Conservative treatment in children with Legg-Calve-Perthes disease, aged up to 7 years

Wyniki leczenia zachowawczego dzieci z chorobą Legg-Calve-Perthes do 7 roku życia

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Abstract


Materials and methods. The inclusion criterion was the age of affected patients: under 7 years at the time of diagnosis. The study population included 50 children (10 girls and 40 boys) in the mean age of 5 years and 1 month. All the children participated in follow up appointments, arranged every 3 months, during which clinical examinations (the range of hip joint motion, limb length) were carried out and radiological parameters (Salter extrusion index, ATD, Wiberg angle) were assessed. All the patients were assessed in the IPSG Modified HOOS-16 Hip Survey.

Results. The patients, qualified to group A by the Herring classification, demonstrated less limits in hip joint inward rotation and no length reduction of the affected limb (p<0.05). In the patients with Wiberg angle and Salter extrusion index in the affected joint being similar to their corresponding values on the healthy side, the intensity of pain was lower, assessed in the IPSG Modified HOOS-16 Hip Survey, as well as their functioning in everyday life and in sport and recreation activities was better, thus their general quality of life was higher (p<0.05).

Conclusions. Children with Perthes disease, diagnosed before the age of 7, do not usually require surgical intervention. In our study, the radiological evaluation of hip joint development showed complete healing of the proximal femoral epiphysis, while preserving femoral head sphericity and good hip functionality. In general, the quality of life of affected patients does not differ from the quality of life of healthy children. The indication for conservative treatment of children with Perthes disease is their age below 7 years. Then a regular monitoring of the course of physiotherapeutic therapy is required, combined with regular radiological controls and clinical assessments of hip joint motion range.

Key words: avascular femoral head necrosis, Perthes disease, conservative treatment, quality of life, radiological evaluation, clinical evaluation

Streszczenie

Wstęp. Choroba Legg-Calve-Perthes’a jest jedną z najczęściej występujących martwic kości u dzieci i młodzieży.

Cele. Ocena rozwoju stawu biodrowego oraz jakości życia dzieci leczonych zachowawczo z powodu choroby Perthes’a oraz ustalenie wskazań do leczenia zachowawczego.

Materiał i metody. Analizę poddano dane 50 dzieci w wieku o średnio 5 lat i 1 miesiąc. Wśród badanych dzieci było 10 dziewczynek i 40 chłopców. Wszyscy pacjenci byli poddawani badaniu kontrolnemu co 3 miesiące podczas którego oceniano parametry kliniczne (zakres ruchomości w stawach biodrowych, długość kończyn) oraz radiologiczne (koeficient ekstruzji Saltera, ATD, kąt Wiberga). Wszyscy pacjenci poddani zostali ocenie w kwestionariuszu IPSG Modified HOOS-16 Hip Survey.

 Wyniki. Pacjenci zakwalifikowani do grupy A klasyfikacji Herringa (30 pacjentów) mieli mniejsze ograniczenie z zakresu ruchu rotacji wewnętrznej w stawie biodrowym oraz brak skrócenia kończyny objętej procesem chorobowym (p<0.05). W ocenie statystycznej pomiędzy wybranymi parametrami klinicznymi a radiologicznymi wykazano, że u pacjentów, u których wartości kąta Wiberga i współczynnika ekstruzji Saltera w stawie objętym chorobą były zbliżone do tych po stronie zdrowej, natężenie dolegliwości bólowych, ocenione w kwestionariuszu IPSG Modified HOOS-16 Hip Survey było mniejsze a także funkcja w życiu codziennym, funkcja podczas zajęć sportowych i rekreacyjnych była lepsza oraz jakość życia wyższa (p<0.05).

