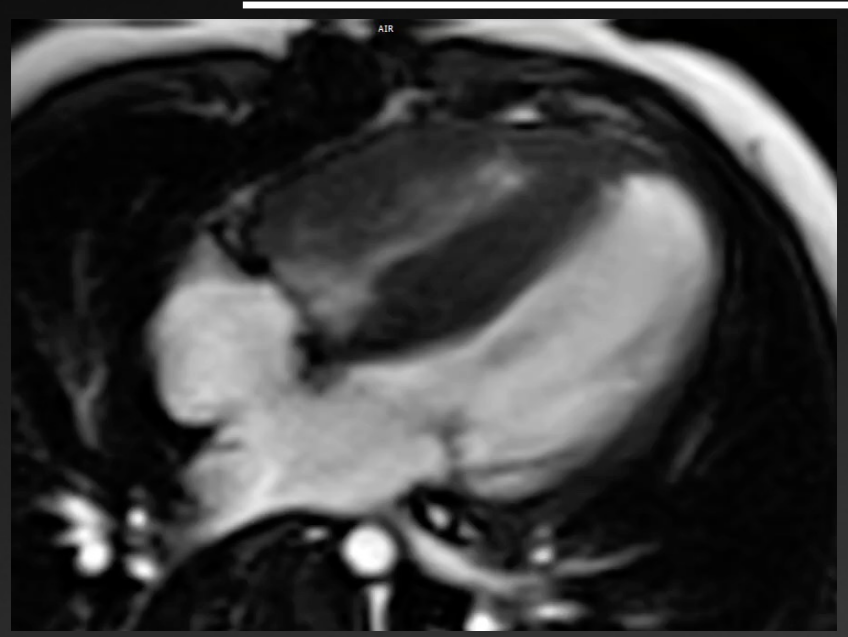
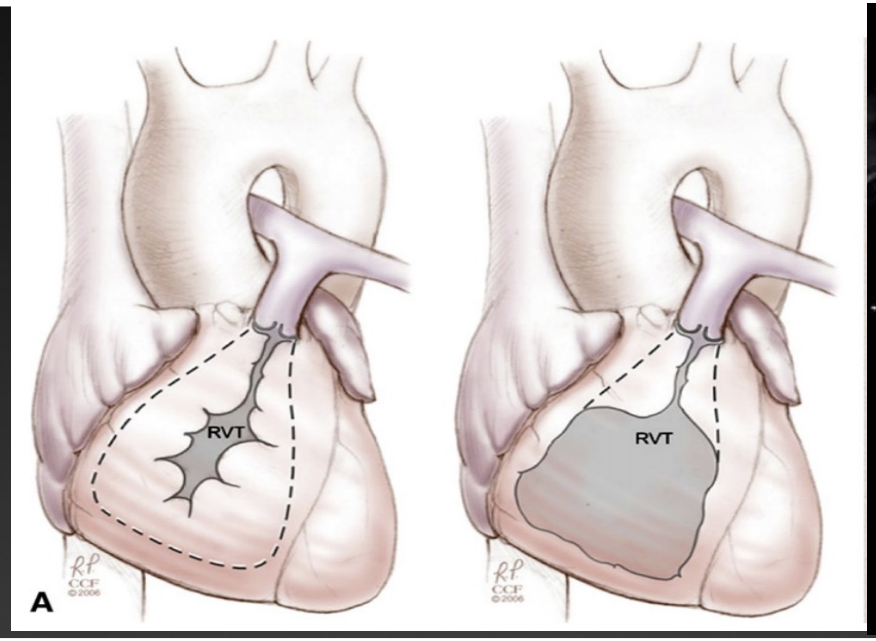
The Borderline Right Ventricle:
determining adequacy of the right heart





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No disclosures

CHOP Cardiology 2023

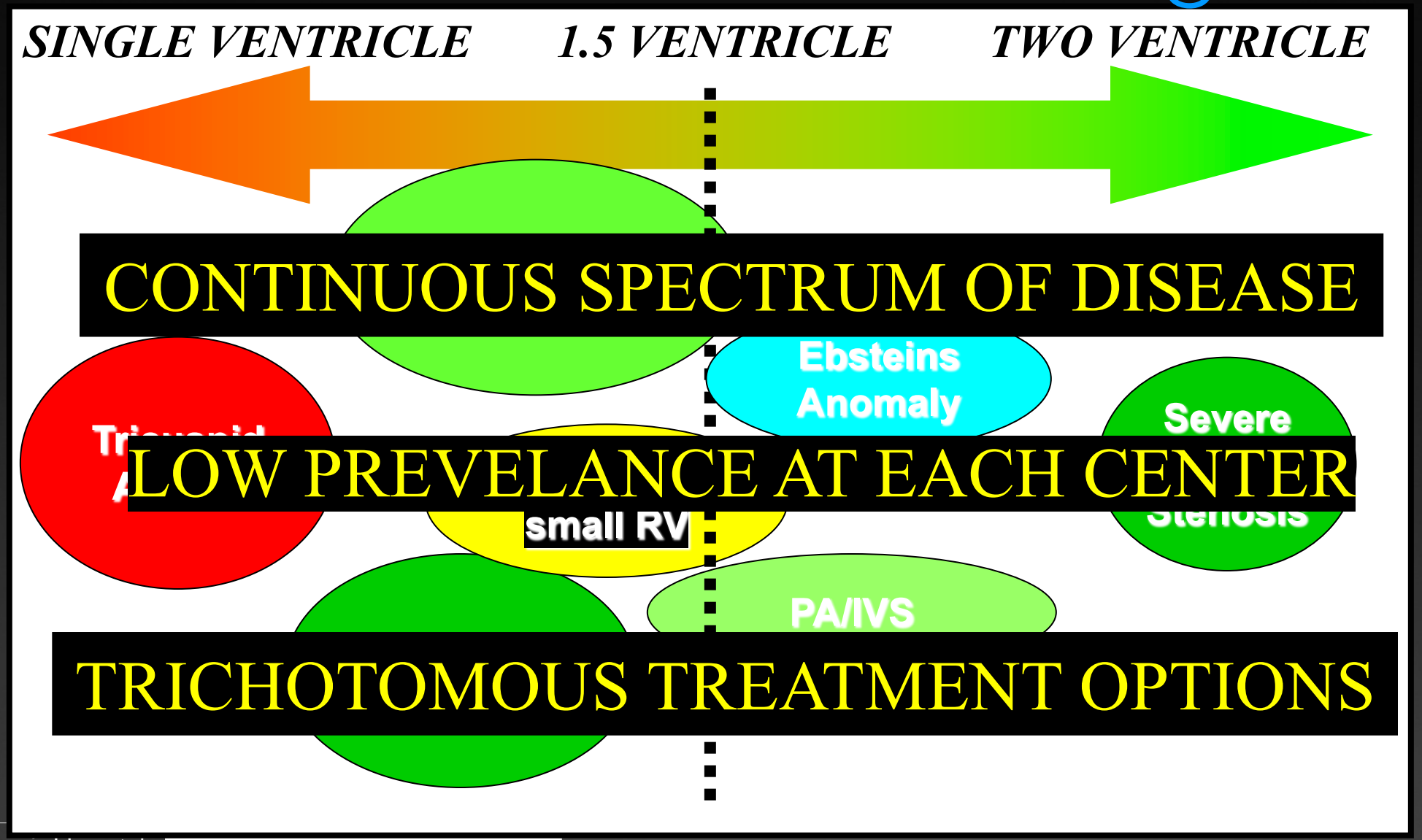
Borderline RV: Overview

1. How do we define the borderline right ventricle?

2. What data do we have to support decision making?

3. How do we determine the optimal surgical pathway?

Borderline RV: the challenges



Differences: small wins

Borderline RV

RVOTO can easily be addressed surgically – RV-PA conduit

PR from RVOT reconstruction well tolerated in childhood

Partial repair (1.5V) is an option for the real borderline cases

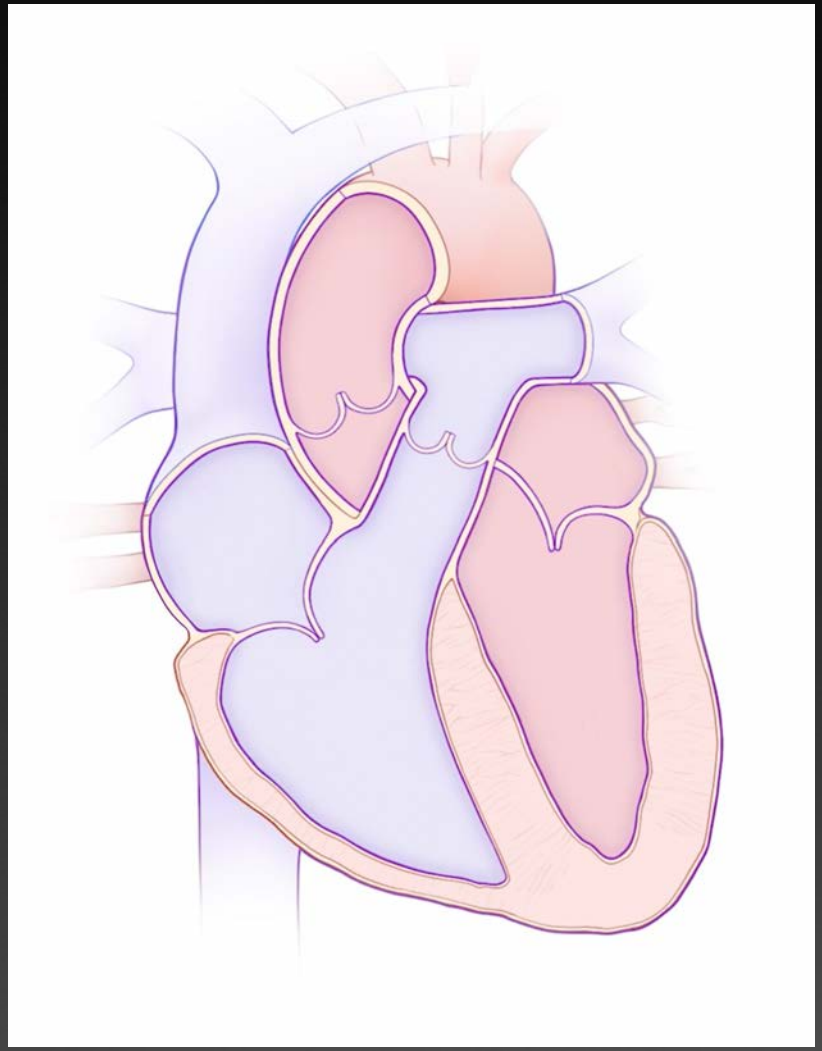
Borderline LV

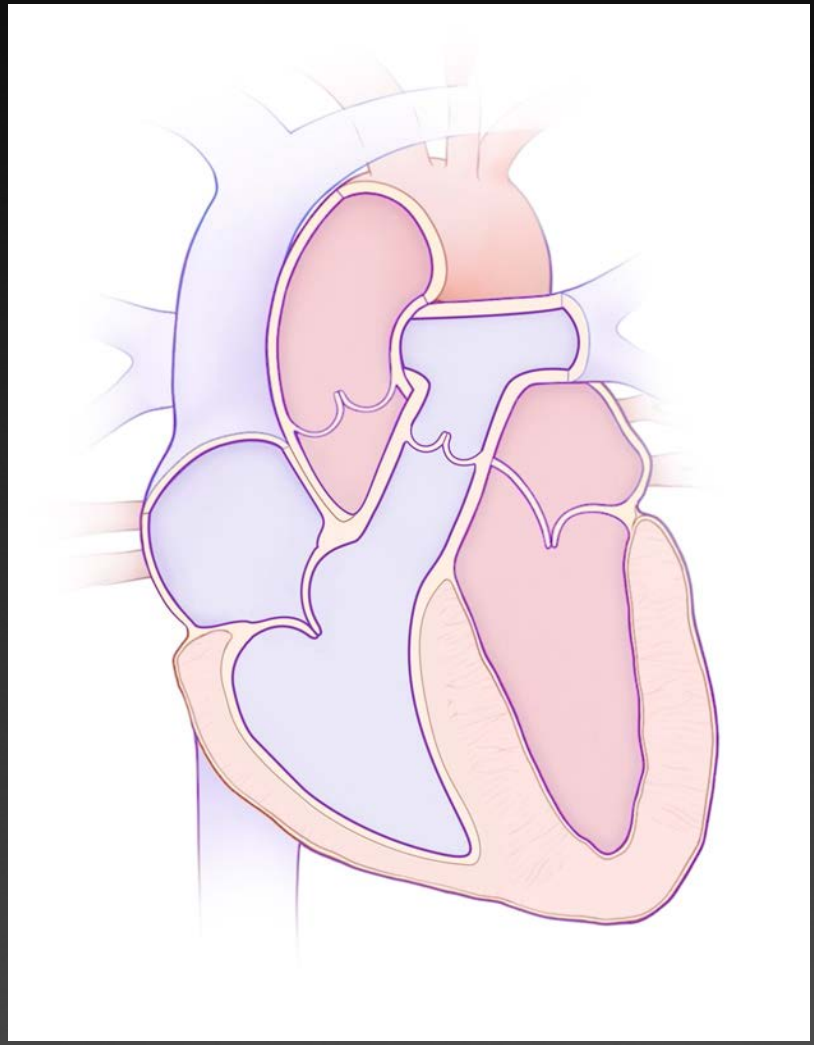
LVOTO much more complex to address (Ross-Kono)

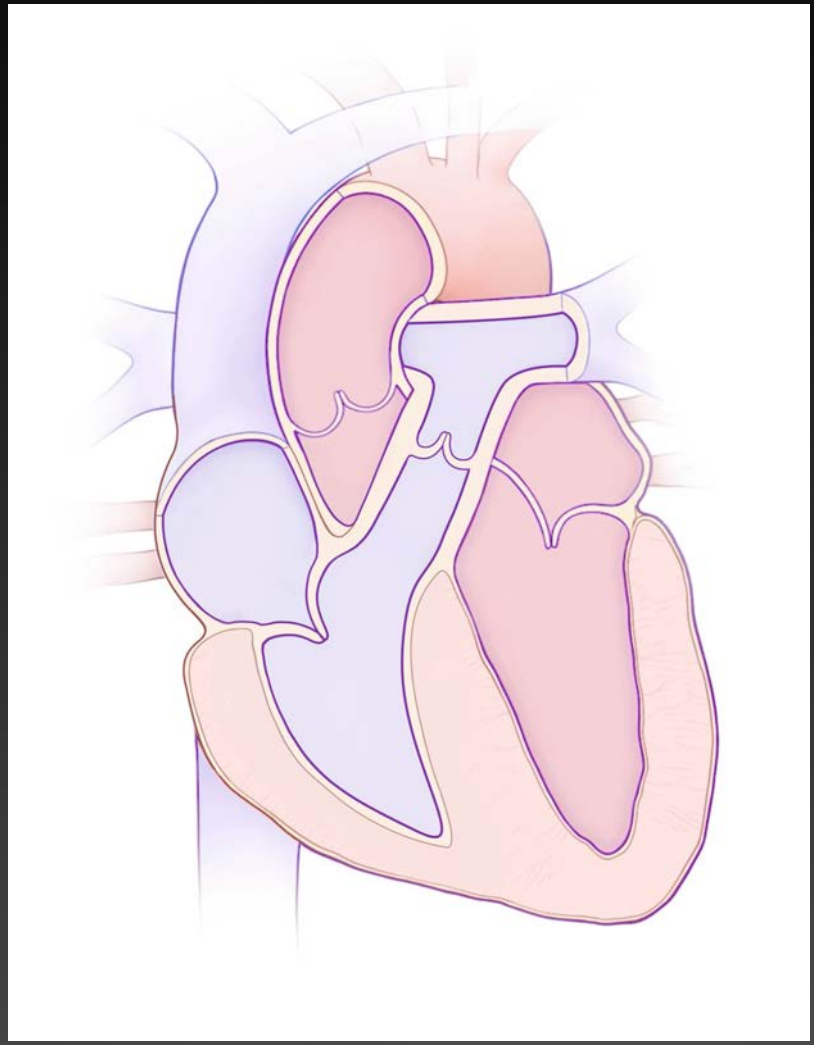
AR from LVOT reconstruction not well tolerated

