Usefulness of magnetic resonance imaging in the prenatal study of intracranial and facial malformations.

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Learning objectives

Magnetic resonance imaging is a complementary test to ultrasound for intrauterine fetal study.

We propose to attend these objectives:

- Describe the usefulness of fetal MRI in the prenatal diagnosis of intracranial and facial malformations.

- Show the most common abnormalities diagnosed in our center.

Background

Ultrasound (U.S) evaluation of some fetal anomalies provides limited information, even in experienced hands. In such cases, fetal MRI is the excellent imaging modality to confirm or rule doubtful ultrasound findings. Also it can reveal others abnormalities undetected by ultrasound with an evident influencing in the gestational management.

At present there is no evidence to suggest that exposure to electromagnetic fields for short periods have harmful effects on the fetus and the mother, however, and because of the potential harmful effects unknown it is not advised to perform MRI studies in the first trimester, while it might have some clear indication of all to study maternal abnormalities.

There are some circumstances with increased risk of birth defects which could be studied with MRI, such as: high maternal age, maternal serum biochemical screening high risk for Down syndrome, intrauterine growth retardation, polyhydramnios or oligohydramnios, rhythm disturbances fetal heart, previous child with chromosomal abnormalities, chromosomal abnormalities parent carrier, X-linked, first degree family history of birth defect, ingestion of certain drugs during pregnancy, exposure to ionizing radiation during pregnancy and exposure to an infection disease and the monochorial twin pregnancy although ultrasound does not show abnormalities.

A number of MRI scans performed to evaluate possible abnormalities of the central nervous system, face or spine. The U.S. may have difficulty in the evaluation of intracranial structures when the fetus is in advanced pregnancy because of the
ossification of the cranial bones collinear or if the fetal head is engaged in the depth of the female pelvis. MRI produces images of good quality regardless of the circumstances.

The evaluation of the structures of the central nervous system, face, neck and spine by MRI is better the higher gestational age.

The establishment of normal fetal brain MRI is difficult because, for ethical reasons, it is impossible to RM normal fetuses.

T1-weighted sequences are useful for detecting brain hemorrhage, but T2-weighted sequences allow better assessment of fetal craniofacial anatomy and spine, showing the circumvolutions and patterns of neuronal migration and cerebrospinal fluid spaces.

Around week 20 of gestation, the cerebral ventricles and look good in any plane. Lower gestational ages its valuation is more difficult, except when they are dilated. An increased the transverse diameter of the atrium of the lateral ventricle above 10 mm, measured at the posterior margin of the choroid plexus through the thalamus, after week 25 is considered pathological. There is a physiological ventricular relative increase until week 25, which persists in the occipital horns until week 30. The ventricles III and IV are often viewed without difficulty by RM.

Ventriculomegaly is the most frequent anomaly of the central nervous system. It is associated with other anomalies (structural or chromosomal) in 70-84% of cases would include: neural tube defects, agenesis of the corpus callosum, Dandy Walker syndrome, holoprosencephaly, cortical malformations, intracranial hemorrhage and porencephaly. MRI is useful in determining the degree and cause of ventriculomegaly and discard additional structural anomalies.

The analysis of the cerebral sulci required to learn patterns of brain maturation. According to gestational age and MRI findings can be compared with the anatomical atlas.

The Silvio sulci is visible at week 18 and at 23, seems more angular, for the formation of the seal.

The parieto-occipital and calcarine fissures are about 18-19 week, the cingulate sulcus at week 24 and the center on the 26.
A delay in the maturation indicates abnormal development.

The corpus callosum is formed between weeks 8 and 20, for the development of axons that form the interhemispheric fissures. Abnormalities of the corpus callosum are quite common.

This structure, discretely hypointense relative to white matter is relatively difficult to assess. Abnormalities of the corpus callosum were mainly detected by morphological alterations of the ventricular system and cerebral sulci typical of this entity.

The fetal cerebellum is displayed well in any gestational age and is relatively small compared to the brain.

The fetal subarachnoid space is more prominent than in the neonate.

The brain stem and spinal cord of the rest are more difficult to assess, independent of gestational age.

The palate is identified as a hypointense linear structure in T2-weighted sequences, which separates the oral and nasal cavities. The sagittal and coronal planes are the best to watch it. The axial is the best plane to study the maxillary and mandibular.

The eyes are observed in all 3 planes of space, like two round structures hyperintense on T2. The crystalline can be identified, in the anterior part, as an ovoid and hypointense structure on T2.

**Facial anomalies.**

**Cleft lip and cleft palate.**

They are the most common congenital anomalies of the face. They are more frequent in male fetuses. The cause is multifactorial, and is related to environmental factors (intrauterine infections, such as rubella; toxins such as alcohol, snuff, drugs and medicines) and genetic (chromosomal syndromes). The risk of other malformations in the case of isolated cleft lip is 8%, if is associated to the cleft palate, the risk increases to 21%.
Cleft lip and cleft palate are 2 different entities embryologically. There are different combinations of these anomalies.

**Cleft lip.**

Cleft lip or cleft primary palate (anterior segment of the hard palate, ahead of the incisive foramen) are the result of the absence of fusion of one or both nasal processes with the maxillary prominences, a fact that occurs between the fourth and sixth weeks of gestation.

It may affect only the soft tissues of the lip (incomplete cleft lip) or extend deep and affect the primary palate (complete cleft lip) but without affecting the secondary. This defect may be unilateral or bilateral. The isolated unilateral cleft lip has very good prognosis and is rarely associated with other malformations. Coronal MR images are useful to demonstrate the cleft lip and distortion of the nose, a finding that guides about a defect of the primary palate. The axial plane shows the loss of continuity of the maxillary.

