“The Ross Procedure for Laubrey-Pezzi Syndrome: Surgical Success and Post-Operative Neurological Challenges”

Abstract : The article discusses a 21-year-old patient with Laubrey-PESI syndrome, characterized by structural cardiac anomalies requiring multiple surgeries, including a Ross procedure after dysfunction of a mechanical aortic valve. The Ross procedure, which uses the patient's own pulmonary valve to replace the diseased aortic valve, is beneficial for young patients due to reduced rejection and thrombosis risks and better hemodynamic adaptation. Despite its advantages, the procedure is complex and carries risks such as prosthesis-patient mismatch and neurological complications, evidenced by the patient's postoperative ischemic stroke, which resolved with anticoagulation. The case underscores the importance of a multidisciplinary approach and close monitoring for optimal outcomes, contributing valuable insights to the management of similar conditions.

1. Introduction

Laubrey-PESI syndrome is a rare and complex congenital disease characterized by structural cardiac abnormalities, often including a ventricular septal defect (VSD) and valvular dysfunctions. Management of this syndrome requires a multidisciplinary approach and repeated surgical interventions throughout the patient's life due to the progressive nature of the cardiac anomalies [1].

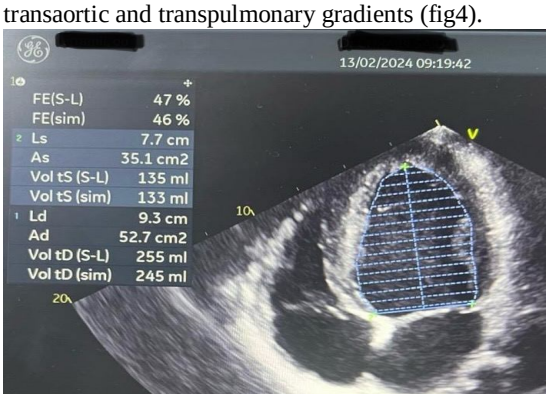
The Ross procedure, which involves replacing the diseased aortic valve with the patient's own pulmonary valve and implanting a bioprosthesis in the pulmonary position, offers several advantages. It uses the patient's own tissues, reducing the risks of rejection and thrombosis, and improves hemodynamic outcomes [2]. This technique is particularly beneficial for young patients as it adapts to growth and increased metabolic needs [3].

Here, we present the case of a 21-year-old patient with Laubrey-PESI syndrome who underwent multiple surgical interventions, including a Ross procedure due to dysfunction of a mechanical aortic valve. This case illustrates the challenges and complications associated with managing this complex syndrome [4].

2. Case Presentation

The patient, followed since birth for Laubrey-PESI syndrome, underwent surgery at the age of 1 year for closure of a ventricular septal defect (VSD). At 20 years old, he underwent reoperation for aortic valve replacement with implantation of a mechanical valve. In the immediate postoperative period, the patient presented with valve mismatch with high transaortic gradients of 65 mmHg, indicating dysfunction of the mechanical valve, necessitating a Ross procedure. At admission for the Ross procedure, the patient was asymptomatic except for exertional dyspnea. Vital signs were stable (BP 148/80 mmHg, HR 90 bpm) and ECG showed a regular sinus rhythm with negative T waves in the anteroseptal-apical territory. Echocardiography revealed a dilated non-hypertrophied left ventricle with mild dysfunction and an ejection fraction of 47% (fig2), a non-dilated right ventricle with dysfunction, and dysfunction of the aortic valve prosthesis with a mean gradient of 65 mmHg (fig2) and moderate paravalvular leak (fig3). Pulmonary artery pressure was 32 mmHg. The workup showed microcytic hypochromic anemia at 11 g/dL with a CRP of 10 mg/L. After the Ross procedure, during which the diseased aortic valve was replaced by the patient's own pulmonary valve and a bioprosthesis was placed in the pulmonary position, the patient developed muscle weakness in the left upper limb. An emergency brain CT scan revealed a right frontal hypodensity of ischemic origin. Curative anticoagulation was initiated. The patient recovered muscle strength, and the follow-up CT

scan was normal. Transthoracic echocardiography performed five weeks after surgery showed normal transaortic and transpulmonary gradients (fig4).



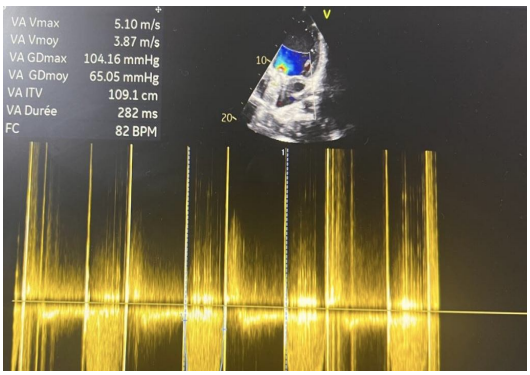


Fig1 :apical 4-chamber echocardiographic view showing a dilated dysfunctional left ventricule with FEVG : 47%

Fig2 :Continuous Doppler on the aortic prosthesisshowing a high mean transprostheticgradient of 65 mm

