

# Hemihelp

## Management of Seizures in Children with Hemiplegia

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The National Centre  
for Young People  
with Epilepsy

**Great Ormond  
Street Hospital**

***NHS***

**Institute of  
Child Health**



**The  
child  
first  
and  
always**

## Predominant brain lesions in congenital hemiplegia related to time of insult

<u>Stage of development</u>	<u>Type of lesion</u>
Early fetal life	Cerebral malformations
Very preterm	Periventricular hemorrhagic infarction (PHI)
Preterm	Periventricular leukomalacia (PVL)
Late preterm	Subcortical leukomalacia (SCL)
Term	Cortical infarctions

# Brain lesions in 111 patients with congenital hemiplegia. CT findings (Wiklund et al. 1990)

<u>CT findings</u>	<u>Preterm (n=28)</u>	<u>Term (n=83)</u>
Maldevelopment	5	14
Periventricular lesion	14	33
Cortical/subcortical lesion	2	11
Miscellaneous	0	3
Normal	7	22

# Extra-uterine

- Meningitis
- Encephalitis
- Head injury
- Rasmussen's encephalitis

# Grey matter impairments

- Cognitive
- Epilepsy
- Behaviour - ADHD  
ASD  
Confrontational

# Poor prognostic factors for seizure remission

- Symptomatic seizures
- Cognitive impairment
- > 1 sz type
- Neurological signs
- Status epilepticus

# Seizures in congenital hemiplegia

- 25% - 35%
- Focal motor
- Startle seizures
- Atonic drop attacks (negative myoclonus)
- Myoclonic jerks
- Infantile spasms (Eg. TS)



# Startle seizures (25-85% with hemiplegia)

- v.common in congenital hemiplegia
- Ischaemic pathology especially perinatal
- Auditory, tactile, stumbling
- Exaggerated startle  $\pm$  posturing

# Epileptic encephalopathies

- Congenital in hemimegalencephaly
- ESES in perisylvian polymicrogyria  
regression (Sleep EEG)  
psychiatric abnormality  
psychosis
- Benzo, corticosteroids, ketogenic diet, surgery

# AED Treatment

- 1 or 2 drugs
- Cbz, Lung, Top, Lev, Val
- Risk of polytherapy
- Behavioural S.E.
- Rescue treatment for CSE

# Surgical treatment

- Resection/disconnection
- Functional hemispherectomy/lobar resections
- Intractable to AEDs. (sz or ESES)
- Identifiable source
  - MRI
  - EEG (?)
- Safely removable/disconnection
  
- Priority of seizures
- Behaviour

# Issues about hemispherectomy

- Early onset assessment (hemimeg/TS)
- Motor deficit
- Visual fields
- Walking/rehab
- Bilateral damage - clinical  
- MRI
- Recurrence of seizures

