Pediatric Vascular Surgery

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No Disclosures
Case – ALI (Iatrogenic Trauma)

- Two-month old boy - Trisomy 21, unbalanced AVSD underwent PA band placement

- Left femoral arterial line – lost left pedal pulses post-op
Arterial Duplex
6-weeks anticoagulation
• Arterial cannulation is a leading cause of acute arterial thrombosis and ischemia

• Femoral artery catheterization may be complicated by a 2% injury

• Long term risks
  – Claudication
  – LLD (33%)
  – Scoliosis (>2cm LLD)
  – Degenerative arthritis

Flanigan et al – Ann Surg - 1983
Lin et al – JVS - 2001
- **Treatment – Anticoagulation**
- **Revasc when limb threatening (<10%)**

**Table I.** Patient demographics and risk factors for iatrogenic femoral artery injury

<table>
<thead>
<tr>
<th>Demographic/risk factor</th>
<th>Acute limb ischemia</th>
<th>Chronic limb ischemia</th>
</tr>
</thead>
<tbody>
<tr>
<td>Total number of patients</td>
<td>74 (91%)</td>
<td>7 (9%)</td>
</tr>
<tr>
<td>Gender</td>
<td>39 female, 35 male</td>
<td>4 female, 3 male</td>
</tr>
<tr>
<td>Age at presentation</td>
<td>1.4 year (1 day–17 years)</td>
<td>3.5 year (15 months–6 years)</td>
</tr>
<tr>
<td>Weight at presentation</td>
<td>8.7 kg (1.0–77.6 kg)</td>
<td>14.8 kg (6.8–19.1 kg)</td>
</tr>
<tr>
<td>Vasopressor use</td>
<td>31 (41.9%)</td>
<td>Unknown</td>
</tr>
<tr>
<td>Venous thrombus</td>
<td>20 (27.0%)</td>
<td>2 (28.6%)</td>
</tr>
</tbody>
</table>

24% neonates

*Andraska et al; Annals of Vasc Surg; 2017*
Risk Factors for Operative Interventions

Table II. Patient risk factors for operative intervention in patients with ALI

<table>
<thead>
<tr>
<th>Risk factors for operative intervention in patients with ALI</th>
<th>P value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Weight</td>
<td>0.077</td>
</tr>
<tr>
<td>Age</td>
<td>0.008*</td>
</tr>
<tr>
<td>Concurrent DVT</td>
<td>0.938</td>
</tr>
<tr>
<td>Vascular consult</td>
<td>0.085</td>
</tr>
<tr>
<td>Proximal extent of injury</td>
<td>0.353</td>
</tr>
<tr>
<td>EIA</td>
<td>0.665</td>
</tr>
<tr>
<td>CFA</td>
<td>0.210</td>
</tr>
<tr>
<td>SFA</td>
<td>0.225</td>
</tr>
<tr>
<td>Distal</td>
<td>0.269</td>
</tr>
<tr>
<td>Mechanism of injury</td>
<td>0.002*</td>
</tr>
<tr>
<td>A-line</td>
<td>0.778</td>
</tr>
<tr>
<td>Cardiac catheterization</td>
<td>&lt;0.001*</td>
</tr>
<tr>
<td>ECMO</td>
<td>&lt;0.001*</td>
</tr>
<tr>
<td>Intraoperative injury</td>
<td></td>
</tr>
<tr>
<td>Multiple</td>
<td>0.214</td>
</tr>
</tbody>
</table>

Fig. 2. Mechanism of injury resulting in acute limb ischemia requiring operation.

Andraska et al; Annals of Vasc Surg; 2017
Medical Management - Outcomes

- Mean f/u 2.5mo
- Complication (7.5%)
  - Compartment syndrome / fasciotomy (1)
  - Contralateral femoral thrombosis (1)
  - Chronic occlusion / LLD (1)
- Mortality = 12.2% (9)
  - 1 death related to ALI (hemorrhagic stroke from anticoag)
- Surveillance inconsistent
- Only 65% - inpatient c/s (half returned for clinic follow-up)
- 58% - ‘‘normal’’ surveillance duplex study confirming arterial patency
- 42% - no repeat study
Mean f/u 4.5mo; 88% limb salvage

Table III. Surgical details and outcomes for pediatric patients presenting with acute limb ischemia following iatrogenic femoral artery injury

