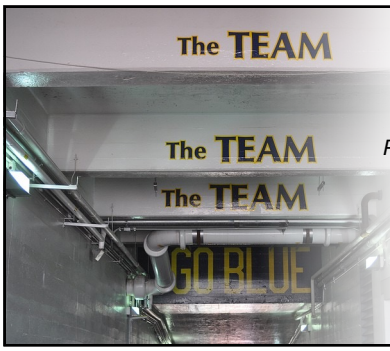
Multidisciplinary Consensus Statement On the Management of Pediatric Renovascular Hypertension, Renal Artery Stenosis and Midaortic Syndrome – Optimal Conduits, Techniques and Results

Dawn M. Coleman, MD
Professor, Surgery
Division Chief, Vascular Surgery and Endovascular Surgery
Duke University

Duke Surgery



Disclosures

Project Lead (PCORI-funded pRVH PCOR Collaborative)

Past Co-Director, pRVH Center, University of Michigan

Leadership (APDVS / VESS)

Consensus Statement:

NO CONSENSUS

Pediatric RV HTN

- 3rd most common cause of HTN in children (5-10%)
- Up to 40% of patients may have a genetic etiology
  - *NF1, ELN/Williams Syndrome, TSC1/2, JAG1, FBN1, Turner Syndrome and Alagille Syndrome*

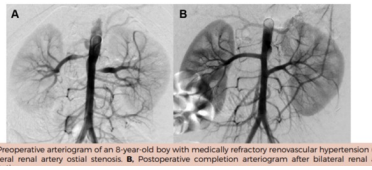
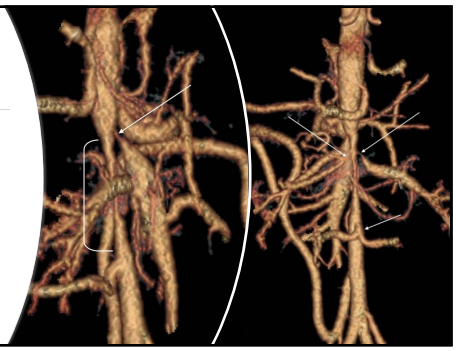


Fig 1. **A.** Preoperative arteriogram of an 8-year-old boy with medically refractory renovascular hypertension (RVH) and bilateral renal artery ostial stenosis. **B.** Postoperative completion arteriogram after bilateral renal artery

Mid-abdominal Aortic Syndrome (MAS)

- Classified by cephalad extent of narrowing
- Renal Involvement 87%
- Splanchnic Involvement 62%



Timely Detection and Treatment Prevents Kidney Injury, Cardiopulmonary Complications and Flash Pulmonary Edema, Stroke and Death

- Medically refractory HTN
- Optimize Renal Perfusion (atrophy, single kidney)
- NICM (concentric LVH)
- Failure to thrive
- Lower extremity sequelae (claudication, exertional fatigue, growth disturbance)
- Consider timing and challenges (patient size and projected growth)

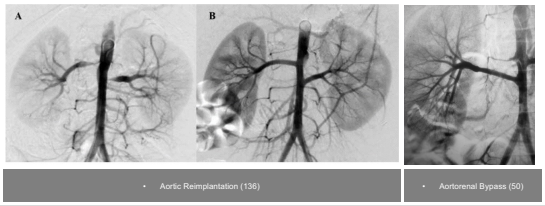
Table I. Demographics and preoperative risk factors

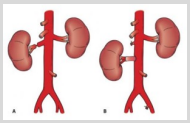
Variable	Total cohort (N = 169)
Male sex	93 (55)
Age at time of surgical intervention at University of Michigan, years	8.96 ± 5.53
Weight at time of intervention at University of Michigan, kg	58.33 ± 16.8
No. of initial antihypertensive medications	2.70 ± 1.58
NF1	31 (18.34)
Abdominal aortic coarctation	76 (44.97)
Prior intervention performed elsewhere for RVH before intervention at University of Michigan	51 (30.18); open surgical (14), endovascular (35), combination (2)

NF1, Neurofibromatosis type 1; *RVH*, renin-mediated renovascular hypertension.
Categorical variables are presented as number (%). Continuous variables are presented as mean ± standard deviation.

Surgical management of pediatric renin-mediated hypertension secondary to renal artery occlusive disease and abdominal aortic coarctation

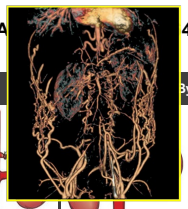
Dawn M. Coleman, MD,[a] Jonathan L. Eliason, MD,[a] Robert Beaulieu, MD,[a] Tatum Jackson, BA,[a] Monita Karmakar, PhD,[a] David B. Kershaw, MD,[a] Zubin J. Modi, MD,[a,b] Sarthi K. Ganesh, MD,[a] Minhaj S. Khaja, MD, MBA,[a] David Williams, MD,[a] and James C. Stanley, MD,[a] on behalf of the University of Michigan Pediatric Renovascular Hypertension Center, *Ann Arbor, Mich*