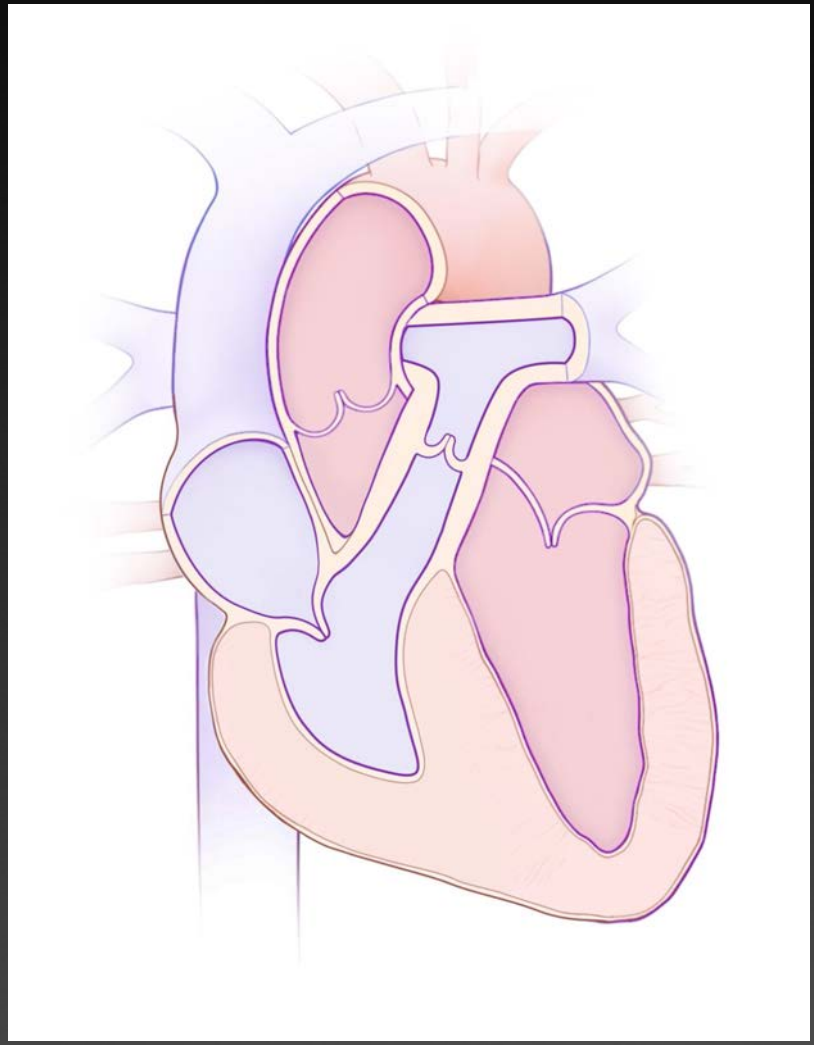
Dichotomous surgical pathways, all or nothing…

What are the morphologic and physiologic criteria we must consider for decision making ?







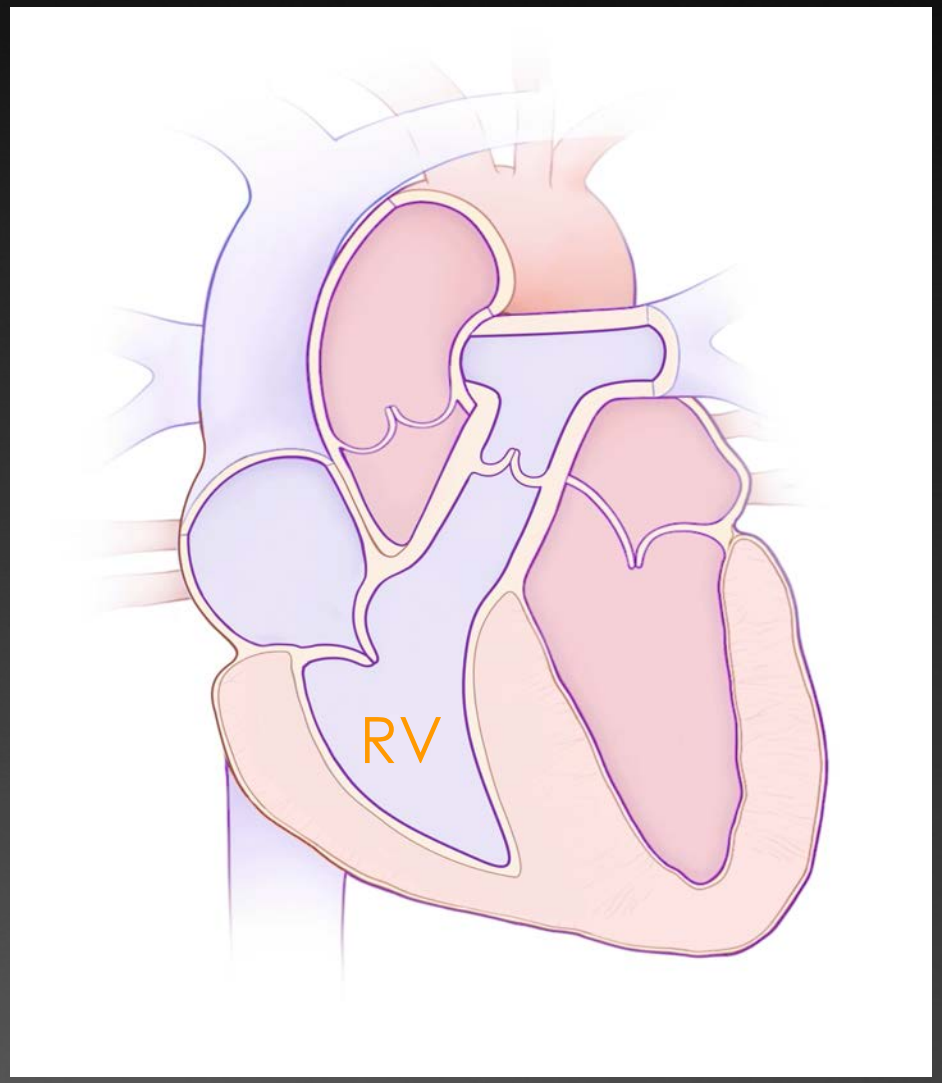


Right Ventricle Characteristics

1. Ventricular End Diastolic Volumes

2. RV morphology (? tripartite, presence of infundibulum)

3. RV systolic function



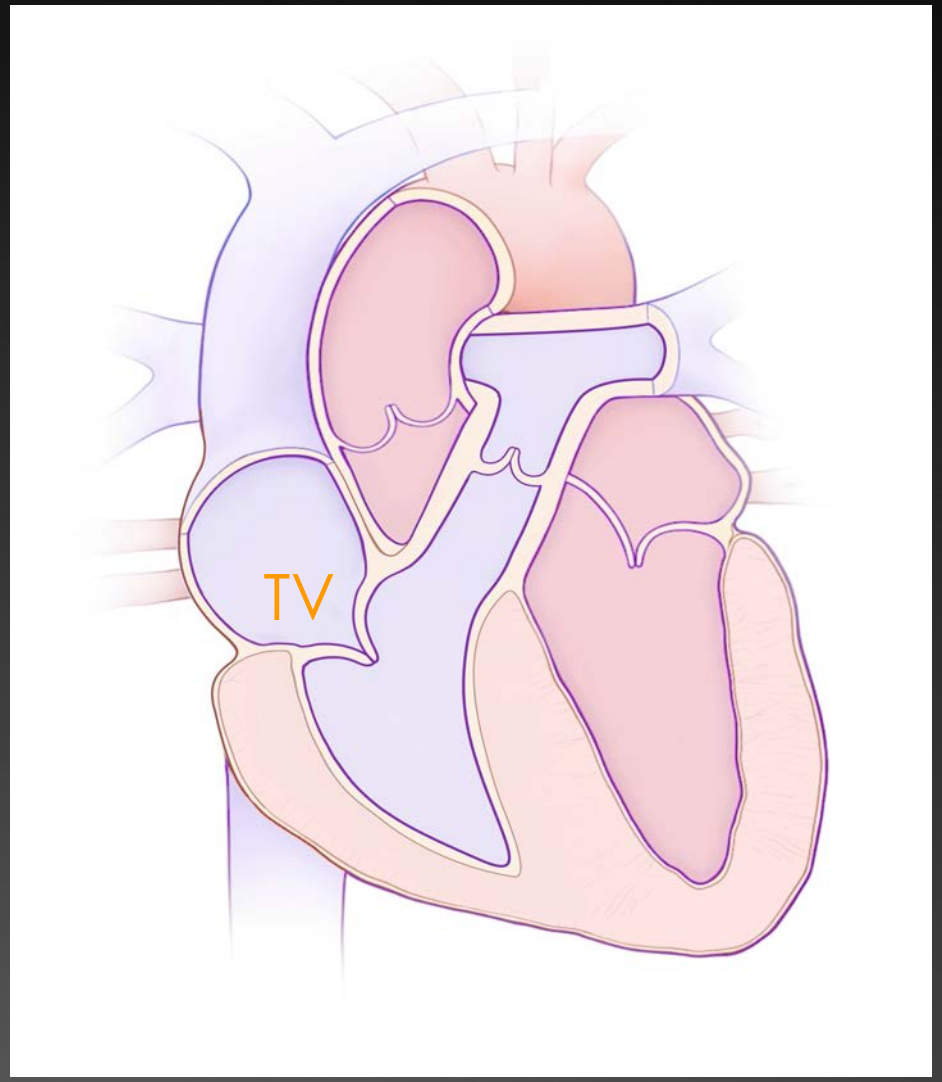
Tricuspid Valve Characteristics

1. Annular size/ Z-score

2. Morphology/degree TR

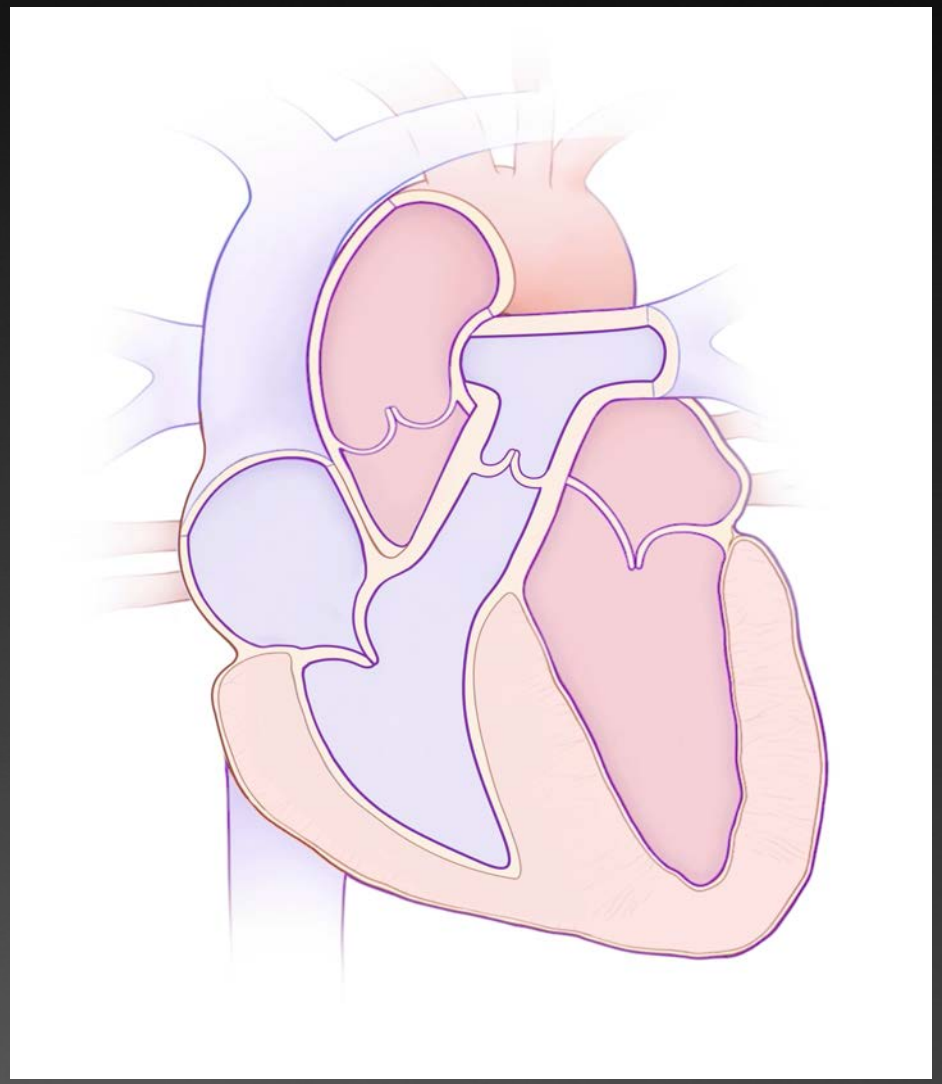
3. Inflow potential in uAVSD

4. Chordal attachments STV



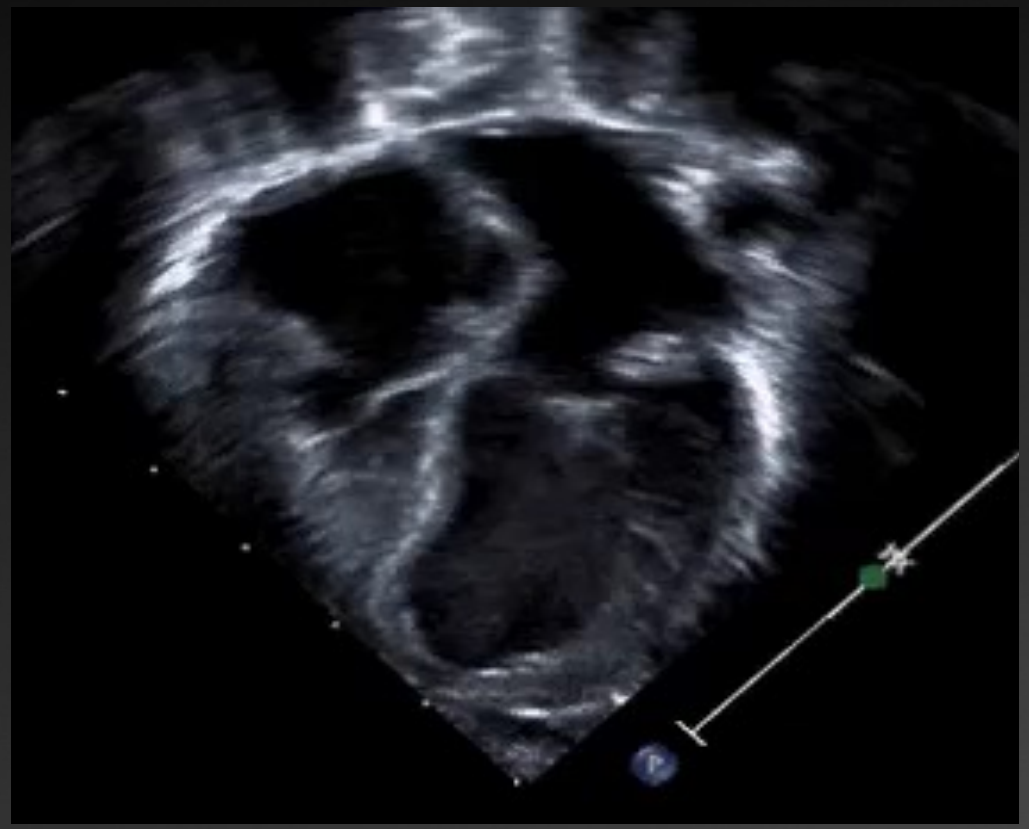
What other factors ?

