

# ULTRASONOGRAPHIC DIAGNOSIS OF DIASTEMATOMYELIA DURING THE 14<sup>TH</sup> WEEK OF GESTATION

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## SUMMARY

**Objective:** We present a case of type I diastematomyelia diagnosed with ultrasonography at the 14<sup>th</sup> week of gestation.

**Case Report:** A 26-year-old primigravida at 14 weeks' gestation was presented to our outpatient department with a complaint of dysuria. Routine antenatal ultrasound revealed disruption of the fetal upper thoracic vertebral column curvature, together with hemivertebrae. Further high-resolution two-dimensional ultrasound examination revealed dilatation of the thoracolumbar vertebrae and extra echogenic foci in the spinal canal. Thoracolumbar meningocele was also observed. These observations (thoracic hemivertebrae, scoliosis and thoracolumbar meningocele) suggested a diagnosis of type I diastematomyelia. The family was counseled regarding the prognosis for the fetus. Their decision to terminate the pregnancy was considered and approved by the ethical council. The postabortion pathologic examination, along with X-ray, three-dimensional computed tomography and magnetic resonance imaging examinations, supported the initial diagnosis.

**Conclusion:** Prenatal diagnosis of diastematomyelia during the 14<sup>th</sup> week of pregnancy is possible using high-resolution ultrasound. To the best of our knowledge, this is the earliest case presented in the literature. [*Taiwan J Obstet Gynecol* 2009;48(2):163-166]

**Key Words:** computed tomography, diastematomyelia, prenatal diagnosis, ultrasound

## Introduction

Diastematomyelia is a rare form of spinal dysraphism characterized by a sagittal cleft in the spinal cord, conus medullaris, and/or filum terminale with splaying of the posterior vertebral elements [1,2]. This condition results from the complete or incomplete sagittal division of the spinal cord into two hemicords owing to the presence of an osseous or fibrocartilaginous septum, caused by abnormal development of the notochord between 15 and 18 days' gestation [3].

Diastematomyelia can be associated with spinal dysraphisms, such as myelomeningocele, meningocele, spinal lipoma, neuroenteric cysts or dermal sinuses; it can also be associated with vertebral corpus abnormalities, such as hemivertebrae, butterfly vertebrae, kyphosis or scoliosis [1,2]. Other internal organ abnormalities may also be associated. Cases of isolated diastematomyelia have been found to have better prognoses [3].

Structural abnormalities in the fetal neurovertebral axis can be diagnosed using prenatal ultrasound. The first prenatal diagnosis of diastematomyelia was reported in 1985 [4], and a total of 22 cases of diastematomyelia with typical features have been reported in the literature [5,6].

This case report is presented along with a review of the current literature. The case was confirmed as thoracolumbar diastematomyelia with accompanying meningocele, thoracic scoliosis and hemivertebrae,



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diagnosed during a prenatal visit (during the 14<sup>th</sup> week of gestation).

