A Review of Avascular Necrosis in Developmental Dysplasia of the Hip and Contemporary Efforts at Prevention

Lael M. Luedtke, M.D., John M. Flynn, M.D., and Stephan G. Pill, M.S., P.T.

Abstract: Avascular necrosis (AVN) can be a devastating complication of the treatment of developmental dysplasia of the hip (DDH) and as such has received much attention. Despite this attention, little consensus exists regarding the definition, prevention, or treatment of AVN when it occurs. The literature is reviewed regarding the incidence, definition, and etiology of AVN. Diagnostic criteria are evaluated and the potential role of magnetic resonance imaging (MRI) is discussed. The currently used classification systems are also surveyed. More in-depth analysis is applied to the controversies regarding efforts at preventing AVN. These include the protective properties of the ossific nucleus, the current role of traction, the importance of position of immobilization, and femoral shortening osteotomies.

Introduction

Avascular necrosis (AVN) of varying severity is a well-known complication of the treatment of developmental dysplasia of the hip (DDH). It can be a devastating complication, potentially resulting in premature debilitating osteoarthritis. A milder end of the spectrum exists as well, with some patients demonstrating minimal residual deformity or dysfunction. Despite the potential significance of this problem, difficulties abound in determining the many features of this complication, which is not part of the natural history of DDH. The reported incidence in the literature ranges widely from 0% to 73% [2,3,5,8,10,19,40,41], reflecting the variability of making the diagnosis. This variability has not changed much over time; in reports of AVN in DDH in the 1990s, the incidence varies from 3% to 60% [2,4,7,17,22,23,34]. Many attempts have been made to understand the relationship between AVN and age at time of treatment, open reduction, closed reduction, prereduction traction, forceful reduction, and position of immobilization. The study of AVN has yielded some information, such as the importance of positioning, yet much essential knowledge remains elusive.

This paper focuses on our current understanding of AVN in DDH and how this relates to its prevention.

Definition

Precisely defining AVN has been difficult. Even use of the term AVN is disputed by some, as it implies a pathology that has not been correlated with an actual pathological specimen [14]. Weinstein [39,40] has coined the term "proximal femoral growth disturbance" instead of AVN; however, this fails to take into account the milder end of the spectrum of AVN, which does not result in significant growth disturbance. However, for convenience, the term AVN is used here and refers to the spectrum of changes seen in the proximal femur as a result of ischemic necrosis.

Etiology

Two commonly proposed mechanisms of ischemia are extrinsic blood vessel compression and excessive pressure on the femoral head, both of which prevent perfusion. Direct vascular obstruction would seem to portend more global effects, with a pressure phenomenon resulting in a more localized effect. Certainly, the proximal femur is vulnerable to an interruption in blood flow due to the particular developmental anatomy [3]. In the first six months of life, the medial circumflex artery supplies the medial and posterior epiphysis, metaphysis, and physis. The lateral circumflex artery supplies the lateral and anterior epiphysis, metaphysis, and physis. The circulation at this stage is predominantly an end-arteriole structure with little anastomotic overlap. After six months of age, the medial circumflex artery provides the blood supply for the entire epiphysis and physis through the posterior superior and posterior inferior branches (Fig. 1). The lateral circumflex goes to the anterior intracapsular metaphysis and the greater trochanter. With so much of the proximal femur dependent on the medial circumflex artery, the proximal femur becomes highly susceptible to any blockage of this vessel.

Excessive pressure on the femoral head has some basis in animal studies as a mechanism for developing AVN. Using a canine model, Schoenecker et al. [32] evaluated blood flow in the femoral head in several different positions as well as with compression applied through an external fixator. Compression dramatically decreased the perfusion of the femoral head in a statistically significant fashion. Gore [10], however, notes that pressure should not account for the physeal and metaphyseal changes. In addition, there are numerous accounts of the contralateral uninvolved hip developing AVN after treatment of the opposite hip [2,3,5,8,10]. There is not an intuitive mechanism for increased pressure in the uninvolved hip, so excessive abduc-
tion would appear to be the culprit in those instances. The likelihood is that both mechanisms play a role, creating the spectrum of changes that are recognized as AVN.

