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Case Report

# The Mechanical Cause of Myocardial Infarction in Eisenmenger Syndrome: Unidimensional Left Main Compression

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## Abstract

Eisenmenger syndrome may rarely present as acute coronary syndrome due to extrinsic left main compression by the dilated and hypertensive pulmonary artery. Though computed tomography (CT scan) has been the investigation of choice for diagnosis of this entity, high-definition intravascular ultrasound (HD-IVUS) may play a crucial role in diagnosis especially in sick patients with a characteristic finding of a "slit-like" ostia. Treatment of this "left main compression syndrome" with a drug eluting stent of high radial strength, evidenced angiographically as well as by HD-IVUS makes percutaneous coronary intervention (PCI) a preferred strategy to coronary artery bypass graft (CABG) in such high-risk patients.

**Keywords:** eisenmenger syndrome; left main compression syndrome; non- atherosclerotic acute coronary syndrome; high-definition intravascular ultrasound

## Introduction

Anginal symptoms may be ignored in patients with Eisenmenger Syndrome because of overlap features like dyspnoea and cyanosis. It is not uncommon to see patients with Atrial Septal Defects (ASD) presenting in their late fifties with heart failure. However, patients presenting with acute coronary syndrome (ACS) secondary to left main coronary artery compression as a mechanical effect of dilated and hypertensive pulmonary artery is an under-suspected and under-reported cause of angina in this subset. CT Aortogram can beautifully depict this entity but in sick patients, intracoronary imaging especially with HD-IVUS can also help in appropriate recognition of this entity.

## Case Presentation

We describe a case report of an unoperated neglected case of Atrial Septal Defect patient with Eisenmenger syndrome. A 62-year diabetic post-menopausal female presented in emergency with severe chest pain for eight hours. She was in cardiogenic shock with blood pressure of 80 mm systolic and heart rate of 134 beats per minute. She was oliguric for ten days. On interrogation, family mentioned that she was diagnosed as a case of atrial septal defect but neglected care. She had New York Heart Association (NYHA) class II exertional dyspnoea for eight years that had progressed to NYHA III in past six months. She had features of right heart congestion with elevated jugular venous pressure and grade three pandigital clubbing. Cardiovascular examination revealed gallop rhythm with marked tachycardia, loud P2, pansystolic murmur at left lower sternal border. Patient was tachypnoeic with SpO2 of 86% on room air, with crepitations in both lung bases (right more than left). The troponin and baseline serum creatinine were elevated (3.14 ng/ml and 2.84 mg/dl respectively).

Case is summarized in Figure 1.

## Clinical Details – 62 year old female, Post menopausal

- Diabetic for 10 years
- DOE II for 8 years
- NYHA III for 6 months
- Oliguria for 10 days
- Severe chest pain for 8 hours
- BP = 80 systolic
- HR = 134/min
- JVP – Raised
- Right heart congestion
- Grade 3 Digital Clubbing
- Diagnosed ASD on Echocardiogram 3 years back but neglected care
- CVS – Gallop Rhythm, Loud P2, PSM at LLSB, Carvallo's Sign
- RR = 36/min
- Extensive Bilateral Crepts
- SpO2 = 86%
- Troponin Positive (3.14 ng/ml)
- Creatinine = 2.84 mg/dl

The ECG (Figure 2) at presentation revealed a narrow QRS tachycardia at 140bpm, possibly atrial tachycardia with 2:1 AV conduction, right axis deviation, right bundle branch block, right ventricular hypertrophy and secondary ST depression and T wave inversion in precordial leads.

