Chordal Systolic Anterior Motion of the Mitral Valve in Dextro-Looped Transposition of the Great Vessels After Mustard Procedure

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Patient: Male, 41-year-old
Final Diagnosis: Chordal SAM
Symptoms: Dyspnea • dyspnea on exertion • fatigue
Medication: —
Clinical Procedure: Alfieri stitch • Mustard procedure • subpulmonic stenosis resection
Specialty: Cardiology

Objective: Congenital defects/diseases
Background: D-transposition of the great vessels (D-TGA) was once a fatal diagnosis within the first year of life. The Mustard and Senning procedures were invented to redirect the blood flow via intra-atrial baffles. The complicated nature of the clinical course and presence of chordal systolic anterior motion of the mitral valve in a patient with D-TGA and prior subpulmonic resection and Alfieri stitching is presented.

Case Report: A 41-year-old man presented to the clinic with a chief concern of dyspnea on exertion and chronic chest pain. Diagnosed with D-TGA as an infant, he underwent balloon septostomy and later a Mustard procedure at 3 months of age and subpulmonic resection and Alfieri stitching as an adolescent. The patient now presented with transthoracic echocardiogram-revealed severe turbulence in native left ventricular outflow tract to the pulmonary circulation. Doppler velocities indicated this was originating from chordal systolic anterior motion of the mitral valve.

Conclusions: This case reinforces the need for practitioners caring for such patients to become familiarized with and educated in the field of adult congenital heart disease, as patients once plagued with shorter life expectancies are living longer. Repeat surgical intervention or catheter-based therapies may be considered in the future should medical therapy fail to control our patient’s symptoms. A multidisciplinary approach and further monitoring of these patients for best practice guidelines would be ideal and beneficial for the patients and practitioners alike.

Keywords: Arterial Switch Operation • Chordae Tendineae • Mitral Valve • Systolic Murmurs • Transposition of the Great Arteries, Dextro-Looped 1 • Ventricular Outflow Obstruction

Abbreviations: D-TGA – D-transposition of the great arteries; SAM – systolic anterior motion; chordal SAM – chordal systolic anterior motion; LVOT – left ventricle outflow tract; RVOT – right ventricle outflow tract; LVOTO – left ventricular outflow obstructions; HCM – hypertrophic cardiomyopathy; MRI – magnetic resonance imaging; EKG – electrocardiogram

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Background

Unpalliated D-transposition of the great vessels (D-TGA) was once a lethal diagnosis reaching upwards of 90% mortality during the first year of life. In the 1970s, the Mustard and Senning procedures were invented to redirect the blood flow from the atria by using pericardium or atrial tissue, respectively [1]. The preservation of the patent ductus arteriosus via prostaglandin E1 and atrial septostomy provided increased survival due to adequate shunting and atrial redirection of blood flow [2]. These techniques were superseded in 1975 with the arterial switch operation, such as the Jatene procedure, which became the criterion standard owing to better survival.

Subpulmonic stenosis can occur due to abnormal ventricular geometry and compression by the enlarged right ventricle. Subpulmonic stenosis is found upon inception of the diagnosis in 5% to 10% of all patients, second to the identification of ventricular septal defects in approximately 40% of all cases [1,2]. Chordal systolic anterior motion (SAM) of the mitral valve, also known as the pulmonary atrioventricular valve, has been reported due to a change in the ventricular geometry but has not been well described in the literature in patients with corrected D-TGA. Management of such cases is an uncommon occurrence in most adult cardiology clinics.

Case Report

A 41-year-old man presented to the outpatient cardiology clinic with a chief concern of shortness of breath with physical exertion and ongoing chronic chest pain that had been going on since childhood. After further conversation, he revealed that he had also been experiencing infrequent palpitations. He had not experienced any loss of consciousness, lower extremity edema, nor abdominal distension. He was diagnosed with D-TGA as an infant and underwent balloon septostomy and later a Mustard procedure at 3 months of age. He was followed at routine intervals during childhood for residual moderate subpulmonic stenosis. In 2003 at age 24, the subpulmonic stenosis was found to require surgical intervention and was subsequently resected, and a concomitant Alfieri stitch was sutured to the anterior and posterior mitral valve leaflets. After the procedure, echocardiographic findings included mild subpulmonic stenosis with trivial severity of mitral and tricuspid regurgitation. Prior cardiac magnetic resonance imaging (MRI) studies from 2010 were reviewed. He was treated withenalapril and digoxin throughout his childhood and from adolescence into adulthood. He had gained about 18 kg above his ideal body weight during this time. Due to relocation, the patient was lost to cardiology follow-up and presented to establish care after about 9 years of stable condition, with the chief concern of increasing dyspnea. On clinical examination, he had a high-pitched, crescendo-decrescendo murmur best heard at the right sternal border and made worse with deep inspiration.

