

The yield of brain imaging in patients with Sturge Weber syndrome presenting with acute neurologic symptoms

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Disclosure

- * nothing to disclose

Neurological Features

- * Signs of brain involvement by abnormal **brain MRI**
- * Signs of abnormal **clinical** neurological symptoms

Neurological symptoms

- * Neurological deficits can be acquired slowly over time
- * Toddlers and young children are prone to “stroke-like events” triggered by falls with mild head

Zolkipli et al 2007

Epilepsy natural history in SWS

Kossoff *et al* *Epilepsia* 2009

- * Seizure control and only mild/moderate developmental impairment
- * Young age of onset is associated with higher level of hemiparesis
- * SE is common
- * Prolonged Todd's paralysis and new onset of visual field deficit

Introduction:

- * Seizures, stroke-like episodes and headaches are common complications in patients with Sturge-Weber syndrome (SWS).
- * We hypothesize that patients with SWS have frequent brain imaging studies when presenting acutely to the ER, which may potentially be not necessary in every case.

The yield of brain imaging in patients with Sturge Weber syndrome presenting with acute neurologic

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Methods:

- * To determine frequency and yield of brain imaging, in patients with SWS who presented with acute neurologic symptoms.
- * we conducted a retrospective chart analysis at Boston Children's Hospital between 1996-2016

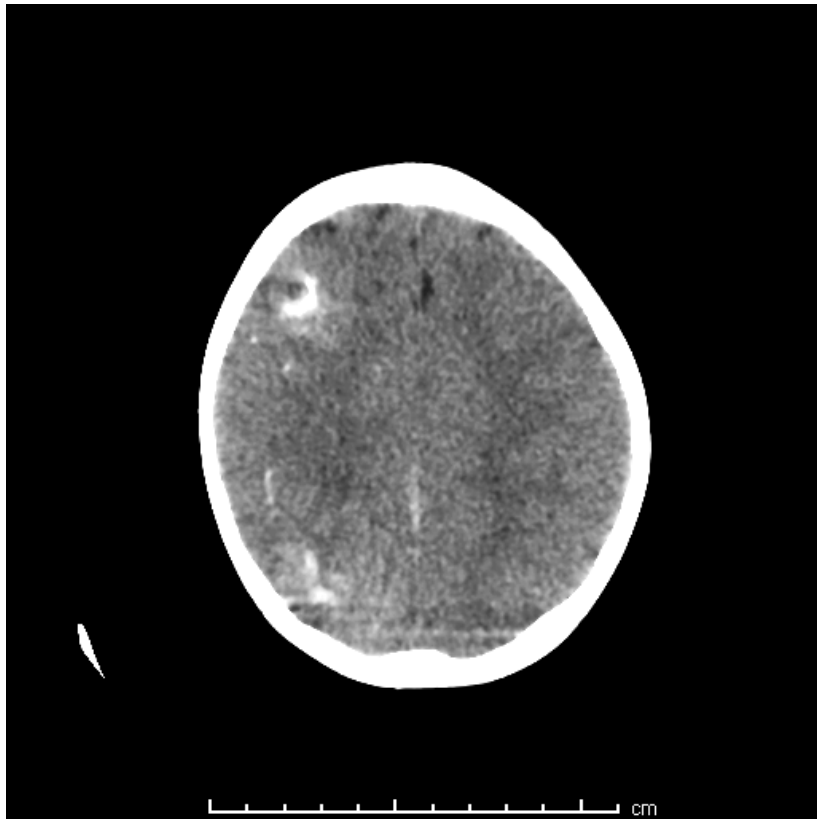
Results:

- * 36 patients with SWS accounted for a total of 141 encounters.
- * Images were obtained in 76/141 (53.9%)
- * total of 96 images (47 head CTs and 49 brain MRIs).
- * 19/76 (25%) - patients had both CT and MRI.