<table>
<thead>
<tr>
<th>Age at operation (years)</th>
<th>Weight (kg)</th>
<th>Mechanism of injury</th>
<th>Time between limb ischemia and operative intervention</th>
<th>Surgery</th>
<th>Operative findings</th>
<th>Postoperative outcome</th>
<th>Length of vascular follow-up (months)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>6</td>
<td>ECMO</td>
<td>&lt;1 day</td>
<td>Thrombectomy, patch angioplasty, and fasciotomy</td>
<td>Acute iliofemoral thrombosis and surrounding hematoma</td>
<td>Chronic EIV DVT</td>
<td>1</td>
</tr>
<tr>
<td>2</td>
<td>12</td>
<td>Surgical trauma</td>
<td>&lt;1 day</td>
<td>Interposition bypass graft with PTFE</td>
<td>Acute thrombosed dissection of left common femoral artery</td>
<td>Graft stenosis requiring balloon angioplasty at 7.5 months postoperative Amputation revisions x2</td>
<td>9</td>
</tr>
<tr>
<td>3</td>
<td>4</td>
<td>Arterial monitor line</td>
<td>2 days</td>
<td>Bilateral below the knee amputation</td>
<td>Concurrent arterial and venous thrombosis; unreconstructable given small size</td>
<td></td>
<td>6</td>
</tr>
<tr>
<td>4</td>
<td>17</td>
<td>ECMO</td>
<td>1 day</td>
<td>Thrombectomy, primary repair</td>
<td>Acute thrombosis of common femoral artery No known complications to date</td>
<td></td>
<td>1</td>
</tr>
<tr>
<td>5</td>
<td>13</td>
<td>ECMO</td>
<td>1 day</td>
<td>Thrombectomy; patch angioplasty, fasciotomy; ultimate amputation</td>
<td>Acute iliofemoral thrombosis Stump wound dehiscence requiring reoperation x2 hemATOMA</td>
<td></td>
<td>3</td>
</tr>
<tr>
<td>6</td>
<td>11</td>
<td>Surgical trauma</td>
<td>&lt;1 day</td>
<td>Bovine patch angioplasty</td>
<td>Acute disruption of the proximal common femoral artery at the junction to the external iliac artery. No known complications to date</td>
<td></td>
<td>16</td>
</tr>
<tr>
<td>7</td>
<td>17</td>
<td>ECMO</td>
<td>2 days</td>
<td>Patch angioplasty with hemashield</td>
<td>Vascular injury and surrounding hematoma. No known complications to date</td>
<td></td>
<td>0</td>
</tr>
<tr>
<td>8</td>
<td>7</td>
<td>Multiple mechanisms of injury</td>
<td>Unknown</td>
<td>Ligation of 2 AVFs and a pseudoaneurysm</td>
<td>Two AVFs, pseudoaneurysm of left common femoral artery. No known complications to date</td>
<td></td>
<td>0</td>
</tr>
</tbody>
</table>

Primary-Assisted Patency 100%; No Vascular-related Mortality

Andraska et al; Annals of Vasc Surg; 2017
Conclusions

• Immediate anticoagulation is mainstay of care after suspected arterial injury / occlusion in this clinical setting
  – 9% required operation for ALI
  – 16% required revascularization during follow-up
  – 97% limb salvage

• Minority of ALI patients had f/u

• Occult EIA occlusion may not be readily identified by infantile duplex

• Pilot data has driven a change in surveillance and standardized arterial duplex studies
Fig. 3. Treatment algorithm summarizing the University of Michigan’s Pediatric Vascular Surgery approach to acute pediatric limb ischemia. ABIs, arterial brachial indices; D/C, discontinue.
Case – CLI (Iatrogenic Trauma)

- 2 yo boy with truncus arteriosus, polydactyly, congestive heart failure, cleft palate

- At least 3 prior cardiac catheterizations (bilateral femoral access) with no documented episodes of ALI

- Referred for leg pain

- Clinical exam – palpable pulses; pedal asymmetry
Arterial Duplex

- Patent vessels – no thrombus
- RLE – triphasic; LLE – biphasic
- 1cm LLD
- At 1y f/u: progressive left leg pain, 1.7cm LLD
LLD – Scoliosis – Later
Degenerative Arthritis

Orthoradiograph or Scanogram
Revascularization

- Left iliofemoral bypass using reversed left great saphenous vein wrapped with Parietex mesh (CIA-femoral bifurcation)
  - Spatulate venous anastomosis
  - Interrupted suture line
  - PTFE in older adolescents
6mo follow-up
8mm LLD
• 33 consecutive children (1974-2016)
  – 26 pre-adolescent (mean 6.1y) / 7 adolescent (mean 13.9y)

