Quebec Cooperative Study
of Friedreich's Ataxia

Scoliosis in Friedreich's Ataxia

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SUMMARY: *The preliminary results based on a three year study on the evolution and management of scoliosis in Friedreich's ataxia are presented. Thirty-two patients were followed in the Neuromuscular Disease Clinic at Sainte-Justine Hospital where standardized spinal radiographs were taken periodically with the Scoliosis Chariot and the Throne.*

Pathomechanics — *Between the age groups I (5 - 10 years) and II (10 - 15 years), a substantial increase in the Cobb values occurs. Associated with it, an increase was observed in the thoracic and thoraco-lumbar projected surface area indices. The relative torsion between the thoracic and lumbar segments was associated with a sudden increase in the Cobb measurements.*

A computer program which generates a tri-dimensional display of the spine from the coordinates obtained from the standardized radiographs has been adapted to study the importance and influence of the thoracic torsion on the progress of the scoliosis and on the evolution of the total spinal deformity.

Management — *Prevention of the progression of established curves was our main objective. Careful examination of the spine, depending on the age of the child, in our preliminary study, stimulated early orthopaedic treatment in any curve of 20⁰ or more. There was always concern for curves of 30⁰ or more. In the growing child, bracing which was recommended is now under a more thorough investigation. The use of spinal pacemakers is being clinically evaluated.*

In the older child, the curve was usually stable after sixteen years of age. Surgery was usually attempted in curves over 40⁰ in the growing child. The same curve was usually stable after the growth period.

For the non-ambulatory patients, the present study suggested the prescription of a molded seat with the following characteristics: i) a posterior lumbar support, ii) low thoracic lateral supports and iii) a slight inclination of the seating system. This was presumed to be beneficial in maintaining stability of the spine.

RÉSUMÉ: *Les résultats de trois années d'étude sur l'évolution et le traitement de la scoliose dans l'ataxie de Friedreich sont présentés dans cet article. Trente-deux patients ont été suivis à l'Hôpital Sainte-Justine aux Cliniques des maladies neuromusculaires où des prises de radiographie de la colonne sont normalisées au moyen du Chariot de la scoliose et du Trône.*

Pathomécanique — *Entre les groupes d'âge I (5 - 10 ans) et II (10 - 15 ans), il existe une augmentation substantielle des valeurs de l'angle de Cobb. Associé à ceci, une augmentation est aussi observée dans les indices de la surface projetée. La torsion relative des segments thoracique et lombaire de la colonne vertébrale semble être associée à une augmentation subite dans les mesures de l'angle de Cobb.*

A la suite des résultats obtenus relativement à un paramètre global de déformation (la surface projetée), une étude de la torsion thoracique a été effectuée. Par le biais de simulations mathématiques, il a été révélé que la surface projetée varie en fonction de l'angle de torsion pour divers angles de Cobb. En comparaison avec les données cliniques obtenues à partir des radiographies normalisées de la colonne, ces simulations ont permis d'évaluer la contribution de la torsion sur l'ensemble des déformations associées à la scoliose.

Traitement — *La prévention de la progression de la scoliose établie représente notre objectif principal. Une évaluation de la colonne des patients de différents groupes d'âge stimulera un traitement orthopédique précoce de toute courbe de 20⁰ ou plus. Une anxiété persiste lorsque des courbes de 30⁰ ou plus sont observées. Chez l'enfant en croissance, le port du corset qui était autrefois recommandé, fait présentement l'objet d'une étude plus approfondie. L'utilisation de stimulation électrique pour la correction de scoliose est présentement étudiée en clinique. Chez l'enfant plus âgé, les courbes sont ordinairement plus stables après l'âge de seize ans. La chirurgie est habituellement entreprise chez les enfants en croissance, possédant une courbe de plus de 40⁰. La même courbe est ordinairement stable après la poussée de croissance.*

Pour les patients non-ambulants, cette étude semble indiquer la prescription d'un siège moulé possédant les caractéristiques suivantes: i) un support lombaire postérieur, ii) des supports latéraux au niveau inférieur de la colonne thoracique et iii) une légère inclinaison du fauteuil de positionnement. Ceci semble être bénéfique dans le maintien de la stabilité de la colonne.

