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Abnormal Spinal Curvature in the Fetus

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We reviewed 20 cases of sonographically detected abnormal fetal spinal curvature to help determine the significance of this finding. Both marked (13 cases) and mild (7 cases) curves were diagnosed sonographically. Sonography showed associated anomalies in 19 fetuses, including neural tube and ventral wall defects. Outcomes were poor, with only three infants surviving. Isolated scoliosis was identified prenatally in one neu-

rologically normal infant with hemivertebrae. We conclude that abnormal spinal curvature in the fetus is a significant finding, whether mild or severe. Even as an isolated finding, it is significant, as prenatal detection assists postnatal management. **KEY WORDS:** Fetus, ultrasound studies; Fetus, abnormalities; Fetus, central nervous system; Fetus, skeletal system; Spine, curvature.

Evaluation of the fetal spine with high-resolution sonography has largely focused on the detection of neural tube defects. This close scrutiny of the developing spine has led to the prenatal diagnosis of a number of other spinal abnormalities. Reports have been made of the identification in utero of vertebral body malformations, such as hemivertebrae and butterfly vertebrae, and of fetal scoliosis.¹⁻³ However, the significance of these other spinal abnormalities when detected in utero has not been explored. Abnormal spinal curvature in infants is recognized as a distinct pathophysiologic entity from the more common idiopathic form presenting in adolescence. Congenital scoliosis is a structural deformity resulting from vertebral malformations, usually with different growth rates on the two sides of the spine. It is known to be associated with other anomalies, particularly those involving the spinal cord, genitourinary system, and gastrointestinal system, as well as with musculoskeletal dysplasias.^{4,5} This study was performed to determine the outcome for fetuses with sonographically detected scoliosis and help elucidate the significance of the finding.

MATERIALS AND METHODS

Twenty fetuses were diagnosed as having abnormal spinal curvature at the University of California, San Diego, Department of Radiology from April 1986 to July 1991. These fetuses comprised our study group. Three cases of mild fetal scoliosis were identified during a short time period, and these prompted our study. Other fetuses were identified by review of the case files of interest, from which seven additional cases with prenatally diagnosed scoliosis or kyphosis were noted. The remaining 10 were identified by a review of ultrasound cases with a known association with scoliosis. All fetal spines are routinely examined in transverse and longitudinal planes for normal configuration and curvature. The sonograms, spinal radiographs, autopsy reports, and medical records were reviewed retrospectively. The sonograms were evaluated by three radiologists. Abnormal spinal curvature was defined as any focal, fixed kyphosis or as any fixed curvature. As the fetal spine is quite mobile, only clearly fixed abnormalities of curvature that persisted over the course of the sonographic study despite fetal movements were included. The anatomic level of the area of abnormal curvature was noted, and the degree of severity was assessed subjectively with the curvatures graded as either mild or marked. The changes in fetal position and the frequent inability to include the entire spine on a single image made it difficult to perform a standardized quantitative measurement of the actual angle of curvature on all fetuses, so this was not performed.

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The sonograms were evaluated for other anomalies, particularly neural tube or abdominal wall defects, amniotic bands, cranial abnormalities, and abnormal amniotic fluid volume. The outcome of each pregnancy was recorded, and the abnormal spinal curvature was confirmed by autopsy findings, physical examination, or postnatal spinal radiographs. The spinal radiographs were interpreted in the same manner as the sonograms, with the spinal curvature graded qualitatively as mild or marked and the anatomic level of deformity noted. The radiographs also were evaluated for the presence of vertebral body malformations. Other data collected, if available, included reason for ultrasound examination referral, fetal age at diagnosis, maternal serum alpha-fetoprotein (AFP) levels, and fetal karyotype.

RESULTS

Reasons for ultrasound referral were varied and included high (8 cases) or low (1 case) maternal serum AFP level, further evaluation of abnormalities noted on sonograms obtained at other institutions (7 cases), history of a previous child with a neural tube defect (3 cases), and gestational dating (1 case). Sonographic examinations were performed at ultrasonically determined gestational ages ranging from 13 to 31 weeks. In two cases of twin gestations, one twin was affected and the other sibling was normal sonographically and at delivery. Karyotypes were available for six fetuses, with only one being abnormal (trisomy 18).

