

Ebstein Anomaly

- Treatment Strategy –
(Use or Abandon the RV ?)

장기영

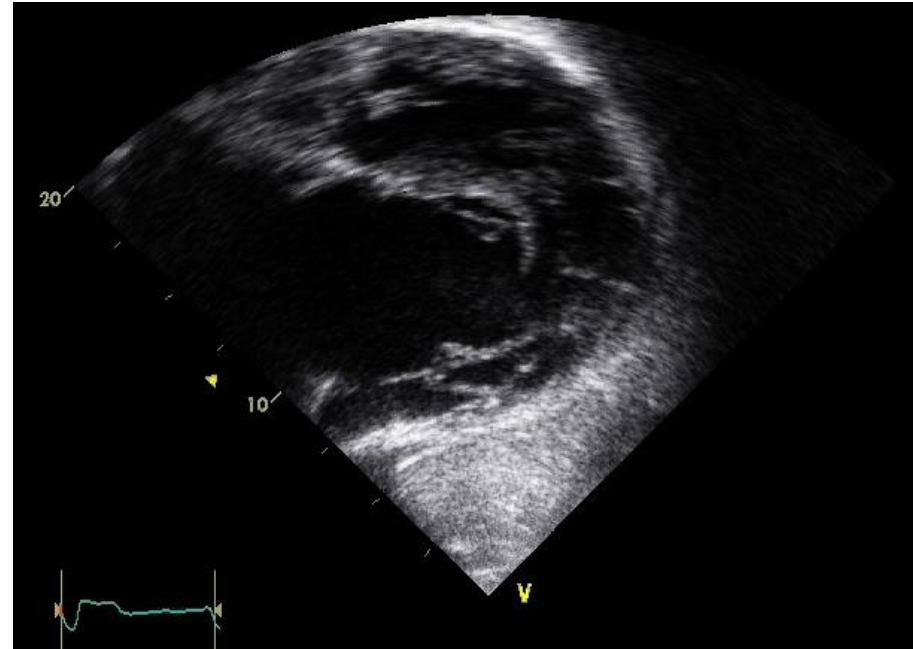
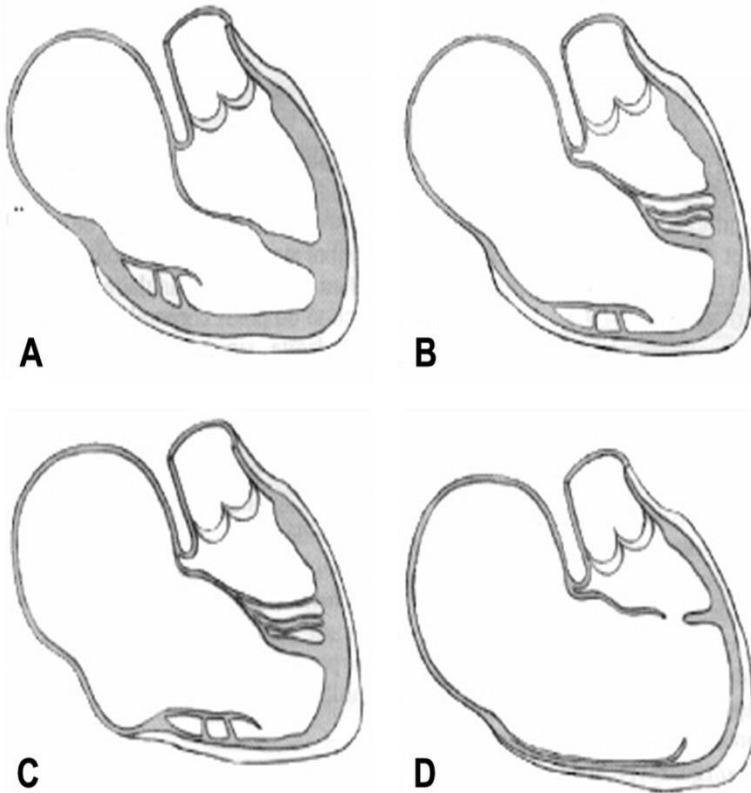
고려대 안산 병원

