

Cortical Dysplasia

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Malformations of Cortical Development

absent or abnormally broad gyri: lissencephaly

Agyria/
pachygyria

excessive folding
of an abnormally
thin cortex

Poly-
micro-
gyria

Areas of
atypical
cortical
structure

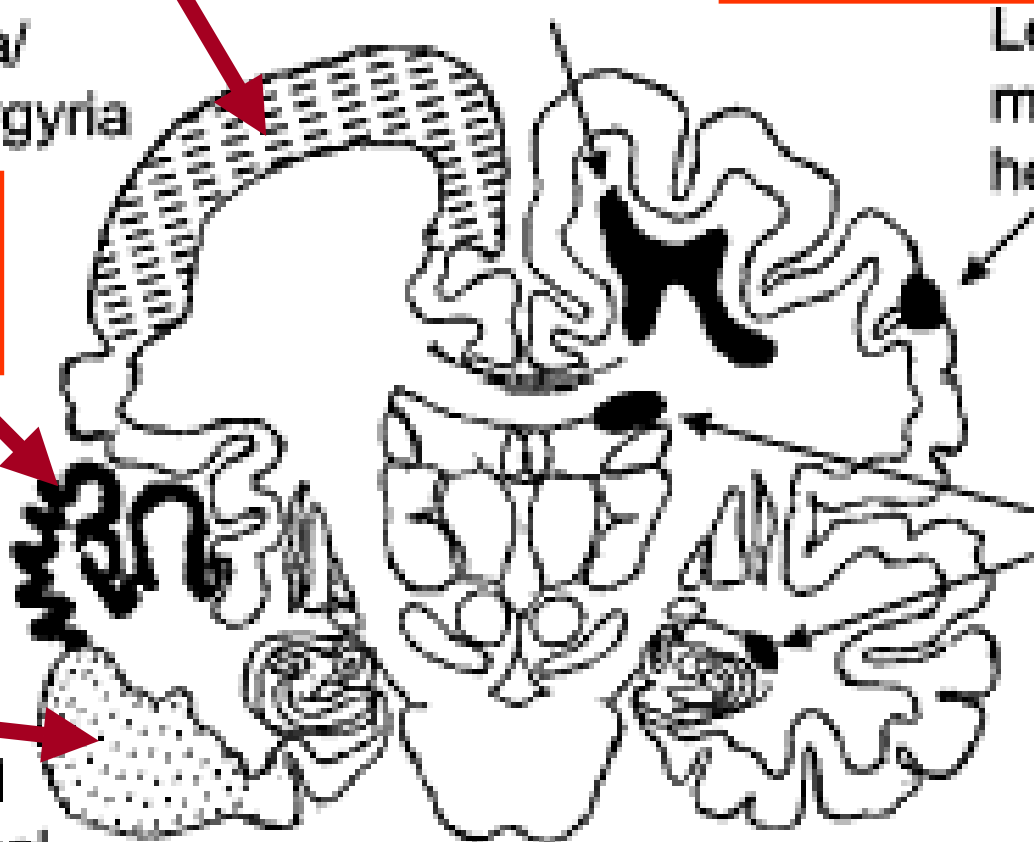
Focal
cortical
dysplasia

Laminar
heterotopia

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

Lepto-
meningeal
heterotopia

Peri-
ventricular
nodular
heterotopia



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. Palmi, 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. Prayson, 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 (dyslaminations)
- 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 epileptogenic, high seizure frequency,

