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ORIGINAL ARTICLE

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Prognostic significance of the Nasca classification for the long-term course of congenital scoliosis

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Abstract *Aim:* The aim of our investigations was to answer the question of whether the radiologic classification according to the investigations of Nasca has prognostic relevance for congenital scoliosis. Based on our results, a therapeutic concept was given. *Patients and methods:* Radiographs of 49 patients with a congenital scoliosis were classified according to Nasca. The progression of the malformation was analyzed by regarding the changes over a period of 8.3 years on average. *Results:* Of the analyzed patients with congenital scoliosis, 73% showed a mean progression rate of 1.8° per year. The most advanced progress of the Cobb angle per year was seen in patients with unilateral unsegmented bars with or without additional hemivertebrae (type 4 according to the Nasca classification) and patients with wedge-formed vertebrae (type 2a according to the Nasca classification). The presence of unilateral bars and the location of the hemivertebra or hemivertebrae (type 4) and wedge-formed vertebrae (type 2a) are the major determinants of deformity. Within the scope of our investigations, the following three factors were seen as deleterious when combined: thoracolumbal and long-distance scoliosis, female gender, and prepubertal growth period. *Conclusion:* From our analysis of 49 patients, we can propose the following therapeutic system for congenital scoliosis. Sole treatment with physiotherapy should only be recommended with congenital scoliosis curves ranging from 0° to 20° according to the Cobb method. With a Cobb angle smaller than 40° and some flexibility remaining in the spine, the congenital scoliosis should be treated additionally with corrective casts. For congenital scoliosis with a Cobb angle of more than 50°, as well as for congenital scoliosis with a poor prognosis (for example unilateral bars), a spondylodesis of the spine should be done before reaching the age of 5 years.

Keywords Congenital scoliosis · Nasca classification · Progression

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Introduction

The share of congenital scoliosis among the total of all scoliosis types is 10% [3, 14, 15]. Congenital scoliosis is defined as a deformity of the developing spine that results from defects in vertebral development. The developmental etiologies may be classified as either a failure of formation, a failure of segmentation, or a mixture of these two modes. Early detection and close surveillance of congenital scoliosis are critical, as a rapidly progressive curve may lead to significant deformity, pulmonary restriction, and neurologic problems if not treated [8]. Congenital scoliosis with a failure of formation and/or segmentation has to be distinguished from those other types of congenital scoliosis which can occur on account of other hereditary causes (e.g., diastematomyelia, myelomeningocele) without additional malformation of the vertebral bodies. Concerning the malformations of the vertebral bodies, we have to distinguish between dysmorphism of the vertebral body and the vertebral arch. According to the classification of Nasca [11], the malformations of the vertebral bodies were subdivided into disturbances of the segmentation and of formation. Nasca et al. [11] rated a total number of 315 patients with congenital scoliosis, who were separated into 6 subtypes. They ascertained that the location of hemivertebrae and the presence of unilateral bars are the major causes of deformity. In this investigation, the rate of progression of the scoliosis was variable, ranging from 1° and 33° per year (average 4° per year). Other authors found out that unilateral unsegmented bars showed the severest progression: the yearly rate of deterioration of the curve averaged 7.6° [4]. Curves with incomplete block vertebrae, clinically recognized as disc space narrowing, were considered to be a specific type of

congenital scoliosis which sometimes revealed a rapid progression during the adolescent growth spurt [16]. Concerning the pathogenesis of congenital scoliosis, the investigations of Tanaka and Uhthoff [17] especially brought more understanding. A hemivertebra or other defect of formation of the vertebral body is considered to be caused by abnormal differential growth of the loose-celled area. Defects of segmentation are due to complete chondrification of the dense-celled area or to an absence of the dense-celled area. Considering the importance of the intersegmental artery in the formation of the definitive vertebral body anlage, it may be concluded that congenital vertebral malformations are likely to occur during the stage of resegmentation and to be related to the abnormal distribution of the intersegmental arteries.

