



Pulmonary arterial hypertension (PAH) with right ventricular failure (RVF) poses major perioperative challenges due to the compromised right ventricle's difficulty with rapid hemodynamic changes. In advanced cases, volume shifts, vasodilation, positive pressure ventilation, and altered sympathetic tone can lead to critical declines in cardiac output and coronary perfusion. Effective management necessitates an interdisciplinary approach focused on thorough preoperative optimization to stabilize hemodynamics and reduce risks.

We present a 67-year-old female with history significant for idiopathic PAH with fulminant RVF, chronic respiratory failure on home oxygen, chronic kidney disease stage III, and HFpEF who was evaluated for skin graft placement for non-healing, bilateral lower extremity skin lesions following a low velocity traumatic injury.

A pre-operative right heart catheterization was performed and significant for worsening PAH (73/26mmHg), RVF, and cardiogenic shock (CI = 1.4) despite active management with remodulin, tadalafil, and ambrisentan. Other pertinent medications included furosemide, metolazone and milrinone for heart failure.

These findings were consistent with an ECHO performed 10-months prior, which also demonstrated normal LV function, severely reduced RVEF, severe RV enlargement w/ septal deviation (D-sign), and severe tricuspid regurgitation with IVC flow reversal (peak TR gradient = 64mmHg).

