

# **SEIZURE MANAGEMENT IN CHILDREN**

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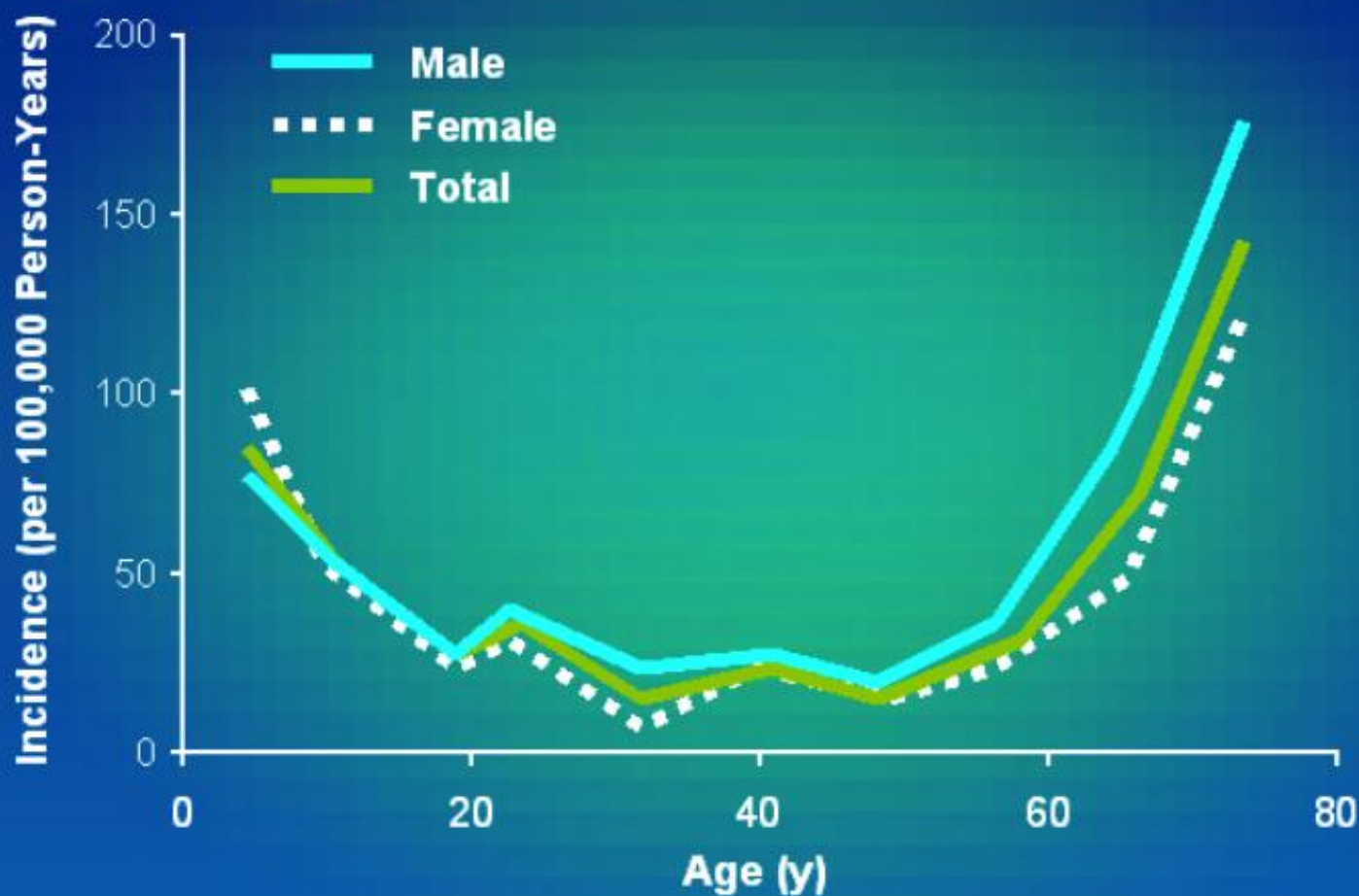
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# EPILEPSY

- **AFFECTS 0.5%-1% OF POPULATION**
- **70% OF EPILEPTICS CAN BE CONTROLLED WITH MEDICATION (USUALLY BY THE 3<sup>rd</sup> 'REASONABLE' MEDICATION ATTEMPTED)**
- **OF THE REMAINING 30%, ABOUT HALF (15%) ARE SURGICAL CANDIDATES**
- **THE OTHER 15% ARE NEITHER CONTROLLED NOR CAN BE HELPED WITH RESECTIVE SURGERY**
- **NEW MEDICATIONS, KETOGENIC DIET, VAGAL NERVE STIMULATOR, OTHER SURGICAL TECHNIQUES**

