

Three-dimensional sonographic description of abnormal metopic suture in second- and third-trimester fetuses

R. CHAOU^{*}, J. M. LEVAILLANT[†], B. BENOIT[‡], C. FARO[§], P. WEGRZYN[§] and K. H. NICOLAIDES[§]

^{*}Center for Prenatal Diagnosis and Human Genetics, Berlin, Germany, [†]Department of Obstetrics and Gynecology, Armand-Brillard-Hospital, Nogent-sur-Marne, France, [‡]Maternite, Hopital Princesse Grace, Monaco and [§]Harris Birthright Research Centre for Fetal Medicine, King's College Hospital Medical School, London, UK

KEYWORDS: 3D ultrasound; fetal syndromes; frontal bones; metopic suture; transparent maximum mode

ABSTRACT

Objective To describe patterns of abnormal development of the metopic suture in association with fetal malformations during the second and third trimesters of pregnancy.

Methods This was a cross-sectional study of the frontal bones and metopic suture in 11 fetuses at 17–32 weeks of gestation. Cases were selected because there were obvious abnormalities in the metopic sutures. In each case, a malformation was detected by two-dimensional (2D) ultrasound and the abnormality of the metopic suture was detected and evaluated on three-dimensional (3D) ultrasound, using transparent maximum mode.

Results There were essentially four patterns of abnormality in the metopic suture: firstly, delayed development with a V- or Y-shaped open suture, which is found in normal fetuses at 12–16 weeks; secondly, a U-shaped open suture, presumably due to upward growth of the frontal bones with delayed closure; thirdly, premature closure of the suture, which is normally observed after 32 weeks; fourthly, the presence of additional bone between the frontal bones. Premature closure of the suture or additional bone between the frontal bones was observed in fetuses with holoprosencephaly and abnormalities of the corpus callosum, whereas the V-, Y- and U-shaped metopic sutures were observed in fetuses with facial defects involving the orbits, nasal bones, lip, palate and mandible, in the absence of holoprosencephaly and abnormal corpus callosum.

Conclusions This preliminary study describes the pattern of possible abnormalities of the metopic suture and should stimulate further investigation to establish the prevalence and evolution of abnormal sutures as well as the incidence

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INTRODUCTION

Examination of the fetal face constitutes an important component of prenatal sonography, because dysmorphic features in the face are commonly found in association with many chromosomal defects and genetic syndromes. In two-dimensional (2D) ultrasound examination the standard views include the mid-sagittal plane for the detection of frontal bossing, nasal bone hypoplasia and micrognathia and a series of transverse planes for the diagnosis of anophthalmia/microphthalmia, hypotelorism and hypertelorism, and facial cleft.

The advent of three-dimensional (3D) ultrasound has now made it possible to assess reliably skull bones with their sutures^{1–5}. In a previous study we described the normal development of the frontal bones and metopic suture⁵. Ossification of the frontal bones starts at 9 weeks in the middle of each supraorbital region and then spreads medially and laterally, so that by 11 weeks the frontal bones appear as ‘thick eyebrows’⁵. Between 11 and 20 weeks, ossification spreads upwards in a radial fashion. At 11 weeks, the frontal bones reach the midline at the nasal area and subsequently extend superiorly towards the future anterior fontanelle. Similarly, the gap between the two frontal bones starts closing at around 16 weeks in the supranasal region and with advancing gestation the two frontal bones enlarge and converge in the midline as if being zipped together. At 32 weeks there is apparent closure of the metopic suture starting from the glabella and then moving upwards towards the anterior fontanelle.

Correspondence to: Prof. K. H. Nicolaidis, Harris Birthright Research Centre for Fetal Medicine, King's College Hospital Medical School, Denmark Hill, London SE5 8RX, UK (e-mail: fmf@fetalmedicine.com)

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In our centers we have incorporated 3D examination of the fetal face into the routine assessment of fetuses with malformations. In this preliminary study we describe certain patterns of abnormal development of the metopic suture in association with fetal malformations during the second and third trimesters of pregnancy.