Review of the sonograms showed scoliosis or kyphosis of both marked (13 cases) and mild (7 cases) severity (Figs. 1 and 2). In 12 fetuses, the abnormal spinal curvature was primarily a scoliosis, whereas in the remaining eight fetuses, the abnormal curve included a component of kyphosis. Abnormal curves involved all anatomic levels, with the most frequent site of involvement being the thoracolumbar spine (5 cases), followed by the thoracic spine (4 cases), lumbosacral spine (4 cases), cervical spine (3 cases), and lumbar spine (3 cases). One fetus had abnormal curvature involving all segments of the spine.

Evaluation of the sonograms showed other abnormalities in 19 of 20 fetuses (95%). Final diagnoses made on the basis of pathologic and sonographic evaluations are summarized in Table 1. The one fetus with isolated scoliosis and no other abnormalities by sonography had mild involvement. The mother of this fetus was referred for a family history of anencephaly. Mild thoracic scoliosis was identified on longitudinal, coronal scans, and this was confirmed on serial studies (Fig. 3A). Although a vertebral body malformation was suspected as the cause of the abnormal curve, it could not definitely be confirmed on sonography. A radio-

graph obtained at 6 months of age showed the mild scoliosis and two balanced hemivertebrae (Fig. 3B). This infant was normal neurologically, and the scoliosis was not detectable by physical examination.

Four of the seven cases of mild scoliosis (57%) were found in fetuses with myelomeningoceles, all of which were diagnosed prenatally. The remaining two fetuses with mild abnormalities of curvature had multiple anomalies. One of these (Fig. 2) had severe dysplasia of the left side of the thoracic cage, a hypoplastic left lung, and left renal agenesis. The other fetus had lower limb agenesis, absent sacrum, and multicystic kidneys, likely representing a severe form of caudal regression syndrome. Vertebral anomalies were suspected in the lumbar spine on prenatal sonography of this fetus, and these were confirmed on postmortem radiographs.

All of the 13 fetuses with marked scoliosis had other sonographically demonstrated abnormalities. Neural tube defects were the most common sonographic finding, seen in eight of the 13 fetuses with marked spinal curvatures and in 12 of the 20 patients overall. One of the anencephalic fetuses also showed spinal dysraphism of the entire spine, a condition known as craniorachischisis. In five fetuses, sonograms showed ventral wall defects of varying size fulfilling criteria for the limb-body wall complex. In one case, the amniotic band syndrome was suspected on the basis of abnormal membranes about the fetus. Cranial abnormalities were frequent and included ventriculomegaly in patients with myelomeningoceles (6 cases) and holoprosencephaly (1 case). The amniotic fluid volume was normal in 14 patients, decreased in 3 patients, and increased in two patients.

The outcome of the 20 pregnancies included pregnancy termination in 12 patients. In the remaining eight patients, three fetuses were delivered stillborn and two neonates died within the first day of life. Three infants are living, including the 6 month old infant with hemivertebrae and isolated scoliosis described earlier. The remaining two living children have neurologic deficits from myelomeningoceles.

Scoliosis diagnosed by sonography was confirmed in 13 of the 20 patients. Confirmation was by postnatal or postmortem radiograph in nine cases. There was good correlation between prenatal diagnosis and radiographic findings in terms of level and severity of abnormal curvature. Discrepancy in severity was seen in only one case, in which prenatal sonography underestimated the severity of thoracic kyphoscoliosis. However, correlation was not good between sonographic findings and radiographic demonstration of vertebral body anomalies. Vertebral body anomalies were seen on radiographs of three patients. Although anomaly was suspected sonographically in two cases, the extent was underestimated as both of these patients had