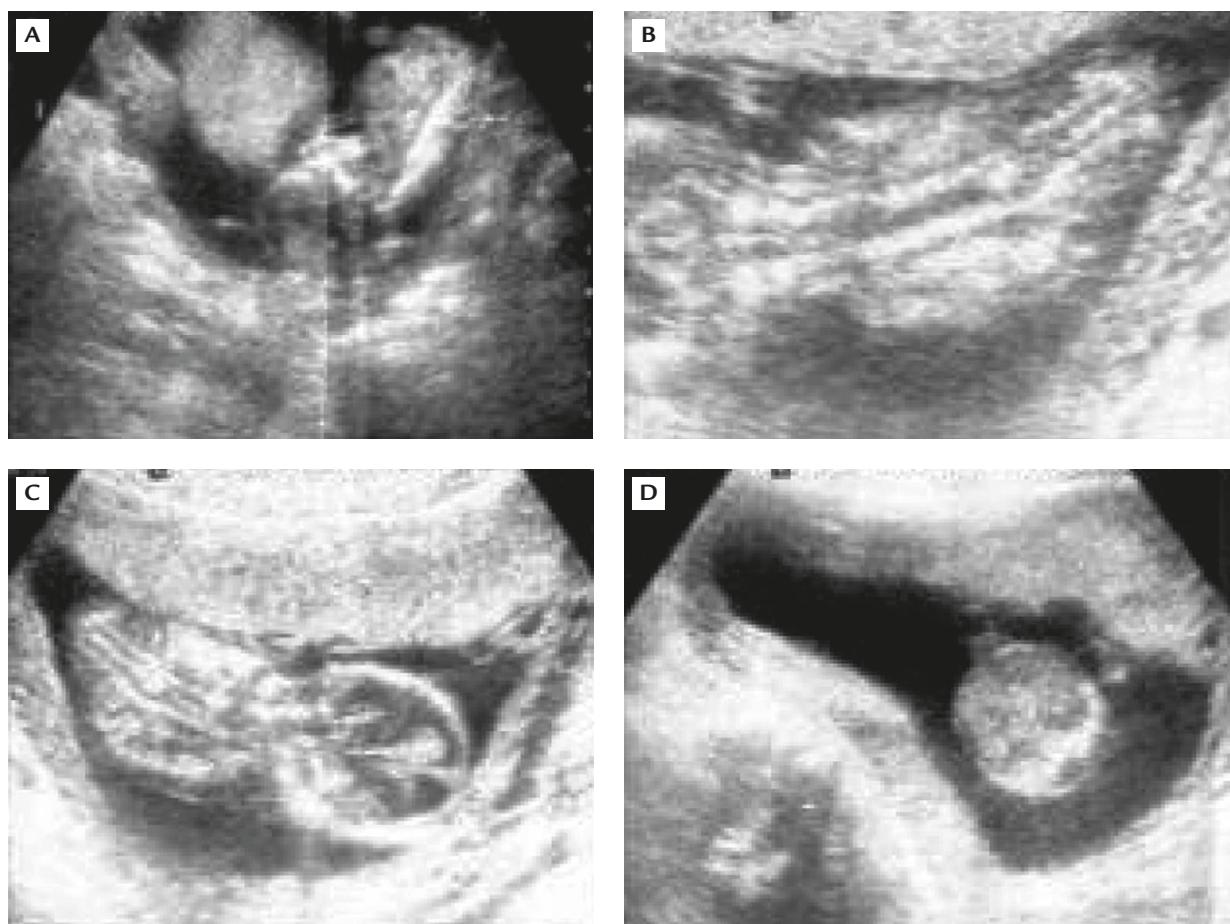
## Case Report

A 26-year-old primigravida at the 14<sup>th</sup> week of gestation was presented to our outpatient clinic with a complaint of dysuria, and was diagnosed with a lower urinary tract infection. Cultures and an antibiogram were obtained, and appropriate antibacterial treatment was started. Routine antenatal ultrasound screening had been performed using high-resolution ultrasound (Sonoline Antares; Siemens AG, Erlangen, Germany). Biparietal diameter, femur length, abdominal circumference and humerus length were 26 mm (14 weeks 4 days), 14 mm (14 weeks 1 day), 77 mm (14 weeks 0 days), and 14 mm (13 weeks 5 days), respectively. Fetal heart motion, lower and upper extremity movements, amniotic fluid, and internal organs were normal. However, cranial examination revealed a “lemon sign” and “banana sign”, arousing suspicion. A subsequent detailed ultrasound

examination focusing on the vertebral column revealed a disturbance in the curvature of the upper thoracic vertebrae, along with hemivertebrae, thoracolumbar spinal canal dilatation, and extra echogenic foci within the spinal canal (Figures 1A–C). A meningocele was observed in the thoracolumbar area (Figures 1A and 1D). A diagnosis of type I diastematomyelia was made, based on these findings of thoracic hemivertebrae, scoliosis, and thoracolumbar meningocele.

A detailed anamnesis revealed that a recent ultrasound scan, when the crown-rump length was 50 mm (12 weeks), showed a 0.9-mm nuchal translucency, and a nasal bone was present. The mother did not report any other medical conditions. She previously used a folic acid supplement of 400 µg per day for 3 months, which she began taking prior to conception. No other extraordinary findings in terms of personal or family history were noted. The parents were not related to each other. A genetic consultation failed to reveal any genetic diseases in the family pedigree.