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INTRODUCTION

The management of scoliosis affecting Friedreich ataxia patients has been limited to body braces and back surgery (Geoffroy et al, 1976). Only recently, a clinical and analytical preliminary retrospective study by Allard et al. (1980a) on the pathomechanics and management of scoliosis in this type of neurological disorder has been reported in the literature. In this study, twenty-one patients having Friedreich's ataxia had been followed over a one year period. Seventeen scoliosis-related parameters have been extracted from forty-seven standardized spinal radiographs. A substantial increase in the Cobb angle values (Cobb, 1948) has been observed at about the age of ten. These patients generally assume a posture with the shoulders rotated, to probably balance the internal forces produced by the increase in the scoliosis. This rotation of the shoulders generates, in turn, an additional deformation of the spine which appears to be concentrated in the thoracic segment. Associated with it, an increase in the projected surface area index (PSA), representing the relative spinal torsion of the thoracic segment with respect to the lumbar segment, has been identified as a probable quantitative index in the prognosis of the spinal deformities in this entity.

The present study was initiated then to evaluate the importance and the influence of this thoracic torsion on the progress of the scoliosis and on the evolution of the total spinal deformity. In particular, it consists of two parts. In the first one, the original study has been expanded to cover a three year period where seventeen boys and fifteen girls were on the average seen once a year in the Neuromuscular Disease Clinic at Sainte-Justine Hospital. It was thought necessary to investigate the progression of scoliosis from the onset when the patients were ambulatory to the time when they were wheelchair-bound with severe structural deformation of the spine. In the second part, a computer program which generates a tri-dimensional display of the spine from the coordinates obtained from the standardized radiographs has been adapted to simulate a thoracic torsion.

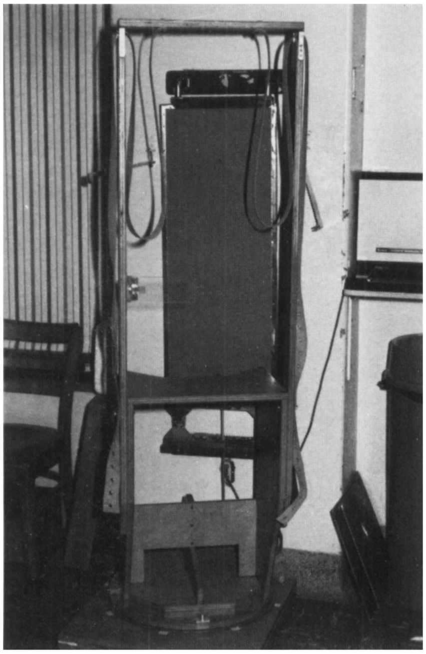

Figure 1 — Scoliosis Chariot

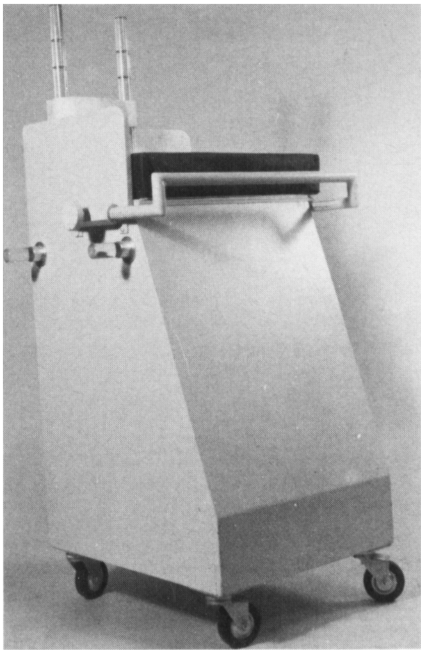

Figure 2 — Throne

Standardized Stereo radiographs Technique

An important consideration in the study of scoliosis by means of radiographs is the accuracy of the measurement method. Traditionally, the antero-posterior spinal radiographs are taken with the patient either in the standing or supine position. If an additional lateral radiograph is taken, it is often viewed separately. The physicians quantify a tri-dimensional or spatial deformity of the spine from its projection in a single plane, namely that of the radiograph. It has been shown (McNeice and Dawson, 1976) that the currently accepted technique of obtaining antero-posterior and lateral radiographs leads to inconsistent measurements in scoliosis.

These systematic errors can be reduced by utilizing two devices, namely the Scoliosis Chariot shown in Fig. 1 and Throne shown in Fig. 2 to obtain standardized spinal radiographs of ambulatory and non-ambulatory patients respectively.