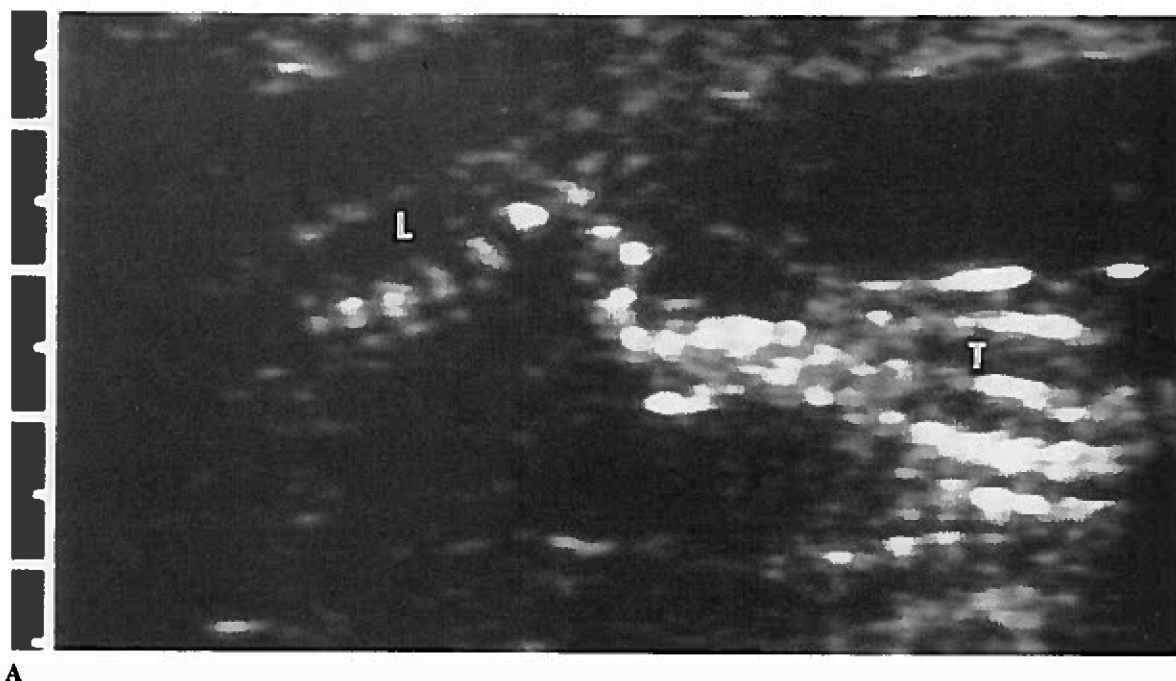


Figure 1 Marked kyphoscoliosis in myelomeningocele. A, Longitudinal scan of the fetal spine at 19 weeks' gestational age shows marked kyphoscoliosis involving the thoracolumbar spine. A thoracolumbar myelomeningocele also was diagnosed. T, Upper thoracic vertebrae; L, lumbar vertebrae. B, Radiograph at 3 years of age demonstrates marked thoracolumbar kyphoscoliosis.

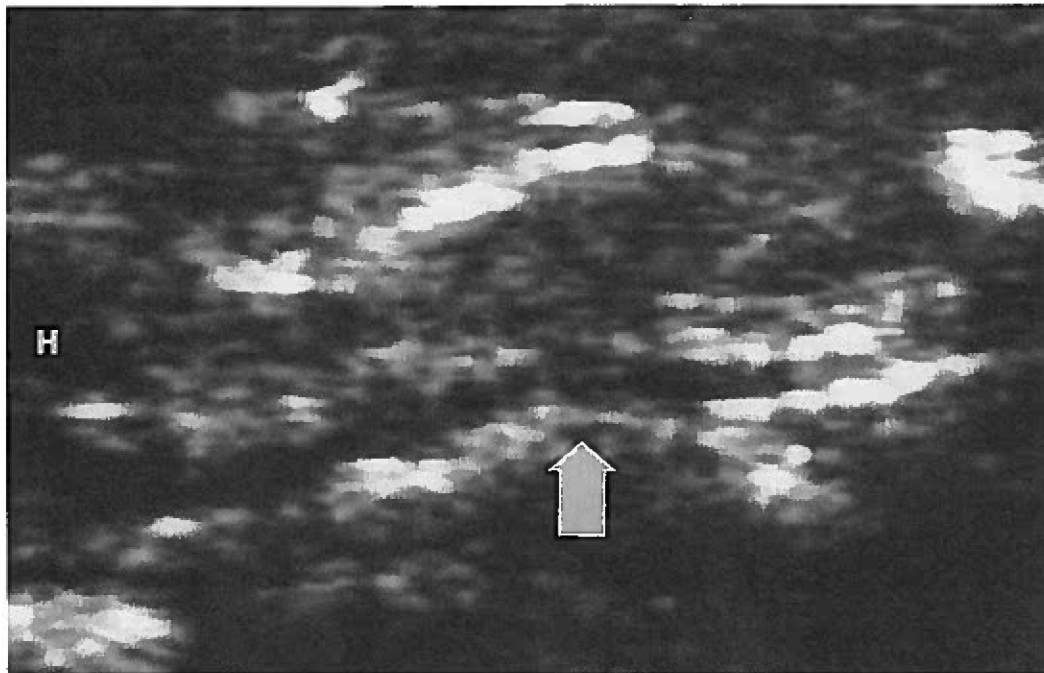
vertebral anomalies at multiple levels on spinal radiographs.