• Mechanism: Iatrogenic (30) > trauma (2) > vasculitis (1)

• Preoperative S/Sx: Claudication/fatigue (25), LLD 1.32 ± 0.62mm (21) and scoliosis (5)

• Time to revascularization: 5.3y (pre-adolescent) / v. 11.2y (adolescent)
Outcomes

• Early complications: Early graft occlusion (1)

• **Primary patency = 69%**

• 1/3 required graft revision for: graft occlusion (6), graft stenosis (3), aneurysmal deterioration (2) and anastomotic pseudoaneurysm (1)

• **Secondary patency = 94%**

• Symptoms resolved in all but two patients (treatment failures)

• Inconsistent bone growth reversal (5%)
Case - Trauma

- 3 yo boy restrained, in a car seat. MVC, vehicle was 'T-boned'.
- Reported decline in mental status at the scene requiring intubation.
- OSH - left open humerus fracture with active arterial bleeding that was ligated
- Transferred - no radial or ulnar signal by Doppler interrogation
Wound Exploration

- Open and complex humerus fracture with extensive soft tissue loss
- Obvious clean ligation of the proximal and distal brachial artery with a long segment of missing vessel (vein and nerve)
- Estimated 5-6h ischemia
Surgical Repair

- Shunting of left brachial artery (6Fr Argyle)
- ORIF (Ortho)
- Left brachial-brachial bypass using reverse GSV [total shunt time = 2h 16 min]
  - Interrupted suture line (7-0 Prolene)
  - Nitroglycerin peri-adventitial
  - Did not reverse heparin
- Deferred venous reconstruction
- Forearm/hand fasciotomies (hand surgery) and complex wound closures
Case Presentation

- 8mo PICU transfer for HTN and FTT
- Unremarkable gestational (39w) and PMH

- Exam: 7.5kg and 66.5cm (19th%)
  - Upper Extremity HTN (185mmHg)
  - Lower Extremity hypotension (70s)
  - Weak femoral pulses
  - Systolic ejection murmur
Case Presentation

Phillips et al; Ann Vasc Surg; 2016(3)
Case Presentation

- Multiple anti-hypertensives

- Progressive abdominal bloating and pain with SBP <120mmHg

- Transient azotemia (Cr to 1.54)

- Progressive cardiopulmonary failure – intubated / vent support
Aorto-aortic bypass
Reimplantation of LRA

Phillips et al; Ann Vasc Surg; 2016(3)
Case Presentation

- Uneventful post-op course
- D/C POD #18
  - Amlodipine – BP 120/50; Cr 0.1
- One year follow-up
  - 12.6kg (86th %)
  - Otherwise stable
  - Reassuring MRA
8yo, single kidney, 5-drug HTN and AKI/IHD
6yo – 4-drug HTN + LVH

3.5x12 Empira Balloon left
LRA Reimplantation – D/C POD 9, normotensive
5yo with long-segment thoraco-abdominal aortic occlusion; 3-drug HTN + LVH

16mm PTFE

Tunnel anterior to main pulmonary artery along left lateral aspect of heart

Modest redundancy

Retroperitoneal, retro-renal tunnel

D/C POD 19 with single drug

2y f/u – LVH resolved; stable 1-drug HTN

Coleman et al. JVS 2012;56:482-5.
Renovascular HTN

Renin-angiotensin system interrelation with aldosterone and bradykinin in regulation of blood pressure.

- Renin substrate
  - Renin
  - Bradykinin
    - Kininase II
    - Bradykinin
- Angiotensin I
  - Kinase II
  - Angiotensin II
  - Angiotensin III
    - Cholesterol
    - Inactive peptides
    - Blood pressure
- Aldosterone
  - Pregnenolone
Pediatric HTN - #3

- **Normal BP:** < 90\(^{th}\) % for sex, age, height

- **Hypertension:** Average SBP or DBP ≥ 95\(^{th}\) % for sex, age, height on at least three separate occasions

- **BP Screening Recommendations:**
  - ≥ 3 years – any medical setting
  - < 3 years - congenital heart disease, renal diseases or urologic malformations, hospitalization
Pediatric Renovascular HTN: 3rd Most Common Cause

- Up to 40% of patients may have a syndromic (genetic) etiology - *NF1*, *ELN/Williams Syndrome, TSC1/2, JAG1, FBN1, Turner Syndrome and Alagille Syndrome*

Mid-abdominal Aortic Syndrome (MAS)

Classified by cephalad extent of narrowing

Supra-renal
Intra-renal
Infra-renal

Renal Involvement 87%
Splanchnic Involvement 62%
Intimal proliferation

Fragmentation of elastic lamina

Medial thinning

Excessive peri-adventitial elastin

Renal Artery Occlusive Disease

Coleman DM; JVS; 2021
Abdominal Aortic Stenosis

Extensive concentric intimal fibroplasia, isolated internal elastic lamina duplication and fragmentation of a segmental abdominal aortic coarctation in a 6-year-old (Movat x4).

