

screening. The heterozygous forms are probably not detectable during the first half of pregnancy.

**References**

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Correspondence to Dr P-H Persson, Department of Obstetrics and Gynaecology, University of Lund, General Hospital, S-214 01 Malmö, Sweden.

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## Subependymal cysts in normal neonates

E-Y SHEN AND F-Y HUANG

*Department of Paediatrics, Mackay Memorial Hospital, Taipei, Taiwan*

**SUMMARY** We undertook a prospective ultrasound study of subependymal cyst formation in normal neonates. Twenty five of 500 normal Chinese neonates (5%) were found to have subependymal cysts. Most of these (18 of 25) showed no obvious perinatal insult. This study suggests a rather high incidence of concealed intrauterine subependymal haemorrhage in normal term neonates.

Subependymal pseudocysts in a neonate suggest an intrauterine pathology. Larroche<sup>1</sup> described these cystic formations, mainly located within the remnant zone germinativa or close to it, and considered that antenatal insults accounted for the cellular destruction in the remnant matrix. A germinal matrix haemorrhage may also develop into a subependymal cyst.<sup>2 3</sup>

In this study, 25 of 500 (5%) normal neonates were found to have subependymal cysts soon after their birth. These neonates were all healthy at birth and during scanning.

We suggest that subependymal haemorrhage may develop in term as well as preterm fetuses at the same gestational age and cause these cystic formations at birth.

**Materials and methods**

Between June and August 1984, 500 of 1701 (29.4%) normal neonates in this hospital were scanned after obtaining informed parental consent. There were no differences in social class or economic status between the parents who gave or refused their consent. There were 265 boys and 235 girls, whose ages ranged from 2 to 9 days.

A commercially available, portable real time sector scanner with 5 MHz rotatory head transducer and a 80° field of vision was used at the bedside according to a standard method.<sup>3</sup>

Physical examination is performed routinely on every neonate at the age 2 or 3 days in this nursery. If a neonate was noted to have subependymal cyst, he or she was then assessed again physically and neurologically soon after scanning.

Patients were asked to come back for repeat ultrasound scans monthly until resolution of cysts.

**Results**

Subependymal cysts were discovered in 25 (5%) of 500 neonates, 12 (4.5%) of whom were boys and 13 (5.5%) girls. Their gestational age ranged from 37 weeks to 43 weeks, mean 39.2 weeks (Fig. 1). Birthweight ranged from 2800 g to 4000 g, mean 3302 g and occipitofrontal diameter ranged from 31 cm to 37 cm, mean 33.6 cm.

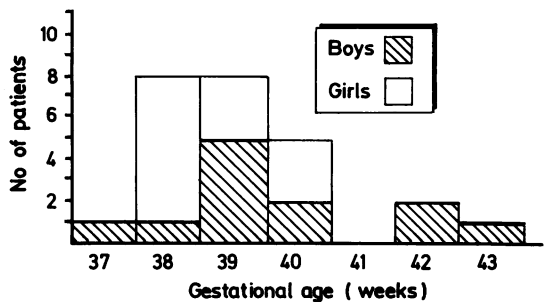


Fig. 1 Gestational age distribution in 25 neonates with subependymal pseudocysts.

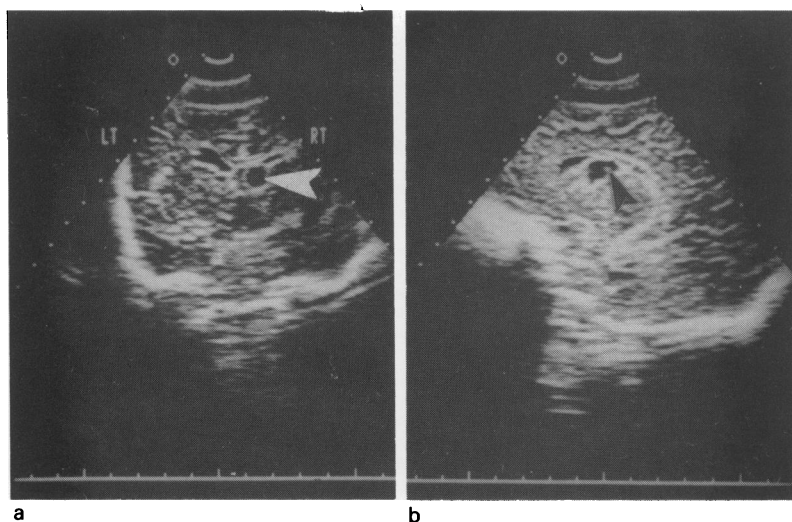


Fig. 2 Subependymal cyst at the caudothalamic notch: (a) coronal view, (b) parasagittal view.

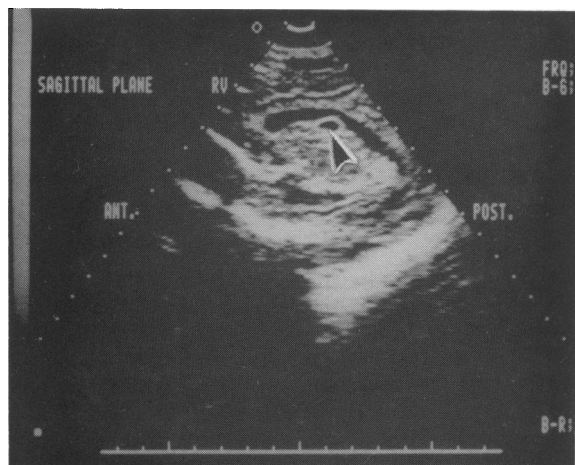


Fig. 3 Subependymal cyst at the inferolateral margin of the lateral ventricles in the area above the thalamus.

