CLINICAL VIGNETTE

Novel Perioperative Management of VSD/Eisenmenger's Syndrome/Severe Pulmonary Hypertension

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Case Report

An 80-year-old male with history of intellectual developmental disorder and Eisenmenger's syndrome secondary to congenital ventricular septal defect (VSD) presented to the emergency department with right hip pain after tripping over a chair while getting up from lunch. The pain was constant, non-radiating in the right hip, and worsened by movement. He was hypertensive on admission with blood pressure of 161/74mmHg, a pulse of 92 beats per minute, and a respiratory rate of 18 breaths per minute with oxygen saturation of 98% on room air. X-rays of hips and femur found a displaced right femoral neck fracture and diffuse osteopenia (Figure 1). ECG (Figure 2) showed atrial flutter and left anterior fasciculate block.

He was scheduled for a right hip hemiarthroplasty that afternoon. Transthoracic echocardiogram found a large (1cm) membranous ventricular septal defect and severe pulmonary hypertension (94mmHg based on a central venous pressure of 10mmHg) (Figure 3). He was classified as high risk (~20% chance of major adverse occurring peri- and post-operatively) due to these findings. The patient and his surrogate decision maker elected to proceed with surgery. Considerations included the high risk of paradoxical stroke and lower quality of life without surgery. Cardiology recommended he continue heparin infusion until one hour prior to surgery; use of protamine sulfate as needed for residual bleeding; spinal anesthesia to avoid intubation; and use of nitrates for shunt related hypoxemia, with addition of rotating tourniquets to control hypotension from nitrates.

The patient underwent right hip hemiarthroplasty (Figure 4) that afternoon under spinal anesthesia and tolerated the procedure well without pulmonary or neurological symptoms. He had some ecchymosis over the surgical site, but patient continued to do well and was discharged home on post-op day 3 and prescribed to enoxaparin.

Discussion

Eisenmenger's syndrome, also known as tardive cyanosis, is a condition characterized by pulmonary vascular sclerosis and a right-to-left shunt resulting from longstanding left-to-right shunt. The left-to-right shunt may be from a VSD as in our patient or atrial septal defect, patent ductus arteriosus, or other cyanotic heart diseases.[1]

A VSD shunts left-to-right early in life due to lower total pulmonary vascular resistance (PVR) than the systemic vascular resistance (SVR). If the shunting is significant enough, the increased pulmonary circuit pressures damage capillaries causing pulmonary vascular sclerosis. The sclerotic tissue does not transfer oxygen and increases vascular resistance causing further pulmonary hypertension and right ventricular hypertrophy (RVH). If RV pressures raise enough to compete with systemic pressures, then the shunt stops or reverses to right-to-left sending deoxygenated blood to the systemic circulation via the VSD, causing cyanosis from the admixture of desaturated blood. This can acutely worsen with increased PVR, which can be caused by intubation, positive end-expiratory pressure (PEEP), continuous positive airway pressure (CPAP), bilevel positive airway pressure (biPAP), or pulmonary emboli. The drop in arterial oxygen is unresponsive to oxygen supplementation.

In some cases, the chronic murmur from VSD would disappear. If the murmur disappears after age 30, it should raise concern the murmur may have stopped for the wrong reason. The right reason would be spontaneous closure of the defect, which can occur over time, typically by age 20. Spontaneous closure occurs in about 37% of membranous VSDs and 50% of muscular VSDs.[2] The wrong reason is increasing pulmonary hypertension reducing the trans-defect flow, typically after age 40. Thus for any patient with a history of murmur but no murmur on exam, shunt flow reversal and Eisenmenger's syndrome should be considered.

Decreased effective oxygen transfer area causes decreased oxygenation and triggers increased erythrocyte production causing polycythemia. Polycythemia predisposes the patient to clot formation and embolism. Due to presence of VSD, patients with Eisenmenger's syndrome are also at risk of paradoxical embolism causing a potentially fatal stroke, for example, in our case during his planned arthroplasty.[3] On the other hand if surgery is cancelled, femur fractures pose high risk of venous thromboembolism (VTE) and delayed fixation of fracture is associated with increased morbidity from infection.[4] Femur fracture is considered a high risk for VTE

with mortality due to pulmonary embolism being as high as 10% before anticoagulant prophylaxis was implemented.[5] Worse still, VTE has been documented in 45% of patients who underwent tibial casting, most of whom were asymptomatic. In a patient with Eisenmenger's syndrome, such embolism could cross the VSD and cause a stroke.