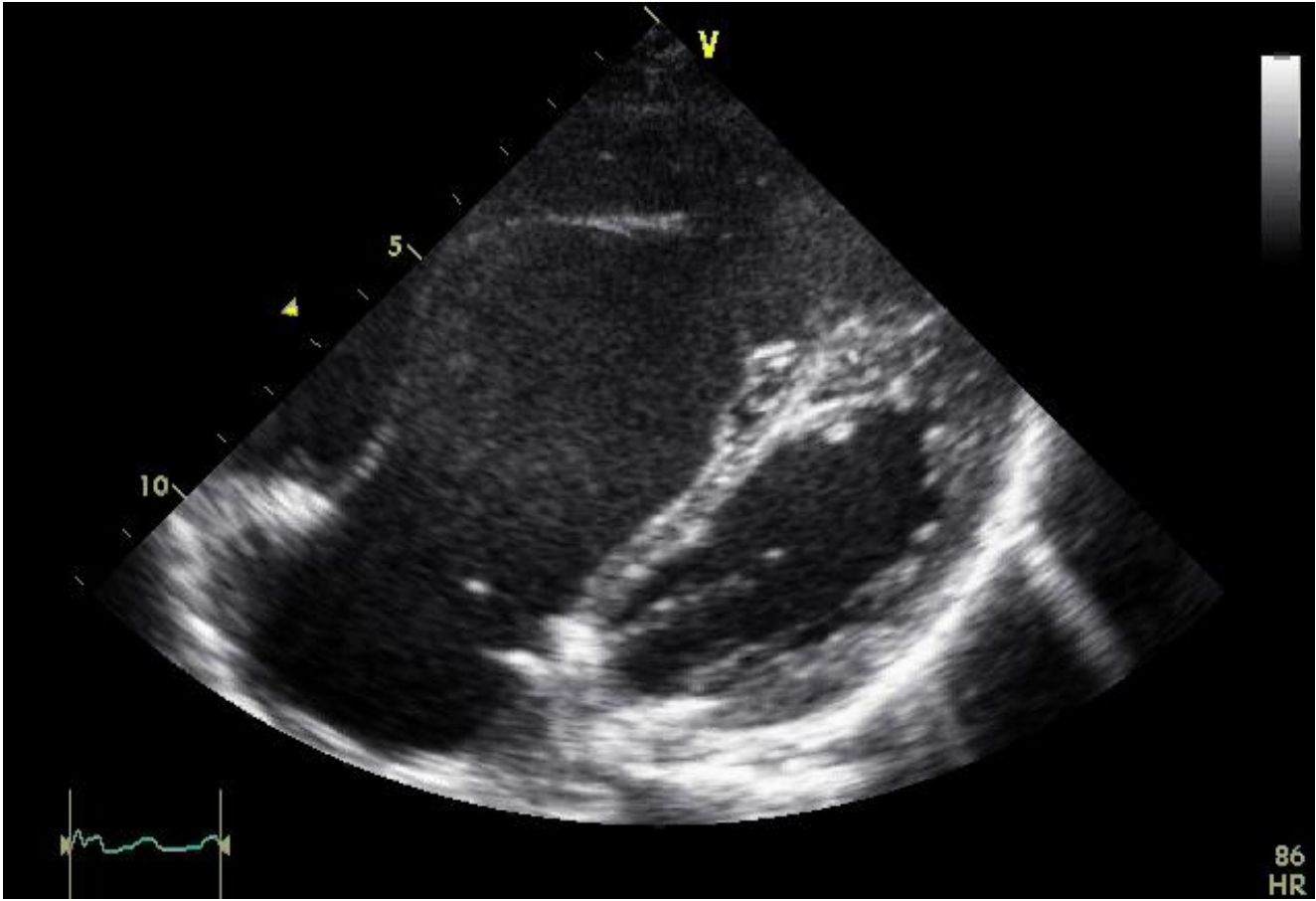
Introduction

- Ebstein Anomaly
 - Wide spectrum of anatomy
; variable abnormalities of
TV dysplasia and RV.
 - => infinite degree of anatomic variabilities.