The aim of our investigation was to answer the question of whether the radiologic classification according to Nasca [11] has prognostic relevance for congenital scoliosis and if, based on our results, a therapeutic concept can be given. Therefore, we analyzed the X-rays of patients with a congenital scoliosis and grouped them according to the Nasca classification. After a long-term follow-up (on average 8.3 years), we measured the Cobb angle of the congenital scoliosis collective again. With these results, we want to make a therapy recommendation according to the progression of the congenital scoliosis.

Patients and methods

From 1956 to 1990, 49 patients were given in-patient treatment for a congenital scoliosis. The assessment of the congenital scoliosis occurred by means of at least two X-rays (anteroposterior projection and side projection) according to the Nasca classification [11]. The classification contains six types of malformations of congenital scoliosis (Figs. 1, 2):

- Type 1 is characterized by an individual surplus half vertebral body (Fig. 3). It is mostly round or oval and localized between two adjoining vertebral bodies. In the course of time, it often merges with one or both adjacent vertebral bodies. In the thoracal region, the surplus vertebral body is associated with an additional rib and a regular vertebral arch oval.

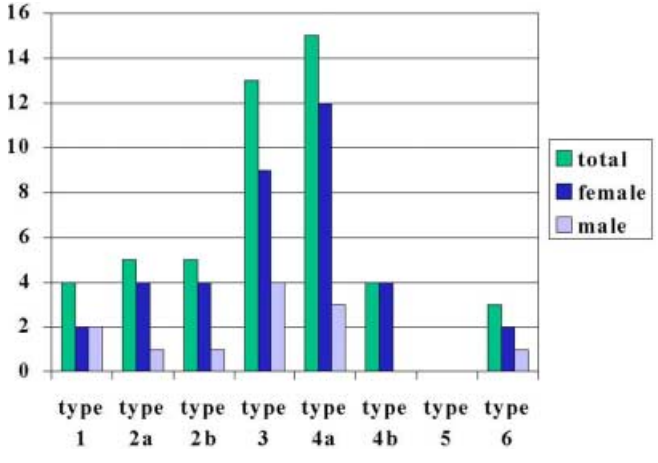

Fig. 1 Distribution of the patient collective analyzed (total number; male/female distribution)

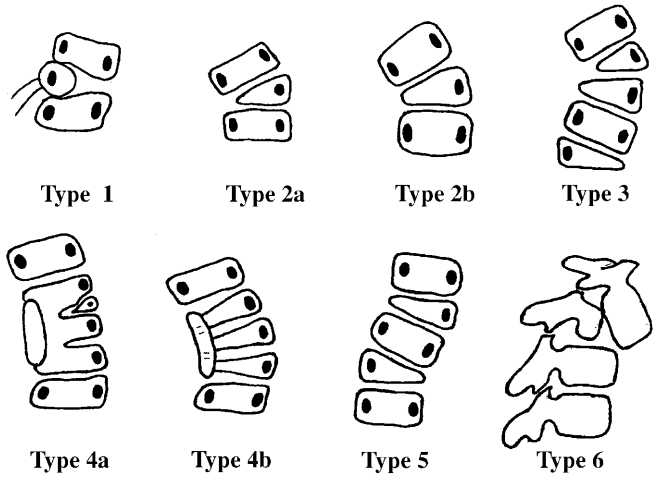

Fig. 2 Drawing with explanation of the different Nasca congenital scoliosis types

