The natural history of acetabular development in developmental dysplasia of the hip

Gelişimsel kalça displazisinde asetabulum gelişiminin doğal seyri

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Normal hip joint development requires a genetically determined balance of growth between the acetabular and triradial cartilages, and a well-centered femoral head. It is unclear how and why this delicate balance becomes disrupted, leading to abnormal development of the hip joint. It is known that, without treatment, a certain number of hips will become problematic later in life. This article outlines normal hip joint development and discusses the natural history of acetabular development in developmental dysplasia of the hip (DDH). The natural history of DDH depends on many factors including age of diagnosis, the growth capacity of the involved structures, and the effects of treatment interventions. Early identification and treatment of hip instability maximizes the chance for normal hip development to occur. However, close observation of the child is crucial to identify individuals where normal development is not occurring. Intervention may become necessary in certain situations to positively affect hip joint development.

Normal hip development

In order to understand the pathoanatomy of developmental dysplasia of the hip joint (DDH) it is beneficial to review the normal development of the hip in the embryo and fetus. The embryonic stage of development occurs in the first two months of life, this phase is followed by the fetal period. Tissue differentiation predominantly occurs during the embryonic phase, while during the fetal period, growth and development are the main events.

For the hip joint to develop normally there must be a balanced and harmonious relationship between the growth of the acetabular and triradial cartilages and the femoral head.

Early in development primitive mesenchymal cells give rise to limb buds that differentiate into an infantile extremity during the embryonic period. The mesoderm layer gives rise to the bony structures, cartilage, muscles and tendons. At the seventh week of intrauterine life a cleft develops in the primitive cartilage, defining the future femoral head and acetabulum. By the end of the embryonic period (8th week of gestation) a cartilaginous model of the hip joint is present and by the 10–11th week all the components of the hip
Joint – a spherical femoral head, a cup shaped acetabulum, ligamentum teres, synovium and capsule are present and fully formed. During the fetal period of development the hip joint enlarges and becomes more mature. With continuous development the femoral head becomes deeply seated in the growing acetabulum.\[2\]

Postnatally the acetabular cartilage complex is interposed between the ilium superiorly, the ischium inferiorly and the pubis anteriorly. The outer two thirds of this structure form the acetabular cartilage. The medial one third is referred to as the non-articular medial wall.

The acetabular cartilage is very cellular hyaline cartilage, and is covered with articular cartilage where it articulates with the femoral head. At the periphery of the acetabular cartilage is a fibrocartilagenous structure or labrum that adds depth to the acetabulum. The joint capsule inserts above the labrum. The capsular insertion is continuous with the labrum below and the periosteum covering the pelvic bones.

The lateral portion of the acetabular cartilage complex is similar to any other epiphyseal cartilages in the skeleton. A growth plate is located on the portion of this structure that faces away from the articular surface of the hip joint and lays down a cartilaginous template that becomes ossified. Growth of the acetabular cartilage complex occurs by interstitial growth within the cartilage and by appositional growth under the perichondrium.\[3\]

The triradiate cartilage is the conjoined physeal plates of the three pelvic bones. It is composed of hyaline cartilage. Interstitial growth occurring in the three limbs of the triradiate cartilage allows the acetabulum to expand during growth.\[4\]

Much data exists, both clinical and experimental, supporting the concept that the main stimulus for concave acetabular growth is the presence of a spherical femoral head within the acetabulum during development.\[5,6\] Other factors such as normal interstitial and appositional growth in the acetabular cartilage, and periosteal new bone formation must occur in the adjacent pelvic bones to allow for the acetabulum to develop normally during growth.

During puberty acetabular depth is further increased by the development of three secondary ossification centers. The os acetabulum is a secondary epiphysis in the pubis and contributes to the anterior wall of the acetabulum. The acetabular epiphysis is the secondary growth centre of the ilium and forms a major portion of the superior edge of the acetabulum. The ischium also contains a small unnamed secondary growth centre that contributes to the acetabular growth.\[6,5\]

Normal development of the proximal femur is crucial in order for normal acetabular growth and development to occur. A delicate balance exists between the growing proximal femur, the acetabular and tri-radiate cartilages and adjacent bones that allows the acetabulum to develop. It is not fully understood what controls this balance, but it likely has some genetic involvement and there is also evidence suggesting that an adverse intrauterine environment may play a role in the pathogenesis of hip dysplasia.\[3,4\]

DDH refers to a spectrum of disorders, ranging from mild dysplastic changes to severe high dislocations associated with abnormal pathoanatomic findings. In the normal hip at birth, there is a tight fit between the femoral head and acetabulum, such that, even after the hip joint capsule is completely sectioned it is difficult to dislocate the femoral head. In the case of a dysplastic hip this tight fit no longer exists, and it is quite easy to push the femoral head out of the acetabulum.\[7\] When this occurs, it is commonly associated with a palpable sensation or clunk as the femoral head glides over an abnormal rim of articular cartilage, the Ortholani sign.\[10\] This ridge of hypertrophied articular cartilage located in the posterior and superior and inferior part of the acetabulum is referred to as the neolimbus.

