Sixteen children with late onset Perthes’ disease were surgically treated at an average age of 12 years 1 month (9 years 4 months to 15 years) by soft tissue release and articulated hip distraction. Two patients had additional diagnosis of Down syndrome and one hip was graded Catterall III Herring B. On preoperative arthrograms, 14 patients had a saddle-shape subluxating femoral head with hinge abduction. The distraction discontinued when Shenton line was corrected. The fixation lasted 4–5 months. At a mean follow-up of 2 years 7 months (1–5 years), an improvement of hip range of motion was found in all patients. Hip joint arthrograms upon removal of the fixator showed disappearance of the saddle-shape femoral head in 10 of the 14 patients with this deformation. Shenton line breakage improved from 7.6 mm to 2.1 mm at last follow-up. The epiphyseal index improved by 14%. The average pain score on an analog scale dropped from a preoperative 7.0 to 1.6 at last follow-up (filled in by patients or parents in case of Down syndrome). Separate analysis of data from the group having only Perthes disease (13 children) showed better results. These preliminary data suggest that soft tissue release combined with unloading of the femoral head and restoration of joint space can improve the function and epiphyseal height.

Introduction
Late onset Perthes’ disease is defined as a disorder that is diagnosed after 9 years of age [1]. Lateness of age at presentation carries less favorable prognosis for the joint. The acetabulum is unable to accommodate the deformed femoral head when the remodeling phase takes place due to the short time until skeletal maturity. Deterioration of the joint is rapid and is accompanied by early arthritic changes [2]. The way to treat late onset Perthes’ disease is controversial. Conservative treatment will not change the shape of the femoral head or joint congruency. Thus, different surgical treatment options were proposed but they had inherent drawbacks. One of them, varus realignment of the hip, may even increase the incongruency to the acetabulum and shorten the leg, and another, valgus osteotomy, will succeed in unloading the deformed part but it may increase the subluxation without influencing the basic avascular process. Acetabular lateral shelf, Salter, Chiari and triple osteotomies are procedures that aim to reorient or increase the size of the acetabulum and produce more support to the head [3]. However, these approaches fail to reduce the pressure or change the shape of the femoral head.

For the past 5 years, we have been using a combination of soft tissue release and joint distraction with a hinged monolateral external fixator to treat patients with severe late onset Perthes’ disease. The procedure had a positive effect on the regeneration of bone and cartilage on both sides of the joint [4]. The regeneration process improved the congruency of the femoral head in the acetabulum and reduced the superolateral subluxation. The distraction treatment did not affect any change in the joint anatomy, thus allowing future performance of bony procedures should they be needed. A description of this surgical technique and our early results for a series of 16 patients are presented.

Patients
The study cohort comprised 13 boys and three girls diagnosed as having severe late onset Perthes’ disease. They all suffered from persistent severe pain and limited range of motion (ROM) of the hip. The radiographs demonstrated 50% or more femoral head involvement with hip subluxation in each of them (Figs 1–3). Two children had Down syndrome and another child had geleophysic epiphyseal dysplasia.

Methods
Clinical assessment
The assessed clinical parameters were hip ROM, limp and Trendelenburg sign. A questionnaire with a visual
analog pain scale was filled in by all patients or parents, both preoperatively and at last follow-up. The assessed radiological parameters were the Catterall and Herring classifications and risk signs [2,5].

All patients underwent hip arthrography before the soft tissue release and before application of the external fixator and after its removal (Fig. 2). The arthrograms provided the following measurements: (1) the epiphyseal index (calculated as the ratio of epiphysis height to width); (2) femoral superior subluxation (measured by the break in the Shenton line) [6,7]; (3) Stulberg classification of the hips (performed when the patient was approaching skeletal maturity as defined by plain radiographs) [8].

