Perthes disease. Etiology. Symptoms. Physiotherapy

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Abstract
Many Pediatric Orthopedic Departments in Europe treat children with aseptic necrosis of femoral head. The illness is known in orthopedics as Legg - Waldenström - Calve - Perthes disease, described as the Perthes disease, or LCP disease. The treatment in all Orthopedics Centers mostly is similar - conservative, by physiotherapy and in cases with deformity of proximal end of femur-like varus deformity, partially dislocation / lateralization of femoral head-is done surgery. In many congresses and symposia-discussions about causes of the Perthes disease are very controversial. In our article we describe the causes of this illness. We are providing a lot of evidence that etiology is connected with trauma and concerned mostly children with symptoms of Minimal Brain Dysfunctions (MBD).

Keywords: Necrosis of femoral head, Etiology, Symptoms, Therapy.

Introduction
The illness-as an “aseptic necrosis of femoral head” was described by Legg, Waldenström, Calve, Perthes-but in the “medical world” is known as the “Perthes disease” (Figure 1). Perthes disease - aseptic necrosis of femoral head - usually occurs in children aged 4 to 11. The main feature of the illness are disturbances in blood supply of femoral head and its gradual destruction leading to hip joint insufficiency and early degenerative changes. The influence factors - trauma - acting on the femoral head - to bone and blood circulation system (Figure 2, 3, 4, 5). The clinical and radiological symptoms are significant and similar or identical in all patients (Figure 6).

Figure 1: Legg - Waldenström - Calve-Perthes Disease. Authors' photos in the chronological order (according to the Internet).

Necrosis of femoral head was described:
(1) By Arthur Thornton Legg in 1908
(2) By Johann Henning Waldenström in 1909
(3) By Jacques Calve in 1910
(4) By Georg Clemens Perthes in 1910

Figure 1: Legg - Waldenström - Calve-Perthes Disease. Authors' photos in the chronological order (according to the Internet). The necrosis of femoral head was described: (1) By Arthur Thornton

Figure 2: Anatomy of the hip joint (picture taken from M. Dudin’s lecture). 1/ Epiphysis 2/ Cervix (collum) femoris 3/ Acetabulum 4/ Trochanter major 5/ Trochanter minor 6/ Capsula articularis.


Figure 5: (A) Anatomy of hip joint in child. Broken spongy bones in subchondral part of femoral head after weeks/months of jumping. In result disturbed blood supply and start of all stages of Perthes disease (pictures taken from M. Dudin’s lecture).

Figure 6: Symptoms and stages of Legg-Waldenström-Calve-Perthes disease. From the first years of activity of University Orthopedic Department in Lublin, Poland- founded on December 7, 1954-we have treated the children with this illness, but therapy has been changed many times. First, we used mostly orthopedic devices, later-children were mostly operated, in last years of XX and beginning of XXI century-kinesiotherapy and physical therapy were used mostly for treatment.

In literature, many various conceptions about etiology can be found, like: hormonal causes, valgus of knees and improper loading of femoral heads, too high “intra-articular pressure in the joint” and other like genetic causes, abnormalities in blood contents, genital disturbance of growth plate / the proximal “discus epiphysealis” of femur [1-14]. In the article we describe the “trauma-causes” of Perthes disease responsible in all cases with presented clinically and radiology symptoms (Figure. 6).
Material
In the years 1995-2009 (in this time Prof. T. Karski was the head of Pediatric Orthopedic and Rehabilitation Department of Medical University in Lublin, Poland) among patients in the Department, there were in every time 5 - 6 children with the Perthes disease. Together in period of 14 years there were 133 children treated with Perthes disease (Figure 7, 8, 9, 10). In years 2010-2020 (material gathered by Dr J. Karski) in the treatment because of Perthes disease there were 83 boys (84 %), 16 girls (16 %) Together-99 patients (100 %) (Table 1). Some children have been admitted to the Department 2 or 3 times-there were 126 hospitalizations altogether in 10 years. Age of children was from 4 to 11 years old.

Figure 7: Clinical example. Adam S. born 20.12.1989. Perthes disease right hip. First Clinical Symptoms-July 1998. In anamnesis - Jumping - 30-50 times per day in school, jumping from couch at home. X rays from years 1998-2000. Phases of illness: (A) sclerotic, (B) & (C) fragmentation, (D) reparation.

Figure 8: Clinical example. Karolina K. born 20.03.1993. Perthes disease right hip. First Clinical Symptoms-July 1998. In anamnesis - Jumping 30-50 times per day in school from steps (from 4th step to the floor). X ray pictures from years 1998-2001. Phases of illness: (A) & (B) sclerotic, (C) fragmentation, (D) reparation.

