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“Syndrome of contractures and deformities” according to Prof. Hans Mau as the primary cause of motoric deformities in children. Case studies including deformities of hips, neck, shank and spine

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Summary

Authors claim that the problem of deformities in children and pain in adult patients can be connected to shortening of soft tissues followed by contractures of joints. The „syndrome of contractures” (SofC) described by Prof. Hans Mau - as “Syndrome of seven contractures”, explains influence of the foetal position during pregnancy to future asymmetrical contractures of multiple joints (an infantile scoliosis, torticollis, limitation of abduction of left hip, feet deformations). The “left sided syndrome of contractures” is more common, since 85% - 95% of pregnancies are cephalic presentations, first position pregnancies.

In years 2005 – 2013 authors examined 818 new-borns and infants with SofC. The control group was 212 healthy children, without signs of the SofC.

Analysis of the material confirmed the presence of syndrome of contracture signs described by Prof. Mau. Effective treatment of children with SofC is possible and it should be used as prophylaxis of future insufficiency of movement of knees, hips and spine in adult age.

Key words: syndrome of contracture, weak posture, torticollis

Introduction

Orthopaedic literature discusses mainly deformities in children and pain in adults in the context of weakness of muscles. Many authors claim that “the muscles are not strong enough”. In our clinical observations, the problem of deformities in children and pain in adult patients proved to be connected with asymmetric or symmetric shortening of soft tissues even causing contractures of joints and not with “the weakness of muscles”.

Causes of „the syndrome of contractures” (SofC). The „syndrome of contractures” (Fig. 1a, 1b): has been described primarily by Prof. Hans Mau - as Sieberner [Kontraktoren] Syndrom” (syndrome of seven con-

tractures) [1]. The elements of SofC has been also described by: Hensinger [2], Howorth [3], Green & Griffin [4], Dega [5], Vizkelety [6], Glauber & Vizkelety [7], Kane [8], Karski J. [9,10,11,12], Karski T. [13,14], Kalkucki [15] and Watson [16]. The origins of the „syndrome of contractures” are influenced by the foetal period of life that is bigger than normal the weight and the length of the foetus, the small size of belly during pregnancy, the reduced amount of the amniotic fluids (oligohydramion), “androidal” or “platypeloidal” anatomical characteristics of the pelvis. In the cases of the asymmetrical contractures of joints also CNS may be an additional cause. The “left sided syndrome of contractures” is more common, since 85% - 95% of pregnancies



Fig. 1a. 1b. Typical clinical symptoms of „Syndrome of contractures and Deformities” in newborns and babies
 Fig. 1b. Asymmetries in head, neck, spine, hips (later dysplasia), pelvis. „Haltungsschwäche according to Hans Mau is equal to the abduction contracture and often external-rotation and flexion contractures of the right hip according T. Karski (1995 - 2007). This phenomenon explains many questions concerning the so-called idiopathic scoliosis and other deformities stated and unanswered till 1995.

are cephalic presentations pregnancies with foetus lying of the left side of uterus (first position) and this position of the foetus influences future deformities. In cases of the “syndrome of contractures and deformities” the following may be observed frequently: an infantile scoliosis, torticollis and limitation of abduction of left hip (Fig. 2a, 2b, 2c) [Barlow 17, Golden 18, Cheng 19].

2, 3, 5, 6, 14],

5. *contracture (shortening) of abductor muscles and soft tissues of the right hip* (acc to T. Karski), described as *Haltungsschwäche* (“weak posture”) by Mau. With time, asymmetry in movement causes asymmetry during gait and loading; causing so-called idiopathic scoliosis (Karski 1995-2006) [9, 10, 11, 12, 13, 21, 22,



Fig. 2a, 2b Typical „syndrome of contractures and deformities”. Fig. 1a and 1b child Anna 1 month old. Changed axis of spine (scoliosis infantilis). Limited abduction of the left hip – first symptoms of dysplasia. Fig. 2c – child Kryspin 2 months old. Bigger limitation of abduction of left hip. Bigger dysplasia.

