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Congenital Coxa Vara: Computed Tomographic Analysis of Femoral Retroversion and the Triangular Metaphyseal Fragment

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Summary: Three patients with congenital coxa vara studied with two- and three-dimensional computed tomographic (2DCT and 3DCT) methods are reported. In all cases, the femoral retroversion was documented and subsequently corrected by proximal femoral osteotomy. In two patients with isolated coxa vara, the physeal-femoral neck angle was decreased as seen in slipped capital femoral epiphysis in adolescents. Our studies suggest that the triangular metaphyseal fragment reflects a Salter-Harris type II separation pattern through the defective femoral neck. The epiphysis and attached triangular fragment slip from the normal superoanterior portion of the neck in an inferior-posterior direction. The treating surgeon should be aware of the often marked femoral retroversion component present in severe congenital coxa vara. This knowledge allows surgical planning for corrective osteotomies that will better normalize hip mechanics. A combination of marked valgus and flexion with internal rotation of the distal fragment are required to fully correct the deformity. **Key Words:** Congenital coxa vara—Femoral head retroversion—Triangular metaphyseal fragment.

Gradual inferoposterior movement of the capital femoral epiphysis, physis, and attached triangular fragment of metaphysis in congenital coxa vara can lead to severe varus deformity associated with femoral retroversion. However, the rather marked degree of femoral retroversion typically present in congenital coxa vara has not been adequately described.

Theories to explain the formation of the triangular femoral neck fragment in congenital or developmental coxa vara have been proposed by many investigators and include a separate ossification center (8), vascular insufficiency (5,18), osteochondritic lesion (2), and insufficiency fracture (3,14).

We studied three cases of congenital coxa vara (one with isolated congenital coxa vara, one with associated cleidocranial dysostosis, and one with congenital short femur and coxa vara) using two- and three-dimensional computed tomographic (2DCT and 3DCT) methods. All patients demonstrated greatly increased femoral retroversion.

The purpose of this report is to clearly define the proximal femoral anatomy in congenital coxa vara using new imaging techniques. Clarification of both degree of varus and the often poorly recognized associated femoral retroversion allows more accurate and biomechanically correct surgical correction of the condition.

CASE REPORTS

Case 1

A 5-year-old girl presented with multiple congenital anomalies, including Pierre-Robin syndrome and a cleft palate. She also had an altered gait. A pelvic radiograph demonstrated a marked coxa vara with a classic inverted Y-shaped triangular fragment in the femoral neck. The Hilgenreiner epiphyseal angle was 73° on the right and 67° on the left (Fig. 1A).

Physical examination demonstrated flexion of both hips to 140°, abduction 30°/45°, external rotation 45°/70°, and internal rotation 15°/30°. She also had a flexion contracture of 20° on the right and 10° on the left. A gait study in the Motion Analysis Laboratory demonstrated external rotation of the limbs and a Trendelenburg gait with increased hip adduction bilaterally.

A 3DCT image of the right hip (Fig. 1B) demonstrated a marked coxa vara with a chronic fracture line through the growth plate of the capital femoral physis that connected with the line in the inferior femoral neck that formed the metaphyseal triangle. The bony fragment of the metaphysis (Thurston-Holland segment) accompany-

