Studies on idiopathic scoliosis*

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In many patients with idiopathic scoliosis, the deformity increases only up to a moderate degree and then remains stable. Even with advanced vertebral curvatures, it is frequently seen that the body's position is not significantly affected. On the other hand, in other cases idiopathic scoliosis develops fast and produces very severe deformities. In order to establish some grounds for prognosis, we studied the evolution of this condition in 394 cases that were not subjected to surgical treatment. Many of these patients were treated by means of postural exercises and physical therapy, which often succeeded in obtaining postural improvements. Nevertheless, vertebral curvatures do not seem to be amenable to any kind of conservative treatment. In our study we found that the prognosis of idiopathic scoliosis can be based on: 1) the type of curvature; 2) the patient's age at the onset of scoliosis; and 3) osteoporosis of the vertebrae and abnormalities in the intervertebral spaces, which are apparent in the x-rays.

1. Type of curvature.–In idiopathic scoliosis the characteristics of the curvatures remain constant throughout the progression of the deformity. Nor do any changes occur as regards the apex and the location of the curvature and the direction in which the vertebrae rotate. The very initial x-rays already show the type of scoliosis the patient is going to suffer. All cases of idiopathic scoliosis can be grouped into five main types, i.e., lumbar, thoracolumbar, combined thoracic and lumbar, thoracic and cervicothoracic curvatures. 5% of patients show combinations of these five main types.

Lumbar curvatures in general comprise five vertebrae, from the eleventh thoracic to the third lumbar vertebrae. The apex is located at the first or second lumbar vertebra. 93 cases of the 394 cases studied (23.6%) had this type of scoliosis. These are the most benign idiopathic curvatures, which often go undetected until the age of 13 and become stabilized at age 15 (fig. 1.a).

Thoracolumbar curvatures comprise from 6 to 8 vertebrae and extend from the sixth or seventh thoracic to the first or second lumbar vertebrae. The apex is located at the eleventh or twelfth thoracic vertebra and the convexity of the curvature is, in general, towards the right. We found this type of scoliosis in 16% of our cases. In general it is not detected until the age of 13 and tends to progress until age 16. Body posture does not appear overly deformed, except in those unusual cases in which the curvature starts developing during childhood. In those cases it can develop considerable and produce significant deformities (fig. 2.a).

Combined thoracic and lumbar curvatures are the most common. We observed them in 37% of our cases. This scoliosis has two main curvatures: a thoracic curvature that extends from the sixth to the tenth thoracic vertebra and has its apex at the seventh thoracic vertebra; the lumbar curvature takes the opposite direction and extends from the eleventh thoracic to the fourth lumbar vertebra. The apex is located at the second lumbar vertebra. The convexity of the thoracic curvature is, in general, towards the right, and that of the lumbar one, towards the left. This scoliosis appears around the age of 12 and progresses until slightly before age 16. As there are two similar curvatures of opposing direction that compensate each other, body posture does not appear excessively altered (fig. 3.a).

Thoracic curvatures comprise, in general, 6 vertebrae, and extend from the sixth to the eleventh thoracic vertebra. The apex is located at the eighth or ninth thoracic vertebra and vertebral rotation is sharply marked. These curvatures were found in 22% of our cases. This type of scoliosis tends to start at an early age and around age 11 are already at an advanced stage, becoming increasingly worse until a few months after age 16. In most cases, the curvature in this type of scoliosis has its convexity towards the right and produces a sharply marked thoracic deformity (fig. 4.a).

We only found 5 cases of cervicothoracic curvature. The apex is located at the third thoracic vertebra and the curvature extends from the seventh cervical to the fourth or fifth thoracic vertebrae. Our patients were already 15 years of age when the deformity was detected, an in none of the cases did it progress too much.

Thoracic curvatures are, therefore, the most dangerous ones since they can progress much further than the others and produce severe deformities. The prognosis of lumbar, thoracolumbar and cervicothoracic curvatures is generally favorable. Combined thoracic and lumbar curvatures have, in general, a good prognosis if they start developing after age 10, and a poor prognosis if they start before.

