CHAPTER 18 Congenital Coxa Vara

Congenital coxa vara is a developmental abnormality characterized by a primary cartilaginous defect in the femoral neck with an abnormal decrease in the femoral neck-shaft angle, shortening of the femoral neck, relative overgrowth of the greater trochanter, and shortening of the affected lower limb. Typically the deformity either is not present at birth or is sufficiently subtle as not to be recognized at that time.[1,2] Affected patients almost invariably present after walking age, and sometimes as late as adolescence, with a limp (Trendelenburg and/or short-leg gait) and, in unilateral cases, relatively mild limb length inequality.

This disorder has specific radiographic characteristics and a clinical presentation unique to it, and should be distinguished from acquired causes of coxa vara deformity and coxa vara associated with congenital femoral deficiency.* Confusion and controversy, however, exist in the literature as to the terminology and classification of this disorder. It has been variously referred to as "congenital," "developmental," "cervical," or "infantile" coxa vara.† To add to the confusion, some authors, in discussing developmental or congenital coxa vara, do not distinguish between the disorder described in this chapter and congenital coxa vara with short femur[3,4,23,39–41]; others include acquired causes of coxa vara. Furthermore, some cases of coxa vara are associated with skeletal dysplasias, especially cleidocranial dysostosis, metaphyseal dysostosis, and some types of spondylometaphyseal dysplasia.[21,33,52,53,55,76,80] Some authors exclude, others include, this skeletal dysplasia–associated type of coxa vara in series describing developmental coxa vara. We believe that the term *developmental coxa vara* should be used to describe the clinical entity characterized by postnatal presentation of coxa vara without other known cause, with typically mild limb shortening and characteristic radiographic features; the deformity may or may not be associated with a generalized skeletal dysplasia. Acquired forms of coxa vara and those associated with significant femoral deficiency should be considered separate entities (Table 18–1).

Fiorani in 1881[35] was the first to publish a clinical description of a case of bending of the neck of the femur. The term *coxa vara* was coined by Hofmeister in 1894.[46] The association of coxa vara with other malformations was noted by Kredel in 1896.[54] Amstutz in 1970 described two patients with coxa vara who had previously had normal radiographs of the hips.[2] The term *developmental coxa vara* was first used by Hoffa[45] in 1905 and later by Duncan.[26] As mentioned previously, subsequent authors have used not only "developmental" but also "congenital," "infantile," or "cervical" coxa vara to describe this condition.

Incidence

Developmental coxa vara is rare; its incidence is estimated by Johanning to be 1 in 25,000 live births in the Scandinavian population.[49] There is no racial predilection. The disorder appears to be equally common in males and females. Various series report the ratio of unilateral to bilateral cases to be between 1 : 2[25] and 3 : 1.[75] Bilateral cases may be more likely to be associated with a generalized skeletal dysplasia, so the examiner should seek further evidence of such dysplasia during the physical and radiographic examination of the patient.

Heredity

In addition to the presence of the disorder in skeletal dysplasias with known genetic cause (cleidocranial dysostosis, metaphyseal dysostosis [especially Jansen type], and spondylometaphyseal dysplasia [especially Kozlowski type], all of which are autosomal dominant disorders), there is a presumed genetic cause in "isolated" developmental coxa vara as well. There are reports of the condition in families and in both homozygous and heterozygous twins.[6,9,26,36,59,72]

Clinical Features

The deformity does not become manifest until after birth and usually not until walking age. Clinically, the child presents with a painless limp due to a combination of true Trendelenburg gait and relatively minor limb length inequality in unilateral cases. Easy fatigability or aching pain

*See references 2, 10, 11, 17, 19, 49, 67–69, 75, 80.

†See references 1, 2, 5, 7, 8, 10, 11, 15, 19, 20, 22, 24–32, 34, 36, 38, 39, 43, 47–49, 57, 58, 60, 64, 65, 67–69, 72, 75, 77, 79–81.

