



Incomplete Shone's Complex, Surgical Resolution Through Minimally Invasive Approach

Carlos A. Jimenez Fernandez, MD1, Victor H. Sainz Escarrega, MD1, Oscar S. Lomeli Sanchez, MD, MSc1, Francisco J A Gutierrez Lopez, MD1, Italo D. Masini Aguilera, MD1, Benigno Ferreira Piña, MD1, Jaime G. Lopez-Taylor MD2, Dalia T. Feliz Alcantara, MD2.

1Instituto Cardiovascular de Minima Invasion, Zapopan, Mexico,
2Hospital Civil de Guadalajara Fray Antonio Alcalde, Guadalajara, Mexico.



Introduction

- Anomalies in the mitral valve are frequently reported in congenital diseases, Shone et al. first described a complex in 1963 that consisted of 4 simultaneous anomalies: parachute mitral valve, supra-annular mitral ring, subaortic stenosis, and coarctation of the aorta (1,2). Since the original description, the “Shone's complex” definition has broadened to include other patients with other congenital obstructive lesions, as the majority of patients have only 2 or 3 of these anomalies (3). We present the **surgical resolution through a minimally invasive approach**.

Case report

- We present the case of a 16-year-old female with **4 previous surgical events** throughout her life; pulmonary artery banding at 9 months old, interventricular septum defect closure at 5 years old, and pulmonary artery reconstruction at age 9.
- She presented to the outpatient clinic with a history of progressive dyspnea, orthopnea, and paroxysmal nocturnal dyspnea. Initial Eco showed a “**hammock sign**” and a mean gradient of 17 mm Hg in the mitral valve with decreased valvular area (1.2 cm²) related to the presence of a supra valvular ring and a hypoplastic posterior leaflet (Figure 2 and 3).
- A minimally invasive approach was proposed due to the high risk of complications related to previous surgeries and the benefits of shorter in-hospital stay, and recovery time.
- An anterolateral right thoracotomy was made with assisted 3D thoracoscopic view, approaching the posterior interatrial septum through 4 intercostal space. Extracorporeal circulation was placed through a right femoral incision, mild hypothermia and anterior cardioplegia were used. Surgical findings include the **presence of a firm supra-annular mitral ring and a hypoplastic posterior leaflet** (Figure 1). We place a mechanical prosthesis 27 mm in an annular position. Post-operative eco showed normal function and no regurgitation. She underwent an uneventful recovery and was discharged on postoperative day 7.

