BRAIN SURGERY IN STURGE-WEBER SYNDROME: When is the right time?

Harry T. Chugani, M.D.
Chief, Pediatric Neurology
Director, Center for Neuroscience
Nemours A.I. duPont Hospital for Children
Wilmington, Delaware, USA

Professor of Pediatrics and Neurology
Thomas Jefferson University Sidney Kimmel College of Medicine, Philadelphia, PA, USA

Jefferson

Nemours Alfred I. duPont Hospital for Children

OUTLINE

- GENERAL CONCEPTS OF EPILEPSY SURGERY
- WHAT IS THE NATURAL COURSE IN SWS?
- HEMISPERECTOMY VS. FOCAL RESECTIONS
- 'AUTOHEMISPHERECTOMY'
- BRAIN PLASTICITY
- CORPUS CALLOSOTOMY IN SWS
- BILATERAL SWS

WHY PERFORM EPILEPSY SURGERY IN INFANTS & YOUNG CHILDREN?

- PERSISTENT SEIZURES IN THIS AGE GROUP OFTEN RESULTS IN AN EPILEPTIC ENCEPHALOPATHY
- MANY SEIZURE MEDICATIONS HAVE COGNITIVE SIDE EFFECTS
- EPILEPSY SURGERY SHOULD BE CONSIDERED WHEN EPILEPSY IS INTRACTABLE, 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)

WHAT IS THE PROCESS?

- ESTABLISH THAT PATIENT IS AN APPROPRIATE CANDIDATE
- VIDEO-EEG ADMISSION ON OR OFF MEDICATIONS TO SHOW THAT SEIZURES ARE FOCAL, COMING FROM ONE HEMISPHERE (INTERICTAL EEG, ICTAL EEG, SEIZURE SEMIOLOGY)
- ESTABLISH FOCALITY ON MRI AND/OR PET
- NEUROPSYCHOLOGICAL BASELINE
- FUNCTIONAL MRI FOR LANGUAGE, MOTOR FUNCTIONS
- SURGERY CONFERENCE TO REACH CONSENSUS
- TYPE OF SURGERY: USUALLY ONE STAGE IN SWS
- IDENTIFY MOTOR CORTEX IF NECESSARY
- SURGICAL RESECTION: FREEZE PORTION OF TISSUE
- POST-RESECTION ELECTROCORTICOGRAPHY

EPILEPSY SURGERY IN SWS

- MOST PATIENTS WITH SWS WILL NOT NEED EPILEPSY SURGERY
- THE BRAIN LESION (LMA) WILL UNDERGO A DYNAMIC COURSE OF ITS OWN AND EVENTUALLY STABILIZE
- THIS DYNAMIC COURSE IS DIFFERENT FROM CHILD TO CHILD
- THE KEY IN UNDERSTANDING SWS IS WHETHER THIS COURSE IS ASSOCIATED WITH UNACCEPTABLE COGNITIVE DEFICITS

LONG TERM FOLLOW-UP IN STURGE-WEBER SYNDROME

- Pascual-Castroviejo et al., 2008: 55 patients long term
- Although cerebral lesions followed a progressive course during early childhood, a plateau was eventually reached and the patient stabilized
- Epilepsy, hemiparesis, intellectual disability and ocular problems were the most frequent and severe features
- Major impediments towards a successful integration into adult society (i.e., gainful employment, quality of life) were limited intelligence, poor social skills, poor aesthetic appearance due to the facial angioma, and poor seizure control

WHO ARE THE SURGICAL CANDIDATES?

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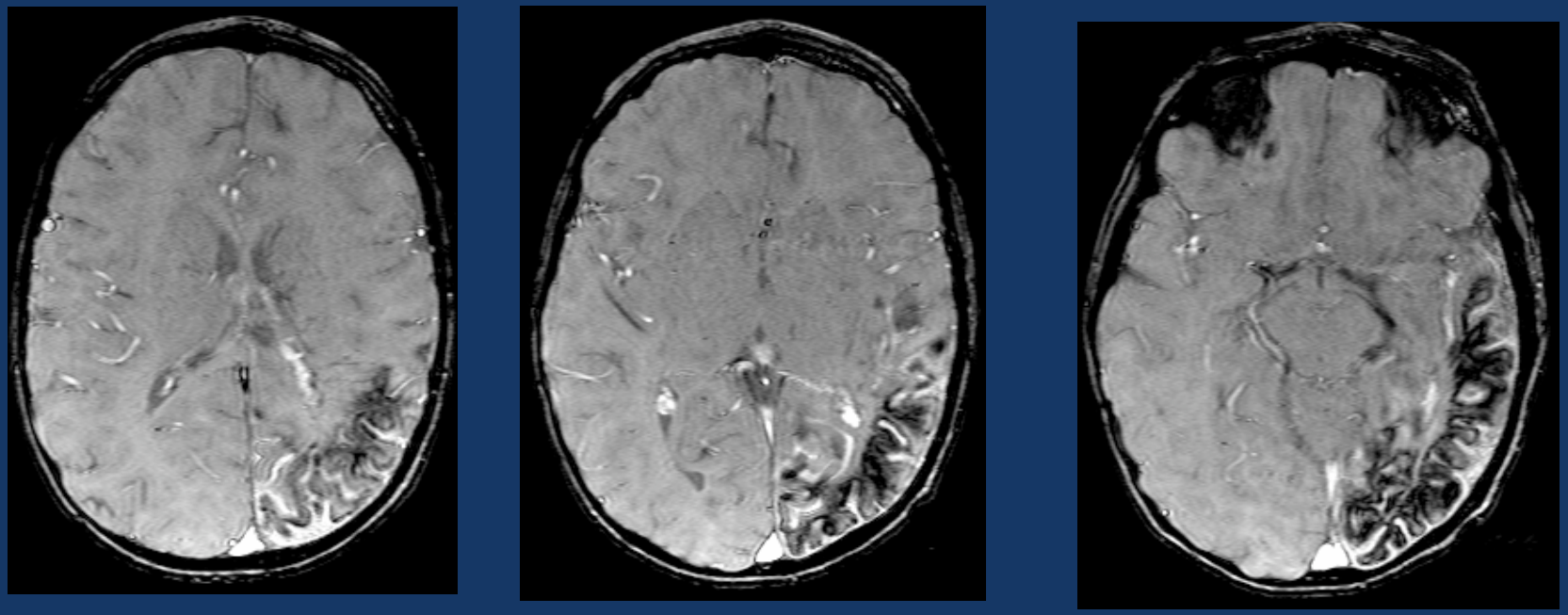
STURGE-WEBER SYNDROME: Surgery

- Probably no more than 20% of children with SWS will require surgery
- Those with unilateral posterior LMA sparing the motor cortex are excellent candidates if they have seizures and developmental delay. No weakness, but will have visual field loss on one side
- Children with unilateral hemispheric LMA with multiple episodes of life-threatening seizures should undergo hemispherectomy

STURGE-WEBER SYNDROME: Surgery

- If having seizures but developmentally at or near target, WAIT!
- Let the developmental status be the main guide as to whether or not to operate
- Make sure the opposite hemisphere is normal
- Very rarely, bilateral cases can be surgical candidates for palliative resection

STURGE-WEBER SYNDROME:
POSTERIOR LEPTOMENINGEAL ANGIOMA
IDEAL CANDIDATE FOR POSTERIOR RESECTION SPARING MOTOR CORTEX



22 months old boy with frequent seizures and developmental delay

SWI post-Gad.

