Pulmonary Atresia and intact ventricular septum:

Anesthetic Considerations for Palliative and Definitive Procedures

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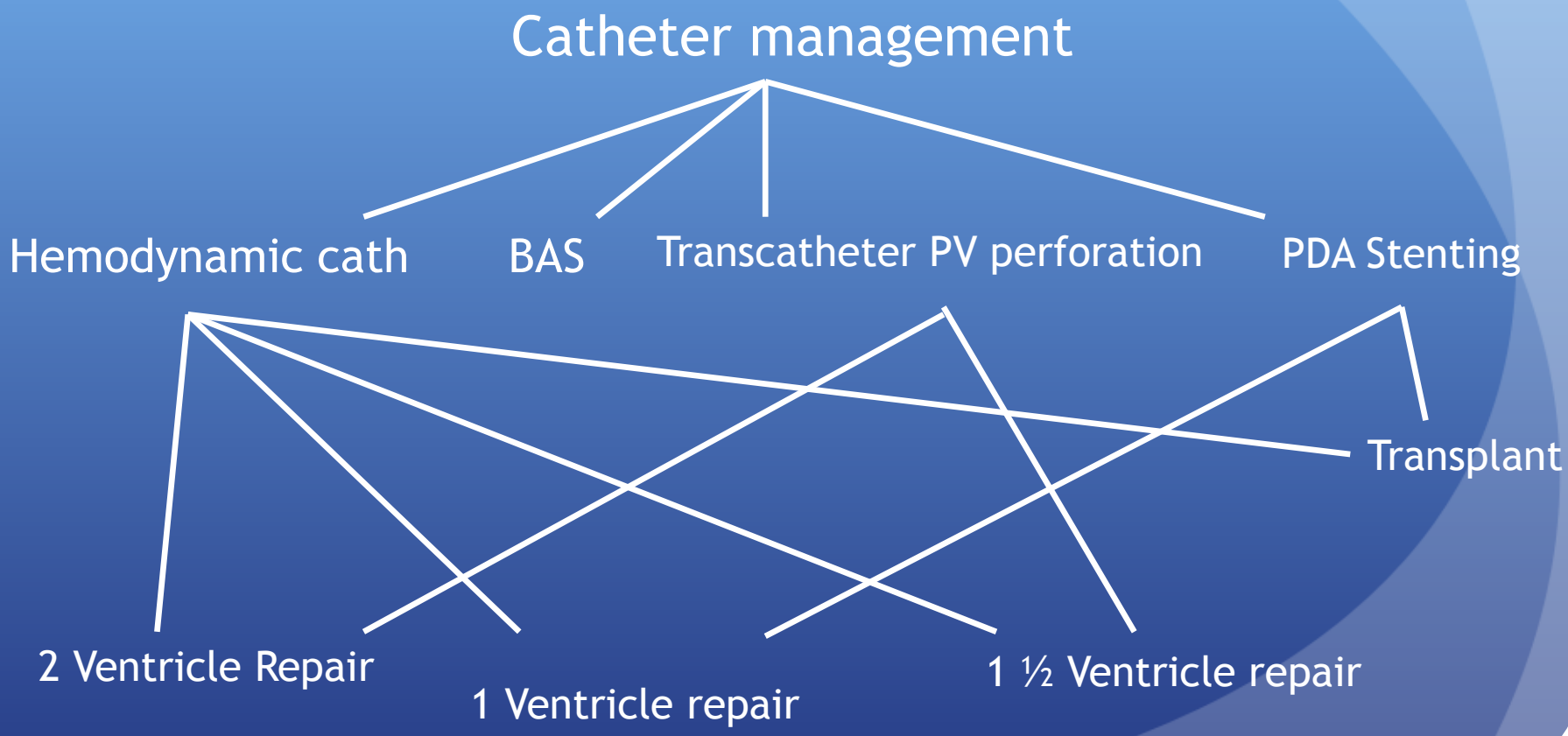
- No disclosures

PAIVS rare lesion yet morphologically heterogeneous lesion

- Anesthetic considerations depends on the surgical management at different stages

