hereditary mechanism in the aetiology of the disorder. Evidence for a genetic component in the aetiology of pyloric stenosis was suggested by Metrakos (whose series of cases were assembled from the literature) who stated that there was a higher concordance rate of the disease in monozygotic twins than in dizygotic pairs of the same sex. In contrast, the study of a consecutive series by MacMahon and McKeown (1955) showed no difference in concordance by zygosity, and an incidence of pyloric stenosis in one or both of twins no different from that in the general population.

Karyotypic abnormality. The twin girls reported in this article have 45,X/46,XX mosaicism. They are phenotypically females and both appear to be normal. Neither show stigmata of the Bonnevie-Ullrich syndrome and it is too early to diagnose gonadal dysgenesis. That both patients have similar karyotypes suggests that twinning occurred after the mosaicism had been established. Benson and King (1964) were the first to report a higher prevalence of pyloric stenosis in patients with gonadal dysgenesis; though the coexistence of these two disorders had previously been reported on two occasions (Keay and Lewis, 1954; Lindsten, 1963).

In 2 of the 4 patients reported by Benson and King (1964) the karyotype was X/XX and in the other 2, 45,X. These authors suggested that the occurrence of pyloric stenosis (which occurs predominantly in males) in subjects with gonadal dysgenesis is related to monosomy-X; the incidence in such patients might be similar to that in males.

The occurrence of pyloric stenosis and X/XX mosaicism in both of twin females supports the hypothesis of Benson and King that there is an aetiological relation between familial pyloric stenosis and the chromosomal anomaly of gonadal dysgenesis.

Studies of the chromosomal pattern in a larger series of patients with pyloric stenosis would be of interest in elucidating this relation. As a corollary, the occurrence of pyloric stenosis in infants with chromosomal anomalies should be investigated.

Summary

A pair of monozygotic twins, both phenotypical females and concordant for congenital hypertrophic pyloric stenosis, are described. Both have X/XX mosaicism. This provides additional evidence for a hereditary factor in the aetiology of congenital hypertrophic pyloric stenosis.

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REFERENCES


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Orbital Blow Out Fracture

Fractures of the orbital floor may occur in association with multiple facial injuries, or in isolation while the orbital rim remains intact. Such isolated injuries may be caused by a blow to the eye from a fist or a tennis ball and present a characteristic picture which has recently been reviewed by Lerman (1970). The condition is rare in childhood.

Case Report

An 8-year-old boy was punched in the left eye after which he was drowsy and vomited. He was admitted to another hospital where plain skull x-rays showed no fracture. There were no external signs of injury around the eye and his symptoms subsided in the course of the next few days and he was discharged. He was subsequently seen at The London Hospital complaining of double vision, and was found to have limitation of upward gaze on the left (Fig. 1) with diplopia plus enophthalmos. Tomograms of the orbit showed the characteristic 'hanging drop' opacity in the left antrum (Fig. 2) produced by herniation of orbital fat and fascia through a fracture of the orbital floor. At operation the inferior rectus muscle was released from the fracture with subsequent improvement in upward gaze. The fracture was small and a graft of bone or Teflon was not required.

Discussion

The floor of the orbit is paper thin and may be damaged by the impact of the globe from an injury which the orbital rim and facial bones can easily withstand. The injury may be slight and leave
Calcification and Loss of Subcutaneous Tissue Following Trauma and Hypothermia

Calcification of areas of fat necrosis in the neonate is a well-recognized condition. Loss of subcutaneous tissue after resorption of the calcium salts appears to be an unusual complication and I have been unable to find a previously recorded case.

Case Report

This baby was admitted to hospital at the age of 7 weeks with a diagnosis of bronchopneumonia. Examination confirmed the diagnosis, but on handling the limbs, hard plaque-like masses were palpable as though the child had a 'coat of armour' within the tissues of the limbs. X-rays confirmed extensive calcification in the subcutaneous tissues of all four limbs (Fig. 1a and b).

The early history was indeed dramatic. The mother was an unmarried 15-year-old girl and though living at home, her parents were unaware that she was pregnant. One winter night she went into labour and, unattended, delivered her baby. He began to cry and in her panic she opened her first floor window and flung him out into the garden. Fortunately that night there was thick snow on the ground, which must have softened his fall. The child lay in the snow for about 2 hours and on being found was admitted to the nearest maternity hospital, where he was thought to be dead, but was fortunately put into an incubator. His temperature was not recorded, being below that of the low reading rectal thermometer. However, he responded to warmth and his weight 12 hours after admission was 2864 g.