Wnioski. Ocena radiologiczna rozwoju stawu biodrowego u dzieci leczonych zachowawczo wykazała, pełną przebudowę nasady kości udowej, w stawie objętym chorobą przed 7 rokiem życia dobrze podają się leczeniu zachowawczemu i nie wymagają zazwyczaj leczenia operacyjnego. Jakość życia dzieci z chorobą Perthes’a nie odbiega od jakości życia dzieci zdrowych, mimo rezimu usprawniającego. Wskazaniem do leczenia zachowawczego dzieci z chorobą Perthes’a, jest wiek poniżej 7 roku życia przy regularnym kontrolowaniu przebiegu leczenia zachowawczego, regularnie i klinicznej ocenie zakresu ruchów w stawie biodrowym.

Słowa kluczowe: jadowata martwica głowy kości udowej, Legg-Calve-Perthes, leczenie zachowawcze, jakość życia, ocena radiologiczna, ocena kliniczna
Introduction

Avascular femoral head necrosis is one of the most common necroses in children and adolescents. It affects one of the largest joints and mat lead to serious anatomical and functional disorders, while the hip joint destruction, formed in the childhood period, may then impact the whole life of affected patients. Despite the passage of almost 100 years since the first description of Legg-Calve-Perthes disease, we still are either not sure, of its exact cause(s) or able to design and implement a fully effective therapy [1-4]. In the aetiology of Perthes disease, multiple, various factors are considered, some being assumed to be important, while treatment protocols are not yet precisely defined. Some doctors prefer conservative treatment, while others choose surgical intervention. It is, however, a well-known fact that the final outcome of treatment is influenced by a number of clinical and radiological factors, out of which, only a small part remain within our precise recognition and knowledge potential, as well as their role in the treatment process is not completely unveiled [3-6].

Despite the direct availability of a huge imaging potential, we are still far from precisely defining the aetiology of Perthes disease [3, 4, 7, 8].

Next to the femoral head necrosis area it is the age, when the first symptoms of the disease occur, which is recognised and emphasised by all researchers as one of the main prognostic factors in Perthes disease. Most authors take the view that the earlier the symptoms of femoral head necrosis appear, the more likely it is to end the disease process without permanent and negative consequences for the hip joint. It seems, however, that one of the most important prognostic factors in Perthes disease is the size and extent of femoral head necrosis. Almost all the authors, who have ever dealt with Perthes disease, agree that a larger area of necrosis and, therefore, a more advanced deformity of the proximal epiphysis of the femur, lead to worse end results.

Perthes disease occurs in children, mainly boys, at the age from 2 to 14 years, usually between the 5th and the 8th year, while the incidence of the disease before the 2nd year and the 14th year of age is rather scarce [7, 8].

Our observations have shown that the early onset of physical exercises and the attempts to keep a full range of movement in affected hip joint, help the joint rebuild much sooner, giving the affected children a chance of faster return to their normal life. We focused our particular attention on a group of the youngest children, diagnosed with the disease before the age of 7 years, and in whom an early improvement and appropriately designed and conducted individual exercises let maintain the sphericity of the femoral head, preventing its deformation.

Research Aim


Material and Methods

The material for this reported study came from the research, carried out in children at the Outpatient Clinic of the Department of Orthopaedics and Paediatric Orthopaedics of the Medical University of Lodz during the years 1996-2014. The study material constituted a group of 190 children, treated for avascular femoral head necrosis. The age under 7 years, at which the disease was diagnosed, was the inclusion criterion for the study. Out of the analysed group of children, 96 patients were finally selected and subjected to further examinations. Ultimately, the data of fifty (50) children with full medical records were used for the final assessment, including radiographic hip joint interpretations. The age of the examined children ranged from 2 years and 11 months to 6 years and 11 months, with the mean age of 5 years and 1 month. There were 10 girls and 40 boys among the examined children.

All the children were followed up every three months until the femoral head was rebuilt. During each follow-up appointment an examined child was subjected to a clinical orthopaedic evaluation with the particular attention towards hip joint movement range, which was each time compared to the corresponding values, recorded at the previous visit. In a unilateral disease, the loss of hip motion range was measured in relation to the corresponding values on the other (healthy) side. In a child with bilateral disease, it was assumed that the minimum range values should be following: abduction – 45°, adduction – 20°, inward rotation – 30°, outward rotation – 45°, flexion – 120° and extension 30°.