TV and RV dysplasia

- wide spectrum -





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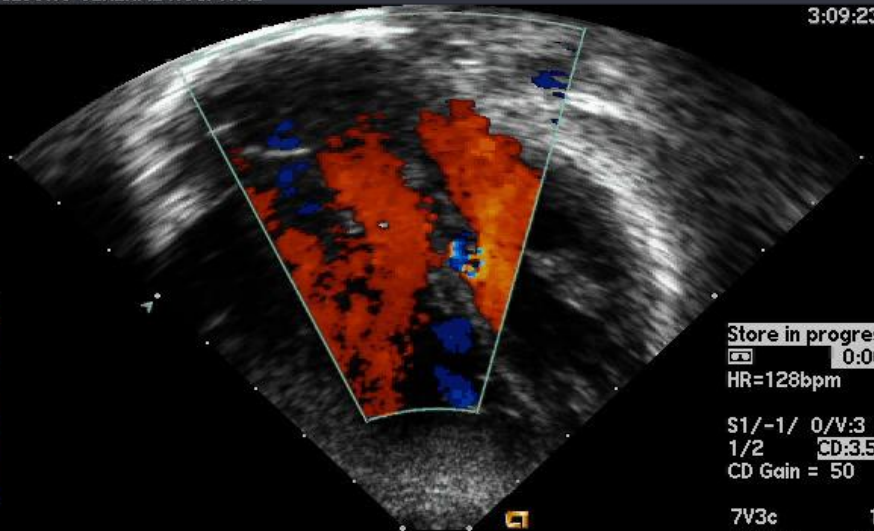
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06 Sep 05

3:09:23 pm



.79
Store
HR=12
60dB
Gain=
7V3c .79
6.0MHz
PED EC
Gener



Store in progress
0:00:13
HR=128bpm
S1/-1/ 0/V:3
1/2 CD:3.5MHz
CD Gain = 50
7V3c 19Hz
6.0MHz 80mm
PED ECHO
General



CD Pan CD Pos/Size

TV and RV dysplasia

- wide spectrum -

- Each heart is different from Pt to pt.
 - definite classifications are difficult
 - ➔ definitive selection of Tx options is not easy.

Ebstein Anomaly

- wide spectrum -

- infinite degree of anatomic variabilities
 - > Various symptoms and sign
 - Asymptomatic,
 - exercise intolerance,
 - cyanosis,
 - increasing cardiomegaly,
 - palpitations....

- Wide spectrum of anatomy

=> several kinds of Treatment options..

Treatment Options

- Observation
- Operation
 - Bivent. Repair
 - 1 1/2 repair
 - single vent. Repair (RV exclusion)
 - transplantaion ...