# Epilepsy Incidence: 1935–1984



# Common Etiologies of Seizures Change With Age

- Children
  - Febrile seizures
  - Congenital causes
  - Metabolic causes
- Young adults
  - Trauma
  - Tumor
- Elderly
  - Stroke
  - Degenerative changes

# PRIMARY GENERALIZED VERSUS PARTIAL (FOCAL) SEIZURES

- PRIMARY GENERALIZED SEIZURES HAVE A GENERALIZED ONSET ON EEG, i.e., NO FOCUS
  - CAN BE CONVULSIVE (e.g., tonic, tonic-clonic) OR NONCONVULSIVE (e.g., petit mal absence)
- PARTIAL SEIZURES HAVE A FOCAL ONSET.
  - CAN BE SIMPLE PARTIAL (no LOA) OR COMPLEX PARTIAL (with LOA)
  - CAN GENERALIZE INTO CONVULSION (i.e., secondary generalization)
- MYOCLONIC SEIZURES AND ATONIC SEIZURES CAN BE EITHER PARTIAL OR PRIMARY GENERALIZED









# SHOULD I TREAT FIRST SEIZURE?

GENERALLY NO, BUT YES IF:

- SEIZURE IS PROLONGED. ALSO PRESCRIBE RESCUE MEDICATION (e.g., DIASTAT, MIDAZOLAM NASAL, CLONAZEPAM ODT)
- FOCAL FEATURES, e.g., TODD'S PARALYSIS or FOCAL ONSET
- STRONG FAMILY HISTORY OF EPILEPSY
- HIGHLY EPILEPTIFORM EEG
- UNDERLYING NEUROLOGICAL DISEASE





# SEIZURE MEDICATIONS

- **FOCAL SEIZURES: OXCARBAZEPINE, LACOSAMIDE, LEVETIRACETAM**
- **PRIMARY GENERALIZED SEIZURES**
  - **CONVULSIVE: VALPROIC ACID, LAMOTRIGINE, LEVETIRACETAM, ZONISAMIDE, CLOBAZAM, PERAMPANEL**
  - **NON-CONVULSIVE (TYPICAL ABSENCE, “PETIT MAL”): ETHOSUXIMIDE, VALPROIC ACID, LAMOTRIGINE**
  - **NON-CONVULSIVE (ATYPICAL ABSENCE): VALPROIC ACID, CLOBAZAM, RUFINAMIDE, KETOGENIC DIET, TOPIRAMATE, ZONISAMIDE**
- **ATONIC SEIZURES: CLOBAZAM, VALPROIC ACID, KETOGENIC DIET, TOPIRAMATE, ZONISAMIDE, RUFINAMIDE**
- **MYOCLONIC SEIZURES: CLOBAZAM, VALPROIC ACID, TOPIRAMATE, CLONAZEPAM, ZONISAMIDE, RUFINAMIDE**

