Spinal and pelvic parameters in Marfan’s syndrome and their relevance to surgical planning

Spinal deformities are a common feature of Marfan’s syndrome and can be a significant cause of morbidity. The morphology of the scoliosis associated with this condition was previously described by Sponseller, but no correlation with the pelvic parameters has been seen. We performed a retrospective radiological study of 58 patients with scoliosis, secondary to Marfan’s syndrome and related the findings in the thoracolumbar spine to the pelvic parameters, including pelvic version (tilt), pelvic incidence and sacral slope. Our results showed marked abnormalities in the pelvic values compared with those found in the unaffected population, with increased retroversion of the pelvis in particular. In addition we found a close correlation between the different patterns of pelvic parameters and scoliosis morphology. We found that pelvic abnormalities may partially dictate the spinal disorders seen in Marfan’s syndrome. Our results supplement the well-established Sponseller classification, as well as stressing the importance of considering the orientation of the pelvis when planning surgery.

Marfan’s syndrome is one of the most common connective tissue disorders and is caused by the mutation of a gene which codes the formation of fibrillin.1 Spinal deformities are frequently encountered in patients with Marfan’s syndrome.2-4 The spinal balance and the pelvic parameters are fundamental elements in both the analysis of such deformities and in the choice of appropriate treatment.5

In the morphological classification defined by Sponseller et al2 for patients with Marfan’s syndrome, two groups of spinal profiles are identified. In type-I, the transition between the kyphosis and the lordosis is situated at the second lumbar vertebra or higher. This group is sub-classified to identify a normal kyphosis (20˚ to 50˚) and lordosis (type-IA), a kyphosis of less than 20˚ (type-IB) or a kyphosis which exceeds 50˚ (type-IC). In the type-II spine, the transition is situated below the level of the second lumbar vertebra. Type-II has two sub-groups; type-IIA represents a spine with an extensive kyphosis which involves the thoracolumbar junction, while type-IIIB has an inversion of the spinal curvature with a thoracic lordosis, thoracolumbar kyphosis and a low-set lumbar lordosis (Fig. 1).

The aim of this study was to establish the spinal and pelvic parameters in a group of patients with a scoliosis which was treated conservatively and who also suffered from Marfan’s syndrome and to determine the relationship between the two conditions.

Patients and Methods

The initial study group comprised 498 patients with Marfan’s syndrome, who were attending a national multidisciplinary specialised centre for review. Of these patients, 279 had a scoliosis with a Cobb angle ≥ 10˚, representing 56% of the initial group. Our exclusion criteria reduced the number of patients for review to 238, because we excluded 31 patients who had undergone surgery at other centres and ten who were under 15 years of age at the time of the clinic visit. Our methodology required a full set of standardised radiographs to be available in the archives. Unfortunately, because of the nature of the clinic, with patients attending from throughout France and overseas, in addition to the mixture of private and public hospitals within the healthcare system, only 58 complete sets of medical records were available for review and it is these patients which form the basis of our study. There were 25 males and 33 females with a mean age of 36 years and one month (16 to 54 years) and a mean weight of 70 kg (52 to 105). The mean maximum Cobb angle was 20˚ (10˚ to 61˚).

Ours was a retrospective clinical and radiological study. We employed the diagnostic criteria agreed in the Berlin and the Gand
conference⁶ to identify abnormalities in the systems affected by Marfan’s syndrome.

In almost three-quarters of the 58 cases, patients presented with one, or several, major criteria involving the locomotor system (44; 76%), cardiac system (47; 81%), neurological system (40; 69%) and the ophthalmological system (43; 74%). The majority of the patients (46; 79%) had involvement of the cutaneous tissues and pulmonary
signs were found in five patients (9%). No major diagnostic
criteria existed for these last two manifestations of
Marfan’s syndrome.