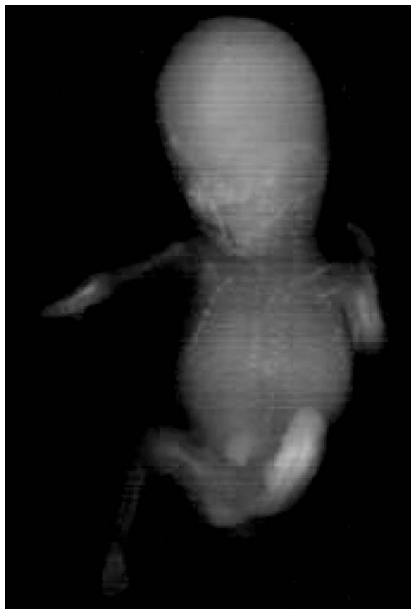
The family was given counseling. Their wish to terminate the pregnancy was considered, and an ethical



**Figure 1.** (A, B, C) Ultrasonographic views of disturbance of the curvature of the upper thoracic vertebrae, along with hemivertebrae, a thoracolumbar spinal canal dilatation, and extra echogenic foci within the spinal canal. (D) Ultrasonographic view of thoracolumbar meningocele.



**Figure 2.** Postmortem examination supported the diagnosis of thoracolumbar meningocele.

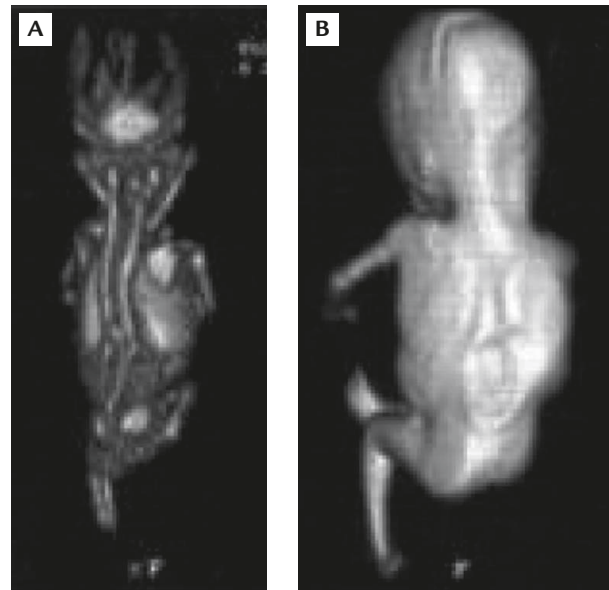


**Figure 3.** Thoracolumbar spinal canal dilatation and hyperdense foci inside the canal were detected.

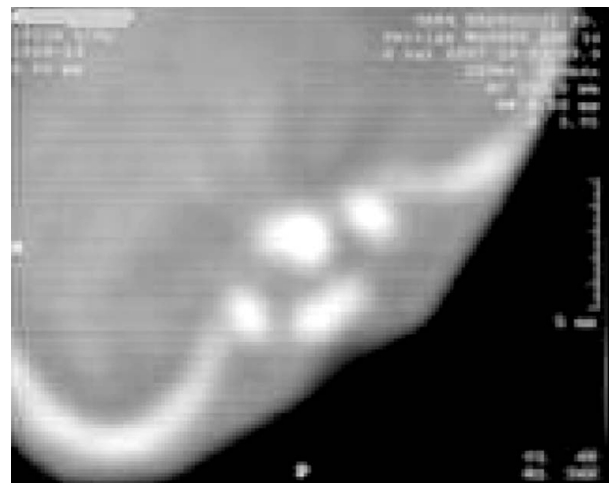
council decision was made. The pregnancy was terminated using vaginal misoprostol induction.

The postmortem examination supported the diagnosis of thoracolumbar meningocele (Figure 2). X-ray examination of the fetus showed upper thoracic hemivertebrae and scoliosis. Thoracolumbar spinal canal dilatation and hyperdense foci within the canal were also found (Figure 3). The extremities were normal. Postmortem magnetic resonance imaging (MRI) results were not satisfactory because of technical difficulties. Three-dimensional computed tomography clearly revealed thoracic scoliosis, hemivertebrae, and meningocele (Figures 4A and 4B).

