The Terrors of the Tricuspid Valve: Care and Management of the Neonate with Ebstein's anomaly or Severe tricuspid valve dysplasia

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DISCLOSURE

NO CONFLICTS OF INTEREST

Agenda

01 Ebstein Anomaly Morphology

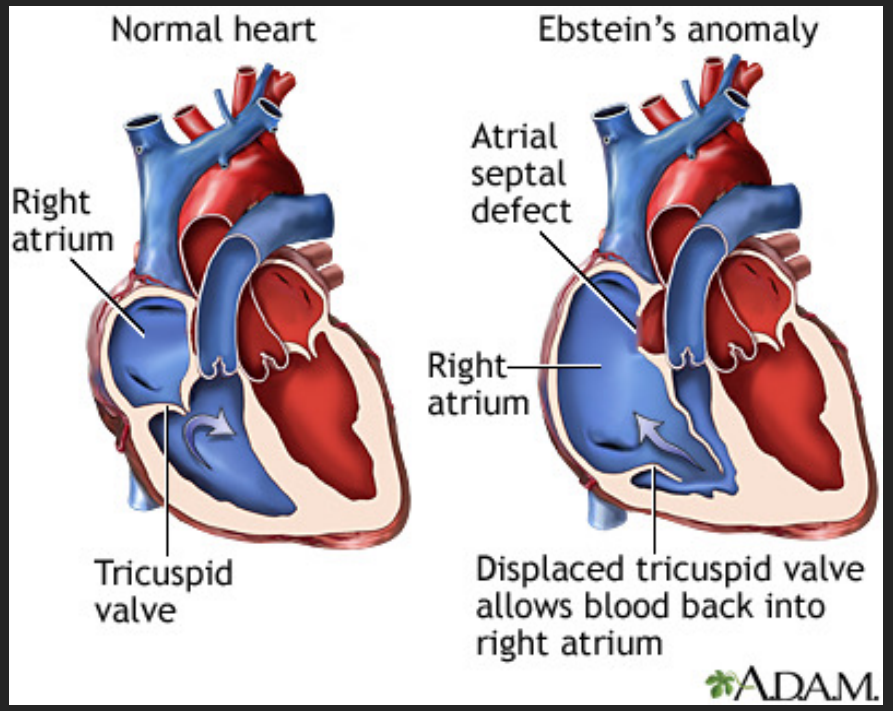
02 Prevalence and associated defects

03 Clinical Presentation/ Pathophysiology

04 Medical Management

05 Surgical Management

06 Conclusions

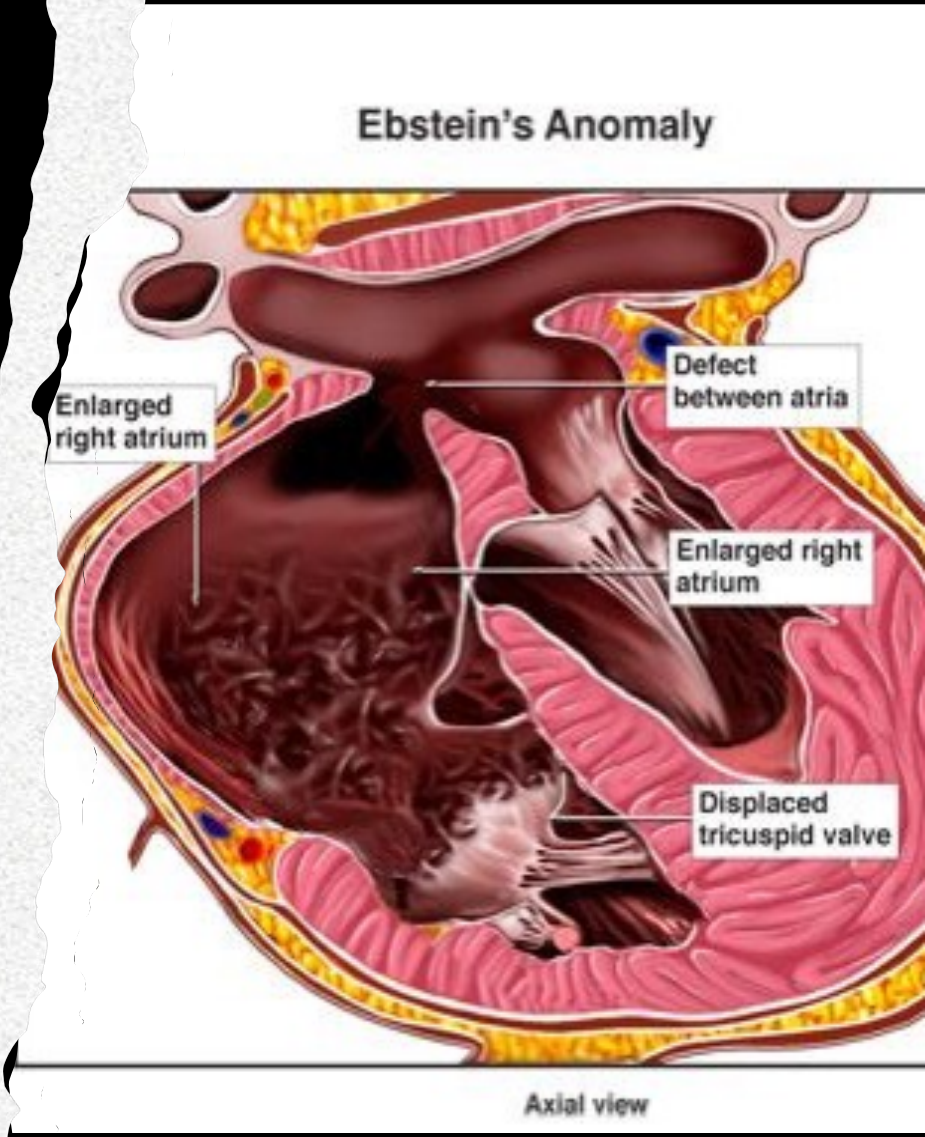


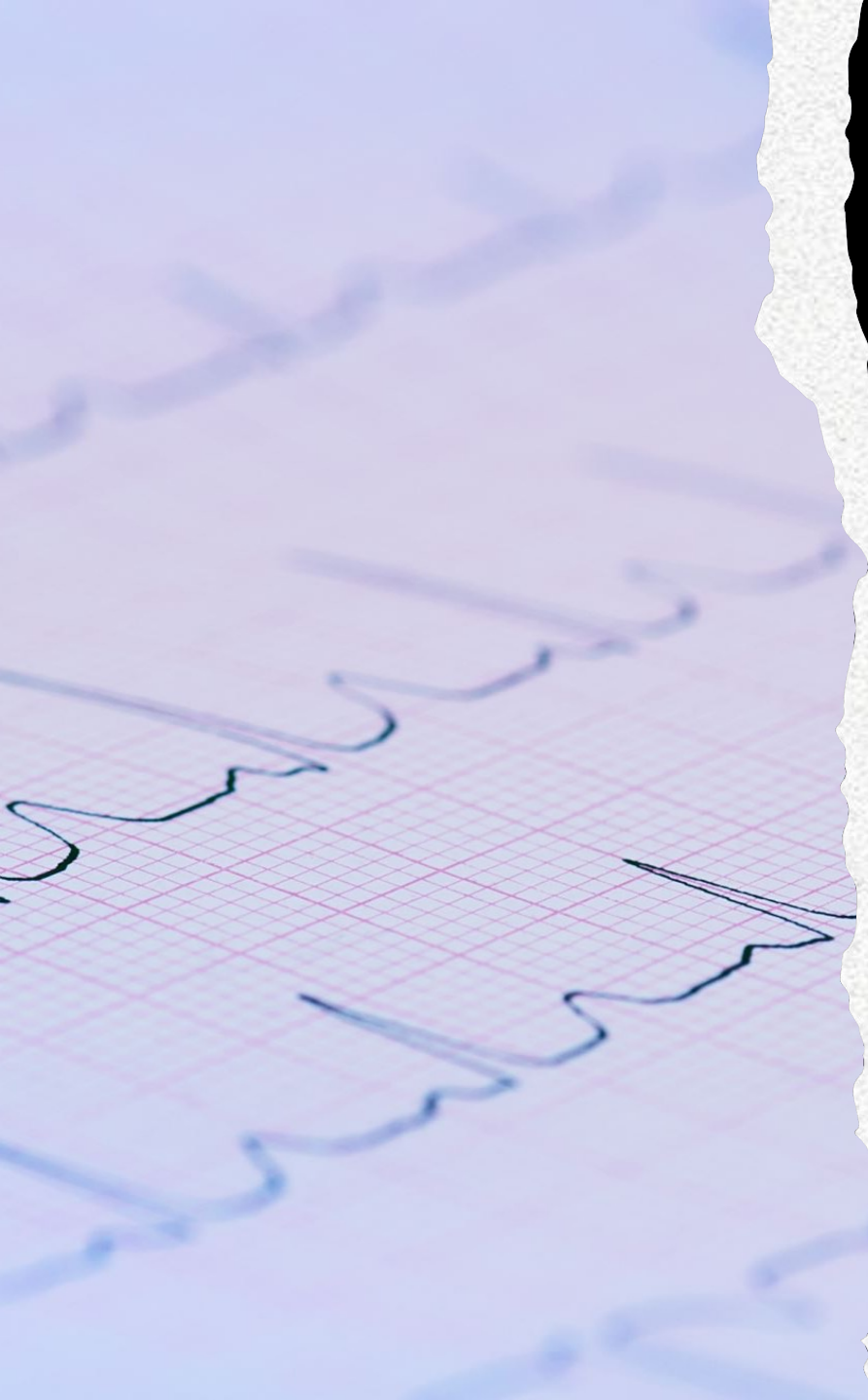
Ebstein's Anomaly (EA)

