Relationship of "syndrome of contractures" in newborns with the development of the so-called idiopathic scoliosis

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**Background:** In this article, we describe early clinical symptoms of "syndrome of contractures" (Mau) in newborns and children as well as its relations to the so-called idiopathic scoliosis.

**Data sources:** Two different groups of children were analyzed. The first group consisted of 60 children. They were first examined in newborn and infancy period with hip dysplasia, torticollis or deformities of feet or others. They were examined again at the age of 5-7 years in 1996. All the 60 examined children, who were found primary symptoms of "syndrome of contractures" in histories, showed first symptoms of the so-called idiopathic scoliosis later at the age of 5-7 years. The other group in our research included 123 newborns born in the period of 1998-2000. In this group we analyzed with emphasis on the mothers' conditions during the pregnancy period, the children's conditions in infancy period, and their connection with symptoms of "syndrome of contractures".

**Results:** "syndrome of contractures" could explain the predominance of female patients with scoliosis, sides of curves (lumbar—left convex, thoracic—right convex). What we see "left sided syndrome of contractures" mostly can explain the clinical picture of so-called idiopathic scoliosis: the side of curves—lumbar left, thoracic right, side of the rib hump (right side of thorax), progression in the acceleration period of growth, and the sensibility to new rehabilitation exercises.

**Conclusions:** Newborns with clinical signs of "syndrome of contractures" require further spine examinations at the age of 3-6 years to detect the danger of oncoming scoliosis and to introduce rehabilitation management in the program of the "causal neo-prophylaxis".

**Key words:** "syndrome of contractures"; so-called idiopathic scoliosis; etiology; abduction contracture of the right hip

**Introduction**

deformations of the skeletal system can be related to "syndrome of contractures", which was described by Mau from Germany as "Siebener Kontrakturen Syndrom" ("Seven Contractures Syndrome").[1,2] This syndrome has also been described by Dega,[3,4] Hensinger,[5] Howorth,[6] Green & Griffin,[7] Vizkelety,[8] Komprda,[9] Karski,[10-15] and Tarczyńska, Karski & Konera.[16,17] Dega[3,4] from Poland described the "asymmetry of child's body" in the fetus period as "ultrapositions" in 1932, and he reported that the asymmetry was related to the problem of congenital dysplasia of the hip. Normelli[18] in 1985 described the relationship between idiopathic scoliosis and anthropometric changes in child's body typical for "syndrome of contractures".

In which situations does the "syndrome of contractures" develop? They are connected with mother and fetus. The causes of the "syndrome of contractures" can be related to fetus itself: heavier weight and longer height of the fetus or to maternal conditions: small belly during pregnancy, lack of amniotic fluids and pelvic bone type including anthropoid and platypelloid type which are inconvenient for fetus growth.[14,16,17] The asymmetry in hips and pelvis region is induced by adduction contracture of the left hip and/or the right hip (shortening of the muscles, tendons, fascias). The abduction contracture of the right hip has great influence on the spine in the period when the child starts to stand and walk.[19] In the following years of growth and development, the already 'started' scoliosis develops into its next stages.[20]

**Early clinical signs of "syndrome of contractures"**
The left-sided "syndrome of contractures" are connected...
with the first fetus position during pregnancy in European countries (80%–90%). Clinical symptoms of the syndrome varied according to Mau (Fig. 1).

1. Skull deformity (plagiocephaly) includes mostly flattening of the left forehead and temporal bone, left chick atrophy, eyes asymmetry, deformations of nose and ears.

2. Torticollis muscularis (wry neck) caused by shortening (contracture) of sterno-cleido-mastoideus muscle is usually related to plagiocephaly and/or traumatic delivery or with congenital tumor neonatorum (fibrous tumor in sterno-cleido-mastoideus muscle).

3. Infantile scoliosis, different than idiopathic scoliosis, is usually recedes spontaneously in 80% of cases or even in 100%. Untreated contracture of adductor muscles of the left hip can lead to the development of hip dysplasia. Only 10% of the affected cases can be observed in the newborn period, the remaining 90% cases are of secondary deformity resulting from the contracture and are classified as cases of developmental hip dysplasia (DDH).

4. Contracture of abductor muscles of the right hip, described in Siebener syndrome as weak posture, may cause oblique positioning of the pelvic bone on X-ray picture of the hip joint. With time, asymmetry in movement of both hips causes asymmetry during gait and loading and asymmetry of growth and development of the spine. The contracture of the right hip leads to the permanent habit of standing at ease only on the right leg (the right leg is stronger and more stable due to the contracture), which leads to the development of I-epg, II/A-epg or II/B-epg so-called idiopathic scoliosis.

The division into three etiopathological groups (epg) was described in 2001 and 2004 with additional information on the new classification in 2006. The influence of "gait" and "standing on the right leg" in I-st epg ("S" double scoliosis, both curves at the same time) is especially decisive. In II-nd/A epg ("C" one curve scoliosis) and II-nd/B epg ("S" double scoliosis—lumbar primary, thoracic secondary) the standing habit alone is decisive.

