Challenge of a therapeutic sequence: rare case of heart failure in mitral valvular disease intensified by extreme mediastinal shift from major diaphragmatic eventration.

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Extreme mediastinal shift due to major diaphragm eventration is complex when mitral valve repair is required. We report the case of a 59-year-old woman with diaphragmatic eventration presenting 2 recent episodes of heart failure due to arrhythmia associated with severe mitral valve regurgitation (SOR 47 mm²). Forced expiratory flow-volume (FEV₁) and vital capacity (VC) were at 32% and 33% respectively, decreasing to 20% and 30% in supine position. We found impossible to first repair the valve because of extreme mediastinal shift and respiratory dysfunction. Therefore, we decided to perform diaphragm plication first followed 3 months later by mitral valve repair. Six months after cardiac surgery, the patient showed significant clinical improvement. FEV₁ and VC increased to 58% and 55% respectively. The choice of thoracic and cardiac surgery sequencing was key to improve the patient’s respiratory function and medialise the heart to safely support cardiac surgery.
Introduction

Severe mediastinal shift due to major left diaphragmatic eventration is complex when mitral valve repair is required. We report the case of a patient with major congenital diaphragmatic eventration surprisingly never treated, presenting two episodes of cardiac failure due to arrhythmia and severe mitral valve regurgitation. The challenge was to propose the optimal sequence to perform riskless valvular and diaphragmatic surgery.

Case

A 59-year-old female with long history of cardiac arrhythmia presented two episodes of cardiac failure revealing prolapsed mitral valve causing severe regurgitation (SOR 47mm²). On echocardiography left ventricular function was 52%, left atrium was dilated to 66 mm in diameter and Limit LV: TD diameter was 32 mm²/m². Mitral valve surgery was inevitable. However, it was not feasible straight away because of major diaphragm eventration, as seen on dynamic magnetic resonance imaging (dMRI) of the diaphragm (Fig 1), leading to complete mediastinal shift. The patient had severe dyspnea for limited efforts as the result of combined cardiac and diaphragmatic diseases. Forced expiratory flow-volume (FEV$_1$) and vital capacity (VC) were measured at 32% and 33% of theoretical values respectively, decreasing to 20% and 30% in supine position. We debated on the best sequence to safely perform the two procedures in this adult patient. One-step surgery (diaphragm and cardiac) including our technique of diaphragm plication added with prosthesis was considered impossible through median sternotomy. We thought that cardiac surgery first was too risky because of major mediastinal shift and important lung compression. So, we decided to perform diaphragm plication first considering the possibility to quickly perform cardiac surgery if necessary. Surgical postoperative course was uneventful apart from left basal pneumonia. Three months later, cardiac surgery was performed including mitral valve repair associated with annuloplasty on both mitral and tricuspid valves. Cryoablatherapy around pulmonary veins was also associated. The postoperative course was marked by reversible cardiogenic shock and which required temporary NIV.
Six months after the second surgical procedure, the patient totally recovered. The cardiac and pulmonary results were excellent with normalised cardiac rhythm and respiration (Film 1). The patient is now able to walk for several hours as her breathing improved.
diaphragm is in a normal position as seen on dMRI of the diaphragm (Film 2). FEV1 and VC were 58% and 55% respectively. The patient gave her written informed consent for this report.

**Discussion**

In this exceptional case we report the clinical strategy to cure the mitral valve disease reducing first the mediastinal shift by diaphragm plication. This sequence was challenging but most likely the only possible strategy. A safe cardiothoracic environment was needed for the first step of diaphragmatic surgery. Similar cases in adult patients were not found in the literature. It is probably the first case of heart failure due to severe mitral valvular disease, complicated by an extreme mediastinal shift due to major left diaphragmatic eventration. The plication rebuilds a strong thoraco-abdominal frontier and removes atelectasis, which increases lung volume even in almost complete collapsed lung [1,2]. Therefore, the breathing is improved with excellent long-term results [1,2]. There are several approaches to perform diaphragm plication but, in our case, major mediastinal shift contraindicated robotic or thoracoscopic surgery [2,3]. This first step was uneventful apart from postoperative non-severe pneumonia. Such a complication is common after diaphragm surgery, due to compressed lung re-expansion [1,2]. No cardiac surgery was immediately required after diaphragm plication. The patient’s mitral valve could therefore be repaired in optimal conditions. Our therapeutic sequence proposal may be applied in any other association of diseases involving hemidiaphragm dysfunction. The main interest of this therapeutic sequence was to improve the patient’s respiratory function to support another major surgery.

**Conclusion**

The choice of thoracic and cardiac surgery sequencing was the key to improve the patient’s respiratory function and medialishe the heart to safely support cardiac surgery 3 months later.
References


Legend

Figure 1: Pre-operative Dynamic Diaphragmatic dynamic MRI (sequence Fiesta gradient echo steady state GE Architect 3T 2019). A: Axial view mediastinal shift with major left atrial and ventricular dilatation. B: Sagittal view Major left diaphragmatic eventration C: Coronal view Mediastinal Shift
Film 1: Pre-operative Dynamic Diaphragmatic dynamic MRI Axial view MRI sequence Fiesta Gradient echo steady state GE Architect 3T 2019. Mediastinal shift with major mitral valve insufficiency (off-center dephasing regurgitation flow) leading to left atrial and ventricular dilatation.