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Early Childhood Outcomes Following Repair of Truncus Arteriosus: A Contemporary Multicenter Analysis

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Disclosures

- None

Background

- **Truncus arteriosus (TA)**
  - Complex defect requiring multiple interventions throughout life
  - Relatively uncommon
    - ~300 US cases per year, 0.7% of CHD, 4% of CCHD
  - Perioperative morbidity and mortality continues to be significant
  - Reports describing intermediate and long-term outcomes are limited to single-center experiences

Aims

- Describe contemporary early childhood outcomes after repair of truncus arteriosus
- Identify risk factors for RV-PA conduit intervention

Methods

- Multicenter, retrospective cohort study
- Inclusion: any patient who underwent surgery for TA
- Study period: 2009 – 2016
- Exclusion:
  - concomitant arch obstruction / interrupted aortic arch
  - “hemitruncus” and “pseudotruncus”

Methods: Institutions

1. Riley Hospital for Children, Indianapolis, IN
2. Cleveland Clinic, Cleveland, OH
3. Children’s Hospital of Michigan, Detroit, MI
4. Morgan Stanley Children’s Hospital of New York, New York, NY
5. North Shore-LIJ Cohen Children's Medical Center, New Hyde Park, NY
6. Medical University of South Carolina, Charleston, SC
7. Arnold Palmer Hospital for Children, Orlando, FL
8. Seattle Children’s Hospital, Seattle, WA
9. Ann & Robert H. Lurie Children’s Hospital of Chicago, IL
10. University of Iowa Stead Family Children’s Hospital, Iowa City, IA
11. Children’s National Health System, Washington, DC
12. University of Utah Health, Salt Lake City, UT
13. Cincinnati Children’s Hospital Medical Center, Cincinnati, OH
14. University of Michigan C.S. Mott Children's Hospital, Ann Arbor, MI
15. American Family Hospital, Madison, WI

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Methods: Definitions and Analysis

- **Operative mortality:** STS-CHSD definition
- **Late mortality:** death after hospital discharge or > 30 days post-op
  - Kaplan-Meier survival analysis and Cox Regression analysis to determine risk factors for overall mortality
- **RV-PA conduit intervention:** cath or surgical intervention
- **RV-PA conduit replacement:** surgical replacement
  - Fine Gray competing risk model used to determine probability of any RV-PA conduit intervention or replacement over time

Results

- 216 patients with TA, 15 centers
- Operative mortality: n = 16 (7.4%)
- Overall mortality: n = 29 (13.4%)
- 207 patients received RV-PA conduits
  - 8 underwent direct anastomosis of main PA
  - 1 underwent systemic-to-PA shunt
- Median follow-up: 2.9 years (range: 0.1 – 8.8 years)

Results: Baseline Characteristics

Variable	n	%
Prenatal diagnosis	135	63%
Prematurity	42	19%
Any chromosomal abnormality	83	38%
DiGeorge syndrome	61	28%
Non-cardiac abnormality	63	29%
Preop mechanical ventilation	45	21%
Preop NEC	17	8%

