

Introduction to Epilepsy Surgery

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Overview

- Drug resistant epilepsy
- Cortical zones
- Presurgical evaluation
- Operative procedures

Drug resistant (intractable) epilepsy

- Failure of adequate trials of two tolerated, appropriately chosen and used antiepileptic drug schedules (whether as monotherapy or in combination) to achieve sustained seizure freedom
- 40% of patients with epilepsy

| Cortical zone | Definition |
|-------------------------|--|
| Symptomatogenic zone | The cortical region which generates the patient's habitual ictal symptoms when activated by the epileptiform discharges |
| Irritative zone | The cortical region which is capable of generating interictal epileptiform discharges on EEG |
| Ictal onset zone | The cortical region from which ictal epileptiform discharges originating |
| Epileptogenic lesion | A structural neuroimaging abnormality responsible for the generation of seizures |
| Functional deficit zone | The cortical region that displays aberrant function in the interictal period |
| Epileptogenic zone | The theoretical zone of cerebral cortex that is able to generate seizures. Complete removal or disconnection of this zone renders the patient seizure free |
| Eloquent cortex | The area of cerebral cortex that, if removed, will bring about a neurologic deficit |

Cortical zones

| Zone | Period | Investigation |
|-------------------------|------------|---|
| Symptomatogenic zone | Ictal | Semiology, video-EEG |
| Irritative zone | Interictal | Interictal EEG (scalp, invasive), ECoG, MEG, fMRI-EEG |
| Ictal onset zone | Ictal | Ictal EEG (scalp, invasive), ictal SPECT |
| Functional deficit zone | Interictal | PE, neuropsychological test, Wada test, PET, interictal SPECT |
| Epileptogenic lesion | - | Neuroimaging |
| Epileptogenic zone | - | None |

Operative procedures

Resective surgery

- **Temporal lobe surgery:** anterior temporal lobectomy, selective amygdalohippocampectomy, lesionectomy, tailored resection
- **Extratemporal lobe surgery:** topectomy, lobectomy, multilobar resection, lesionectomy, tailored resection
- **Hemispheric surgery:** hemispherectomy, hemispherotomy

Palliative surgery

- **Disconnection:** corpus callosotomy, multiple subpial transections, hippocampal transections
- **Neurostimulation:** vagus nerve stimulation, deep brain stimulation, responsive cortical stimulation

Standard anterior temporal lobectomy

Anterior 2/3 temporal lobectomy

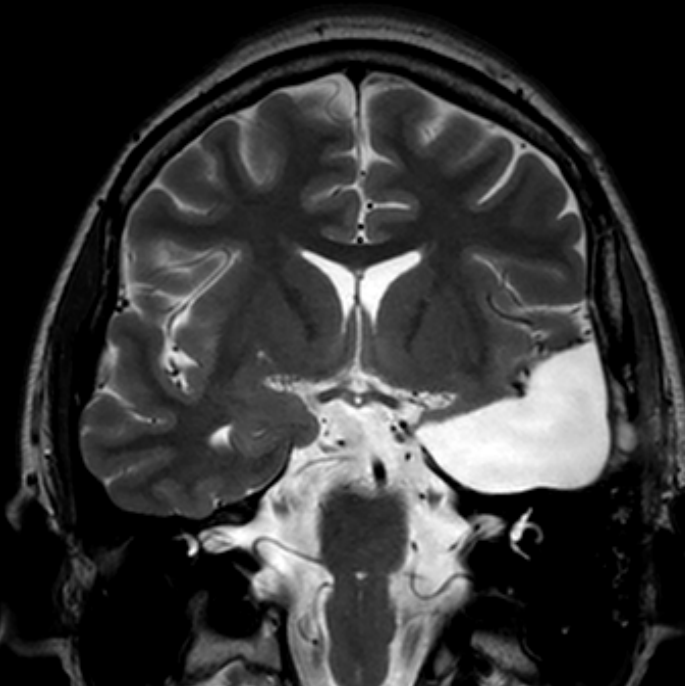
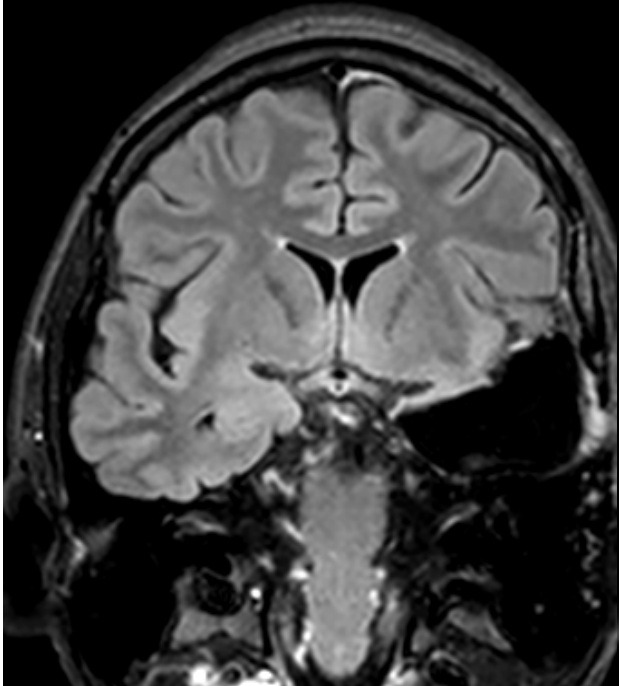
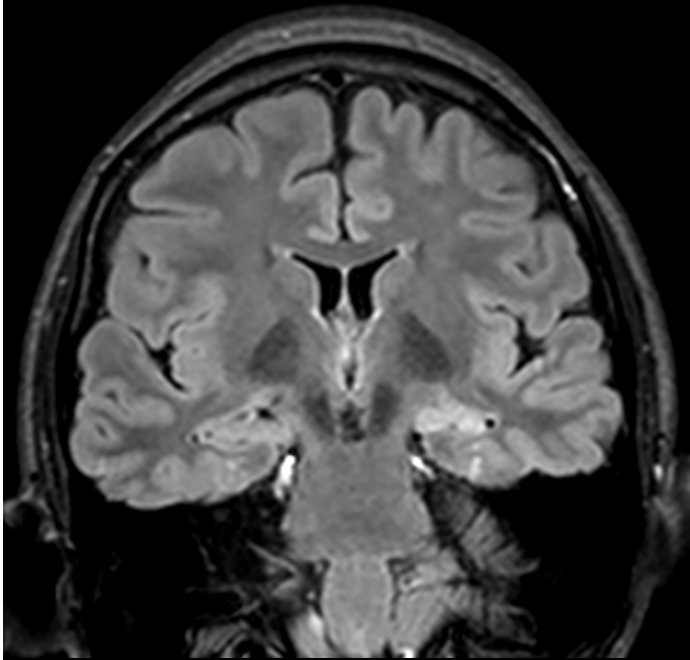
Neocortical resection + amygdalohippocampectomy

