Coxa Vara in Childhood: Evaluation and Management

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Abstract

Coxa vara in childhood may be clinically classified as developmental, congenital, dysplastic, or traumatic and may occur at the physis or in the trochanteric or subtrochanteric area. Evaluation should include a search for a family history of similar deformity, a history of trauma or infection, and evidence of associated skeletal abnormality. Radiographs will illustrate whether the deformity is unilateral or bilateral and whether it occurs at or below the physis. With this information, coxa vara can be classified, and the optimal treatment can be selected. Surgical treatment of coxa vara in childhood is usually indicated when the disease is progressive, painful, unilateral, or associated with leg-length discrepancy.

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Coxa vara is an uncommon childhood deformity. It may be clinically classified as developmental, congenital, dysplastic, or traumatic. It may also be classified by the anatomic site, which may be at the physis or in the trochanteric or subtrochanteric area. Determination of both the location and the cause is important to prognosis. Coxa vara occurring at the physis is often progressive and rarely improves. Coxa vara that occurs distal to the physis is usually traumatic or congenital. The traumatic type improves; the congenital type does not. Thus, it is important to classify coxa vara before selecting treatment.

Anatomy

In early skeletal development, a common physis serves the greater trochanter and the capital femoral epiphysis. This physis divides as growth continues in a balance that favors the capital femoral epiphysis and creates the normal neck-shaft angle.[1] The cervicofemoral angle is approximately 35 degrees in infancy and increases to 45 degrees by skeletal maturity. The corresponding angle of inclination is 135 degrees at skeletal maturity. Coxa vara refers to an increase in the cervicofemoral angle or a decrease in the angle of inclination (Fig. 1).

Radiographic Measurements

Measurement of coxa vara or the neck-shaft angle is complicated by the presence of anteversion or torsion of the femur. In most clinical situations, an anteroposterior (AP) radiograph with the limb internally rotated to place the femoral neck parallel to the film is adequate for diagnosis and treatment. Hilgenreiner's epiphyseal angle is used to measure the severity of deformity. In developmental coxa vara, this is the angle formed by Hilgenreiner's line and an intersecting line through the physis (Fig. 2).

If more accurate measurement of the angle of inclination and anteversion is required, standard AP and lateral radiographs are taken without moving the femur. The apparent anteversion and apparent cervicofemoral angle are then measured and converted to the true anteversion and the true cervicofemoral angle with the use of tables.[2] Anteversion may also be measured with computed tomography.[3]

Further Evaluation and Management

In addition to radiographic measurements, evaluation should include a

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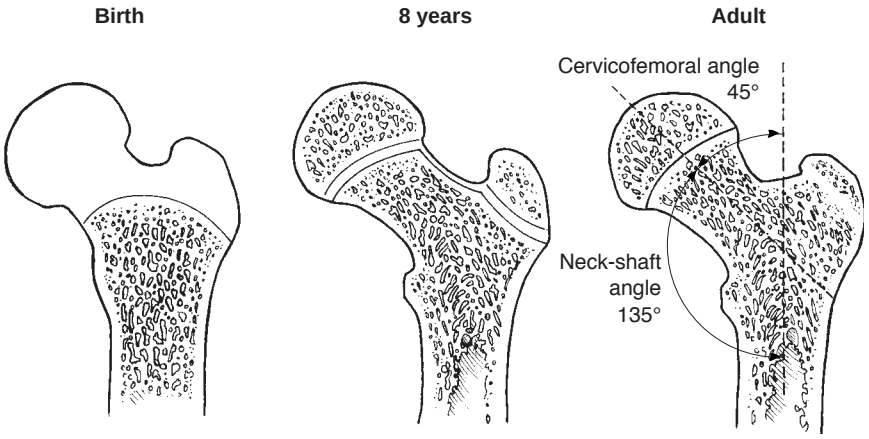


Fig. 1 At birth, there is a common physis for the proximal femur. The physis divides to create separate physes for the capital epiphysis and the greater trochanter. Different rates of growth create the normal neck-shaft angle. The normal adult neck-shaft angle (angle of inclination) is about 135 degrees. The normal cervicofemoral angle is about 45 degrees.

search for a family history of similar deformity, a history of trauma or infection, and evidence of associated skeletal abnormality. Laboratory studies may help to identify patients with metabolic disorders.