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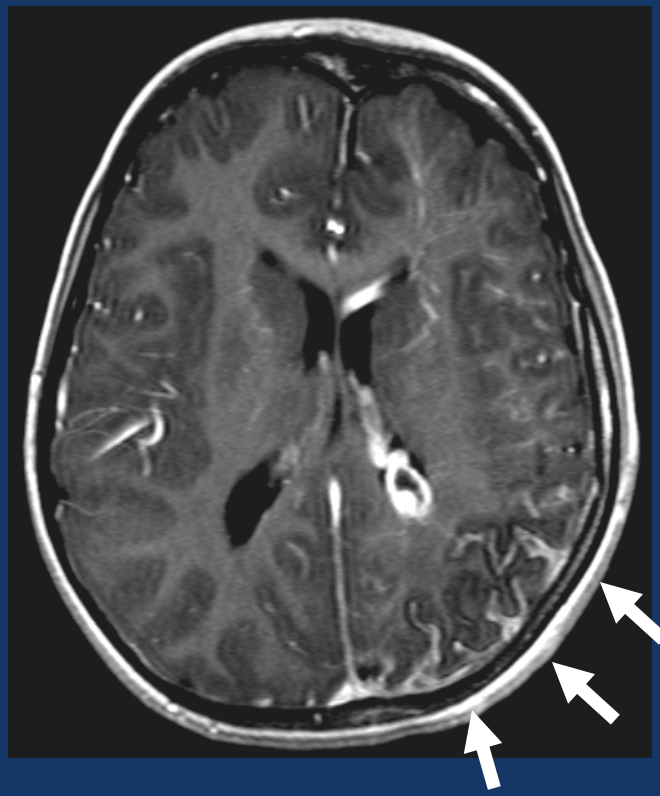
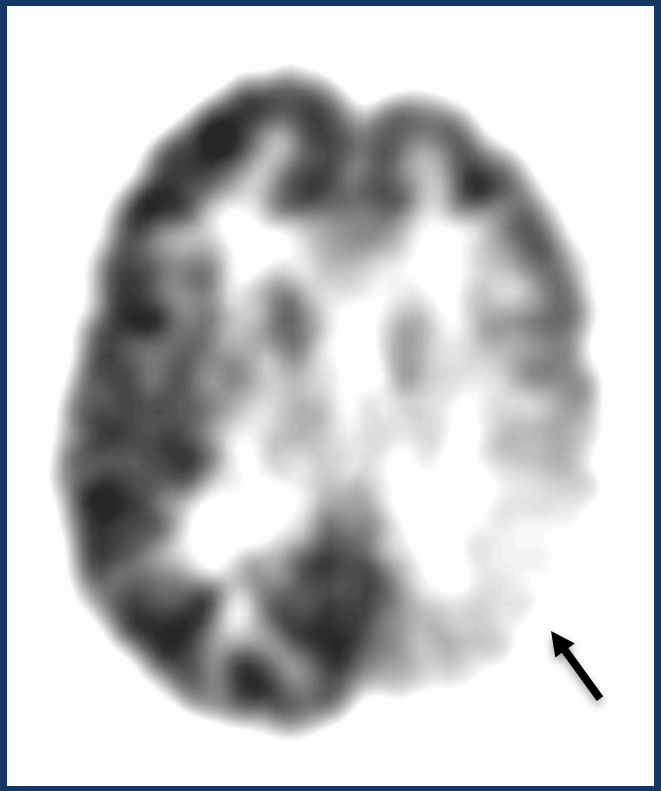
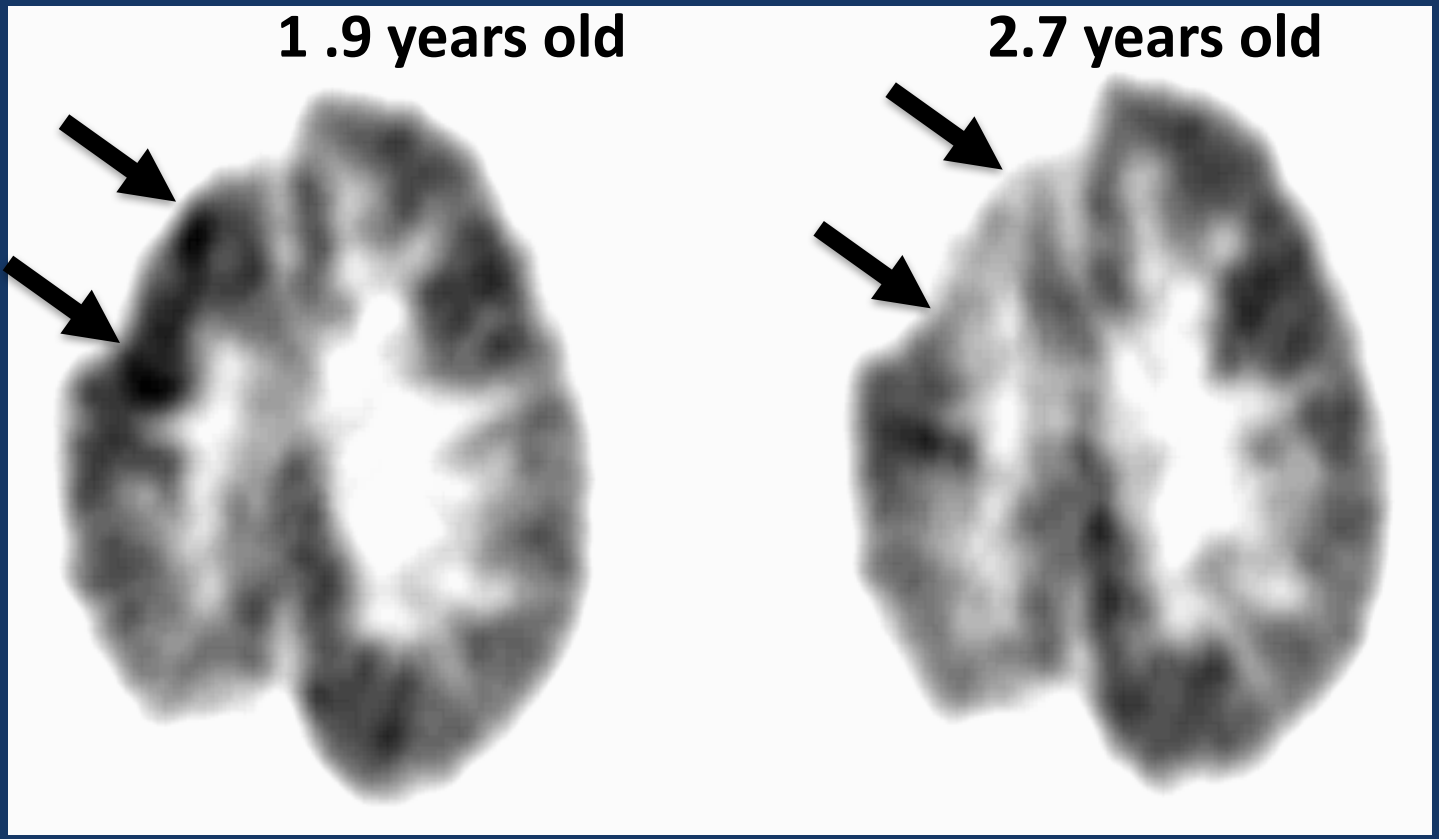




EARLY METABOLIC CHANGES IN STURGE-WEBER SYNDROME



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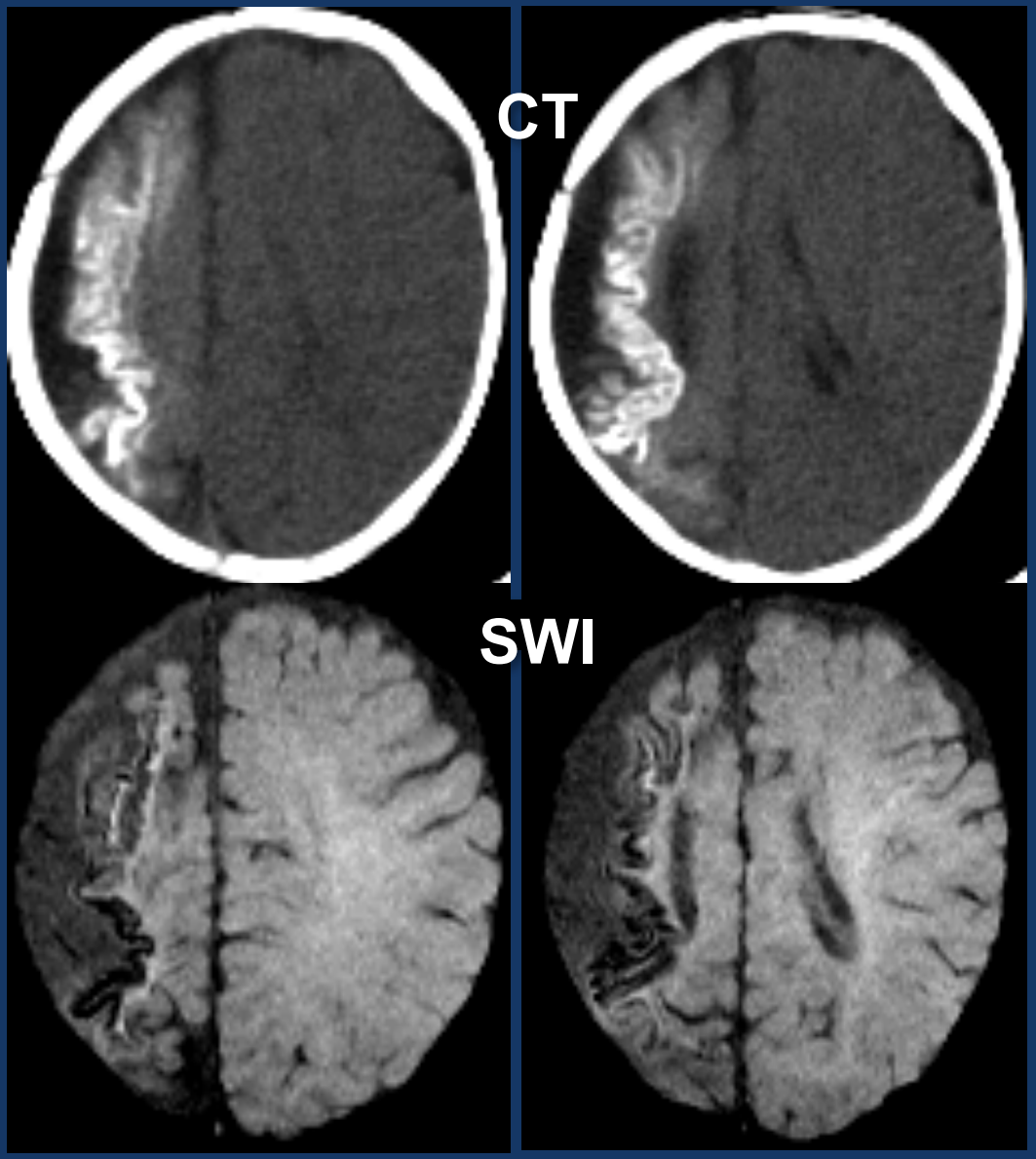