Apart from motion range assessments, the length of lower limbs and the quality of gait were evaluated. Each child was submitted to multi-dimensional sonography of both hip joints and radiography in two projections: AP and Lauenstein, to assess the extent of epiphyseal necrosis, the sphericity of the femoral head, changes in the proximal metaphysis of the femoral bone and the joint congruence. During the last follow-up examination I assessed radiographs for the epiphyseal extrusion index after Salter osteotomy, acc. to the formula: AC/ABx100%, where extrusion > 20% was considered a poor and > 10% a satisfactory outcome, as well as the ratio of the articulo-trochanteric distance (ATTD) to that in the healthy joint (46). I also analysed the Wiberg angle (CE center edge angle), where the angle above 25° classified the hip joint as a very good outcome, the values between 20 and 25° as satisfactory results and the values below 20° as a poor (unsatisfactory) result.
Our evaluation of the clinical results was based on the classification, adopted in 2011 by the International Perthes Study Group (IPSG), called the IPSG Modified HOOS-16 Hip Survey (Hip Disability and Osteoarthritis Outcome Score). This classification evaluates such parameters as clinical symptoms, joint stiffness, pain, daily life functions, sports activities and the quality of life.

The obtained results served as input for statistical analysis, carried out by the Dell Statistica 13 Pl software.

The study was approved by the Bioethics Committee of the Lodz Medical University, Consent No. RNN/22/14/KE, issued on 11th February 2014.

Results

The statistical analysis of clinical and radiographic results showed that the patients, classified to group A by the Herring classification (30 patients), demonstrated less restriction in the internal rotation range of their hip joints plus no shortening of the limb, affected by the Perthes disease process (p<0.05). The statistical evaluation of selected clinical and radiological parameters showed that the patients with the Wiberg angle and the extrusion index in the affected joint, being similar to those on the healthy side, reported a lower intensity of pain, confirmed in the IPSG Modified HOOS-16 Hip Survey, their functioning in everyday life and in sports and recreational activities apparently improved, indirectly communicating a much better general quality of their life (p<0.05).

Regarding the majority of clinical parameters, the study did not show any statistical relationship among the group of patients with Perthes A disease, in the Herring classification, group B and group C. Statistical significance was demonstrated in group A, according to the Herring classification, where the patients presented both smaller differences in limb lengths (p<0.001) and less limited internal rotation ranges (p=0.04).

Patients with the femoral head extrusion index in the hip joint, affected by LCP, revealed better overall results in the IPSG - HOOS Modified Hip Survey. The results of those patients were much better in terms of their overall quality of life, the severity of pain, functioning in everyday life, as well as during sports and recreational activities.

The Wiberg angle and femoral head extrusion index, AC/AB x 100%, are the most important parameters which positively correlate with the clinical results. The ATD index shows certain relationships only with certain functional parameters.

Discussion

Avascular femoral head necrosis is one of the most common necroses in children and adolescents. It affects the largest joints and may lead to serious anatomical and functional disorders, while hip joint destructions, formed in the childhood period, may impact the whole life of affected patients. Despite the passage of almost 100 years since the first description of Legg-Calve-Perthes disease, we still do not completely know its exact cause(s) or are able to design and implement fully effective therapeutic methods. In the aetiology of Perthes disease, multiple, various factors are considered and assumed to be important, while treatment protocols are still not yet precisely defined. Some doctors prefer conservative treatment, while others choose surgical intervention. It is, however, a well-known fact that the final outcome of treatment is influenced by a number of clinical and radiological factors, out of which, only a small part remain within our recognition and knowledge, as well as their role in the treatment process is only partially understood.

The basic rules in the treatment of Perthes' disease include the prevention of femoral head deformities, providing optimal conditions for epiphysial reconstruction, maintenance of congruent hip joint and femoroacetabular conflict reduction.