Surgical management of coxa vara is usually indicated when the deformity is progressive or is associated with limb-length inequality, pain, or limp. Surgical management is not often indicated if the deformity is moderate, nonprogressive, and not associated with a limp. Although moderate coxa vara alters the mechanical axis of the hip, it is not known to cause protrusio acetabuli, osteoarthritis of the hip, or ipsilateral knee disorders.

Developmental Coxa Vara

Developmental coxa vara occurs at the physis. It has historically been called congenital or infantile coxa vara and occurs in 1:25,000 births.[4] It is not part of a generalized bone dysplasia, nor is it congenital, as was once believed. The deformity is bilateral in one third to one half of affected patients. It has been reported in siblings, but its occurrence is usually sporadic. Progression is common and is thought to be related to biomechanical stress on the physis, analogous to Blount's disease at the knee.

Developmental coxa vara is radiographically characterized by development of a dense triangular portion of the femoral calcar. Because it is not possible to predict progression in developmental coxa vara with certainty, all affected patients need to be monitored. Those with a Hilgenreiner's epiphyseal angle less than 45 degrees are unlikely to worsen, and those with an angle over 60 degrees usually worsen; in those with an angle between 45 and 60 degrees, the outcome is less predictable.[5]

Early surgery is not indicated for developmental coxa vara, but a valgus-producing osteotomy is indicated for progressive or severe deformity.[6,7] The adage "Overcorrect or correct over" is useful, as it indicates the importance of placing the physis in a nearly horizontal position to reduce shear stress and prevent recurrence of deformity (Fig. 3). Premature closure of the capital physis may complicate treatment and subsequently necessitate trochanteric epiphysiodesis or transfer.[8]

Congenital Coxa Vara

Congenital coxa vara is usually associated with a congenitally short femur, but may be associated with all degrees of proximal femoral focal deficiency.[9,10] It is nearly always unilateral. Femoral deformity is present in the subtrochanteric area where the bone is bent, the cor-

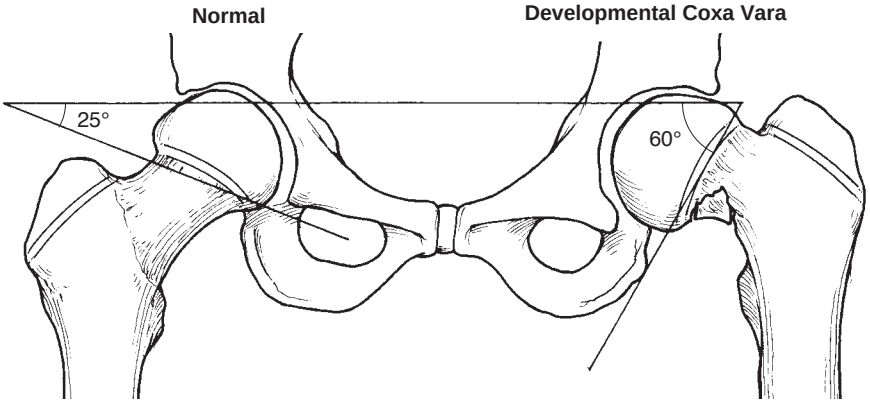


Fig. 2 Hilgenreiner's epiphyseal angle is created by a line through the triradiate cartilage and its intersection with a line through the physes. The normal angle is about 25 degrees.

