

# WHERE IS THE CONCEALED SHUNT?

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## ***History :***

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A 15 year old child presented to the cardiology department 2 months ago by central cyanosis that improved by oxygen.

The condition started 3 years ago by rapidly progressive dyspnea that started 2 months after performing an appendectomy operation .

The father also noticed cyanosis of his son that was first intermittent and appeared with marked exertion but later it appeared with minimal exertion and even at rest.

During the 3 year period he sought medical advice and was hospitalized several times .

7 months ago the patient was admitted with an attack of hematemesis and bleeding per rectum.

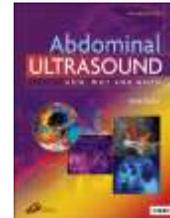
He performed upper GIT endoscopy that showed: **grade II oesophageal varices** ,and severe **portal hypertensive gastropathy** with active bleeding spots,

Duodenum also showed **multiple scattered ulcers**

## ***Investigations :***

Hb:10 g/dl  
PLT: 79 ,000/cm,  
ALT: 48 U/L  
AST: 91 U/L  
Cr.: 0.5 mg/dl  
Albumin:2 g/dl  
BUN: 6.0 mg/dl  
HCV antibodies are positive.

## ***ABDOMINAL ULTRASOUND :***



\*\**Liver*: Showing coarse echopattern ,irregular nodular surface , measuring 11.9 cm in MCL .  
No  
evidence of hepatic focal leisons .

\*\**Spleen*: is enlarged in size measuring 17.7 mm with homogenous parenchymal splenic vein 8.7 .mm in caliber with splenic collaterals .

ECG: within normal ,

Chest X-Ray: normal ,

Echo : Moderate PHT ,

*contrast echocardiography* was done showing passage of microbubbles from the right to the left side with the third beat.

CT- angiogram and MRI were done showing no abnormalities in both lungs and pulmonary arteries



## Abdominal CT-Angiogram

- ▶ No Intrahepatic shunts, with normal portal venous system.

**PULMONARY ANGIOGRAPHY WAS  
DONE:**

- ▶ Pulmonary angiogram of RT. And LT.

Pulmonary arteries shows multiple micro pulmonary AV fistulae of both lungs mainly on the right side appearing as ground glass appearance.

### Selective PA branches injection:

- ▶ No big or sizable fistulae to be closed
- ▶ Pulmonary veins are in place without anomalous insertion (no other cause of desaturation).

- ▶ **Test occlusion of RPA & LPA by balloon (mechanical pneumonectomy): marked improving in O2 saturation was noticed and marked reduction in PAP (more on Rt. Side) indicating a contribution of both lungs.**

**Test occlusion of a Small PFO by balloon: No improvement in saturations.**

Provisional diagnosis:  
**Hepato-pulmonary Syndrome**

## **MECHANISM:**

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The hepato-pulmonary syndrome results from the formation of microscopic intrapulmonary arteriovenous dilatations in patients with both chronic and acute liver cell failure.

The mechanism is unknown but is thought to be due to increased hepatic production or decreased hepatic clearance of **vasodilators** possibly involving **nitric oxide**.

## TREATMENT:

Hepatopulmonary syndrome (HPS), affects 47% of patients with end-stage liver disease

The main treatment is supplemental oxygen for symptoms.

Other therapies, such as **somatostatin** to inhibit vasodilation, are of modest benefit in only some patients.



Inhaled **nitric oxide** synthesis inhibitors may be a future treatment option.

Results of HPS management have been disappointing.

Liver transplantation may result in the resolution of HPS, and it is not a contraindication to liver transplantation.

## PROGNOSIS:

Prognosis is poor without treatment: the presence of hepato-pulmonary syndrome worsens the prognosis , of the underlying disease.

