Gen.Medicine

INDEX COPERNICUS INTERNATIONAL

ICV value for 2023 = 74.57

Case Report

International Journal of Medical Case Report

# Presentation of Pulmonary Hypertension Treated with Sotatercept

**Mohamed Zaki[1], Mutia Al Jabi[2], Ahmed Khaled Aboeldahab[3], Hani Sabbour[4]**

[1,2]College of Medicine, RAK Medical and Health Sciences University, Ras Al Khaimah, UAE.

[3] University of Sharjah, United Arab Emirates.

[4]Department of Cardiology, Mediclinic Hospital, United Arab Emirates.

INTERNATIONAL JOURNAL OF MEDICAL CASE REPORTS ISSN 2455-0574

## Abstract

Pulmonary hypertension (PH) is a complex, progressive disease often leading to acute kidney injury (AKI), right heart failure (RHF), and multi-organ dysfunction. This case involves a 64-year-old woman with severe pulmonary arterial hypertension who required intensive care after repeated episodes of AKI and RHF. The case explores the interconnected pathophysiology of reduced renal perfusion, ascites-induced intra-abdominal hypertension, and venous congestion. Management included continuous renal replacement therapy (CRRT), non-invasive ventilation (NIV) to avoid hemodynamic compromise, and later conventional dialysis for volume overload and worsening renal function. Oxygen therapy was carefully titrated due to significant right-to-left shunting through a patent foramen ovale (PFO). After conventional treatments failed, she was started on Sotatercept, a novel therapy targeting pulmonary vascular remodeling.

**Keywords:- Acute kidney injury (AKI), Pulmonary Hypertension (PH), Right Heart Failure (RHF), Sotatercept**



## INTRODUCTION

Clinical improvements in the patient, such as improved oxygenation and renal recovery, point to the potential advantages of Sotatercept in patients that are resistant. The significance of interdisciplinary, pathophysiology-driven decision-making in critically ill patients with PH and RHF is emphasized in this article

Access this Journal Online

Quick Response Code

Website: www.ijomcr.net

Email:ijomcr@gmail.com

**Corresponding Author: Ahmed Khaled Aboeldahab[3]**

University of Sharjah, United Arab Emirates.

Email: aboeldahaba32@yahoo.com

# CASE REPORT

A 64-year-old woman with pulmonary hypertension (diagnosed January 2022) presented with worsening dyspnea and low oxygen saturation ($SpO_2$ 76%). She had a history of acute-on-chronic right heart failure (February 2023), sarcoidosis with generalized lymphadenopathy, and newly diagnosed hyperlipidemia. On admission, her vitals showed elevated BP (153/71 mmHg) and MAP of 98 mmHg. Labs revealed elevated troponin T, BNP, urea, and mild renal dysfunction (creatinine 1.1 mg/dL, eGFR 54 mL/min). In the ICU, she was started on milrinone, Lasix, tazocin, and lactulose. BiPAP was initiated, and a chest X-ray confirmed a right-sided pleural effusion (figure 1).

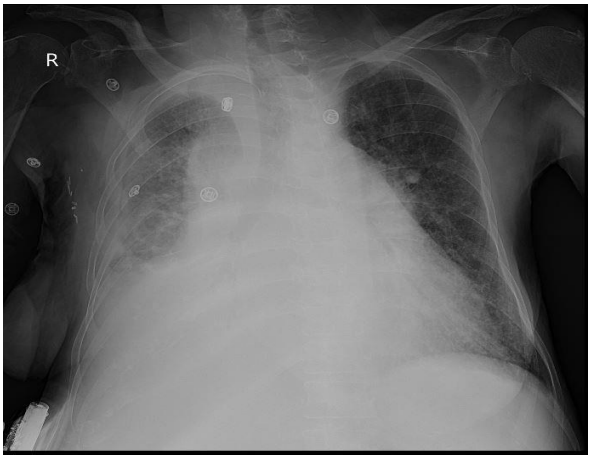


**Figure – 1 : Chest radiograph showing left sided lung opacification.**

Flexible bronchoscopy was performed, revealing a large foreign body (38 mm x 10 mm) lodged in the left main bronchus. Given its size and location, the patient had to be decannulated, and the foreign body was successfully retrieved using a controlled radial expansion (CRE) balloon catheter and artery forceps.

Over the following days, her condition worsened, with reduced urine output and further renal impairment. The milrinone dose was adjusted, and norepinephrine was added to maintain a MAP greater than 65 mmHg. A pulmonology consultation diagnosed her with chronic respiratory failure (Type 2) with hypoxemia and hypercapnia. The pleural effusion was attributed to cardiac and renal impairment. Management included nebulization, consideration for long-term oxygen therapy (LTOT), and long-term BiPAP.

