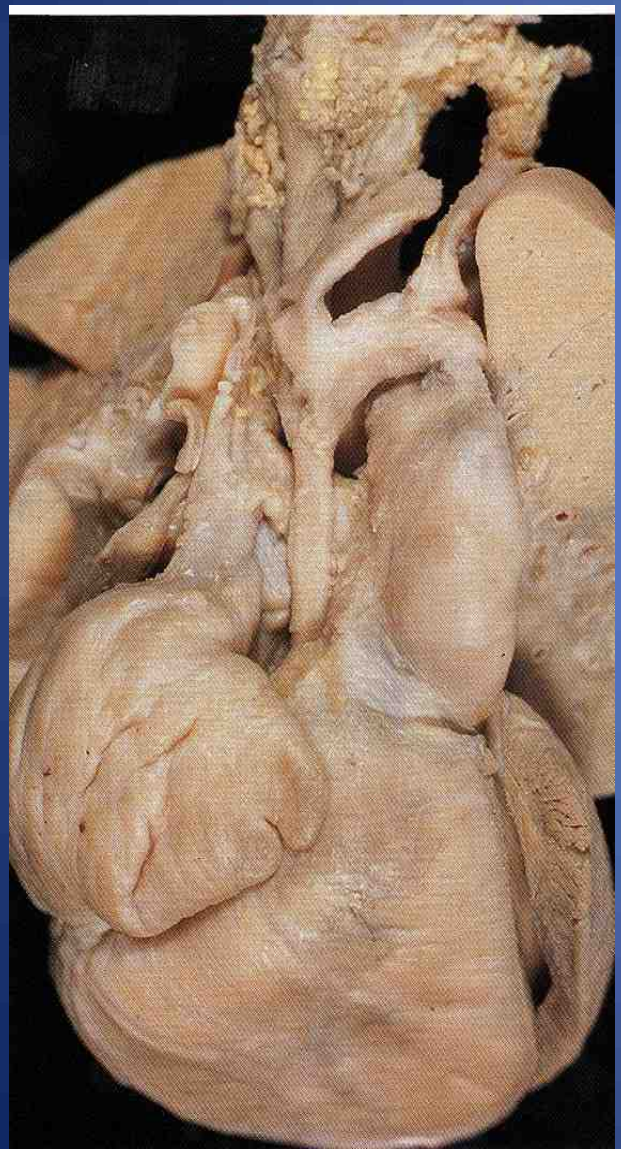
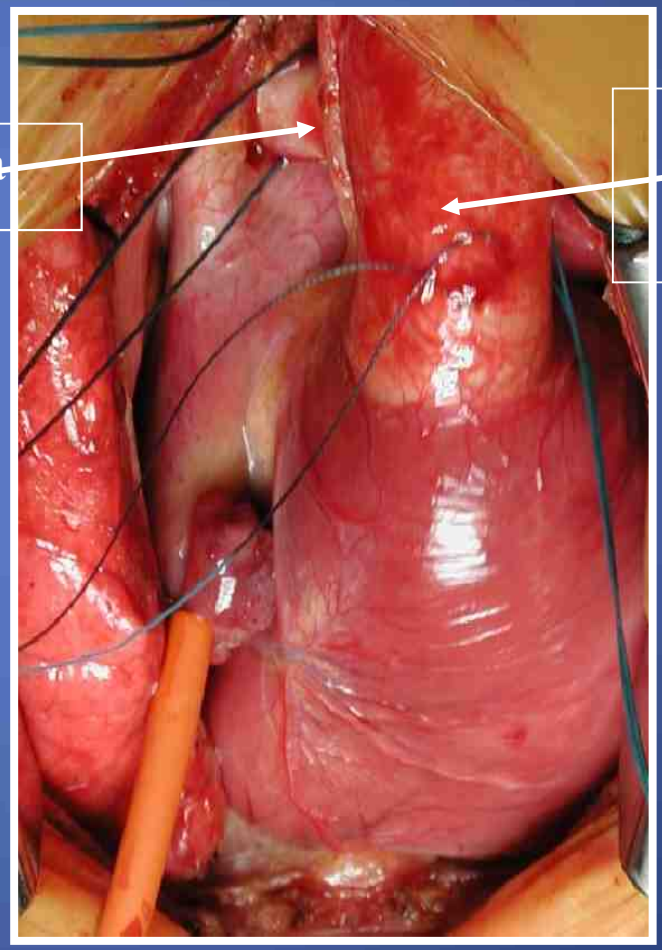
Hybrid Procedure for HLHS

Teiji Akagi, Shinichi Otsuki, Shunji Sano
Okayama University Hospital,
Okayama, Japan

Hypoplastic Left Heart Syndrome





Atrial Morphology

A) Large left atrium, thick prominent septum secondum with thin septum primum adherent
B) Small left atrium with thick, muscular atrial septum
C) Giant left atrium, thin atrial septum with severe mitral regurgitation

Rychik J, et al. JACC 1999

Atrial Morphology and Pulmonary Vascular Histopathology

	Type A	Type B
Arteries	Muscular extension into intra-acinar arteries	Muscular extension into intra-acinar arteries
Veins	Normal	Thick and dilated with arterialized, ≥2 elastic laminae
Lymphatics	Normal or mildly dilated	Severely dilated