Most of the primary pathological changes observed in DDH are found on the acetabular side of the joint. Abnormal growth and development of the acetabulum is related to primary pathology of the acetabular growth plates and/or secondary changes due to abnormal pressures from the femoral head. Secondary changes in the acetabulum may also occur as a result of treatment interventions.\[7,11\]

Changes on the femoral side are believed to be due to secondary changes that occur as a result of pressure changes on the proximal femoral physis usually associated with treatment interventions.

**Natural history of acetabular development in DDH**

The natural history of DDH results in one of four possible outcomes.
The hip may become normal, the hip may dislocate, the hip may become subluxated (retain some partial joint contact) or finally the hip may stay located but go on to become dysplastic.

Most unstable hips identified in the newborn period will stabilize. Barlows original paper reported that it is possible to demonstrate hip instability in 1 in 60 newborns. 60% or more of these “unstable” hips will stabilize in the first week without any treatment, and 88% will stabilize within the first 2 months. The remaining 12% are true congenital dislocations and will persist without treatment.[12] Other authors have also reported normalization of previously documented unstable hips. Coleman reported on 23 Navajo children with unstable hips. 5 of the 23 hips spontaneously corrected, 18 remained abnormal. Over the following 3 years, 9 hips in this group of 18 remained dysplastic, 3 were subluxated and 6 were dislocated.[13]

Because it is not possible on an individual basis to say which of the four outcomes outlined above will occur for a specific patient, all identified cases of hip instability detected at birth are treated. Empiric evidence exists suggesting that most abnormalities of the hip joint are reversible if detected early in the newborn period. Devices such as the Pavlik harness or Von Rosen splint are successful in treating this condition in 95% of cases if used early in the course of the disease.[14]

If there is a delay in the diagnosis of DDH in a child, reduction can become more difficult, and the results of treatment more unpredictable. Both extrarticular and intra-articular obstacles to reduction exist that make successful reduction more difficult to achieve. As the child becomes older non-operative methods of obtaining a reduction are less likely to be successful.[15,16] Extra articular obstacles include the adductor longus and iliopsoas muscles, while intra-articular obstacles include the antero-medial joint capsule, the ligamentum teres, the transverse acetabular ligament and rarely, an in folding of the acetabular rim or neolimbus.

The neolimbus occurs as a result of the dislocated/subluxed femoral head abutting against the cartilaginous acetabulum and distorting it. The true acetabular labrum is a thin cartilaginous rim around the periphery of the acetabulum that rarely if ever is a barrier to reduction. It is important to realize that the main restriction to reduction of the femoral head is due to constriction of the hip capsule, not because of the infolded acetabular labrum or the neolimbus. The cartilaginous acetabulum and labrum are crucial structures for normal acetabular development and should not be excised during reduction attempts.

Obtaining and maintaining an early reduction maximizes the chance for normal acetabular development.[17,18] However, the maximum age of reduction, which will result in normal acetabular development is not known. Other factors apart from the age at which reduction occurred, such as the inherent growth capacity of the developing acetabular cartilage and the developing proximal femur, whether either side of the hip joint sustained any growth disturbance as a result of being subluxed or dislocated or was damaged during attempts at closed or open reduction, will all influence how the joint develops.

Dynamic arthrography with fluoroscopy at the time of attempted closed reduction is useful to assess the quality of reduction, amount of femoral head coverage and the stable zone of reduction. Debate exists regarding whether soft tissue interposition between the femoral head and acetabulum adversely affects future acetabular development. If the femoral head is not fully reduced in the acetabulum contrast material will collect medially, this is referred to as “medial dye pooling” A study by Race and Herring[19] reported that pooling of between 5 to 7 millimeters was associated with good outcome in 11 of 13 hips while 5 out of 23 hips with a larger dye pool had an unacceptable outcome. However, there are limitations in using the size of the medial dye pool of contrast material to assess quality of reduction in DDH. It is prone to subjectivity, and it is also affected by image magnification and quality.