Figure 9: Clinical example. Miroslaw M. born 16.02.1989. Perthes disease right hip. First Clinical Symptoms-May 1997. In anamnesis - jumping 30 - 40 times per day in school from steps (from 4th step to the floor). X ray pictures from years 1997-2000. Phases of illness: (A) & (B) sclerotic, (C) fragmentation, (D) reparation.

Epidemiology and Clinical Symptoms

In the years 1995-2020 in our material the Perthes disease occurs from 3 to 5 times more frequently in boys. Children usually complain about pain in inguinal region affected hip. In some cases, the pain was in distal - medial part of thigh or even in knee region. As one of the first symptoms, except for pain, children had problems with walking or even they limp. In the beginning of illness, the pain of the hip joint was not dominating and not permanent but disappeared for some days or weeks and again appeared - it was erroneous for parents and the cause of delayed diagnosis. In clinical examination, limitation of abduction and internal rotation of the affected hip joint have been noted in all children. Additional symptoms were fatigue of function of lower extremities and atrophy in thigh but especially in buttck muscle. In some children, as a result of “the painful adductor contracture” a slight leg length discrepancy appeared - shortening of the affected leg.

Our Observations About Etiology in The Perthes Disease

In our Pediatric Orthopedic and Rehabilitation Department in 1980 - 1990 we found that the Perthes disease is connected with trauma-jumping, many times every day, many weeks or months of the children in age 4-11 years. It concerns the children with symptoms of Minimal Brain Dysfunction (MBD) and is connected with special “psychological changes” of this group of patients [15-25]. From a psychological point of view - these children are nervous, they cry frequently, are easily offended and also they like to jump, the trauma is minimal but repeated many times gives “cumulative / summarized overloading” of the femoral head or even fractures of the spongy bones and therefore influences the blood circulation in form of restriction to the femoral head. It is the cause of all aseptic changes and all stages of femoral head necrosis, called “Perthes disease”. In article we describe clinical symptoms, X-ray changes and present examples of this illness. We also are giving the method of physiotherapy. The presented material is mostly from the years 1995-2009.

Psychological and Clinical Symptoms of Perthes Disease in Connection with Minimal Brain Dysfunction (MBD)

As mentioned above this illness we observe in children in age of 4-11 years. More frequently among boys. In many of these patients we had to see not only above described psychological symptoms of Minimal Brain Dysfunction (MBD) but also “orthopedic symptoms” as sign of MBD [15-25]. The MBD is more frequent in boys [14-25]. In years 1970-1980 in all children with Perthes disease we noticed the limitation of dorsal flexion of the feet but in this time these symptoms were “secrete for us and their origin unknown”. Over time, we started to perceive these symptoms as a sub-spastic contracture of m. triceps surae. They are typical for children with MBD. At these patients we see also very often sum-spastic contracture of flexors of knees-m. semitendinosus, m. semimembranosus, m. biceps femurs and m. gracilize. What is more, precise anamnisis from the parents confirms all psychological changes in the child’s behavior. The children are very sensible, with variable emotions, and-what was in every case confirmed - they like to jump. Hyperactive and impulsive boys are the most endangerd to the disease. Frequent jumping-many times per day, many weeks, or months causes the fractures in sub-chondral part of the femoral head, it disturbs the blood circulation in bone, what means the first stage of the Perthes illness. About traumatic theory of etiology in the Perthes disease had spoken himself Perthes and Calve, also Dimeglio, Dudin in St. Petersburg and in Lublin-Karski T., Karski J., Kandzierski G. and Kalakucki J. All these authors underline the importance of repeated „micro-traumas” causing mechanical overloading and over-stressing of hip joint leading to sub-chondral fractures of spongy bones with first symptoms in the region of the growth zones, leading to disturbances in blood supply of whole femoral head.

Information about physiotherapy. Since 1991 till now in the treatment we have used physiotherapy methods- kinesiotherapy and physical methods as well as for the Perthes disease as for other illnesses of femoral head (see Discussion). Limiting the loading of affected leg in first months of the therapy is especially important. Exercises aiming to recover the movement of affected hip, and in the first place, the abduction and internal rotation are essential. Exercises in warm water, especially in geothermal water are highly efficient. All the exercises should be done in prone position of the child's body-only in this position hip / hips receive also full extension. Resting during the day and sleeping - at least partially - should be in prone position with flexed and abducted affected hip / leg. Laser therapy is very important, what has been confirmed in many cases in Lublin and in St. Petersburg (Figure 11). Here, we would like to emphasize, that in the therapy of the children with MBD - not only physiotherapy but also permanent positive psychological stimulation is very important-parents' love for the child in every day, over years.

