MRI findings of cortical malformations in infants presenting with intractable epilepsy- The subtle and the obvious.

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Learning objectives

- To provide a brief overview of the MR imaging approach in infants presenting with intractable epilepsy.
- To present a simplified and clinically relevant classification of congenital cortical malformations.
- To emphasise the points that should be kept in mind while searching for potential sites of subtle cortical malformations, pattern recognition of common cortical malformations and associated features.
- To discuss MRI protocols and special sequences for optimising grey-white matter contrast and topographical clarity in a limited tertiary care set-up.
Background

Cortical malformations are responsible for up to 40% of childhood intractable epilepsy, that is, epilepsy not controlled by three drugs(1). The following is a simplified version of Barkovich 2012 classification of cortical malformations (2):

**Group I: Malformations secondary to abnormal neuronal and gyral proliferation or apoptosis:**

A) Severe congenital **microcephalies**.

B) **Megalencephalies**

C) Cortical dysgenesis with abnormal cell proliferation without neoplasia- **Hemimegalencephaly, Focal cortical dysplasia type II, Tuberous sclerosis**

D) Cortical dysplasias with abnormal cell proliferation and neoplasms.

**Group II: Malformations due to abnormal neuronal migration:**

A) **Periventricular heterotopia**

B) Malformations due to generalised abnormal transmantle migration- **Diffuse classic lissencephaly, posterior predominant, X-linked, Reelin type and Variant lissencephalies**.

C) Malformations due to localised abnormal late radial or tangential transmantle migration- **Subcortical heterotopia, sublobar dysplasia**

D) Malformations due to abnormal terminal migration and defects in pial limiting membrane- **Cobblestone malformation complex**.

**Group III: Malformations due to abnormal post-migrational development**

A) **Malformations with polymicrogyria- with or without schizencephaly**.

B) Cortical dysgenesis secondary to inborn errors of metabolism.

C) **Focal cortical dysplasias**.

D) Post-migrational developmental microcephaly.
Polymicrogyria is cerebral cortex with excessive microscopic gyration. Pachygyria is reduced, broad, small gyration and shallow sulcation. Agyria is complete absence of gyration. Lissencephaly is a term used to denote a smooth contour of brain. Focal cortical dysplasia is a localised region of non-neoplastic malformed cortical grey matter. Hemimegalencephaly is enlargement and cyto-architectural abnormality of one cerebral hemisphere. Microcephaly is the term used when head circumference is more than 3 SD below the mean for age and sex. Schizencephaly is the term used for dysplastic grey matter lined clefts extending from ventricular ependyma to periphery. Grey matter heterotopia is the presence of grey matter in abnormal locations.
Findings and procedure details

In a child presenting with *intractable epilepsy*, a more rigorous search for subtle cortical malformations must be made. The standard imaging anatomy must be kept in mind while examining the brain in all 3 planes;

- **Sagittal**- For lateral convexity, midline structures (commissures, 3rd ventricles, pineal region, brainstem, vermis)
- **Coronal**- For cerebellum, temporal and parietal lobes and superior convexities.
- **Axial**- For sylvian fissures, mid-brain, frontal and occipital lobes.

While setting an **Imaging protocol**-

- Must know normal appearance of brain at patient's age to recognise abnormal. Entire brain to be scanned from nasion to inion.
- Both T1WI and T2WI images should be obtained. During the process of myelination, for a variable period of time in first year of life, the sub-cortical white matter becomes isointense with grey matter on T1WI. This loss of grey/white matter differentiation obscures structural detail and makes identification of subtle abnormalities such as polymicrogyria difficult to detect within the first 6 months of life. The white matter shows maturity later, between 6 months and 2 years, on T2-W images and at this stage T1-W images are essential to evaluate structural abnormalities.
- Routine fast spin echo T2WI imaging is not recommended. T2WI-fast multiplanar inversion recovery with inversion time of 200 ms results in additive effect of T1 and T2 prolongation and a maximal discrimination of grey and white matter.
- Heavily weighted T1 Inversion recovery also optimises grey/white matter contrast.
- 3D volume acquisition by T1 weighted MPRAGE or 3D-FSPGR with multiplanar reconstruction and slice thickness 1.5 mm with no intervening gap. Reformation useful to correct for asymmetrical positioning of head. Surface rendering for topographical delineation of cortical abnormalities and their relation to adjacent gyri and sulci.
- Coronal and axial FLAIR sequences with 2-3 mm slice thickness and 0-1 mm interslice gap.
- Phased array and surface coils- Improve signal-to-noise ratio.
- Specialised protocol of quantitative volumetry, T2 relaxometry, Magnetic resonance spectroscopy, functional MRI, Diffusion weighted imaging, Diffusion tensor imaging, Magnetic source imaging should be considered as per availability and specific indications.

