Approach to Focal Cortical Dysplasia in Epilepsy, Challenges and Lessons

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Focal dysplasia of the cerebral cortex in epilepsy

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From the Neurosurgical Unit of Guy's, Maudsley, and King's College Hospitals, London
and
C. J. BRUTON AND J. A. N. CORSELLIS
From the Department of Neuropathology, Runwell Hospital, Wickford, Essex

• Pathology reports 1951-1960

• Unusual findings in 10 individuals undergoing lobar resection for epilepsy

• 'consisted of congregations of large, bizarre neurones which were littered through all but the first cortical layer. In most, but not in all cases, grotesque cells, probably of glial origin, were also present in the depths of the affected cortex and in the subjacent white matter.......reminiscent of tuberous sclerosis'

• 3% operative cases
Classification of MCD

I. Malformations secondary to abnormal neuronal and glial proliferation of apoptosis
   1A Microcephaly
   1B Megalancephalacies
   1C Cortical dysgeneseses with abnormal cell proliferation

II. Malformations due to abnormal neuronal migrations
   IIA Heterotopia
   IIB Lissencephaly
   IIC subcortical heterotopia and sublobar dysplasia
   IID Cobblestone malformations

III. Malformations secondary to abnormal postmigrational development
   IIIA. Polymicrogyria and schizencephaly
   IIIC Focal cortical dysplasia
   IIID Postmigrational microcephaly

Barkovich et al 1996,2002
Neurology 2005;65:1873–1887
Brain 2012;135:1348-1369
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I. Malformations secondary to abnormal neuronal and glial proliferation of apoptosis
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   1B Megalancephalies
   1C Cortical dysgeneses with abnormal cell proliferation
      *Tuberous sclerosis*
      *Focal cortical dysplasia*

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*References*

- Barkovich et al 1996,2002
  *Neurology* 2005;65:1873–1887
- Brain 2012;135:1348-1369
Focal cortical dysplasias

- Type I: No dysmorphic neurons or balloon cells
  - IA: isolated architectural abnormalities (dyslamination)
  - IB: architectural abnormalities + giant of immature neurons
  - Imaging: ?can be seen by current techniques

- Type II: Taylor type FCD (dysmorphic neurons with or without balloon cells)
  - IIA: architectural abnormalities with dysmorphic neurons without balloon cells
  - IIB: architectural abnormalities with dysmorphic neurons & balloon cells
  - Imaging: commonly identified on MRI

*Palmini et al
 Neurology 2004;62(Suppl 3):S2–S8*
The clinicopathologic spectrum of focal cortical dysplasias: A consensus classification proposed by an ad hoc Task Force of the ILAE Diagnostic Methods Commission

*2Ingmar Blümcke, †Maria Thom, ‡Eleonora Aronica, §§Dawna D. Armstrong, ¶Harry V. Vinters, #Andre Palmini, **Thomas S. Jacques, ††Giuliano Avanzini, †††A. James Barkovich, §§§Giorgio Battaglia, ¶¶Albert Becker, ###Carlos Cepeda, ####Fernando Cendes, ††††Nadia Colombo, ††‡‡Peter Crino, §§§§J. Helen Cross, ¶¶¶Olivier Delalande, ######François Dubeau, ****John Duncan, †††††Renzo Guerrini, ††††‡Philippe Kahane, §§§§Gary Mathern, ¶¶¶¶Imad Najm, ††††††Çiğdem Özkara, *****Charles Raybaud, ††††††Alfonso Represa, †††††‡Steven N. Roper, §§§§§Noriko Salamon, ¶¶¶¶¶Andreas Schulze-Bonhage, †††††††Laura Tassi, ***Annamaria Vezzani, and ††Roberto Spreafico

Table 1. The three-tiered ILAE classification system of focal cortical dysplasia (FCD) distinguishes isolated forms (FCD Types I and II) from those associated with another principal lesion (FCD Type III).