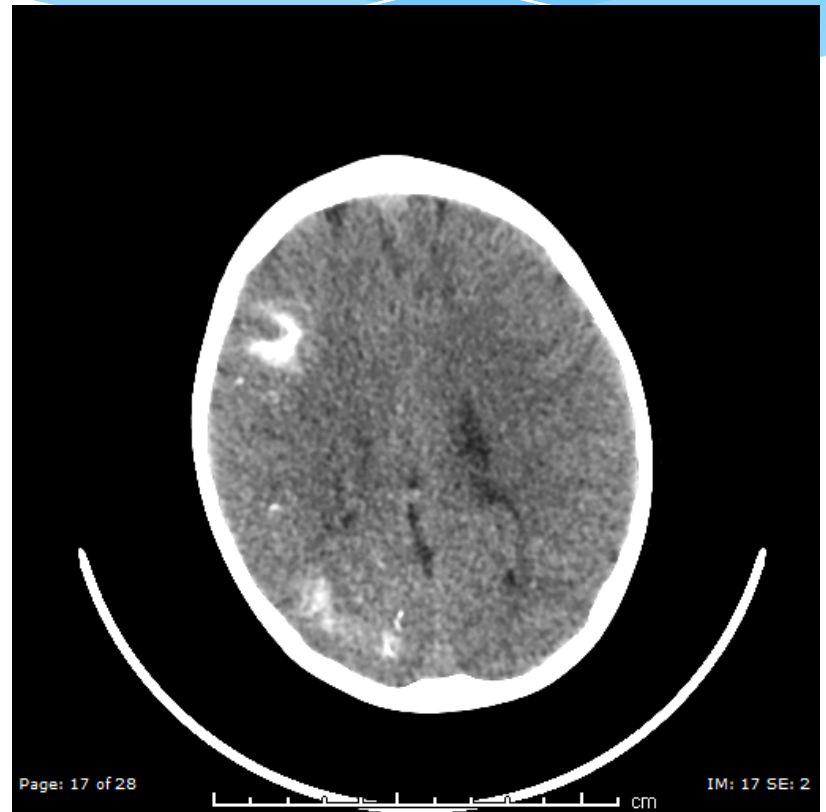
Results:

- * **None of the images showed acute hemorrhage**
- * Patients with stroke-like episodes were more likely to be imaged compared to patients presenting with seizures (89.7% vs 44.7%)
- * None of the images performed for seizures showed acute changes
- * One patient who presented with stroke-like episodes was found to have a new M1 stenosis.

