



# A Case Study on a Differential Diagnosis of a Restrictive Cardiomyopathy

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## Background

- Restrictive cardiomyopathy (RCM) is a type of cardiomyopathy with a predominance of severe diastolic dysfunction, normal or mildly increased ventricular wall thickness, and either normal or mildly reduced ejection fraction.
- RCM is typically diagnosed with less invasive Doppler and Echocardiogram-2-dimensional procedures. Thus, in this case, the combined left-right heart catheterization allowed for a definitive answer in the face of a rapidly debilitating patient. While echocardiograms and Dopplers could have reached the same conclusion, they could have also prolonged the diagnostic timeline, delaying proper treatment delivery.

## Case Presentation

- A 52-year-old female with extensive cardiac history initially presents to the emergency department with complaints of left-sided chest pain, worsening shortness of breath, and nausea. Her prior medical history consists of rheumatic heart disease with multiple mitral valve repairs, heart failure with preserved ejection fraction (HFpEF), atrial fibrillation on rivaroxaban medication, and chronic obstructive pulmonary disease (COPD) on 2L of oxygen.
- An initial cardiology consult performed a right heart catheterization which prompted a transfer to the heart failure (HF) unit. In the HF unit, the patient was placed on a urinary catheter for diuresis measurement with guideline-directed medical therapy (GDMT) but went on to develop shock and acute kidney injury (AKI). Given this, the patient was transferred to the critical care unit (CCU) where the etiology was deemed to be hypovolemia from over diuresis; improvement in volume resuscitation hypotension resulted in transfer out of the CCU. After three days, the rapid response team was called for hypoxia and imaging revealed diffuse bilateral infiltrates, causing a transfer into the ICU. While there, the patient had a right heart catheterization and echocardiogram that showed severe pulmonary hypertension and severely elevated filling pressures.
- After two days in the unit, the patient was intubated and shortly after started on continuous renal replacement therapy (CRRT) for AKI. The patient had a combination left-right heart catheterization (L/RHC) completed, confirming restrictive cardiomyopathy. Despite aggressive attempts to reduce volume, the patient exhibited persistent bilateral ground-glass opacity. For three days, the patient continued to decline and ultimately became septic and went into shock. Given her pre-existing and acute comorbidities, the family opted for comfort measures and the patient expired shortly after.

