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Multidirectional Habitual Bilateral Hip Dislocation in a Patient with Down Syndrome

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A multidirectional (anterior and posterior) bilateral habitual nontraumatic dislocation of the hip in a 3-year-old girl with Down syndrome is reported. The treatment was conservative with application of a spica cast for 12 weeks to shrink the capsule. During the first 6 weeks of treatment, the hips were immobilized in abduction and full external rotation. During the last 6 weeks, the hips were in abduction and slight internal rotation. After cast removal, the hips seemed to have been stabilized, and the child has remained asymptomatic for the last 12 months.

Habitual nontraumatic dislocation of the hip in children is rare. It usually occurs in patients with syndromes that affect the connective tissue (eg, Down syndrome, Ehlers-Danlos syndrome, Marfan's syndrome, homocystinuria, and osteogenesis imperfecta),[12] in which generalized ligamentous laxity seems to be the single common denominator.[4] This is the first patient with Down syndrome, to our knowledge, with multidirectional (anterior and posterior), bilateral, habitual, nontraumatic dislocation of the hip. Patients with Down syndrome tend to have habitual hip dislocation develop.[2,4,7,12,14] We suspect the global or multidirectional instability of both hips, such as in our patient, is a result of generalized ligamentous laxity that exists in patients with Down syndrome. This means that other patients with Down syndrome may have global instability of their hips. Therefore, a thorough physical examination, as far as multidirectional hip instability is concerned, should be done for every patient with Down syndrome, especially patients who manifest joint laxity or who already have posterior instability of the hip.

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Each author certifies that his or her institution has approved the reporting of this case report, that all investigations were conducted in conformity with ethical principles of research, and that informed consent was obtained.
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CASE REPORT

The patient was the 3-year-old daughter of a 34-year-old Caucasian woman and a 37-year-old Caucasian man, with no significant medical history. The pregnancy was full term and uncomplicated, and the child was delivered vaginally. The patient's clinical features were consistent with the diagnosis of Down syndrome (trisomy 21). One older sibling did not have Down syndrome.

According to the mother, the dislocation of the hips first occurred when the child was approximately 4 months of age. The parents sought medical advice when they noticed an audible click when the child was angry and irritated and movement in both hips was restricted.

On first medical examination, the patient showed generalized ligamentous laxity, which according to the mother existed since birth. She was unable to walk and had never walked before. Her mental abilities and skills were extremely limited. When she was irritated, both hips were dislocated anteriorly by extension and lateral rotation of the lower limbs. The posterior dislocations occurred simultaneously every time she tried to flex and internally rotate her hips. Reduction of the anterior and posterior dislocations occurred automatically after termination of muscle contraction of the lower limbs. Because the child was used to extending and externally rotating her hips, anterior dislocation occurred more often. The latter was diagnosed not only by the audible click, but also by the palpable femoral heads that protruded in the groin area. Anterior instability of the hips was confirmed by a modified Ortolani maneuver (abduction and external rotation of the hips) and with simple full external rotation in neutral

position of the hips, which resulted in the anterior dislocation. Posterior dislocation of the hips was confirmed by the Barlow maneuver. According to information provided by the child's parents, at first the hips only dislocated anteriorly. After a period of rapid deterioration and multiple daily dislocations, the hips began dislocating posteriorly as well. Multidirectional bilateral hip dislocation was confirmed by standard anteroposterior (AP) radiographs of the hips (Figs 1, 2). Because of the child's lack of cooperation, and because the parents refused to allow administration of anesthesia, the images obtained by computed tomography (CT) were not high quality (Fig 3).

The treatment options are reported to be conservative or surgical.[4,8,12] The first usually includes application of a hip spica cast for approximately 3 weeks and then the use of a special hip abduction splint.[8] The latter can be either a soft tissue operation or bone procedure of the innominate or the femur or both.[8]

We initiated conservative treatment because the patient's bilateral, multidirectional (anterior and posterior) dislocation meant that a soft tissue operation implied bilateral plication of the anterior and posterior elements of the hips' capsules. These procedures likely would endanger the vascular supply of the femoral heads. Also, because we observed no acetabular dysplasia or insufficiency on the radiographs, no bony surgical procedure seemed necessary.

We began treatment by application of a spica cast with the hips in extension, abduction, and full external rotation. The aim of the initial stabilization of the hips by a hip spica cast in external rotation was to shrink the posterior elements of the capsule without jeopardizing the vascular supply of the femoral heads. When this had been achieved, we intended to do anterior plication.