CT scan



September 2015

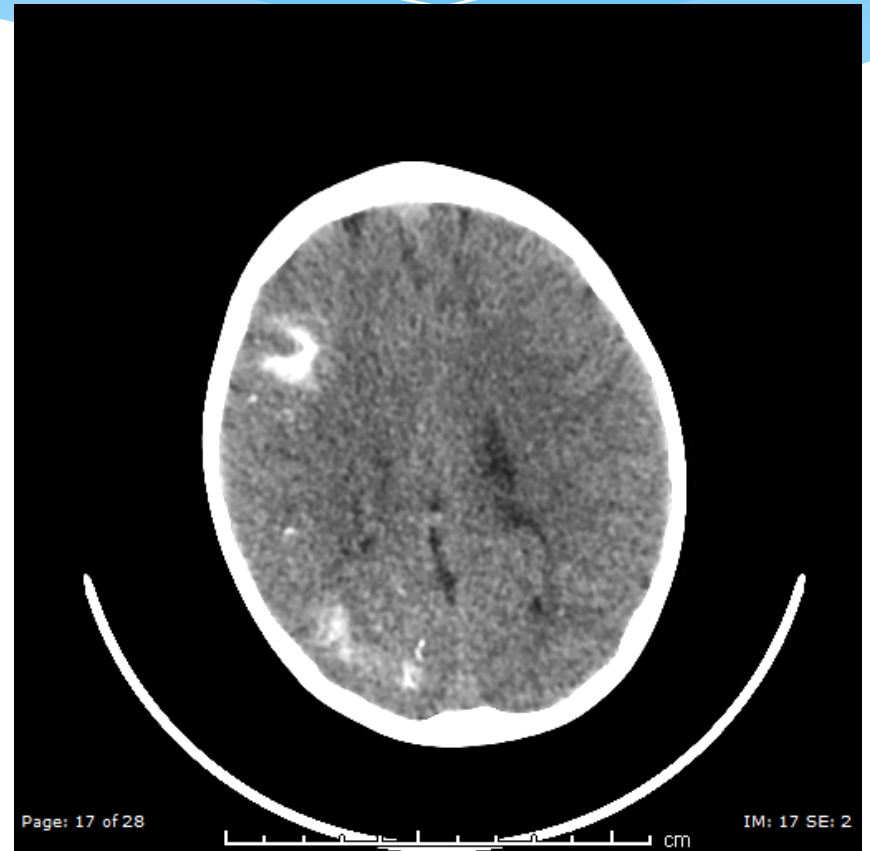


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Calcifications

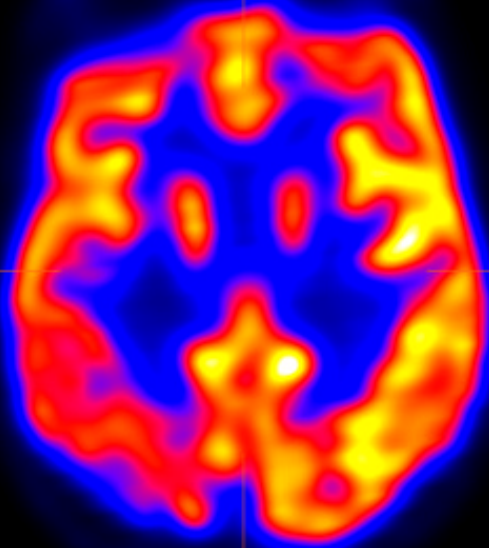


MRI - SWI

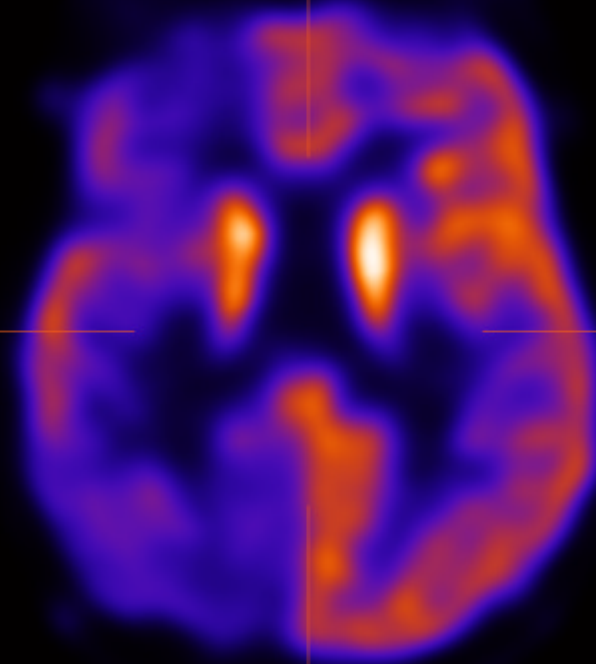


