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Mirror-Image Coxa Vara in Identical Twins*

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Many theories have been put forth to explain the development of congenital coxa vara. Some of the more popular concepts that have been expressed are that coxa vara occurs secondary to: (1) a developmental error⁵; (2) a kind of juvenile osteochondrosis resembling Legg-Calvé-Perthes disease²; (3) a pathological ossification of the femoral neck¹¹; or (4) an embryonic interference with the blood supply to the femoral neck¹⁰. Familial occurrences of congenital coxa vara have been reported^{1,4,11}.

The occurrence of coxa vara in identical twins with mirror-image localization is here reported along with genetic investigations.



FIG. 1-A

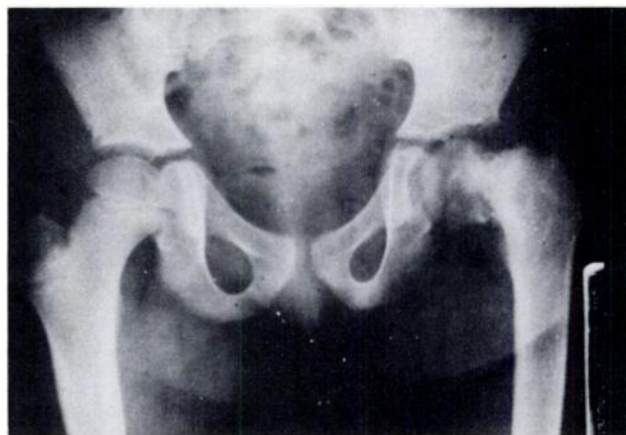


FIG. 1-B

Figs. 1-A and 1-B: Roentgenograms of the hips illustrating mirror-image coxa vara occurring in identical twin girls. Fig. 1-A is patient I.C.; Fig. 1-B is patient E.C.

Case Report

I.C. and E.C., identical twin girls, were born one month prematurely weighing 1,979 grams and 2,097.5 grams, respectively. The delivery was normal and the mother had taken no drugs during the pregnancy. There were two other siblings in the family, four years and three years older than the twins. There was no family history of coxa vara.

Investigation of a limp noted in both children revealed coxa vara affecting the left hip in E.C. and the right hip in I.C. This was a mirror-image coxa vara deformity (Figs. 1-A and 1-B). Both girls later underwent a valgus osteotomy which improved their varus deformities as well as their gait.

I.C. and E.C. were of identical height, weight, features, complexion, eye color, and texture and color of hair. The features were compared for assessment of resemblance, assisted by studies of photographs. The ridge counts were obtained and virtual identity established, because the digits of I.C.'s left hand had a total ridge count of seventy-seven and those on the

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right hand, eighty, for a total of 157; the counts on E.C. were seventy-seven and seventy-eight on the digits of the left and right hands, respectively, for a total of 155. Based on Nixon's discriminant function for the diagnosis of zygosity, a probability estimate of 0.9695 for monozygosity of E.C. and I.C. was obtained⁸.

Laboratory Investigations

Chromosome studies using G-banding (Giesma) and reverse R-banding revealed identical karyotype in both twins. Based on thirteen red cell antigen systems surveyed, complete identity was shown between the two co-twins. No discrepancy could be detected in plasma protein phenotypes between the two girls. Both starch gel electrophoresis and immunoelectrophoresis were utilized in these studies.

Panorex views of both jaws of I.C. and E.C. failed to reveal any disparity.

Comment

The use of all the genetic markers whereby the identities of I.C. and E.C. were explored enabled us to establish their homozygosity.

The presence of identical coxa vara deformity on opposite sides of a pair of ostensibly monozygotic twins strongly suggests that the deformity, or an antecedent local cause for it, occurred early in fetal life or even in the genetic material of the conceptus. The monozygosity amply demonstrated by extensive and detailed investigations in the twins presented possibly points to a genetic etiology of coxa vara deformity.

NOTE: The surgical management of these children was supervised by Dr. David McQueen, Children's Centre, Winnipeg.

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