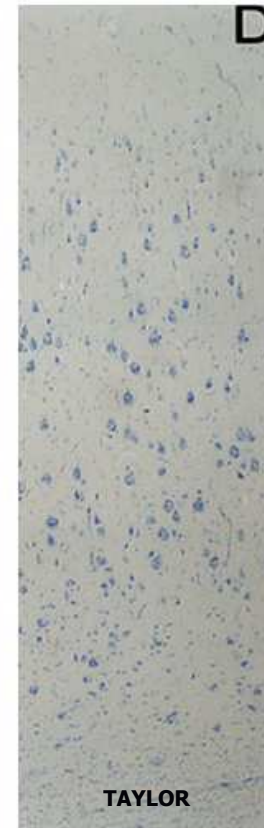
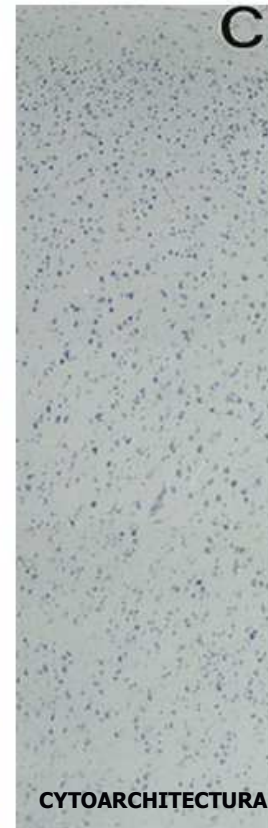
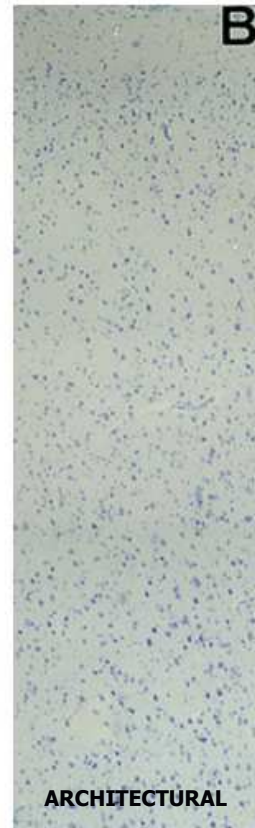
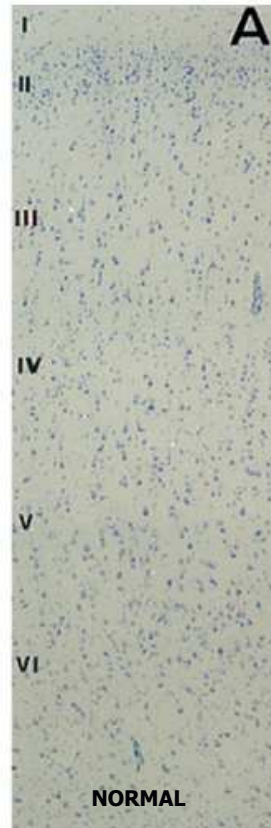


Normal

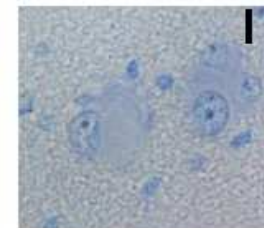
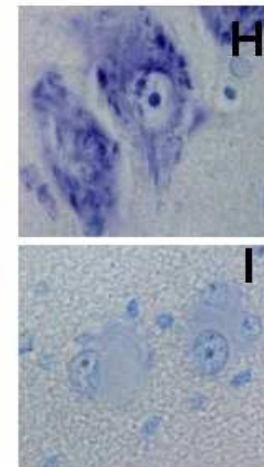
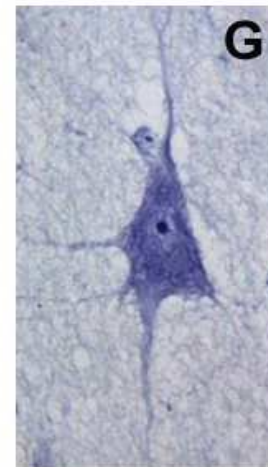
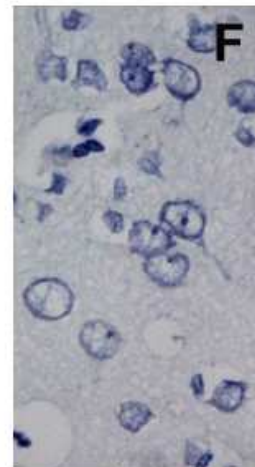
Type IA and IB