Many different methods have been used to treat Perthes disease. A currently and commonly used treatment method restores the hip joint congruency, with the aim to cover the femoral head by the acetabulum and restore the sphericity of the proximal femoral bone epiphysis. Nowadays, the methods that restore congruence of the hip joint include, among others, lying in bed with indirect traction devices for the lower limbs in abduction, exercise plasters acc. to Petri, abducting orthoses, an appropriate set of physical exercises, as well as various methods of surgical intervention, which include the release of soft tissues and bone surgery (varus osteotomy, femoral bone intertrochanteric osteotomy, pelvic osteotomy by the Salter method, acetabular roof plasty and others) [4, 9-14].

In the treatment of Perthes disease, it is extremely important to implement a proper rehabilitation treatment, consisting in the selection of appropriate exercises, especially in the initial period of the disease. These exercises are aimed to maintain joint mobility and prevent contractions in the joint. The main emphasis in physiotherapy should be the improvement of abduction and external rotation. Maintaining these movements in particular within a good range in the course of the disease ensures that the femoral head remains spherical and better nourished and that the gait performance is becoming better and better, regarding its functional aspects [15, 16].
The starting point for the reported study was the publication of Sharma et al. from 2009 [15], confirmed two years later by Herring [17].

Both Sharma and later Herring in his publication assessed and compared the results of surgical and non-surgical treatment of children with Perthes disease on the basis of multi-centre studies. Non-surgical treatment included, untreated, orthoses, physical exercises and a combination of those measures. The applied surgical interventions included varus osteotomy of the femoral bone, pelvic osteotomy by the Salter method or a combination of these two methods. Analysing many-year observations from multiple centres in the world, the researchers did not find any significant differences between surgical and conservative treatment in the final examination of children, diagnosed with Perthes disease under the 8th year of age.

The conservative treatment of Perthes’ disease is often an issue for the entire family. The beginnings bring, of course, some anxiety, associated with the first pain sensations in hip or knee joint area, often accompanied by limping. Such typical symptoms of Perthes disease can very often be underestimated. This is especially true for boys, who are very active, in general, and such symptoms may often be not unusual for their parents. It is only when the pain lasts longer or recurs that parents start seeking for help from specialists. The first suspicion and diagnosis that this is avascular femoral head necrosis is a problem for the whole family and, therefore, parents should accompany their child in the illness and significantly support doctors’ recommendations. The parents should support their child in difficult moments, help them understand the difficult situation. Children very often stay at home under the care of one of their parents for the duration of their illness. This is inextricably associated with their general development and may adversely affect their cognitive skills, education and social situation among peer groups (especially school-age children) [18-20].

Our interest targeted the youngest group of children who were diagnosed with the disease before the age of 7 years. Similarly as in literature reports, boys prevailed over girls (40 boys vs. 10 girls) in our material. In the group of analysed children, most of them had not been previously treated for hip disease (32 children). Those children were either largely treatment-naive for any condition or their treatment was directed towards knee joint conditions (inflammation, sprain, other injury). Sixteen children had previously been treated with gypsum dressings, on the average, for several months, and Thomas splint had been applied in two patients, on the average, for 4 weeks.

The same physiotherapy protocol was applied in all the children. Physiotherapeutic exercises of the hip joint, according to instructions (see the description at the end of the paper), were repeated 3-5 times a day, depending on the degree of hip joint motion limitations, supported by physical pro-
cures once per 3 months. Follow-up visits of the children took place at the Clinic every 3 months, during which the range of movements and radiographs of affected joints, obtained in 2 projections, were thoroughly evaluated.

Assessing the clinical outcome of the treatment, most children achieved a full range of movement in the affected joint, while inward rotation remained slightly limited in a few cases. A comparison of radiographs with the clinical picture showed that the observed limited internal rotation had mostly been identified in those children who had been diagnosed with coxa magna on radiographs. In addition, 9 children presented a slight shortening of the limb, which did not require the use of an orthopaedic inner sole and, only in 1 case, the limb length reduction was 1 cm.