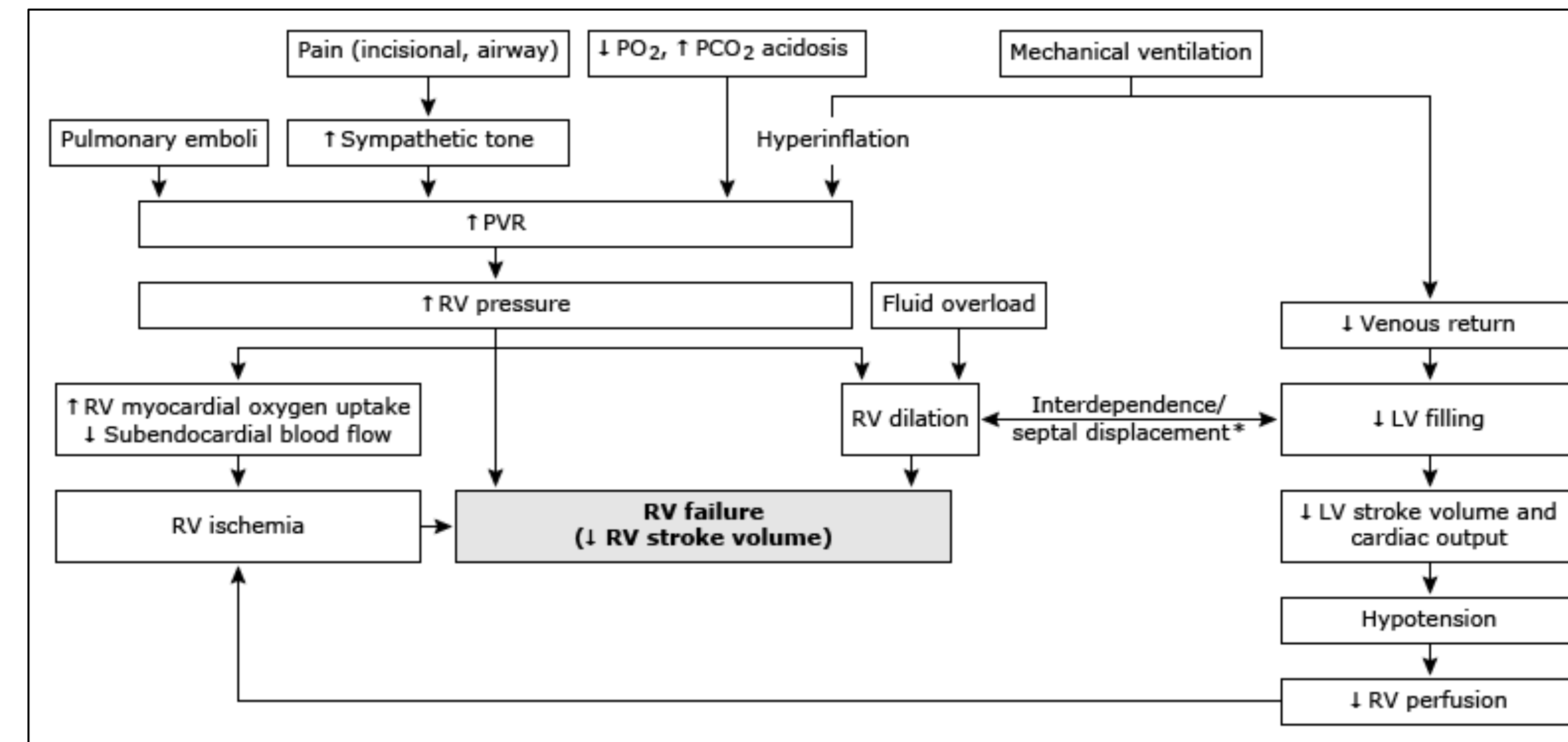


Figure 1: Pathophysiology of RV Failure in Patients with PAH during the Preoperative Period. This diagram illustrates how acute hemodynamic stressors lead to elevated pulmonary vascular resistance (PVR) and RV pressure, promoting RV dilation and ischemia. The resulting RV failure diminishes RV stroke volume, causing a cascade of effects on left ventricular filling, cardiac output, and systemic perfusion - ultimately exacerbating hypotension and RV perfusion deficits^[1].

