changes in the placenta has been recognised for many years, but the precise correlation between the origin of additional haploid complements and these changes has been made only recently: all paternally derived triploids are partial moles whereas a few maternally derived triploids are molar.8 Our observation is not an exception to this general rule. In these conditions it is difficult to implicate the HLA system in the mechanism leading to the development of a partial hydatidiform mole in the presence of two paternal and one maternal set.

The survival or expulsion of an aneuploid fetus seems to depend on various factors, such as the existence of serious malformations which interrupt embryogenesis at an early stage1 or hormonal factors. In chromosomal abnormalities the placenta has varying degrees of defective endocrine function which results in embryonic death.9 Here it should be observed that the development of hydatidiform mole in triploid conceptuses does not change the gestational age of fetal death but favours the retention of the fetus after death.10 Another factor is maternal age, in that it is possible that older mothers are more tolerant of the aneuploid zygote.11 We are thus led to consider the possibility that immunogenetic similarity between the aneuploid fetus and the mother may also favour the continuation of the pregnancy.

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Concordant monozygotic twins with bilateral renal agenesis

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SUMMARY We report the unique observation of monozygotic twins concordant for bilateral renal agenesis.

Case report

Both parents were 25 years of age and in good health. They were unrelated and there was no family history of renal tract abnormalities. The mother’s first pregnancy had resulted in a healthy male infant born by Caesarean section after failed forceps delivery. In this, her second pregnancy, twins were diagnosed by ultrasound at 15 weeks gestation and the decision was taken to deliver them by elective Caesarean section. She remained well throughout an uneventful pregnancy and there was no history of exposure to drugs or other known teratogens. A second ultrasound examination at 20...
weeks failed to detect any abnormality. Following
the spontaneous onset of labour at 36 weeks'
gestation an emergency Caesarean section was
carried out. Both twins were breech presentation
and very little liquor was present. The first twin, a
girl, weighed 2020 g and died after 20 minutes. The
second twin, also a girl, weighed 2050 g and was
stillborn. Both had typical Potter's facies, large
spade-like hands, and talipes equinovarus. Twin 1
had severe flattening of the right side of the skull and
posterior elongation of the occiput. Twin 2 had a
similar deformity affecting the left side of the skull.
The single placenta weighed 530 g and microscopy
confirmed that the membranes were diaminotic and
monochorionic. Amnion nodosum was present.

Necropsy findings were similar in both infants.
There was bilateral renal agenesis with complete
absence of the ureters and bladder. The fallopian
tubes and uterus were hypoplastic. Normal ovaries
and female external genitalia were present. The lungs
were hypoplastic and in twin 1 the right middle lobe
was absent. Naked eye and radiological appearances
suggested frontal craniostenosis but this was not
confirmed histologically. Twin 1 had a bony defect in
the parietal region of the skull. Both twins were
blood group O Rhesus positive.

Discussion

Levin1 in 1952 described uniovular male twins dis-
cordant for bilateral renal agenesis and, reviewing
the literature in 1954, Davidson and Ross2 found
another six discordant twin pairs of unstated
zygosity. A large family study of bilateral renal
agenesis by Carter et al3 included five male index
cases who were twin-born. One co-twin with a form
of unilateral renal agenesis was thought to be
monochorionic. The remaining four co-twins were
unaffected. Two of these were probably monozygotic
and two were of unstated zygosity. A bilateral case
whose monozygotic co-twin had unilateral agenesis
has also been reported by Mauer et al.4 Concordant
monozygotic twins with unilateral renal agenesis
have been described,5 but we are not aware of any
reports of twins discordant for bilateral renal
agenesis. At least 24 families have been described
in which a patient with bilateral renal agenesis has
had a non-twin sib or other near relative with
bilateral or unilateral renal agenesis or total renal
dysplasia.3 6 7 This suggests a genetic component
in the aetiology of bilateral renal agenesis and also
a genetic relationship with unilateral renal agenesis
and total renal dysplasia.

Carter et al3 have pointed out that the recurrence
rate in bilateral renal agenesis is too high to be
explained on the basis of multifactorial inheritance,
unless estimates of the birth frequency are much too
low. Single gene defects may account for some cases
and non-genetic factors may well be involved in
others. Schinzel et al7 reported bilateral renal
agenesis affecting two male infants whose parents
were cousins. However, increased consanguinity is
not usually a feature of this condition and there is
little evidence for autosomal recessive inheritance
unless the renal defect is part of a distinct syndrome.
Kohn and Borns5 described a man with an absent
right kidney whose daughter and first cousin had
bilateral renal agenesis and whose sister had uni-
ilateral renal agenesis. Further evidence in favour of
an autosomal dominant entity with incomplete
penetrance being responsible for at least some cases
of bilateral renal agenesis comes from the finding of
unilateral renal agenesis or hypoplasia in the parents
of index cases screened for renal anomalies by
ultrasound.3

The finding of bilateral renal agenesis in monozy-
gotic twins is most readily explained on a genetic
basis, but identical twins share more than their
genome and exposure to a common environmental
factor during fetal life cannot be excluded.

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case.

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