Management of PAIVS

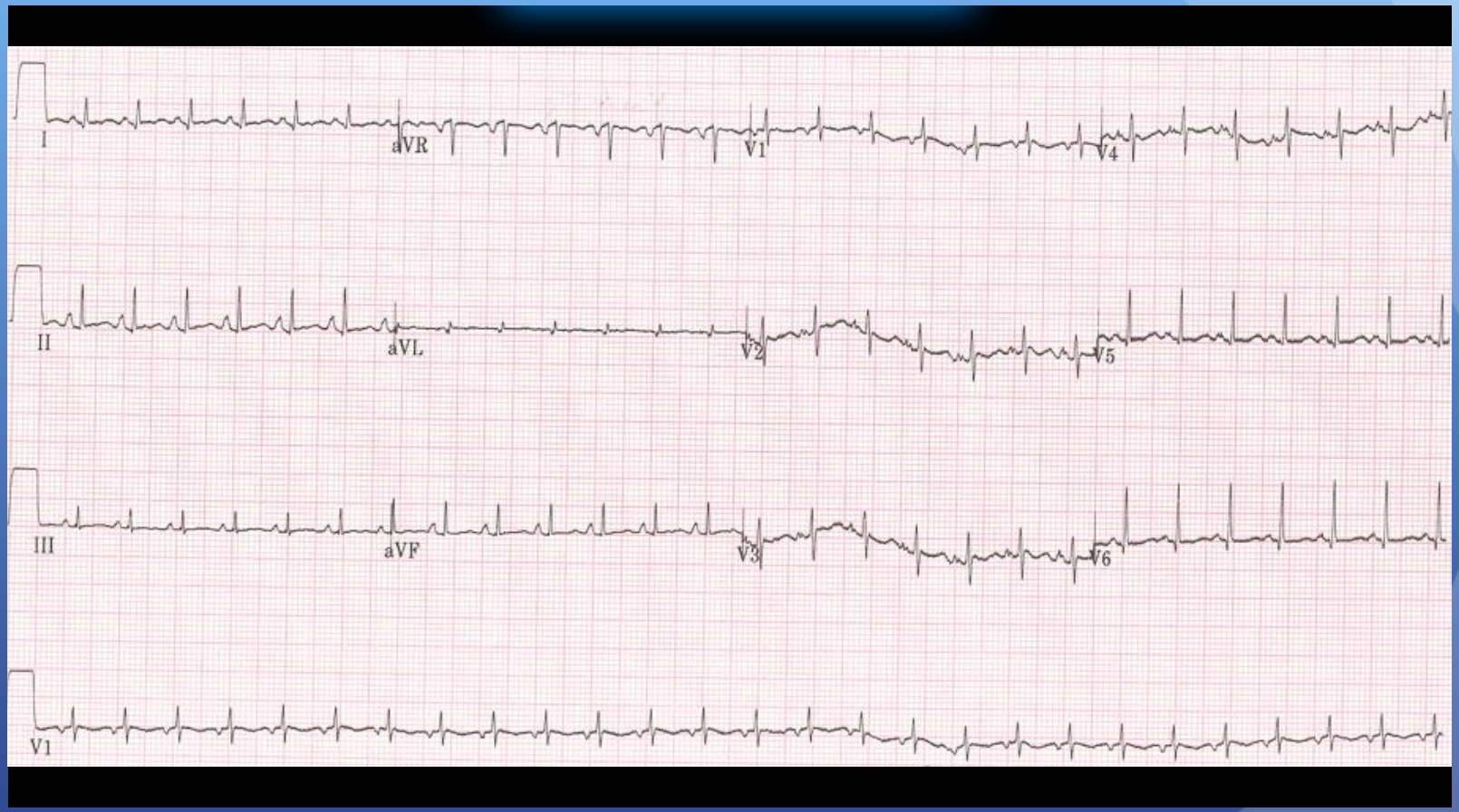
Initial management ----PGE1



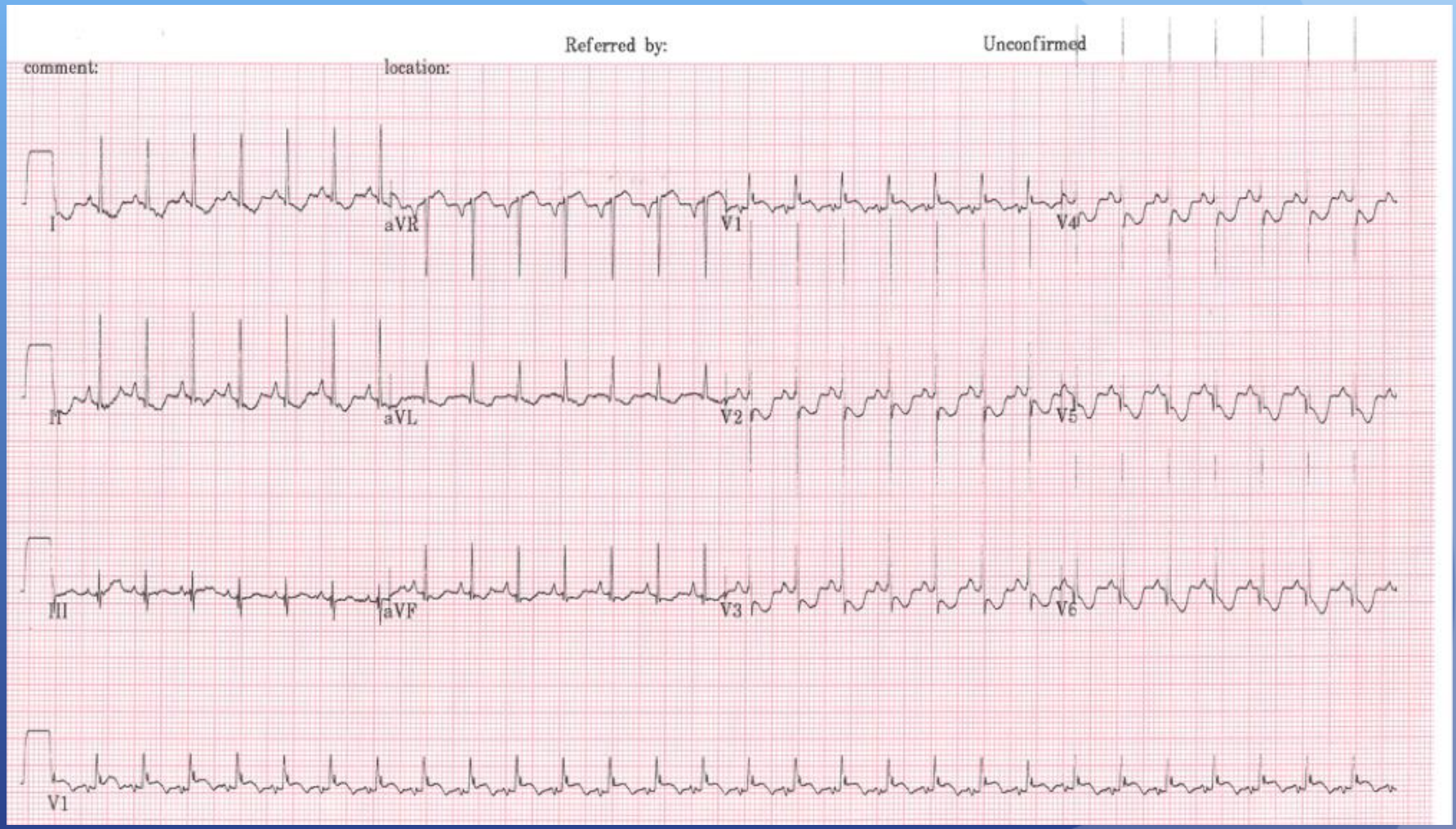
In the CICU with PAIVS

- *History of present illness:*
- 2.1 Kg 36 week newborn Infant with a prenatal diagnosis of PAIVS. Following birth started on PGE1 and admitted to the CICU