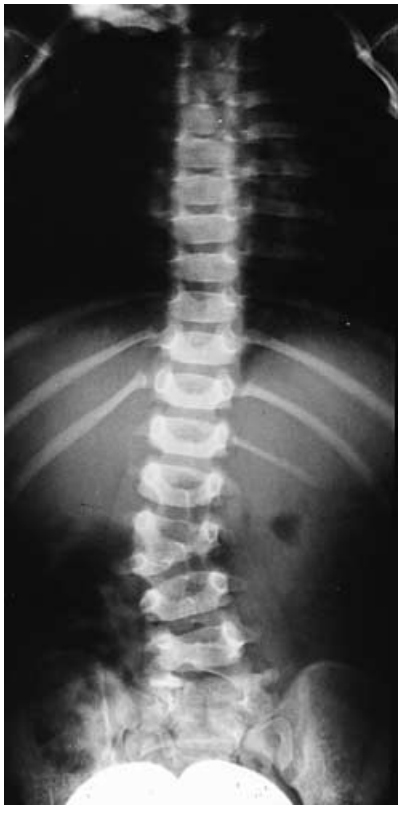

Fig. 3 X-ray of a type 1 scoliosis according to the Nasca classification [11]

- Type 2 is a wedge-shaped hemivertebra and a wedge-shaped vertebra (Fig. 4). It mostly has a triangular configuration. It is not associated with an additional rib at the thoracic spine. Subtype 2a represents a hemivertebra for which the ontogenesis of the other side of the vertebral body as well as of the neural tube has completely failed. It is designated by definition as a wedge-shaped hemivertebra. Subtype 2b is a hemivertebra in which one side of the vertebral body and of the neural tube is underdeveloped. It is designated a wedge-shaped vertebra.
- Type 3 is characterized by multiple hemivertebrae (Fig. 5) that can be round, oval, or wedge-shaped.

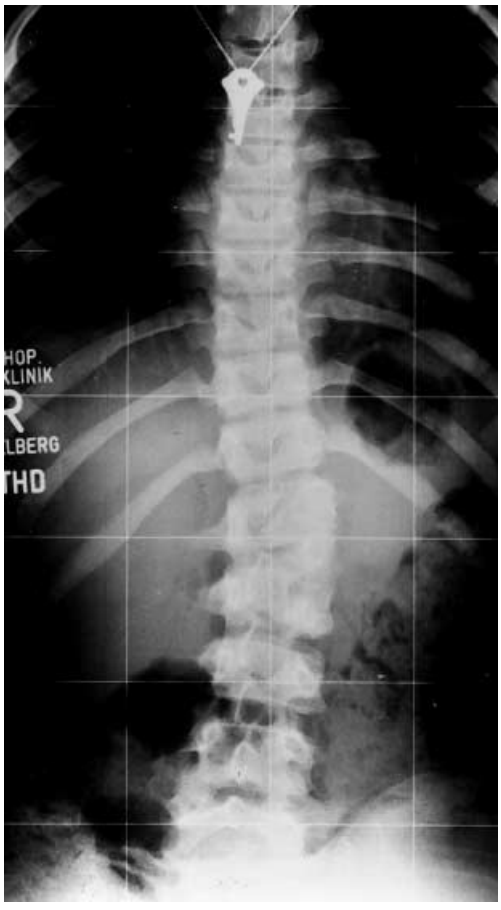

Fig. 4 X-ray of a type 2 scoliosis according to the Nasca classification [11]

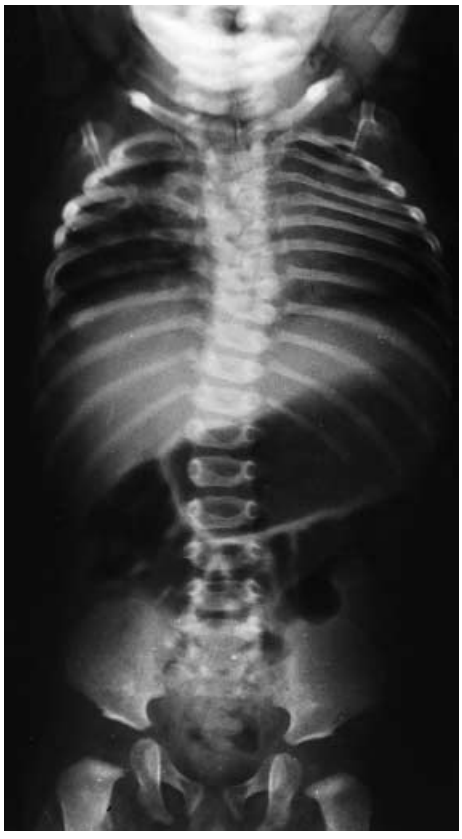

Fig. 5 X-ray of a type 3 scoliosis according to the Nasca classification [11]