The median length of resection from temporal tip

- Non-dominant: 5.5 cm
- Dominant: 4.5 cm
- Hippocampus: 3 cm

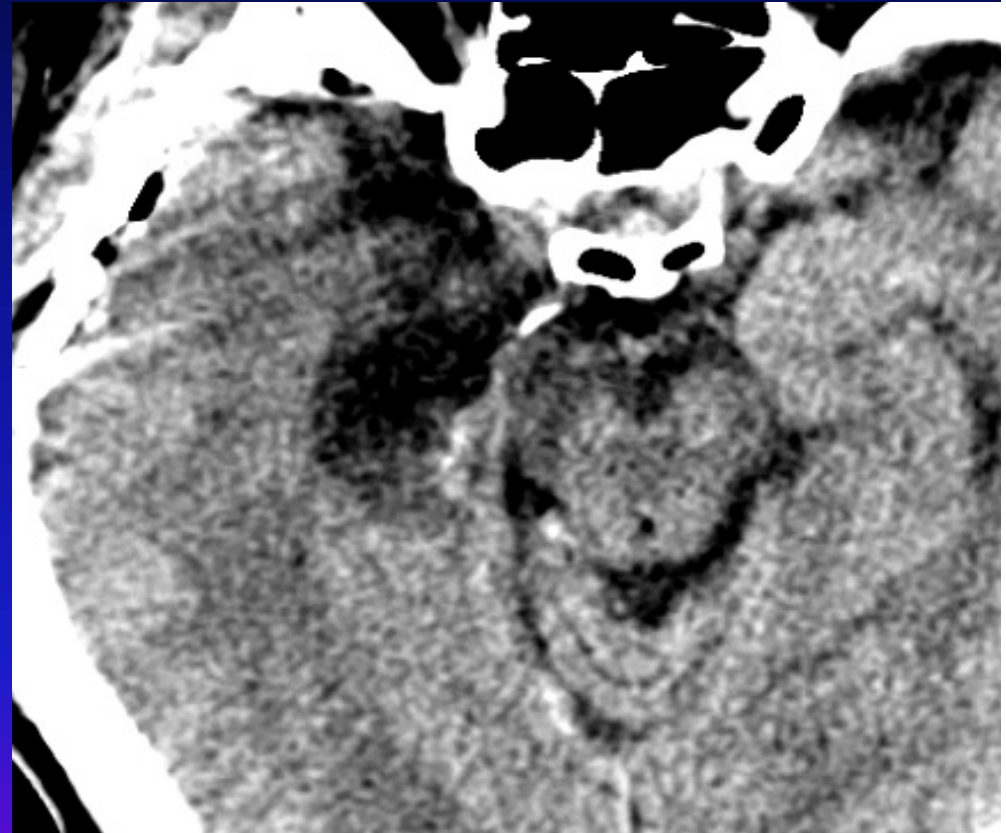
Hippocampal sclerosis

Anterior temporal lobectomy



Selective amygdalohippocampectomy

- Resection of mesial temporal structures without neocortical resection
- Indication: clear evidence of MTLE without lateral temporal involvement



Dual pathology

- Hippocampal sclerosis + extra-hippocampal lesion
- Extra-hippocampal lesion: temporal or extratemporal
- Which one is (are) epileptogenic lesion?
- Removal of both lesions: the greatest chance of seizure freedom
- Seizure free is less than single pathology

Tailored resection

- ✓ Intraoperative electrocorticography (ECoG) recording on the cerebral cortex
- ✓ Identification of epileptogenic area
- ✓ Useful in cortical lesion e.g. cortical dysplasia
- ✓ Limitations
 - Inter-ictal EEG (irritative zone): cannot demarcate the epileptogenic zone definitely
 - Need a period of time during the operation

Surgery for hemispheric epilepsy

Hemispheric epilepsy

- Hemimegalencephaly
- Rasmussen's encephalitis
- Struge-Weber syndrome
- Ischemic/anoxic brain injury

Goal: early control of catastrophic epilepsy

Hemispherectomy techniques

Resection

Anatomical
hemispherectomy

Modified anatomical
hemispherectomy
(Oxford modification)

Hemicortectomy
(hemidecortication)

Disconnection

Functional
hemispherectomy

Modified
hemispheric
disconnection

Hemispheric
deafferentiation

Keyhole
hemispherectomy

Transopercular
hemispherotomy

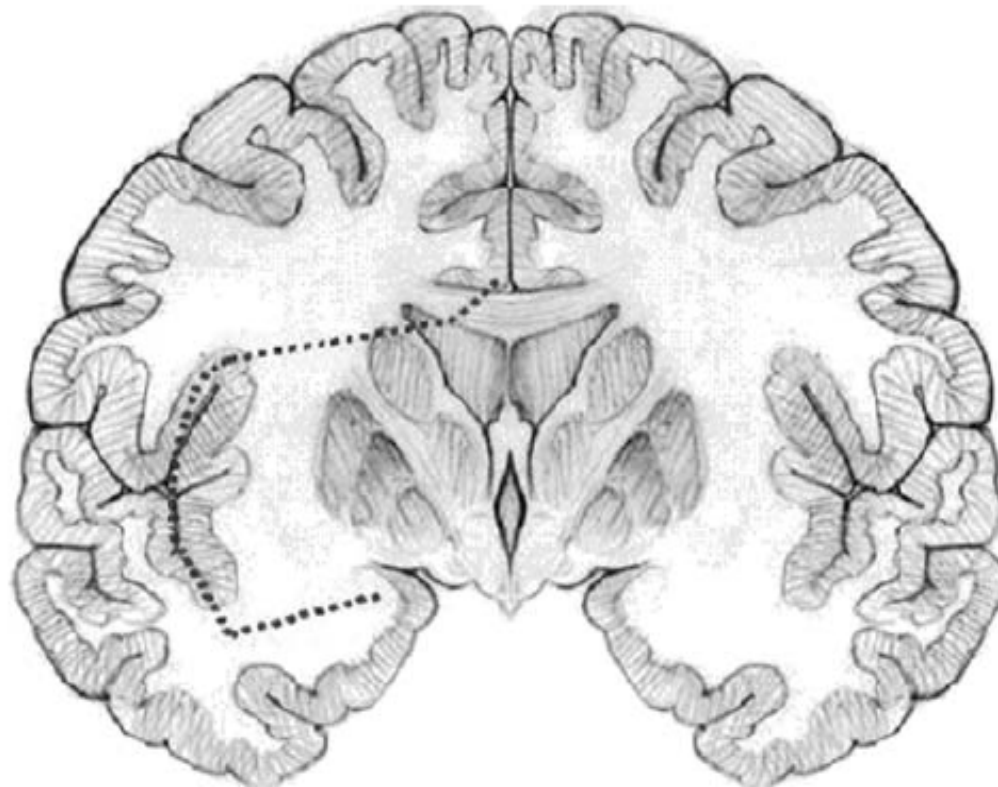
Hemispherotomy

Peri-insular (lateral)
hemispherotomy

Vertical
hemispherotomy

Anatomical hemispherectomy

- McKenzie 1938
- Indications: hemimegalencephaly, reoperation after failed functional hemispherectomy
- En bloc resection vs. piecemeal resection



