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EDITORIAL COMMENT

Mitral Valve Repair

When Routine Is Not Routine*

Dimosthenis Pandis, MD,[a] Eric Isselbacher, MD[b]

Remember what a difference has been made by modern anaesthesia, blood transfusion and antibiotics. Looking back on it coldly I was perhaps too adventurous—but it did come off!..

—Sir Henry S. Souttar, CBE, MCh, Correspondence to Brian Blades, MD (1)

When Sir Henry Souttar wrote these lines to Brian Brewer Blades, MD, the 37th president of the American Association for Thoracic Surgery, it was 3 decades after he had successfully split the fused mitral commissures of a 19-year-old girl with his index finger through a controlled incision to the left atrial appendage. It took another 60 years for cardiac surgery to achieve the safe, reproducible, and predictably durable operative outcomes observed in our era of modern surgery. The leap from sporadic digital valve exploration, often fraught with devastating complications, to quaternary specialist centers with dedicated heart teams has been remarkable for allowing the adjudication of patient- and procedure-related risk, which is translated to a care plan adjusted to individual patient characteristics. In the case of complex degenerative mitral valve disease, careful patient selection and a lesion-specific operative approach can achieve near 100% successful repair rates (2), with excellent long-term outcomes, due mainly to judicious preoperative screening and available teams specializing in cardiac anesthesia, intensive care, heart failure, interventional cardiology, and advanced, multimodality imaging (3).

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In this issue of *JACC: Case Reports*, Fabozzo et al. (4) present a case of what initially appeared to be a routine elective repair of degenerative mitral prolapse (if "routine" could describe the appropriate nuance of diversity and unique complexity inherent to degenerative valve pathology; it does not) (4). As such, the vignette describes a young (49 years of age), asymptomatic man recently made aware during a routine physical examination of an incidental, previously unknown systolic heart murmur, subsequently referred to the investigators' service for surgical consultation and further management. The vignette proceeds to report the subsequent echocardiographic evaluation of the mitral valve and left ventricle, which was pertinent for severe mitral regurgitation (MR), left ventricular dilatation and an ejection fraction of 65%, and an anomalous origin and trajectory of the left circumflex coronary artery (LCx), the latter emerging as the key player in the clinical case reported herein. Although the first synoptic lines of the vignette aim to clearly frame the context of the case: an asymptomatic young man with severe MR, leaflet prolapse, a dilated left ventricle, preserved ejection fraction, and the twist (anomalous coronary anatomy), it is important to note that what the investigators are attempting, perhaps fleetingly, is to describe the patient's indication(s) for mitral surgery according to current recommendations for the diagnosis and management of mitral valve disease (5). Although the focus (of this case report) is on the role of the coronary anomaly in the operative planning and surgical strategy, a clear description of the reason to contemplate surgery, as opposed to the alternative of active surveillance, is necessary to educate but also limit any attempt of speculative interpretation by the

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From the [a]Department of Cardiovascular Surgery, Icahn School of Medicine at Mount Sinai, New York, New York; and the [b]Department of Cardiology, Massachusetts General Hospital, Harvard Medical School, Boston, Massachusetts. The authors have reported that they have no relationships relevant to the contents of this paper to disclose.

