in association with poliomyelitis. The spine has failed at the neuromuscular level. (B) Scoliosis in association with osteogenesis imperfecta. The spine has failed at the bone level. (C) Scoliosis in association with Marfan syndrome. The spine has failed at the soft-tissue level.
be vulnerable to the development of idiopathic scoliosis (Fig. 3.18). Boys do not go through their growth spurt until much later, when the thoracic kyphosis is maximizing, which is why boys are more vulnerable to the opposite condition of idiopathic thoracic hyperkyphosis (Scheuermann’s disease) (Fig. 3.19).

That a thoracic lordosis is the primary event in the generation of idiopathic thoracic scoliosis was conclusively shown in the Leeds epidemiological survey. A sensitive positive test of an angle of trunk inclination of 5 degrees or more was the criterion for admission to the study, and of the 16,000 Leeds schoolchildren surveyed, 1000 were harvested and subsequently radiographed on an annual basis for 6 years with AP and lateral low dose films. With such a sensitive entry criterion many children had straight backs to begin with, but some developed true idiopathic scoliosis during the course of the study. This afforded the opportunity of going back to look at the lateral profile when the spine was straight in the frontal plane, and children who developed idiopathic scoliosis already had a flat thoracic spine with an apical lordosis (Fig. 3.20).

Transverse plane geometry is also important in the normal as well as the scoliotic spine. This became apparent when the specimens of idiopathic scoliosis in the Royal College of Surgeons of Edinburgh Museum were first examined. More detailed studies of the same specimens confirmed this. In the cervical and lumbar regions, where the spine is naturally lordotic, the cross-sectional vertebral shape is prismatic, with the base pointing anteriorly. This is most obvious in the lumbar region where the vertebrae in cross-section are typically described as broad and kidney-shaped (Fig. 3.21). When prisms are flexed toward their bases, they are much more stable because of the second moment of area, and the potentially vulnerable cervical and lumbar lordoses are therefore countered by having a stable transverse-planar shape. By contrast, vertebrae in the thoracic region are typically heart-shaped in the transverse plane, with the apex of the prism pointing anteriorly. This is
a dangerous configuration, favoring buckling with flexion, and the thoracic spine is therefore protected by having a safe kyphosis in the sagittal plane.

However, more thoracic curves are convex to the right and more lumbar curves convex to the left, and this is because of a pre-existing asymmetry of vertebral shape in the transverse plane. Anatomists have shown that in the thoracic spine, the T4 to T9 vertebrae are constantly deformed on the left side by the descending thoracic aorta (Figs. 3.21C and 3.22), whereas a dynamic form of transverse-planar asymmetry exists in the lumbar region, where the left-sided abdominal aorta provides a restraint on curves tending to go to the right (Fig. 3.23). This has been confirmed more recently with axial computed tomography (CT) scans of normal human spines. Not surprisingly, the situation was opposite this in individuals with situs inversus.

Clearly, however, the preponderance of right-sided thoracic curves and left-sided lumbar curves does not equate with the prevalence rate of situs inversus. This is because among small thoracic curves (<20 degrees), more are left-sided than right-sided, and because lumbar curves do not really have a left predominance until they reach or exceed 15 degrees, according to data of the Oxford School Screening Study (Table 3.1).
Experimental Scoliosis

Following Adams’s original dissections of cadavers with idiopathic scoliosis, showing the essential lordosis at the curve apex, Somerville in Oxford produced an experimental model of progressive idiopathic-type scoliosis in the growing rabbit. He tethered the back of the spine into lordosis and then performed a soft-tissue release posteriorly on one side to direct subsequent buckling that would cause the typical lordoscoliotic deformity seen in patients with idiopathic scoliosis. Our group in Leeds conducted extensive experimental work that consistently caused an idiopathic-type deformity in rabbits by producing an asymmetrical lordosis similar to what Somerville achieved with his methods (Fig. 3.24). Importantly, no buckling occurred if the lordosis wasn’t given directional instability. Moreover, if the lordosis was released before the end of spinal growth, and the deformity had not progressed beyond approximately 20 to 30 degrees, the spine grew straight again, suggesting that addressing the sagittal-plane component of the scoliotic deformity in children might be beneficial. If, however, the tether is not released, the deformity progresses relentlessly (Fig. 3.25).

We also extensively studied the three-dimensional nature of the deformity in idiopathic scoliosis and in particular its transverse-plane component in both animals.

