Treatment of Pediatric Intracranial Vascular Malformations using Onyx-18

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Background

- AVMs cause 30-50% of all ICHs in children
- Estimated risk of hemorrhage is 2-4% per year
- Estimated M&M for each hemorrhagic event in children is 50% and 5-10% respectively
- Consensus on aggressive treatment of pediatric AVMs

Onyx Liquid Embolic System

• Approved in 2005 by the US FDA for the pre-surgical embolization of brain AVMs
• Cohesive polymer of ethylene vinyl alcohol (EVOH) and dimethyl sulfoxide (DMSO)
• Mixed with tantalum particles for visualization
Role of Endovascular Treatment for AVMs

- **Primary treatment** (goal of complete embolization)
- **Adjuvant treatment**
  - Pre-operative embolization
    - Reduce blood loss
    - Eliminate deep feeders with difficult surgical access
  - Pre-radiosurgery embolization
    - Goal to reduce nidus volume
    - Improved obliteration rates with smaller volumes
    - Reduce radiation exposure and radiation induced edema

Pittsburgh Experience with Endovascular Treatment of Pediatric Vascular Malformations

• 6 children
  – 3 female
  – 3 male
• Age range
  – 1 day to 12 years
• 4 AVM
• 2 VOGM
<table>
<thead>
<tr>
<th>Case No.</th>
<th>Age, Sex</th>
<th>Location</th>
<th>AVM Size (cm)</th>
<th>AVM Drainage</th>
<th>Clinical Presentation</th>
<th>Treatment</th>
<th>Angiographic Outcome (% residual)</th>
<th>Clinical/Radiographic FU (mos)</th>
<th>mRS/FIM Score</th>
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<tbody>
<tr>
<td>1</td>
<td>10 yrs, M</td>
<td>rt cerebellar</td>
<td>2.5</td>
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<td>papilledema</td>
<td>1 endovascular embolization, 1 GKS</td>
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<td>6</td>
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<td>1 endovascular embolization</td>
<td>NA</td>
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* FU = follow up; NA = not applicable.
Methods

• Intentional staging
  – Limit renal toxicity
    • Attempt to keep dye load < 5 mL/kg
    • Attempt to keep DMSO dose below 600 ug/kg
  – Allow brain adaptation to altered hemodynamics
• General anesthesia
  – Neurophysiological monitoring
    • Upper and lower ext. SSEPs, EEG, BSAERs
• Modified WADA prior to embolization
  – 1 mL lidocaine or 3 mL methohexital sodium
DMSO dose administered in relation to patient's weight and volume of Onyx

<table>
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<tr>
<th>Volume of Onyx (ml)</th>
<th>Patient Weight (lb/kg)</th>
<th>DMSO Dose (μg/kg)</th>
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<tr>
<td>10.0</td>
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</table>
Methods

• We typically use Onyx-18
  – Less viscous than Onyx-34
  – May allow deeper penetration
• Remember DMSO is a universal solvent
  – Must use DMSO compatible catheters
    • ev3 – Marathon (1.3 F)  Echelon (1.7 or 1.9 F)
    • BALT – SONIC (1.2 F)
Caveats for very young patients

• Consider asking general surgery colleagues to obtain peripheral arterial access via cut-down
  – Allows direct repair of vessel after removal of sheath
• Consider leaving in sheath between stages to avoid repeat arteriotomy
• Warn families of DMSO aroma post procedure
Case 1

- 10 y.o. boy noted to have papilledema on routine eye exam
- CT, MR, and DSA show
  - 2.5 cm right frontal AVM
  - 2.5 cm left frontal AVM
  - 2.5 cm right cerebellar AVM
Right Frontal AVM

• 2 endovascular embolizations
  – Pericallosal and aberrant proximal ACA feeders embolized in 2 procedures over 48 hours
  – 60% nidus reduction
• 1 Gamma Knife Radiosurgery (GKRS) treatment
Left Frontal AVM

• 1 endovascular embolization
  – 3 distal left MCA feeders, 2 distal left ACA feeders
  – 60% nidus reduction
• 2 staged GKRS treatments
Right Cerebellar

- 1 endovascular embolization
  - Right SCA and PICA feeders
  - 80% nidus reduction
- 1 GKRS treatment
Case 2

- 20 day old female presented with high output heart failure
- Cranial Ultrasound, MR, and DSA show
  - VOGM
  - Fed by bilateral PCAs and AChA
  - Drainage into falcine and straight sinus
Treatment

• 5 endovascular procedures slowed flow enough to eliminate heart failure and improve cardiac hemodynamics, allowing patient to return home
  – Stage I  – tranvenous coil embolization of fistula
  – Stage II  – transarterial coil embolization of left PCA feeders
  – Stage III  – transarterial coil sacrifice of distal left PCA
  – Stage IV  – transvenous coil embolization of venous varix fed by right PCa
  – Stage V  – transarterial coil embolization right AChA
One year later

• Patient returned with shortness of breath
• DSA shows residual flow through fistula
• Underwent Stage VI transvenous Onyx embolization of residual fistula resulting in eradication of flow through the VOGM
Case 3

• 12 y.o. boy presented with sudden LOC and IVH
• CT, MR, and DSA show
  – 3.5 cm right parieto-occipital AVM
Right Parieto-occipital AVM

• 1 endovascular embolization
  – Onyx embolization of the medial posterior choroidal artery
• No residual on 6 month f/u DSA
Conclusions

- Pediatric vascular malformations often require multimodality treatment
- Onyx embolization is one therapy that can provide safe and durable treatment
- Should be considered an adjuvant treatment; complete embolization is rare
- Toxicity and recanalization remain primary concerns
Thanks to ev3 for providing an unrestricted educational grant.

Acknowledgements