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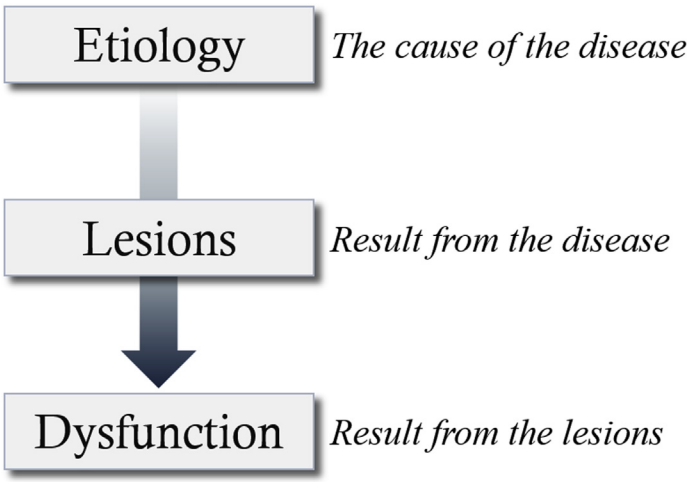
reader. Starting from the referral, it is important to convey whether the interpretation of MR severity from the history, clinical examination, and evaluation of the echocardiographic study by the surgical team concurs with the impression of the referring physician. This is a crucial first step, as discrepancies may result from the highly dynamic nature of the transmitral pressure gradient (lesions notwithstanding), which is susceptible to heart rate, systemic blood pressure, compliance of the left atrium and ventricle, conduction delays, and arrhythmic activity both ventricular and supraventricular. Echocardiographic assessment will also reveal the pathoanatomic lesions and the primary mechanism of MR. And although "prolapse of the x-leaflet segment" may appear to be a sufficiently specific (and commonly used) description, it is not. Although it does describe the primary mechanism (leaflet prolapse) responsible for the observed predominant type of hemodynamic dysfunction (here being regurgitation; the other type is stenosis, not reported here but a possible coexisting finding), it does not convey any information of the valve lesions that cause the leaflet to prolapse, leading to incompetence and ultimately regurgitation. A simple and fail-safe approach to effectively communicate the observed pathology is to use Carpentier's pathophysiologic triad of mitral valve disease (6) (**Figure 1**). An example would be "severe degenerative MR (etiology; also implies primary or secondary etiology) with diffuse thickening, excess leaflet height and volume P2P3, chordal elongation P2P3, chordal rupture P2, chordal thickening and restriction P1 (lesions), with prolapse P2 and restriction P1 (dysfunction)." This approach ensures the accurate description of the lesions, the elements of the mitral valve apparatus involved in the disease, and the functional type of valve dysfunction on the basis of leaflet mobility (normal, excess, or restricted) that may coexist. Echocardiographic assessment can be very useful in detecting most lesions and in determining the mechanism of regurgitation and left ventricular function (on standard 2-dimensional grayscale views), whereas color Doppler 3-dimensional reconstructions can help assess the hemodynamic characteristics of the MR (origin, trajectory, and severity of the regurgitant jet). For the purpose of reporting the findings, it is always useful to include clear still frames of a grayscale view (optimal for anatomy), a color Doppler view (optimal for MR severity), and, when available, a midsystolic 3-dimensional volume rendering of the atrial aspect of the mitral valve (optimal for operative planning), preferably zoomed to avoid obscuring the commissures or cropping or dropouts of the annular circumference.

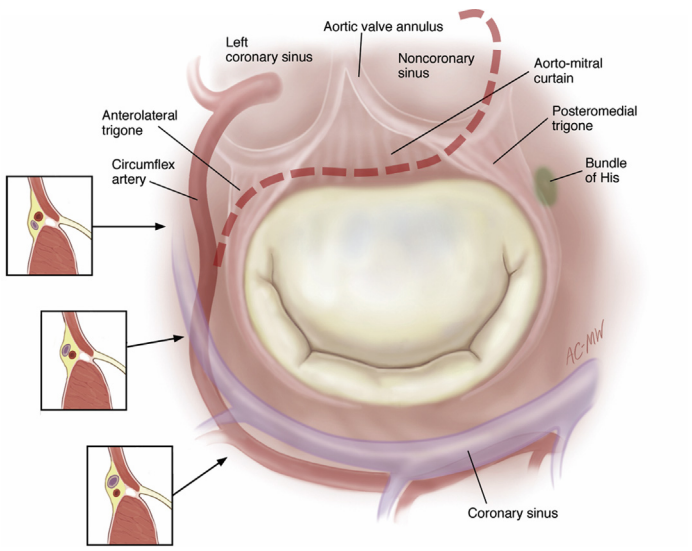
FIGURE 1 Carpentier's Pathophysiologic Triad of Mitral Valve Disease



See text for details.