**Surgical technique**

The patient is placed supine on a transparent operating table; hip arthrogram is performed via a medial approach to assess cartilage architecture and the extent of hinge abduction. An adductor and iliopsoas tenotomy is performed via a medial approach. Using image intensifier, a 1.6 mm K-wire is inserted into the femoral head at the center of rotation of the hip, while the leg is kept in 15° of abduction with the patella pointing forward. An articulated body for the hip (Cat. No. 10022; Orthofix,
Patient 6. (a,b) Perthes’ disease right hip (Catterall IV, Herring C), radiographs at age 12 years, 1 year after disease onset: hinge abduction. (c,d) Hip arthrogram at surgery with superolateral pulling of dye versus the arthrogram on removal of frame, the pulling of dye disappeared. (e) At early distraction phase with joint space opening and over correction of Shenton line. (f,g) Follow-up radiographs at 21 months post-operation: superolaterally reformed femoral head with congruency to the acetabulum, hinge abduction resolved.
Guilford, Surey, UK) is then applied on the K-wire and a standard model kit body (Cat. No. 10000 or 90000; Orthofix) is attached to the hinge distally. The proximal part is fixed to the supra-acetabular area with a T-clamp (Cat. No. 10007; Orthofix) using two or three 5–6 mm Orthofix screws (Cat. No. 10115 or 10144; Orthofix). A standard side clamp is used distally with three 5–6 mm screws in the femur (Cat. No. 10142; Orthofix); the procedure is performed with the proper template (Cat. No. 11101; Orthofix) that is replaced by the above pieces. The joint space is immediately distracted 4–5 mm under image control. Distraction is continued 1 mm per day until the Shenton line is overcorrected. Flexion extension exercises are encouraged with the fixator in place, and the patient is kept non-weight bearing. The fixator is left in place for 4–5 months until lateral pillar reossification appears. The fixator is removed in the operating room and hip arthrogram is repeated. After removal of the frame, the patient continues a program of protective non-weight bearing and intensive physical and hydrotherapy for an additional 6 weeks. At this stage full weight bearing is allowed with continued physiotherapy for another 6 months.

**Statistical analysis**

The Wilcoxon non-parametric paired test was used to evaluate the changes in pain, the arthrographic epiphyseal index and the Shenton line breakage following treatment. We used SPSS; Orthofix version 11.0 (for windows SPSS Inc., Chicago, Illinois, USA) for analysis and the statistical significance value ($P$) was set to 0.05.
Results

Our 16 patients (Table 1) had symptoms for an average of 19 months (range 6–48) before surgery and all required the use of crutches or a wheelchair. Fifteen hips were graded Catterall IV and Herring C and one hip Catterall III and Herring B. There were 12 hips with four and four with three Catterall head-at-risk signs (see Figs 1–4 for the imaging studies of patients 3, 6 and 7).

The average age at surgery was 12 years 1 month (9 years 4 months to 15 years), and the mean follow-up was 2 years 7 months (1–5 years) (Table 2).

Postoperatively, all patients had improved ROM, flexion by a mean of $20^\circ$, abduction by $16^\circ$, and internal rotation by $17^\circ$. At the last follow-up all patients were able to walk without support. The Trendelenburg sign disappeared in eight patients but persisted in the other eight.

On their preoperative arthrograms, 14 patients had saddle-shaped and subluxating femoral heads with hinge abduction and two patients had extrusion only. The most recent arthrograms showed that the saddle shape disappeared in 10 of those 14 hips. The break in the Shenton line that measured, on average, 7.69 mm before the operation dropped to 2.19 mm ($P < 0.05$). The epiphyseal index, as a measurement of femoral head sphericity, increased from an average of 0.7 to 0.80 ($P < 0.05$) (Table 2). The Stulberg classifications on the last follow-up (Figs 1–3) were type III for three hips and type IV for the other 13 hips (Table 1).

The average pain score (analog scale) dropped from 7.0 before surgery to 1.6 at the last follow-up ($P < 0.05$) (Table 2). Thirteen patients had pin site infection (all treated successfully with oral or intravenous antibiotics) and one patient’s Orthofix clamp broke and was replaced in the ward.

