Pediatric Aortic Aneurysms

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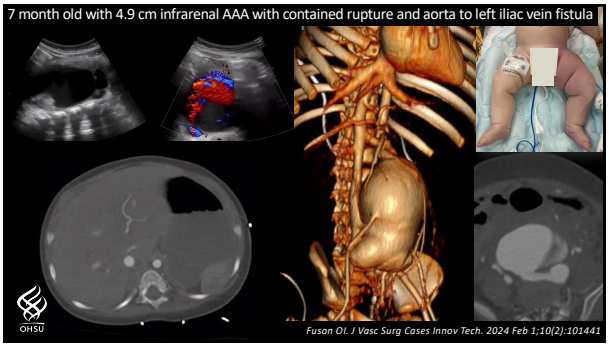
Disclosures

None

7 month old with 4.9 cm infrarenal AAA with contained rupture and aorta to left iliac vein fistula



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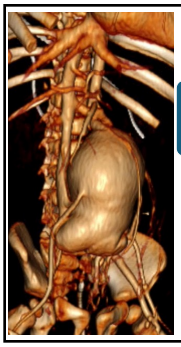


Pediatric AAA

113 cases from 1967-2024

Exceedingly rare

?? Prevalence: Most cases are case reports/series

Data synthesized

RESEARCH Open Access

Diagnosis and treatment of congenital abdominal aortic aneurysm: a systematic review of reported cases

Orphanet Journal of Rare Diseases (2015) 10:4

surgical treatment of abdominal aortic aneurysms n infancy and early childhood 2014

N=26

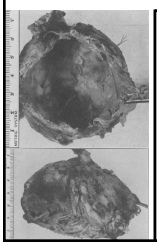
N=11
5 years
follow up

Case Reports

First documented case in 1946

ANEURYSM OF THE LOWER ABDOMINAL AORTA WITH RUPTURE IN A SIXTEEN MONTH OLD INFANT

THOMAS A. GIBSON, M.D., WINCHESTER, VA.



ANEURYSM OF ABDOMINAL AORTA IN THE NEWBORN INFANT

Report of Case

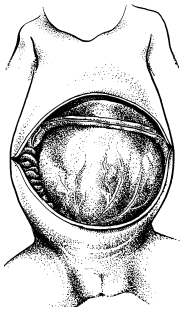
M. BECKETT HOWORTH, JR., M.D.

OXFORD, MISSISSIPPI

ANEURYSM of the aorta in children is rare. Aneurysm of the abdominal aorta at birth of a living infant has not been reported. In the following

THE NEW ENGLAND JOURNAL OF MEDICINE

May. 18, 1967

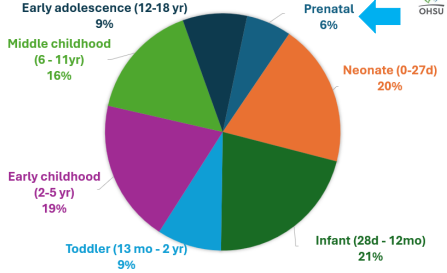


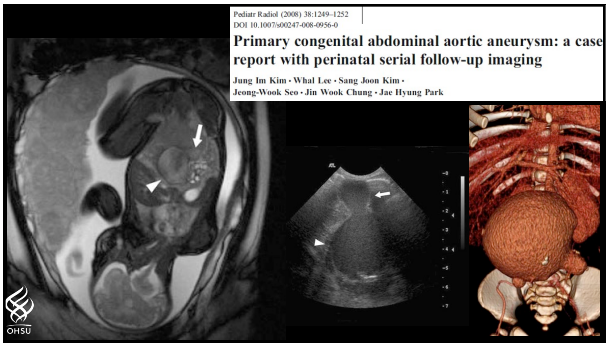
Age of presentation (N=113)

