Lesson of the Week

Apparent microcephaly caused by a bicornuate uterus

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Because a reduced biparietal diameter in a fetus during pregnancy may be due to a misshapen uterus, advice to parents regarding prognosis should be carefully considered

Microcephaly is usually defined as a head circumference less than two standard deviations below the mean for age and sex. Intellect in microcephalic children is strongly correlated with head circumference,[1] and those with an appreciably reduced measurement usually have a poor prognosis. Microcephaly at birth can be secondary to intrauterine infection, drugs, or radiation or may be part of many chromosomal and malformation syndromes.[2] True microcephaly can be inherited as an autosomal recessive disorder. Since autosomal dominant forms of microcephaly have recently been reported[3] parental head circumferences should be taken into account when a child is assessed.[4] While in most cases microcephaly is due to an underlying defect of brain development and growth, the possibility of external constraint on a normal brain must also be considered. Though craniostenosis may cause an apparent microcephaly by limiting skull growth in an anteroposterior or lateral plane, the cranial volume may still be normal owing to expansion of the normal brain in other directions. We report a case of cranial constraint in a fetus with apparent microcephaly in utero and at birth.

Case report

Both parents were white, non-consanguineous, and aged 27. The mother was a primigravida and gave no history during early pregnancy of ingestion of drugs or alcohol, viral infection, or exposure to radiation. She had had episodes of vaginal bleeding throughout the pregnancy. Though at 11 weeks the combination of bleeding and a small uterus suggested a missed abortion, an ultrasound scan confirmed the presence of a live fetus. At 16 weeks an ultrasound scan showed a slightly small fetal biparietal diameter. Bleeding stopped at 20 weeks, but the mother reported appreciably less fetal movement. At 33 weeks the biparietal diameter was consistent with 27 weeks' gestation. A more detailed real-time scan showed a biparietal diameter of 75 mm, a head circumference of 26·2 cm (both equivalent to 27 weeks), and an abdominal circumference of 30·6 cm (equivalent to 34 weeks). When she asked about the result of the scan the mother was told that the baby had a very small head and that the prognosis for intelligence and survival was poor.

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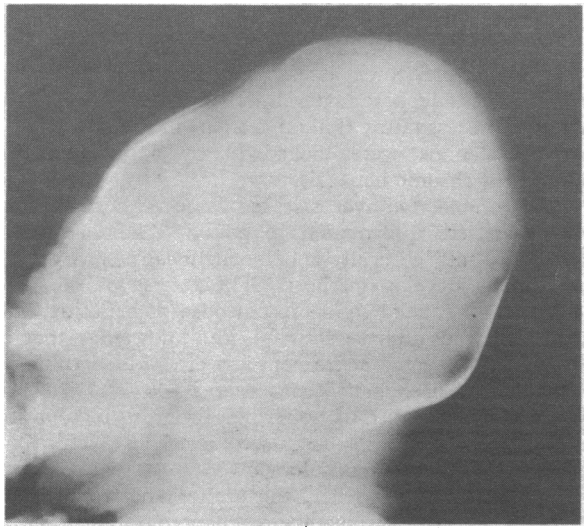
She was induced at 37 weeks, and an assisted breech delivery was performed. The baby, a boy, was born with Apgar scores of 1 at five and 5 at nine minutes and breathed spontaneously. Neither parent wanted to see the baby, but eventually the mother turned to the baby but remained unwilling to handle him. The baby appeared reasonably well, was not grossly malformed, and had no defects except for an abnormally sloping forehead and apparent microcephaly with a head circumference of 29·5 cm. The birth weight was 266 g, gestational age 37 weeks, and results of neurological examination were normal. The skull x ray films showed a small cranial vault, no craniostenosis, no convolutional markings (figure), and no intracranial calcification. Serological investigations for cytomegalovirus, toxoplasmosis, and rubella and Epstein-Barr virus were negative. Primary microcephaly was diagnosed, though the head shape was unusual for this condition and some degree of distortion was apparent, suggesting abnormal intrauterine compression.

A paediatrician saw the mother on several occasions and she and the baby were discharged when the head circumference was still well below the third percentile. The parents failed to keep two follow up appointments and sought fostering for the baby. The foster mother found the baby difficult to feed and at the age of 3 weeks he was admitted to another hospital. At this time, though the baby's head circumference was small, the shape of the skull was deformed with a receding forehead and pointed apex. Possible intrauterine constraint was considered. Computed tomography, an electroencephalogram, electroretinogram, and visual evoked response and evoked response audiograms were carried out; none showed any abnormality.

Meanwhile, the parents received genetic counselling three weeks after the delivery. A detailed history indicated that the fetus had always remained in the same position with the head under the mother's right costal margin. Movements had been very slight and only on the left side. This raised the possibility of intrauterine constraint due to a bicornuate or septate uterus. The mother was referred back to her obstetrician for a hysterosalpingogram, which confirmed the presence of a bicornuate uterus, the right horn being larger than the left. Radiographs of the baby taken at birth (figure) showed an unusually shaped skull with extremely hypoplastic supraorbital ridges, a flattened occipital bone, and an abnormal protrusion at the vertex. A radiograph taken before delivery confirmed that the head was in the right upper quadrant and appeared compressed, presumably in the apex of the right uterine horn. The hands and feet appeared to be compressed into the left uterine horn.