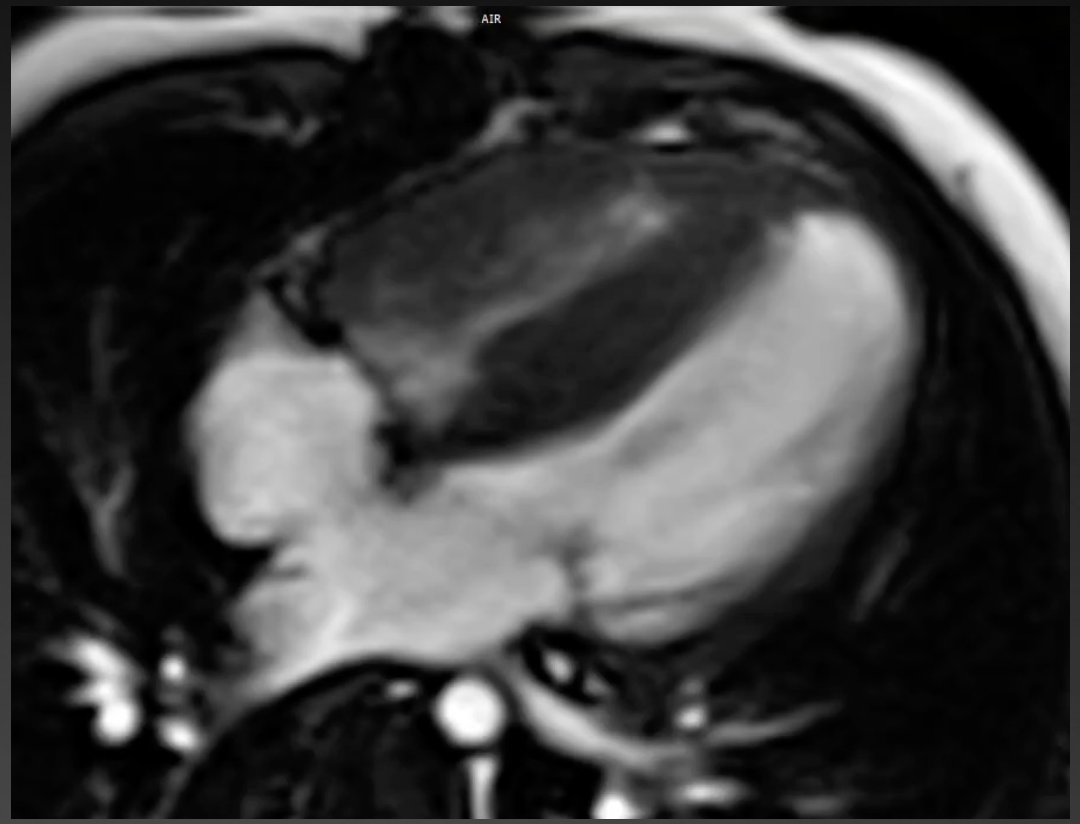
- Coronary abnormalities (PA-IVS)
- Ability of the ventricles be separated
- PVR



What data exists to guide decision making?

PA/IVS





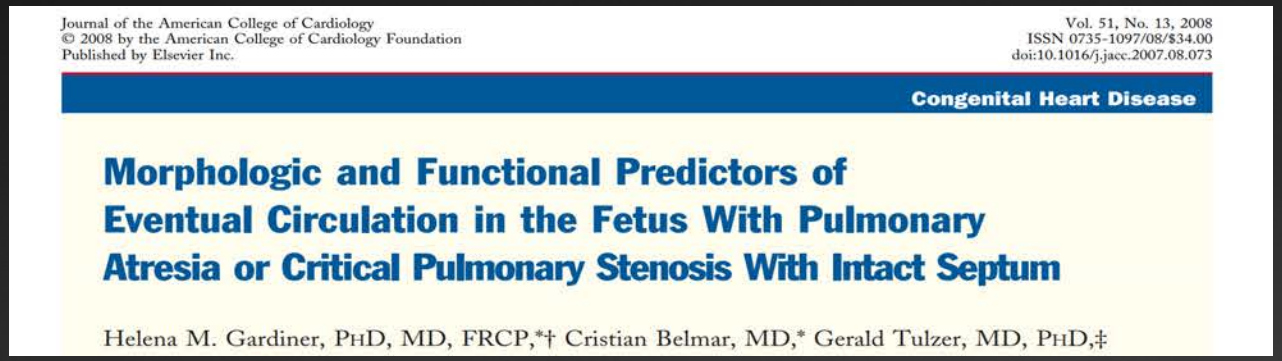
It Begins Before Birth

Fetal Tricuspid Valve Size and Growth as Predictors of Outcome in Pulmonary Atresia With Intact Ventricular Septum

Joshua W. Salvin, MD[a,b], Doff B. McElhinney, MD[a,b], Steven D. Colan, MD[a,b], Kimberlee Gauvreau, ScD[a], Pedro J. del Nido, MD[c,d], Kathy J. Jenkins, MD, MPH[a,b], James E. Lock, MD[a,b], Wayne Tworetzky, MD[a,b]

Departments of [a]Cardiology and [c]Cardiac Surgery, Children's Hospital Boston, Boston, Massachusetts; Departments of [b]Pediatrics and [d]Surgery, Harvard Medical School, Boston, Massachusetts

Z score > -3 = BiV



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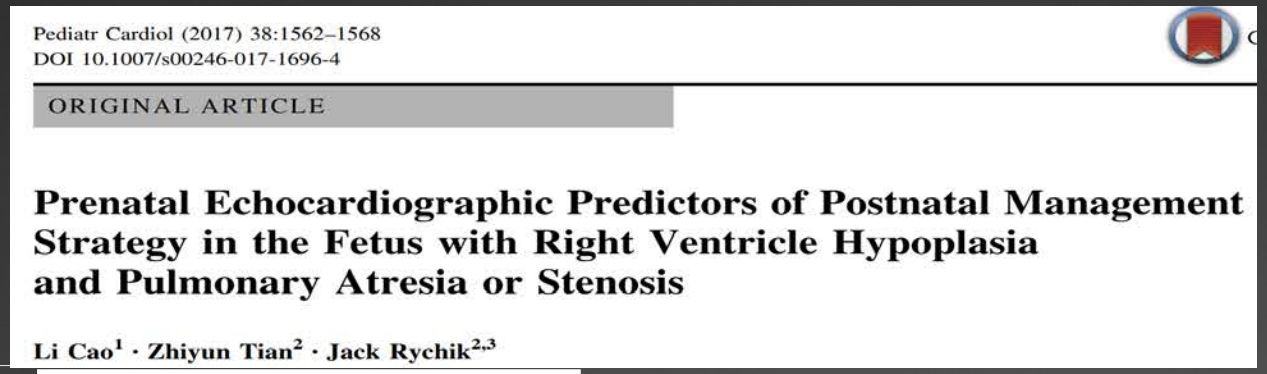
ISSN 0735-1097/08/$34.00
doi:10.1016/j.jacc.2007.08.073

Congenital Heart Disease

Morphologic and Functional Predictors of Eventual Circulation in the Fetus With Pulmonary Atresia or Critical Pulmonary Stenosis With Intact Septum

Helena M. Gardiner, PHD, MD, FRCP,*† Cristian Belmar, MD,* Gerald Tulzer, MD, PHD,‡

Z score > -3 = BiV +
Functional parameters



Pediatr Cardiol (2017) 38:1562–1568
DOI 10.1007/s00246-017-1696-4

ORIGINAL ARTICLE

Prenatal Echocardiographic Predictors of Postnatal Management Strategy in the Fetus with Right Ventricle Hypoplasia and Pulmonary Atresia or Stenosis

Li Cao[1] · Zhiyun Tian[2] · Jack Rychik[2,3]

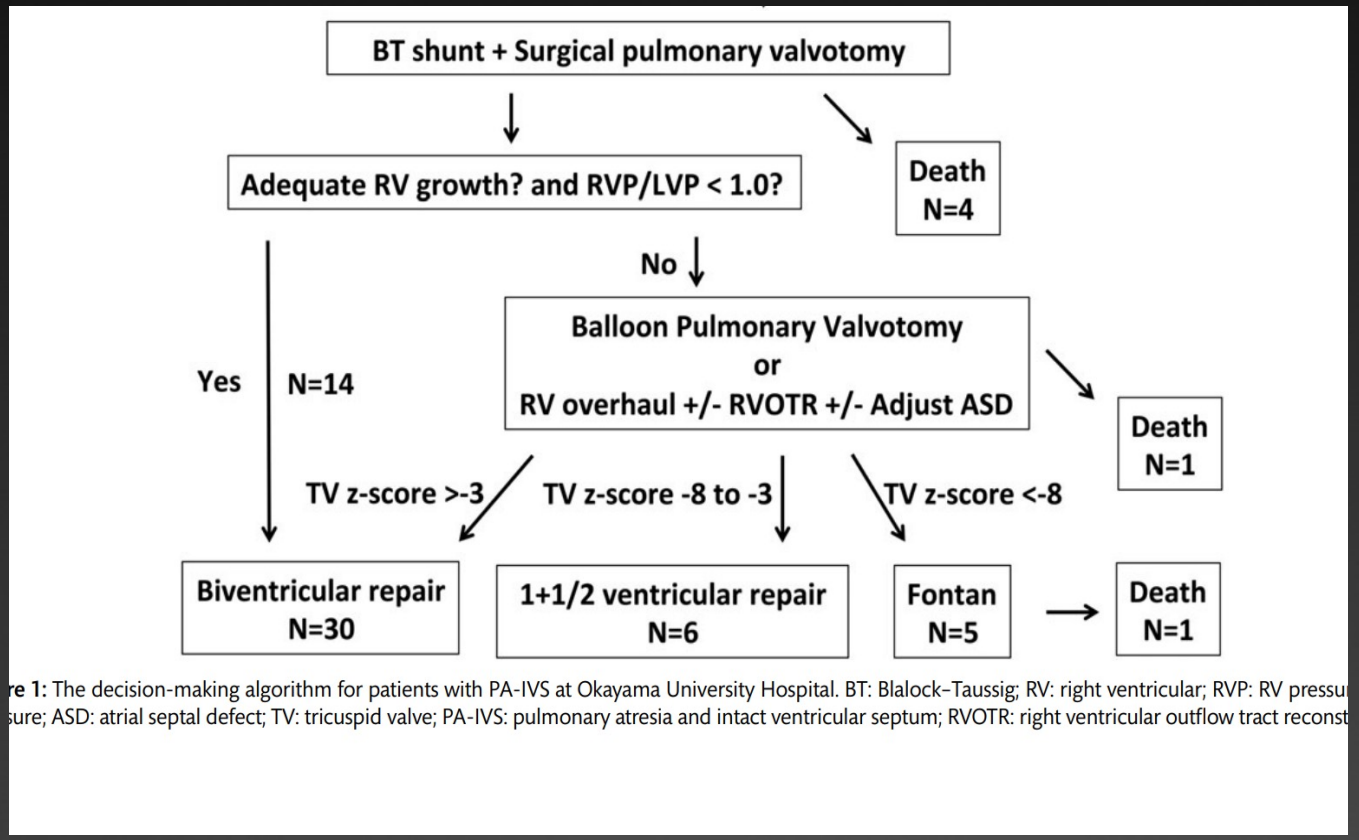
Z score > -3 = BiV
Presence of TR
Absence of CCF

Pulmonary Atresia with IVS

- Potential for growth of the RV if antegrade flow persists
  - *Lewis AB et al. JTCVS 1986;91:835-840*
  - *Ovaert C et al. JTCVS 1998;115:1055-1062*
  - *Sano S et al. Ann Thoracic Surg 2000; 70:1501*

A staged decompression of right ventricle allows growth of right ventricle and subsequent biventricular repair in patients with pulmonary atresia and intact ventricular septum†