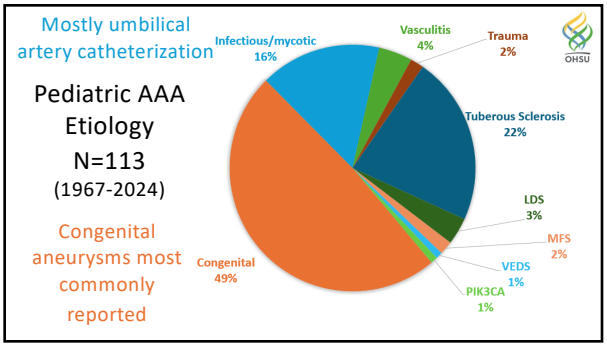
55% < 2 years

59% males
41% females

Most case reports have short-term follow-up

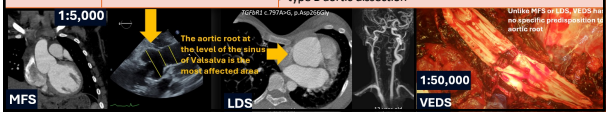






Heritable thoracic aortic disease does not commonly present with pediatric AAA

	Pediatric AAA	Pediatric Thoracic Aortic Disease
Prevalence	Extremely rare, case reports	Rare
Etiology	Congenital, genetic infections, trauma	Syndromic genetic aortopathies Hypertension
Genetics	Tuberous sclerosis >> Syndromic genetic aortopathies	Marfan, Loeys-Dietz, VEDS, Smooth muscle cell aortopathies
Age of Onset	Infants or young children Prenatal	Adolescence or young adulthood Neonatal Marfan syndrome
Anatomy	Infrarenal > juxtarenal > TAAA	Aortic root aneurysm/type A aortic dissection > type B aortic dissection



Tuberous Sclerosis

Major Criteria	Minor Criteria
Hypomelanotic macules (≥3; at least 5 mm diameter)	"Confetti" skin lesions
Angiofibroma (≥3) or fibrous cephalic plaque	Dental enamel pits (≥3)
Ungual fibromas (≥2)	Intraoral fibromas (≥2)
Shagreen patch	Retinal achromic patch
Multiple retinal hamartomas	Multiple renal cysts
Multiple cortical tubers and/or radial migration lines	Nonrenal hamartomas
Subependymal nodule (≥2)	Sclerotic bone lesions
Subependymal giant cell astrocytoma	
Cardiac rhabdomyoma	
LAM*	
Angiomyolipomas (≥2)*	

1:6,000-10,000

Updated International Tuberous Sclerosis Complex Diagnostic Criteria and Surveillance and Management Recommendations

Insufficient evidence to support routine evaluation for AAA at time of diagnosis unless there are clinical symptoms....

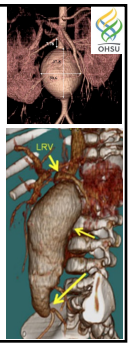
Northrup et al, *Pediatric Neurology 123 (2021) 50e66*

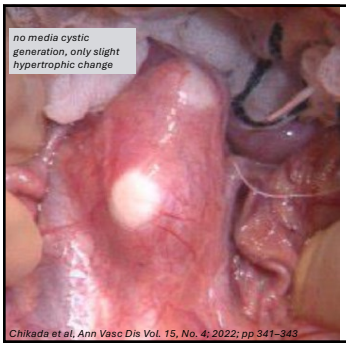
Autosomal dominant pathogenic variant in the genes coding for TSC1 or 2 (*TSC1, TSC2*) → mTOR overactivation

deletion of TSC1 in smooth muscle cells → ? degradative smooth muscle phenotype

Triad: seizures, intellectual disability, and angiofibromas

Multisystem: dermatologic, neurological, heart, lungs, and kidneys





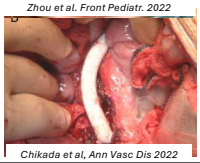
Congenital AAA

Developmental defect during embryogenesis

→focal narrowing of the abdominal aorta
→ post-stenotic turbulent blood flow
→increased stress on the aortic wall
→ AAA

Surgical Challenges

Zhou et al. Front Pediatr. 2022



Chikada et al, Ann Vasc Dis 2022

Need for growth accommodation

Age (years)	Proximal Aorta (mm)	Distal Aorta (mm)	Iliac Artery (mm)
1–3	8.8	6.7	4.5
3–7	10.0	8.1	5.3
7–11	12.3	10.8	7.1
11–15	14.5	12.1	8.5
15–17	16.5	14.0	9.3

What do we know?

