

# *Serious complications in Adult cyanotic congenital heart disease .....a clinical dilemma*

*Case Presentation  
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## **BACKGROUND:**

- Our patient is a 32 years old male married, presented to NHI GUCH clinic for follow up.
- His condition started since birth with cyanosis and difficulty of breathing during feeding and repeated attacks of chest infection, when he was diagnosed to have congenital cyanotic heart disease and his mother was told that he will need to do surgical repair which was unfortunately postponed several times.



- After that the patient started to develop worsening of cyanosis and dyspnea with exertion.
- At age of eighteen years he sought medical advice again echocardiography and hemodynamic study were done followed by **pulmonary artery banding**, but his symptoms didn't improve.
- Since then he had several attacks of chest infection.

## GENERAL EXAMINATION:

- Patient is alert, conscious, oriented to time place and person.
- He has an average built, and is lying flat.
- **Severe central cyanosis, So2 72% ( At room air)**
- **Drum stick clubbing** in upper and lower limbs.
- His ABP was 95/70 on presentation.
- HR was 90 bpm with regular average volume. With no special character and well felt peripheral pulsations.
- Abdominal Examination: Lax, no organomegally
- No lower limb oedema
- Chest Examination : showed **median sternotomy** scar, equal air entry at both sides, no additional adventitious sounds.



## CARDIAC EXAMINATION:

### Inspection and palpation:

Apex in the 6<sup>th</sup> intercostal space outside the midclavicular line, systolic bulge, diffuse.

### Auscultation:

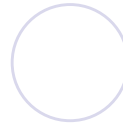
Muffled S1, Single accentuated S2  
Soft ejection systolic murmur grade 3/6,  
Early diastolic murmur



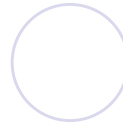
## LABS.:

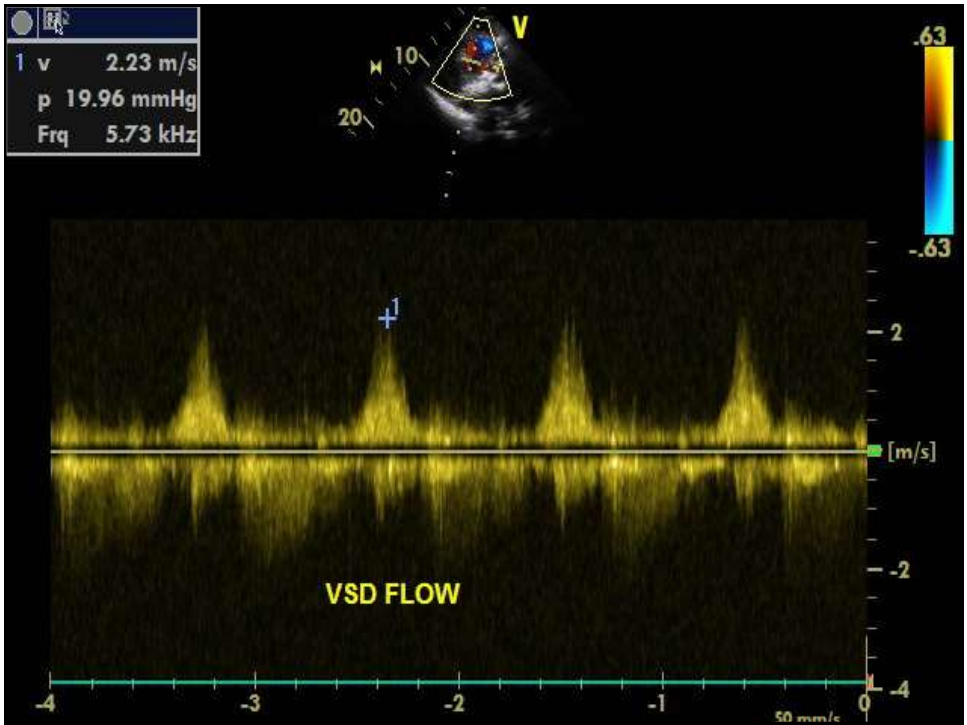
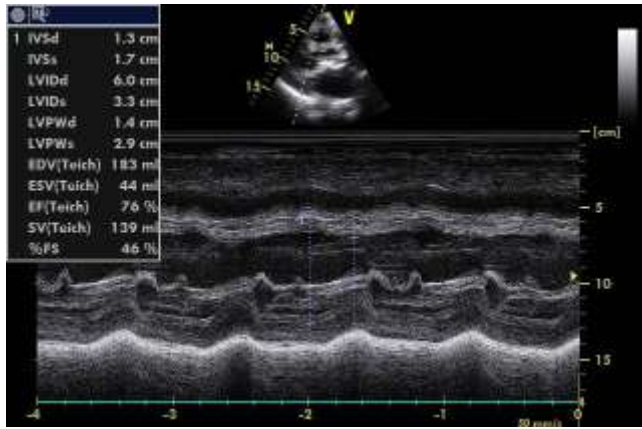
- HGB 21.7 g/dL .
- HCT 68.5 %
- WBC  $3.0 \times 10^3/\mu\text{L}$ .
- PLT  $129 \times 10^3/\mu\text{L}$ .
- RBS  $7.64 \times 10^3/\mu\text{L}$ .
- BUN 34 mg/dL.
- Creatinine 0.75 mg/dL.
- Sodium 165 mmol/L.
- Potassium 5.1 mmol/L.
- T.Bilirubin 0.97mg/dL.
- D.Bilirubin 0.1 mg/dL.
- U.A 7.5
- AST 36 IU/L.
- ALT 13 IU/L.
- Albumin 3.6 g/dL.
- INR 1.18

