Supratentorial Abnormalities in the Chiari II Malformation, II

Tectal Morphologic Changes

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Objective. The sonographic diagnosis of fetal myelomeningocele has improved mainly because the diagnostic focus has shifted from observation of spinal manifestations to observation of cranial abnormalities. Intracranial diagnostic criteria rely on abnormalities in the posterior fossa. We describe abnormalities in tectal morphologic characteristics that, although well described on magnetic resonance imaging, have received little attention in the sonographic literature. This study analyzed the frequency of this observation and technical aspects that improve its visualization. ***Methods.*** From a database of obstetric sonograms, we identified all cases of sonographically detected myelomeningocele. The search covered the years 1999 to 2007. We retrospectively reviewed the fetal intracranial findings with special attention to elongation or "beaking" of the tectum. ***Results.*** A total of 89 fetuses were identified. The mean and median gestational ages were 22 weeks 4 days and 22 weeks 2 days, respectively. Of the 89 cases, 59 (66%) had an abnormal tectal shape. The abnormality was seen in 77% of cases judged to be suboptimally visualized and 62% of cases with good visualization. Tectal abnormalities were seen equally well in fetuses before and after 24 weeks. Finally, tectal abnormalities were seen more frequently as the severity of posterior fossa findings increased. ***Conclusions.*** Tectal morphologic alteration is a common supratentorial feature of the Chiari II malformation on prenatal sonography both before and after 24 weeks' gestation. Its frequency increases with the severity of posterior fossa abnormalities. Therefore, it may be useful as a supratentorial indicator of both the presence and, potentially, the severity of the Chiari II malformation. ***Key words:*** Chiari II malformation; fetal sonography; myelomeningocele; tectum shape.

Abbreviations
MRI, magnetic resonance imaging

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The features of the Chiari II malformation that have proven to be most useful and have received the most attention in the sonographic literature are the infratentorial findings. These include effacement of the cisterna magna and deformation of the cerebellum, known as the "banana" sign.[1] Other infratentorial abnormalities are commonly observed postnatally.[2] Although the dominant features of the Chiari II malformation relate to the hindbrain, many supratentorial features have been described.[2,3] Because posterior fossa manifestations of the Chiari II malformation can be obscured in severe cases because of shadowing from the petrous ridges,[1,4] our goal in this series has been to identify the sonographic appearance and frequency of supratentorial findings that can provide the interpreting physician with clues to the diagnosis of the Chiari II malformation in cases in which the posterior fossa is poorly visualized.

In the first article of this series, we described a supratentorial finding that has received scant attention in the sonographic literature, the ventricular "point," a misshapen occipital horn that is pointed rather than rounded.[5] In this article, we address tectal morphologic abnormalities. The normal tectum has a rounded or, sometimes, a squared appearance of the colliculi and a short cranial caudal length (Figure 1). In the Chiari II malformation, variable degrees of fusion of the colliculi and upward deflection of the tectum result in prominent beaking and elongation of the tectum. These abnormalities have been well described on magnetic resonance imaging (MRI) of infants with the Chiari II malformation.[2,6] Our review of the previous literature identified 2 articles that discussed tectal beaking in the Chiari malformation.[7,8] In both, the discussion regarded the Chiari III malformation associated with encephaloceles. We were unable to identify reports in the sonographic literature regarding tectal beaking in the Chiari II malformation.