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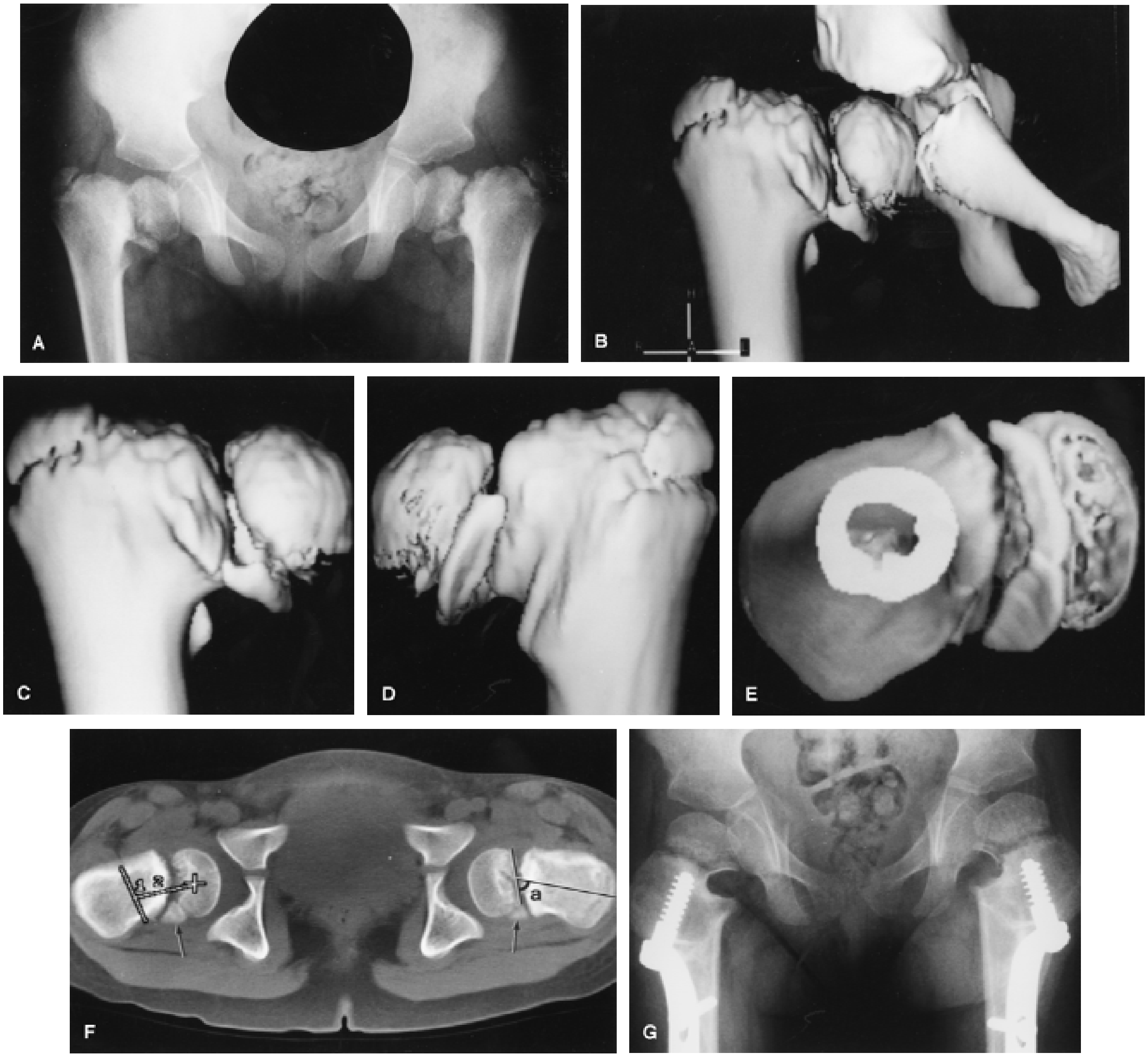