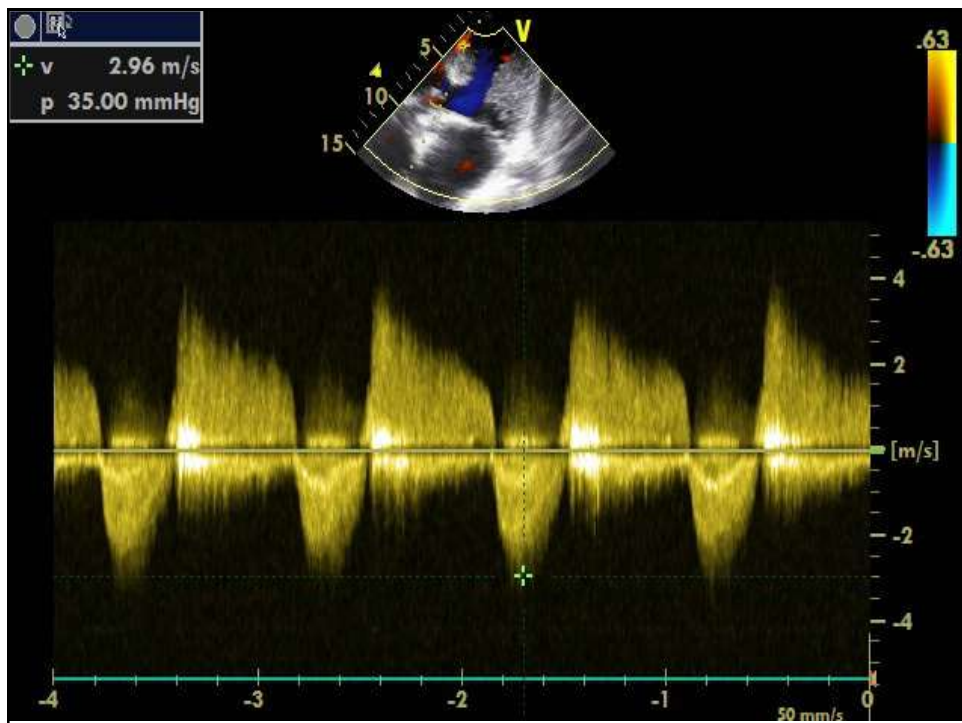
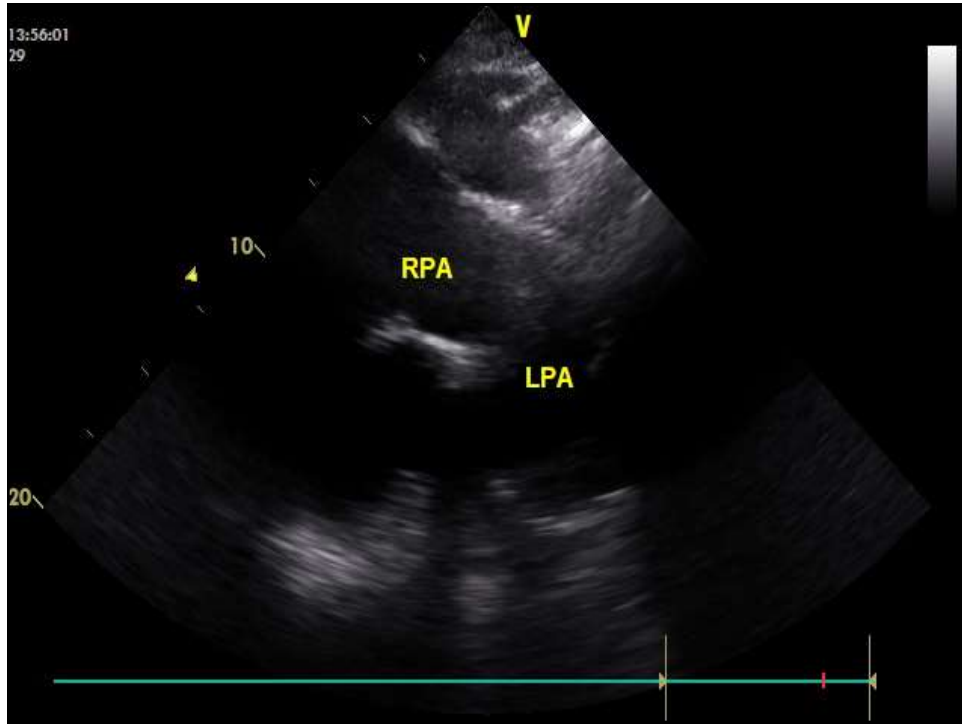
**ECG:**



