Feature Article

Epidemiology, Demographics, and Natural History of Congenital Hip Disease in Adults

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ABSTRACT

This study examined the epidemiology and demographics of congenital hip disease in 468 (660 hips) patients who were examined between 1970 and 1996. In 356 (54%) hips, the diagnosis was secondary osteoarthritis due to congenital hip disease, and in 272 (41%) hips, the diagnosis was idiopathic osteoarthritis. In the remaining 32 (5%) hips, the diagnosis was uncertain. Of the hips with congenital hip disease, 170 (47.7%) hips were dysplastic, 85 (23.9%) had low dislocation, and 101 (28.4%) high dislocation. The majority of patients with congenital hip disease were women (338 [95.6%] hips). The natural history of the three types of congenital hip disease was studied in 157 patients (202 hips: 102 dysplastic, 42 low dislocation, and 58 high dislocation) who had received no treatment before the initial examination. Average length of follow-up was 17 years. In dysplastic hips, the disease remained undiagnosed until the onset of symptoms at an average age of 34.5 years. In patients with low dislocation, pain had started at an average of 32.5 years due to progressive degenerative arthritis within the false acetabulum. In patients with high dislocation, in the presence of a false acetabulum, pain started at an average age of 31.2 years, while in its absence, pain started at an average age of 46.4 years due to muscle fatigue. These findings suggest dysplasia, low dislocation, and high dislocation in adults are the results of untreated dysplasia, subluxation, and complete dislocation in infancy, respectively.

The term congenital hip disease was first used by Wedge and Wasylenko in 1979 to describe the wide spectrum of anatomic hip abnormalities seen at birth. Other authors subsequently described three radiographic types of the disease present after the neonatal period: 1) dysplasia, in which poor acetabular and femoral head development, with an intact Shenton’s line, is found, 2) subluxation, in which Shenton’s line is broken due to the proximal and lateral migration of the femoral head, without the latter passing the upper edge of the acetabulum, and 3) dislocation, in which the femoral head is completely out of the acetabulum.

In our previous studies of congenital hip disease in adult patients, we noted three distinct types of the disease: 1) dysplasia, in which the femoral head, despite some degree of subluxation, is contained within the original acetabulum, 2) low dislocation, in which the femoral head articulates with a false acetabulum that partially covers the true acetabulum, and 3) high dislocation, in which the femoral head has migrated superiorly and posteriorly and articulates with a hollow in the iliac wing, which may have the appearance of a false acetabulum. Acetabular deficiencies also have been noted in all three types.

This study examined the epidemiology and demographics of congenital hip disease in adults as well as the natural history of its three types: dysplasia, low dislocation, and high dislocation.

MATERIALS AND METHODS

From 1970-1996, a total of 468 adult patients (660 hips) with osteoarthritis were examined and underwent follow-
up at regular intervals by the senior author. In a recent revaluation of the records of these patients, it was confirmed that in 356 (54%) hips, osteoarthritis was secondary to congenital hip disease; in 272 (41%) hips, idiopathic osteoarthritis was diagnosed; and in 32 (5%) hips, the diagnosis was uncertain.

The total number of patients in our series with osteoarthritis secondary to congenital hip disease, 231 patients (356 hips), were used to obtain epidemiologic and demographic data. For the definition of the natural history of the disease, 157 patients (202 hips) who had received no previous treatment prior to their initial examination were separated from the total series. Of these untreated hips 102 were dysplastic, 42 had low dislocation, and 58 had high dislocation. For all of these hips, clinical data and long-term sequential radiographs during adulthood with an average follow-up of 17 years (range: 5-30 years) were available for study.

For the evaluation of the natural history, the following parameters were investigated:
- age at onset of symptoms (pain and limping),
- evolution of radiographic degenerative changes, and
- type of subsequent reconstructive surgery performed.

RESULTS
Epidemiologic and Demographic Data

Of the 356 hips (231 patients) with congenital hip disease, 170 (47.7%) hips were dysplastic, 85 (23.9%) had low dislocation, and 101 (28.4%) had high dislocation. Three hundred thirty-eight (95%) of the hips were in women. Left and right hips were equally involved in dysplasia and low dislocation, and left-to-right hip ratio was approximately 2:1 in high dislocation. Of all 231 patients, 125 (54%) had bilateral congenital hip disease, 17 (7%) had idiopathic osteoarthritis of the contralateral hip, and 89 (39%) had a normal contralateral hip.

Natural History

In the 102 dysplastic hips that had received no previous treatment, no history of hip disease in childhood was recorded. The disease had been undiagnosed until the onset of symptoms at an average age of 34.5 years (range: 18-40 years). Dysfunction, moderate pain during exertion, and a mild limp were the initial symptoms.

Degenerative arthritis developed progressively in three stages following biomechanical patterns (Figure 1).8,11 At stage I (prearthritic), the weight-bearing surface of the acetabulum showed abnormal obliquity with the femoral head being spherical without osteophytes and cysts. Average age of patients at this prearthritic stage was 37.1 years (range: 23-49 years).