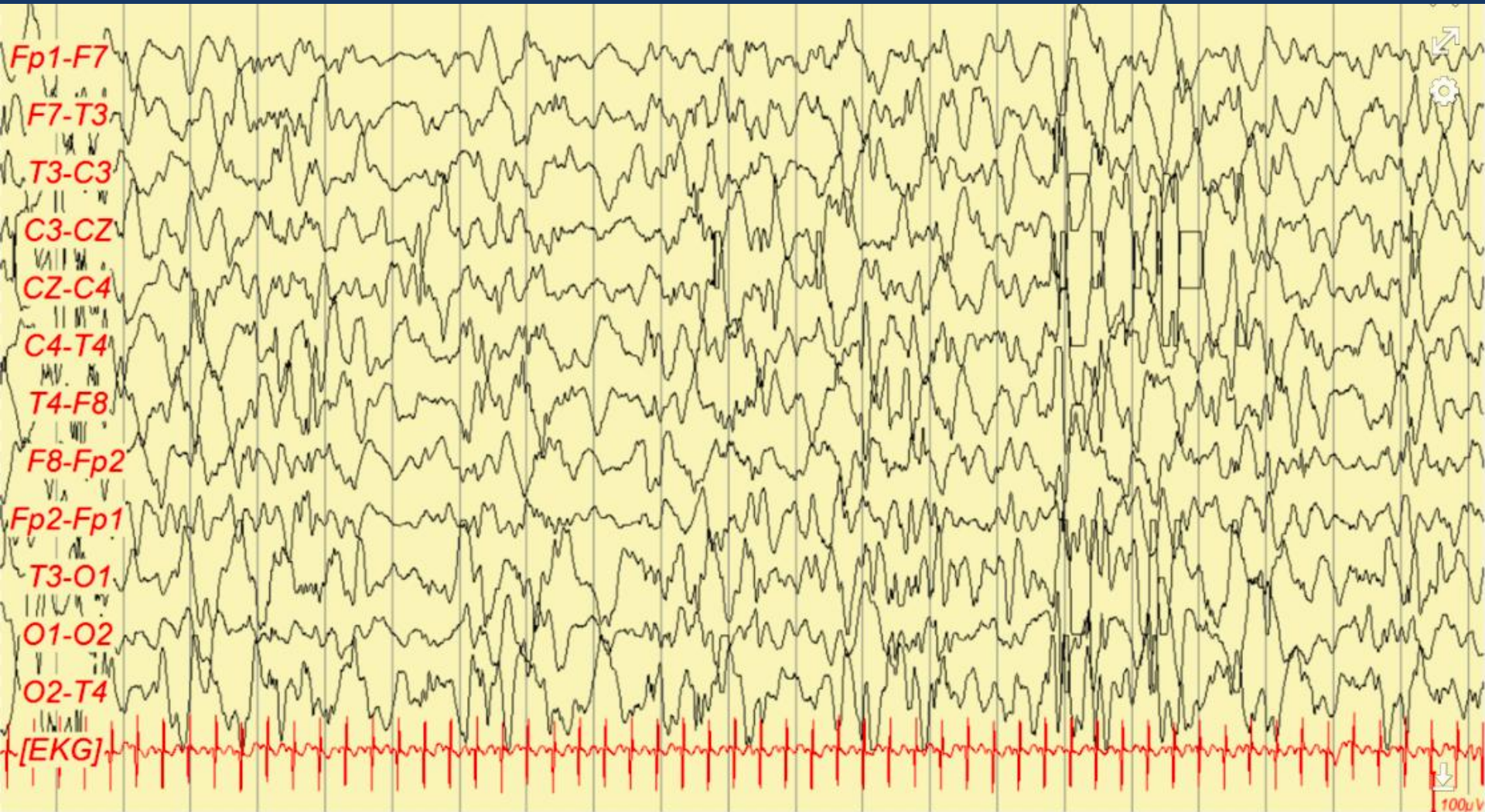
# INFANTILE SPASMS

- **SUDDEN FLEXION, EXTENSION, OR MIXED FLEXION-EXTENSION JERKS OF THE ARMS, HEAD, TRUNK, LEGS**
- **ONSET 3-8 MONTHS, USUALLY DISAPPEARS BY 3 YEARS**
- **OFTEN MISTAKEN FOR COLIC, REFLUX, STARTLE, HICCUPS**
- **COMES IN CLUSTERS OFTEN WHEN WAKING UP**
- **SPASMS + HYPERSARRHYTHMIA ON EEG + DEVELOPMENTAL ARREST = WEST SYNDROME**

