Pump Fiction: A Tale of Acute Promyelocytic Leukemia Mimicking Pump Thrombosis

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Introduction:

- An abrupt increase in the incidence of pump thrombosis (PT) has been noted during the last years.
- Elevated lactate dehydrogenase (LDH) level is a clinical biomarker (indicating hemolysis) for PT, however five isoenzymes exist that represent different tissue distribution.
- We present a case of chronic elevated LDH secondary to acute promyelocytic leukemia (APL).

Case Report:

A 36-year-old obese male with non-ischemic cardiomyopathy underwent HeartMate II implantation at an outside hospital as destination therapy. Within a year his LDH increased and he underwent Pump exchange revealing a laminated thrombus in the outflow conduit.

Two years later, his LDH gradually increased and he was kept on 325mg of aspirin, 75mg of dipyridamole, and warfarin (INR goal 3). He transferred his care to us and during his first visit, he was thrombocytopenic with total LDH of 1100 units/L but no power spikes or high flows were noted. Echocardiogram revealed a dilated left ventricle (LV) and a speed ramp study failed to close his aortic valve (Figure 1).

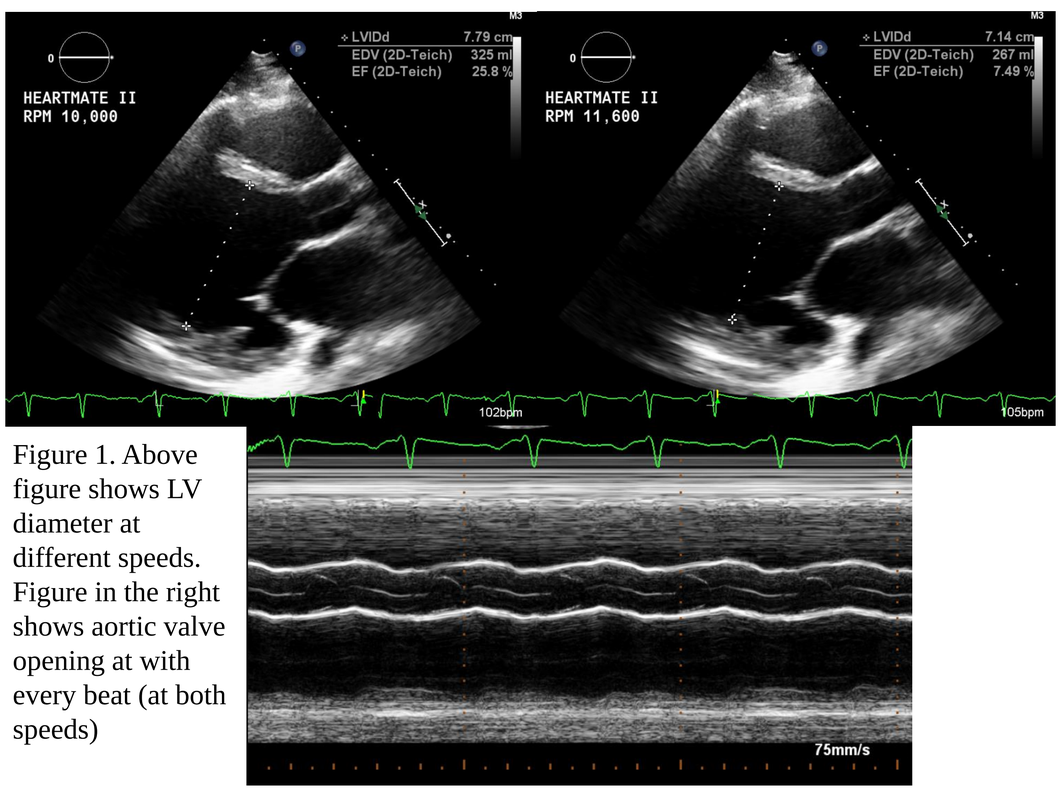


Figure 1. Above figure shows LV diameter at different speeds. Figure in the right shows aortic valve opening at with every beat (at both speeds)

He was admitted for further work-up of suspected recurrent of pump thrombosis. CT angiogram showed no evidence of thrombus in the inflow or outflow cannulas, but a distal outflow anastomosis kink was seen with patent lumen and smallest diameter of 9mm (Figure 2, white arrow).

