CARDIOLOGY 2025

28th Annual Update on Pediatric and Congenital Cardiovascular Disease

NEONATAL EBSTEIN'S ANOMALY

Surgical treatment

Staged Cone Repair

Luciana da Fonseca da Silva, MD

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UPMC Children's Hospital of Pittsburgh –Da Silva Center for Ebstein's Anomaly

Surgical Management and Outcomes of Ebstein Anomaly in Neonates and Infants: A Society of Thoracic Surgeons Congenital Heart Surgery Database Analysis

Kimberly A. Holst, MD, Joseph A. Dearani, MD, Sameh M. Said, MD, Ryan R. Davies, MD, Christian Pizarro, MD, Christopher Knott-Craig, MD, T. K. Susheel Kumar, MD, Vaughn A. Starnes, MD, S. Ram Kumar, MD, PhD, Sara K. Pasquali, MD, MHS, Dylan P. Thibault, MS, James M. Meza, MD, MSc, Kevin D. Hill, MD, MS, Karen Chiswell, PhD, Jeffrey P. Jacobs, MD, and Marshall L. Jacobs, MD

The Society of Thoracic Surgeons Congenital Heart Surgery Database

2010 to 2016 at 95 centers - 239 infants and 255 neonates

NEONATES

- median 7 days
- preoperative ventilation 61.6%
- primary operation
  - Ebstein repair 39.6% (101)
  - Systemic-to-pulmonary shunt 20.4% (52)
  - Tricuspid valve closure 9.4% (24).
  - PA banding 5.5% (12)
  - Other 25%

Overall neonatal operative mortality was 27.4% (n 70) and 9.2% in infants

Mortality for neonates requiring preoperative ECMO was 75.0% (12 of 16)

The Annals of Thoracic Surgery, May 2018

Neonatal Ebstein´s Anomaly
Cone Procedure (N=8)

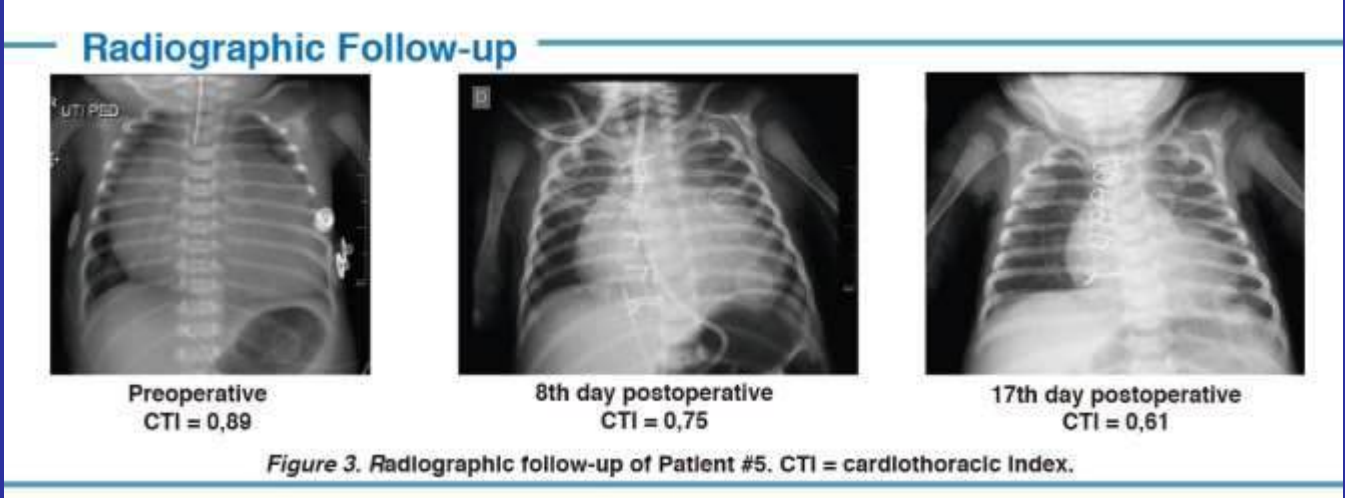


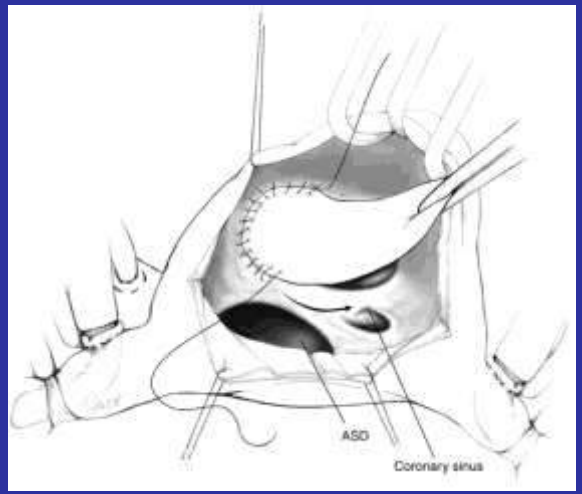
Figure 3. Radiographic follow-up of Patient #5. CTI = cardiothoracic index.

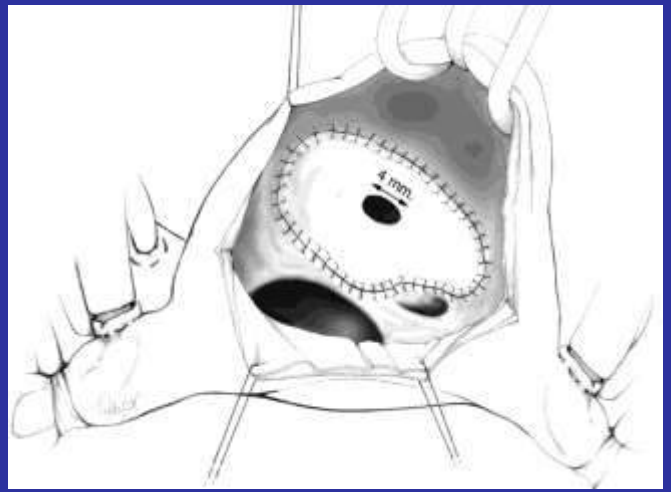
Good results depending on RV function and degree of displacement

AATS Annual meeting 2014, poster presentation. Da Silva et al

Neonatal Ebstein's Anomaly
Starnes (1991) – RV exclusion and BT shunt

UNIVENTRICULAR PHYSIOLOGY





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Reemtsen BL, … Starnes VA; J TCVS 2006;132:1285-9

Neonatal Ebstein's Anomaly

The Starnes procedure is REPRODUCIBLE, and effective early palliative treatment of the neo nate in critical condition. (Univentricular heart)

Before 2019: decision between Biventricular repair x Single ventricle at neonatal period.

The right ventricle undergoes progressive involution after the Starnes procedure[1]

At a point in this reverse remodeling process, the right ventricle might be a suitable pump for a biventricular repair.[2]

1. *Reemtsen BL et al. J Thorac Cardiovasc Surg 2007,134 (6):1410-1412*

2. *Da Silva JP, Viegas M, MD, Castro-Medina M, Da Fonseca Da Silva L. JTCVS Techniques , 2020. 3:281–283*

Neonatal Ebstein's Anomaly
Cone after Starnes

The Da Silva cone operation after the Starnes procedure for Ebstein's anomaly: New surgical strategy and initial results

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JTCVS Techniques 2020;■:1-3

2666-2507

https://doi.org/10.1016/j.xjtc.2020.05.011

The completed Da Silva cone repair after a neonatal Starnes procedure.

CENTRAL MESSAGE

Tricuspid valve repair using the cone technique after a neonatal Starnes procedure is feasible, allowing right ventricle rehabilitation and restoring a biventricular physiology.

▶ Video clip is available online.