Clinical symptoms of the “syndrome of contractures” according to Mau (Fig. 1a, 1b, 2a, 2b, 2c):

1. *scull deformity /plagiocephaly/*,
2. *torticollis muscularis (wry neck)* [8, 12, 15, 16, 18, 19, 20],
3. *scoliosis infantilis (infantile scoliosis)* – other than idiopathic scoliosis [1, 9, 10, 13, 16, 21],
4. *contracture (shortening) of adductor muscles of the left hip*. Untreated contracture can lead to development of developmental hip dysplasia (DDH acc to Klisič) [1,

6. *pelvic bone asymmetry* – the oblique pelvis positioning visible during X-ray examination for hip joint screening – [see above points 4 & 5];

7. *Foot deformities* – such as: *pes equino-varus*, *pes equino-valgus*, *pes calcaneo-valgus* [1, 15, 23, 24].

In 2006, the research conducted in Lublin allowed to add one more symptom to the series of symptoms enumerated above (T. Karski) and now we speak about “Syndrome of Contractures and Deformities” (SofCD). The studies proved that newborns and babies may have



Fig. 3a, 3b Proper nursing of the child. Hips in maximal abduction. Head and neck turned to the left side for treatment of left sided wry neck.

Fig. 3c, 3d Improper / incorrect nursing of the child. Hips without abduction. Danger of the development of dysplasia.

8. *excessive shank deformity (crura vara)* which later can lead under special conditions to Blount disease [25].

Material

In the years 2005 – 2013 authors examined 818 newborns and babies with the signs of the “syndrome of contractures”. The control group included 212 children presented for examination, without signs of the “syndrome of contractures”.

Results

Hips

The dysplasia of the hips can be connected with a/ Syndrome of Contractures and Deformities (SofCD), with b/ laxity of joints and with c/ disorders of central nerve system influencing muscular system in the hip region. The dysplasia of the hips in SofCD shows in babies as a deficit of abduction of hip, mostly on the left side (Fig. 2b, 2c). The physiological treatment of patients in this group of deformation is simply, easy and gives good results. In therapy, it is important to carry the child in full abduction and flexion of hips permanently, for as long as the baby requires carrying (Fig. 3a, 3b)

as opposed to caring completely incorrectly that is without abduction of the hips. The wrong way of carrying is frequently recommended by “wrong-educated physiotherapies and orthopaedic surgeons (Fig. 3c, 3d). To help achieve this proper position of hips joints we use orthopaedic devices (Fig. 4a, 4b, 4c, 4d, 4e). or successful treatment and for full “restitutio ad integrum” a period of a few months or sometimes years is needed if the child suffers from dysplasia accompanied by laxity of joints. Our recommendation is not to allow the child walk before the age of 1. Standing and walking before the age of 1 is a mistaken therapy and should be forbidden. If dysplasia of the hips in not cured by the time the child is 1 year, we suggest prolonged therapy of spongy triangle for walking or Weickert treasures – model for walking (Fig. 4c).

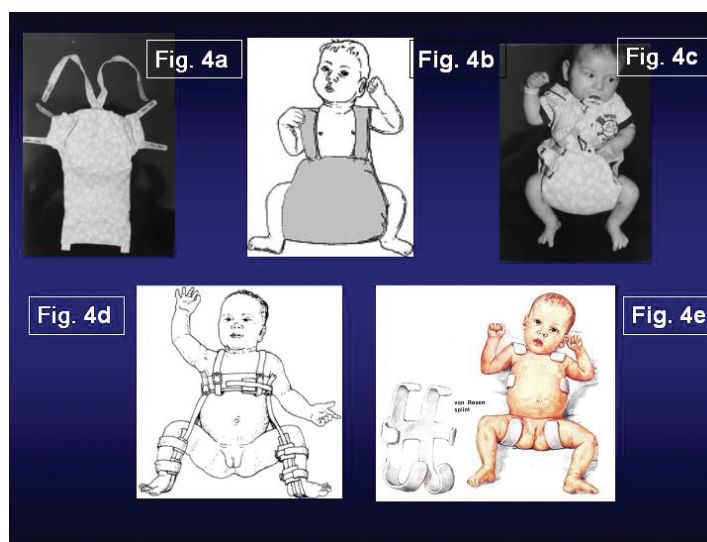


Fig. 4a, 4b, 4c, 4d, 4e Orthopaedic devices for the treatment of the hip dysplasia. Frejka pillow (4a, 4b), Weickert treasures (4c). Pavlik splints (4d), Von Rosen splint (4e).