<table>
<thead>
<tr>
<th>FCD Type I (isolated)</th>
<th>Focal cortical dysplasia with abnormal radial cortical lamination (FCD Type Ia)</th>
<th>Focal cortical dysplasia with abnormal tangential cortical lamination (FCD Type Ib)</th>
<th>Focal cortical dysplasia with abnormal radial and tangential cortical lamination (FCD Type Ic)</th>
</tr>
</thead>
<tbody>
<tr>
<td>FCD Type II (isolated)</td>
<td>Focal cortical dysplasia with dysmorphic neurons (FCD Type IIa)</td>
<td>Focal cortical dysplasia with dysmorphic neurons and balloon cells (FCD Type IIb)</td>
<td></td>
</tr>
<tr>
<td>FCD Type III (associated with principal lesion)</td>
<td>Cortical lamination abnormalities in the temporal lobe associated with hippocampal sclerosis (FCD Type IIIa)</td>
<td>Cortical lamination abnormalities adjacent to a glial or glioneuronal tumor (FCD Type IIIb)</td>
<td>Cortical lamination abnormalities adjacent to vascular malformation (FCD Type IIIc)</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Cortical lamination abnormalities adjacent to any other lesion acquired during early life, e.g., trauma, ischemic injury, encephalitis (FCD Type IIId)</td>
</tr>
</tbody>
</table>

FCD Type III (not otherwise specified, NOS): if clinically/radiologically suspected principal lesion is not available for microscopic inspection. Please note that the rare association between FCD Types IIa and IIb with hippocampal sclerosis, tumors, or vascular malformations should not be classified as FCD Type III variant.
Three types of FCD

- **Type 1**: Architectural Dysplasia
- **Type 2**: Architectural and Cytological Dysplasia
- **Type 3**: Architectural Dysplasia + 2nd pathology
Imaging Characteristics

FCD Type I

- Atrophy lobar/sublobar
- Volume loss of Subcortical WM
- Increased T2 signal in WM
- Blurring of GM/WM border
- May have abnormal gyral/sulcal pattern
Imaging Characteristics

Type II

- Increased cortical thickness
- Blurring of the GM/WM junction
- Increased subcortical WM signal on T2
- WM signal alterations taper to ventricle
- Abnormal gyration and sulcation
- Focal enlargement of the subarachnoid spaces adjacent to lesion
Focal Cortical Dysplasia Type III

IIIa
Associated with HS

IIIb
Adjacent to a tumour

IIIc
Adjacent to a vascular malformation

IIId
Adjacent to an acquired lesion from early life
Clinical Characteristics of FCD

- Present early
- Vary in size and location
- May be multilobar
- Seizures very resistant to treatment
- Minimal focal neurology
- Neuropsychological and developmental impact
- Focal rhythmic electrical discharges on scalp EEG
Age of onset of epilepsy

• Most series suggest early onset epilepsy in the majority

• Cascino et al 2005, surgical series, 7 centres; 21/213 (10%) onset >18 years

• Fauser et al 2006, 120 patients surgical series, 61% <5 yrs, 92.5% <16 years
Medical Treatment

Stephan, Kwan and Brodie, *Epilepsia* 2001; 42:357-362
550 patients; 70% newly diagnosed focal epilepsy over 13 years  *Minimum 2yr review*
63 (12%) cortical dysplasia
34 (54%) seizure free  AEDs (none) 5, (1) 22

Semah et al, *Neurology* 1998; 51: 1256-1262
2200 patients, 8% first seizure, over 7 years
96 (8%) cortical dysgenesis
23 (24%) seizure free
Clinical characteristics in focal cortical dysplasia: a retrospective evaluation in a series of 120 patients

Susanne Fauser, Hans-Juergen Huppertz, Thomas Bast, Karl Strobl, Georgios Pantazis, Dirk-Matthias Altenmueller, Bertram Feil, Sabine Rona, Christoph Kurth, Dietz Rating, Rudolf Korinthenberg, Bernhard J. Steinhoff, Benedikt Volk and Andreas Schulze-Bonhage

Responsiveness to antiepileptic drugs
## Medical Treatment

Vigevano & Koivikko *Epilepsia* 1997;38:1275-1282

Vigabatrin vs ACTH for Infantile Spasms  \textbf{N=47}

<table>
<thead>
<tr>
<th>Condition</th>
<th>VGB</th>
<th>ACTH</th>
</tr>
</thead>
<tbody>
<tr>
<td>Cessation of spasms</td>
<td>11/23 (48%)</td>
<td>14/19 (74%)</td>
</tr>
<tr>
<td>Cerebral malformation</td>
<td>3/4 (75%)</td>
<td>0/3 (0%)</td>
</tr>
<tr>
<td>Tuberous sclerosis</td>
<td>3/3 (100%)</td>
<td>1/1 (100%)</td>
</tr>
</tbody>
</table>
Ketogenic diet

Long term outcome of the ketogenic diet for intractable childhood epilepsy with focal malformation of cortical development  *Jung et al Paediatrics 2008;122:e330-3*

