NATURAL HISTORY AND PATHOGENESIS OF IDIOPATHIC SCOLIOSIS

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ABSTRACT :

In order to set forth a treatment plan of idiopathic scoliosis, predictors of progression should be known. Progression factors are well studied in the literature, however, still one can not pre-determine precisely which curve will progress or up to what extent it will progress. Family history, height-weight ratio, lumbosacral transitional anomalies, thoracic kyphosis, lumbar lordosis, balance are found to be non-prognostic factors, whereas, gender, curve pattern, curve severity, age at diagnosis, menarch, Risser sign are considered as prognostic factors. The most important of these are, curve magnitude, Risser sign, skeletal age at diagnosis.

Theories concerning the etiology and pathogenesis of idiopathic scoliosis are numerous. As yet, none is proven to be correct. Recent well accepted concepts are biplanar asymmetry (Leeds group) and Nottingham concept based on the "pathology" of central pattern generators of central nerveous system.

In this review article, issues of progression and etio-pathogenesis are undertaken.

Key Words : *Progression factors, biplanar asymmetry, Nottingham concept.*

Increasing use of schoolscreening for the early detection of spinal deformities shows that a large number of patients with idiopathic scoliosis that have a minimum curve (16). In order to evaluate this curves and make a rational treatment program, we have to know which factors will influence these curves. For this reason we have to answer some of these questions : (15, 2, 7, 19, 20)

1. How many of these minimum curves will progress? (What's the incidence of progression?)
2. What factors are related to progression of the curve?
3. Is it possible to use the factors in order to predict which curve will progress and which will not?

In 1974; Clarisse (6) attempted to distinguish the progressive from the non-progressive curve by investigating 110 patients with 10 to 29 degree idiopathic curves presenting during growth. Patients were not treated unless the curve progressed above 30 degrees. The patients presenting between the ages of 3 and 11 showed progression 53 percent. The progression rate was 15 percent between the age of 11 and the onset of menses. Also Clarisse found that thoracic curves progressed in 42 percent of the patients whereas 12 percent at lumbar and 67 percent at thoracolumbar curves.

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Bunnell (2) investigated 123 patients who met the following criteria :

1. The presence of a curve showing less than 50 degrees at the time of the diagnosis.
2. A radiograph taken at skeletal maturity.
3. A minimum of one year follow up of one year prior to skeletal maturity.

In this study it was shown that the risk of at least 5 degrees of progression was correlated with the curve pattern. 77 percent of thoracic, 67 percent of thoracolumbar, 66 percent of double major, 30 percent of lumbar curves progressed. Patients diagnosed prior to age 10 had a progression of 10 degrees and more, while patients older than 15 years when diagnosed had a 14 per cent risk. Patients diagnosed prior to menarche progressed in 53 percent of cases, whereas only 11 percent of patients showed progression after menarche. Patients had Risser 0 had a 68 per cent of progression, while Risser 3-4 had an 18 per cent of progression. According to this study Bunnell explained the prognostic and non prognostic indicators of curve progression :

Non-Prognostic factors :

1. Family history.
2. Height - weight ratio.
3. Lumbosacral transitional anomalies.
4. Thoracic kyphosis

5. Lumbar lordosis
6. Balance

Prognostic factors :

1. Gender : The curves that have 10 degree or below have the same progression rate between boys and girls, while are greater at girls that have 10-30 degrees. At curves that have a 30 degrees or mare have the same progression rate between boys and girls. According to Bunnell 53 % of boys and 35 % of girls progressed 10 degrees or more before skeletal maturity.

2. Curve Pattern : According to Clarisse (6) the progression rate was 67 % at double major, 42 % at thoracic, 12 % at lumbar curves. Bunnell showed the progression rate 66 %, 77 %, 30 % respectively. He also found that the progression rate at thoracolumbar curves were 67 %.