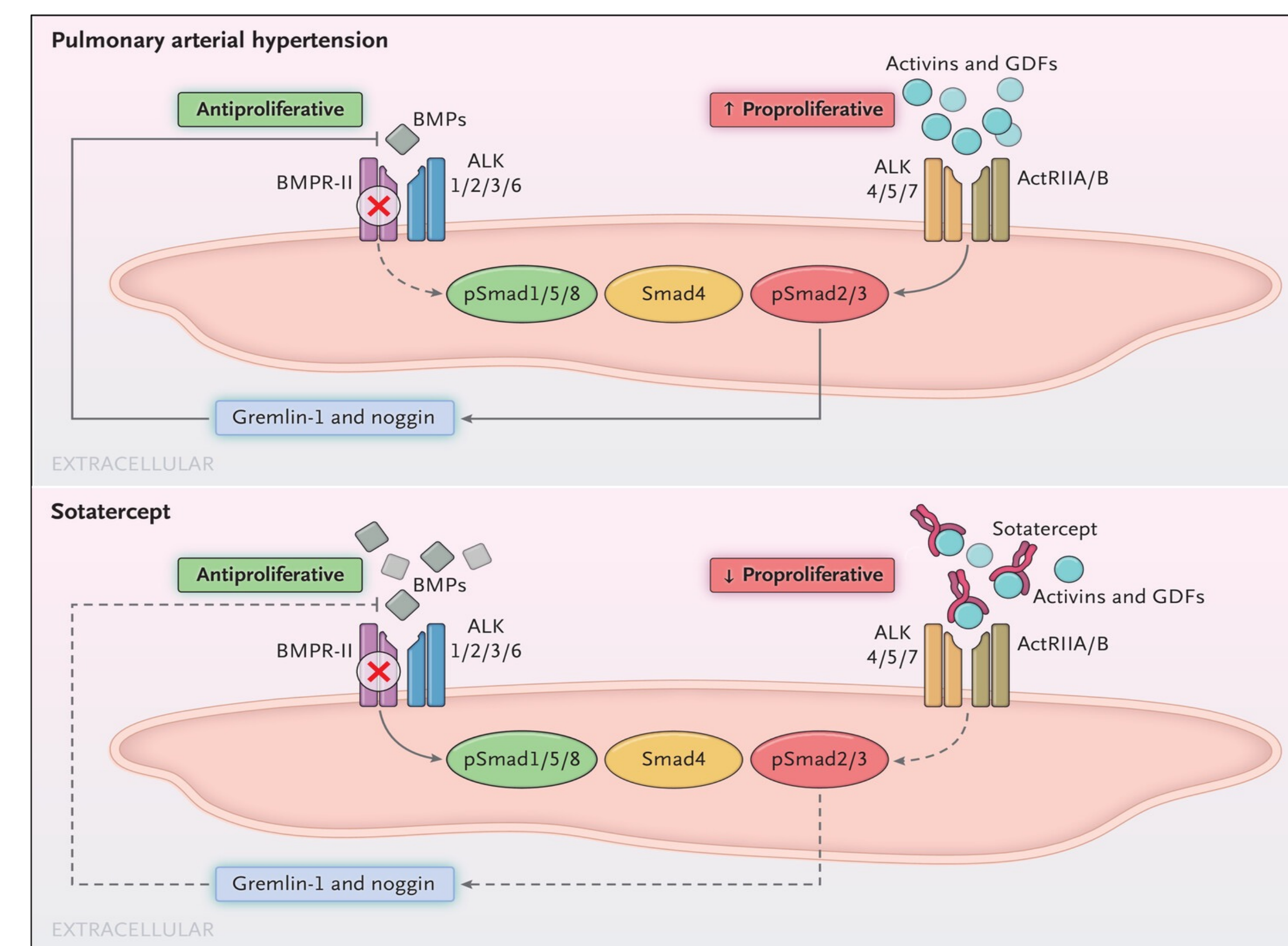


Figure 2: Proposed Mechanism of Action of Sotatercept. In PAH, reduced BMPR-II signaling leads to decreased BMP activity, resulting in increased pro-proliferative signals and vascular remodeling via unregulated activin and growth differentiation factor (GDF) pathways. Sotatercept - a ligand trap composed of a fusion between the Fc portion of IgG and the extracellular domain of the human activin receptor type IIA - selectively binds to activins and GDFs, reducing their pro-proliferative effects and rebalancing growth signals towards the antiproliferative BMP pathway. Overall, the downstream effects helps to restore endothelial function and vascular integrity in PAH patients.^[4]

Discussion and Plans for Management

The greatest risk for surgical patients with RVF secondary to PAH is the inability of a pathologic RV to accommodate rapid changes in hemodynamics. Volume shifts, vasodilation from anesthetic agents, positive pressure mechanical ventilation, and altered sympathetic tone can all precipitate a cascade of downstream effects that decrease cardiac output and coronary perfusion, which ultimately increases risk for severe morbidity or mortality^[2].

Ultimately, a multidisciplinary risk-based discussion between cardiac anesthesiology, cardiology, pulmonology, and plastic surgery determined the patient was not optimized and at a prohibitively high-risk for experiencing adverse cardiovascular events during surgery. The patient's most recent right heart catheterization revealed severely elevated pulmonary artery pressures and markedly low cardiac index despite aggressive management, suggesting further increases in RV afterload could precipitate spiraling hemodynamic decline^[3]. With continued plans for wound care, antibiotics, and frequent close monitoring, the team concluded that the risks of cardiovascular collapse during the perioperative period overshadow concerns for wound infection. Therefore, the decision to delay surgery and start Sotatercept – an activin signaling inhibitor approved by the FDA in March 2024 – for better preoperative optimization was warranted considering all conventional treatment of PAH has proved unsuccessful thus far^[4]. In the end, patient was continued on frequent wound assessments and an expedited initiation of Sotatercept, with a reconsideration for intervention in 4-6 weeks pending responsiveness to new medication.

This case illustrates the imperative for anesthesiologists to recognize the delicate balance between surgical necessity and hemodynamic stability in patients with severe PAH and RVF. The strategic postponement of surgery to optimize PAH management serves as a critical learning point in prioritizing patient safety over procedural urgency.

References

- [1] Baumgartner N, De Jesus Perez V. Anesthesia for noncardiac surgery in patients with pulmonary hypertension or right heart failure. UpToDate.
- [2] McGlothlin, Dana P et al. "ISHLT consensus statement: Perioperative management of patients with pulmonary hypertension and right heart failure undergoing surgery." *The Journal of heart and lung transplantation : the official publication of the International Society for Heart Transplantation* vol. 41,9 (2022): 1135-1194. doi:10.1016/j.healun.2022.06.013
- [3] Caddigan S, Granlund B. Anesthesia for Patients With Pulmonary Hypertension or Right Heart Failure. [Updated 2023 Jan 29]. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2024 Jan-. Available from: <https://www.ncbi.nlm.nih.gov/books/NBK572071/>
- [4] Humbert, Marc et al. "Sotatercept for the Treatment of Pulmonary Arterial Hypertension." *The New England journal of medicine* vol. 384,13 (2021): 1204-1215. doi:10.1056/NEJMoa2024277