If a generalized skeletal dysplasia is suspected on the basis of the family history or short stature in the affected individual, a skeletal survey will reveal features characteristic of those conditions. These features include deficiencies of the clavicle and medial portions of the pubic rami in cleidocranial dysostosis and generalized physeal widening and angular deformity in metaphyseal dysostosis. The radiographic differential diagnosis should also include coxa vara produced by avascular necrosis (trauma, infection, or associated with developmental dysplasia of the hip or Legg-Perthes disease), and pathologic bone-associated conditions such as osteogenesis imperfecta, fibrous dysplasia, osteopetrosis, or renal osteodystrophy.

The amount of varus deformity of an affected hip may be quantified on AP radiographs by measuring the neck-shaft angle, the head-shaft angle, or the Hilgenreiner-epiphyseal angle (H-E angle) index as described by Weinstein and colleagues (Fig. 18–3). Because the pathophysiology and rationale for operative treatment imply a sagging or slippage of the femoral epiphysis (and the triangular neck fragment so characteristic of this condition), some authors have found that traditional measurement of the neck-shaft angle does not adequately reflect the amount of deformity, anticipated surgical correction, or prognosis for spontaneous progression or recurrence after surgery in this disorder.[80] Measurement of the angle between the axis of the femoral shaft and a line perpendicular to the base of the femoral epiphysis, the head-shaft angle, more accurately reflects the severity of the deformity and its likely progression or correction.[17,22] Weinstein and colleagues[80] identified the prognostic value of the H-E angle, the angle between Hilgenreiner's line and a line drawn parallel to the physis of the proximal femur. On an AP radiograph of the pelvis with the hips in neutral position, this angle is usually between 0 and 25 degrees (average of 16 degrees, in Weinstein's series of 100 normal hips). Weinstein and associates in a study of 22 patients with coxa vara, found that in hips with an H-E angle greater than 60 degrees the deformity invariably progressed and