The analysis of data of patients presenting with Perthes disease only, without any known genetic skeletal disorders showed much better results compared with the series taken as a whole, both on the postoperative pain score (1.38 versus 1.63) and on the postoperative radiologic assessment (epiphyseal index 0.81 versus 0.8 and Shenton line breakage 1.31 versus 2.19) (Table 2).

Discussion

The long-term prognosis of late-onset Perthes’ disease is unfavorable for several reasons: the patients being older than 9 years at the start of the disease or at the healing phase, the involvement of 50% or more of the femoral head or collapse of the lateral pillar, lateral subluxation of the femoral head with hinge abduction and the combination of an aspherical head and incongruent joint at maturity [9–11]. Of these factors, a meta-analysis of long-term prognosis in Perthes’ disease clearly showed that age at disease onset and the shape of the femoral head at maturity are the foremost reliable factors to predict outcome [12,13]. The Herring class in combination with arthrographic sphericity was shown to be the best predictive factor for the radiological outcome at maturity [7,14]. Herring type B hips in children older than 7 years indicated poor results in terms of the Stulberg class at maturity.

Table 1 Clinical and radiographic parameters of patients

<table>
<thead>
<tr>
<th>Patient</th>
<th>Sex</th>
<th>Other conditions</th>
<th>Age at surgery (years, months)</th>
<th>Follow-up (years, months)</th>
<th>Pain</th>
<th>Trendelenburg sign resolution</th>
<th>Classification</th>
<th>Head-at-risk signs</th>
<th>Arthrographic epiphyseal index</th>
<th>Shenton line breakage</th>
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<tr>
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Pre, preoperative; Post, postoperative. Fracture.
skeletal maturity, while Herring type C hips resulted in uniformly poor results. The worst long-term results were seen in Stulberg V hips at maturity, with a flat femoral head and an incongruent acetabulum [10,15]. These patients tended to have a debilitating hip disease when they reached their early forties. The radiographic results of a congruent joint even in the presence of deformed femoral heads (congruent incongruency) may conform to a good outcome in the long term [15,16].

Another significant prognostic factor after the age of 12 years is the potential for remaining growth according to the Oxford system: remaining growth of less than 25% in the presence of a closed acetabular growth plate yielded poor radiographic outcome at skeletal maturity, while remaining growth of more than 35% produced better hips even in older children [16]. The presence of some form of a generalized chondrogenesis defect with other skeletal abnormalities in patients with Perthes disease may also have an effect on healing potential [17].

Various authors studied the anatomical alterations that occur in Perthes’ disease. Plane radiograph parameters and three-dimensional computed tomography scans of the diseased hip could show that the femoral head overgrows the acetabular boundaries, leading to lateral

![Fig. 4](image)

Patient 6. Perthes’ disease left hip, magnetic resonance imaging scan at age 11.5 years, four coronal views: coxa magna with flattening, fragmentation, subluxation and fluid collection in the joint. High signal intensity in the head and neck reflecting local edema. (a,b,c) T2-weighted images. (d) Short tau version recovery (STIR) image.

| Table 2 Comparative analysis of cumulative results with and without the genetically affected patients |
|---------------------------------|-----------------|-----------------|-----------------|-----------------|
| Age at surgery                  | Follow-up       | Pain Pre        | Pain Post       |
| Mean patients 1–16              | 12 years 1 month| 2 years 7 months| 7               |
| Exact significance (2-tailed)   |                 | 1.63            | 0.7             |
| Mean patients 1–16              | 12 years 3 months| 2 years 9 months| 7               |
| Exact significance (2-tailed)   |                 | 1.38            | 0.72            |
| Mean patients 1–16              | 12 years 3 months| 2 years 9 months| 7               |
| Exact significance (2-tailed)   |                 | 1.63            | 0.7             |