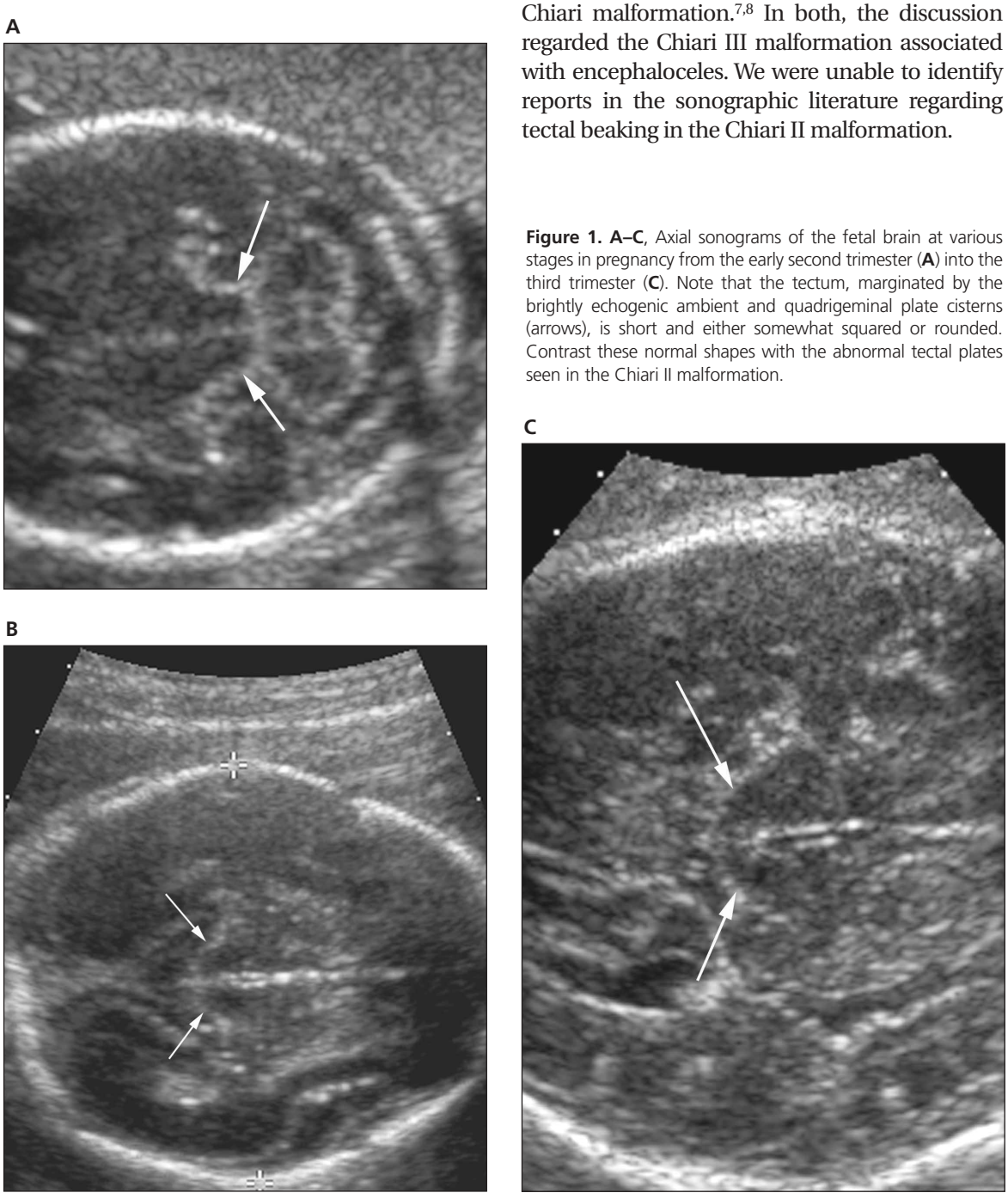


Figure 1. A–C, Axial sonograms of the fetal brain at various stages in pregnancy from the early second trimester (**A**) into the third trimester (**C**). Note that the tectum, marginated by the brightly echogenic ambient and quadrigeminal plate cisterns (arrows), is short and either somewhat squared or rounded. Contrast these normal shapes with the abnormal tectal plates seen in the Chiari II malformation.

Materials and Methods

From a database of obstetric sonograms, we searched for all cases in which a fetal myelomeningocele was sonographically detected. The search covered the years 1999 to 2007. We then retrospectively reviewed the fetal intracranial findings in each case.