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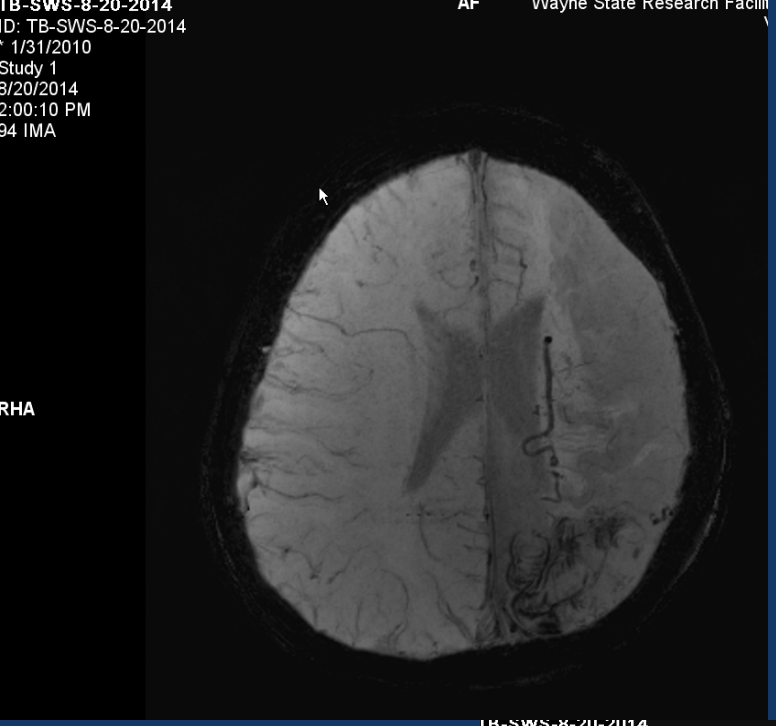

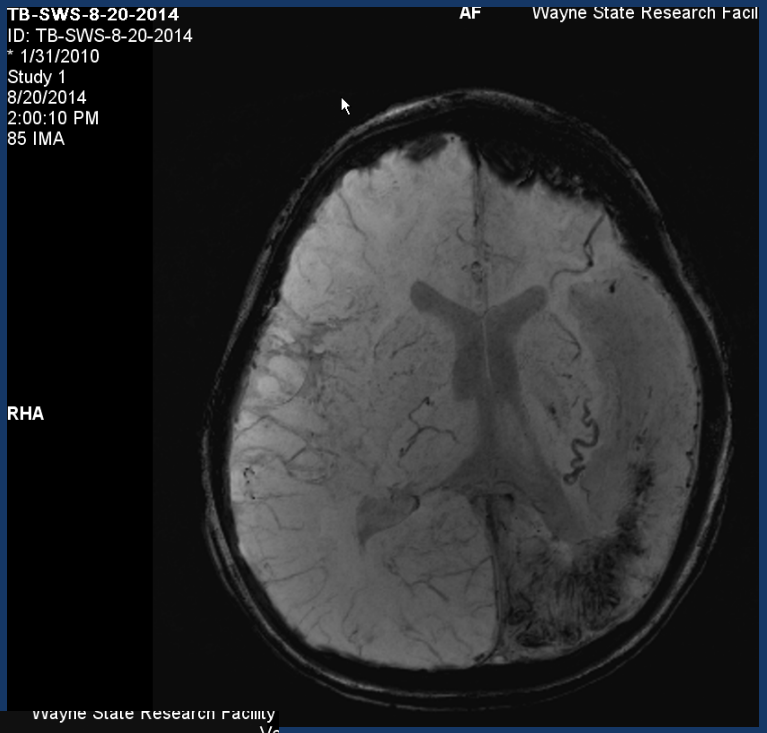

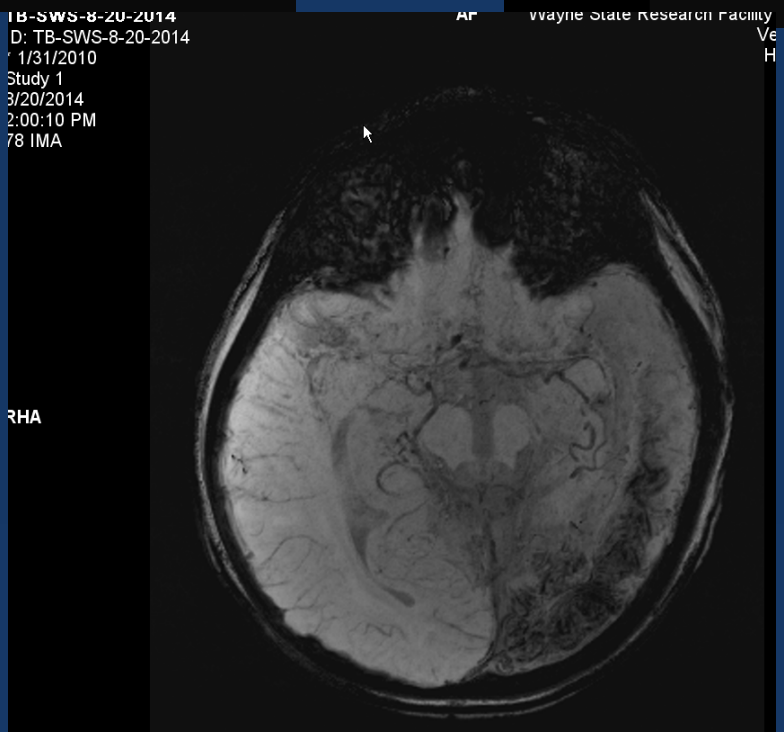

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STURGE-WEBER SYNDROME: Surgery

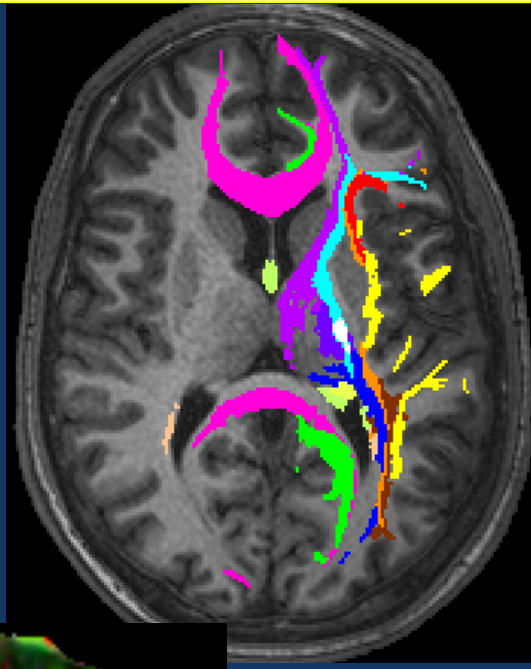
- Although controversial, we believe that children with progressive developmental delay, but whose seizures are not all that intractable, should also be evaluated for surgery to preserve cognitive function by ‘forcing’ reorganization in the opposite hemisphere