Heider et al; MVSS Presentation; 2021 (unpublished)
Diagnostics

- Inflammatory work-up (ESR/CRP)
- Blood hormone levels
- Echocardiogram
- Renal Duplex
- Cross-sectional Imaging (MRA)
- Diagnostic arteriography and renal vein renin sampling
Renal Artery Duplex

Renal sonography with Doppler for detecting suspected pediatric renin-mediated hypertension – is it adequate?

Patricia K. Castelli · Jonathan R. Dillman · David B. Kershaw · Shokoufeh Khalatbari · James C. Stanley · Ethan A. Smith

- Renal Doppler sonography reliably detects renin-mediated HTN caused by aortic or main renal artery narrowing in children
- 90% Sensitive, 68% Specific
- Additional imaging required to detect intra-renal / accessory renal artery stenoses
14yo with mid-renal focal stenosis (80mmHg gradient) – web by angiogram
Indications for Revascularization

- Medically refractory HTN
- Progressive renal insufficiency
- NICM (concentric LVH)
- Failure to thrive
- Lower extremity sequelae (claudication, exertional fatigue, growth disturbance)

- Consider timing and challenges (patient size and projected growth)
19 hypertensive patients (ages 2-18)- underwent PTA
Neurofibromatosis N=7
Technical success 29 out of 32 lesions (91%)
39% cure, 17% improved, 44% clinical failures
PTA failures (OSH): Restenosis, thrombosis and rupture

Time to Failure < 2 years

Remedial Operations:
- Aortic reimplantation (13), Semgental reimplantation (1), Aorto-renal bypass (10), Arterioplasty (1), Iliorenal bypass (1), Nephrectomy (7)

Postoperative HTN cured (24%), improved (60%), unchanged (16%)
Remedial Operations

- Risk of nephrectomy following PTA and stenting was 31% (compared to 15% following PTA alone).
- Failures in EV therapy in patients aged <10 years resulted in a nephrectomy rate of 44%.
  - No nephrectomy following failures in patients ≥10y.
- PTA +/- stenting complicated remedial surgery in 56% of patients.
- No major morbidity, operative mortality or late deaths.
Indications for Endovascular Interventions

- Endovascular therapy as a “bridge” to surgical therapy
  - Allowed delay of surgical reconstruction > 1 year

- Small vessel size and fibrotic nature of stenoses limits endovascular utility in OSTIAL lesions

- Endovascular therapy for renal artery stenoses in pediatric patients should be undertaken with caution (high-volume center)

- No role for routine stenting in children!
Surgical management of pediatric renin-mediated hypertension secondary to renal artery occlusive disease and abdominal aortic coarctation

Dawn M. Coleman, MD, Jonathan L. Eliason, MD, Robert Beaulieu, MD, Tatum Jackson, BA, Santhi K. Ganesan, MD, Stanley, MD, on behalf of the University of Michigan, Mich

Table I. Demographics and preoperative risk factors

<table>
<thead>
<tr>
<th>Variable</th>
<th>Total cohort (N = 169)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Male sex</td>
<td>93 (55)</td>
</tr>
<tr>
<td>Age at time of surgical intervention at University of Michigan, years</td>
<td>8.96 ± 5.53</td>
</tr>
<tr>
<td>Weight at time of intervention at University of Michigan, kg</td>
<td>58.33 ± 16.8</td>
</tr>
<tr>
<td>No. of initial antihypertensive medications</td>
<td>2.70 ± 1.58</td>
</tr>
<tr>
<td>NFI</td>
<td>31 (18.34)</td>
</tr>
<tr>
<td>Abdominal aortic coarctation</td>
<td>76 (44.97)</td>
</tr>
<tr>
<td>Prior intervention performed elsewhere for RVH before intervention at University of Michigan</td>
<td>51 (30.18); open surgical (14), endovascular (35), combination (2)</td>
</tr>
</tbody>
</table>

NFI, Neurofibromatosis type 1; RVH, renin-mediated renovascular hypertension.
Categorical variables are presented as number (%). Continuous variables are presented as mean ± standard deviation.