# INFANTILE SPASMS



# HYPARRHYTHMIA



# INFANTILE SPASMS: Etiology

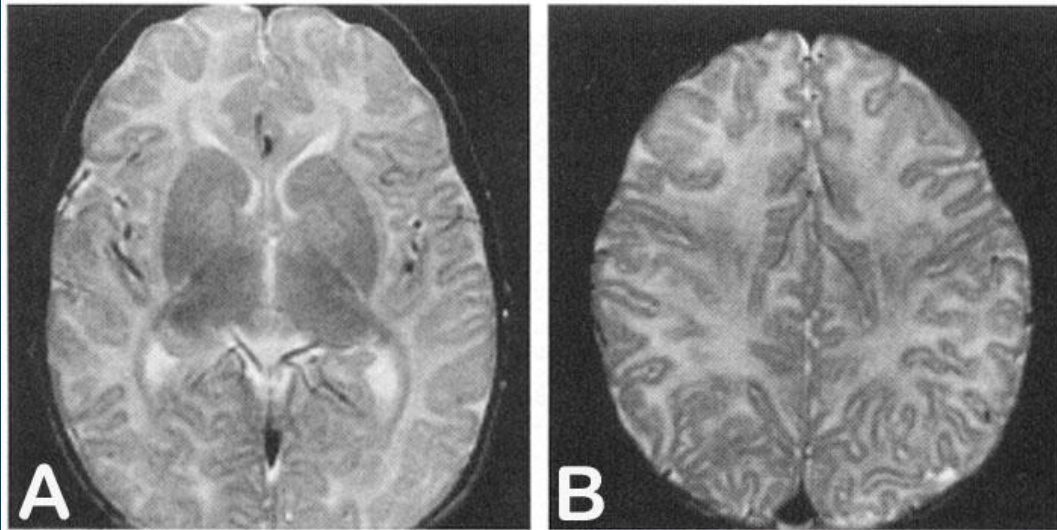
- MALFORMATIONS OF CORTICAL DEVELOPMENT
- PRENATAL, PERINATAL AND POSTNATAL INJURIES (stroke, infections, hypoxia-ischemia)
- CHROMOSOMAL DISORDERS, e.g., DOWN SYNDROME
- NEUROCUTANEOUS DISORDERS, e.g., TUBEROUS SCLEROSIS, NF1, STURGE-WEBER
- INBORN ERRORS OF METABOLISM
- TUMORS
- VARIOUS SYNDROMES, e.g., AICARDI, PEHO
- PYRIDOXINE DEFICIENCY
- ARX, CDKL5, MANY MUTATIONS

# INFANTILE SPASMS:

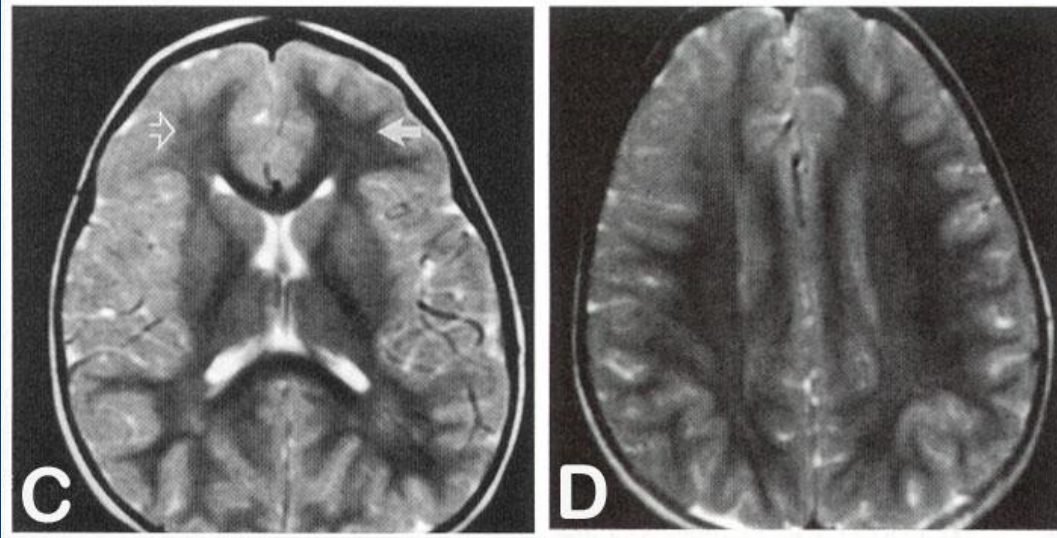
## Partial list of gene mutations

- ARX
- CDKL5
- DNM1
- EEF1A2
- FOXP1
- GRIN1
- GRIN2A
- KCNQ2
- KCNT1
- SCN2A
- SCN8A
- SIK1
- SLC25A22
- SPTAN1
- STXBP1
- TSC1
- TSC2
- MEF2C
- DCX
- LIS1
- TUBA1A
- MAG12
- ATP7A
- KCNJ11
- GLDC
- PAH