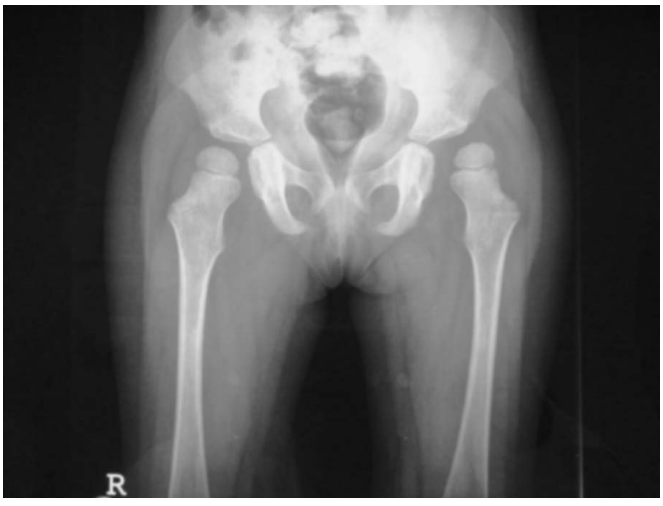

Fig 1. A standard AP radiograph of the hips shows both hips are anteriorly dislocated, although no acetabular dysplasia is evident.

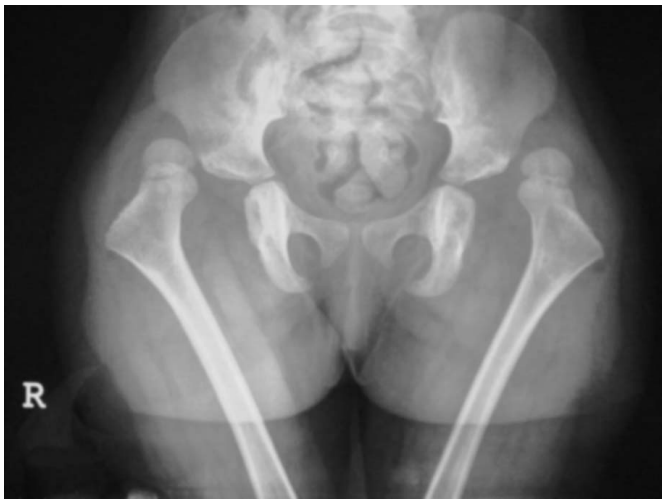

Fig 2. A standard AP radiograph shows both hips are posteriorly dislocated.

The cast remained in place for 6 weeks, and it then was replaced by another spica cast with the hips in abduction and slight internal rotation for another 6 weeks. After cast removal, the hips seemed to stabilize, and we decided the anterior plication would not be necessary. The child has remained free of symptoms or dislocation for the past 12 months (Fig 4). However, the child is still under close medical supervision, and the parents have been informed that an operation might be necessary in the future should a dislocation recur.

DISCUSSION

Habitual hip dislocation in children is rare and usually is associated with trauma,[5,6,10] developmental dysplasia of

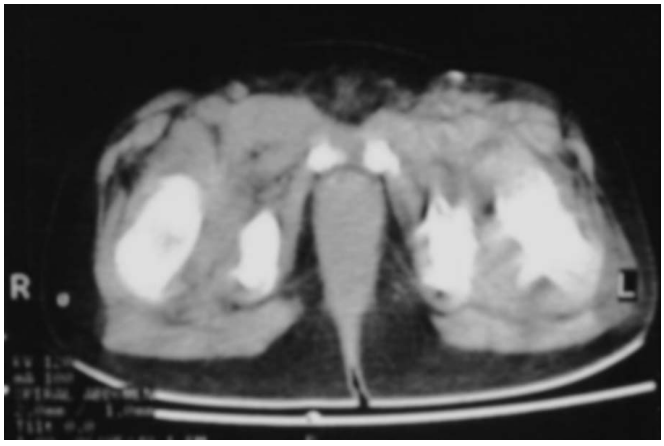

Fig 3. The CT scan confirms anterior dislocation of both hips.