Perioperative Approach

Spinal anesthesia can avoid the complication of intubation and mechanical ventilation. Intubation causes an increase in sympathetic tone resulting in increased heart rate and blood pressure[6] and can increase PVR directly, especially if PEEP is required. In a patient with Eisenmenger's syndrome, the increased blood pressure increases the right-to-left shunt due to pulmonary vascular sclerosis. This increased right-to-left shunting causes hypercapnia and hypoxia refractory to supplemental oxygen. In case of hypoxia due to shunt, inhaled nitric oxide (NO) and, indirectly, NO elevation via nitroglycerin have been shown to reduce PVR and thus alleviate the right-to-left shunt.[7] Nitroglycerin lowers PVR, but it can also lower systemic vascular resistance, hence the recommendation to compensate for low SVR if needed via rotating tourniquets. The efficacy of Nitroglycerin can be monitored with pulse oximetry, which will show whether the patient is responding to supplemental oxygen.[8]

The treatment goal for hypotensive shock is to restore adequate blood pressure usually by volume repletion with the use of vasopressors like dopamine, phenylephrine, or norepinephrine to bridge the treatment.[9] In a patient with Eisenmenger's syndrome, however, the use of vasopressors can increase right-to-left shunt similarly or even in excess of the increase in systemic effects, producing hypoxemia not responsive to supplemental oxygen. In order to avoid increasing preload on right ventricle, triple rotating tourniquets, which are usually used to pool blood in extremities[10] at low (venous) pressure, can also be used to increase SVR under high (arterial) pressure, thus counteracting the drive for right-to-left shunt.

Figures

Figure 1. Initial radiograph of right hip and femur.

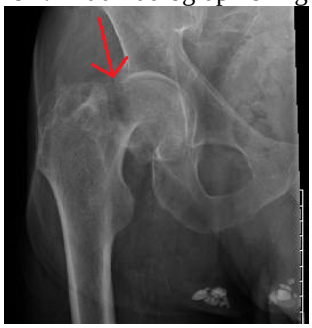


Figure 2. ECG showing atrial flutter and left anterior fasciculate block.

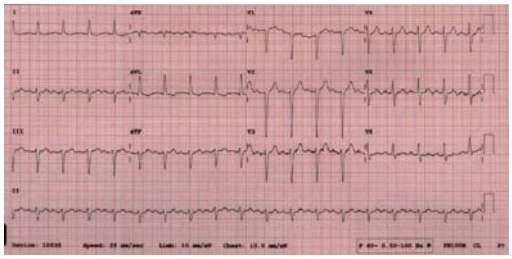


Figure 3. Transthoracic echocardiogram showed a large (1cm) membranous ventricular septal defect, severe pulmonary hypertension (94mmHg based on a central venous pressure of 10mmHg), mild concentric left ventricular hypertrophy, mild diastolic relaxation impairment, and an ejection fraction of 65%.

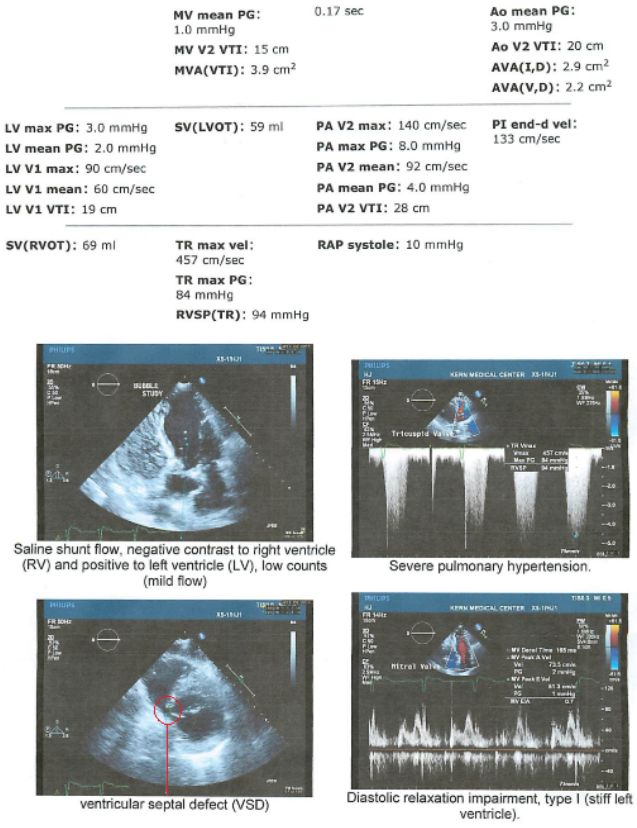
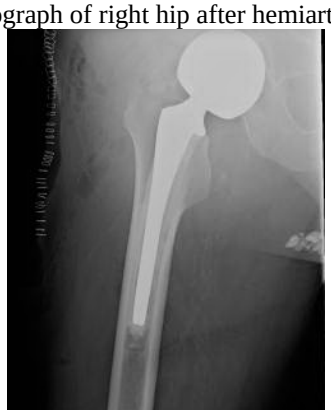


Figure 4. Radiograph of right hip after hemiarthroplasty.



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