The head circumference increased from below the third percentile at birth to the tenth percentile at 7 weeks and to the

fiftieth percentile at 9 weeks. So far development has been normal for age and gestation. The increase in head circumference has been due not only to the normal increase in volume but also to a change in the shape of the skull. The parents were told that the small head size during pregnancy was due to moulding by the abnormally shaped uterus and that the normal head size at 9 weeks, together with the explanation for the intrauterine measurements, indicated a good prognosis. They were persuaded to take the child home, and since then there have been no apparent problems.



Neonatal anteroposterior radiograph of the skull.

Comment

Unusual findings on a routine ultrasound scan should not automatically be interpreted as indicating a rare malformation of the fetus. For example, by definition about 2% of infants will have a head circumference two standard deviations below the mean for gestational age. Primary microcephaly is nevertheless rare, with incidences ranging from 1 in 25 000[5] to 1 in 250 000.[6] In the absence of any obvious cause for a small biparietal diameter—for example, intrauterine infection—a diagnosis of primary microcephaly must be considered in relation to other possible causes. Gross uterine malformations are reported in 1 in 1000 pregnant women at term.[7] Compression of the fetal head by a malformed uterus may therefore be at least as common a cause for a reduced biparietal diameter as primary microcephaly.

Any prognosis based on abnormal ultrasound findings should be very carefully considered. The clinicians in charge should decide whether or not the parents should be told of possible abnormalities. It is debatable whether they should be told of a poor prognosis at a late stage of pregnancy when intervention is impossible, as this may lead, as in the present case, to rejection of the child at birth. The cranial volume may be calculated from measurements of length, height, and width[8] if there is any doubt about the diagnosis of microcephaly at birth.

The sequence of events in this report throw a fascinating perspective on current views on mother to baby bonding. The mother's expectations in the last months of pregnancy led to her rejection of the baby. After the baby's birth the paediatric care concentrated on trying to persuade the parents to accept and care for a possibly handicapped child. Despite these efforts, the parents had the baby placed in foster care. Once the correct diagnosis became clear, however, they established a successful relationship with their baby.

Though the present case is unusual the increasing use of ultrasound throughout pregnancy may make detection of reduced biparietal diameters due to fetal constraint more common. Constraint on a fetus by a misshapen uterus, fibroids, or oligohydramnios is now well recognised as a cause of specific deformities.[9,10]

References

1 Pryor HB, Thelander H. Abnormally small head size and intellect in children. *J Pediatr* 1968;**73**:593-8.
2 Smith DW. *Recognizable patterns of human malformation.* 3rd ed. Philadelphia: WB Saunders, 1982.
3 Haslam RHA, Smith DW. Autosomal dominant microcephaly. *J Pediatr* 1979;**95**:701-5.
4 Weaver DD, Christian JC. Familial variation of head size and adjustment for parental head circumference. *J Pediatr* 1980;**96**:990-4.
5 Böök JA, Schut JW, Reed SC. A clinical and genetic study of microcephaly. *Am J Ment Defic* 1953;**57**:637-60.
6 Van den Bosch J. Microcephaly in the Netherlands: a clinical and genetical study. *Ann Hum Genet* 1959;**23**:91-116.
7 Blair RG. Pregnancy associated with congenital malformations of the reproductive tract. *Journal of Obstetrics and Gynaecology of the British Empire* 1960;**67**:36-42.
8 Austin JHM, Gooding CA. Roentgenographic measurement of skull size in children. *Pediatr Radiol* 1971;**99**:641-6.
9 Miller ME, Dunn PM, Smith DW. Uterine malformation and fetal deformation. *J Pediatr* 1979;**94**:387.
10 Smith DW. *Recognizable patterns of human deformation.* Philadelphia: WB Saunders, 1981.

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Do women with menorrhagia need iron? The finding by Lewis[1] that women with menorrhagia have an appreciable lowering of iron stores is not surprising, the conclusion that they do not need iron is. Surely the presence or absence of anaemia is not the best criterion for treatment with iron. Is there some danger in replenishing iron stores in women with menorrhagia or is prevention of anaemia in these cases not worth while?

There is no evidence that any benefit is obtained from treating patients with a low serum ferritin concentration unless they are also anaemic.[2] The article referred to in the question[1] showed that there was no significant difference in the haemoglobin concentration between a group of women with subjective menorrhagia and a group with normal menstrual function, and that fewer than 15% of women with heavy menstrual loss were anaemic. Furthermore, there was no direct correlation between the haemoglobin concentration and the number of months that the patients had complained of menorrhagia. This suggests that these women do not require prophylactic iron supplements but should always have their haemoglobin concentration checked and treated if found to be anaemic. Nevertheless, in countries where nutritional sources of iron are poor and intake or absorption of iron is inadequate the risk of iron deficiency anaemia would be present even with menstrual blood loss only moderately above amounts generally considered average.[3] In such circumstances prescribing prophylactic iron supplements may be justified.—G J LEWIS, senior registrar in obstetrics and gynaecology, London.

1 Lewis GJ. Do women with menorrhagia need iron? *Br Med J* 1982;**284**:1158.
2 Anonymous. Serum ferritin. *Lancet* 1979;i:533-4.
3 Rybo G. Clinical and experimental studies on menstrual blood loss. *Acta Obstet Gynecol Scand* 1966;**45**, suppl 7:1-23.

If a baby has its first triple immunisation plus oral polio vaccine at the age of 4 months what is the longest interval that may be allowed without having to start a full course of immunisation again?

There is no need to start the full course again if the normal immunisation schedule is interrupted; the second and third doses should be given as if there had been no interruption.—D P ADDY, consultant paediatrician, Birmingham.