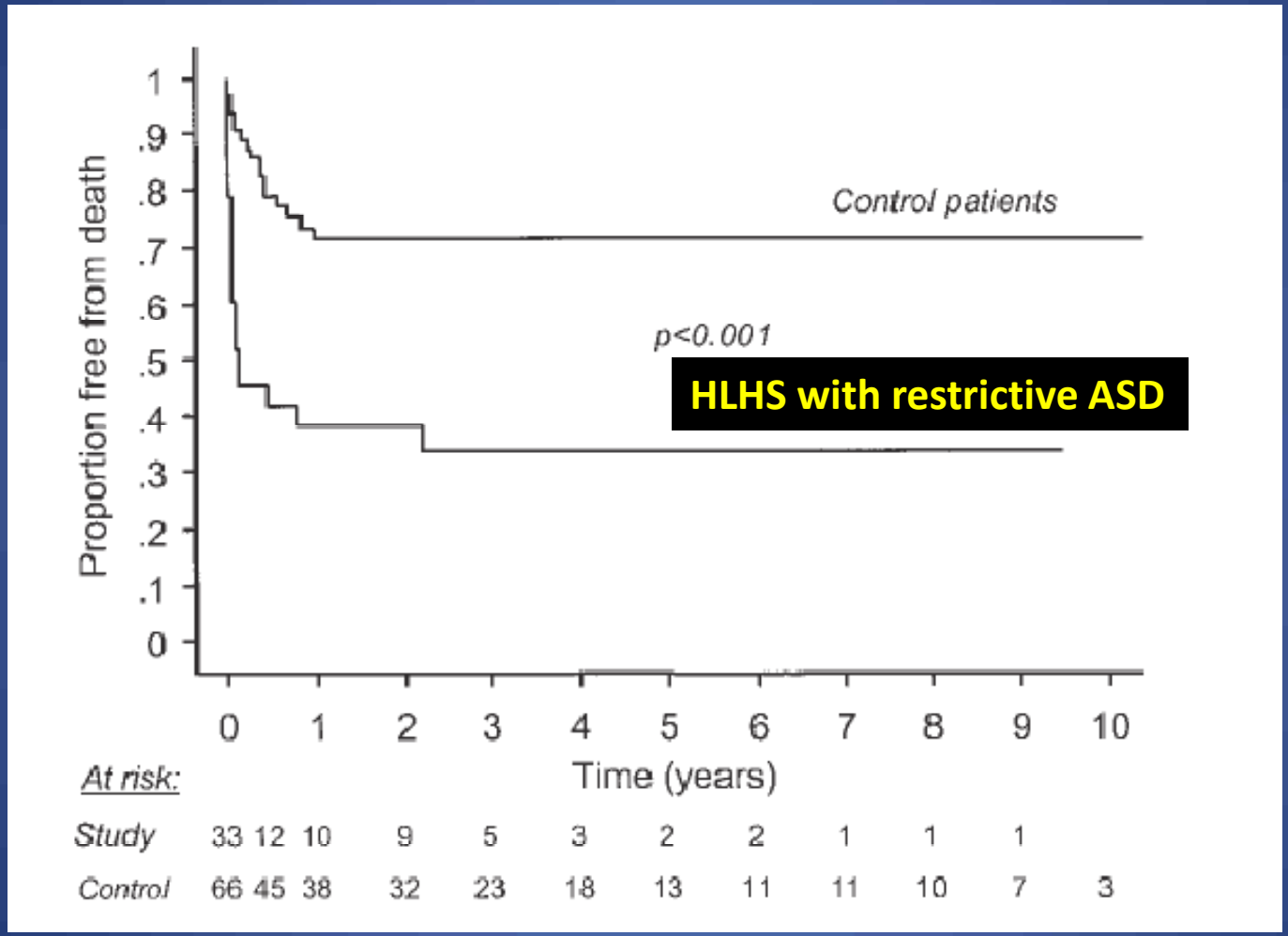
Rychik J, et al. JACC 1999

Atrial Morphology

Type A:	n=12	Survive: 6
Type B:	n=4	Survive: 0
Type C:	n=2	Survive: 0

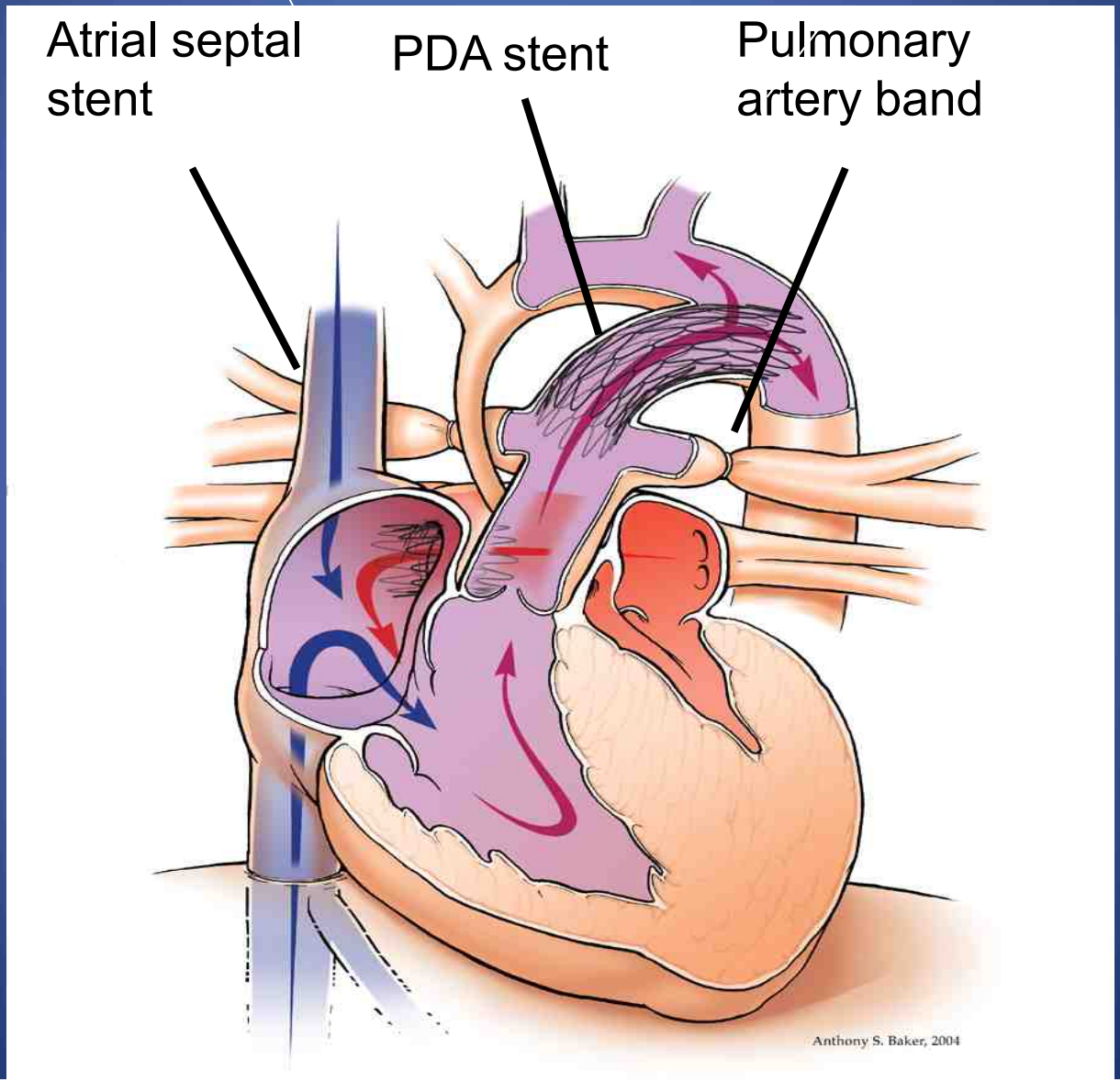
Rychik J, et al. JACC 1999

HLHS with Restrictive Atrial Septum



Vlahos AP, et al. Circulation 2004

Stage I Palliation



RV-PA Shunt to HLHS

The "Sano" modification

- **Initially reported ↑ survival from 53% to 89%**

  Sano, et al, JTCVS, Vol 126: 504-, 2003

- **Recent multi-institutional report from Japan**

  84% survival after Sano, but 1 year survival was 65% & 2 year 63%

  Sano, et al, ATS, Vol 78: 1951-, 2004

- **More recent report 92% survival after Sano, and 5 year survival was 73%**

  Sano, et al, ATS, Vol 87: 178-, 2009

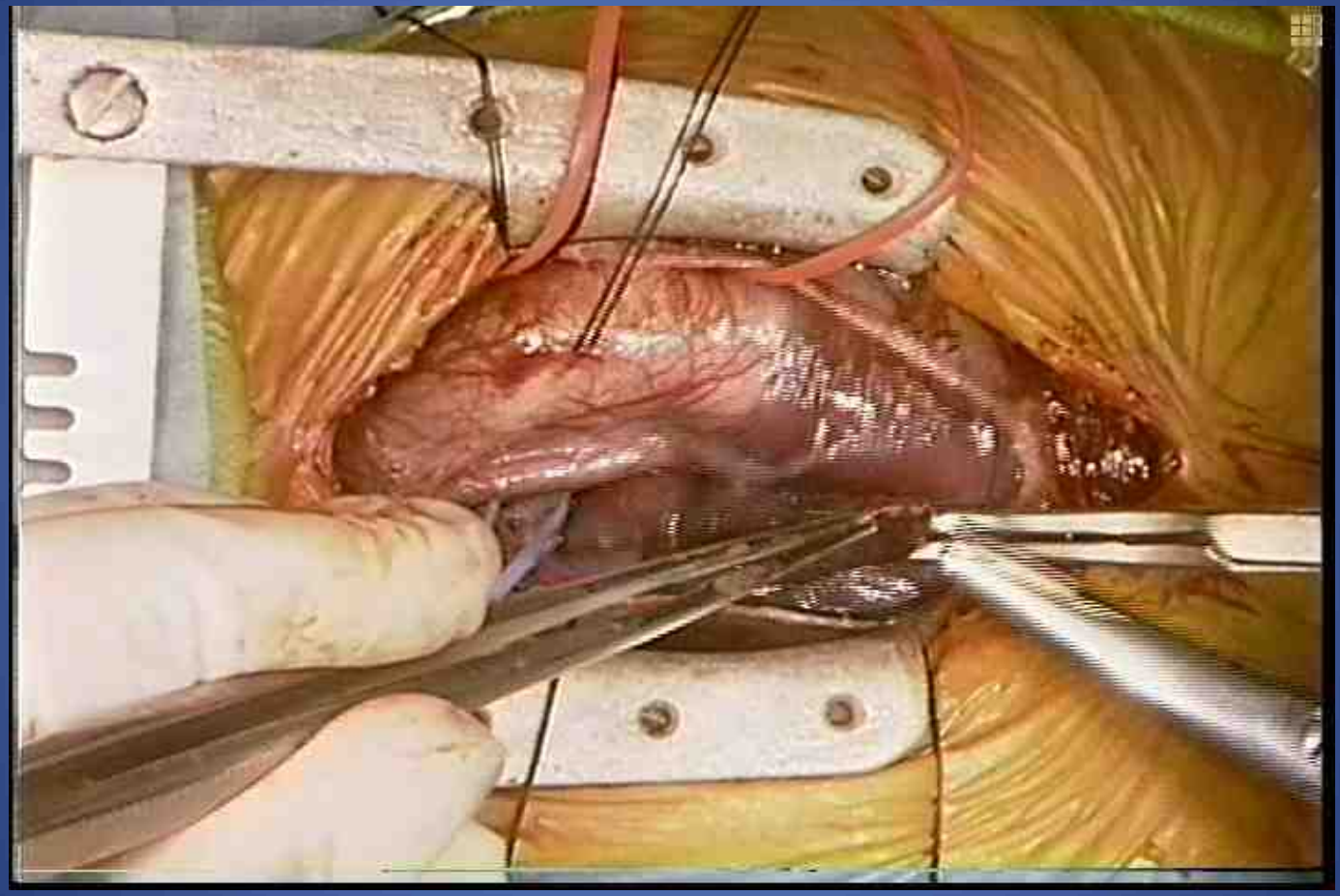
Risk Factor after Repair of HLHS

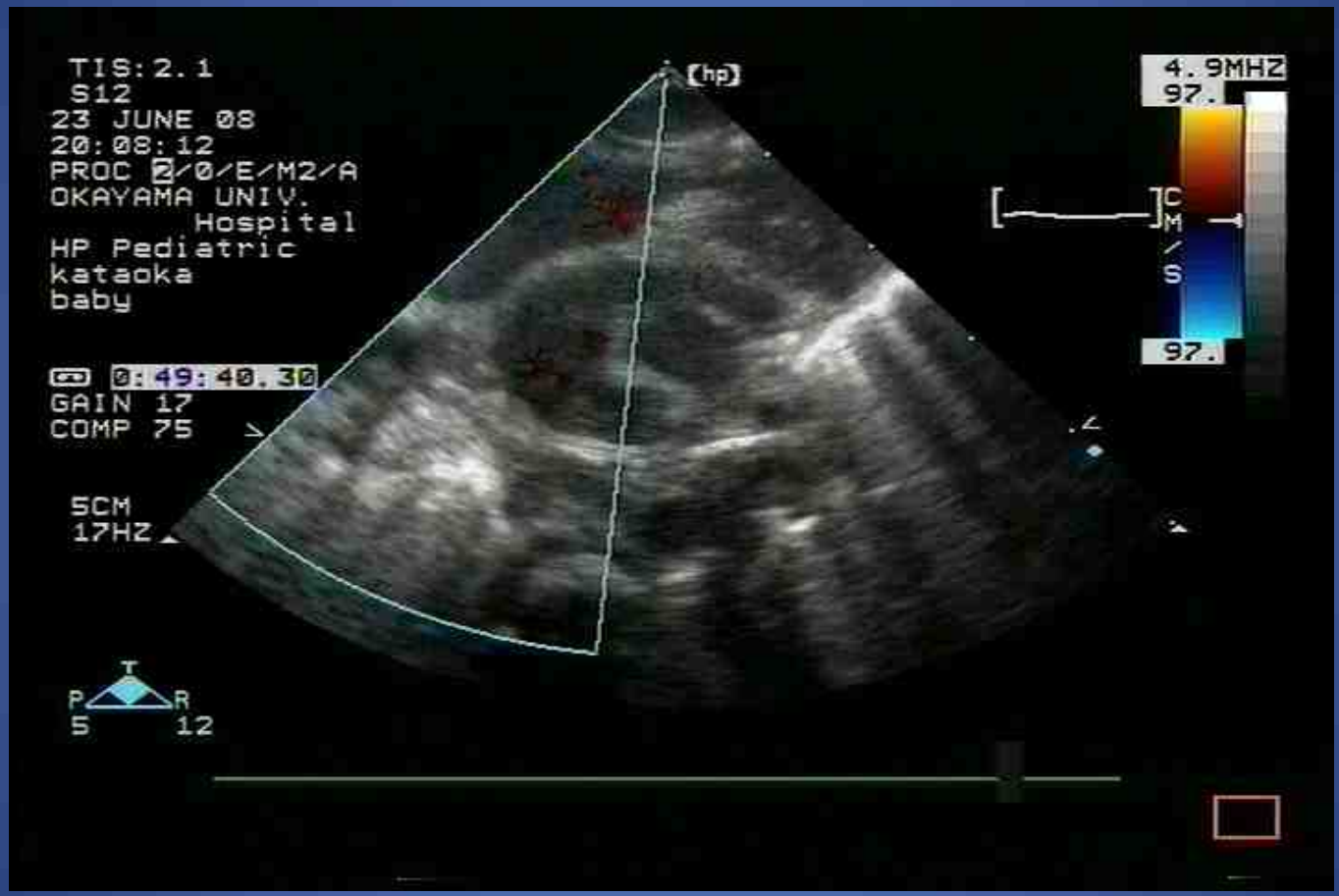
- Mortality after Stage I Palliation using RV-PA Shunt is 7-8% in our institution and more than 90% of the patients with HLHS reached Stage II BDG
- No risk factor after Stage I (RV-PA Shunt)

- Risk factor in mid-long term result
  - Intact atrial septum/ restrictve ASD,
  - BW less than 2.5kg
  - Associated with Non–Cardiac Anomalies

Difficulties of catheter intervention for restrictive ASD

- Critical condition
- Thick muscular atrial septum
- Small left atrium
- Limited echo window (space)
- Limited vascular access
- Less efficacy if conventional technique

