

DIASEMATOMYELIA: PRE- AND POSTNATAL MULTIMODAL DIAGNOSTIC APPROACH

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Diastematomyelia is a relatively rare congenital abnormality presenting as a sagittal separation of the spinal cord. Although cases of diastematomyelia have been previously reported, fully documented approaches by both prenatal and postnatal diagnostic workup are rare in the literature. We present a fully studied case of diastematomyelia type I investigated by prenatal US and MRI and postnatal US, MRI and radiography.

Key-word: Diastematomyelia.

Diastematomyelia is a form of spinal dysraphism characterised by a cleft in the spinal cord. This abnormality may be diagnosed at first by the obstetric intrauterine US and is confirmed by prenatal MRI which also provides complementary information about the fetal nervous system. Postnatal diagnostic approach refines the diagnosis, providing additional information about skeletal and nervous lesions. We describe a fully documented case of diastematomyelia type I investigated using prenatal US and MRI and postnatal US, MRI and Radiography.

Case report

At the 25th week of an uncomplicated gestation the routine US raised the suspicion of spina bifida as it showed a spinal abnormality of the fetus consisting of widening of the spinal canal and an abnormal angulation of the spine. Four days later, a fetal MRI was performed on a 1.5 Tesla scanner. The examination acknowledged a split of the spinal cord at the level of lumbosacral junction. A thick septum which separated the spinal cord was present at the same level (Fig. 1, 2A). No brain abnormality was detected.

At birth examination a hairy patch at the level of the lower spine was observed. There was no motor abnormality of the extremities. Three months later an ultrasound examination, radiography of the spine and a MRI were performed. The X-Ray exam showed scoliosis of the dorso-lumbar region, multiple costovertebral malformations and widening of the spinal canal (Fig. 2B). The ultrasonography of the spine showed precisely the divided spinal cord and the thick septum (Fig. 3).

MRI confirmed the presence of diastematomyelia from the T12 to the L5 level with a thick band separating the two hemicords (Fig. 4, 5). Several abnormalities of the vertebrae and agenesis of the sacrum were also detected



Fig. 1. — Prenatal sagittal T2 weighted magnetic resonance image showing the thick septum (arrow) which divides the spinal cord at the lower lumbar spine region.

Discussion

Diastematomyelia is a rare form of spinal dysraphism characterised by a cleft in the spinal cord. As a result is the separation of the spinal cord in a sagittal direction. There are two types of diastematomyelia. Type 1 consists of two hemicords each being surrounded by a separate dural sac and divided by a bony-cartilaginous septum. Type 2 consists of a single dural sac which contains

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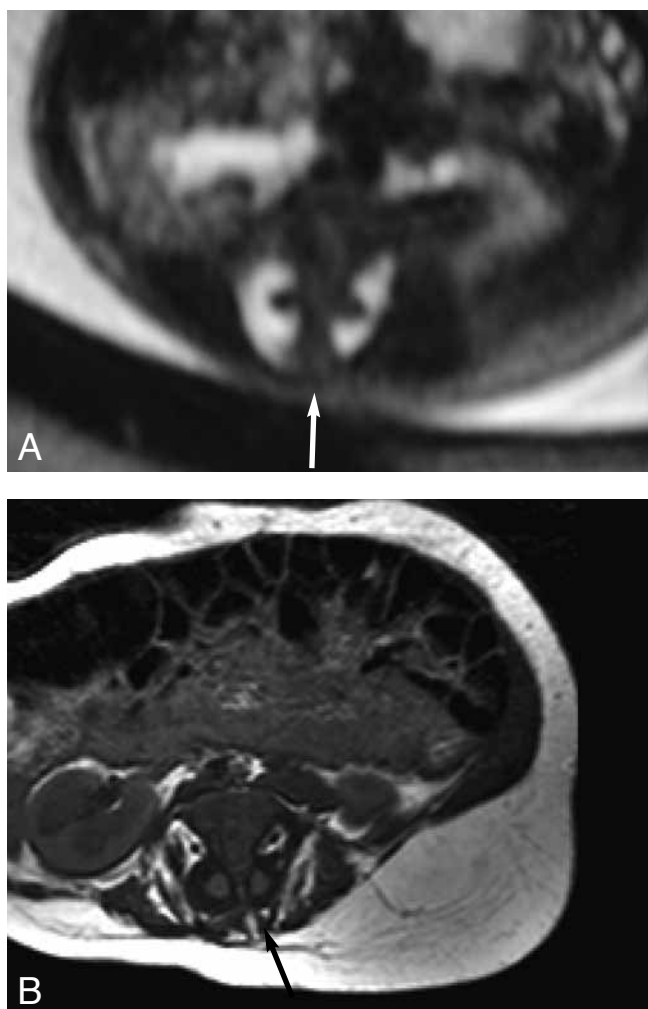


Fig. 2. — Prenatal axial T2 (A) and postnatal axial T1 (B) weighted magnetic resonance image (lower lumbar spine level) (arrow) showing a thick septum dividing the spinal cord.