JTCVS Techniques 2020. 3:281–283

The Da Silva cone operation after the Starnes procedure for Ebstein's anomaly: New surgical strategy and initial results

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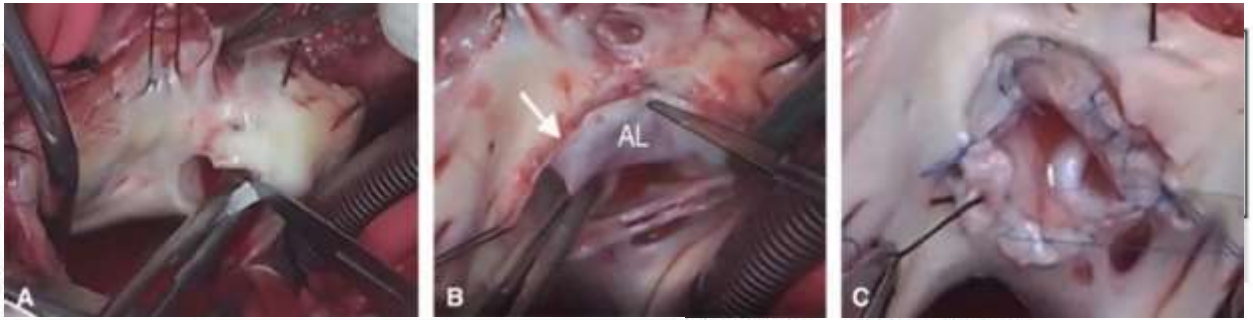
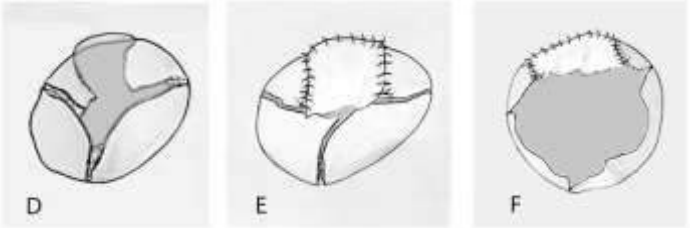




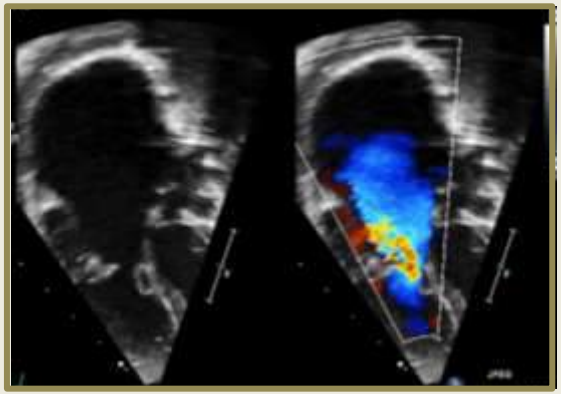
CENTRAL MESSAGE

Tricuspid valve repair using the cone technique after a neonatal Starnes procedure is feasible, allowing right ventricle rehabilitation and restoring a biventricular physiology.

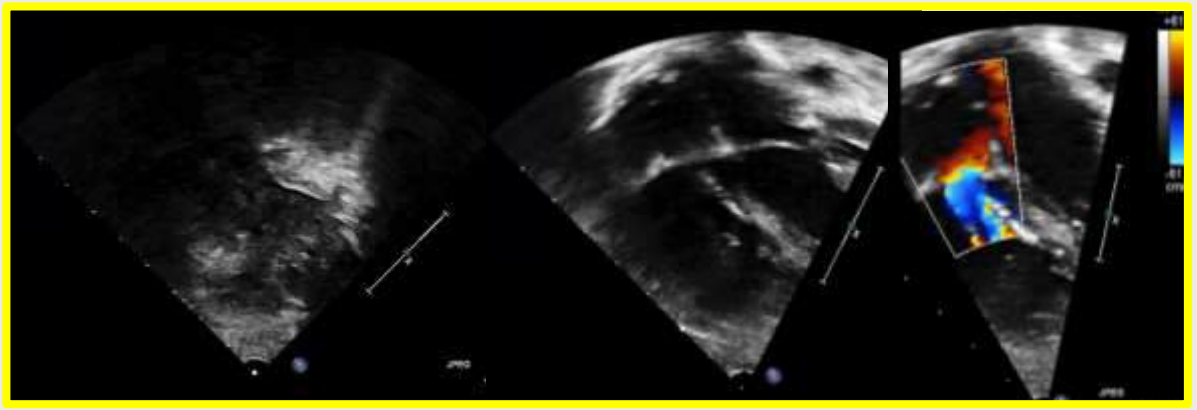
▶ Video clip is available online.

JTCVS Techniques 2020. 3:281–283

Biventricular repair with Cone after Starnes

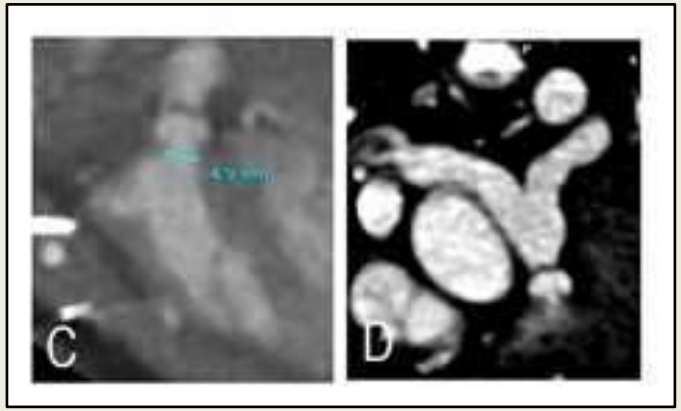


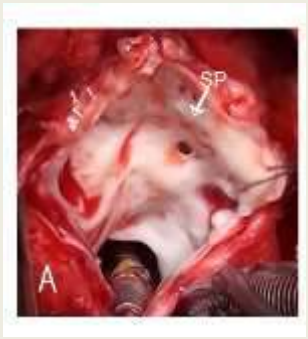
Newborn

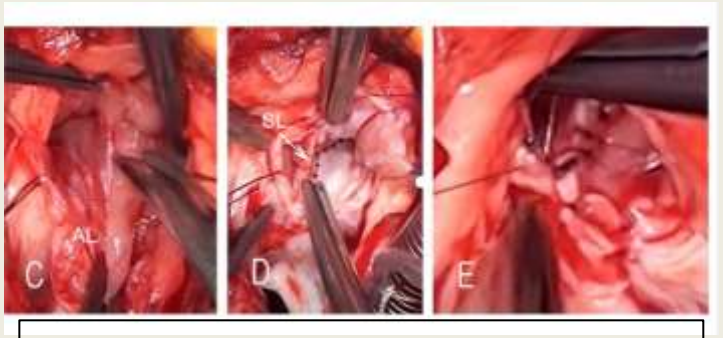


Cone biventricular repair 5 months after ECMO+Starnes

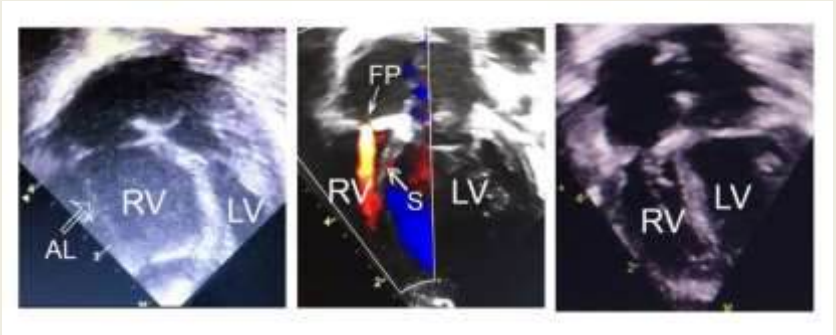
Neonatal circular shunt → Starnes +PA patch → Cone +PA patch removal
5-6 months