The Chariot, designed by McNeice and Dawson (1976), consists of a booth positioned in such a way that when the patient stands in it, he is in a true lateral position to the X-ray tube. After the first spinal radiograph is taken, the Chariot is rotated through 90^0 and positioned in such a way that the patient now stands relatively to the X-ray tube in a true antero-posterior position in which the second radiograph is taken.

The Throne, designed by Koreska et al. (1978a), consists of a seat on which the patient is placed with his back resting against two referenced acrylic rods and with the side of his pelvis leaning against a lateral support. Antero-posterior and lateral radiographs are taken by positioning the Throne accordingly against the film cassette.

From these radiographs, the centroid of each vertebra between the 7th cervical (C7) and the 1st sacral (S1) as well as the reference scales were located and then traced on an overlay transparent sheet of paper. These tracings were sent to the Assessment Department, Glenrose Hospital, Alberta, where they were digitized. The information was fed into a computer program which generated a graphical display of the spine and computed tri-dimensional indices (Koreska et al., 1978b). This information was mailed to Montreal for clinical analysis.

Simulation of Thoracic Torsion

A computer geometric reconstruction program based on a spline function algorithm (McNeice et al. 1975,

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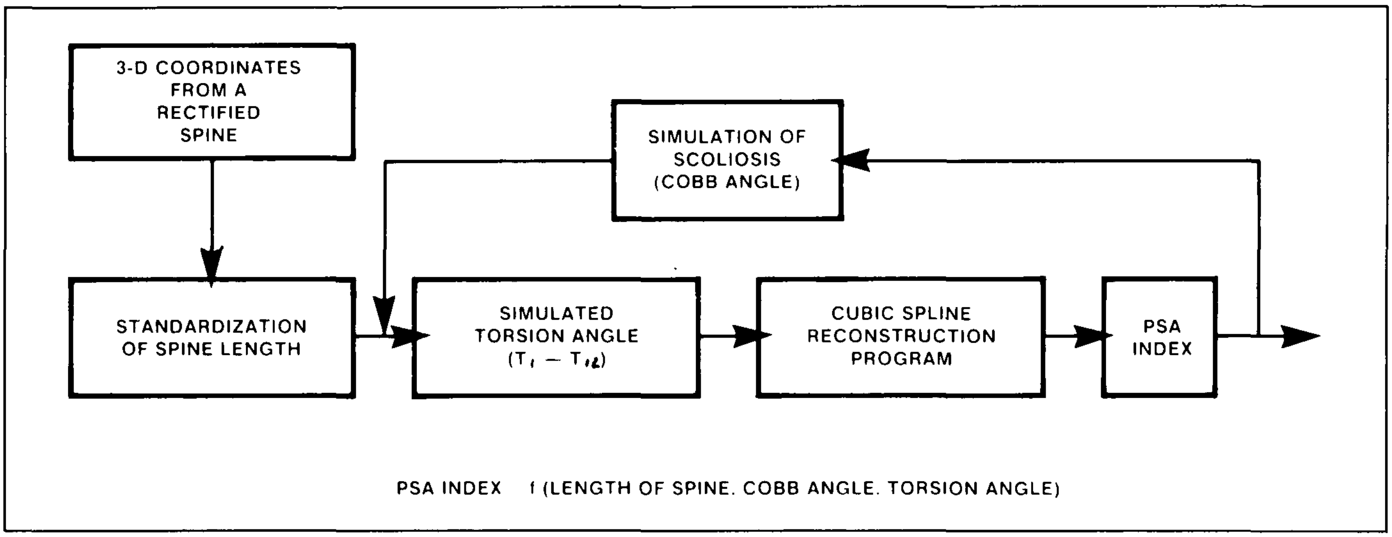

Figure 3 — Schematic illustration presenting the methodology used to carry out the mathematical simulation.

Raso, 1977) has been adapted to study the thoracic torsion by means of the projected surface area index. This index is a function of the length of the spine, the Cobb angle measurement and the torsion angle. To determine its impact on the thoracic torsion, the other two parameters have to be known (Dansereau, et al. 1982).

Figure 3 presents a schematic description illustrating the analytical methodology utilized to carry out the desired thoracic torsion simulation. A normal reference spine configuration with no scoliosis has been mathematically rectified to align the vertebral centroids with the vertical. Afterwards, the length of the reference spine has been standardized in order to compare it with that of some Friedreich ataxia patients. A torsion angle, equally distributed along the length of the thoracic spine, as schematically illustrated in fig. 4 (90^0 torsion), was simulated respectively for values varying between 0^0 and 50^0. By means of a cubic spline function reconstruction program, the projected surface area index was calculated. The reference rectified normal spine was then modified into order to generate a pure scoliosis of 10^0. The thoracic torsion angles were again simulated for this new spine configuration and their respective projected surface area indices were calculated. This procedure was repeated until an induced scoliosis of 50^0 had been generated.

