Entire Hemispheric Proton In Vivo MRS Examination of Sturge-Weber Syndrome at 3 T

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Introduction
Because N-acetylaspartate (NAA) is found predominantly in neurons, a decrease in NAA can be used as a marker for neuronal damage [1] e.g. in Sturge-Weber syndrome (SWS). Leptomeningeal angiomatosis is the essential pathologic feature in SWS. Cortical calcification is usually found where the cortex is degenerated underneath the vascular malformation. The presence of defects in perfusion and glucose metabolism has been reported [2], as well as the correlation between NAA and glucose metabolism (rCGM) for SWS for the region of apparent involvement and its contralateral hemisphere [3].

In this study, not only contra-lateral comparison in the affected region, such as parieto-occipital lobe, but also in the region where the neuronal damage may not be evident, such as frontal lobe, was examined for SWS patients. The purpose was the evaluation of the relevance of the correlation regarding metabolites for the whole cerebrum. Healthy volunteers were also examined with the same technique in order to evaluate inter-subject variability.

Methods
In vivo proton MR spectroscopy was done over entire hemisphere for four SWS patients and six healthy volunteers at 3T GE SIGNA system. One of the patients had bihemisphric disease, and the other three patients had unilateral involvement based on the presence of the angiomatosis.

Four contralateral voxels were selected from the frontal lobe, and the parieto-occipital lobe. The water suppressed proton spectra (TE=45ms or 136ms, TR=2s) were obtained from the 2x2x1.5 cm³ voxels using the PROBE-SV (PRESS). Additional scans were performed in thalamic region in case that decreased perfusion (HMPAO) and glucose metabolism (FDG) were found by the SPECT examination (Fig. 1). For each spectrum, 128 acquisitions were accumulated for 5 minutes. The NAA concentrations were evaluated using internal water reference from the same voxel [4]. T1 and T2 relaxation corrections were performed for quantitation of the regional NAA level.

Results
HEALTHY VOLUNTEERS: The NAA level of normal volunteers (32 ± 5 y.o.; 2 female; 4 male) are 12.36 ± 0.71 and 11.4 ± 1.36 (mmol/Kg wet wt) for white matter in parieto-occipital lobe and frontal lobe, respectively. The contralateral differences in NAA level among healthy groups are 4.7 ± 2.1% in the parieto-occipital region, and 7.5 ± 5.5% in the frontal region.

SWS PATIENTS: The NAA level in affected region of SWS patients is 6.32 ± 3.25 (mmol/Kg wet wt), whereas it is 9.49 ± 3.23 (mmol/Kg wet wt) in unaffected region.

(A) NAA reduction in unilateral cases: In three unilateral cases (1 female, 2 male; 2, 15, 17 y.o.), decreased NAA level by 21.0 ± 10.8 % in the affected hemisphere were observed in the parieto-occipital lobe in all (Fig. 1). Reductions of the NAA concentration were observed in the affected hemisphere independent of the severity of the atrophy and calcification. This is consistent with hemispheric cerebral dysfunction. For frontal lobe, however, a significant drops was observed among two patients (40 ± 27%). A reduction in NAA was also found in the thalamus in two cases (by 72% and 27%), a region that showed no calcification or atrophy, but had decreased perfusion (HMPAO) and glucose metabolism (FDG).

(B) NAA reduction in bilateral case: In the single case of bihemispheric disease (6 y.o. female), reduction of NAA in the occipital region was found in both hemispheres where calcifications were present (48.5% reduction, compared with frontal lobe). Unlike the unilateral SWS cases, the reduction in NAA was not significantly different between left and right hemispheres.

Discussion
Compared with the normal control group, contralateral reduction in NAA concentration in the affected parieto-occipital region of SWS patients is significant (21% vs. 4.7%, p < 0.02). The reduction in NAA in the frontal lobe is larger than normal variation, but is not significant when all cases are included (27.7% vs. 7.5%, p=0.18). This reduction in NAA in the frontal lobe also shows statistical significance for the cases where the reduction is larger than the variation observed in normal controls (40% vs. 7.5%, p<0.05) (Fig. 2).

Regional changes in NAA that correspond to morphological change observed in both unilateral and bilateral disease patients suggest that the NAA level may have the potential to be a neuronal marker for SWS. The changes in NAA levels in regions where no morphological changes were seen are less clear. Not only contralateral but ipsilateral variations were also observed using entire hemispheric examination. From the data of healthy volunteers, it is apparent that the variation between the two hemispheres is small. These data can be used to determine inter-patient and repeat measurement variability.

References

Acknowledgement
Partial financial support was provided by the University of Florida Brain Institute.