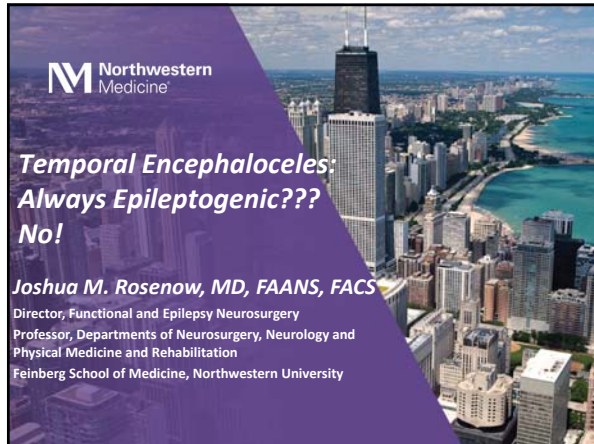


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**Temporal Encephaloceles:
Always Epileptogenic???**
No!

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Evidence Overview

- Most found incidentally or via other symptoms/signs
 - CSF leak, proptosis
- We actually do very little temporal lobe encephalocele surgery
- Peer reviewed literature only has small case series
- Often used as cases on neurosurgery oral board exams

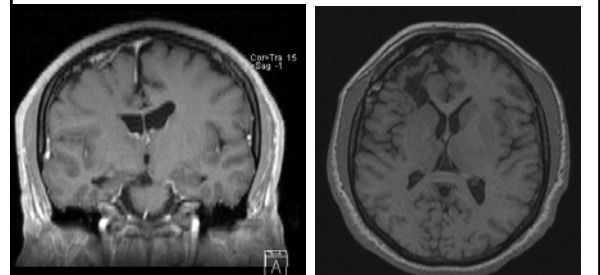
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Disclosures

Corporate Ownership, Equity, Stocks, Bonds	None
Corporate Consultant Contracts – Boston Scientific Neuromodulation, GLG	Yes
Corporate Fiduciary or Board Positions	None
Corporate research - Boston Scientific, SanBio/Sunovion, SPR Therapeutics	Yes
Non-Profit Board Positions –Medical Advisory Board, Epilepsy Foundation of Greater Chicago	Yes
Grants – Co-investigator on grants from NIH, Brain Research Foundation, NMH Dixon Fund, DoD, NIDDR, VA	Yes
Patents	None

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Where is the focus?



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Disclosure (Mea Culpa)

- I perform epilepsy surgery as often as my epileptologists allow me to do so
- I really (really) like epilepsy surgery
- I *especially* like surgery for encephaloceles

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Temporal Encephaloceles

- Spontaneous
 - Congenital
 - Acquired
 - Infection
 - Inflammation
 - Neoplasm
- Traumatic

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Temporal Encephaloceles

• Temporal

- Lateral (defect involves the pterion/asterion/cranial vault)
- Anteroinferior/transalar (defect involves the anteroinferior portion of the middle fossa)
- Posteroinferior (defect involves the tegmen tympani)

• Sphenoidal

- Sphenoorbital/posterior-orbital (defect involves the sphenoid wing)
- Anteromedial (defect involves the anteromedial portion of the middle fossa)



Epileptogenic zone

• Lüders et al. 1993

- The area of cortex that is necessary and sufficient for initiating seizures and whose removal (or disconnection) is necessary for complete abolition of seizures

• Epileptogenic Lesion

- Radiographic lesion that causes seizures



Encephalocele Location and Seizures

• Temporal

- Typically present with CSF leak, hearing loss
- Much less common to present with seizures
 - Stucken, 2012 – 20 consecutive temporal tegmen encephaloceles, no epilepsy
 - Marone 2015 - 5 patients with TLE and encephaloceles, 2/5 defect in tegmentum

• Sphenoidal

- Sphenoorbital/posterior-orbital (defect involves the sphenoid wing)
- Anteromedial (defect involves the anteromedial portion of the middle fossa)



Peer Reviewed Encephaloceles/Epilepsy

• Fewer than 20 reported series

• Largest series – 23 patients

- Saavalainen, *neurology* 2015

• If these lesions are *always* epileptogenic, why are there not more/larger series

• Larger epilepsy centers would collect the cases, publish them

– Cavernomas –

- Ferrer 2008 – locations of 690 cavernomas
- Rate of epilepsy with cavernoma approx. 50%
- Baumann 2007 – 168 patients with epilepsy surgery for cavernomas
- Dammann – 79 consecutive patients with cavernoma-related epilepsy
- Englot 2011 – systematic review 1226 pts with cavernoma/sz, 361 intractable sz



Traumatic Encephaloceles

- More often present with CSF leak than with seizures
- No significant case series of surgery for epileptogenic traumatic encephaloceles



Where are the Encephalocele Epileptics?