3. Curve severity : 70 % of curves less than 20 degrees at the time of the diagnosis progressed greater than 5 degrees, and 44 % of these progressed more than 10 degrees. Curvatures of 20-30 degrees progressed 5 degrees in 52 % of patients and 10 degrees or more in 48 %. 78 % of the curvatures of 40-50 degrees at the time of the diagnosis progressed more than 5 degrees and 62 % of these progressed more than 10 degrees.

4. Age at diagnosis : Patients diagnosed before 10 years had a 88 % risk of progression of 5 degrees or more. The risk was the same between the ages of 10 and 12. It was reduced to 56 % for the patients between ages 12 and 15 and to 29 % for those diagnosed after age 15. 76 % of those diagnosed before age 12 progressed 10 or more, as did 33 % of those diagnosed between ages 12 and 15 and 12 % of those diagnosed after age 15.

5. Menarche : 53 % of the patients diagnosed before the onset of menses progressed 10 or more, whereas 11 % of those diagnosed after menarche progressed 10 degrees or more. Also it was noted that there was no correlation between the age at menarche and the risk of the progression but there was a very strong correlation between the age at menarche and the age at which the deformity was noted by an unskilled observer.

6. Risser sign : Patients with Risser 0 at the time of diagnosis had a 68 % risk of progressing 10 or more. This risk was decreased to 52 % in Risser 1 or 2 and 18 % in Risser 3 or 4.

Lonstein and Carlson (15) further investigated the risk factors for curve progression in their 1984 review of 727 patients with idiopathic scoliosis presenting initially 5 to 29 degree curves. The patients were followed until the end of the growth or until curve progression detected. Progression were defined as a curve increase of 10 degrees or more for less than 19 degrees and a curve increase of 5 degrees or more for curves of 20 to 29 degrees. They found 23 % progression rate. In order to predict the progression; they used a formula that included many factors that had a high correlation with progression.

Progression Factor : Cobb Angle – 3 x Risser Sign / Chronological Age.

While no single factor determines curve progression, one is able to predict with some accuracy which curve will progress by combining different pieces of information (18, 19, 20, 25). The most important of these are curve magnitude, Risser sign, skeletal age at diagnosis, secondary are knowledge of curve pattern and menarchal status (18, 25, 27).

ETIOLOGY AND PATHOGENESIS OF IDIOPATHIC SCOLIOSIS

There are many theories have been put forward about the pathogenesis of idiopathic scoliosis, however most of these theories can explain the secondary changes in idiopathic scoliosis. Although there is no clinical evidence of nerve or muscle dysfunction, most workers support this theory. Recent studies show us that there isn't any specific abnormality found at electroencephelographic and laybrinthine studies. After Lerique and Le Coeur in 1951 (14) demonstrated action potentials differences on the two sides of the scoliotic spine, muscle imbalance was believed to be the main cause of the idiopathic scoliosis. The findings at electromyography (1) only show the secondary changes is reported by many authors. These changes are the increased activity of the convex side and this shows us that these are the secondary changes (4). Also it was shown that the increased elektromyographic changes at the convex side of the curve was secondary to the presence of the curve or patient improper positioning. The denervation potentials of the end plates at idio-

pathic scoliosis was found to be the end-plate noise (1, 28).

Collagen abnormalities of the intervertebral discs in the region of the scoliosis is an another theory but these changes are considered to be secondary to the spinal deformity (13, 22). Virus like particles of a glycogenic nature at intervertebral discs is a common finding but they are also shown at normal human vertebra (5, 24). Metachromasia of fibroblasts has been an contributory phenomenon but is has still in suspense. The experimental studies are done by many workers in order to back up neuromuscular theory but it is shown that by dividing the soft tissues on the side of the spine at muscle, ligament or nerve level can cause a scoliosis but this kind of scoliosis is similar to paralytic scoliosis that has little rotation and risk of progression. This suggest that an idiopathic scoliosis has not been produced. By cutting either anterior or posterior nerve root produces poliomyelitis type curve. The division of nerve roots, the costotransverse ligament and rib resection in the rabbit all cause a structural neuromuscular type of scoliosis because of associated blood vessel damage that cause spinal cord damage. De Salis demonstrated that when the rabbit intercostal artery was coagulated at four consecutive levels, a structural scoliosis was developed at two thirds of the patients but histological examinations showed that there were characteristic ischaemic changes at spinal cord which caused abnormal neuromuscular responses in the perevertebral muscles that results scoliosis.

