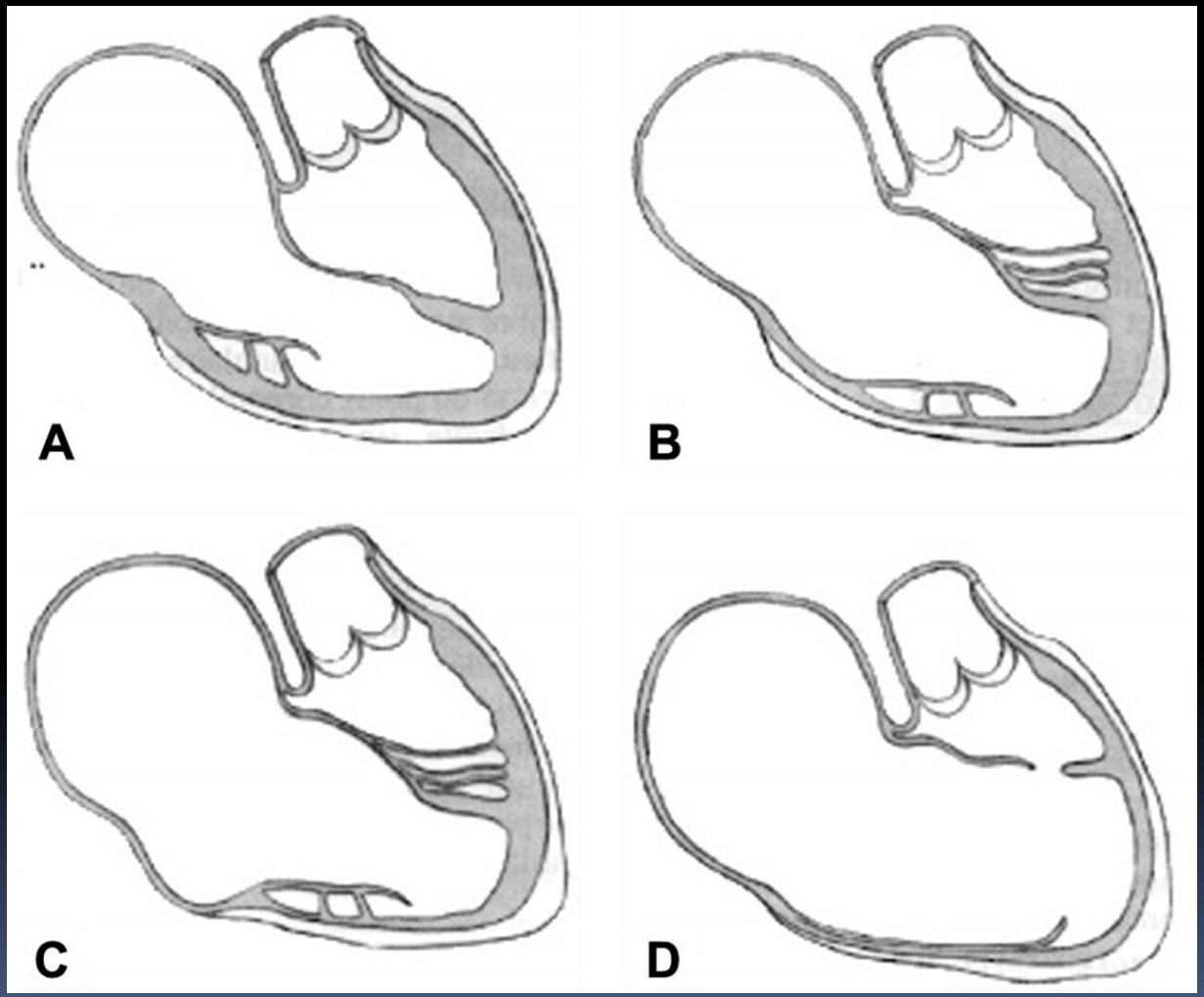
Deok Young Choi, Gil Hospital, Gachon University