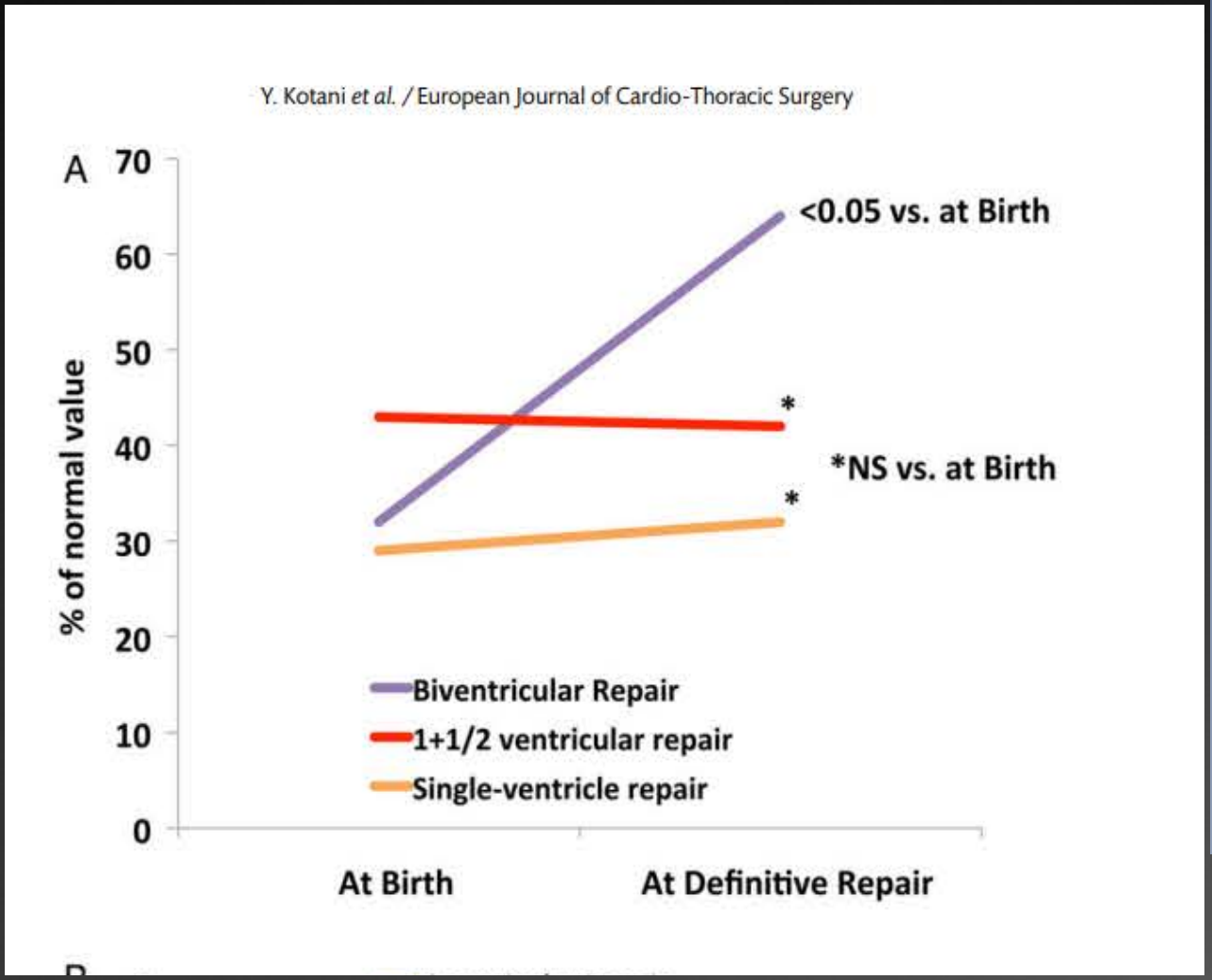
Yasuhiro Kotani[a], Shingo Kasahara[a], Yasuhiro Fujii[a], Takahiro Eitoku[b], Kenji Baba[b], Shin-ichi Otsuki[b], Yosuke Kuroko[a], Sadahiko Arai[a] and Shunji Sano[a,*]



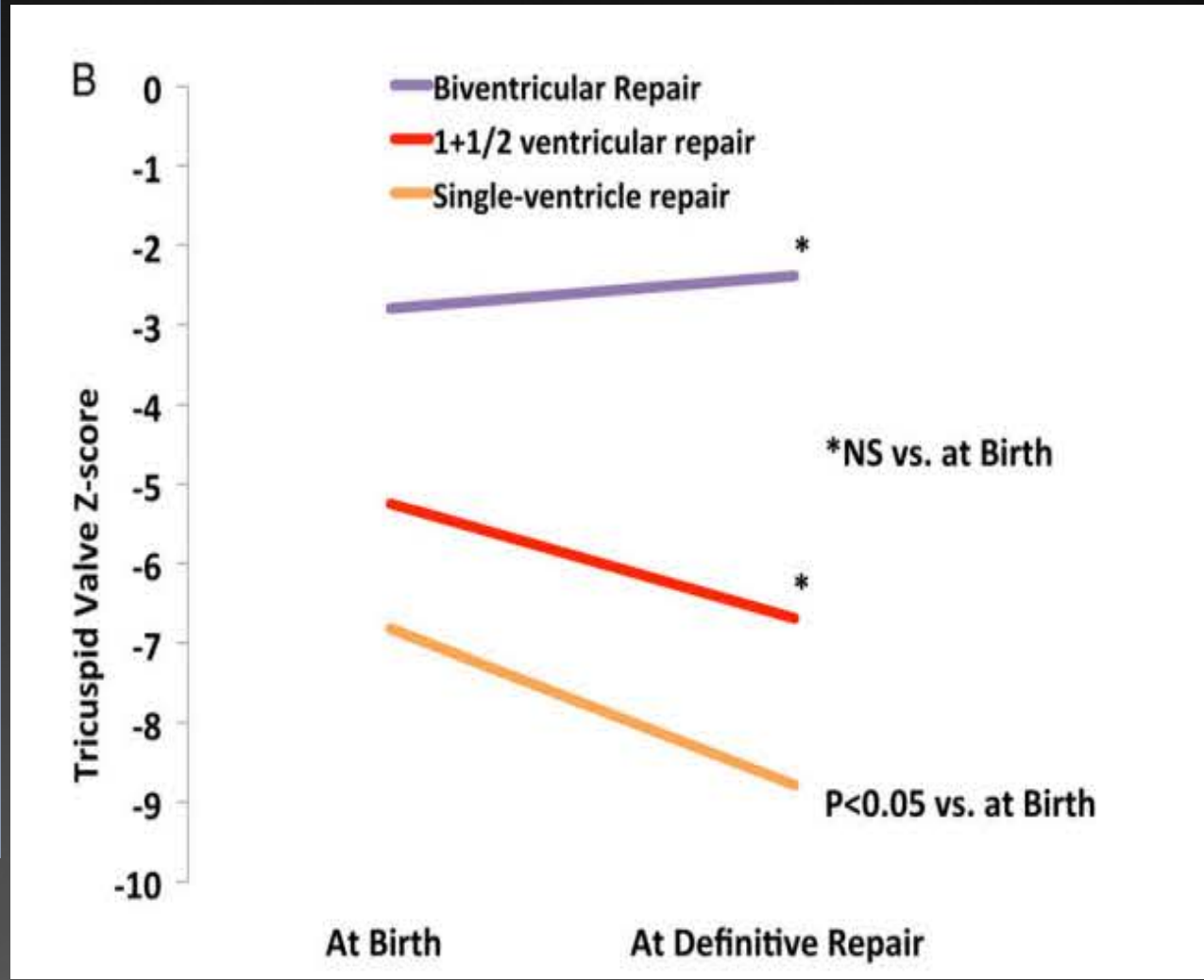
re 1: The decision-making algorithm for patients with PA-IVS at Okayama University Hospital. BT: Blalock–Taussig; RV: right ventricular; RVP: RV pressu
sure; ASD: atrial septal defect; TV: tricuspid valve; PA-IVS: pulmonary atresia and intact ventricular septum; RVOTR: right ventricular outflow tract reconst

European Journal of Cardio-Thoracic Surgery 50 (2016) 298–303
doi:10.1093/ejcts/ezw124 Advance Access publication 26 April 2016

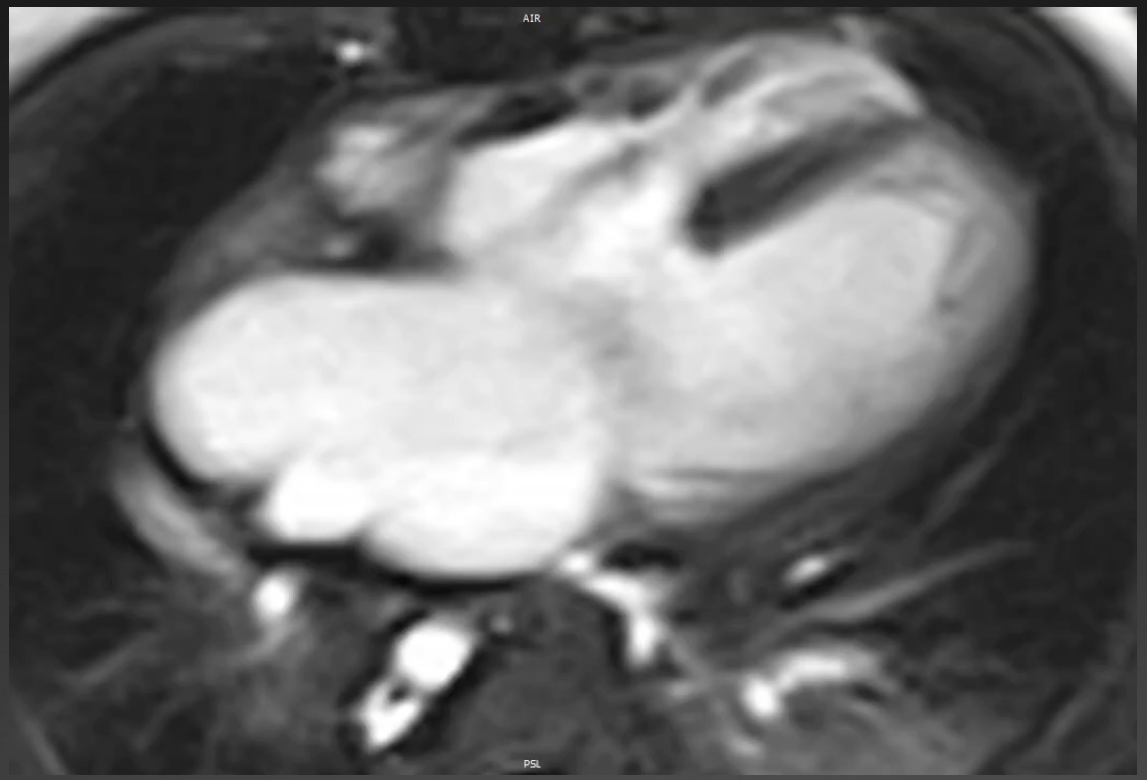
RV Growth



TV Growth



Unbalanced AV Canal to the Left (with small RV)



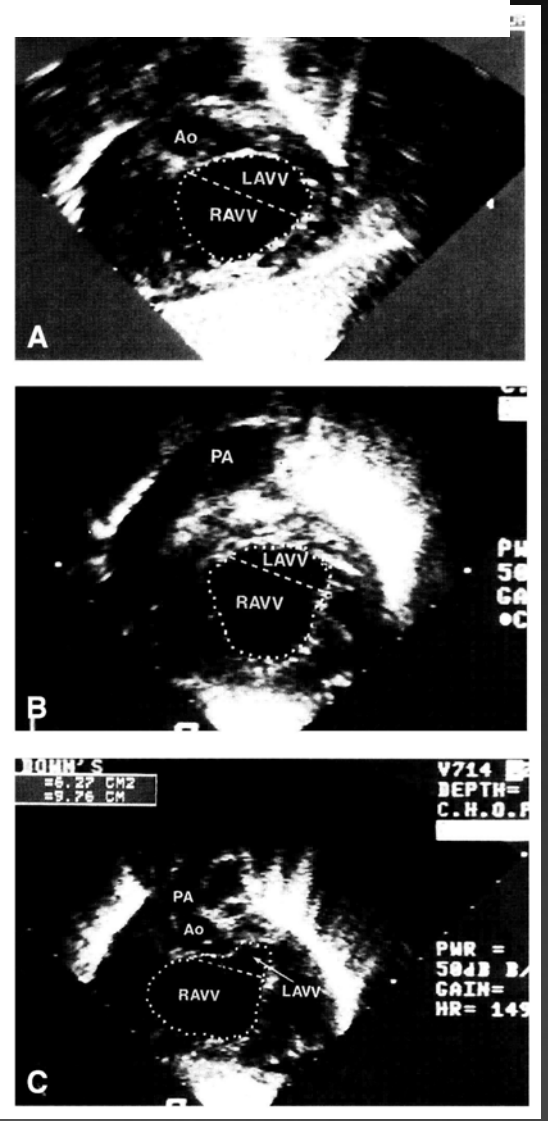
uAVC to left data

- Even less common…
- Some centers have attempted aggressive BiV repairs
  - especially in Trisomy 21 pts/Significant AVVR

Morphometric Analysis of Unbalanced Common Atrioventricular Canal Using Two-Dimensional Echocardiography

MERYL S. COHEN, MD, MARSHALL L. JACOBS, MD, PAUL M. WEINBERG, MD, FACC, JACK RYCHIK, MD, FACC

- Use of AVVI and Ventricular Cavity Ratio
- AVV Index (AVVI):
  - smaller valve area/larger valve area
  - AVVI = 1 is a perfectly balanced AVC
  - AVVI <0.67= unbalanced
  - Only 3 pts with uAVC to left with small RV



JACC 1996(28:);1017-23

Echocardiographic Definition and Surgical Decision-Making in Unbalanced Atrioventricular Septal Defect

A Congenital Heart Surgeons' Society Multiinstitutional Study

Anusha Jegatheeswaran, MD; Christian Pizarro, MD; Christopher A. Caldarone, MD; Meryl S. Cohen, MD; Jeanne M. Baffa, MD; David B. Gremmels, MD; Luc Mertens, MD, PhD; Victor O. Morell, MD; William G. Williams, MD; Eugene H. Blackstone, MD; Brian W. McCrindle, MD, MPH; David M. Overman, MD

Table 2. Operative Strategy Versus AVVI Determination of "Balancedness"

		Unbalanced		
	Balanced	Total	R	L
All patients (n=305)				
Total	247	58	50	8
Single ventricle repair				
Total	1	22	22	0
Alive	1	15	15	0
Dead	0	7	7	0
Biventricular repair				
Total	245	34	28	6
Alive	229	29	24	5
Dead	16	5	4	1
Intermediate repair				
Total	1	2	0	2
Alive	0	1	0	1
Dead	1	1	0	1

-Unbalanced to L with small RV

Circulation 2010;122[suppl I]:S209

Biventricular repair in children with atrioventricular septal defects and a small right ventricle: Anatomic and surgical considerations

JTCVS 2005;130:250

Nilto C. De Oliveira, MD,[a*] Rekwan Sittiwangkul, MD,[a] Brian W. McCrindle, MD, MPH,[b] Anne Dipchand, MD,[b] Tae-Jin Yun, MD,[c] John G. Coles, MD,[a] Christopher Caldarone, MD,[a] William G. Williams, MD,[a] and Glen S. Van Arsdell, MD[a]

- BVR was successful in
  - AVVI as low as 0.50
  - RV/LV cavity ratio of 0.66
- BVR with AVVI < 0.50 perform with caution !
- At 10 years, survival 87% for BVR

Where does this leave us ?
Possible Treatment Algorithms

- There remains controversy as to what constitutes an appropriate right heart size for what pathway

Anatomical Criteria for Borderline RV

TV z-score	RV Vol	Ultimate operation
> -3 (ish)	> 75%	BiV
-3 to -5	50-75%	1.5 repair (+/- ASD)
< -6	< 30-50%	Fontan

