

Ebstein's Anomaly

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History

- A 24 year old male was playing soccer, turned blue and passed out.
- His brother brought him to the hospital saying something similar had happened one year ago while in Mexico.
- His only other complain was palpitations.

History Continued

- No other medical problems.
- Social & Family History : No Tobacco or illicit drugs,occasional ETOH.
- Mother on some Psychiatric medications at the time of pregnancy per older brother.

Differential Diagnosis

Cardiovascular

Congenital
Arrhythmias
Valvular

Neurological

Stroke
SAH
Seizures

Pulmonary

Embolism

Hematological

Polycythemia
Hypercoagulability

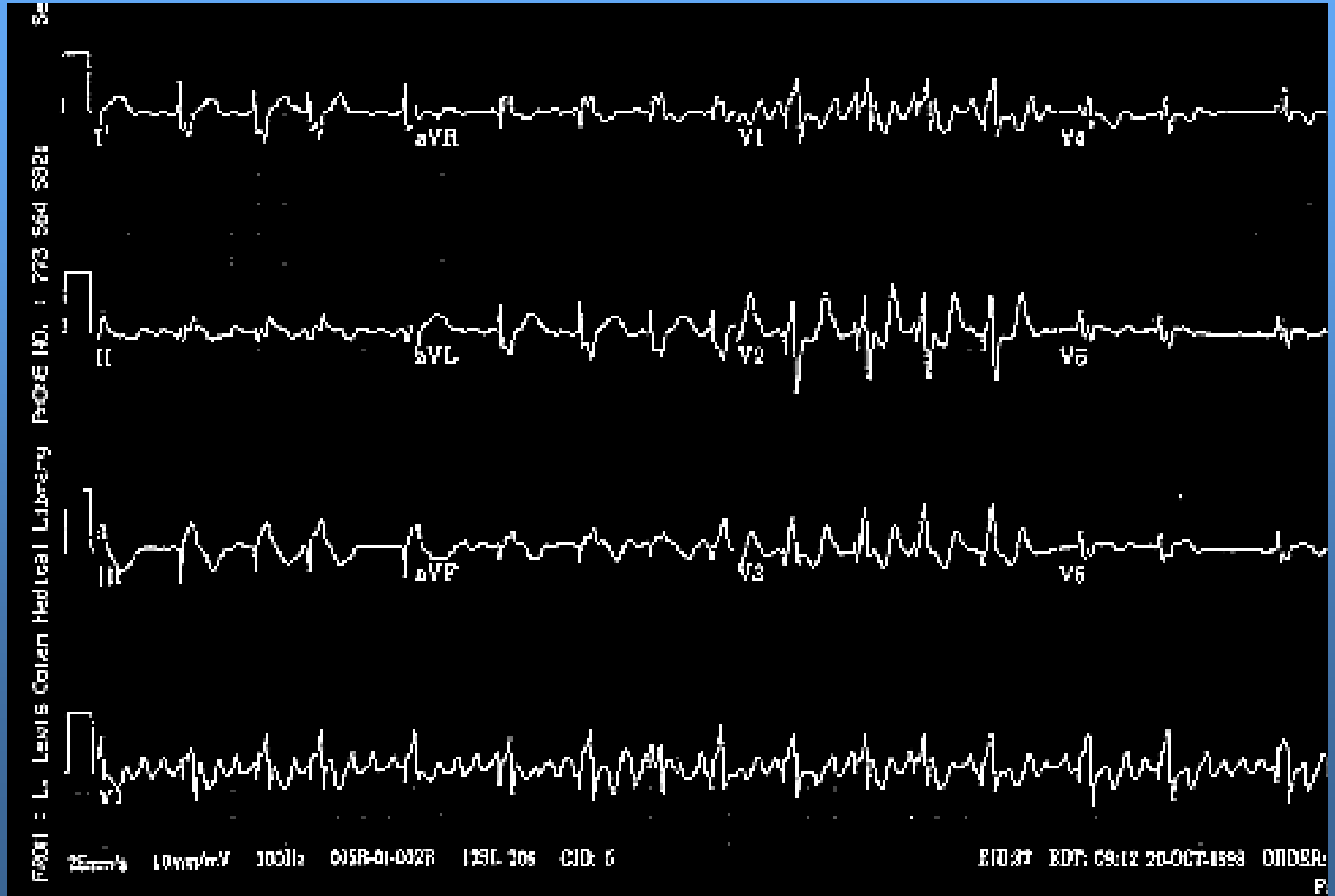
Cardiac Exam

- Cardiac:
 - Irregularly irregular rhythm.
 - Diffuse PMI, systolic thrill
 - S1 loud & split
 - S2 fixed split.
 - Holosystolic murmur 4/6 over the LSB.
 - Clubbing & Cyanosis in all extremities
 - No edema, good peripheral pulses.