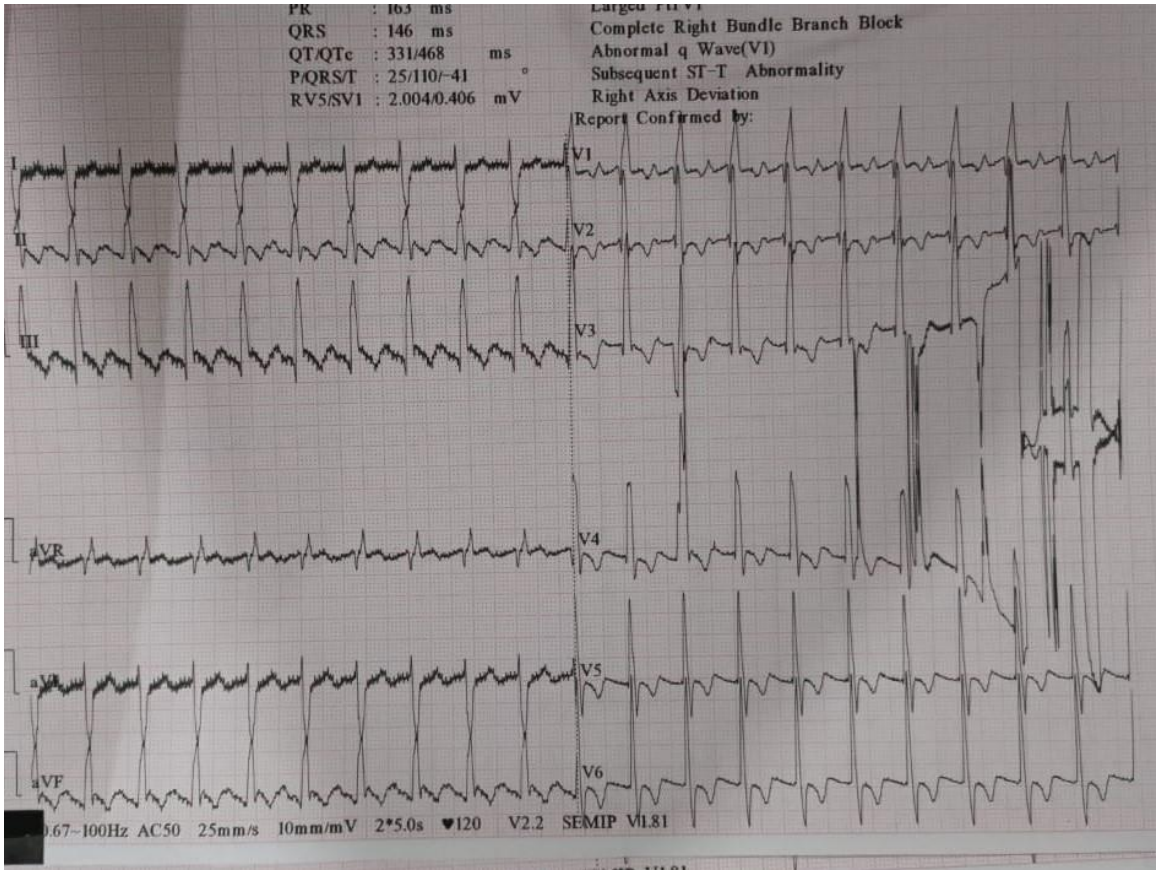



**Figure 2** – Baseline ECG

Transthoracic Echocardiogram (Figure 3a,b,c,d,e) showed a large 34 mm Ostium secundum Atrial Septal defect with bidirectional shunt, predominantly right to left shunt with severe tricuspid regurgitation, dilated non collapsible inferior vena cava and severe pulmonary artery hypertension, predicted right ventricular systolic pressure of 76 mm of mercury against a systemic systolic pressure of 80 mm of mercury and MPA diameter 2.8 times that of aorta.

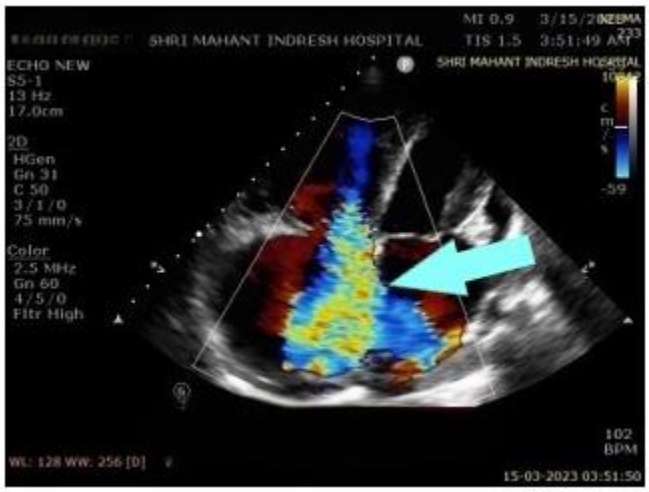



**Figure 3(a) –** Apical 4 chamber view showing large ostium secundum atrial septal defect with severe tricuspid regurgitation and predominantly right to left shunt across the defect.