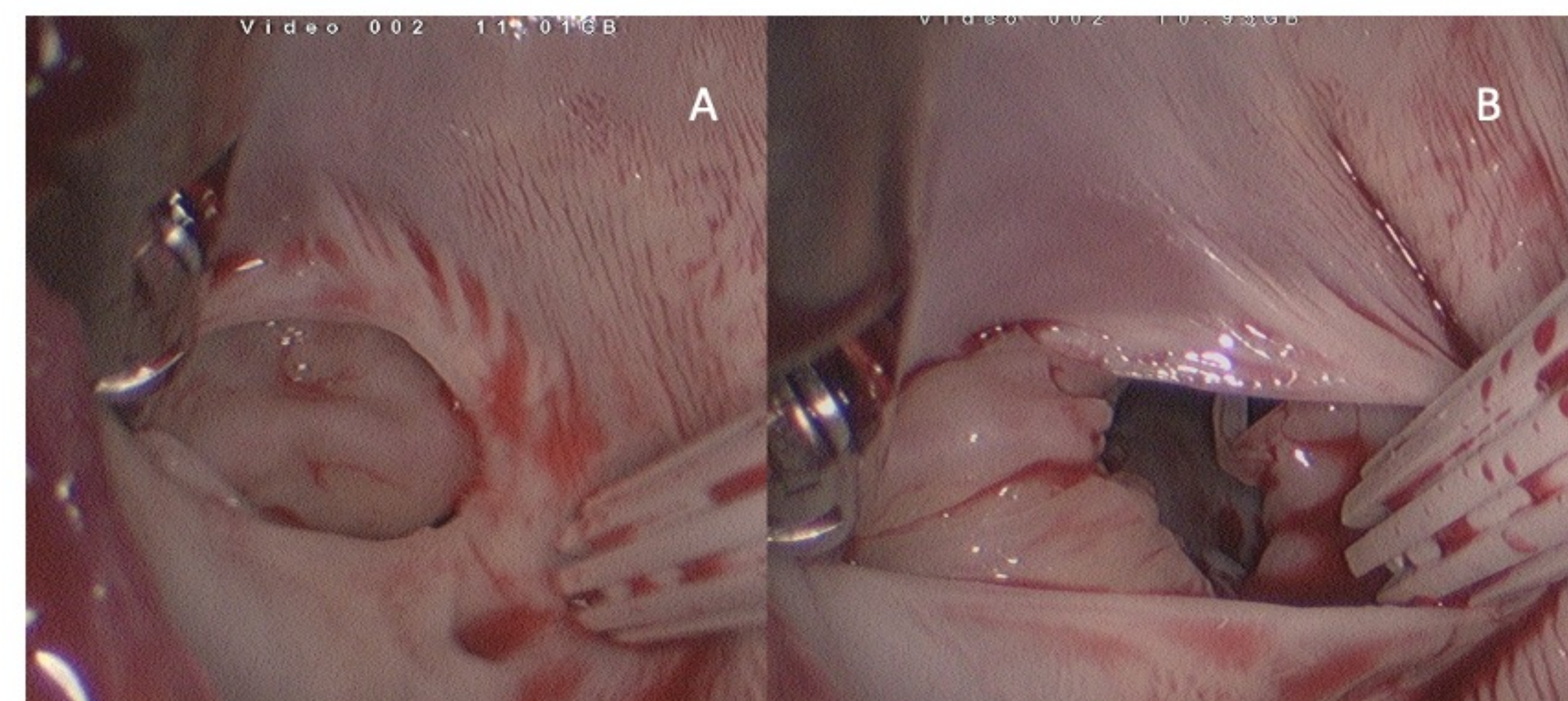


Figure 1. Endoscopic Vision of Mitral Valve A) Supra-annular annular fibrous ring B) Exposure of thickened anterior leaflet and hypoplastic posterior leaflet