NEONATES WITH EBSTEIN'S ANOMALY: PROBLEMS AND SOLUTION

Carpentier classification



Chauvaud S, Carpentier A. Multimedia Manual of Cardiothoracic Surgery 2007

Newborn with Ebstein’s anomaly

- Symptomatic
- Asymptomatic

Asymptomatic Neonates

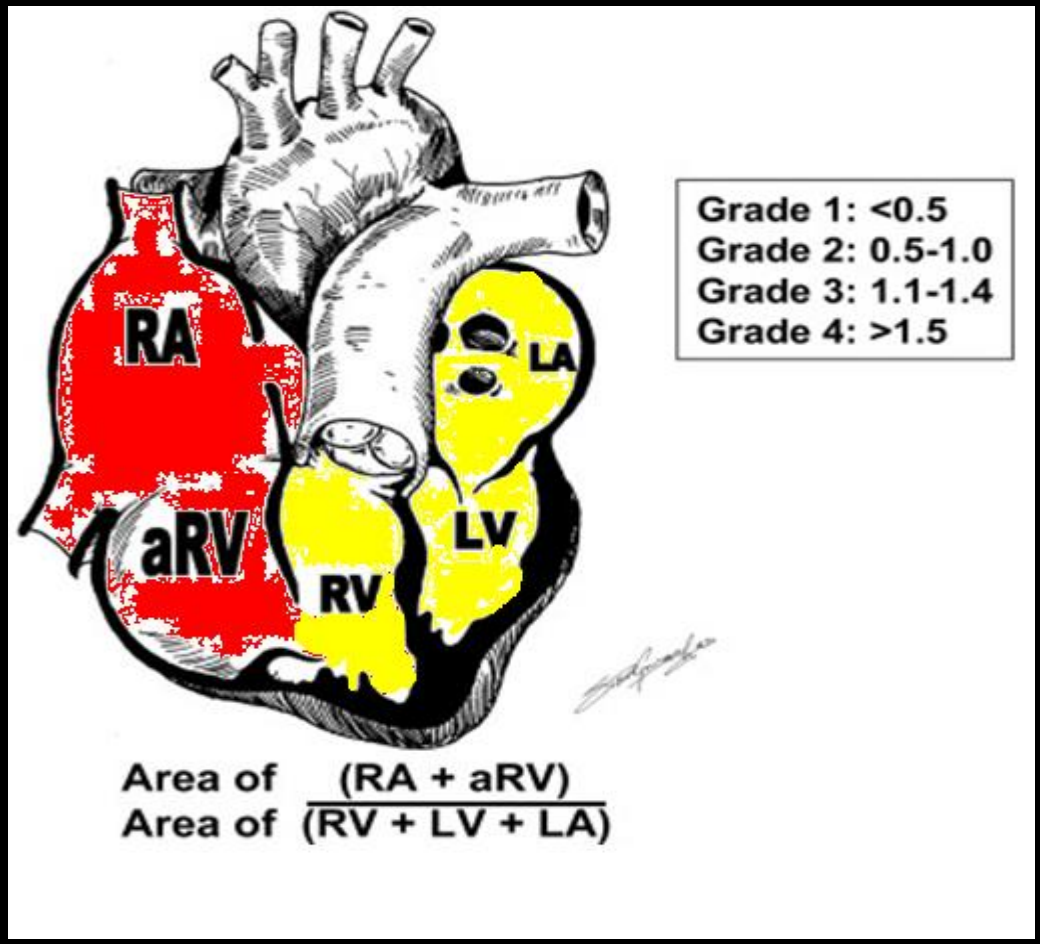
- Type A or B
- No blood flow disturbance (from RA to PA)
- Sx develop with age

  ; Rt heart failure, arrhythmia

Symptomatic Neonates

- Type C or D
- Moderate or severe flow disturbance (to PA)
- High pulmonary vascular resistance (aggravate the flow disturbance)
- Right heart failure (edema, ascites, hepatomegaly)
- Cyanosis (Rt to Lt shunt through the ASD, PFO)

Risk of Ebstein's Anomaly



GOSE Score	Ratio	Mortality
1–2	<1.0	8%
3 (acyanotic)	1.1–1.4	10% early, 45% late
3 (cyanotic)	1.1–1.4	100%
4	>1.5	100%

Christopher J. Seminars in Thoracic and Cardiovascular Surgery: Pediatric Cardiac Surgery Annual Volume 10, Issue 1 2007 112 - 116

- GOSE score ≥ 1.5 or ≥ 1.1 with cyanosis
  ; approaching 100% mortality
- Cardiomegaly (CT ratio > 0.8) with severe TR, acyanotic neonates with GOSE score > 1.0 with functional PA and large ASD
  ; dismal prognosis

Boston US et al. J Thorac Cardiovasc Surg 2011;141:1163-9

Fetal & neonatal study

- Boston children's hospital
- 1984~2004
- 66 patients – Ebstein (61), TV malformation(5)
- 33 – fetal echo (16 fetus ; survived, 49%)
- 49 neonates ; 35(71%) survived more than 1 month
- Independent predictors of death

  - RA area index > 1

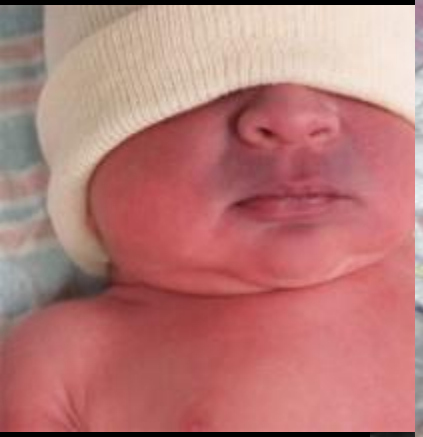
  - the absence of antegrade flow across the pulmonary valve