The following is a comprehensive description of non-neoplastic cortical dysplasias encountered commonly in clinical practice:
1. Polymicrogyria

How to identify-

- Multiple small delicate gyri without intervening sulci or with intervening sulci obliterated by fusion of their superficial layers or
- Multiple small, thick, irregular, widened gyri separated by shallow sulci
- Associated corrugated appearance of grey-white matter junction.
- Unilateral / Bilateral, symmetrical / asymmetrical, focal / hemispheric / diffuse

Where to look-

- **Perisylvian cortex** - In congenital bilateral perisylvian syndrome, cortical thickness may of normal range. However, sylvian fissures are deep and extend farther back than normal. In suspicious cases, the degree of depth of sylvian fissures should be assessed on a sagittal image and compared with normal subjects.
- **Cerebral convexities** - Superficial with cortex appearing flat and congruent to the arc of normal cortex, coursing radially inwards. (frontal, fronto-parietal> lateral parieto-occipital>temporal> parasagittal mesial occipital with typical sparing of striate cortex, hippocampus, gyrus rectus, cingulate gyrus and visual cortex).

What else to expect-

- Focal enlargement of sub-arachnoid space, focal parenchymal atrophy, calcifications, uncondensed cortical veins (Figure 1)
- T2WI hyperintense sub-cortical zone representing gliosis/ischemia/dilated perivascular spaces.
- Thinner (2-3 mm) and bumpy inner surface of affected cortex in unmyelinated areas and paradoxical appearance of thicker (5-8 mm) and smooth surface in myelinated brain.(3)

What else to consider-

- Large vessels common in regions where there is infolding of thickened cortex should not be mistaken for vascular malformations.
- The width of cortex in polymicrogyria is 5-7 mm, with irregular grey-white matter junction whereas in pachgyria, width of the cotex is > 8mm with relatively smooth or indistinct grey-white matter junction(Figure 2). In contrast to pachgyria, in perisylvian polymicrogyria, there is scalloping or inversion of lateral ventricles.

2. Lissencephaly spectrum.
How to identify-

- Smooth abnormal contour of brain devoid of sulci (agyria)(Figure 3) or few patchy areas of small, broad, thick gyri (pachygyria) or both, (Figure 4) abnormal opercularisation with oblique/vertically oriented shallow, wide sylvian fissures giving rise to hour-glass/figure-of-8 appearance of brain. *(Classical lissencephaly).*
- Homogeneous gray matter band between cerebral cortex and lateral ventricles, characteristically surrounded by a zone of white matter, with abnormal overlying cortex (polymicrogyria / pachygyria) and shallow sulci. *(Subcortical band heterotopia).*

Where to look-

- In incomplete form of classical lissencephaly, some sulcation is present in inferior frontal and temporal region with antero-posterior gradation of disease severity.
- In *LIS1* mutation, fronto-temporal gyri are more developed than parieto-occipital gyri.
- In *X-linked lissencephaly*, posterior gyri are more developed(4).
- *Subcortical band heterotopia* is typically seen symmetrically in frontal region.