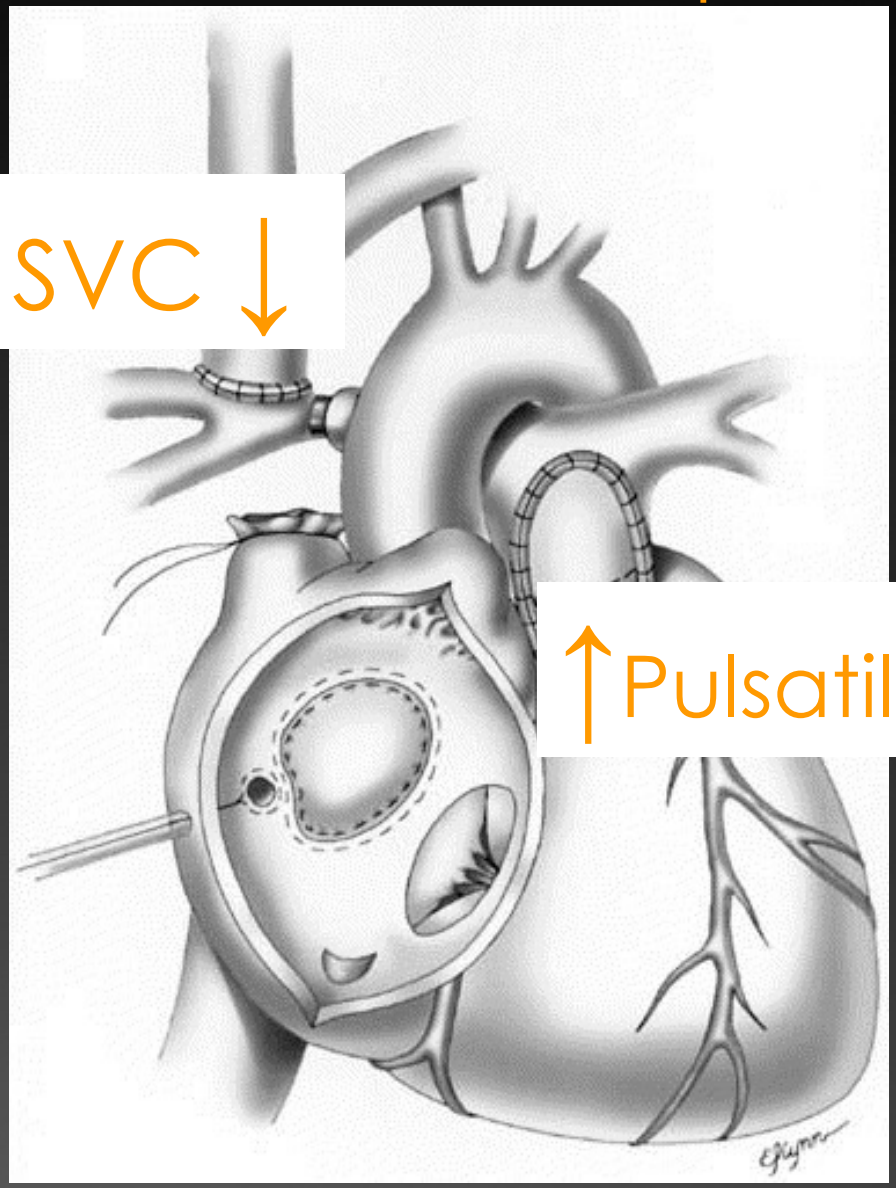
- Must rule out RVDCC

To Be Determined

- Can the 1.5V approach allow for avoidance of early problems of high risk BiV repair and avoid long term complications of the Fontan procedure?

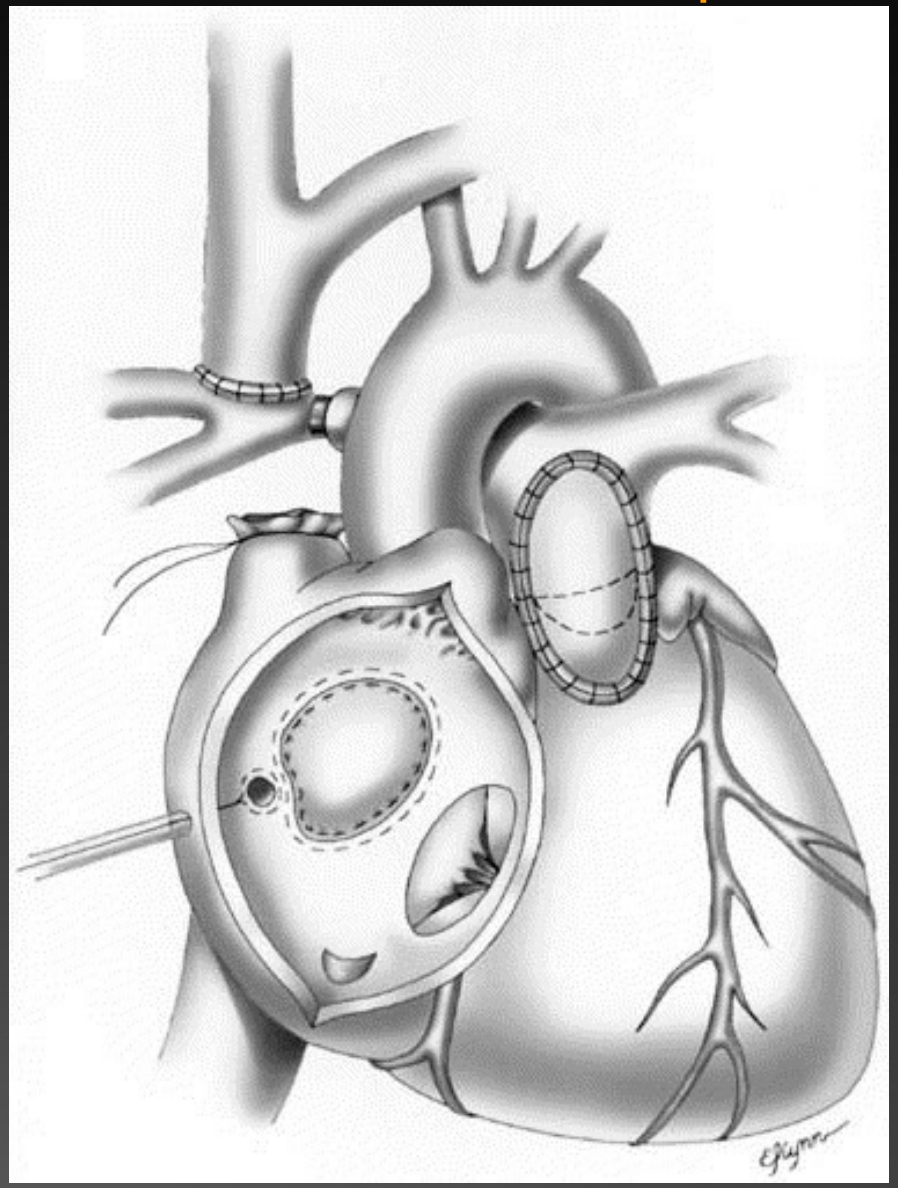
One-and-half repair

The pulmonary circulation receives a full cardiac output



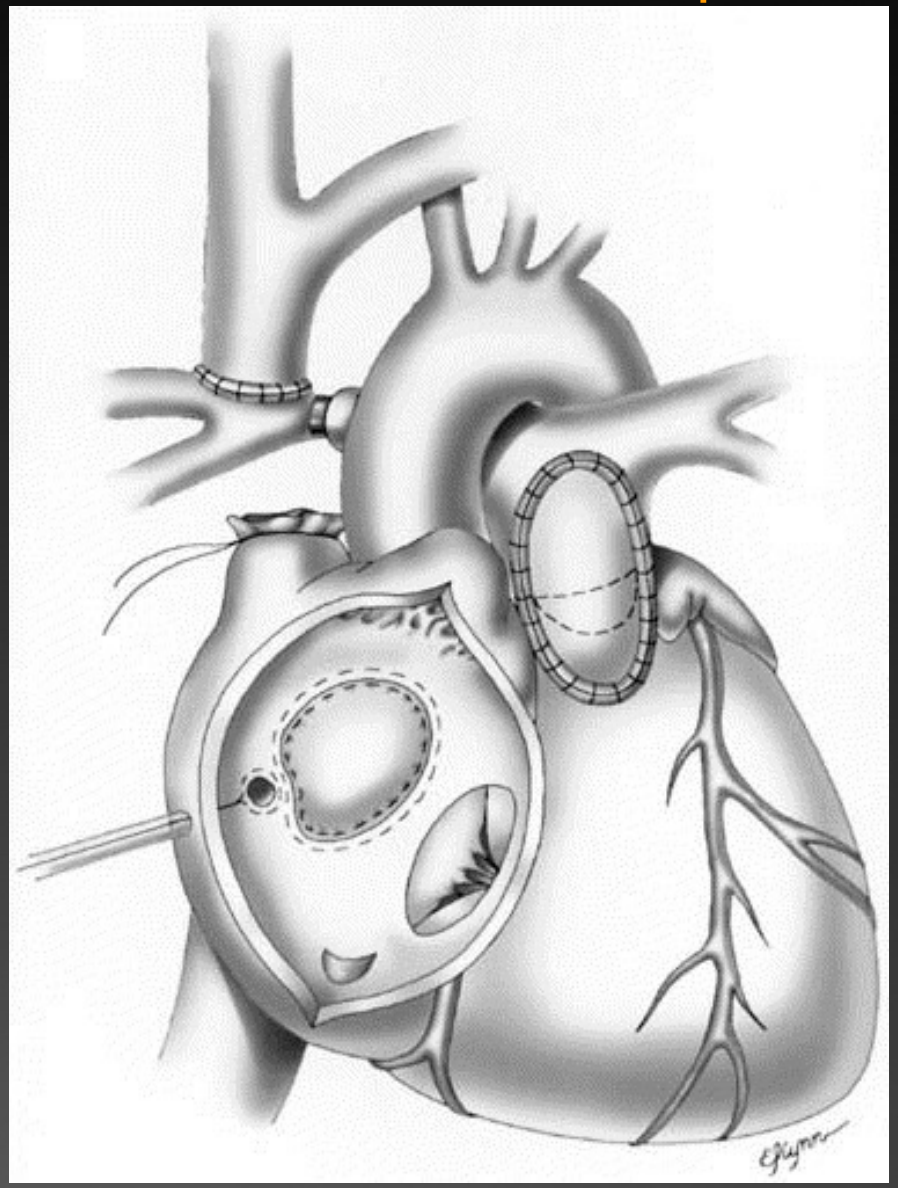
One-and-half repair

Creates a modified in series circulation with no shunts (Qp=Qs)

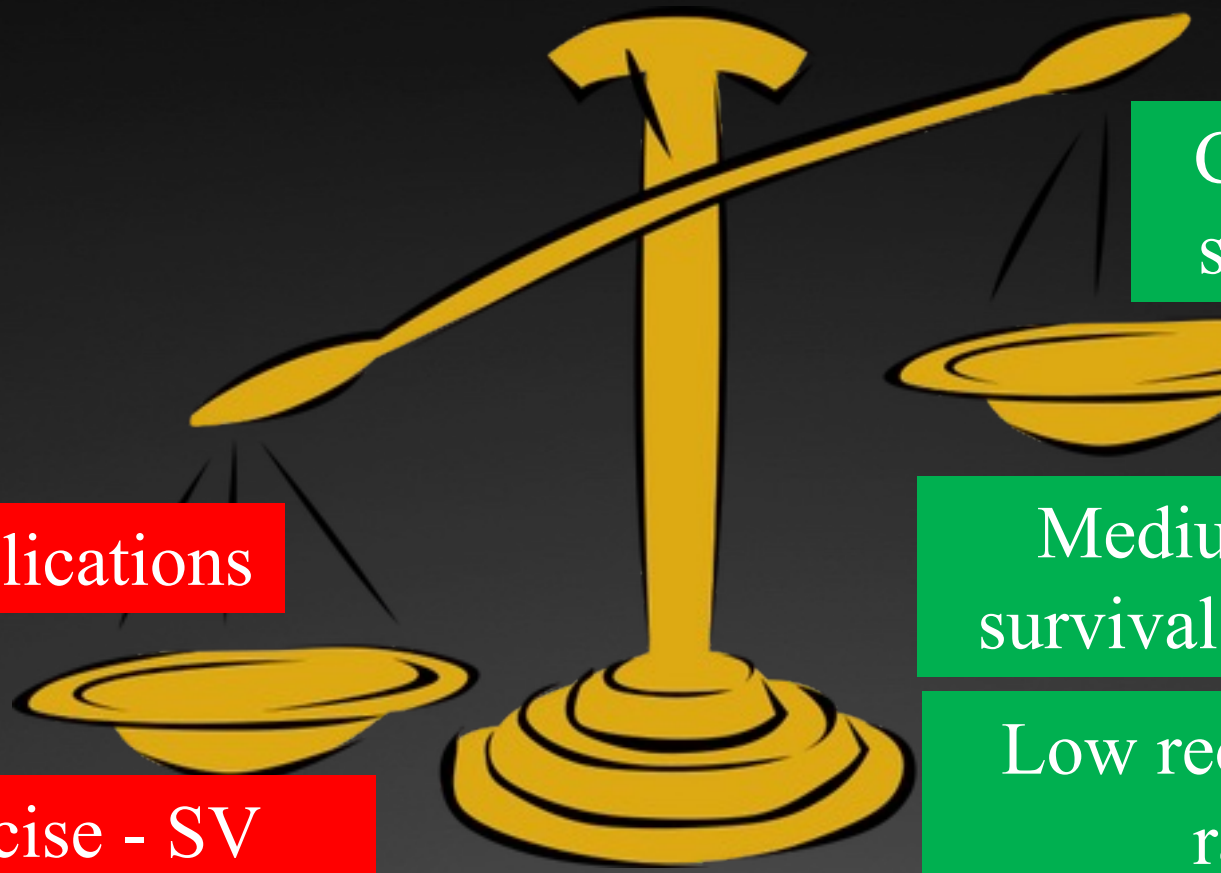


One-and-half repair

The Left Ventricle pumps single cardiac output



One and Half Ventricle Repair



Can be done successfully

Medium-term survival 77-100%

Low reoperation rate

Complications

Exercise - SV

Pulsatility in SVC

Arrhythmia

Case Examples to Highlight some strategies to The Borderline RV

Case 1: PA/IVS

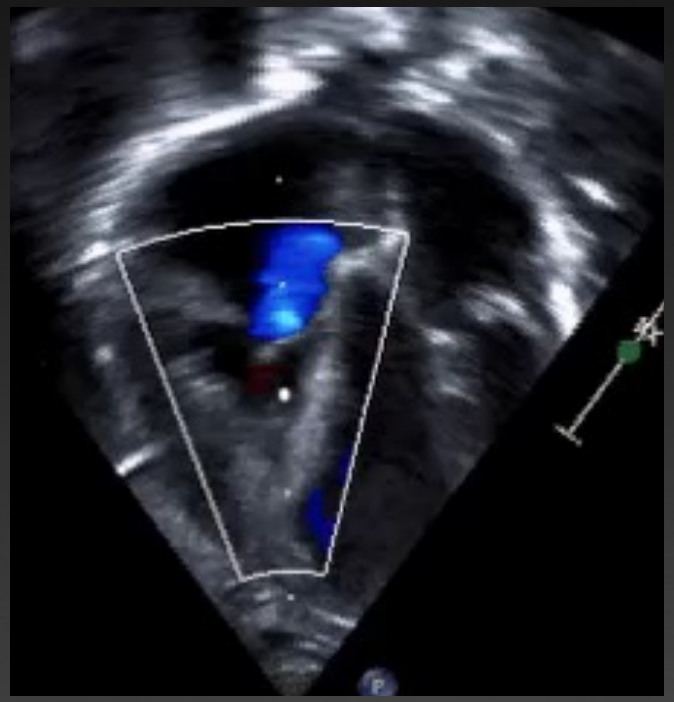
29 weeks GA
No ECA
TV 4.8mm
Z- score = -3.7