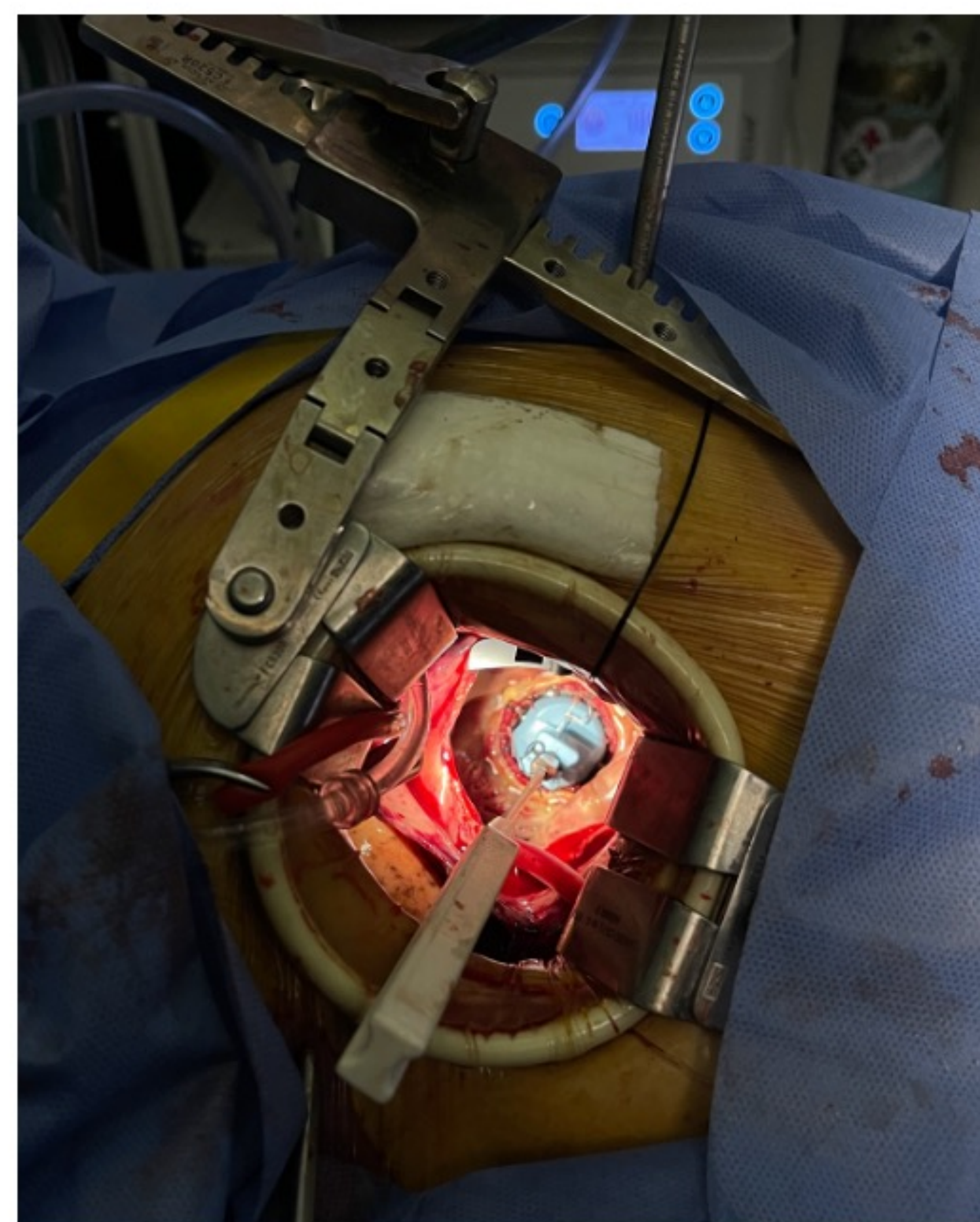


Figure 4. Mechanical prosthetic valve placed through right anterolateral minithoracotomy fixed with Cor Knot® fasteners.

Discussion

- The initial definition of Shone's complex described a multilevel obstruction due to a mitral deformity (parachute), the supra-annular mitral valve ring, sub-aortic stenosis and coarctation of the aorta. Since then individuals with some of these features are referred to as having a **partial or incomplete form of this anomaly**, which is very well recognized in children. However, reports of this partial form are not so commonly reported in adults (4).
- In children with this condition pulmonary hypertension has been associated with worse prognosis. Jain et al described a case series with invasive hemodynamic data in patients that survive to adulthood, they described a high prevalence of diastolic dysfunction and pulmonary hypertension, which had a significant pre-capillary and post-capillary component (5).
- This is important because diastolic dysfunction is associated with an **increased number of reinterventions**. Moreover, repeated surgical procedures might be related to more injury to the myocardium during cardioplegia periods affecting hypertrophic hearts significantly more (5).
- Is important to note **the high prevalence of atrial arrhythmias**, reported in almost 25% of all patients, likely related to the prolonged exposure to increase pressure in the LA and LVEDP, as well as redo surgeries(5)
- Because of these particularities and the uncommon presentation of these patients, and also variability and severity of the anatomic features, **surgical outcomes in adults are not well established and are mostly limited to case reports** (4,6). The patient described in this report had several previous surgeries related to congenital defects but was never classified as a variant of the Shone's complex, fortunately she did not have the aortic component and so underwent a successful minimally invasive mitral valve replacement.