5. Pelvic bone asymmetry is caused by contracture of the adductors of the left hip and/or the right hip which can influence the pelvis positioning visible during X-ray examination for hip joint (Fig. 2) (see above points 4 & 5).

6. Feet deformities include pes equino-varus (club foot), pes equino-valgus (equino-valgus deformity of foot) and pes calcaneo-valgus (calcaneo-valgus deformity of foot).

7. The last clinical observations from Lublin inform that varus axis of shank which can lead (with others influences) to Blount's disease (congenital tibia vara) should also be included into "syndrome of contractures and deformities".

Relation of "syndrome of contractures" to so-called idiopathic scoliosis

The first observations about relationships between "syndrome of contractures" and idiopathic scoliosis in our department were performed in years 1995-1996. We studied medical records of 60 children previously treated in our outpatient clinic because of congenital hip dysplasia, torticollis, feet deformities, plagiocephaly (signs of "syndrome of contractures") in the newborn and infancy period. We recalled these children for another examination including spine examination at age of 5-7 years. In these children we found "first symptoms of early scoliosis". Spine examination demonstrated loss of spine flexion, shifting of spinous processes under the skin Th6-Th12, flattening of the thoracic spine, and tendency in these children of standing at ease on the right leg. According
to biomechanical etiology these are signs of rotation deformity which is the first step in three dimensional (3-D) development of scoliosis and characteristic for early symptoms in the 1-st epg or III-rd epg. In these 5-7 years old children, the angles of curves on X-ray examination were only 5-10 degrees (Cobb degree). The children of these groups were subjected to prophylaxis programs (Fig. 3). We noticed that the percentage of the children with this first symptom of scoliosis was similar to that of those with DDH since both were connected with "syndrome of contractures".1-3,5,14 We concluded from this research that children with clinical signs of "syndrome of contractures" in infancy period can be endangered with scoliosis.

We conducted another study on another group in the years 1998-2000. In this study, we analyzed not only the children (we examined newborns at age of 3-5 days) but also the mothers at the pregnancy period. The aim of the research was to describe the development conditions for "syndrome of contractures" in context of mother and child. Emphasis was put on the pregnancy period and mothers’ conditions. We analyzed the medical records of 123 children (62 male and 61 female) and their mothers in detail. The age of the mothers varied from 18 to 39 years. Syndrome of contractures were noted in 54 children (44%): left-sided accounting for 82% (44 children) and right-sided for 18% (10). Seventy-three (60%) of the 123 children were born at the first pregnancy (60%), 49 mothers had small bellies, of which 38 (71%, 38/49) were "flattened" during pregnancy, and 9 (7%, 9/123) mothers were informed of lack of amniotic fluids. Fifty-nine newborns (48%) at birth were heavier or taller than the normals. These examinations of mothers and infants can indicate the conditions leading to "syndrome of contractures".

"Syndrome of contractures" and biomechanical etiology of so-called idiopathic scoliosis

"Syndrome of contractures" can explain some pending questions about the etiology of idiopathic scoliosis: 1) why scoliosis occurs mostly in girls? 2) why the lumbar left convex and thoracic right convex curve occur? 3) why the rib hump is on the right side? 4) why some scoliosis is associated with progression but some not? 5) why there is rapid progression of scoliosis in the acceleration period of growth?

Scoliosis occurs mostly in girls because the contracture of the right hip that was connected with the "syndrome of contractures" mostly seen in girls (the ratio of boys to girls is 1:5).1,2 In our study, girls predominated for 73%.

The curves of the lumbar left convex and thoracic right convex as well as the rib hump on the right side are related to the "left-sided syndrome of contractures" in 85%-90% of pregnancies when the fetus is on the left side (the first longitudinal position of fetus) of the belly of the mother.21-23

The types of scoliosis ie, "S" (I-st epg), "C" (II/A epg), "S" (II/B epg) and "I" (III-rd epg) are dependent on the range of abduction contracture or limited adduction of the right hip in comparison with the range of adduction of the left hip20 and other causes.11,15 The co-factors for development of scoliosis include walking and standing position on the right leg.15

Progression of scoliosis in the acceleration period of child's growth is related to asymmetrical growth of
borders and soft tissues. "Syndrome of contractures" limits the proper growth of bones. Thus fast progression of scoliosis is due to biomechanical influences especially in the I-st epg. The asymmetry of movement of both hips, which leads to the asymmetry of loading during gait, next leads to the asymmetry of growth of the child and to the asymmetry of development of the spine, for instance, the asymmetry of growth of vertebra bodies and processes. The asymmetry of forces/loading due to compression on the concave side of curve decreases the growth ("vicious cycle") according to Heuter-Volkmann effect. The explanation is as the following: asymmetrical loading causes asymmetrical growth, then the wedging of vertebra and discs and later the next stages of spinal curvature. The greatest influences of this "vicious cycle" is in "S" scoliosis from the I-st epg (double curves scoliosis as 3D deformity of spine), the smaller influence is in "C" II/A-epg and "S" II/B-epg scoliosis.