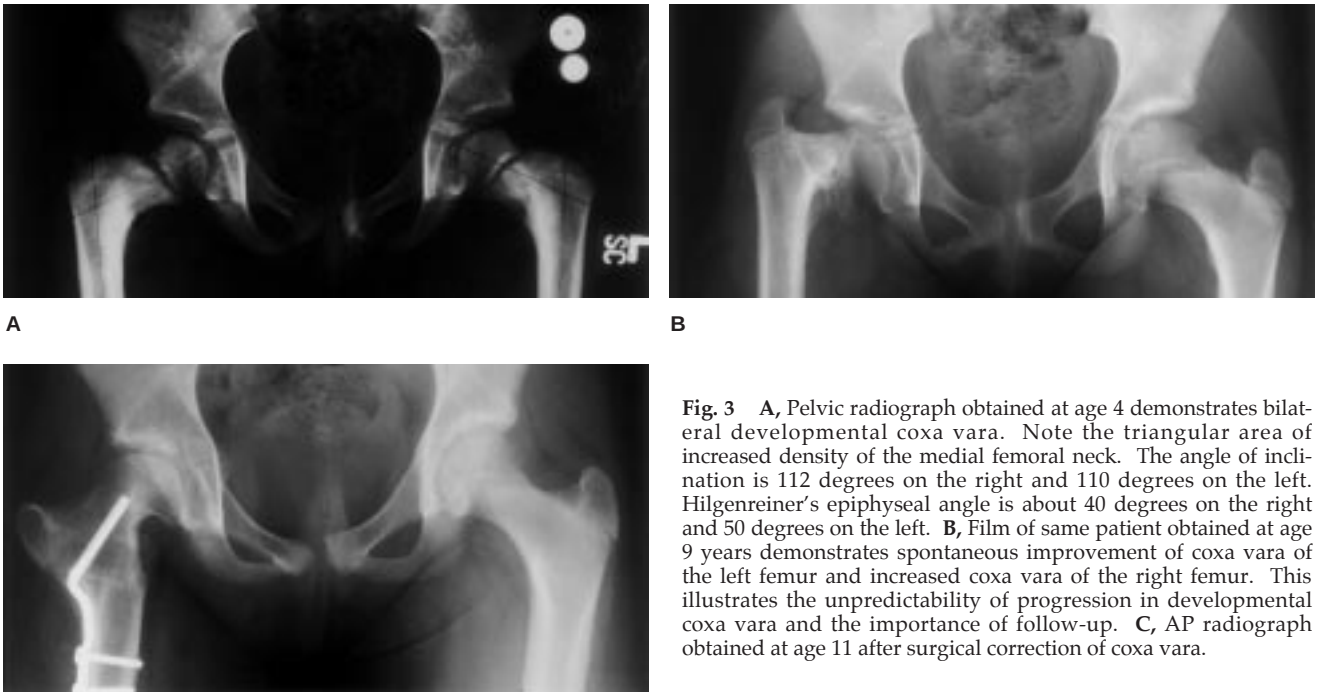


Fig. 3 **A,** Pelvic radiograph obtained at age 4 demonstrates bilateral developmental coxa vara. Note the triangular area of increased density of the medial femoral neck. The angle of inclination is 112 degrees on the right and 110 degrees on the left. Hilgenreiner's epiphyseal angle is about 40 degrees on the right and 50 degrees on the left. **B,** Film of same patient obtained at age 9 years demonstrates spontaneous improvement of coxa vara of the left femur and increased coxa vara of the right femur. This illustrates the unpredictability of progression in developmental coxa vara and the importance of follow-up. **C,** AP radiograph obtained at age 11 after surgical correction of coxa vara.

tices are thickened, and there may be an associated overlying skin dimple (Fig. 4). It is associated with external rotation of the femur (retroversion) and often with valgus of the knee. There may be other associated skeletal anomalies, most commonly fibular deficiency. The congenitally short femur–abnormal facies syndrome is a rare condition associated with bilateral congenital short femora and coxa vara.[11]