CONE + Pulmonary valvoplasty at 5 months

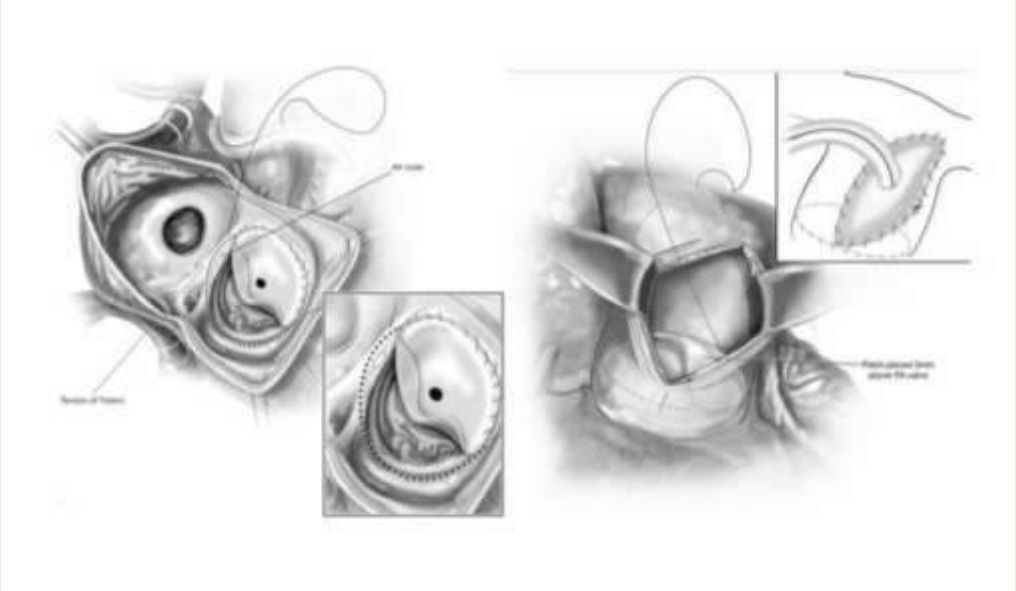


Sequential echocardiograms

Lopes LM, Da Silva JP, Freire R, Da Silva LF.
Cardiology in the young, published on line on 03/8/2021

Surgical details

At Starnes Procedure



Adequate Pulm Valve or PI

Cone at 5-6 mo
Biventricular repair

Avoid:
-heart block
-damage to TV leaflets and PV

Bezerra RF, da Silva JP, da Silva LdF. Op Tech Thorac Cardiovasc Surg 2024
Da Fonseca Da Silva et al. Seminars in Thorac Card Surg 2025, in press

Surgical details

Anatomical Pulm Atresia

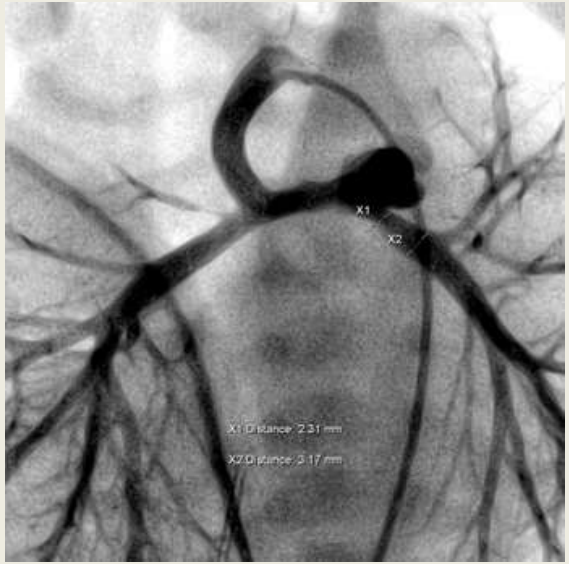
↓

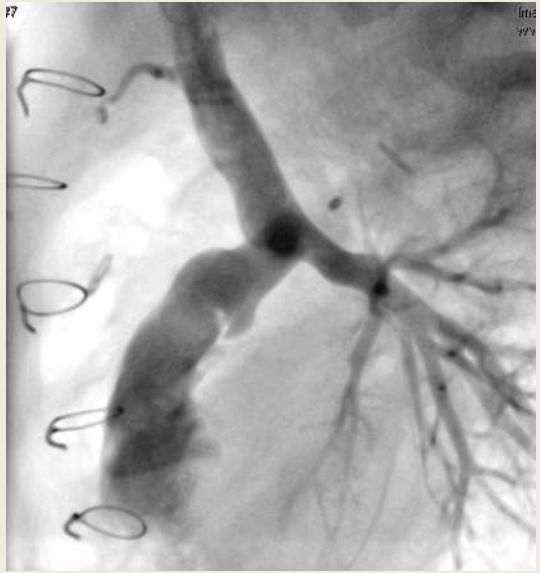
Glenn at 5-6 mo

↓

Cone at 12-18 mo
1 ½ ventricular repair

At Glenn





Pulmonary commissurotomy
-Keep the RV and PV flow/stimulus to grow

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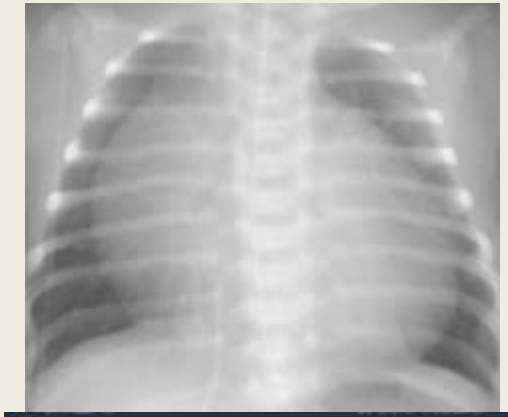
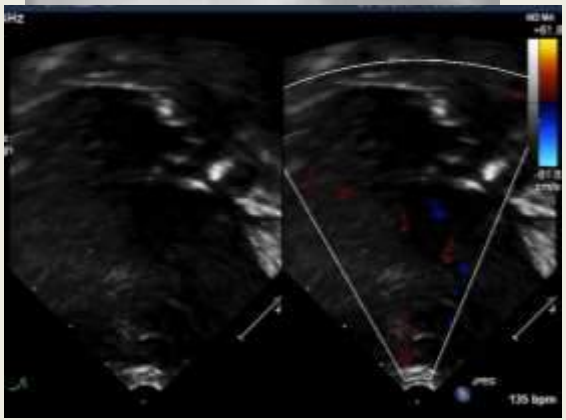
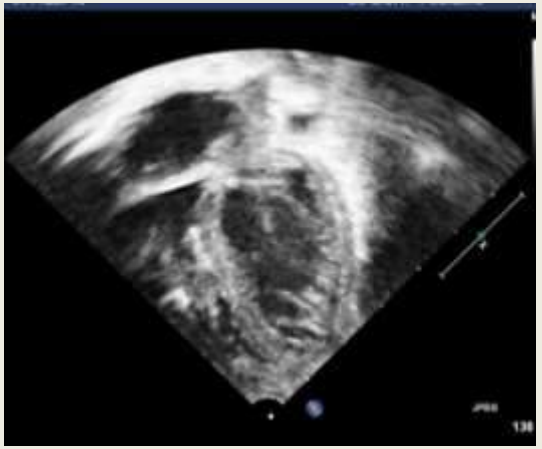
Ebstein's anomaly with membranous pulmonary atresia
BT shunt, ECMO, Starnes, Glenn 3.5 mo, Cone 17mo

