Learning objectives

- To become familiar with Magnetic resonance imaging techniques and findings in children epilepsy.
- To be aware of the main etiologies of children epilepsy.
Background

We report a retrospective study that evaluates 86 cases of epilepsy: 55 boys and 31 girls with a mean age of 7 years (range, 8months -15 years) seen during four year period from January 2010 to December 2013.

Patients were admitted with variant clinical features: typical absences or petit mal seizures (n=40), infantile spasms (n=20), juvenile myoclonic epilepsy(n=14), benign Rolandic epilepsy (n=12).

OVERVIEW

- A seizure represents the clinical expression of abnormal, excessive, synchronous discharges of neurons residing primarily in the cerebral cortex. This abnormal paroxysmal activity is intermittent and usually self-limited, lasting seconds to a few minutes.
- Newborns and young children have seizures most frequently. Their seizures are often caused by problems around the time of birth, including injuries during birth or infections.
- Epilepsy is a condition in which a person is more susceptible to having unprovoked seizures. A child may have epilepsy if she has had two or more unprovoked seizures.

CLINICAL PATTERN

SYMPTOMS

A child may have a wide variety of symptoms including:

- staring
- stiffening of the body
- loss of consciousness
- breathing problems
- loss of bowel or bladder control
- not responding to noise or words for short periods of time
- appearing confused or in a haze
- extreme sleepiness and irritability when waking up in the morning
- periods of rapid eye blinking and staring

TYPE OF SEIZURES
The type of seizure depends on which part and how much of the brain is affected and what happens during the seizure. The two broad categories of epileptic seizures in children are Focal seizures and generalized seizures:

**Focal seizures.**

Focal seizures take place when abnormal electrical brain function occurs in one or more areas of one side of the brain. Two types of partial seizures include:

- **Simple focal seizures.** The child may show different symptoms depending upon which area of the brain is involved. The seizure activity is limited to an isolated muscle group, such as fingers or to larger muscles in the arms and legs. Consciousness is not lost in this type of seizure.

- **Complex focal seizures.** This type of seizure commonly occurs in the temporal lobe of the brain, the area of the brain that controls emotion and memory function. This seizure usually lasts one to two minutes. Consciousness is usually lost during these seizures.

**Generalized seizures.**

Generalized seizures involve both sides of the brain. There is loss of consciousness and a postictal state after the seizure occurs.

Types of generalized seizures include the following:

- **Absence seizures (also called petit mal seizures).** These seizures are characterized by a brief altered state of consciousness and staring episodes. Absence seizures almost always start between ages 4 to 12 years.

- **Atonic (also called drop attacks).** With atonic seizures, there is a sudden loss of muscle tone and the child may fall from a standing position or suddenly drop his or her head.

- **Generalized tonic-clonic seizures (also called grand mal seizures).** The classic form of this kind of seizure, which may not occur in every case, is characterized by five distinct phases. The body, arms, and legs will flex (contract), extend (straighten out), tremor (shake), a clonic period (contraction and relaxation of the muscles), followed by the postictal period. Not all of these phases may be seen with every one of this type of seizure.

- **Myoclonic seizures.** This type of seizure refers to quick movements or sudden jerking of a group of muscles.

- **Infantile spasms.** This rare type of seizure disorder occurs in infants from before six months of age. The infant usually has brief periods of movement of the neck, trunk, or legs that lasts for a few seconds.

- **Febrile seizures.** This type of seizure is associated with fever and is not epilepsy, although a fever may trigger a seizure in a child who has epilepsy.
These seizures are more commonly seen in children between 6 months and 5 years of age and there may be a family history of this type of seizure.

**DIAGNOSIS TOOLS**

**Brain imaging tools**

Magnetic resonance imaging is superior in identifying congenital or developmental abnormalities and should be performed in preference to CT. MRI has revolutionized the detection of structural abnormalities in pediatric patients with epilepsy. It is important to use the proper protocol to age.