An exercise treadmill test was performed given his dyspnea and angina. The patient was found to have good exercise capacity, completing 8 min and 6 s on the Bruce protocol, achieving 10.10 metabolic equivalents. An abnormal baseline electrocardiogram (EKG) was maintained throughout the exercise treadmill test, most likely because of his congenital heart disease. The baseline EKG revealed normal sinus rhythm with a first-degree atrioventricular block and a right ventricular strain pattern (Figure 1). He was then sent for a Lexiscan nuclear stress test, which revealed appropriate myocardial perfusion with uniform uptake of Technetium 99m in all walls. A formal sleep study was completed, and there was no overt evidence of obstructive sleep apnea; however, he did have some overnight nocturnal desaturations to an oxygen saturation of 87%.

Transthoracic echocardiography revealed a significantly elevated peak gradient of 90 mmHg and mean gradient of 40 mmHg.
originating from chordal SAM (Figure 2). This was differentiated from recurrent subpulmonic stenosis and the possibility of systemic ventricular expansion of the septum into the pulmonary ventricular outflow tract (Figures 1, 3). Biventricular function was found to be preserved. The Mustard baffle was inadequately visualized. The Pedoff measurements from the right sternal border and suprasternal notch suggested the pulmonary circuit ventricle or morphologic left ventricle pressure was severely elevated. The peak velocity was over 4 m/s, and the peak gradient was over 64 mmHg. The reason for SAM was most likely postoperative changes that brought the chordae toward the left ventricle outflow tract (LVOT), as there was no evidence of significant recurrent subpulmonic stenosis.

He was referred out to the adult congenital heart disease clinic for further evaluation, and a cardiac MRI was ordered. Once all diagnostic studies have been ascertained, the decision for catheter-based therapies and repeat open-heart surgery would both be potential valid options if conservative/medical therapy fails to manage his symptoms. Unfortunately, the patient failed to obtain his cardiac MRI due to the inability to have his MRI scan performed locally.

Discussion

It is likely that the symptoms experienced by our patient were a result of the outflow tract obstruction due to the chordal SAM of the pulmonary circulation atrioventricular valve. Management of chordal SAM of the mitral valve in corrected D-TGA adult cases is still an area of controversy. The prevalence of D-TGA in today’s population is 0.2/1000, exhibiting a male predominance, with approximately 80% being D-TGA [1,2]. Genetic implications have been postulated, such as Zic3, CFC1, and FoxH1, mutations resulting in altered neural crest migrations and altered anatomical positioning. Nodal signaling, neural crest migration, and left-right patterning have all been postulated as causes [2]. Patients who underwent atrial septostomy and Mustard and Senning procedures are now being seen in adulthood, as demonstrated in our patient’s case.

Cuypers et al reported the long-term survival of the Mustard procedure to be 68% at 39 years after the procedure [3]. The main complication reported in their study was impaired right
ventricular systolic function with an increasing incidence of heart failure and arrhythmias, with no specific mention of morphologic left ventricular outflow obstructions (LVOTO) [3]. Stewart et al reviewed data on 8 patients with transposition of the great arteries and LVOTO. Preoperative evaluation via a left ventricular angiogram and postoperative echocardiogram on a subset of these 8 patients was significant for posterior or septal bulging and SAM of the mitral valve [4]. SAM of the mitral valve or chordae tendineae can be a potential cause of LVOTO [5,6]. When SAM occurs, the LVOT narrows, which results in blood being transported through a smaller cross-sectional area, decreasing static pressure. A proposed mechanism of SAM of the mitral valve is the increased forces acting to draw the mitral valve anteriorly, which can be explained by the Venturi effect [5]. The exact mechanism of development of SAM following congenitally corrected transposition of the great arteries has not been completely explained; however, there is evidence that the Mustard procedure itself increases the risk, compared to anatomical surgical correction [7]. The driver of isolated chordal SAM is not known [5]. This phenomenon is most commonly observed in hypertrophic cardiomyopathy (HCM), with the mitral valve more likely to be the culprit than the chordae tendineae [6]. Causes of SAM of the mitral valve not due to HCM include diabetes mellitus, post-myocardial infarction, and hypertension [5]. Pearson et al studied the prevalence of chordal SAM in patients without HCM and found that one-third of patients had idiopathic chordal SAM and another third had associated mitral valve prolapse [8].

Symptoms of chordal SAM are analogous to those of LVOTO, including fatigue, dyspnea on exertion, and chest pain, all of which our patient had been experiencing. The altered pressure hemodynamics cause the septum to protrude posteriorly, while the anterior mitral leaflet moves anteriorly during systole. Our patient had not experienced any loss of consciousness, lower extremity edema, or abdominal distension. It is likely that the symptoms experienced by our patient were a result of the chordal SAM in the absence of obstructive coronary artery disease or HCM. Aortic insufficiency and coronary obstruction are also concerning for all patients with D-TGA. In patients with moderate to severe chordal SAM, there may be inherent consequences if left uncorrected. Exercise-induced hemodynamic abnormalities would include lower cardiac indices before and after exercise due to the increased gradient depending on the severity of the ventricular outflow obstruction, much like what one would expect in significant HCM. Long-term pressure overload, much like what would be expected with a chronic native aortic stenosis to the systemic aortic valve, can lead to myocardial damage evident as myocardial fibrosis.

