Renal Artery Reconstruction in Children and Young Adults

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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 (cont)

- Echocardiogram: Severe concentric LVH (preserved systolic function).

- Abdominal ultrasonography: Tardus parvus renal artery waveforms, a very small right kidney, and infrarenal aortic stenosis.

- Cross-sectional Imaging:
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; re-implantation of LRA

Uneventful post-op course

D/C POD #18
  – Amlodipine – BP 120/50;

One year follow-up
  – 12.6kg (86th %)
  – Otherwise stable
  – Reassuring MRA

Phillips et al; Ann Vasc Surg; 2016(3)
**Renovascular HTN**

![Diagram showing the renin-angiotensin system interaction withaldosterone and bradykinin in regulation of blood pressure.]

**Pediatric HTN**

- Normal BP: < 90th % for sex, age, height
- Hypertension: Average SBP or DBP ≥ 95th % 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 RV HTN**

- 3rd most common cause of HTN in children (5-10%)
- Associations with:
  - NF-1, William’s Syndrome, Alagille Syndrome, Tuberous Sclerosis and maternal infection
  - Arteritis
  - Umbilical artery catheterization

**Histopathology**

- Intimal proliferation
- Fragmentation of elastic lamina
- Medial thinning
- Excessive perivascular elastin
**Natural History**

- Medically refractory HTN, failure to thrive, renal insufficiency and progression to LVH
- Life expectancy (untreated) <40 years
- COD: Heart failure, flash pulmonary edema, hypertensive encephalopathy, stroke/ICH

**Mid-abdominal Syndrome (MAS)**

- Classified by cephalad extent of narrowing
  - Suprarenal (69%)
  - Intrarenal (23%)
  - Infra-renal (8%)
- Thought to arise from embryonic overfusion of the paired dorsal aortas during the 4th week of development
  - 87% renal involvement
  - 62% splanchnic involvement

**Diagnostics**

- Inflammatory work-up (ESR/CRP)
- Blood hormone levels (renin)
- Echocardiogram
- Renal Duplex
  - 90% Sensitive, 68% Specific for main renal/Ao
- Cross-sectional Imaging (MRA)
- Diagnostic arteriography and renal vein renin sampling

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

*Castelli et al; Pediatr Radiol; 2014*
**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)

**From the Society for Vascular Surgery**

Pediatric renovascular hypertension: 132 primary and 30 secondary operations in 97 children

James C. Stanley, MD,* Enrique Créado, MD,* Gilbert R. Upchurch, Jr, MD,* Patrick D. Brophy, MD,* Kyung J. Cho, MD,* and John E. Reutenwald, MD,* for the Michigan Pediatric Renovascular Group,* Ann Arbor, Mich

- 97 patients (58 boys, 39 girls), 3 mos to 17 years (1963-2006)
- 80% developmental renal artery disease
- Concurrent disease:
  - Splanchnic arterial occlusive lesions 24%
  - Abdominal coarctations 33%

_J Vasc Surg 2006; 44: 1219-29_

**UM Experience with RVH - 2006**

- 132 primary operations
  - 13 primary nephrectomies
- 30 secondary operations
  - 9 primary nephrectomies
- 17 mesenteric revascularizations
- 30 aortic reconstructions

**Options for Reconstruction**

- Renal artery aortic reimplantation (49)
- Aorto-renal or ilio-renal bypass (40)
- Extra-anatomic bypass (2)
- Segmental reimplantations (7)
- Arterioplasty (10)
- Resection with re-anastomosis (4)
- Nephrectomy – partial / complete (sub-capsular)
6yo with bilateral ostial RAS s/p bilateral renal artery reimplantation

Aorto-aortic bypass (PTFE); retro-renal tunnel with visceral revasc.

Patch aortoplasty (PTFE) with visceral revasc.

Results

<table>
<thead>
<tr>
<th>Time period (Patients Treated)</th>
<th>1968-72 (21)</th>
<th>1973-80 (13)</th>
<th>1981-93 (24)</th>
<th>1994-06 (40)</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>Cured</td>
<td>81% (17)</td>
<td>77% (10)</td>
<td>78% (19)</td>
<td>58% (23)</td>
<td>78% (68)</td>
</tr>
<tr>
<td>Improved</td>
<td>14% (3)</td>
<td>29% (3)</td>
<td>22% (5)</td>
<td>37% (15)</td>
<td>27% (26)</td>
</tr>
<tr>
<td>Unchanged</td>
<td>5% (1)</td>
<td>4% (0)</td>
<td>0% (0)</td>
<td>5% (2)</td>
<td>3% (3)</td>
</tr>
</tbody>
</table>

*See text for outcome definitions.

