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Prenatal Sonographic Diagnosis of Congenital Hemivertebra

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The prenatal sonographic diagnosis of spinal abnormalities associated with neural tube element malformations has been well described; however, the diagnosis of fetal skeletal anomalies of the vertebral bodies with a neurologically intact spine has received little attention. Three cases of congenital hemivertebra identified sonographically in fetuses between 17 and 28 weeks' gestation are reported. One of the fetuses also had associated abnormalities involving the head and extremities. The prenatal diagnosis of congenital hemivertebra should alert the sonographer to search for other congenital defects and for orthopedic complications of vertebral body malformations in childhood. Prior knowledge of the presence of an abnormal vertebral body can alert the pediatrician to watch for early signs of problems such as scoliosis or kyphoscoliosis, and to plan surgery if necessary. (Key words: ultrasonography; obstetrics; pediatrics; vertebrae, anomalies of; prenatal diagnosis)

Sonography has been extremely useful in the prenatal diagnosis of neural tube defects and is currently a promising diagnostic modality for examination of the fetal spine.^{1,2} There are many reports describing the appearance of such fetal spine defects as meningocele and spinal dysraphism. Although the sonographic diagnosis of spinal abnormalities associated with neural tube element malformations is well described, the diagnosis of fetal skeletal anomalies of the vertebral bodies with a neurologically intact spine has received little attention.

We describe three cases of congenital hemivertebra identified sonographically in fetuses between 17 and 28 weeks' gestation. The prenatal diagnosis of this finding should alert the sonographer to look for other associated fetal anomalies and will prepare the pediatrician for possible subsequent complications.

REPORTS OF THREE CASES

Case 1. A 29-year-old gravida 1, para 0, woman was first diagnosed with a twin pregnancy at eight weeks.

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Past medical history included pseudoxanthoma elasticum, diagnosed at 7 years of age; and duodenal ulcer, complicated by a hemorrhage on two occasions requiring transfusions. Her twin pregnancy was uncomplicated.

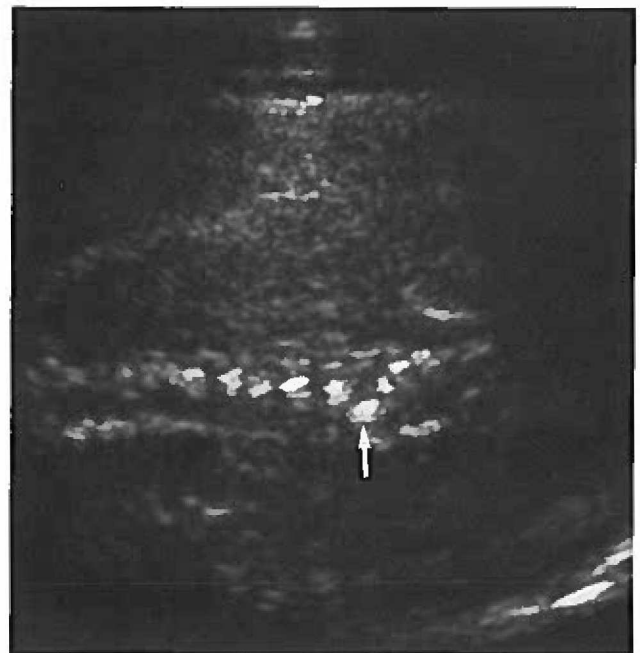


Figure 1. Case 1. Longitudinal view of the lumbar spine at 26 weeks in an oblique projection. Notice the abnormal ossification center of the hemivertebra (arrow). There was no visible scoliosis at this time, and the scoliosis appeared several weeks after birth. This and subsequent images were made using an Acuson 128 imaging system.