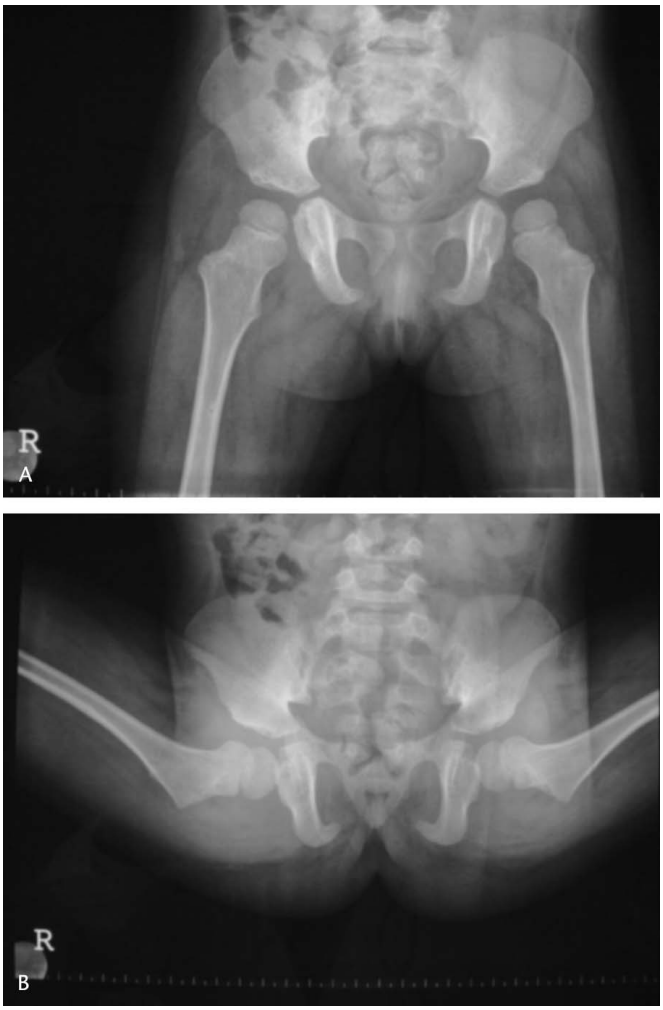

Fig 4A–B. (A) This standard AP radiograph shows the hips after cast removal. The dislocations have been reduced and the hips are stable. (B) A lateral (frog leg) radiograph of the hips is shown after the cast removal. The femoral heads are concentric.

the hip, or connective tissue diseases and syndromes.[4] Patients with habitual hip dislocation related to connective tissue diseases and syndromes usually present with generalized ligamentous laxity, which to some extent may be the actual cause of the dislocation. It has been established that 10–16%[2,11,13] of patients with Down syndrome have significant ligamentous laxity.[4,11] This means generalized ligamentous laxity occurs twice as often in children with Down syndrome as in those without.[3] Cristofaro and Heskiaoff[4] reported that the ability to dislocate the hip in children with Down syndrome is probably secondary to ligamentous laxity. Ligamentous laxity may contribute to the development of multidirectional dislocations that can be easily misdiagnosed; therefore, all the patients with Down syndrome who have posterior dislocation should examined for anterior instability. However, Carter and Wilkinson[3] attribute the commonly occurring orthopaedic problems (eg, dislocation or subluxation of the hip) to the muscle hypotonia that is observed in these children. In our patient, although muscle hypotonia was apparent, the nonsurgical posterior plication of the hips' capsules seems to have treated the dislocation for the time being. This can be accredited to the fact that the actual cause of the dislocation was the generalized ligamentous laxity. However, not all the orthopaedic occurrences in the hips of children with Down syndrome can be attributed to soft tissue disorders. Shaw and Beals[14] found that in patients with Down syndrome, the bone structures have anatomic alterations that tend to destabilize the hip, whereas Woolf and Gross[17] attribute the development of posterior dislocation in two of their patients to a deficiency of the posterior acetabular wall. In our patient, the actual radiographic findings did not predispose hip instability.

Treatment strategies should be tailored according to the underlying causes leading to the dislocation. This means that all the possible causes that contribute to the development of the dislocation must be examined thoroughly before initiation of treatment. Song et al[15] and Greene[9] stated that treatment should first be conservative with immobilization with a cast or a brace. This treatment principle was effective in our patient with multidirectional or global instability of the hip and might be effective in others. Therefore, treatment approaches should not be altered. Generalized ligamentous laxity or muscle hypotonia often is treated with soft tissue operations such as capsular plication,[4] but substantial acetabular dysplasia or disorientation is treated with osteotomy (either of the innominate or the femur).[12,16] Beguiristain et al[1] proposed doing a femoral derotational osteotomy and an anterosuperior capsule plication simultaneously for treatment of recurrent posterior hip dislocation in patients with Down syndrome. However, we think it is reasonable to attempt early immobilization of the hips for a substantial time (more than 6 weeks) with the hips in a posture that keeps the capsule lax either anteriorly or posteriorly according to the axis of the dislocation. We presume this will lead to capsular shrinkage and stabilization of the joint. However, patients should be kept under close medical observation, as recurrence can occur many years later even in patients who were surgically treated.[7]

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