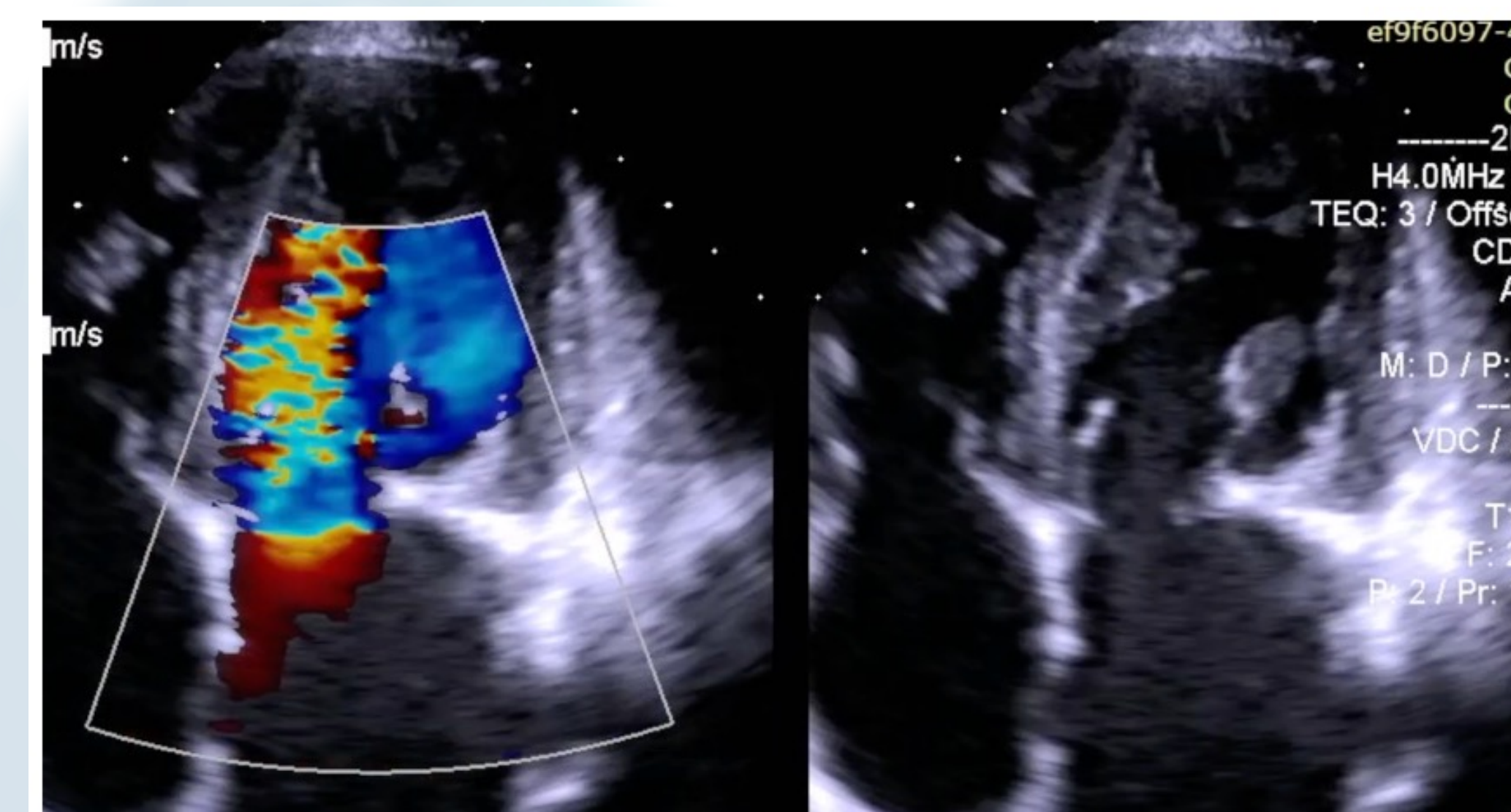


Figure 2. TTE 4ch showing hammock sign and supra-annular mitral stenosis