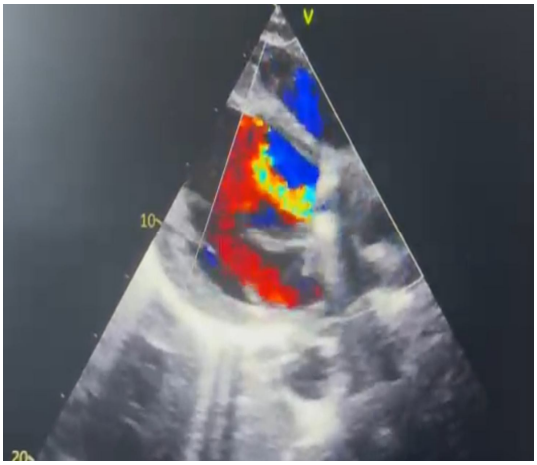


Fig 3:Moderate Aortic Paraprosthetic Leak

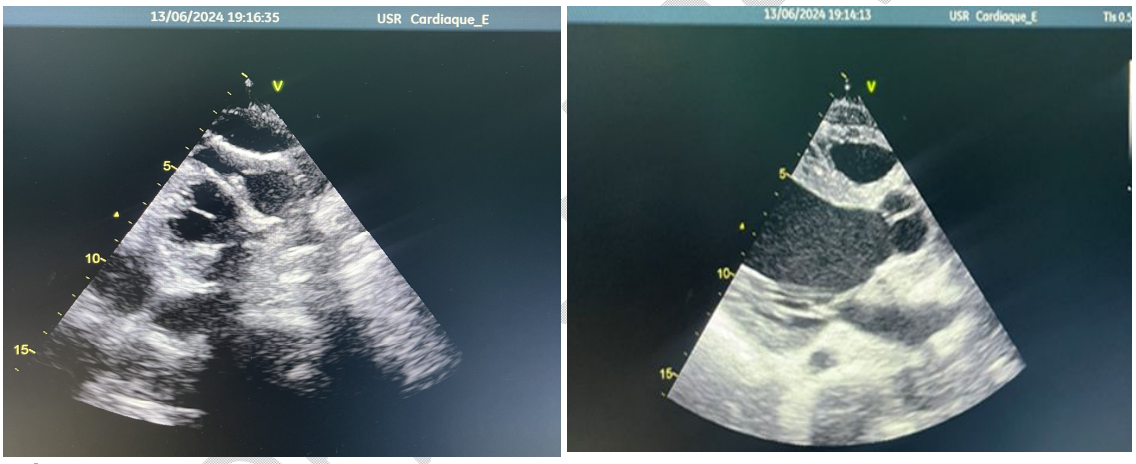


Fig4:Post-operative evaluation of theaortic valve: parasternal long-axis and short axis views

3. Discussion

The Ross procedure, developed by Donald Ross in 1967, is a complex surgical technique but particularly beneficial for young patients requiring aortic valve replacement [5]. This technique involves harvesting the patient's pulmonary valve and implanting it in the aortic position, with a bioprosthesis placed in the pulmonary position. Using the patient's own tissues to replace the aortic valve offers several advantages, including better adaptation to hemodynamic needs, reduced risks of rejection and thrombosis, and increased durability [6]. The autologous pulmonary valve can better tolerate the high systemic pressures of the aortic circulation, thereby reducing the risks of long-term complications [2]. Prosthesis-patient mismatch is a well-documented complication of aortic valve replacements, where the size of the prosthesis is inadequate relative to the patient's aortic annulus, leading to high transvalvular gradients and valve dysfunction [7]. In the reported case, the patient presented with a transaortic gradient of 65 mmHg after the placement of a mechanical valve, indicating severe mismatch. This problem is critical as it can increase the risk of cardiac complications, including heart failure and prosthetic failure [8]. The Ross procedure, although more complex, offers an effective solution to this problem by using autologous tissues, thus reducing the risks of mismatch and thromboembolic complications [3].

The development of a cerebrovascular accident (CVA) after cardiac surgery is a dreaded complication, although rare, especially in a young patient of 21 years. The incidence of postoperative CVA varies, but factors such as aortic manipulations, emboli from thrombus or prosthetic material, and episodes of cerebral

hypoperfusion can increase this risk [9]. In the presented case, the patient developed muscle weakness in the left upper limb, diagnosed as a right frontal hypodensity of ischemic origin on a brain CT scan. Management of this complication involved initiating curative anticoagulation, which allowed complete recovery of muscle strength and normalization of images on follow-up CT scan .

Prevention of neurological complications relies on close monitoring of high-risk patients, optimization of surgical and anesthetic techniques, and rapid management of postoperative neurological signs [10].

4. Conclusion

This case illustrates the clinical challenges and potential complications in managing patients with Laubrey-PESI syndrome requiring multiple valvular interventions. The Ross procedure appears to be a viable solution for young patients, despite the associated risks. Close postoperative monitoring and a multidisciplinary management approach are essential to optimize long-term outcomes. The rarity of this case and the complexity of the medical and surgical management underscore the value of this report. By documenting specific challenges and applied solutions, this article contributes to enriching the scientific literature on Laubrey-PESI syndrome and postoperative complications of the Ross procedure, providing valuable information for the future management of patients with similar conditions.

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