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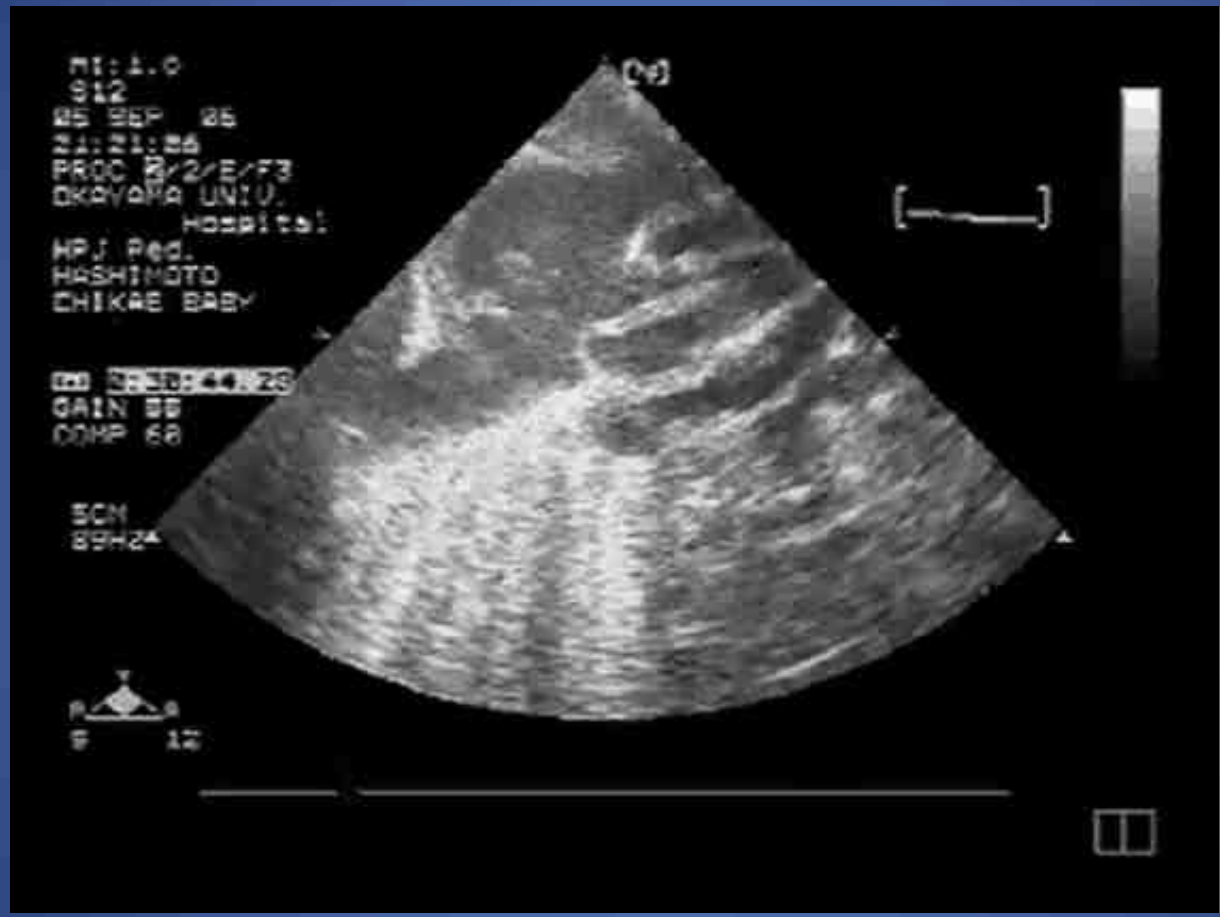
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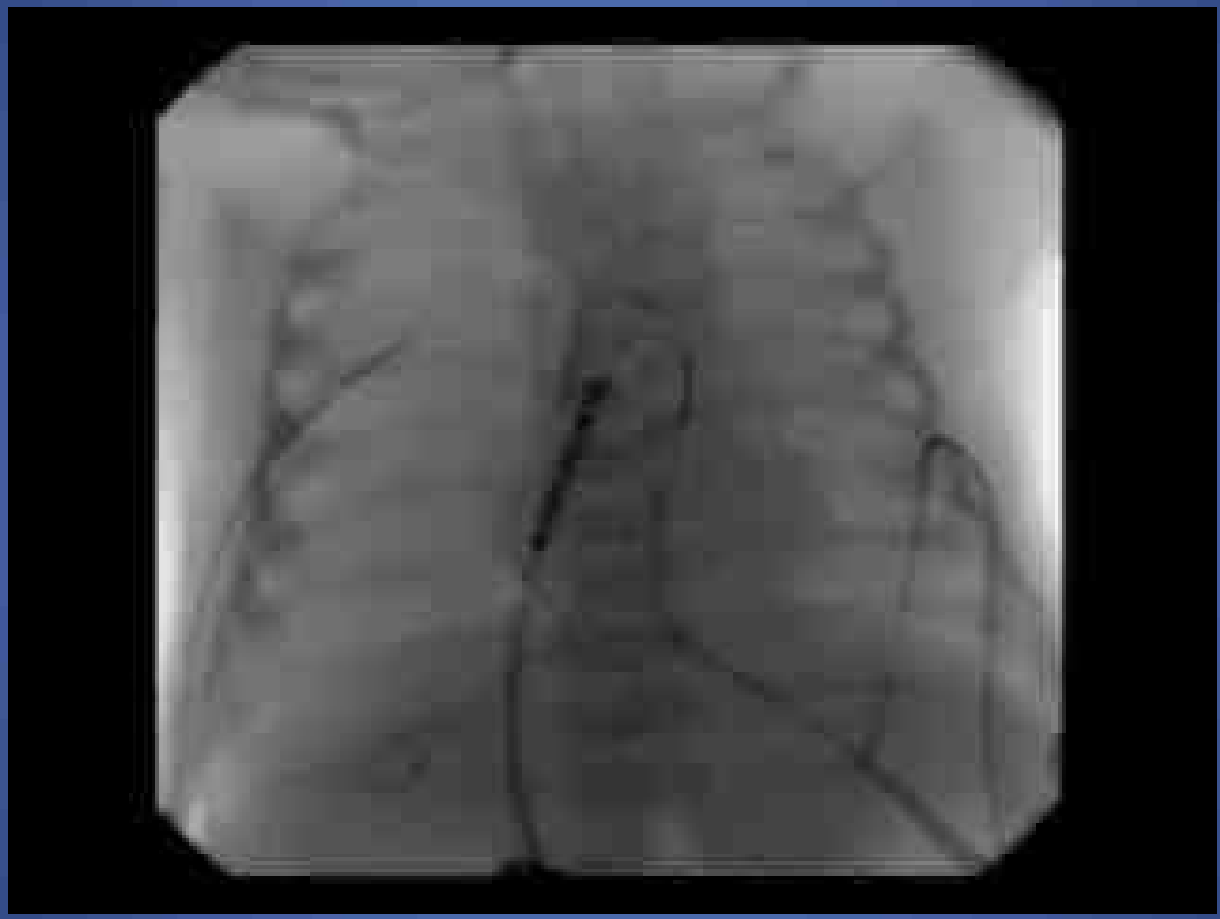
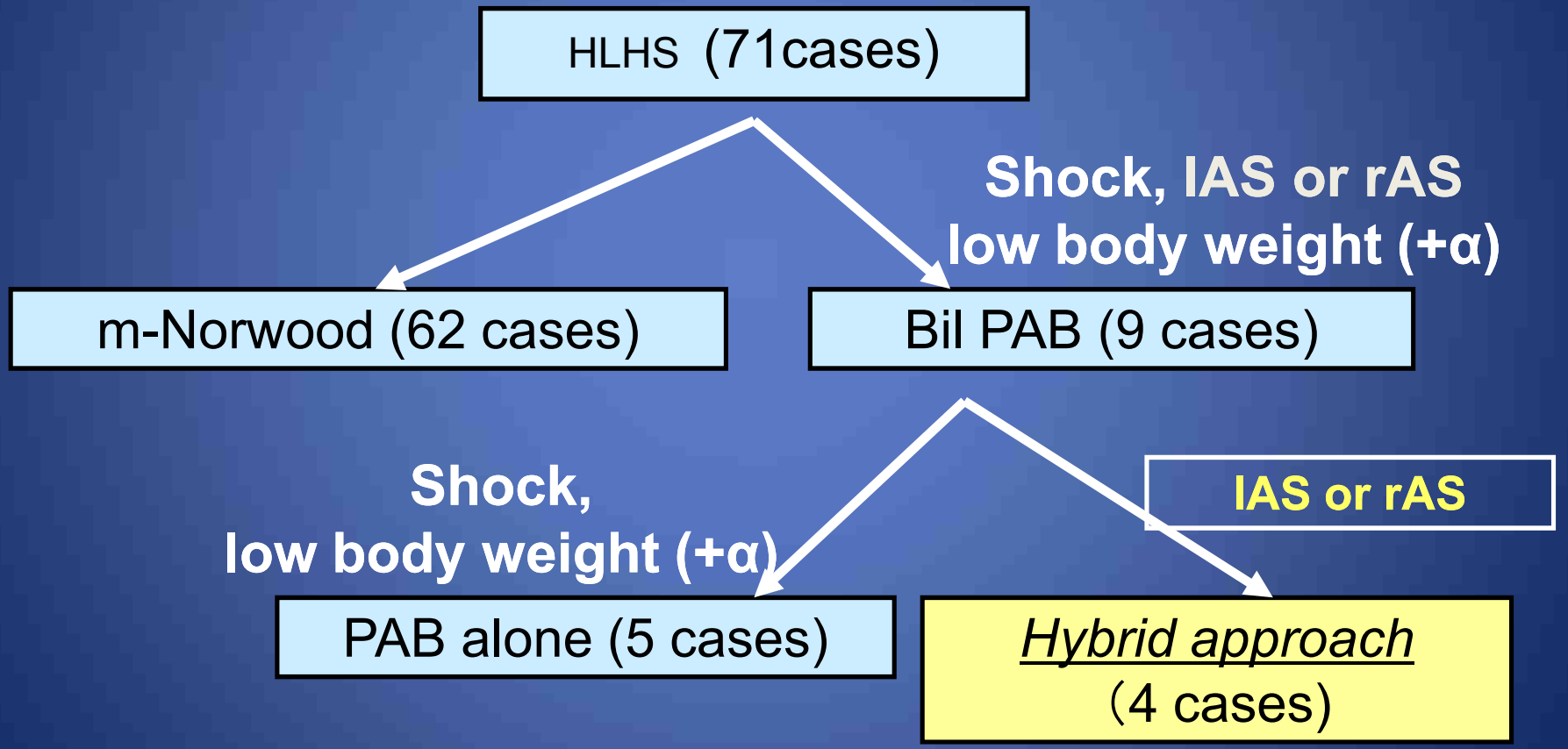


Recent Treatment Strategy to HLHS

(2005.8~2008.8)



Demographic Data

Case	BW	Diagnosis	ASD	Fetal Diag.	Time to op. (hrs)
1	2700g	HLHS/IAS	-	-	23
2	2660g	HLHS/IAS	-	+	0
3	1650g	HLHS/rAS	2.6m/s (L→R)	-	4 d
4	2470g	HLHS/IAS	-	+	0

Hybrid approach: Post Op Course

Case	PAB size(mm)	Baloon size(mm)	Post op ASD	re-BAS
1	3.5	7	2.9mm, 1.2m/s	× 2
2	3.0	10	2.3mm, 2.0m/s	stent
3	2.8	7	3.8mm, 0.9m/s	failure
4	3.0	7	2.4mm, 1.3m/s	× 1