Fig. 3.21  **(A)** In the cervical region and **(B)** the lumbar region, vertebral shape in the transverse plane resembles a prism with its base facing anteriorly. **(C)** In the thoracic region, however, the shape of the transverse plane resembles a prism with its apex pointing anteriorly, which is a much more unstable configuration. Moreover, it can be clearly seen that midthoracic vertebrae are asymmetric, being flattened on the left by the descending thoracic aorta, thereby putting the apex of the prism to right of the midline.
and humans. We clearly found the most asymmetrical vertebra at the curve apex, where the pedicle on the convex side was short and stout and that on the concave side was long and slender (**Fig. 3.26**). The transverse-plane geometry changed above and below the apex of the curve, first becoming neutral before becoming the opposite in the compensatory kyphoses that balance the central lordotic

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**Fig. 3.22**  CT scan at the T8 level, showing transverse-plane asymmetry caused by the descending thoracic aorta.

**Fig. 3.23**  In the lumbar region the abdominal aorta is to the left of the midline and thus rests against the left side of the base of the lumbar prism, favoring left-sided rotation in the lumbar spine.

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**Table 3.1** Direction of Idiopathic Curves with Curve Size

<table>
<thead>
<tr>
<th>Curve Size</th>
<th>No. of Curves</th>
<th>Direction (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>R</td>
<td>L</td>
</tr>
<tr>
<td>Thoracic</td>
<td></td>
<td></td>
</tr>
<tr>
<td>5–9</td>
<td>93</td>
<td>38</td>
</tr>
<tr>
<td>10–14</td>
<td>36</td>
<td>47</td>
</tr>
<tr>
<td>15–19</td>
<td>9</td>
<td>33</td>
</tr>
<tr>
<td>20+</td>
<td>6</td>
<td>67</td>
</tr>
<tr>
<td>Total</td>
<td>144</td>
<td>41</td>
</tr>
<tr>
<td>Lumbar</td>
<td></td>
<td></td>
</tr>
<tr>
<td>5–9</td>
<td>50</td>
<td>28</td>
</tr>
<tr>
<td>10–14</td>
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<td>43</td>
</tr>
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<td>15–19</td>
<td>8</td>
<td>-</td>
</tr>
<tr>
<td>20+</td>
<td>6</td>
<td>-</td>
</tr>
<tr>
<td>Total</td>
<td>94</td>
<td>29</td>
</tr>
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</table>

**Fig. 3.24**  When the growing rabbit spine is tethered into lordosis (**A**), a progressive lordoscoliosis develops over the next few weeks (**B**). If the tether is released at this stage, in which there is a mild curve, the spine will subsequently straighten with growth because the Heueter–Volkmann Law has not irreversibly deformed the apical vertebrae.
Fig. 3.25  With further growth the scoliosis progresses (A) at 6 weeks and (B) at 12 weeks. (C) Axial CT scan of the spine in B, showing significant rotation at the curve apex. (D) Looking inside the chest of this specimen shows exactly the same changes as during anterior thoracic surgery for idiopathic scoliosis. There is no way in which this deformity can be created experimentally other than through rotation of a primary lordosis.

area. In these regions the pedicle on the convex side was long and slender and that on the concave side short and stout.

The same pattern of apical vertebral-body deformation was seen in the rabbit as in the human, and by labeling areas of active vertebral growth with a dye similar to tetracycline, we observed bone drift toward the curve concavity, indicating that the spine was trying to correct the deformity imposed upon it (Fig. 3.27).81

When the segmental blood supply to the spinal cord was occluded at the curve apex, a cord infarct was produced, and this led to a significant deformity, in excess of 40 degrees, as soon as the procedure was completed, resembling what was observed in experimental neuromuscular scoliosis.
Fig. 3.26  Transverse-plane asymmetry at the curve apex, with a short, stout pedicle on the convex side and a longer, thinner pedicle on the concave side. (A) Human, (B) rabbit.