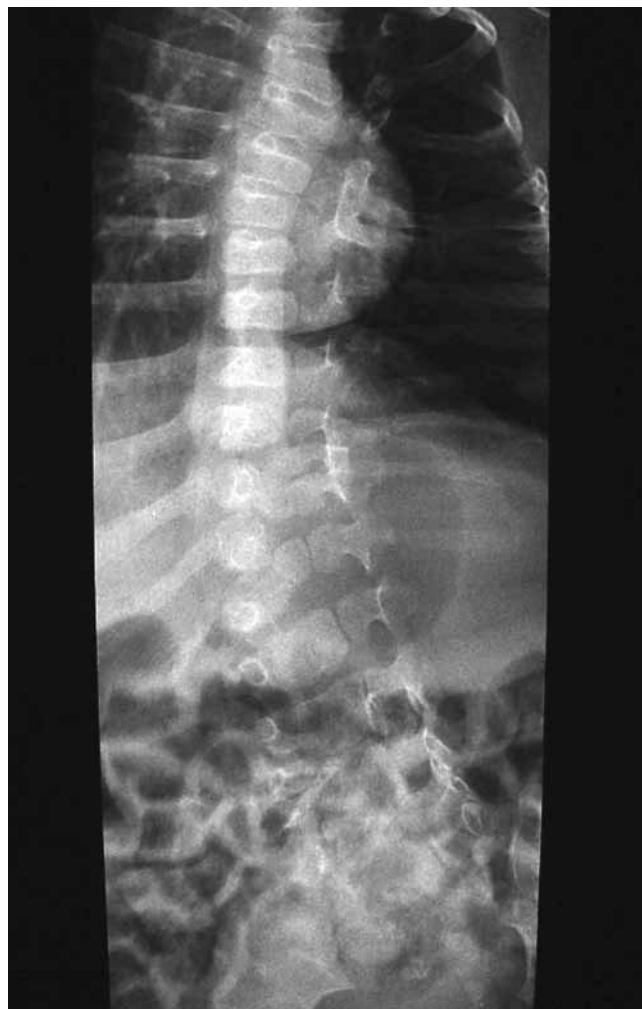


Fig. 3. — Radiography of the spine which demonstrates multiple vertebral defects at lower dorsal and lumbar level: hemivertebrae, costovertebral anomalies, widening of the spinal canal and scoliosis.

both of the hemicords. The two hemicords are separated by a non-rigid septum (1).

The first evidence of diastematomyelia is usually described during the intrauterine life by the routine ultrasound. The two main sonographic signs are the widening of the spinal canal in coronal view and the observation of an additional echogenic focus in the posterior part of the spinal column in the axial view (2, 3). The magnetic resonance confirms the diagnosis and sometimes provides important additional information regarding the fetal nervous system although there are some limitations at the description of the exact bone anatomy (4). This is the main reason for which some centers have described the use of intrauterine 3D CT for the diagnosis of fetal anomalies (5-8).

This dysraphism is an abnormal development of the notochord

between the 15th and 18th week of intrauterine life (9). All the reported individuals, including our patient, are females. This may indicate an X-linked inheritance (10, 11).

The signs of diastematomyelia may appear at any time of life. Diastematomyelia has been diagnosed in adult patients with progressive sensimotor symptoms (12, 13). Nowadays, thanks to organized prenatal control in addition to the advanced imaging techniques, diastematomyelia is usually diagnosed during intrauterine life or childhood. Patients with diastematomyelia may be asymptomatic (10) or present symptoms, usually progressive.

The symptoms of diastematomyelia are classified in 4 categories: cutaneous (hypertrichosis, pilonidal cyst, lipoma, naevus, angioma, meningocele and fistula), orthopedic (feet deformities, scoliosis, and asymmetric limbs), neurological and

radiological. The neurological signs are usually progressive, appear late and are non-specific. The slowly progressive neurological damage is explained by the injury of the spinal cord resulting from tethering or traction (14). It is estimated that diastematomyelia is responsible for 5% of congenital scoliosis (15).

The treatment of diastematomyelia necessitates the decompression of neural elements and removal of bony spur. This may be accomplished with or without resection and repair of the duplicated dural sacs (16). Surgical treatment is recommended in patients with progressive neurological symptoms. The reduction of the associated scoliosis in these patients is a complicated and ambiguous issue.

The prognosis of diastematomyelia depends on the associated anomalies (congenital scoliosis, kyphosis, spina bifida, myelomeningocele).