In congenital coxa vara, the deformity does not spontaneously resolve. The percentage of shortening of the femur compared with the normal side is a constant (i.e., if the femur is 15% shorter than normal in infancy, it will be 15% shorter when the patient is an adult). Ultimate shortening can be estimated to assist in selection of treatment. Surgical treatment of the femur in congenital coxa vara may include a valgus-producing osteotomy to improve hip mechanics and length and a rotational osteotomy to correct retroversion and lengthening.[10,12]

Dysplastic Coxa Vara

Many generalized skeletal dysplasias and diseases are associated with coxa vara, including vitamin D–resistant rickets, fibrous dysplasia, Paget's disease, and osteopetrosis. In these conditions, the deformity is primarily in the subtrochanteric area and is usually part of generalized bowing of the femur. Laboratory studies may be helpful in diagnosing vitamin D–resistant rickets and Paget's disease, but are normal in most bone dysplasias.

Most generalized bone dysplasias associated with coxa vara are metaphyseal dysplasias.[13] In these conditions, varus deformity at the physis is often progressive and may be associated with sym-

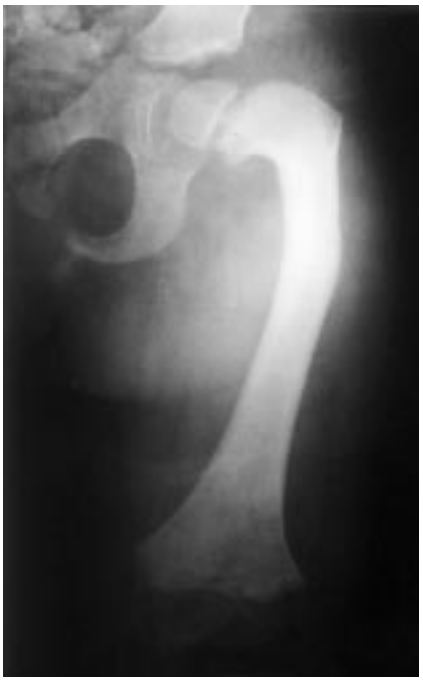


Fig. 4 Radiograph of a child with congenital coxa vara demonstrates that the subtrochanteric area and diaphysis is the principal site of varus deformity. The femur is also short.

metrical delay in ossification of the capital femoral epiphysis. Examples of generalized dysplasias associated with coxa vara include those described by Jansen, Schmid, and Strudwick, as well as spondylometaphyseal dysplasia, spondyloepiphyseal dysplasia, sponastrime dysplasia, and cleidocranial dysplasia.[14-16] The presence of a generalized skeletal dysplasia is suggested by short stature, deformity, dysmorphism, abnormality in other parts of the skeleton, and bilateral coxa vara.

Surgical treatment of dysplastic coxa vara should await ossification of the femoral head and determination of the rate and magnitude of progression of varus. The prognosis varies with the dysplasia, but surgery is often beneficial. Subtrochanteric osteotomy to correct varus angulation may have to be repeated if the deformity recurs (Fig. 5).

Traumatic Coxa Vara

Traumatic coxa vara may result from physeal separation in the newborn or from fracture or osteotomy of the proximal femur in childhood. Coxa vara has also been demonstrated experimentally to develop when there is an injury to the proximal femoral physis while the trochanteric physis continues to grow.[17] Clinically, injury to the proximal femoral physis leading to coxa vara may be due to vascular injury or infection.

