Cortical Dysplasia

Roberto Spreafico
**Malformations of Cortical Development**

**Areas of atypical cortical structure**

- **absent or abnormally broad gyri:** lissencephaly
- **excessive folding of an abnormally thin cortex**
- **Focal cortical dysplasia**
- **Polymicrogyria**
- **Agyria/pachygyria**
- **Heterotopia**
- **Laminar heterotopia**
- **Leptomeningeal heterotopia**
- **Periventricular nodular heterotopia**

**Misplaced neurons; may serve as foci for epileptogenic activity; wide spectrum from isolated nodules to “band” heterotopia**

*Source:* Copp and Harding 1999
Classification Schemes

- **Mischel et al. (1995)** J. Neuropathol Exp Neurol. 54; 137-153
  
  Based entirely on pathology. Impractical and impossible to apply to the majority of patients with MCD who never undergo surgical resection or biopsy

- **Raymond et al. (1995)** Brain 118; 629-660
  
  Clinical, neuropsychological, EEGraphic, neuroradiological and pathological features with various cortical dysgenesis

- **Barkovich et al. (2001)** Neurology 57;2168-2178
  
  Too heavily weighted on imaging and not enough on pathology. However data imaging are available for essentially all patients whereas pathological material and genetic data are available for very few.

- **Tassi et al. (2002)** Brain 125; 1719-1732
  
  Limited to FCS and based on neuropathological, electroclinical and MRI findings

- **Sarnat and Flores–Sarnat (2003)** Epileptic Disord. 5 (2); S35-S43
  
  Based entirely on molecular genetics

- **Palmini et al. (2004)** Neurology 62 (suppl. 3) S2-S8
  
  A review of current terminology and classification issues of potential clinical relevance to epileptologists, neuroradiologists and neuropathologists dealing with FCD

- **Barkovich et al. (2005)** Neurology 65;1873- 1887
  
  A revised version of the classification published in 2001
Terminology and classification of the cortical dysplasias

A. Palmini, MD, PhD; I. Najm, MD; G. Avanzini, MD; T. Babb, PhD; R. Guerrini, MD; N. Foldvary-Schaefer, DO; G. Jackson, MD; H.O. Lüders, MD, PhD; R. Frayson, MD, PhD; R. Spreafico, MD, PhD; and H.V. Vinters, MD

1. Mild MCD
   - **Type I**: with ectopically placed neurons in or adjacent to Layer I
   - **Type II**: with microscopic neuronal heterotopias outside Layer I
   **Structural Imaging**: both types probably are not detectable by current MRI
   **Clinical relevance**: no specific data delineating clinical or epileptic profile

2. FCD
   - **Type IA**: isolated architectural abnormalities (dyslamination)
   - **Type IB**: architectural abnormalities and giant or immature neurons
   **Structural Imaging**: unclear in type I B
   **Clinical relevance**: no specific data delineating clinical or epileptic profile
   - **Type IIA**: architectural abnormalities with dysmorphic neurons without balloon cells
   - **Type IIB**: architectural abnormalities with dysmorphic neurons with balloon cells
   **Structural imaging**: increased cortical thickness, blurring grey/white matter, increased T2-weighted signal, mainly extra temporal
   **Clinical relevance**: highly intrinsic epileptogenetic, high seizure frequency,
NORMAL ARCHITECTURAL CYTOARCHITECTURAL TAYLOR CYTOLOGIC ABNORMALITIES

<table>
<thead>
<tr>
<th>Normal</th>
<th>Type IA and IB</th>
<th>Type IIA/B</th>
</tr>
</thead>
<tbody>
<tr>
<td>A</td>
<td>B</td>
<td>C</td>
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</tbody>
</table>

STRUCTURAL ABNORMALITIES

CYTOLOGIC ABNORMALITIES
Seizure onset; FCD in 198 patients
Type IA: isolated architectural abnormalities (dyslamination)
M.M.
Interictal activity
during wakefulness,
electrode J
500 patients operated on for intractable epilepsy at the “C. Munari “ Epilepsy Surgery Centre – Milano -Italy

Malformations of Cortical Development (MCD-Site of surgery)

<table>
<thead>
<tr>
<th>Lesion</th>
<th>N°</th>
<th>T</th>
<th>Fr</th>
<th>P</th>
<th>C</th>
<th>O</th>
<th>Multilobar/T</th>
<th>Multilobar</th>
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<tr>
<td>Hamartoma</td>
<td>10 (4%)</td>
<td>9</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>1</td>
<td>0</td>
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<tr>
<td>Arachnoid cyst</td>
<td>2</td>
<td>1</td>
<td>1</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
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<tr>
<td>FCD I A/B</td>
<td>102 (39%)</td>
<td>64</td>
<td>18</td>
<td>1</td>
<td>0</td>
<td>1</td>
<td>12</td>
<td>6</td>
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<tr>
<td>FCD II A/B</td>
<td>69 (26%)</td>
<td>15</td>
<td>27</td>
<td>4</td>
<td>1</td>
<td>0</td>
<td>10</td>
<td>12</td>
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<tr>
<td>Neuronal Heter.</td>
<td>44 (17%)</td>
<td>39</td>
<td>1</td>
<td>0</td>
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<td>0</td>
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<td>3</td>
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<tr>
<td>DCX</td>
<td>3</td>
<td>0</td>
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<td>0</td>
<td>0</td>
<td>0</td>
<td>2</td>
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<tr>
<td>HMEG</td>
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<td>0</td>
<td>0</td>
<td>0</td>
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<tr>
<td>PNH</td>
<td>14 (5%)</td>
<td>6</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>7</td>
<td>1</td>
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<tr>
<td>Polymicrogyria</td>
<td>3</td>
<td>1</td>
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<td>0</td>
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<tr>
<td>T.S.</td>
<td>14 (5%)</td>
<td>0</td>
<td>11</td>
<td>1</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>2</td>
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<tr>
<td>TOTAL</td>
<td>262 (52%)</td>
<td>135 (23%)</td>
<td>59</td>
<td>6</td>
<td>1</td>
<td>1</td>
<td>33 (13%)</td>
<td>27 (10%)</td>
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