In the clinical assessment, we used the HOOS-16-Hip Survey classification, modified by IPSG. In our opinion, this classification allows for a very thorough analysis of treatment outcome, as confirmed by our results.

Analysing the radiological parameters, we found out that maintained good and constant movement of the sick hip joint resulted in its better nutrition and faster reconstruction.

Having analysed the publication of Nakamura et al., proving that the prognosis in children under 7 years of age was fairly promising, we fully agree with their thesis. The authors evaluated 114 children, with very good outcome observed in 72 out of 114 (63%). Patients with of Perthes disease, diagnosed at onset as A and B in the Herring classification and demonstrating a good range of motion in each case, achieved good results at treatment completion. However, even under 7 years of age, when the area of necrosis is very extensive, the results may be unfavourable [20].

Our study was also confirmed in the report by Kamegaya, who concluded that a non-operative treatment in children under 7 years of age should bring about very good results [21].

The most important issue in treatment implementation, both conservative and surgical, is its long-term outcome.

Aalrson et al., while analysing patients after non-operative treatment, showed in their study that patients could be functioning very well up to 5-6 decades of their age. Later on, the incidence of degenerative changes may be higher and higher. In a 20-year observation, more than 50% of our patients demonstrated clinical features of FAI, and 40% reported pain which, in 6%, required medical intervention [16, 22].

Rich et al. obtained similar results. The authors showed that a treatment with exercises (physiotherapy) was broadly accepted, bringing good, long-term results. They obtained spherical femoral head in 78% of the cases (B and C in the Herring classification) and 93% of the treated children demonstrated congruent joints [10].

Herring also draws our attention to the results of conservative treatment, obtained in the younger age group of children. In his study, based on a multi-centre trial, 59% of children under 8 years of age had a final score in the Stulberg II classification, while only 39% of children at the age above 8 years could present such a result. The author also proved that the Herring classification closely correlated with the final result. The score in Stulberg I or II classification was 100% in children with group A, 62% in those with group B and 28% with group C [17].

Although many authors emphasise the importance of modern imaging techniques in the diagnostics and monitoring of treatment in Perthes disease, standard X-ray still seems to be the examination of choice in the initial stage of the diagnostic process.

In recent years, however, modern imaging techniques have been increasingly used, with perfusion MRI scanning being at the forefront of diagnostic innovations. This technique, developed by the IPSG group in Dallas, enables a quick and accurate assessment of blood circulation disorders in the femoral head epiphysis [23]. The previously used imaging methods, such as scintigraphy or arthrography, seem to have fewer and fewer supporters in the world of today, although arthrography is still a valuable diagnostic method if dynamic mobility of the hip joint is to be assessed, especially in cases of hinge dislocation [16].

In our study, we evaluated radiograms, finding out that the major parameters, which positively correlated with clinical results, included the Wiberg angle and the femoral bone head extrusion index (AC/AB x 100%). The ATD index shows certain relationships only with certain functional parameters only.

Conclusions

1. The radiographic evaluation of hip joint development in the conservatively treated children showed a full reconstruction of the femoral epiphysis with preserved femoral head sphericity and good hip joint functionality. I demonstrated a statistical relationship between individual radiological parameters and the assessment of life quality in the patients.

2. Children with Perthes disease, diagnosed before the 7th year of age, are well susceptible to conservative treatment and, usually, they do not require surgical intervention.

3. The quality of life of children with Perthes disease does not, in fact, differ from that of healthy children, despite the necessary physiotherapy regimes. Most of the children, assessed in our study by the IPSG Modified HOOS-16 Hip Survey, characterised their quality of life as very high.

4. The indication for conservative treatment of children with Perthes disease is the age below the 7th year, with regular monitoring of physiotherapy, regular radiographic control and clinical evaluations of hip joint motion range.
References