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The course and treatment of avascular necrosis of the femoral head in developmental dysplasia of the hip

Gelişimsel kalça displazisinde femur başı avasküler nekrozunun seyri ve tedavisi

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Literatürde sıklıkla femur başı avasküler nekrozu (AVN) olarak adlandırılan proksimal femoral büyüme bozukluğu, gelişimsel kalça displazisinin tedavisinde potansiyel olarak yıkıcı bir komplikasyondur. Tedavi seçeneklerinin sınırlı olduğu bir dönemde kalça eklemindeki dejeneratif değişikliklerin erken başlangıcı en büyük korku olarak kalmaktadır. Bu sorunun kesin tanımını yapıp tanısını koymak kadar, hastalığın patofizyolojisi hakkında da anlaşmazlıklar vardır. Gelişimsel kalça displazisi zemininde gelişen AVN'nin doğal seyri oldukça değişkendir. Günümüzdeki uygulamalar öncelikle bu sorunun önlenmesinde odaklanmaktadır; çünkü, değişikliklerin potansiyel olarak geri dönebilir olduğu erken evrede girişimde bulunabilmek için güvenilir bir yol bulunmamaktadır. Manyetik rezonans görüntülemenin kullanıldığı yeni teknikler, geri dönüşümsüz hasar oluşmadan önce bu durumun erken tanısında rol oynayabilir ve AVN'nin doğal seyrini olumlu etkileyecek girişimlerin önünü açabilir. Gelişimsel kalça displazisinde AVN'nin tedavisindeki seçenekler proksimal femurun biyomekaniğini iyileştirmede yoğunlaşmaktadır; ne yazık ki, daha sonraki evrelerde artrodez ya da artroplasti ihtiyacını ortadan kaldırmamaktadır.

Proximal femoral growth disturbance, commonly referred to as avascular necrosis (AVN) of the femoral head in the literature, is a potentially devastating complication in the treatment of developmental dysplasia of the hip (DDH). The early onset of debilitating degenerative changes in the hip joint when treatment options are limited remains the biggest fear. Controversy exists regarding the pathophysiology of this disorder, as well as exactly defining and diagnosing this problem. The natural history of AVN in the setting of DDH is very variable. Current practice concentrates primarily on prevention of this disorder, as there are no reliable ways to intervene at an early stage when changes are potentially reversible. Newer techniques using magnetic resonance imaging may have a role in early diagnosis of this condition before irreversible damage has occurred, and allow for interventions that will favorably affect the natural history of AVN. Treatment options for established AVN in DDH concentrate on improving the biomechanics of the proximal femur but unfortunately do not obviate the need for arthrodesis or arthroplasty at a later date.

Embryology of the proximal femur

The proximal femur develops from a single chondroepiphysis that encompasses the femoral head, the femoral neck and the greater and lesser trochanters.[1] Growth and development occur by appositional growth at the surface of the upper femur and epiphyseal growth at the junction of the cartilagenous upper femur and the femoral shaft. Between the fourth and seventh months of life the proximal femoral ossification center develops. This center continues to enlarge until skeletal maturity, when it is covered with only a thin layer of articular cartilage. With progressive maturation of the proximal femur three main growth centers develop. These

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are the physeal plate of the proximal femur, the growth plate of the greater trochanter and the growth plate along the femoral neck isthmus. Growth patterns in these three physes will determine the adult proximal femoral shape. Growth disturbances of any of these three centers, by whatever mechanism, will alter the shape of the proximal part of the femur.

Other factors also influence the development of the proximal femur. Forces across the joint that occur during weight bearing, muscle forces around the joint, joint nutrition and circulation all affect proximal femur development. Abnormal or excessive pressure on the femoral head will adversely affect cartilage perfusion and the subsequent development of this portion of the proximal femur. Abnormal growth patterns that occur depend on the portion of the proximal physis that is injured. Usually the physis of the greater trochanter is unaffected by these processes and it continues to grow normally resulting in relative trochanteric overgrowth.[2, 3]

Growth disturbances of the proximal femur

Growth disturbances of the proximal femoral epiphysis, also referred to as avascular necrosis (AVN) of the femoral head, remains a major complication following treatment of developmental hip dysplasia (DDH).[4,5,6] Potential sequelae that result from this problem include femoral head deformity, acetabular dysplasia and early onset of degenerative changes in the affected hip.[7] The incidence of this complication varies from 3% to 60%[8] Part of this large variability relates to the fact that the definition of what constitutes avascular necrosis is not agreed upon. Even the term AVN is disputed as it implies a pathophysiology that has not been correlated with any pathological specimen.[9]