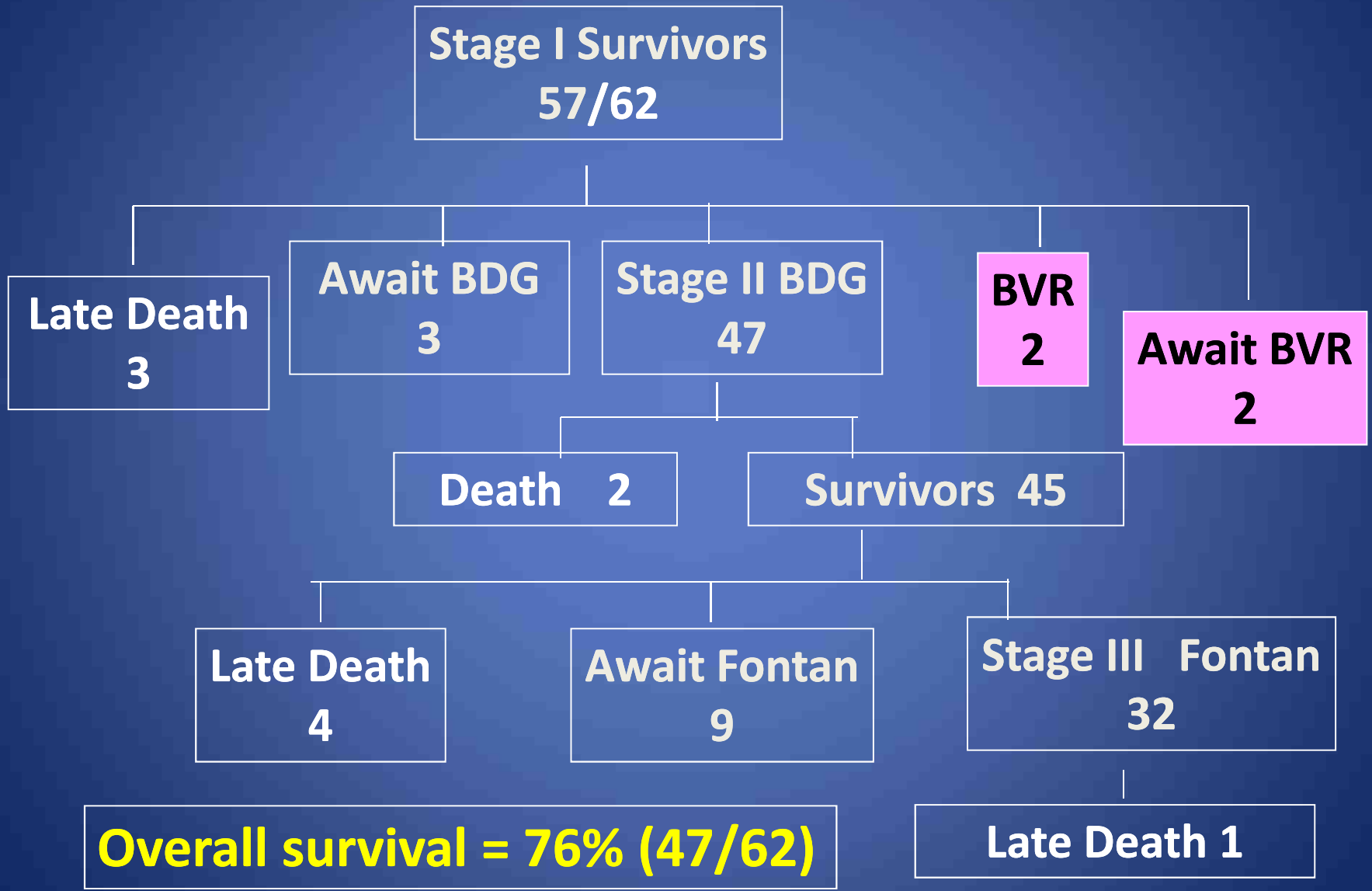
Result of Hybrid Approach

Case	Age (m)	ASD	TR	Post-op ECMO	Outcome
1	3	3.5mm, 1.5m/s	trivial	+ PH desaturation	HD due to PH
2	1	8.0mm, 1.3m/s	trivial	-	BDG
3	1	2.7mm, 1.9m/s	trivial	+ LOS	HD Due to PR
4	2	2.7mm, 2.0m/s	moderate	-	BDG

Conclusions

- Transcatheter decompression of the LA for patients with HLHS is still challenging procedure. However, procedure can be performed safely, reduces the transatrial gradient, and improves oxygenation.
- Catheter intervention can contribute survival of this condition compared to conventional emergent Norwood procedure.

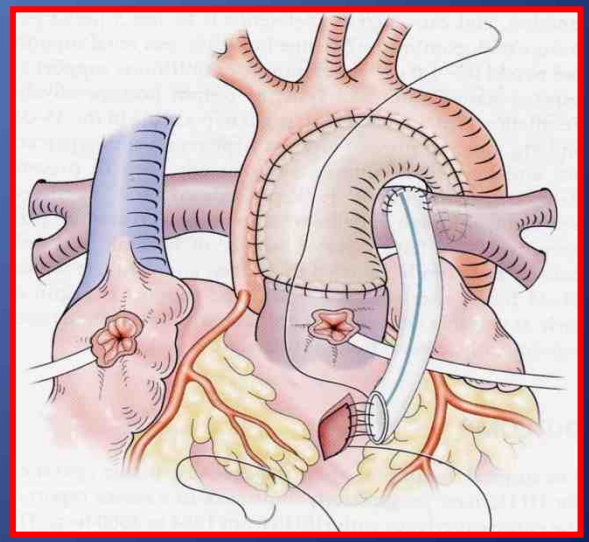
Current Status of Survivors



Okayama Experience - Patients

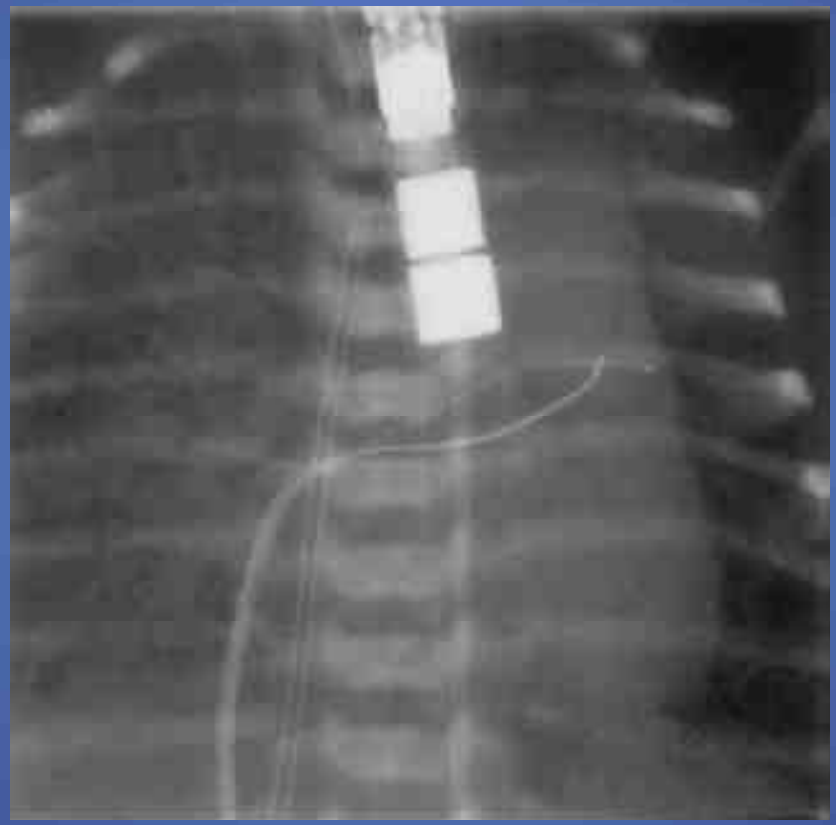
Okayama University Hospital

- February 1998 – June 2007
- 62 infants (36 boys & 26 girls)
- Age : 3 – 57 days (median, 9 days)
  >14 days : 11 infants
- Weight : 1.6 – 3.9 kg (median, 2.7 kg)
  7 infants < 2.0 kg
  18 infants < 2.5 kg
- Prematurity <37w : 4 infants