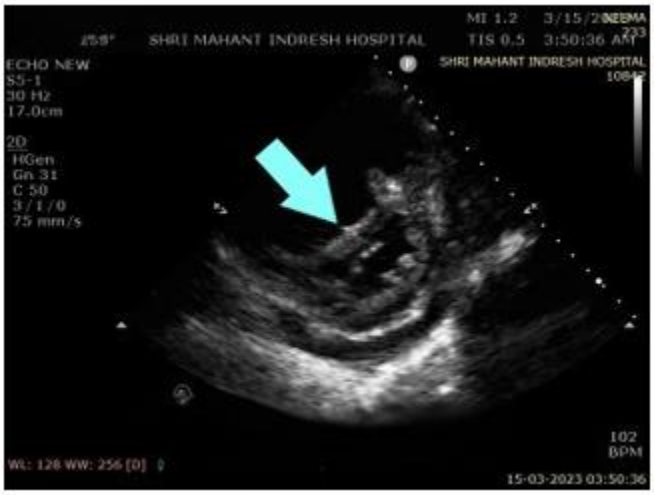



**Figure 3(b) –** Transthoracic short axis view at ventricular level showing D-shaped LV because of severely elevated right sided pressures.

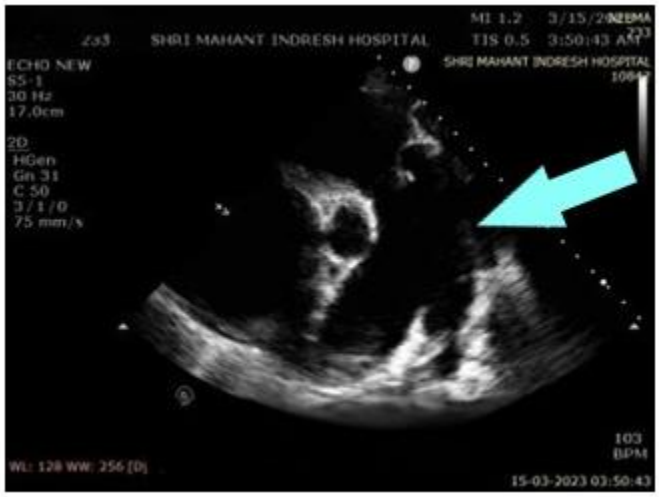



**Figure 3 (c)** – Transthoracic short axis view at arterial level showing dilated pulmonary artery with PA to Aorta ratio of 2.8.

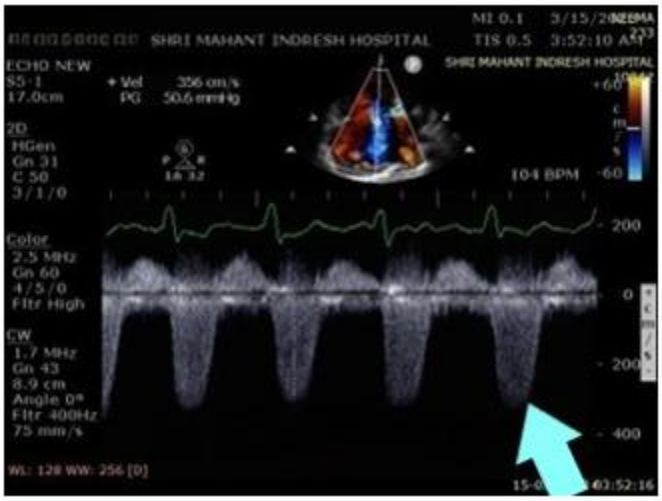



**Figure 3 (d) –** Continuous Wave Doppler at tricuspid valve showing markedly elevated right ventricular systolic pressures.