CT scan

PET scan

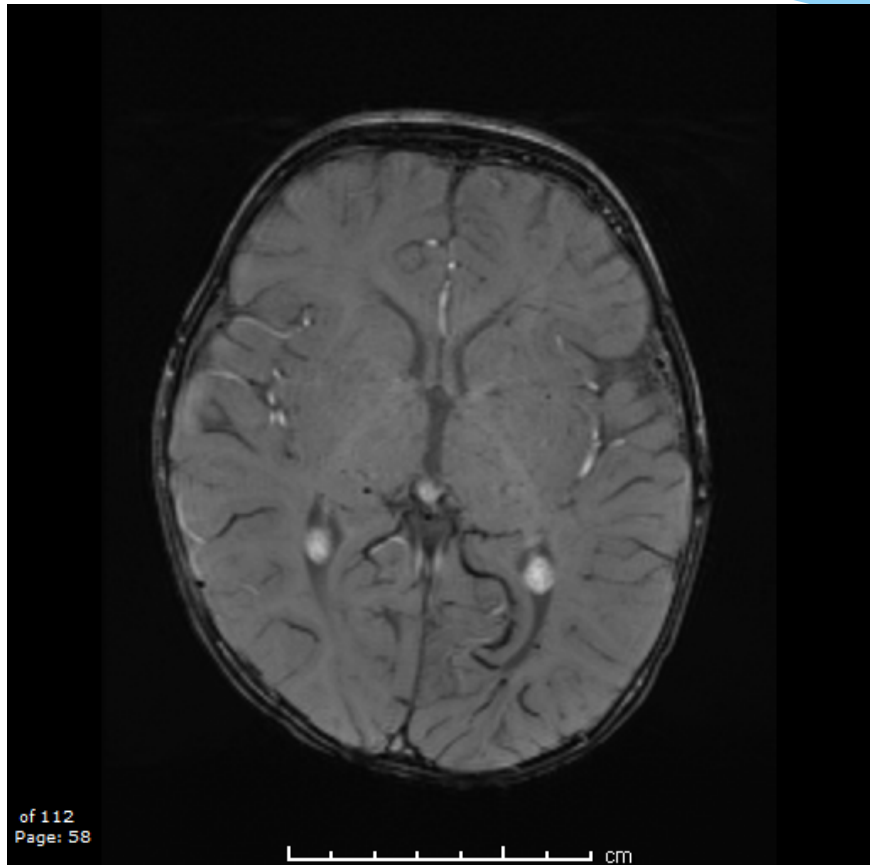


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Prominent deep venous anomaly

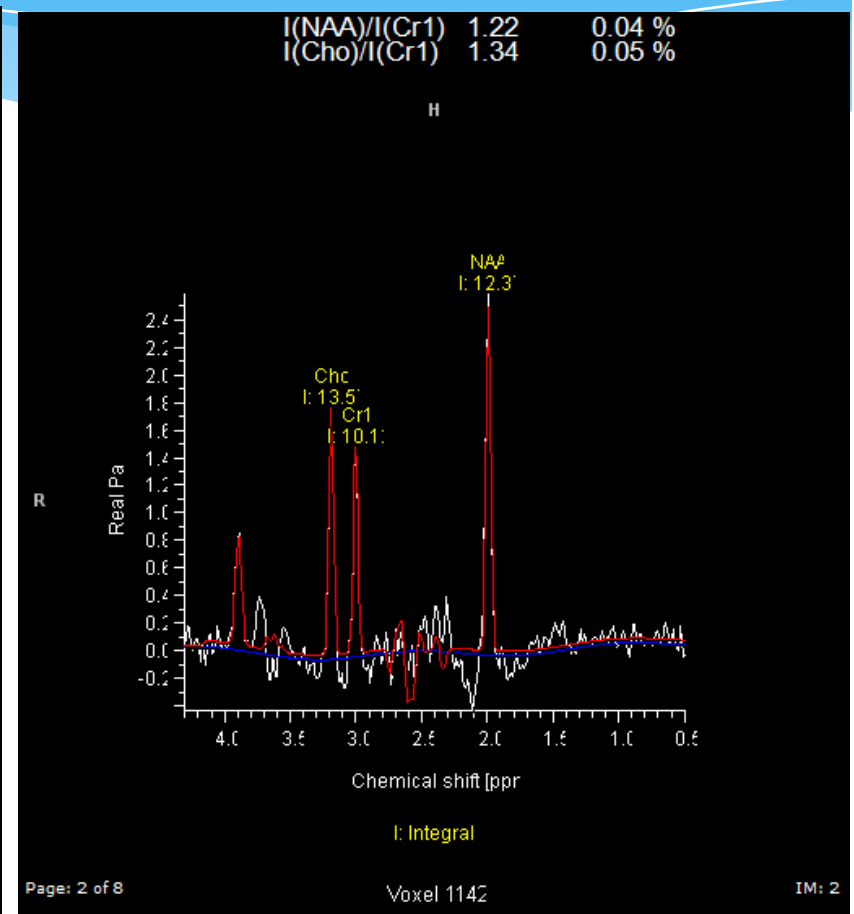
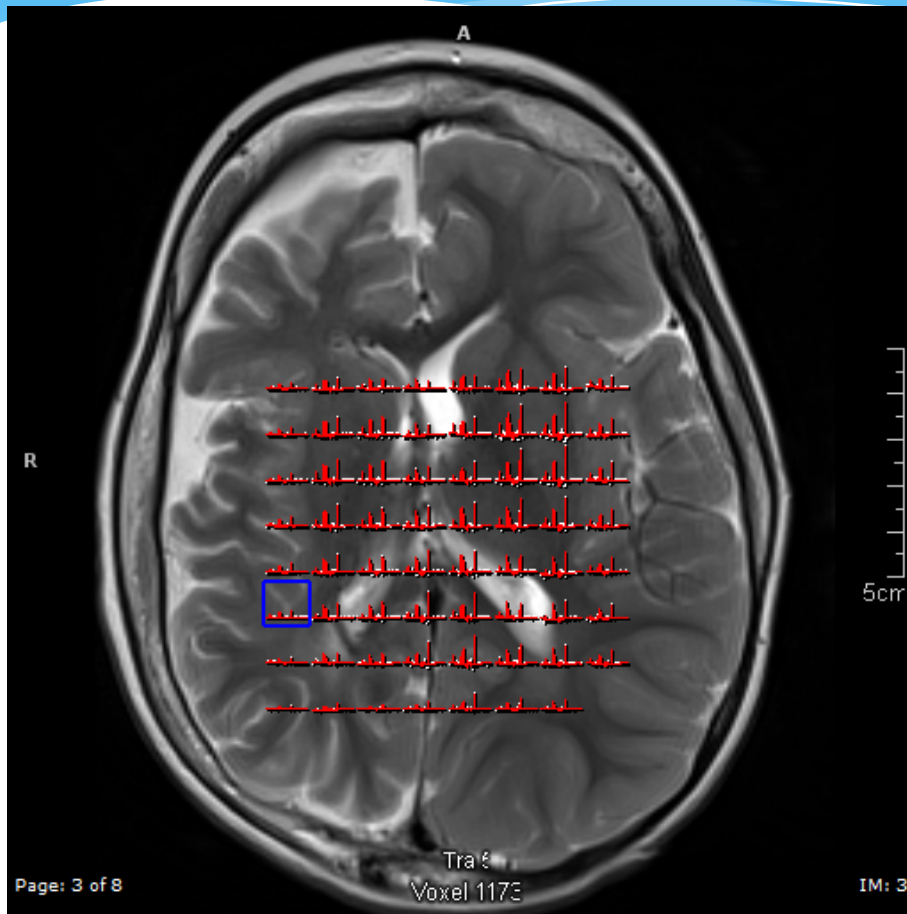