At 3 mo
- Seizure free: $n = 21$ (44.7%)

At 2 y Completion KD
- Seizure free: $n = 16$ (34%)

Follow-up range after completion KD: 13 mo–7 y
- Seizure free: $n = 9$ (19.1%)
  - Seizure free on additional AED: $n = 1$ (2.1%)
    - Seizure relapse: $n = 7$ (14.9%)
      - Partial or no response to KD: $n = 26$ (55.3%)
        - Partial or no response to KD: $n = 37$ (78.7%)
          - Surgery: $n = 22$ (46.8%)
          - AED: $n = 15$ (31.9%)
Major aetiological categories

European Epilepsy Brain Bank 1990-2014, N = 7286, Blumcke et al 2017
Surgical resection

Evaluation required will depend on extent & location of FCD, as well as age of child

Epileptogenic zone often from around rather than within lesion

?Role for ECoG

Role of invasive EEG – grids/SEEG

- Limits of lesion
- Dysplastic tissue often located in eloquent cortical regions
Optimised imaging

Protocols

• Anatomic thin slice volumetric T1
• Axial & coronal T2
• 3D FLAIR

Children <2yrs

• 3D data set,
• Sagittal, axial & coronal T1
• Axial & coronal T2
Are the MRI Findings Specific?

<table>
<thead>
<tr>
<th>MRI Appearance</th>
<th>+</th>
<th>-</th>
<th>Positive %</th>
</tr>
</thead>
<tbody>
<tr>
<td>Subcortical white matter signal change</td>
<td>24</td>
<td>0</td>
<td>100</td>
</tr>
<tr>
<td>Well-defined margins</td>
<td>21</td>
<td>3</td>
<td>87.5</td>
</tr>
<tr>
<td>Blurring of gray-white matter junction</td>
<td>20</td>
<td>4</td>
<td>83.3</td>
</tr>
<tr>
<td>Abnormal cortical gyration/sulcation</td>
<td>20</td>
<td>4</td>
<td>83.3</td>
</tr>
<tr>
<td>Single lobe involvement</td>
<td>20</td>
<td>4</td>
<td>83.3</td>
</tr>
<tr>
<td>Apparent cortical thickening</td>
<td>13</td>
<td>11</td>
<td>54.2</td>
</tr>
<tr>
<td>Signal intensities on MRI scans</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>• Hyperintense on T2W &amp; Hypointense on T1W images</td>
<td>10</td>
<td>14</td>
<td>42</td>
</tr>
<tr>
<td>• Hypointense on T2W &amp; T1W images</td>
<td>8</td>
<td>16</td>
<td>33</td>
</tr>
</tbody>
</table>
Timing of Scan & Maturation

Taylor-type Focal Cortical Dysplasia in Infants: Some MRI Lesions Almost Disappear with Maturation of Myelination

*Christin M. Eltze, †Wui K. Chong, †Sanjay Bhate, †Brian Harding, †Brian G. R. Neville, and *†J. Helen Cross

*Institute of Child Health, University College London, United Kingdom; and †Great Ormond Street Hospital for Children, London, United Kingdom
7T
F 12 yr R frontal seizures
Doughnut Method

1. FreeSurfer Reconstruction
2. Calculation of Cortical Thickness, GM/WM Intensity
   Contrast & FLAIR Signal Intensity

Neural Network

A. 1) feature measurement
   2) intrasubject z-score
   3) registration to average space
   4) control group

B. 1) lesion masks on volumetric T1 and FLAIR
   2) masks in brain template space

Classifier Results

A R L
B R L
C RH
D LH
E RH
Histopathology in MRI Negative Cases
2004 ILAE Pediatric Outcome Survey (N=100)

Positive Etiology 66%

- Cortical Dysplasia: 57%
- Hippocampal Sclerosis: 5%
- Tumor: 2%
- Rasmussen: 2%
- Gliosis/Normal: 34%
FDG-PET/MRI coregistration improves detection of cortical dysplasia in patients with epilepsy

*Salamon et al Neurology 2008;71:1594-1601*
### The role of additional investigations