The diagnosis of AVN is also a controversial area. The most commonly used criteria for diagnosing AVN are those established by Salter in 1969.[10] AVN is said to be present when there is failure of the femoral head to ossify or grow within one year of being reduced. Other radiological findings suggestive of a growth disturbance are widening of the femoral neck, changes in bone density of the femoral head and residual deformity suggestive of a growth disturbance. Postulated mechanisms for proximal femoral growth disturbances associated with DDH treatment include, excessive pressure applied to the femoral head that adversely affects the germinal cell layer of the physis. This is believed to be caused by sustained compressive forces transmitted through the epiphysis.[11] Another theory suggests possible extrinsic compression of the capsular blood supply of the hip thereby inhibiting perfusion.[5]

Two of the most commonly cited classification systems of AVN of the proximal femur associated with DDH treatment are those by Bucholz and Ogden[5], and Kalamachi and MacEwen.[12] The Bucholz/Ogden system is a 4-point classification system. Type 1 refers to irregular ossification of femoral head. No significant growth disturbance occurs and a good outcome results. Occasionally growth arrest lines may be visualized in the metaphysis that demonstrates growth is occurring.[10,13] Type II refers to a lateral metaphyseal injury. The femoral neck and head grow in to a valgus position because of premature lateral physeal closure. With a type III injury the entire metaphysis is affected and the result is a shortened femoral neck. Type IV refers to a medial growth plate injury where a radiolucent defect is observed along the medial metaphyseal area. This type of injury results in a varus deformity of the proximal femur. In types II, III and IV the greater trochanteric physis continues to grow normally resulting in relative trochanteric overgrowth.

Kalamachi and MacEwen's classification is also a four point scale. Similar to the Bucholz and Ogden classification, grade 1 refers to changes confined to the femoral head only and grade 2 involves a lateral growth plate arrest. A grade 3 injury refers to damage to the central physis and grade 4 indicates total physeal and head involvement.

Natural history

The natural history of proximal femoral growth disturbances varies enormously and it is difficult to separate it out from the underlying DDH process. Currently, it is only possible to diagnose AVN after a period of time has elapsed from the time of a specific intervention to the time radiologic evidence of AVN is present. Most reports in the literature have not controlled for variables that may affect the natural history of this diseas eprocess, such as age at presentation and the effects concomitant treatment have

on DDH. Cooperman et al.[14] reviewed the long-term results of AVN in the setting of DDH treated by closed reduction. 30 hips (25 patients) were followed for an average of thirty-nine years. Twenty-four of the thirty hips had moderate or severe osteoarthritis and twenty-two of the twenty-five patients had significant pain or loss of function, or both, by the time they were forty-two years old. The authors reported that the deformities produced by avascular necrosis that were related to osteoarthritis included loss of sphericity of the femoral head, persistent lateral and proximal subluxation, irregularity of the medial part of the femoral head, and acetabular dysplasia. Other authors suggest that patients with more extensive damage to the physis and/or persistent acetabular dysplasia have an increased risk of a poor outcome.[15,16]

Kim et al.[17] reported on acetabular development in DDH complicated by the most common type of proximal femoral growth arrest, Bucholz/Ogden type II. Their report studied forty-eight patients (fifty eight hips) treated by both open and closed methods. The average age at reduction was twenty-two months (range three to ninety seven months) and average age at follow up was twenty-one years (range ten to fifty years). Based on Severin classification thirty-four hips or 59% had a satisfactory outcome and were rated as Severin Class I or II. Twenty-four hips were rated as unsatisfactory, severin class III or IV. The authors concluded that proximal femoral growth disturbances are not always associated with poor acetabular development, those that are likely to have poor acetabular development are usually identifiable by age seven. They recommended close follow up of children who had treatment for DDH to identify those who may benefit from further intervention.