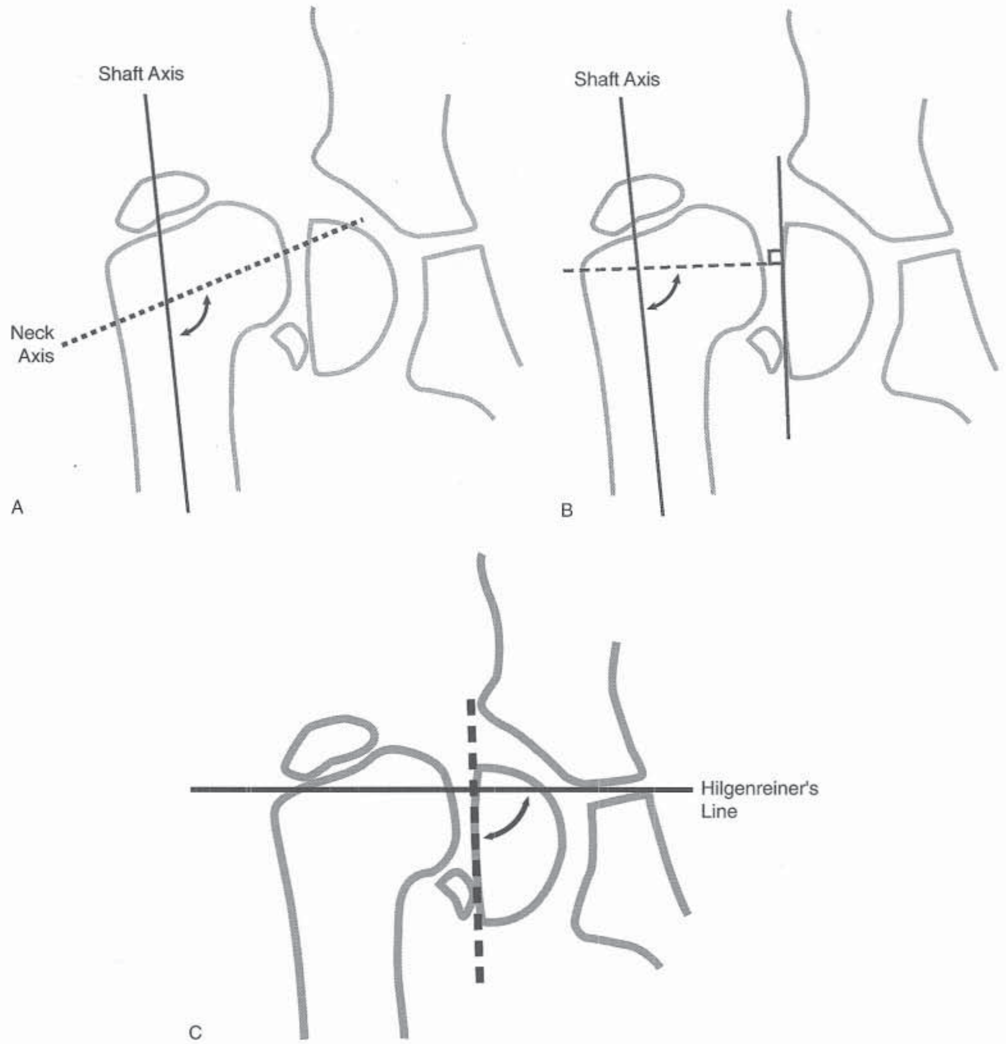

FIGURE 18–3 Quantification of the extent of radiographic deformity of the proximal femur in developmental coxa vara. **A,** The neck-shaft angle is the angle between the axis of the femoral shaft and the axis of the femoral neck. **B,** The head-shaft angle is the angle between the axis of the femoral shaft and a perpendicular line drawn to the base of the capital femoral epiphysis. **C,** The Hilgenreiner-epiphyseal angle is the angle between Hilgenreiner's line and a line drawn parallel to the capital femoral physis.