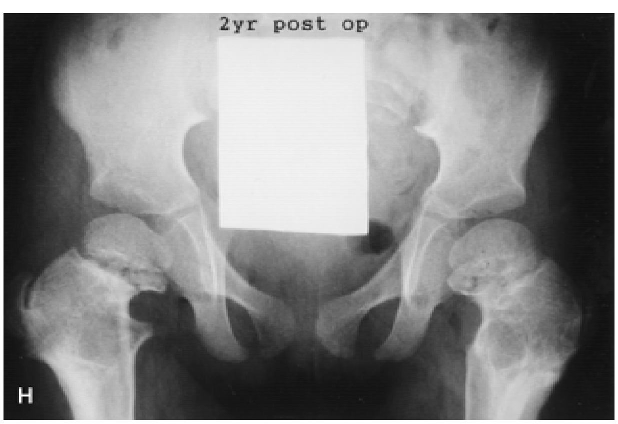


FIG. 1. A: Plain radiograph of a 5-year-old girl demonstrates bilateral coxa vara with a triangular metaphyseal fragment. **B:** Anteroposterior 3DCT image of the right hip demonstrates in greater detail the nature of the coxa vara and its relationship with the acetabulum. **C–E:** Detailed views of the proximal femur with the acetabulum subtracted viewed from anteriorly **(C)**, posteriorly **(D)**, and below **(E)** demonstrate clearly the nature of the metaphyseal fragment. The overhang noted on the inferior aspect of the neck suggests a chronic inferiorly slipping Salter-Harris type II fracture through the capital femoral physis. **F:** 2DCT image demonstrates a posteriorly located triangular metaphyseal fragments (arrows). The physeal neck–femoral neck angle is decreased (angle a). This indicates posterior slipping of the capital femoral epiphysis along with the metaphyseal fragment. **G:** Anteroposterior pelvis radiograph taken 6 months after corrective valgus, internal rotation osteotomy of the proximal femur. The physis is returned to a more normal position. **H:** Anteroposterior pelvis radiograph taken 2 years after corrective osteotomies (patient now age 6 years 10 months).

ing the displaced epiphyseal growth center suggested a Salter-Harris type II separation pattern.

3DCT images of the right proximal femur viewed from anteriorly (Fig. 1C), posteriorly (Fig. 1D), and below (Fig. 1E) demonstrated more clearly the nature of the metaphyseal fragment. The triangular metaphyseal fragments on the plain radiograph (Fig. 1A) and 2DCT view (Fig. 1F indicates a posteriorly located triangular metaphyseal fragment) correspond to the overhang noted on the inferior aspect of the neck on the 3DCT images.

On the 3DCT images, the anterior portion of the metaphyseal fragment was less visible than that seen in the posterior view. This finding coincided with the 2DCT image (Fig. 1F) that demonstrated only a posteriorly located metaphyseal fragment in certain cuts. We believe that this finding confirms an inferoposterior rotatory slipping of the capital femoral epiphysis along with the physeal plate and associated triangular metaphyseal fragment.

The physeal neck-femoral neck angle on 2DCT images (Fig. 1F) was decreased, demonstrating posterior displacement of the capital femoral epiphysis. The angle of femoral anteversion was +2° on the right side and −5° on the left side (5° retroversion). Surgical treatment included bilateral valgus-internal rotational osteotomies and adductor lengthenings. The radiographs performed at 6 months (Fig. 1G) and 2 years (Fig. 1H) postoperatively demonstrated correction of the Hilgenreiner-epiphyseal angle to 36° and correction of the neck-shaft angle to 150° in both hips.

Case 2

A 5-year-old boy with cleidocranial dysostosis presented with a waddling gait and bilateral coxa vara (Fig. 2A). His mother, who had the same condition, had undergone three operations on each hip in childhood. The patient was treated with bilateral proximal femoral valgus-flexion osteotomies plus pinning of the physes using a custom smooth-tipped pin to allow continued physeal growth. Early postoperative films demonstrated a neck-shaft angle of 130° (right) and 155° (left) (Fig. 2B).

Four years postoperatively, radiographs (Fig. 2C) demonstrated a neck-shaft angle of 120° on the right and 125° on the left. The femoral head was growing off the tip of the physeal pins on both sides. The neck-shaft angle was reduced as compared to the 6-month postoperative radiograph. He had a mild abductor lurch bilaterally and external rotation of 85° bilaterally with only 15° internal rotation. His foot progression angle was 30°/30°.

A 3DCT image of right hip viewed from the front (Fig. 2D) with the acetabulum subtracted demonstrated posterior slipping of the capital femoral epiphysis. The angle of femoral anteversion on 2DCT images was right, −10° (10° retroversion) and left, −20° (20° retroversion). The calculation of retroversion is dependent on whether one considers the anatomical neck-head axis in alignment with the original head position or the slipped position (Fig. 2E). The physeal neck-femoral neck angle was decreased, suggesting posterior slipping of the capital femoral epiphysis. He was further treated with repeat bilateral valgus-flexion-internal rotational osteotomies (Fig. 2F).

With further growth, the neck-shaft angle again returned to varus, and at age 11 years an additional proximal femoral valgus osteotomy was performed on the right side (Fig. 2G). At age 13 years, he required a proximal valgus osteotomy of the left femur. At age 16+5 years, the physes were noted to be closed in both proximal femora and the neck-shaft angle was well maintained (Fig. 2H).