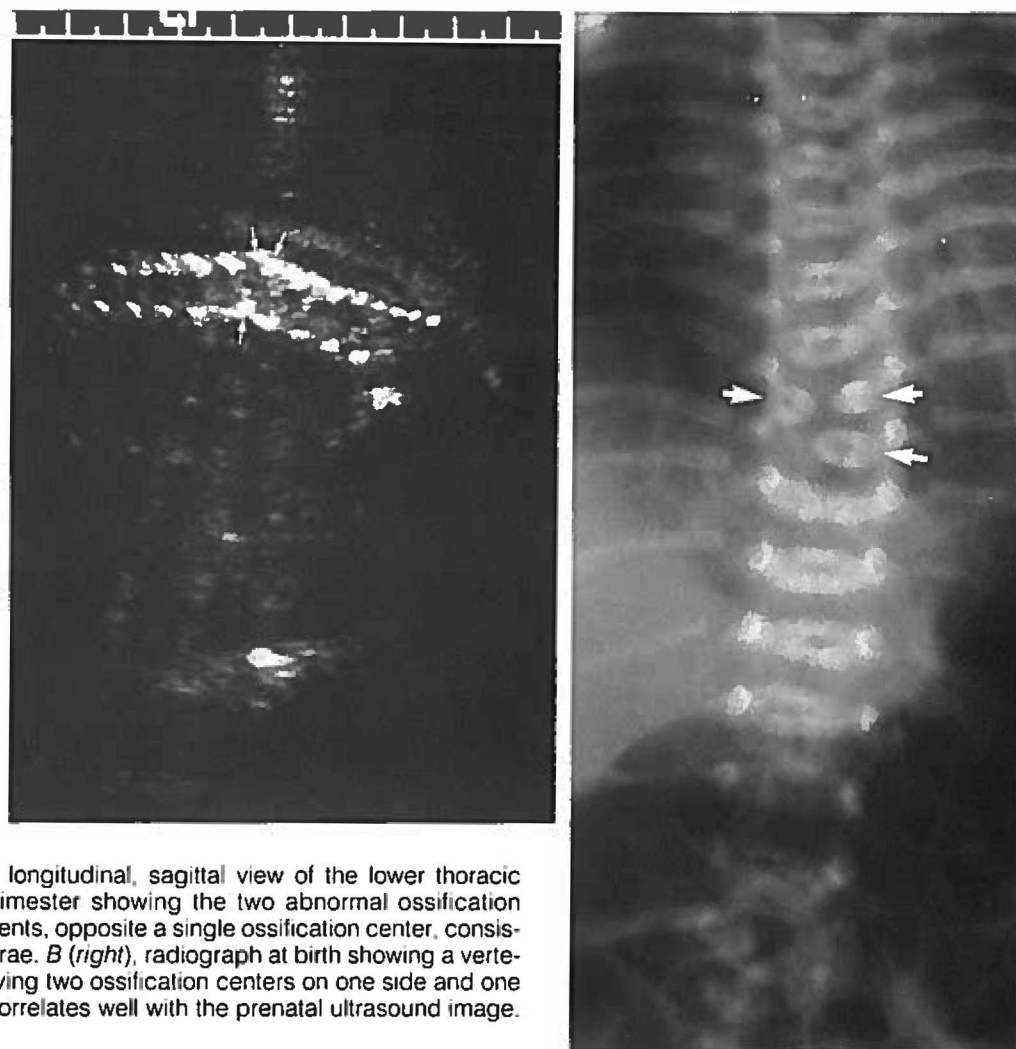


Figure 2. Case 2. A (left), longitudinal, sagittal view of the lower thoracic spine in the late second trimester showing the two abnormal ossification centers of the posterior elements, opposite a single ossification center, consistent with an abnormal vertebrae. B (right), radiograph at birth showing a vertebral abnormality at T-9 involving two ossification centers on one side and one on the other (arrows). This correlates well with the prenatal ultrasound image.