Some considerations on the sequences we use in MRI in children:

- T2 FSE: demonstration of myelin in the brain premature;
- FLAIR: great contrast between white and gray matter, as well as assessment of myelination
- Under 18 months is not necessary to use the FLAIR sequence.
- Volumetric 3D T1 sequence: Important MR sequence to facilitate the study of several cortical defects. Obtained from three-dimensional acquisitions, which makes it possible multiplanar reconstructions
- Diffusion weight Images related to differences in molecular mobility or diffusivity of water between the tissues. Cerebral infarction and hypoxic ischemic encephalopathy.

Protocol recommended in pediatric patients with a history of epilepsy:

- T1 volumetric (slice thickness 1 mm or less)
- Coronal FLAIR 3mm thin slices
- TSE T2 axial and coronal
- DWI

**Other tests**

- **Blood tests** (such as blood sugar, complete blood count, electrolytes and liver and kidney function tests)

- **Electroencephalography(EEG)**, a test that records electrical activity in your child's brain using tiny wires attached to her head. The instruments are so sensitive that they pick up even small seizures that don't lead to physical symptoms.
- Lumbar Puncture to see if there is an infection or other problems
Findings and procedure details

Imaging and Spectrum of Etiologies in children epilepsy

Malformations of cortical development

Development of the cortex can be roughly separated into three steps: cell proliferation and differentiation, cell migration and cortical organization. Any abnormality during the process of cortical formation can lead to malformations of cortical development.

- Based on pertinent aspects of central nervous system embryology, the 2005 classification of MCD classifying them according to the developmental stage during which disruption is considered to be the cause of the anomaly. There are four groups of malformations.

Group I comprises disorders with decreased/increased proliferation or the proliferation of abnormal cells, and includes microcephaly, focal cortical dysplasia with balloon cells, hemimegalencephaly, tuberous sclerosis, dysembryoblastic neuroepithelial tumor and ganglioglioma/gangliocytoma.

Group II includes malformations due to abnormal neuronal migration, such as lissencephaly, dystroglycanopathy and heterotopia.

Group III comprises disorders due to abnormal cortical organization (including late neuronal migration), and also includes polymicrogyria, schizencephaly, cortical dysplasia without balloon cells and microdysgenesis.

Group IV includes all other MCD that are not otherwise classified, such as mitochondrial disorders and sublobar dysplasia.

Groupe I Abnormal cell proliferation

Focal cortical dysplasia (Fig. 1)
Focal cortical dysplasia is a congenital abnormality where the neurons fail to migrate in the proper formation in utero.

The most common findings are cortical or subcortical hyperintensities especially seen on FLAIR-images. These are often found at the bottom of a deep sulcus.

**Hemimegalencephaly (Fig. 2)**

is a rare but well-known congenital malformation, consists of enlarged hemisphere with ipsilateral ventriculomegaly.

**Tuberous sclerosis (Fig. 3, 4)**

is a rare autosomal-dominant neurocutaneous syndrome affecting about 1 in 6000 live births and is characterized by the presence of benign congenital tumors in multiple organs, and including cerebral, cardiovascular, pulmonary, renal, retroperitoneal, hepatic, gastrointestinal and skeletal involvement. Common cerebral involvement are cortical tubers, subependymal nodules, subependymal giant-cell astrocytomas and whitematter abnormalities. Other rare cerebral disorders include: mild dilatation of lateral ventricles, cerebellar atrophy, dysgenesis of the corpus callosum, Chiari malformation, microcephaly, macroencephaly and arachnoid cysts.

**Dysembryoblastic neuroepithelial tumor (Fig. 5)**

DNET in typical cases present as a bubbly mass which expands the affected gyri. The bubbly cystic appearance is seen as small cyst-like intratumoral structures that are very hyperintense on T2WI.