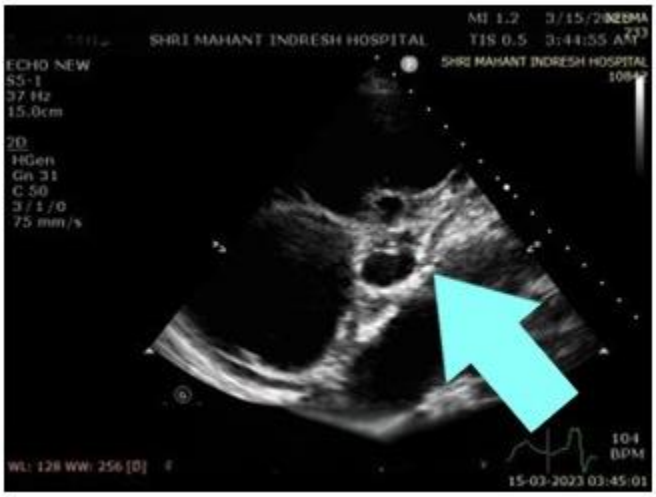



**Figure 3 (e) –** Modified short axis view at arterial level showing the course of left main arising from aorta

Coronary Angiogram (Figure 4a,b,c) showed eccentric left main stenosis that was not evident in caudal views but only in cranial views.

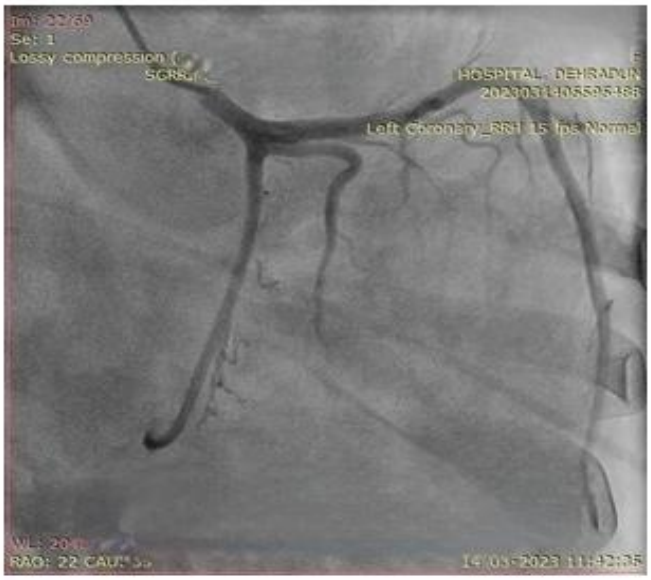



**Figure 4 (a)** – AP Caudal view showing no significant stenosis in left main.

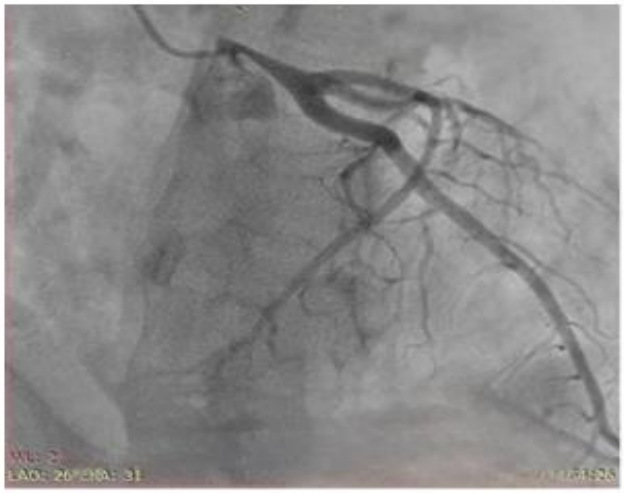


**Figure 4 (b) –** AP Cranial view showing significant stenosis in ostial left main (Unidimensional stenosis).

Right coronary artery was anomalous and arising high from right coronary cusp with downpointing ostia and aorta was diminutive.