Pre, preoperative; Post, postoperative.
subluxation and loss of containment [18,19]. Treatment of these hips by conservative means is disappointing and the surgical options are limited. Results of varus osteotomies were reported to be good below the age of 9 years, but a rapid decline of clinical and radiographic parameters was found to occur after age 10 years [1,20]. Lateral shelf augmentation, Salter, Chiari and triple osteotomies were reported to increase the superolateral head cover and the capacity of the acetabulum to contain the head: these procedures do not affect the joint cartilage and no long-term results are available to date [3,21–23]. Valgus extension osteotomies have an effect on the remaining remodeling and can be considered salvage procedures: in one large reported series, it was not advocated in older children or after closure of the triradiate cartilage [24,25].

Reports on the value of hip distraction started to appear in the early 1990s, when the technique was applied to stiff hips with Perthes’ disease and other conditions. This method achieved improvement of hip ROM and joint space [4]. The Ilizarov technique for distraction was used to treat a group of children with Perthes’ disease whose average age was 7.5 years. The applied fixator allowed hip movement in two of the 11 patients, and soft tissue release was done only in the presence of flexion contracture. The time in the fixator did not exceed 4 months and the results were mixed [26]. On the other hand, the results of an articulated hinge distraction with the Ilizarov fixator for patients having Perthes’ disease (average age 7.9 years) were reported to be good in terms of hip sphericity and joint congruency [27].

The 16 patients in our series had a poor prognosis due to their age and the severity of the disease at presentation: severe femoral head deformities with secondary acetabulum changes, incongruent and subluxating hip joints mostly moving in a pattern of hinge abduction. In addition, two of our patients had Down syndrome and one had geleophysic epiphyseal dysplasia.

While the extent of femoral subluxation is underestimated by plain radiographs, measurement of femoral subluxation by hip arthrography on adduction was shown to be very reliable [6]. Our patients demonstrated a significant improvement of these parameters when their preoperative and postoperative arthograms were compared. We measured the arthrographic sphericity by calculating the ‘epiphyseal index’ on the preoperative arthograms and those carried out on the day the fixator was removed. Arthrographic sphericity combined with the Herring grade had been shown to be a good predictor of final outcome [7], and the postoperative improvement of femoral head sphericity may be one of the pivotal factors responsible for the good clinical results in our group.

All our study patients had needed either crutches or wheelchairs and had been suffering from pain for many months before the operation. Considering the clinical situation and the radiographic appearance on presentation, each was considered as having a severe form of the disease and qualified for a salvage procedure. The alternative of using an approach of soft tissue release and articulated hip distraction was based on a number of considerations. The combination of iliopsoas and adductor tenotomy with continuous distraction of the hip for 4–5 months allows osteochondral regeneration. The unloading of the cartilage inside the joints induced growth of this tissue on both sides of the joint. Experiments in vivo on animal models have demonstrated that the cartilage tissue, which is maintained under hydrostatic pressures, transforms itself into bone when the compressive force is switched to distraction. The unloaded cartilage proliferates on the side of the joint space while endochondral ossification allows the epiphysis to reshape on the subchondral side [28,29]. The newly transformed subchondral bone may now build itself under a condition of almost total unloading. Maintaining hip joint motion in the apparatus further assists in the remodeling process. We believe that our good short-term clinical and radiological results confirm the validity of applying these lines of evidence in the clinical setting. Furthermore, this treatment is not limited by femoral head deformity and burns no future bridges for the patients. Finally, the significant drop in the average level of pain can also be included among the indications of surgical success (Table 2).

We should raise a word of caution regarding our two patients with Down syndrome and the one with geleophysic epiphyseal dysplasia. This small subgroup had results significantly inferior to those having only Perthes’ disease (13 children). For the latter, the statistical analysis of results showed that these children had better results when compared with the whole group regarding pain relief and postoperative improvement of Shenton line (Table 2). The worse results for the subgroup may be related to the genetic skeletal condition they have and this should be taken into account when considering hip distraction for them.

We continue to monitor our patients to determine their long-term outcome.

Acknowledgements

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References