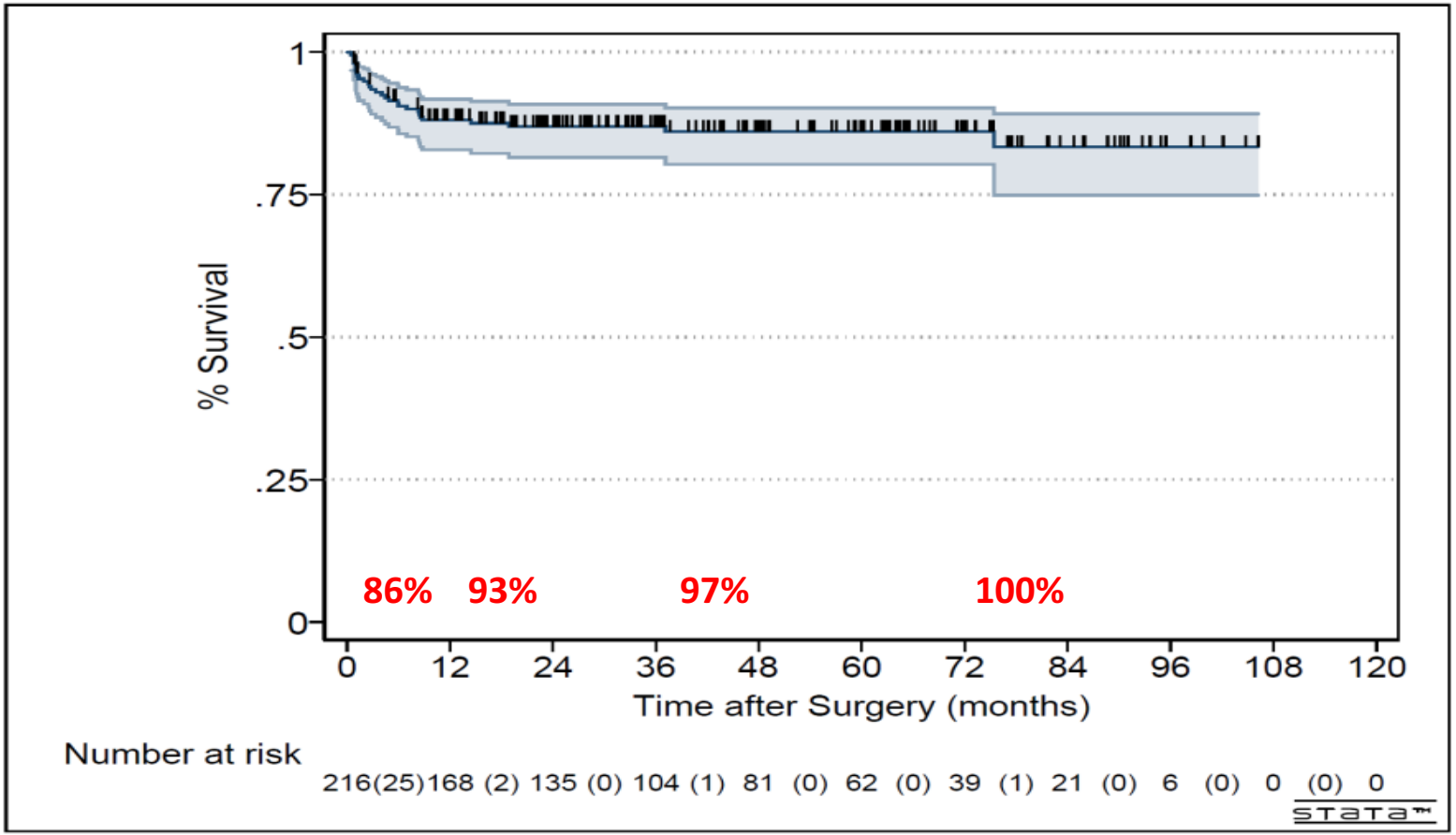
Results: Operative Data

Variable	n / median	% / (IQR)
Truncus Type (Collette-Edwards)		
Type 1	112	52%
Type 2	90	42%
Type 3	14	6%
Age at surgery, days	10	(7, 24)
CPB Duration, min	150	(124, 186)
Corticosteroids	161	75%
Truncal valve repair	36	17%

Results: Postoperative Data

Variable	n	%
Delayed sternal closure	126	58%
ECMO	22	10%
iNO utilization	102	47%
Postoperative infection	43	20%
Postoperative arrhythmia	78	36%
Reoperations	47	22%
Operative mortality	16	7%

Results: Overall Mortality



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Cox Regression Analysis: Overall Mortality

Risk Factor	Hazard Ratio	95% CI		p-value
DiGeorge syndrome	2.2	1.1	4.7	0.04
Preoperative ventilation	2.4	1.2	5.4	0.02
Delayed sternal closure	3.3	1.1	9.8	0.04
Postoperative ECMO	5.3	2.3	11.8	<0.01

Bivariate Analysis: Risk Factors for Late Mortality

Risk Factor	Operative mortality	p-value	Overall mortality	p-value
DiGeorge syndrome	**3 (19%)**	**0.38**	**13 (45%)**	**0.03**
Preoperative ventilation	**5 (31%)**	**0.34**	**13 (45%)**	**0.02**
ECMO	8 (50%)	<0.01	11 (38%)	<0.01
Delayed sternal closure	15 (94%)	<0.01	25 (86%)	<0.01