As her renal function declined further, with creatinine rising to 387 µmol/L and urea exceeding 30 mmol/L, continuous renal replacement therapy (CRRT) was initiated due to severe acidosis and fluid overload. Despite minimal improvement in renal function, CRRT was discontinued after a few days due to very low urine output. Conventional hemodialysis (HD) was started and continued for several days, with ultrafiltration and drainage of pleural effusion. Over time, improvements were noted, including stable oxygenation, decreasing BNP levels, and better urine output. Renal recovery was observed, with creatinine decreasing to 60 µmol/L. The patient was gradually weaned off the medications, and BiPAP was replaced with nasal cannula oxygen. The patient's condition stabilized, and she was able to tolerate regular dialysis. A right heart catheterization confirmed severe pulmonary arterial hypertension (mPAP 48 mmHg, RVSP 80+15 mmHg) and the presence of a right-to-left shunt (figure 2).

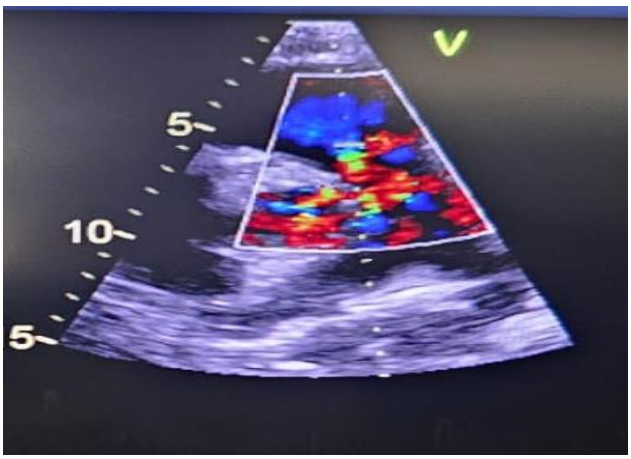


**Figure 2:- Echo showing right-to-left shunt**

The patient was later re-admitted due to severe acute-on-chronic decompensation. On re-admission, she presented with low blood pressure, a low MAP, and worsening hypoxia. Additional vasopressors and dialysis were required to manage fluid overload and renal failure. Repeat right heart catheterization showed severe pulmonary hypertension with elevated pulmonary vascular resistance, reduced cardiac index, and confirmed right-to-left shunting.

At the time of discharge, the patient was stable on triple therapy for pulmonary arterial hypertension, including Treprostinil, selexipag, and an endothelin

receptor antagonist. She was discharged with a plan to continue outpatient therapy and potential initiation of Sotatercept for advanced pulmonary hypertension.

Subsequently the patient had a fever, cough, dyspnea, and rectal bleeding when she was recently admitted to the intensive care unit. She was treated with norepinephrine, BiPAP, and Sotatercept (45 mg subcutaneously every three weeks for 90 days) due to her severe hemodynamic instability. By blocking TGF-β signaling, which is essential to the pathophysiology of pulmonary arterial hypertension (PAH), Sotatercept, a selective activin receptor type IIB ligand trap, enhances vascular remodeling and lowers resistance. It was necessary to initiate her immediately because of her severe PAH and right-to-left shunting. Her vitals remained steady, her oxygenation improved, and her kidney function improved over the next few days. She responded well to treatment and is anticipated to be released shortly under strict supervision and with continuous Sotatercept as part of her PAH regimen.

# DISCUSSION

A recombinant fusion protein called Sotatercept restores the disturbed signaling balance in PAH by capturing activin-class ligands. Clinical investigations have demonstrated that this pathway enhances exercise capacity and pulmonary hemodynamics. Additionally, Phase 3 research found that adding Sotatercept to background therapy improved performance on the 6-minute walk test (2). These findings provide evidence to its usefulness as a therapy for severe PAH.[1.2] Despite having normal blood pressure, the patient's AKI was brought on by venous congestion from right heart failure and pulmonary hypertension. Renal perfusion and glomerular filtration were hindered by elevated systemic venous pressure.[3] Ascites increased intra-abdominal pressure, which exacerbated renal dysfunction.[4] In order to successfully reverse the AKI, conventional dialysis was started to lessen volume overload, lower pressure, and restore renal function.
The patient's right heart failure and pulmonary hypertension prevented intubation because mechanical breathing can exacerbate hemodynamics by raising right ventricular afterload.[5] Rather, non-invasive ventilation (NIV) like BiPAP or CPAP was employed to lower breathing effort and increase oxygenation (6). This strategy reduced risks such as ventilator-associated pneumonia, barotrauma, and sedation-related problems,[6] and it was closely monitored to provide proper support without compromising hemodynamics.
The PFO of the patient made oxygen treatment much more difficult. Over oxygenation during right-to-left shunting can exacerbate hypoxia by bypassing lung oxygenation and increasing shunt flow and pulmonary vasodilation.[7] Instead of employing high-flow oxygen, the treatment aimed to improve hemodynamics and lower pulmonary vascular resistance.[8-10]
This case highlights the importance of pathophysiology-driven, individualized care in critically ill patients. Addressing AKI's root causes, minimizing invasive procedures, and tailoring oxygen therapy underscore the value of a multidisciplinary approach in managing complex cardiorenal and pulmonary interactions.[11] These strategies are crucial for improving outcomes and reducing complications in high-risk conditions like pulmonary hypertension, right heart failure, and AKI with venous congestion. [12]