**Ganglioglioma (Fig. 6)**

Ganglioglioma is the most common tumor associated with temporal lobe epilepsy. Calcification is common in ganglioglioma and is an important distinguishing factor from DNET

**Groupe II Abnormal cell migration**

**Classic (Type I) Lissencephaly (Fig. 7, 8)**

Patients with classic lissencephaly may have a smooth brain surface in the complete form, or more commonly, they have a smooth surface with some gyral formation along the inferior frontal and temporal lobes in the incomplete form.
Cobblestone (Type II) Lissencephaly (Congenital Muscular Dystrophy)(Fig. 9)

Cobblestone lissencephaly is characterized by a nodular brain surface, ocular anomalies, and congenital muscular disorders. Cobblestone cortex results from overmigration of the neuroblasts and glial cells beyond the external glial limitations into the subarachnoid space.

Heterotopia(Fig. 10, 11, 12, 13)

Heterotopic Grey Matter results from an arrested migration of normal neurons along the radial path between the ventricular walls (ependyma) and the subcortical regions. There are two types of heterotopia: subependymal and subcortical. Heterotopia present as nodular foci of grey matter intensity on all sequences. They do not enhance.

Group III abnormal cortical organization

dopolymicrogyria (Fig. 14)

Polymicrogyria is a malformation due to an alteration of the cortical development in the late stage of neuronal migration.

It is characterized by small fine undulating gyri, very

similar to an undulating cortical ribbon, which cannot be detected on imaging. The cortex is also slightly thickened (5- 7 mm) with an abnormal increased T2 signal intensity of the underlying white matter, which is noted in 25% of cases.

When PMG is due to congenital infection, the areas of T2 prolongation are present bilaterally in the cerebral white matter.

Schizencephaly (Fig. 15, 16)

Schizencephaly is a cleft in the brain that connects the lateral ventricle to the subarachnoid space. The cleft is lined by polymicrogyric gray matter. 

*Open-lip* schizencephaly is characterized by separation of the cleft walls. 

*Closed-lip* schizencephaly is characterized by cleft walls in apposition to each other. 

Patients have seizures and hemiparesis, which is proportional to the size of the cleft and are more common in the open-lip type.
Group IV Not otherwise classified

malformations secondary to inborn errors of metabolism

- mitochondrial and pyruvate metabolic disorders
- peroxisomal disorders

other unclassified malformations

- sublobar dysplasia

Vascular abnormalities

Cavernoma (Fig. 17)

Cavernoma is also known as cavernous malformation or cavernous angioma. It is a benign low flow vascular malformation with a tendency to bleed. 75 percent occur as solitary sporadic lesions and 10-30 percent occur as multiple lesions.

Radiographic features

MRI

- MRI is the modality of choice, demonstrating a characteristic "popcorn" or "berry" appearance with a rim of signal loss due to haemosiderin, which demonstrates prominent blooming on susceptibility weighted sequences.
- T1 and T2 signal is varied internally depending on the age of the blood produces and small fluid fluid levels may be evident.
- Gradient Echo or T2* sequences are able to delineate these lesions better than T1 or T2 weighted images.
- In patients with familial or multiple cavernous angiomas GRE T2* sequences are very important in identifying the number of lesions missed by conventional Spin echo sequences.
- The lesions generally do not enhance, although enhancement is possible.

CT

- Unless large, these lesions are difficult to see on CT.
- They do not enhance.
- If large then a region of hyperdensity can be seen.
- If there has been a recent bleed then it is more conspicuous and may be surrounded by a mantle of oedema.
Sturge-Weber Syndrome

Sturge-Weber is also called encephalotrigeminal angiomatosis. It is a vascular malformation with capillary venous angiomas in the face (port-wine stain), choroid of the eye and leptomeninges. Venous occlusion and ischemia lead to angiomatosis with cortical calcium deposition and atrophy. Clinical features are seizures, hemiparesis, anopsia, mental retardation and port-wine stain.

