A YOUNG BOY PRESENTED WITH SEVERE AORTIC STENOSIS ALONG WITH COARCTATION OF AORTA, SEVERE LV DYSFUNCTION AND CML FIRST-IN-HUMAN EXPERIENCE WITH REAL-TIME 4D HOLOGRAPHIC THERAPY GUIDANCE AS AN INTRAPROCEDURAL TOOL FOR STRUCTURAL HEART INTERVENTIONS

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History and Examination:

18 years old male presented to our OPD after visiting multiple institution with complaints of Headache, Shortness of breath for the past few months. Headache was moderate intensity remains there for most of the daytime with no aggravating and reliving factors. Currently he developed shortness of breath with walking along with chest pain. He falls in NYHA class II. He had multiple episodes of syncope for last 6 months before coming to hospital. No other constitutional symptoms. He is vaccinated developmentally normal and student of grade 11 with no significant family history.

A young markedly pale-looking thin lean boy with no facial dysmorphism. His Heart rate was 110/min, RR 28/min, Blood pressure right arm 145/67, left arm 140/60, right leg 95/57, and left leg 100/54. Having low volume pulses and radio-femoral delay. On precordial examination apex beat in 6th ICS lateral to mid clavicular line. S1+S2+ grade 4/6 ESM best audible at right upper sternal border. In addition to the above sign and symptom there was 4 cm palpable firm liver and spleen.

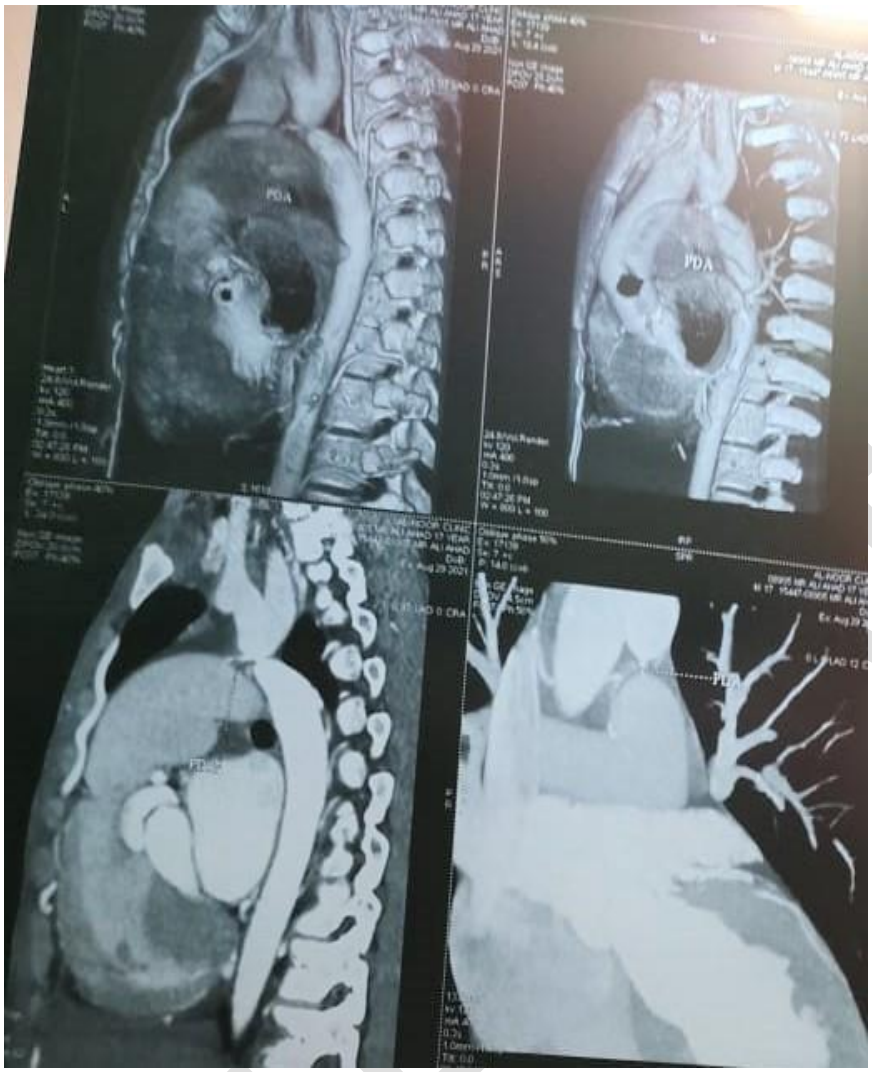
Imaging:

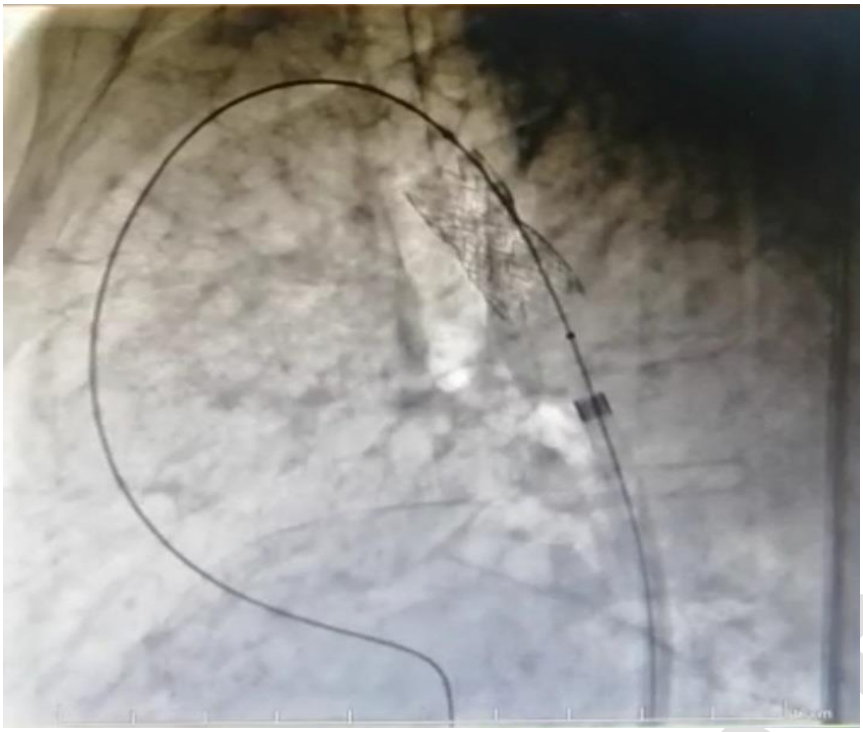
1. X RAY chest was done which showed marked cardiomegaly.
2. ECG showed sinus rhythm, Left axis with marked LVH.
3. Echocardiography was performed which revealed Severe valvular Aortic stenosis with peak gradient 107 mmHg and annulus of 22mm with no regurgitation. Severe juxta ductal discrete Coarctation of aorta, Small PDA, Severe LV dysfunction with an ejection fraction of 34%

Blood report and Bone marrow

1- CBC : Hb =12g%,TLC =42 (10 3 /ul) ,PLT = 502 (103 /uL)

2- Bone marrow finding were Chronic Myeloid Leukemia.





Indication of intervention:

Although it was a high-risk case as EF was 34% patient was discussed with multidisciplinary team and decided to perform simultaneously aortic balloon valvuloplasty and Coarctation stenting. Patient was admitted and baseline blood investigation were performed. His CBC revealed TLC of 57,000. HB 10 and platelets of 610,000. Pt was referred back to hematologist for this derangement. The Imatinib (Glivec) 100 mg twice daily recommended from hematologist. This ailment further added risk to the above-mentioned interventions due to raised TLC and abnormal platelets count.

Pt was having severe Aortic stenosis, Severe coarctation of aorta with symptoms. According to AHA guidelines there was indication of intervention only

Interventions:

In collaboration with multidisciplinary team colleagues, intervention was planned. On Fluoroscopy Aortic valve annulus was measured 22mm. An 18 x40 mm balloon was selected and full inflation was done. Post procedure angiogram and pressures showed mild residual stenosis with the good opening of valve and Trivial AR.

Coarctation measured 4mm at the narrowest point proximal segment was 15mm. Bentley covered stent 38 x 16mm was taken and positioned across the coarctation segment after multiple check injections balloon inflated to deploy the stent. Post-procedure angiogram showed well-dilated coarctation segment with PDA closure and good flow in subclavian artery with almost no pressure gradient between ascending and descending aorta.

Post procedure echocardiography was performed after 24 hrs which showed satisfactory procedure results. Patient was discharged after 48 hrs of procedure. Came for follow up and performing well.

Learning points / challenges :

It was a difficult case and challenging due to CML and high TLC count and Platelet count. The use of chemotherapy in severe LV dysfunction further worsens dysfunction and high platelet count increases risk of thrombo-embolic phenomenon.

Important learning point is that such patients can be salvaged by this intervention only because the surgical outcome has a very high mortality.