Coxa Vara Secondary to Perinatal Epiphyseal Separation

Traction and external rotation of the lower extremity during delivery may cause traumatic displacement of the proximal femoral epiphysis. It is rare and is most often associated with breech delivery. About one third of such injuries result in coxa vara. The diagnosis should be suggested by a history of difficult breech delivery, lack of spontaneous movement of the limb, and swelling and ecchymosis of the proximal thigh. The limb is of normal length but lies in external rotation.[18]

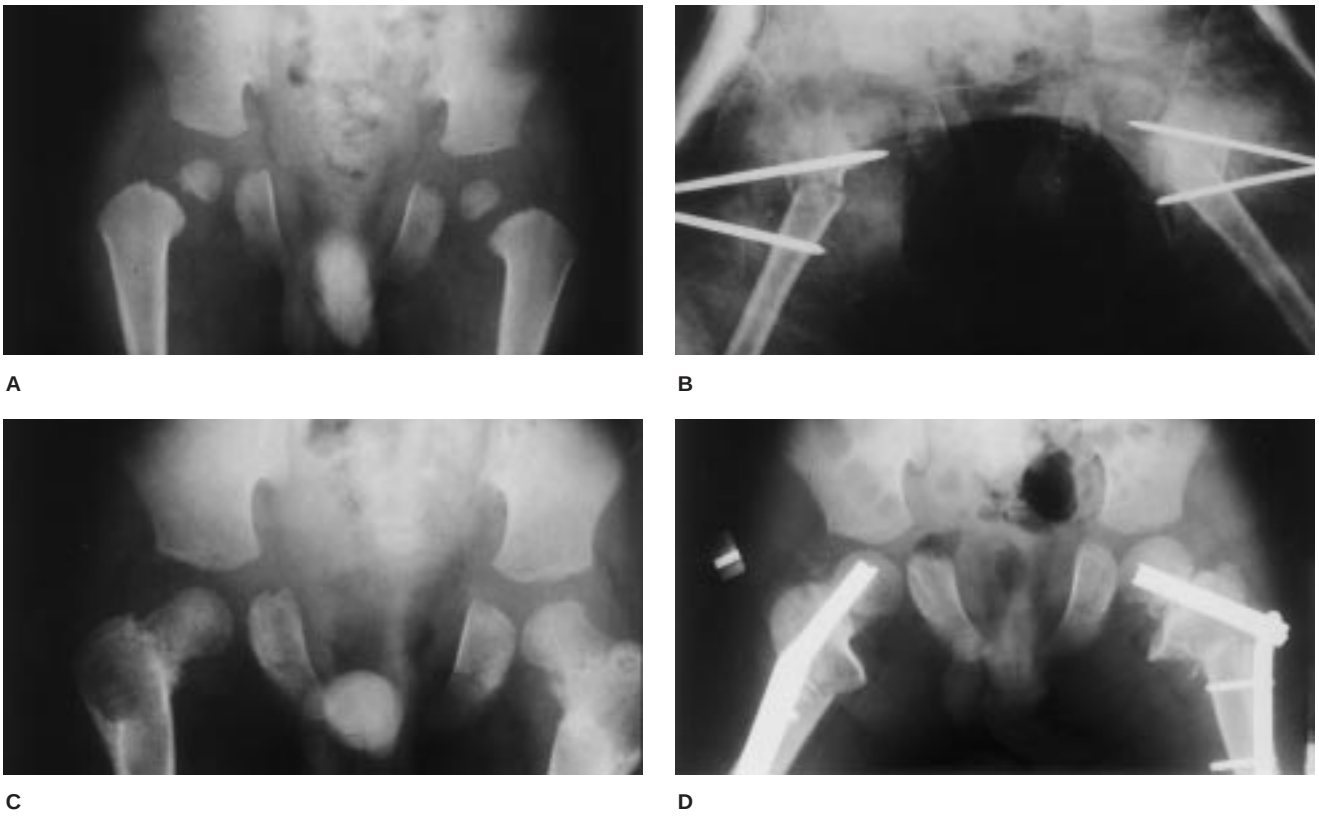
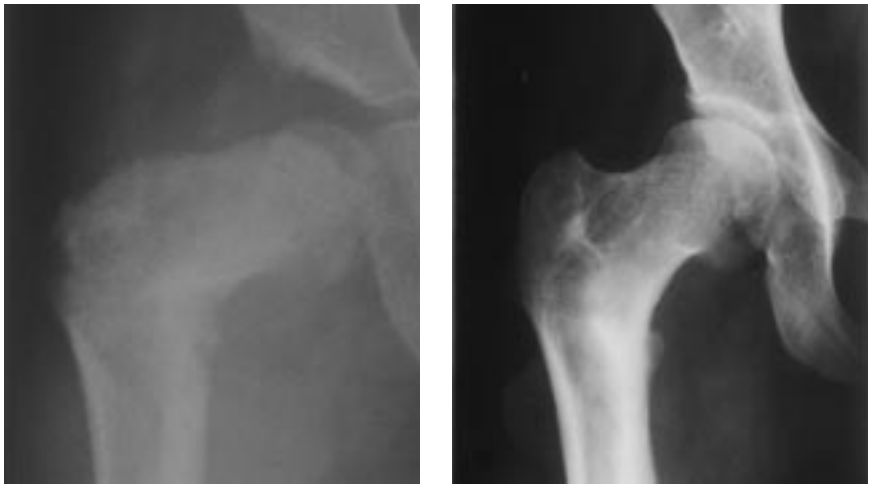