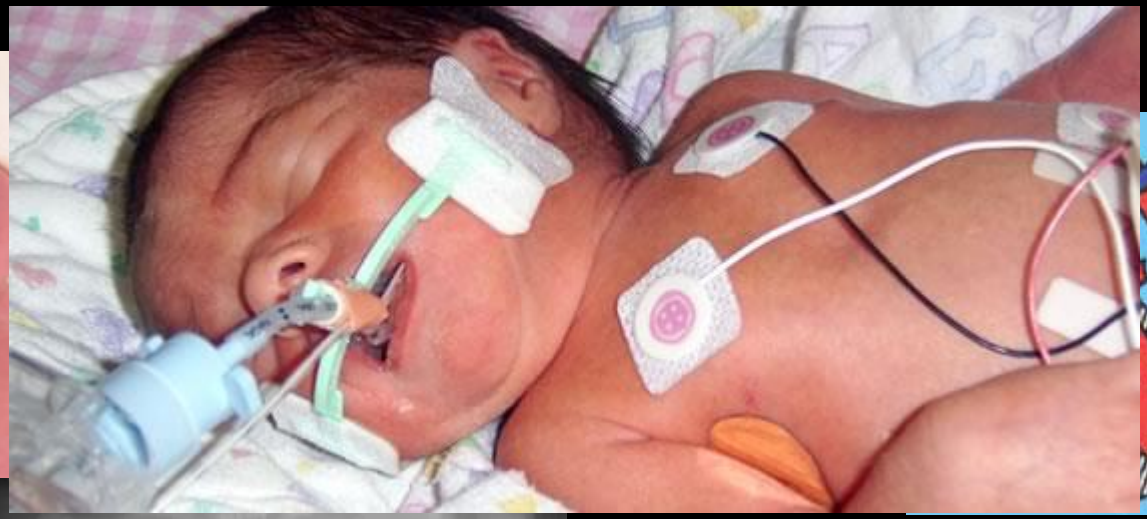
McElhinney DB et al. Am J Cardiol 2005;96:582

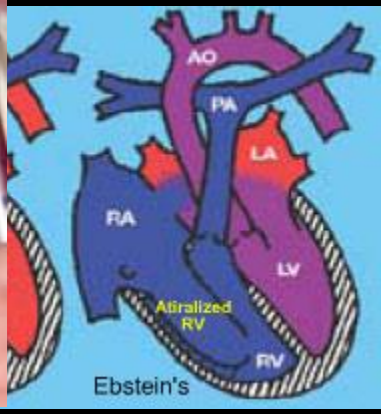
Problem of neonates with EA

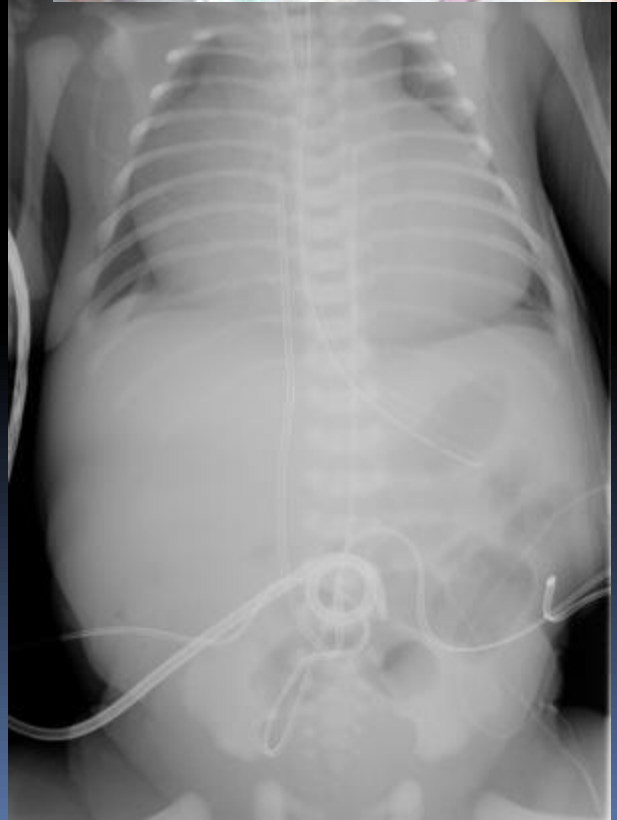
- Inadequate pulmonary blood flow (functional or anatomical PA)
- Tricuspid valve anomaly (include TR)
- RV dysfunction
- Inadequate left ventricular filling (due to septal bowing)
- Arrhythmia