2. *Age of the patient at the onset of scoliosis*.— Idiopathic curvatures almost inevitably progress during the period of development of the spine and stop developing at the end of puberty. The center of ossification of the iliac

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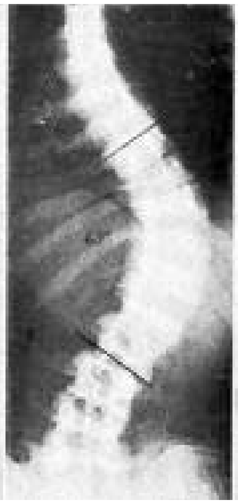

Fig. 1.[a] *Fig. 2.*[a] *Fig. 3.*[a] *Fig. 4.*[a]

Figure 1. *Spine radiograph of a 17-year-old girl showing lumbar curvature. The deformity became noticeable at age 12.*
Figure 2. *X-ray of a 17-year-old girl with thoracolumbar curvature. The deformity became noticeable at age 11.*
Figure 3. *Combined thoracic and lumbar curvatures in a 17-year-old girl. The deformity became noticeable at age 10.*
Figure 4. *Thoracic scoliosis in a 17-year-old girl. The deformity started showing at age 13.*

processes is a good index of vertebral growth. The spine stops growing once the ossification of these processes is completed.

In general, scolioses that develop during childhood end up producing severe deformities (fig. 5.a). On the other hand, if scoliosis develops after skeletal maturity, progression is very slight or even inexistent, only giving rise to minor deformities. The patient's age at the onset of scoliosis varies as a function of the type of curvature. Thoracic curvatures appear, in general, at an earlier age than the others. Lumbar, thoracolumbar and cervicothoracic scolioses appear later, often after age 13.

Idiopathic scolioses are much more frequent in females than in males. Nevertheless, in cases with thoracic curvatures there are quite a few men. We had 87 cases with these types of curvatures, 25 of which were in males.

3. *Vertebral osteoporosis and irregularities of intervertebral spaces.*— Vertebrae adjacent to the apex of dorsal curvatures seem to be the most severely affected. Shortly after scoliosis started progressing, vertebral bodies already showed osteoporosis and malalignment and soon formed into the shape of a wedge (fig. 6.a). Intervertebral spaces are irregular and narrow. There is a direct relationship between the intensity of these changes and the increase in the curvature. Vertebral osteoporosis disappears a few months before the curvature stabilizes. For this reason, vertebral changes are inconsequential from a prognostic point of view. The bone structure of lumbar vertebrae has quite a normal appearance even in the most severe lumbar curvatures.

From these Studies we learned to recognize that malignant idiopathic scolioses are, in general, those of the thoracic type, which are always accompanied by severe spinal changes that point to a metabolic origin. For this reason, we have spent the last three years, together with doctor Genevieve Stearns, studying the metabolism of a large group of patients with progressive dorsal idiopathic scoliosis dorsal. These studies have shown that the metabolism of minerals, hydrocarbonates and lipids is normal. On the other hand, protein metabolism remains abnormal for as long as scoliosis progresses.

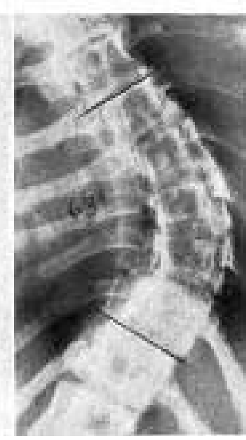

Fig. 5.[a] *Fig. 6.*[a]

Figure 5. *Thoracic scoliosis in a 15-year-old girl. The deformity became noticeable at age 6.*
Figure 6. *Osteoporosis and irregularity of the intervertebral discs in the wedge-shaped vertebrae of a thoracic curvature.*

It6 is interesting to see that the only known laboratory procedure capable of inducing scoliosis in animals is a diet with certain pulses. The Spanish school of Professor Jiménez Díaz has worked extensively on this aspect of the problem since lathyrisim is a fairly common problem in certain areas of Spain. Lathyrism epidemics have been a common occurrence for many years in regions where food scarcity has forced the population to consume large amounts of legumes. Humans and animals with lathyrism have, among other symptoms, weakness and spasticity in their lower limbs and urinary and fecal incontinency. Rats and other animals with lathyrism are apt to develop scoliosis.

There are many similarities between the radiographic characteristics of scoliosis in rats with lathyrism and idiopathic scoliosis in children. Induction of scoliosis in animals offers an excellent opportunity to perform a hystopathologic (and maybe pathogenic) study of this disease, which has not been researched into previously.