What else to expect-

- Marked reduction in white matter volume with smooth gray-white matter junction in classical lissencephaly. However, linear disruptions of gray-white matter junction are noted in individuals with LIS 1 mutations, irregular disruptions of gray-white matter junction with nodular heterotopia in DCX mutations.(5)
- Multilayered cortex with thin outer layer(grey matter), underlying thin 'cell sparse zone'(white matter) and thicker broad band (grey matter) in classical lissencephaly.
- Laterally placed middle cerebral arterial branches due to shallow sylvian fissures, wavy vessels on surface of brain in classical lissencephaly.
- Posterior fossa structures characteristically spared in complete lissencephaly.
- Associated features- agenesis of corpus callosum, cerebellar hypoplasia, peri-sylvian pachgyria, absent anterior limb of internal capsule and dysplastic basal ganglia in lissencephaly variants and septum pellucidum calcifications in Miller Dieker syndrome.

What else to consider-

- *Isolated pachygyria*- Usually focal areas of thick, small, broad gyri with shallow sulci and indistinct gray-white matter junction.
- Brain of classical lissencephaly may resemble *normal fetal brain at 23-24 weeks.*
3. Focal cortical dysplasia.

How to identify-

<table>
<thead>
<tr>
<th>FCD I</th>
<th>FCD II</th>
</tr>
</thead>
<tbody>
<tr>
<td>• Significant segmental or lobar hypoplasia/atrophy with variable cortical thickness. (Figure 5)</td>
<td>• Localised area of cortical thickening</td>
</tr>
<tr>
<td>• Decreased volume of subcortical white matter, increased signal intensity of subcortical white matter on T2 WI, marked increase on FLAIR, decreased on T1 WI, marked decrease on T1 IR.</td>
<td>• More pronounced increased signal intensity on T2 WI of grey matter and subcortical white matter. Altered white matter signal extends towards suprolateral margin of lateral ventricle (transmantle sign)</td>
</tr>
<tr>
<td>• Slight blurring of grey-white matter junction</td>
<td>• More evident blurring of grey-white matter junction</td>
</tr>
<tr>
<td>• Not usually associated with gyral abnormalities, may be associated with mesial temporal sclerosis.</td>
<td>• Often associated with abnormal gyral and sulcal pattern and enlarged perivascular spaces</td>
</tr>
<tr>
<td>• Occasional contrast enhancement</td>
<td>• Not associated with contrast enhancement</td>
</tr>
</tbody>
</table>

Where to look-

• Temporal lobes- type IA
• Extra temporal location (frontal lobes, hemispheric, multilobar) type IB, type II
• Rarely seen in medial temporal lobe

What else to expect-

• Anterior temporal lobe volume loss with hippocampal sclerosis, abnormal white matter hyperintensity on T2 WI/FLAIR with otherwise normal appearing cortex (FCD type III)
• FCD with developmental tumor characterised by positive mass effect, cystic component, calcifications, contrast enhancement (type IIIb). Dysplastic lesions usually located parallel to tumor(6).
• Focal cortical thickening at the bottom of sulcus and funnel shaped structure directed at ependymal surface surrounded by stellate gyral complex (Bottom of sulcus dysplasia)
• Sublobar region of dysplastic brain with abnormal ventricle and involvement of entire thickness of cortex (sublobar dysplasia)

**What else to consider**-

• Blurring of grey-white matter junction absent in polymicrogyria.
• Incomplete enhancing ring in solitary demyelinating lesions.
• Other stigmata (subependymal nodules) in tuberous sclerosis.
• Normal variation with distinct transmantle vein to be differentiated from bottom of sulcus dysplasia.

**4. Hemimegalencephaly.**

**How to identify**-

• Enlarged, dysplastic unilateral cerebral hemisphere, extreme asymmetry not corresponding to any normal stage of human brain development. May be focal/localised/lobar.
• Cortex thickened with abnormal gyration, 'lumpy-bumpy' on T1WI, associated with focal areas of agyria, pachygyria, polymicrogyria and heterotopias in affected hemisphere.
• Heterogeneous white matter signal intensity on T2WI and FLAIR- gliosis and cyst-like hyperintensities.
• Myelination is often disordered and accelerated with T1 shortening.
• In severe cases, no normal hemispheric architecture.
• Ipsilateral lateral ventricle is most often enlarged and deformed, with straightening of frontal horn and extension of posterior horn across midline. Occasionally may be small.
• Macrocephaly is often initial presenting feature.
• *Divided into three types: isolated, syndromic and total.*