Fig. 5 **A,** AP radiograph of the pelvis of a 3-year-old child with cleidocranial dysplasia shows delay in ossification of the pubis, widening of the triradiate cartilage and femoral physis, and early coxa vara. **B,** Progression of deformity was treated at age 4 by valgus-producing osteotomies. **C,** Radiograph at age 5 demonstrates healed subtrochanteric osteotomies with early redevelopment of coxa vara and narrow physes. **D,** Radiograph at age 8 depicts repeat valgus osteotomy. Transphyseal fixation was used because of recurrence of deformity.

In the newborn, the presence of the deformity can be inferred from radiographic demonstration of lateral displacement of the proximal ossified femur or can be demonstrated early with ultrasound[19] or magnetic resonance imaging. Metaphyseal periosteal ossification occurs about 7 to 10 days after birth.

The treatment of perinatal traumatic physeal separation is spica casting. Healing is prompt, and progressive coxa vara does not occur. However, accelerated local skeletal maturation does occur and may be associated with persistent external rotation. Limb lengths are equal, as this injury does not affect the growth of the proximal femoral physis.



A B

Fig. 6 **A,** Radiograph demonstrates a healed varus-producing subtrochanteric osteotomy performed for acetabular dysplasia in a child. **B,** Film obtained at skeletal maturity depicts remodeling with restoration of the nearly normal cervicofemoral angle.

Coxa Vara Due to Fracture or Osteotomy of the Proximal Femur

Traumatic coxa vara in childhood may result from malunion after fracture of the femoral neck or trochanter. More often, it is the result of a varus-producing osteotomy of the proximal femur performed as part of the treatment of developmental hip dislocation or of hip subluxation and dislocation associated with cerebral palsy. Traumatic coxa vara of this type undergoes spontaneous remodeling. The mild associated leg-length discrepancy tends to correct spontaneously (Fig. 6).

Coxa Vara Secondary to Physeal Sepsis and Trochanteric Overgrowth

Coxa vara may result from injury to the proximal femoral epiphysis and/or physis due to neonatal or childhood sepsis. In infancy, the septic process may destroy the entire femoral head or may be associated with separation of the head and neck. The options for treatment when there is no femoral head include placing the greater trochanter into the acetabulum or the use of pelvic-support osteotomies.[20]

The septic process may destroy only the epiphysis, leaving a residual nubbin of calcar, which may act as a femoral head. Continued growth of the greater trochanter produces coxa vara. Limb shortening by 3 to 4 inches at skeletal maturity can be expected, as 10% of the growth of the lower limb comes from the proximal femoral physis. Treatment may include a valgus osteotomy, taking care to keep the calcar remnant in the acetabulum; trochanteric epiphysiodesis or transfer; and femoral lengthening.[12]

