

A challenging case of coronary intervention in Ehlers-Danlos Syndrome, dextrocardia and Situs Inversus

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Background:

- Ehlers-Danlos syndrome (EDS) has a low prevalence, and is even rarer in conjunction with dextrocardia and situs inversus
- Collagen tissue disorders (CTD) result in higher rates of vascular complications.
- We present a case of coronary atherosclerosis with the above comorbidities that resulted in successful outcomes.

Case

- A 70-year-old female with a history of Ehlers-Danlos syndrome, situs inversus, dextrocardia, and Kartagener's syndrome, referred for left heart catheterization (LHC) for exertional dyspnea with a positive myocardial perfusion stress test (reversible perfusion defects in the left anterior descending artery [LAD] territory).
- Her electrocardiogram (EKG) revealed normal sinus rhythm (Figure 1). Transthoracic echocardiogram revealed situs inversus with dextrocardia, preserved left and right ventricular ejection and normal valves (figure 2).
- Due to increased risk of vascular complications with CTD and dextrocardia, standard catheters but transfemoral approach with reverse torquing technique and setting of mirrored angles to image the coronary arteries were used.
- LHC demonstrated a left-dominant circulation with a diffuse 80% proximal LAD lesion at the bifurcation with a large diagonal branch (Figure 3). Percutaneous coronary intervention was successful with intravascular ultrasound (IVUS) imaging guidance to ensure appropriate stent size, good stent apposition while avoiding over-aggressive post-stent dilation, as well as absence of stent edge dissections.
- Adjunctive heparin peri-procedurally followed by dual anti-platelet therapy was prescribed without bleeding complications.



Figure 1. Reversed-lead EKG; normal.

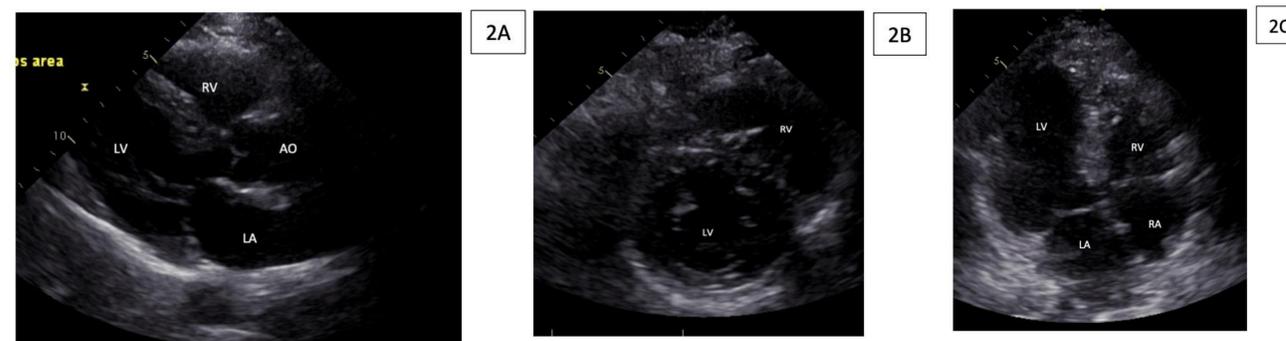


Figure 2 Transthoracic echocardiogram revealing situs inversus with dextrocardia (LV= left ventricle, LA = left atrium, RA = right atrium, RV = right ventricle, AO = aorta).

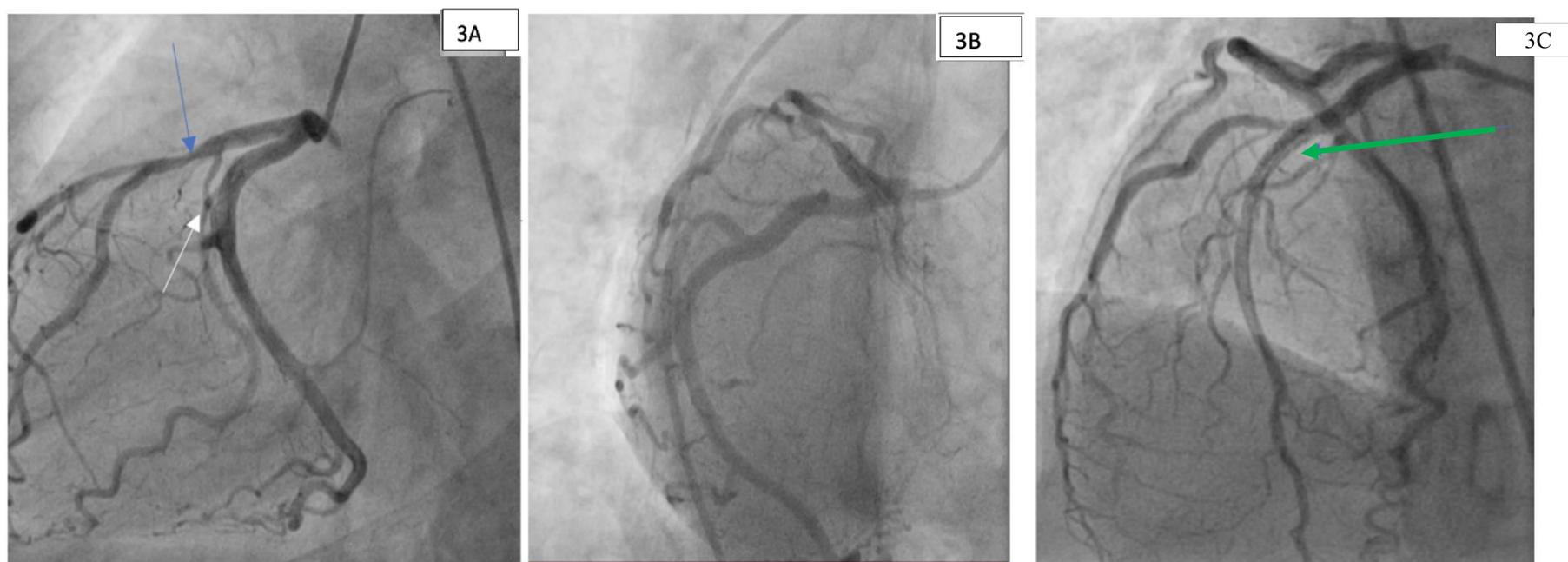


Figure 3. Coronary catheterization demonstrating left-dominant coronary circulation with a long, diffuse 80% stenosis of the left anterior descending (LAD) artery at the bifurcation with a large Diagonal branch (Blue Arrow) and diagonal branch (White Arrow) in figure 3A and 3B. Cardiac catheterization post-revascularization with drug eluting stent at LAD (green arrow, 3C).

Conclusion:

In patients with CTD pre-disposing to vascular complications, as well as anatomical aberrations, trans-femoral approach with anti-torquing maneuvers and mirrored image angles as well as IVUS-guided intervention should be employed to optimize results and minimize iatrogenic complications.