**Diagnosis**

Given that a definition of AVN is not universally accepted and the underlying mechanism is not yet fully understood, making the diagnosis can be challenging. The most commonly used diagnostic criteria for AVN were established by Salter et al. in 1969 [31]. Their criteria (Table 1), however, referred only to what they termed “total” AVN. Fragmentation of the femoral head or “temporary irregular ossification” when not preceded by increased density was not included in their definition of AVN, as it was believed to represent a revascularization phenomenon. This has resulted in at least one article that analyzed the cases with and without these hips [22], further complicating the literature of AVN. In 1972, Gage and Winter [8] provided a definition of partial AVN (Table 2). Many subsequent articles on AVN have used one or both of these systems to include or exclude cases in their studies [1,2,6,10]. Other studies do not explain their diagnostic criteria [1,11]. These criteria require subjective judgements and neither have been evaluated by the modern standards of interobserver or intraobserver reliability, thus calling into question much of the literature based on these criteria.

An essential element in making the diagnosis of AVN is allowing passage of sufficient time for changes to be manifest on a radiograph. In some instances, AVN did not become evident until 12 years following treatment [23]. Bucholz and Ogden [3] noted that their type II form of AVN was not evident until five years after treatment and on av-

<table>
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<th>Table 1. Criteria for total AVN</th>
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<td>AVN present if any of the following present:</td>
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<td>1. Failure of initial appearance of the capital femoral ossification center during one year or longer following reduction.</td>
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<td>2. Failure of continued chondro-osseous transformation and maturation of an existing epiphyseal ossification center during one year or longer following reduction.</td>
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<td>3. Broadening of the femoral neck (metaphysis) during one year following reduction.</td>
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<td>4. Increased radiographic density of the capital femoral ossification center followed by the radiographic appearance of fragmentation.</td>
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<td>5. Residual deformity of the femoral head and neck when ossification is complete.</td>
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(Data from Salter et al. [31]).

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Fig. 1. The blood supply to the proximal femur is superimposed over the immature hip. B: With the hip abducted, the posterior superior branch of the medial circumflex artery can become compressed [24].
Although lesser known and more recently published, Kruezynski systems are quite similar, yet each has its shortcomings. For the Kalamchi and MacEwen [18] (Table 4) systems. The time delay between the episode of ischemia and diagnosis presents another hurdle in the treatment of AVN. Diagnosing AVN long after the actual event occurs prevents intervention at an early phase when the necrosis might be reversible or even at a stage when further damage might be mitigated. O’Brien [26] proposed using the growth disturbance lines, commonly known as Harris growth arrest lines, as indicators of the health of the physis. The analysis by O’Brien et al. [27] in 1986 showed that the diagnosis of a physeal growth arrest could be made within six months of treatment. Other researchers, however, have not yet confirmed this work.

One possibility on the horizon is the use of gadolinium-enhanced magnetic resonance imaging (MRI). MRI has been used to study piglets positioned in extreme hip abduction, demonstrating femoral head ischemia at a reversible phase [5]. A preliminary study of immediate postreduction MRI in children [16] suggests that this is a reasonable option for imaging in lieu of the usual postreduction computed tomography (CT) scan. The MRI is performed immediately postreduction and is accomplished without further sedation. Jaramillo et al. [15,16] showed asymmetry of enhancement evident in those patients immobilized in wide abduction. However, this has yet to be correlated with evidence of AVN on plain films. Thus, MRI could provide valuable early information regarding ischemia of the femoral head as well as yield the same information about reduction and amount of abduction as the usual postreduction CT scan. In addition, MRI can also demonstrate any soft tissue obstacles to reduction.