<table>
<thead>
<tr>
<th>Cohort</th>
<th>II EEG</th>
<th>Video EEG</th>
<th>MRI</th>
<th>3D EEG/MEG</th>
<th>PET</th>
<th>SPECT</th>
<th>ECoG</th>
<th>IEM</th>
<th>Comments</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Single Lesion</strong></td>
<td></td>
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<td></td>
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<tr>
<td><em>Dev Tumors</em></td>
<td>M*</td>
<td>H</td>
<td>M*</td>
<td>O</td>
<td>O</td>
<td>O</td>
<td>O</td>
<td>O</td>
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<tr>
<td><em>FCD I</em></td>
<td>M*</td>
<td>M*</td>
<td>M*</td>
<td>H</td>
<td>H</td>
<td>H</td>
<td>H</td>
<td>H</td>
<td></td>
</tr>
<tr>
<td><em>FCD II</em></td>
<td>M*</td>
<td>M*</td>
<td>M*</td>
<td>O</td>
<td>O</td>
<td>O</td>
<td>O</td>
<td>M/H</td>
<td>O</td>
</tr>
<tr>
<td><em>HS</em></td>
<td>M*</td>
<td>M</td>
<td>M*</td>
<td>O</td>
<td>O</td>
<td>O</td>
<td>O</td>
<td>O</td>
<td>Consider possibility of dual path</td>
</tr>
<tr>
<td><em>SWS</em></td>
<td>M*</td>
<td>M</td>
<td>M*</td>
<td>L</td>
<td>O</td>
<td>O/L</td>
<td>O/L</td>
<td>L</td>
<td></td>
</tr>
<tr>
<td><em>Hyph Hamar</em></td>
<td>M*</td>
<td>H</td>
<td>M*</td>
<td>L</td>
<td>L</td>
<td>L</td>
<td>L</td>
<td>L</td>
<td>IEM not justified</td>
</tr>
<tr>
<td><em>Vascular</em></td>
<td>M*</td>
<td>M</td>
<td>M*</td>
<td>O</td>
<td>O</td>
<td>O</td>
<td>O</td>
<td>O</td>
<td></td>
</tr>
<tr>
<td><em>Post-infec/Ischemic</em></td>
<td>M*</td>
<td>M</td>
<td>M*</td>
<td>O</td>
<td>O</td>
<td>O</td>
<td>O</td>
<td>O</td>
<td>Lesions may be bilateral</td>
</tr>
<tr>
<td><strong>Hemispheric</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
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</tr>
<tr>
<td><em>No Function</em></td>
<td>M*</td>
<td>H</td>
<td>M*</td>
<td>L</td>
<td>L</td>
<td>L</td>
<td>L</td>
<td>L</td>
<td>Possible EEG false lateralization</td>
</tr>
<tr>
<td><em>Function ++</em></td>
<td>M*</td>
<td>M*</td>
<td>M*</td>
<td>H</td>
<td>H</td>
<td>H</td>
<td>H</td>
<td>H</td>
<td>Tailored resection</td>
</tr>
<tr>
<td><em>PMG</em></td>
<td>M*</td>
<td>M*</td>
<td>M*</td>
<td>H</td>
<td>H</td>
<td>O</td>
<td>O</td>
<td>O/H</td>
<td>Tailored resection</td>
</tr>
<tr>
<td><em>Rasmussen</em></td>
<td>M*</td>
<td>M</td>
<td>M*</td>
<td>L</td>
<td>L</td>
<td>L</td>
<td>L</td>
<td>L</td>
<td>Serial MRI required</td>
</tr>
<tr>
<td><em>TS</em></td>
<td>M*</td>
<td>M*</td>
<td>M*</td>
<td>H/O</td>
<td>O</td>
<td>H</td>
<td>H</td>
<td>H/O</td>
<td>AMT PET useful</td>
</tr>
<tr>
<td><strong>MRI negative</strong></td>
<td>M*</td>
<td>M*</td>
<td>M*</td>
<td>H</td>
<td>H</td>
<td>H</td>
<td>H</td>
<td>H</td>
<td>Serial Tests</td>
</tr>
</tbody>
</table>

*Jayakar et al Epilepsia 2014; 55(4):507–518,*
Treatment Paradigm GOSH

- Evidence of structural lesion?
  - MRI, SPECT, PET, MRS

- Is the lesion focal or diffuse?
  - Imaging & Neurophysiology (EEG, MEG, fMRI)

- Is intracranial monitoring required?