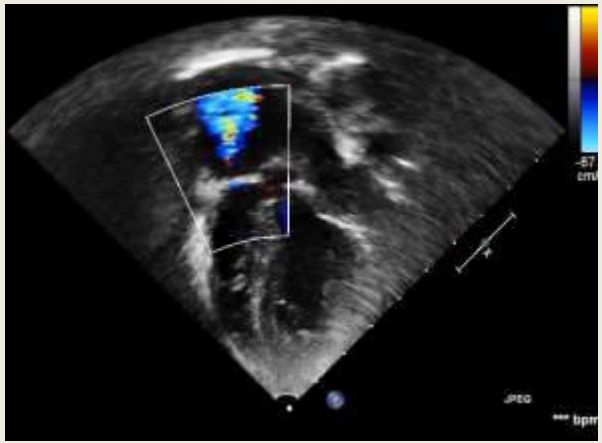


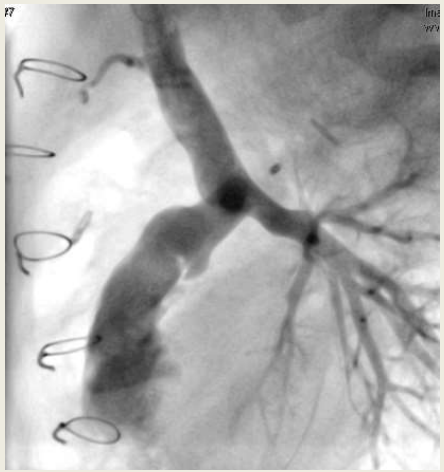


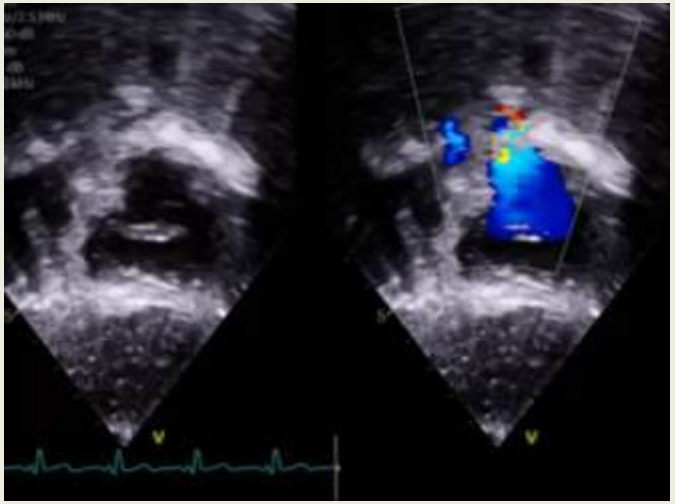


5 weeks after Starnes TR grad 11mmHg

Ebstein's anomaly with membranous pulmonary atresia
BT shunt, ECMO, Starnes, Glenn, Cone







After Glenn 19 mmHg TR gradient

2 years After Cone and PV valvoplasty

The American Association for Thoracic Surgery (AATS) 2024 expert consensus document: Management of neonates and infants with Ebstein anomaly

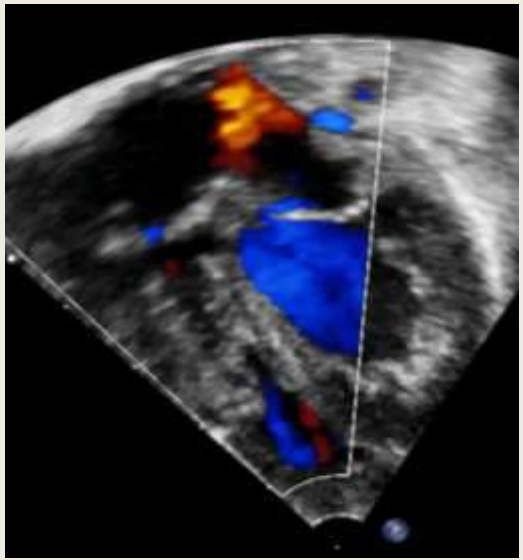
Igor E. Konstantinov, MD, PhD, FRACS,[a,b] Paul Chai, MD,[c] Emile Bacha, MD,[d] Christopher A. Caldarone, MD,[e] Jose Pedro Da Silva, MD,[f] Luciana Da Fonseca Da Silva, MD,[f] Joseph Dearani, MD,[g] Lisa Hornberger, MD,[h] Christopher Knott-Craig, MD, FACS,[i] Pedro del Nido, MD,[j] Muhammad Qureshi, MD,[k] George Sarris, MD,[l] Vaughn Starnes, MD,[m] and Victor Tsang, MBBS[n]

Results: When evaluating fetuses with EA, those with severe cardiomegaly, retrograde or bidirectional shunt at the ductal level, pulmonary valve atresia, circular shunt, left ventricular dysfunction, or fetal hydrops should be considered high risk for intrauterine demise and postnatal morbidity and mortality. Neonates with EA and severe cardiomegaly, prematurity (<32 weeks), intrauterine growth restriction, pulmonary valve atresia, circular shunt, left ventricular dysfunction, or cardiogenic shock should be considered high risk for morbidity and mortality. Hemodynamically unstable neonates with a circular shunt should have emergent interruption of the circular shunt. Neonates in refractory cardiogenic shock may be palliated with the Starnes procedure. Children may be assessed for later biventricular repair after the Starnes procedure. Neonates without high-risk features of EA may be monitored for spontaneous closure of the patent ductus arteriosus (PDA). Hemodynamically stable neonates with significant pulmonary regurgitation at risk for circular shunt with normal right ventricular systolic pressure should have an attempt at medical closure of the PDA. A medical trial of PDA closure in neonates with functional pulmonary atresia and normal right ventricular systolic pressure (>20-25 mm Hg) should be performed. Neonates who are hemodynamically stable without pulmonary regurgitation but inadequate antegrade pulmonary blood flow may be considered for a PDA stent or systemic to pulmonary artery shunt.