- Wihelm Ebstein 1866
- Incidence 0.5% of all CHD
- Overall Mortality related to presenting age
  - Severe Neonatal Forms 20-80%
  - All age groups 35-45%
- Most common related lesion
  - ASD, PFO
  - Bicuspid Ao valve
  - Pulmonary atresia
- Equal sex incidence

Ebstein's Anomaly

- Congenital anomaly of the tricuspid valve and right ventricle due to incomplete delamination of tricuspid valve leaflets
- Apical displacement of tricuspid valve, leads to decreased size of functional right ventricle
- Right ventricular outflow and degree of pulmonary blood flow is variable and unpredictable





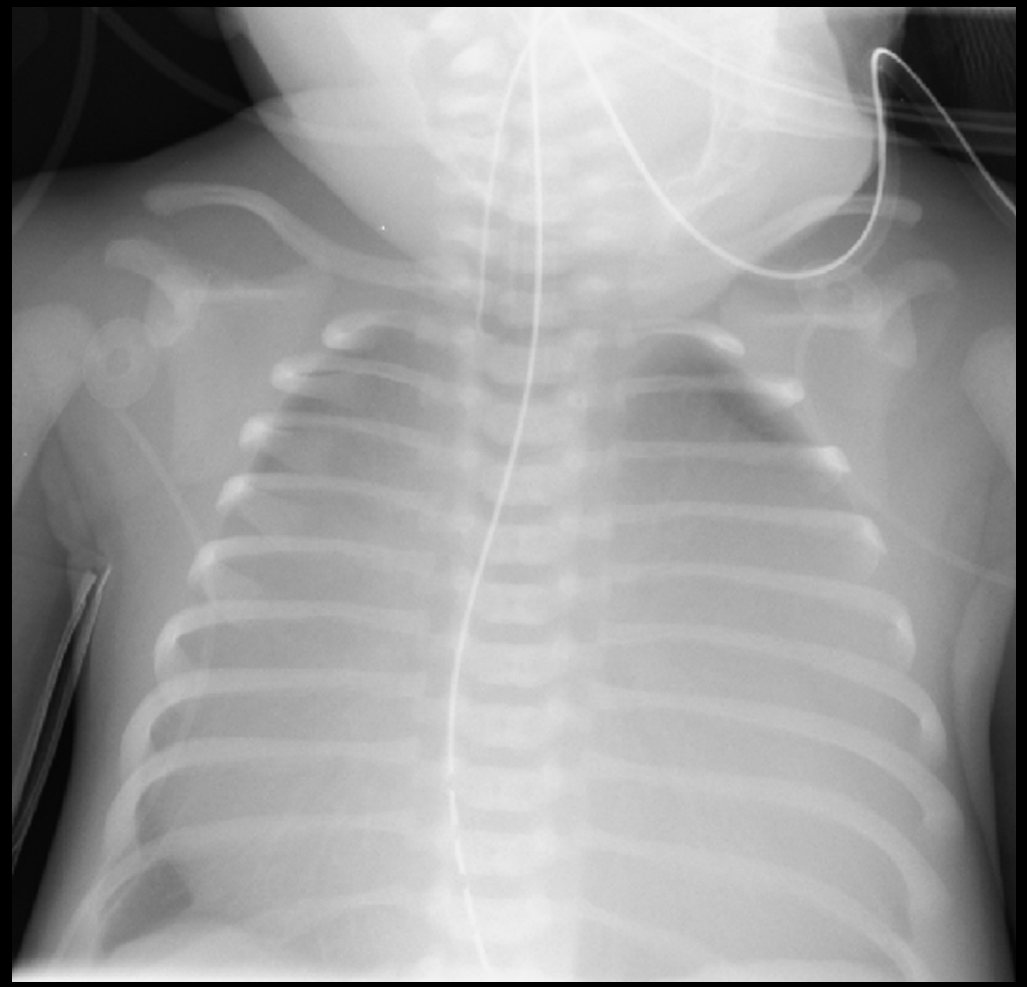
Clinical presentation

- Neonatal Period
  - Severe Cyanosis
  - Congestive Heart Failure
  - Metabolic Acidosis