Anatomical hemispherectomy

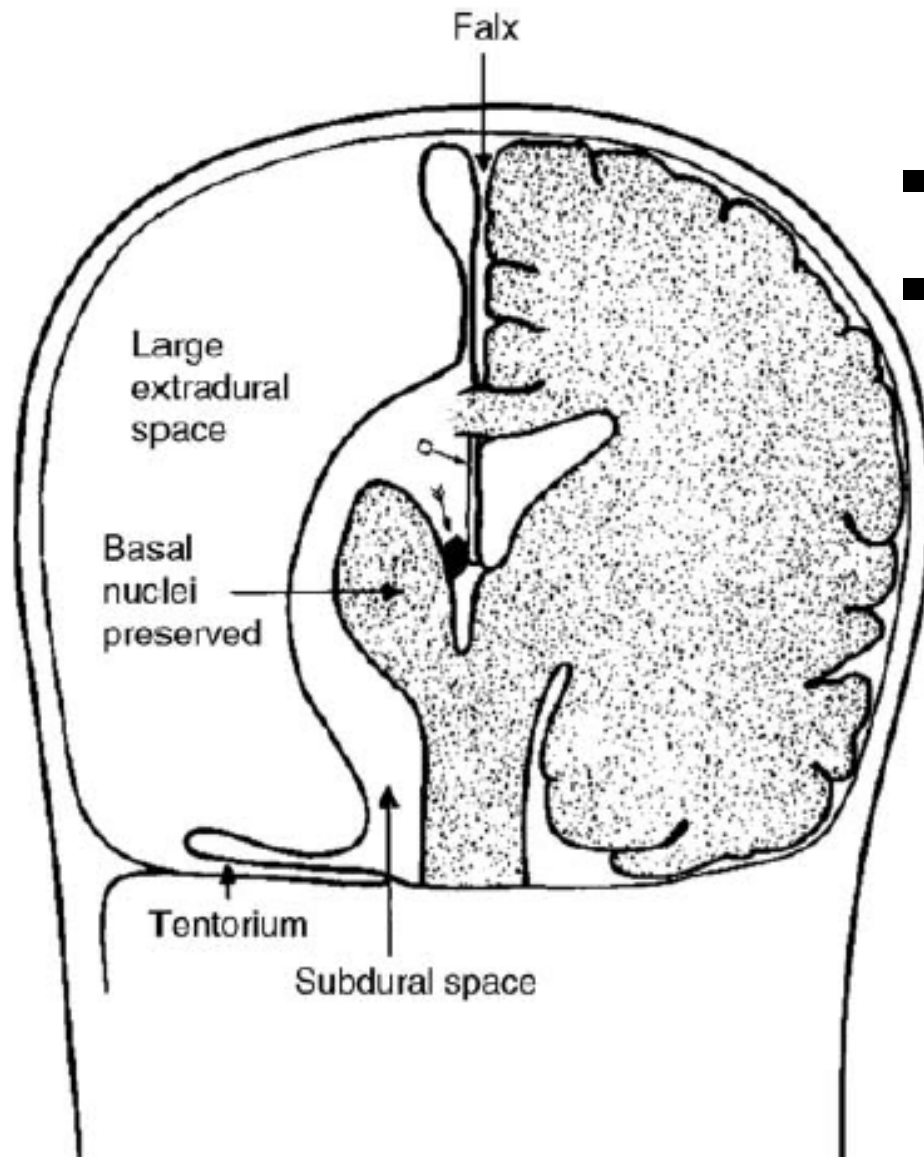
Early postoperative complications

- Aseptic meningitis
- Brain shift and herniation
- Hydrocephalus

Late postoperative complications

- Superficial cerebral hemosiderosis (SCH)
- Spontaneous or post-traumatic hemorrhage

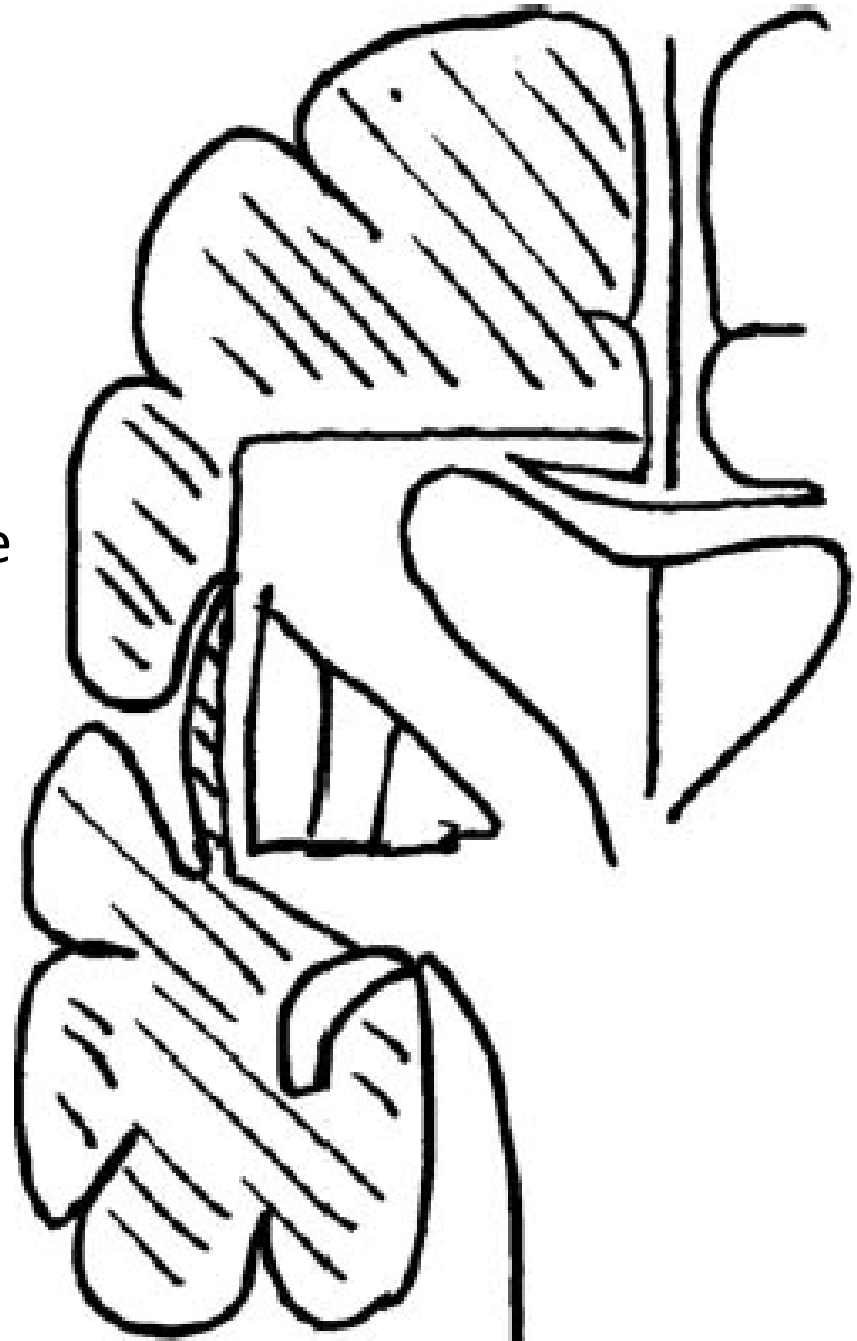
Modified anatomical hemispherectomy (Oxford modification)



- Adams 1968
- Procedures
 1. Anatomical hemispherectomy
 2. Muscle plug at foramen of Monroe
 3. Dural plication for reducing subdural space
 4. Expansion extradural cavity

Hemicortectomy (hemidecortication)

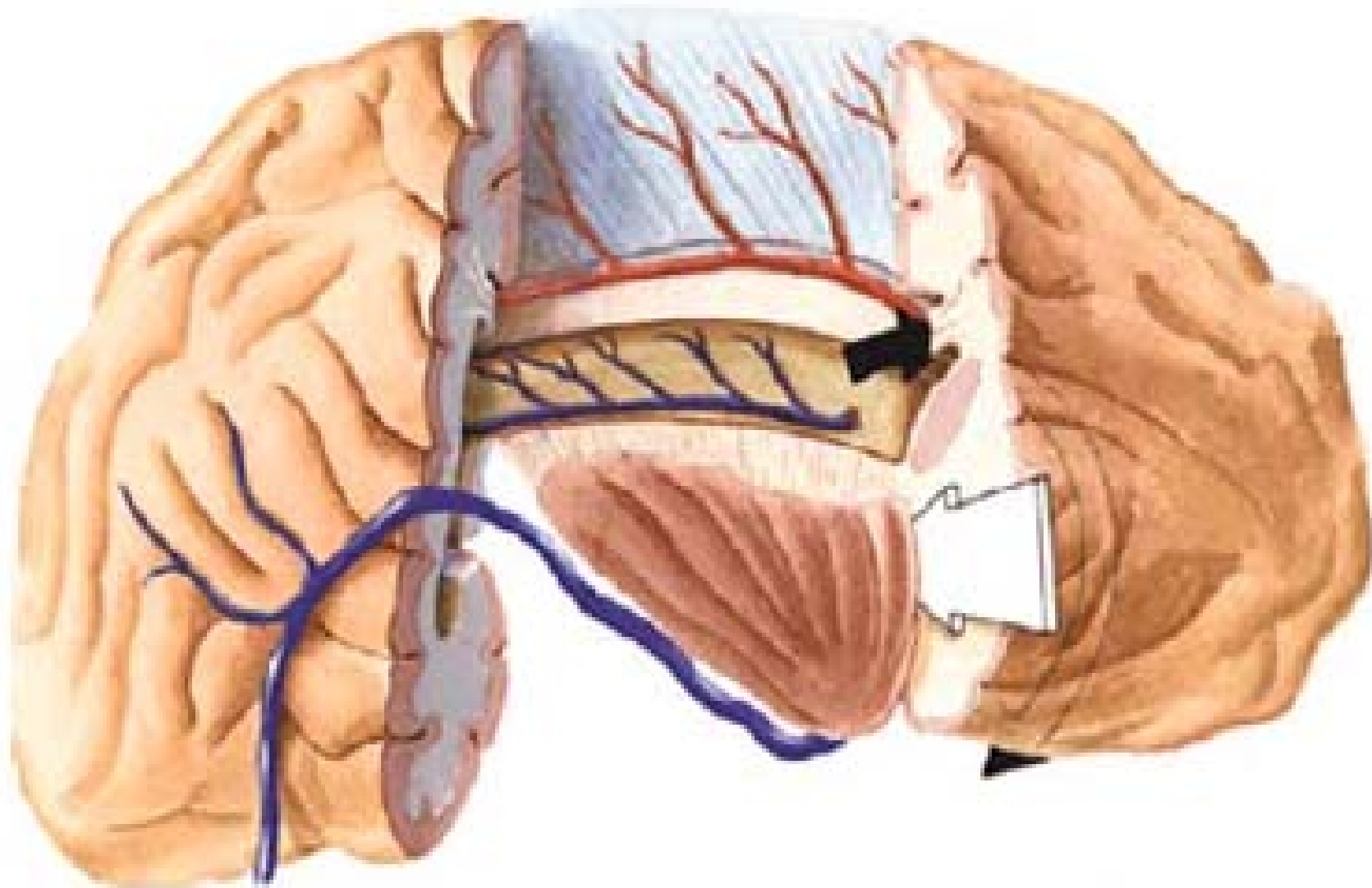
- ✓ Ignelzi and Bucy 1968
- ✓ Degloving of the cortex from the white matter
- ✓ Minimize ventricular exposure
- ✓ Disadvantages
 - Blood loss
 - Obliteration of arachnoid granulation: hydrocephalus
 - Incomplete disconnection