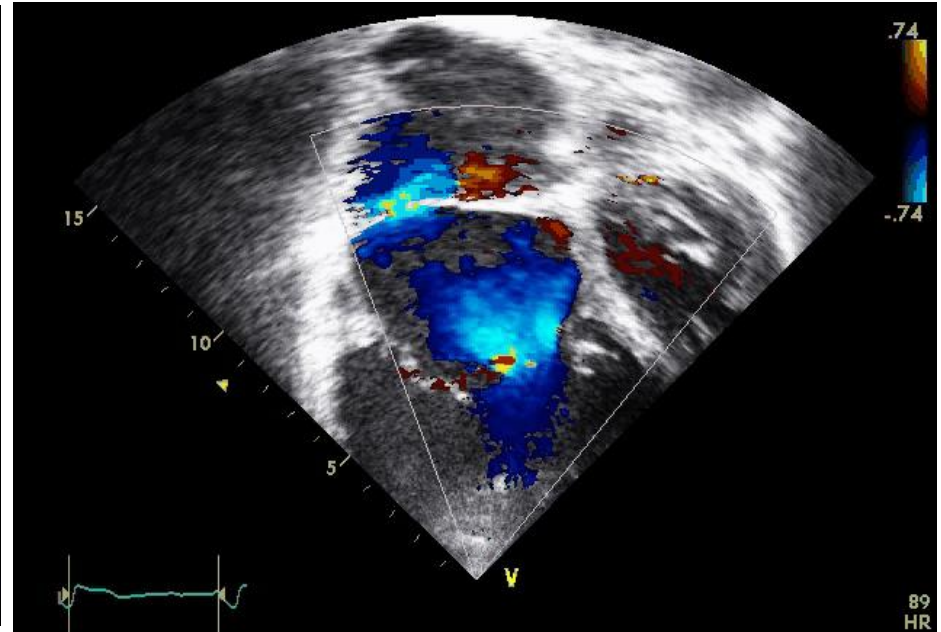
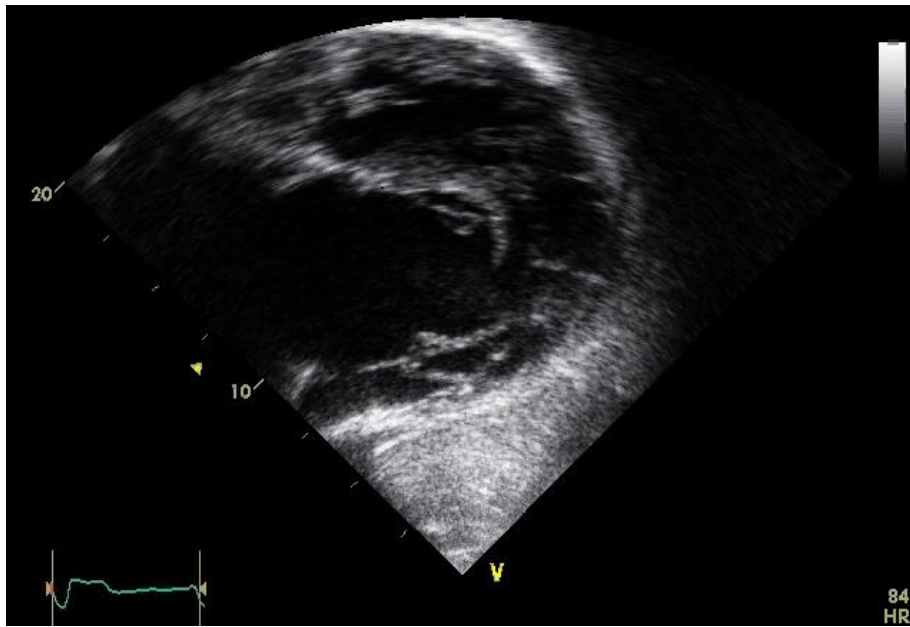
Op. Indication

- Progressive symptoms
- Exercise intolerance
- Increasing cyanosis
- Increasing cardiomegaly
- Worsening RV dilatation
- Deteriorating RV dysfunction
- Progression of atrial or vent. arrhythmia

Biventricular Repair

- No significant RVOTO
- Good –sized RV (esp, functional RV)
- Reasonably functioning tricuspid valve
(esp. anterior leaflet)

One-and-a-half Vent. repair



One-and-a-half Vent. repair

- Severe Ebstein Anomaly
 - massive TR
 - severely enlarged RV
 - dysfunctional RV
 - small, squashed LV
 - long standing at. Fib.

One-and-a-half Vent. repair

- add **BCPS** + intracardiac repair
 - **BCPS** : Advantage
 - unloading the enlarged, dysfunctional RV
 - optimize preload to LV
- #. Severe Ebstein, (severely impaired RV)
-→ high risk for surg. (in 2-vent. repair)

One-and-a-half Vent. repair

- BCPS parameter in Ebstein
 - m PAP < 18-20 mmHg
 - LVEDP < 15 mmHg
 - transpul. PG < 10 mmHg

One-and-a-half Vent. repair

- Disadvantage
 - facial suffusion
 - pulsation of head / Neck
 - collateral vein , pul A-V fistula
 - limited access (EP, ablation, PPM)

Bi-directional cavopulmonary shunt associated with ventriculo and valvuloplasty in Ebstein's anomaly: benefits in high risk patients

S. Chauvaud*, J.F. Fuzellier, A. Berrebi, P. Lajos, J.P. Marino, S. Mihaileanu, A. Carpentier

- 60 pts (severe Ebstein A)

 - massive TR, enlarged and dysfunctional RV, or long standing A.fib.