Fetal imaging was conducted with 2- to 4-MHz vector and 2- to 6-MHz curved array transducer formats with both selectable focus and frequency as appropriate for the size of the pregnancy (Siemens Medical Solutions, Mountain View, CA). Images included both transverse axial planes of the section of the intracranial anatomy as well as coronal and sagittal planes of the section when feasible. Features of both normal intracranial anatomy and those associated with the Chiari II malformation were evaluated. In particular, an experienced sonologist (R.A.F.) individually evaluated each case to determine the morphologic characteristics of the tectum. Notations were made as to whether the contour was rounded or squared (normal) or "beaked," a feature of the Chiari II malformation.

Other features that were analyzed and recorded included the highest vertebral level of the myelomeningocele, the diameter of the ventricular atrium, the grade of the Chiari II malformation (mild, moderate, or severe), the gestational age of the fetus at the time of the study, whether appropriate images of the tectum were seen only on a digital video clip, and whether there were technical difficulties due to fetal position or patient body habitus. Inward curvature of the frontal bones, known as the "lemon sign," was not evaluated. All statistical comparisons used the Fisher exact test. The Committee on Human Research at the University of California, San Francisco, approved this research project (approval number H7798-31138-01).

Results

A total of 89 fetuses with a myelomeningocele were identified. Fetuses with skin-covered spina bifida lesions were not evaluated. Gestational ages ranged from 17 weeks 1 day to 34 weeks 5 days. The mean gestational age was 22 weeks 4 days, and the median gestational age was 22 weeks 2 days. Because the study was retrospective, no specific effort had been made to assess the shape of the tectum during real-time sonography. Therefore, the quality of the imaging in each case was judged relevant to the ease of assessing the tectal shape. Sixty-three cases were judged to have good visualization, and 26 had suboptimal visualization.

Of the 89 cases, 59 (66%) had a beaked or abnormally elongated tectal shape (Figure 2). No attempt was made to grade the severity of the tectal beaking. In those studies with good visualization, 39 of 63 (62%) had a beaked tectum. Among those studies with suboptimal visualization, 20 of 26 (77%) had a beaked tectum. In 5 of the studies, the beaked tectum was seen only on a digital video clip. In 7 of 89 studies (8%), the tectum could not be visualized retrospectively.

We further divided cases into 2 gestational age ranges and determined the occurrence of the beaked tectum within each group. The 2 groups were 17 weeks 1 day through 23 weeks 6 days and 24 weeks 0 days through 34 weeks 5 days. In the earlier group, a beaked tectum was identified in 46 of 71 cases (65%). In the later group, a beaked tectum was seen in 13 of 18 cases (72%). There was no difference in the likelihood of seeing the abnormality in these 2 groups ($P = .8$)

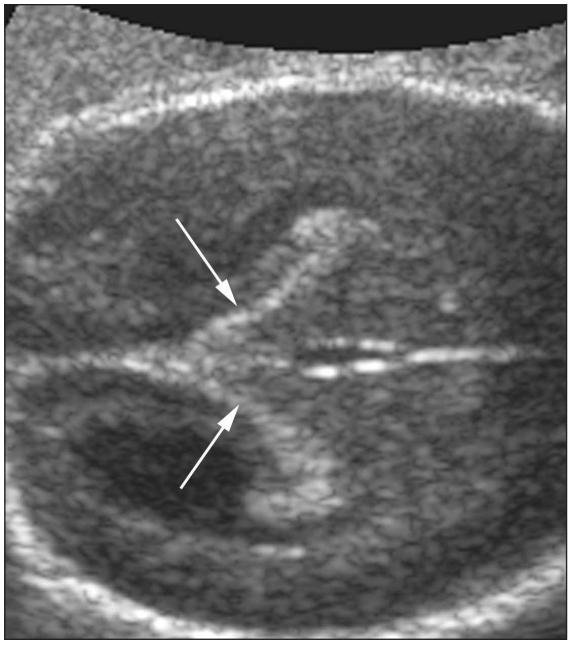
The severity of the posterior fossa manifestations of the Chiari II malformation was categorized as mild, moderate, or severe in each case. Fifty-two cases were severe, 27 were moderate, and 6 were mild. In 4 cases, because of technical factors, the severity of the Chiari malformation could not be adequately assessed. Of the 4 cases in which the severity of the Chiari malformation could not be assessed, all (100%) had a beaked tectum. Of the 6 mild cases, 3 (50%) had a beaked tectum. Of the 27 moderate cases, 19 (70%) had a beaked tectum. Of the 52 severe cases, 46 (88%) had a beaked tectum. The finding was more likely to be observed in cases with severe malformation than with mild malformation ($P = .04$). There was no statistical difference in the likelihood of observing tectal beaking between the mild and moderate groups ($P = .37$) or the moderate and severe groups, although the difference in the latter group approached statistical significance ($P = .063$).