Functional hemispherectomy

- Rasmussen 1974
- Modern technique for hemispheric epilepsy
- Avoidance of SCH
- Procedures
 1. Temporal lobectomy (neocortex + hippocampus + amygdala)
 2. Central (frontoparietal operculum) resection
 3. Transventricular total corpus callosotomy
 4. Disconnection of mesial frontal and parieto-occipital lobes
 5. Insular cortex aspiration

Functional hemispherectomy



Peri-insular hemispherotomy

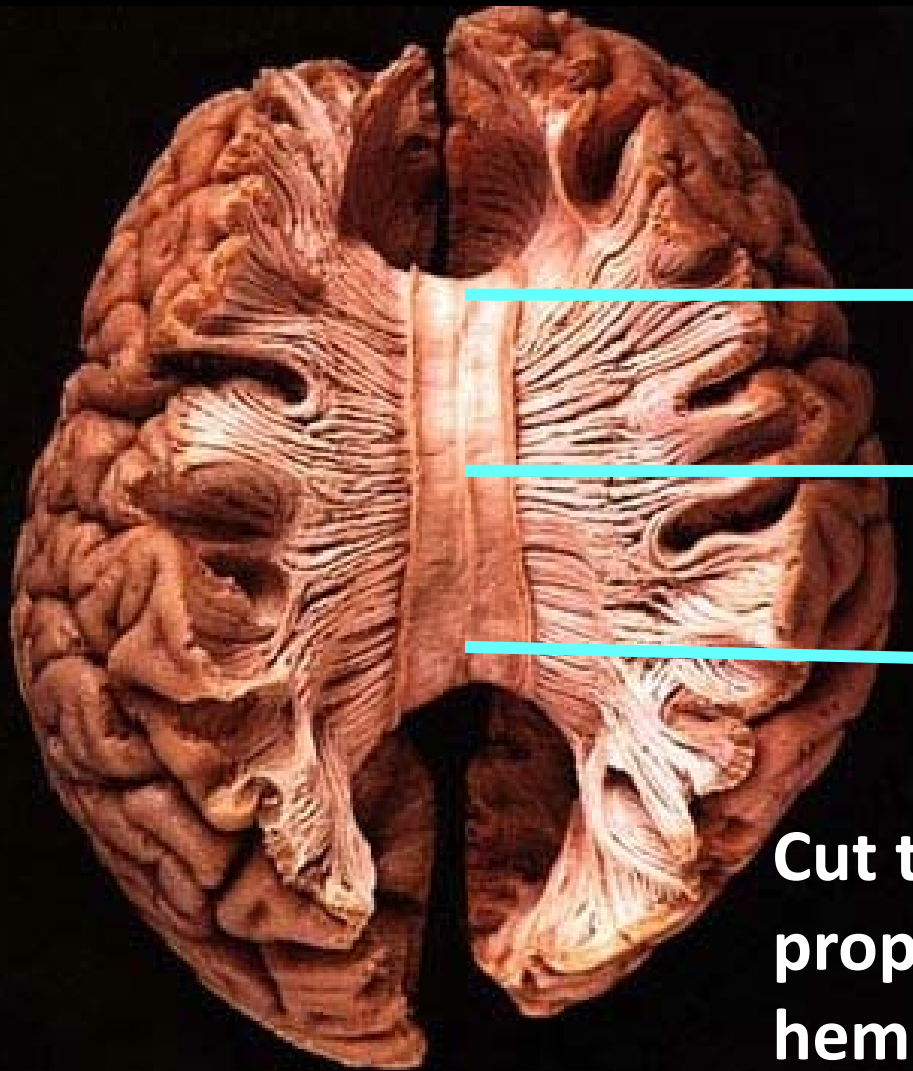
- Villemure 1995
- Minimal brain removal to achieve complete hemispheric disconnection
- Procedures
 1. **Suprasylvian window:** removal of frontoparietal operculum, transventricular total corpus callosotomy, disconnection of frontal lobe

Peri-insular hemispherotomy

- Procedures

2. **Infrasylvian window:** removal of temporal operculum, total resection of temporal stem from trigone to anterior temporal horn, amygdalectomy, hippocampectomy (or posterior hippocampotomy)
3. **Insular resection:** two techniques
 - 3.1 Insular cortex aspiration
 - 3.2 Insular disconnection

Corpus callosotomy (interhemispheric disconnection)



Rostrum and genu

Body

Splenium

Cut the pathway of seizure
propagation from one to the other
hemisphere

Corpus callosotomy

Intractable generalized seizures

- Atonic seizure (drop attack)
- Lennox-Gastaut syndrome
- West syndrome

Anterior vs. total corpus callosotomy

Anterior vs. total corpus callosotomy

Anterior corpus callosotomy

- Spare of splenium
- Diminish risk of complications, esp. callosal disconnection syndrome

Total corpus callosotomy

- Second stage procedure following failed anterior corpus callosotomy
- Initial procedure for patients with severe cognitive impairment or significant unilateral hemispheric disease (low risk of callosal disconnection syndrome)