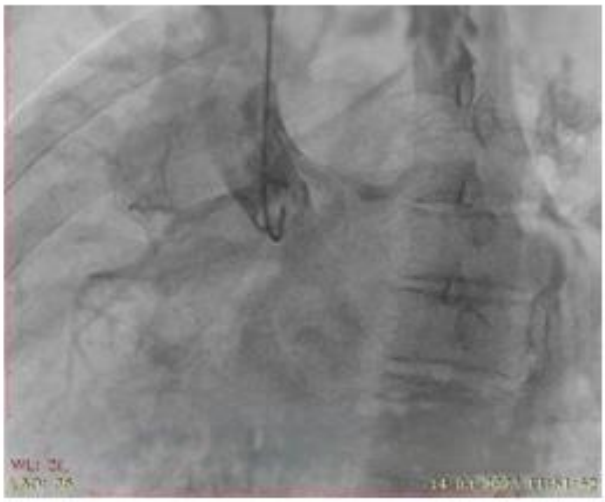
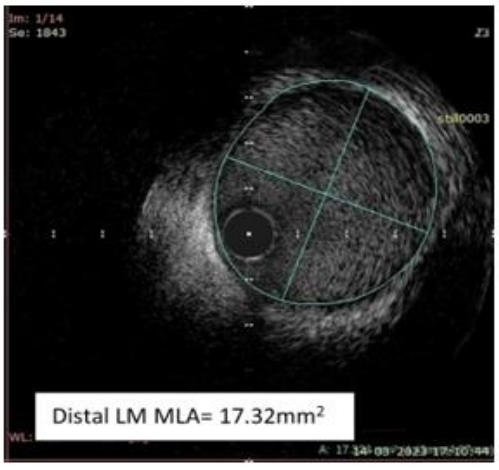


**Figure 4 (c)** – LAO view for RCA.

Since patient was sick and in Killip III/IV with baseline deranged renal function, CT Aortogram was omitted and intravascular imaging with HD-IVUS (Figure 5a,b) was done to evaluate this left main stenosis that revealed virtually no atherosclerosis in any segment, but just dynamic severe compression of the left main coronary artery with a slit like ostia of the left main coronary artery.



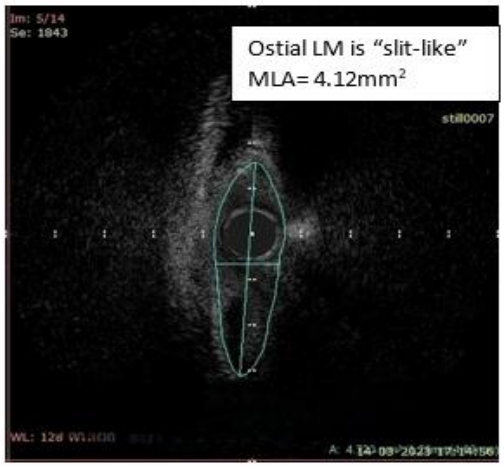




**Figure 5a)** IVUS of distal left main showing virtually no atherosclerosis and MLA of 17.32 $mm^2$.
**Figure 5b)** IVUS at ostial left main showing extrinsic compression with "slit-like" ostia and MLA of 4.12 $mm^2$.

PCI with Zotarolimus eluting stent 5 X 22 mm was deployed in ostial left main with post dilatation with 6 x 8 mm NC balloon with good angiographic result (Figure 6) and achieved MLA of 20.61 $mm^2$ and few struts hanging in aorto-ostial region with MLA of 25.72 $mm^2$ (Figure 7a and b). The Resolute Onyx stent was chosen that has the highest radial strength (5.75 $N/mm^2$) to withstand the extrinsic compression force of approximately 26 WU of pulmonary vascular resistance (calculated non-invasively by echocardiographic doppler estimation in view of critical illness).