At stage II, narrowing of the superolateral articular space, elliptical femoral head due to formation of the capital drop, and progressive subluxation were found. Average age of patients with stage II degenerative arthritis was 43.7 years (range: 34-50 years).

- At stage III, the involved joint showed advanced degenerative arthritis. Large cysts and osteophytes were present both in the acetabulum and the femoral head. Subluxation of the femoral head was more prominent.
Average age of patients at this stage of arthritis was 54.8 years (range: 34-75 years).

In 10 stage I hips, varus intertrochanteric osteotomy was performed, and in 4 stage II hips, valgus intertrochanteric osteotomy was performed (Figures 2 and 3). At final follow-up, all hips with stage III degenerative arthritis, except 16 that had not been operated on at all, had undergone total hip arthroplasty (THA) at an average age of 54.9 years (range: 34-75 years).

Patients with low dislocation (42 hips) who had received no previous treatment reported having a limp since early childhood and pain at an average age of 32.5 years (range: 17-50 years). Degenerative arthritis developed within the false acetabulum, following the same biomechanical patterns as in the dysplastic hips (Figure 4).

Three radiographic types of low dislocation according to the extent of the covering of the true acetabulum were recognized. In type I (13 hips, 31%), two thirds of the true acetabulum were covered by the false one; in type II (17 hips, 40%), one half of the true acetabulum was covered; and in type III (12 hips, 29%), one third of the true acetabulum was covered.

All hips with low dislocation, except two, had undergone THA by final follow-up. Average age of patients at the time of surgery was 51.4 years (range: 34-74 years).

Patients with high dislocation (58 hips) who had received no previous treatment also reported having a limp since early childhood. Limping was more severe in those patients with unilateral involvement. In 30 (52%) hips in which a false acetabulum was present, pain had started at an average age of 31.2 years (range: 18-40 years), while in 28 (48%) hips without a false acetabulum, pain had started at an average age of 46.4 years (range: 30-65 years).

In all patients with high dislocation, a $10^\circ-25^\circ$ valgus deformity of the ipsilateral knee was found; in unilateral cases, a coexisting thoracolumbar scoliosis was present, and in all patients with bilateral high dislocation, increased lordosis of the lumbar spine also was found. Degenerative arthritis had developed in all patients with a false acetabulum (Figure 5).

In the 30 hips with a false acetabulum, THA was performed at an average age of 48.8 years (range: 23-63 years), while in 22 hips without a false acetabulum, THA was performed at an average age of 54.7 years (range: 37-68 years). Six hips without a false acetabulum have not been operated on yet.

**DISCUSSION**

There is little in the literature referring to the natural history of congenital hip disease. Stulberg and Harris studied the relationship between acetabular dysplasia and the development of osteoarthritis of the hip. Wedge and Wasylenko referred particularly to adult hips with no previous treatment to demonstrate the relationship between a patient’s functional level and the development of osteoarthritis at the time of examination.

Cooperman et al conducted follow-up on 32 hips in adults with acetabular dysplasia for an average of 22 years to determine the natural history of this particular type of disease (dysplasia). Weinstein, in two class-
The natural history of completely dislocated hips depends mainly on two factors: the unilateral or bilateral nature of the disease and whether a false acetabulum is present. In unilateral involvement, functional disability is greater. Limp is more severe from infancy, and early in adult life, patients complain of muscle fatigue pain, low back pain, and pain from the ipsilateral knee, which presents a progressively increasing valgus deformity. These patients need early treatment by THA.

The false acetabulum is not a constant finding in high dislocation, but when present, leads to the development of degenerative changes and early THA is necessary (Figure 5). When a false acetabulum is not present, the height of the dislocation, measured by the method of Crowe et al., increases gradually and causes pain mainly from muscle fatigue. We do not know why a false acetabulum is formed in some hips and not in others.

In this study, the natural history of congenital hip disease was assessed in patients who had received no previous treatment prior to their initial examination only as previous treatment alters the natural history of the disease (Figure 6).

CONCLUSION

Knowledge of the natural history of congenital hip disease facilitates the understanding of the potential development and progress of the disease, which differs among the three types. It can lead to a better understanding of the anatomical abnormalities found in the different types and thus facilitate preoperative planning and choice of the most appropriate therapeutic measures for adult patients.

REFERENCES

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Figure 6: AP radiographs of a patient who in infancy had a right hip with dysplasia and a left hip with subluxation. Closed reduction followed by the application of a hip-spica cast for both hips was the initial treatment at age 2 years. An intertrochanteric osteotomy was performed at age 12 years on the left hip. Radiograph before treatment at 2 years (A). Radiograph taken at age 12 years shows the right hip is normal while the left hip is in subluxation (B). Radiograph taken at age 16 years (4 years post-osteotomy) shows the left hip remains unreduced (C). Radiograph taken at age 38 years shows the right hip is normal while the left hip has developed a type I low dislocation (two-thirds coverage of the true acetabulum) with osteoarthritic changes within the false acetabulum (D).