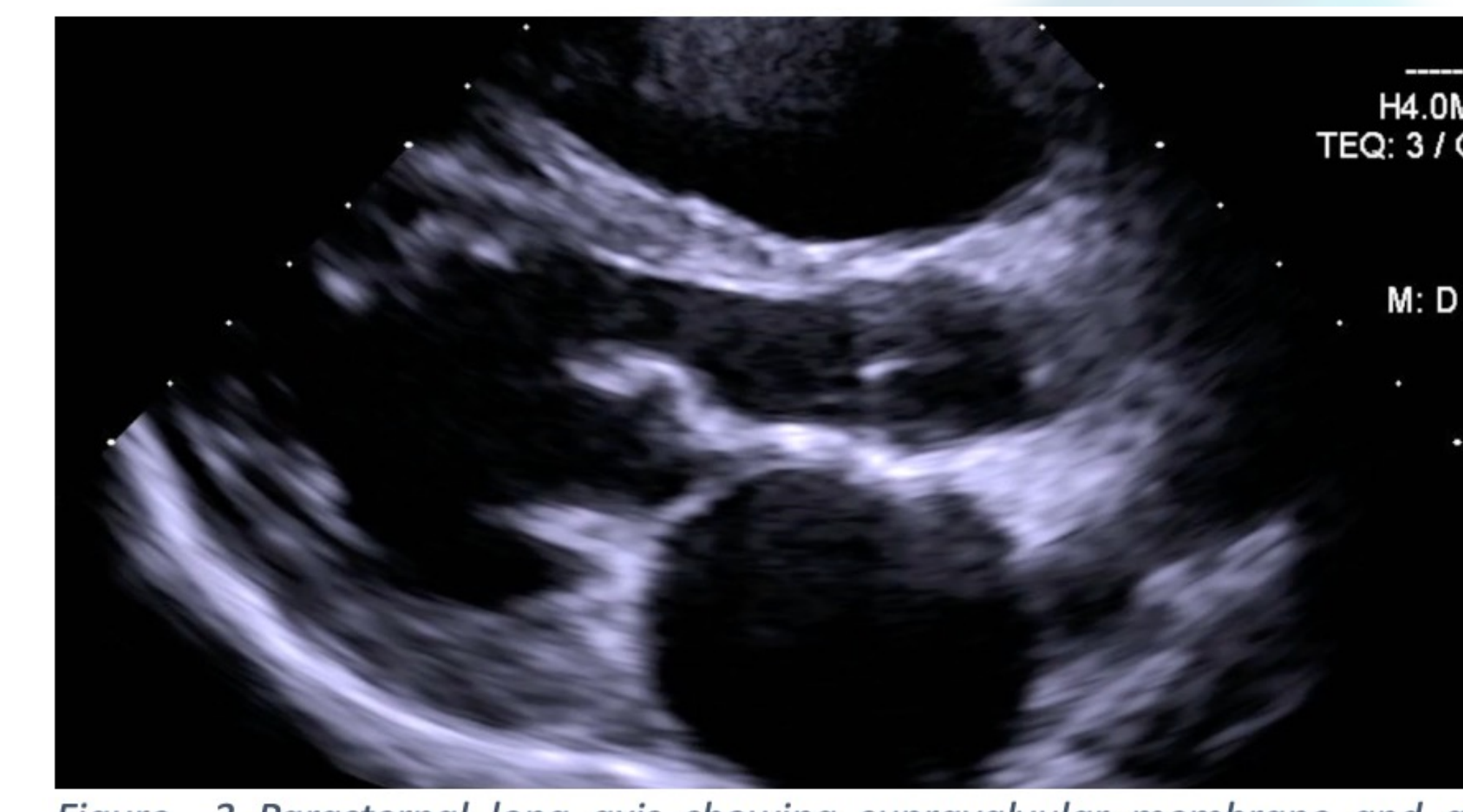


Figure 3 Parasternal long axis showing supra-annular membrane and an enlarged LA