Ventricular size ranged from 0.6 to 3.43 cm. The mean ventricular size was 1.24 cm. Fifty-three cases had ventriculomegaly; 28 had a normal ventricular size; and in 8 of the studies, suboptimal visualization prevented a ventricular measurement from being made. Twenty-two of the 28 fetuses with a normal ventricular size (79%) had a beaked tectum. Of the 53 cases with ventriculomegaly, 36 (68%) had a beaked tectum. These differences were not statistically significant (P = .43). Of the 8 studies in which suboptimal visualization prevented an accurate ventricular measurement from being made, assessment of the tectal shape was still possible. Seven of the 8 (88%) had a beaked tectum.

The most cephalad observed level of a myelomeningocele was at the T9 vertebral level; the most caudad was at the S3 vertebral level. Occurrence of a beaked tectum was not assessed relative to the spinal level of the myelomeningocele.

Figure 2. A–C, Three examples of beaked or, if one prefers, elongated tectal plates (arrows). Note the easily recognized difference in shape when compared with the normal tectal plates seen in Figure 1. The normal tectum appears much shorter and does not taper at the quadrigeminal plate.

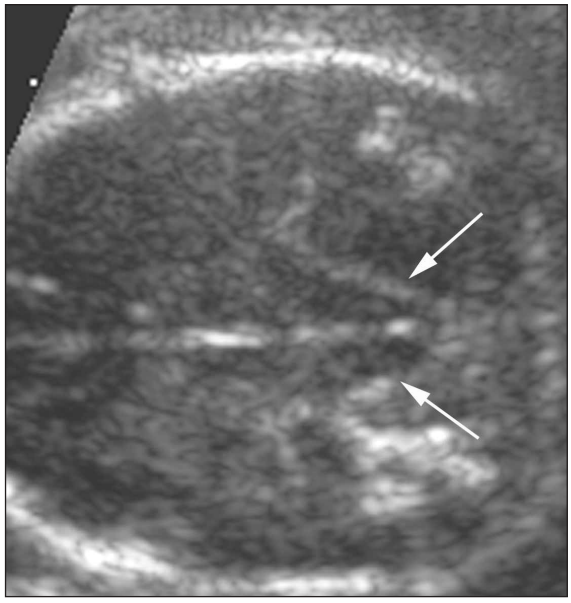
B



Discussion

The diagnosis of myelomeningocele in a fetus is important for many reasons. It provides the parents with an opportunity to consider pregnancy termination. Among parents electing to continue the pregnancy, adequate counseling and psychological preparation can be provided. Plans for delivery at a tertiary center and consideration of surgical delivery and its potential benefit can be addressed. Some studies have suggested that early reversal of a myelomeningocele may lead to