 - . Gr I (bi-vent. Repair, 45 pts)

 - Gr II (1 ½ repair, 15 pts)

	Gr. I	Gr. II	
Op. mortality	24%	0%	P<0.05
5-YSR	66.1%	80%	NS
Re-op	11%	0%	
Residual TR	26%	26%	

Bi-directional cavopulmonary shunt associated with ventriculo and valvuloplasty in Ebstein's anomaly: benefits in high risk patients

- Cause of mortality in Gr I (11/45 pts)
 - RV failure 5
 - LV failure 1
 - arrhythmia 2
 - infection 2
 - MOF 1

Bi-directional cavopulmonary shunt associated with ventriculo and valvuloplasty in Ebstein's anomaly: benefits in high risk patients

- in high risk Ebstein A.
 - : 1 ½ repair
 - decrease op. mortality and re- operation.
 - reasonable strategy in high risk pt

Results of the 1.5-ventricle repair for Ebstein anomaly and the failing right ventricle

Luis G. Quinonez, MD,^a Joseph A. Dearani, MD,^a Francisco J. Puga, MD,^a Patrick W. O'Leary, MD,^{b,c} David J. Driscoll, MD,^{b,c} Heidi M. Connolly, MD,^c and Gordon K. Danielson, MD^a

- 14 pts (severe EA and severe RV dysfn.)

; TV repair or replace , RV resection + BCPS

(3pts : transplantation candidate)

-> 9 pts; planned BCPS

5 pts: intraop. Salvage procedure

➔ mortality (1)

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#. In severely enlarged & dysfunctional RV

: 1 ½ repair

- planned procedure in high risk Ebstein.
- intraop- salvage procedure for RV failure
- improve LV function

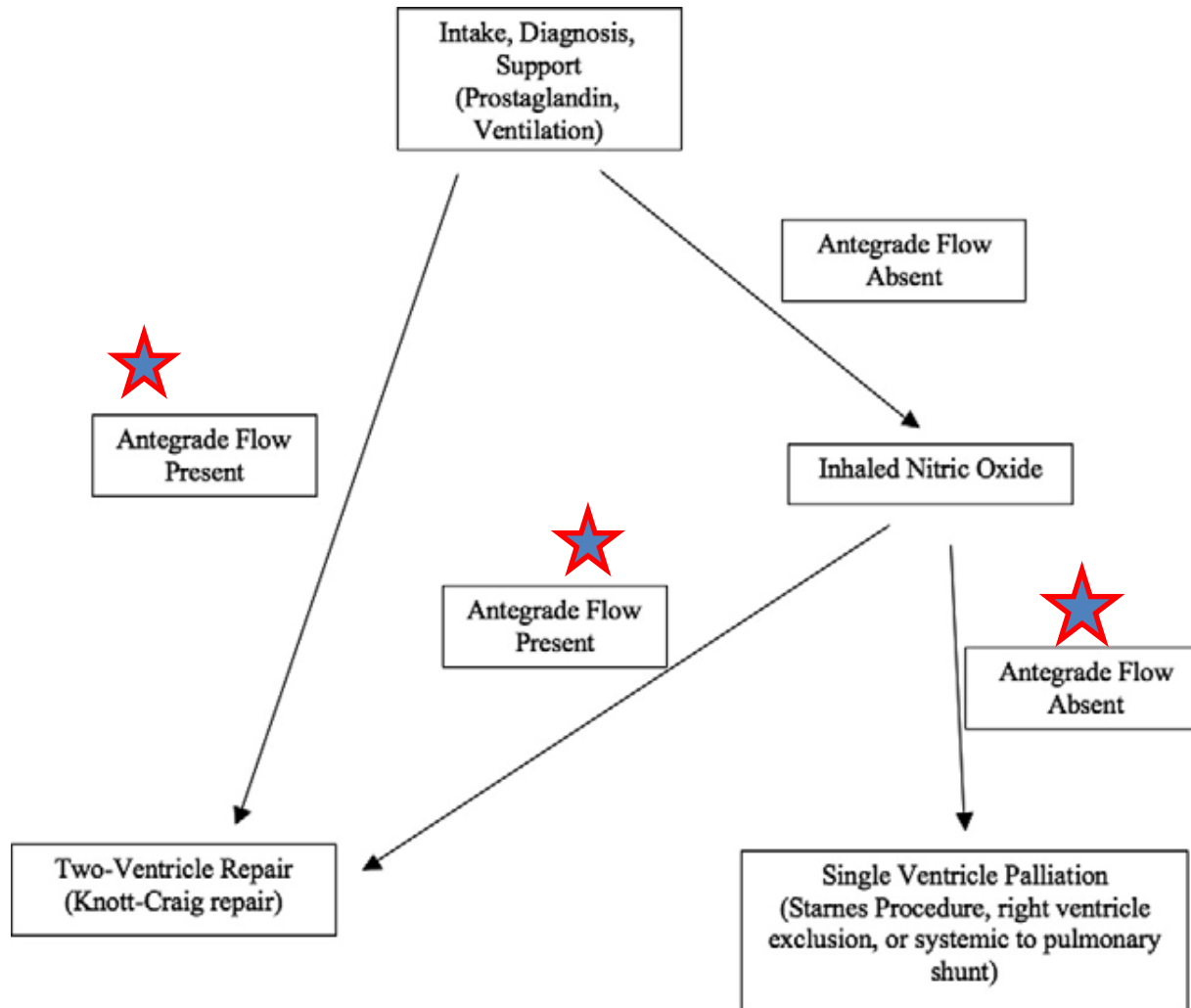
RV Exclusion (1-vent. Repair)