**Cleft palate.**

Cleft palate or cleft in the secondary palate (posterior segment of the hard palate) is a consequence of the lack of fusion of the palatal shelves, between the eighth and twelfth weeks of gestation. MRI shows this lesion in the midsagittal plane, also can be observed in the coronal plane the direct communication between the oral cavity and the nostrils.

The isolated cleft of the secondary palate without cleft lip is a rare anomaly that is usually secondary to processes which prevent a correct development of the palate such as: tumors, macroglossia or micro / retrognathia

**Central cleft lip and cleft palate.**

This anomaly is often associated with chromosomal abnormalities. Cleft palate is identified in the mid-sagittal image and the central cleft lip in the coronal and axial planes.

**Facial cleft**

Associated a defect of the soft tissues and the bones facial.
**Micrognathia / retrognathia**

Micrognathia is the mandibular hypoplasia and retrognathia is the posterior location of a normal sized jaw, both disorders can be associated.

These changes tend to displace the tongue posterior and superiorly, interfering with the normal development of the hard palate, so it's not uncommon the association with cleft palate. The midsagittal plane is the best to identify this anomaly in MRI studies.

The characteristic feature is the protrusion of the upper lip relative to the lower in the midsagittal image, due to a small jaw in posterior position. The alteration in the jaw can hamper fetal swallowing causing polyhydramnios and a stomach in low repletion.

**Oculo -auriculo- vertebral spectrum.**

It consists of a group of craniofacial anomalies of multifactorial cause. The clinical spectrum is broad, ranging from mild forms with only with preauricular appendices to more severe forms such as Goldenhar syndrome (hemifacial microsomia, vertebral anomalies and epibulbar dermoid).

The hemifacial microsomia is the second most common craniofacial anomaly after cleft lip. It is characterized by facial asymmetry and mandibular hypoplasia, usually unilateral.

**Ocular abnormalities.**

The anophthalmia is a complete absence of the eyeball with integrity of the eyelids, conjunctiva and lacrimal apparatus. It may be primary or secondary. The primary form is usually associated with chromosomal abnormalities or genetic disorders and the secondary may be due to infections (rubella), toxic-metabolic, or vascular / ischemic injury.

The anophthalmia can be diagnosed without difficulty by RM also we can evaluate the crystalline and the vitreous humor, allowing the study of entities as persistent hyaloid artery and congenital cataract.

**Tumors**
Fetal intracranial tumors are rare and account for 0.5-1.9% of all pediatric tumors. The most common types found are teratomas, with 25-50% of all cases, followed by primitive neuroepithelial tumors, mesenchymal tumors, craniopharyngiomas and hemangioblastomas.

The prenatal tumors are usually supratentorial unlike the tumors that are detected during late childhood which are infratentorial.

In fetuses with prenatal diagnosis of tumor we should look for signs of heart failure, hydrops, enlarged liver, extramedullary hematopoiesis, polyhydramnios, and hydropic placenta. The differential diagnosis of intracranial tumors includes vascular malformations of the brain, infection and bleeding. Calcifications are best detected by CT scan and MRI better defines the extent of the tumor, especially if the mass is located in the posterior fossa.

Images for this section:
Fig. 1

Bilateral ventriculomegaly. Colpocephaly.

Fig. 2
Ventriculomegaly, porencephalic cyst.
Volume loss of the left temporal and parietal lobes.
➢ Possible hypoxic-ischemic encephalopathy or sequelae of infection.
Cyst of the septum pellucidum

Fig. 4
Fig. 5

Agenesis of corpus callosum

Colpocephaly, extension of the interhemispheric fissure to the roof of the third ventricle.
Fig. 6

Cystic enlargement of the posterior fossa with dilatation of the fourth ventricle and dorsal rotation of the vermis with elevation of the tentorium, also we can see the hypogenesis of right cerebellar hemisphere.
Fig. 7
Increased extracerebral subarachnoid space and moderate ventriculomegaly in fetus of 34 weeks' gestation.
Polyhydramnios, alteration of sulcation with incomplete lissencephaly, pachygyria.
Fig. 9

Unilateral cleft lip without cleft palate
Complete unilateral cleft lip, affectation of primary and secondary palate

Fig. 10
Findings and procedure details

Fetal MR imaging is routinely performed on 1.5-T Siemens scanners.

Patient supine, entering feet first.

The left lateral decubitus position is optional for advanced stages of pregnancy.

We manage sublingual 5mgr Valium 15 minutes before the test.

An eight-channel torso phased-array coil is used to allow increased coverage of the fetal head and an increased signal-to-noise ratio over more standard pelvic phased-array coils.

MR imaging is performed primarily with ultrafast MR imaging, routinely techniques HASTE (Siemens), in some cases TRUE FISP (Siemens)

Using these techniques, a single T2-weighted image can be acquired in less than 1 second, decreasing sensitivity to fetal motion. Typically an initial localizer is obtained in three orthogonal planes with respect to the mother using 6- to 8-mm thick HASTE T2-weighted slices with a 1- to 2-mm gap and a large field of view.

The localizer is useful for visualizing the position of the fetus and determining fetal sidedness.

We also use this localizer to ensure that maximal signal is obtained from the area of interest.

For imaging the fetal brain, HASTE T2-weighted images are obtained. using a slice thickness of 3 mm with no gap. For the fetal spine, a 2-mm slice thickness is use.

The most common indication in the intracranial abnormality group for taking fetal MRI was ventriculomegaly. The congenital anomalies of the face most frequently identified are the cleft lip and the cleft palate and different combinations of these anomalies.
Conclusion

The magnetic resonance is helpful when ultrasound findings are inconclusive or insufficient for choosing proper management and prenatal counseling, for that reason is essential an adequate knowledge of the MRI features of the different fetal malformations.

Personal information

References