- Type 4 is defined by the appearance of multiple hemivertebrae which are fused on one side (so-called unilateral bar) (Fig. 6). There is a further differentiation into two subtypes. Subtype 4a is characterized by multiple hemivertebrae with one-sided fused vertebrae of the vertebral bodies and of the posterior elements of the vertebral body on the concave side. Subtype 4b is characterized by multiple wedge-shaped vertebrae with developing one-sided fused vertebrae.
- Type 5 represents balanced hemivertebrae, localized in such a way that the deforming effects are neutralized by each other. In this way, an extreme scoliosis form can be avoided.
- Type 6 is defined by posterior hemivertebrae (Fig. 7) that lead to a progressive kyphosis rather than to a scoliosis. A kyphosis arises when the anterior part of the vertebral body does not develop as an independent unit.

Cases with butterfly-shaped vertebrae, completely fused vertebrae, or spina bifida were excluded, because they do not belong to the Nasca classification. The measurement of the scoliosis angle was performed according to Cobb [5]. The judgement of the rotational component of the vertebral body was carried out by the method of Nash and Moe [12]. The deviation of the solder was measured by means of X-rays with a comparative raster. The progression of the malformation was analyzed by regarding the changes over a period of on average 8.3 years.

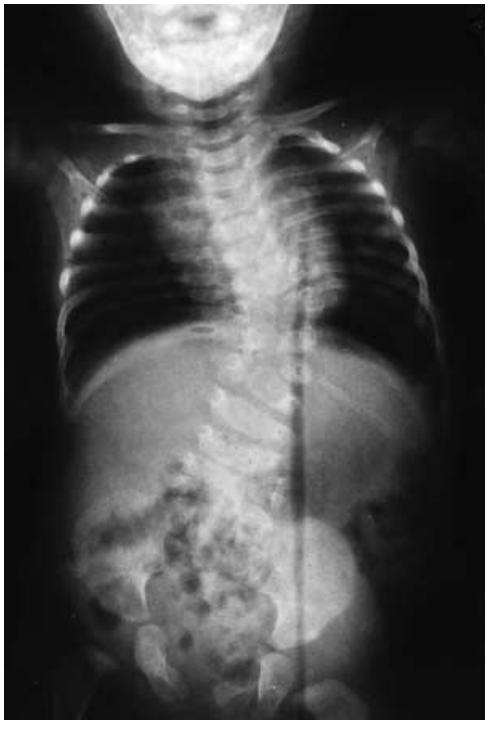

Fig. 6 X-ray of a type 4 scoliosis according to the Nasca classification [11]

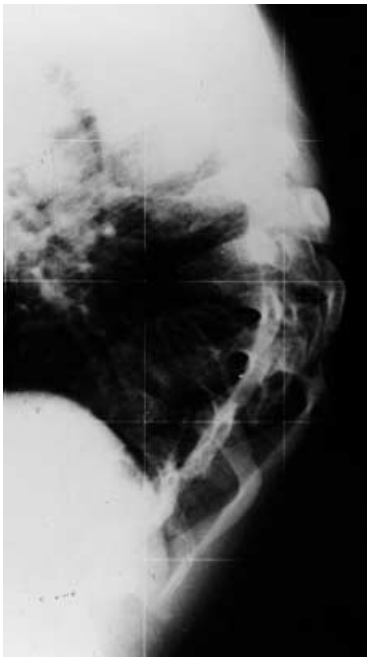

Fig. 7 X-ray of a type 6 scoliosis according to the Nasca classification [11]