- Indication
 - severe Ebstein A (esp. neonate)
(severely enlarged & dysfunctional RV)
; overt heart failure, cyanosis, severe cardiomegaly
 - + Pul. Atresia / hypoplastic PA
(anatomic or functional)
 - ; functional PA (→ NO evaluation)

RV Exclusion (1-vent. Repair)

- Op strategy
 - to assess the possibility of TV repair,
f-RV status,
RVOT status.
 - consider RV exclusion : Starnes procedure
(TV closure c fenestration,
reduction atrioplasty,
B-T shunt)

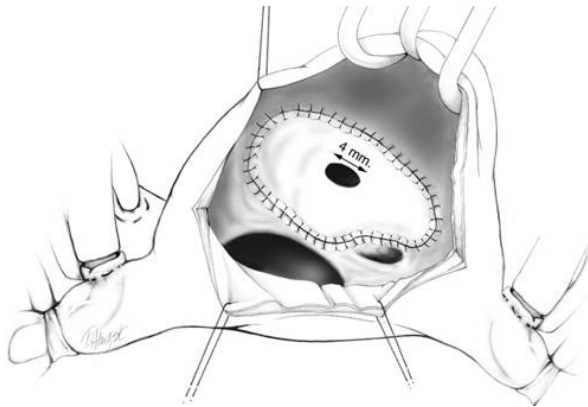
depends on RVOT flow



RV Exclusion (1-vent. Repair)

- Neonatal Ebstein with
severe ht failure, cyanosis, acidosis,
severe cardiomegaly
 - ➔ most will die w/o intervention.
(TV repair ➔ high mortality)
 - =➔ consider RV exclusion.

RV Exclusion (1-vent. Repair)



➔ Big impact at severe end of Ebstein.

- diminutive true RV
- highly laminated leaf.
- unguarded TV orifice
- RVOTO
- marked Rt chamber dilatation
- squashed LV

#. RV exclusion

➔ simplify initial palliation
(no need for RVOT repair
TV repair..)

RV Exclusion (1-vent. Repair)

- In severe neonatal Ebstein,
 - good single vent. Repair is better than bad 2-vent. Repair.
 - RV exclusion allow the LV to function effectively.

RV Exclusion (1-vent. Repair)

- Disadvantage
 - single-vent. Fontan pathway
 - at least 2- subsequent op.
 - limited access (EP, ablation, PPM)