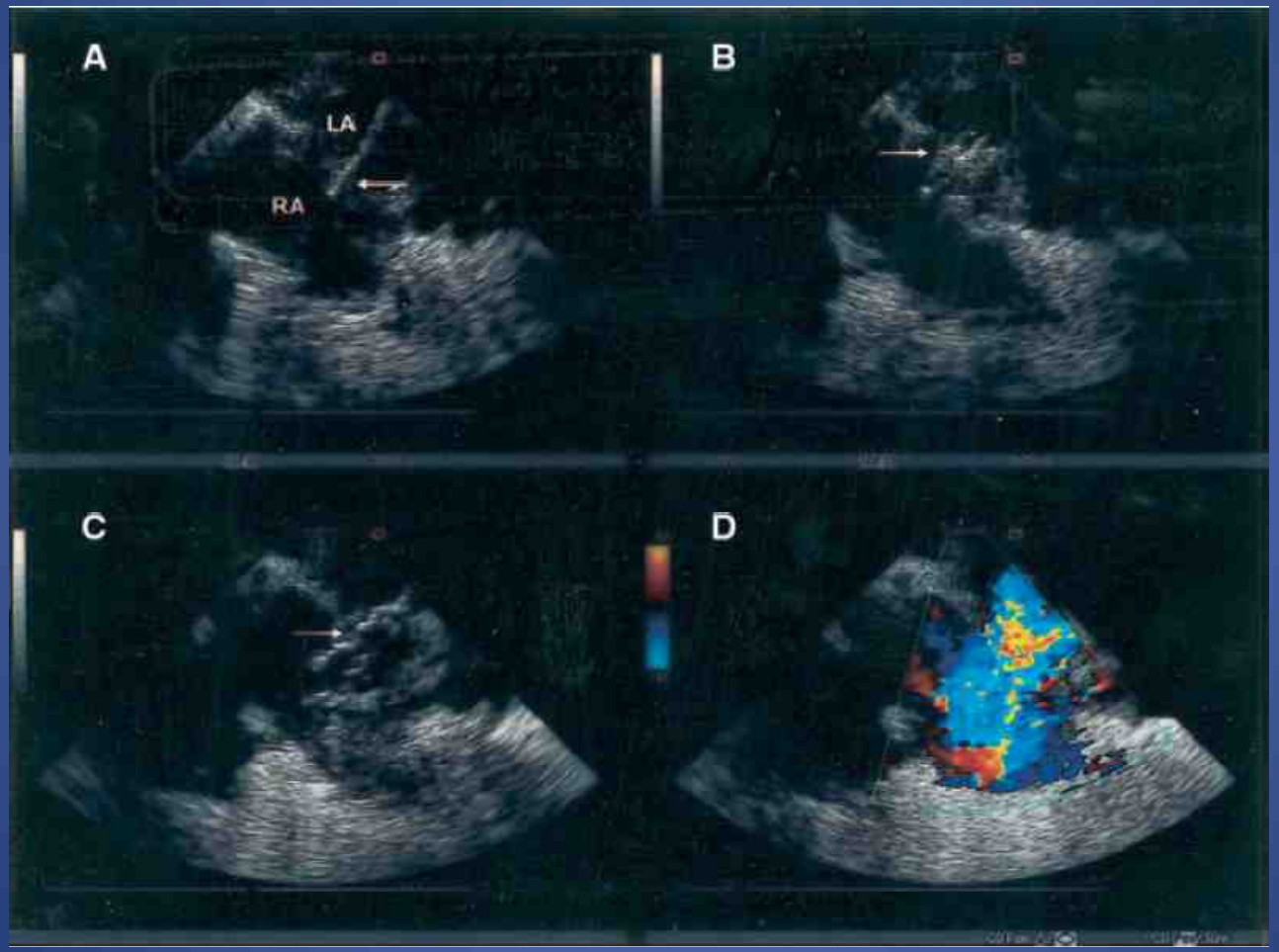
exclude Bilateral PAB : 7 infants

Radiofrequency-Assisted Atrial Septoplasty



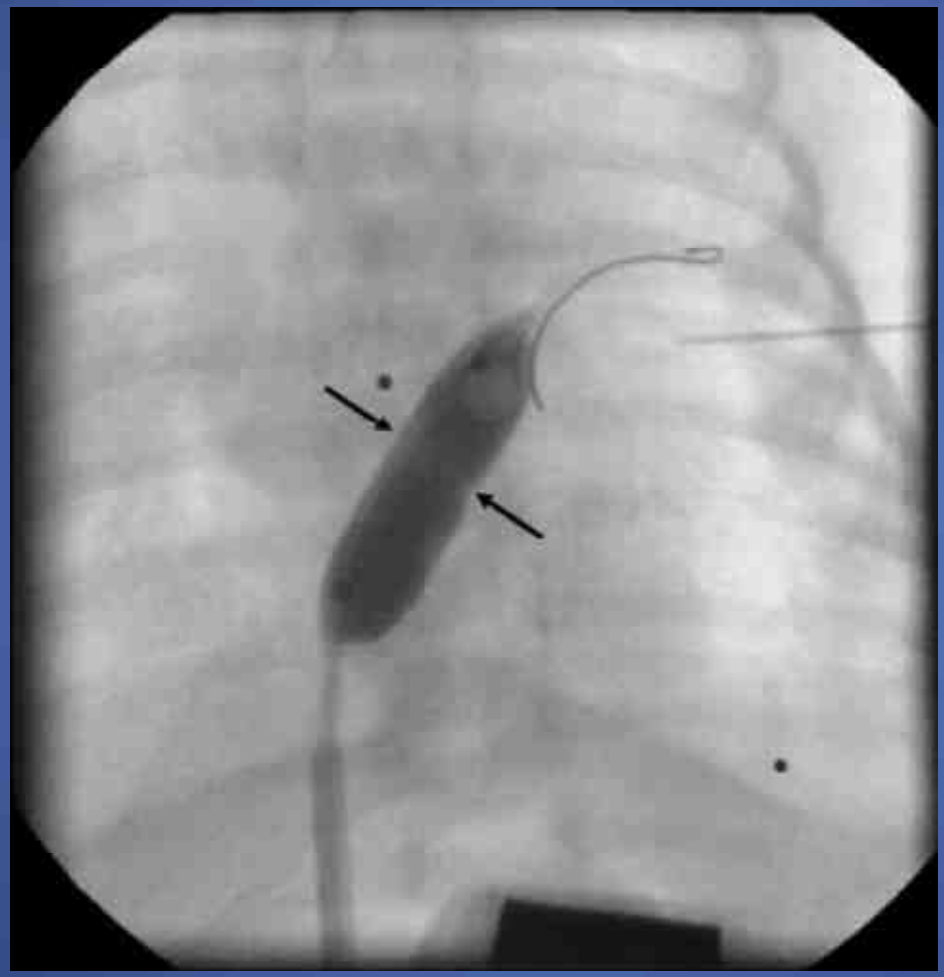
Du Marchie Sarvaas et al. CCI 2002

Radiofrequency perforation and Cutting balloon septoplasty



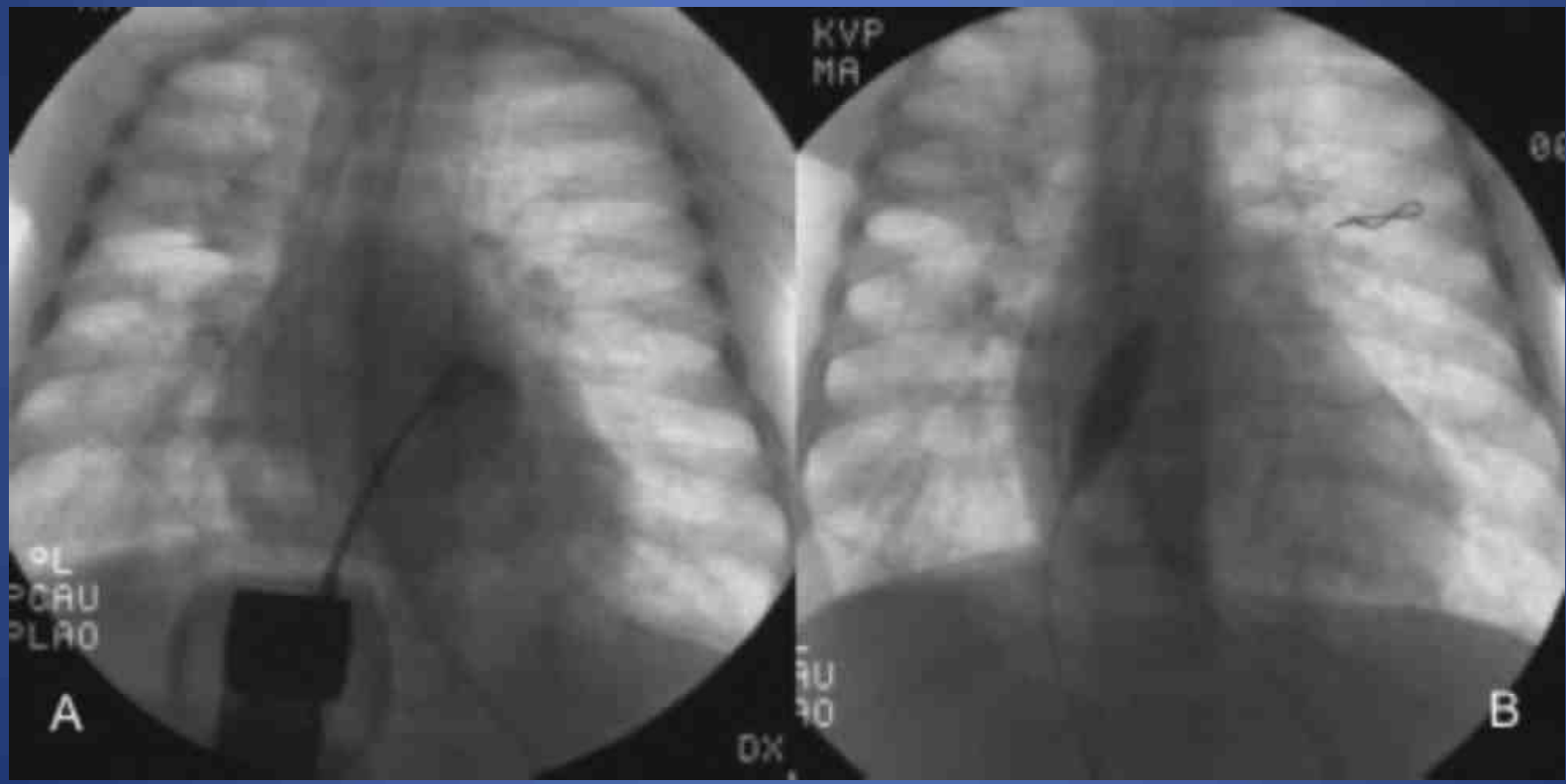
Hill et al. CCI 2005

Umbilical vein approach



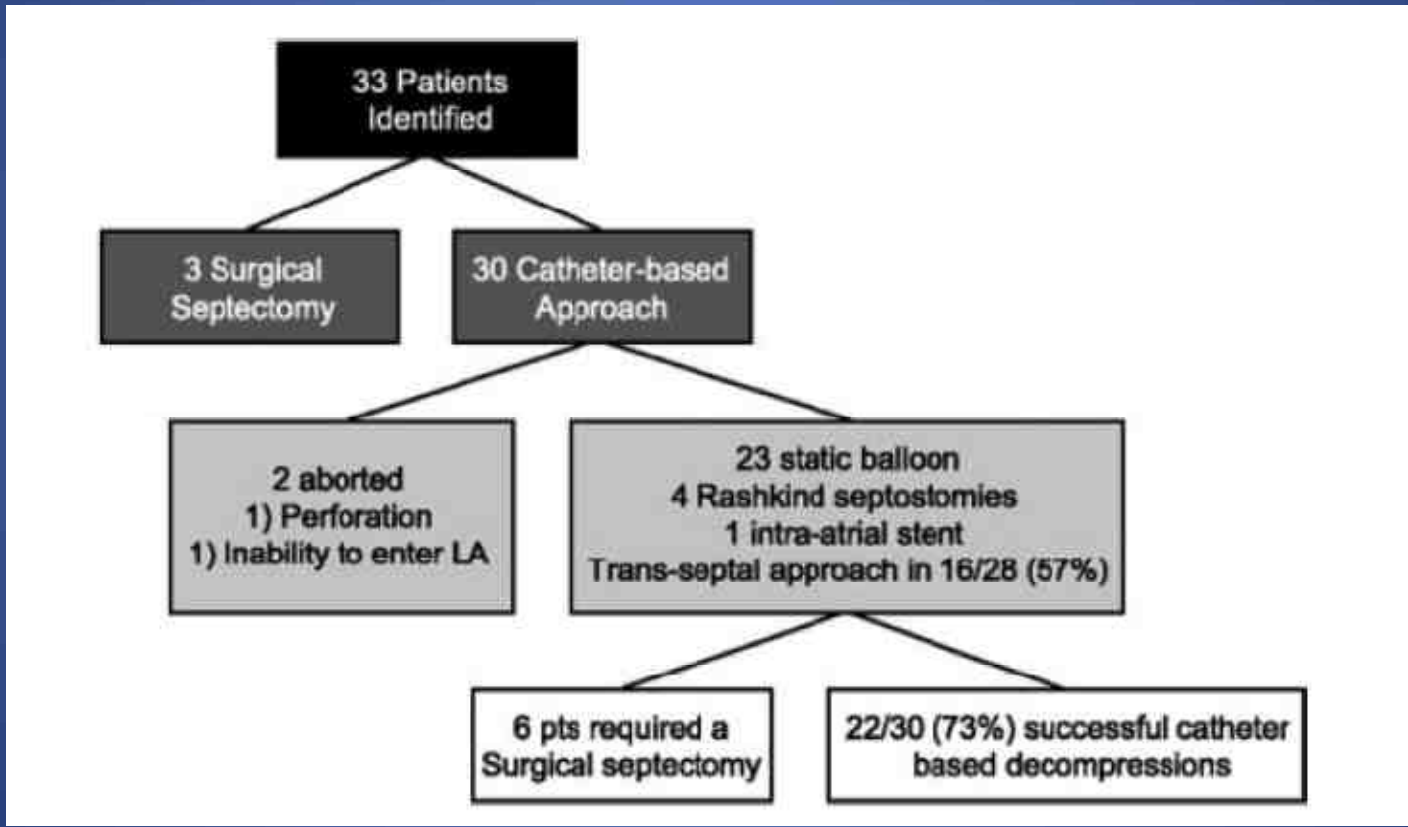
Javois et al. CCI 2005

Transhepatic approach



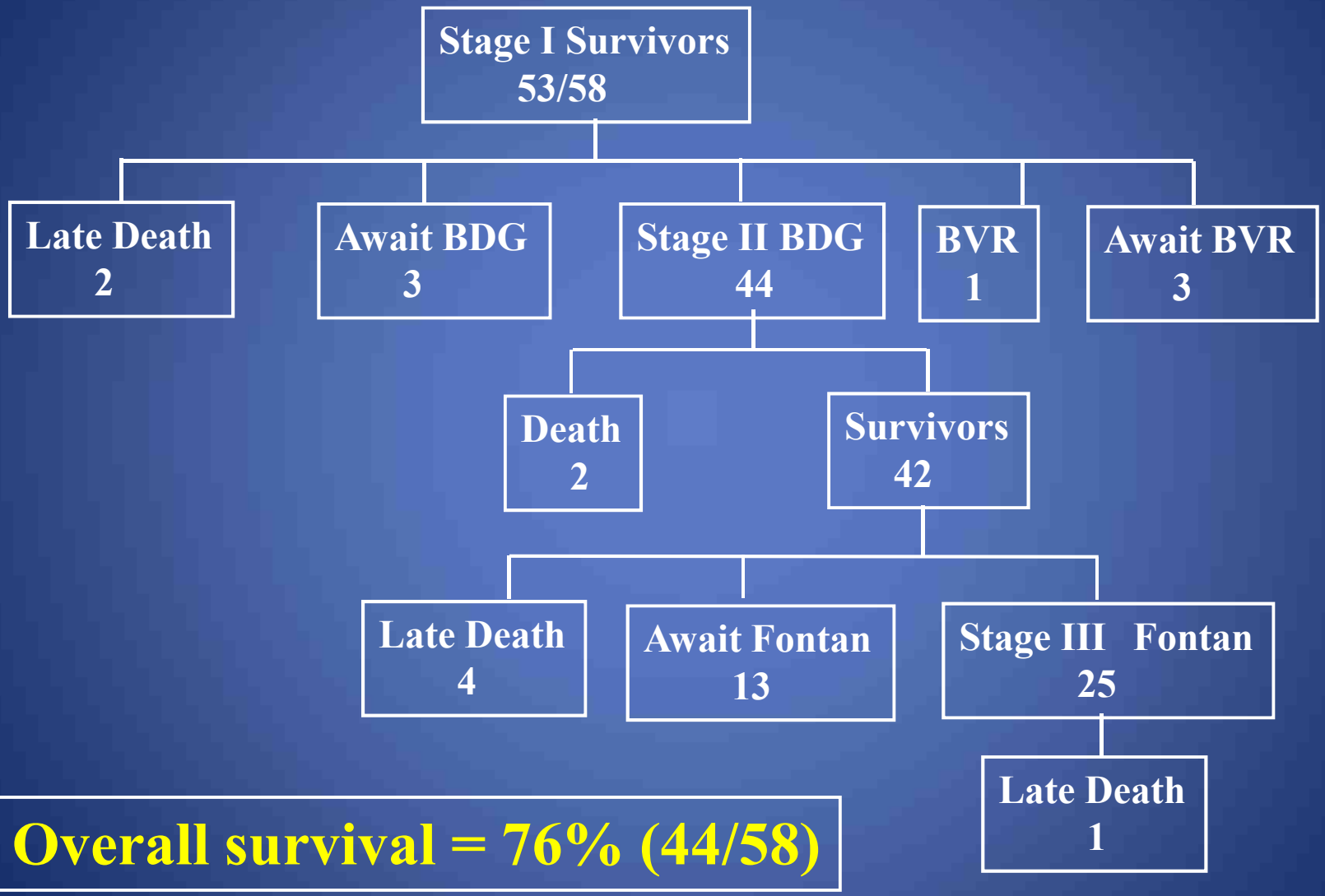
Pedra et al. CCI 2007

Role of catheter intervention for HLHS



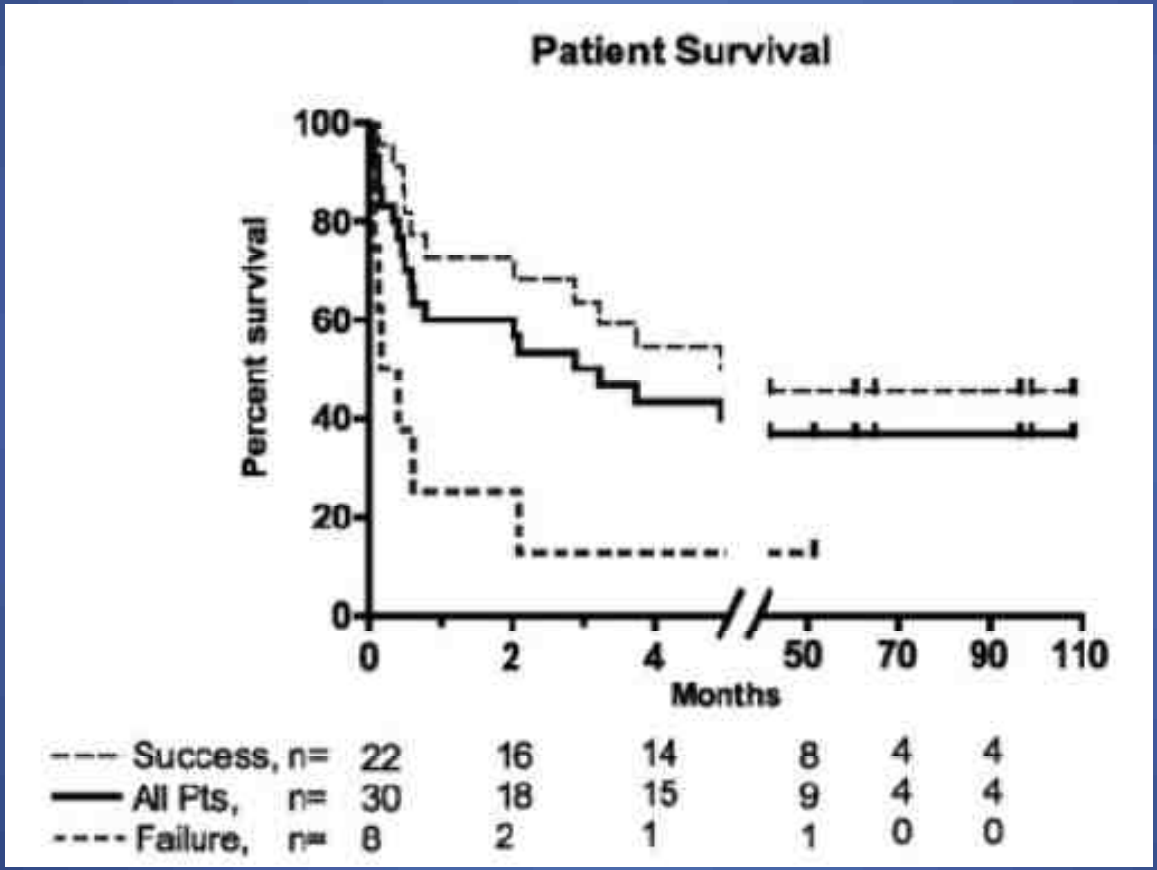
Gossett et al. CCI 2006

Current Surgical Outcome



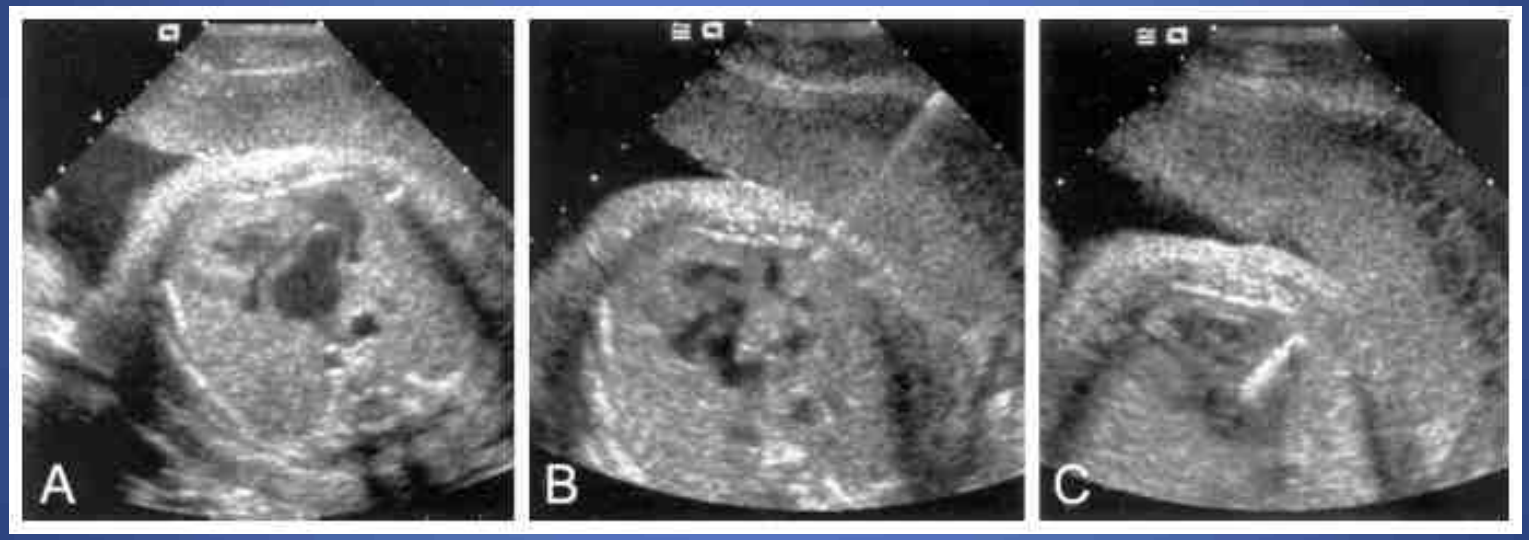
Overall survival = 76% (44/58)

Role of catheter intervention for HLHS



Gossett et al. CCI 2006

Creation of ASD in Utero



Marshall AC, et al. Circulation 2004

Background

- Historically, surgeons and interventionalists have had a somewhat competitive relationship, especially in adult cardiac disease
- Each have been thinking of how to treat patients by their own speciality
- The management of CHD is evolving due to advances in transcatheter therapies that coincide with surgical strategies to improve outcomes

Hybrid Approach – What is it ?

- Collaborative effort between surgeons and interventional cardiologists
- Collaborative effort between physicians and industry
- Sharing of ideas, expertise, equipment, & techniques
- Development of novel treatment strategies

Why ?