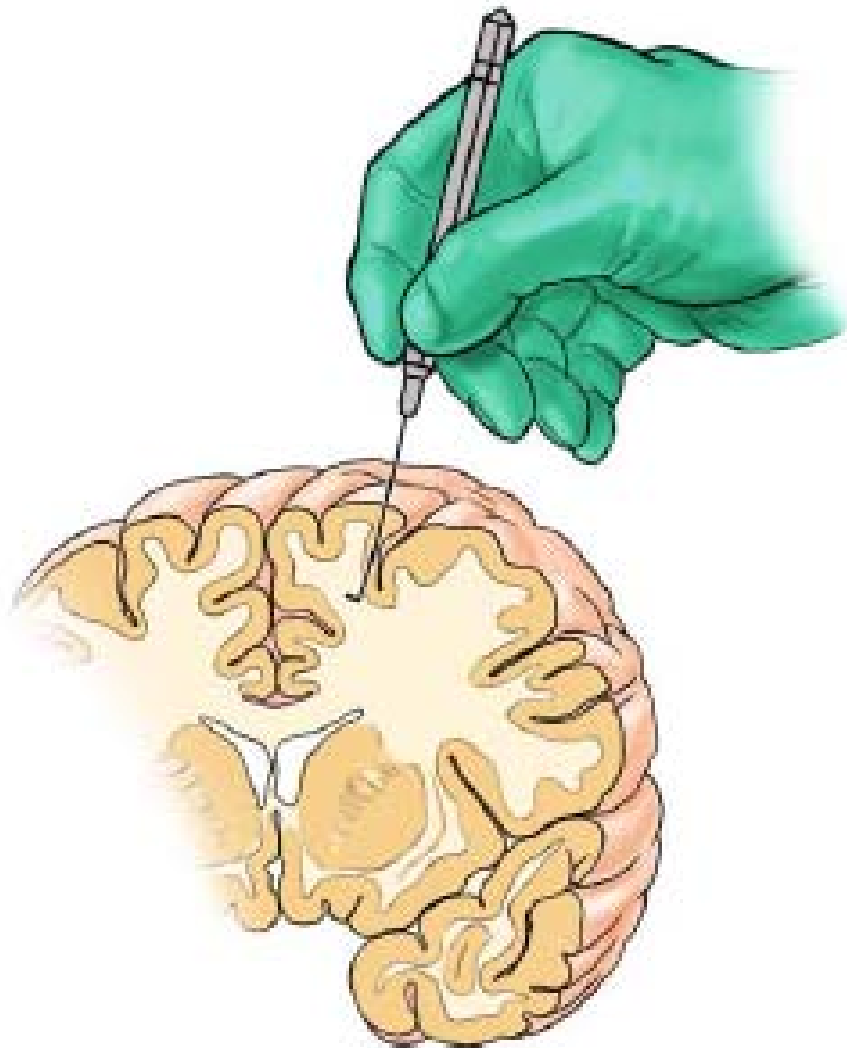
Complications

- Retraction brain edema
- Callosal disconnection syndrome
- Language impairment and transient aphasia
- Transient akinetic state
- Memory deficits
- New types of seizure: esp. simple partial seizure
- Aseptic meningitis and ventriculitis after exposure of 3rd ventricle (prevent by extraependymal disconnection and short-term steroid)

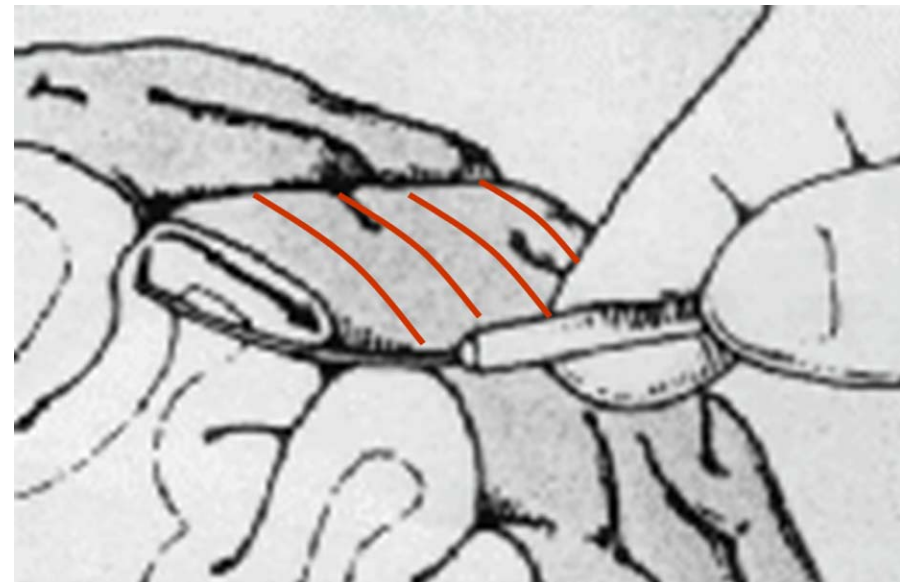
Callosal disconnection syndrome (split-brain syndrome)

- Alien hand syndrome and intermanual conflict
- Left hand apraxia
- Left hand agraphia
- Left tactile anomia
- Right constructional apraxia
- Visual anomia
- Hemineglect

Multiple subpial transections



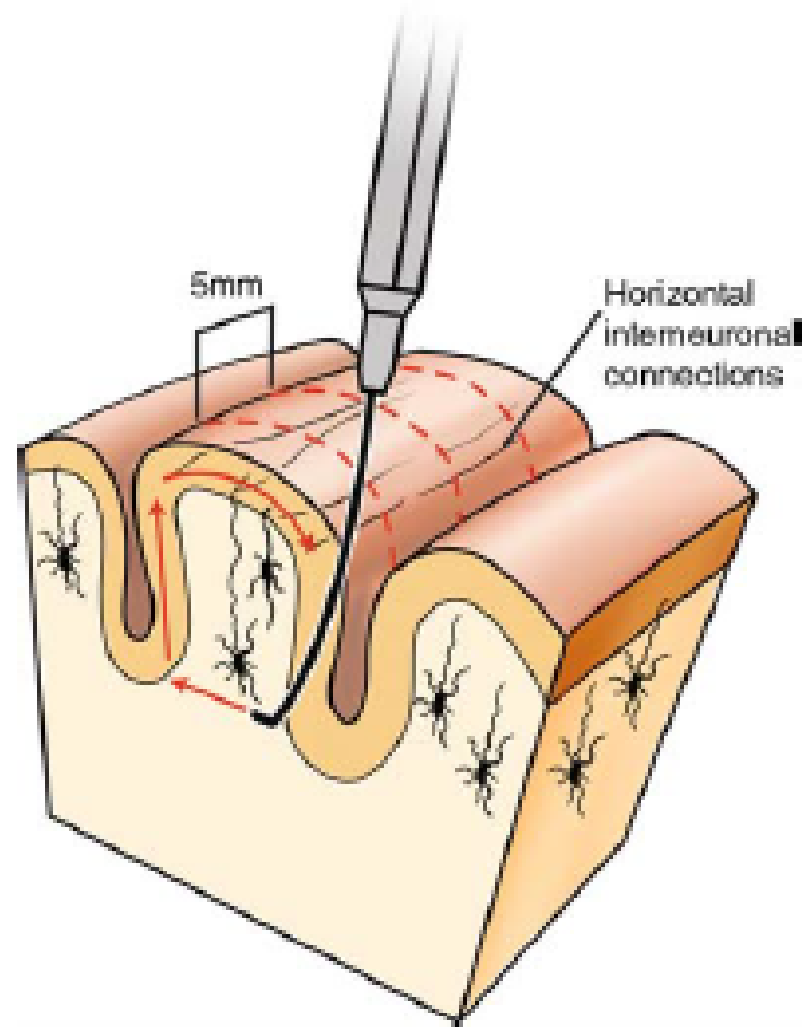
- Disconnection of epileptogenic focus in the eloquent cortex to avoid permanent neurological deficit