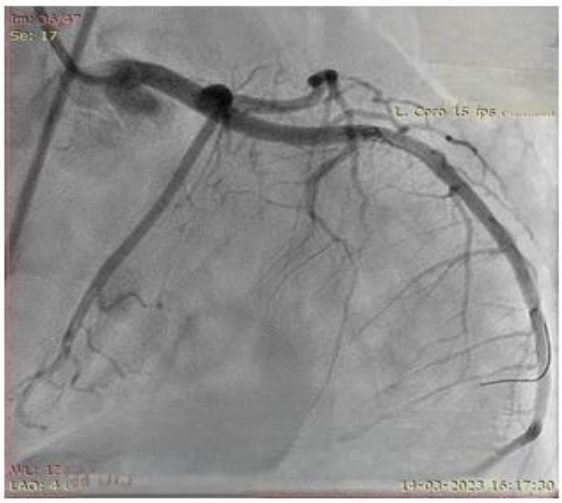


**Figure 6** – Post angioplasty angiogram revealing no residual stenosis in left main in all views.

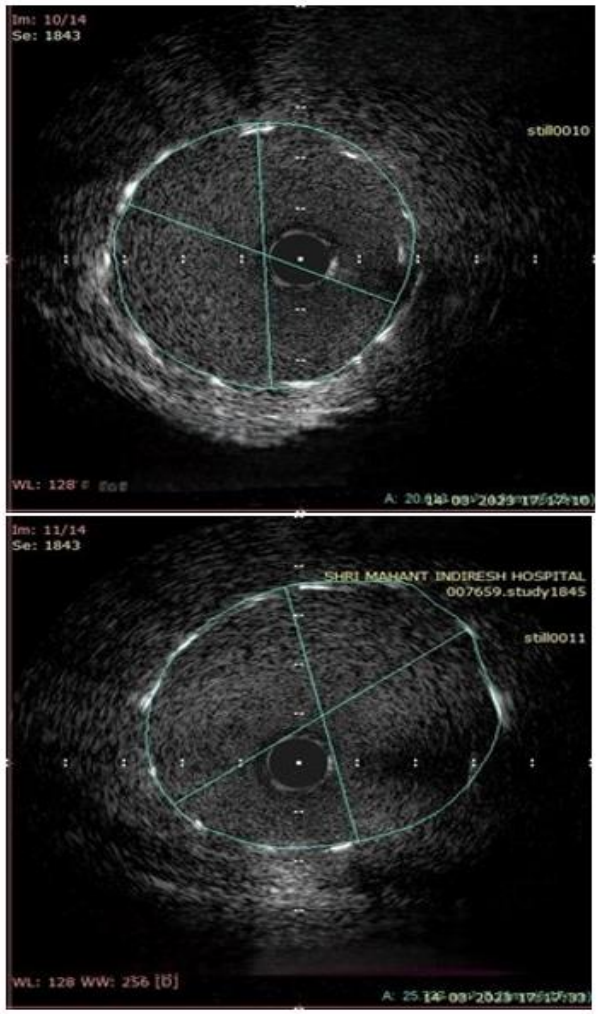



**Figure 7 –** Post angioplasty IVUS revealing good stent expansion in left main (Figure 7 a) with few struts protruding at aorto-ostial level (Figure 7 b)

## Discussion

Coronary artery disease in pulmonary hypertension patients presenting with Coronary artery disease in pulmonary hypertension patients presenting with angina is often under suspected and usually diagnosed as a coincidence finding. [1] It is now recognized as a serious mechanical complication of PH due to extrinsic compression of the LMCA due to dilation of the PA first described in 1957 by Corday et al [2] with a prevalence rate was around 40% [3, especially if accompanied by angina symptoms.

The relationship between LMCA and dilated PA has been described as normal, contiguity, dislocation, and significant stenosis [4]. The compression risk is related to some strong predictors measured by contrast enhanced CT [4,5] such as PA diameter > 40 mm, ratio of the MPA and aortic root ≥ 1.5, and take-off angle < 45° (formed between the longitudinal line of the LMCA and orthogonal line of the aortic valve) [5,6]. In our case, similar inferences may be derived from the course of left main artery seen in modified short axis view on transthoracic echocardiogram and the from the "slit" on HD-IVUS at the left main ostia.

Coronary artery bypass graft (CABG) as a treatment option of this syndrome involves a higher risk in general anaesthesia and cardiopulmonary bypass (CPB) in sick patients with deranged renal and liver function, so Percutaneous Coronary Intervention (PCI) with a drug eluting stent (DES) of good radial strength may be the preferred strategy in such selected individuals.[7-9]

## Conclusion

The prognosis of Eisenmenger syndrome is poor and though the classical presentation is right heart failure culminating in death, the course in such patients may be punctuated by an acute coronary event precipitated by extrinsic left main compression or arrhythmic events. Early diagnosis of "left main compression syndrome" can be difficult because of non-specific symptoms. However, a high index of suspicion, a careful look at the angiogram in orthogonal views to diagnose this unidimensional compression and utilization of HD-IVUS in sick individuals where CT scan is not feasible may help us recognize this entity. This classical "slit-like" ostia of left main coronary artery on HD-IVUS is an important finding and is potentially correctable with the use of DES wherein we achieved an excellent minimal luminal area (MLA) post intervention, with no evidence of stent deformation. Invasive modalities are particularly useful in high-risk individuals presenting with ACS who need urgent treatment besides mere diagnosis.

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