- Adulthood
  - Arrythmias
  - Heart Failure
  - Fatigue
  - Cyanosis

Pathophysiology

- Presentations and symptoms related to:
  - Tricuspid regurgitation
  - RVOT obstruction
  - Cyanosis
  - Pulmonary hypertension
  - Circular shunt
  - RV failure
    - Congestive heart Failure



EA: Classification

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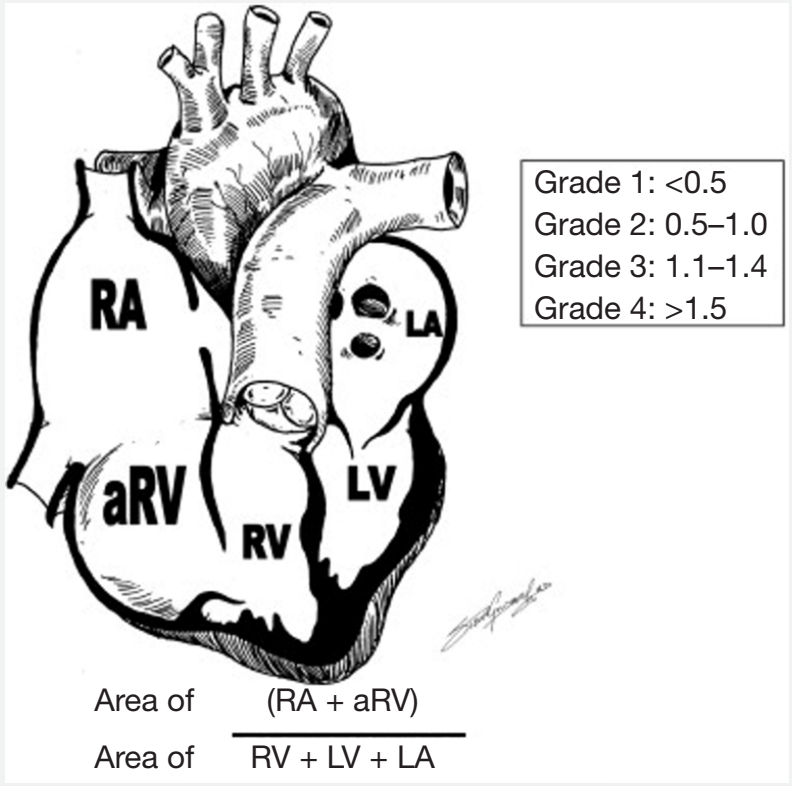


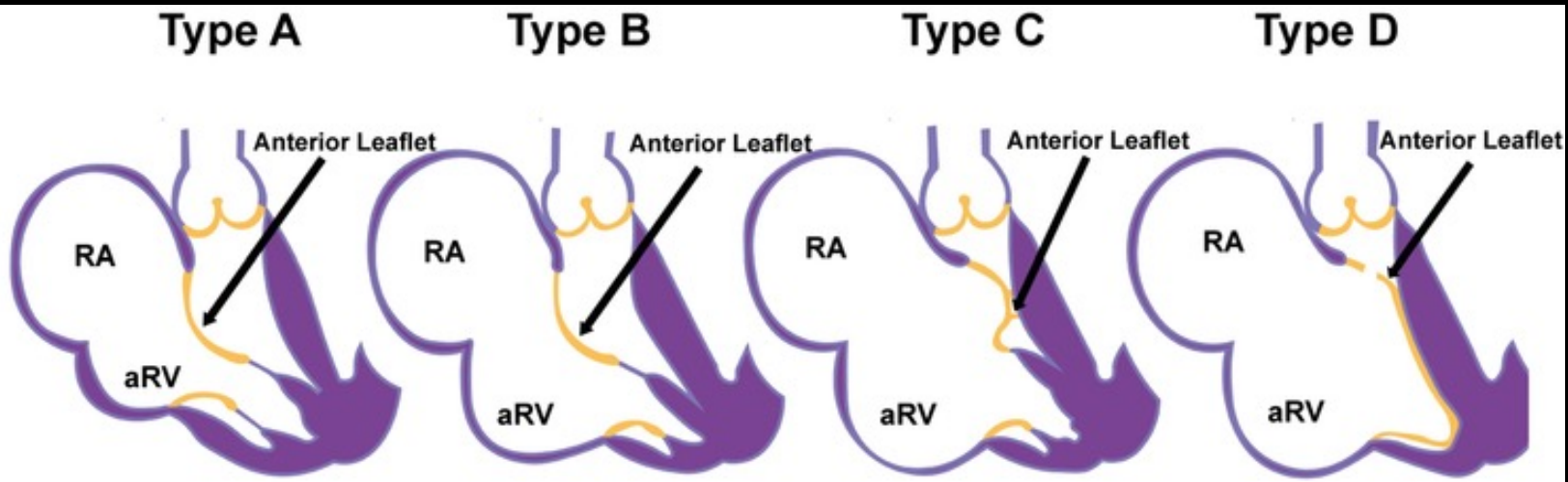
Figure 2 Celermajer index. RA, right atrial; aRV, atrialized right ventricle; RV, right ventricle; LA, left atrium; LV, left ventricle.