Type IIA/B

STRUCTURAL
ABNORMALITIES

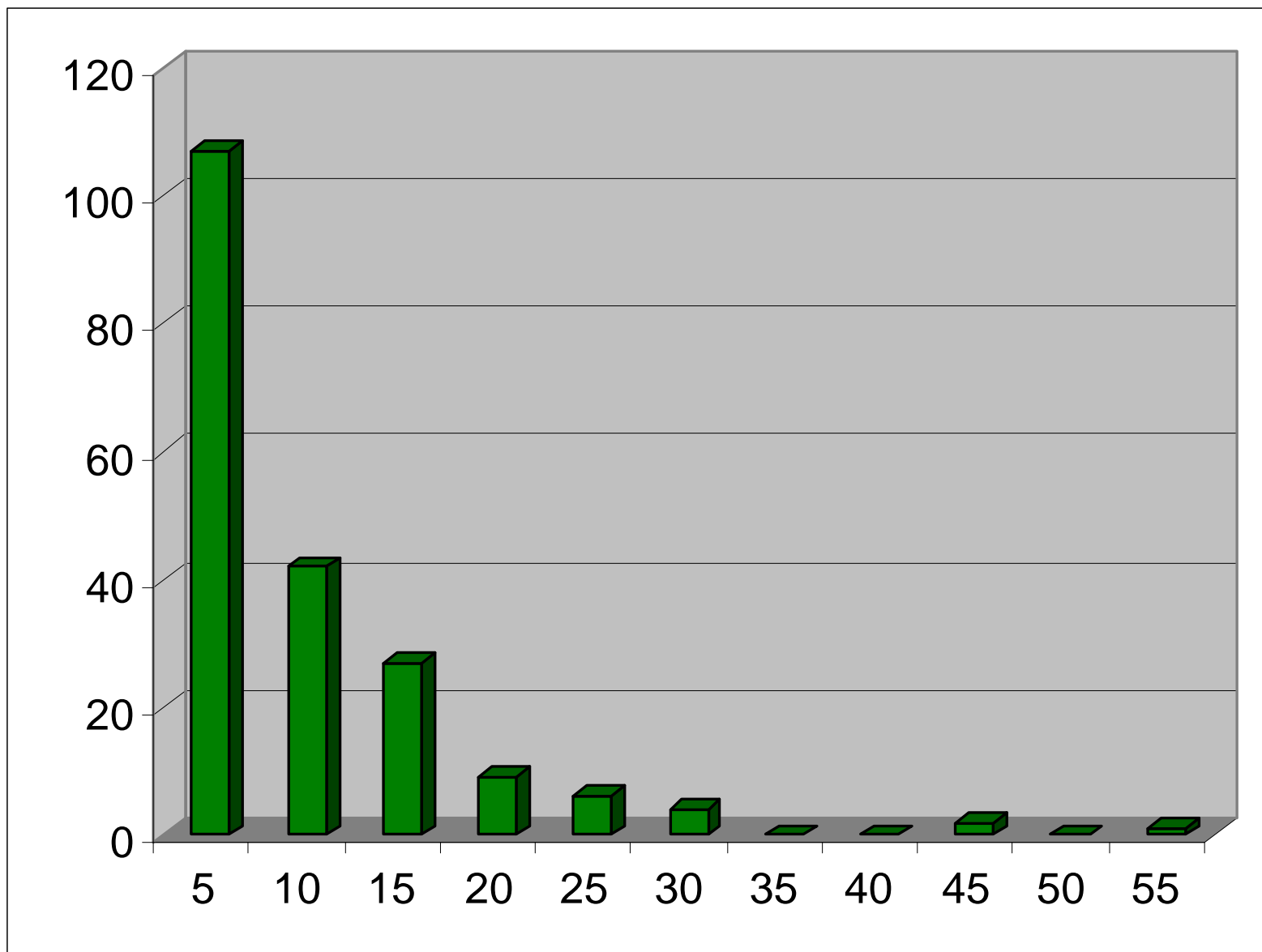


CYTOLOGIC
ABNORMALITIES