It is the senior author’s current practice not to accept any medial dye pooling during arthrography when performing a closed reduction. If a concentric reduction is not achieved, open reduction is performed through a modified medial approach utilizing the interval between the pectineus and the neurovascular bundle.[20] This approach can be safely and predictably used up to age eighteen months.[21] After this results may be more unpredictable.
In the case of late diagnosis of DDH acetabular development may differ significantly from that observed in the normal hip. The normal stimulus of the reduced femoral head is lacking and growth and development of the acetabulum will be abnormal. In this setting secondary centers of ossification contribute to acetabular development. These centers are seen in 2-3% of normal hips and rarely occur before age 11 years. In patients treated for DDH they may be present in upwards of 60% of cases most often appearing between 6 months and 10 years after reduction has been achieved. These accessory centers of ossification most likely represent areas where ossification has occurred in the peripheral acetabular cartilage as a result of pressure from the subluxated or dislocated femoral head, or during attempts at closed or open reduction. These centers should be actively sought on sequential radiographs of cases of DDH, as their presence is indicative of progressive acetabular development. However, their presence does not guarantee that normal acetabular development will occur, and therefore it is essential that all patients with DDH be followed until skeletal maturity.

If a hip remains dislocated additional changes occur in the growth and development of the acetabulum. The roof of the acetabulum becomes more oblique, and acetabular depth fails to develop. The medial wall thickens. This can be observed on radiographs by alteration in the shape of the teardrop (24). To a point these changes are reversible, but the exact age at which hip reduction will result in normal acetabular development is not known.

The natural history of untreated complete dislocations depends on two factors: bilaterality and on whether or not a false acetabulum develops.

Reports exist documenting that bilateral completely dislocated hips in a “high riding” position may remain free of degenerative changes for many years or even for the individual’s life. These patients may develop low back pain over time. In cases where the femoral head retains some contact with the acetabulum (subluxated), or a dislocated hip forms a “false acetabulum”, degenerative changes are more likely to occur and become symptomatic. Also, patients with unilateral hip dislocations will have difficulties with leg length discrepancies and possible ipsilateral knee problems.

Natural history of dysplasia and subluxation

It is imperative that terminology is accurately defined when discussing the natural history of dysplasia and subluxation of the hip. Dysplasia has both an anatomical and radiological definition. Anatomic dysplasia refers to inadequate development of the femoral head and/or acetabulum. The radiographic definition is determined by the presence or absence of an intact Shenton’s line. Anatomic dysplasia exists where there are abnormalities of the femoral head and/or acetabulum but where Shenton’s line is intact. Subluxation occurs when the patient has these abnormalities of the femoral head and/or acetabulum as well as having disruption of Shenton’s line.

The natural history of hip subluxation is usually the occurrence of significant degenerative changes around the third or fourth decade of life.

The natural history of hip dysplasia is not fully known. Patients usually present because of an incidental finding of dysplasia on a radiograph, or because they become symptomatic. Evidence exists supporting the idea that dysplasia will result in degenerative joint disease in adults particularly in females. Increased contact stresses at the joint interface are postulated as being the cause of articular degeneration.

Albinana et al. reported that the Severin classification of hip dysplasia at skeletal maturity could be used to predict long-term radiological and functional outcomes of dysplastic hips treated by closed or open reduction. In this study of 72 hips 47 (65%) were classified as Severin classes I/II and 25 (35%) as Severin class III/IV. With follow up of 40 years in this patient group, the probability of having a total hip replacement was 7% for Severin class I/II hips, 29% for class III hips and 49% for Severin class IV hips. The authors also noted that the age at which reduction was achieved was the most important factor in predicting the Severin class of the affected hip at maturity, and postulated that an earlier age at reduction allowed more acetabular remodeling to occur.

Conclusion

The fate of an unstable hip remains impossible to predict. It is known that every attempt should be
made to reduce the hip as early as possible to allow for restoration of a normal environment for hip joint development to occur. The resumption and adequacy of acetabular development depend on the age at reduction and the growth potential of the acetabular cartilage and the proximal femur. An appreciation of the natural history of acetabular development in DDH is helpful in understanding the pathophysiology of this condition. Close monitoring of patients with hip instability and of patients who have undergone reduction of the hip joint by either a closed or open approach is crucial to assess the development of the joint as the individual ages. Occasionally, some intervention is necessary to positively influence the development of the hip joint and ideally allow normal hip joint development to occur.

References