HIGH DIAGNOSTIC YIELD OF NON-CONTRAST BRAIN MRI WITH SWI IN NON-SEDATED CHILDREN WITH STURGE-WEBER SYNDROME

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INTRODUCTION

Sturge-Weber syndrome (SWS) is a sporadic neurocutaneous disorder, characterized by a vascular facial birthmark ("port-wine stain"), leptomeningeal venous malformation, and glaucoma (in 50%). The majority (cca. 85%) of SWS brain involvement is unilateral.

Common neurological manifestations of SWS include seizures, cognitive impairment, stroke-like episodes, hemiparesis, and visual field deficits. Behavioral issues are common.

In clinical practice, pre- and post-contrast MRI obtained under anesthesia is typically used to diagnose SWS and follow brain abnormalities in children with Sturge-Weber syndrome (SWS). However, anesthesia and contrast administration carry risks. Recent MRI acquisitions can include **susceptibility-weighted imaging (SWI)**, that can visualize normal and abnormal cerebral veins and calcifications, even without contrast administration.

OBJECTIVES

In this prospective imaging research study, we explored the feasibility and diagnostic utility of a safe non-contrast, non-sedate MRI acquisition with SWI in children with SWS and their healthy siblings with a wide range of cognitive and behavioral functioning.

STUDY SUBJECTS

Thirty children were recruited prospectively for the study, including:

- **15 with unilateral SWS** (8 F, 7 M; age range: 2.6-19 years, mean age: 11.8 years); all SWS children had a previous contrast-enhanced clinical brain MRI to verify SWS brain involvement.
- **15 healthy siblings** (11 M, 4 F; age range: 0.7-18 years, mean age: 11.1 years).

A total of 5 children (3 with SWS, 2 siblings) were below 6 years of age.

METHODS

Study procedures included:

1. **Non-sedate (15-minute) native brain MRI** on a 3T scanner, with the acquisition of: SWI, axial T1, T2, and fluid-attenuated inversion recovery (FLAIR) sequences;
2. **Formal neurocognitive and behavioral assessment** using age-appropriate neuropsychology batteries.

Young children were given extra time in the MRI scanner and/or scanned in natural sleep.

All MRIs were evaluated by two investigators and abnormalities were compared to those identified on previous clinical MRI.

RESULTS

- Twenty-nine children (96%) completed the MRI with good (n=28) or adequate (N=1) quality, including all SWS children, and all seven children <8 years of age.

- The SWS group had lower cognitive functions than the siblings (full-scale IQ: 85±14 vs. 108±14, p<0.001).

- 11/30 children (37%; 5 SWS, 6 siblings) had clinically significant behavioral issues without an effect on the MRI success rate and quality.

- SWI detected normal veins, enlarged deep veins and calcifications (**Figures 1** and **2**). Together with the other MRI sequences, other SWS vascular and parenchymal abnormalities, including leptomeningeal venous malformations, enlarged choroid plexus, and atrophy were also detected comparably to previous clinical MRI.

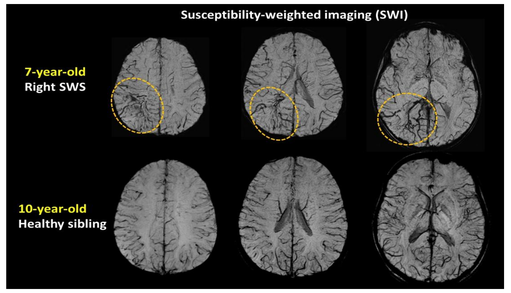

Figure 1. SWI MIP images showing enlarged (circles) and normal deep medullary veins. [MIP: minimal intensity projection]

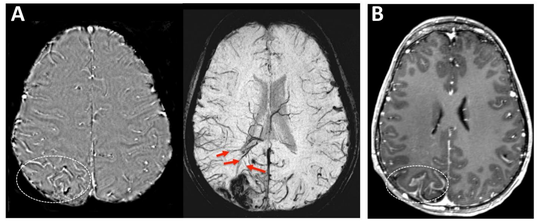

Figure 2. (**A**) Right posterior leptomeningeal venous malformation (LVM) on SWI phase image (left panel) and enlarged deep medullary veins (on SWI MIP image, right panel), as well as calcifications, in a 13-year-old child with SWS. Veins, including the LVM (area within the dotted circle), are seen with high signal intensity on the SWI phase images. The SWI MIP images (right panel in (**A**)) visualized multiple enlarged deep medullary veins (arrows) connecting the affected cortex with the periventricular veins. (**B**) Previous clinical post-contrast T1-weighted MRI at age 9.2 years, obtained with sedation, showed abnormal leptomeningeal enhancement in the same region.

CONCLUSIONS

- Non-sedate, non-contrast MRI can be acquired with a high success rate (>90%) even in young children, including those with cognitive impairment and behavioral issues.
- Brain MRI with native SWI can provide high diagnostic yield in children with SWS, as it can detect a range of vascular and parenchymal abnormalities without contrast enhancement.
- While contrast-enhanced MRI remains useful for the initial diagnosis of SWS, follow-up studies may utilize non-contrast MRI acquisition without sedation.