To understand the pathology of idiopathic scoliosis, two important teories are accepted. One of them is the concept of biplanar asymmetry which is developed by Leeds group. (Dickson et al.) The other is Nottingham concept.

Biplanar Asymmetry : A clinical, cadaveric, biomechanical and radiological investigation indicates that biplanar asymmetry is the essential lesion of the idiopathic scoliosis (9, 10, 11). Many children have median plane asymmetry and all have vertebral plane asymmetry in the transverse plane, but when median plane asymmetry (flattening or reversal of thoracic kyphosis at the apex of the scoliosis) is superimposed during growth, an idiopathic scoliosis occurs. According to Leeds theory median (sagital) plane asymmetry is the important factor that causes idiopathic scoliosis and lateral profile of the spine should always be taken into consideration while evaluating the patients. Median plane asymmetry is crucial for progression. It is also noted that increased anterior vertebral height at the apex of the curve with posterior end-plate irregularity characterizes the median plane asymmetry and suggests that idiopathic scoliosis is the reverse Scheuermann's disease.

Idiopathic scoliosis is a lateral curvature of the spine in the absence of congenital spinal anomaly or associated musculoskeletal condition (9). Rotation and progression potential both characterize the clinical deformity. Normal children have neither straight nor symmetrical spines. 15 % of children show evidence of a lateral curvature on a crude visual test and as many as 30 % with a more accurate screening method. It has been known that the patients with thoracic curves show median plane asymmetry in the nature of lordosis at the apex (26). And normal thoracic kyphosis in normal children is reduced in early adolescence (12). Dickson also showed experimentally that a combination of lordosis and asymmetry of the spine produce scoliosis (9, 10).

In this study (9); it was mentioned that there was a significant correlation between the Cobb angle of the scoliosis and that of the overall kyphosis; and the apical lateral Cobb angle was significantly negatively correlated with the angle of overall kyphosis. While the apical vertebrae had spun furthest from the neutral position they were least rotated one to another. Maximum intervertebral rotation occurred at the junctions of the structural curve and its upper and lower compensatory curves. These curves brought the spine neutral above and below. When true anteroposterior and lateral projections of the apex were measured, the mean Cobb angle of scoliosis was increased and there was a mean lordosis at the apex.

In biomechanical studies; it was demonstrated that the effect of flexion of a lordotic segment of the spine with pre-existing coronal plane asymmetry created a force of forward flexion (F). This force tightened the posterior structures and created reactive force (f). The horizontal component of f acting at a distance (d) from the midline would produce a spining moment (M). The magnitude of this spinning moment was therefore a function of the force of forward flexion and the degree of biplanar asymmetry ($M = f . d$)

The investigations at the rabbits showed that if the rabbit's spine flexed beyond the limit, a lateral curvature of the spine would produce a lateral curvature and the thoracic spinous processes are directed towards the convexity. If posterior elements of the rabbits tethered and created mid-thoracic lordosis, a thoracic scoliosis would produce and the spinous processes were directed toward concavity. It was a lordosis that had buckled to the side.

