Cortical formation

7th week gestational age

Mitotic activity in the subependymal layer of the ventricular wall, cells begin to migrate to form the cerebral cortex.

[Atlas SW, 1996, MR imaging of brain and spine]
Any insult during the first two trimesters will lead to migrational anomalies:

- Infections
- Metabolic
- Ischemia
- Genetic
- Chromosomal

Patients usually present with:

- Seizures
- Developmental delay
- Mental retardation
Disorders of Cortical Formation

- Abnormal organization
- Abnormal migration
- Abnormal cell proliferation
Disorders of Cortical Formation

Abnormal cell proliferation

Malformation
- Hemimegalencephaly
- Cortical dysplasia

Hamartomatous
- Tuberous sclerosis
  [Forme furste]

Neoplastic
- Gangliogliomas
- Dysemmbryoblastic NET
Hemimegalencephaly

- **Isolated form**
  - Commonest
  - One hemisphere is affected

- **Syndromic form**
  - One cerebral hemisphere
  - Hemihypertrophy of a part or all one side of the **BODY**

- **Total form**
  - Least common
  - Cerebellar+ cerebral Hemispheres + Brainstem on one side
Hemimegalencephaly

- Enlargement of all or part of the cerebral hemisphere and ventricle.
- Focal or extensive calcification in both white and grey matter.
- Gliosis of the white matter.
- Straightening of the frontal horn of the enlarged ventricle.
- Agyria, pachygyria, polymicrogyria, heterotopia.
- MRS: ↓ NAA, ↑ creatine, choline, myoinsitol (glial proliferation)
Hemimegalencephaly

Hamartomatous overgrowth of a part or all one hemisphere

Imaging Findings:
- Enlarged hemisphere
- Lateral ventricle is often enlarged
- Dysplastic cortex [thick, calcified,..]
- Heterotopia
- Loss of gray-white matter interface.

Broumandi DD et al., Radio graphics, 2004
Hemimegalencephaly M 3M
Hemimegalencephaly

M 3M
Hemimegalencephaly

Brouandi DD et al., Radiographics, 2004
Hemimegalencephaly

Blaser & Jay, Neuroimaging Clinics, 1999
**Focal Cortical dysplasia (FCD)**

- Presence of abnormal neurons and glia arranged abnormally in focal areas in the cerebral cortex
- The incidence of FCD is 5-15% in patients with seizures.

**MRI findings**

- Abnormal high signal intensity in the gray matter on T2-weighted Images
- Blurring of the gray/white matter junction
- Thickening of the affected cortex
Focal Cortical dysplasia (FCD)
Dysembryoblastic Epithelial Tumors  DNET

Old names [ Hamartoma, mixed glioma] [ WHO I ]

Supratentorial [ temporal lobe] 60%
Well defined wedge shaped
Extends from the CORTEX to the ventricle
calcium
No edema
No enhancement
Multinodular cystic appearance
Low T1  High T2
May remodel the calverium
No recurrence after removal
DD Infarct

Blaser & Jay, Neuroimaging Clinics, 1999
Ganglioglioma

- Temporal lobe 85% of cases
- 80% occur before the age of 30 Y
- Calcifications 40% of cases
- Benign with calverial remodeling
- A solid enhancing part is the rule in all cases
- Non specific MR findings

In children < 10Y the tumor is large & more cystic with more edema around. The lesion may convert into the aggressive form, anaplastic ganglioglioma which is suspected by the degree of edema

Blaser & Jay, Neuroimaging Clinics, 1999
ANAPLASTIC GANGLIOGLIOMA
Disorders of migration

Migration occurs during 8th week along radial glial fibers [RGF]

- Damage of RGF
- Ischemia
- Infection
- Trauma
- Metabolic errors

Migration disorders
**Band Heterotopia** [double cortex]

- Three layers
- The cortex may be normal or pachygyric
- Seizures with mild-moderate mental retardation
Band Heterotopia

The severity of symptoms correlates with

- Degree of cortical disorganization
- Thickness of the heterotopic band
- High signal on T2 WIs
- Degree of ventriculomegaly
Band heterotopia