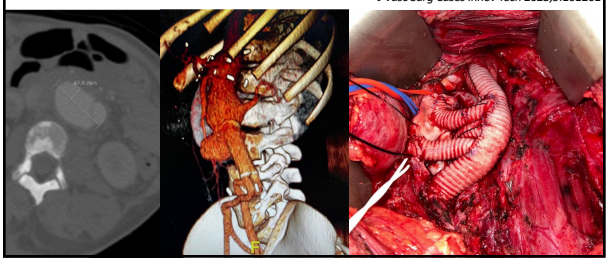
- Infant graft sizes ranged 4-10 mm
  - Small vessel size limits graft options
  - Patency of grafts at diameters < 6 mm have poor *(can you delay?)*
- Reintervention likely as they grow

Recurrent symptomatic aortic aneurysm in a young girl with tuberous sclerosis complex and review of the literature

9 years old

Ayman M. Alrazim, MD,[a] Joshua K. Burk, MD, MBA,[a] Travis J. Wilder, MD,[b] Woosup Michael Park, MD,[a] William J. Yoon, MD,[a] and Jae S. Cho, MD,[a] Cleveland, OH

J Vasc Surg Cases Innov Tech 2023;9:101261



Pediatric AAA: Operative Strategies

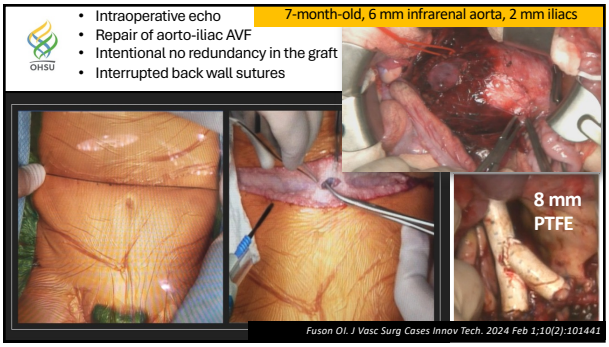
APPROACH	INDICATION	MATERIALS USED	KEY CONSIDERATIONS
Interposition Bypass	Most common approach	**Synthetic grafts (Dacron, PTFE), cryopreserved or autologous grafts**	**Sizing to accommodate future growth, C-shaped configuration**

Graft Materials Selection

Material	Advantages	Disadvantages
Synthetic (Dacron, PTFE)	Durable, widely available	Fixed size
Cryopreserved Allograft	Size flexibility, reduced infection risk	Aneurysmal Degeneration, limited supply
Autologous Tissue	Possibly reduces need for reintervention	Limited availability in infants

- Intraoperative echo
- Repair of aorto-iliac AVF
- Intentional no redundancy in the graft
- Interrupted back wall sutures

7-month-old, 6 mm infrarenal aorta, 2 mm iliacs



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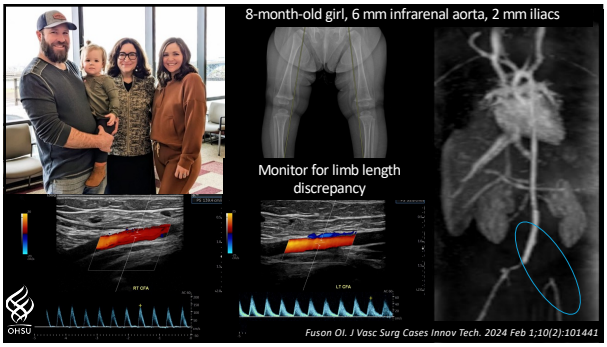
8-month-old girl, 6 mm infrarenal aorta, 2 mm iliacs



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Pediatric AAA: Operative Strategies

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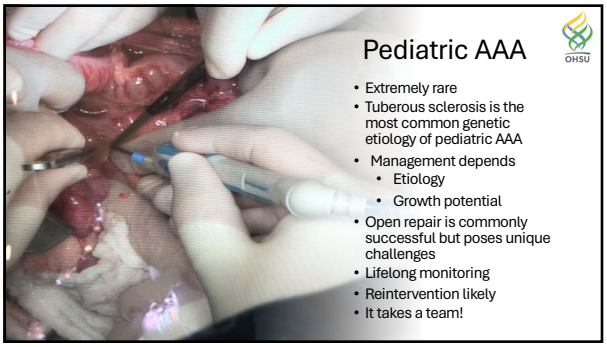
Aneurysmorrhaphy	Small aortic size or post-stenotic dilation	n/a	Preserves native tissue; not for infections/genetic AAA
Resection & Patch Angioplasty	Saccular aneurysms	Prosthetic, autologous, or cryopreserved patches	May need reintervention as patient grows
Aortic Ligation	Complex anatomy or infection	n/a	Collateral perfusion essential
Endovascular Techniques	Adolescents or temporizing measure	Palmaz stents reported	Limited role due to growth concerns



Pediatric AAA

- Extremely rare
- Tuberous sclerosis is the most common genetic etiology of pediatric AAA
- Management depends
  - Etiology
  - Growth potential
- Open repair is commonly successful but poses unique challenges
- Lifelong monitoring
- Reintervention likely
- It takes a team!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