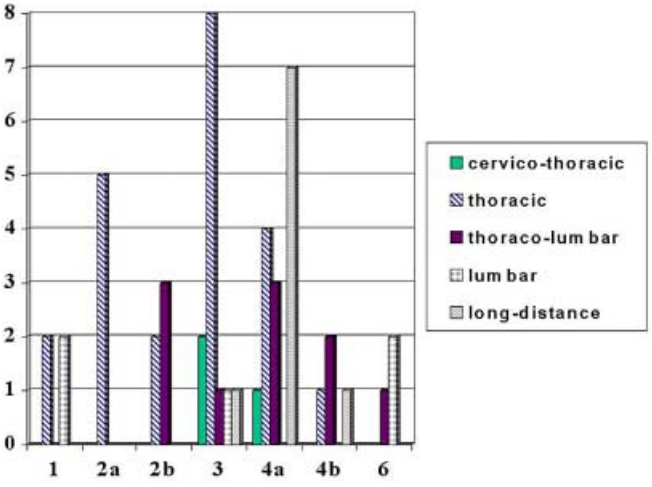

Fig. 8 Localization of the congenital scoliosis curves

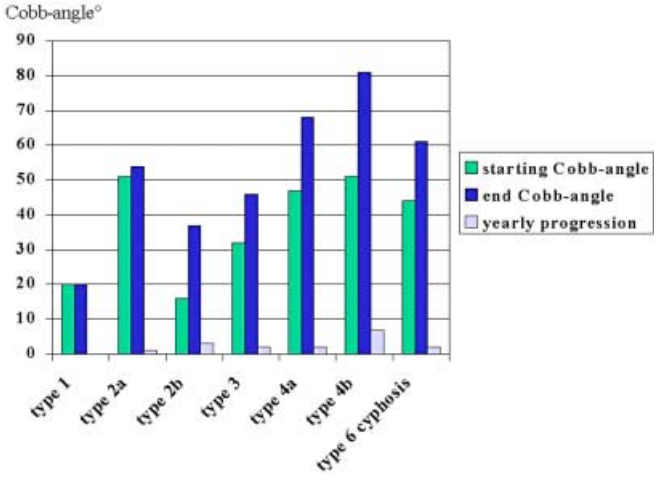

Fig. 9 Variations of the Cobb angle according to the different congenital scoliosis types of the Nasca classification [11] over an observation period of on average 8.3 years

Results

Distribution of the malformation types according to the Nasca classification

The largest group was type 4 [multiple hemivertebrae with developing one-sided fused vertebrae (so-called unilateral bar)] with 19 patients (Fig. 8). Type 5 (balanced hemivertebrae) was nonexistent in our patient collective. Type 6 (posterior hemivertebrae) was localized only thoracolumbar or lumbar. All congenital scoliosis type 2a patterns were localized on the thoracic level.

Development of the scoliosis according to the malformation type

With a Cobb angle of 51°, types 2a and 4b showed the highest scoliosis angles at the beginning of the observation period (Fig. 9). At the end of the observation period, type 4b showed the highest scoliosis angle and the greatest annual progression rate of 6.7°.

Solder deviation and rotation degree according to the malformation type

The solder could be determined in 41 patients (Table 1). Types 4a and 6 showed the strongest solder deviation. A maximal angle of torsion with a degree of one according to the classification of Nash and Moe [12] was seen in types 1 and 6. Types 2a, 3, 4a, and 4b showed stronger angles of spinal rotation.