Clinical Results

Although seventeen scoliosis-related parameters are included in the analysis of the standardized radiographs, six of them presented in Table I are relevant to this study. The results obtained for this three year retrospective study on the pathomechanics of spinal deformities in Friedreich's ataxia patients are presented.

TABLE I

Scoliosis — Related Parameters

1. Type of curve
2. Spinal deformity angle
3. Length of spine
4. Sacral angle
5. Lumbo-sacral angle
6. Projected surface area

1. *Type of Curve.*

The type of thoraco-lumbar deformity varies from patient to patient and sometimes from one clinical visit to another. This latter observation was often noticed shortly after a sudden increase in the deformity. A typical case, presented a right thoracic spinal deformity which was fairly stable at 28^0 between the ages of 10 to 15 years, to increase dramatically as evidenced by a Cobb angle rise of 24^0. Shortly afterwards, the spine developed an additional compensatory left lumbar curve. Of ninety-three observations, 70% were identified as right thoracic type, half of them with left lumbar deformity.

2. *Spinal Deformity Angle*

The spinal deformity angle was measured by means of the Cobb method. Table II presents the average values of spinal deformities for ambulant and non-ambulant patients. These deformities are classified in four age groups consisting each of a five year interval.

For the ambulant and non-ambulant boys an increase of 8^0 in the Cobb angle values was observed between the age groups I and II. However, for the ambulant patients, the average Cobb angle measurements in the age group II is only 21^0 as compared to 38^0 for the non-ambulant male patients. For the boys, an increase of 12% in the Cobb angle values was observed between the age groups II and III. This increase does not reflect the substantial evolution of the scoliosis (34^0) in the ambulatory patients and the fairly stable scoliosis (41^0) for the non-ambulant patients.

For the girls, an average increase of 13^0 in the Cobb angle values was observed between the age groups I and II. Between the age groups II and III, another substantial average increase of 21^0 in the Cobb angle measurements was observed. For the ambulant patients, the increase between the last two age groups is much less (8^0) than that

TABLE II
Average Spinal Deformity
(Cobb Angle)

Age Group (years)	Ambulant	Non Ambulant	All
BOYS			
I (5-10)	16	—	16
II (10-15)	21	38	24
III (15-20)	34	41	36
IV (20-25)	17	31	27
GIRLS			
I (5-10)	13	—	13
II (10-15)	25	—	25
III (15-20)	33	57	46
IV (20-25)	15	37	33

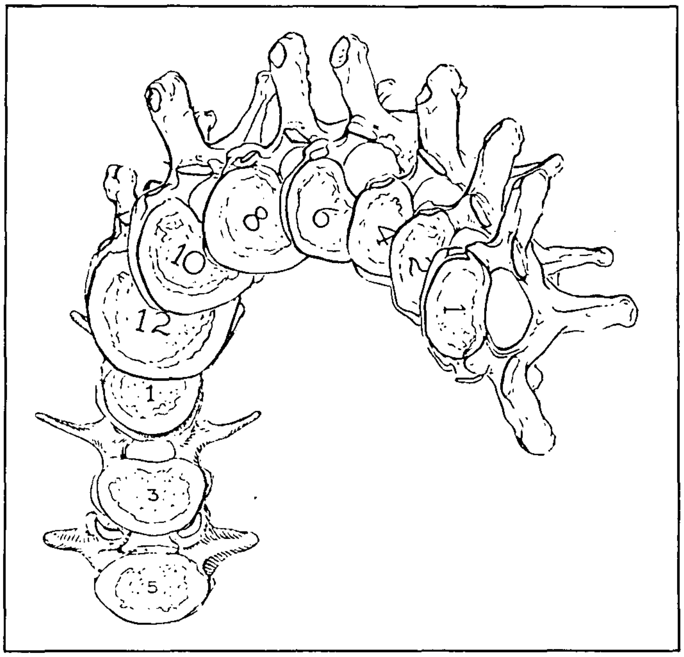

Figure 4 — Exploded view of a spine after a 90^0 torsion.

noticed for the non-ambulant patients (27^0).