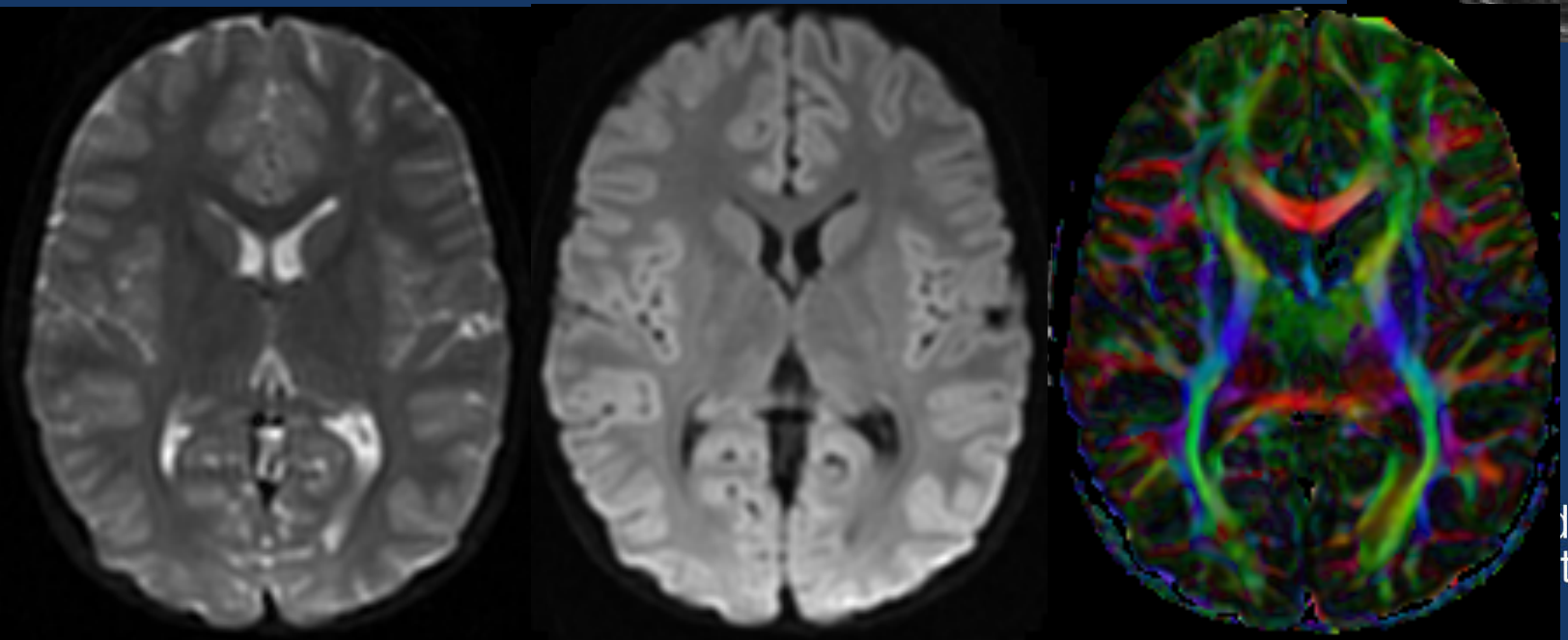
- Surgery is most successful in first 3-4 years when brain plasticity is at high capacity

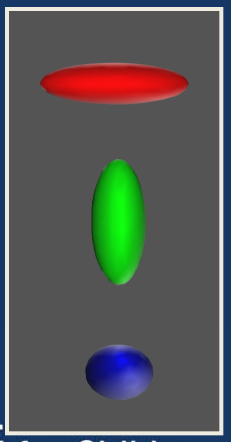
3T whole-body PET/MR scanner allowing simultaneous acquisition of PET and MR data

Diffusion Tensor MRI

- Brain water diffusion
  - Integrity of white matter fibers
  - Intactness of brain connectivity
  - Rapidly evolving technique
  - Fractional anisotropy (FA)
  - Apparent diffusion coefficient (ADC)







2 YEAR OLD BOY WITH STURGE-WEBER SYNDROME AFFECTING ENTIRE RIGHT BRAIN, UNDERGOING STROKE-LIKE EPISODES

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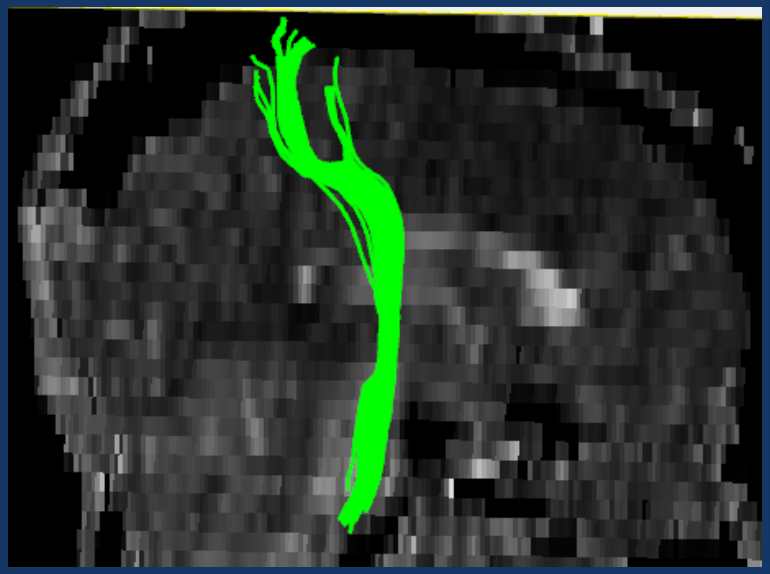
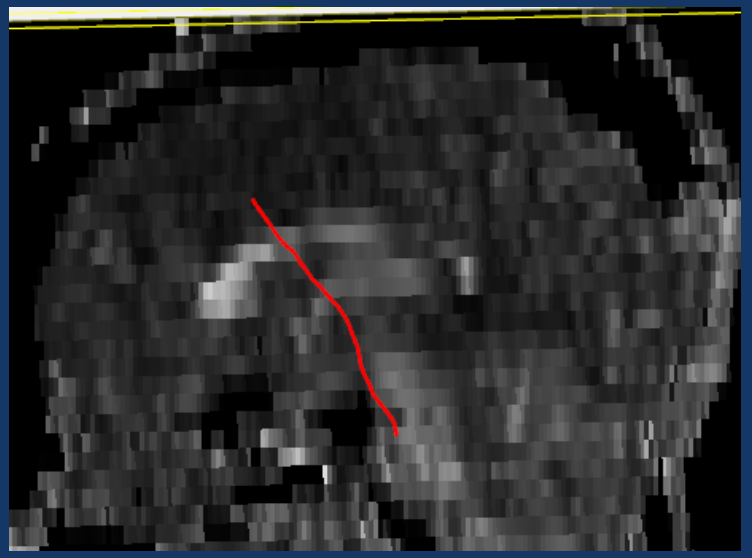
Loss of left corticospinal tract in child with hemiplegia