For our study, we used four-week old white rats. One group of rats was given the following diet:

Lathyrus odoratus flour	50 per 100
Corn starch	28
Sucrose	6
Saline mixture	4
Corn oil	2
Dry yeast	10
Halibut oil, 0.2 cc per 100 g	

Another Group of rats was given the diet above, plus 10% casein, and a third group was given this plus 0.75% methionine. Rats were weighed every day and x-rays taken every week with the animals under ether anesthesia. Rats were put down with ether, at intervals, after having developed scoliosis. All animals were autopsied.

Radiographs taken two or three weeks after starting a diet with *Lathyrus odoratus* show generalized skeletal demineralization. Shortly afterwards, areas of new periosteal bone formation appear, mainly at the level of the femoral metaphysis. Long bones soon appeared arched and deformed. From the fourth to the sixth week into the diet intervertebral space irregularities and narrowing were observed at the level of the last dorsal and the first lumbar vertebrae. Soon afterwards, a slight forward displacement is observed at this same level of one vertebra over the other. A week later, a collapse of two or three vertebral bodies is seen at this level, which results in kyphosis. One or two weeks later, there is lateral displacement and rotation of the vertebrae, which produces thoracolumbar kyphoscoliosis (figs. 7.a y 8.a).

Two or three weeks later, similar changes occurred in the fourth to eighth thoracic vertebrae, resulting in thoracic kyphoscoliosis with rib deformation. All rats developed progressive scoliosis, with the severity of the curvature related to the time during which the experimental diet was administered. Thoracolumbar scolioses with convexity to the right are the most usual. Lumbar curvatures are rare.

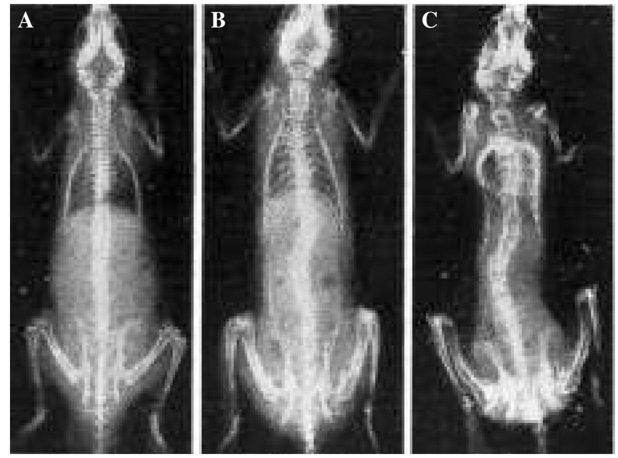

Figure 7. *Rat I AL. Posterior-anterior x-ray:* A) *at 11 weeks of age, there is slight lumbar scoliosis towards the left, generalized skeletal osteoporosis and ligated curvatures of the femora;* B) *at 15 weeks of age there is an increase of lumbar scoliosis and femoral curvature. Periosteal ossifications can be seen along both femora;* C) *at 14 weeks, thoracic scoliosis can be seen towards the right accompanied by rib deformity. Periosteal ossifications along the femora and the lumbar vertebrae.*

When rats with well developed kyphoscoliosis are given a normal diet, skeletal reossification occurs in two to three weeks. Deformed long bones straighten up and periosteal ossification disappears. Kyphoscoliosis stops progressing but vertebral curvatures do not go away but rather stay in the same condition as they were when the diet was changed.

Addition of casein and methionine to the diet with *Lathyrus odoratus* brings about faster growth in the rats, but

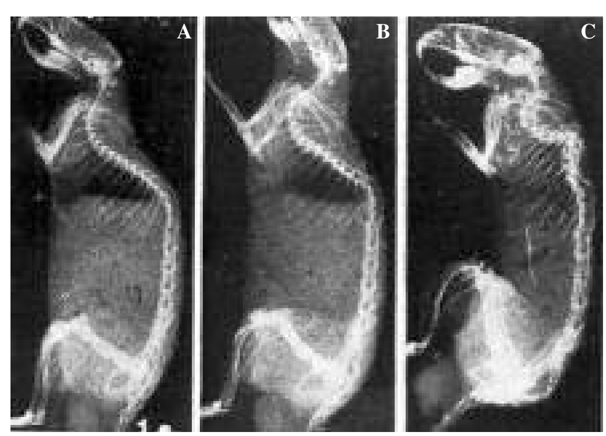

Figure 8.a *Rat I AL. Lateral x-rays:* A) *At 11 weeks, narrowing of intervertebral spaces at the level of the thoracolumbar region;* B) *at 15 weeks of age, sliding of the 11th thoracic vertebra over the 12th. Slight kyphosis can be seen at this level;* C) *at 24 weeks, marked kyphosis can be seen.*