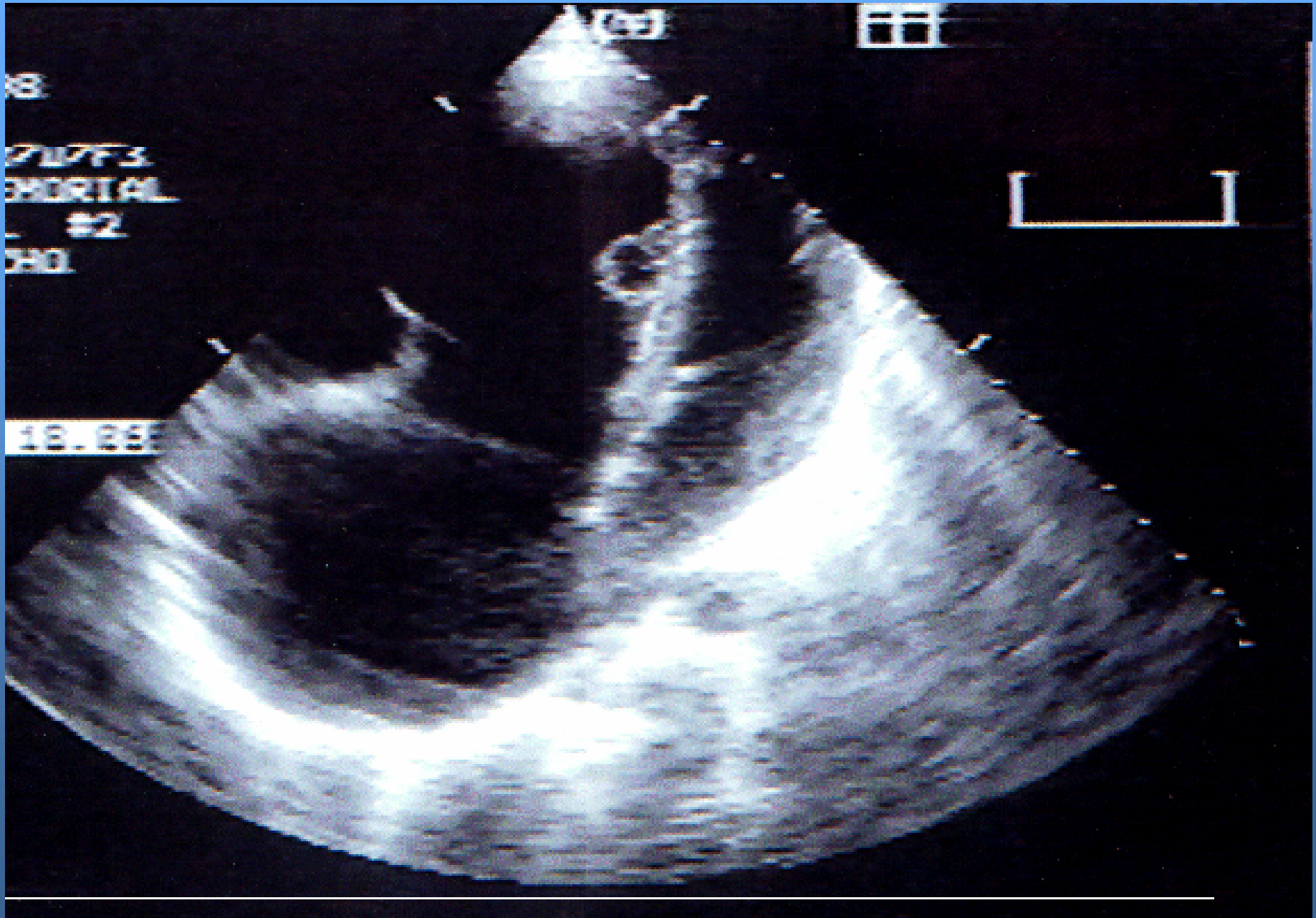
Lab

- Hematology: Hb 22 Hct: 69
 - normal WBC and platelet counts
- Chemistry: normal.
- ABG: hypoxemia REFRACTORY to O₂.
- CXR: Cardiomegaly.
- Spiral chest CT: normal.
- Brain CT: normal.

ECG



Echo



Cardiac Catheterization:

- Clean coronaries.
- Normal LV EF, normal LVEDP. No evidence of VSD or Right Ventricular outflow tract obstruction.
- Normal PA, LA & RA pressures.
- Evidence of Right to Left intra cardiac shunt through a PFO.

Ebstein's Anomaly

1. Definition:

- Apical Displacement of tricuspid valve.
- Right Heart has 3 parts: atrium, an atrialized ventricle and a ventricle.
- Majority have ASD or PFO.
- Some may have accessory tracts (WPW) or pulmonary valve atresia.

Ebstein's Anomaly

2. Epidemiology:

- Incidence: 1/20,000 live births.
- Children born to mothers exposed to lithium in the first trimester have an increase incidence (28-500 fold).

Ebstein's Anomaly

3. Clinical Features:

- History: syncope, cyanosis, palpitations.
- Physical Exam:
 - Neck veins: occasional A waves, no V waves.
 - CVS Exam: Systolic thrill; loud, split S1; fixed split S2. Pan systolic murmur along LSB.

Ebstein's Anomaly

4. Diagnostics:

- PHYSICAL EXAM.
- EKG:
 - Usually sinus. Q waves V1- 4 and RBBB.
 - SVT in 25-30 %.
 - Delta wave and pre-excitation common.
- *Echo is gold standard.*

Ebstein's Anomaly

5. Prognosis:

- Only 5% survive beyond 5th decade.
- Bad prognostic signs are:
 - Right Ventricular failure
 - NYHA class 3 or 4
 - Cardiomegaly
 - Cyanosis

6. Treatment:

- Tricuspid valve repair or replacement.