Coleman et al; JVS; 2020

18% NF 1 (N31)  
WS (6)  
Moya Moya (5)  
TA (1)  
Alagille (1)
Renal Artery Occlusive Disease
Anatomic Location

Ostial > Segmental > Mid
66% 10% 8%

54% Bilateral (91), 18% Solitary Kidney (30)
Primary Surgical Interventions

342 Primary Surgical Interventions
248 directed at renal artery disease

- Partial Nephrectomy: 21
- Complete Nephrectomy: 4
- Segmental Reimplantation: 10
- Endarterectomy/Intimectomy: 2
- Segmental Embolization: 10
- Segmental Reimplantation (another instance): 4
- Patch arterioplasty: 8
- Bypass: 50
- Reimplantation: 136
Primary Surgical Procedures

Aortic Reimplantation
136

Aortorenal Bypass
50
Concurrent Aortic Procedures 64

Patch Aortoplasty 32

Aortic Bypass 32
Technical Pearls

- Single-stage operation
- In-vivo reconstruction
- Spatulate renal artery-aortic implantations
- Single interrupted sutures with fine monofilament suture (allows for growth)
  - Avoid vein grafts (late aneurysm)
    - Hypogastric Artery

Stanley et al; J Vasc Surg 2006(44)
**LOS** averaged 13.6 days

Perioperative Morbidity = 31%

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**Table V. Perioperative complications**

<table>
<thead>
<tr>
<th>System</th>
<th>Complication</th>
</tr>
</thead>
<tbody>
<tr>
<td>Reoperation</td>
<td>Renal graft thrombosis requiring nephrectomy (2) and revision (1), postoperative hemorrhage (4), fascial dehiscence (1), delayed abdominal closure (1)</td>
</tr>
<tr>
<td>Infectious (no cases of graft infection or wound infection)</td>
<td>Bacteremia (2), UTI (4), <em>Clostridium difficile</em> colitis (3), vaginal yeast infection (1), thrush (1), cellulitis associated with peripheral intravenous line (1)</td>
</tr>
<tr>
<td>Gastrointestinal</td>
<td>Chemical pancreatitis or prolonged (&gt;7 days) ileus (20), gastrointestinal bleed (2)</td>
</tr>
<tr>
<td>Respiratory</td>
<td>Pneumonia (7, of which 3 patients required prolonged intubation or reintubation), ARDS with prolonged intubation (1), viral URI (1), pneumothorax requiring tube thoracostomy (1), pleural effusion requiring tube thoracostomy (1)</td>
</tr>
<tr>
<td>Hematologic</td>
<td>Perinephric hematoma requiring transfusion (2), phlebitis (3), DVT (2), PE (1), digital thromboembolism (1), HIT (1)</td>
</tr>
<tr>
<td>Miscellaneous</td>
<td>Partial-thickness skin loss (1), decubitus ulcer (1), MCA stroke (1), chylous ascites requiring delayed thoracic duct ligation (1), intraoperative splenic laceration requiring splenectomy (1)</td>
</tr>
</tbody>
</table>

ARDS, Acute respiratory distress syndrome; DVT, deep venous thrombosis; HIT, heparin-induced thrombocytopenia; MCA, middle cerebral artery; PE, pulmonary embolism; URI, upper respiratory infection; UTI, urinary tract infection.

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- Reoperation (5%, N=9)
- Pancreatitis/Prolonged Ileus (12%)
- Infections (7%) / Pulmonary (7%)
- No RRT / No Mortality
Mean f/u 49mo

- 35 children (21%) required at least one reoperation directed at the renal arteries (36) or aorta (15):
  - Open (37) / EV (14)
  - Secondary interventions to preserve primary patency were required following index operation in 22 children (13%)
    - Median time to secondary intervention = 14mo (Range 2 to 159mo, SD ±38.43)

- Relative incidence of reoperation ↑↑ in patients with MAS (N=19, 24%) and NF1 (N=9, 29%)
Independent Predictors of Reoperation (logistic regression)

As age increased by one year, the rate of reoperation decreased by ~10% (HR=0.90; 95% CI: 0.83-0.97)