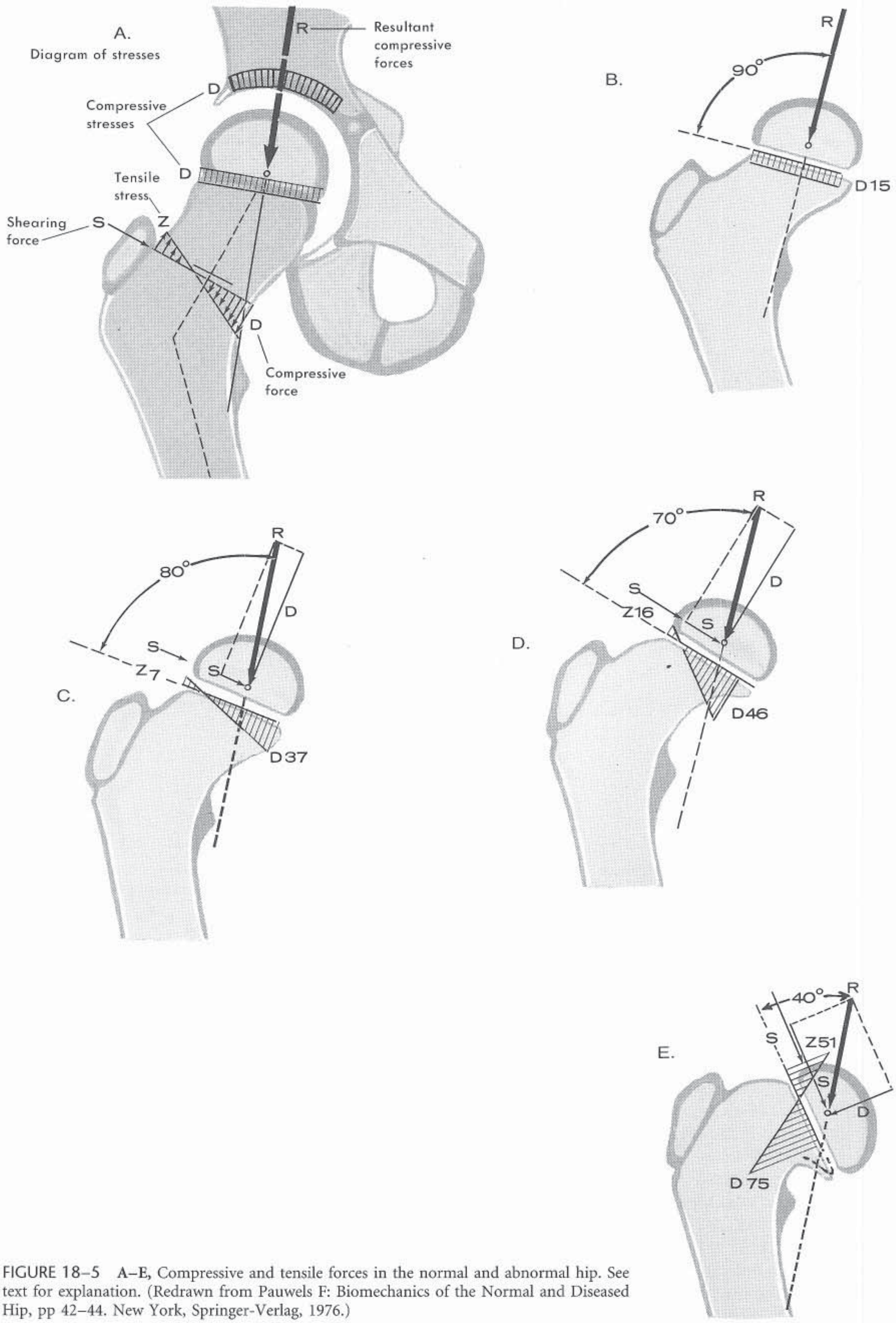

FIGURE 18–5 **A–E,** Compressive and tensile forces in the normal and abnormal hip. See text for explanation. (Redrawn from Pauwels F: Biomechanics of the Normal and Diseased Hip, pp 42–44. New York, Springer-Verlag, 1976.)