Treatment of DDH has evolved over the years in an attempt to improve on the natural history of the disease process. The main course of treatment of proximal femoral growth disturbances has been to try and prevent this problem from occurring. Earlier treatments using the Frejka pillow or immobilization in a position of forced internal rotation and/or wide abduction have been abandoned due to their association with higher incidences of AVN.[18,19,20]

Another controversial issue affecting the natural history of DDH is the timing of closed or open reduction with regard to the presence or absence of the capital femoral ossific nucleus. Segal et al.[21] in 1999 suggested that the ossific nucleus might have a protective role in decreasing the incidence of AVN when it is present on radiographs. In their study, they noted 1 case of AVN out of 25 treated hips when the ossific nucleus was present. While the incidence of AVN was 17 out of 32 hips when there was no ossific nucleus visible on x-ray. This group developed a porcine model in an attempt to demonstrate the protective role of the ossific nucleus.[22] Other authors have contradicted Segal's clinical findings. Both Luhmann et al.[23] and Ilfeld et al.[20] in their studies looking at 124 and 166 patients respectively failed to demonstrate any statistical difference in the rate of AVN that occurs between hips treated regardless of whether the ossific nucleus was present or not.

A major argument proposed against delaying reduction of a hip in the setting of DDH, is the idea that the greatest capacity for acetabular remodeling occurs when a hip can be reduced at an early age. A concentrically reduced femoral head acts as a stimulus for the acetabular cartilage to develop a spherical acetabulum. Delaying reduction to allow the ossific nucleus to develop may adversely affect the long-term outcome of acetabular development by delaying this opportunity for modeling to occur. In a study by Malvitz and Weinstein.[6] an increased incidence of AVN was seen when reduction was delayed. It is generally accepted now that early treatment of DDH is more important than waiting for the ossific nucleus to develop.

Traction in DDH

Pre-reduction traction was commonly used in the past in an attempt to avoid or reduce the risk of AVN in DDH. The theory behind this mode of treatment was that traction would allow gradual stretching of soft tissues that potentially create excessive pressure on the femoral head when it is reduced into the acetabulum, thereby reducing the risk of AVN. The arguments for, and against pre-reduction traction have been discussed by Weinstein.[24] He pointed out that traction in the setting of a dislocated hip has no real effect on intra-articular obstacles to reduction, and, as commonly used, probably has little effect on extra-articular obstacles to reduction. Previous studies reporting on the results of pre-reduction traction in DDH were poorly controlled for multiple parameters

such as position of immobilization, duration in traction, direction of traction and weight of traction used.[9,25] Other more rigorous studies have shown no real benefit in terms of a decreased incidence of AVN by using pre-reduction traction.[4,26] For a variety of reasons, including cost implications, pre-reduction traction is not as widely utilized in North America by paediatric orthopaedists as it has been in the past. Many now prefer to proceed directly to open reduction. Prereduction traction continues to be used in some European centers.[27]

Another method utilized by surgeons in the operative treatment of DDH in an attempt to minimize the risk of AVN occurring is concomitant femoral shortening at the time of performing an open reduction. This procedure has been demonstrated to decrease the incidence of AVN when performed in older children by Schoenecker and Strecker.[28] These authors compared femoral shortening directly with traction and demonstrated in a small series that femoral shortening was more effective in preventing AVN. Wenger has expanded the role of femoral shortening by utilizing this option in children as young as 5 months.[29] The indications for this procedure are evolving, but it clearly allows for reduction to be achieved without undue pressure being exerted on the femoral head. It can also be combined with varus or derotation to improve coverage of the femoral head.

A current major difficulty in treating AVN of the femoral head in the setting of DDH is the time delay that occurs in making the diagnosis. Unfortunately, a significant amount of time must elapse from the time an ischaemic insult occurs to the onset of radiologic changes that allow the diagnosis to be made. This situation makes treatment interventions at a time when ischaemia is potentially reversible very difficult to implement. It is theoretically possible to alter the course of AVN of the femoral head in DDH if the lesion could be identified at an early stage, before irreversible damage occurs. Preliminary work by Jaramillio et al. using MRI scans and diffusion weighted images in a piglet model have demonstrated differences in perfusion of the normal and ischaemic epiphysis.[30] With refinement, this work may become applicable in the human setting in helping to identify at an early stage, the reduced hip that may be at risk of developing ischaemia and AVN thereby allowing treatment to be altered appropriately.

Treatment of Established AVN

Treatment options for established AVN in the setting of DDH are limited and primarily attempt to optimize the biomechanics of the affected hip. The relative height of the greater trochanter and the length of the femoral neck determine the mechanical function of the hip abductor musculature. When the tip of the greater trohcanter reaches the level of the femoral head the patient usually develops a limp because the functional distance over which the hip abductor muscle fibers act is shortened, and the tension that is generated by these muscle during contraction is decreased.