TABLE 1. Mortality prediction based on Great Ormond Street Echocardiography (GOSE) score*

	GOSE score	
GOSE score	**Ratio**	**Mortality**
I	<0.5	8%
II	0.5-1.0	8%
III (acyanotic)	1.1-1.4	10% early, 45% late
III (cyanotic)	1.1-1.4	100%
IV	>1.5	100%

*From: Celermajer DS, Bull C, Till JA, Cullen S, Vassillikos VP, Sullivan ID, et al. Ebstein's anomaly: presentation and outcome from fetus to adult. *J Am Coll Cardiol*. 1994:22:170-6.

EA: Classification



	A	B	C	D
Septal/Posterior Leaflet Displacement	Mild	Moderate	Severe	Extremely Severe (Tricuspid Valve Sac)
Anterior Leaflet	Normal Morphology and Mobility	Abnormal Attachments with Normal Mobility	Partial Adhesions to the Ventricular Free Wall and Restricted Motion	Severe Adhesions to the Ventricular Wall
"Atrialized" Ventricle	Small/Preserved Function	Moderate/Reduced Function	Large/Reduced Function	Very Large/Minimal Function
Functional Ventricle	Normal/Preserved Function	Reduced/Preserved Function	Small/Reduced Function	Infundibulum/Depressed Function

Review Article on Management of Congenital Heart Disease

Ebstein's anomaly: contemporary management strategies

Sandeep Sainathan1, Luciana da Fonseca da Silva2, Jose Pedro da Silva

J Thorac Dis 2020;12(3):1161-1173 | http://dx.doi.org/10.21037/jtd.2020.01.18

Tricuspid Valve Malfunction

- RA Dilatation
- Volume Overload

- Decreased Contractility

Tricuspid Valve Regurgitation

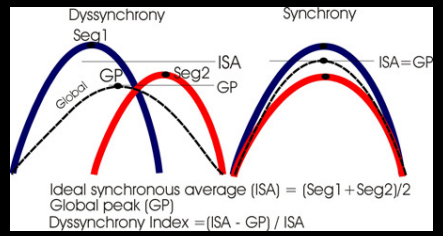
RV Systolic Impairment

RV Dilatation

Decreased RV Stroke Volume

- RV Ischemia
- IVS Shifting
- Arrythmias
- LV Dysfunction

- Cyanosis
- Increased Rt-Lt Shunt



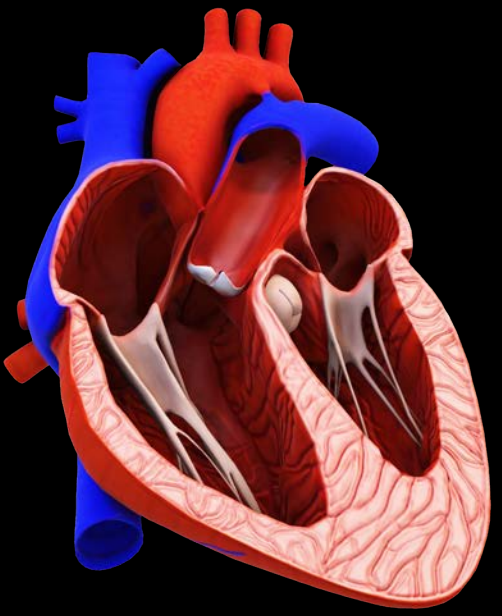
Decreased Cardiac Output

Multiorgan Failure

DEATH

Neonatal Management

- Goals of management
  - Decrease RV afterload
  - Decrease degree of regurgitation
  - Correct metabolic acidosis and avoid arrythmias
  - Avoid further atrial RV dilatation
  - Avoid Recirculation syndrome



Medical Management

1

Decrease RV afterload

- Intubation and sedation
- Nitric oxide
- Optimal tidal volume with low PEEP
- Prostaglandins