Lissencephaly

A rare congenital brain malformation that means Smooth brain (agyria), represents a spectrum of disorders ranging from total agyria to mixed agyria and pachygyria

Exact cause is not known
Lissencephaly

Complete form
Some gyral formation along the inferior frontal and temporal lobes

Imaging
- broad flat gyri with thickened cortex
- primitive sylvian fissures → hourglass configuration
Lissencephaly

MRI FINDINGS

- Agyric brain with or without areas of pachygyria
- Hour glass configuration of the brain
- Primitive vertical Sylvian fissures
- Small temporal lobes
- Associated anomalies:
  - Corpus callosum hypoplasia
  - Small brain stem
  - Gray matter heterotopia
Lissencephaly
Pachygyria / Lissencephaly
PACHYGYRIA

F 1Y

F 5Y
Focal gyral thickening

Surface reconstruction from a 3D data set [ TR / TE =15 / 8 MSEC ]
Blaser & Jay, Neuroimaging Clinics, 1999
GRAY MATTER HETEROOTOPIA

Neurons in abnormal location due to arrest of radial migration to the surface of the brain

Types:

- Subependymal heterotopia
- Focal heterotopia
- Diffuse band heterotopia
  [Double cortex syndrome]
Late-onset seizures with normal milestones
Focal isolated lesions or diffuse form
Diffuse bilateral form is associated with cerebellar hypoplasia
DD tuberous sclerosis
Subependymal heterotopia
Subependymal heterotopia
Subependymal heterotopia

Tuberous sclerosis

Subependymal heterotopia
FOCAL HETEROTOPIA

MRI FINDINGS

- Focal masses within the deep white matter
- Isointense to the cortex in all sequences
- No perilesional edema
- No mass effect
- Ipsilateral dysmorphology of the lateral ventricle
Focal heterotopia
Disorders of cortical organization

Abnormal organization

14-20 weeks  Sylvian and parieto-occipital sulci

32-33 week  Large number of sulci are formed

38-40 week  normal adult sulcal pattern is reached

Disruption of the process of gyral formation
  - Polymicrogyria
  - Schizencephaly
Thickened cortex with many small gayri
Gayri may be so small to be identified on imaging
Flat thick cortex similar to pachygyria or agyria
3D reformatted images

Bilateral symmetric PMG of the operculum
Primitive sylvian fissures
POLYMICROGYRIA
Schizencephaly
[gray matter-lined clefts]

- Small clefts with coapted walls [Closed lip type]
- Large clefts with free communication between the ventricle and subarachnoid spaces [Open lip type]

Schizencephaly

- Originally described as bilateral symmetrical full thickness cleft within the cerebral hemispheres.
- The cleft is lined by gray matter and commonly involves the parasylvian regions.
- The cause is unknown.
- The severity of disease correlates with the extent of schizencephaly.

**Type I** ([closed lip]):
- Fused cleft lined with gray matter.

**Type II** ([open lip]):
- Large true hemispheric cleft lined by a membrane of two layers of gray matter.
Schizencephaly

TYPE II SCHIZENCEPHALY

Open lip schizencephaly

surface reconstruction from a 3-D data set

Blaser & Jay Neuroimaging Clinics, 1999
Pachygyria
THE END

THANK YOU
MAMDOUH MAHFOUZ MD
Migrational Disorders of the Brain

- Gyral abnormalities
  - Agyria [Lissencephaly]
  - Pacchgyria
  - Polymicrogyria
- Schizencephaly
- Gray matter heterotopia
- Unilateral megalencephaly
Subependymal heterotopia
Pachygyria

- Broad flat gyria
- Commonly coexist with agyria
- May be focal or diffuse
venous angioma
Type I Schizencephaly
Megalencephaly
Megalencephaly
Megalencephaly
Megalencephaly
Megalencephaly
Megalencephaly
Hemimegalencephaly
Subependymal heterotopia
Polymicrogyria  PMG
Schizencephaly
Schizencephaly