6 months old



4 years old

MR spectroscopy



Author /year	Premorbid neurological status	Location of hemorrhage	Signs and symptoms
Anderson 1974	Normal	Subarachnoid hemorrhage	<i>Headache nuchal rigidity</i>
Pozzati 1983	Epilepsy	left post cerebral artery aneurysm	<i>Headache hemiparesis</i>
Dolkart 1995	Epilepsy	Intraventricular hemorrhage – angioma of choroid plexus	<i>Seizure exacerbation</i>
Aguglia 2008	Hemiparesis headache	Left temporal hemorrhage	<i>New onset of seizures</i>
Lopez 2013	normal	Right subdural hemorrhage Injury of the occipital region	<i>Altered mental status</i>
Nakajima 2014	Epilepsy	Right thalamic hemorrhage Venous occlusion	<i>Seizures and altered mental status</i>

In summary

- * Acute neurologic manifestations of SWS frequently lead to acute brain imaging.
- * In our cohort, no patient had any evidence of acute hemorrhage,
- * And no patient who presented with seizures had any acute imaging findings resulting in a change in management.

Clinical indication for neuroimaging

- * New seizure type
- * Severe headache – or changes in headache pattern
- * Progressively worsening neurological deficits

Clinical indication

- * Altered mental status
- * Severe headache – not responsive to treatment and/or associated with other signs
- * **Nuchal rigidity, anisocoria, trauma, altered mental status**

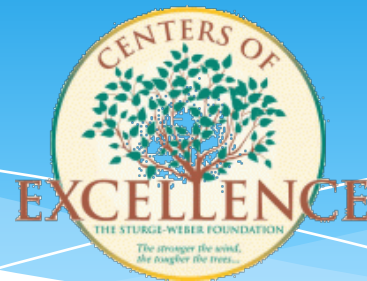
If neuroimaging is necessary, what modality to use?

- * Brain MRI with contrast (?) and additional sequences:
 - * Perfusion, calcification, white matter changes
- * PET scan
- * SPECT – ictal and interictal (?)
- * MR spectroscopy

Challenge: How to interpret the findings?

- * Consider cerebral “steal phenomenon”
- * “Paradoxical signals”
- * Changes caused by chronic hypoxic regimen (ex: “accelerated myelination”)
- * Active seizures – changes in metabolism
- * Consider limitations on the images technique and use of sedation

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