METHODS

This was a cross-sectional study of the frontal bones and metopic suture in 11 fetuses at 17–32 weeks of gestation, based on the last menstrual period and confirmed by crown–rump length measurement in early pregnancy. The cases were selected because there were obvious abnormalities in the metopic sutures. In each case, a malformation was detected by 2D ultrasound and the management of the pregnancies, including further investigations and counseling concerning expectant management or pregnancy termination, were not affected by the 3D ultrasound findings.

3D volumes were obtained in the mid-sagittal plane, the transducer being parallel to the direction of the fetal nose. The 3D examinations were carried out transabdominally (RAB 4-8L probe, Voluson 730 Expert, GE Medical Systems, Milwaukee, WI, USA). The sweep angle was selected to include the whole fetal skull. For post-processing the 3D volumes were analyzed using transparent maximum mode with the three orthogonal planes (Voluson 730 Expert Operation Manual). Having selected the A-plane showing the fetal profile, fine rotational adjustments were made to show the frontal bones aligned horizontally with the *x*-axis. The rendering box was then narrowed to include only the fetal face

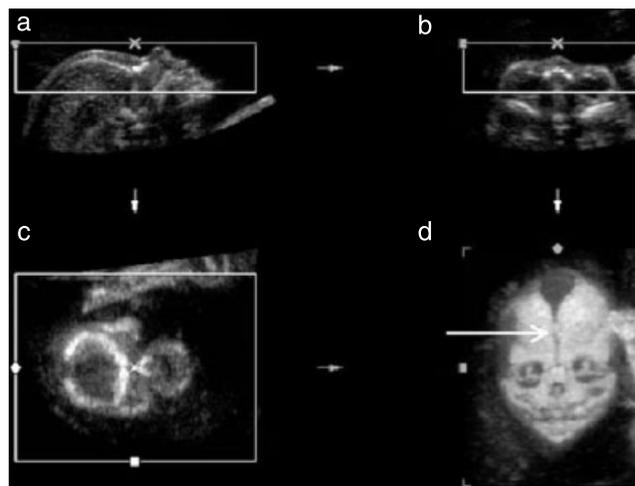


Figure 1 Three-dimensional (3D) volume of the normal fetal face at 22 weeks displayed in orthogonal planes A, B and C (a, b and c, respectively). The rendering box is narrow over the face with the render view direction from ventral (upper horizontal line). To avoid artifacts the forehead is completely in the box. (d) The bony face is rendered in 3D transparent maximum mode, demonstrating the metopic suture (arrow).

(Figure 1). The size of the rendering box varied according to the gestational age, and the image was displayed with a combination of transparent maximum mode and surface texture mode in order to achieve a clear view of the frontal bones and the metopic suture.

RESULTS

2D ultrasound findings, prenatal diagnosis and pregnancy

Table 1 Findings in fetuses with abnormal metopic suture

| Case | Metopic suture | GA (weeks) | Intracranial and facial anomalies | Extracranial anomalies | Outcome | Genetic syndrome/ abnormal karyotype |
|------|-------------------|------------|--|--|-------------|--------------------------------------|
| 1 | V- or Y-shaped | 24 | Dandy–Walker variant, cleft lip and palate | Hepatic arteriovenous fistula, echogenic kidneys | Termination | |
| 2 | V- or Y-shaped | 17 | Hydrocephaly, microphthalmia, cleft lip and palate | Ventricular septal defect, short femur | Termination | |
| 3 | V- or Y-shaped | 22 | Hypoplastic nasal bones | Bowed long bones, hypomineralized vertebral bodies | Termination | Stuve–Wiedemann syndrome |
| 4 | V- or Y-shaped | 22 | Dandy–Walker variant, hypoplastic nasal bones, facial tag | Bladder extrophy, ambiguous genitalia | Termination | |
| 5 | U-shaped | 22 | Coronal craniosynostosis, hypoplastic nasal bones | Bilateral syndactylies | Termination | Apert syndrome |
| 6 | U-shaped | 32 | Dandy–Walker malformation | — | Termination | |
| 7 | U-shaped | 23 | Ventriculomegaly, hypoplastic nasal bones | Tetralogy of Fallot, growth restriction, echogenic bowel | Fetal death | |
| 8 | Premature closure | 17 | Holoprosencephaly, hypertelorism, cleft lip and palate | Tetralogy of Fallot | Termination | |
| 9 | Additional bone | 23 | Absent corpus callosum, cerebellar hypoplasia | Right clubfoot, pre-axial polydactyly | Termination | |
| 10 | Additional bone | 22 | Dandy–Walker malformation, absent corpus callosum, cleft lip and palate, hypertelorism | Pulmonary atresia, ventricular septal defect | Fetal death | Patau syndrome |
| 11 | Additional bone | 28 | Facial cleft from right nostril to eyelids, lipoma of corpus callosum | Unilateral clubfoot | Termination | Goldenhar syndrome |

GA, gestational age.