For both boys and girls, a slight decrease in the spinal deformity in the age group IV may be explained by the small number of observations for that group.

3. *Length of Spine.*

The length of the spine, obtained from the tri-dimensional geometric reconstruction program, is defined as the summation of the intervertebral distances from S1 to C7 calculated along the length of the cubic spline function. Figures 5 and 6 respectively present the values of the spine's length computed for boys and girls. Due to a greater number of patients followed as compared to that of the previous study (Allard et al., 1980a), a growth spurt which was not evident before was noticed for boys and girls at approximately the ages of 12 and 9 respectively. The growth spurt mainly occurred during the age groups II and III. It is during that same period that the progression of the scoliosis is most evident.

4. *Sacral and Lumbo-Sacral Angles.*

The sacral and the lumbo-sacral angles were measured in the lateral radiographs. The sacral angles is defined as the angle subtended by the horizontal plane and an axis formed by joining the mid-points of the superior surface of S1 and S2. The lumbo-sacral angle is defined as the angle subtended by the axis (S1-S2) and another one formed by joining the centroids of the vertebral bodies of L4 and L5.

The value of sacral angle varied between 20^0 and 50^0 until the age of

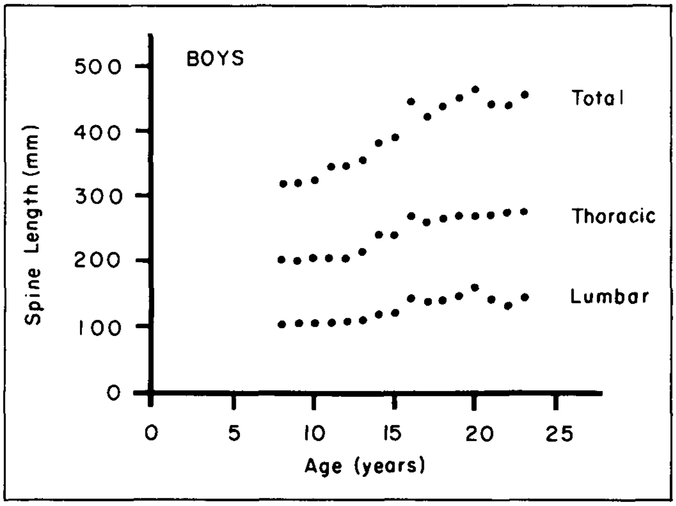

Figure 5 — Computerized length of the spine in boys.

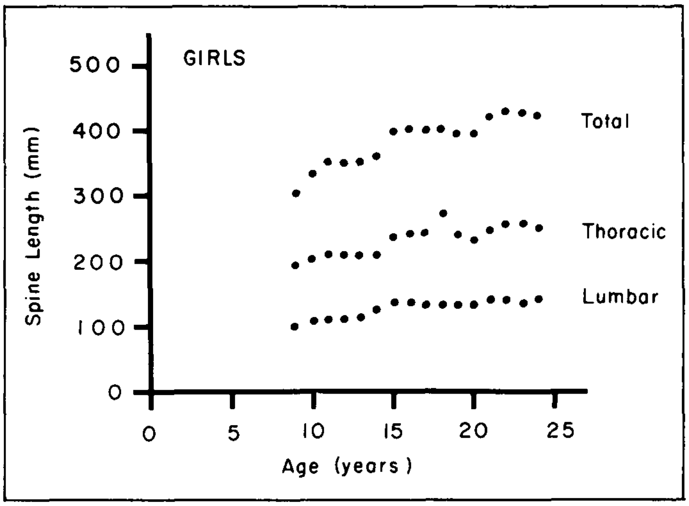

Figure 6 — Computerized length of the spine in girls.

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eighteen when the angle increased to values above 80⁰. The value of the lumbo-sacral angle varied between 35⁰ and 60⁰ until the age of eighteen and afterwards decreased to 20⁰. Both these phenomena coincided with a change in posture of the patient who was becoming non-ambulant.