In the remaining four confirmed cases, confirmation was obtained by autopsy examination. Of the seven cases for which confirmation was unavailable, five were in pregnancies terminated by dilation and extraction. Severe fragmentation prevented confirmation of the sonographic findings. Confirmation of scoliosis was possible in three other fetuses aborted by this method. In the two remaining cases that were not confirmed, mild scoliosis was not noted on physical examination of stillborns with myelomeningoceles, and no radiographs were obtained.

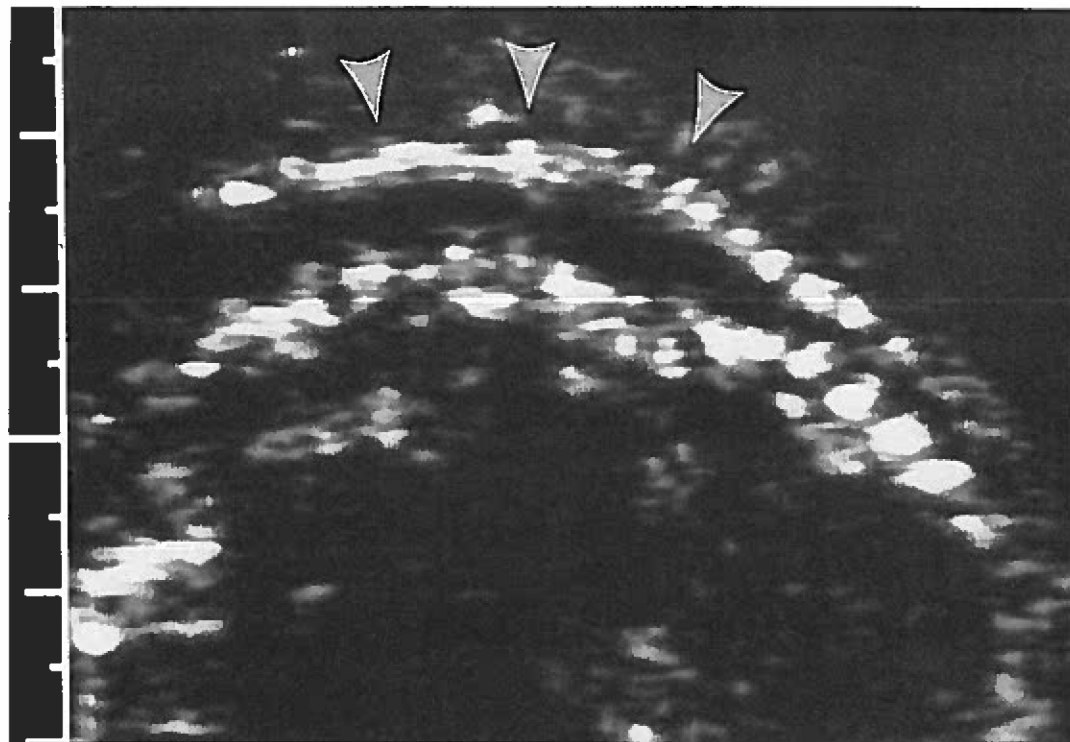
DISCUSSION

Advancements in real-time sonography have allowed an unprecedented view of the developing fetus. A number of abnormalities, including abnormal spinal curvature, are being detected prenatally with little knowledge of their significance when identified in utero. Although we may be tempted to equate prog-