1.Konstantinov, Igor E. et al. JTCVS 2024.168(2), 311 - 324

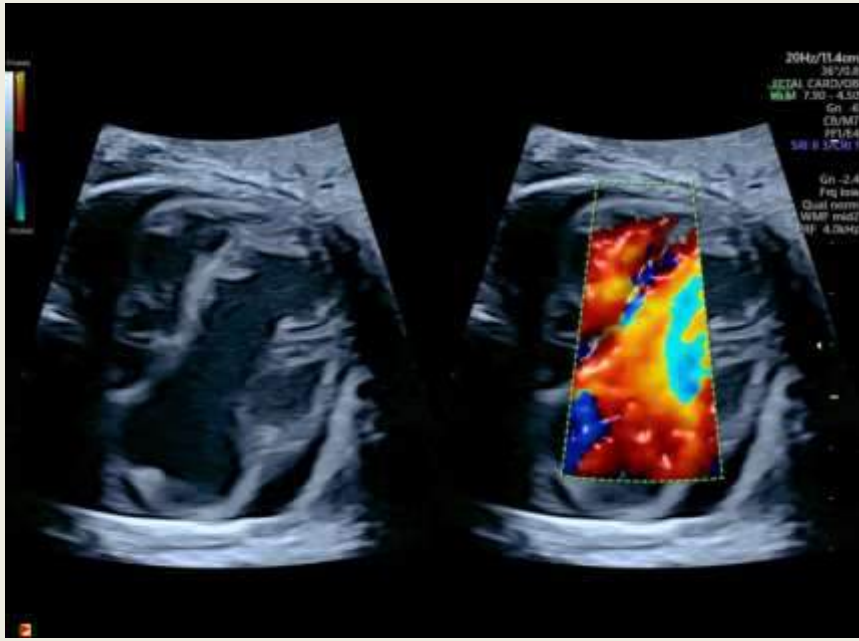
Pre-Cone Evaluation

Echocardiogram



After Starnes

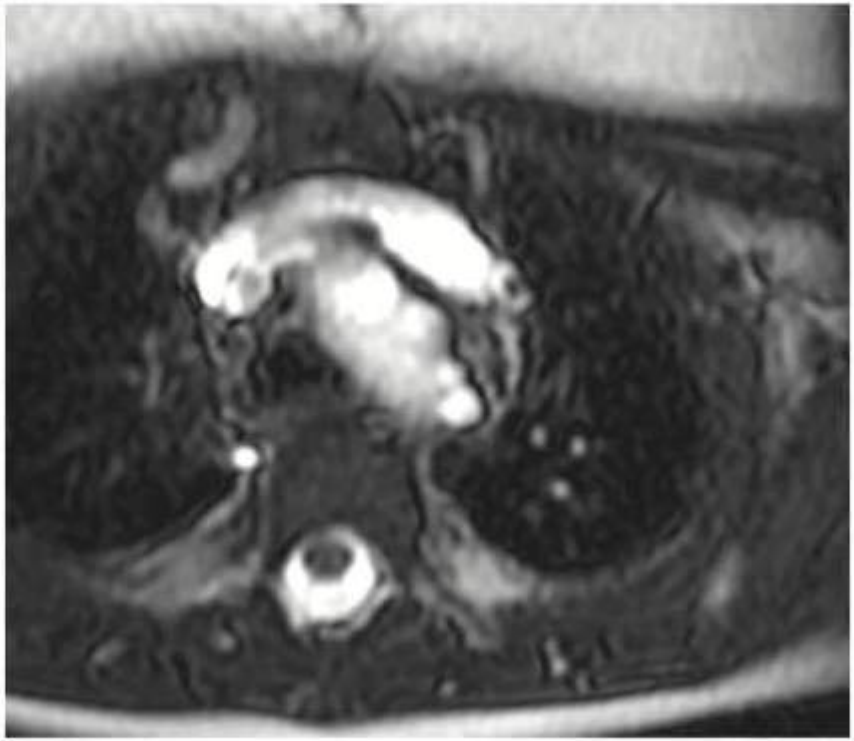
Neonatal



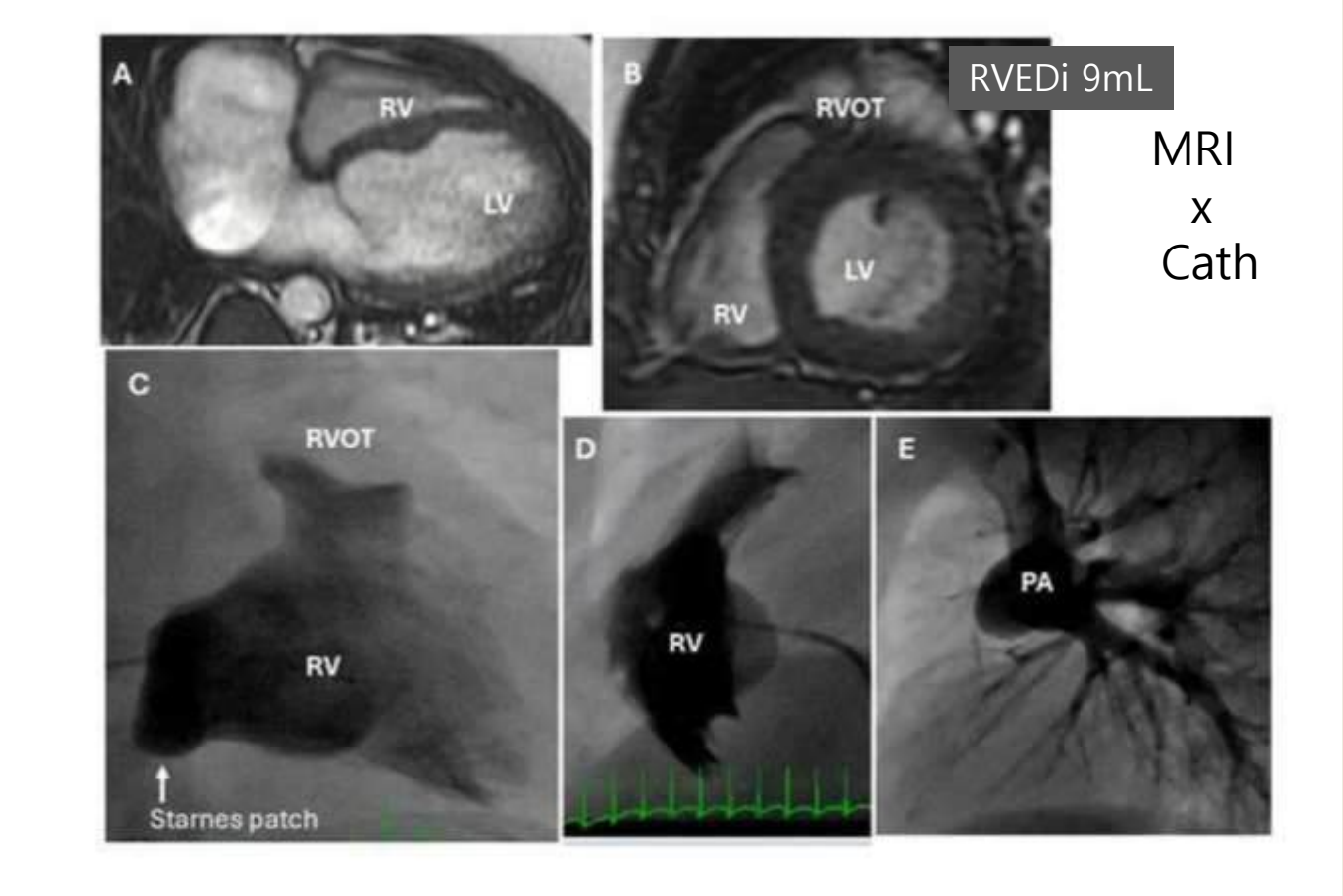
Fetal

Pre Cone Evaluation

MRI



Pre Cone Evaluation



Da Fonseca Da Silva et al. Seminars in Thorac Card Surg 2025, in press

The cone repair allows right ventricle rehabilitation with excellent tricuspid valve function following the Starnes procedure

Syed Faaz Ashraf, MD,[a] Jose Pedro Da Silva, MD,[a] Mario Castro-Medina, MD,[a] Melita Viegas, MD,[a] Tarek Alsaied, MD,[b] Laura Seese, MD,[c] Victor O. Morell, MD,[a] and Luciana Da Fonseca Da Silva, MD[a]

TABLE 5. Postoperative imaging

			Discharge echocardiography		Follow-up echocardiography			
Patient	Anatomy	Valves	RV function	TR	Time since surgery, mo	TR (late)	RV function	LVEF, %
1	EA + PA	1.5	Moderately depressed	Trivial	27	Mild	Moderately depressed	75
2	EA + PA	2	Normal	Mild	53	Trivial	Normal	66
3	EA + PA	1.5	Normal	Mild	43	Mild	Normal	58
4	EA + PVS	1.5	Severely depressed	Mild	16	Trivial	Severely depressed	53
5	EA + severe PI	1.5	Severely depressed	Trivial	17	Mild	Moderately depressed	58
6	EA	2	Mildly depressed	Trivial	N/A	N/A	N/A	N/A
7	E + PA	1.5	Normal	Trivial	N/A	N/A	N/A	N/A
8	EA + moderate PI	2	Normal	Trivial	N/A	N/A	N/A	N/A
9	EA + severe PVS + VSD	2	Normal	Moderate	N/A	N/A	N/A	N/A
10	EA	2	Normal	Trivial	N/A	N/A	N/A	N/A
11	EA + PA	1.5	Normal	Trivial	N/A	N/A	N/A	N/A

RV, Right ventricle; *TR*, tricuspid regurgitation; *LVEF*, left ventricular ejection fraction; *EA*, Ebstein anomaly; *PA*, pulmonary atresia; *PVS*, pulmonary valve stenosis; *PI*, pulmonary valve insufficiency; *N/A*, not available; *VSD*, ventricular septal defect.