## Hospital Course

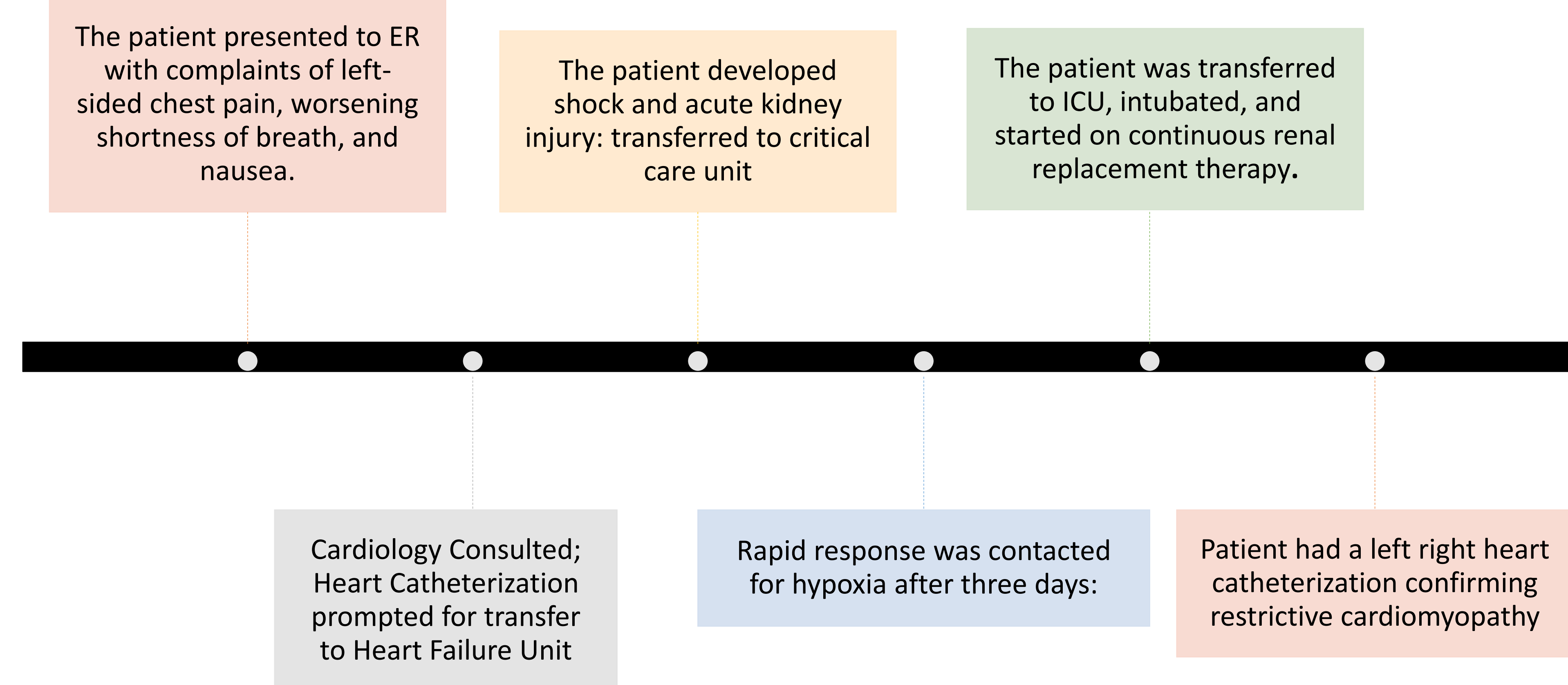


Figure 1: Hospital Course Timeline.