Mod + TR
Rvp= 110mmHg

- Counseled to all 3 approaches

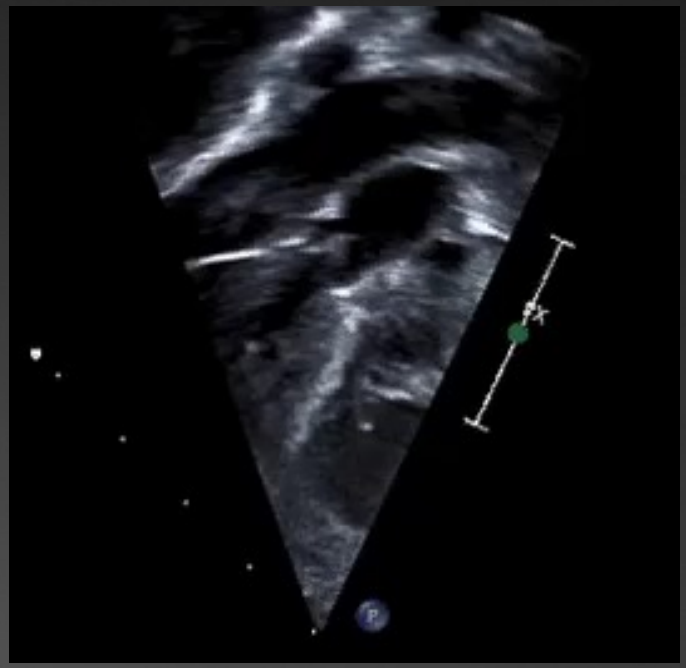
At Birth

Full term 3.3 kg
TV 5.5 mm
Z- score = -3.8



Moderate TR
RVp 2x systemic

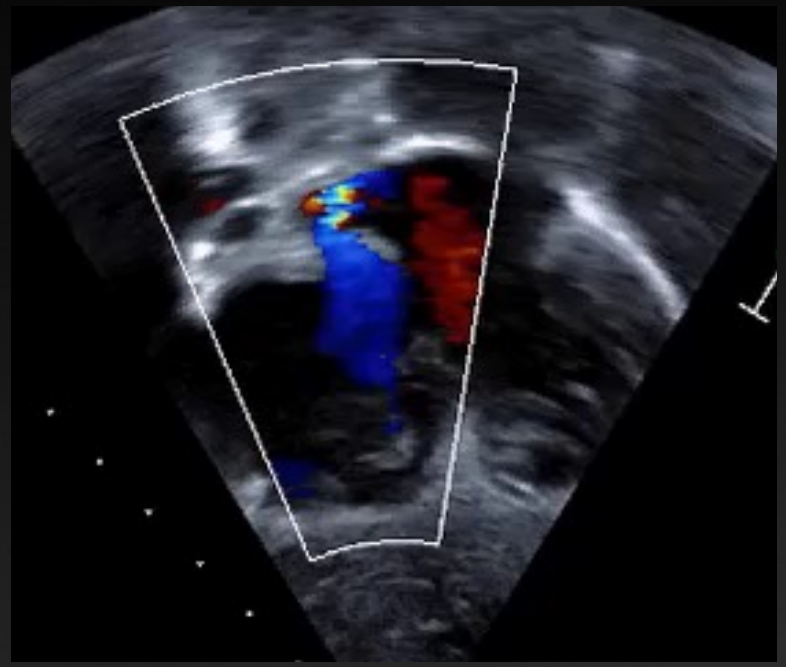
Tripartite RV
Well formed PV (7mm)



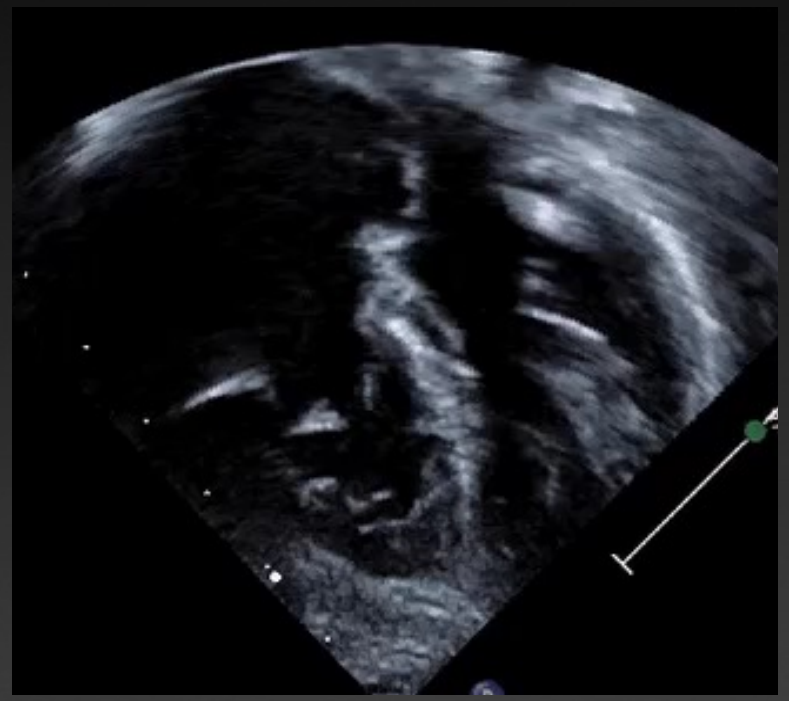
Clinical story….

- Cath lab on DOL #2 : CCF but no RVDCC
  - RF perforation
  - PDA stenting
- Discharged home
- 12 months had decline in saturations, returned to cath lab
  - Good hemodynamics, no RVOTO, minimal flow thru PDA

Echo 12 months age

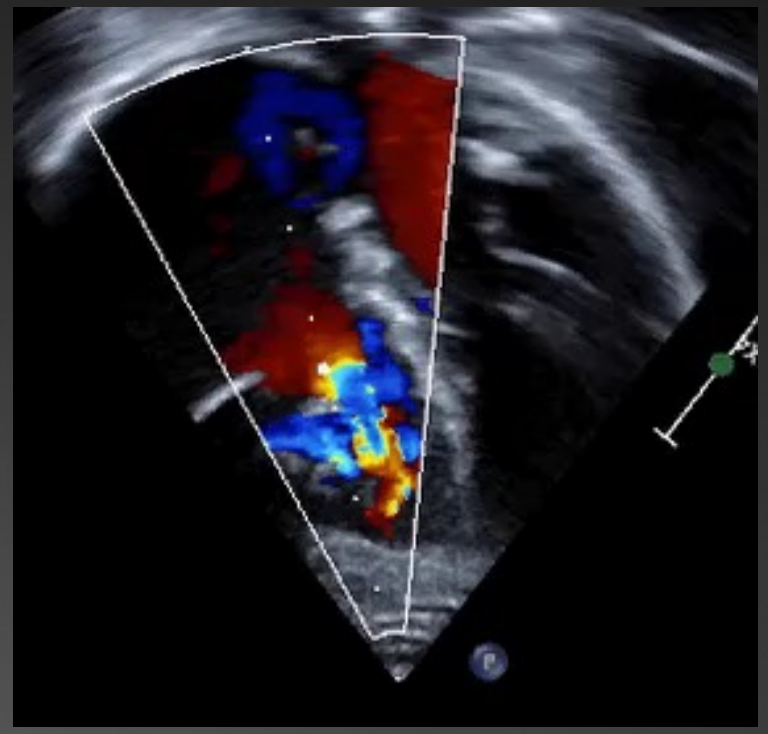


Rt to Left PFO



TV 10 mm
Z- score = -3
RV vol 70% normal

Mn grad= 10 mmHg

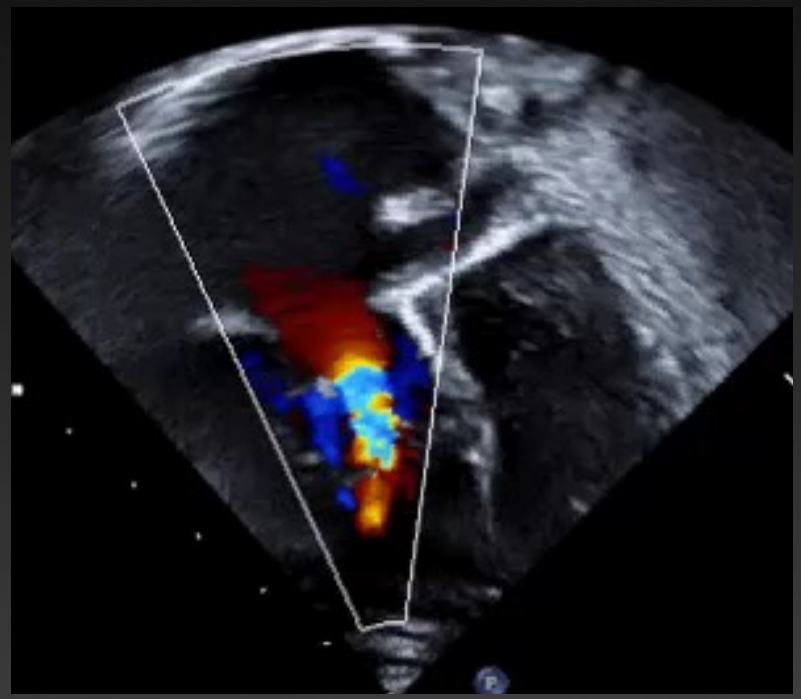


To OR for One and Half Ventricle

- At 1 yr of age: BDG, RV muscle resection, division of TV papillary muscle and fenestrated (4mm) ASD
- Now 2 years of age, he has normal growth and clinically well, Pox 96%
  - No SVC dilation
  - No arrythmia

2 years of age

TV 12mm
Z score -2.6



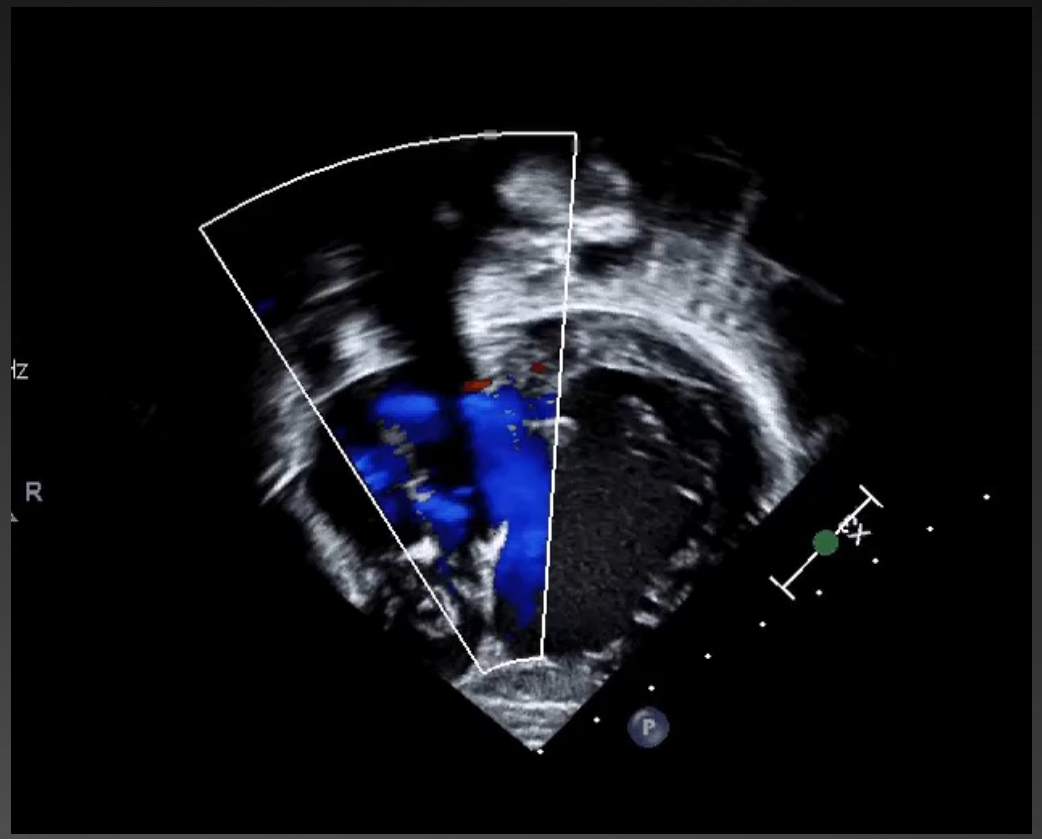
Case 2; DORV, PS, RV hypoplasia, Straddling TV

- BT shunt at 2 months 2/2 cyanosis
- At 6 months of age, growing well and Saturation 80%

Pre-operative Assessment

- Cardiac cath performed to assess hemodynamics and PA anatomy
- Referred to OR to for bidirectional Glenn at 6 months
- Clinically did well and at 3 years of age for planning of next surgery underwent repeat echo and cardiac MRI
  - MRI performed for assessment of RV volume and LV to aortic pathway

1.5V Repair: VSD closure, TV chordae translocation, RV-PA conduit



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BDG left 2/2 RV hypoplasia

Summary- where does this leave us?

- B-RV has a wide spectrum of disease with low prevalence creating challenges

- Some algorithms do exist, remains limited data for guiding decision making
  - Utilize multi-imaging modalities
  - Give time for growth
  - 1.5V repair for real borderline cases

THANK YOU

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Cardiac Echo Lab at CHOP – Faculty and Sonographers

April 27, 2017

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One and half ventricle repair

- Avoid early mortality seen when extending limits of small RV and later outcomes of Fontan circulation
- Performed biventricular repairs along with a BDG
  - Applied to pts with small or poorly functioning RV
  - Reports in Tv z score as small as -10 and Rv volumes as low as 30%
- Reduce the volume load on the RV, the pulmonary circulation receives a full CO (BDG flow + pulsatile RV)
- No shunts, equal systemic and pulmonary flows
- Avoid early problems of BiV repairs and the long term sequele of Fontan circulation

CHOP Criteria for Borderline RV

TV z-score	RV Vol	Ultimate operation
> -3	> 75%	BiV
< -3	< 75%	1.5 repair
< -4	< 50%	1.5 repair +ASD
< -5 to -7	< 30-50%	Fontan

- RV function, morphology (tripartite), PA size and PVR all important contributors, Coronaries (RVDCC), Degree of TR.