**CHEST X-RAY:**








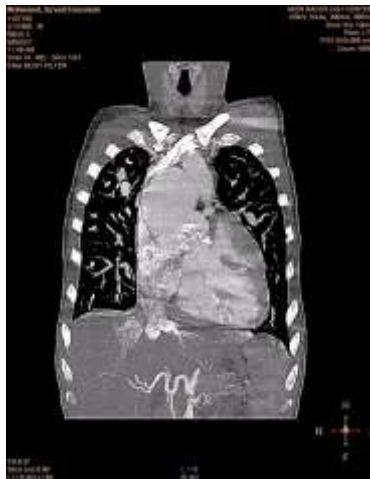
## CARDIAC MAGNETIC RESONANCE

- **Left ventricle:** dilated with good function  
EF ~55%, EDV = 333 ml, ESV =151 ml,  
SV=182 ml
- **Right ventricle:** fair function EF ~ 47%,  
EDV =111ml, ESV= 59ml, SV = 52 ml
- RVEDV:LVEDV=1.47:1
- Significant **pulmonary regurgitation**  
(regurgitation fraction ~57%)

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- TGA
  - Large nonrestrictive high muscular subpulmonic VSD
  - Dilated LV with good systolic function, supplying PA (posterior & right)
  - Small hypertrophied RV supplying AO (anterior & left)
  - Hugely dilated MPA and both branches, peak PG across LVOT ~ 35 mmHg (**lose band**), **significant PR**
  - **Unprotected pulmonary circulation.**
  - **Large ASD**
  - No PDA, no CoA
  - Normal systemic and pulmonary venous drainage