Right CST



Left CST

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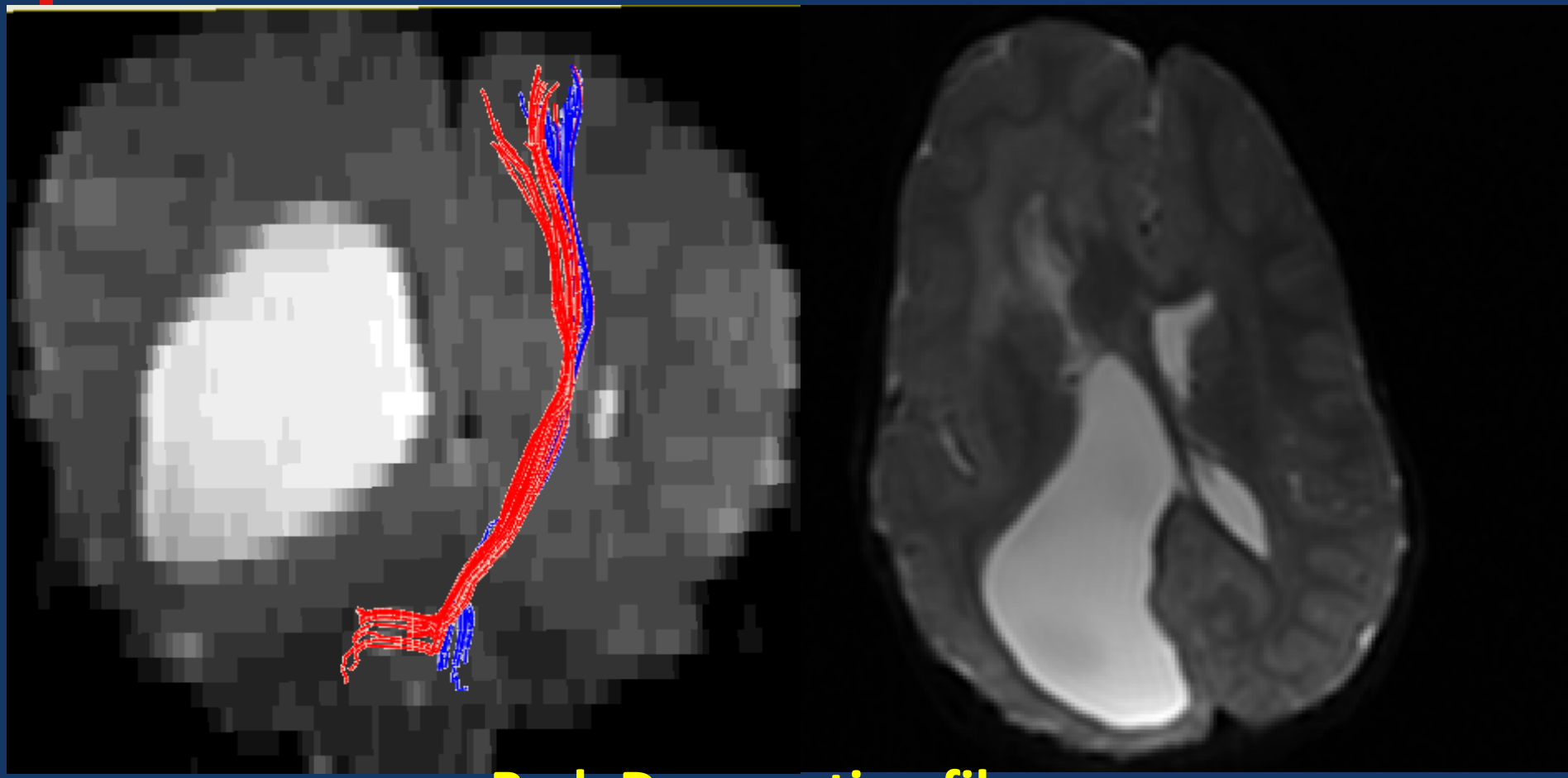
3 year old boy with hemimegalencephaly



Red: Decussating fibers

Blue: Non-decussating fibers

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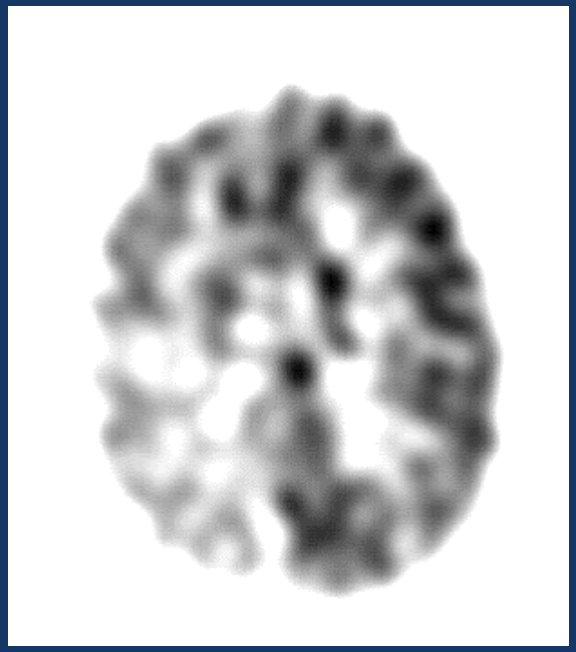
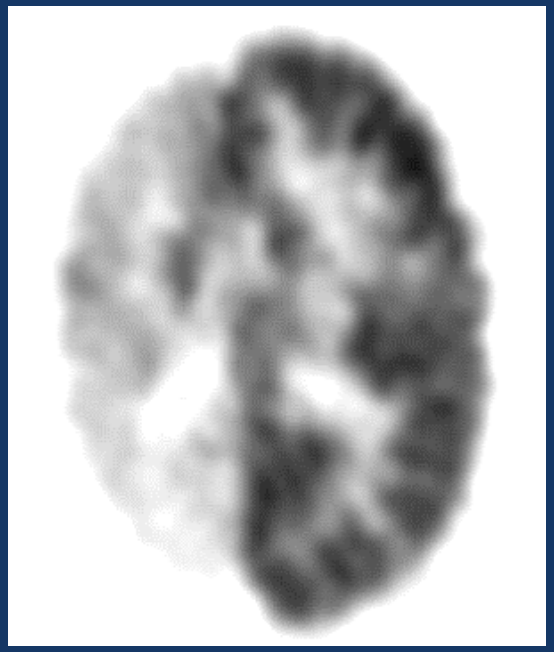
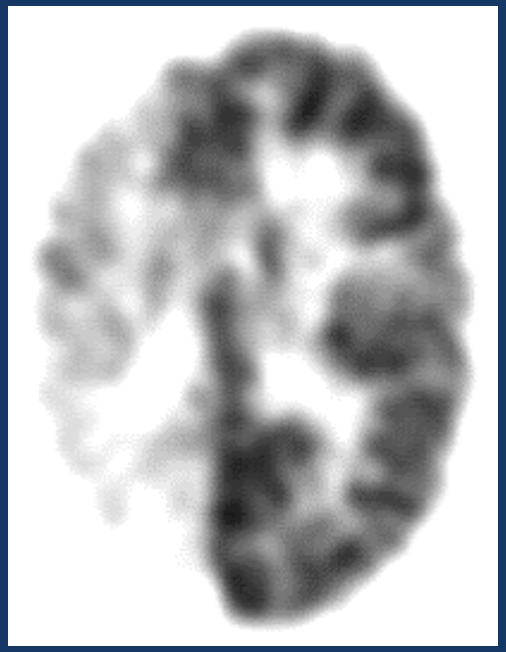
STURGE-WEBER SYNDROME:
Rapid Progression of Severe Hypometabolism is Associated with Good Cognitive Outcome



5 months



38 months



5 years

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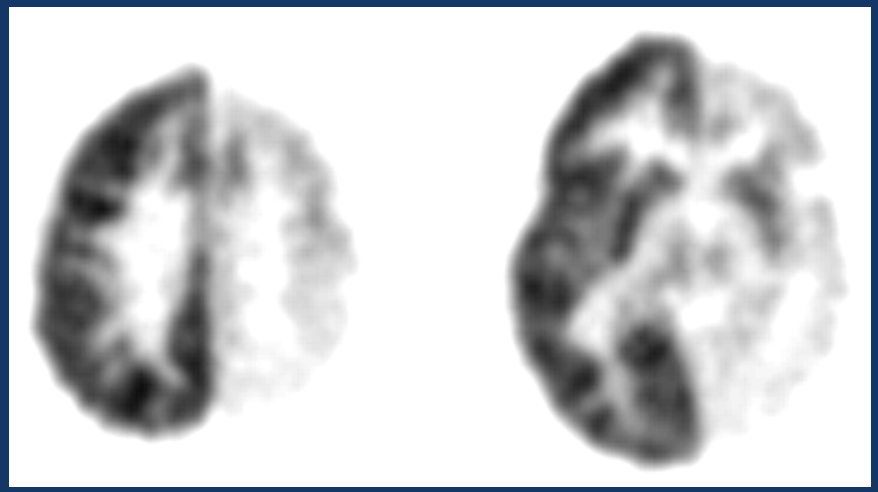
STURGE-WEBER SYNDROME

- Behen et al., 2011: Stabilization of lesion consistent with our own studies
- ***Early*** demise of affected hemisphere associated with better cognitive function
- Forces reorganization in contralateral hemisphere when developmental plasticity is at a maximum