Numerous procedures have been described to alter the anatomy of the proximal femur to improve the biomechanical function of the hip in the treatment of AVN associated with DDH. Procedures such as greater trochanteric epiphysiodesis, trochanteric advancement, and varus and valgus osteotomies are among the more commonly performed procedures attempting to address this complex problem. Trochanteric epiphysiodesis addresses relative overgrowth of the greater trochanter and is ideally performed when the child is around five years old. It is less effective if the child is more than eight years old.[31,32] The option of trochanteric advancement is available for the older child/adult when there is concentric reduction of the hip and a positive Trendelenberg sign due to relative trochanteric overgrowth. This procedure should be avoided if there is any radiological evidence of subluxation or significant dysplasia.[33]

A proximal femoral varus osteotomy can address the common deformity of increased hip valgus seen in Bucholz-Ogden type II injuries. Valgus osteotomies may be indicated for increased hip varus, this procedure can be combined with a greater trochanter transfer to improve hip biomechanics.

A theoretical intervention in the treatment of type II injuries that are recognized early is some form of growth arrest of the medial proximal femoral physis. However, this procedure has not been reported in the literature.

Other more complicated proximal femoral osteotomies such as the Wagner intertrochanteric

double osteotomy have been described to address the biomechanical problems associated with AVN of the hip in the setting of DDH.[34] Procedures combining both pelvic osteotomies and osteotomies of the proximal femur also attempt to positively influence the natural outcome of AVN by addressing biomechanical issues and improving coverage of the femoral head. Despite these interventions, the long-term prognosis of hips significantly affected by AVN in DDH is guarded. Ideally, the procedures described to treat AVN in this setting would prevent the need for hip arthrodesis or arthroplasty at a later date, but unfortunately, they may only defer this need.

Conclusion

Currently the best treatment of AVN associated with DDH is prevention. Research attempting to identify ischaemia of the femoral head before irreversible damage has occurred is at an early stage and is not yet applicable in the clinical setting. Treatments to primarily influence early proximal femoral growth disturbances by repairing physeal injuries or altering the blood supply to the physis also are not yet available. As a result, our current treatments can only secondarily address biomechanical issues that occur because of growth disturbances of the proximal femur in DDH, and not reverse these problems.