EKG in CICU



EKG in CICU

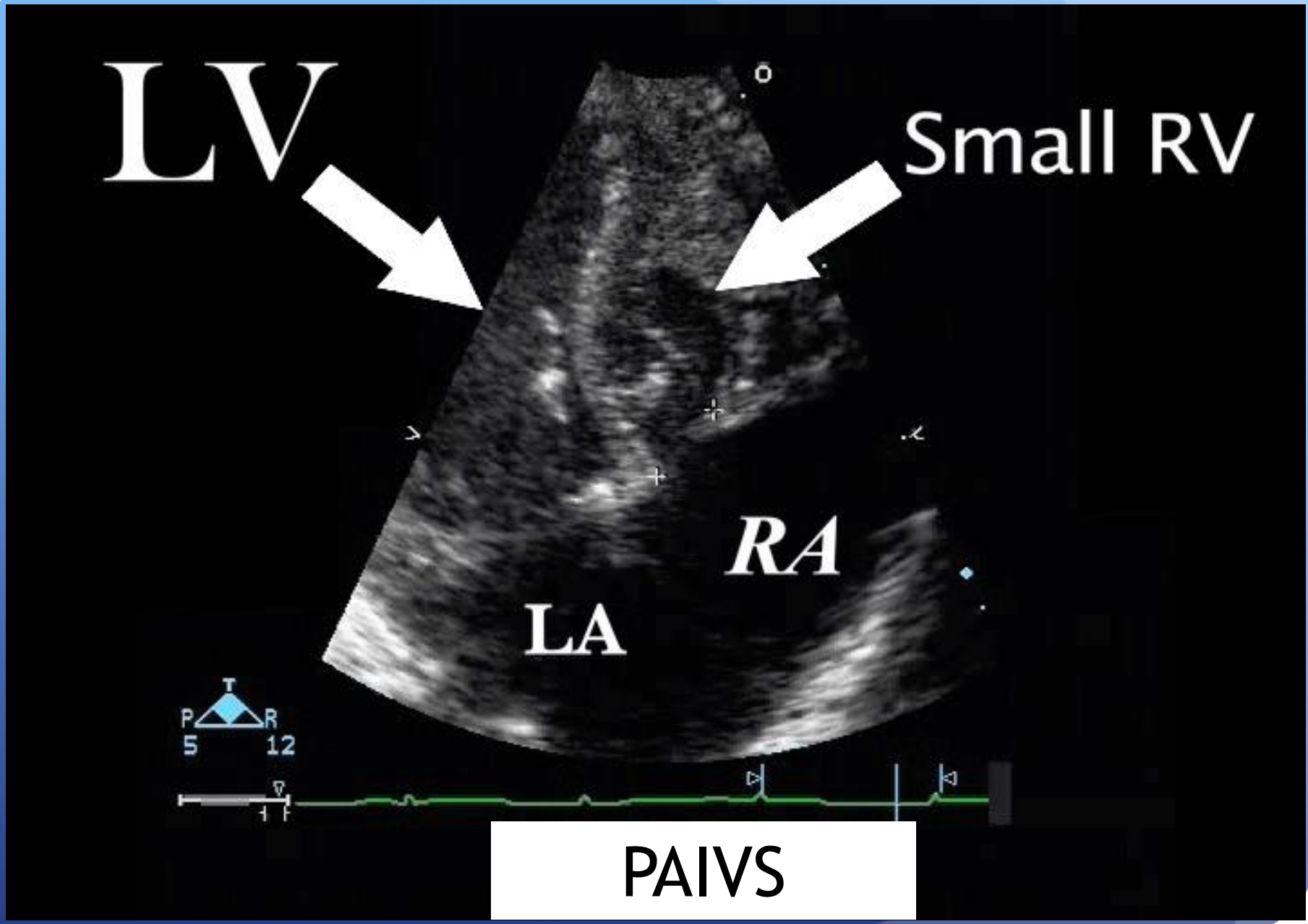

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Management of PAIVS on PGE1

- Dependent on R-L shunt on the atrial level
- Dependent on L-R shunt on the ductal level
- Small RV → CO ↓
- Restrictive physiology
- Increasing PVR to maintain CO
- Large PDA → increased "run off" with low Diastolic pressure → ischemia