cated and she had sonograms at 8 weeks, 12.5 weeks, and 16 weeks, where no structural abnormality could be identified. During the 26-week sonogram, an abnormality in one of the lumbar vertebral bodies was identified on the left twin (fig. 1). No other anomaly could be seen. This abnormal vertebral body was noted again at 30 weeks and 32.5 weeks. Her pregnancy was otherwise uneventful until her membranes ruptured spontaneously at 36 weeks. Labor was induced with oxytocin and twin 2 developed monitor evidence of fetal distress. A primary cesarean section was performed for delivery of twins with Apgar scores of 9/9 and 8/9 and fetal weights of 2330 grams and 1850 grams, respectively. A hemivertebra was confirmed radiographically in the upper lumbar spine of twin 2, with no other congenital abnormality noted.

Case 2. A 23-year-old gravida 2, TAB 1, para 0, woman was a Class R diabetic with the onset of her diabetes at 9 years of age. Her initial visit was at eight weeks of gestation, where her hemoglobin A1C was noted to be 14 (normal up to 7.6). Her first sonogram at 17 weeks revealed a blighted twin and a single live fetus with an abnormal vertebral body in the lower thoracic spine (fig. 2A). A repeat scan was performed at 20 weeks

and again at 22 weeks, where the sonographic diagnosis of a hemivertebra was made in the distal thoracic spine. No other congenital abnormality could be seen. Spontaneous rupture of membranes occurred at 37 weeks and a primary cesarean section was done for active proliferative retinopathy. A female fetus, 2380 grams, with Apgar scores of 7 and 8 and one and five minutes, respectively, was delivered and the vertebral abnormality noted sonographically was confirmed by radiograph (fig. 2B).

Case 3. A 37-year-old gravida 2, TAB 1, para 0 had her first sonogram at 28 weeks. The diagnoses of hydrocephalus, vertebral abnormality in the lower thoracic spine, and clubfoot were made at that time. These findings were confirmed again at 30, 32, and 34 weeks sonographically. At 35.5 weeks, premature rupture of the membranes occurred, with subsequent premature labor. A normal vaginal delivery ensued, yielding a 2180-gram baby with Apgar scores 1/4/5. On physical examination, the following abnormalities were noted: dysmorphic features, wide-set eyes, right epibulbar dermoid, hypoplastic mandible, microtia of the left ear, asymmetrical chest wall, bilateral clubfeet, scoliosis with a thoracic hemivertebra and accompanying rib ab-

normality just below the ninth rib, 13 ribs on the right and 12 on the left, and a sacral dimple. The skeletal abnormalities were confirmed radiographically.

DISCUSSION

Sonography has been used extensively for the prenatal diagnosis of neural tube defects.¹ Other vertebral body abnormalities can cause significant orthopedic problems, although the neural elements are normal. To our knowledge, the literature contains only sporadic case reports of vertebral malformation diagnosed prenatally by ultrasonography.^{3,4} We describe three cases of congenital hemivertebra where the diagnosis was made prenatally, by sonography, between 17 and 28 weeks of gestation. One of our patients also had associated anomalies involving the head and extremities and it is well known the abnormalities of the vertebral bodies can be associated with a variety of other congenital defects such as the VATER or VACTERL syndromes.^{5,6} When a vertebral malformation is discovered sonographically, a careful search should be made for other malformations. Anomalies of the extremities, gastrointestinal tract, central nervous system, and other organ systems may also be present. The second patient was a diabetic whose pregnancy was at high risk for a congenital spinal anomaly because of her initially high hemoglobin A1C. According to Miller et al.,⁷ she had a 22 per

cent chance of carrying a fetus with a major malformation.

Although congenital hemivertebra is not a life-threatening malformation, the prenatal diagnosis of this abnormality of segmentation should raise the index of suspicion for other congenital defects and for orthopedic complications of vertebral body malformations in childhood. Severe scoliosis or kyphoscoliosis can develop and prior knowledge of the presence of an abnormal vertebral body can alert the pediatrician to watch for early signs of problems and plan surgery if necessary.

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