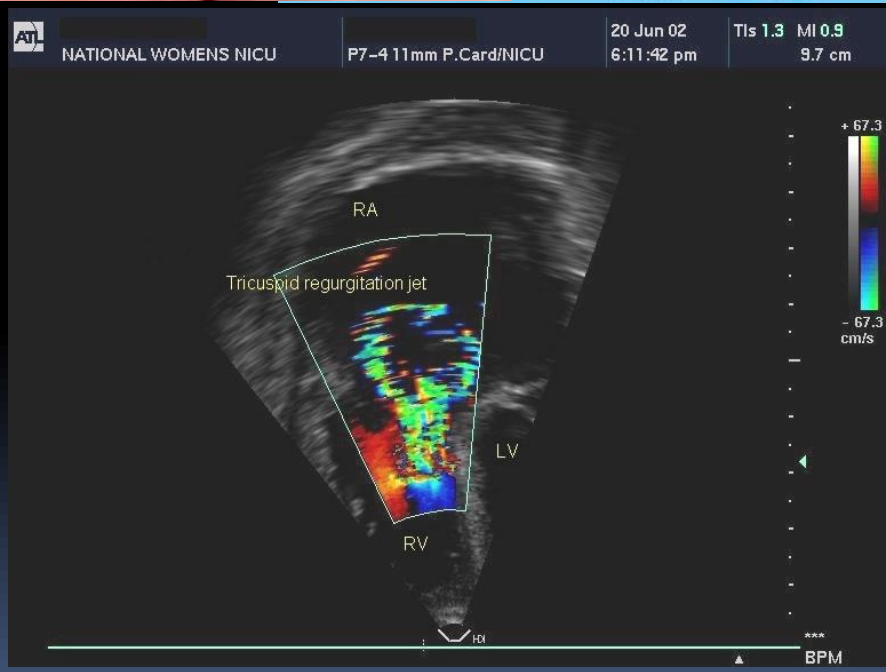
Worst case







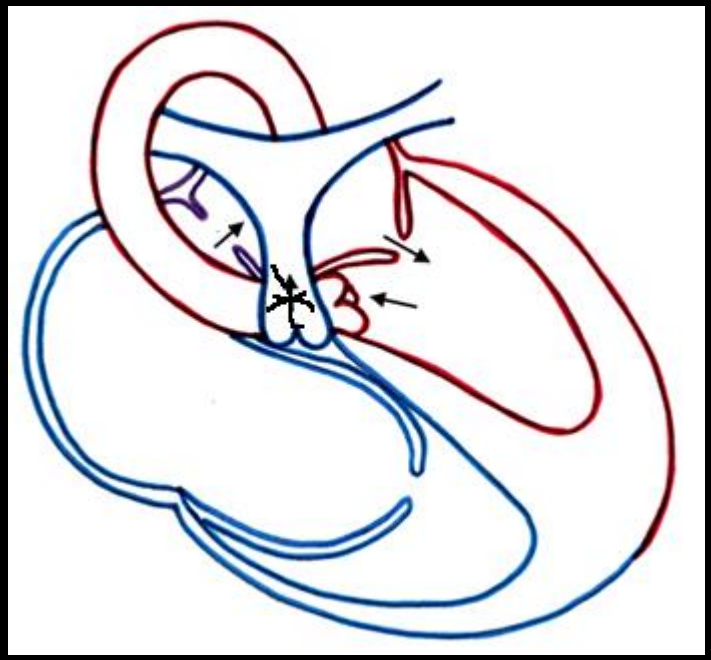




Donnelly L et al. Diagnostic Imaging: Pediatrics First Edition
Pediatricimaging.wikispaces.com/EbsteinsAnomaly
www.adhb.govt.nz/newborn/teachingresources
Easypediatrics.com, McNearly group medical consultation

Echocardiography

- Complete anatomic evaluation
- Tricuspid valve morphology, severity of TR
- Accompanied anomaly
- ASD flow direction
- Pulmonary valve anatomy, blood flow (anatomical atresia or functional atresia)



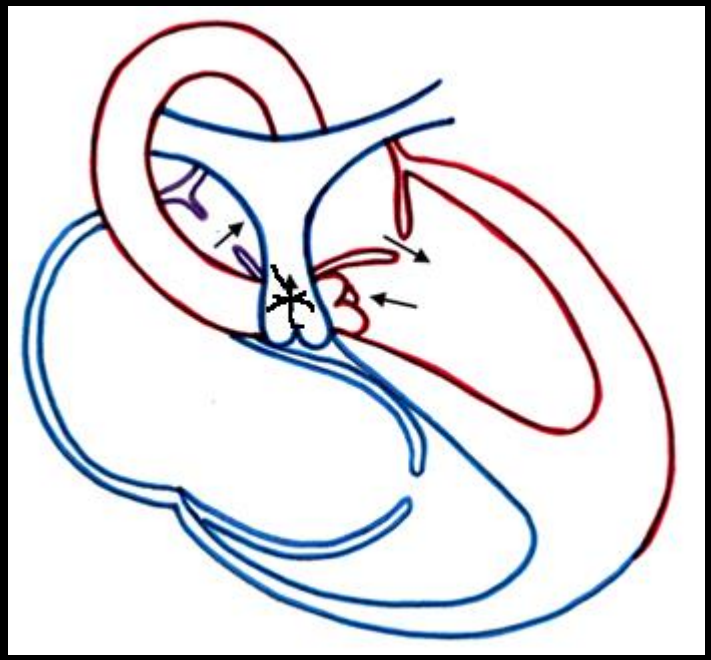
- RVOT obstruction
  - elevated PVR
  - severe TR
  - RV dysfunction