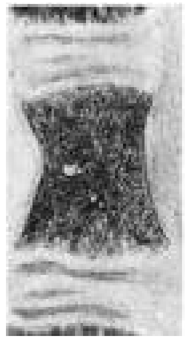

Fig. 9.ª Fig. 10.ª Fig. 11.ª

Figure 9. *Photomicrograph of a vertebra showing osteoporosis and cortical thinning. (from a rat put down at the age of 12 weeks).*
Figure 10. *Photomicrograph of the lower thoracic vertebrae, showing the collapse of the anterior portion of the vertebral bodies and a forward sliding of the upper over the lower vertebra. (this rat died of a dissecting aortic aneurysm at 13 weeks of age).*
Figure 11. *Photomicrograph of the thoracic vertebrae of a rat of 18 weeks of age. Vertebral sliding and intervertebral space irregularity are at an advanced stage.*

does not alter the development of osteoporosis or the spinal deformities. These changes are therefore not due to protein deficiencies but rather to a toxic factor in the diet.

A microscopic analysis of the skeletal system reveals at the beginning generalized osteoporosis. Growth cartilage does not seem to be overly affected and endocondral ossification proceeds almost unaltered. The cortex of the vertebrae is very thin and the bone trabeculae of the vertebral body are scarce and very thin (fig. 9.a). The bone marrow has a normal appearance. No abnormal amounts of osteoid tissue are to be observed. Similar changes are seen in the long bones, in whose metaphyseal areas there is abundant subperiosteal bone formation. The periostium is thick and highly cellular, and covers a layer of highly vascular connective tissue with irregular areas of incipient intermembranous bone (fig. 14.a). Similar periosteal bone formation areas can be observed on the anterior aspect of some vertebrae; the new trabeculae are arranged perpendicularly to the vertical vertebral axis.

At the beginning no final histological changes can be observed in the intervertebral discs. The pathogenesis of scoliosis seems to be as follows: as a result of severe osteoporosis the anterior portion of the vertebral bodies collapses. This happens, in general, at the anterosuperior portion of the vertebral body, below the growth plate. Because of this collapse, the upper vertebra slides forward over the caudal vertebra. This displacement occurs, partly, in the intervertebral disc, but it is intensified by the collapse of the caudal vertebra. Subsequently, there may be a certain degree of bone hypertrophy at the site of the collapse, which does not alter the evolution of the deformity. In the studies on the most severe cases of scoliosis, the vertebral bodies involved appear wedge-shaped and highly osteoporotic, without signs of new bone formation. Masses of fibrous tissue partly replace the bone in the vertebral body. In these cases intervertebral discs show large distortions and are extremely narrow (figs. 10.a and 11.a).

Three months after the start of the experimental diet, when the kyphoscoliosis is already highly advanced, some animals develop paraplegia and drag their hind limbs along. Autopsy shows medullary compression at the level of the kyphosis, where scar tissue can be seen attached to the dura

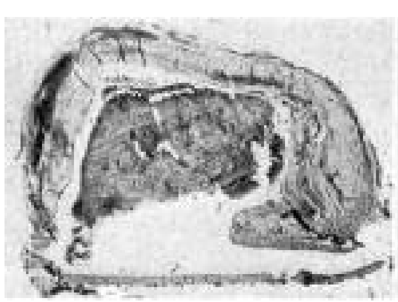
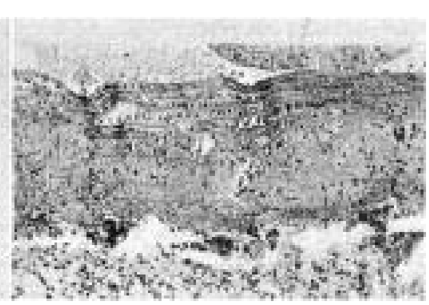