# Outcome

- Seizure relief      50-60% malformations  
80-90% mca

Behaviour

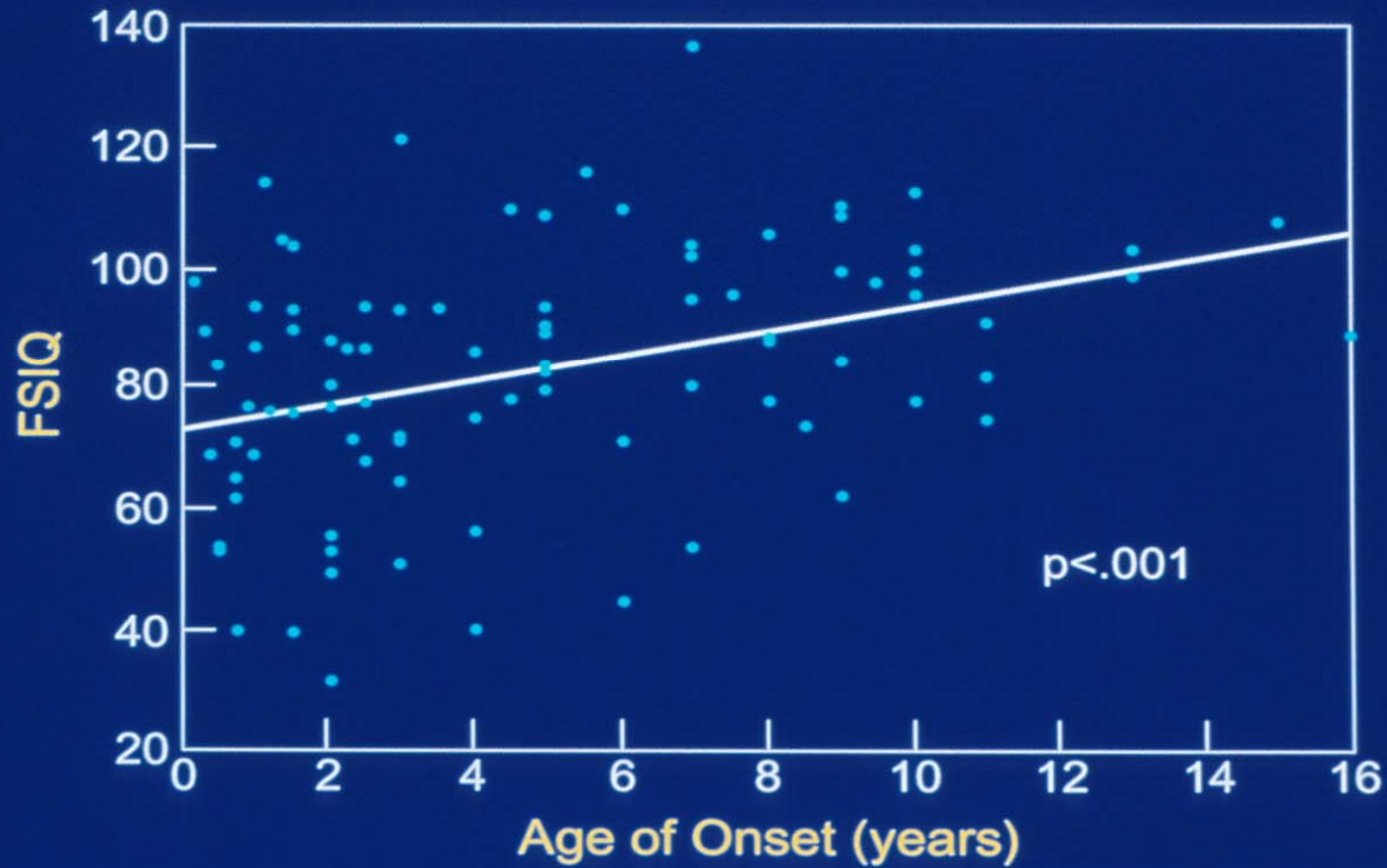
Cognition

Complications