Figure 2 Mild scoliosis in thoracic dysplasia. **A**, Longitudinal, coronal scan of the fetal spine at 16 weeks' gestational age shows mild thoracic scoliosis (*arrow*). H, Head. **B**, A follow-up study at 18 weeks confirms the mild scoliosis, and this longitudinal, parasagittal scan also shows splaying of the spine at this level (*arrowheads*). **C**, Postmortem radiograph shows marked thoracic cage deformity, particularly on the left, with mild thoracic scoliosis. Multiple other congenital anomalies were identified in this fetus.



A



B



C

Table 1: Diagnoses in Fetuses with Abnormal Spinal Curvature

Diagnosis	Number (N = 20)
Neural tube defect	12
Myelomeningocele	6
Frontal encephalocele with ectrodactyly	1
Exencephaly	1
Anencephaly	1
Anencephaly with trisomy 18	1
Iniencephaly	1
Craniorachischisis	1
Limb-body wall complex (LBWC)	2
Amniotic band syndrome (ABS)	1
Multiple congenital anomalies, likely LBWC or ABS	2
Caudal regression syndrome	1
Thoracic dysplasia with multiple anomalies	1
Hemivertebrae, no other anomalies	1

nosis of in utero abnormalities with that of the same conditions in neonates, this may be erroneous.

Abnormal curvature of the fetal spine can be detected by sonography on routine longitudinal views of the fetal spine. We were able to identify fetal scoliosis and kyphosis of both marked and mild severity. Severe

deviations from normal spinal curvature are readily identified and often are seen in fetuses with marked dysmorphology. In these fetuses, scoliosis represents one aspect of a more complex congenital malformation. Marked scoliosis often is manifested as an inability to obtain longitudinal images of the entire spine.

Mild degrees of scoliosis also are detectable prenatally. Detection requires careful longitudinal scanning in both coronal and parasagittal planes. The middle of the second to early third trimester is the ideal time to identify abnormal spinal curvature as there is sufficient spinal ossification without the fetal crowding late in pregnancy that makes the spine difficult to image. When the fetal spine is evaluated carefully, so as to exclude neural tube defects, even mild deviations from normal spine curvature are detectable. Real-time scanning should demonstrate persistence of the abnormal spinal curvature despite fetal movements. Serial scanning often is helpful in subtle cases to confirm the fixed nature of the abnormal curve. Vertebral malformations such as hemivertebrae or butterfly vertebrae can be identified by noting asymmetry in the number of ossification centers on high-resolution equipment.^{1,2} However, our study indicates that despite careful scanning, vertebral malformations may be recognized only by their disruption of normal spinal curvature. Although sonography's true sensitivity for the detection of abnormal spinal curvature and of vertebral body malformations cannot be determined from this retrospective study, it is clear that mild scoliosis can be diagnosed prenatally.

