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# Pulmonary hypertension: Tortuous route to diagnosis

Giovanna Riolo, Bader Al Ghamdi & Christine L. D'Arsigny

Division of Respiratory and Critical Care Medicine, Queen's University, Kingston, Ontario, Canada

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**Correspondence**
Giovanna Riolo, Queen's University, Kingston, Ontario, Canada, Division of Respiratory and Critical Care Medicine, 102 Stuart St. Kingston ONT K7L 2V6.
E-mail: 7gr8@queensu.ca

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## Abstract

A 47 year-old woman, on hemodialysis via an arteriovenous (AV) fistula, was assessed for severe dyspnea and presyncope secondary to pulmonary hypertension. Right heart catheterization confirmed a mean pulmonary arterial pressure of 85 mm Hg. She had a normal wedge pressure. Investigations revealed that the total high cardiac output AV fistula, 8.3 L/min, resulted in pulmonary arteropathy and increased pulmonary vascular resistance at 674 dyne.sec.cm$^{-5}$. The AV fistula was banded and Sildenafil was prescribed, which resulted in improvement of pulmonary hypertension within one week.

## Introduction

Approximately 40–60% of patients on hemodialysis (HD) develop pulmonary hypertension (PH), usually due to left sided heart failure [1, 2]. Our patient manifested a rare cause of PH, due to her high flow arteriovenous (AV) dialysis fistula.

## Case Report

A 47 year old female was admitted to hospital with severe progressive dyspnea for over one year, associated with pre-syncope and intermittent chest discomfort on minimal exertion. She was a WHO class 4 on presentation to hospital.

Her past history was relevant for having a renal transplant at age 25. However, 6 years prior to this admission, she developed gastric lymphoma, necessitating stopping her immunosuppressive therapy. Ultimately, her transplanted kidney failed and she has an AV fistula inserted for dialysis.

On examination, her oxygen saturation was 98% on room air. Her respiratory rate was 14 breaths per minute. She had a loud P2 but no right ventricular (RV) heave or extra heart sounds. Her JVP was not distended, and there was no ascites, abdominal organomegaly or peripheral edema. She did have a large tortuous left brachial AV fistula (Figure 1).

Given the suspicion for PH, a transthoracic echocardiogram (TTE) was completed showing a severely elevated RV systolic pressure at 101 mmHg. There was no intracardiac shunt on bubble study and the left ventricular (LV) ejection fraction was 62%.

A right heart catheterization done on the morning of dialysis confirmed severe PH with a mean pulmonary arterial pressure (mPAP) of 85 mmHg (PH defined as ≥25 mmHg at rest). She had an elevated pulmonary vascular resistance (PVR) at 674 dyne.sec.cm$^{-5}$. (PH: >240 dyne.sec.cm$^{-5}$). Her pulmonary capillary wedge pressure (PCWP) was 14 mmHg (non-cardiac PH defined as <15 mmHg). Her cardiac output (CO) was elevated at 8.3 L/min (N: 4–7 L/min). Her LV diastolic pressure was 8 mmHg.

Work up for PH was normal and included a CT chest, pulmonary function tests (normal except for low DLCO), V/Q scan, HIV, connective tissue disease (ANA, ENA, RF), liver profile, TSH, polysomnogram, abdominal ultrasound with Doppler flow and TTE with bubble study. She was deemed a WHO class 4. As she could not walk a few meters without assistance and had presyncope, we could not perform a baseline 6 MWD. Her brain naturetic peptide (NTproBNP) was elevated at 35,620 ng/L (normal: less than 125 ng/L).

Our patient had severe pulmonary hypertension with normal LV function, along with an elevated CO. We measured flow through the fistula by sodium-dilution technique, and calculated the flow to be a minimum of 2 L/min. We proposed that the chronic high flow through her AV fistula likely resulted in pulmonary artery vascular remodeling and the development of PH.

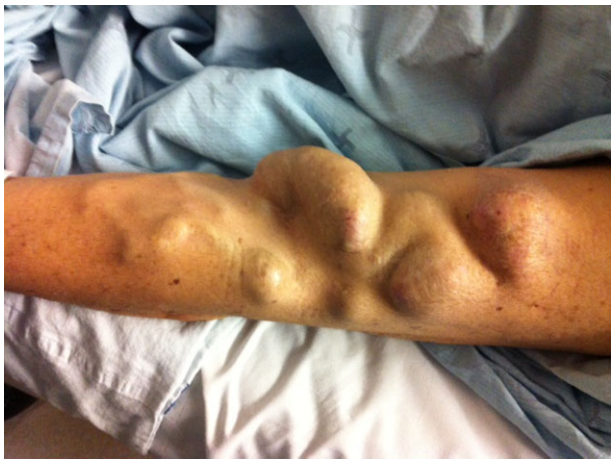

**Figure 1.** She had a large tortuous left brachial AV fistula, resulting in high cardiac output.

The high CO and increased PVR were managed simultaneously by banding the fistula, decreasing its flow to 959 ml/min and prescribing oral Sildenafil 20 mg, three times per day. It was intended to start her on intravenous Epoprostenol but she improved within one week from a WHO class 4 to a WHO class 2.

Results of a repeat right heart catheterization 4 months later correlated with her improved exercise ability. Her mPAP decreased to 47 mmHg, her PVR decreased to 422 dyne.sec.cm$^{-5}$, her PCWP decreased to 10 mmHg and her CO was lower at 7 L/min. Her fistula was then compressed with a sphygmomanometer (140 mmHg for 15 minutes). The total CO decreased to 5.3 L/min but her mPAP and PVR remained unchanged, confirming a fixed vasculopathy. Follow up 6 MWD was 540 meters (90% predicted) and her NTproBNP decreased to 10 305 ng/L. Follow up TTE showed only mild RV dilatation, with mild tricuspid regurgitation and a reduced RV systolic pressure at 69 mmHg.

## Discussion

High CO failure in HD patients usually results in LV dilation and/or hypertrophy, myocardial damage and an ejection fraction decline with a resultant increase in PCWP/ left ventricular end diastolic pressure (LVEDP) and mPAP. These patients often present in clinical left heart failure [1, 2]. BNP release in these patients is due to myocardial ischemia as a consequence of an imbalance between increased left sided cardiac oxygen demand and decreased oxygen supply [2].

Our patient, however, did not have any evidence of LV failure. Her rise in mPAP and PVR was due to either Group 1 or 5 PH. We favor the etiology being secondary to her high output AV fistula (group 5) as she acutely improved with banding of her fistula with a drop in her pulmonary pressures and BNP levels and an increase in 6MWD and functional status.

The proposed mechanism for an elevated PVR in dialysis patients is not completely understood. Theories include vascular remodeling and smooth muscle proliferation [3], an increased production of endothelin-1 [4–5], and/or extraosseous pulmonary calcification, which is a proposed but unproven theory for PH in HD patients [5].

This case is unique due to the presentation of severe PH, which was most likely triggered by the chronic exposure to a high CO from her tortuous AV fistula. Unlike most dialysis patients, she had normal LV function. Her functional status and PH improved with banding of her fistula and the addition of Sildenafil but she was found to have fixed elevation in her PVR, most likely from residual pulmonary artery remodeling. Her high BNP levels were likely due to an inability of the RV to cope with the high pulmonary pressures, with resultant RV failure and ischemia. As her PH has improved and her RV has decreased in size, her BNP levels have dropped.

### Disclosure Statements

No conflict of interest declared.

Appropriate written informed consent was obtained for publication of this case report and accompanying images.

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