Figura 12.ª Figura 13.ª

Figure 12. *Photomicrograph of a saccular thoracic aorta aneurysm.*
Figure 13. *Photomicrograph of the aortic arch wall, showing necrosis and separation of the elastic fibers. This rat died of a dissecting aortic aneurysm at 11 weeks of age.*

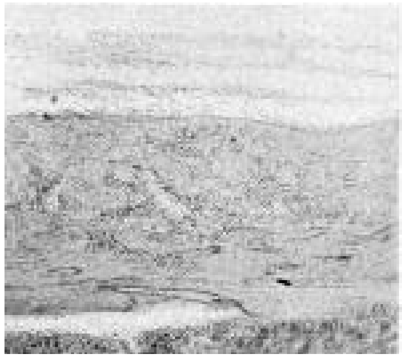

Fig. 14.ª

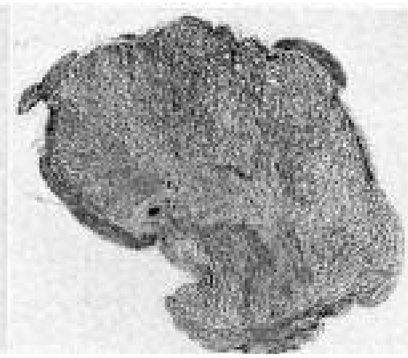

Fig. 15.ª

Figure 14. *Photomicrograph showing periosteal ossification along a femur.*
Figure 15. *Rat I AL (put down at 24 weeks of age). Photomicrograph of the spinal cord at the level of a severe kyphoscoliosis. The posterolateral region of the cord was compressed.*

mater on the posterior or lateral side of the spinal cord. Histological spinal cord sections at this level show a severe distortion of the posterior funiculi and horns. Many of the cells of the posterior horns and some of the cells of the anterior horns are missing. Gliosis is observed in the posterior funiculi as well as a loss of myelin throughout the white matter. Above and below the compression level the spinal cord has a normal appearance. Histologically, there are no abnormal findings in the spinal cord of animals without paraplegia (fig. 15.a).

25% of rats died one or one and a half months after starting the diet, when kyphoscoliosis was minimal or nonexistent. In some of these rats no injury was found to account for their death, but the majority show either hemopericardium or a bilateral hemothorax. A microscopic aortic examination of these animals showed necrotic areas in the tunica media, mainly at the level of the aortic arch. Thus dissecting aneurysms are formed in the aorta with necrosis of the elastic fibers (fig. 13.a). Finally, the blood propagates to the mitral valve where large hematomas are formed and the vessel breaks. Sometimes, if the toxic diet is discontinued after one or one and a half months and replaced by a normal diet, when the experimental diet is resumed after a few months it is possible to see the formation of saccular aneurysms in the aortic arch (fig. 12.a). No atheromatous plaques are observed in the vessels.

Histological sections of the other thoracic and abdominal organs do not reveal any significant lesion. The skeletal muscles appear histologically normal. However, from the second week into the experimental diet, the musculature of these rats consumes more oxygen than the musculature of the control rats.

From these Studies one could infer that the seeds of *Lathyrus odoratus* contain a toxic factor that affects the formation of the bone organic matrix, the aortic elastic and collagenous fibers and possibly other mesodermal structures.

When a careful study is made of the diet of patients with idiopathic scoliosis, one sees that protein intake is poor throughout the growth period. These patients seem to be averse to protein and there is a direct relationship between aversion to protein and the severity of scoliosis. Although some of these patients are overweight, the majority have flaccid musculature and are indifferent or apathetic.

Nitrogen retention is poor in all children and adolescents with idiopathic scoliosis. Protein digestion and absorption in the digestive tract is not abnormal, but a certain amount of protein is lost because of the increased secretion of urea per gram of protein assimilated; there is also a sharp increase in the total aminoacid excretion in urine. Likewise, aminoacid distribution in urine is abnormal. These metabolic abnormalities disappear when scoliosis stops progressing.

In cases of severe progressive scoliosis, the abnormal elimination of aminoacids continues and it increases when a protein-rich diet is administered. Conversely, in moderate cases an increase is seen in protein retention and the progression of the deformity seems to halt. In the last year we have tried to increase protein retention by administering vitamin B12 and testosterone. Too little time has elapsed to be able to provide details about the results of these studies.