LETTER TO THE EDITOR

Limb body wall defects the result of trauma?

EDITOR,—A 20 year old woman, pregnant for the second time, had a serious car accident in her fifth week of pregnancy. She developed backache, shin pains, and difficulty in micturition, suggesting pelvic and uterine injury. After an ultrasound scan a termination for gross fetal abnormalities was carried out at 17 weeks of gestation. A post mortem examination showed a growth retarded female fetus weighing 50 g. There were multiple abnormalities, including an extensive defect of the anterior abdominal wall with eversion of the abdominal organs, severe kyphoscoliosis, absence of the umbilical cord, a left diaphragmatic hernia, and underdeveloped legs. Strands of inflamed and calcified amnion adherent to the retroperitoneum were also identified. Absence of the corpus callosum was seen on ultrasonography and confirmed at necropsy.

Limb body wall defects are considered to be part of the spectrum of the early amnion rupture sequence. It has been estimated that about 1% (0.6–1.45%) of all stillborn fetuses die because of early amnion rupture.¹ The varying presentations of this condition, it has

been suggested, are related to the timing of the insult, with injuries sustained at 7 weeks of gestation causing amniotic bands, those at 3 weeks causing amniotic sheets resulting in anencephaly, and those at 5 weeks causing the limb body wall complex.²

The aetiology of this condition, or group of conditions, remains far from clear, however, with different authors offering different hypotheses, including vascular aetiology, an underlying genetic abnormality, or amnion rupture as the primary event.¹ Vascular damage seems to be the mechanism in several animal models of the disease, leading to the disruption of the embryonic tissue because of haemorrhage and ischaemia.³ Although trauma has been suspected as the possible aetiological factor in the production of this condition, an association with trauma has not been shown in most reported cases. On the contrary, most cases seem to occur spontaneously, although isolated case reports describe associations with amniocentesis, intrauterine devices, and hereditary collagen diseases.^{3,4}

The additional abnormality present in this case was agenesis of the corpus callosum. Several craniocerebral abnormalities have been associated with the early amnion rupture sequence, but agenesis of the corpus callosum is not typical. Structures that form the precursors of the corpus callosum become visible from about the sixth week of gestation, and the anterior portion of the corpus callosum is itself visible by 12 weeks. The cause of the agenesis is unknown, although it would be reasonable to assume that this occurs quite early, and that disruption to the

vascular supply of the developing brain might produce such an anomaly.

In this case the timing of the accident and the presence of an addition intracranial abnormality supports the concept that in at least a percentage of cases the limb body wall defect may be the result of trauma.

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- 1 Luebke HJ, Reiser CA, Pauli M. Fetal disruptions: assessments of frequency, heterogeneity, and embryologic mechanisms in a population referred to a community-based stillbirth assessment program. *Am J Med Genet* 1990; 36: 56–72.
- 2 Gilbert-Barnes EP, Opitz JM. Congenital anomalies-malformation syndromes. In: Wigglesworth JS, Singer DB, eds. *Textbook of fetal and perinatal pathology*. Boston: Blackwell Scientific, 1991: 381–427.
- 3 Van Allen MJ, Curry C, Walden CE, Gallagher L, Patten RM. Limb body wall complex. II. Limb and spine defects. *Am J Med Genet* 1987; 28: 549–65.
- 4 Moerman P, Fryns J-P, Vandenberghe K, Lauweryns M. Constrictive amniotic bands, amniotic adhesions and limb body wall complex: discrete disruption sequences with pathogenetic overlap. *Am J Med Genet* 1992; 42: 470–9.

BOOK REVIEWS

Diseases of the Fetus and Newborn. 2nd Edn. Vols 1 & 2. Edited by G B Reed *et al* (Pp 854.) Chapman & Hall Medical, 1995 ISBN 0-412-39 160-0

Fetal medicine is such a rapidly expanding and important area that the ambitious aims of the editors are clearly sensible. Unfortunately, I am not sure that they have achieved these, because in the attempt to cover such a vast canvas they have inevitably lost the necessary detail required by each potential user.

The material is of interest to perinatal pathologists, surgeons, radiologists, neonatologists and obstetricians. The book is beautifully illustrated, mainly with histological specimens, and there are eloquent pathological descriptions throughout, but the text is light on clinical advice. From my own perspective as a practising neonatologist this meant that my attempts to review the book by consulting it regularly over a period of six months led to disappointment: perhaps an obstetrician or perinatal pathologist would get more help than I did.

The neonatal clinical coverage is patchy, with a whole chapter on ECMO but no advice on treatment or investigation of neonatal seizures, very little on RDS and chronic lung disease. Many of the references are very out of date – neural tube defects may have accounted for 15% of perinatal deaths

in 1985, but this was not the case in 1995, and there is no mention of periconceptional folate supplementation. Periventricular leucomalacia did occur in 35% of very low birthweight infants in 1982, but few would report such a high incidence now. There is a large section on the physics of ultrasound and its use in fetal diagnosis, but there are no illustrations of its use postnatally in the discussions of neonatal brain, heart, liver or kidney disease. That is not to say that there are not nuggets of information which would be difficult to find in such a handy form elsewhere. One example is Nick Fisk's clear chapter on oligo- and polyhydramnios and another, Ryan and Whittle's, on hydrops. In general, the emphasis is on fetal disease rather than neonatal disease and if this is indeed the editors' intent then a different title might have helped.

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Prenatal Diagnosis in Obstetric Practice. 2nd edn. Edited by Martin J Whittle and J Michael Connor. (Pp 362, hardback.) Blackwell Science, 1995. ISBN 0-632-03838-1

This is the second edition of a very popular book for obstetricians. While retaining much of the very practical and easy reference style of the first edition, the new one has taken into account the many developments in prenatal diagnosis. It is a book of high quality and considerable usefulness for obstetricians dealing with congenital abnormalities and their attendant risks.

Each section deals with a different area of prenatal diagnosis and its congenital prob-

lems, while maintaining an integrated approach to their management. There are the expected chapters covering such subjects as general assessment, pre-pregnancy counselling, the philosophy and practice of prenatal screening by both biochemical and ultrasound methods, specific problems such as chromosome disorders, haematological disorders and structural abnormalities. In particular, I was most impressed with the chapter by Bryan Beattie, on the termination of pregnancy for fetal abnormality. Although the title suggests a very single minded, single option approach to congenital abnormality, it gives a very balanced view of how to present options to patients, and advice about non-directive counselling in particular. The other excellent feature of the chapter is that there is a section on caring for the carers, an area often overlooked in such texts.

The other section that is exceptionally good and of great practical value is the compendium of prenatally diagnosable diseases, an amalgam of previous appendices, brought together to facilitate reference access.

It seems invidious to make any negative comments and my criticisms are few and minor. For example, the chart used in the chapter on rhesus disease is now out of date. I would also have liked to have seen a section on the ethical issues of whether we should be screening for congenital abnormality, and the implications—both positive and negative—of such programmes for parents, professionals, and the health service. But, overall, I enthusiastically commend this book.

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