Coxa Vara Due to Vascular Injury to the Physis and Trochanteric Overgrowth

Injury to the vessels supplying the cartilaginous femoral head may lead to a delay in ossification and failure of ossification of the epiphysis and growth of the proximal physis. Normal growth of the greater trochanteric physis with impaired growth of the physis of the femoral head results in coxa vara. This is most often seen as a manifestation of Perthes disease or occurs after treatment of hip dislocation, in which case it may occur on both the nonaffected and affected sides. It has been reported to occur more often when traction has not been used before reduction of a dislocated hip and after wide abduction of the hip, which may cause obstruction of the lateral epiphyseal vessels.

The radiographic manifestations of vascular injury to the capital femoral epiphysis and physis are failure of growth of the ossific nucleus, fragmentation and deformity of the femoral head, and failure of growth of the femoral neck. Coxa vara associated with vascular injury is usually asymmetrical.

This type of coxa vara is assessed radiographically by comparing the growth of the femoral physis with the growth of the greater trochanter (Fig. 7). The distance from the lesser trochanter to the top of the femoral head reflects the growth of the capital femoral epiphysis; the distance from the lesser trochanter to the top of the greater trochanter reflects the

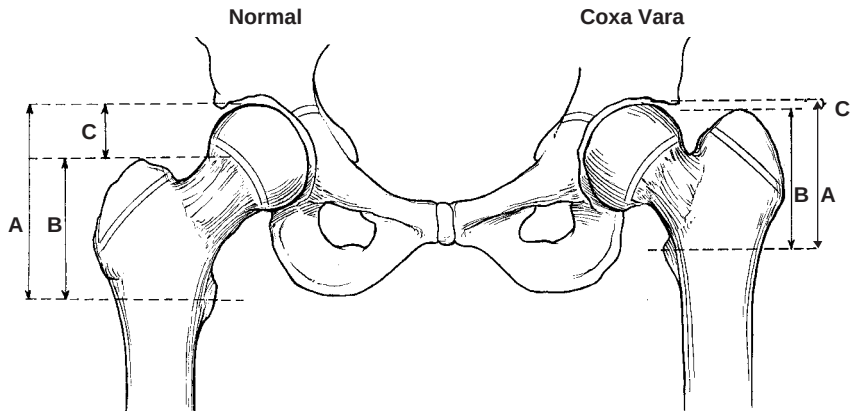


Fig. 7 Coxa vara due to impaired growth of the capital femoral physis is measured by comparing the growth of the femoral physis and the trochanteric physis. The measurement from the lesser trochanter to the top of the femoral head **(A)** reflects growth of the capital physis, and the measurement from the lesser trochanter to the top of the greater trochanter **(B)** reflects trochanteric growth. In the normal hip **(left),** the distance from the top of the greater trochanter to the top of the femoral head **(C)** is about 20±5 mm. In the hip with coxa vara due to impaired growth of the capital femoral physis **(right), A** decreases and **B** is normal, thereby decreasing **C**.

growth of the greater trochanter.[21] The distance from the top of the femoral head to the top of the greater trochanter is normally 20±5 mm. When this distance decreases, there is associated abductor weakness, limited abduction, and limp.[21]

Hip mechanics after this type of coxa vara can be stabilized by performing an epiphysiodesis of the greater trochanteric physis when the apophysis becomes visible radiographically, which usually occurs about age 5 years. After age 9, distal transfer of the greater trochanter is necessary to improve hip mechanics (Fig. 8).

Summary

Coxa vara may be clinically classified as developmental, congenital, dysplastic, or traumatic (Table 1). It may also be classified by the anatomic site, which may be at the physis or in the trochanteric or subtrochanteric area. Determination of both the location and the cause is important to prognosis. Coxa vara occurring at the physis is often progressive and rarely improves. Coxa vara that occurs distal to the physis is usually traumatic or congenital. The traumatic type improves; the congenital type does not.