• If these were *always* epileptogenic, there would be much more epilepsy surgery occurring for these lesions

• Saavalainen, *neurology* 2015

• Finland MRI review over 7 years (approx. 2900 MRI)

- Encephalocele freq 0.3% - MRI for new dx epilepsy (out of 250pts/yr)
- Encephalocele freq 1.9% - refractory patients referred for surgical eval (out of 110 pts/yr)

• Why is the frequency of refractory patients with encephaloceles so low?

• Contrast –

- MTS –
- FCD –
- MCD –



Acquired Encephaloceles

- If these were *always* epileptogenic, we would see more people with these lesions and epilepsy as time progressed and more people developed them
- Stucken 2012
 - Obesity → increased ICP → encephalocele
 - Should mean we are operating on all these large seizing people with onset later in life
 - *Do any of you have a clinic full of these people?*



Why remove more than encephalocele?

- Encephalocele is not the entire lesion
 - Panov, *epilepsia* 2016
 - ECoG in 6 patients
 - Hippocampus with abnormal EEG in 4/5 patients recorded
 - Either synchronous or rapid spread to the hippocampus
 - Santos de Souza *neurosurg rev* 2018
 - sEEG in patient with temporal encephalocele
 - frequent SW left temporal pole, amygdala, hippocampus
 - Martinoni, *acta neurochir* 2017
 - Temporopolar encephalocele and HS
 - Aquilina, *JNS peds* 2010
 - Ictal subdural EEG - 2 sz – anterior/middle temporal cortex (quiet subtemporal/pole) and anterior subtemporal region
 - Diffuse temporal lobe disorganization



Encephalocele = Epileptogenic zone/lesion?

- If they are *always* epileptogenic, then lesion removal should result in cessation of seizures
- Saavalainen, *neurology* 2015
 - 12 patients underwent surgery (5 ATL/AH, 7 TPR)
 - ATL – 4 Engel 1A, 1 Engel 1B
 - TPR – 4 Engel 1A, 3 others - Engel 2A/2/3A



Why remove more than encephalocele?

- The epilepsy surgery teams are doing the right thing
 - treating the case as a true epilepsy case and not just a lesion resection case
- The nondominant hippocampus has no place being in the head



What is the “lesion?”

- Darn surgeons keep taking out more than the encephalocele
- Saavalainen, *neurology* 2015
 - 12 patients underwent surgery (5 ATL/AH, 7 TPR)
 - ATL – 4 Engel 1A, 1 Engel 1B
 - TPR – 4 Engel 1A, 3 others - Engel 2A/2/3A
- Toledano – *epilepsia* 2016
 - 22 patients, only 5 with surgery but 3 patients had standard right ATL
- Abou-Hamden, *epilepsia* 2010
 - 3 patients, all had temporal polar resections larger than encephalocele
- Aquilina, *JNS peds* 2010
 - Standard left ATL with temporopolar encephalocele
- Byrne, *world nsgy* 2010
 - 3 cases, 2 ATL
- Panov, *epilepsia* 2016
 - 6 patients, 4 standard ATL



Conclusions

- Are encephaloceles epileptogenic?
 - At times, yes
- Are encephaloceles ALWAYS epileptogenic?
 - No
 - Dual pathology
 - Wider issues such as cortical dysplasia



Conclusions

- Don't be suckered in like my residents and focus on the obvious MRI abnormality
 - These are epilepsy patients, not glioma patients
- Always have a working hypothesis but maintain an open mind as to the location of the epileptogenic zone
- Treat the patient, not the MRI
- Treat your neurosurgeon nicely, we have fragile egos



Principle

Everyone is entitled to his own opinion, but not his own facts.
-Ronald Reagan
-Daniel Patrick Moynihan



Thank you!



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