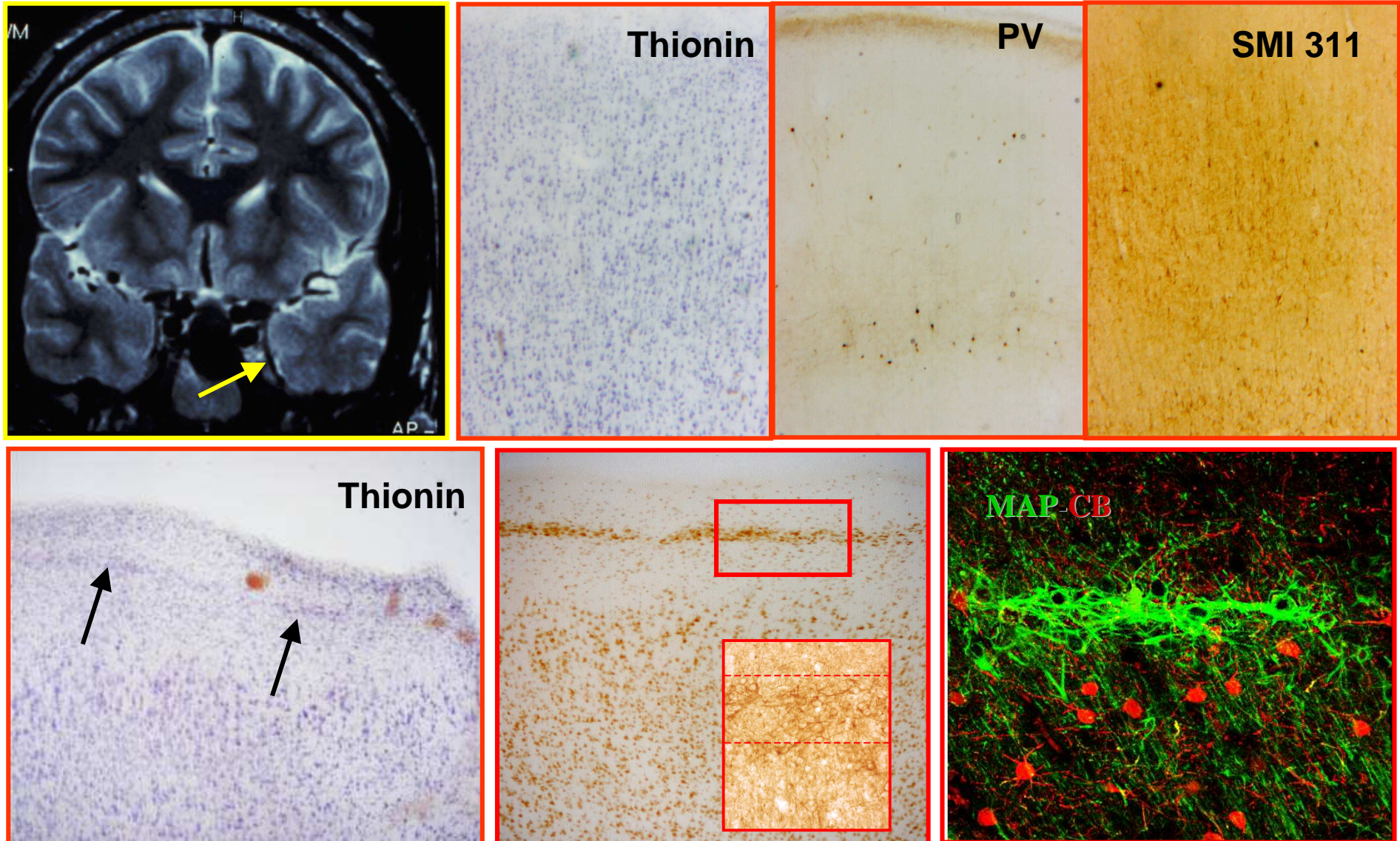


Seizure onset; FCD in 198 patients



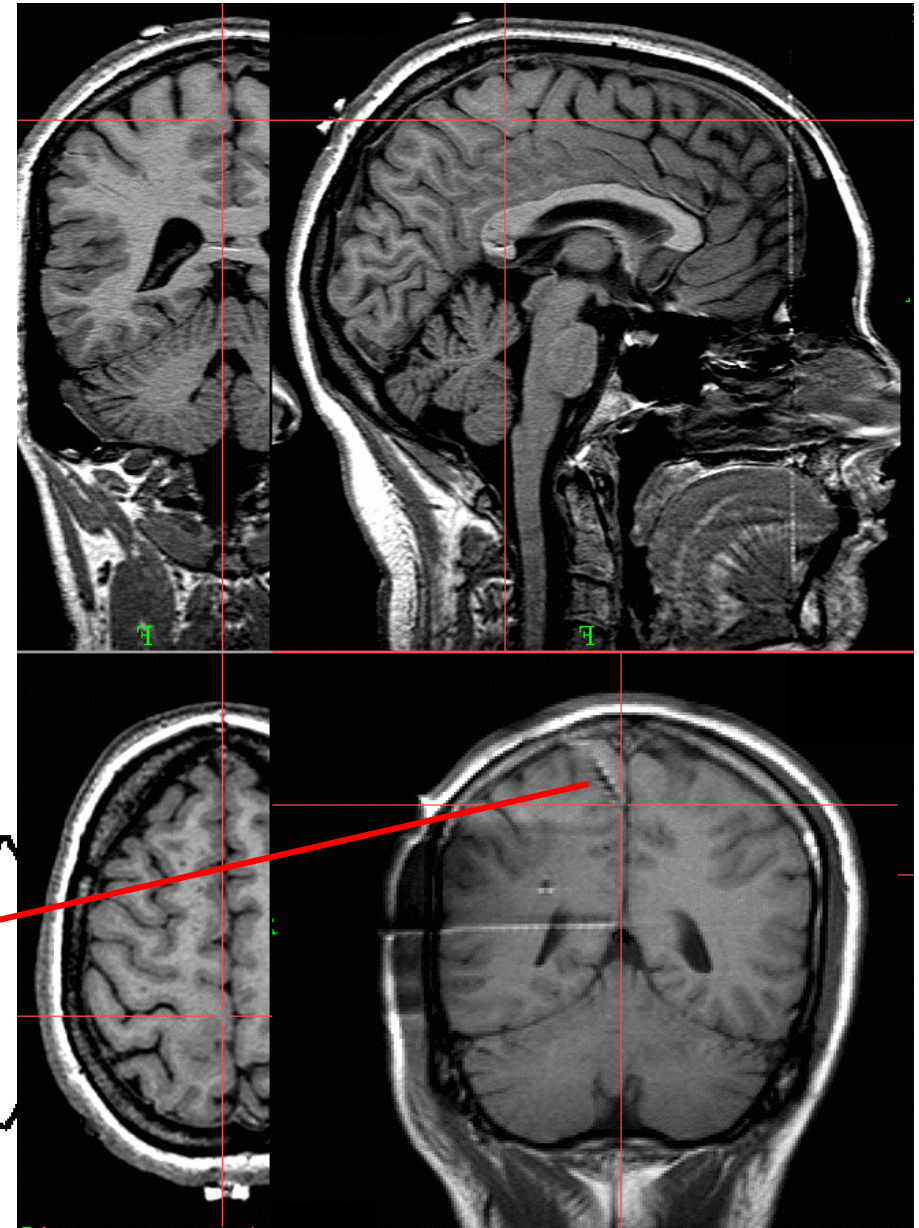
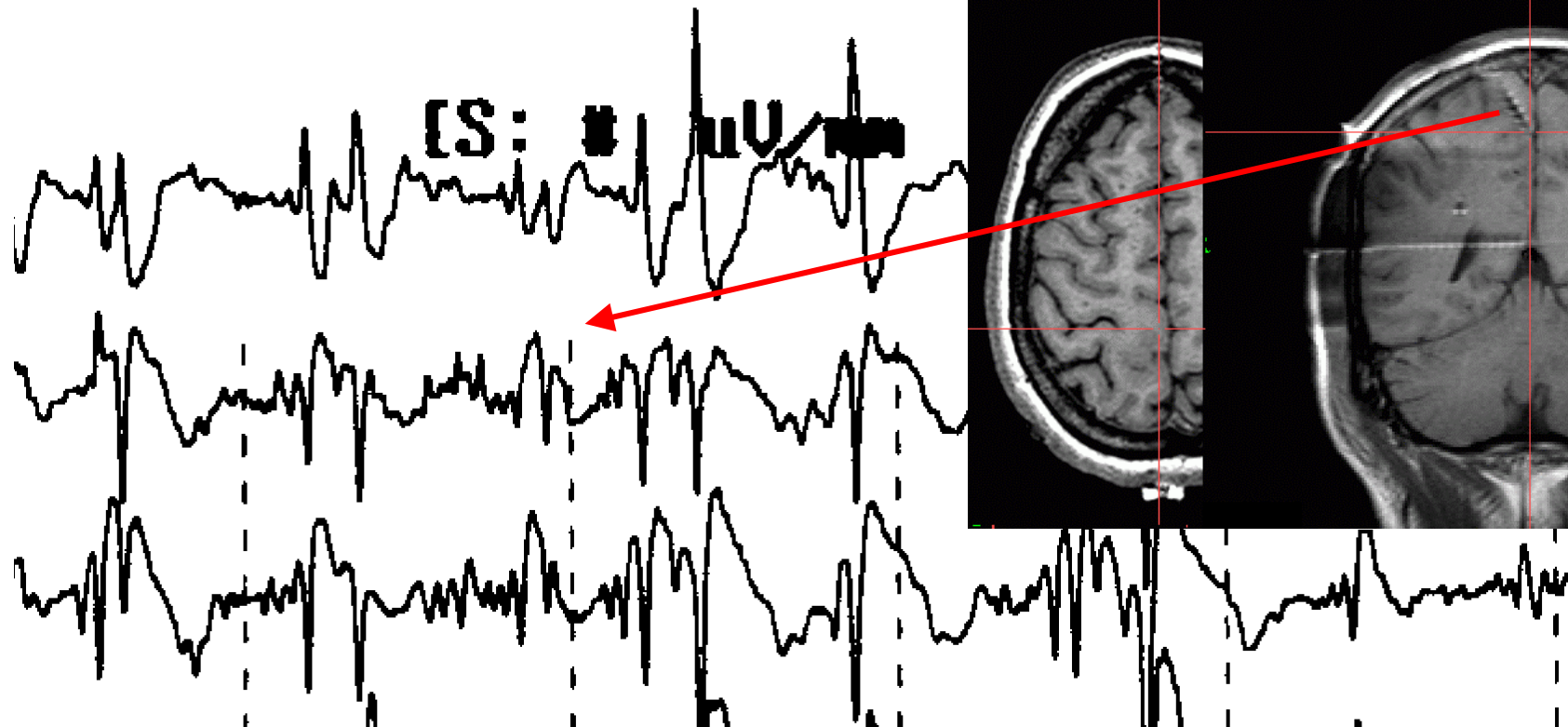


Type IA : isolated architectural abnormalities (dyslamination)





**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)

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



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