**What else to expect**-

• White matter may be calcified.
• Enlargement of ipsilateral half of brainstem and cerebellum in total hemimegalencephaly.
• Roughly inverse relationship between grey-white matter abnormalities and size of cerebral hemisphere.(7)
• Mild to moderate enlargement when associated with agyria, severe enlargement when associated with polymicrogyria.
• Contralateral cerebral hemisphere is normal except for being compressed.
• May be associated with developmental venous anomalies, olfactory nerve enlargement, bilateral abnormal cerebellar folia.

**What else to consider**-
• Unilateral enlarged hemisphere- *Gliomatosis cerebri.*
• Unilateral focal enlargement- *Tuberous sclerosis with widespread cortical dysplasia, gangliocytoma.*
• Small hemisphere, making normal hemisphere appear large- *Rasmussen encephalitis, Dyke-Davidoff Masson syndrome, Sturge-weber syndrome.*

5. Microcephaly-

How to identify-

• Reduced number of gyri and shallow sulci (25-50% of normal depth) with normal or thinned cortex (microcephaly with simplified gyral pattern)(Figure 6).
• Severe microcephaly, abnormal sulcation, greatly simplified or almost completely smooth cortex with thickness more than 3mm (microlissencephaly).
• Craniofacial ratio less than or equal to 1.5:1, slant forehead, calverial sutures may appear over-riding.

What else to expect-

• Secondary insults with haemorrhagic residues on T2\(^*\) (GRE,SWI)
• May be associated with diffuse/asymmetric polymicrogyria/callosoal agenesis or hypogenesis/atyical cortical dysgenesis/ cerebellar and brainstem hypoplasia/enlarged extra-axial spaces/hydrocephalus ex vacuo/arachnoid cysts.(8)

What else to consider-

• Syndromes associated with primary microcephaly.
• *Hypoxic ischaemic encephalopathy* with or without cortical, white matter or basal ganglia volume loss.
• *TORCH infection*: calcification, abnormal white matter, neuronal migration anomalies, germinolytic cysts.
• *Fetal alcohol syndromes*: callosal abnormalities, ventriculomegaly.
• *Non accidental head injury*: encephalomalacia, chronic subdural collection, parenchymal laceration.

6. Schizencephaly

How to identify-

• Heterotopic grey matter lined cleft extending from ventricular ependyma to pial surface of brain traversing through white matter (Figure 7).
• Closed lip (type I): walls of cleft appose each other with no intervening CSF, outpouching or nippling at epenymal surface at cleft. Smaller in size than open lip schizencephaly.
• Open lip (type II): cleft walls widely separated, wide and wedge shaped/parallel walled intervening canal of CSF. Severe form gives appearance of "Basket brain".

Where to look-
• Unilateral or bilateral.
• Posterior frontal and parietal lobe near central sulcus (most common).
• Large clefts may extend to temporal and occipital lobes. Isolated involvement is rare.

What else to expect-
• Grey matter lining cleft in closed lip schizencephaly may be cobblestone/dysplastic with indistinct/irregular grey-white matter junction.
• On T2 WI grey-white matter distinction more clear in unmyelinated brain. Abnormal white matter is typically hypointense.
• Calcifications on T2* in cytomegalovirus infections.
• Heterotopias, Septo-optic dysplasias,Absent septum pellucidum, cortical dysgenesis, developmental venous anomalies (9).
• MCA branches can fall into large clefts or may be displaced along cleft walls.

What else to consider-
• Encephaloclastic porencephaly- Lined by gliotic white matter
• Hydranencephaly- Destruction of cerebral tissue in anterior and middle cerebral artery territory. Residual parenchyma displaced inferiorly and posteriorly.
• Holoprosencephaly- Semilobar can mimic bilateral open lip schizencephaly. Residual parenchyma displaced anteriorly, midline fusion anomalies.
• Agenesis of corpus callosum with inter-hemispheric cyst- Type 1 cyst: Diverticulum of third ventricle. Residual parenchyma displaced laterally.
• Post-operative cavity.
• Transmantle heterotopia may actually represent a form of closed lip schizencephaly.