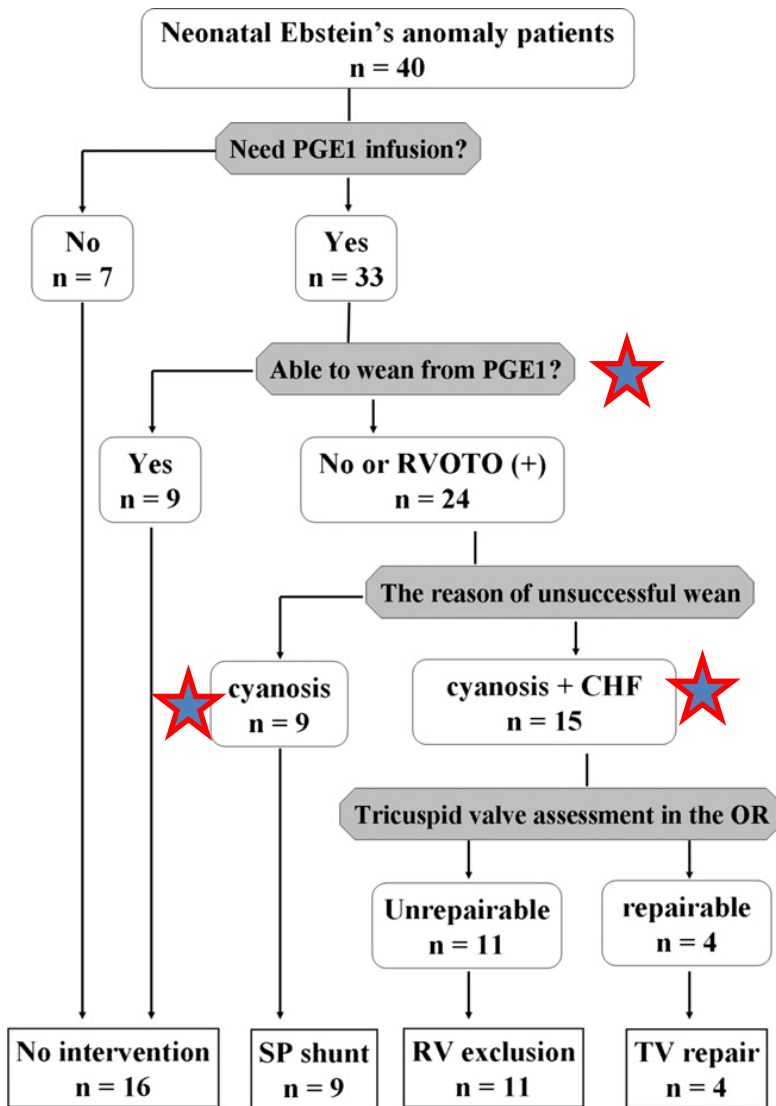
Management and long-term outcome of neonatal Ebstein anomaly

Takeshi Shinkawa, MD,^a Anastasios C. Polimenakos, MD,^a Carlen A. Gomez-Fifer, MD,^b
John R. Charpie, MD,^b Jennifer C. Hirsch, MD,^a Eric J. Devaney, MD,^a Edward L. Bove, MD,

- 40 pts (neonatal Ebstein)
 - symptomatic neonate; challenging
 - optimal surgical intervention; controversial

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• Decision making guideline

1) **PGE 1 weaning** ?

. yes → observation

. no →

2) symptom

. **cyanosis** →

. **cyanosis + CHF** →

Current surgical therapy for Ebstein anomaly in neonates

Brian L. Reemtsen, MD, Brian T. Fagan, MD, Winfield J. Wells, MD, and Vaughn A. Starnes, MD

- 16 pts, symptomatic neonate c Ebstein
- B.wt; 3.1 kg, age; 8 days (±10)

{ . 1 → transplantation
{ . 3 → TV repair { -> 1: dead
 -> 1: alived
 -> 1: RV exclusion
{ . 12 → RV exclusion

=> Tot: 13 RV exclusion (-> 4 : dead)

Transplantation

- Severe Ebstein A

(severely enlarged & dysfunctional RV)

+ [significant LV dysfunction (LV EF < 25%)
or
left heart obst. Defect
(Coa., MV hypoplasia)

Conclusions

- In Ebstein anomaly, there are wide spectrum of anatomy, diverse symptoms and signs, and several kinds of Treatment options.
- It's not easy to select definitive treatment option in symptomatic pt.
- So, we have to find optimal treatment options according to symptoms, degree of anatomic severity, and hospital environments.

Timing & Choice of Tx

- Depends on
 - clinical presentation,
 - TV morphology,
 - R→L shunting,
 - severity of Rt chamber dilatation,
 - severity of RV dysfunction,
 - atrial or vent. Arrhythmia.