; difficult to DDx

anatomic obstruction

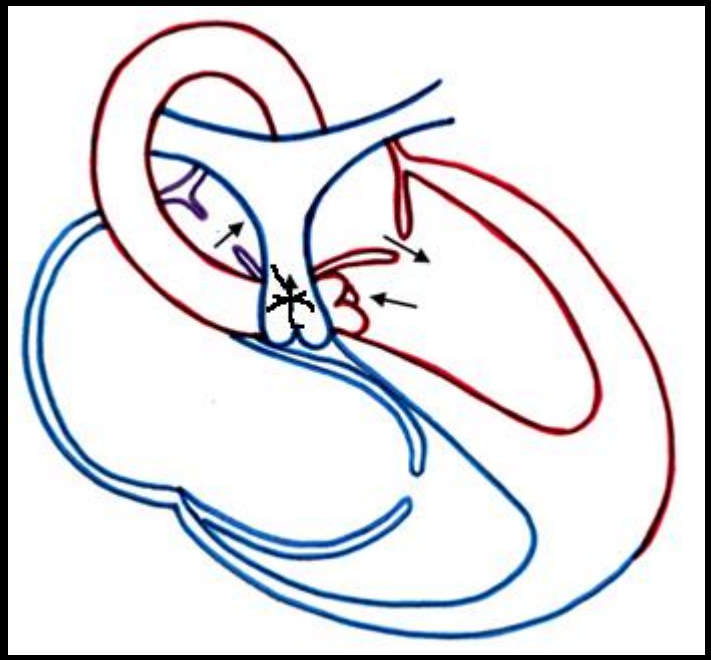
functional obstruction

Ferguson EC et al. RadioGraphics 2007;27:1323



- Anatomical PA
  -pulmonary valve anomaly
  -PDA dependent pulmonary circulation
  -PGE1
  -ductal stent
  -systemic to pulmonary shunt
  -RVOT reconstruction

Ferguson EC et al. RadioGraphics 2007;27:1323



- Functional PA

  -pulmonary valve ; intact

  -cannot generate blood flow from RV to PA

  -PGE1 (±)

  -oxygen (decrease the PVR)

  -NO gas (selective pulmonary vasodilators)

  -try to weaning the PGE1

Ferguson EC et al. RadioGraphics 2007;27:1323

Tip) Prostaglandin E1

- Overdose

  ; increase the pulmonary blood flow

  → increase the pulmonary blood pressure

  → RVOT flow disturbance

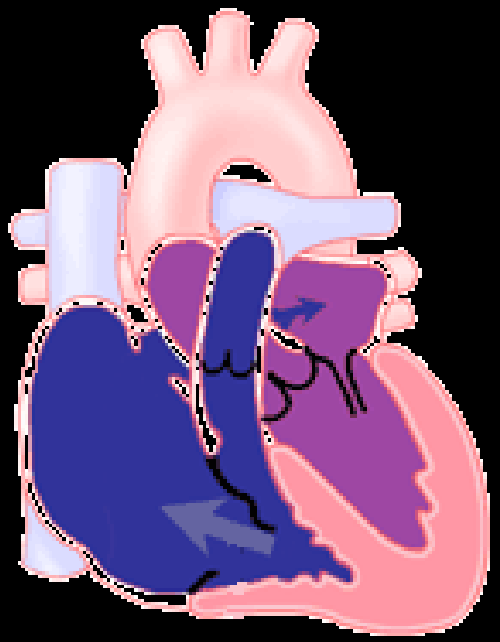
  → aggravate the TR, increase the RA size and Rt to Lt shunt