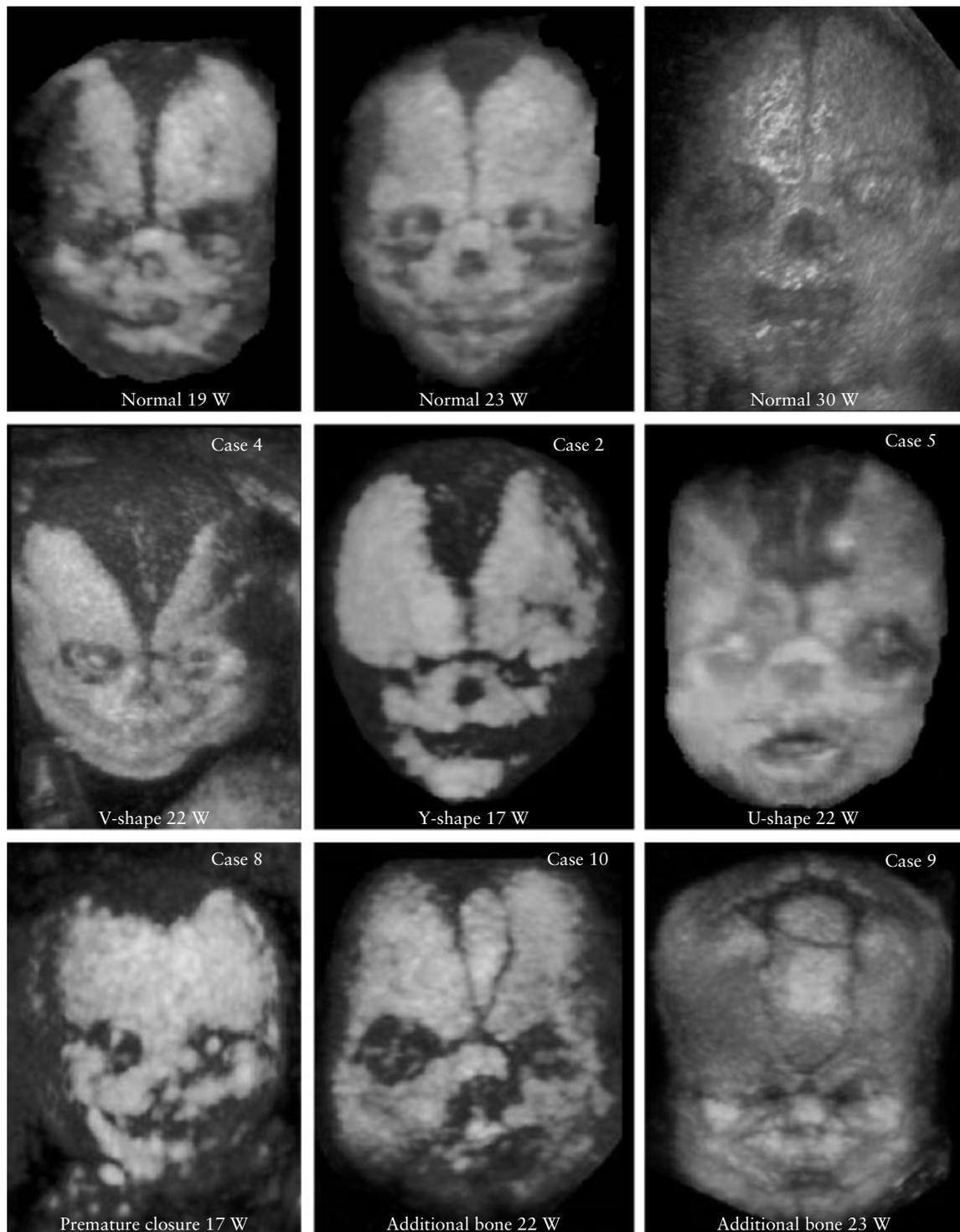


Figure 2 Normal development of the metopic suture (upper three images), delayed development with V-, Y- and U-shaped metopic sutures (middle three images), and accelerated development of the metopic suture with premature closure or additional bones (lower three images). Case numbers correspond to those in Table 1. W, gestational weeks.