Fig. 3.27  (A) The diagram in the center shows that growth of a normal vertebra in terms of the spinal canal and the vertebral body is outward. Consequently, the orange-stained growth area in the canal above and the vertebral body below is facing outward.
This is how Langenskiold and Michelsson accidentally produced scoliosis by dividing the costotransverse ligament, because they damaged the segmental blood supply to the spinal cord, as De Salis and colleagues demonstrated. Interestingly, growth and pulmonary function were considerably impaired with a rapidly progressive thoracic deformity, rather like the situation in progressive infantile idiopathic thoracic scoliosis (Fig. 3.29). Much interest in experimental scoliosis was rekindled by observations of pinealectomized chickens and rats popularized by Dubousset et al and Machida et al. The pineal gland produces melatonin from tryptophan through a series of enzyme reactions, and serotonin is intermediary in this pathway. In 1959 Thillard first produced scoliosis in pinealectomized chickens to assess the role of melatonin and its associated compounds in the disorder. If chickens are pinealectomized shortly after they hatch, a scoliosis similar to human idiopathic scoliosis is consistently produced. If melatonin supplements are given after pinealectomy, a scoliosis does not develop. The precise reason why pinealectomy produces this deformity is uncertain, and research translated to the human situation has shown conflicting results with regard to melatonin levels in patients with idiopathic scoliosis and those in controls, with some careful studies involving diurnal variation showing no differences in the two groups. It is thought that melatonin activity may be mediated by growth hormone as the common denominator.

However, the biomechanics of this experimental model are also interesting. Even with the pinealectomized animal model it is accepted that the “primary abnormality is a lordosis,” which subsequently buckles to produce the typical three-dimensional lordoscoliotic deformity, as confirmed by Machida. This does not occur spontaneously in quadrupeds, and chickens are bipedal. Consequently, Dubousset and Machida and colleagues went on to investigate the effects of pinealectomy in rats. If rats were initially rendered bipedal and then pinealectomized, they developed a scoliosis comparable to that in chickens, whereas the spine remained straight in rats that underwent a sham operation after being rendered bipedal (Fig. 3.30). Quadrupedal rats when pinealectomized did not develop a spinal deformity.
Fig. 3.28  (A) Experimental scoliosis. PA radiograph of a rabbit spine. The spine has been tethered into lordosis and the spinal cord damaged by thermal ablation of the facet joints. There was a curvature of 70 degrees immediately after the animal awakened from anesthesia. (B) Within a couple of weeks the deformity was gross, as in progressive infantile malignant idiopathic scoliosis. (C) Transverse section of the spinal cord at this level showing a dorso-lateral infarct.

Fig. 3.29  Two rabbits, one with experimental idiopathic scoliosis (normal eye) (A) and the other (B) with experimental neuromuscular scoliosis, resembling progressive infantile idiopathic malignant scoliosis with respiratory malfunction and cyanosis (cyanotic eye).
In other words, the effect of being bipedal was to exaggerate the existing thoracic lordosis, but no buckling into a lordoscoliosis was produced unless the bipedal rat was pinealectomized, suggesting that the hyperlordosis rendered by the upright posture was destabilized by pinealectomy to produce the scoliosis.

Furthermore, a scoliosis was more easily produced when the tails of bipedal rats were removed, allowing them to have a more upright posture. The sagittal profiles of these rats showed that the pinealectomized quadrupedal rat had a physiological thoracic lordosis, whereas a thoracic hyperlordosis was produced in both the sham-operated and pinealectomized bipedal rats. In other words, the effect of being bipedal was to exaggerate the existing thoracic lordosis, but no buckling into a lordoscoliosis was produced unless the bipedal rat was pinealectomized, suggesting that the hyperlordosis rendered by the upright posture was destabilized by pinealectomy to produce the scoliosis.
Interestingly, having tethered rabbits’ spines into a lordosis, neither Somerville nor the Leeds Group could make it buckle unless the lordosis was rendered asymmetrical by producing a few degrees of scoliosis as well. Perhaps pinealectomy would have done the same. In the bipedal chickens and rats that developed scoliosis, there was no preferentiality in its developing either on the right or the left side.

There doesn’t seem to be any other explanation for the effect of the pinealectomy performed on rats, because the rats were of much the same weight at the end of the experiment and were not constitutionally disadvantaged. However, scoliosis was also noted in the thoracolumbar region, and this is where the lateral radiographs clearly showed a kyphosis of the order of 40 or 50 degrees. Perhaps these were the slightly asymmetrical kyphoses seen with severe Scheuermann’s disease, in which there is a concomitant mild coronal–plane deformity with the opposite direction of rotation to that in idiopathic scoliosis, with the vertebral bodies turning into the curve concavity. This is simple right–left growth asymmetry rather than mechanical buckling.

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