Results: RV-PA Conduit Characteristics

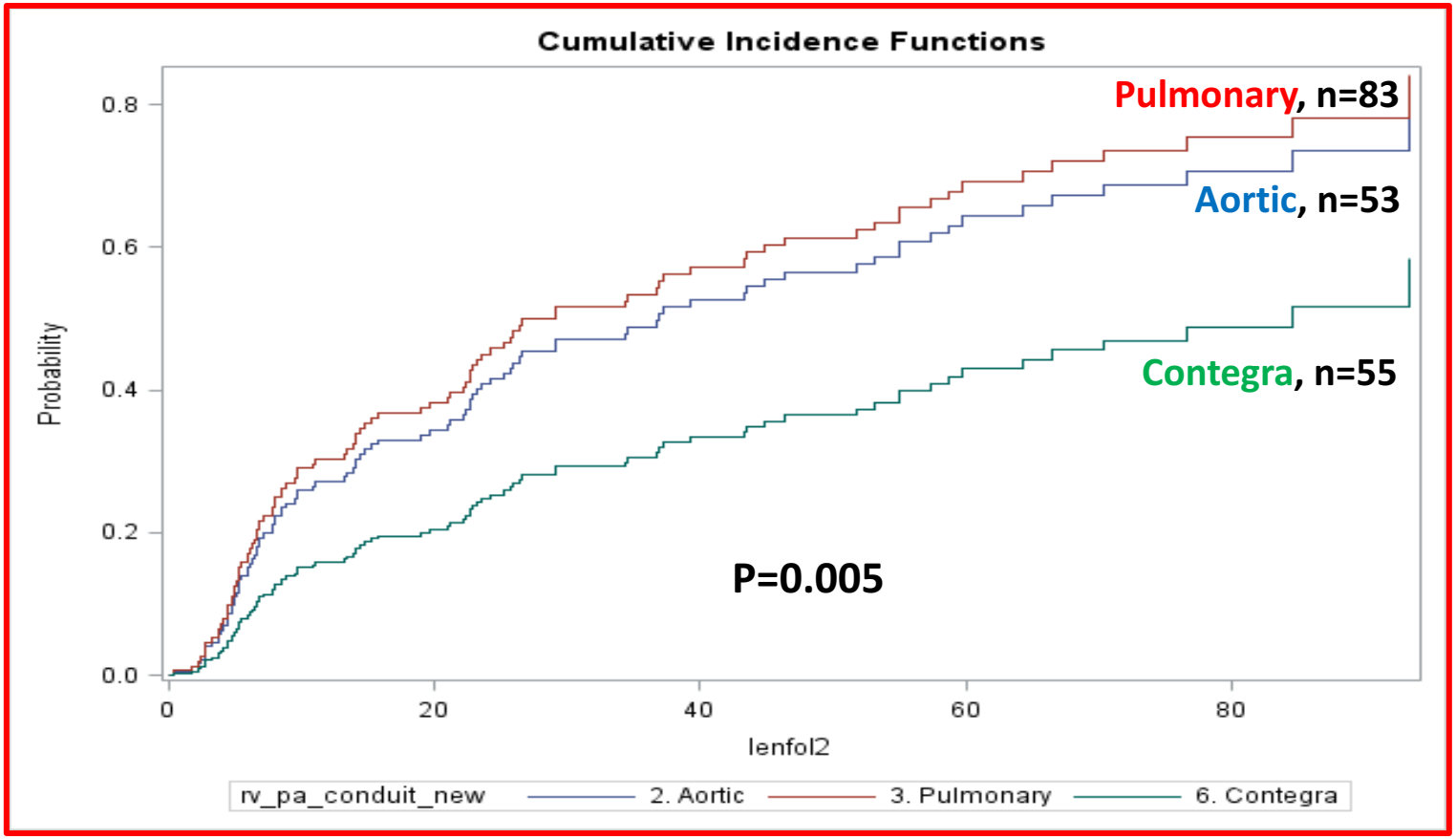
Variable	n / median	% / (IQR)
RV-PA conduit type		
Pulmonary homograft	83	38%
Contegra	55	25%
Aortic homograft	53	24%
Femoral vein	13	6%
Direct anastomosis / patch	8	4%
Other	4	2%
RV-PA Conduit Size, mm	11	(9, 12)

Results: RV-PA Conduit Intervention

Conduit	Pulmonary (n=83)	Aortic (n=53)	Contegra (n=53)
Duration of Follow-up (months)	36 (18, 60)	36 (22, 63)	42 (15, 77)
# of conduit interventions	48 (58%)	29 (55%)	**20 (36%)**
Time to first intervention (months)	11 (6, 24)	14 (6, 27)	**20 (13, 46)**

*Data represented as median (IQR) or absolute counts (%) as appropriate

Results: Probability of Conduit Intervention



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Results: Probability of Conduit Intervention

Conduit Comparison	Hazard Ratio	95% CI		p-value
Pulmonary vs. Aortic	1.14	0.72	1.80	0.57
Aortic vs. Contegra	**1.84**	**1.05**	**3.20**	**0.03**
Pulmonary vs. Contegra	**2.10**	**1.27**	**3.48**	**< 0.01**