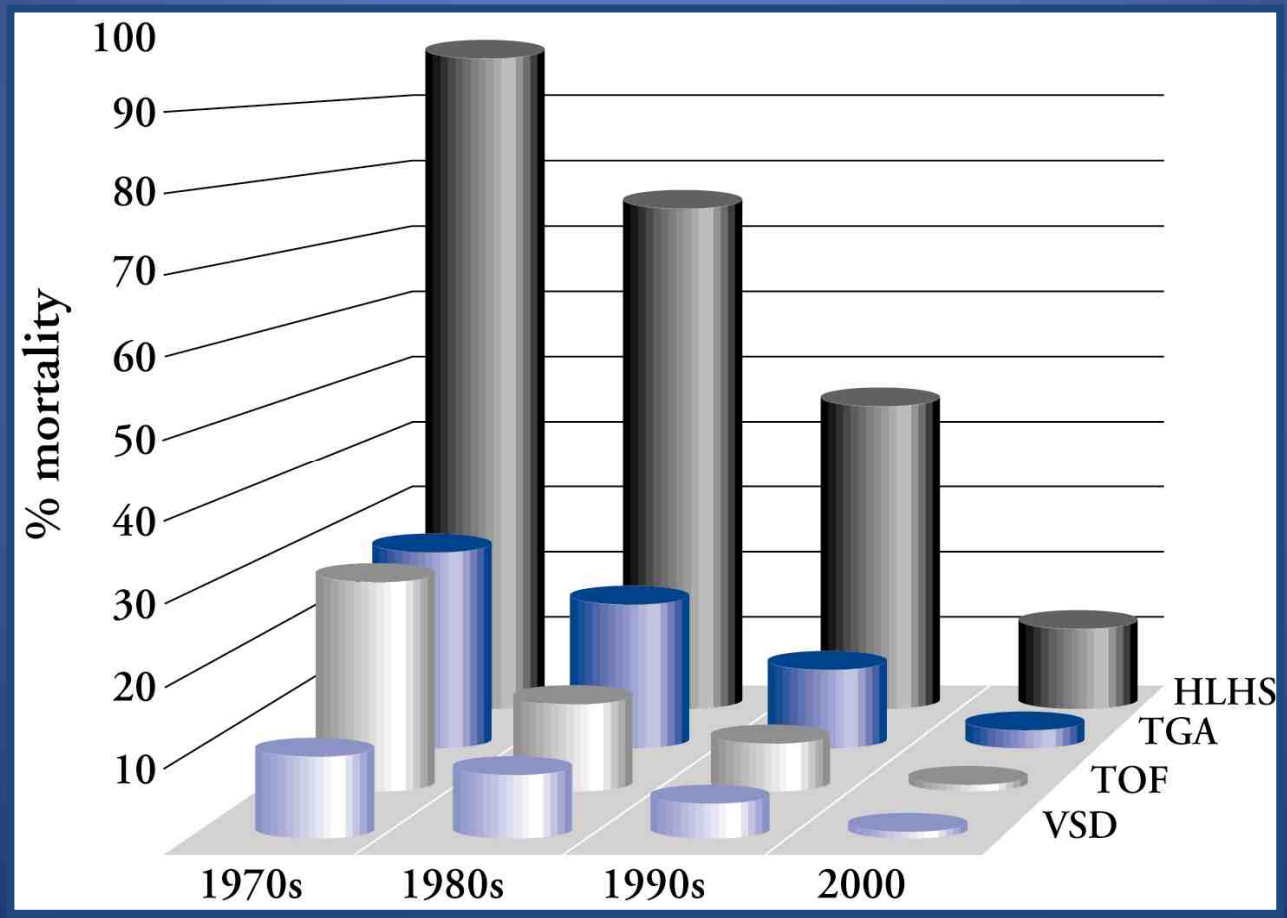
- Offer best treatment to the patients
- Reduce morbidity & mortality
- Improve quality of life
- Deliver more efficient & cost effective care

Hybrid Cardiac Procedures for CHD

- Group I: intraoperative stents
- Group II: perventricular muscular VSD
- Group III: ***PA bands & PDA stent***
- Group IV: young adults requiring combined interventional & EPS
- Group V: unusual Hybrid procedures

Hypoplastic Left Heart Syndrome

-A Benchmark for the Surgical Treatment of Congenital Heart Disease-



Hybrid Approach to HLHS

So What's The Big Deal ?

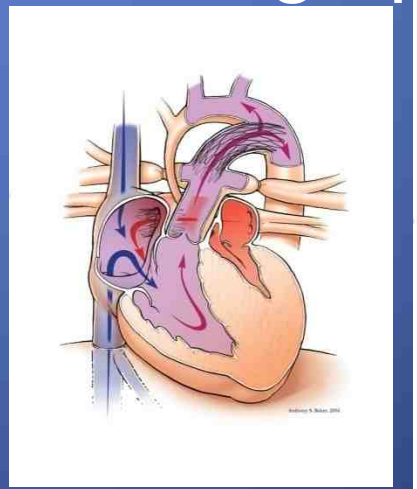
- *Neurologic & Developmental Morbidity*
  - After Norwood repair: Full Scale IQ
    - Kern, et al: 91
    - Mahle, et al: 86
    - Goldberg, et al: 94
  - Abnormalities of speech & language, oral aversion & poor feeding, poor adaptive behavior, & growth failure
  - Later, there is significant emotional & behavior dysfunction, low self esteem, & psychosocial and physical health issues

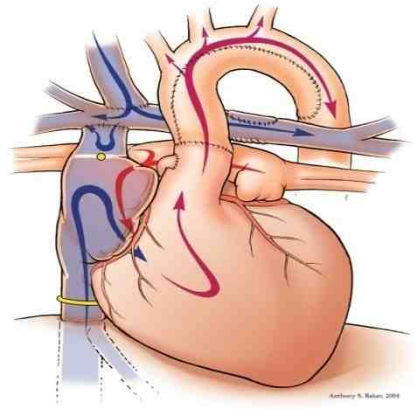
Wernovsky & Newburger, J Peds, Vol 142: Jan, 2003

Hybrid Approach to HLHS

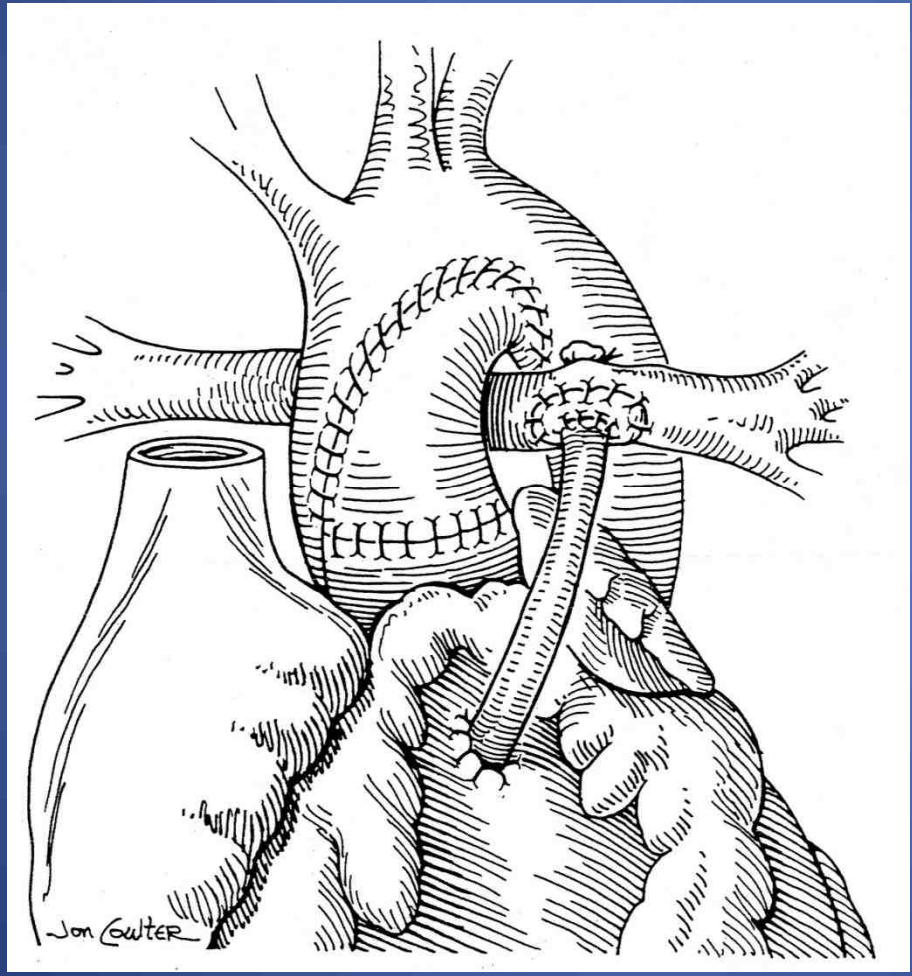
"Hybrid" Concept Of HLHS Repair

- Less invasive procedures (Bilateral PAB +/- Stent)
- Avoid open heart surgery in Neonate , DHCA
- One comprehensive open heart procedure at an age appropriate for the "big operation"
  - Stage 1 Neo-aortic reconstruction
  - Bidirectional Glenn