*One patient listed twice with opposite renal artery disease treated in 1968-72 and in 1981-90.
Technical Pearls

- Single-stage operation
- Favor reimplantation
- 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)

Late vein graft aneurysm (8 years)

PTA

Angioplasty for Renal Artery Stenosis in Pediatric Patients: An 11-year Retrospective Experience

Abhay Srinivasan, MD, Ganesh Krishnamurthy, MD, Lucia Fontalvo-Herazo, MD, Elis Nijp, MD, Marc S. Keller, MD, Kevin Meyers, MD, Bernard Kaplan, MD, and Anne-Marie Cahill, MD

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


UM Experience: Endovascular Failures
18yo – single-drug HTN, failed PTA and early stent

Stent Extraction and LRA reimplantation

10yo, stented for dissection complicated PTA

Stent extraction, syndactalization and LRA reimplantation

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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

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

**2017 Contemporary Update**

- 218 children – 55% last decade
  - 29% previous open or EV intervention prior to transfer to UM

- 19% risk of reoperation
  - Age < 5 years at index surgery

- 43% cure, 42% improvement, 14% unchanged
  - Remedial operations and MAS – less likely to be cured of hypertension

**UM Experience with Renal Artery Aneurysms (N=15, 26 aneurysms)**

- 3-25mm (avg 9mm)
- 50% segmental
- Treatment:
  - Resection with primary anastomosis
  - Resection with reimplantation
  - Angioplastic 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

*Davis et al; JVS; 2016; 63.1
Coleman and Stanley; JVS; 2015*
Conclusions

• Open surgical revascularization may be performed with negligible M&M

• Completion imaging and surveillance critical

• Multidisciplinary team important

Pediatric Vascular Program

• Pediatric Nephrology
  – David Kershaw
  – Neal Blatt
  – David Selewski
  – Kera Luckritz

• Pediatric Urology
  – John Park

• PICU

• Pediatric Anesthesia

• Social Work
  – Matt Butler

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  – James Stanley
  – Jonathan Eliason
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• Interventional Radiology
  – David Williams
  – Ravi Srinavasa

• Nurse Coordinators
  – Char Minard
  – Susan Young

Thank You
Fibromuscular Dysplasia (FMD)

- Non-atherosclerotic, non-inflammatory vascular disease that may result in arterial stenosis, occlusion, aneurysm or dissection
- Cause unknown
- Prevalence unclear
- Clinical manifestations dependent on vascular bed involved

Histopathological Classification

<table>
<thead>
<tr>
<th>Histologic (1971)</th>
<th>Angiographic</th>
</tr>
</thead>
<tbody>
<tr>
<td>Medial: Medial fibroplasia (60-70%)</td>
<td>Multifocal</td>
</tr>
<tr>
<td>Perimedial fibroplasia (15-25%)</td>
<td></td>
</tr>
<tr>
<td>Medial hyperplasia (1-2%)</td>
<td></td>
</tr>
<tr>
<td>Intimal fibroplasia (1-2)</td>
<td>Unifocal (&lt;1cm)</td>
</tr>
<tr>
<td>Tubular (≥ 1cm)</td>
<td></td>
</tr>
<tr>
<td>Adventitial (&lt;1%)</td>
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Focal FMD:
- Younger at diagnosis and onset of HTN (26 v 40y)
- Male:Female (2:1)
- More likely to undergo revascularization (90 v 35%)
- Higher rate of HTN cure (54 v 26%)  
  Savard et al, Circulation, 2012

Renal FMD

- Suspect FMD when:
  - Early onset HTN (<35 years)
  - Medically-refractory HTN
- Average age of onset ~ 43.1 years
- Renal insufficiency, dissection, infarction and CKD uncommon
- H/A common

**Renal Artery Revascularization - INDICATIONS**

- Medically refractory HTN (failure to reach goal BP with appropriate 3-drug regimen that includes a diuretic)
- HTN of short duration with goal of cure
- Dissection
- Aneurysm
- Preservation of renal function

- **NO RANDOMIZED, CONTROLLED TRIALS**

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**Renal Angioplasty**

- Diagnostic: Ostia, main artery, branches and parenchyma
- 0.014 pressure wire (pressure gradient)
- +/- IVUS
- Size balloon to normal vessel – semi-compliant (avoid cutting/scoring)
- Post-PTA arteriogram and PRESSURE gradient
- Heparin, Papaverine / Nitro available

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**Renal PTA Outcomes**

- Data Limited (retrospective, short-term f/u and variable definitions of cure)
- Suggested 50% cure rate (up to 86% improvement)
- Re-intervention ~20%
- Complications typically minor
- Surgical revascularization (selective patients) offers HTN cure 33-72%

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**Young woman + FMD + HTN**

Coleman and Stanley; JVS; 2015
Clinical Pearls (AHA updates)

- Screen for occult aortic / arterial aneurysm
- Reserve genetic testing for suspected CTD
- Anti-platelet therapy
- Reserve renal PTA for select patients with significant likelihood of success and significant pressure gradient
- SELECTIVE renal artery stenting
- Do not intervene on ASx patients (exclusive of aneurysm)