Standardised anteroposterior (AP) and lateral weight-
bearing radiographs were taken of the whole spine and pel-
vis using a single film. The lateral view was taken in the
clavicle/arm position. The radiographs were digitalised and
analysed by the senior author (CGL), using Spineview soft-
ware (Surgiview, Paris, France). For each radiograph, the
spinal curvatures were quantified in the AP and lateral
views. In the same manner, the pelvic views were analysed
and pelvic incidence, pelvic version (pelvic tilt) and sacral
slope7-9 measured (Fig. 2).

We compared the results obtained with spinal parame-
ters of an unaffected population.10 Previous studies10,11
have shown a strong statistical correlation between the pel-
vic incidence with the sacral slope and the spinal curvature
and have allowed the establishment of parameters for the
unaffected population. For the patients with Marfan’s syn-
drome, we calculated the expected values for the sacral
slope, the lumbar lordosis and the thoracic kyphosis on the
basis of the pelvic incidence using the methodology out-
lined in previous publications.10,11

**Statistical analysis.** We performed a regression analysis
between the various parameters both expected and mea-
sured. Statistical analysis was performed using analysis of
variance with p < 0.01 considered significant.

**Results**

In the frontal plane, the morphology of the curves was
varied, with three superior thoracic, nine inferior thoracic,
11 double thoracic, seven thoracolumbar, 13 lumbar, ten
combined thoracic and lumbar and five triple curvatures.

In the lateral plane, the mean thoracic kyphosis was
41.3˚ (SD 14.8˚; 15.9˚ to 85.3˚) and the mean lumbar lordo-
sis was 45.7˚ (SD 12.2˚; 6.5˚ to 70.3˚). The mean pelvic inci-
dence was 47.1˚ (SD 14.2˚; 20˚ to 87˚), with a mean sacral
slope of 32.7˚ (SD 8.6˚; 19˚ to 53˚) and a mean pelvic version
of 14.3˚ (SD 10.1˚; -8˚ to 45˚).

A study of the relationship between these parameters
showed a strong correlation between pelvic incidence and
sacral slope (Pearson’s correlation; bilateral significance
p < 0.001; regression coefficient R = 0.704; significant at
p < 0.001) (Fig. 3).

We analysed the curvatures in the lateral view in order to
classify the patients into subgroups. Of the studied patients,
32 (55.1%) were type-IA, three (5.1%) were type-IB and 15
(25.8%) were type-IC, while four (7%) were classified as
type-IIA and a further four (7%) were type-IIB. The pelvic
incidence and the measured sacral slope, the expected sac-
ral slope, the lumbar lordosis and kyphosis of each type are
summarised in Table I.

By comparing type-I and type-II patients, significant dif-
fences in the various parameters were found. In addition,
the comparison between the five sub-groups demonstrated
significant differences for the different variables (Table II).

In our study of the lateral radiographs and the pelvic
parameters, the mean pelvic incidence of 47.1˚ was smaller
than the 51.44˚ obtained from studies of unaffected subjects. In addition, the sacral slope is very different from the expected values for the unaffected population. The measured mean is 32.7˚, compared with the expected value of 40.3˚, a mean difference of -7.6˚ (F = 2.968, p = 0.005).

It is important to stress that in a normal population, the average sacral slope is approximately 40.6˚, while our study showed that for patients with Marfan’s syndrome there is a large increase in retroversion, which has an effect on the lumbar curvature. The measured lordosis was close to the expected values for a given sacral slope, with no significant difference found. Nevertheless, the expected lordosis calculated from the expected sacral slope, itself defined by the pelvic incidence, showed a significant difference (mean lumbar lordosis in our patients, 45.7˚, lumbar lordosis corresponding to the measured sacral slope, 48.4˚; theoretical lumbar lordosis in the normal population, 59.6˚). In addition, the measured thoracic kyphosis was slightly higher than the expected value for a given lordosis in these patients; a mean of 41.3˚ for an expected value of 38.5˚.