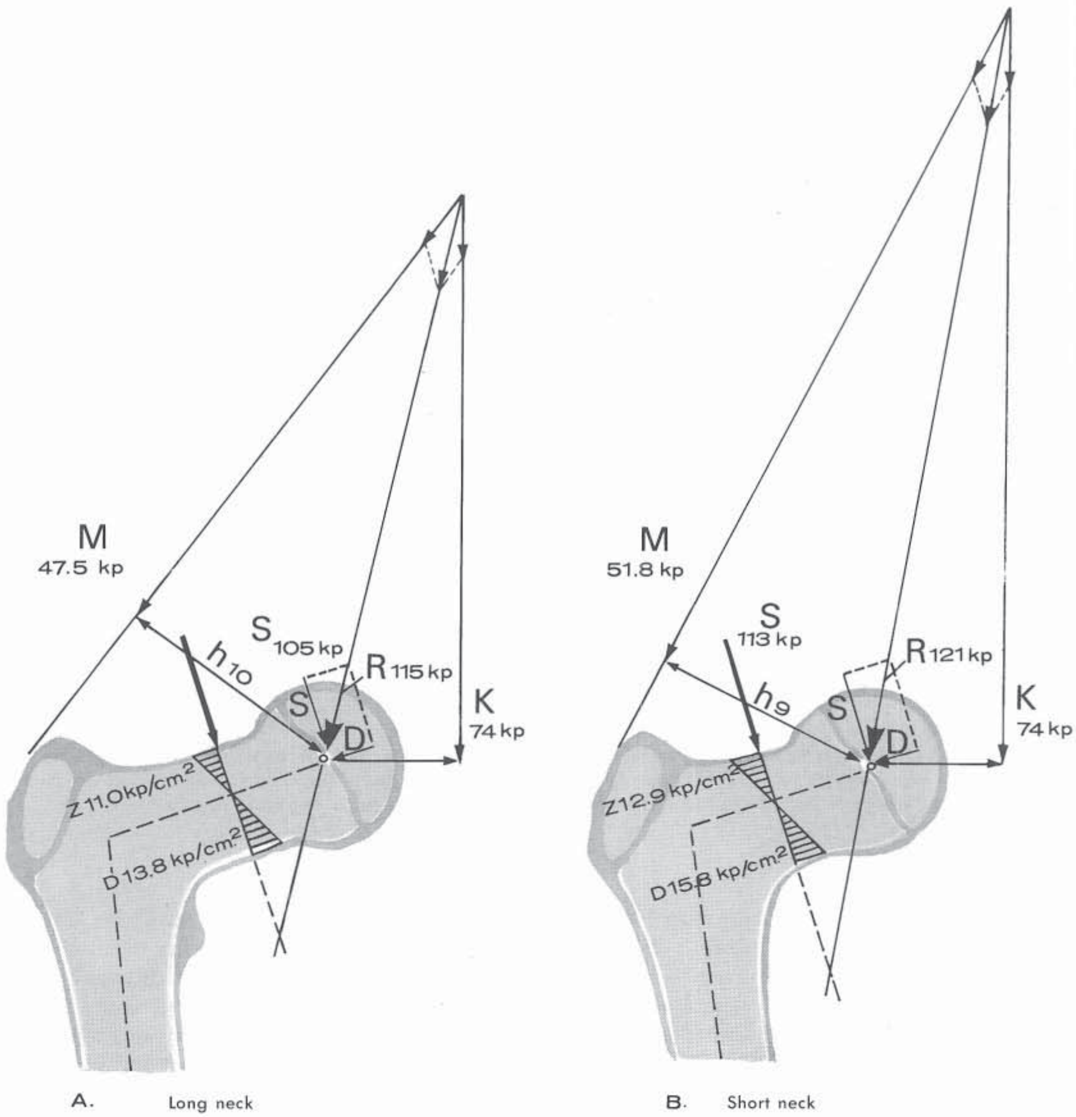

FIGURE 18–7 The effect of the length of the femoral neck on the magnitude of mechanical stressing of the upper end of the femur. **A,** Long neck. **B,** Short neck. See text for explanation.

primary determinants of the need for surgical correction of the deformity. Valgus osteotomy of some form is recommended in hips with an H-E angle of 60 degrees or greater, will not usually be required in patients with an angle less than 45 degrees, and may or may not be required in patients with angles between 45 and 59 degrees. The last group of patients must be observed for evidence of progression of deformity with serial radiography. In addition, patients with symptomatic limp, Trendelenburg gait, or progressive deformity merit valgus osteotomy.

Patients with an H-E angle less than 45 degrees who are asymptomatic need only be assessed for limb length inequality (in unilateral cases) and for evidence of skeletal dysplasia. Periodic radiographic assessment for evidence of progression of the deformity, the development of symptoms, and evidence of limb length inequality is prudent in skeletally immature patients. Limb length inequality is typically minor and can be treated by observation, shoe lift, or contralateral epiphysiodesis as indicated on an individual basis (see Chapter 23, Limb Length Discrepancy). Patients with an H-E angle between 45 and 59 degrees will require this assessment and management as well. Patients in this group who develop hip symptoms or progressive deformity will require surgical treatment.