In the thoracic region the vertebrae are heart shaped and they are like a prism with the apex anteriorly but the prism is rendered asymmetric from D4 to D9 by the pressure effect of the left-sided aorta. On forward flexion this has the effect of rotating the apex of the prism to the right. The thoracic vertebrae are vulnerable to rotation but thoracic kyphosis protected the thoracic vertebrae to rotation. If the thoracic vertebrae are flexed forward beyond the limits, spinous processes will be seen towards the convexity and slight coronal plane curvature is produced (1). This pathology is similarly seen in a rotated kyphosis. This is a normal buckling of the spine. When lordosis is present at the thoracic vertebrae, the asymmetrical prisms spin out to the side to produce the typical deformity with the spinous processes now in concavity. On the other hand; in the cervical and lumbar regions these vertebrae are lordotic in the sagital plane and prismatic in the transverse plane with the bases of the prisms directed anteriorly and this position is a rotationally stable configuration. In the lumbar region the abdominal aorta is to the left of the median sagital plane and this creates left-sided of the lumbar prism (8).

The deformity of idiopathic scoliosis appears as a curvature in the coronal plane but this deformity is entirely secondary. It's the lordosis plus rotation and rotation occurs with the spinous processes are always directed towards the convexity.

When a child is born there is a smooth kyphosis from the foramen magnum to the lumbosacral junction which is lordotic at birth. When head control is developed by neck extension a cervical lordosis appears and then the child assumes the sitting and then standing positions the lumbar lordosis is fully developed. At the thoracic vertebrae; the median plane shape change during growth. In normal children the thoracic kyphosis reduces in size from the age of 8 to 14 years, it is minimum at age of 12. This change occurs at the same time in boys and girls at the same time. It shows that it is independent from growth velocity. When the thoracic kyphosis is at its minimum in girls, they are going to peak adolescent growth velocity and this explains greater progression potential in girls. When the thoracic kyphosis is at its minimum in boys their growth velocity is constant but when their thoracic kyphosis becomes maximal in later adolescence. That't why boys are more prone to Scheurmann's disease.

The concept of short-segment lordosis is being an important factor and shown by Somerville et al (21). It is therefore the lateral profile of the spine which is important in progression of idiopathic scoliosis. The flat lateral profile of patients with idiopathic scoliosis also explains the observation that these children are taller than their peers but not growing faster. This is more likely the effect of the lateral profile than genetic tall stature as the condition effects children off all sizes. The patients of idiopathic scoliosis represent the ends of lateral profile and have a similar familiar trend and community prevalence rate (23).

As a result; Leeds group mention that idiopathic scoliosis is a three-dimensional deformity and in the thoracic region the essential lesion lies in the sagital plane in the form of an area of inappropriate lordosis (11). The thoracic kyphosis is normally protected from buckling by being behind the axis of spinal column rotation but when the thoracic lordosis develops it brings the apical region anterior to this axis and thus under compression with resultant buckling failure of the spinal column. A number factors favor column buckling (Euler's law) and thus the bigger a deformity the more likely it will be continue progressing and the taller and more slender the column the more likely it will be to fail and this is seen in idiopathic scoliosis. Not only is lordosis the essential lesion but it is also the primary abnormality which can be demonstrated in children before lateral curvature and rotation develop (11).

Nottingham Concept : (3) According to this theory; they evaluated data from studies of the hips, pelvis, spine, rib cage and trunk muscles in scoliotic (pre and post operative) and control patients, cadavers in order to formulate a new theory of etiology for idiopathic scoliosis. Idiopathic scoliosis results, in part, from a developmental abnormality in the central nervous system creating rib vertebra nagle asymmetry which leads

to a cyclical failure of mechanisms of rotation control in the trunk; these involve rotation-inducing (pelvic) and rotation defending (discal, ligamentous and costal) mechanisms acting mainly in gait. The mechanical break down of rotation occurs in association with a lateral spinal curvature and a lordotic segment to create the initial deformity of idiopathic scoliosis. Then, growth, both abnormal (secondary to vertebral high-pressures) and normal (linear spinal growth) with gravity adds to the initiating and continuing neuromuscular mechanisms to augment curve progression. This theory views the spine in the wider prespective of function in the trunk, evolution and development, all in relation to bipedalism.

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