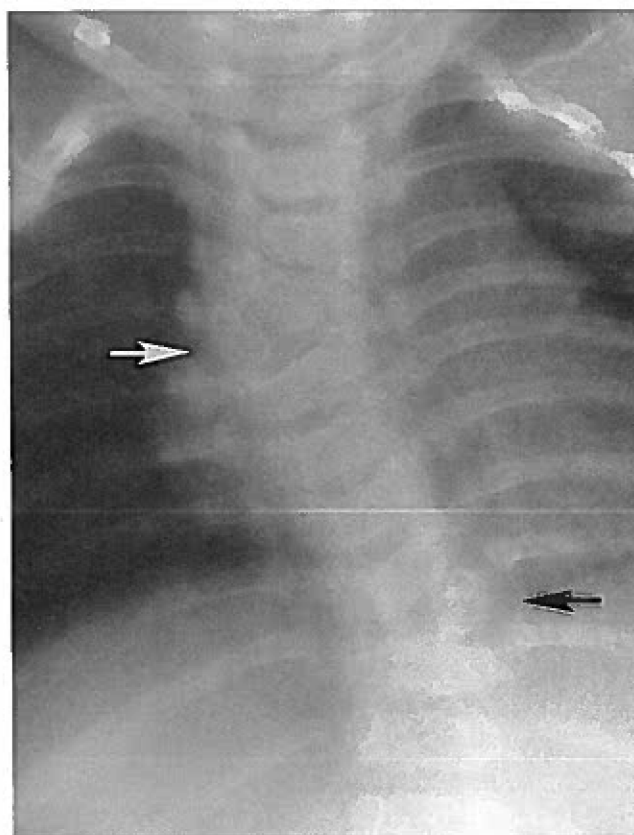
Fetal scoliosis is associated with a number of congenital abnormalities, as seen in 19 of 20 cases in our study group. These include neural tube defects, limb-body wall complex, amniotic band syndrome, caudal regression syndrome, arthrogryposis, skeletal dysplasias, and the VACTERL association.⁶ The most frequently associated anomaly in our study group was neural tube defects (60%). In all of the cases of marked fetal scoliosis or kyphosis, other anomalies were present. However, the high incidence of associated anomalies in our study must be interpreted in light of our retrospective study design and the possibility that the study was biased by our means of patient selection.

The prognosis for fetuses with abnormal spinal curvature generally is determined from the prognosis of the underlying malformation, and usually it is poor. For example, anencephaly, the anencephalic variants, and the limb-body wall complex were uniformly fatal disorders, and these represented 45% of our study group. Again, selection bias may have contributed to the high percentage of fatal disorders. Because of the poor prognosis of many of the associated anomalies, 12 families (60%) elected pregnancy termination. Even with continuation of pregnancy, outcomes were poor,

Figure 3 Isolated vertebral body malformations. **A**, Longitudinal coronal scan of fetal spine at 21 weeks' gestational age. Note the mild scoliosis (*arrow*) in the thoracic spine. H, Head. **B**, Radiograph at 6 months of age demonstrates mild thoracic scoliosis with hemivertebrae (*arrows*) at the T5-6 and T9-10 levels.



A



B

with five of eight ending in stillbirth or neonatal death. Perhaps the most important finding of our review is that fetuses with only mild abnormalities of curvature also have poor outcome. Of the seven fetuses with mild scoliosis, two families elected pregnancy termi-

nation because of the severity of associated anomalies, and three fetuses suffered in utero or early neonatal death.

Isolated fetal scoliosis without associated anomalies is most likely a result of vertebral body malformations.

The diagnosis of scoliosis from vertebral body malformation can be presumed when other anomalies, particularly myelomeningoceles, have been excluded. Identification of this type of anomaly in utero is important for appropriate postnatal management. As with the infant in our study, physical examination alone may fail to identify the abnormal spinal curvature. Scoliosis present at birth carries a significant risk of progression of the curve, and evaluation by orthopedic specialists is indicated once it is identified.⁷ Hemivertebrae and other vertebral malformations may be associated with other, unsuspected spinal anomalies, such as myelomeningoceles or diastematomyelia.⁴ Some experts also advocate routine evaluation of the genitourinary system of these infants because of the high frequency of associated anomalies.⁸

Once abnormal spinal curvature is identified, a thorough evaluation to exclude associated anomalies should be performed. Attention should be directed to excluding neural tube defects, abdominal wall defects, amniotic bands, and cranial abnormalities. The fetal limbs should be measured to assess for skeletal dysplasias. Prenatal sonography plays a particularly important role when the pregnancy is terminated by dilation and extraction of the fetus. In these cases, sonography may be the only means of determining a diagnosis, as fragmentation of the fetus may preclude adequate pathologic evaluation.

In conclusion, abnormal spinal curvature in the fetus is a significant finding. When severe, it is often associated with other, frequently lethal anomalies and when mild it has a high association with spinal dys-

raphism. The identification of abnormal spinal curvature in utero should prompt a thorough evaluation to exclude other abnormalities. Even when isolated, abnormal curvature is significant, and the obstetrician and pediatrician should be alerted to ensure appropriate postnatal management.

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