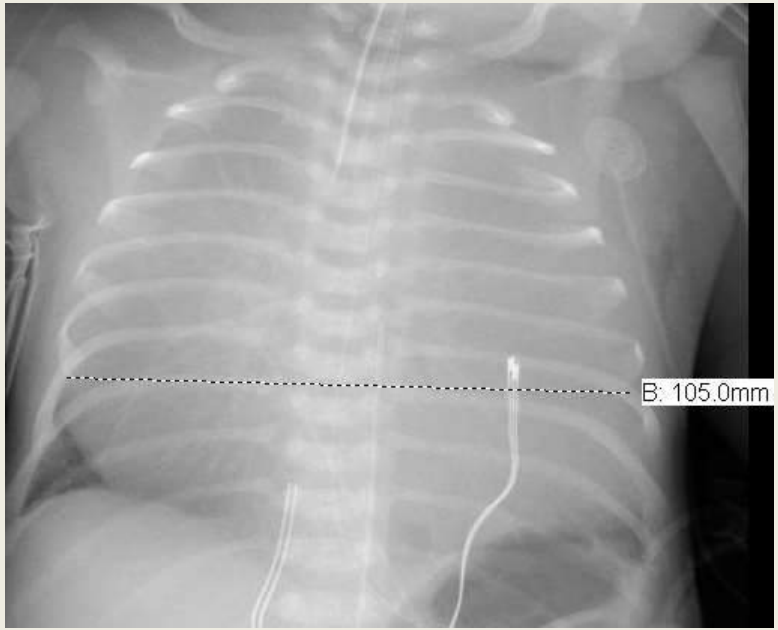
J Thorac Cardiovasc Surg 2025. 169(2) 354-61

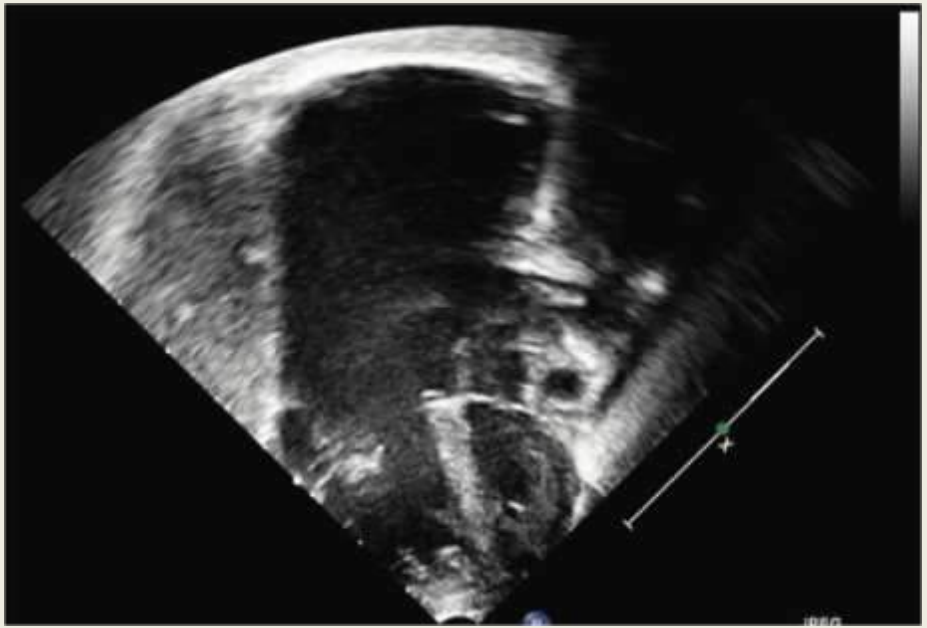
OUR CURRENT EXPERIENCE
18 sequential cases- Cone after Starnes

				Echo	Cardiac catheterization			Cardiac MRI		Discharge Echo	
Patient	Initial Anatomy	Age (mo)	Glenn Age	Veloc(m/s)	RVSp (mmHg)	PVR(wu x m2)	O2 Sat	RVEDVi(ml/m2)	RV EF %	RV function	Tricusp Regurg
1	EA+PA	17	3	2.1	24/5	1.5	81	Not done	Not done	Mod depr	Trivial
2	EA+PA	5	Not done	2.6	42/8	0.7	76	35	31	Normal	Mild
3	EA+PA	29	4	4	Not done	2.0	79	55	10	Normal	Mild
4	EA+PVS	61	9	1.1	11/6	0.9	78	105	12	Severe depr	Mild
5	EA+PI Severe	33	4	0.7	13/6	1.2	77	Not done	Not done	Severe depr	Trivial
6	EA	27	Not done	1.7	12/1	3.3	82	Not done	Not done	Mild depr	Trivial
7	EA + PA	41	6	2.8	25/6	2.0	80	24	42	Normal	Trivial
8	EA + PI Moder.	18	11	1.6	11/4	2.1	73	22	34	Normal	Trivial
9	EA+PS+VSDs	16	Not done	Non-fenest	80/9	1.9	71	Not done	Not done	Normal	Moderate
10	EA	8	Not done	1.4	N/A	2.8	88	Not done	Not done	Normal	Trivial
11	EA+PA	55	4	0.9	28/8	1.5	82	Not done	Not done	Normal	Trivial
12	EA + PI	26	13	2	Not done	2.1	78	CT scan normal RV	Not done	Mild depr	Mild
13	EA +PI	28	5	2	Not done	1.79	76	Not done	Not done	Normal	Mild
14	EA+ PS suprav	96	7 + Fontan	1.4	Not done	2.49	92	25	10	Normal	Mild
15	EA + PA memb	28	7	1.3	Not done	0.9	77	Not Done	Not done	Mod depr	Mod/severe
16	EA+ PA memb	22	7	2.5	47/6	2.68	73	Not done	Not done	Mild depr	Mild/mod
17	EA+ PI Severe	19	5	2.6	25/4	0.82	79	20	Not done	Mild depr	Mild
18	EA+ PI Severe	15	8	1.2	15/3	1.7	79	23	24	Normal	Mild

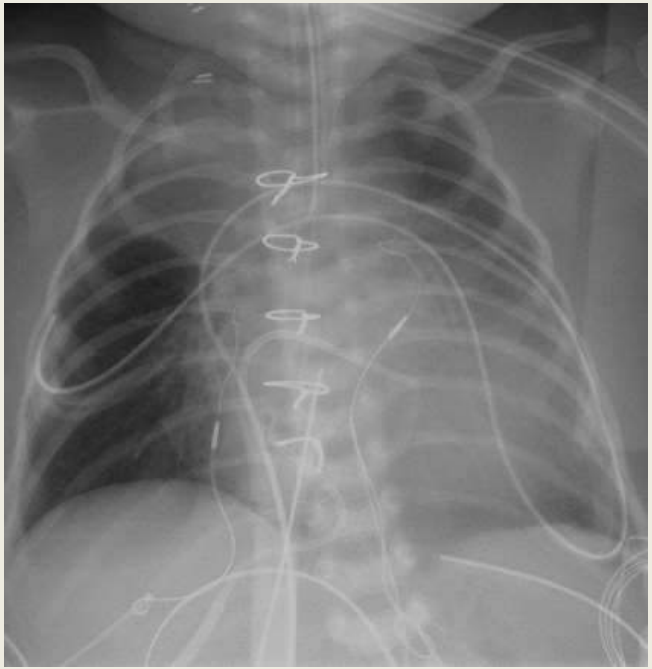
Da Fonseca Da Silva et al. Seminars in Thorac Card Surg 2025, in press