Multiple subpial transections

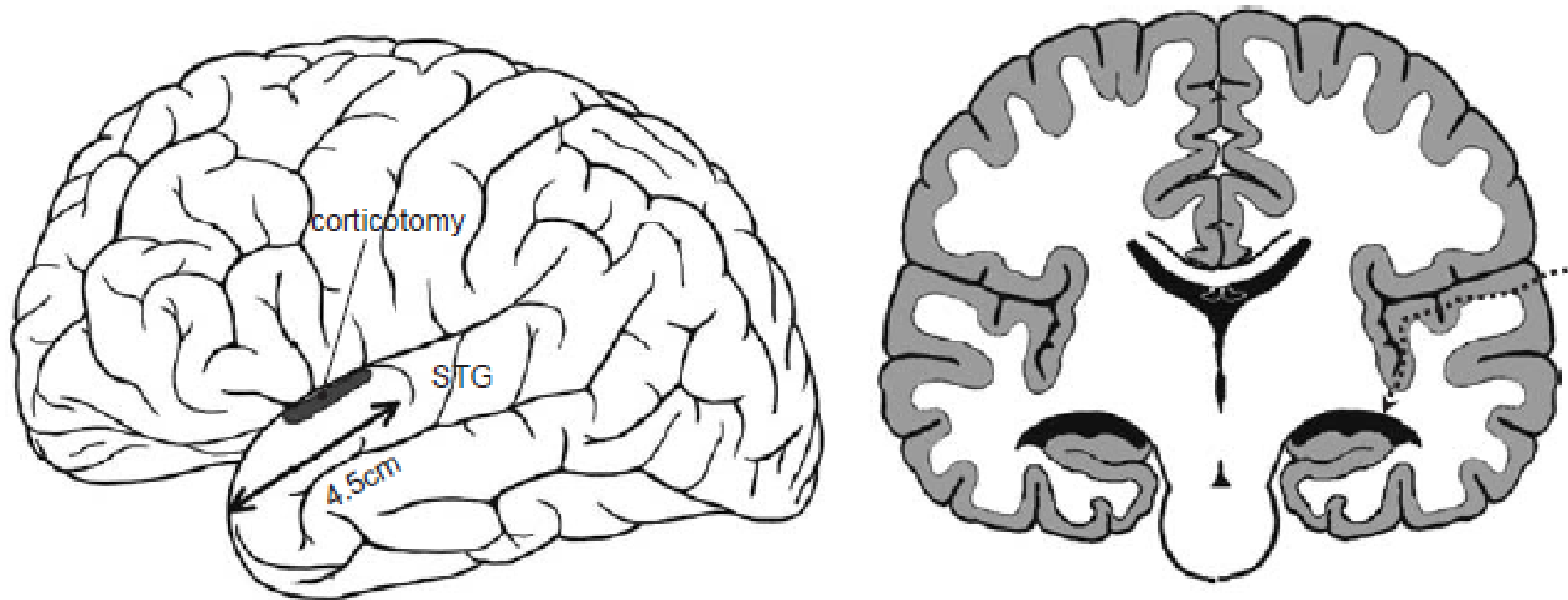
Indications

- Focal seizure arising in eloquent cortex (motor, sensory, language, memory)
- Landau-Kleffner syndrome (LKS)
- Malignant rolandic-sylvian epilepsy syndrome (MRSE)
- Others: cortical dysplasia, epilepsy partialis continua (EPC) caused by Rasmussen's encephalitis



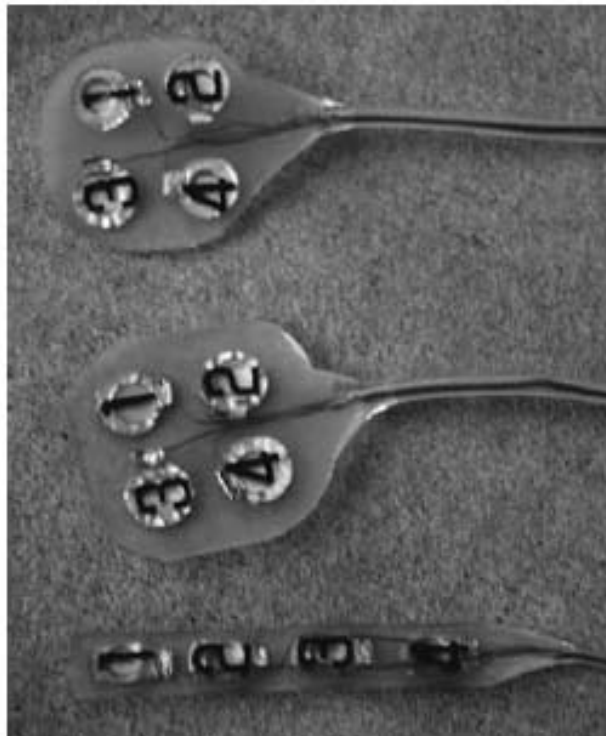
Multiple hippocampal transections

- Seizure control with preservation of verbal memory
- Surgical candidate: intractable MTLE without hippocampal atrophy or sclerosis on MRI



Multiple hippocampal transections

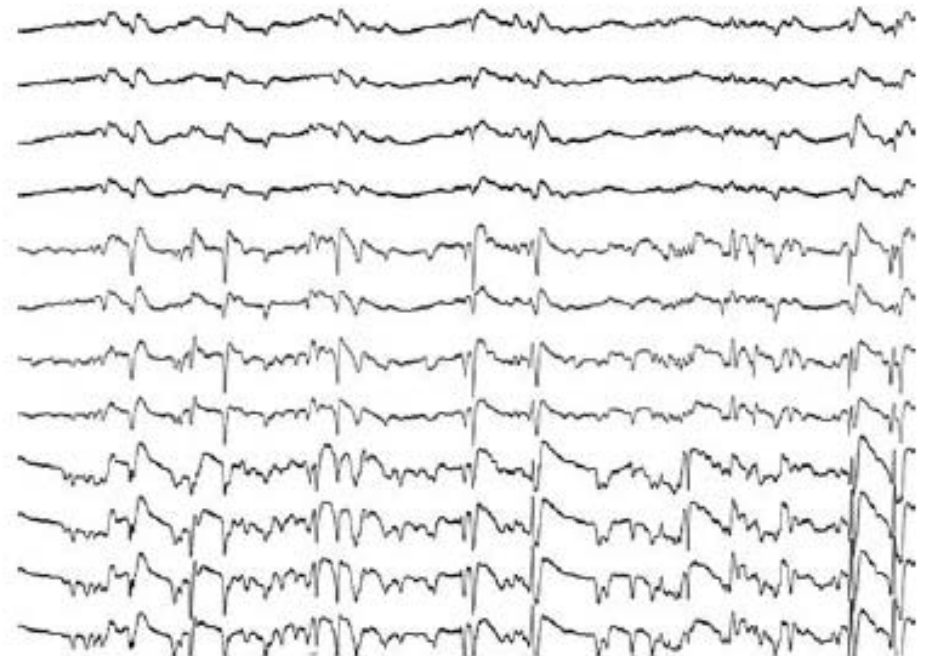
- Intraoperative ECoG before and after transections
- If no spike on amygdala: partial resection of amygdala to expose the hippocampal head
- If there is active spikes on amygdala: amygdalectomy