5. *Projected Surface Area.*

The projected surface area (PSA), is a measurement of a surface observed when the segment is projected on a plane perpendicular to the axis formed by superimposing its extremities (McNeice et al, 1975). Thus for a normal spine, the PSA obtained by superimposing S1/C7 reduces to a straight line of zero area. Figure 7 and 8 show the results obtained for the lumbar, thoracic and total PSA values for boys and girls respectively. The lumbar PSA was negligible (values less than 50 mm^2). For the boys, the thoracic PSA index reached a value fo 200 mm^2 at approximately the age of nine. Afterwards, it fluctuates over this value before stabilizing at a value above 300 mm^2 by the age of fifteen. The total PSA index closely followed those thoracic PSA values to the age of eleven. Later on, they increased substantially and reached values of around 550 mm^2. For the girls, a more evident pattern was observed. The thoracic PSA index reached a value of 100 mm^2 at approximately the age of eleven. Afterwards, it increased to values of 200 mm^2 where it fluctuated. The total PSA values closely followed those thoracic PSA values to the age of thirteen. Then they increased substantially and stabilized at values of 900 mm^2. These results agree with those reported by Allard et al. (1980a). It was then assumed that up to the age of eleven the thoracic deformity increased in the same plane as that of the total spine. Afterwards, a sharp rise in the total PSA index seemed to indicate the occurrence of a relative torsion of the thoracic segment with respect to the lumbar segment.

To further investigate the PSA index, a simulation program was developed. The results obtained from the simulation were applied to determine the importance and influence of the thoracic torsion on the progress of scoliosis and

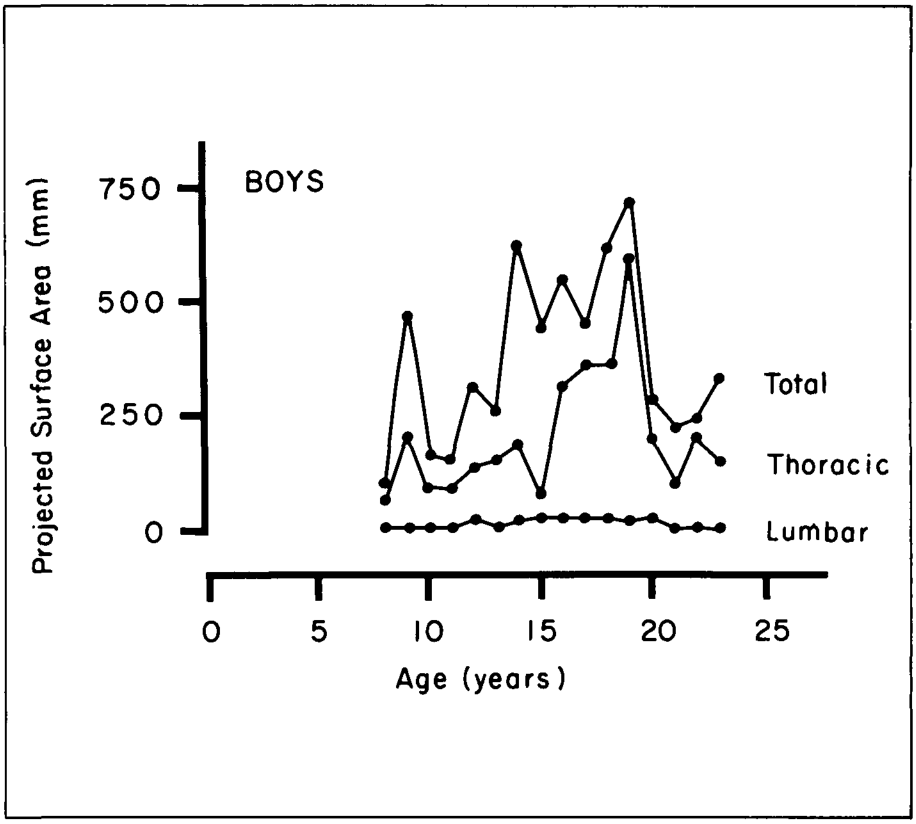

Figure 7 — Computerized projected surface area of the spine in boys.