- Acute vs Chronic

- Map Eloquent Cortex
Evaluation Protocol

Clinical history/exam
   Video EEG capture
      Ictal & interictal
   High resolution MRI
   Neuropsychology

Preliminary review

Optional tests
   HS
   Sturge Weber
   Discrete tumours
   FCD II
   AVM Stroke

Highly recommended testing
   FCDI
   TS
   Hemispheric lesion w/function

Mandatory testing
   Non-lesional
   Divergent vEEG/MRI
   Multilesional
   Malformation by eloquent cortex (minimal resection)

No additional test
   HT Hamartoma
   Progressed Rasmussens
   Hemisphere lesion w/no residual function
   Non resective cases

*Jayakar et al Epilepsia 2014; 55(4):507–518,*
No additional tests

**PET**
- Yes: Non lesional
  - Divergent data
  - MRI are sufficient
- No: vEEG and MRI are sufficient
  - Lesion extent is uncertain
  - Prior resection
  - Large lesion

**SPECT**
- Yes: TS
  - Prior resection
  - Large lesion & preserved function
- No: PET sufficient
  - Multifocal
  - Not feasible
  - PET/MRI/vEEG are discordant

**EEG Source localisation**
- Yes: Focal interictal
  - Ictal onset discharges
- No: Hemispherectomy
  - Generalised discharges

**MEG**
- Yes: Focal tangential
  - Interictal spikes
  - Post-op cases
- No: 3D source is sufficient

**fMRI or MEG**
- Yes: If need to define eloquent cortex

**Wada**
- Yes: If older child and memory is a concern

**Epilepsy Conference**
Non resective or no surgery

- Multifocal
- Generalised
- Eloquent cortex

One stage

- No ECog
  - Hemispherectomy
  - HT hamartoma
  - Non-FCD lesions without a clear penumbra

- ECog
  - FCD
  - Non FCD lesions without a clear penumbra

Two stage

- Inconsistent localisation
- Divergent localisation

Mapping

- Extra-operative
- Intra-operative

Jayakar et al Epilepsia 2014; 55(4):507–518,
Threshold for invasive evaluation

Extent of resection
Plasticity
Choosing an invasive strategy

<table>
<thead>
<tr>
<th></th>
<th>SUB –DURAL GRID</th>
<th>sEEG</th>
</tr>
</thead>
<tbody>
<tr>
<td>MRI negative</td>
<td>✗</td>
<td>✓</td>
</tr>
<tr>
<td>Multiple lesions</td>
<td>✓</td>
<td>✓✓</td>
</tr>
<tr>
<td>Deep structures involved</td>
<td>✗</td>
<td>✓</td>
</tr>
<tr>
<td>Defining limits of cortical malformations</td>
<td>✓</td>
<td>✗</td>
</tr>
<tr>
<td>Functional mapping</td>
<td>✓</td>
<td>✗</td>
</tr>
<tr>
<td>Morbidity</td>
<td>✗</td>
<td>✓✓</td>
</tr>
</tbody>
</table>
• FTND
• Day 1: Twitching right arm and leg
• Day 6: Jerking right, spread to involve both sides
• Short, frequent. Need for rescue medication
• Further seizures subtle behaviour change, eye flickering, deviation, some with right upper limb involvement; 50-100/day
• PB, CBZ, VPA, CLB, PHT, VGB, LVT
• Clonazepam infusion x2
• At 10 weeks unable to wean CLN infusion
• When well, fixing, following, smiling
Interictal

Ictal
• Age 8 years
• FTND; no early concerns
• First seizure age 21m; prolonged
  • Right focal UL>LL, speech affected, 2 to 4/night
  • Right focal with sec Generalised, 1 / fortnight
  • Right focal (face), mild and with aura, 1 to 2/week
• Variable upper limb function but no fine finger movement since presentation
• Multiple medications
• MRI: cortical dysplasia
• Decision made ‘not surgical candidate’
• VNS inserted
  • No benefit

• Continued seizures; cognitively low average but days where less interactive, poor oral intake and drooling
Ictal Findings

- Most consistent ictal onset zone
- Broader field of ictal onset at times
- Marked flattening post-ictally
- Post-ictal fast (+/- spasms) in bursts and at times longer trains of self-sustained re-activation of fast
Functional Stim & Ictal Onset

Motor Hand/Arm
Motor Face/mouth
Sensory face/mouth
Ill-defined sensory face?
Minor clinical or electrographic seizure
Nil

ABSENT SEP
Posterior to motor hand
14 yr old boy
- First seizure 2 yrs
- Warning, appears agitated, fumbles, noncommunicative
- 4 previous AEDs
- Mainstream education
- Increasing difficulty
Surface EEG

- Mild background slowing
- Occ discharges over the left temporal region in sleep
- Three similar seizures; EEG changes lagged behind clinical change
- Attenuation at onset in two & right fronto temporal discharges late in event in third
Right hand motor

Hospital Number: 819690
Date of Birth: 24/03/92
Date of exam: 12/05/06
No lesion?