Neck

Wry neck, called in Latin torticollis muscularis or torticollis myogenes, is a very common problem (Cheng JC and coll., Golden KA and coll., Kane AA and coll., Watson GH., Rogers GF. & Mulliken J.) in newborns and babies (6% to 8 % in Polish population) (Fig. 5a) There are three types of this deformity:

- a/ connected with SofCD
- b/ following traumatic delivery
- c/ congenital wry neck with tumor neonaorum

The treatment of wry neck is frequently performed wrongly with the old method of physiotherapy (Fig. 5b, 5c). Consequently, the therapy never gives sufficient results. For proper treatment permanent “ro-

tation and stretching” is advised (Fig. 5a, 6b, 7a, 7b, 8). This method of therapy was successfully introduced in Lublin in 1974 (T. Karski) for the treatment of children aged from 1 to 6 – 8. It is worth mentioning that before 1974 we were operating 2-3 cases of wry neck each week in Paediatric Orthopaedic and Rehabilitation Department of the Medical University of Lublin. After 1974, only 2-3 cases were operated annually (data from the Department documents; Prof. T. Karski was the Head of this Department in years 1995 - 2009).



Fig. 5a, 5b, 5c Julia, 3 months old. Right sided wry neck (5a). Improper method of physiotherapy performed in one of Centres of Rehabilitation in Poland (5b, 5c). In result there is only paresis plexus brachialis - on the right (arrow). Such treatment for the wry neck brings no results.



Fig. 6a, 6b Julia, 3 months old. Right sided wry neck. Proper method of physiotherapy in Lublin. Permanent stretching exercise of the head and neck by twisting to the right side and keeping of the upper right extremity in abduction and in external rotation. Such non-invasive therapy gives good results after the period of 8 month.

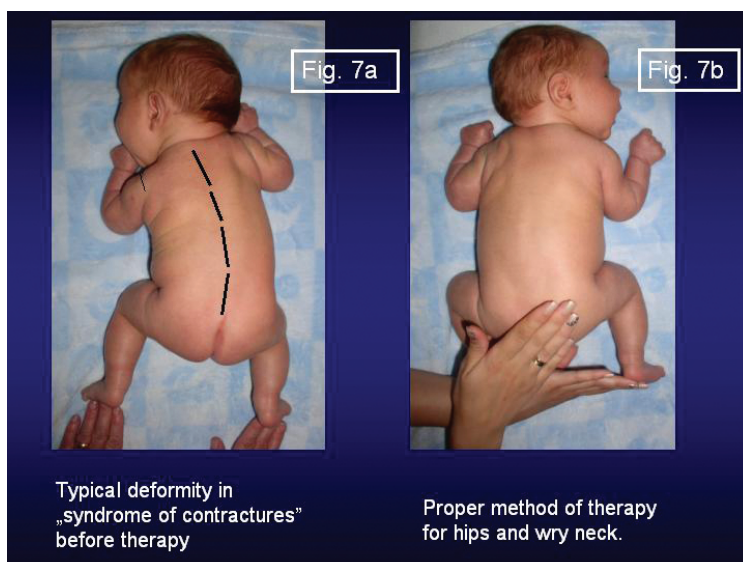


Fig. 7a, 7b Bartosz 6 weeks old. Typical „syndrome of contractures to the right side”. Infantile scoliosis. Right sided wry neck. On the left - before treatment. On the right - during treatment (7b). Proper position for the hips, neck and spine.