Appropriate selection of treatment is dependent on accurate clas-

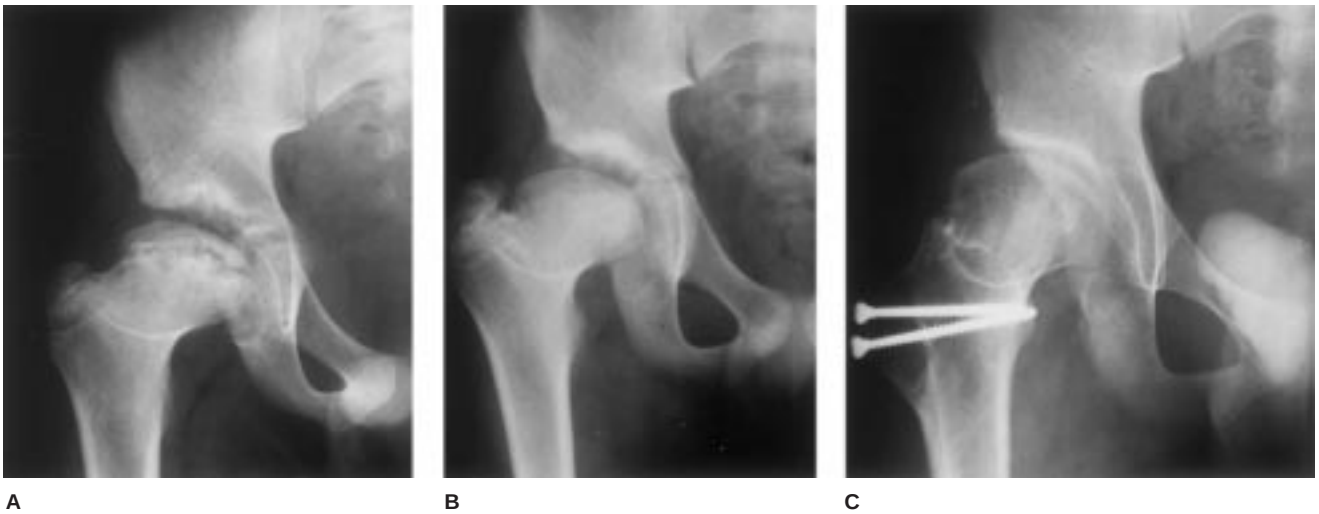


Fig. 8 Coxa vara due to vascular injury and trochanteric overgrowth. **A,** Radiograph obtained after closed treatment of hip dislocation demonstrates coxa breva, plana, and vara. The capital physis is narrowed and irregular. **B,** Coxa vara progressed as the greater trochanteric physis continued to grow and the femoral physis closed. There was no change in the distance from the lesser trochanter to the top of the femoral head, but the distance from the lesser trochanter to the top of the greater trochanter increased. **C,** Distal transfer of the greater trochanter improved hip mechanics.

Table 1
Classification of Coxa Vara

Type	Physeal Location	Progressive	Remodels	Generalized Dysplasia
Developmental	Yes	Often	No	No
Congenital	No	No	No	No
Dysplastic	Usually	Often	No	Yes
Traumatic				
Perinatal injury	Yes	No	Yes	No
Fracture/osteotomy	No	No	Yes	No
Vascular injury	Yes	Yes, trochanteric overgrowth	No	No
Septic injury	Yes	Yes, trochanteric overgrowth	No	No

sification, which in turn is dependent on a thorough evaluation. Evaluation should include a search for a history of trauma or sepsis, a family history, and identification of associated abnormalities. Radiographs will indicate whether the condition is unilateral or bilateral, whether the bone abnormalities are generalized or localized, and whether the deformity is physeal or nonphyseal. Laboratory studies may help to identify patients with metabolic disorders. With this information, the diagnosis and prognosis can usually be established, and an appropriate treatment plan can be devised.

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