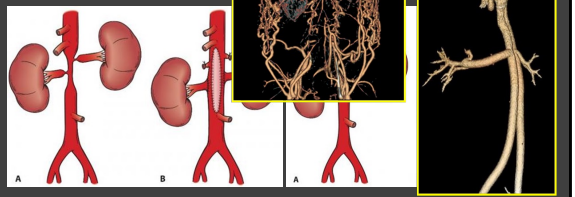


Primary Surgical Procedures

Concurrent A **4)**



Patch Aortoplasty **By**



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%)

Hypertension Benefits

- **44% Cured, 46% Improved, 10% No change**
- Mean Number of Anti-hypertensives Post-op = 0.99 ±1.16 *(v. 2.7 ±0.58 preop)*
- Mean Post-op Cr = 0.51 ± 0.30
- No Dialysis

Independent Predictors of Reoperation (logistic regression)

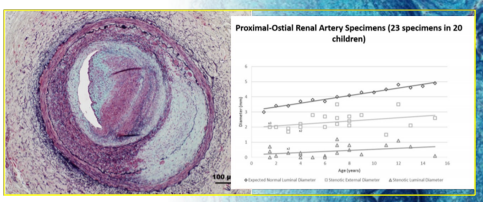
Table VII. Postoperative hypertension benefits

	Cure	Improved	Failure (unchanged)
Total cohort (N = 169)*	74 (44)	78 (46)	17 (10)
Among patients with concurrent abdominal aortic coarctation (n = 76)	28 (37)	37 (49)	11 (14)
Among patients without abdominal aortic coarctation (n = 93)	46 (49)	41 (44)	6 (6)
Among patients with a prior intervention for RVH (n = 51)	17 (33)	29 (57)	5 (10)
Among patients without a prior intervention for RVH (n = 118)	57 (48)	49 (42)	12 (10)
Among patients with NF1 (n = 31)	9 (29)	16 (52)	6 (19)
Among patients without NF1 (n = 138)	65 (47)	62 (45)	11 (8)
Among patients with NF1 and concurrent aortic treatment (n = 19)	3 (16)	11 (58)	5 (26)
Among patients <3 years at operation (n = 21)	7 (33)	12 (57)	2 (10)
Among patients ≥3 years of age at operation (n = 148)	67 (45)	66 (45)	15 (10)

NF1, Neurofibromatosis type 1; *RVH*, renin-mediated renovascular hypertension.
Values are reported as number (%).
*Includes primary index and secondary operations.

As age increased by one year, the rate of reoperation decreased by ~10% (HR=0.90; 95% CI: 23 0.83-0.97)

Renal Artery Occlusive Disease



Coleman DM; JVS; 2021

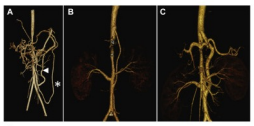
Endovascular Therapy

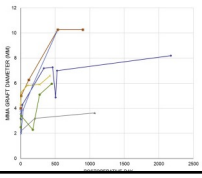
- Catheter-based interventions increasingly reported
  - Restenosis well described (at least 15%)
    - More frequently reported in children with defined genetic syndromes
    - Benefits from secondary (salvage) open surgical revascularization may be limited
      - Nephrectomy rates 20%, blunted HTN improvement
  - **Outcomes ~1/3 cured, 1/3 improved, 1/3 no improvement**
  - Serial angioplasty often required
  - Early recurrence portends poor outcomes
- No role for stenting (routinely)

Alexander, J Pedi Surg, 2017
Agrawal, Congen Heart Dis, 2018
Kari, Arch Dis Child, 2015
Patel, CVIR, 2021

Innovative Surgica
Need for Pro

- TESLA (n=4, 5.7y follow-up)
  - Tissue-Expander Stimulated Le
- MAGIK (n
  - Margina
  - Graft hy
- Normoten
  (mean 1-2
- Both assc
  catheter)





Kim, JVS, 2020

Conclusions

- Surgical reconstructions must offer durability with acceptable morbidity
- Surveillance is critical
- Leverage a multi-disciplinary team
- Phenotype ("genotype") should drive individualized surgical decision making / treatment plan

Duke pRVH Clinical Program: Stakeholders

Vascular Surgery: D. Coleman
Transplant Surgery: D. Sudan, D. Vikraman-Sushama
Congenital Cardiac Surgery: J. Turek
Radiology and IR: J. Cao, W. Pabon-Ramos
Pedi Nephrology: A. Chua, C. Sheldon, R. Gbadegesin
Anesthesia: W. Ames, E. Jooste, M. McDaniel
Critical Care: O. Alibrahim
Genetics: J. Cohen, E. Nading
APP: J. Prescott, B. Kozak

THANK YOU!

ganeshlab.org/contact/
Ganesh Lab

ColemanDM_vasc

(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

PCORI pRVH PCOR Collaborative