Classification

Once the diagnosis has been made, classification of AVN usually follows either the Bucholz and Ogden [3] (Table 3) or the Kalamchi and MacEwen [18] (Table 4) systems. The systems are quite similar, yet each has its shortcomings. Although lesser known and more recently published, Kruezynski (21) has synthesized the elements of both into a more comprehensive system:

1. Involvement of the epiphysis, no fragmentation, mild changes.
2. Involvement of the epiphysis with fragmentation, moderate changes.
3. Involvement of the epiphysis and lateral metaphysis under the physis, severe changes.
4. Involvement of the epiphysis and the medial metaphysis under the physis, severe changes.
5. Involvement of the epiphysis and the entire metaphysis under the physis, severe changes.

Kruezynski’s system highlights the significance of changes in the physis and metaphysis as the determinants of the severity of changes seen in AVN. Limited involvement of the epiphysis alone results in minimal deformity and little long-term dysfunction [2,3,18,25,37]. Regardless of the system preferred, none of the classification systems has undergone the rigors of interobserver and intraobserver validation, as noted with the diagnostic criteria above. Thomas et al. [37] failed to correlate either the Bucholz and Ogden or Kalamchi and MacEwen classification schemes with outcome. However, Robinson and Shannon [29] did find the Kalamchi and MacEwen classification related to long-term results.

Table 2. Criteria for partial AVN

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<th>Each of the three following conditions must exist:</th>
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<td>1. Residual deformity of the femoral head two years or more after closed reduction (usually mild flattening of the medial aspect of the femoral epiphysis).</td>
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<td>2. Abnormalities in a specific area of the epiphysis visible on roentgenograms made within 12 months of reduction; occasional progression to fragmentation but most often manifest by failure of ossification of the nucleus.</td>
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<td>3. Roentgenographic evidence of viability of the remainder of the femoral head.</td>
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(Data from Gage and Winter [8].)

Table 3. Bucholz and Ogden classification

| 1. Complete fragmentation of the capital femoral ossific nucleus, slight widening of the femoral neck, and little long-term residual deformity. |
| 2. Fragmentation of the capital femoral epiphysis plus changes in the lateral aspect of the metaphysis and physis, often very delayed in presentation. |
| 3. Fragmentation of the capital femoral epiphysis plus the entire proximal femur affected. |
| 4. Fragmentation of the capital femoral epiphysis plus changes affecting the medial epiphysis. |

(Data from Bucholz and Ogden [3].)

Table 4. Kalamchi and MacEwen classification

| 1. Delay in the appearance of ossific nucleus or mottling of the cartilage model with little effect on the neck, minimal loss of height of the ossific nucleus, occasional coxa magna. |
| 2. Changes in ossific nucleus plus lateral physeal damage. |
| 3. Changes in ossific nucleus plus central physeal damage. |
| 4. Total damage to the head and physis. |

(Data from Kalamchi and MacEwen [18].)
cohort included patients whose age at time of closed reduction ranged from 1 month to 33 months. The natural history of DDH varies significantly in that range of ages. In Cooperman et al.’s evaluation of long-term results of AVN [6], the prognosis was quite grim, with 24 of 30 hips having moderate to severe osteoarthritis at an average follow-up of 39 years. However, none of these patients were under one year of age at the time of initial treatment for their DDH. This group already had a worse prognosis for their hips as established by Malvitz and Weinstein [23] before the AVN developed. Although a much smaller cohort, Gore’s long-term follow-up study [10] showed no degenerative changes in six cases at an average follow-up of 33 years. From the literature, two groups of patients appear to be at an increased risk of a poorer outcome: those with more extensive damage to the physis and those who have persistent acetabular dysplasia and subluxation [18,19,21,37].

**Prevention Efforts**

Treatment of DDH has evolved over the years toward the goal of safely improving on the natural history of the disease. Obviously, minimizing complications, such as AVN, is important in making the treatment less risky. Because the ischemic necrosis occurs occultly, little can be done to intervene in the process directly. Therefore, efforts have been concentrated on prevention to make treatment safer. Treatments such as the Frejka pillow and positioning in internal rotation have been abandoned due to their association with higher rates of AVN. Current issues relating to prevention of AVN include the importance of the presence of the ossific nucleus at the time of reduction, prereduction traction, positioning in immobilization, and femoral shortening.