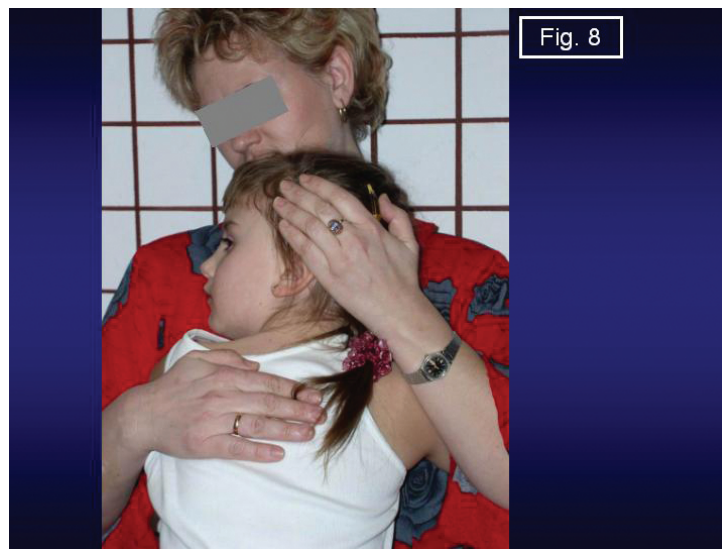


Fig. 8. Stretching-rotation treatment to the left side for left sided wry neck is a successful method even for older children, requiring a longer treatment period even 2 – 3 years.

Shank

The varus deformity of shank was added to the Syndrome of Contractures and Deformities in 2006 - T. Karski (Fig, 9). When the varus of shank is bigger than “normal”, the doctor should be alarmed. In result of adaptation to the space in uterus every newborn is born with varus deformity of the lower extremities. Conditions for the development of varus deformity called Blount disease are: a/ standing and walking before the age of 1, and b/ insufficient vitamin D supply which may lead to easier development of the Blount disease, that is varus of shank at the knee level. Initially, the Blount disease in all cases is bilateral. In the following years one shank, mostly left one can get corrected spontaneously (Fig. 10a, 10b, 10c).

All cases of Blount disease till 1981 were operated. After long consideration, we concluded that the deformity develops according the Heuter Volkmann law. Yet, there are cases when the law does not apply or is not in function, the axis of shank is spontaneously corrected. Until 1981, for therapeutic reasons, we were advising parents not to allow the child to stand or walk for 2-3 months, but to carry the child. After this time the axis of leg comes to normal in all treated cases.

Spine – scoliosis

The biomechanical aetiology of scoliosis has been presented since 1995 (Karski T., Karski J. and coll., Kałakucki and coll.) and now (2014) is accepted in many countries and by many orthopaedic surgeons, rehabilitation doctors and physiotherapists. At the beginning, in development of the so-called idiopathic scoliosis there were: 1/ asymmetry in hips movements connected with SofCD, 2/ the influences connected with walking. Our explanation is as follows: the restricted movement of right hips is transmitted to the pelvis and to the spine and causes the rotation deformity and stiffness of spine, next

the second cause works that is the permanent standing ‘at ease’ on the right leg. In the new classification of the so-called idiopathic scoliosis there are three group and four types of spine deformity (Fig. 11, 12, 13, 14). During the therapy of every types of scoliosis new proper exercises include only stretching, never strengthening.

Discussion:

Authors as Hensinger, Howorth, Green and Griffin, Dega and Vizkelety described some elements of SofC as plagiocephaly, dysplasia of the hip, feet contracture. The first who connected all those asymmetrical deformations into one syndrome was Prof. H. Mau. In our observations we confirmed presence of syndrome of contractures and its influence on locomotor system in children like dysplasia of the hip, Blount disease and wry neck, but also in youth and adults usually as painful contractures and deformations. Additionally we found influence of SofC in the biomechanical aetiology of the so-called idiopathic scoliosis.

Conclusions.