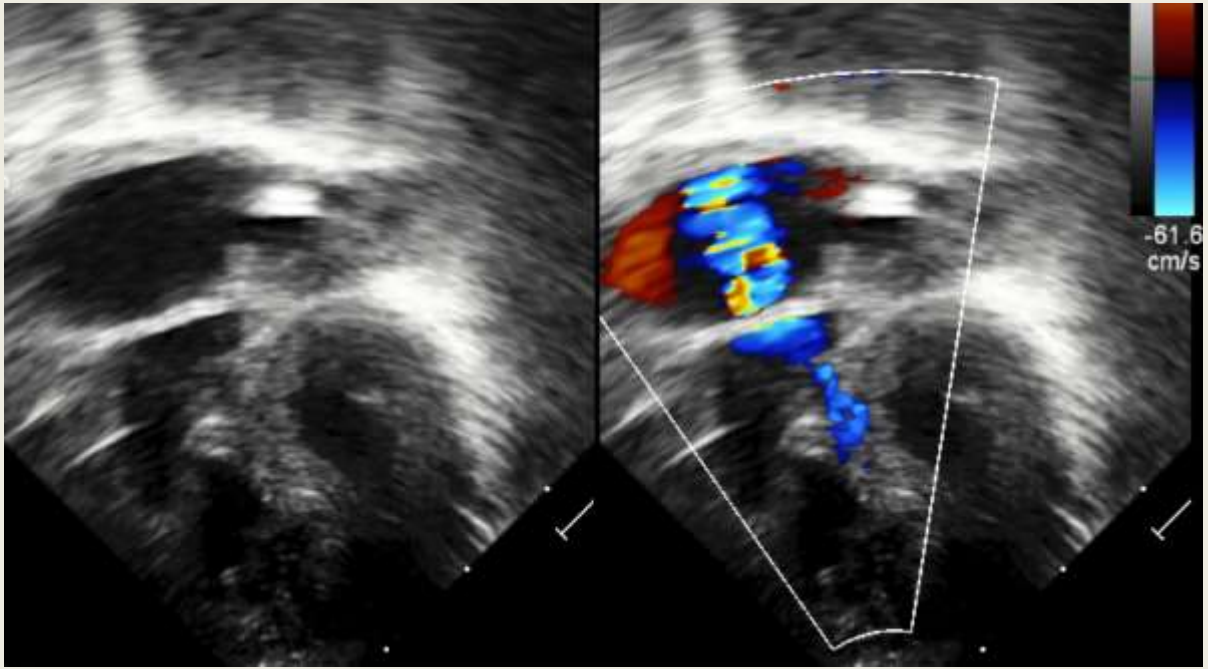
Newborn with severe EA and PA



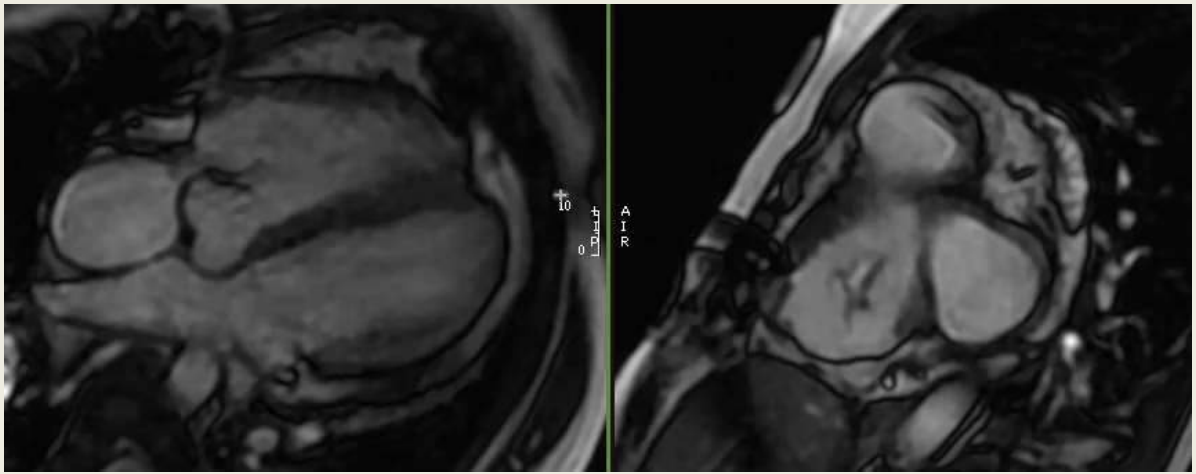


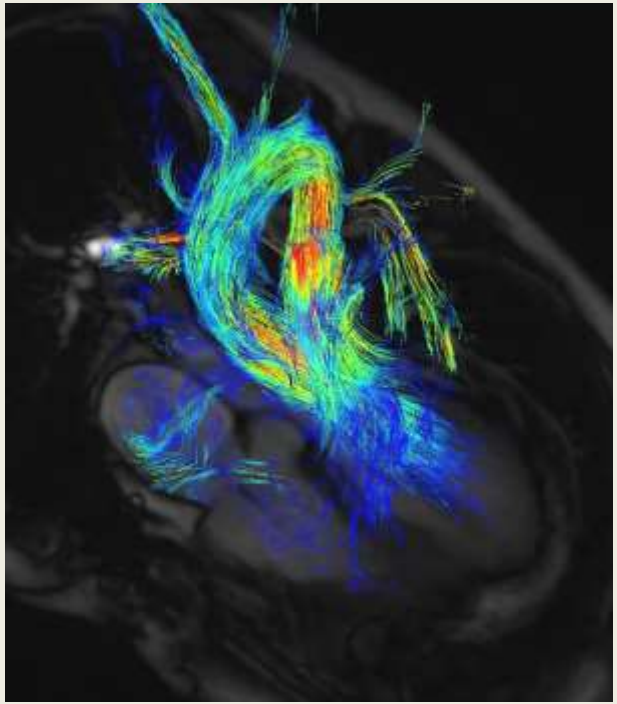
1 wk s/p Starnes, RA reduction, BTTS





Six-year-old, good clinical condition, attending kindergarten





NEONATAL EBSTEIN'S ANOMALY
Summary

The right ventricle rehabilitation with the Cone repair after the Starnes proce dure provides excellent clinical outcomes, encouraging the use of the Starnes procedure on critically ill Ebstein's anomaly neonates.

Preserving the pulmonary and tricuspid valves during the neonatal interventi on is essential to facilitate the subsequent steps in this staged approach.

Thank you!

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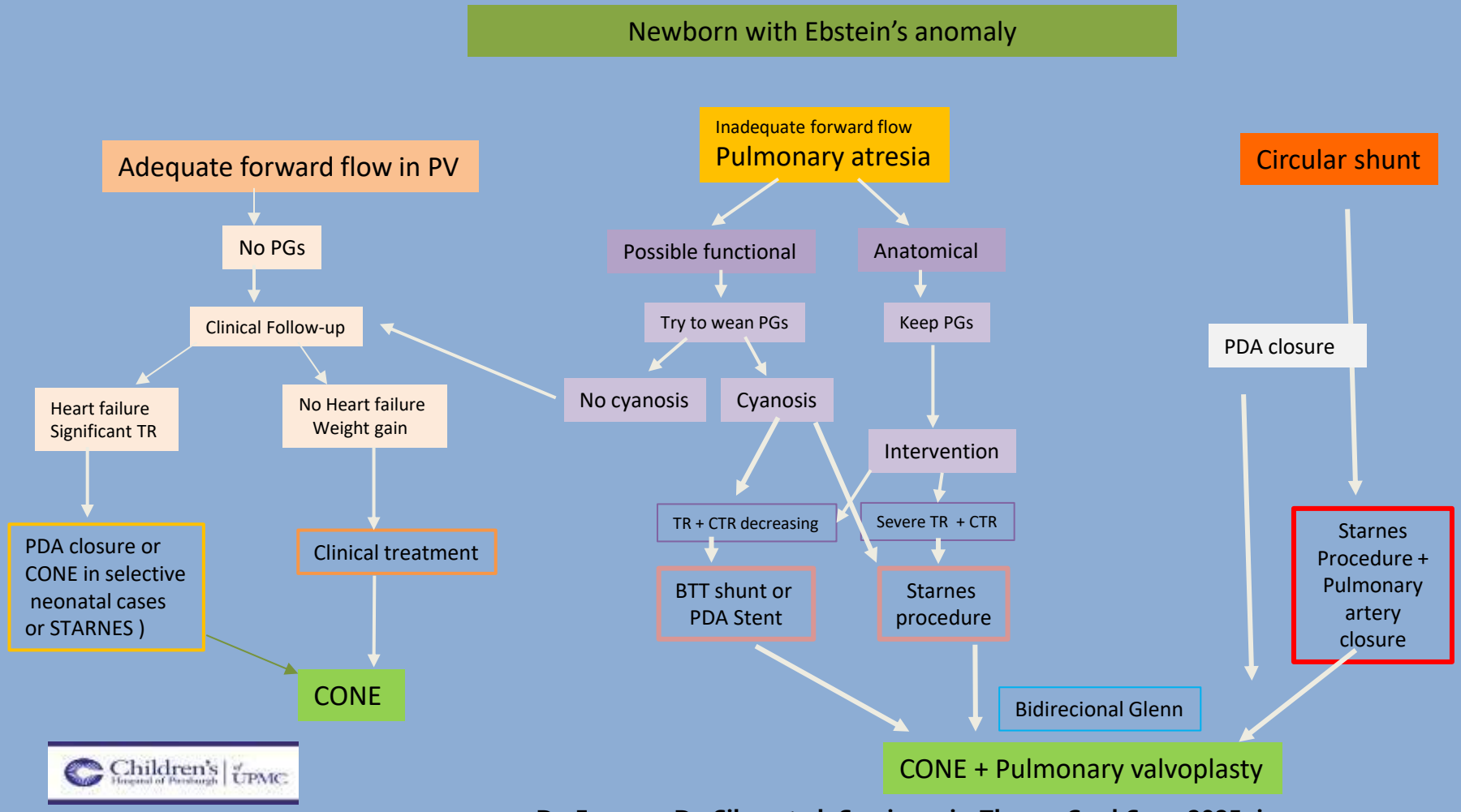
UPMC | CHILDREN'S HOSPITAL OF PITTSBURGH