# Focal CD, Right Frontal Lobe



**At 6 months of age**



**At 3 years of age**

**Sankar R, AJNR, 1995**

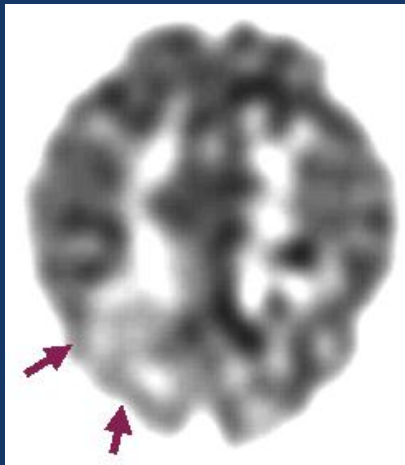




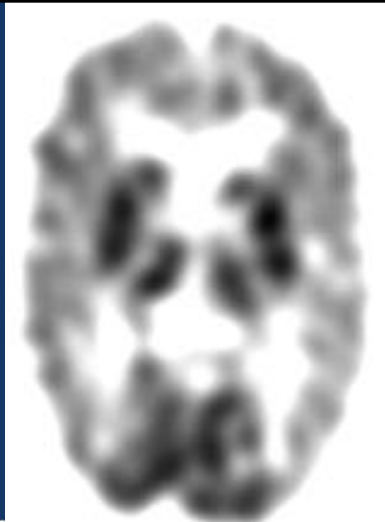
# INFANTILE SPASMS: MEDICAL TREATMENT

- PYRIDOXINE
- ACTH V. PREDNISILONE
- VIGABATRIN
- KETOGENIC DIET
- ZONISAMIDE
- BENZODIAZEPINE (NITRAZEPAM, CLOBAZAM)
- TOPIRAMATE
- VALPROIC ACID
- GOOD RESPONSE: PREMATURE INFANTS,  
NEUROFIBROMATOSIS 1,  
DOWN SYNDROME WITH HYPARRHYTHMIA

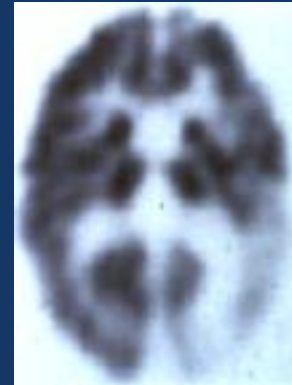
# Infantile Spasms: Metabolic Patterns



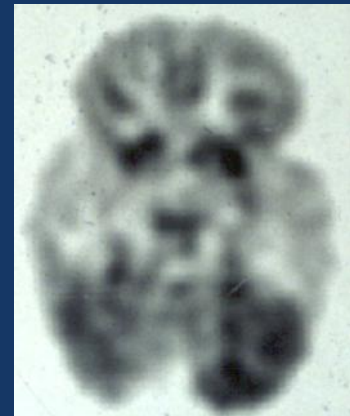
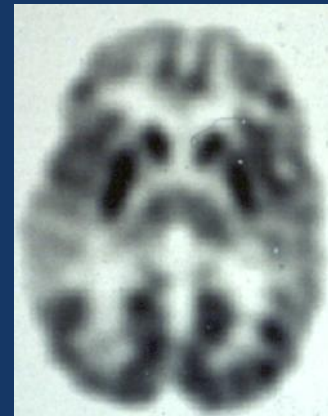
**Surgical Candidate - 20%**