40 months old boy Verbal IQ: 93



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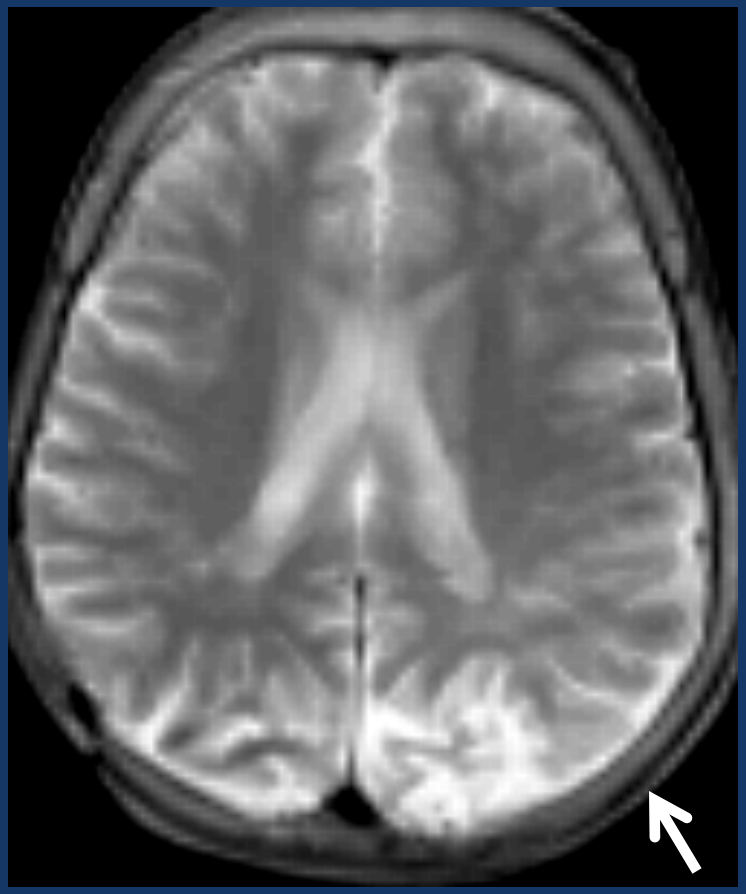
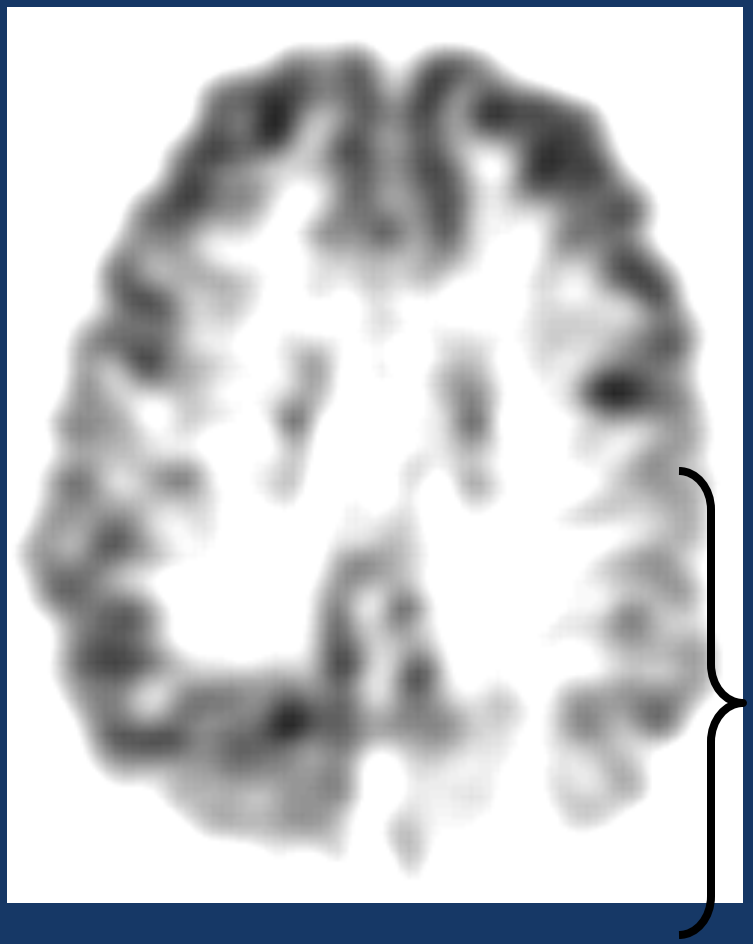
STURGE-WEBER SYNDROME:
Large Area of Mild Glucose Hypometabolism is Associated with Poor Cognitive Outcome

MRI



FDG PET



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Extent of severe unilateral cortical damage vs. IQ

U-shaped correlation

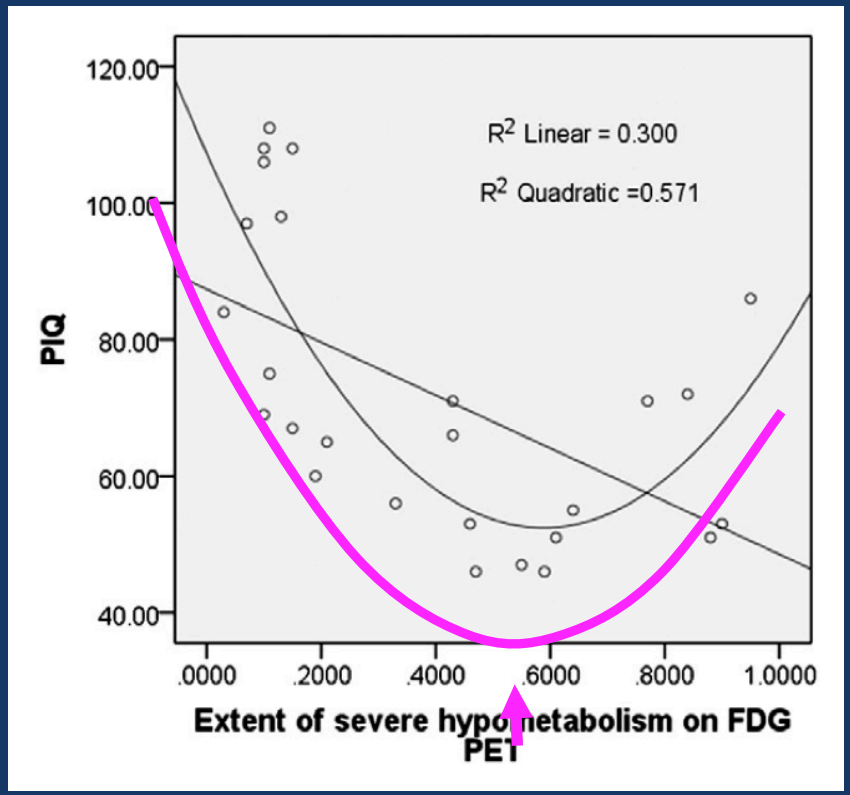
- Children with unilateral SWS (n=35)

- Extent of severe hypometabolism vs. IQ: quadratic correlation ($P < 0.001$)

- Both PIQ and VIQ

- Also affected by epilepsy severity (seizure frequency)

- Bottom of quadratic curve: ≈60% of severely hypometabolic hemisphere



Behen et al. Epilepsy Behav, 2011

Support for "fresh-start hypothesis (Bates): switch from intra- to interhemispheric reorganization

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LANGUAGE TRACT

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BRAIN PLASTICITY

- INTERHEMISPHERIC TRANSFER OF LANGUAGE FUNCTIONS HAS BEEN WELL STUDIED

- FUNCTIONAL MRI STUDIES IN EARLY LEFT HEMISPHERE INJURY SUPPORT TRANSFER OF LANGUAGE TO RIGHT HEMISPHERE IF THAT HEMISPHERE IS INTACT

- HOWEVER, EFFECTIVE LANGUAGE TRANSFER TO RIGHT HEMISPHERE MAY INTERFERE WITH NON-VERBAL ACTIVITIES (e.g., VISUAL-SPATIAL), KNOWN AS 'CROWDING EFFECT'

BRAIN PLASTICITY (contd)

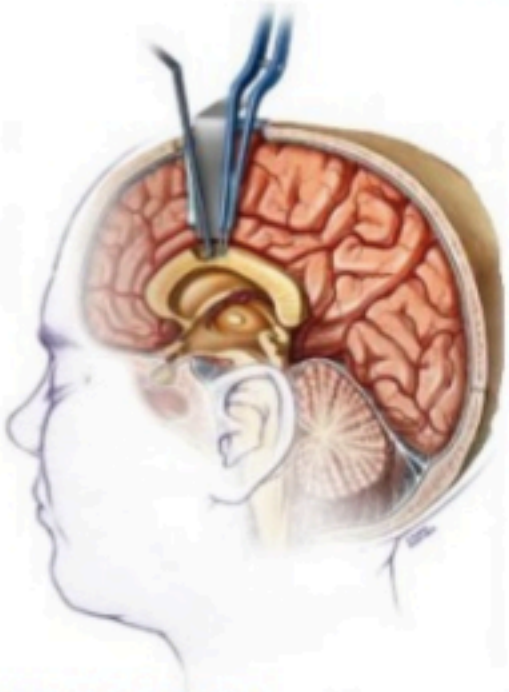
- OUR RECENT STUDY USING PET, MRI AND NEUROPSYCHOLOGICAL TESTING ON 27 CHILDREN WITH SWS FOUND THE MAJORITY HAD HIGHER VERBAL THAN NON-VERBAL IQ, REGARDLESS OF THE SIDE OF THE LMA