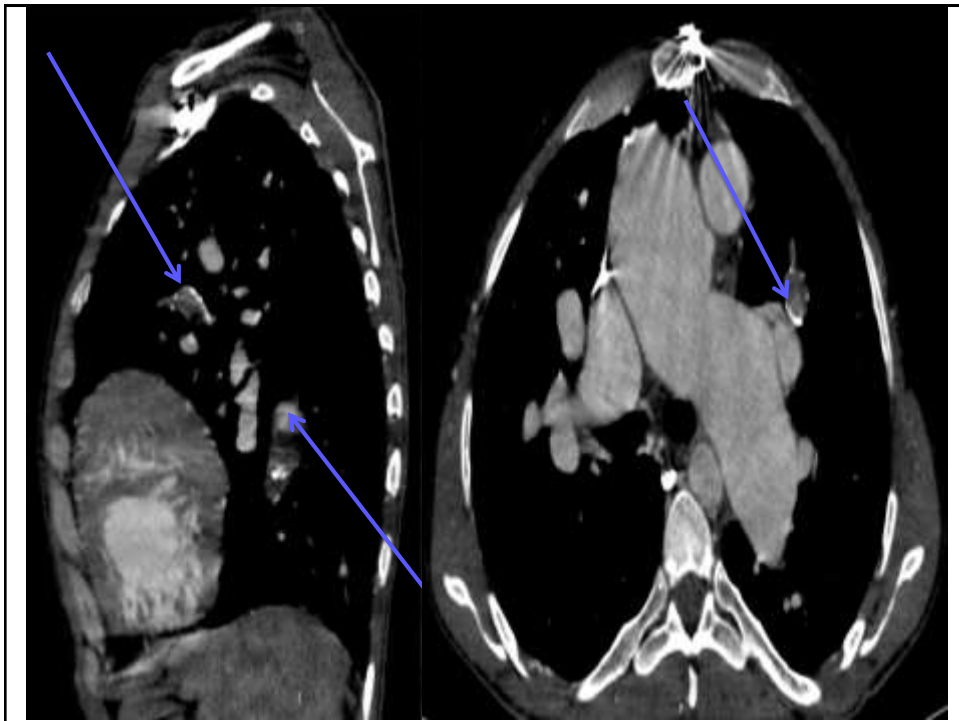


- One and half year ago he started to have attacks of blood tinged sputum followed by frank hemoptysis of about ½ cup which was repeated two times, progressive dyspnea, and chest pain.
- He went to E/R, he was hemodynamically stable, FFP was given.
- new echocardiography and laboratory investigations were done revealing same previous data, CT pulmonary angiography was done and accordingly bronchoscopy.