**Supplemental Table 1: Logistic regression model to predict the odds of reoperation**

<table>
<thead>
<tr>
<th>Independent Variables</th>
<th>Unadjusted Odds Ratio (95% CI)</th>
<th>p-value</th>
<th>Adjusted Odds Ratio (95% CI)</th>
<th>p-value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age at OR</td>
<td>0.88 (0.81 - 0.95)</td>
<td>0.002</td>
<td>0.86 (0.77 - 0.95)</td>
<td>0.003</td>
</tr>
<tr>
<td>Number of Initial Medications</td>
<td>1.09 (0.87 - 1.38)</td>
<td>0.440</td>
<td>0.90 (0.68 - 1.10)</td>
<td>0.450</td>
</tr>
<tr>
<td>Aortic reconstruction</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>None</td>
<td>Reference</td>
<td></td>
<td>reference</td>
<td></td>
</tr>
<tr>
<td>Patch</td>
<td>0.46 (0.15 - 1.43)</td>
<td>0.178</td>
<td>0.11 (0.02 - 0.54)</td>
<td>0.007</td>
</tr>
<tr>
<td>Bypass</td>
<td>0.74 (0.27 - 2.00)</td>
<td>0.550</td>
<td>0.27 (0.06 - 1.19)</td>
<td>0.084</td>
</tr>
<tr>
<td>NF1</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>No</td>
<td>Reference</td>
<td></td>
<td>reference</td>
<td></td>
</tr>
<tr>
<td>Yes</td>
<td>1.76 (0.73 - 4.27)</td>
<td>0.210</td>
<td>1.26 (0.43 - 3.68)</td>
<td>0.671</td>
</tr>
<tr>
<td>Abdominal Aortic Coarctation</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>No</td>
<td>Reference</td>
<td></td>
<td>reference</td>
<td></td>
</tr>
<tr>
<td>Yes</td>
<td>1.60 (0.76 - 3.39)</td>
<td>0.216</td>
<td>6.01 (1.70 - 21.24)</td>
<td>0.005</td>
</tr>
<tr>
<td>Had a prior intervention</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>No</td>
<td>Reference</td>
<td></td>
<td>reference</td>
<td></td>
</tr>
<tr>
<td>Yes</td>
<td>1.27 (0.58 - 2.80)</td>
<td>0.553</td>
<td>1.61 (0.60 - 4.31)</td>
<td>0.309</td>
</tr>
</tbody>
</table>
Hypertension Benefits

44% Cured, 46% Improved, 10% No change

Mean Number of Anti-hypertensives Post-op = $0.99 \pm 1.16$

(v. $2.7 \pm 0.58$ preop)

Mean Post-op Cr = $0.51 \pm 0.30$

No Dialysis
Table VII. Postoperative hypertension benefits

<table>
<thead>
<tr>
<th></th>
<th>Cure</th>
<th>Improved</th>
<th>Failure (unchanged)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Total cohort (N = 169)&lt;sup&gt;a&lt;/sup&gt;</td>
<td>74 (44)</td>
<td>78 (46)</td>
<td>17 (10)</td>
</tr>
<tr>
<td>Among patients with concurrent abdominal aortic coarctation (n = 76)</td>
<td>28 (37)</td>
<td>37 (49)</td>
<td>11 (14)</td>
</tr>
<tr>
<td>Among patients without abdominal aortic coarctation (n = 93)</td>
<td>46 (49)</td>
<td>41 (44)</td>
<td>6 (6)</td>
</tr>
<tr>
<td>Among patients with a prior intervention for RVH (n = 51)</td>
<td>17 (33)</td>
<td>29 (57)</td>
<td>5 (10)</td>
</tr>
<tr>
<td>Among patients without a prior intervention for RVH (n = 118)</td>
<td>57 (48)</td>
<td>49 (42)</td>
<td>12 (10)</td>
</tr>
<tr>
<td>Among patients with NFI (n = 31)</td>
<td>9 (29)</td>
<td>16 (52)</td>
<td>6 (19)</td>
</tr>
<tr>
<td>Among patients without NFI (n = 138)</td>
<td>65 (47)</td>
<td>62 (43)</td>
<td>11 (8)</td>
</tr>
<tr>
<td>Among patients with NFI and concurrent aortic treatment (n = 19)</td>
<td>3 (16)</td>
<td>11 (58)</td>
<td>5 (26)</td>
</tr>
<tr>
<td>Among patients &lt;3 years at operation (n = 21)</td>
<td>7 (33)</td>
<td>12 (57)</td>
<td>2 (10)</td>
</tr>
<tr>
<td>Among patients ≥3 years of age at operation (n = 148)</td>
<td>67 (45)</td>
<td>66 (45)</td>
<td>15 (10)</td>
</tr>
</tbody>
</table>

NFI, Neurofibromatosis type 1; RVH, renin-mediated renovascular hypertension. Values are reported as number (%).