One of the more recent dilemmas in the treatment of DDH is the timing of closed or open reduction with regard to the presence or absence of the ossific nucleus. Segal et al. [34] brought this issue to the forefront when they analyzed their patients. They noted that of the 25 hips with an ossific nucleus present radiographically, only one patient developed AVN. This contrasts with the 17 of 32 hips without an ossific nucleus that developed AVN. Segal et al. [35] used a porcine model to explain the protective role of the ossific nucleus. They demonstrated a significant increase in the compressive strain in the epiphysis when the ossific nucleus was present. This was noted particularly in the area of the posterior superior branch of the medial circumflex artery, which they believed accounted for the protective effect. Accordingly, their recommendation was to delay reduction until the ossific nucleus was evident or until the patient was 12 months of age. The clinical findings are contradicted, however, by Luhrmann et al. [22] and Ilfeld et al. [13]. These two studies of 124 and 166 patients, respectively, could not demonstrate a statistical difference in the rate of AVN between those with and without an ossific nucleus.

Delaying the reduction fails to take into consideration the findings of Malvitz and Weinstein [23] on the long-term outcome of DDH, which showed the rate of AVN to be significantly higher the longer reduction is delayed [38]. Those who do develop AVN at a younger age do seem to have an increased severity [13,18,30]. However, this is offset by the much greater capacity of the acetabulum to remodel in a younger child, which allows improved treatment of the underlying condition. Malvitz and Weinstein [23] note: “The younger the patient at the time of the reduction (if the reduction was maintained), the better the over-all function and radiographic appearance of the hip.” The ultimate long-term function of the hip affected by DDH has clearly been shown to be related to persistent acetabular dysplasia and subluxation. Long-term analysis shows a consistent decline in radiographic results as the age at treatment increases [13]. This has been reaffirmed by other studies [14,17]. Current data support the prompt treatment of DDH in infants rather than waiting in hopes of avoiding AVN.

Prereduction traction, another effort at avoiding AVN, has fallen out of favor with many orthopaedists. Conventional wisdom had been that the use of traction would stretch the soft tissues that create excessive pressure on the femoral head after reduction, resulting in AVN. Weinstein [39] summarized the arguments for and against the effectiveness of traction, pointing out that the intraarticular obstacles would not be affected by traction and that traction, as used, also failed to have much of an effect on the extraarticular obstacles. Studies advocating traction to avoid AVN were poorly controlled for direction, time in traction, and weight [3,4,7,14,18]. In addition, they failed to control variables that affect the rate of AVN (e.g., position of immobilization and age at time of reduction) [8]. More rigorous studies have subsequently shown no significant change in the rate of AVN when employing traction [2,17]. One explanation of the apparently contradictory information regarding traction could be the difference between inpatient traction and home traction used most often today. Certainly, in some communities, prereduction traction remains the standard of care. For some surgeons, prereduction traction may be little more than a gesture toward a treatment tradition or a medicolegal environment. Many pediatric orthopaedists have ceased using traction and proceed directly to reduction.

Unlike the questionable benefit of traction, the importance of the postreduction immobilization position is strongly supported in the literature [8,11,15,27,31,41]. The hip is clearly at risk in wide abduction and internal rotation. Despite this knowledge, Smith et al. [36] showed that postreduction CT scans demonstrate that patients are still immobilized in extremes of abduction. Follow-up of these patients demonstrates increased rates of AVN with wide abduction. Their study further demonstrated that no patient developed AVN if the abduction was under 55 degrees. They were not able to show a limit beyond which AVN always developed. Thus, the knowledge we do possess about preventing AVN must be more consistently applied in order to minimize this complication (Fig. 2).