- hydrocephalus
- haemosiderosis

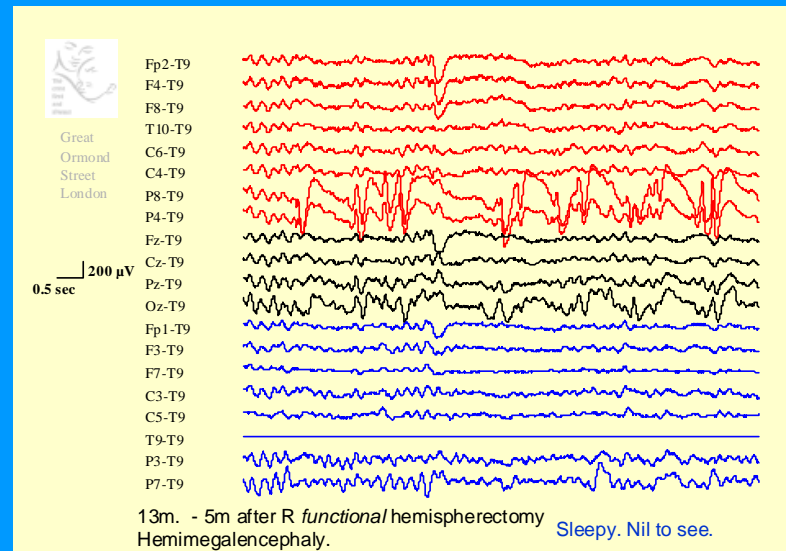
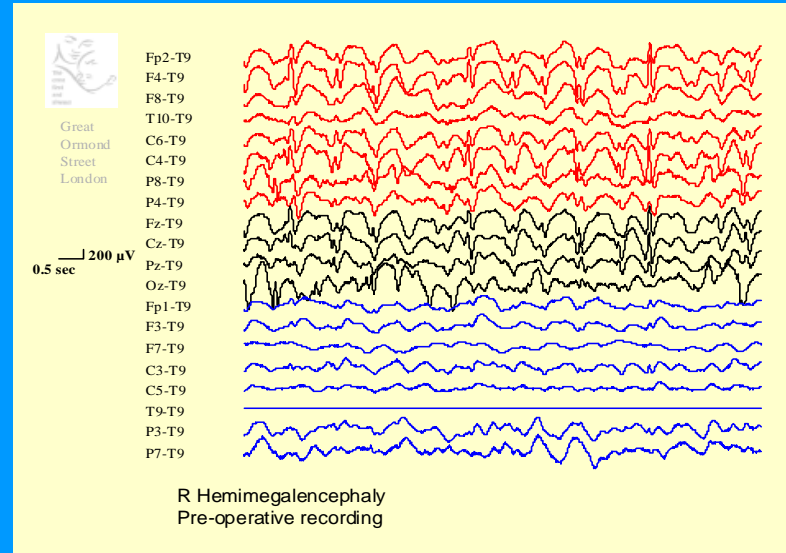
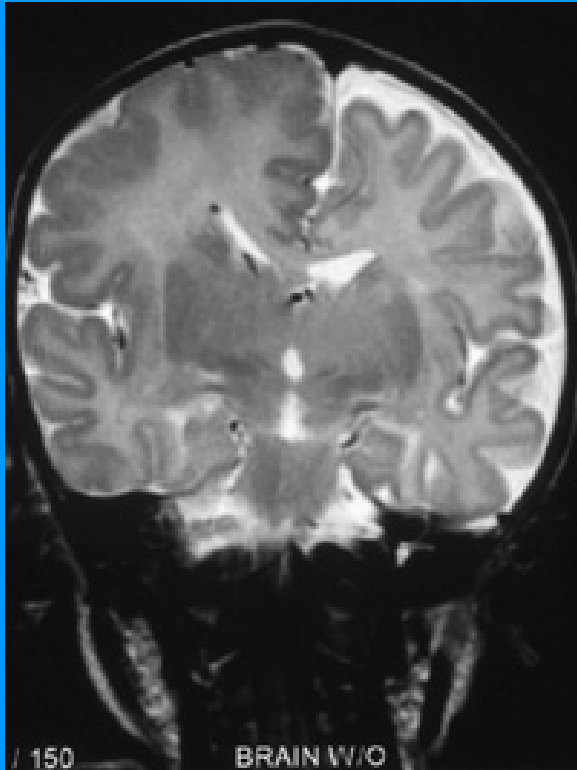