The clinical vignette becomes intriguing when, during screening transthoracic echocardiography, the investigators noted the presence of an anomalous LCx, which upon further evaluation using fluoroscopy

FIGURE 2 Graphical Illustration of the Anatomic Structures Adjacent to the Mitral Valve With Emphasis on the LCx (Insets)



Red dashed line indicates retroaortic anomalous left circumflex coronary artery (LCx) from case vignette.

and contrast computed tomography was confirmed to be a nondominant circumflex vessel emerging from the opposite aortic sinus, adjacent to a dominant right coronary artery ostium and following a retroaortic course around the anterolateral aspect of the mitral annulus. And although a separate ostium of the left anterior descending coronary artery and LCx from the left sinus of Valsalva is considered a normal variant, an LCx emerging from the right sinus of Valsalva or right coronary artery is the most frequent coronary artery origin and course anomaly observed in published series, with a prevalence of <2% of analyzed angiograms (7). In the absence of additional ostial pathology (i.e., fistula or atresia), this coronary anomaly should not impose limitations on resting or maximal blood flow (8). It does, however, rise to relevance in the context of operative planning, especially in the case of heart valve procedures and most notably in mitral valve surgery, given the proximity of the LCx to the posterior mitral annulus or, in the case of the presented vignette, the anterior mitral annulus as the LCx courses across the aortomitral curtain (**Figure 2**). This exposes the LCx to incremental risk for iatrogenic injury from placement of annuloplasty sutures or because of inadvertent distortion (kinking) of the annulus from undersized complete rings or annular plication, which can be easily avoided with careful intraoperative monitoring (9). Knowledge of the coronary anatomy is, of course, a sine qua non in bypass-assisted cardiac surgery in the context of a safe and effective myocardial protection strategy, as it relates to potential regions of deficient perfusion and/or retrograde filling as well as the anatomic position and structure of the coronary ostia and sinus for the effective delivery of antegrade and retrograde cardioplegia on the basis of the coronary dominance and regional wall distribution. The investigators, following a detailed angiographic assessment and confirming the nonischemic nature of the coronary anomaly (as opposed to a left coronary origin from the pulmonary artery), elected to proceed with a median sternotomy for optimal exposure, which is important not only because of the complex degenerative valve disease but also to allow judicious perioperative assessment for possible additional congenital structural heart lesions in light of the hereditary signature of mitral valve prolapse, even with nonsyndromic cases (10).

Apropos of the concept of lesion-specific strategy, mitral reconstruction requires a plan to stabilize the repair with an annuloplasty prosthesis. The choice of prosthesis should consider the true height of the anterior leaflet, while allowing a deep coaptation away from the left ventricular outflow tract (preventing systolic anterior motion of the anterior leaflet and chordae) and to avoid undue stress on the native structures. And although a variety of options are currently available, a growing body of evidence advocates the use of annuloplasty prostheses with the lowest strain impact on both the annulus and leaflets, depending on the desired effect and native pathology (11). Either way, an overzealous reduction of either the anteroposterior or intercommissural distance (i.e., with rigid complete rings) should be avoided for this reason (12). An additional incentive favoring the use of an annuloplasty band is the presence of a coronary vessel along the anterior or anterolateral annulus, which was underappreciated by the investigators, ultimately leading to the inadvertent ligation of the vessel by an annuloplasty suture. Fortunately, after a period of peripheral circulatory support, the patient made a full recovery, albeit at the expense of a small ischemic sequela. Alas, the scenario may not have been as favorable had an air embolus (even transiently) compromised the right coronary-dependent posterior or inferior wall region, a known perioperative risk during valve surgery that would have been compounded by the suboptimal flow contribution from the anomalous LCx marginal branches. The simple yet effective solution of an annuloplasty band would have minimized the risk for coronary injury without sacrificing the durability of mitral repair.