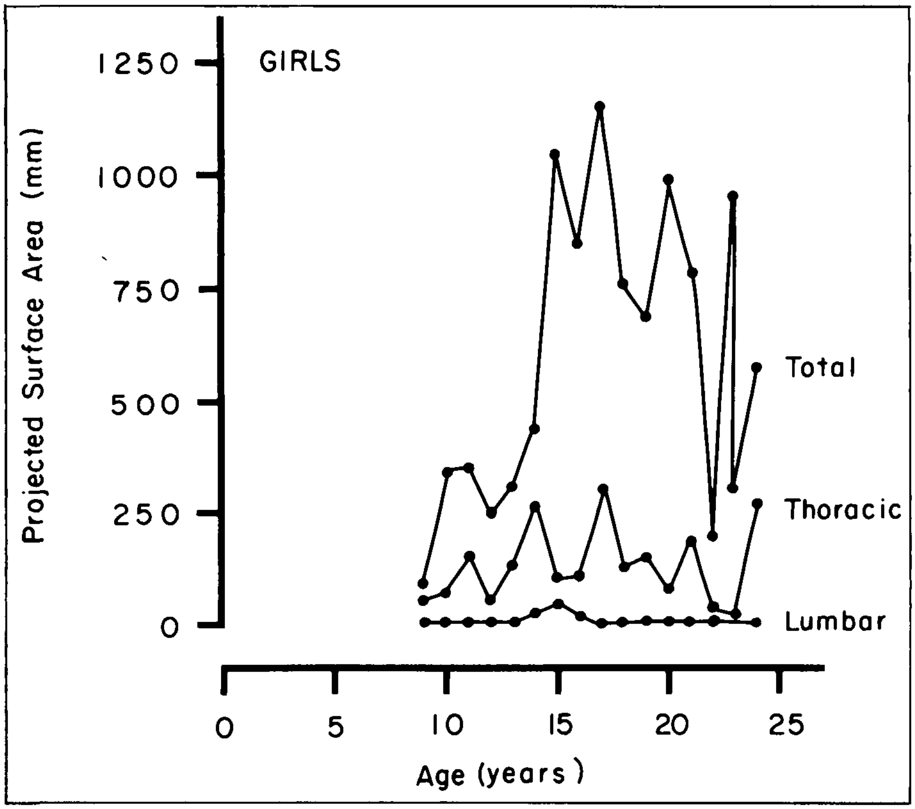

Figure 8 — Computerized projected surface area of the spine in girls.

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on the evolution of the total spinal deformity of a particular patient.

Results of the Simulation.

The results of the thoracic torsion simulation are graphically presented in Fig. 9 where the total PSA index is plotted as a function of the total torsion angle of the thoracic segment for various initial Cobb angles. Three main remarks are in order in view of these results:

(i) for a given torsion angle, the PSA index increases for increasing Cobb angles;

(ii) for zero torsion angle and zero Cobb angle, the PSA index is also zero; and

(iii) the curves for various Cobb angles are quasi-parallel.

This leads to the conclusion that the PSA index can be decomposed into two parts: one due to the scoliosis as measured by the given Cobb angle which seems to remain constant, regardless of the value of the total PSA and the other which increases monotonously with the torsion angle. Therefore, for a given PSA index, Cobb angle and spine length, it seems possible to obtain a measure of a "simulated torsion angle". The contribution of the pure torsion on the total PSA index is directly seen from the relationship between the total PSA index and thoracic torsion angle for a zero value Cobb angle, as shown in Fig. 9. For a given Cobb angle measurement the contribution of the scoliosis on the total PSA index is calculated by subtracting from its total PSA value, its corresponding total PSA value taken from the zero degree Cobb relationship at the same torsion angle. It is important to note that this "simulated torsion angle" can be reproduced by various combinations of intervertebral relative torsions and that this new information is, at the present, only a qualitative indicator of the deformity.

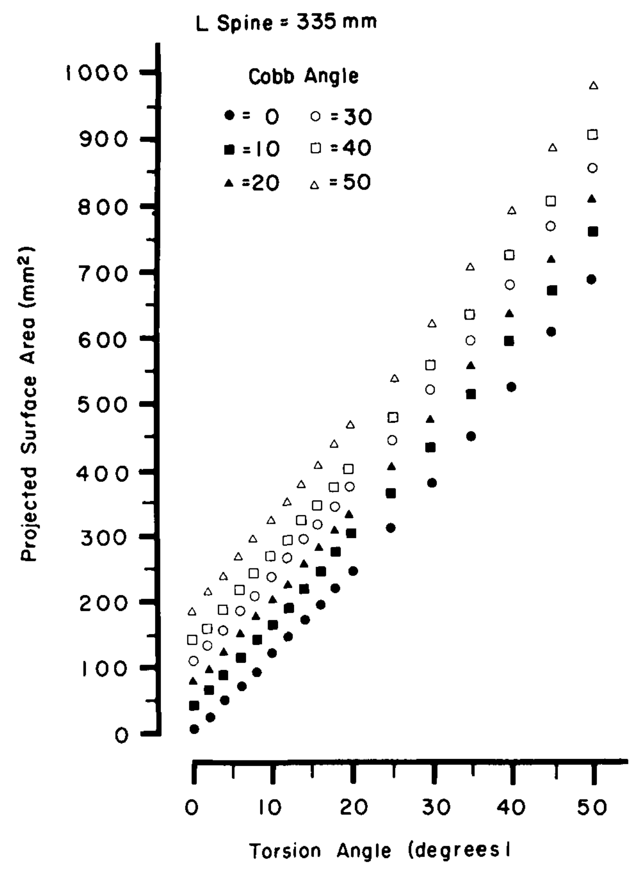

Figure 9 — Relationship between the total PSA index and thoracic torsion for various Cobb angles.

