Serial Endovascular Embolization as a Novel Treatment for Refractory Epilepsy in Hemispheric Overgrowth: A Case Series

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INTRODUCTION

In children with congenital hemispheric overgrowth, hemispherotomy is a standard treatment approach for severe, medically refractory epilepsy. However, performing this surgery in early infancy presents significant risk. Prior centers[1,2,3] have reported a few cases of endovascular trans-arterial embolization as part of management in these patients. As reports of this method in the literature are limited, we describe clinical and surgical details of two infants from our center with hemispheric overgrowth and intractable epilepsy who underwent serial endovascular trans-arterial embolization as part of comprehensive epilepsy management. With this approach, invasive surgery was delayed to allow somatic growth and reduction of surgical morbidity.

OBJECTIVES

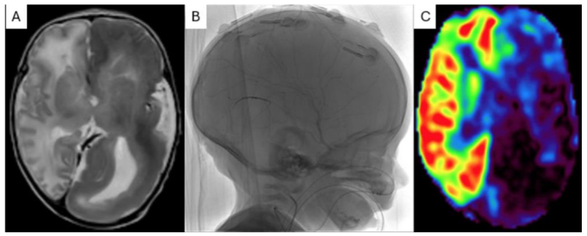
- Describe surgical and medical management approach
- Provide peri-surgical imaging and EEG considerations

METHODS

Pre- & intra-operative EEG monitoring was performed. Postoperative evaluations included MRI & EEG in order to correlate ischemic and electrographic evolution.

METHODS, CONT'D

	Patient A	Patient B
DOB	1/31/2023	3/4/2023
Pathology	Left hemispheric hemimegalencephaly	Large right parieto-occipital cortical malformation
Genetics	AKT3 pathogenic variant (+skin biopsy; serum without variant due to mosaicism)	NPRL3 pathogenic variant
Arterial Embolization Sites	• 2 mo: L distal PCA • 3 mo: L temporo-occipital, superior occipital, post. temporal, inferior frontal basal lateral • 5 mo: L superior occipital, angular, inferior posterior parietal, superior posterior parietal; L posterior, middle, anterior, and superior prefrontal; L post-central, anterior parietal, medial occipital, basal occipital • 9 mo: Distal L posterior, posterior insular, middle insular, inferior frontal, anterior insular • 11 mo: L frontal middle meningeal; L prefrontal, orbito-frontal, L distal pericallosal parieto-occipital; L parieto-occipital middle meningeal (for pre-hemispherotomy bleeding control)	• 5 mo: R superior parietal, R anterior parietal, and R parietal-occipital • 6 mo: R parieto-occipital and R splenial
Complications	Bronchospasm during first embolization; embolizations delayed due to viral illness	Embolizations delayed due to viral illness
Hemispherotomy	Yes, performed at 12 months of age	Yes, performed at 11 months of age
Medical treatment	ASM polypharmacy (including sirolimus & VGB) + ketogenic diet	ASM polypharmacy (including sirolimus & VGB)



Images shown for patient A:
(A) Pre-operative axial MRI showing left hemispheric hemimegalencephaly
(B) Intra-operative unsubtracted lateral view demonstrating first embolization of left distal PCA via Onyx™
(C) Perfusion imaging demonstrating cumulative result of serial embolizations (prior to hemispherotomy)

RESULTS

Patient A: Stabilization of clinical seizures following embolization reduced requirement for IV seizure rescue medications and allowed for NICU discharge. The only unplanned hospital admissions for seizures occurred in the context of embolizations delayed due to illness. Due to continued refractory seizures, hemispherotomy performed, with no clinical seizures reported afterwards.

Patient B: Both embolizations had a temporizing effect on seizures that allowed for stabilization of anti-seizure medications. Due to continued refractory seizures (as well as anterior migration of patient's epileptogenic network over time), hemispherotomy performed, with no clinical seizures reported afterwards.

Both: Sirolimus trialed due to affected genes' involvement in the mTOR pathway, but consistent use of this medication was difficult due to frequent infections and procedures (Patient A) as well as frequent infections and elevated lipid levels (Patient B).

CONCLUSIONS

- In congenital hemispheric overgrowth, endovascular embolization shows promise for palliative reduction of seizure burden and reduced hospitalization as a bridge to more traditional surgical intervention.
- These adjunctive embolizations have been safely performed in a multi-disciplinary setting without significant reported complications thus far.
- Rarity of these conditions would require national or international networks to accumulate sufficient patient numbers to make statistically significant conclusions.
- Sirolimus is mechanistically promising for use in these infants, though logistical barriers present challenges for consistent administration.

REFERENCES

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