<sup>a</sup>Includes primary index and secondary operations.
Conclusions

Contemporary Surgical Treatment of Pediatric RVH Must be Individualized

High Volume Center, Multi-disciplinary

Age at Operation & MAS Increases Likelihood of Reoperation

Children Undergoing Remedial Surgery Less Likely to be Cured of HTN

Sustainable Hypertension Benefit in ~90% with Acceptable Morbidity/Mortality

Post-operative Surveillance Imperative!
# Pediatric Arterial Aneurysms

<table>
<thead>
<tr>
<th>Etiology</th>
<th>Principal Artery Affected</th>
<th>Histologic and Morphologic Character</th>
<th>Clinical Characteristics</th>
</tr>
</thead>
<tbody>
<tr>
<td>I: Infection</td>
<td>Aorta, iliac arteries</td>
<td>Acute inflammatory infiltrates present initially, then chronic inflammation and fibrotic changes; saccular aneurysms</td>
<td>Umbilical artery catheterization, endocarditis, predisposing factors; fever, systemic infectious symptoms; untreated aneurysms often progress to rupture</td>
</tr>
<tr>
<td>II: Atherosclerosis</td>
<td>Aorta</td>
<td>Chronic inflammation with giant cells, vessel wall necrosis; saccular aneurysms</td>
<td>Signs and symptoms more often relate to stenotic disease than aneurysms, with the latter often remaining asymptomatic</td>
</tr>
<tr>
<td>III: Arteritis</td>
<td>Renal, splanchnic, axillobrachial, iliofemoral arteries (Kawasaki: coronary arteries)</td>
<td>Chronic parmural inflammation and degeneration, late fibrosis; multiple small saccular aneurysms</td>
<td>Usually asymptomatic but may cause hematuria, perirenal hematomas or death with rupture (lupus, periarteritis nodosa); myocardial infarction or tamponade (Kawasaki), aneurysm thrombosis more common than rupture</td>
</tr>
<tr>
<td>IV: Genetic</td>
<td>Aorta, muscular arteries</td>
<td>Medial elastic tissue disorganization, mucinous deposits (cystic medial necrosis); solitary fusiform and saccular aneurysms</td>
<td>Aortic rupture or dissection relatively common; arteriography and vascular reconstructions may be hazardous in the face of active disease</td>
</tr>
<tr>
<td>V: Developmental: Idiopathic</td>
<td>Aorta, renal, iliofemoral arteries</td>
<td>Medial thinning and fibroplasia; solitary and multiple saccular aneurysms affecting arterial bifurcations most often</td>
<td>Usually asymptomatic; extremity aneurysms often present as painless, pulsatile mass</td>
</tr>
<tr>
<td>VI: Traumatic: Extravascular</td>
<td>Aorta, extremity arteries</td>
<td>Disruption of usually three layers of artery (pseudoaneurysm), thrombosis of small aneurysms Common; saccular aneurysms</td>
<td>Protean manifestations; aortic aneurysms often rupture; peripheral lesions often asymptomatic</td>
</tr>
</tbody>
</table>
Frequently – etiology unclear

- Developmental (idiopathic)
- Post-stenotic dilation

6mo – 2cm AAA with occlusive coarctation
7y s/p open aneurysmorrhaphy,
primary aorto-aortic reanatomosis and reimplant
Developmental AAA Repair Principles

- Aneurysmorrhaphy with interposition graft v non-anatomic reconstructions
  - NB 6-10mm; young children 12-14mm; 14-20mm for larger children
  - Favor PTFE

- Role for closed aneurysmorrhaphy if cause of dilation can be eliminated (ie: coarctation) – assumes residual wall will remain stable
  - C/I = thrombus and large size
- 5yo with >3cm idiopathic AAA

- Treated with resection and primary repair

- No complications at 13y f/u
• 2002-2014 (N=11) aged 2wk – 6y
• 5 = infra-renal
• Perioperative death occurred in one child (preoperative heart and renal failure)
• Aortic graft occlusion affected two children at 1 month and 3 years postoperatively.
• The remaining children incurred no aortic reconstruction-related morbidity (mean f/u = 4.9y)
**AAA Conclusions:**