2

Decrease degree of regurgitation

- Milrinone
- Euvolemia

3

Correct metabolic acidosis and avoid arrythmias

- Lactate levels
- Urine output
- Diuretics

Medical Management

Avoid Recirculation Syndrome

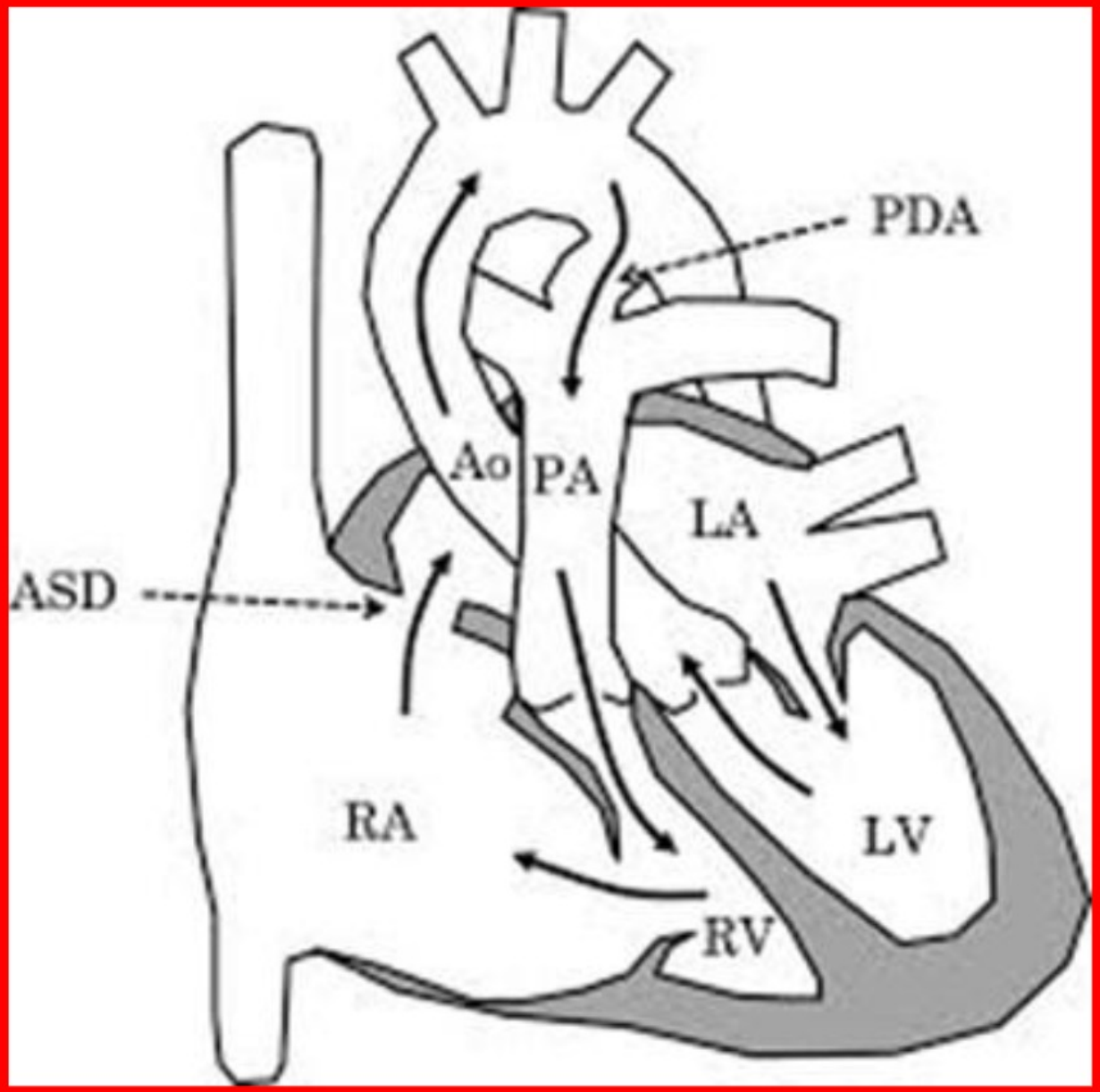


Image source: icvts.oxfordjournals .org

Failure

Failure in Medical Management may need Surgical Alternatives

Failure defined as:

Unable to wean inotropes	Persistent cyanosis	Decompensated CHF with organ dysfunction	Metabolic acidosis	Low TR Jet	PR

Surgical management

Neonatal surgical management associated with HIGH mortality

Starnes Procedure (1991,2008)	• RV exclusion • Atrial Septostomy, TV closure, AP shunt creation
Knott-Craig monocusp technique (1994)	• Mobilization of Anterior leaflet to achieve functional TV and biventricular repair
Carpentier Procedure (1988)	• TV repair based on delamination of the three leaflets based on the anatomy
Da Silva Procedure	• Cone Modification of Carpentier procedure (2007,2020) in neonates