Case 3

A 5-year-old girl presented with the Pierre-Robin syndrome and bilateral coxa vara with slightly short femora and radial head dislocation. A radiograph of the hips (Fig. 3A) demonstrated severe bilateral coxa vara with poor acetabular development. The left femoral head was dislocated superiorly. Both femoral heads were conically deformed and the femoral shafts demonstrated angulation and cortical thickening. We diagnosed these femoral abnormalities as Hamanishi type III congenital short femur with coxa vara (right side, type IIIa; left side, type IIIb).

The 3DCT images of the right hip viewed from the front (Fig. 3B) and back (Fig. 3C) and the acetabulum (lateral view [subtracted femur] Fig. 3D) demonstrated a marked coxa vara associated with global deficiency of the acetabulum. The angle of femoral anteversion on 2DCT images was −50° (right, 50° retroversion [Fig. 3E]). The left side also demonstrated −20° femoral anteversion (20° retroversion). The physeal neck–femoral neck angle on 2DCT images (Fig. 3E) was not as decreased in this case compared to that seen in isolated congenital coxa vara. An inferior view of the 3DCT image of right hip combined with the 2DCT image of the distal femoral condyles more clearly demonstrated the torsional relationship between the proximal femur and the distal femoral condyles (Fig. 3F).

DISCUSSION

The definition of congenital coxa vara includes a decrease of the angle between the neck and shaft of the femur in the presence of a primary neck defect. In congenital coxa vara and slipped capital epiphysis (adolescent coxa vara), the physis cannot withstand the applied mechanical shear forces for a variety of reasons (6,12). These include a metabolic defect causing abnormal composition of the physis (11), excessive stress in sport (7,16), and endocrine or connective tissue disorders (4). It is our opinion, as has been reported by other investigators, that in congenital coxa vara the varus deformity occurs as result of a defect in enchondral ossification (17,18) and that shearing forces through this weakened area produce a triangular fragment (14) in a pattern similar to that seen in a Salter-Harris type II fracture.

Controversy continues concerning the plane of the physeal movement in congenital coxa vara. It has been suggested that the slip occurs in only one plane and that