A



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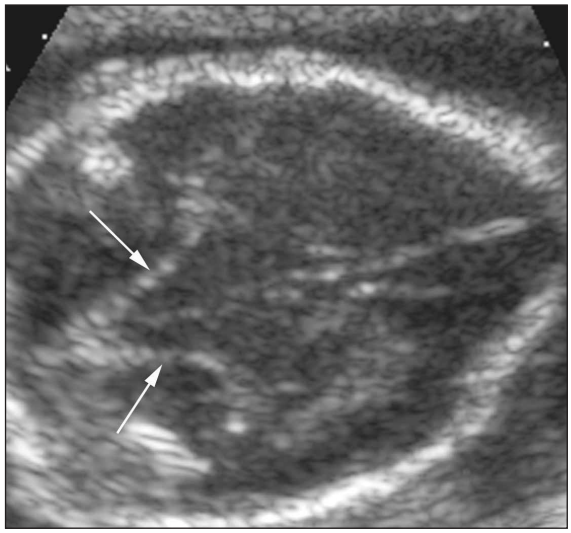

regression of posterior fossa abnormalities, but the long-term effect of prenatal surgery on outcome is still unknown.[9] Currently, patients may be eligible for fetal surgery in the Management of Myelomeningocele Study trial, a randomized prospective trial currently being conducted by 3 centers in the United States. This trial is expected to be completed in 2010. Indeed, if this trial shows a positive benefit, then prenatal diagnosis of myelomeningocele will be even more important.

Fortunately, detection of fetal myelomeningocele is possible in an extremely high percentage of cases.[10] As noted earlier, the greatest improvement in sonographic diagnosis occurred when the focus shifted from observation of the spina bifida abnormality to the cranial findings associated with the Chiari II malformation. Because the intracranial findings associated with the Chiari II malformation are seen exclusively in fetuses with a myelomeningocele, identification of features of the Chiari II malformation virtually ensures that a myelomeningocele is present.

Although the hindbrain findings are paramount among the features of the Chiari II malformation, the posterior fossa is sometimes difficult to appropriately observe because, in the most severe cases, shadowing from the petrous ridges interferes with visualization.[1,4] This may then be mistaken for a technical problem rather than a major abnormality. As well, the posterior fossa deformation may be mild, as was noted in 6 of our cases.

For these reasons, recognition of supratentorial abnormalities can prove valuable in the detection of a myelomeningocele. The abnormalities that have received the most attention in the sonographic literature are the so-called lemon sign (inward scalloping of the frontal bones) and ventriculomegaly. However, the lemon sign is frequently not present in later pregnancies[3] and can be seen in healthy fetuses and in other conditions.[11] Ventriculomegaly may be absent, particularly before 24 weeks, and is associated with many other conditions in addition to myelomeningocele.[12] However, there are multiple supratentorial features of the Chiari II malformation, as already noted, that can also be visualized on prenatal sonograms.

In 1891, Hans Chiari described only the hindbrain manifestations of the deformity that bears his name. Nearly a decade earlier, Cleland[13] had noted the curious beaklike deformity of the corpora quadringemina in his description of what would later be called the Chiari II malformation. In a study of 100 brain stem specimens, Adeloye[14] found that the "mesencephalic spur" was a "uniform occurrence" in the Chiari II malformation. Other pathologic series have placed the incidence of tectal beaking in patients born with a myelomeningocele at nearer to 75%.[15] The finding is seen with similar frequency on postnatal MRI.[16] This apparent discrepancy is likely the result of the stringent criteria used by Adeloye,[14] who divided beaking into 4 varieties ranging from slight fusion of the inferior colliculi to fusion of all colliculi into a pointed mass of unrecognizable tissue.

