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THE RITZ-CARLTON REYNOLDS, LAKE OCONEE
GREENSBORO, GA
Multidisciplinary Approach to the Management of Complex Vascular Malformations

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Duke University School of Medicine, Durham, NC
Disclosure(s):

• None
Vascular Malformations?

- Complex group of vascular lesions
- Arise by embryologic dysmorphogenesis *without* increased endothelial proliferation
Vascular Malformations?

• Lead to structural and functional anomalies of the vascular system
• Characterized by wide range of presenting symptoms and often unpredictable clinical course
Incidence: 1.2%-1.5% in general population

2/3 Low Flow

1/3 High Flow
The most recent genetic analysis data: CVMs are caused by Deregulation of VEGFA/VEGFR2/AKT signaling system controlled by cell signaling and flow-mediated shear forces.
Multidisciplinary Approach Rationale:

- The management of CVMs exceeds the level of expertise of any single medical specialty
- Dedicated navigator required to coordinate care
- Care requires advanced resources in many disciplines
Multidisciplinary Approach: Patient centered and lesion driven
Treatment Modalities
(every patient is assigned a primary treating physician and therapeutic goals are formulated before the treatment)

r/o Vascular tumors
(Hemangiomas most common)

dceMRI +/- Arteriogram
(diagnostic and treatment planning)

Duke Multi-D Protocol:

CVMs

HFVMs
- Conservative
- Embolization
- Embolization + Sclerotherapy
- Resection

LFVMs
- Conservative
- Sclerotherapy
- Resection
- Multimodal
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- LFVMs

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Duke Multi-D Protocol:
CVMs vs. Hemangiomas:

- Initial step in the diagnostic algorithm
- Distinction between vascular neoplasms (most frequently infantile hemangiomas) and CVMs can be made based on history and clinical assessment in most cases
Duke Multi-D Protocol:

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High Flow Vascular Malformations (HFVMs)

- “arteriovenous malformations”
- Direct connection between arterial and venous system caused by congenital malformations resulting in fistulous A-V tracts
• HFVM - Clinical findings:
  • Can be aggressive; cutaneous ischemia with ulceration, infection or hemorrhage
  • Can be painful
  • High output cardiac failure when extensive (rare)
18 year-old male patient congestive heart failure secondary to HFVM; CT demonstrated cardiomegaly and multiple coils in the AVM, percutaneously inserted during multiple coil embolization sessions.
US: Arterio-venous malformations demonstrate arterialized venous waveform and spectral broadening
HFVM - Diagnosis with Conventional MRI:
HFVM Diagnosis with Catheter Angiography:
LFVM Clinical Presentation:

- Affects both superficial and deep underlying anatomic structures (skin, muscles, abdominal viscera, CNS)
- Rarely asymptomatic
- May be isolated or part of a syndrome
Venous malformations:

- No increase in local skin temperature
- No thrill or bruit

Venous malformations demonstrate mixed (or monophasic) waveform on US
Coronal T2-weighted MRI demonstrates hyperintense signal in the venous malformation involving the anterolateral aspect of the right upper extremity.

LFVM - Diagnosis with Conventional MRI:

Axial T2-weighted MRI of the same patient

Time-Resolved MRA Acquisition

- Inflow phase
- Arterial phase
- Venous phase

**Gd**

2-5 sec

**ROBUST HEMODYNAMIC DATA and relationship to vital structures**

(MIP)

2-5 min

Courtesy of Charles Kim, MD
Arterial phase image demonstrates no vascular abnormality = LFVM.

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Duke Multi-D Protocol:
Treatment Options:

• Conservative management: Observation, Compression therapy

• Sclerotherapy:
  – Foam: Polidocanol (Aethoxysklerol,® Asclera,®) or Sodium Tetradecyl Sulfate (Sotradecol®)
  – Ethanol

• Embolization
2 yo F w/ KTS
Presented s/p EtOH Tx at OSH (complicated by cellulitis Tx w/ i.v. Abx)
dceMRI: diagnosis and treatment planning
4 w s/p ligation and sclerotherapy
Surgical Resection: Lymphatic LFVM

Surgical Resection: Lymphatic LFVM

HFVMs Principals of treatment:

- Often multimodal
- Lesion structure dictates therapy
- Aim for obliteration of nidus to avoid loss of access and recruitment
Treatment of HFVM:

- Coil embolization
Treatment of HFVM:

• Glue, ETOH chemoablation

Treatment of HFVM:

• Glue embolization

No matter what you use, treat the nidus and outflow, not the inflow!
Embolization and surgical resection

Courtesy of William Eward, M.D., Department of Orthopedic Surgery, Duke University Medical Center
Treatment Efficacy
Treatment Outcomes:

• 135 patients with 136 lesions

• Patient and lesion demographics comparable to prior studies (LFVM:HFVM = 3.5:1, M=F)
<table>
<thead>
<tr>
<th>Type N (%)</th>
<th>Treatment</th>
<th>Treated N (%)</th>
<th>Symptoms Improvement</th>
<th>Complications N (%)</th>
<th>Specific Complications</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td></td>
<td>Worse</td>
<td>No Change</td>
<td>Significantly Improved</td>
</tr>
<tr>
<td>LFVM 105</td>
<td>Conservative management</td>
<td>23 (21.9%)</td>
<td>2</td>
<td>18</td>
<td>2</td>
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<tr>
<td></td>
<td>Foam Sclerotherapy</td>
<td>31 (29.5%)</td>
<td>0</td>
<td>1</td>
<td>8</td>
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<tr>
<td></td>
<td>Ethanol Sclerotherapy</td>
<td>7 (6.7%)</td>
<td>0</td>
<td>2</td>
<td>3</td>
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<tr>
<td></td>
<td>Resection</td>
<td>18 (17.1%)</td>
<td>0</td>
<td>2</td>
<td>3</td>
</tr>
<tr>
<td></td>
<td>Combination therapy</td>
<td>8 (7.6%)</td>
<td>0</td>
<td>0</td>
<td>7</td>
</tr>
<tr>
<td>HFVM 31</td>
<td>Conservative management</td>
<td>8 (25.8%)</td>
<td>1</td>
<td>0</td>
<td>1</td>
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<td></td>
<td>Embolization</td>
<td>8 (25.8%)</td>
<td>0</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td></td>
<td>Embolization + Sclerotherapy</td>
<td>6 (19.4%)</td>
<td>0</td>
<td>0</td>
<td>5</td>
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<tr>
<td></td>
<td>Resection</td>
<td>5 (16.1%)</td>
<td>0</td>
<td>1</td>
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</tbody>
</table>

*Tabular data do not include 18 pts lost to follow up (4 HFVM, 14 LFVM); 2 pts treated with laser, and 1 pt who died prior to initiation of treatment.
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<tr>
<td>LFVM 105</td>
<td>Conservative management</td>
<td>23 (21.9%)</td>
<td>Worse: 2 (8.7%)</td>
<td>0</td>
<td>0 (0%)</td>
</tr>
<tr>
<td></td>
<td>Foam Sclerotherapy</td>
<td>31 (29.5%)</td>
<td>No Change: 18</td>
<td>0</td>
<td>4 (57.1%)</td>
</tr>
<tr>
<td></td>
<td>Ethanol Sclerotherapy</td>
<td>7 (6.7%)</td>
<td>Significantly Improved: 2</td>
<td>0</td>
<td>2 (11.1%)</td>
</tr>
<tr>
<td></td>
<td>Resection</td>
<td>18 (17.1%)</td>
<td>Completely Resolved: 1</td>
<td>0</td>
<td>0 (0%)</td>
</tr>
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<td>8 (7.6%)</td>
<td></td>
<td></td>
<td>0 (0%)</td>
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<td></td>
<td></td>
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<td>Symptoms improvement</td>
<td></td>
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<tr>
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<td>Sclerotherapy: 84.2%</td>
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<td>Resection: 88.9%</td>
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<td></td>
<td></td>
<td></td>
<td>Combination: 100%</td>
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<td>HFVM 31</td>
<td>Conservative management</td>
<td>8 (25.8%)</td>
<td>Worse: 0 (0%)</td>
<td>0</td>
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<tr>
<td></td>
<td>Embolization</td>
<td>8 (25.8%)</td>
<td>No Change: 1</td>
<td>0</td>
<td>1 (12.5%)</td>
</tr>
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<td></td>
<td>Embolization + Sclerotherapy</td>
<td>6 (19.4%)</td>
<td>Significantly Improved: 5</td>
<td>0</td>
<td>1 (16.7%)</td>
</tr>
<tr>
<td></td>
<td>Resection</td>
<td>5 (16.1%)</td>
<td>Completely Resolved: 3</td>
<td>0</td>
<td>0 (0%)</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Symptoms improvement</td>
<td></td>
<td></td>
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<td></td>
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<td>Embolization: 87.5%</td>
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<tr>
<td></td>
<td></td>
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<td>Resection: 80.0%</td>
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<td>Ethanol Sclerotherapy</td>
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<td>Significantly Improved: 2 (6.7%)</td>
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<td>0 (0%)</td>
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<td></td>
<td>Resection</td>
<td>18 (17.1%)</td>
<td>Completely Resolved: 1 (4.3%)</td>
<td>8 (25.8%)</td>
<td>4 (57.1%)</td>
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<td>Symptoms improvement Sclerotherapy= 84.2% Resection = 88.9% Combination= 100%</td>
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<td>6 (19.4%)</td>
<td>Significantly Improved: 3 (50.0%)</td>
<td>5 (83.3%)</td>
<td>0 (0%)</td>
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<td></td>
<td>Resection</td>
<td>5 (16.1%)</td>
<td>Completely Resolved: 3 (60.0%)</td>
<td>1 (20%)</td>
<td>1 (12.5%)</td>
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Summary:

• Recent advancements in the diagnostic and treatment modalities have resulted in a better understanding of the pathophysiology and natural history of CVMs and improved management strategies for these lesions
Summary:

- Multidisciplinary approach affords the opportunity to streamline the evaluation process for CVM patients and facilitate individualization and integration of treatment strategies
Summary:

- Diagnostic algorithm utilized to distinguish vascular tumors from CVMs and HFVM from LFVM has been validated as clinically applicable for making an accurate anatomical and hemodynamic diagnosis of CVMs
- Serve as a basis for proper treatment selection, and significantly facilitate communication among different medical specialists
Summary:

• Data shows that implementation of the described therapeutic protocols results in favorable outcomes with acceptable complication rates in this frequently challenging patient population
Summary:

- Duke Team approach and management protocols have been fully incorporated in the 2009 and 2013 International Guidelines for the Management of Congenital Vascular Malformations
Summary:

• The evaluation of novel, recently developed medications that may prove even safer and more effective than current treatment modalities
• Moving forward, we must create a fertile environment for the development of innovative therapeutic interventions based on our understanding of the genetic and molecular and physiologic mechanisms responsible for the development and progression of CVMs
Duke Vascular Malformation Team Roster 2019/2020