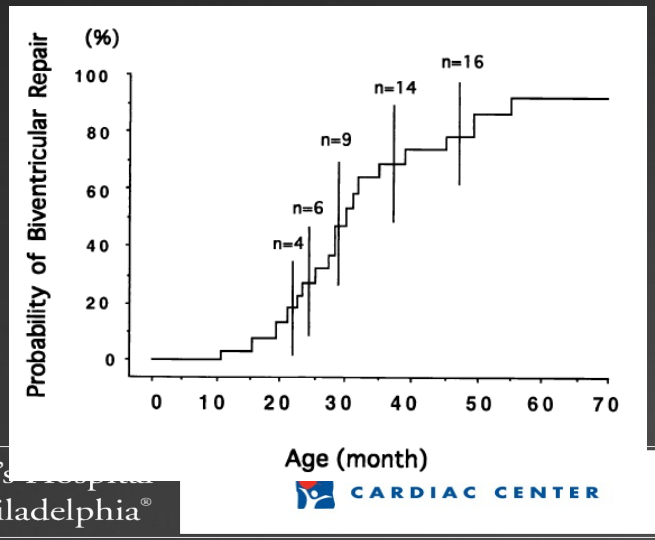
Pulmonary Atresia with IVS

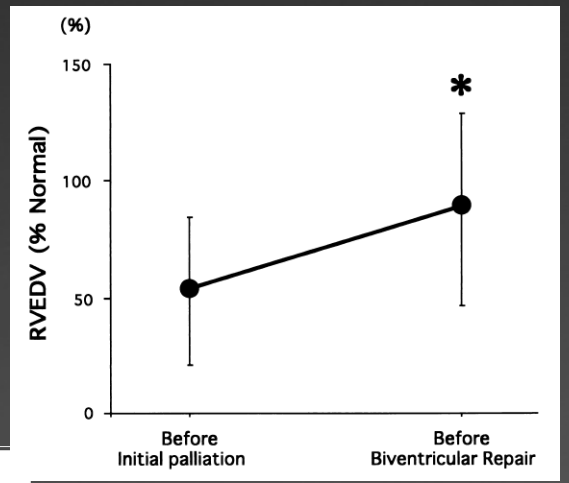
- Potential for growth of the RV if antegrade flow persists – staged reconstruction

Staged Biventricular Repair of Pulmonary Atresia or Stenosis With Intact Ventricular Septum

Shunji Sano, MD, Kozo Ishino, MD, Masaaki Kawada MD, Emi Fujisawa, MD, Masahiro Kamada, MD, and Shin-ichi Ohtsuki, MD

Ann Thoracic Surg 2000;70:1501





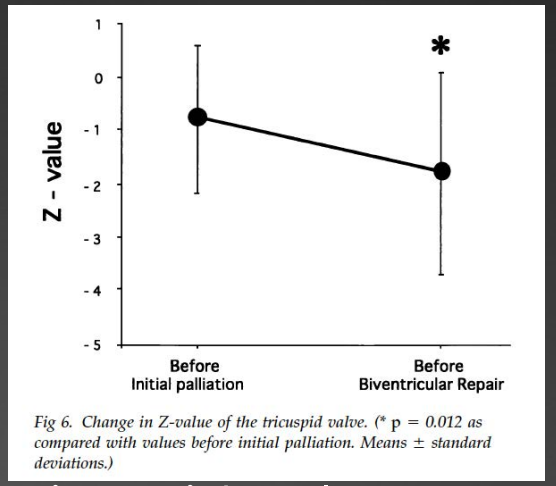


Fig 6. Change in Z-value of the tricuspid valve. (p = 0.012 as compared with values before initial palliation. Means ± standard deviations.)*

Tricuspid Valve

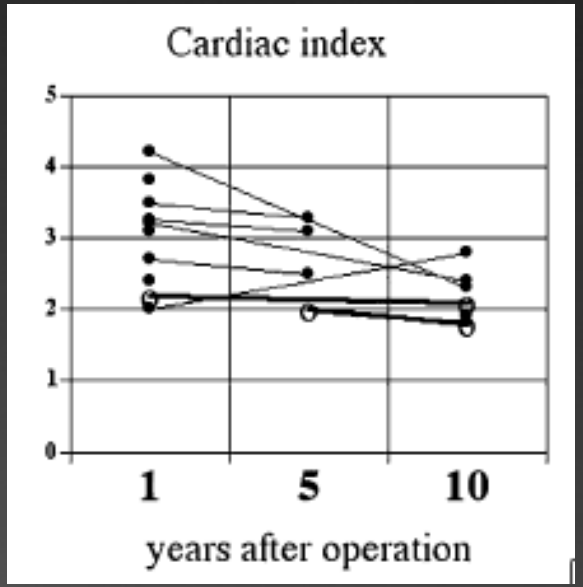
One and Half Ventricle

Long-term functional results of the one and one half ventricular repair for the spectrum of patients with pulmonary atresia/stenosis with intact ventricular septum☆

European Journal of Cardio-thoracic Surgery 24 (2003) 516–520

Satoshi Numata, Hideki Uemura*, Toshikatsu Yagihara, Koji Kagisaki, Masashi Takahashi, Hideo Ohuchi

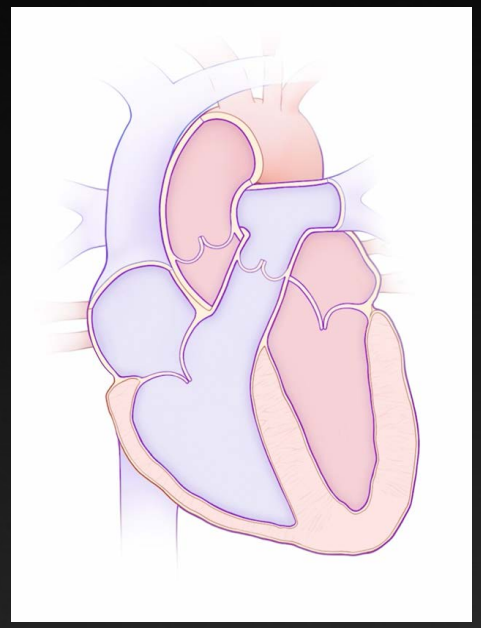
- N=13 (UAVC+PA/IVS), follow-up 3-15 yrs (10±4 yrs)
  - Freedom from arrhythmia 20% @ 12 yrs
  - No change in RVEDVi or tricuspid valve size @ 10 yrs
  - RA pressure 12±2 mm Hg @ 10 yrs
  - Exercise tolerance similar to Fontan



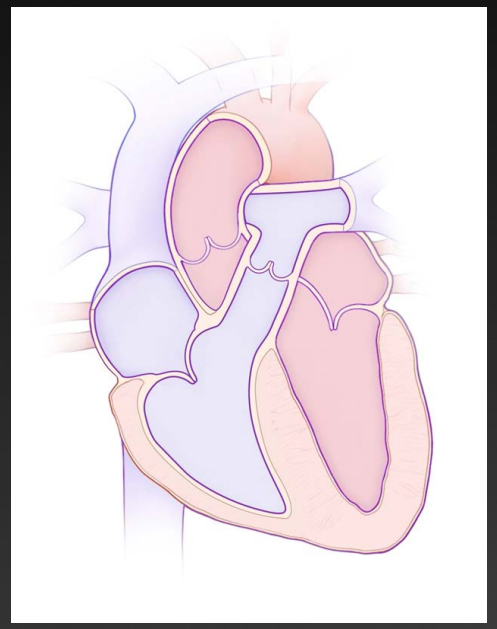
B-RV: what factors to consider

- Right Ventricle
  -Ventricular End Diastolic Volumes
  -RV morphology (? tripartite, presence of infundibulum)
  -RV systolic function (? Cardiomyopathic changes)
- Tricuspid Valve
  -Annular size/ Z-score (as a surrogate for volume)
  -Morphology: Dysplasia, Ebsteins, and tricuspid insufficiency
  -Inflow potential in malaligned AV canal
  -Chordal attachments
- RV outflows and PAs (not usually as important, can be surgically treated)
- Ability of the ventricles be separated
  -VSD too large "Swiss cheese " ventricular septum
- Pulmonary vascular resistance

Spectrum of right heart hypoplasia



2 Ventricle



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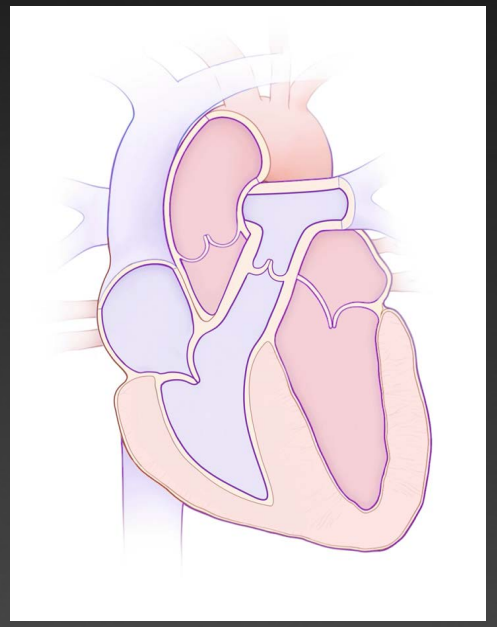

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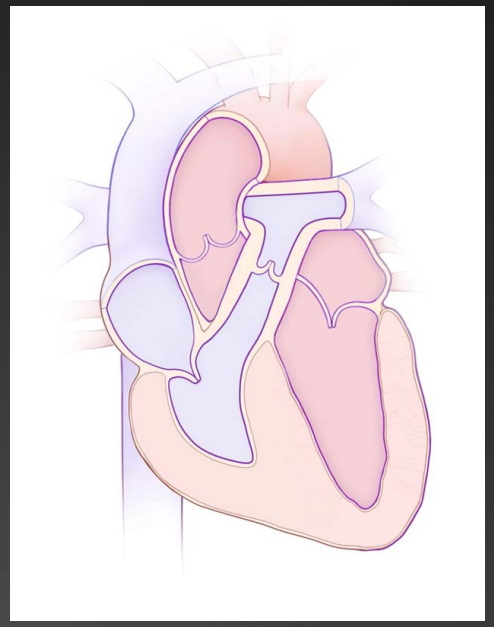
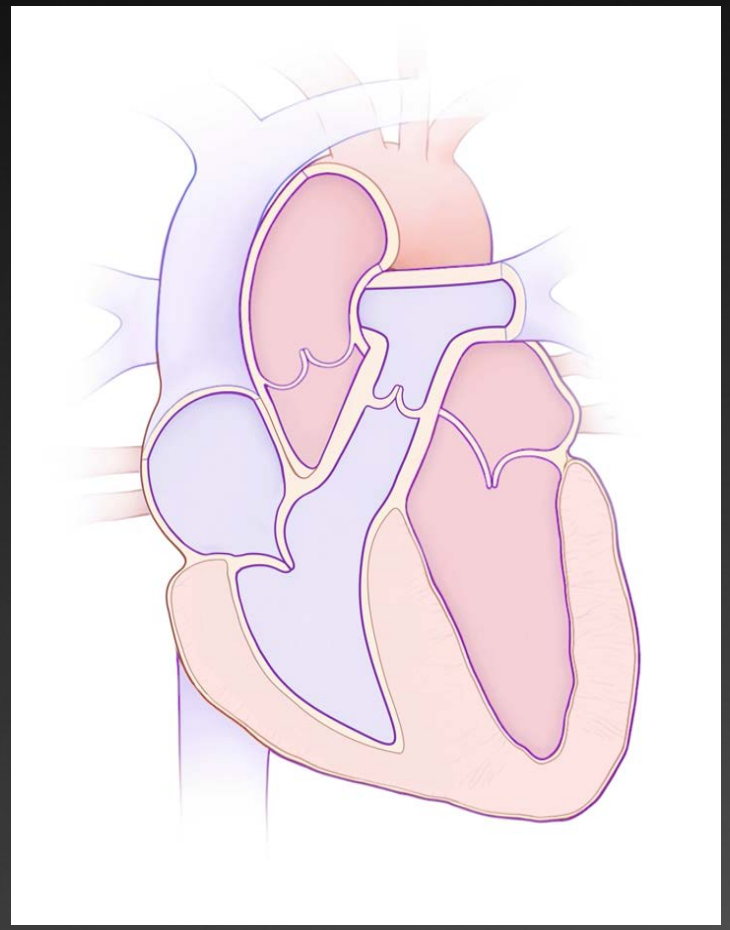
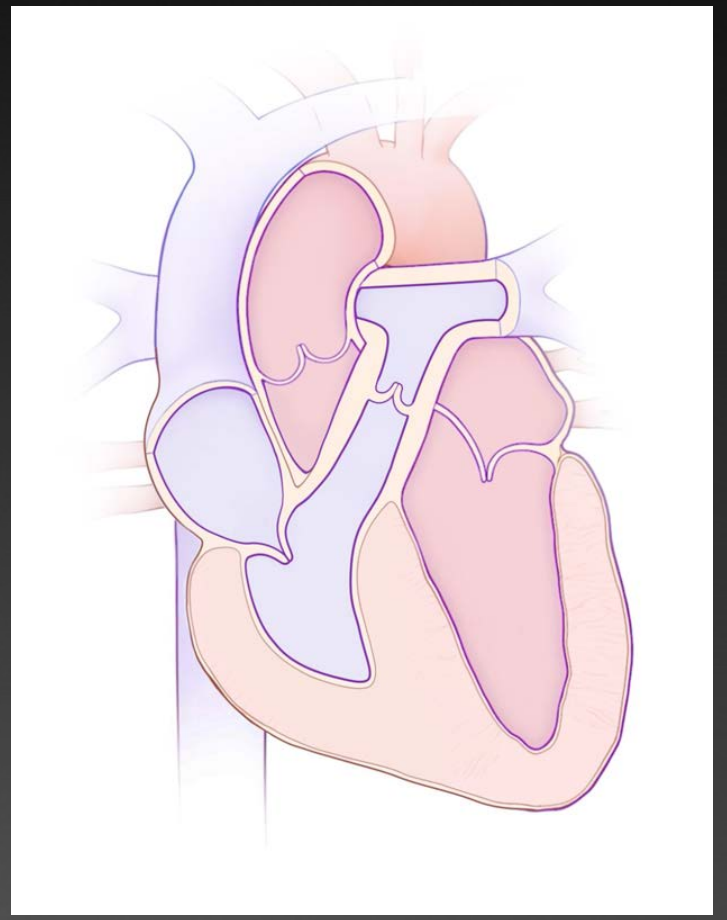