TTE findings:

- Hypoplastic RV, TS, RVH
- No VSD
- Retrograde Coronary Blood Flow
- PDA dependent Pulmonary Blood Flow



PAIVS

In the Cathlab with PAIVS



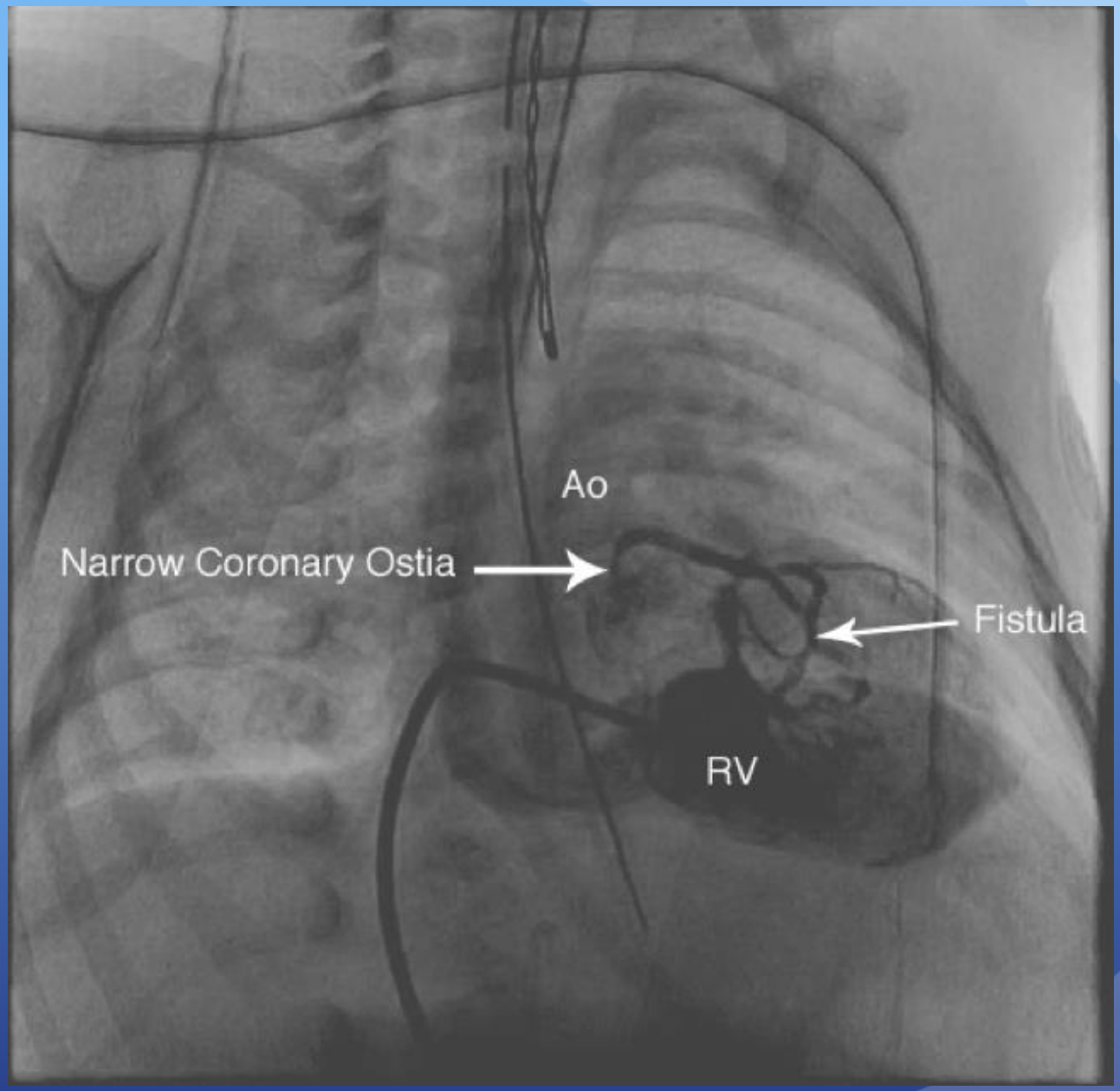
Hemodynamic cath

- Treat them as they would have RVDCC
- Maintain RVp and systemic pressure
- Keep them euvolemic to hyervolemic
- Fistulas without obstruction risk for ischemia via run off
- Keep Hct up