7. Heterotopia

How to identify-

Subependymal heterotopia Subcortical heterotopia
• Unilateral/Bilateral focal/diffuse nodular/smooth confluent masses isointense to grey matter indenting ependyma of ventricles. (Figure 8)

• Little or no distortion of remaining brain

• Surrounding white matter normal

• Nodular/curvilinear swirls of grey matter originating from deep sulci which wind their way through cerebral mantle to ependyma. Few cases show a mixed nodular-curvilinear pattern

• Focal sub-cortical heterotopia: marked distortion of ventricles and diminished hemispheric size.

• Qualititavely decreased surrounding white matter

**Where to look**-

• Trigone and occipital horns (most common) followed by body and frontal horns of lateral ventricles in sub-ependymal heterotopia.

**What else to expect**-

• No contiguity of nodular type of sub-cortical heterotopia with cerebral cortex. Affected area reduced in size.

• In curvilinear subcortical heterotopia, heterotopic tissue gives appearance of enfolded cortex having contiguity with cortex. Overlying cortex is often thin with shallow sulci. May contain blood vessels or CSF traversing along with it.

• Subcortical heterotopia often associated with schizencephaly, microcephaly, polymicrogyria, dysgenesis of corpus callosum and absent septum pellucidum.

**What else to consider**-

<table>
<thead>
<tr>
<th>Sub-ependymal heterotopia</th>
<th>Sub-ependymal nodules of Tuberous sclerosis</th>
</tr>
</thead>
<tbody>
<tr>
<td>Iso-intense to grey matter</td>
<td>Not precisely iso-intense to grey matter</td>
</tr>
<tr>
<td>No contrast enhancement</td>
<td>Occasionally shows contrast enhancement.</td>
</tr>
<tr>
<td>Smooth, ovoid, with long axis parallel to ependyma</td>
<td>Irregular, with long axis perpendicular to ependyma.</td>
</tr>
<tr>
<td>Large dysplastic and disorganised masses of ectopic grey matter may simulate <em>intra-cranial mass</em>.</td>
<td></td>
</tr>
</tbody>
</table>
8. Cobblestone lissencephaly.

How to identify-

- Bumpy or pebbly cortical surface phenotypically associated with congenital muscular dystrophies with laminin alpha-2 chain deficiency and those with hypoglycosylation of alpha-dystroglycan.

Where to look-

- Antero-posterior gradient in Fukuyama disease and muscle-eye-brain disease.
- Polymicrogyria with thick cortex/cobblestone cortex predominantly in frontal lobes (Figure 9). In some cases may involve fronto-parietal lobes. Occipito-temporal lobes when involved show pachygyria/agyria/lissencephaly with thin cortex.

What else to expect-

- More severe gyral malformations, diffuse cerebral cobblestone cortex, retinal and cerebellar malformations in Walker-Warburg disease associated with hydrocephalus, small pyramids, flat pons with midline ventral cleft resulting in Z shaped brainstem, abnormal white matter, agenesis of corpus callosum, occipital encephalocele, dandy-walker malformation.
- Delayed myelination, ventriculomegaly, hypoplasia of pons, cerebellar polymicrogyria, disorganised cerebellar foliation accompanying cysts in mid-portion and dorsal surface of cerebellar hemisphere particularly in superior semilunar lobule in Fukuyama Congenital muscular dystrophy.
- Cerebellar polymicrogyria(with or without cysts), absence of septum pellucidum, pontine and cerebellar vermian hypoplasia, diffuse cortical dyslastic changes, patchy hypomyelination and variable callosal hypogenesis and hydrocephalus in muscle-eye-brain disease(11).