1 Ventricle

The dilemma arises when determining optimal pathways for pts that fall between the extremes



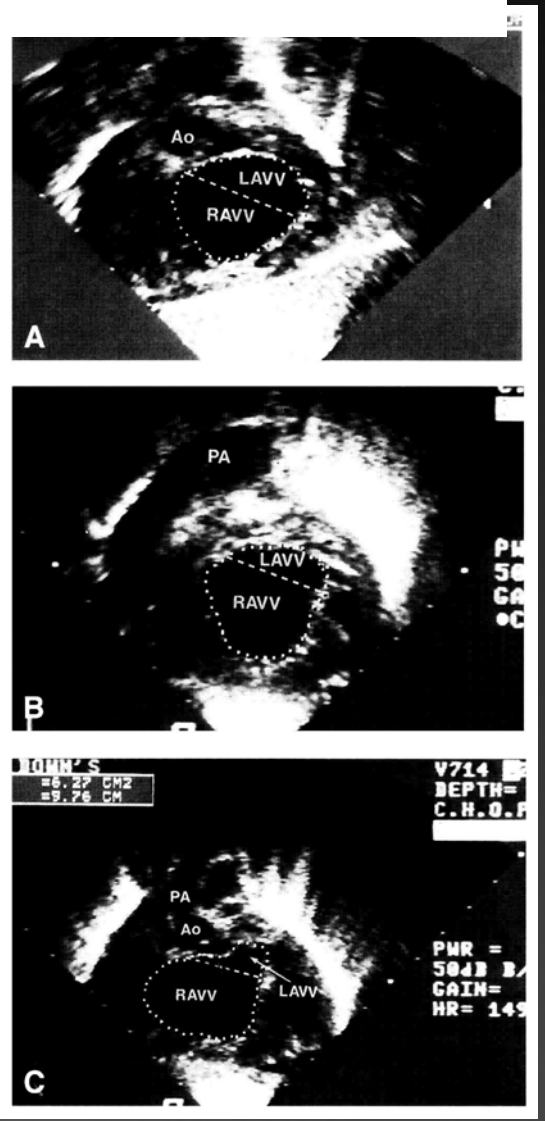


Morphometric Analysis of Unbalanced Common Atrioventricular Canal Using Two-Dimensional Echocardiography

MERYL S. COHEN, MD, MARSHALL L. JACOBS, MD, PAUL M. WEINBERG, MD, FACC, JACK RYCHIK, MD, FACC

- Use of AVVI and Ventricular Cavity Ratio
- AVV Index (AVVI):
  - smaller valve area/larger valve area
  - AVVI = 1 is a perfectly balanced AVC
  - AVVI > 0.67 =balanced, 2V repair
- Ventricular cavity ratio

$$\frac{\text{LV length x LV width}}{\text{RV length x RV width}}$$



JACC 1996(28:);1017-23

The Children's Hospital of Philadelphia®

CARDIAC CENTER

Biventricular repair in children with atrioventricular septal defects and a small right ventricle: Anatomic and surgical considerations

JTCVS 2005;130:250

Nilto C. De Oliveira, MD,[a*] Rekwan Sittiwangkul, MD,[a] Brian W. McCrindle, MD, MPH,[b] Anne Dipchand, MD,[b] Tae-Jin Yun, MD,[c] John G. Coles, MD,[a] Christopher Caldarone, MD,[a] William G. Williams, MD,[a] and Glen S. Van Arsdell, MD[a]

TABLE 4. Morphometric measures in 11 patients who underwent creation of a fenestrated atrial septal defect*

Patient no.	AVVI	RV/LV ratio	Patient no.	AVVI	RV/LV ratio
1	0.20	1.04	7	0.53	0.69
2	0.29	0.96	8*	0.48	0.62
3	0.42	0.74	9	0.65	0.74
4*	0.26	0.98	10	0.82	0.87
5	0.52	0.79	11	0.85	0.75
6	0.87	0.70			

Values are absolute numbers for each patient. *AVVI,* Atrioventricular valve index; *RV/LV ratio,* right ventricular length/left ventricular length ratio. *Patients 4 and 8 had atrial septal defect creation during the first 48 hours after biventricular repair.

- At 10 years, survival 87% for BVR and 100% for control

- Need both AVVI and the RV/LV cavity ratio
- BVR was successful in
  - AVVI as low as 0.50
  - RV/LV cavity ratio of 0.66
- BVR with AVVI < 0.50 perform with caution
- Use of adjustable ASD can

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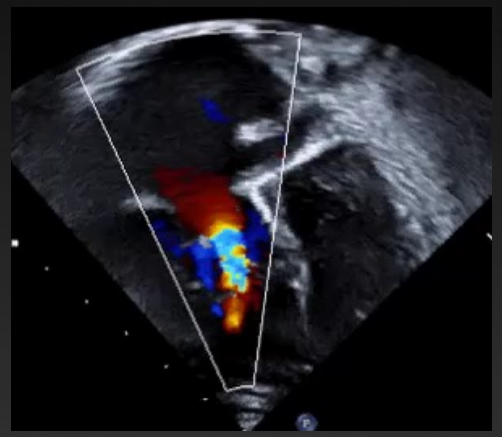
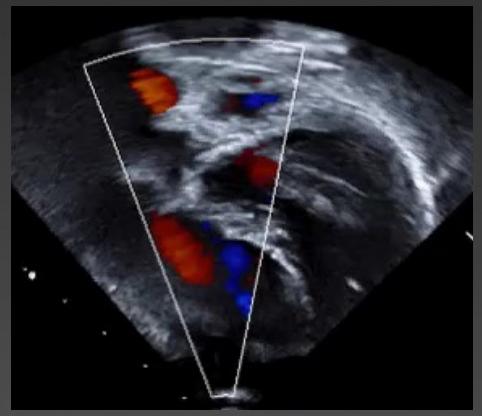
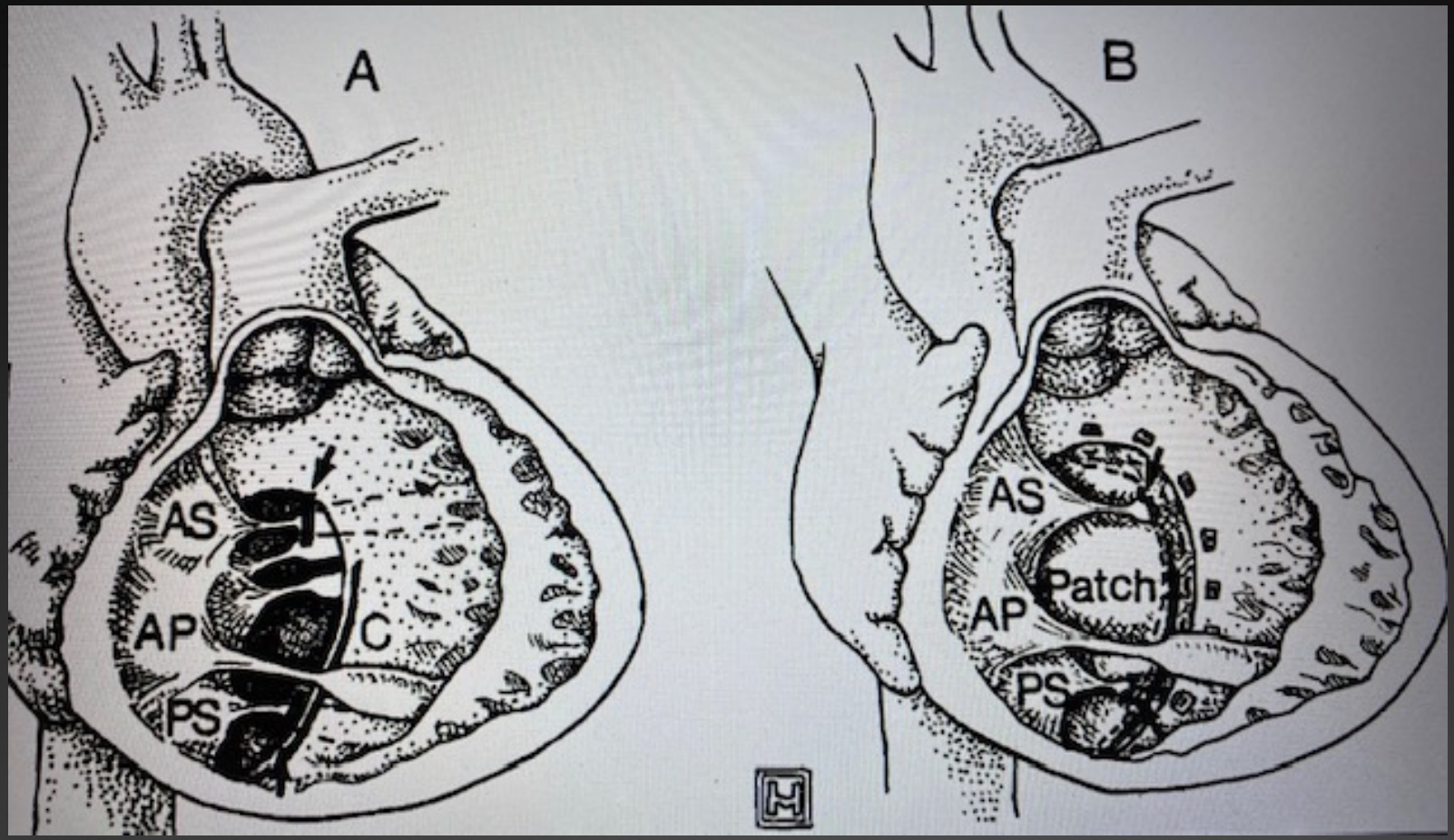
- Early data utilizing AVVI and RV/LV volume ratios
- AVVI < 0.5 had re-ops plus deaths
- Adjustable atrial fenestration utilized
- But aggressively pushed pts to BiV

2 years of age, Pox 97% and clinically well





Reddy, V.M., Brook, M, et al. Cardiology in the Young. April 1997



Pulmonary Atresia with IVS

- Potential for growth of the RV if antegrade flow persists
  - *Lewis AB et al. JTCVS 1986;91:835-840*
  - *Ovaert C et al. JTCVS 1998;115:1055-1062*
  - *Sano S et al. Ann Thoracic Surg 2000; 70:1501*
- *Surgical reconstruction performed in stages for biventricular repair*
  - RVOT opened surgically or via catheter
  - Add PBF
  - Resect RV muscle
  - Adjust ASD to promote flow across the TV
  - Close systemic to pulmonary shunt/ASD

PA-IVS: the problem is the TV

- Prospective, multi-institutional study if 171 neonates, TV z-score:
  - Highly correlated with RV size
  - Negatively correlated with CCF and RV dependence
  - <- 3 Risk factor for not receiving biventricular repair

- Tricuspid valve z score <-3 predicts poor outcome

Hanley FL et al. Outcomes in neonatal pulmonary atresia with intact ventricular septum. A multi-institutional study. JTCVS 1993;105:406

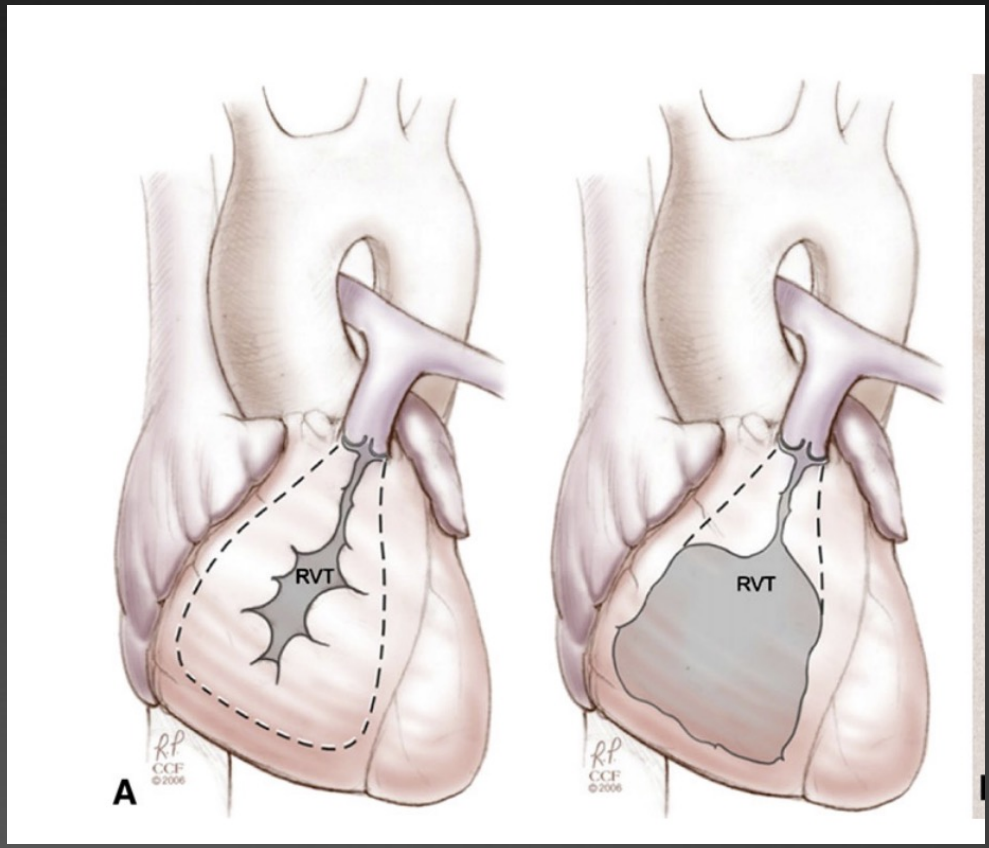
Anatomic Selection Criteria for Borderline RV

TV z-score	RV Vol	Operation
> -2	> 80%	BiV
<-2	< 80%	1.5 repair
<-5	< 50%	1.5 repair +ASD
<-7	< 30%	Fontan

Van Arsdell et al. Ann Thorc Surg, 1998

"RV Overhaul"

- In an effort to extend BiV repairs to pts with more severe forms of right heart hypoplasia
  - RV sinus myomectomy
  - Described by Mee et al. 1986, has refinements



Success and limitations of right ventricular sinus myectomy for pulmonary atresia with intact ventricular septum

Roosevelt Bryant, III, MD,[a] Edward R. Nowicki, MD,[a] Roger B. B. Mee, MB, ChB, FRACS,[b] Jeevanantham Rajeswaran, MSc,[c] Brian W. Duncan, MD,[d] Geoffrey L. Rosenthal, MD, PhD,[b] Uthara Mohan, MD,[d] Muhammad Mumtaz, MD,[b] and Eugene H. Blackstone, MD[a,c]

- 16 pts PA-IVS that underwent RV overhaul
  - Mean TV z score = -4.9
  - All had RV infindibulum

- Early increases in RV volume and 87% pts had BiV repairs
  - TV growth was minimal, rate limiting factor