  → aggravated hypoxia

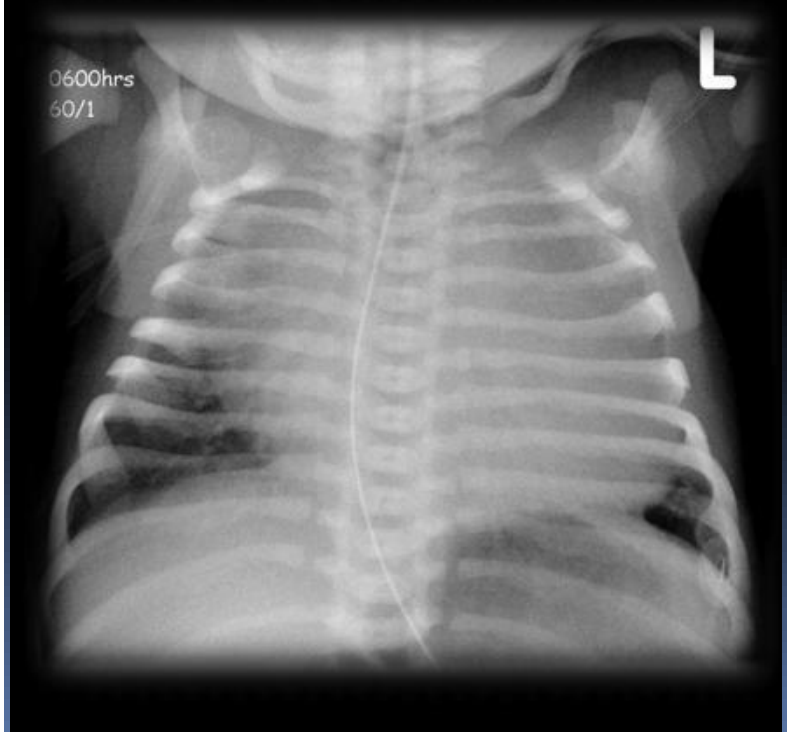
Anatomical vs Functional PA

- Early postoperative results
- 94% early survival rate – without anatomic PA
- 60% (with anatomic PA)

Boston US et al. J Thorac Cardiovasc Surg 2011;141:1163



www.Cincinnatichildrens.org/health



- Cyanosis (alone and dominant sx)

  ; systemic to pulmonary shunt only
- Cyanosis + RV dysfunction (severe TR)

  ; consider RV exclusion

University of Michigan Neonatal Experience

- 1988 ~ 2008
- 40 neonates with Ebstein's anomaly – 24 underwent surgery
- Overall survival ; 66.7% at 1 year
- Op indication ; symptomatic neonate with fail to weaning the PGE1

- *Bove EL, et al. Semin Thorac Cardiovasc Surg Pediatr Card Surg Ann 2009;12:63*