Cathlab findings

- No Antegrade Pulmonary BF
- RV-Coronary Fistulae
- Narrow Coronary Ostia

➔ Listed for transplant


Ao
Narrow Coronary Ostia
Fistula
RV

General risks for catheterization

- Vascular access
- Vascular injury
- Air embolism
- Arrhythmias
- Cardiac perforation
- Significant blood loss
- Brachial plexus injury

Transcatheter approach for PAIVS

- Patients on PGE1
- arrhythmias
- Perforation of the RVOT/PA
- Bleeding
- Tamponade
- Decreased pulmonary blood flow
- Increased pulmonary blood flow

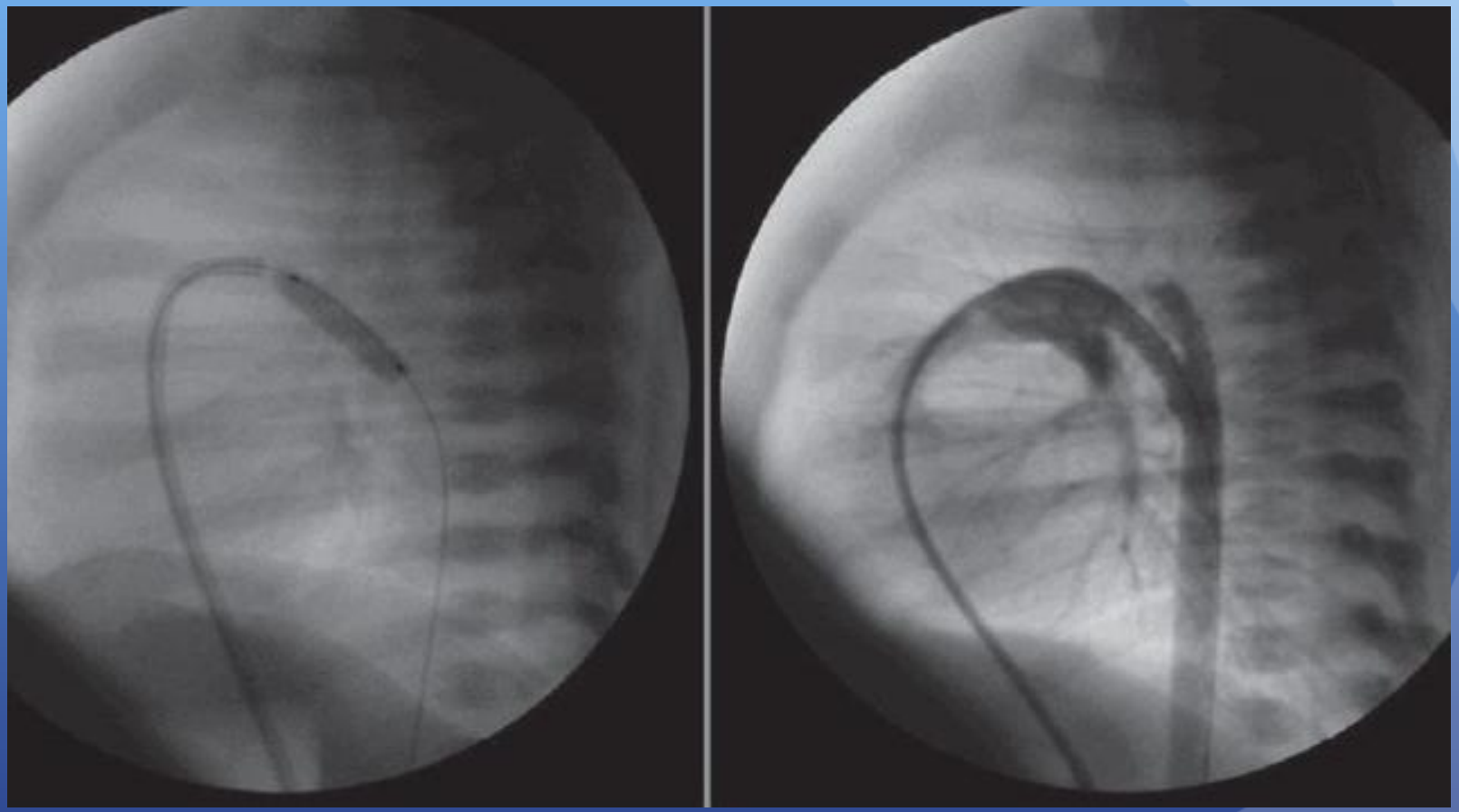
Catheter Cardiovasc Interv. 2013 Jan 1;81(1)