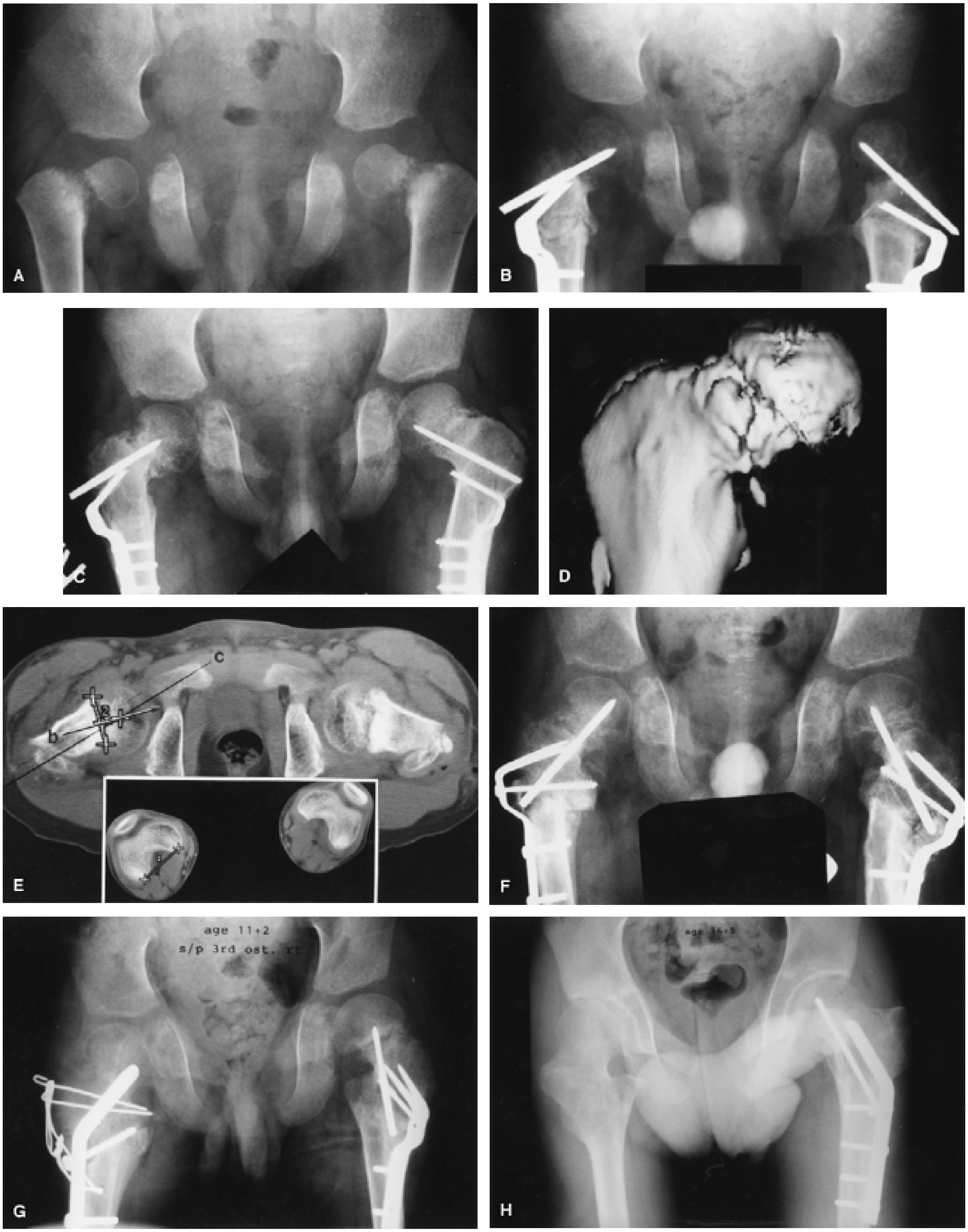
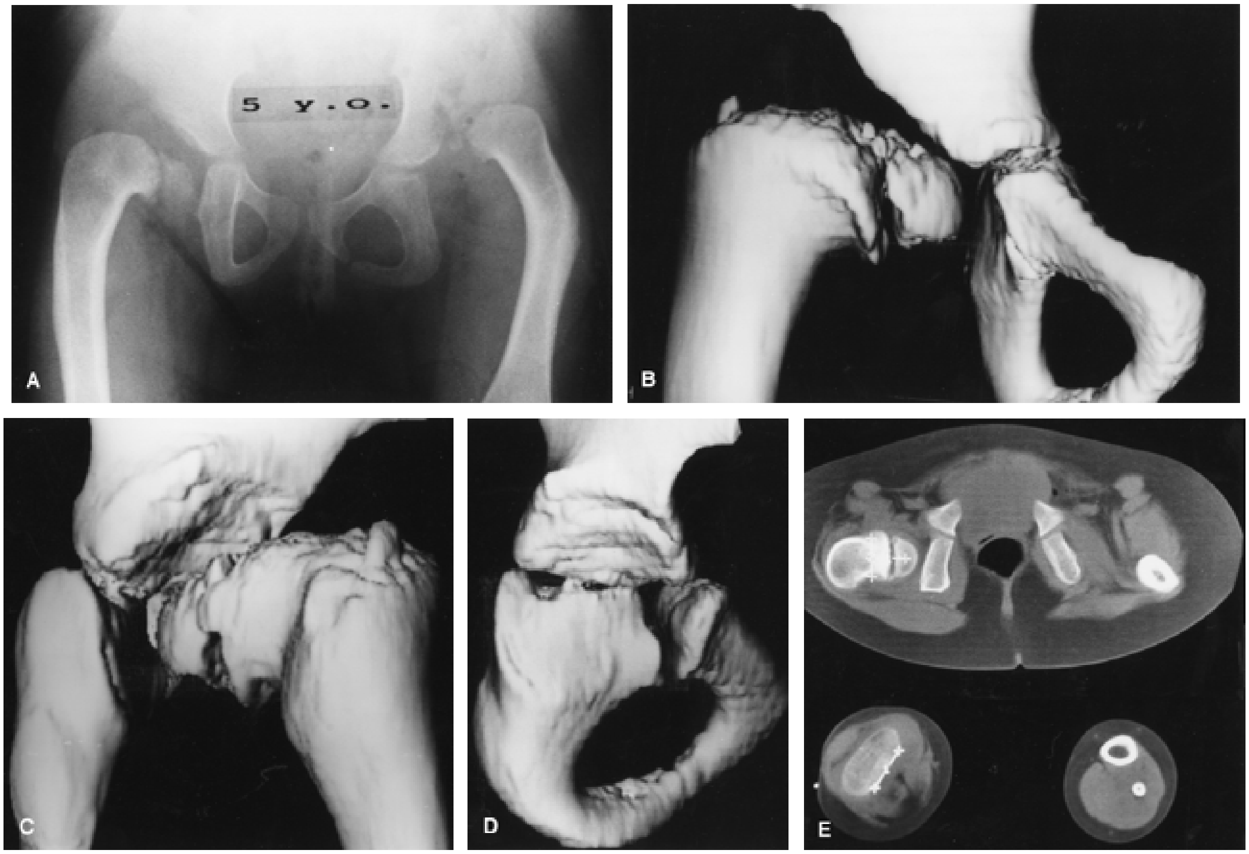