Although the precise embryogenesis of the Chiari II malformation is not known, the hindbrain manifestations likely result from formation and development within an abnormally small posterior fossa.[15] Subsequent hydrodynamic effects and developmental arrest of the embryonic tectal plate result in varying degrees of fusion of the colliculi with elongation and superior deflection of the tectum. Several authors have concluded that the severity of tectal beaking correlates with the size and extent of the myelomeningocele and that a severe tectal deformity is the likely cause of upward gaze paresis and other oculomotor problems in children with the Chiari II malformation.[6,14,15] A recent study in patients with a repaired myelomeningocele showed that the severity of nystagmus correlates with the severity of tectal beaking, an observation that is not surprising when one considers the anatomic proximity of the third and fourth cranial nerves to the tectum and the function of the superior colliculi in controlling saccadic eye movements.[17]

Our results indicate that tectal morphologic changes are common observations in the Chiari II malformation on fetal sonograms. We observed elongation or beaking of the tectum in 66% of cases with the Chiari II malformation. The frequency of tectal beaking noted in our study was similar to that previously reported in postnatal imaging and pathologic series.[2,15] Importantly, it is seen with equal frequency in fetuses before 24 weeks and in fetuses after 24 weeks. The finding is also seen with equal fre-

quency in fetuses with normal-sized ventricles and those with ventriculomegaly. Therefore, even in the absence of ventriculomegaly, one of the most reliable clues to the presence of a myelomeningocele,[12] tectal beaking, may alert the interpreting physician to the presence of the malformation. Furthermore, tectal beaking is seen more commonly among fetuses with severe posterior fossa deformations than in those with mild distortions, confirming the previous conclusion drawn from pathologic and MRI studies that the presence of tectal beaking correlates with the overall severity of the Chiari II malformation.[6,14,15] We did note the spinal level of the myelomeningoceles in the study cohort but did not compare this parameter with the presence or absence of tectal beaking. Our purpose in noting the spinal level was to document that the range of spinal levels was broad, as was indeed the case. The most proximal involved segment ranged from a cephalad location of the T9 vertebral level to the sacrum.

Importantly, our study was not prospectively constructed to detect tectal beaking. Therefore, although we cannot definitely confirm this, in all likelihood, we underestimated the prevalence of tectal morphologic changes in fetuses with the Chiari II malformation. Furthermore, we observed no significant difference in visualization of tectal beaking among cases judged to be of suboptimal quality versus those of good quality. This indicates that the feature is relatively easily observed even in technically challenging cases.

There were several weaknesses in our study, which should be addressed. First, a retrospective study design starting with known Chiari II malformations was used. No controls were identified. It is unclear how often similarly appearing tectal morphologic changes occur in fetuses without the Chiari II malformation or how accurate sonography is at prospectively identifying these changes. In 8% of our cases, we could not visualize the tectum retrospectively. A prospective case-control study is needed to address these weaknesses. In addition, our study relied on the subjective determination of elongation or pointing of the tectum by a single experienced reviewer. No attempt was made to grade the severity. More readily quantifiable methods should be sought. Pathologic studies have used the depth of the grove between the colliculi as the determinant of the severity of the deformity.[15] This measurement does not seem possible given the resolution of current gray scale sonography, but the resolution of sonographic systems does improve progressively with advances by manufacturers. A recent MRI study defined the morphologic appearance of the tectum as seen on midsagittal MRI of the head as normal (type I), mild beaking with primarily the inferior colliculi involved (type II), and severe beaking with both colliculi involved (type III).[17] It is not clear whether grading tectal changes using a similar scale on sonography would be possible.

Although no attempt was made to retrospectively grade the severity of tectal morphologic changes in our study, if future studies can establish the feasibility and accuracy of sonographic grading of tectal beaking, tectal changes on prenatal sonography may be able to independently predict the severity of the hindbrain findings and the severity of future oculomotor impairment.

In conclusion, tectal morphologic alteration is a common supratentorial feature of the Chiari II malformation on prenatal sonography both before and after 24 weeks' gestation and in the presence or absence of ventriculomegaly. Its frequency increases with the severity of posterior fossa abnormalities, and its presence has been shown to correlate with long-term oculomotor impairment. Therefore, it is useful as a supratentorial indicator of the presence, the severity, and potentially the prognosis of the Chiari II malformation.

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