Discussion

The manifestations of Marfan’s syndrome shown in our study compares with previously published literature and serve to emphasise the body systems affected by this condition.

The incidence of scoliosis in this group of patients with Marfan’s syndrome is similar to that previously reported. Our 56% incidence of scoliosis compares with 55% reported by Robbins et al, 63% by Sponseller et al and 64% by Tallroth et al.

The distribution of the different types of scoliosis was diverse, but with a predilection for thoracic localisation. The distribution of thoracic and double thoracic curvatures is comparable with those previously reported. However, our study identified fewer thoracolumbar but more lumbar curves (12% and 21.5%, respectively) when compared with 22% and 14% in the Sponseller et al series (Table III). A weakness of our study is that the patients were not randomly selected. However, the fact that there is no statistically significant difference between our sample and previously published series, would suggest that our patients are fairly representative of the Marfan’s population as a whole.

The early puberty-associated peak in growth velocity in patients with Marfan’s syndrome, combined with the effect of the syndrome on connective tissue, might explain the pelvic malpositioning and spinal repercussions, which are seen with this disease.

Our study of the groups and sub-groups on the lateral radiographic views, according to the level of the kyphosis-lordosis junction and the type of thoracic curvature, was revealing. It showed statistical differences between the two groups of spinal profile, particularly for pelvic incidence. One possible explanation for these differences is that pelvic incidence is the initial factor which determines the transition level between a lumbar lordosis and a thoracic kyphosis. In our study, the small values for pelvic incidence were...
accompanied by transition levels below the second lumbar vertebra, the highest values being above this level.

In type-I, the values for pelvic incidence were higher than those for the group as a whole and were nearer to normal values. However, the sacral slope was much smaller than its expected value (Table I). The larger the difference, the more the retroversion of the pelvis and the less the capacity for the lumbar spine to maintain a good balance. The increasing mean sacral slope was reflected by similar increases in the values for lumbar lordosis. This might explain the morphology of the thoracic spine in a normal kyphosis, hypokyphosis, or hyperkyphosis, respectively, for the subgroups IA, IB and IC. Therefore, we believe that the morphological criteria for the thoracic spine classification proposed by Sponseller et al. are, in part, a consequence of the pelvic anomalies.

In type-II cases, the value of the sacral slope corresponded to the value of the pelvic incidence. However, if the patient had the ability to maintain a balance with the lordosis corresponding to the sacral slope, this was achieved by a long superior kyphosis and resulted in a type-IIA curvature. If the lordosis was much smaller than its expected value, the imbalance was characterised by the reversal of the superior curvatures as seen in a type-IIB curve.

We suggest that these findings highlight the difference in the morphology of the curves for the two Sponseller groups and that the pelvic parameters of patients with Marfan’s syndrome should be added to the well-established Sponseller system as a further level of refinement. In addition, our data enable a new correlation, between pelvic incidence and sacral slope, for patients with Marfan’s syndrome to be identified.

Our study is the first in the literature to assess spinal balance based upon the measurement of the pelvic parameters, in a population with Marfan’s syndrome. We have highlighted the anomalies encountered in the sagittal plane, mainly in the measurement of pelvic incidence and sacral slope. We consider that any deviation from the normal expected values for the parameters we have measured is linked to underlying abnormalities of pelvic orientation and spinal balance. This may explain the poor results of bracing in this condition.3,14

Our study was initially conceived with the aim of investigating spinal and pelvic parameters and their relationship to patients suffering from Marfan’s syndrome and to further expand the Sponseller classification. As a result of our findings we have changed our practice so that even more attention is paid to pelvic orientation when performing scoliosis surgery for patients with Marfan’s syndrome. Correct spinal balance must be obtained in the sagittal plane. In addition, our study documents the high frequency of associated pathology which can be a possible cause of postoperative surgical complications5 and which must be considered when surgical intervention becomes necessary.

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References