One of the more effective means of reducing the rate of AVN, particularly in the older child, is concomitant femoral shortening at the time of reduction (Fig. 3). Schoenecker
and Strecker [33] compared femoral shortening directly with traction and showed that femoral shortening was significantly more effective in preventing AVN in a small series. Galpin et al. [9] and Wenger et al. [41] have further explored this approach and expanded its indications to children as young as five months. Furthermore, the procedure appears to have few negative consequences [9,20]. The theoretical advantage is that the shortening allows an excellent reduction without excessive pressure on the femoral head. The shortening can also be combined with varus and or derotation, if needed, in the older child [12]. The indications for femoral shortening are evolving. Heinrich et al. [12] recommend open reduction with femoral shortening for any patient over the age of four years. For patients younger than four, the surgeon’s judgement must be used to determine if the reduction will be under excessive pressure.

Fig. 2. CT following closed reduction of the right hip of a 12-month-old. Preoperative traction was used. The ossific nucleus was present. The reduction was easy and without force. The CT shows excessive abduction of 62 degrees. The hip is well reduced. B: Plain radiograph of the pelvis at the time of the first cast change, six weeks after closed reduction, shows a concentric reduction. A well-formed ossific nucleus appears normal. C: Plain radiograph one year after closed reduction. Fragmentation of the ossific nucleus and widening of the proximal femoral metaphysis are consistent with AVN/proximal femoral growth disturbance.

Fig. 3. Plain radiograph of a 20-month-old with a high dislocation of the right hip and a well-formed false acetabulum and femoral ossific nucleus. B: Plain radiograph one year after open reduction with femoral shortening and Pemberton osteotomy. There is no evidence of AVN/proximal femoral growth disturbance. Note the Harris growth line in the proximal femur, suggesting that there has been no significant disturbance in growth [26, 27].
AVN remains a poorly understood complication of the treatment of DDH. Conclusions about prevention and treatment are difficult to reach when the diagnostic and classification criteria have not been validated. The long-term studies of Malvitz and Weinstein [23] are helpful in establishing priorities, such as in the case of the role of the ossific nucleus. The elements that determine the long-term prognosis, such as persistent acetabular dysplasia and subluxation, must be balanced with the intent to minimize complications. The current practice of unsupervised home traction has failed to prevent AVN. However, avoiding wide abduction and judicious use of femoral shortening can have an impact on the occurrence of AVN while treating DDH. With advances in technology such as gadolinium-enhanced MRI, early diagnosis and intervention will be the future approach to AVN in DDH.

The authors’ treatment recommendations based on the review of the literature include reduction of hips that have failed standard Pavlik harness treatment as soon as possible to maximize the time remaining for acetabular remodeling. This reduction should be performed regardless of whether the ossific nucleus is present or absent. Although prereduction home traction is still the standard in many communities, its effect on preventing AVN is not supported by the literature. If there is significant pressure on the head after open reduction in the judgement of the surgeon or in the child older than four years, femoral shortening should be used at the time of open reduction. Careful attention should be paid to the position of the patient at the time of spica cast application, perhaps using a goniometer, to avoid immobilization in abduction greater than 55 degrees. Postreduction CT scans should be analyzed for the extent of abduction, as well as for reduction of the dislocation.

Summary

AVN remains a poorly understood complication of the treatment of DDH. Conclusions about prevention and treatment are difficult to reach when the diagnostic and classification criteria have not been validated. The long-term studies of Malvitz and Weinstein [23] are helpful in establishing priorities, such as in the case of the role of the ossific nucleus. The elements that determine the long-term prognosis, such as persistent acetabular dysplasia and subluxation, must be balanced with the intent to minimize complications. The current practice of unsupervised home traction has failed to prevent AVN. However, avoiding wide abduction and judicious use of femoral shortening can have an impact on the occurrence of AVN while treating DDH. With advances in technology such as gadolinium-enhanced MRI, early diagnosis and intervention will be the future approach to AVN in DDH.

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References