CONGENITAL DISLOCATION OF THE HIP IN IDENTICAL TWINS

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As early as 1678 Paré stated that lame people beget lame children, and in 1882 Krönlein compiled the family trees of many generations of children with congenital dislocation of the hip. In series of patients with congenital hip dislocation a hereditary factor was noted in 20.2 per cent by Isigkeit (1928), in 22.7 per cent by Scagletti (1932) and in 6.1 per cent by Francillon (1937). Mercer (1950) stressed the importance of the hereditary influence.

A different emphasis is given by those who believe that the essential cause of the dislocation is intra-uterine pressure from oligohydramnios with unusual transverse lie, breech presentation and prolonged abnormality of position of the femoral head in relation to the pelvis during intra-uterine life. The particular relation of this to anteversion of the upper end of the femur was discussed by Krida (1928), Le Damany (1908), Badgley (1943) and Somerville (1953b). Lorenz and Reiner (1905) ascribed an important role in the etiology to hereditary influences, but at the same time tried to develop a mechanical theory.

It was expected that a study of twin children might yield a decisive contribution, and this was in fact possible in the remarkable series reported by Idelberger (1951), who himself examined 138 living identical twins with congenital dislocation of the hip. Before discussing his conclusions we report an example of congenital dislocation of the hip in Italian twin sisters who came north to Switzerland with the influx of Italian workers.

CASE REPORT

Maria and Giacomina B. were born in 1932 in the neighbourhood of Brescia in Northern Italy. They were identical twins as shown by the identical pheno-type, the genotypical identity of the ABO and Rhesus systems, and the Kell, NN and Duffy blood factors which were present or absent in like quantities. The twins had found difficulty in learning to walk but were first seen by a doctor at the age of three years. They limped but had no pain. Even now at the age of twenty-five years Maria has no pain and cleverly conceals her limp; but Giacomina developed pain in the dislocated hip after an appendix operation and has recently been treated by subtrochanteric abduction osteotomy.

Figures 1 to 4 show the clinical and radiographic appearances. These are mirrored almost exactly in the two girls. Giacomina has congenital dislocation of the right hip with a false joint above the acetabulum, a stress fracture of the right pubic bone and a left-sided convergent strabismus. Maria has a congenital dislocation of the left hip with a false joint, stress fracture of the right pubic bone and right-sided strabismus concomitans convergens.

DISCUSSION

These mirrored defects in twins are well recognised as favouring a vitiurn primac formationis, as demonstrated for example in Speemann's string experiments on salamander eggs, in which the resultant identical twins showed bilateral symmetry in the 50 per cent occurrence of situs inversus. Ophthalmologists report that when strabismus occurs in identical twins it affects opposite eyes in 70 per cent. We cannot explain the fact that the fatigue fractures occurred in the right pubic bone in each child. Their occurrence on the same side
does not suggest a direct hereditary influence; but on the other hand if the fractures developed from simple mechanical stress it seems likely that they should both have developed on the side of the normal hip with its greater weight bearing, or both on the side of the dislocated hip with its porotic bones.

Congenital dislocation of the hip in twins has been reported by Hale (1926, one case in identical twins), Hooff (1928, one case in identical twins), Nitsche and Arkmknecht (1933, one case in identical twins and one in non-identical twins) and Ihlenfeldt (1948, one case in identical twins). Idelberger (1951) reviewed the reports of 22,004 congenital dislocations of the hip and found among them 236 pairs of twin children. He personally examined 138 living identical twins with congenital dislocation of the hip. From his investigations he showed that there was no increased incidence of this congenital disorder in twin children; in fact the incidence tended to be less in twins than in the average population. A similar conclusion was reached by Hass (1951). Clearly such intra-uterine constriction or pressure as may arise in twin pregnancy has no decisive influence in the occurrence of congenital dislocation of the hip as has been asserted by the supporters of mechanical theories of etiology. Furthermore, there was no greater incidence of breech presentation or other abnormal position at birth in Idelberger’s twins than is the average with twin births. The frequency of other malformations corresponded in these twins to that recorded in single babies. Of the identical twins with congenitally dislocated hips 42.7 per cent were concordant—that is, both twins had a dislocated hip; whereas the concordance in binovular twins is only 2.8 per cent.*

The proof that mechanical influences of the twin pregnancy do not play a decisive part in the occurrence of dislocation of the hip, and the great concordance in identical twins, show that hereditary factors are of prime importance in congenital dislocation. The hereditary process seems, according to Idelberger, to be dependent on sex, and to be an irregular dominant and of varying penetration. In Idelberger’s large series of identical twins there were many cases in which the correspondence extended even to anatomical details of the dysplasia and even to some of the secondary osteoarthritic lesions. However, the fact that congenital dislocation of hip is determined by hereditary factors does not mean that genetic factors alone bring about dislocation. The part played by external factors, such as extension of the hips resulting from upright posture and gait, is quite plausible. It is known that many hips do not dislocate until the child begins to stand up and walk. But these external factors can cause dislocation only if dysplasia of the hip is present. The dysplasia of genetic origin seems to be the prerequisite for the development of typical congenital dislocation of hip. This inherited dysplasia can be present in varying degree: often the dysplasia is so slight that the external causes are not enough to produce dislocation, yet it is sufficiently marked to predispose to osteoarthritic changes at a relatively early age.

The idea that congenital dislocation of the hip develops as an inherited dysplasia of the acetabulum and upper end of the femur, whereby the upright posture cannot be supported, has an influence on our choice of treatment. The inherited dysplasia prejudices every form of treatment, as experience has proved. Reduction of the dislocation alone suffices in a few cases—namely those in which the dysplasia has reached only moderate proportions or is discovered very early. Reduction alone does not correct the disturbance in the mechanics of the joint which is associated with the dysplasia. The aim of treatment therefore consists in restoring functional conditions as nearly normal as possible in the dysplastic joint and in allowing function of the joint as soon as possible. Early use has a favourable effect on dysplasia, whereas prolonged immobility has an unfavourable influence. Even with treatment that fulfils the requirement of restoring early function (Somerville and Scott 1957) there remains a risk that osteoarthritis will develop years later in consequence of the original dysplasia.

* The concordance of congenital dislocation of the hip in identical twins thus exceeds that in the case of congenital club foot. Idelberger found in 242 twin pairs with congenital club feet a concordance of 22.9 per cent in identical twins and of 2.3 per cent in binovular twins.
Fig. 1—Giacomina

Fig. 2—Giacomina.
FIG. 3—Maria

FIG. 4—Maria.
SUMMARY

1. Congenital dislocation of the hip in identical twins is reported.
2. The heredity of congenital dislocation of hip is discussed. Studies in twins show that congenital dislocation of hip is probably a hereditary dysplasia of the acetabulum and upper end of the femur, and that external factors play a less important role.

We wish to thank Dr. A. Haessig, of the Central Blood Laboratory of the Swiss Red Cross, for the blood examinations.

REFERENCES

PARE, Ambroise (1678): In Lorenz and Reiner.