Outcomes of transcatheter approach for initial treatment of pulmonary atresia with intact ventricular septum

Hasan BS, Bautista-Hernandez V, McElhinney DB, Salvin J, Laussen PC, Prakash A, Geggel RL, Pigula FA.

- PVP attempted in (30 of 50) 60%, 87% (26) were successful
- 5 (17%) had a myocardial perforation (one needed to go to the OR, the other managed in cathlab)
- 2 patients developed NEC post cath
- 10 did not have surgery and 16 did have surgery prior to discharge
- Success in patients with greater TV valve Z-score

Stenting of the PDA



Stenting of the PDA

- Awaiting Transplant to avoid PGE1
- As only procedure if RVDCC
- After transcatheter perforation to maintain PB
- Complications / anesthetic considerations:
- PGE1 stopped in advance
- Acute thrombosis
- Spasm of the PDA
- Migration of the expanded stent

The Frequency of Anesthesia-Related Cardiac Arrests in Patients with Congenital Heart Disease Undergoing Cardiac Surgery (Odegard, Anesth Analg 2007)

- Results demonstrated 41 CA in 40 patients (5213 patients) overall frequency of 0.79%
- 30 CAs procedure related
- 11 CAs likely(6) possible related (5)
- No mortality
- Highest procedure for CA was Truncus arterious (frequency of 17.4 pr 100 procedures) and *mBTS for PAIVS* (3.6 pr 100 procedures)

BT shunt for PAIVS

- Good size RV with nl coronaries, normally do well
- Hypotension on induction
  - "steal" during diastole into the coronaries with oxygenated blood into the RV, the "runoff" is during systole from the RV to the coronaries with venous blood ➔ ischemia
  - Maintain BP, ↓ PBF to ↑ Aortic root pressure and better coronary perfusion
  - Optimize oxygen delivery by decreasing oxygen demand (Levophed, high dose Fentanyl)
  - Maintain Hct

BT shunt for PAIVS

- Critical moment when the surgeon opens the BTS, due to hypotension and low diastolic pressure
- After the shunt is open, low diastolic pressure with decreased coronary perfusion may persist
- Patients who are hypoxic after RVOT patch/dilation might need a BTS
- Development of pulmonary regurgitation can cause a retrograde circular shunt (RV-RA-LA-LV-Ao-BTS-PA-RV)

What if you need ECMO in patients with PAIVS and a BTS ?

Indication for initiation of mechanical circulatory support impacts survival of infants with shunted single-ventricle circulation supported with extracorporeal membrane oxygenation (J Thorac and Cardiovasc Surg 2007, Allan and Laussen)

- Evaluated outcome for patients with shunted single-ventricle physiology who required ECMO
- 44 patients (24 HLHS, 12 other, 8 PAIVS) Overall survival 48%, survive dependent on indictaion for ECMO – 81% for hypoxia and 29% for low CO shunt obstruction 83% survival
- 7 of the 8 with PAIVS did not survive

ECMO and BTS

- 3 of the 7 patients had PAIVS with RVDCC (I pt with aortocoronary atresia)
- Significant myocardial ischemia developed after BTS
- ECMO will cause further myocardial injury because drainage of the right atrium via ECMO cannula will cause decompression of the RV – reducing coronary perfusion pressure possible precluding ECMO weaning
- Retrograde circular shunt will also cause difficulty in weaning from ECMO
- Despite adequate technical repair physiology would not allow weaning from ECMO