**Metabolic/Neurogenetic - 5%**



**Multifocal - 65%**

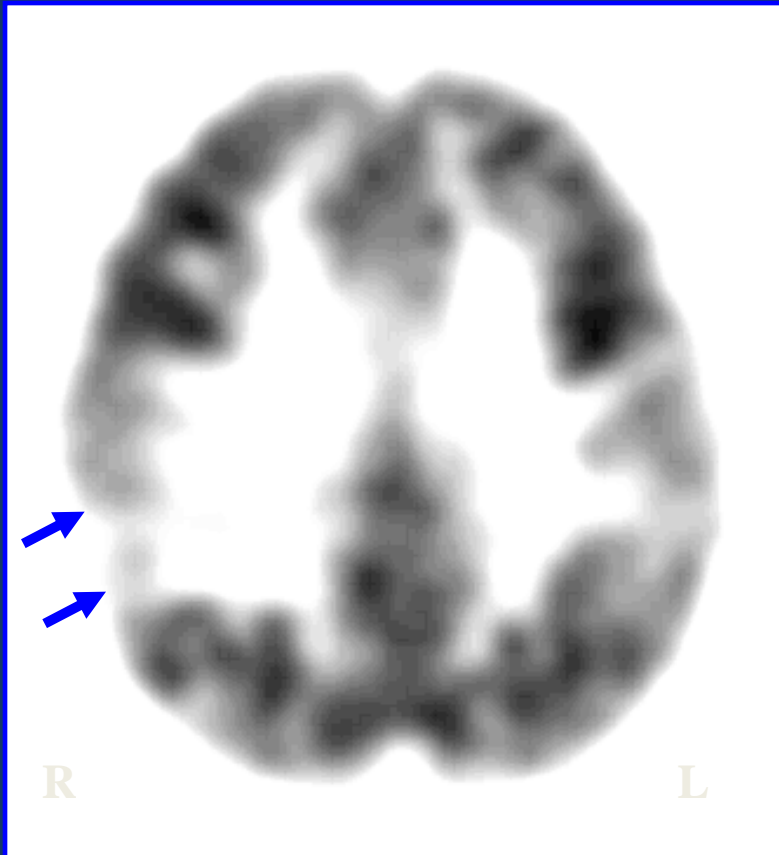


**Bitemporal/Autism - 10%**

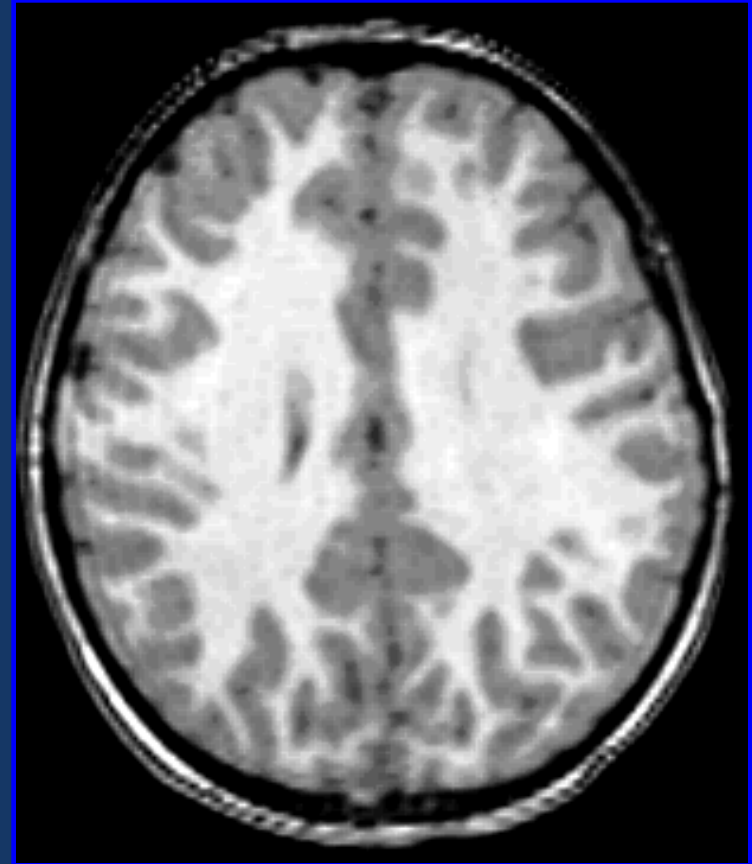
# Normal MRI

Scalp EEG: ictal onset in right central region (C4-F4)

**FDG PET**



**MRI**



# WHY PERFORM EPILEPSY SURGERY IN INFANTS & YOUNG CHILDREN?

- **INTRACTABLE EPILEPSY IN THIS AGE GROUP OFTEN RESULTS IN AN EPILEPTIC ENCEPHALOPATHY**
- **SEIZURE MEDICATIONS ALSO HAVE COGNITIVE SIDE EFFECTS**
- **EPILEPSY SURGERY SHOULD BE CONSIDERED PARTICULARLY WHEN THE DEVELOPMENTAL LAG IS WIDENING**
- **SURGICAL RESULTS ARE BEST WHEN THERE IS CONCORDANCE BETWEEN EEG AND NEUROIMAGING LOCALIZATION, AND WHEN THE SURROUNDING AND CONTRALATERAL BRAIN REGIONS ARE NORMAL**

# EPILEPSY SURGERY

- TEMPORAL LOBECTOMY
- EXTRATEMPORAL FOCAL RESECTION
- MULTILOBAR RESECTION
- SUBTOTAL HEMISPHERECTOMY
- HEMISPHERECTOMY
- CORPUS CALLOSOTOMY
- MULTIPLE SUBPIAL TRANSECTIONS
- DEEP BRAIN STIMULATION
- RESPONSIVE NEUROSTIMULATION  
(NEUROPACE)

**THE FUTURE IS HERE!**

3T whole-body PET/MR scanner allowing simultaneous acquisition of PET and MR data