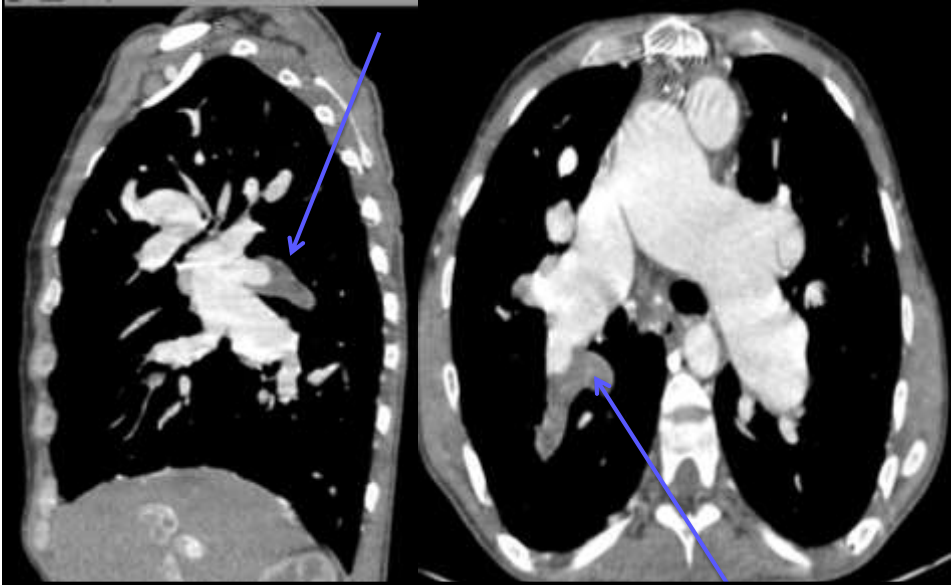




● ○ Left lung ● ○ ●



## Right lung



## Bronchoscopy with bronchial lavage

- No bronchial lesions
- No intrabronchial source of bleeding.
- C & S revealed pseudomonas spp and antibiotic were given according to the culture and Rutin, ascorbic acid combination (decrease capillary fragility)



- four months later patient developed another attack of frank hemoptysis of about ¼ cup associated with stitching chest pain and worsening of dyspnea, another CT pulmonary angiography was done which revealed





## REVIEW OF LITERATURE

**Massive pulmonary artery thrombosis with haemoptysis in adults with Eisenmenger's syndrome: a clinical dilemma**

C Broberg, M Ujita, [...], and M A Gatzoulis

Had case records with CHD with hemoptysis, CTPA showed pulmonary thrombosis. **Bleeding stopped spontaneously after observation in hospital** in the **first case**. The **second case** had experienced **several** episodes of **small** volume haemoptysis.. **Warfarin was instigated**. The patient returned five months later with **large** volume haemoptysis. His **I.N.R 2.3**, After cessation of warfarin he has had no further hemoptysis..

## Usefulness of Direct Oral Anticoagulants in Adult Congenital Heart Disease.

Pujol C, et al. Am J Cardiol. 2016.

- The study included 75 patients. Most common CHD were pre-tricuspid shunts(40%), complex CHD ( 21%), Mean follow-up was 12 months.
- Rivaroxaban was administered in 55 patients, apixaban in 13 and dabigatran in 7 patients for TEE prophylaxis in atrial arrhythmias, stroke/transient ischemic attacks, deep vein thrombosis, pulmonary embolism (n = 1) and atrial thrombi,. Some patients had >1 indication for adequate oral anticoagulation. CHA2-DS2-VASc score was  $\geq 2$  in 23 (31%), and 9 (12%) had a HAS-BLED score  $\geq 2$ .
- There were neither thrombotic or major bleeding events nor major side effects. In conclusion, direct oral anticoagulants appear to be safe and effective in ACHD. Long-term follow-up is needed.

## Clinical Implications of Hemoptysis in Patients with Pulmonary Arterial Hypertension

Jose Cantu, MD, Degang Wang, PhD, and Zeenat Safdar, MD, FCCP

- Hemoptysis is the result of engorged bronchial arteries with abnormal collateral vessel formation and systemic arterial pressures similar to that described in patients with CTEPH<sup>Kauczor HU, Schwickert HC, Mayer E, et al.</sup>
- Bronchial artery embolization is highly successful resulting in rapid cessation of hemoptysis with low complication rates for patients with CHD-APAH, However, re-bleeding is common, and referral for transplant consideration is prudent following the initial hemoptysis episode.



## TAKE HOME MESSAGE

- Pulmonary hypertension is a diverse disease with variety of causes and complications
- Pulmonary embolism is not uncommon complication of Eisenmenger complex and should be taken into consideration especially in **longstanding** cyanotic CHD.
- Clinical management of pulmonary artery thrombus and haemoptysis is an important and difficult problem which needs further data researches.
- When surgery is needed, it should be done in a timely manner or else....patient will suffer.



***THANK YOU***