Heterotopias: classification and differential diagnosis in pediatric MRI

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Learning objectives

To check the main types of gray matter heterotopia, common malformations of cortical development characterized by interruption of normal neuronal migration from near the ventricle to the cortex ("normal neurons in abnormal locations"), and to make an accurate description of their differential characteristics in paediatric MRI.
Background

Grey matter heterotopias are collections of nerve cells in abnormal locations secondary to arrest of radial migration of neurons. During the seventh week of gestation, a proliferation of young neurons occurs in the subependymal layer of the walls of the lateral ventricles, known as the germinal matrix or germinal zone. Most of the neurons that will form the cerebral cortex migrate to their destinations along specialized radial glial cells that span the entire thickness of the hemispheres from the ventricular surface to the pia (Fig. 1). Any abnormality that inhibits neuronal or glial proliferation, neuronal migration, or subsequent cortical organization can result in a cortical malformation.

Gray matter heterotopias are common malformations of cortical development and can be isolated or can be seen in association with other structural anomalies (agenesis of the corpus callosum, aqueductal stenosis, microcephaly and schizencephaly). They are characterized as a type of cortical dysplasia and cause a variety of symptoms, but usually include some degree of epilepsy or recurring seizures. MRI is the modality of choice in assessing heterotopic grey matter due to its as yet unsurpassed contrast resolution, not only in suspected cases, but also in prenatal stage, being more effective than ultrasound.
Fig. 1: Schematic drawing of neuronal migration in the developing cerebral cortex. Normal neuronal migration requires proteins that regulate and help to reach their final destination.

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Imaging findings OR Procedure details

Heterotopias are generally divided into three groups, depending on the location of the ectopic formations: **Subependymal (Figs. 2 and Fig. 3):** large number of heterotopic nodules that completely or nearly completely line the walls of the lateral ventricles, specially the trigones and the temporal and occipital horns. Patients usually present with a seizure disorder in the second or third decade of life. On MRI subependymal heterotopias are seen as small nodules immediately deep to the ependymal layer, isointense to grey matter. The nodules follow grey matter on all sequences including on post contrast sequences where they do not enhance. X-linked forms have a high incidence of associated brain abnormalities as tuberous sclerosis (but theses nodules enhance on post contrast sequence), or subependymal metastases.

**Focal subcortical:** distinct nodes in the white matter, "focal" indicating specific area. Ectopic nodules extend from the cortex into the underlying white matter (Figs. 4 and 5) or from the ventricle into the white matter (Figs. 6 and 7). Subcortical heterotopias are frequently accompanied by other structural abnormalities and frequently patients present fixed neurologic deficits and develop partial epilepsy during the first decade. On MRI focal nodes are seen as isointense to gray matter lesions often containing blood vessels and CSF. They are always contiguous with the overlying cortex and the underlying ventricular surface. The cortex itself often suffers from an absence of gray matter and may be unusually thin or lack deep sulci, including an overall decrease in cortical mass.

**Laminar or band heterotopia (double cortex) (Figs. 8 and 9):** is a form of diffuse heterotopia, resulting in heterotopic grey matter, deep to the cortex, but the gray matter is more diffuse and is symmetric between the hemispheres. On MRI band heterotopia consist of smooth layers of gray matter that often follow the curvature of the overlying cortex. They are not convoluted nor are they contiguous with the overlying cortex and do not contain blood vessels or CSF.