1/ Every paediatric orthopaedic surgeon and paediatric doctor as well every physiotherapist should be acquainted with the knowledge about the “Syndrome of Contractures and Deformities” (SofCD) according Prof. Hans Mau,

2/ Every newborn and baby with SofCD should be treated very early by proper nursing and proper orthopaedic devices, older children should perform stretching exercises in order to reach symmetry of joints’ movements. A surgery is sometimes necessary and performed in orthopaedic wards,

3/ Effective treatment of small children with “residual changes of SofCD”, should be treated as an ef-

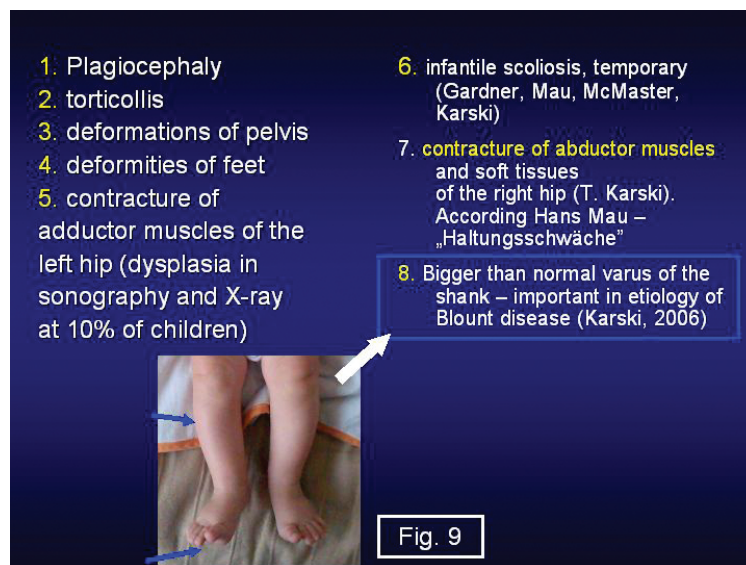


Fig. 9 Clinical symptoms of the „syndrome of contractures and deformities” according to H. Mau and T. Karski

fective prophylaxis of movement insufficiency of knees, hips and spine in adult age.

4/ The physiotherapy of problems of hips, neck, spine should aim at achieving the symmetry of position, symmetry of movement of joints and also symmetry of growth. Stretching exercises should be used as a treatment method for achieving the symmetry of the shortened side or part of body, and region of joints. Strengthening exercises proved to worsen the asymmetry!

5/ Additionally, thermotherapy, magnetic fields, biodynamic (diadynamic), laser, massage can be used for the treatment.

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Fig. 10a, 10b, 10c. Varus deformity of shank (crura vara). On the left (Fig. 10a) typical Blount disease, In the middle (Fig. 10b) Blount disease accompanied by rickets. On the right (Fig. 10c) one-side Blount disease. Left leg - self correction. Treatment of children aged 1 – 3 only by physiotherapy (see the article above). Older children – surgery.



Fig. 11. A healthy child. No deformation of spine. Symmetry of hips and movements. Symmetry of the movement of the trunk both the left and the right side during gait. Symmetry of standing time left vs. right leg.

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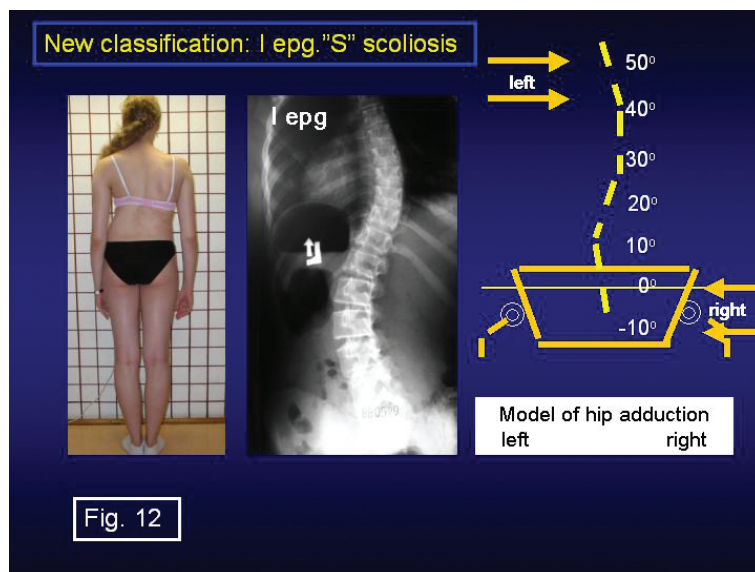