Amygdala

Hippocampal Head

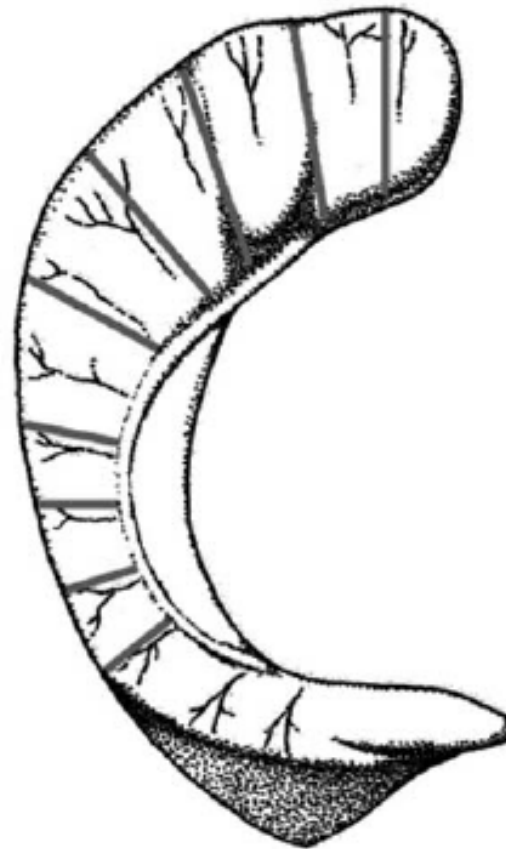
Hippocampal body



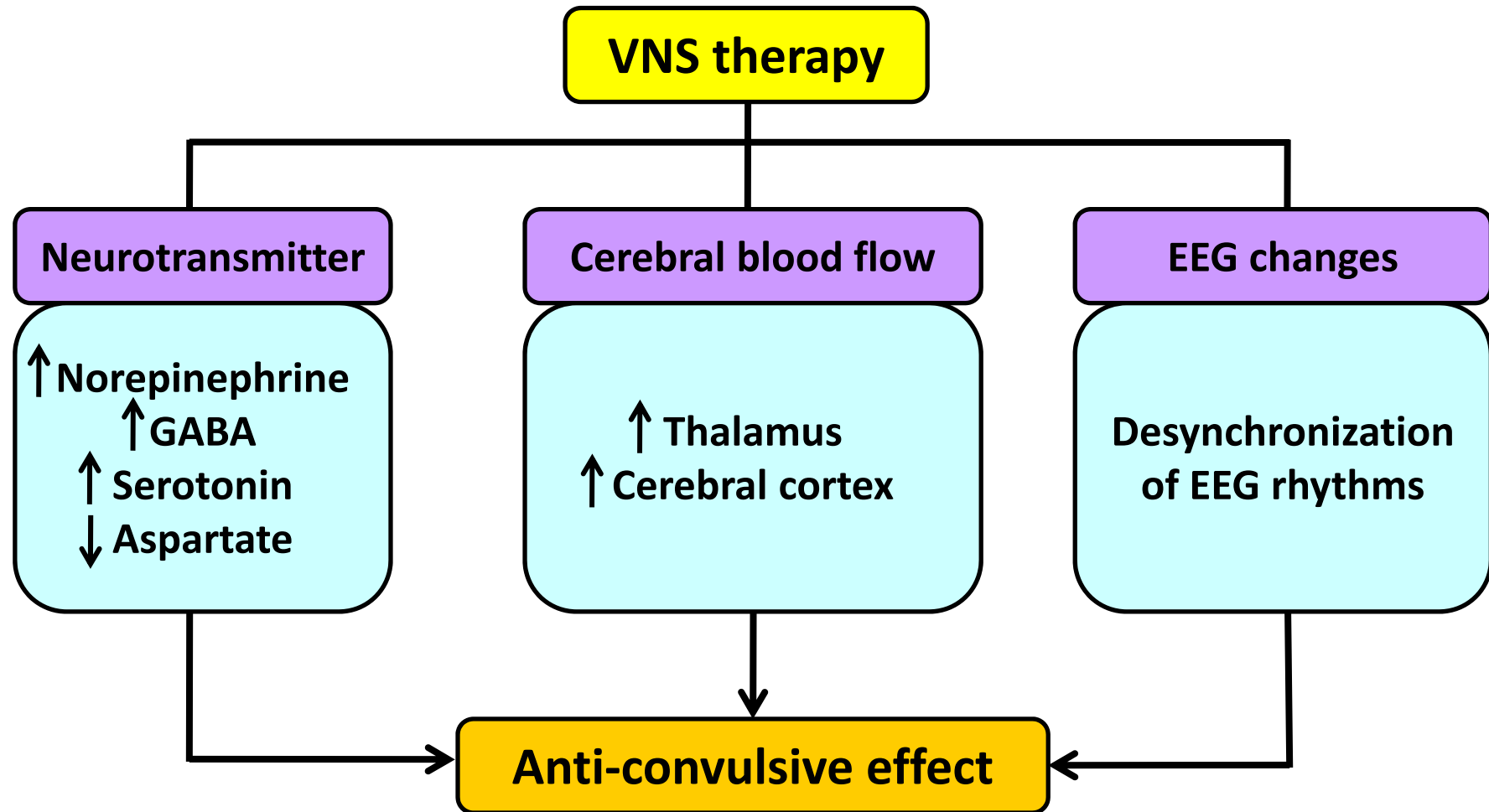
Multiple hippocampal transections



5 mm distance to the next transection



Vagus nerve stimulation



Vagus nerve stimulation

- Unknown mechanism of action
- Vagal afferents project to diffuse areas of CNS, many of them are potential sites of epileptogenesis

- Key structures of VNS action

Locus ceruleus (LC)

Dorsal raphe nucleus (DRN)

**Release norepinephrine
and serotonin**

Anti-convulsive effect

Anti-depressive effect

Vagus nerve stimulation

Indications

- Patients who are not candidates for resective surgical procedure

Not possible to predict response to VNS therapy

VNS can decrease seizure duration, frequency or severity, may reduce amount of AEDs

Vagus nerve stimulation

- The right vagus nerve preferentially supplies the sino-atrial (SA) node
- VNS electrode is usually applied to the left side to avoid stimulation-related asystole or bradycardia
- Implantation of VNS electrode at the midcervical level because the vagus nerve is relatively free of branches

VNS complications

- Vocal cord abnormalities

 - Transient vocal cord paralysis 0.7 %

 - Spontaneous remission

- Infection

- Bradycardia/asystole (very rare)