**Differential diagnosis** of neuroglial heterotopias include not only **tuberous sclerosis (Fig. 10),** but also the **lissencephaly-pachygyria spectrum (Fig. 11 y Fig. 12),** a group of diseases that cause relative smoothness of the brain surface and includes agyria (no gyri), pachygyria (broad gyri) and lissencephaly (smooth brain surface). **Polymicrogyria (Fig. 13)** is an abnormal development of the deeper layers of the cerebral cortex and the formation of multiples small gyri, thus, is not a real migration disorder, it is a disorder of neuronal organization. The most common location is around the Sylvian fissure, particularly the posterior aspect of the fissure; however any area can be affected. The numerous small gyri that lend their name to the condition are very small and only seen on thin section high resolution MRI. Polymicrogyric cortex usually has signal characteristics similar to normal grey matter. **Schizencephaly (Figs. 14 and 15)** is the
term used to describe gray matter lined clefts that extend through the entire hemisphere from the ependymal lining of the lateral ventricles to the pial covering of the cortex. Patients with schizencephaly typically present with seizures, haemiparesis, and variable developmental delay. The severity of the symptoms is related to the amount of involved brain. And finally hemimegalencephaly (Fig. 16), disorder that seems to be the result of abnormal stem cell proliferation. Affected hemispheres contain areas of pachygyria, polymicrogyria and heterotopia. On MRI part or all of a hemisphere may be affected, and typical features include: increased lateral ventricle size, shallow sulci, enlarged gyri, enlarged or thickened calvaria and contralateral displacement of the posterior falx.
Fig. 2: Figure 2: Subependymal Heterotopia. Axial T2-weighted MR image shows gray matter nodules located in the left periventricular region that protrude into the ventricular wall. This is the most common location for heterotopia.

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Fig. 3: Figure 3: Subependymal Heterotopia. Coronal T2-weighted MR image shows gray matter nodules located in the left periventricular region that protrude into the ventricular wall. This is the most common location for heterotopia.

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Fig. 4: Subcortical heterotopias are continuous with the overlying cortex or underlying ventricle, and they can be divided into nodular form (extend from the ventricle into the white matter) or curvilinear form (extend from the cortex into the underlying white matter).

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**Fig. 5:** Figure 5: Focal subcortical Heterotopia.

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**Fig. 6:** Figure 6: Axial inversion-recovery (IR) weighted MRI showing gray matter focal subcortical and bilateral periventricular heterotopia.

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**Fig. 7:** Figure 7: Coronal inversion-recovery (IR) weighted MRI showing gray matter focal subcortical and bilateral periventricular heterotopia.

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Fig. 8: Figure 8: Laminar or band heterotopia (double cortex). Neuronal migration disorder characterized by the presence of bands of grey matter located between the ventricular walls and the cerebral cortex, and separated from both by a layer of normal appearing white matter.

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Fig. 9: Figure 9: Laminar or band heterotopia (double cortex).

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Fig. 10: Figure 10: Tuberous Sclerosis. Hyperintense cortical and subcortical lesions represent white matter nodes or tubers.

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**Fig. 11:** Figure 11: Lissencephaly. Axial T1-weighted MRI shows generalized pachygyria that results in thick convolutions of the cerebral cortex.

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Fig. 12: Figure 12: Lissencephaly. Sagittal T1-weighted MRI.

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**Fig. 13:** Figure 13: Polymicrogyria. Cerebral cortical malformation characterized by excessive cortical folding and by shallow sulci. Bilateral symmetrical perisylvian polymicrogyria is the most frequent form.

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Fig. 14: Figure 14: Unilateral open lip schizencephaly. Axial MRI demonstrates a cleft that extends across the entire cerebral hemisphere, from the ventricular surface to the cortex.

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**Fig. 15:** Figure 15: Unilateral open lip schizencephaly. Coronal T2-weighted MRI demonstrates a cleft that extends across the entire cerebral hemisphere, from the ventricular surface to the cortex.

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Fig. 16: Figure 16: Hemimegalencephaly. Axial T2-weighted MRI shows pachygyria and enlargement of the right cerebral hemisphere.

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Conclusion

Gray matter heterotopia is a neurological disorder that causes a variety of symptoms, but usually includes some degree of epilepsy, recurring seizures and developmental delay. Detection and characterization of these structural malformations is important because in some cases symptoms may require neurosurgical treatment.
References