outcome of the 11 fetuses with abnormal metopic sutures are summarized in Table 1.

In all cases the metopic suture was grossly abnormal in comparison with healthy fetuses at similar gestational ages (Figure 2)⁵. There were essentially four patterns of abnormality in the metopic suture: firstly, delayed development with a V- or Y-shaped open suture, which is found in normal fetuses at 12–16 weeks⁵; secondly,

U-shaped open suture, presumably due to upward growth of the frontal bones with delayed closure; thirdly, premature closure of the suture, which is normally observed after 32 weeks⁵; fourthly, presence of additional bone between the frontal bones (Figure 2).

There was similarity of associated malformations according to the shape of the metopic suture. Premature closure of the suture or additional bone between the

frontal bones was observed in fetuses with holoprosencephaly and abnormalities of the corpus callosum. In contrast, the V-, Y- and U-shaped metopic sutures were observed in fetuses with facial defects involving the orbits, nasal bones, lip, palate and mandible, in the absence of holoprosencephaly or abnormal corpus callosum.

DISCUSSION

This 3D ultrasound study, with the use of transparent maximum mode, describes abnormal patterns in the development of the metopic suture in association with fetal brain malformations, chromosomal defects and genetic syndromes.

In many of our fetuses with abnormal metopic suture there was an associated brain defect. This finding is not surprising, because anatomical studies have demonstrated that the development of cranial sutures is driven by the development of the brain⁶. The sutures are believed to grow in response to the centrifugal pressure from the expanding brain and proximity of the dura to the suture is critical in maintaining its patency^{7,8}. The dura supplies osteoinductive growth factors and cellular elements, such as osteoblast-like cells, to the overlying osteogenic fronts in the suture, regulating the complex process of growth and fusion of this space^{9–11}. Interestingly, in many of our fetuses the brain abnormality was a midline defect, including holoprosencephaly, abnormal corpus callosum and Dandy–Walker malformation or variant. However, in the case of holoprosencephaly and abnormal corpus callosum, there was premature closure of the suture or additional bone between the frontal bones, whereas with cerebellar abnormalities, there was delayed ossification of the frontal bones. The premature closure of the metopic suture in association with holoprosencephaly, which was previously reported in a postmortem pathological study¹² and was confirmed in a systematic 3D ultrasound investigation of affected fetuses at 11 + 0 to 13 + 6 weeks¹³, may be a consequence of abnormal or incomplete development of the dura.

The V-, Y- and U-shaped metopic sutures, which presumably represent delayed development of the metopic suture, were observed in fetuses with facial defects involving the orbits, nasal bones, lip, palate and mandible, in the absence of holoprosencephaly and abnormalities of the corpus callosum. The development of the facial and frontal bones is related intimately to the migration of neuroectodermal cells from the crest of the neural tube. These cells migrate forwards and downwards towards, firstly, the mandible, secondly, the maxilla, and thirdly, the frontonasal region^{14,15}. Many facial abnormalities are thought to be a consequence of impaired migration or inadequate function of neural crest cells¹⁴.

This is a preliminary study and does not constitute a systematic investigation of the incidence of abnormal metopic suture in various fetal abnormalities. What it does is to describe patterns of possible abnormalities and this should stimulate further investigation to establish the

prevalence and evolution of the abnormal sutures as well as the incidence and pattern of other associated defects. In many aspects of fetal medicine knowledge has been acquired by the sonographic examination of features that have been well described in postnatal medicine. However, in the case of the metopic suture the reverse is likely to be true, because, except in the case of the craniosynostoses, systematic examination of the metopic suture does not currently constitute an integral part of pediatric examination or assessment of genetic syndromes.

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