## Diagnostics and Imaging

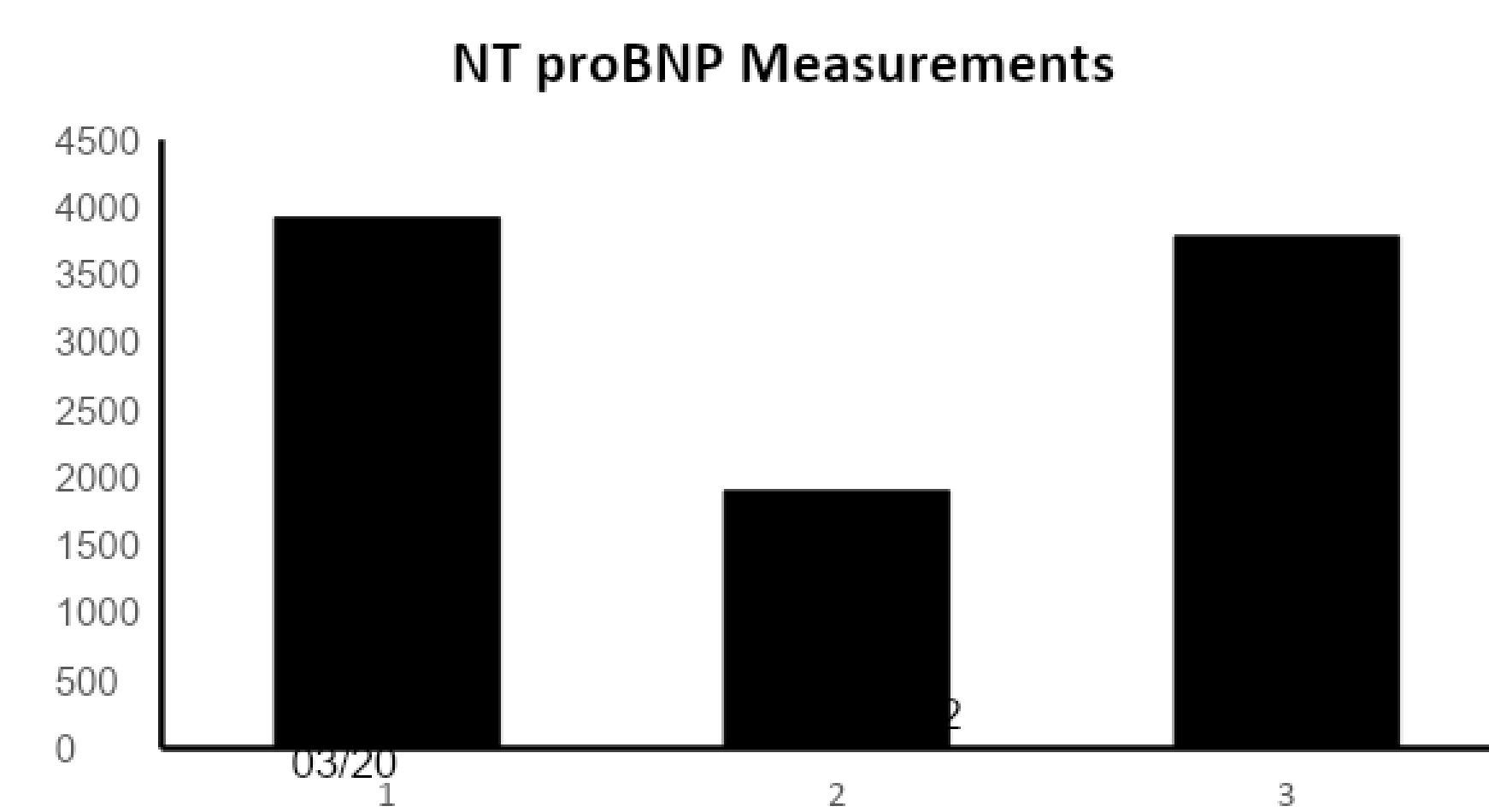


Figure 2. Dates Feb of 2023, March 12, 2023, March 20, 2023, NT proBNP Measurements