The mainstay of modern treatment for SAM is medical management. In a review of 174 cases of SAM in a cohort of 2076 patients, all but 4 were able to be managed medically with beta-adrenoceptor blockade, volume loading, and phenylephrine during the study period. Late follow-up showed almost complete resolution without residual SAM [9]. In patients requiring surgical management, there are many potential techniques in the setting of chordal SAM; however, there are 4 primary techniques: the loop technique, chordal translocation, resection-plication-release, and myectomy-loop technique. In the loop technique, the height of the mitral leaflets is adjusted. In chordal translocation, the coaptation point is adjusted. In the myectomy-loop technique, the height of the mitral leaflets is adjusted and the coaptation point is moved. In resection-plication-release, the area of the mitral leaflets is adjusted and the height and coaptation point is moved [5].

The type of surgery depends on the anatomic lesion responsible as well as the severity. Procedures targeting the individual leaflets, coaptation point, and annulus can all be used. Some can be performed at the time of mitral valve repair, after mitral valve repair, and with specific medical conditions, like HCM and Barlow disease. The release of the old Alfieri stitch and suturing of an asymmetric Alfieri stitch may be an option for patients after mitral valve repair. Also, edge to edge repair, posterior leaflet shortening, annular enlargement or plication, and the Pomeroy procedure can be considered as additional alternatives to the asymmetric Alfieri stitch approach. However, since the pathology is predominantly chordal, the loop technique would have been beneficial at the time of the initial mitral valve repair. Aortic insufficiency and coronary obstruction are also concerning for all patients with D-TGA.

The chordal SAM in D-TGA creates an outflow tract obstruction to the pulmonary circuit. This chordal SAM would then look much like pulmonic stenosis or tricuspid SAM into the right ventricular outflow tract (RVOT) in a morphologically normal, situs solitus heart. Although SAM is traditionally associated with the mitral valve, tricuspid SAM has rarely been reported in patients with hypertrophic cardiomyopathy and systemic tricuspid valve replacement after surgically corrected L-GTA [10,11]. In these cases, the SAM of the tricuspid valve caused RVOT obstruction (morphologic LVOT in the case of L-TGA), which is thought to be primarily attributable to drag forces instead of the Venturi effect, as in mitral SAM. This pattern mimics subpulmonic stenosis hemodynamically and typically requires high quality images on echocardiography to differentiate.

Cardiac MRI can help evaluate the obstruction that occurs during systolic excursion of the mitral apparatus. This imaging modality can help differentiate hypertrophic cardiomyopathy, elongated anterior mitral valve leaflet, hyperdynamic left ventricle contractility, and other causes of basal septal hypertrophy or stenosis, such as subpulmonic stenosis. The narrowing can be attempted to be visualized as turbulent flow draws the anterior mitral leaflet or chordae into the LVOT due to the
Venturi effect. The short-axis imaging plane of the narrowed LVOT can be directly measured by planimetry, and peak velocities can also be obtained using through-plane or in-plane phase contrast imaging techniques.

Given the findings on current testing and symptomatology, recommendations of continuing self-restricted activities, prevention of dehydration, and continuation of his current medication regimen were suggested to our patient. If the hemodynamic obstruction of the chordal SAM continues to progress toward higher pressures and severity, despite medical management, the morphologic left ventricle is inherently programmed to sustain such high pressures, unlike the right ventricle in the “S,D,S” normally configured heart. These considerations have not been fully investigated in adults to determine when repeat surgery should be considered for alleviating chordal SAM in patients with previously corrected D-TGA.

Conclusions

In our patient, given that the outflow tract gradient was greater than 50 mmHg, beta-blockade would be prudent to implement as well as preventing dehydration, and lifestyle modifications, including diet and exercise. Cardiac MRI would be indicated; however, this was never obtained by the patient. With the present data, surgical intervention or catheter-based therapies (still experimental) are the 2 most foreseeable options in the future should medical therapy fail to control his symptoms. Further investigations should include a cardiac MRI to aid in further management.

This case reinforces the need for practitioners caring for such patients to become familiarized and educated in the field of adult congenital heart disease; as patients once facing shorter life expectancies are living longer, more functional lives. As most adult cardiologists are not always confident nor equipped or educated to handle such decisions, a multidisciplinary approach and further monitoring of these patients for best practice guidelines would be both ideal and beneficial for the patients and practitioners alike.

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