References

1. Watanabe RS. Embryology of the human hip. Clin Orthop Relat Res 1974;(98):8-26.
2. Gage JR, Cary JM. The effects of trochanteric epiphyseodesis on growth of the proximal end of the femur following necrosis of the capital femoral epiphysis. J Bone Joint Surg [Am] 1980;62:785-94.
3. Osborne D, Effmann E, Broda K, Harrelson J. The development of the upper end of the femur, with special reference to its internal architecture. Radiology 1980;137(1 Pt 1):71-6.
4. Brougham DI, Broughton NS, Cole WG, Menelaus MB. Avascular necrosis following closed reduction of congenital dislocation of the hip. Review of influencing factors and long-term follow-up. J Bone Joint Surg [Br] 1990;72:557-62.
5. Bucholz RW, Ogden JA. Patterns of ischemic necrosis of the proximal femur in nonoperatively treated congenital hip disease. In: The hip: Proceedings of the 6th Open Scientific Meeting of the Hip Society. St. Louis: Mosby; 1978. p. 43-6.
6. Malvitz TA, Weinstein SL. Closed reduction for congenital dysplasia of the hip. Functional and radiographic results after an average of thirty years. J Bone Joint Surg [Am] 1994;76:1777-92.
7. Thomas IH, Dunin AJ, Cole WG, Menelaus MB. Avascular necrosis after open reduction for congenital dislocation of the hip: analysis of causative factors and natural history. J Pediatr Orthop 1989;9:525-31.
8. Camp J, Herring JA, Dworezynski C. Comparison of inpatient and outpatient traction in developmental dislocation of the hip. J Pediatr Orthop 1994;14:9-12.
9. Ishii Y, Ponseti IV. Long-term results of closed reduction of complete congenital dislocation of the hip in children under one year of age. Clin Orthop Relat Res 1978;(137):167-74.
10. Salter RB, Kostuik J, Dallas S. Avascular necrosis of the femoral head as a complication of treatment for congenital dislocation of the hip in young children: a clinical and experimental investigation. Can J Surg 1969;12:44-61.
11. Schoenecker PL, Bitz M, Witeside LA. The acute effect of position of immobilization on capital femoral epiphyseal blood flow. A quantitative study using the hydrogen washout technique. J Bone Joint Surg [Am] 1978;60:899-904.
12. Kalamchi A, MacEwen GD. Avascular necrosis following treatment of congenital dislocation of the hip. J Bone Joint Surg [Am] 1980;62:876-88.
13. O'Brien T. Growth-disturbance lines in congenital dislocation of the hip. J Bone Joint Surg [Am] 1985;67:626-32.
14. Cooperman DR, Wallensten R, Stulberg SD. Post-reduction avascular necrosis in congenital dislocation of the hip. J Bone Joint Surg [Am] 1980;62:247-58.
15. Thomas IH, Dunin AJ, Cole WG, Menelaus MB. Avascular necrosis after open reduction for congenital dislocation of the hip: analysis of causative factors and natural history. J Pediatr Orthop 1989;9:525-31.
16. Weinstein SL. Natural history of congenital hip dislocation (CDH) and hip dysplasia. Clin Orthop Relat Res 1987; (225):62-76.
17. Kim HW, Morcuende JA, Dolan LA, Weinstein SL. Acetabular development in developmental dysplasia of the hip complicated by lateral growth disturbance of the capital femoral epiphysis. J Bone Joint Surg [Am] 2000;82: 1692-700.
18. Campbell P, Tarlow SD. Lateral tethering of the proximal femoral physis complicating the treatment of congenital hip dysplasia. J Pediatr Orthop 1990;10:6-8.
19. Thomas IH, Dunin AJ, Cole WG, Menelaus MB. Avascular necrosis after open reduction for congenital dislocation of the hip: analysis of causative factors and natural history. J Pediatr Orthop 1989;9:525-31.
20. Ilfeld FW, Makin M. Damage to the capital femoral epiphysis due to Frejka pillow treatment. J Bone Joint Surg [Am] 1977;59:654-8.
21. Segal LS, Boal DK, Borthwick L, Clark MW, Localio AR, Schwentker EP. Avascular necrosis after treatment of DDH: the protective influence of the ossific nucleus. J Pediatr Orthop 1999;19:177-84.
22. Segal LS, Schneider DJ, Berlin JM, Bruno A, Davis BR, Jacobs CR. The contribution of the ossific nucleus to the structural stiffness of the capital femoral epiphysis: a porcine model for DDH. J Pediatr Orthop 1999;19:433-7.
23. Luhmann SJ, Schoenecker PL, Anderson AM, Bassett GS. The prognostic importance of the ossific nucleus in the treatment of congenital dysplasia of the hip. J Bone Joint Surg [Am] 1998;80:1719-27.
24. Weinstein SL. Traction in developmental dislocation of the hip. Is its use justified? Clin Orthop Relat Res 1997; (338):79-85.
25. Kahle WK, Anderson MB, Alpert J, Stevens PM, Coleman SS. The value of preliminary traction in the treatment of congenital dislocation of the hip. J Bone Joint Surg [Am] 1990; 72:1043-7.

26. Sibinski M, Murnaghan C, Synder M. The value of preliminary overhead traction in the closed management of DDH. Int Orthop 2006;30:268-71.
27. Schoenecker PL, Strecker WB. Congenital dislocation of the hip in children. Comparison of the effects of femoral shortening and of skeletal traction in treatment. J Bone Joint Surg [Am] 1984;66:21-7.
28. Wenger DR, Lee CS, Kolman B. Derotational femoral shortening for developmental dislocation of the hip: special indications and results in the child younger than 2 years. J Pediatr Orthop 1995;15:768-79.
29. Jaramillo D, Connolly SA, Vajapeyam S, Robertson RL, Dunning PS, Mulkern RV, et al. Normal and ischemic epiphysis of the femur: diffusion MR imaging study in piglets. Radiolog 2003;227:825-32.
30. Gage JR, Cary JM. The effects of trochanteric epiphyseodesis on growth of the proximal end of the femur following necrosis of the capital femoral epiphysis. J Bone Joint Surg [Am] 1980;62:785-94.
31. Iwersen LJ, Kalen V, Eberle C. Relative trochanteric overgrowth after ischemic necrosis in congenital dislocation of the hip. J Pediatr Orthop 1989;9:381-5.
32. Lloyd-Roberts GC, Wetherill MH, Fraser M. Trochanteric advancement for premature arrest of the femoral capital growth plate. J Bone Joint Surg [Br] 1985;67:21-4.
33. Wagner H. Femoral osteotomies for congenital hip dislocations. In: Weil UH, editor. Progress in orthopaedic surgery. No 2, Acetabular dysplasia and skeletal dysplasia in childhood. New York: Springer-Verlag; 1978. p. 85-105.childhood. New York: Springer-Verlag; 1978. p. 85-105.