## Seizure Onset Early in Life Risk Factor for Lower IQ





# Hemimegalencephaly

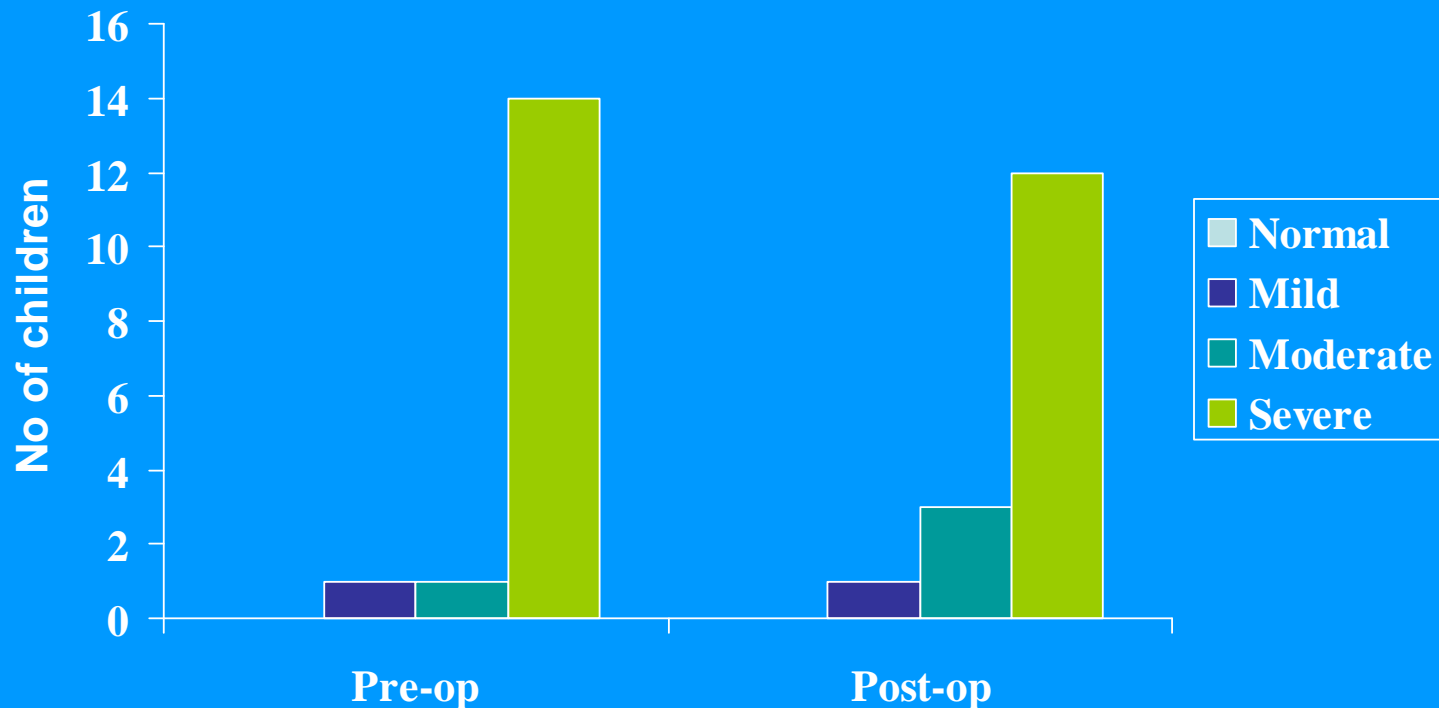


# Developmental outcome following surgery for hemidysplasia

16/33 children undergoing hemispherectomy; age 0.33-16 (2.8yrs)

31% Seizure free; 56% >75% reduction.

Developmental follow-up 1-8yr (3.2yr)



# DN

1. Linear sebaceous naevus syndrome
2. R. Hemimegalencephaly
3. Seizures 0.8y apnoea

motor and status

High rate until phenobarbitone and phenytoin 0.11y

Very severe R EEG abnormality

4. No cognitive progress to 2.6y

Seizures returned

5. Hemispherectomy 3.0y

with loss of seizures and developmental progress

# Sturge-Weber Syndrome

- Leptomeningeal angioma  
(lack of regression of primitive vascular plexus)
- Sz in 80% ? Bias
- 86% in 1<sup>st</sup> 2 years
- 50% CSE + motor and cognitive regression
- 60% intractible
- Surgery, but when