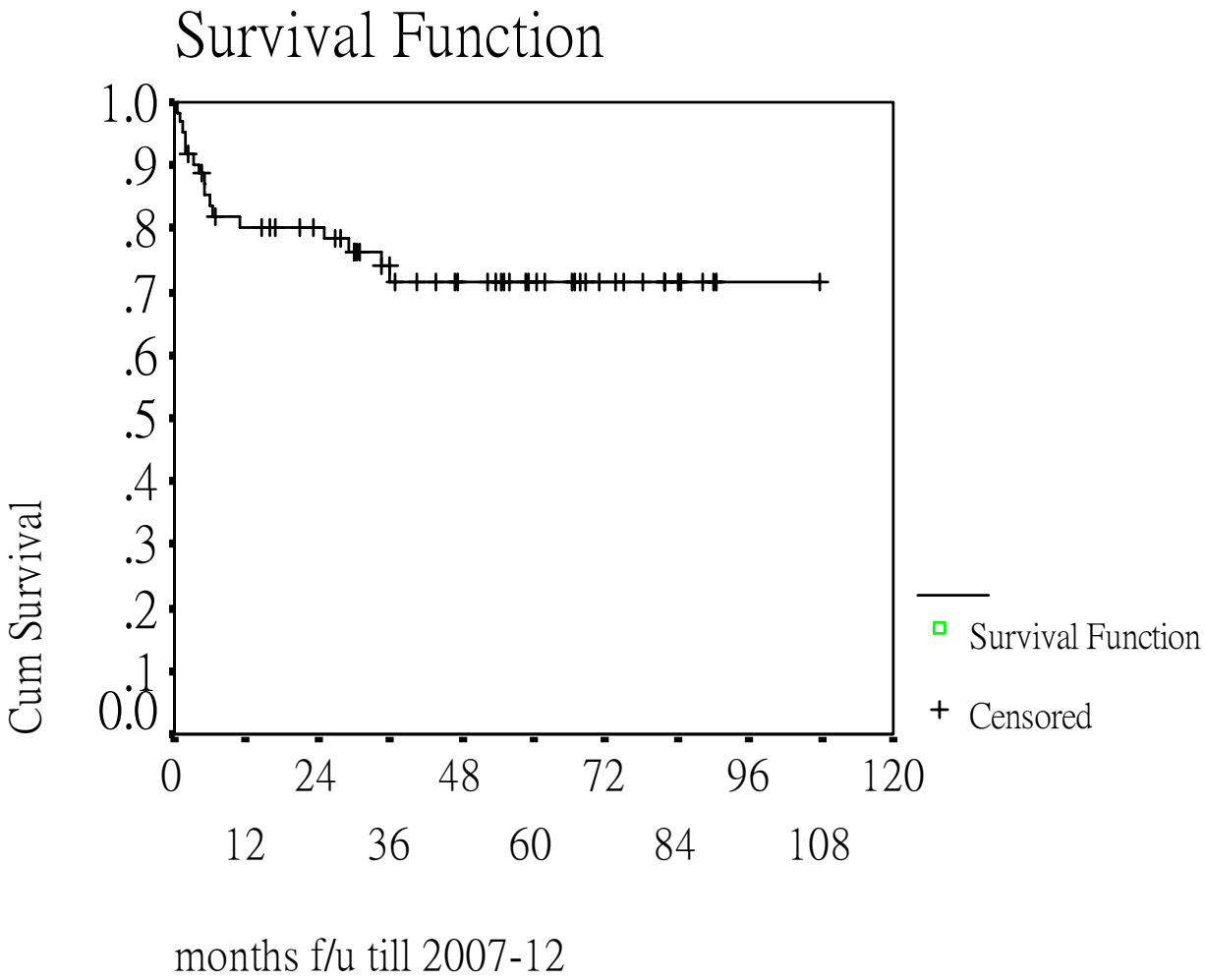




Challenge in the management of HLHS



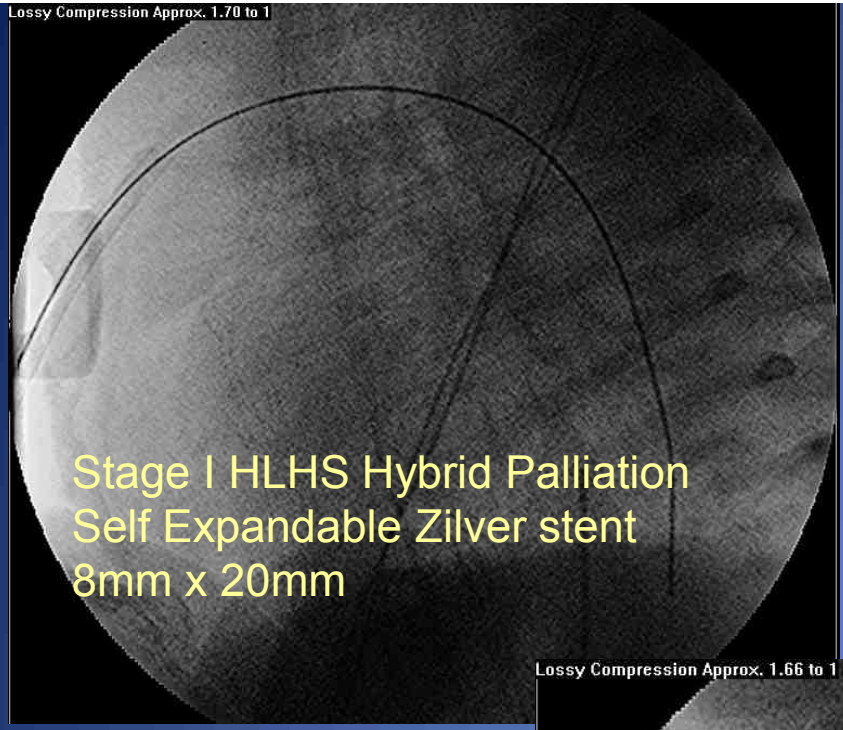
Overall survival rate

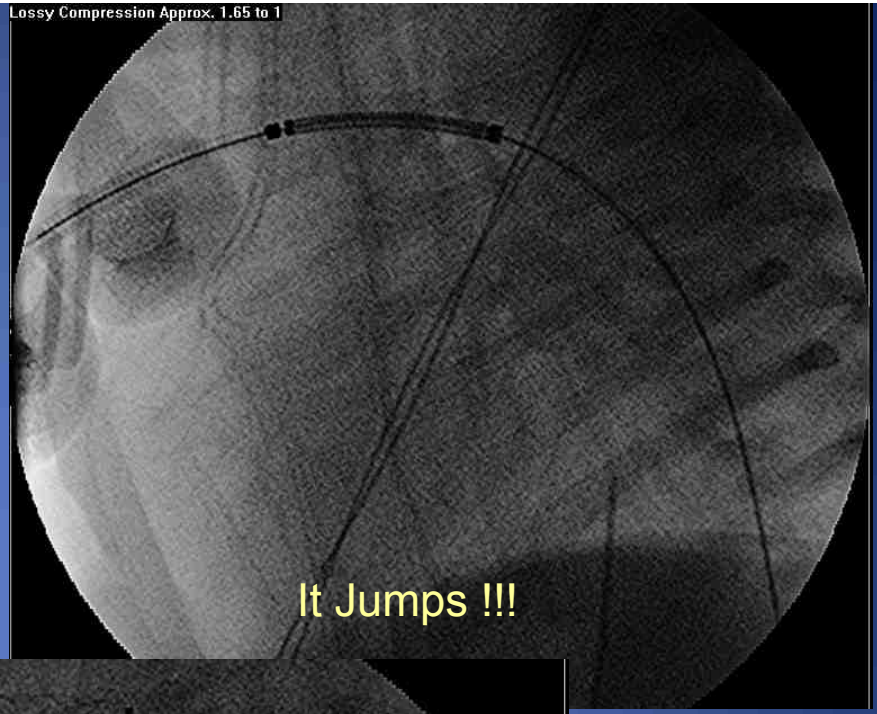


one-year survival rate : 80%

Five-year survival rate : 73%


Hybrid Approach
Hybrid Cathe / OR


Lossy Compression Approx. 1.70 to 1
Stage I HLHS Hybrid Palliation
Self Expandable Zilver stent
8mm x 20mm


Lossy Compression Approx. 1.65 to 1
It Jumps !!!


Lossy Compression Approx. 1.66 to 1

Ductal stent overriding the isthmus at 5 months

Indication of Hybrid Procedure

- **HLHS/IAS, HLHS/rAS**
- **BW< 2.0-2.5 Kg**
- **Poor preoperative condition**
- **Shock**
- **Severe renal failure , liver failure**
- **Sepsis**

New Hybrid Strategy

Okayama University

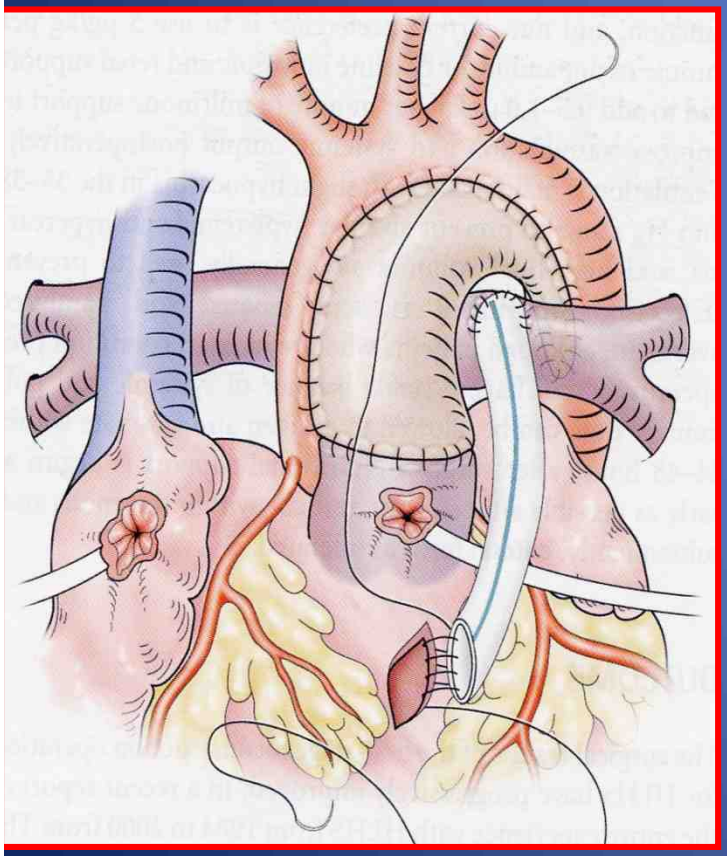
New Hybrid Procedure

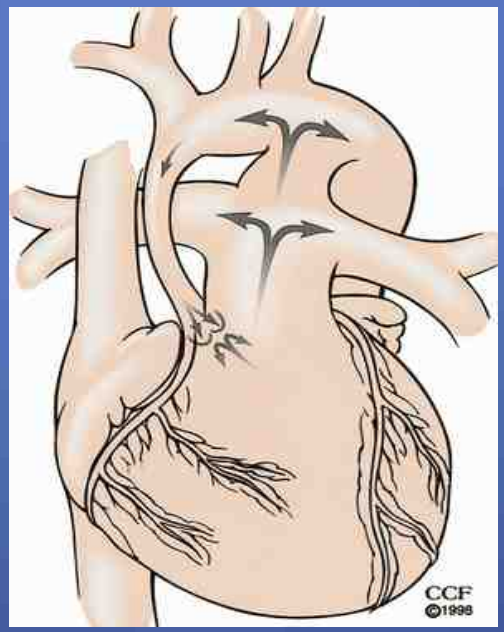
- Bilateral PAB
- ASD creation

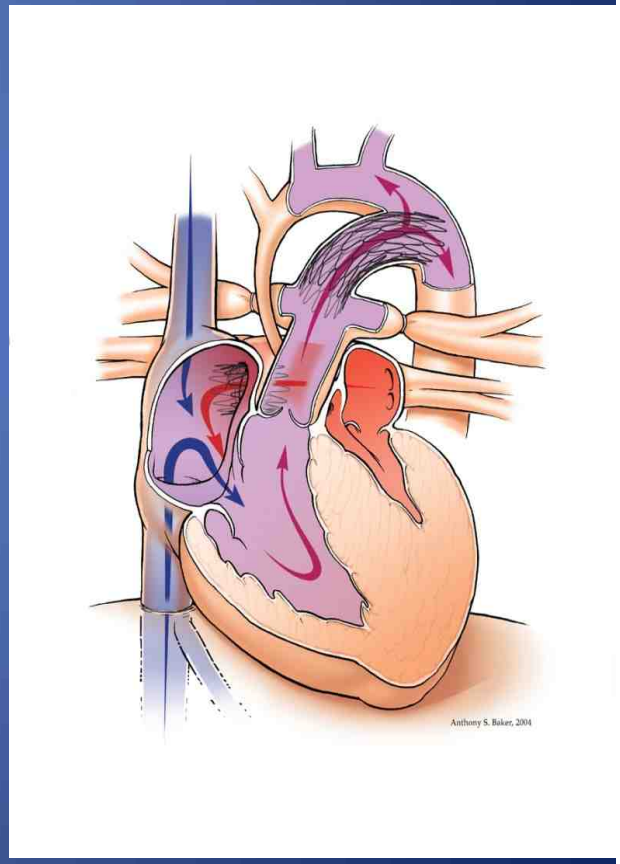
Without using CPB

Strategy in the management of HLHS- Recent Era

RV-PA Shunt Bilateral PAB ± Stent







Strategy in the management of HLHS

Recent Era ~ Near Future

Fetal Intervention

- To create atrial septal defect
- To decrease PVR
- To stimulate growth of LV and Aorta