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Review article

Fetal central nervous system malformations on MR images

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Abstract

Sonography is the method of choice for prenatal malformation screening but it does not always provide sufficient information for correct diagnosis or adequate abnormality evaluation. Fetal magnetic resonance imaging (MRI) is considered as a valuable second line imaging tool for confirmation, completion and correction of sonographic findings. Fetal MRI has proven its value in the evaluation of central nervous system pathologies, especially of midline and posterior fossa malformations. The role of MRI is not only to confirm or exclude possible lesions but also to define their full extent, aiding in their characterization, and to demonstrate associated abnormalities. The authors describe the most common anomalies of CNS revealed by fetal MRI in a chronological way related to the age of pregnancy, with a review of own MR images and with reference to the literature and own experience.

Keywords: Fetal MRI; Central nervous system; Malformations

1. Introduction

The frequency of developmental defects ranges from 2% to 3% of all foetuses. Thirty percentages of cases of the abnormal fetal development are related to genetic disorders. Congenital anomalies are the most severe disorders of the central nervous system. They account for 40% of deaths of all children in the first year of life and in survivors they cause a variety of neurological disorders, mental retardation or drug-resistant epilepsy.

Sonography is the most important imaging method for prenatal malformation screening. US examination is noninvasive, widely available, and safe for both mother and child. Its usefulness in evaluating fetal malformations does however depend on the examiner's experience and equipment quality that significantly influence the sensitivity of this method in detecting pathologies. In addition, this modality is much less useful, regardless of equipment quality and experience of the examiner, in case of oligohydramnios, maternal obesity and complex abnormalities – in particular, when it is performed in late pregnancy. With a passive system of screening, US allows to detect severe anomalies in 2–3% of newborns. In active systems (specially trained physicians perform fetal US according to the standardized protocol), sonography reveals defects in 7.3% of newborns.

As MRI is the best known method for imaging brain and spine structures, for several years attempts have been made to introduce MRI in fetal screening. The first publication concerning fetal examinations dates back to 1983 [1]. At the beginning, long time of acquisition made it impossible to utilize MRI for fetal imaging. Implementation of fast and ultra-fast sequences allowed to reduce acquisition time to 1 s, which subsequently eliminated artefacts resulting from fetal movement and allowed to obtain good quality images [2]. MRI examination is noninvasive. Initially, negative influence of magnetic field on developing embryo and fetus was apprehended. Ten-year observation of women employed in MRI units who, unaware of pregnancy, had been

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working during the first trimester, allowed to exclude high gradient magnetic field influence on increased number of miscarriages. MRI was then introduced to fetal screening: at first mainly in cases of suspicion of the central nervous system pathologies and afterwards also in anomalies of other systems and organs.

2. Fetal MR imaging – methods and contraindications

Fetal MR imaging is usually performed using 1.5 T static magnetic fields. Fast T2-weighted sequences are used at most times [3]. One of them is the fast spin-echo sequence that allows for one layer acquisition in about 1 s, which decreases artefacts related to fetal movement. The signal of fluid (including amniotic fluid, cerebrospinal fluid and other organ fluids) is very high (bright) allowing obtaining good contrast between tissues and excellent, detailed visualization of fetal anatomy [4–6]. The imaging involves scanning the fetus in orthogonal axial, sagittal, and coronal planes, variable in relation to mother's body depending on how the fetus is lying [7]. Standard slice thickness varies with centre from 3 to 8 mm and interslice gap from 1 to 2 mm. Fetal MRI scan is usually performed without intravenous contrast agent administration to the mother, because detection of the most commonly observed pathologies does not require the use of contrast medium. Furthermore, gadolinium chelates cross placenta into fetal circulation system.

Examination is performed with the patient lying down on her back, and in 3rd trimester – to avoid compression of the inferior vena cava – oblique position on the left side may be used. The time of examination varies from 20 to 40 min depending on the number and complexity of the observed anomalies. Contraindications for MRI include claustrophobia, cardiac pacemaker, ferromagnetic hemostatic clips on or in direct neighbourhood of cerebral vessels, metallic splinters in the orbit or in any other vital organ, nerve stimulator, metal or electronic ear implants and miniature subepidermal hearing aid.

3. Indications for fetal MRI

The leading indication to perform fetal MRI is the suspicion of central nervous system abnormalities [8,9]. CNS malformations are observed in about 3% of all newborn babies. Majority of them is detected in US examination. Almost all publications dealing with CSN developmental abnormalities stated MR predominance over US [10,11], especially in the assessment of posterior cranial fossa defects, corpus callosum malformations, in the evaluation of complex brain and spine anomalies, detecting causes for cerebral fluid spaces dilatation, brain and spine tumors, defining cortex maturation and the extent of cortical dysplasia and anomalies related to improper neuronal migration, ischemic lesions, intracerebral hemorrhage and in the evaluation of brain and spine hernias' contents [12–14]. Therefore MRI is necessary in order to detect these fetuses, in whom corrective neurosurgery could be performed.

4. Classification of CNS abnormalities

Central nervous system development is a complex process which may be disrupted by many extrinsic factors, genetical determination or genetic load manifestation evoked by the influence of an extrinsic factor. Depending on the time of occurrence, length and intensity of the exposure to nociceptive agent various morphological lesions occur. On the basis of successive phases of CNS formation and maturation, the classification of CNS abnormalities has been introduced [15]. In this paper the authors present this classification related to the age of pregnancy with a review of own MR images of the selected defects.

4.1. Dorsal induction abnormalities

During the primary and secondary neurulation period (the first 4–5 weeks of gestation) neural tube, notochord, spine, and cranium are formed. Malformations occurring in this period are named the dorsal induction abnormalities. Among them dysraphic disorders of brain and skull and dysraphic defects of spine and spinal column may be distinguished. The most frequently observed anomalies of this group include: anencephalia, meningocele, encephalo-meningocele, Chiari malformations (I–III), myelomeningocele, myelocele, hydromyelia, hydromyelic cystocele, lipomeningomyelocele, epi-/subdural lipoma, dermoid sinus, dermoid and epidermoid cyst, caudal developmental disorders, anterior dysraphies – neurenteric cyst, caudal regression disorders (caudal regression syndrome) [16,17].

Anencephaly (Fig. 1) is a lethal brain developmental abnormality resulting in unevolved hemispheres or their complete absence, commonly occurring together with acrania–cranium undevelopment. In some cases only the bones of cranial roof are absent and bones of skull base show structural defects. In about half cases anencephaly is associated with rachischisis. This defect is caused by failure of the neural tube to close in the rostral region. Total anencephalia is caused by absence of induction or by neural plate damage between 2nd and 3rd week of gestational age. This leads to absence of all three primary brain vesicles. Absence of the hindbrain explains incapability of the fetus to survive. Osseous cranium roof and optic vesicles fail to develop (anopia). Partial anencephaly develops between 3rd and 4th week of gestation. Optic vesicles situated on lateral sides of the head may already be present. Third vesicle, which forms the hindbrain, is also formed.