heart institute



Da Fonseca Da Silva et al. Seminars in Thorac Card Surg 2025, in press

Fetal diagnosis of Ebstein's anomaly

If Circular shunt: DA closure NSAIDs transplacentary

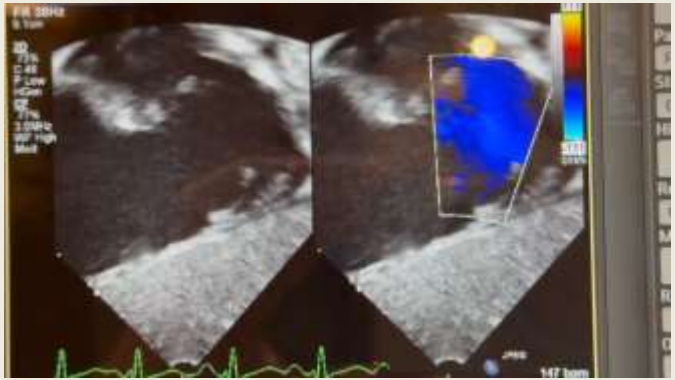
Newborn with Ebstein's anomaly

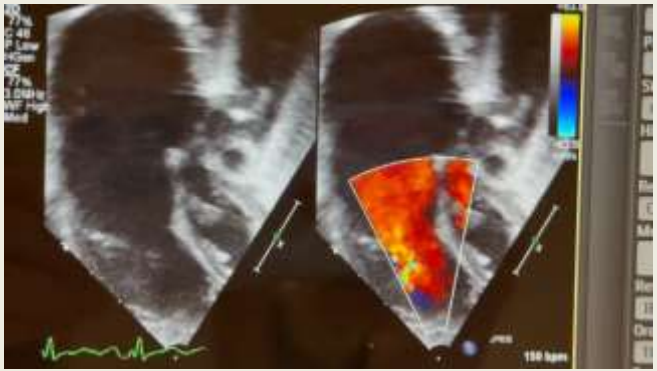
Clinically stable:
Avoid interventions
-Elevated pulmonary vascular resistance
-Risks of AV valve surgery in children
< 3 months and <3,5 Kg.

Unstable:
-Entubation/Ventilation
-***Prostaglandin*** (PDA dependent + reduction of Pulmonary vascular resistance)
-Nitric Oxide
-Inotropic support – Milrinone
-Rhythm control
-ECMO

Children's Hospital of Pittsburgh | of UPMC

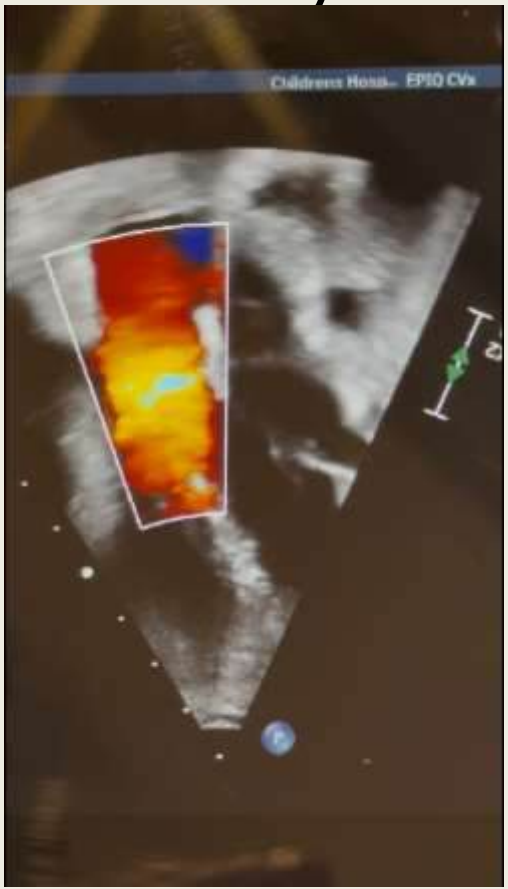
FOLLOW-UP



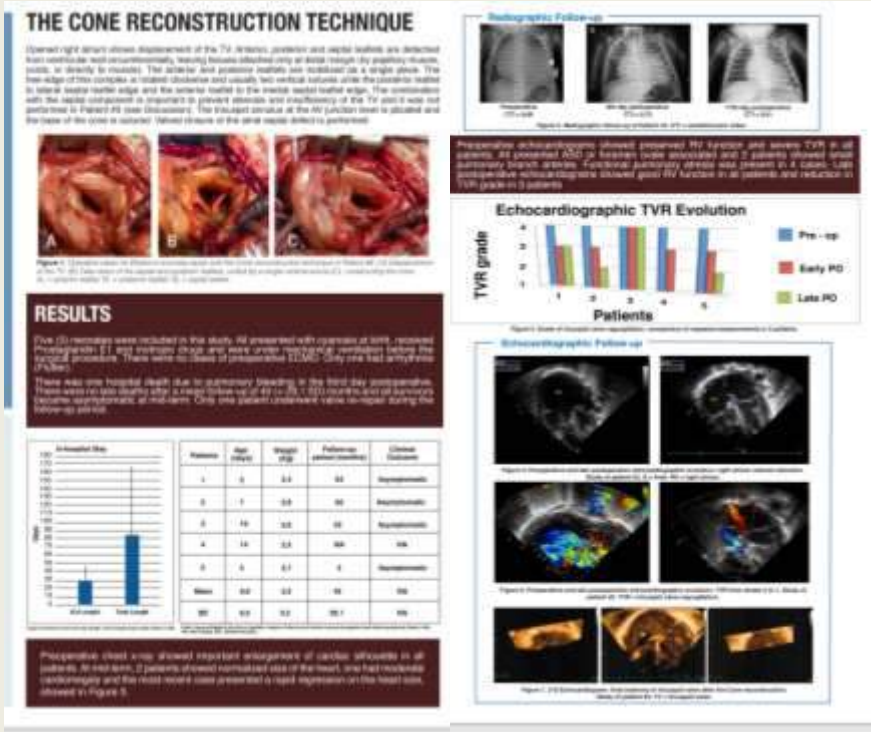


Feb 2019

After 6 years



Cone procedure applied to 5 Newborns – AATS 2014



The Cone Technique for anatomic repair of Ebstein's anomaly of the tricuspid valve in newborn infants: patient selection criteria.

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Hospital Beneficência Portuguesa - São Paulo, Brazil.

CONCLUSION

Although the optimal surgical management of critically ill neonates with Ebstein's anomaly of the TV remains controversial, the Cone reconstruction technique can be used with good short and mid-term results if there is favorable TV anatomy and reasonable RV function. For patients who do not attend to these criteria, other surgical intervention must be performed.

Neonatal Ebstein's anomaly Treatment

Ebstein's Anomaly: Surgical Treatment of 9 Neonates
UPMC Children's Hospital of Pittsburgh, 2016 - 2021

Procedure	Procedures	Special Characteristics	Death/Reoperation
Primary Neonatal Cone Repair	2	1 in preop ECMO	0/1
Neonatal Starnes	4	2 in preop ECMO	0/0
Cone repair after Starnes procedure	3	Age: 5 to 17 months 2 BD Glenn	0/0
Neonatal PDA stenting	2	1-Cone done at 6 months 1-LVnoncompaction - death	0/1 1/x
Neonatal medical management	1	Received the cone repair at 3-yo	0