TABLE III

CLINICAL DATA AND DATA OBTAINED FROM THE SIMULATION FOR A FRIEDREICH ATAXIA PATIENT

PATIENT D.D.

CLINICAL DATA			SIMULATION				
AGE (YEARS)	COBB ANGLE (°)	PSA (mm²)	SIMULATED TORSION ANGLE (°)	POSSIBLE PSA DUE TO SCOLIOSIS mm²	POSSIBLE PSA DUE TO SCOLIOSIS % OF TOTAL	POSSIBLE PSA DUE TO PURE TORSION mm²	POSSIBLE PSA DUE TO PURE TORSION % OF TOTAL
8 1/2	13	115	4	55	48%	60	52%
8 2/3	12	81	3	40	49%	41	51%
9 1/2	23	473	29	108	23%	365	77%
10 1/4	29	650	40	160	25%	490	75%

Application.

To illustrate the possible use of the relationship between the total PSA Index and the thoracic torsion angle, the evolution of the Cobb angle and the PSA index of a Friedreich ataxia patient seen over a two year period is shown in Table III. It can be noted that in less than one year the total PSA index values have increased more than five fold while the Cobb angle increased only by 11^0. The corresponding simulated torsion angle, for that same period of observation, increased from 4^0 to 29^0.

In the early stages of scoliosis (12^0) the total PSA index values seems to indicate that the contribution of the scoliosis and pure torsion on the total spinal deformity are approximately equal. Later on, when the scoliosis progresses (23^0) the total PSA index values seems to illustrate that the con-

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tribution of the pure torsion (75%) on the spinal deformity is much more important than that of the scoliosis.

Management of Scoliosis in Friedreich's Ataxia Patients as Carried out at Sainte-Justine Hospital.

Based on this biomechanical study of scoliosis which confirms most of the observations reported in an earlier work (Allard et al. 1981), a brief description of the type of treatment provided for the ambulant and non-ambulant patients at Sainte-Justine Hospital is given.

1. *Ambulatory Patients*

Patients were reviewed at regular intervals in the Neuro-Muscular Disease Clinic of our institution. The ambulatory patients with evidence of scoliosis were closely followed. Careful observations were recommended for curves under 20^0 in the adolescent child. If a curve progressed beyond the 20^0 limit, an orthopaedic treatment was recommended. Standard bracing which was the usual treatment is now being revised. In view of the new findings on the importance of the thoracic torsion in this type of spinal deformity, a new approach based on spinal electrical stimulation is now being investigated.

Surgery is still suggested for curves in the 40^0 range or more in the growing child. Posterior and anterior fusion have been performed with satisfactory results in both types of approach.

2. *Non-Ambulatory Patients.*

For non-ambulatory patients with a mild scoliosis, a special molded seat was prescribed on an experimental basis (Allard et al., 1980b). The main feature of the seating system was based on the preliminary results from the study of scoliosis detailed above, in particular the observations made on the location of the apex of the scoliosis, the kypho-scoliosis index (KSI) and the projected surface area.

To restrain the progression of the lateral deviation of the spine as well as its rotation as shown respectively by the location of the apex of the deformity and the evolution of the projected surface area indices, lateral supports at the low thoracic levels should be helpful. To manage the evolution of the lumbar lordosis into a kyphosis as shown by the increase in the lumbar KSI a posterior lumbar support was included in the seating system. Finally, since the scoliosis encountered in Friedreich's ataxia is paralytic, an inclined seat such as the Hospital for Sick Children Spinal Support System (Koreska et al., 1975) equipped with low thoracic lateral and posterior lumbar supports should delay the progression of the scoliosis.

If the results with the spinal electrical stimulator are proven beneficial for the ambulant patients, the use and application of molded seats for non-ambulant patients may then be revised.

ACKNOWLEDGEMENTS

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