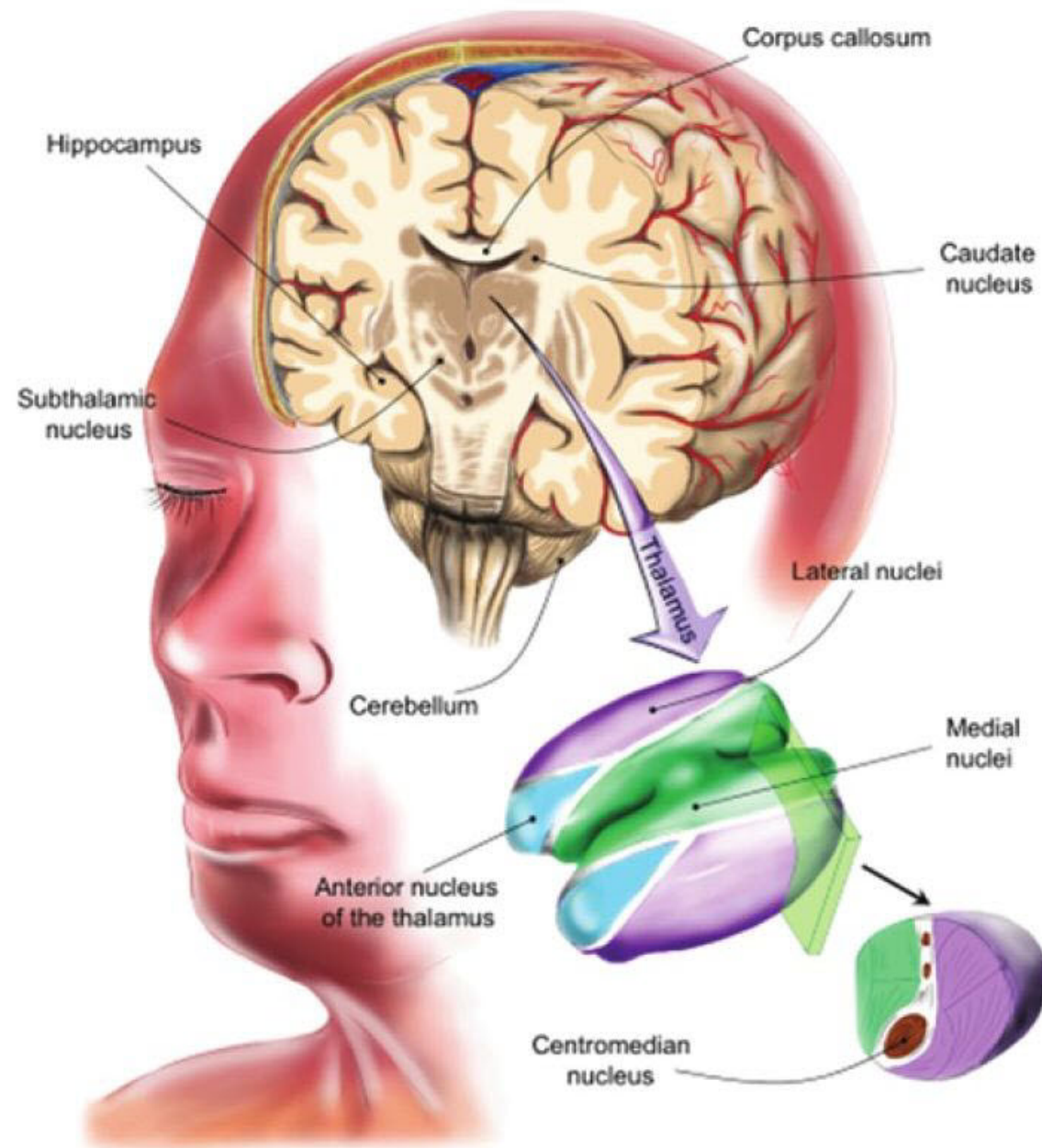
- Sleep-related breathing disorders

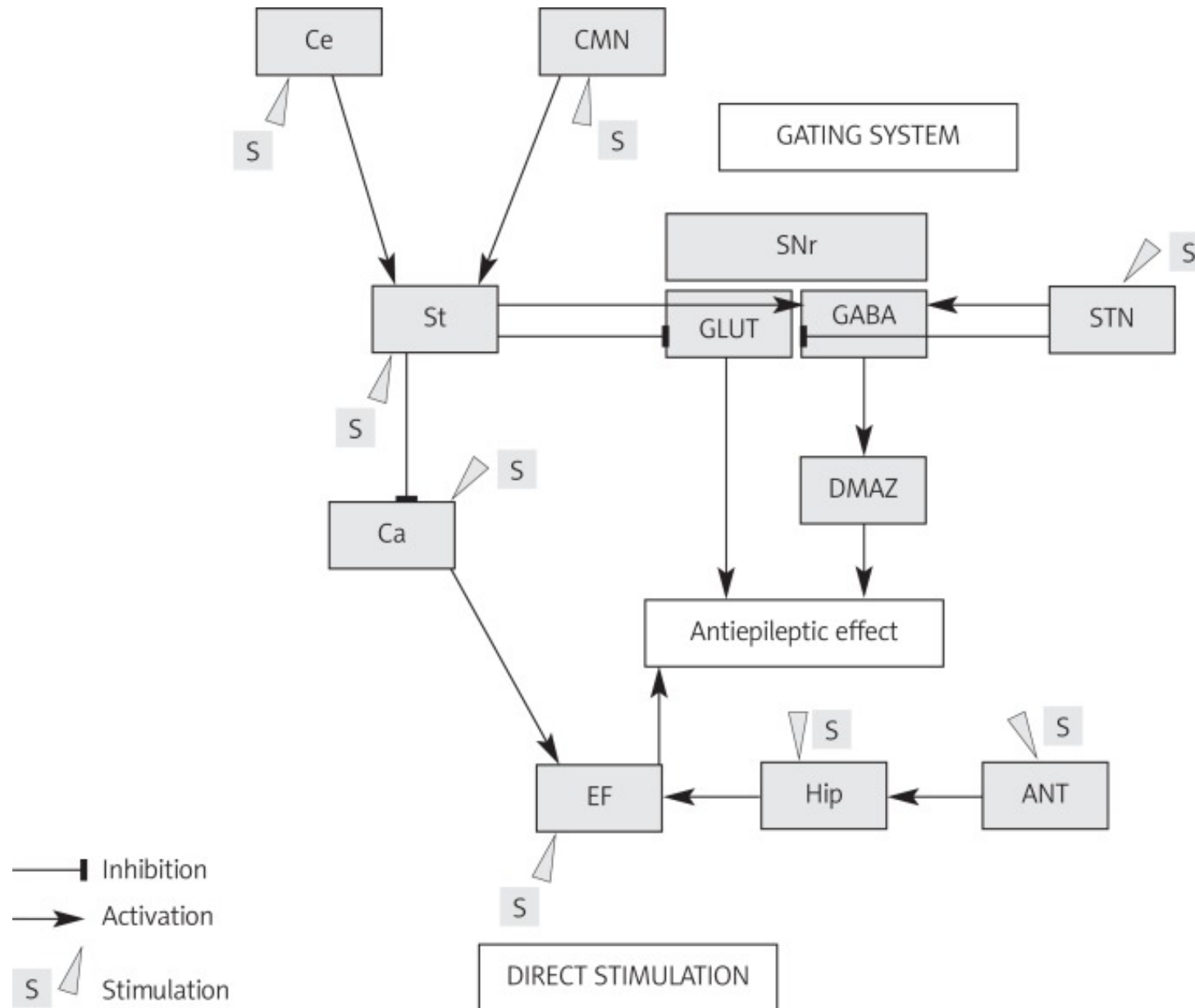
 - Decreased respiratory airflow, OSA

 - Resolve with decrease VNS setting

 - Patients with known OSA should be monitored carefully after VNS implantation

Anterior thalamic deep brain stimulation





Ce – cerebellum, CMN – central-medial nucleus of the thalamus, St – striatum, SNr – pars reticularis of substantia nigra, STN – subthalamic nucleus, Ca – corpus amygdaloideum, DMAZ – dorsal midbrain anticonvulsant zone, EF – epileptic focus, Hip – hippocampus, ANT – anterior nucleus of thalamus

Responsive cortical stimulation

- Treatment of intractable partial-onset epilepsy
- Subdural cortical strip or depth electrode is placed in the brain according to the seizure focus
- Responsive stimulation directly to the seizure focus when epileptic activity is detected
- Aim to disrupt epileptiform activity before seizure developed

Intracranial EEG

Indications

- Disconcordant results of non-invasive methods
- Absent structural lesion on MRI
- Scalp EEG cannot detect seizure origin
- Localization of eloquent cortex by functional brain mapping

Types (subdural, depth, stereo EEG)

Postoperative seizure outcome

Engel vs. ILAE classification

| Engel classification | ILAE classification |
|--|--|
| <p>Class I. Free from disabling seizures</p> <p>A. Completely seizure free since surgery B. Nondisabling simple partial seizures only since surgery C. Some disabling seizures after surgery, but free from disabling seizures for ≥ 2 years D. Generalized convulsions w/AED discontinuation only</p> <p>Class II. Rare disabling seizures (almost seizure free) A. Initially free from disabling seizures, but still has rare seizures B. Rare disabling seizures since surgery C. Occasional disabling seizures since surgery, but rare seizures for the last 2 years D. Nocturnal seizures only</p> <p>Class III. Worthwhile improvement A. Worthwhile seizure reduction B. Prolonged seizure-free intervals amounting to $>50\%$ of follow-up period, but not <2 years</p> <p>Class IV. No worthwhile improvement A. Significant seizure reduction B. No appreciable change C. Seizures worse</p> | <p>Class 1. Completely seizure free; no auras Class 1a. Completely seizure free since surgery; no auras Class 2. Only auras; no other seizures</p> <p>Class 3. 1–3 seizure days/yr; \pmauras</p> <p>Class 4. 4 seizure days/yr–50% reduction in baseline no. of seizure days; \pmauras</p> <p>Class 5. $<50\%$ reduction in baseline no. of seizure days – 100% increase in baseline no. of seizure days; \pmauras</p> <p>Class 6. $>100\%$ increase in baseline no. of seizure days; \pmauras</p> |

Timing of AED withdrawal after successful epilepsy surgery

- Controversial
- Most studies: tapering off AED after one year of postoperative seizure freedom is safe
- Early AED withdrawal may increase risk of seizure recurrence
- An important predictor of seizure outcome and cure is incomplete resection of epileptogenic lesion
- Early AED withdrawal seems safe in patients with presumed complete resection of epileptogenic lesion

Intractable epilepsy

Candidate for epilepsy surgery ?

Neuroimaging (high resolution MRI) + video-EEG

Lesional epilepsy
(MRI positive)

Non-lesional epilepsy
(MRI negative)

Concordant with
video-EEG

Discordant with
video-EEG

Further studies

SPECT (ictal/ interictal)

PET, functional MRI

Neuropsychological test

Wada test

Intracranial EEG monitoring

Functional brain mapping

Resective
surgery

Yes

Candidate for
resective surgery

No

Medication, palliative surgery (corpus callosotomy, vagus nerve stimulation)

Complete resection or disconnection of **epileptogenic zone**



Seizure free

Seizure semiology



Structural
neuroimaging
(MRI)



**Epileptogenic
zone**



Functional
neuroimaging
(SPECT, PET,
fMRI, SISCOM)

Video-EEG



Neuropsychological test

Visible MRI lesion \neq **Epileptogenic lesion**