CONGENITAL DISLOCATION OF THE HIP IN EXTRATER UTERINE PREGNANCY

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Only two cases have been reported of congenital dislocation of the hip in infants born after extrauterine pregnancies. We report a further two and discuss the management and the variable outcome. These cases seem to confirm that congenital dislocation of the hip is associated with moulding forces rather than being a teratological abnormality.

Received 29 March 1996; Accepted 25 April 1996

Extraterine pregnancy is rare, with a reported incidence ranging from 1 in 1100 (Du Toit and De Villiers 1961) to 1 in 50 000 pregnancies (Tan, Goon and Wee 1969), depending on race and other factors. The perinatal mortality is 85% to 95%; the maternal mortality rate is about 6% (Beacham et al 1962). Because of the high fetal mortality little has been reported about the neonate, but the deformities observed include facial asymmetry, torticollis, plagiocephaly, joint contractures, and talipes.

In viable extraterine pregnancy, congenital dislocation of the hip (CDH) has been reported only twice (Jarcho 1949; Burke 1951). We describe another two infants with this condition.

CASE REPORTS

Case 1. A female infant was delivered by an emergency laparotomy at 31 weeks’ gestation after ultrasound at 29 weeks had shown a symmetrically small fetus and oligohydramnios. It had developed near, but outside, the fallopian tube, and the infant had a moulded face, talipes equinovarus and a dislocated left hip (Fig. 1a).

Open reduction of the left hip and an acetabuloplasty were performed at the age of 13 months with a plaster spica applied for eight weeks. After this the hip remained stiff, and at four years of age it was virtually ankylosed with a 70° flexion contracture and deformity of the proximal femur secondary to severe avascular necrosis of the femoral head (Fig. 1b). The deformity was later corrected by a subtrochanteric valgus extension femoral osteotomy and an adductor release.

Case 2. A male infant was delivered by laparotomy at 37 weeks’ gestation. The fetus had been free in the peritoneal cavity, with a placenta attached over the iliac vessels. There was marked plagioccephaly, overlapping cranial sutures, multiple limb contractures and a dislocated right hip. The chest was also severely deformed and the lungs were hypoplastic (Figs 2a and 2b).

The hip dislocation was treated by open reduction at 13 months and a subsequent pelvic osteotomy with a good outcome (Fig. 2c).

DISCUSSION

The most recent review of extraterine pregnancy reports an incidence of 1 in 6389 to 1 in 24 000 pregnancies (Golan et al 1985), but there is clearly a higher incidence in the Bantu population of South Africa (Du Toit and De Villiers 1961). It is commoner in non-caucasians and has been reported to be rising in incidence in the UK because of immigration (Fitzgerald and Goldthorp 1968). The wide variation in incidence is due in part to differing definitions, particularly of the stage of pregnancy at which the fetus is considered viable.

Fetal mortality is high and the optimum time for surgical intervention is still uncertain (Johnson 1967; Strafford and Ragan 1977). After 20 weeks’ gestation, only 25% of extraterine pregnancies will be viable, and three-quarters of these will have deformities (Golan et al 1985). Only 50% will survive for more than one week.

Facial asymmetry, plagioccephaly, torticollis, neck webbing and joint contractures are frequently recorded. Both our cases and most others which have been reported had either talipes equinovarus or talipes calcaneovalgus, but these deformities are reported to be easily correctable, although there has been little long-term follow-up. More severe malformations include myelomeningocele (Golan et al 1985), ectromelia, complete absence of the lower limbs (Johnson 1967), microcephaly with cerebral palsy (Tan et al 1969), and absence of the mouth, arms and eyes (Cornell and Lash 1933).

In surviving infants, CDH has been recorded only twice.
One infant had bilateral CDH and bilateral talipes calcaneovalgus (Jarcho 1949); the other was treated at 20 months of age for left CDH (Burke 1951) after delivery by laparotomy (Lesk 1948). In non-surviving fetuses, Tubby (1912) reported bilateral CDH in a five-month pregnancy and unilateral CDH in an infant ten inches long.

The presumed cause of some of the deformities is the pressure effect of oligohydramnios and amniotic bands (Drury 1960). The amniotic membranes must be intact for the fetus to survive, but the reduced volume of amniotic fluid provides little protection from moulding forces inside the peritoneal cavity in the absence of the protected environment provided by the uterus.

CDH after normal intrauterine pregnancy is associated with other postural deformities such as torticollis, plagiocephaly and talipes (Dunn 1976), and has also been reported in association with ectopia vesicae (Thomas and Wilkinson 1989). Dunn (1976) discussed the aetiology of CDH as either a deformation or a malformation, but it seems that in most cases it is a deformation occurring in the later stages of pregnancy in response to moulding forces applied to an already formed joint.

Teratological malformations are more common in extrauterine pregnancies. Coleman (1978) discussed teratological dislocation, describing the hip abnormalities as very advanced at the time of birth and frequently accompanied by other congenital abnormalities. Lack of contact between the femoral head and the acetabulum during early development is thought to be responsible for these gross anatomical changes (Stanisavljevic and Mitchell 1963) and it seems that teratological dislocation occurs early in fetal life and, in severe cases, the joint may never have been located. Because of gross anatomical abnormalities in teratological dislocation, treatment gives poor results.

The outcome varies in the four reported cases of non-teratological CDH in extrauterine pregnancy. Burke (1951) started treatment at 20 months of age and did not report long-term progress. Jarcho (1949) performed open reduction at 18 months and reported satisfactory mental and physical development. Our case 1 has been difficult to manage and has developed avascular necrosis. Case 2, however, has responded well to conventional treatment.

CDH in extrauterine pregnancy seems to be the consequence of severe moulding forces and to have a spectrum
of severity. A ‘teratological’ type of hip dislocation may be due to the much earlier application of similar forces.

The authors wish to thank Mr M. H. M. Harrison, Emeritus Consultant Orthopaedic Surgeon, The Royal Orthopaedic Hospital, Birmingham, for permission to publish details of case 2.

No benefits in any form have been received or will be received from a commercial party related directly or indirectly to the subject of this article.

REFERENCES


