

Pericardial Decompression Syndrome in a Pediatric Patient

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Background:

Pericardial decompression syndrome (PDS) is a rare but critical complication of pericardial drainage. It is most associated with pericardial drainage to treat cardiac tamponade but can also occur without preceding tamponade physiology.¹ Pericardial drainage is typically performed to relieve external compression on the heart in order to help restore normal cardiac function. In PDS, patients paradoxically develop hemodynamic instability and pulmonary edema from ventricular dysfunction after drainage of a pericardial effusion. PDS can present with isolated right, isolated left, or biventricular dysfunction.² While there are many reports of PDS in adults, there is only one previously reported pediatric case which occurred in a teenager.³

Case summary:

A 4-year-old boy with history of chronic granulomatous disease who underwent haploidentical hematopoietic stem cell transplant required prolonged hospitalization for multiple infections and graft-versus-host disease. His clinical course was complicated by the development of a pericardial effusion which ultimately required pericardiocentesis. Shortly after undergoing uncomplicated pericardiocentesis, the patient developed pulmonary edema and shock. Echocardiogram demonstrated a severely dilated right ventricle (RV) with a flattened interventricular septum and diminished RV systolic function. The patient was diagnosed with PDS. Aggressive supportive care with high dose vasopressors and mechanical ventilation with inhaled nitric oxide was initiated. Over the course of three days the patient's RV function gradually returned to normal and he was weaned off all vasoactive medications and extubated.

Discussion:

PDS is an acute hemodynamic sequela of pericardiocentesis or surgical pericardiostomy that carries significant risk of morbidity and mortality. The mechanism of PDS is not well understood, and there are multiple proposed hypotheses. The hemodynamic hypothesis is most consistent with our patient's presentation; it proposes increased systemic venous return to a more compliant RV after pericardiocentesis causes increased RV preload and RV dilation.^{2,4} This in turn causes LV compression from interventricular septum bowing, which limits LV preload and leads to decreased cardiac output and pulmonary edema.^{2,4} Management of PDS is focused on supportive care, including interventions to promote cardiac function and maintain appropriate blood pressure. This case highlights the importance of hemodynamic monitoring, ideally in an intensive care unit, after pericardiocentesis. Early detection and initiation of treatments centered on supportive care are critical aspects of managing PDS. The paucity of reported pediatric cases of PDS may be attributed to it being an under-recognized and under-reported entity. Further investigation into PDS in pediatric patients is needed to guide risk assessment, prevention strategies, and optimal management.

References

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