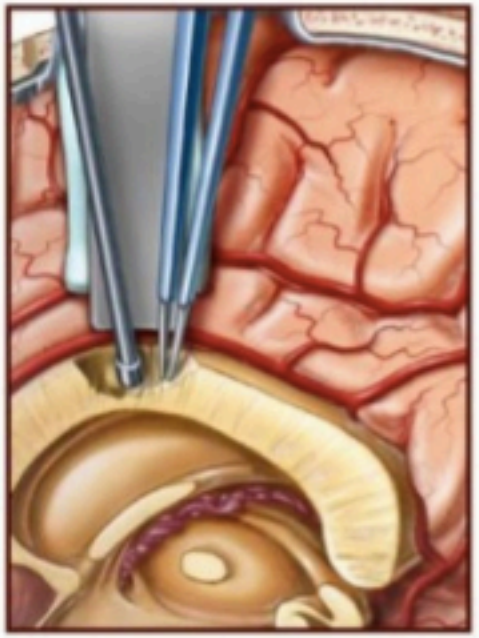
- OVER 70% OF THE LEFT HEMISPHERIC GROUP SHOWED A WEAKNESS OF NON-VERBAL IQ EVEN THOUGH THE RIGHT HEMISPHERE WAS NORMAL

- 'COMPETITIVE EDGE' OF VERBAL OVER NON-VERBAL (SPATIAL) FUNCTIONS

- PET CORRELATIONS SHOWED A MORE ROBUST FUNCTIONAL EFFECT OF LEFT HEMISPHERIC SWS LESIONS ON THE RIGHT HEMISPHERE THAN VICE-VERSA, THUS MAKING PRESERVATION OF SPEECH A HIGH PRIORITY, EVEN AT THE EXPENSE OF 'CROWDING OUT' NON-VERBAL FUNCTIONS

CORPUS CALLOSOTOMY





•**CORPUS CALLOSOTOMY** or the so called Split-Brain Surgery disconnects the 2 hemispheres of the brain by cutting the corpus callosum, which is the band of nerve fibers connecting the 2. This is mainly performed in patients with uncontrollable seizures with a potential of severe injury due to falls.

CORPUS CALLOSOTOMY

- ONLY 2 REPORTS FOUND
- AVILA ET AL., 1980: 18 YR OLD GIRL, SEVERELY DELAYED, ANTERIOR CALLOSOTOMY, MUCH IMPROVEMENT
- RAPPAPORT ZH, 1988: TWO CHILDREN WITH INTRACTABLE GENERALIZED EPILEPSY WITH EXCELLENT RESULTS

Pediatr Neurol. 2011 Jun;44(6):443-9. doi: 10.1016/j.pediatrneurol.2011.01.005.

Clinical outcomes in bilateral Sturge-Weber syndrome.

Alkonyi B[1], Chugani HT, Karia S, Behen ME, Juhász C.

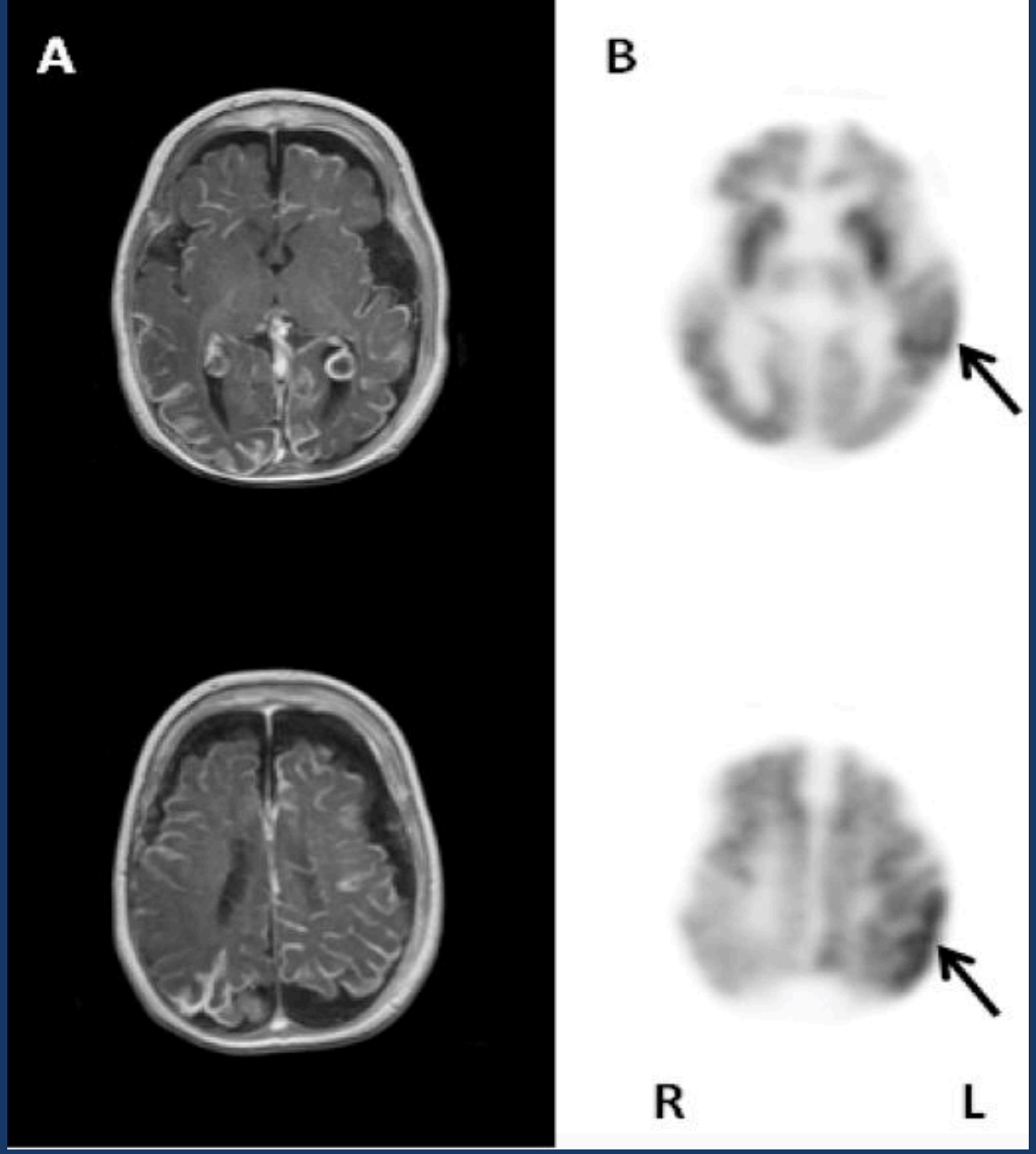
Author information

Abstract

Approximately 15% of patients with Sturge-Weber syndrome demonstrate bilateral intracranial involvement, and the prognosis of these patients is considered particularly unfavorable. We reviewed the clinical and neuroimaging features of patients with Sturge-Weber syndrome and bilateral intracranial involvement. Seizure variables, the presence of hemiparesis, and the degree of developmental impairment at most recent follow-up were compared with imaging abnormalities. Of 110 Sturge-Weber syndrome patients, 14 demonstrated bilateral brain involvement, with an asymmetric pattern on glucose metabolism positron emission tomography. Although most patients manifested frequent seizures initially, associated with frontal hypometabolism on positron emission tomography, six (43%) had achieved good seizure control during follow-up. Bilateral frontal hypometabolism was associated with severe developmental impairment. Two children with bitemporal hypometabolism exhibited autistic features. Hemiparesis was associated with superior frontal (motor cortex) hypometabolism. Three patients underwent resective surgery, resulting in improved seizure control and developmental outcomes. The severity of neurologic complications and clinical course depend on the extent of cortical dysfunction in bilateral Sturge-Weber syndrome. Bilateral frontal and temporal hypometabolism is associated with poor developmental outcomes. Good seizure control and only mild/moderate developmental impairment can be achieved in about 50% of patients with bilateral Sturge-Weber syndrome, with or without resective surgery.