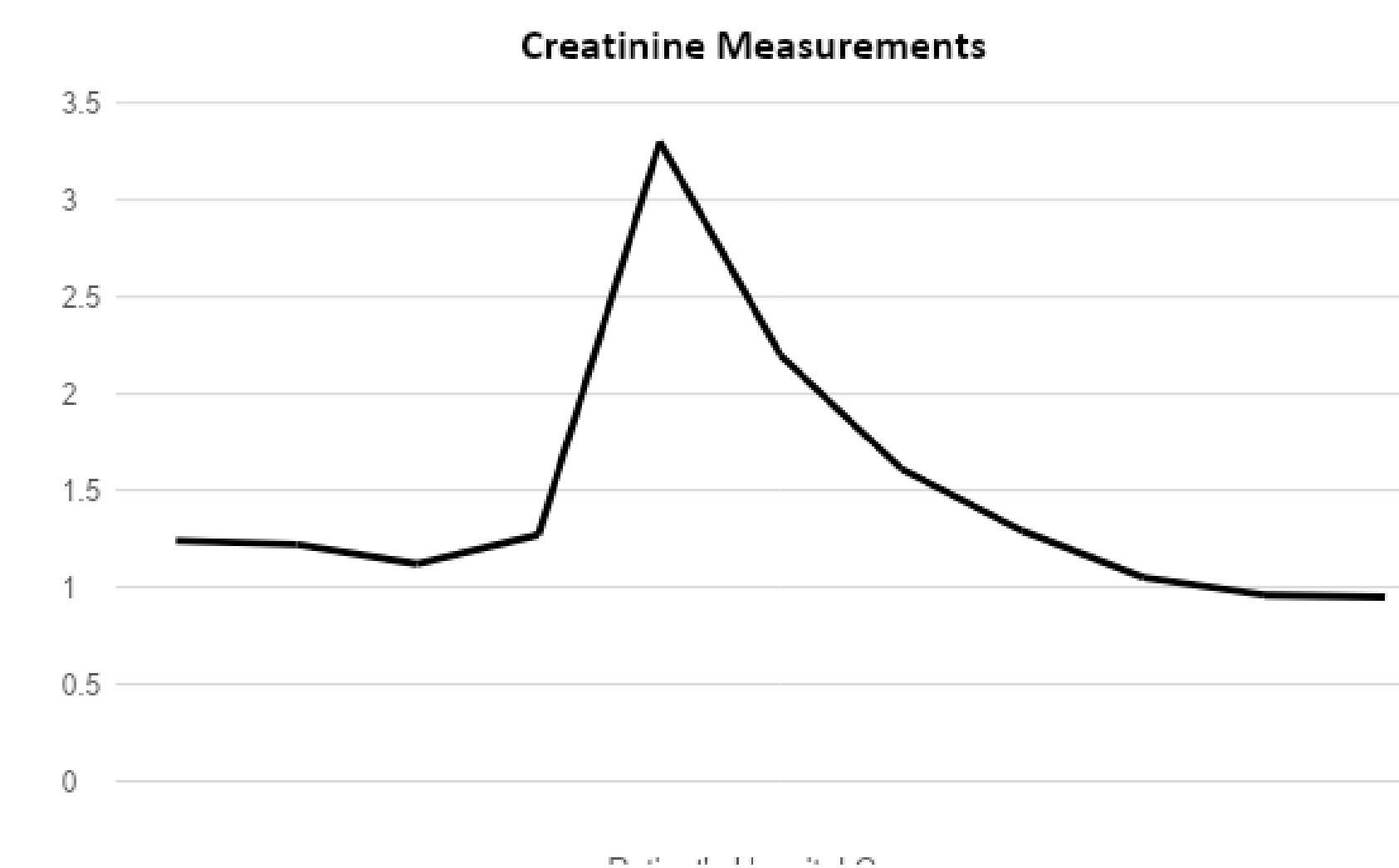


Figure 3. The patient's creatine measures throughout the hospital course stay

### L/R Heart Catheterization Conclusions

Pressures (in mmHg)	Patient	Normal
Right Atrium (RA)	27	3-7
Right Ventricle (RV)	75/30	20-30/3-7
Pulmonary Artery (PA)	68/42	20-30/8-12
Left Ventricle End Diastolic Pressure (LVEDP)	30	3-8
Mean Wedge	33	12 mmHg
PA saturation	56%	70-75%
Arterial saturation	93%	95-100%
Fick Cardiac Index	3	2.5-4
Simultaneous LV/RV pressure tracing during respiration		

Table 1. The right atrium, right and left ventricle, Simultaneous LV/RV pressure tracing during respiration

## Discussion

- In this case, the patient was diagnosed with RCM via a combined left-right heart catheterization. RCM is typically diagnosed with less invasive Doppler and Echocardiogram-2-dimensional procedures. However, the more invasive heart catheterization offers direct measurements of hemodynamic data. These direct findings allow a clearer picture of the issues present, thus distinguishing between restrictive cardiomyopathy vs constrictive pericarditis, the latter demonstrating differences in pressures similar to RCM. The heart catheter data in the presence of RCM is often increased left/right filling pressure with equalization of the diastolic filling pressures of the heart chambers and ventricular concordance. The findings from the patient's L/RHC align with the presentation of restrictive cardiomyopathy, specifically the elevated RA and LVEDP with simultaneous ventricle pressures. Therefore, the heart catheterization procedure is an effective and reliable option when determining cardiovascular disease/disorders.
- An L/RHC procedure was done on the patient as well as a right catheterization and echocardiogram that did not reveal the restrictive cardiomyopathy. The right heart catheterization combined with the L/RHC revealed severely elevated filling pressures with pulmonary hypertension. A day before the L/RHC, the patient had an echocardiogram done that showed normal ejection fraction and elevated filling pressures. The observed ejection fractions align with the expectations in an RCM presentation: normal levels. However, the echocardiogram did not appear to show biatrial enlargement or non-dilated ventricles, features expected in RCM cases. The findings from these two diagnostic tools kept the etiology of the patient's heart failure ambiguous.

## Conclusions

The combined left-right heart catheterization allowed for a definitive answer in the face of a rapidly debilitating patient. While echocardiograms and Dopplers could have reached the same conclusion, it could have also prolonged the diagnostic timeline, delaying proper treatment delivery.

## References

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