FIG. 2. A: Plain radiograph of a 5-year-old boy demonstrates bilateral coxa vara. The film also demonstrates the markedly widened symphysis pubis characteristic of cleidocranial dysostosis. **B:** Radiograph taken 6 months after bilateral valgus osteotomies with pinning of the physes using a custom smooth-tipped pin to avoid physeal closure. **C:** Radiograph taken 4 years after the operation demonstrates recurrence of coxa vara. The neck-shaft angle is reduced from 130°/155° to 120°/125°. **D:** 3DCT image of the right proximal femur viewed from anteriorly suggests posterior slipping of the capital femoral epiphysis. **E:** 2DCT image of the femoral neck-head and distal femoral condylar axes demonstrates a decreased physeal-femoral neck angle and femoral retroversion on both sides (−10°/−20°). The femoral head is slipping posteriorly (line b: true functional neck-head axis owing to posterior slipping of the epiphysis; line c: anatomical neck-head axis in alignment with original head position). **F:** Radiograph taken 6 months after repeat bilateral valgus-internal rotation osteotomies with pinning of the physes, again using a custom pin (threaded shaft–smooth tip). **G:** Pelvis radiograph taken at age 11 years after a third corrective valgus-internal rotation osteotomy on the right hip. **H:** Pelvis radiograph taken at age 16+5 years after a third valgus osteotomy performed on the left proximal femur at age 13 years. Both hips are well centered and the proximal femoral physes are closed with a satisfactory neck-shaft angle. This sequence clarifies that persistence is required to have good hip structure and function at skeletal maturity.



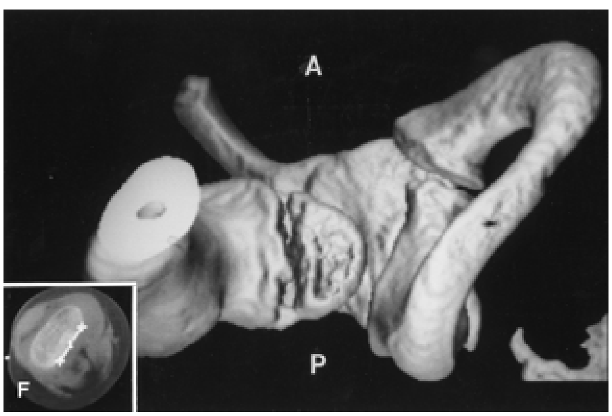


FIG. 3. A: Plain radiograph of a 5-year-old girl demonstrates bilateral coxa vara with dislocation of the left hip. The acetabulae are dysplastic and the proximal femora demonstrate bowing plus cortical thickening. **B:** Anteroposterior 3DCT image of the right hip demonstrates in greater detail the nature of the coxa vara and its relationship with the acetabulum. The femoral head has a conical shape and the superior aspect of the neck is flattened, suggesting that the neck abuts on the acetabular rim when the femur is abducted. **C,D:** Posterior 3DCT view of the same hip in **C** and lateral view of the acetabulum with the proximal femur subtracted **(D)** demonstrate severe acetabular dysplasia. The acetabular dysplasia may have resulted from the long-standing compression force from abutment of the superior neck on the acetabular rim. **E:** 2DCT images (femoral neck-head and distal femoral condylar axis) demonstrate femoral retroversion on the right side (−50°). However, the physeal-femoral neck angle is not decreased. **F:** Composite photograph of a 3DCT image of the proximal femur and acetabulum (inferior view) and a 2DCT image through the transcondylar axis of the distal femur (both are in the same plane) demonstrates a posteriorly bent neck (A, anterior; P, posterior). The retroversion likely results from the bent neck and angulation of the proximal femur. Posterior slipping of the capital femoral epiphysis is not evident.