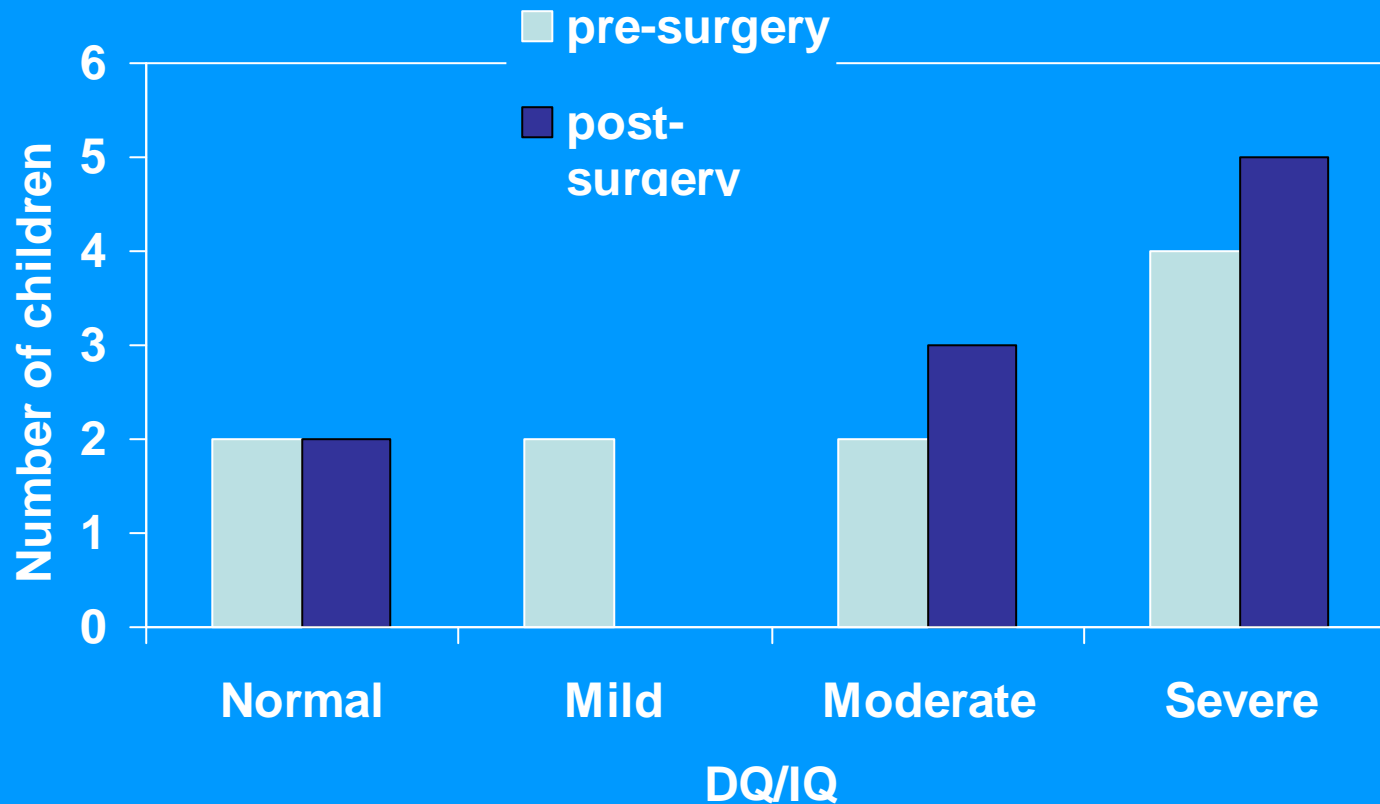
# Sturge Weber syndrome

- Retrospective series 47%, IQ<60 (*Castroviejo et al Ped Neurol 1993;9:283-8*)
- Normal range IQ 25-30%
- Cognitive deterioration with seizures
- Early surgery improved cognitive outcome (*Oluwole et al Can J Neurol Sci 1989;16:78-80, Arzimanoglou et al Neurology 2000 55; 1472-1479*)