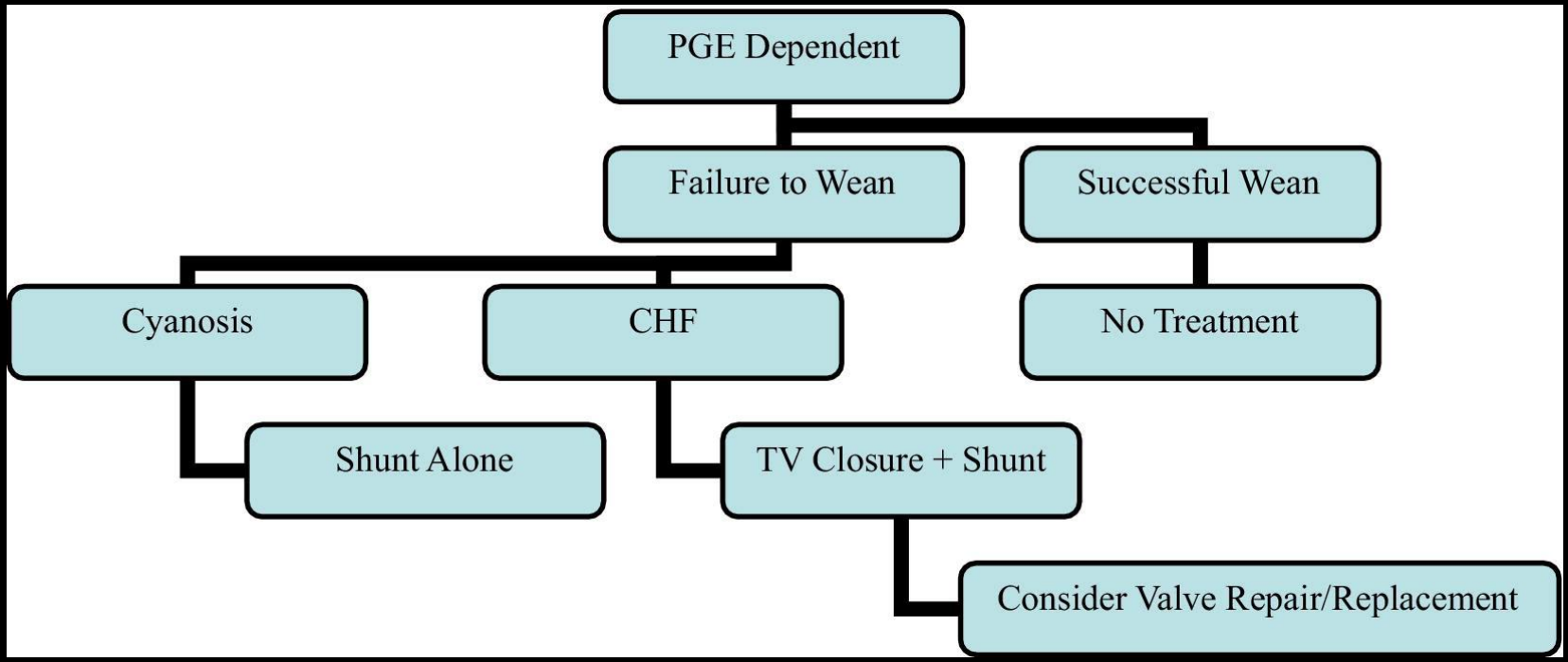


Figure 1 Treatment protocol for Ebstein's anomaly in the neonate.

Edward L. Bove , Jennifer C. Hirsch , Richard G. Ohye , Eric J. Devaney

How I Manage Neonatal Ebstein's Anomaly

Seminars in Thoracic and Cardiovascular Surgery: Pediatric Cardiac Surgery Annual Volume 12, Issue 1 2009 63 - 65

University of Tennessee, Oklahoma

- 1994 ~ 2010
- 23 neonates, 9 young infants (m-wt ; 3.9kg)
- GOSE score ＞1.5 (22 of 23 neonates)
- Preop. Management

  ; adequate sedation, inotropic support, PGE1, inhaled NO, echocardiographic F/U

Boston US, Ward KE et al. J Thorac Cardiovasc Surg 2011;141:1163

University of Tennessee, Oklahoma

- Indication for early surgical intervention
  - persistent ventilator dependency
  - RHF ; unresponsive to medical tx
  - severe TR associated with cyanosis
  - persistent significant inotropic support
  - persistent PGE1 dependent circulation