BILATERAL LEPTOMENINGEAL ANGIOMAS IN STURGE-WEBER SYNDROME

Severe developmental delay and seizures



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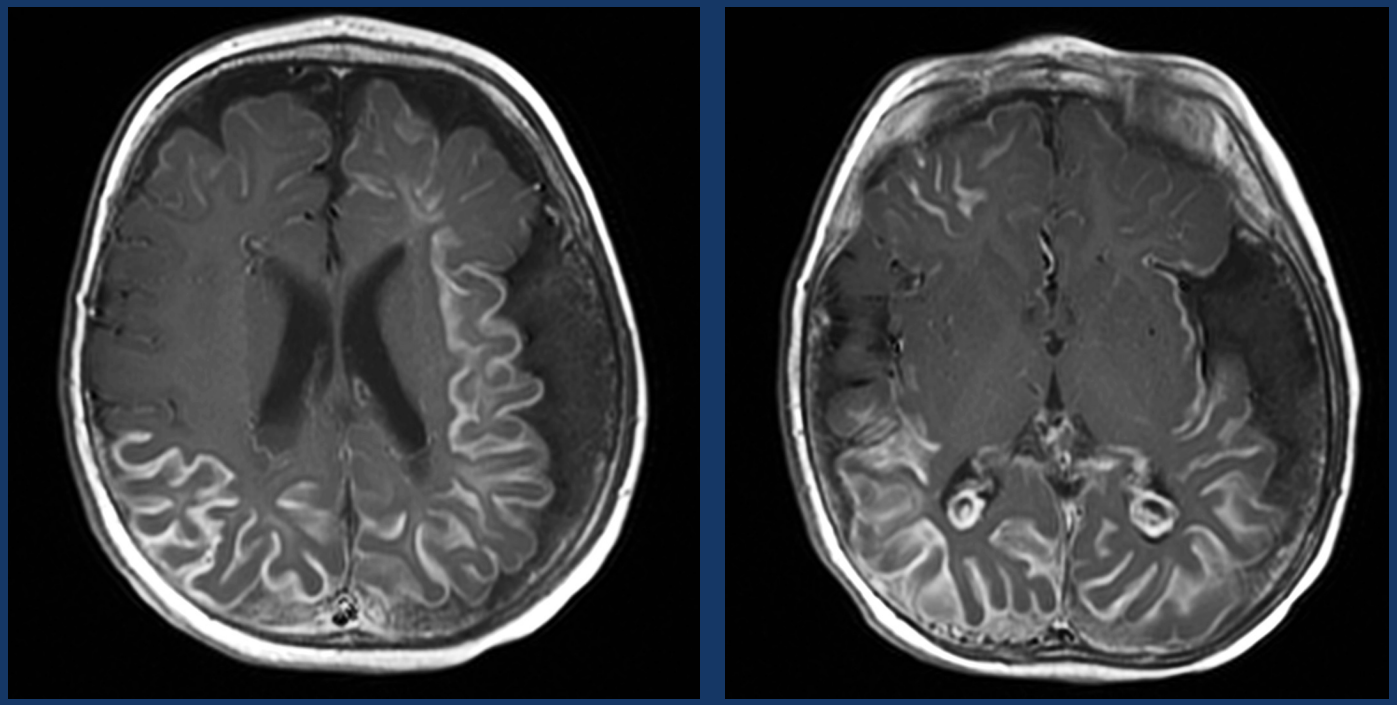
4 year old boy, bilateral LMA, severe delay, seizures



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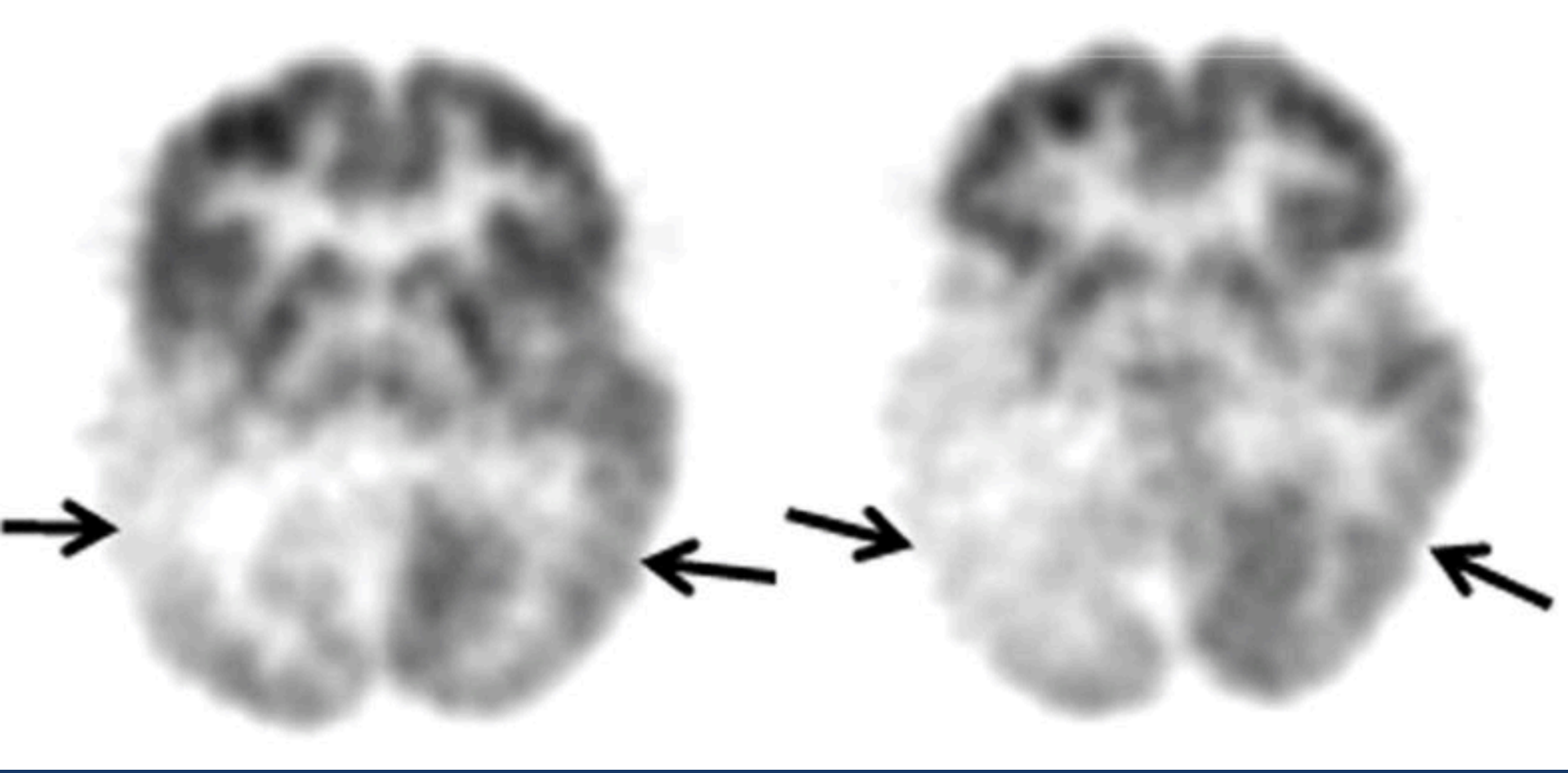
BILATERAL LEPTOMENINGEAL ANGIOMAS IN STURGE-WEBER SYNDROME

Acute development of autism following a stroke-like episode



TAKE HOME MESSAGES

- SWS IS A PROGRESSIVE DISORDER DUE TO THE BRAIN LESION UNDERGOING CHANGES IT MUST GO THROUGH
- WE WANT THESE CHANGES TO BE DONE EARLY IN LIFE (BEFORE 3 TO 4 YEARS) SO THAT THE OTHER HEMISPHERE CAN TAKE OVER AT A TIME OF HIGH BRAIN PLASTICITY
- WHY THEN WOULD YOU WANT TO SLOW THIS DOWN WITH ASPIRIN? (EVEN IF ASPIRIN DOES SOMETHING, FOR WHICH THERE ARE NO SCIENTIFIC DATA)
- MANY PATIENTS WILL UNDERGO AN 'AUTOHEMISPHERECTOMY' EARLY IN LIFE
- IF THEY TAKE THEIR TIME AND SHOW WORSENING DELAYS, WE SHOULD OPERATE
- THE MAIN DRIVER SHOULD BE THE COGNITIVE FACTOR

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