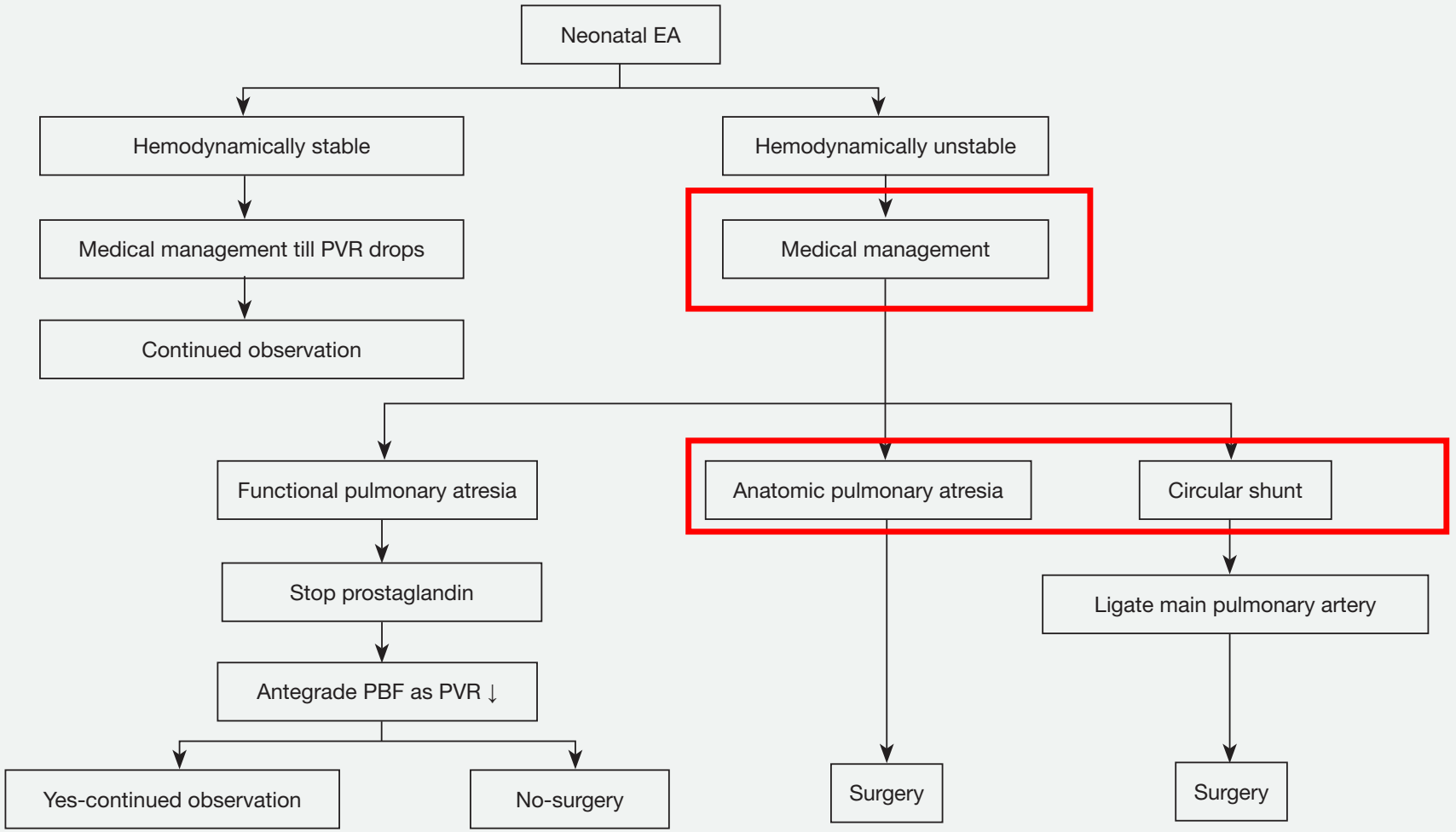


Figure 3 Management algorithm for neonatal EA. EA, Ebstein's anomaly; PVR, pulmonary vascular resistance; PBF, pulmonary blood flow.