What else to consider-

- Lissencephaly type 1: Cortex is thicker and smooth.
- GPR56 associated Bilateral fronto-parietal polymicrogyria: shares many overlapping features with congenital muscular dystrophies.
Fig. 1: Polymicrogyria. Focal area of closely packed numerous gyri and shallow sulci (blue arrow) associated with focal sub-cortical white matter atrophy and enlarged sub-arachnoid space, absent septum pellucidum (black arrow). Corrugated gray white matter junction on 3D-MPRAGE (white box). Prominent anomalous cortical veins in sub-arachnoid space on T1 weighted IR. (red arrow)

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Fig. 2: Bilateral fronto-parietal pachygyria (blue arrow) with parieto-occipital polymicrogyria (white arrow), absent septum pellucidum, thin corpus callosum and fronto-parietal leukodystrophy. Hypoplastic pons (red arrow), cerebellar vermis with enlarged retrocerebellar space communicating with fourth ventricle. Blooming on GRE (black arrow) suggestive of peri-ventricular calcifications. Better appreciation of grey white matter junction on IR coronal (white circle).

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**Fig. 3:** Incomplete Lissencephaly. Smooth contour of brain with few broad thick gyri (blue arrow) with antero-posterior gradient and bilateral frontal leukodystrophy. Wide, posteriorly directed bilateral sylvian fissures (black arrow), smooth underlying grey white matter junction better appreciated on 3D-MPRAGE (red arrow) and coronal IR (white circle).

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**Fig. 4:** On T2WI, dysplastic frontal cortex (blue arrow) appears blurred with indistinct grey-white matter junction due to marked white matter volume loss. Better appreciation of grey white matter junction on IR reveals bilateral frontal polymicrogyria (white circle) and posterior pachygyria. There is associated marked enlargement of sub arachnoid space (white arrow) and ventriculomegaly. Poor contrast discrimination of grey-white matter on FLAIR (white circle) for comparison.

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Fig. 5: Asymmetrical gyral cortical thickening in left frontal lobe on T2WI and T1WI (red arrow) with no evidence of underlying sub-cortical hyper-intensity. Mild thickening of cortex along the depth of sulcus on coronal IR (blue arrow). No contrast enhancement on contrast T1 MPR axial (white square). Findings were suspicious of focal cortical dysplasia.

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**Fig. 6:** Microcephaly with simplified gyral pattern. Small head size, bifrontal agyria with thin cortex (blue arrow), bilateral posterior pachgyria with smooth grey white matter junction, better appreciated on IR coronal image (black arrow).

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Fig. 7: Bilateral open lipped Schizencephaly. Wide CSF containing cleft in right parietal region lined by dysplastic grey matter (blue arrow). On T2WI, it is a bit difficult to delineate a communicating CSF channel on left side due to closely apposed lips (black arrow). The open lipped configuration of schizencephaly on left side confirmed on coronal IR image (red box). Associated dilatation of extra-axial CSF spaces in left anterior temporal region and posterior fossa, ? arachnoid cysts (black arrows).

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Fig. 8: Subependymal heterotopia. Nodular masses of grey matter abutting and scalloping ependyma of body of bilateral lateral ventricles on T2WI (white arrow) and T1WI (black arrow). Subtle lesions can be easily mistaken for caudate nuclei on axial scans. Coronal IR image can be used to better delineate the lesions from surrounding deep gray matter. (red square).

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Fig. 9: Pebbly and thickened cortical surface of bilateral fronto-parietal region with leukodystrophy (blue arrow). Mildly enlarged bilateral basal ganglia and thalamus with normal signal intensity (red arrow). Tiny cerebellar cysts in bilateral dorsal cerebellar hemispheres (white square). Hypoplasia of pons with mid-line cleft (black arrow). Findings suggestive of cobblestone lissencephaly in Fukuyama congenital muscular dystrophy.

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Conclusion

In infants presenting with intractable epilepsy, congenital cortical malformations, especially when subtle, may be missed or misinterpreted if not actively looked for. For an unequivocal diagnosis, a thorough knowledge of imaging anatomy and a standard MRI protocol is essential. With the ever expanding etiological and histopathological classification of cortical malformations, it is important for the radiologist to provide a diagnosis as clinically relevant as possible with emphasis on associated neurological abnormalities.
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