a valgus osteotomy will make the physis horizontal, correct the shear force, and prevent further slipping (4). However, our three-dimensional studies suggest an inferoposterior movement of the capital femoral epiphysis not only in the coronal plane (varus) but also in the sagittal plane (retroversion).

In slipped capital femoral epiphysis, the slipping is usually posterior (1). This is because the physis forms an arc in the anteroposterior plane with the epiphysis moving posteriorly as it is forced to follow the contour of the physeal arc (9). Three-dimensional study confirms that the displacement actually occurs in three planes. The extended metaphysis produces an extended epiphysis in the sagittal plane, the externally rotated metaphysis produces a retroverted epiphysis in the horizontal plane, and the craniolaterally displaced metaphysis produces a varus epiphysis in the coronal plane (4). The same explanation can be applied to congenital coxa vara in which abnormal shearing forces produce the fissure defect of the femoral neck (triangular fragment), which represents an insufficiency fracture (3,14).

In cases 1 and 2 with isolated congenital coxa vara, the physeal neck–femoral neck angle on 2DCT images is decreased, which demonstrates posterior displacement of capital femoral epiphysis. This finding is identical to that seen in a slipped capital femoral epiphysis in adolescents.

In hip disorders of children with open physes, especially in disorders such as congenital coxa vara and slipped capital femoral epiphysis, the nature of the three-dimensional deformity must be clearly understood to allow appropriate surgical correction. Traditionally these disorders have been treated surgically with relatively simplistic uniplanar correction (valgus osteotomy). For correction of congenital coxa vara or a severe slipped capital femoral epiphysis, theoretically intertrochanteric osteotomy (which corrects coxa vara and associated retroversion at the same time) (13) is a more ideal osteotomy. However, controversy remains concerning the varus deformity in slipped capital epiphysis. Valgus correction may not be necessary in chronic severe slipped capital epiphysis because the varus deformity may be an apparent one (4,9,15). However, many surgeons still use a valgus correction via intertrochanteric osteotomy in severe slipping of epiphysis to correct varus deformity. In coxa vara, internal rotation is more clearly an essential component of corrective osteotomy.

Characteristic differences between isolated congenital coxa vara and congenital short femur with coxa vara have been described. Hamanishi (10) noted that one of these differences is the presence of a short, horizontal neck with a triangular fragment in isolated congenital coxa vara. Congenital short femur with coxa vara is characterized by a retroverted, bent neck.

In our limited series, all cases demonstrated considerable retroversion and were therefore treated with valgus-flexion-internal rotation osteotomy with the distal femur rotated internally. The physeal-femoral neck angle was decreased in cases 1 and 2, but not in case 3. These findings suggest that inferoposterior slipping of the capital femoral epiphysis is a major cause of femoral retroversion in isolated coxa vara. Valgus osteotomy without correction of the retroversion may cause some recurrence of the deformity postoperatively owing to the remaining sagittal tilting of the epiphysis. However, in our case of coxa vara with a short femur, the torsional-bending deformity of the neck and shaft rather than posterior displacement of capital femoral epiphysis seemed to be the major cause of femoral retroversion.

CONCLUSION

In congenital coxa vara in young children, whether isolated or associated with bowing or shortening of the femur, the femoral head moves into an inferoposterior position with or without slipping of capital femoral epiphysis. 2DCT and 3DCT studies clearly detail the complex anatomy of the hip deformity including femoral retroversion. The orthopedic surgeon should be aware of the femoral retroversion component of severe congenital coxa vara when planning surgical correction. A valgus-flexion-internal rotation will most reasonably correct the abnormal hip mechanics. Even with this type of osteotomy, patients may require repeat osteotomies as the varus and retroversion may recur with growth.

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