Results: Probability of Conduit Intervention

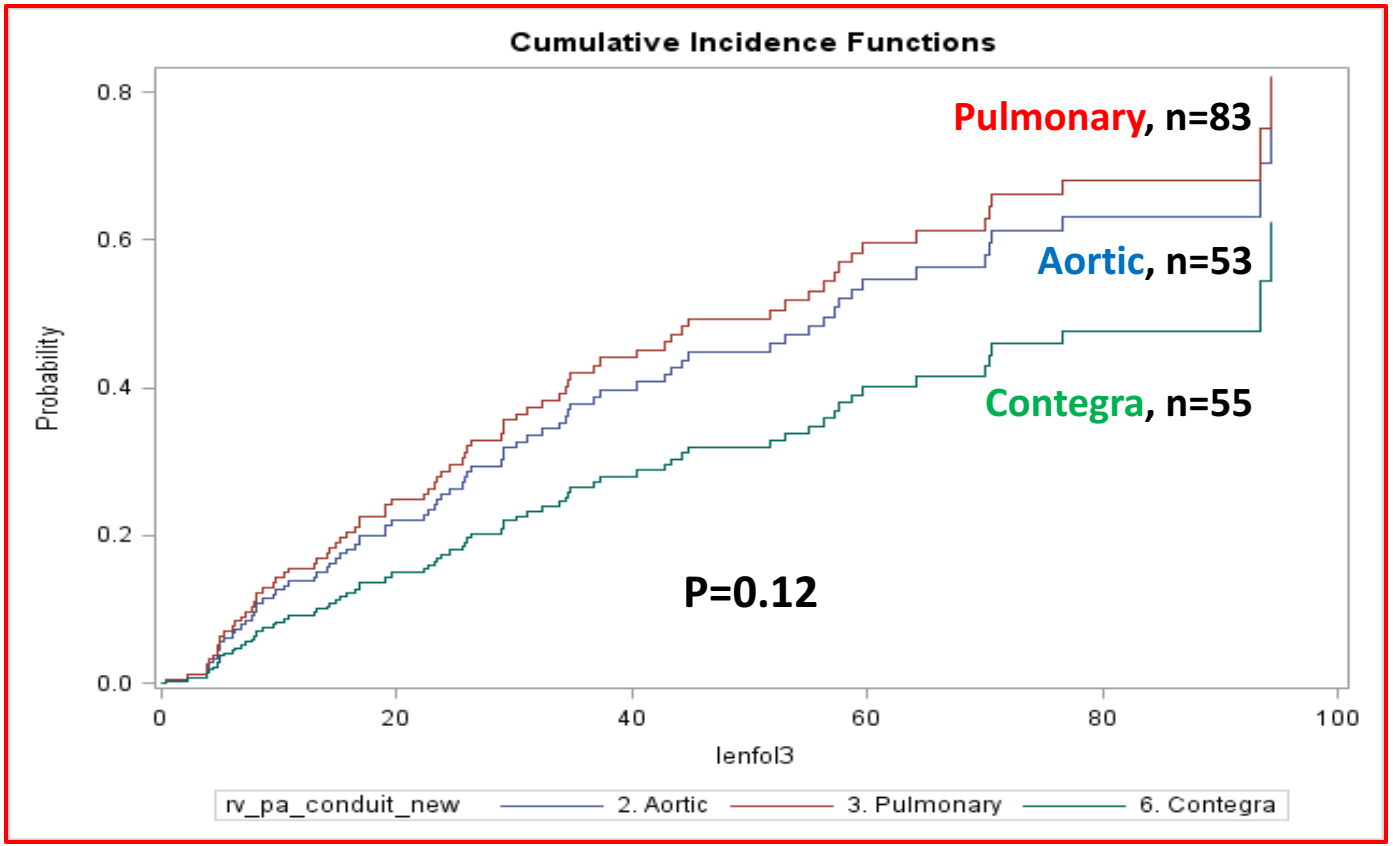
Conduit Comparison	Hazard Ratio	95% CI		p-value
Pulmonary/Aortic vs. Contegra	1.8	1.1	3.0	**0.03**
Conduit Size (mm/m^2)	1.05	1.05	3.20	**<0.01**

Results: RV-PA Conduit Replacement

Conduit	Pulmonary (n=83)	Aortic (n=53)	Contegra (n=53)
Duration of Follow-up (months)	36 (18, 60)	36 (22, 63)	42 (15, 77)
# of conduit replacements	39 (47%)	23 (43%)	**19 (35%)**
Time to replacement (months)	23 (7, 38)	27 (14, 37)	24 (9, 55)

*Data represented as median (IQR) or absolute counts (%) as appropriate

Results: Probability of Conduit Replacement



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Results: Probability of Conduit Replacement

Conduit Comparison	Hazard Ratio	95% CI		p-value
Pulmonary/Aortic vs. Contegra	1.5	0.9	2.6	0.12
Conduit Size (mm/m^2)	1.05	1.01	1.08	**<0.01**

Limitations

- Retrospective analysis
  - Differential losses to follow-up?
- Relatively small number of mortalities
- No specific criteria for conduit interventions
  - May vary across centers
- Data on additional long-term outcomes not recorded
  - RV function, quality of life, neurologic development

Conclusions

- Truncus arteriosus continues to be associated with significant morbidity and mortality
- “Late” mortality predominates within the 1st year of life
- DiGeorge syndrome and preoperative ventilation are risk factors for late mortality – higher surveillance?
- Probability of post-discharge Contegra conduit intervention over time is lower than that of aortic and pulmonary homografts

Thank you for your attention.