6 year old boy, seizure onset 3 years, cluster of seizures, aura with partial awareness – long seizure free periods. Developmentally normal

• Seizures fairly stereotypic

Short events
• Behavioural arrest
• Leans to side and grabs parents

Long events
• Behavioural arrest
• Rubbing nose in the pillow
• Flipping over
• Thrashing movements.
Onset of seizure - slow activity over Right frontal and Ant. Temporal region.

EEG lateralises to R and localises to R frontal/ant. temporal region.
Magnetoencephalography

Virtual Electrode Time Series
Ictal EEG
Abnormality found in Surgery – adjacent to DA03 and running posteriorly

DA03 with Abnormality (greyish colour of cortex)
3D Reconstruction

A – A-IFG 1-10
B – M-MFG-CING 1-18
C – M-IFG-INS 1-10
D – P-MFG 1-8
E – P-IFG-INS 1-10
F – Parietal 1-18
G – S1 1-15
H – SMA 1-12
I – A-SFG 1-8
J – A-MFG 1-15
Seizure From sleep: D1-2 rhythmic
Seizure continued – spread H1-7, B6-10, G8-10
end
Functional Stim & Ictal Findings

A – Ant IFG 1-10

B - Mid MFG-Cing 1-18

C – Mid IFG-Ins 1-10

D – Post MFG 1-8

E – Post IFG-INS 1-10

F – Parietal 1-18

G – S1 1-15

H – SMA 1-12

I – Ant SFG 1-8

J – Ant MFG 1-15

Faulty contact/outside of cortex

Ictal Findings
- Initial change
- Enhanced discharges
- Subsequent spread

Motor
Seizure outcome following surgery

<table>
<thead>
<tr>
<th></th>
<th>FCD type 1 (%)</th>
<th>FCD type 2 (%)</th>
<th>FCD type 3a (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Engel class I</strong></td>
<td>37 (56)</td>
<td>52 (61)</td>
<td>34 (64)</td>
</tr>
<tr>
<td><strong>Engel class Ia</strong></td>
<td>32 (48)</td>
<td>42 (49)</td>
<td>26 (49)</td>
</tr>
<tr>
<td><strong>Engel class II</strong></td>
<td>11 (17)</td>
<td>13 (15)</td>
<td>10 (19)</td>
</tr>
<tr>
<td><strong>Engel class III</strong></td>
<td>12 (18)</td>
<td>13 (15)</td>
<td>3 (6)</td>
</tr>
<tr>
<td><strong>Engel class IV</strong></td>
<td>6 (9)</td>
<td>7 (8)</td>
<td>6 (11)</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th></th>
<th>FCD type 1 (%)</th>
<th>FCD type 2 (%)</th>
<th>FCD type 3a (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Engel class I</strong></td>
<td>17 (61)</td>
<td>26 (67)</td>
<td>17 (65)</td>
</tr>
<tr>
<td><strong>Engel class Ia</strong></td>
<td>13 (46)</td>
<td>22 (56)</td>
<td>15 (58)</td>
</tr>
<tr>
<td><strong>Engel class II</strong></td>
<td>4 (14)</td>
<td>4 (10)</td>
<td>4 (15)</td>
</tr>
<tr>
<td><strong>Engel class III</strong></td>
<td>5 (18)</td>
<td>7 (18)</td>
<td>2 (8)</td>
</tr>
<tr>
<td><strong>Engel class IV</strong></td>
<td>2 (7)</td>
<td>2 (5)</td>
<td>3 (11)</td>
</tr>
</tbody>
</table>

There was no statistically significant difference between FCD types 1, 2, and 3a concerning postoperative outcome (log-rank test p = 0.46). *Statistically significant value.

Fauser et al Epilepsia 2015;56:66-76
Mutations in Mammalian Target of Rapamycin Regulator DEPDC5 Cause Focal Epilepsy with Brain Malformations

Scheffer et al Ann Neurol 2014;75: 782-787

Familial Focal Epilepsy with Focal Cortical Dysplasia Due to DEPDC5 Mutations

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Ann Neurol 2015;77:675–683
Conclusions

• Focal cortical dysplasia most common pathology in paediatric surgical series
  • Challenges & rewards
  • Early referral required for consideration of surgery

• Structured approach to evaluation within complex epilepsy team

• Optimise information available prior to surgical decision

• Specific consideration to need or type of invasive evaluation that may be required