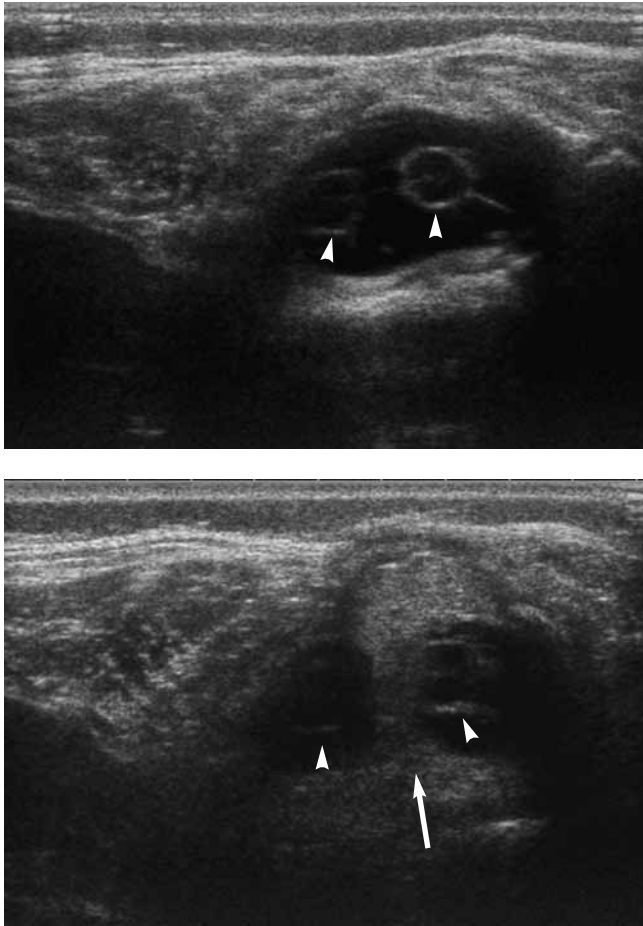


Fig. 4. — Post-natal ultrasound of the spine demonstrates a thick septum (arrow) which divides the spinal canal in two hemi-cords (arrowheads) and confirms the non-bony nature of this structure.

gocoele, hemivertebra). The neurological function may be improved by early surgical removal of the septum. Anderson et al support the opinion that the outcome is excellent in the isolated forms of diastematomyelia (2).

Conclusion

Diastematomyelia is a rare but potentially serious congenital abnormality of the spinal cord. Prenatal and postnatal multimodal imaging is necessary for the early diagnosis and accurate characterization of this complex abnormality.

References

1. Pang D.: Split cord malformation: Part II: Clinical syndrome. *Neurosurgery*, 1992, 31: 481-500.
2. Anderson N.G., Jordan S., MacFarlane M.R., Lovell-Smith M.: Diastematomyelia: diagnosis by pre-

- natal sonography. *AJR*, 1994, 163: 911-914.
3. Boulot P., Ferran J.L., Charlier C., et al.: Prenatal diagnosis of diastematomyelia. *Pediatr Radiol*, 1993, 23: 67-68.
4. Yuh W.T., Nguyen H.D., Fisher D.J., et al.: MR of fetal central nervous system abnormalities. *AJNR Am J Neuroradiol*, 1994, 15: 459-464.
5. Harwood-Nash D.C., McHugh K.: Diastematomyelia in 172 children: the impact of modern neuroradiology. *Pediatr Neurosurg*, 1990, 16: 247-251.
6. Skalej M., Duffner F., Stefanou A., Petersen D.: 3D spiral CT imaging of bone anomalies in a case of diastematomyelia. *Eur J Radiol*, 1999, 29: 262-265.
7. Jaspan T., Worthington B.S., Holland I.M.: A comparative study of magnetic resonance imaging and computed tomography-assisted myelography in spinal dysraphism. *Br J Radiol*, 1988, 61: 445-453.
8. Ruano R., Molho M., Roume J., Ville Y.: Prenatal diagnosis of fetal skeletal dysplasias by combining



Fig. 5. — Postnatal coronal T2 weighted magnetic resonance image showing the division of the spinal cord—already visible at the dorsal level (A) - above the septum (B). The dural sac appears divided below the level of the spur (C).

two-dimensional and three-dimensional ultrasound and intrauterine three-dimensional helical computer tomography. *Ultrasound Obstet Gynecol*, 2004, 24: 134-140.

9. Duczowska A., Bekiesinska-Figatowska M., Herman-Sucharska I., et al.: Magnetic resonance imaging in the evaluation of the fetal spinal canal contents. *Brain Dev*.
10. Kapsalakis Z.: Diastematomyelia in Two Sisters. *J Neurosurg*, 1964, 21: 66-67.
11. Balci S., Caglar K., Eryilmaz M.: Diastematomyelia in two sisters. *Am J Med Genet*, 1999, 86: 180-182.
12. Kramer J.L., Dvorak M., Curt A.: Thoracic disc herniation in a patient with tethered cord and lumbar syringomyelia and diastematomyelia: magnetic resonance imaging and neurophysiological findings. *Spine (Phila Pa, 1976)*, 2009, 34: E484-487.
13. Porensky P., Muro K., Ganju A.: Adult presentation of spinal dysraphism and tandem diastematomyelia. *Spine J*, 2007, 7: 622-626.
14. Hensinger R.N., Jones E.T.: Developmental orthopaedics. II: The spine, trauma and infection. *Dev Med Child Neurol*, 1982, 24: 202-218.
15. Barkovich A.J.: Congenital anomalies of the spine. In: Barkovich A.J., ed. *Pediatric neuroimaging*. Lippincott Williams & Wilkins, 2000.
16. Miller A., Guille J.T., Bowen J.R.: Evaluation and treatment of diastematomyelia. *J Bone Joint Surg Am*, 1993, 75: 1308-1317.