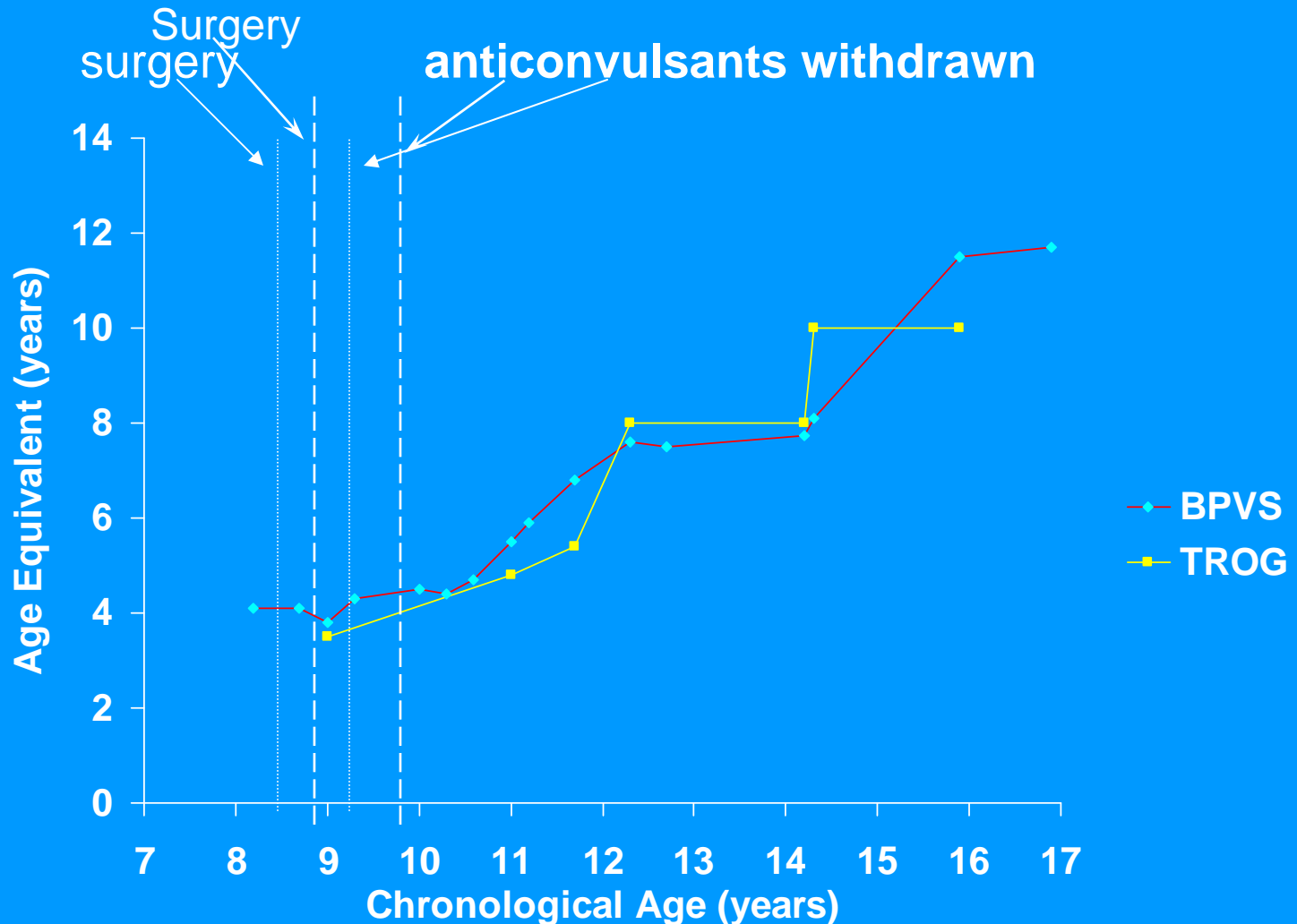


# Developmental Outcome in SWS

10 children; 6 hemispherectomy, 4 focal resection  
Age 31m (10m-17yr)



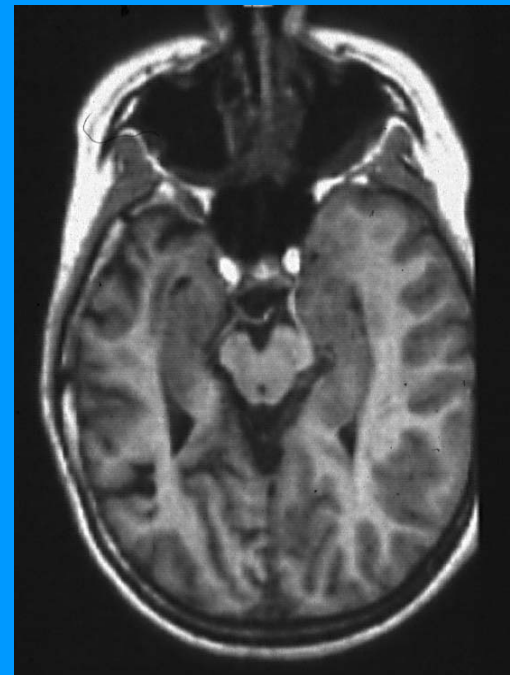
# Receptive language development following hemispherectomy at 9 years



# Rasmussen Syndrome

- Presentation 5-10 yrs
- 60% EPC
- Progressive hemiparesis & cognitive deterioration
- ? Autoimmune aetiology

Surgery curative - when to proceed?