Table 1 Average solder deviation according to the congenital scoliosis types of the Nasca classification [11]

Nasca classification	Patients	Solder deviation (cm)
1	3	1.4
2a	5	0.9
2b	4	1.4
3	11	1.2
4a	14	2.5
4b	2	1.8
6	2	2.6

Development of the scoliosis according to the localization

The annual progression rate of congenital scoliosis depended on its localization (Table 2). Long-distance scoliosis (43°) and lumbar scoliosis (42°) showed the highest Cobb angles at the beginning of our observation period. The lowest values were noted for the cervicothoracic sco-

Table 2 Average Cobb angle (measured in degrees) according to the localization of the major curve at the beginning and the end period of observation

Localization	Starting point	End point	Average annual progression
Cervicothoracic	27	33	0.7
Thoracic	38	50	1.5
Thoracolumbar	36	55	2.6
Lumbar	42	44	0.3
Long-distance	43	73	3.1

Table 3 Average Cobb angle according to the curve type of the congenital scoliosis at the beginning and the end of the observation period

Curve type	Starting point	End point	Average annual progression
C-shaped	33	55	2.5
S-shaped	43	53	1.4
Multicurved	43	73	3.1

Table 4 Distribution of the different conservative treatment possibilities of congenital scoliosis with respect to the Nasca classification [11]

Nasca classification	Physiotherapy	Spine plaster	Corset	Operation
1	Ø	4	Ø	Ø
2a	Ø	Ø	5	4
2b	2	2	2	Ø
3	2	6	4	4
4a	1	10	11	7
4b	Ø	1	3	3
6	1	1	1	Ø

Table 5 Development of the Cobb angle under conservative treatment at the beginning and the end of the observation period

Therapy	Patients (*n*)	Beginning	End	Average annual progression
Physiotherapy	5	20°	25°	3.6
Spine plaster	18	33°	41°	1.0
Corset	14	57°	64°	8.5

liosis (27°). At the end of our investigations, long-distance scoliosis showed the biggest deviation with a Cobb angle of 73°. This value corresponded with the annual progression rate of 3.1° of the long-distance scoliosis. Lumbar scoliosis showed an annual progression rate of only 0.3° in spite of the high output values. Thoracolumbar scoliosis showed an annual progression rate of 2.6°.

Development of the scoliosis according to the curvature form

By means of the curvature form, the 46 patients with types 1–4 scoliosis were subdivided according to the Nasca classification [5] into 3 groups: S-formed scoliosis, followed by C-shaped or one-curved scoliosis were most frequently seen. Three or more scoliosis curvatures had developed by themselves in 5 patients. The C-shaped scoliosis led to the strongest progression and the greatest curves compared with the other forms (Table 3).

Changes of the Cobb angle by conservative treatment

Physiotherapy. Six patients were exclusively treated with physiotherapy. They had a flexible congenital scoliosis with a Cobb angle of less than 20° (Tables 4 and 5).

Plaster shell or plaster jacket. Twenty-four patients were treated with a plaster jacket or shell. Eighteen of them could be evaluated. The duration of therapy was on average 4.0 years. Follow-up was between 0.6 and 10.4 years. During this time period, the scoliosis of 10 patients increased up to 45°. In 1 patient, the Cobb angle was similar to that at the beginning of the observation period, while in 7 patients it decreased under conservative treatment up to 11°. The average annual progression rate for all patients treated with plaster shell or jacket was 8°.

Corset treatment. Twenty-six of 49 patients were treated with a brace during the observation period (Milwaukee, Ducroquet, or Boston corset). Fourteen of them could be evaluated at the end of our investigations. The average treatment time was 5.1 years. The period of observation was between 1.0 and 10.6 years. The average annual progression of the scoliosis amounted to 8.5°. We saw an amelioration of the Cobb angle in 5 patients of up to 11°. In 1 patient, the Cobb angle remained constant, and in 8 patients we saw a decrease of the Cobb angle of up to 35°.

Operative treatment. Eighteen out of 49 patients were operated on. A total of 19 operations was done (spinal fusion according to the Risser, Harrington, or Cotrel-Dubousset technique). The operations were done on Nasca types 2a (*n*=4), 3 (*n*=4), 4a (*n*=7), and 4b (*n*=3).

Discussion

In their investigation including assessment of congenital scoliosis, Winter et al. [18] found no progression in 25%, a slow progression in 50%, and a fast progression in 25%. We found comparable results in our patient collective. In 36 of 49 patients (73%), we noted a progression of the congenital scoliosis. One patient had an identical Cobb angle at the end of our observation period, and 12 patients (24%) even showed a decrease in the Cobb angle with conservative treatment.