The neonates with subependymal cysts were all normal physically, and had normal primitive reflexes. Most had a full Apgar score at 1 minute; one had an Apgar score of 7 at 1 minute and 9 at 5 minutes and three had a score 9 at 1 minute.

The maternal history showed that three (12%) had had a viral infection during pregnancy, two (8%) had had threatened abortions, one (4%) had placenta praevia, and one had had frequent fainting spells during third trimester. The other 18 (72%) pregnancies were normal.

The subependymal cysts were noted in the caudothalamic notch in 15 infants (Fig. 2), at the level of foramen of Monro in one, and at the inferolateral margin of the lateral ventricle in the area above the thalamus in nine (Fig. 3). The cysts were located in the right ventricle in three infants, in the left ventricle in 10 and bilaterally in 12. The size of the cysts ranged from 2 mm to 7 mm in diameter as measured by a caliper mark on the monitor screen.

Twenty two infants underwent ultrasound follow up monthly. Their physical and motor development were all normal up to the last follow up. In 13 the cysts resolved in two months, in seven after three months, and in two by four months. Three infants were lost to follow up.

### Discussion

There are two pathogeneses in pseudocyst formation, focal cerebral destruction<sup>1</sup> and old haemorrhage.<sup>2,3</sup> Larroche described these honeycomb-like cavities lined with immature, undifferentiated cells.<sup>1</sup> Burstein described a thick-walled cystic lesion at the site of the germinal matrix haemorrhage on necropsy.<sup>2</sup> Sauerbrei found a small subependymal cyst which developed several weeks after a previous subependymal haemorrhage.<sup>3</sup>

The incidence of subependymal and intraventricular haemorrhage in infants with a birthweight of less than 1500 g ranged from 43% to 55%.<sup>4,5</sup> The subependymal haemorrhage may eventually progress to cystic formation.<sup>2-4,6</sup> Levene reported two cases of subependymal haemorrhage with subse-

quent pseudocyst formation at 29 days and 10 weeks of age.<sup>6</sup>

The obstetric history was normal in 18 (72%) of the infants with cyst formation in this study. It is unlikely that any noxious cellular agent caused destruction of subependymal cell population in utero in all of these. An hypothetical explanation is that concealed subependymal haemorrhage caused by intrauterine hypoxia or another unknown factor may occur several weeks before delivery and result in cyst formation.

Neurological and physical examination of these neonates were within normal limits during the neonatal period as well as during their monthly follow up. Although the cysts were reabsorbed in all cases followed, it takes time to predict the integrity of the infant's high cortical function and the normality of psychomotor development.

In summary, subependymal cyst in normal neonates may suggest intrauterine subependymal haemorrhage. Its aetiology needs further investigation. Various races and communities with different qualities of prenatal care may have a different

incidence of cyst formation. We look forward to further reports for comparison.

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Correspondence to Dr E-Y Shen, Department of Paediatrics, Mackay Memorial Hospital, No. 92, Sec. 2, Chung-San North Road, Taipei, Taiwan 104, ROC.

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## Marfan's syndrome presenting as an intrapartum death

R BUCHANAN AND G P WYATT

*Departments of Pathology and Paediatrics, St Mary's Hospital, Portsmouth*

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**SUMMARY** A rare case of Marfan's syndrome presenting as an intrapartum death is described. Recognisable mitral valve lesions were present.

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Marfan's syndrome is a generalised connective tissue disorder, and cardiovascular symptoms result from a loss in tensile strength of the supporting tissue of the aorta and cardiac valves. With more sophisticated diagnostic procedures it is realised that cardiovascular defects occur in virtually all patients with this syndrome,<sup>1</sup> but they do not usually become manifest before the second decade. They are rare in childhood but have been described in infants, and very rarely, recognisable defects are present at birth.

#### Case presentation

This baby was the second child of a 25 year old mother. The antenatal period was uneventful apart from the lie, which was a breech. The mother's pelvis was adequate. She was admitted, in labour, at

39 weeks and under a pudendal block an assisted breech delivery was performed. Low forceps were used and the head was delivered with no undue delay or force.

The fetal heart had been heard during labour but on delivery the apex beat was not felt, although the baby gasped once. Intensive resuscitation was unsuccessful.

The infant was a girl weighing 2800 g. Her length crown to rump was 34 cm, and crown to heel 50 cm. Her head circumference was 35 cm, and foot length 8.5 cm. The feet were long and narrow and the hands were long with tapering fingers (Fig. 1). The palate was not high arched, there were no flexion deformities of the limbs, and no signs of injury to the head or scalp were seen.

A small pericardial effusion was present. The heart was enlarged due to right and left ventricular dilatation. The great vessels were macroscopically normal but the tricuspid and mitral valve rings were dilated, the cusps of both valves were voluminous and pale (Fig. 2).

The pulmonary and aortic valves were normal. There was free blood over the surface of the brain,