Recent reports on long-term outcomes from high-volume specialist heart valve centers have evaluated earlier operative practices of nonresection techniques, and/or nonannuloplasty techniques, showing higher incidence of recurrent MR and/or reoperation rates due to disease progression (i.e., chordal elongation, leaflet expansion, and annular dilatation) (13). These results, combined with the limited mid-term and absent long-term data on the remodeling effect of catheter-based therapies on the mitral annulus in unselected samples of degenerative MR patients, should inform the safest approach to ensure the best possible evidence-based repair durability and long-term event-free survival, while mitigating the risk for perioperative injury without exchanging one problem (coronary injury, incomplete repair) for another (unknown outcome without annuloplasty), let alone in a young patient with an expected 25-year (at least) survival, or to phrase it differently, exposure to post-operative morbidity risk from recurrent valve disease. And although the investigators have extensive experience and excellent outcomes with

transcatheter mitral chordae replacement in select patients, the central tenet of surgical repair in the present paradigm should be to choose the strategy with the proven record of patient safety and long-term durability, as opposed to mitigating one risk now in exchange for another, currently unpredictable risk, later.

ADDRESS FOR CORRESPONDENCE: Dr. Dimosthenis Pandis, Icahn School of Medicine at Mount Sinai, Department of Cardiovascular Surgery, 1190 Fifth Avenue, Guggenheim Pavilion, Two-West, New York, New York 10029. E-mail: dimosthenis.pandis@mountsinai.org.

REFERENCES

1. Souttar HS. Correspondence to Blades, B.: intra-thoracic surgery (lungs, heart, and great vessels: surgical management of diseases of the esophagus) 1905–1955. Int Abstr Surg 1955;100:413.

2. Castillo JG, Anyanwu AC, Fuster V, Adams DH. A near 100% repair rate for mitral valve prolapse is achievable in a reference center: implications for future guidelines. J Thorac Cardiovasc Surg 2012;144:308–12.

3. El-Eshmawi A, Castillo JG, Tang GHL, Adams DH. Developing a mitral valve center of excellence. Curr Opin Cardiol 2018;33:155–61.

4. Fabozzo A, Rossi E, Facci G, Bottio T, Gerosa G. Mitral valve repair and anomalous origin of circumflex artery: to ring or not to ring. J Am Coll Cardiol Case Rep 2019;1:503–7.

5. Nishimura RA, Otto CM, Bonow RO, et al. 2017 AHA/ACC focused update of the 2014 AHA/ACC guideline for the management of patients with valvular heart disease: a report of the American College of Cardiology/American Heart Association Task Force on Clinical Practice Guidelines. Circulation 2017;135e1159–95.

6. Carpentier A, Adams DH, Filsoufi F. Carpentier's Reconstructive Valve Surgery: From Valve Analysis to Valve Reconstruction. Philadelphia: Saunders/Elsevier, 2010.

7. Yuksel S, Meric M, Soylu K, et al. The primary anomalies of coronary artery origin and course: a coronary angiographic analysis of 16,573 patients. Exp Clin Cardiol 2013;18:121–3.

8. Angelini P, Velasco JA, Flamm S. Coronary anomalies: incidence, pathophysiology, and clinical relevance. Circulation 2002;105:2449–54.

9. Castillo JG, Sanz J, Fischer GW, Bowman K, Filsoufi F. Management of anomalous left circumflex artery encircling the aortic annulus in a patient undergoing multivalvular surgery. J Card Surg 2009;24:667–9.

10. le Tourneau T, Mérot J, Rimbert A, et al. Genetics of syndromic and non-syndromic mitral valve prolapse. Heart 2018;104:978–84.

11. Bothe W, Rausch MK, Escobar Kvitting J-P, et al. How do annuloplasty rings affect mitral annular strains in the normal beating ovine heart? Circulation 2012;126 11 suppl 1:S231–8.

12. Suh YJ, Chang B-C, Im DJ, et al. Assessment of mitral annuloplasty ring by cardiac computed tomography: correlation with echocardiographic parameters and comparison between two different ring types. J Thorac Cardiovasc Surg 2015;150:1082–90.

13. David TE, David CM, Tsang W, Lafreniere-Roula M, Manlhiot C. Long-term results of mitral valve repair for regurgitation due to leaflet prolapse. J Am Coll Cardiol 2019;74:1044–53.

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