Natural history of pulmonary atresia with intact ventricular septum and right-ventricle-dependent coronary circulation managed by the single-ventricle approach (Guleserian;2006 Ann Thorac Surg)

- Retrospective review 1989-2004
- 32 patients with PAIVS-RVDCC all underwent initial BTS
- Overall mortality 18.8%, all deaths within 3 months of BTS
- Aortocoronary atresia 100% mortality (3 of 3)
- No late mortality after 3 months of age
- Survivor 19 had undergone the Fontan and 7 BDG
- *Conclusion of the study*: early mortality related to coronary ischemia at the time of BTS. Single-ventricle palliation yields excellent longterm survival and patients with aortocoronary atresia should undergo transplant

Fontan patients

- Progressive ventricular dysfunction
- Systemic venous hypertension
  - Pleuro-pericardial effusions
  - Ascites
  - PLE
- Atrial flutter, atrial fibrillation, sinus node dysfunction, heart block
- Thrombosis

Fontan and Anesthesia

- Airway, Airway, Airway
- Hemodynamic goals
- Rate : N → ↑
- Rhythm : Sinus
- Preload : N → ↑ (↑ VEDP)
- Contractility : Usually tolerate volatile
- Afterload : N → ↑
- PVR : N → ↓

Fontan with PAIVS

- Guleserian‘s study from 2006, did not observe regression of the ventriculocoronary fistulae or coronary stenoses to the point that an RV dependent circulation has evolved into a „non-RV-dependent circulation
- This means that you still have to take into account that the Fontan pt still has RVDCC

Fontan with PAIVS

- Avoid Hypotension, optimize oxygen delivery and decrease oxygen demand
- All BDG shunts (at BCH) performed on CPB, with moderate hypothermia (28-30C) with RV adequately filled and heart ejecting
- Fontan on CPB either extra cardiac with heart beating with mild hypothermia or lateral tunnel with Aortic cross clamp, cardiolplegia with moderate hypothermia

PAIVS with Biventricular repair

- Late outcome and functional physiology depend on the size of the RV
- Transannular patch → Free PR and (TR), volume loaded and decreased function
- Small RV might have a restrictive physiology with R - L shunt at the atrial level

Late clinical features of patients with pulmonary atresia or critical pulmonary stenosis with intact ventricular septum after biventricular repair *(Hoashi,Ann Thorac Surg. 2012)*

- Aim to reveal late clinical features of patients with PAIVS after BVR based on preop RV volume
- 23 of 73 underwent BVR (no sinusoids)
- Overall survival at 20 years 90.6%, reintervention free 75.4% and Arrhythmia free 50.4%
- RVEDV between 60-120% showed well maintained Rv fn at 10 years, dilated in 16 (88.9%) due to TR and PR, which caused arrhythmias in 3 patients
- Conclusion, good late clinical features, but RV dilation and arrhythmias might occur

Exercise capacity and cardiac reserve in children and adolescents with corrected pulmonary atresia with intact ventricular septum after univentricular palliation and biventricular repair

(Romeih, J Thorac and Cardiovasc Surg 2012)

- Investigation of response to physical and dobutamine stress and delayed contrast-enhanced MRI
- 16 pts (7 SVR, 7BVR, 2 1.5VR)
- SVR showed impaired exercise capacity compare to BVR, LV EF increased in both groups, with dobutamine LV SV increased only in the BVR, HR increase inadequate in the SVR group
- Conclusion: Impaired Exercise capacity in SVR is related to decreased cardiac reserve and inadequate chronotropic response

Late clinical features in PAIVS:

- Depends on the repair Univentricular or BVR
- Despite BVR might have decreased RV function (at rest, stress or both) volume loaded ventricle, R-L shunt on atrial level
- Univentricular repair, might still have RVDCC, decreased exercise tolerance

Transplantation

- After 3 months of close calls with ischemia in the ICU the patient underwent cardiac transplantation
- Still doing well now 5 years later!