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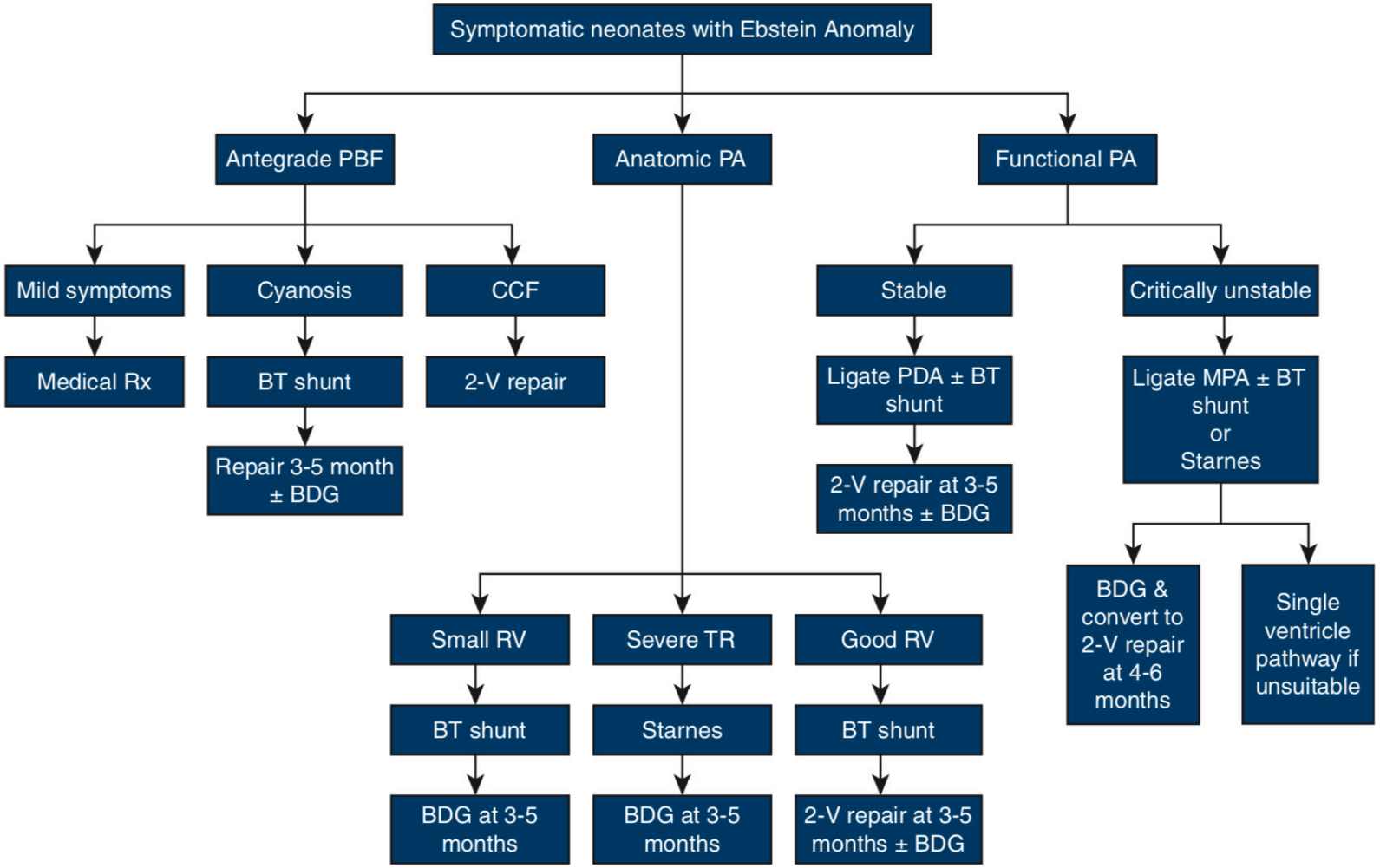

FIGURE 6. Algorithmic approach to the severely symptomatic neonate with Ebstein anomaly. *PBF*, Pulmonary blood flow; *PA*, pulmonary atresia; *CCF*, congestive cardiac failure; *BT*, Blalock-Taussig; *BDG*, Bidirectional GLenn; *RV*, right ventricle; *TR*, tricuspid regurgitation; *PDA*, patent ductus arteriosus; *MPA*, main pulomonary artery.

Current surgical techniques in the management of
the symptomatic neonate with severe Ebstein anomaly: Too much, too little, or just enough?
Christopher J. Knott-Craig, MD, FACS, and Umar S. Boston, MD

Conclusions

Prenatal diagnosis of EA difficult

- Neonatal Presentation usually sicker patients

Neonatal presentation with profound cyanosis and heart failure is associated with increased mortality

Neonatal medical management directed to decrease PVR, decrease RV afterload, improve cardiac output and optimize oxygenation

Surgical correction considered when medical management fails, better outcome in older patients

A multidisciplinary approach that transcends individual institutions' walls will be necessary to lead to improved outcomes


GRACIAS