- Surgical treatment of AAA successful in > 90% of infants and young children
- Often requires complex operative techniques that consider:
  - growth potential
  - aneurysm location (splanchnic/renal involvement)
- Patients may derive great benefit from a multi-disciplinary approach
- Long-term follow-up strongly recommended for all surgical interventions
Pediatric Non-aortic Aneurysms

- 27 boys > 14 girls
- Mean age 9.8y (2mo – 18y)
- Mean weight 31kg
- 14 children had multiple aneurysms
- Majority developmental (14 post-stenotic) > trauma (12) > arteritis (4) > CTD (1)

Davis et al; JVS; 2016; 63.1
Axillary

Internal Carotid

Brachial

Ulnar

James C. Stanley, 2020
UM Experience with Pediatric Renal Artery Aneurysms (N=15, 26 aneurysms)

- 3-25mm (avg 9mm)
- 50% segmental
- Treatment:
  - Resection with primary anastomosis
  - Resection with reimplantation
  - Angioplastastic closure
  - Nephrectomy (N=4)
- 15% reintervention
14yo, NF1, 2-drug HTN + FTT

LRA resection, ex-vivo reconstruction w/ syndactylization of 3 segmental branches, aorto-renal bypass
UM Experience with Pediatric Splanchnic Aneurysms (6 patients with 7 aneurysms)

<table>
<thead>
<tr>
<th>Patient</th>
<th>Age (years)/sex</th>
<th>Affected artery</th>
<th>Size (mm)-morphological features; histological features</th>
<th>Associated diagnosis</th>
<th>Treatment, follow-up</th>
<th>Class</th>
</tr>
</thead>
<tbody>
<tr>
<td>4</td>
<td>13 M</td>
<td>Celiac</td>
<td>10 - Saccular</td>
<td>Superior mesenteric artery stenosis</td>
<td>Resection with reanastomosis, 1 month</td>
<td>VII</td>
</tr>
<tr>
<td>5</td>
<td>6 M</td>
<td>Celiac, superior mesenteric</td>
<td>4 and 4 - Saccular</td>
<td>AAA, bilateral renal artery stenosis, heterotaxy</td>
<td>Plication twice, 2 years</td>
<td>VI</td>
</tr>
<tr>
<td>6</td>
<td>4 M</td>
<td>Common hepatic</td>
<td>40 - Saccular; medial derangement</td>
<td>Kawasaki disease</td>
<td>Ligation, 1 month</td>
<td>IV</td>
</tr>
<tr>
<td>7</td>
<td>10 M</td>
<td>Superior mesenteric</td>
<td>40 - Fusiform; medial degeneration</td>
<td>Ehler-Danlos</td>
<td>Ligation, 2 years</td>
<td>V</td>
</tr>
<tr>
<td>8</td>
<td>14 M</td>
<td>Superior mesenteric</td>
<td>46 - Saccular; supplicative inflammation with microorganisms</td>
<td>Endocarditis, (bicuspid aortic valve)</td>
<td>Ligation, 1 years</td>
<td>I</td>
</tr>
<tr>
<td>9</td>
<td>15 F</td>
<td>Mesenteric branch</td>
<td>8 - Fusiform</td>
<td>—</td>
<td>Ligation, 1 month</td>
<td>VII</td>
</tr>
</tbody>
</table>

AAA, Abdominal aortic aneurysm; F, female; M, male.

*Pediatric Aneurysm Classification System defined according to Sarkar et al.11
13yo with post-stenotic 10mm celiac aneurysm
**Outcomes**

**Morbidity/Mortality**
- No postoperative mortality
- No major 30-day morbidity

**Freedom from Reintervention**
(mean f/u = 47mo)
- 83% at 1 year
- 69% at 3 years

5% required Secondary Intervention
Pediatric Renovascular HTN Center

University of Michigan Health System
C.S. Mott Children's Hospital

Pediatric Renovascular HTN

Pediatric Nephrology

David Kershaw

*Zubin Modi

Pediatric Urology

John Park

Anesthesia

Critical Care

Social Work

Matt Butler

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*Dawn Coleman

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Char Minard

Susan Young

*Co-Directors
Contact the pRVH PCOR Collaborative

(NIH R01 – HL139672-04) Genetic and Genomic Analysis of Arterial Dysplasia
(DOD NF190071) - Genetic Mechanisms of Neurofibromatosis-Related Arteriopathy and Renovascular Hypertension
(PCORI 19976-UM) – Pediatric Renovascular HTN – a pRVH PCOR Collaborative Taubman Institute - Michigan Medicine Dysplasia-Associated Arterial Disease (DAAD) Precision Medicine Network