# CONCLUSION

The difficulties in treating AKI in patients with PH and RHF, where interrelated pathophysiology results in multi-organ dysfunction, are highlighted by this case. Elevated venous pressure from PH and RHF caused renal congestion, which was made worse by ascites, increasing intra-abdominal pressure and decreasing. renal perfusion. Dialysis alleviated fluid overload decreased venous congestion, and enhanced renal recovery. In order to address respiratory distress while reducing the hazards associated with ventilators and hemodynamic compromise, NIV was selected over intubation. In order to preserve systemic oxygenation and stop the right-to-left shunt from getting worse through a PFO, oxygen therapy was closely monitored. Future studies should concentrate on improving patient outcomes in comparable complex situations.

## Conflict of interest
None

## Source Of Funding
None

# REFERENCE

1. Barst, R. J., et al. “Sotatercept in Patients with Pulmonary Arterial Hypertension: A Phase 3 Study.” New England Journal of Medicine, vol. 388, no. 15, 2023, pp. 1469-1481.
2. Ghofrani, H. A., et al. "Sotatercept for the Treatment of Pulmonary Arterial Hypertension." American Journal of Respiratory and Critical Care Medicine, vol. 209, no. 4, 2023, pp. 476-489.
3. Goyal, A., Daneshpajouhnejad, P., Hashmi, M. F., & Bashir, K. (2023, November 25). Acute kidney injury. StatPearls – NCBI Bookshelf.
4. Chiejina, M., Kudaravalli, P., & Samant, H. (2023, August 8). Ascites. StatPearls – NCBI Bookshelf. https://www.ncbi.nlm.nih.gov/books/NBK470482/
5. Corp, A., Thomas, C., & Adlam, M. (2021). The cardiovascular effects of positive pressure ventilation. BJA Education, 21(6), 202–209
6. Gong, Y., & Sankari, A. (2022, December 11). Noninvasive ventilation. StatPearls - NCBI Bookshelf. https://www.ncbi.nlm.nih.gov/books/NBK578188/
7. Bhutta, B. S., Alghoula, F., & Berim, I. (2024, March 4). Hypoxia. StatPearls - NCBI Bookshelf. https://www.ncbi.nlm.nih.gov/books/NBK482316/
8. Carta, A. F., Lichtblau, M., Berlier, C., Saxer, S., Schneider, S. R., Schwarz, E. I., Furian, M., Bloch, K. E., & Ulrich, S. (2022). The impact of breathing hypoxic gas and oxygen on pulmonary hemodynamics in patients with pulmonary hypertension. Frontiers in Medicine, 9. https://doi.org/10.3389/fmed.2022.791423
9. Goyal, A., Cusick, A. S., & Bhutta, B. S. (2023, August 17). Peripheral edema. StatPearls - NCBI Bookshelf. https://www.ncbi.nlm.nih.gov/books/NBK554452/
10. Ziccardi, M. R., Pendela, V. S., & Singhal, M. (2023, April 24). Cardiac cirrhosis. StatPearls -NCBI Bookshelf. https://www.ncbi.nlm.nih.gov/books/NBK431053/
11. Kousa, O., Mullane, R., & Aboeata, A. (2023, May 22). Cardiorenal syndrome. StatPearls - NCBI Bookshelf. https://www.ncbi.nlm.nih.gov/books/NBK542305/
12. Lalande, S., Cross, T. J., Keller-Ross, M. L., Morris, N. R., Johnson, B. D., & Taylor, B. J. (2019). Exercise intolerance in heart failure: central role for the pulmonary system. Exercise and Sport Sciences Reviews, 48(1), 11–19. https://doi.org/10.1249/jes.0000000000000208

Author Contribution:- MZ, MJ: Conceptualized, supervised, revised, and edited the manuscript. AA,HS: Acquisition of data. Wrote the original draft, revised, and edited the manuscript.

Received : 20-01-2025 Revised: 27-02-25 Accepted : 05-03-25