A great number of surgical treatments have been recommended for developmental coxa vara over the years. Many are of historical interest only, as valgus osteotomy has been demonstrated to be the only appropriate definitive surgical management of this disorder. Langenskiöld and Salenius,[56] Mau,[61] and Pylkkänen[68] performed epiphysiodesis of the greater trochanter in an attempt to modify the growth pattern of the upper femur. Langenskiöld and Salenius,[56] however, concluded that the results of greater trochanteric epiphysiodesis in coxa vara were unreliable, and ultimately recommended against this procedure as the sole surgical treatment for this condition. Greater trochanteric epiphysiradiographic control a guide wire is inserted into the center of the superior half of the femoral neck parallel to its upper border. The guide pin is used as a landmark while the blade of a blade plate of appropriate size with an angle of 140 degrees is inserted into the neck. The blade should be parallel to the long axis of the femoral neck. Predrilling a slot will facilitate insertion of the blade. Next, an intertrochanteric transverse osteotomy is made under radiographic control. The level of osteotomy should be at a distance of 2 to 2.5 cm (the diameter of the femoral shaft at that level) distal to the angle of the blade. The lateral surface of the proximal fragment is roughened. The head and neck of the femur are adducted by using the blade as a lever, and the femoral shaft is abducted. The lateral cortex of the upper fragment is thus approximated to the upper end of the lower fragment. Adductor tenotomy or muscle release may be necessary to facilitate correction of the deformity. The plate of the blade plate is fixed to the shaft with screws. The guide pin is removed. The wound is closed, and a one-and-one-half-hip spica cast is applied. The cast is removed when the osteotomy has healed. An alternative form of internal fixation suggested by Wagner is performed with a bifurcated plate driven through the intramedullary surface of the proximal fragment and secured to the distal fragment with screws (Fig. 18–9). A variation of this technique, and one I prefer, is to perform the osteotomy slightly more proximally after insertion of an appropriate-sized screw of a dynamic compression hip screw device, insert the lateral distal edge of the proximal fragment into the medullary canal of the distal fragment, and secure the distal fragment to the plate portion of the device (Fig. 18–10).

A more complex osteotomy is the cuneiform Y-shaped intertrochanteric osteotomy as described by Pauwels.[66] The objectives of the intertrochanteric Y-osteotomy are to place the capital femoral physis perpendicular to the resultant compressive force and to decrease the bending stress in the femoral neck, as described previously. Medial displacement of the upper end of the femoral shaft widens the femoral neck, shifts the compressive force within the core of the cross section of the neck to eliminate the tension stresses caused by bending, and "supports" the medial neck with the upwardly displaced medial portion of the distal fragment. The operation must be executed precisely to achieve the objectives as determined from preoperative drawings from the radiographs.

The operation is diagramed as shown in Figure 18–11. On a transparent paper placed on the radiograph, draw the hip joint, the physis, and the axis of the shaft of the femur. Draw a horizontal line (*H*, parallel to Hilgenreiner's line) to transect the axis of the femoral shaft 4 to 6 cm below the lesser trochanter. Next, draw a line (*Ps*, corresponding to the epiphyseal line) through the physis and extend it inferiorly until it intersects the horizontal line *H*. From the point of intersection of the lines *H* and *Ps*, draw a third line inclined upward 16 degrees from the horizontal (this angle corresponds to the average H-E angle in the normal hip, and at this inclination, the forces across the physis are presumably purely compressive). The angle formed between the third line (inclined upward) and the line *Ps* is the size of the wedge to be resected (50 degrees in this example) (Fig. 18–11A). Next, draw the upper line of the intertrochanteric osteotomy to extend from the base of the greater trochanter medially to transect the capital physis at the zone of resorption in the femoral neck. Then draw the wedge to be resected from a point *x*. The point *x* is selected so that the length of the medial portion of the upper end of the distal femoral fragment (*A*) equals the length of the base of the femoral neck fragment (*B*)(Fig. 18–11B).

Superimpose a new sheet of transparent paper on the first and trace the inferior fragment of the osteotomy with its axis (Fig. 18–11C). Rotate the superimposed tracing sheet with the distal fragment until the osteotomy lines of the two fragments coincide, to simulate removal of the wedge and closing of the osteotomy site. Then trace the upper fragment. The two axes of the femoral shaft should form an angle of 50 degrees (Fig. 18–11D).

Rotate the upper tracing sheet back to the neutral position for the hip, slide it upward parallel to the femoral axis until the femoral head lies in the acetabular socket of the original sheet, and trace the acetabulum (Fig. 18–11E). This represents the desired radiographic appearance after the osteotomy.

The cuneiform Y-shaped intertrochanteric osteotomy of Pauwels is described in Plate 18–1. This procedure is technically demanding, and I have had limited experience with it. Pauwels described using a loop of wire as a tension band to connect the lateral aspects of the proximal and distal fragments for internal fixation, but fixation with a contoured plate or compression screw-plate is probably more desirable.

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PLATE 18–1. Pauwels' Intertrochanteric Y-Osteotomy