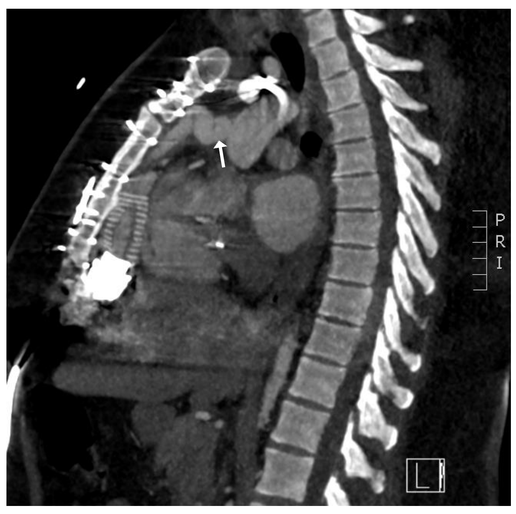
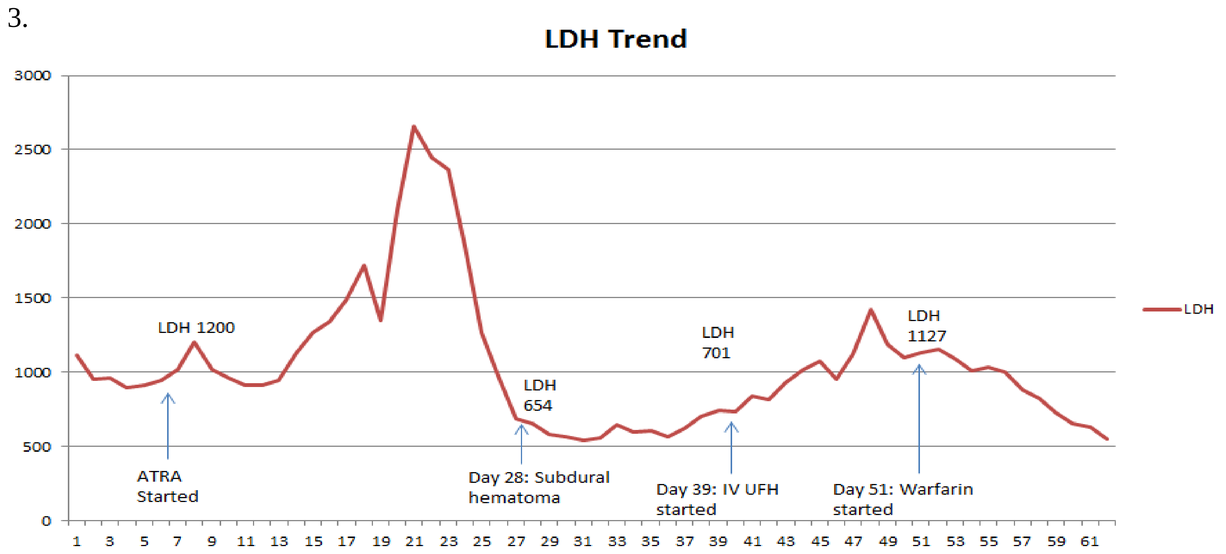


Figure 2.

LDH isoenzymes showed predominantly LDH-2 (reticuloendothelial system) and a peripheral smear revealed circulating blasts. Bone marrow biopsy with FISH confirmed the diagnosis of APL and he was started on all-trans retinoic acid and arsenic acid. Chemotherapy was complicated by disseminated intravascular coagulation, differentiation syndrome and subdural hematoma. Anticoagulation was held but then restarted following an LDH rise. After a month of inpatient treatment, his warfarin was resumed and LDH continued to trend down (Figure 3).

Figure 3.



Discussion:

Left ventricular assist devices have become an important strategy for the treatment of end-stage heart failure. With increased duration of support, especially in destination therapy, a new spectrum of complications has arisen. The most common are disorders of coagulation, including both bleeding to thrombosis.

PT implies the formation (or deposition) of a clot within the flow path of the device, including the inflow cannula, outflow graft, and rotor housing. As our case illustrates, a broad differential diagnosis should be considered with an atypical presentation.

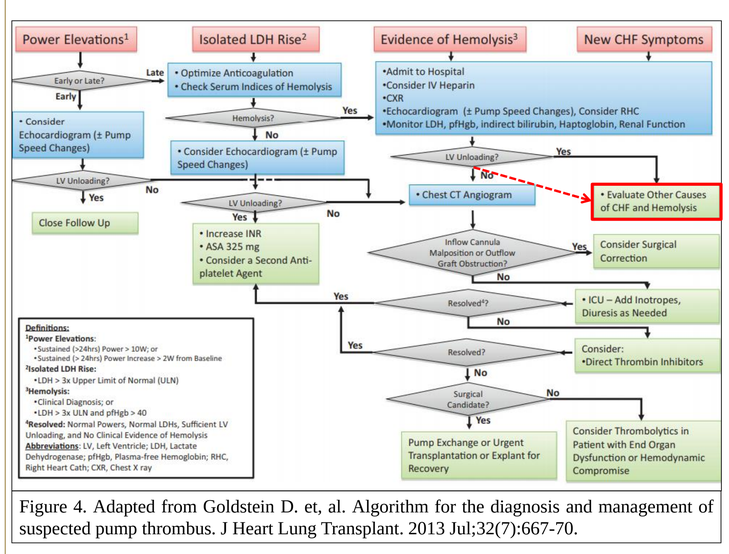


Figure 4. Adapted from Goldstein D. et, al. Algorithm for the diagnosis and management of suspected pump thrombus. J Heart Lung Transplant. 2013 Jul;32(7):667-70.

A current algorithm to diagnose and manage PT, recommends evaluating both pump parameters (power and flow) and markers of red blood cells destruction. Further imaging is useful to identify whether the LV is unloaded (ramp study) and abnormalities in the circuit (CT angiogram).

In this case, the inability to fully unload the LV and maintain aortic valve closure was likely due to the outflow kink and not thrombosis. Measuring LDH isoenzymes were crucial to further understand the pathophysiologic process. This test was extremely valuable, since inappropriate diagnosis could have led to pump exchange or use of thrombolytic, increasing the risks of adverse outcomes in APL. Furthermore, prompt diagnosis of his hematologic condition was facilitated and appropriate therapy was initiated.

Although APL is associated with high rate of early mortality, often due to hemorrhage from disseminated intravascular coagulopathy, treatment with all-trans retinoic acid promotes the terminal differentiation of malignant pro-myelocytes to mature neutrophils and improves long term survival.

Conclusion:

- PT remains a therapeutic prevalent diagnostic challenge, especially in destination therapy patients.
- LDH is a widely available marker for PT, but specific isoenzyme testing may be useful to detect if it is primarily from red blood cell lysis (LDH-1) or other organs.
- Multidisciplinary management with cardiothoracic surgeons, advanced heart failure cardiologist, hematologists and other specialists is crucial for favorable outcomes.

Disclosures:

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- Remaining authors: No disclosures.