Table 1: Patients with Perthes disease in period of 2010 till 2020 in Lublin Pediatric Orthopedic and Rehabilitation Department

<table>
<thead>
<tr>
<th>Perthes disease</th>
<th>Boys</th>
<th>Girls</th>
<th>Together</th>
</tr>
</thead>
<tbody>
<tr>
<td>2010</td>
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<td>0</td>
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<tr>
<td>2011</td>
<td>6</td>
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<td>2012</td>
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<td>1</td>
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<tr>
<td>2020</td>
<td>5</td>
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<td>6</td>
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<tr>
<td><strong>Together</strong></td>
<td>83</td>
<td>16</td>
<td>99</td>
</tr>
</tbody>
</table>
Figure 11: Methods of therapy of Legg-Waldenström - Calve-Perthes disease and DEM. Physiotherapy. Kinesiotherapy. Physical therapy.

Discussion. In our material, without any doubt, we can confirm that etiology of the Perthes disease is trauma. Clinical findings indicate that jumping from even not big heights to a lower hard level repeated many times for long periods of time is especially dangerous. We noticed jumping from trees, stairs, pieces of furniture. Such frequent jumping, which becomes a habit, constitutes the cause of aseptic necrosis of femoral head in many children. The second conclusion - also confirmed in all cases - the endangered group of children are cases with symptoms of Minimal Brain Dysfunction (MBD). In our countries, in the last 20 years of our activity we have encouraged prophylaxis. It is easy - children should avoid “jumping on hard surface” because frequent jumping - many times every day for weeks, or months is the cause of the Perthes disease. Such presented observations about etiology have been made not only in Lublin but also in Russia and in Germany. This is also noted - the seasonal occurrence of the Perthes disease - more frequent in the spring and summer months.

Here we would like to draw your attention - the Perthes disease requires differential diagnosis with Dysplasia Epiphysealis Multiplex (DEM), other name Morbus Meyer (Figure 12, 13). The radiological symptoms are similar, but concern both hips - both femoral heads. The Perthes disease is in all our cases only one sided. In clinical examination - in DEM - the limitation of hips movement is not so strict big - but only mild - only little limited. In DEM cases the pain of the hip is not severe. Very often the diagnosis is made too late and very frequently - “it is mistaken” - diagnosed firstly as Perthes disease. Here we would underline that the illnesses need the differential diagnosis, but therapy often is similar or the same. The authors underline significance of early detection of MBD in children coming for consultation or therapy with problems of feet, knees, spine. The parents of such group of patients should be informed about prophylaxis “of hip illness” by modification of children’s physical activities home and outdoor - no to jump on hard floor.

Figure 12: Differential diagnosis. Perthes disease. Dysplasia Epiphysealis.

Figure 13: Clinical example of Dysplasia Epiphysealis Multiplex (DEM). Boy born 22nd June 2004. First symptoms of hips illness at the age of 4. Primary therapy out of Lublin. Diagnosis - Perthes disease - not correct (!). Consultation in Lublin in 2008: Diagnosis DEM. X ray pictures from 2008 - 2017. Figures (A) & (B) signs of necrosis of both femoral heads, (C) & (D) lower both heads, lateralization, proper shape.

The next group needed the deferential diagnosis are children with necrosis of femoral head-treated for a long time in Pediatric Departments with cortisone or hydrocortisone because of internal diseases. These children were diagnosed with hematology processes or allergies. We had treated small group (6 children in period of 10 years / 1995 - 2009) with heavy necrosis symptoms of femoral head after such therapy in Pediatric Departments.

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Conclusions

1. Etiology of Perthes disease is strictly traumatic. It concerns children with the symptoms of Minimal Brain Dysfunction (MBD).
2. This group of children likes to jump-and it is the cause of femoral head necrosis called the Perthes disease.
3. After long lasting -2-3 months-jumping on hard floor / surface - fractures of spongy bones in femoral head appear, disturbing the blood circulation. At this moment, the necrosis of the femoral head starts to develop.
4. In first few weeks of clinical symptom of Perthes disease it is possible to stop the development of illness though proper physiotherapy and completely excluding of loading of the affected leg for 1-2 months.
5. In physiotherapy in every stage of illness, the following elements are important:
   a. Relieve of loading of affected leg,
   b. Exercise in warm water, the best in geothermal water,
   c. Exercise of abduction in prone position of the child,
   d. Resting and sleeping in flexion and abduction of affected hip / leg,
   e. Sitting in “butterfly position” (term taken from karate language) - feet together, flexed knees maximal lateral, spine relaxed.
   f. Laser therapy and other physical therapy.

References