Vertebral body rotation and solter deviation

In our investigation, the average solter deviation increased with the number of vertebral bodies that were involved in the scoliosis profile. Furthermore, we ascertained a clear increase of the Cobb angle with an increase in the rotational degree of the scoliosis profile. A trunk deviation at the side of the unilateral bar is known for thoracolumbar and lumbar curves [9]. This correlated with the results of our investigation, especially for the type 4a according to the Nasca classification [5], where we primarily saw great solter deviations.

Localization and profile of the scoliosis

Since the investigations of Winter et al. [18], it has been thought that the localization and the type of malformation of the vertebral body have a decisive influence on the progression of congenital scoliosis. Both the thoracic and thoracolumbar localization are characterized by an especially poor outcome. Several authors found that the thoracolumbar localization of congenital scoliosis is combined with the greatest risk of progression and a deteriorating course [9, 10, 16], closely followed by congenital scoliosis in the lower thoracic spine. These facts were confirmed by our investigations, too. A favorable prognosis was described in most cases for the cervicothoracic localization of congenital scoliosis [9, 10, 11]. But in some cases, unforeseeable strong progression was found [18]. In our investigations, a cervicothoracic localization showed a favorable prognosis, too. However, we did have a patient with a distinct thoracic counterbending with a Cobb angle of 104°. According to our results, we may state that the thoracolumbar and especially the distinct congenital scoliosis were often combined with a unilateral bar. Surprisingly, patients with a type 2b and partial malformation (wedge-shaped vertebra), when three vertebral bodies were localized in the thoracolumbar area and two in the thoracic spine, showed a higher progression rate in comparison with patients with congenital scoliosis type 2a with complete half-sided malformation (hemivertebra), where all malformations were localized at the thoracic spine. Indeed, we found the greatest average progression (3.1°) and greatest Cobb angles (73°) for multicurved congenital scoliosis. Despite this fact, it is worth mentioning that the different scoliosis profiles with their average Cobb angles were similar: C-shaped 55°, S-shaped 53°, and multicurved congenital scoliosis 73°, so that the scoliosis profile alone is not an expressive predicting factor for the congenital scoliosis.

Age dependence

Considering the age of patients as a possible performance influencing factor for the progression of scoliosis, some authors state that the prepubertal growth phase is the main influencing factor for progression of congenital scoliosis [9, 10, 18]. With our patients, this evolution could not be proved.

The time of most pronounced physical growth is in the first 5 years. During this period, there is particularly strong growth in the upper part of the body [6]. In our investigations, we saw the highest progression rate for congenital scoliosis in patients aged between 0 and 5 years old (annual progression rate 2.8°). After the pubertal growth spurt, a progression of the Cobb angle could not be recognized in most cases [2].

Conclusions

1. Predominantly congenital scoliosis has to be classified as progressive. Nevertheless, we saw in our investigation all gradations of progression from minimal to rapid, independent of the initial Cobb angle.
2. Both the rotational component and the solter deviation are influenced by the malformation type of congenital scoliosis. A long-distance congenital scoliosis is a negative predictor.
3. Most congenital scoliosis types were conservatively treated (for example brace) during the first period of observation. Physiotherapy is the optimal therapy for a nonprogressive congenital scoliosis up to a Cobb angle of 20° [1, 3].
4. For children younger than 2 years with a progressive congenital scoliosis, a plaster shell or jacket should be tried [13, 18]. For brace treatment, a progressive but still flexible congenital scoliosis with a Cobb angle between 25° and 40° is the optimal indication [7, 19].
5. Congenital scoliosis with a Cobb angle over 50°, that shows a tendency to progression, as well as congenital scoliosis with an obviously poor prognosis (for example unilateral bar) should be operated on up to 5 years of age.

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