The growth of legs in scoliotic patients is faster than the growth of trunk and because of this the scoliosis in I-epg is with great progression in the acceleration period of growth. But we did not observe the progression of curves in the II/A epg ("C" scoliosis), II/B epg ("S" scoliosis) and III-rd epg ("I" scoliosis—stiffness of spine but without curves and without rib hump). In II/A epg and in II/B epg the standing position is an isolated causative factor for the development of curves but it depends on the time for standing on the right leg (hours, days, years) and other influences like laxity of joints, etc. The reacting influences on the spine are much smaller than those on the I-st epg where the dominating factor is gait together with standing. In the III-rd epg the gait is the main factor for the development of stiffness of the spine but the additional factor of standing at ease on the right leg is not present since the adduction of the left hip is also limited and the time of standing at ease on the left and the right leg is the same or very similar (Fig. 4).

Other important factors for biomechanical etiology of "scoliosis"

Progression in the I-st epg is especially fast in children with joint laxity, rickets, and anatomy anomalies of the pelvis and lumbar spine (spina bifida occulta), deformities of chest and ribs (pectus infundibuliforme). Early clinical signs in very young children (1-3 year of life) with danger of scoliosis are "straight position/axis of the spine" or later "stiffness of the spine" with "flat back" and the habit of permanent sitting straight up and standing "at ease" only on the right leg. Our observations in the last 20 years showed that rapid progression of scoliosis is also related to extension-strengthening exercises which are completely wrong in our opinion and experience.

Discussion

In many countries the search for the etiology of idiopathic scoliosis is going on. In the literature about idiopathic scoliosis and at the last meeting of International Research Society for Spinal Deformities (IRSSD in June 2006) the following opinions about etiology were presented: genetic (Brown), primary anatomical disorders (Adams, Świderski), biochemical factors (Skogland, Lowe), hormonal (melatonin, prostaglandin [Skogland]), neurogenic/muscles (calmodulin [Lowe]), imbalance of muscles / left & right side (Zuk, Wejsflog), pineal gland (K. Bagnall, F. Nette, J. Mahood, X. Wang, H. Jiang), labirynitus (G. Kapetanos, Potoupnis M. Dangilas Ang., Markou, K. Pournaras, J. Aristotle), faster growth of "convex side of scoliosis", and asymmetry in growth of vertebrae and the spinal cord.

We found the asymmetrical movement of the right and left hip, and that the asymmetrical movement between the right and left sides of the pelvis during gait disturbed the growth and function of the spine since a child starts to walk. Clinical symptoms of scoliosis in the I-st epg developed many years before the deformity was clearly visible on X-ray examination. In children with scoliosis, deformities in "syndrome of contractures" include plagioccephaly, torticolis, asymmetry of the temporal bone, functional shortening of the left lower extremity, tilting of the pelvis, and asymmetry of the whole body, which confirm the relations between "syndrome of contractures" and the so-called idiopathic scoliosis.

We noted the citations in their reports by Normell and others:

1. Willner (1972): in general the left leg tends to be shorter than the right in childhood and this leads to development of the left convex lumbar curve. Pelvic obliquity has been observed in structural scoliosis.

2. Magoun (1974): asymmetry of temporal bones has also been associated with scoliosis.

3. Wynne-Davies (1975): plagioccephaly has been considered to be closely related to infantile idiopathic scoliosis.

4. Dangerfield and Col (1995): as with the plagiocephaly, the body asymmetry in children with scoliosis is as yet unexplained.


6. Tylman (1995): tilt of the pelvis is an important
sign of the development of scoliosis.


All these observations confirm the relationship between "syndrome of contractures" and scoliosis, which in our research also explained many secret "geographic" signs in scoliosis.

The sensitivity to new rehabilitation exercises[38] underlines the biomechanical influences coming from the asymmetry of movement of both hips which primarily comes from "syndrome of contractures" in early development of so-called idiopathic scoliosis.

Conclusion

1. Detailed examination for newborns and infants helps to discover symptoms of "syndrome of contractures". Early prophylaxis should be introduced in these children in accordance to the types of deformation of the skull, neck, spine, hips and feet.

2. Children at age of 3-6 years should be examined to discover if there is any difference of adduction of the hip. In cases of asymmetrical adduction (in a straight position of the hip) and habit of standing position on the right leg, they should undergo periodically clinical and radiological spine examination.

3. Asymmetry of the pelvis shown by X-ray images of infants (in DDH screening) should be recognized as a possible sign for pathological changes of the spine in children of 3-4 years old or more.

4. According to our observations, infantile scoliosis is not a so-called idiopathic scoliosis, or even "the first stage of so-called idiopathic scoliosis". The occurrence of the so-called idiopathic scoliosis is accidental in such children.

5. All children should sit physiologically, never straight up, should sleep in fetal position and stand at ease on the left leg as an important protection against scoliosis. Early prophylactic programs should be implemented in children as young as 3-5 years old. They should make flexion exercises of the spine in laying position. The best exercises are stretching like "warm-up" in martial art techniques: kungfu, karate, taekwondo, taichi, aikido, yoga, etc. Walking in a manner of "toes in" also can protect against scoliosis.

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