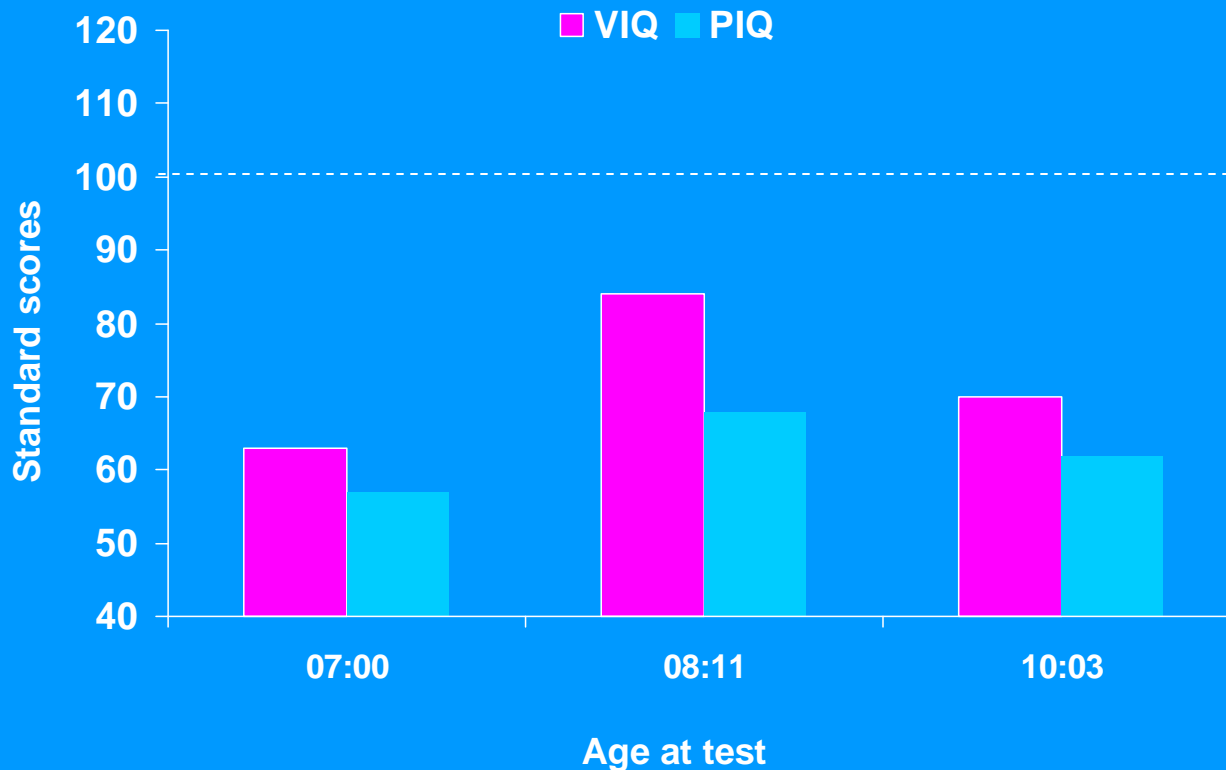




**Age 3.5 Right focal motor seizures EPC; increasing hemiplegia, slurred speech. Became L handed.**

**Age 3.10-4.0 Continued seizures, mute during seizures**

**Age 4.2 Short 2.3 word sentences ictally. L hemispherectomy**



# Oscar Wilde in Lord Arthur Savile's Crime (a crime chosen for him out of context)

He had suddenly been called upon to bear an intolerable burden

Actors are so fortunate

they can chose whether they appear in tragedy or in  
comedy

Whether they will suffer or make merry

laugh or shed tears

But in real life it is different

Most men and women are asked to perform parts for which

they have no qualifications

Our Guildensterns play Hamlet and our Hamlets

have to jest like Prince Hal

The world is a stage

But the play is badly cast.