A transverse computed tomography section revealed three ossification centers, which was specific



**Figure 4.** (A, B) Three-dimensional computed tomographic views of thoracic scoliosis, hemivertebrae, and meningocele.



**Figure 5.** Transverse computed tomography section showing three ossification centers, which are a specific finding for diastematomyelia.

for diastematomyelia (Figure 5). Fetal karyotype was reported as 46,XY.

## Discussion

Diastematomyelia results from the dorsoventral division of the spinal cord and/or filum terminale into two parts, frequently with a septum. This septum is fibrous in 25% and osseous in 75% of cases [7].

Ultrasonographic diagnosis of diastematomyelia is made using the criteria of hemivertebrae, vertebral fusion defects, dorsal midline cutaneous lesions, enlargement of the spinal cord, and echogenic masses in the spinal canal [8,9].

In the current case, after suspicion was aroused by the presence of “lemon” and “banana” signs, spinal canal dilatation in coronal plans, spinal canal hyper-echogenic foci, and meningocele were also detected by ultrasonography. Hyperechogenic foci in the spinal canal are the most specific finding for the diagnosis of diastematomyelia, with an incidence of 0.06% in 10,070 prospective ultrasound examinations [3]. The finding of three ossification centers in transverse plans is an important supportive finding for the diagnosis of diastematomyelia.

Interestingly, although this condition is more common in female fetuses, our case was phenotypically and genotypically male. To the best of our knowledge, this is the earliest diagnosed case in the literature.

Diastematomyelia is subdivided into type I and type II [3]. Type I is characterized by two separate arachnoid and dural sheaths, and a bony spur is frequently observed. These cases are usually symptomatic. Type II is characterized by a single arachnoid and single dural sheath, and a bony spur is frequently not observed. These cases are usually asymptomatic.

These two types of diastematomyelia are called split cord malformations. Hypertrichosis (56%), capillary hemangiomas (26%), dermal sinuses (22%), and subcutaneous lipomas (11%) over the affected area are also encountered. These are the main reasons for admission to hospital in newborns [10]. The most frequent abnormality is midline hypertrichosis over the affected region.

Initial postnatal imaging for diastematomyelia should involve X-ray examination [10,11]. Segmental vertebral abnormalities, hemivertebrae, spinal canal dilatation, and midline spurs can be visualized. Although MRI has been suggested to be the best imaging modality, we were unable to visualize our case using 1.5-tesla MRI because of technical difficulties. Spiral CT scan with three-dimensional reconstruction is useful postnatally, and can be performed in selected cases for precise visualization of the spinal anatomy [10]. Seventy percent of the cases of diastematomyelia diagnosed prenatally and reported in the literature were terminated [11].

In the past, a diagnosis of diastematomyelia was frequently made in the neonatal period, prompted by the presence of skin abnormalities over the vertebral column. The neurologic features of diastematomyelia can worsen during the postpartum period as the body grows [11]. In cases with neurologic deficits, one-third have motor deficits. Other neurologic defects include sensorial deficits, pes talus, and vesico-sphincter dysfunction. Neurologic findings are frequently observed at birth [5].

In parallel with advances in ultrasonography, the likelihood of antenatal diagnosis has increased. The obstetric management of these cases, however, is controversial because of their rarity.

Sepulveda et al [12] analyzed 15 cases and identified isolated diastematomyelia in six cases. Out of these, one had serious spinal deformity with orthopedic problems, small feet, and short legs. Four had no obvious problems. One case underwent spinal reconstructive surgery.

It is difficult to counsel and inform parents regarding prenatally diagnosed cases of isolated diastematomyelia cases. Early spinal and orthopedic surgical interventions may be able to prevent neurologic deficits. However, pregnancy termination could be an option for diastematomyelia in the presence of other spinal or extremity deformities. Standard obstetric management should be adequate for families who wish to continue with the pregnancy.

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