Fig. 12 „S” scoliosis in I epg, 3D, primary double curves – connected with gait & standing ‘at ease’ on right leg. First rotation deformity, next curves. Stiffness of spine. Progression.

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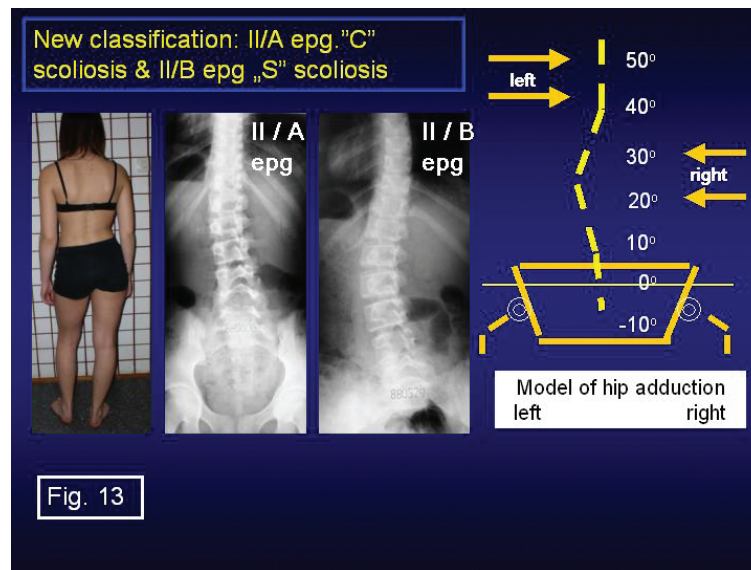


Fig. 13 "C" scoliosis - II / A epg, 1D, connected with standing 'at ease' on the right leg, "S" scol. in II / B, 2D or mix / flexible spine. Thoracic curve - secondary. Aetiology for "S" II/B: standing 'at ease' on the right leg, additional laxity of joints and / or wrong exercises in therapy. No progression or small.

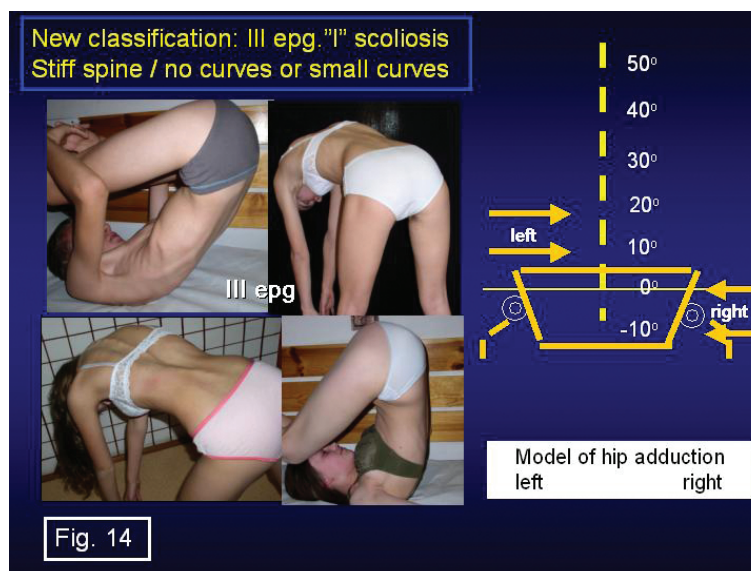


Fig. 14. "I" scoliosis III epg – connected with gait only. 2D or mix. Stiffness of spine. Small curves or no curves. In adulthood - pain. No progression.

