Antenatal and postnatal imaging of a double diastematomyelia without other malformation

Published on 28.10.2015

DOI: 10.1594/EURORAD/CASE.13043
ISSN: 1563-4086
Section: Paediatric radiology
Area of Interest: Paediatric
Procedure: Screening
Procedure: Diagnostic procedure
Imaging Technique: Ultrasound
Imaging Technique: MR
Special Focus: Congenital Case Type: Clinical Cases
Authors: Habre Céline, Toso Seema, Hanquinet Sylviane
Patient: 2 days, female

Clinical History:

Female newborn at term presented for postnatal imaging of a thoracic diastematomyelia with bony septum revealed by routine fetal ultrasound at 28 weeks gestation. Neurological examination was unremarkable and the baby had no cutaneous dysraphism markers.

Imaging Findings:

Prenatal ultrasound at 28 weeks gestation demonstrated a hyperechogenic focus with acoustic shadowing in the thoracic spinal cord within two hemicords and intact overlying soft tissues. At birth, ultrasound showed two hemicords from T10 to L2 with a bone septum in between. A second enlargement of the spinal cord with partial double spinal cord was visible at the level of the conus medullaris, which ended at L3. There was no cord tethering or intradural mass and real-time scanning showed adequate motion of nerve roots. Additional findings included a 3 mm large syrinx extending from T5 to T7. Postnatal MRI obtained at 2 weeks confirmed the presence of a bony spur at level T12, dividing the spinal canal in two compartments each containing a hemicord from level T10 and joining at level L2. MRI did not identify the more caudal incomplete double hemicords.

Discussion:

Diastematomyelia results from a persistent endomesenchymal tract connecting the endoderm and ectoderm, with consequent splitting of the notochord and the overlying folding neural plate and formation of two separate neural tubes and hemicords. [1] In type I diastematomyelia, both hemicords are enclosed in distinct dural sacs separated by extradural cartilage and bony spur in between. In type 2 diastematomyelia, there is no dural compartmentalization of the spinal cord and the two hemicords lie in the same dural sac with a partial or total fibrous midline septum.

Thorough examination for associated dysraphism abnormalities and concurrent cord tethering is necessary since early surgery can decrease secondary neurologic deterioration, including worsening scoliosis, lower limb deformities and sphincter dysfunction. [2] Surgical management consists of untethering the hemicords by resecting the bony spur and the fibrous septum, dural plasty and conus medullaris release, in order to enable longitudinal growth of the spinal cord in the growing spine. [2]

Classic antenatal ultrasound findings of type I diastematomyelia include widening of the spinal canal in the coronal...
view and echogenic focus traversing the spinal column in the axial view. [3] Fetal MRI does not provide more information to the diagnosis unless mother abdominal habitus, oligohydramnios and posteriorly facing fetal spine limit screening. [4, 5]

Postnatal ultrasound confirmed the finding of type I thoracic diastematomyelia and further revealed a lower type II diastematomyelia and a more rostral syrinx, thereby being part of the rare cases of composite-type diastematomyelia previously described [1, 6-8] and illustrating the common reported association of diastematomyelia and syringomyelia. [9, 10]

Noteworthy, only early postnatal ultrasound readily depicted the lower type II diastematomyelia, supporting the use of high resolution ultrasound for scanning of the newborn spinal cord during the window of non-ossified cartilaginous posterior processes. [11] In our patient, postnatal MRI was less accurate than ultrasound. It was unable to identify the additional lower site of cord tethering resulting from a less rigid but potentially harmful fibrous band that could have threatened the surgery outcome of the upper lesion. Furthermore, it did not add any contribution to the diagnosis made by ultrasound workup.

Our case illustrates a diastematomyelia diagnostic on antenatal ultrasound and the importance of postnatal ultrasound to achieve as much as possible just after birth. Postnatal MRI should be reserved for pre-operative assessment of the dysraphism.

**Differential Diagnosis List:** Type I and type II diastematomyelia with associated syringomyelia., Closed spinal dysraphism, Tethered cord syndrome

**Final Diagnosis:** Type I and type II diastematomyelia with associated syringomyelia.

**References:**


Figure 1

**Description:** Fetal ultrasound sagittal view demonstrating a hyperechogenic focus with acoustic shadowing at the level of the caudal thoracic spine. **Origin:** Romaine Robyr, Hôpital de la Tour, Geneva, Switzerland
Description: Axial view at level T5-T7 of the thoracic spine demonstrating a syrinx. Origin: Hôpitaux Universitaires de Genève (HUG), Geneva, Switzerland
Description: Axial view at level T10-L2 of the dorso-lumbar junction demonstrating two hemicords separated by hyperechogenic spur with acoustic shadowing. **Origin:** Hôpitaux Universitaires de Genève (HUG), Geneva, Switzerland
Description: Axial view of the spine at level of the conus medullaris demonstrating an incomplete split of the spinal cord with two ependymal canals. Origin: Hôpitaux Universitaires de Genève (HUG), Geneva, Switzerland
Figure 3

Description: Axial ultrasound in the supine position and axial T2-weighted TSE MR demonstrating two hemicords in separate dural sacs with an incomplete bony spur emerging between the hemicords from the posterior wall of the vertebra. Origin: Hôpitaux Universitaires de Genève (HUG), Geneva, Switzerland