Boston US, Ward KE et al. J Thorac Cardiovasc Surg 2011;141:1163

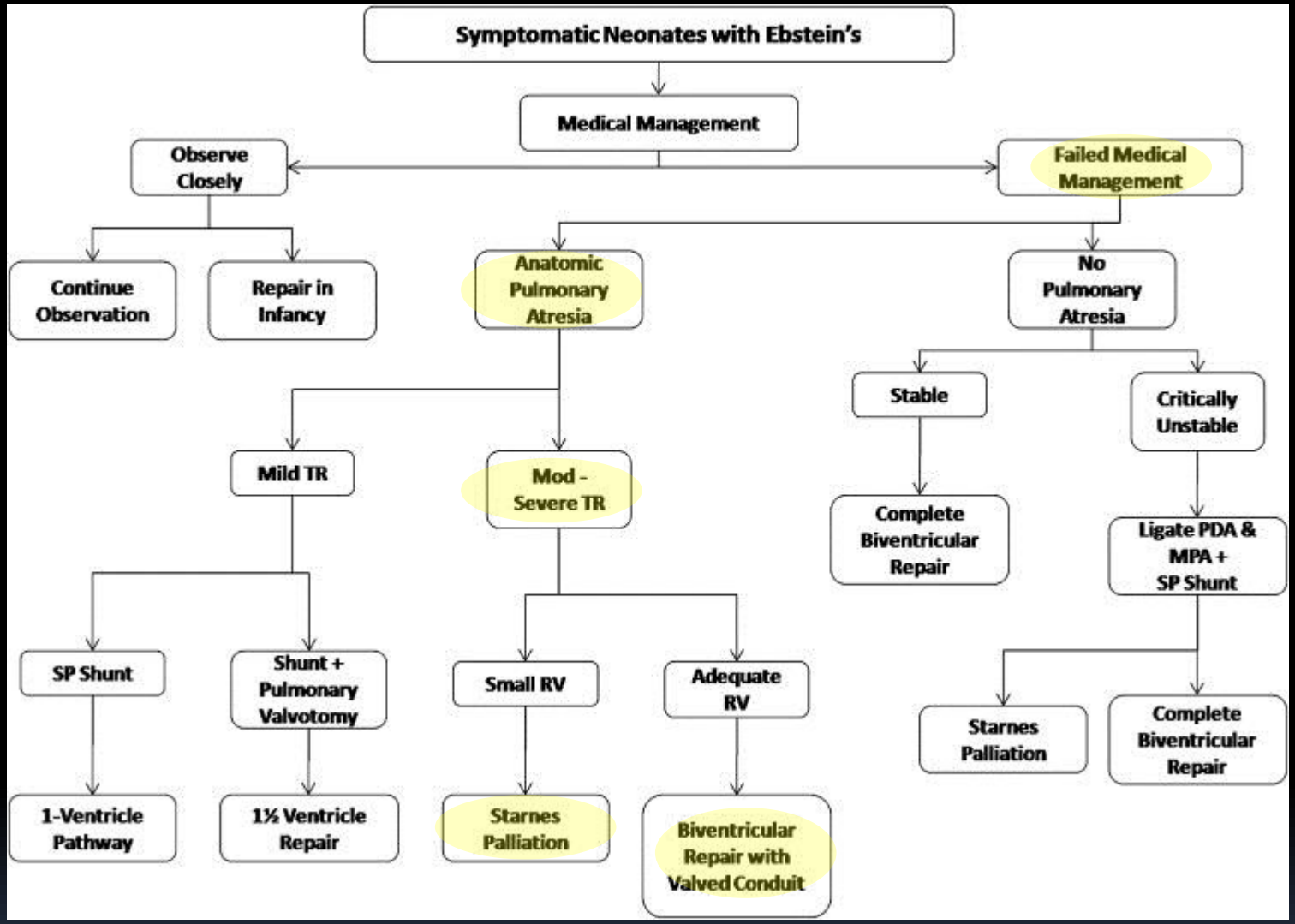


Figure 6 Surgical algorithm for symptomatic neonates with Ebstein anomaly. TR, Tricuspid regurgitation; PDA, patent ductus arteriosus; MPA, main pulmonary artery;

Umar S. Boston , Steven P. Goldberg , Kent E. Ward , Edward D. Overholt , Thomas Spentzas , Thomas K. Chin , Chris...

Complete repair of Ebstein anomaly in neonates and young infants: A?16-year follow-up

The Journal of Thoracic and Cardiovascular Surgery Volume 141, Issue 5 2011 1163 - 1169

Perinatal course of Ebstein's anomaly and tricuspid valve dysplasia in the fetus

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ABSTRACT

Objective We sought to better define echocardiographic predictors of perinatal mortality in fetuses with Ebstein's anomaly (EA) or tricuspid valve dysplasia (TVD).

Method Parameters included measured chamber size, the presence of hydrops, and Doppler recordings of the left ventricular (LV) myocardial performance index (MPI).

Results Between 1 January 2000 and 31 December 2008, 21 fetuses were diagnosed with either EA (17) or TVD (4). Five fetuses were lost to follow-up, and 12 of 16 fetuses were born live (75%). Survivors were found to have smaller right atrial area index scores when compared with non-survivors (1.025 ± 0312 vs 1.502 ± 0.105, respectively, $p = 0.013$) and were less likely to present with hydrops (0% survivors vs 75% of non-survivors, $p < 0.01$). LV MPI sub-analysis revealed a shorter combined isovolemic contraction and relaxation time for non-survivors compared wth survivors (46.5 ± 8.2 ms vs 82.3 ± 21.2 ms, respectively, $p = 0.004$) although no difference was observed for LV ejection times or overall LV MPI between survivors and non-survivors.

Conclusion Physiologic analysis of left ventricular function via the LV Tei index and its component measurements demonstrates potentially novel insights into hemodynamic derangements and their association with outcomes in patients with EA/TVD.

Prenat Diagn 2012;32:245

Outcome in neonates with Ebstein's anomaly

- 1961~1990, 50 neonates
- 9 (18%) – died in the neonatal period
- 15 late death (mean age 4.5yr)
- 10yr actuarial survival rate ; 61%
- Echocardiographic grading (GOSE score) ; prognostic factor

- *Celermajer DS, et al. J Am Coll Cardiol 1992;19:1041*

Survival and mortality

- 53.8% - any surgical intervention during the neonatal period

European Congenital Heart Surgeons Association database (2002~2006)

- Early mortality 24% (16 neonatal Ebstein)

Reemtsen BL et al. J Thorac Cardiovasc Surg 2006;132:1285

- 63% - 10 yr survival, single ventricle palliation of 24 neonates

Shinakawa T et al. J Thorac Cardiovasc Surg 2010;139:354

- Case report of Ebstein anomaly in a fetus

  ; cardiomegaly, severe TR and PI, marked increased RA

  → elective preterm delivery

  MPA and ductal ligation, central shunt, plication of RA (4hrs after birth)

  Starnes procedure (16 days)

  uneventful postoperative course

Fukuoka Children's Hospital

Tsukimori K et al. Pediatr Cardiol 2012;33:343

- 23 neonates, 9 young infants

  early survival ; 78.1%, 15 yrs survival ; 74%

Boston US, Ward KE et al. J Thorac Cardiovasc Surg 2011;141:1163

Summery

- Early detection (fetal echocardiography)
- Noble neonatal care
  mechanical ventilation, inotrophic support
  adequate PGE1 infusion
  try to decrease the PVR (O_2, NO gas, med.)
  echocardiographic evaluation
- Aggressive surgical intervention
  → more improve the survival rate of critically ill neonatal Ebstein's anomaly !!