Radiographic features

MRI (Fig. 18)

T1:
- signal of affected region largely normal, with anatomic volume loss evident at older age

T1 C+ (Gd)
- prominent leptomeningeal enhancement in affected area
- much later in life the angioma may 'burn out' losing enhancement
- enlarged ipsilateral choroid plexus

T2: low signal in white matter subjacent to angioma representing
- postulated accelerated myelination in neonate
- calcification later in life
- abnormal deep venous drainage seen as flow voids

GE/SWI/EPI:
- sensitive to calcification, seen as regions of signal drop out

MR spectroscopy:
- decreased NAA

CT
- detects subcortical calcification and can also demonstrate associated parenchymal volume loss
- appearance liken to 'tram-track' in nature
- calvarial and regional sinus enlargement may be evident
• ipsilateral choroid plexus may be enlarged
**Fig. 1:** The images show typical focal cortical dysplasia. There is cortical thickening and blurring of the grey/white matter junction on T1WI (blue arrow). The FLAIR image shows the subcortical hyperintensity (red arrow).

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**Fig. 2:** 1 year-old girl with refractory nocturnal epilepsy. MRI shows overgrowth of the left cerebral hemisphere with ipsilateral ventriculomegaly.
**Fig. 3:** FLAIR image shows multiple tubers and white matter abnormalities (blue arrows) and subependymal nodules (red arrows)
Fig. 4: Coronal FLAIR image and T1WI-CE show a giant cell astrocytoma at the level of the left foramen of Monro causing obstructive hydrocephalus. Note the dual component of the tumor: solid component with intense enhancement and cystic component.

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Fig. 5: DNET: MR Images show characteristic bubbly appearance and swelling of affected gyri.

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Fig. 6: Ganglioglioma in the left temporal lobe presenting as a cystic mass without enhancement. Notice calcification on T2*.

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Fig. 7: Lissencephaly type I: T1 WI and FLAIR image show diffuse abnormality of the overlying supratentorial mantle with reduction in number gyri and marked thickening of the cortex.

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**Fig. 8:** Lissencephaly type I: T2 WI shows partial absence of sulci with a thick cortex

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Fig. 9: Cobblestone lissencephaly. Axial and coronal T2-weighted images show an irregular nodular (blue arrows) with cortex with hypomyelination of the white matter (yellow arrows). Note the hypoplasia of the vermis and brain stem (red arrows)

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**Fig. 10:** Heterotopia: typical subependymal nodules (red arrows).

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**Fig. 11:** MRI showed Subcortical band heterotopia giving rise to a "double cortex" appearance
**Fig. 12:** Another case of typical Subcortical band heterotopia

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Fig. 13: Another case of heterotopia with typical parietal subcortical nodules.

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Fig. 14: 15-y-old boy with refractory nocturnal epilepsy The coronal FLAIR image shows multiple small gyri with derangement of the normal lamination and sulcation of the right temporo-parietal region (red arrows).

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**Fig. 15:** 15-year-old boy with Open-lip schizencephaly Axial T2 WI and coronal FLAIR image show a wide CSF-filled cleft connecting the right lateral ventricle with the subarachnoid space, which is lined with gray matter parenchyma (red arrows).

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**Fig. 16:** Sagittal T1WI, coronal T2 and FLAIR images show a closed-lip right-sided schizencephalic defect lined by pachygyria (red arrows).

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**Fig. 17:** T2WI and T2* gradient echo show cavernoma of the left temporal lobe. Notice the popcorn appearance with peripheral rim of hemosiderin on the T2WI. The lesion is almost completely black on the gradient echo due to blooming artefacts.

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**Fig. 18:** 1-year-old boy with Sturge-Weber syndrome. Axial FLAIR image shows atrophy of the left posterior cerebral hemisphere(yellow arrow) Axial T1WI post contrast image shows leptomeningeal enhancement and thickening (yellow asterisk).

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Conclusion

It is imperative that radiologists and clinicians understand the spectrum of brain MRI features of children epilepsy to aid in making an early diagnosis, thereby enabling and improving appropriate therapy planning.
Personal information

MD Badreeddine Alami

E-Mail: badr.alami@ymail.com

Department of radiology, university hospital Hassan II, Fes, Morocco
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