From the transducer to the scalpel: Anatomic correlation between echocardiographic images and surgery findings in native and prosthetic mitral pathology

Del transductor ecográfico al bisturí: correlación anatómica entre las imágenes ecocardiográficas y los hallazgos quirúrgicos en la enfermedad de la válvula mitral nativa y protésica

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SUMMARY

Assessing the anatomy and dynamics of the mitral valve (MV) is essential to the understanding of pathology and planning of therapeutic interventions to improve patient morbidity and mortality. Transthoracic echocardiography (TTE) allows a thorough structural and functional assessment of the MV in real time, and thanks to its reproducibility, and near universal availability, it has become the primary modality employed for MV evaluation.

Due to the tomographic nature of TTE, it is necessary to assess the MV in several transducer positions in order to perform a complete anatomical and functional evaluation. Advanced echocardiographic modalities, such as transesophageal (TEE), are often crucial to determine the etiology, mechanism and severity of mitral valve disease (MVD), as well as to decide the appropriate timing and method of intervention. (1) Moreover, guidelines have proposed that further expansion of this technique would be useful to facilitate surgical decision-making. (2)

The use of three-dimensional echocardiography (3DE) allows rotating and visualizing cardiac structures from any desired perspective, avoiding geometrical assumptions, and providing while providing information of both anatomy and function. (1)

A wide range of structural abnormalities and diverse etiologies can affect the MV. Due to the complex structure and physiology of the MV apparatus, identifying the mechanisms resulting in MV disease is often challenging. Although percutaneous procedures have been developed as an alternative therapy for selected patients with suitable anatomy, surgery remains the cornerstone therapy for native MVD and prosthetic-related complications. (3)

We hereby present a comprehensive case series of different etiologies of native MVD, as well as prosthetic pathology, in which we aim to depict the correlation between the clinical information provided by the different echocardiographic modalities and the anatomical findings seen in the operation room, thereby depicting a "full-picture" of the pathology, from transducer to scalpel.

CASE 1: Degenerative mitral valve disease

50 year-old woman with no prior medical history (PMH), presented to the emergency department with oneweek shortness of breath. She was suspected of having a non-ST elevation myocardial infarction (NSTEMI), but urgent coronary angiography showed no significant epicardial coronary artery disease. During that same procedure, right heart catheterization showed elevated right and left filling pressures, and severe mitral regurgitation (MR). Due to a bad acoustic TTE window a TEE was performed.

In a mid-esophageal 4-chamber view (4ch), the TEE shows a MV flail posterior leaflet (**Fig 1A**, **white arrow**). In a 3DE Zoom acquisition of the MV (en-face view), a P2 flail scallop is depicted (**Fig 1B**, **yellow arrow**), associated with myxomatous changes of the remaining segments (**Fig 1B**, **yellow asterisk**). Color Doppler shows the presence of an anteriorly directed eccentric jet of severe MR (**Fig 1C**, **red arrow**), and corresponding 3D Color Doppler image identifies the exact origin of the MR jet, between the P2-P3 scallops (**Fig 1D**, **red arrow**). In accordance with the echocardiographic findings, surgical images show the mixomatous appearance of the MV (**Fig 1E**), as well as the presence of a ruptured chord (**Fig 1F**, **white arrow**) corresponding to the P2 segment.

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Teaching points: these findings are characteristic of degenerative MVD spectrum, in which two different morphologic phenotypes of have been described: diffuse myxomatous degeneration (Barlow disease) and fibroe-lastic deficiency (FED). (4) This last one is characterized by leaflet prolapse, often isolated to one segment, with chordal rupture or flail leaflet. This isolated segment is often thickened and redundant, whereas the remainder of the leaflet is thin and translucent. (4) From an echocardiographic perspective, measurements of the anterior and posterior leaflet heights and annular diameter are important to determine the appropriate size of the mitral annular band or ring to use. (5) 3D TEE en-face views of the MV allows to assess billowing, prolapse or flail of any leaflet segment or even at the commissures. (4) Although prolapse of the P2 segment is observed in over 60% of patients undergoing surgery, there might be other concomitant prolapsing segments2. Some of the MV repair techniques used in these patients include leaflet resection, plication, artificial neochords, leaflet reduction and annuloplasty band or ring. (5)

CASE 2: Severe mitral annular calcification

63 year-old man with PMH of end-stage renal disease (ESRD), long-term hemodialysis and cerebrovascular accident 2 years prior to admission. One year prior to this echocardiographic study, he was admitted with heart failure symptoms but with preserved ejection fraction. He now presents with progressive shortness of breath and dry cough. TTE and TEE were performed.

TTE parasternal long axis (PLAX) view shows a severely dilated left atrium (LA), and severe mitral annular calcification (MAC) affecting both the anterior and posterior MV annulus (MVA) (**Fig 2A, yellow arrows**). Color Doppler on that same view, shows the presence of flow acceleration towards the MV orifice (Fig 2B, white arrow). This is further corroborated on TEE images, where the flow convergence towards the MV orifice depicts a large proximal isovelocity surface area (PISA) on a mid-esophageal 4ch view (**Fig 2C, white arrow**). These findings, and the presence of a mean MV gradient of 16 mmHg on pulsed-wave Doppler (PW) (**Fig 2D**) are consistent with the presence of severe MS. En-face 3D Zoom rendering of the MV, allows visualization of the circumferential MAC distribution (**Fig 2E, dashed white line**) and the severely reduced size of the MV orifice in early-to-mid diastole (white asterisk). Fig 2F demonstrates the high correlation between surgical findings and echocardiographic images, as seen by the anteriorly and posteriorly extended MAC (**dashed white lines**), the commissural involvement (**white arrow**) and the severely stenotic MV orifice (**white asterisk**).

Teaching points: MAC is a chronic degenerative and metabolically regulated process resulting in calcium deposition especially in the posterior annulus, related not only to hemodynamic stress, but also to inflammation, lipid, bone, and mineral altered metabolism. (6) Its prevalence varies from 5% to 42% depending on the imaging modality used, the characteristics of the studied population and their associated risk factors. (7) Patients with ESRD have a particular high risk of MAC (7), which has been found to be independently associated with all-cause and cardiovascular mortality, stroke and arrhythmias (specially atrial fibrillation). (7) Although frequently detected by TTE and TEE, cardiac tomography is useful to qualify and quantify the density, severity, and extension of MAC and its relationship to other cardiac structures. (6) Medical treatment options are limited. Surgical MV repair or replacement can be challenging, as it usually requires extensive operating time for decalcification and reconstruction of the MVA.



Case.1



Case.2

CASE 3: Mitral Fibroelastoma

38-years-old man with no PMH presents with two transient episodes of right arm and leg weakness 48 hours prior to admission. Brain tomography and magnetic resonance images were negative, and a carotid Doppler showed no abnormalities. An electrocardiogram depicted normal sinus rhythm without any rhythm disturbances. A TTE and further TEE were performed.

PLAX and apical 4-chamber (A4ch) views on TTE, show the presence of a thin, highly mobile, echodense structure attached to the ventricular side of the anterior MV leaflet (**Fig 3A and B respectively, yellow arrows**). This is also visualized on TEE imaging, as seen in a mid-esophageal 4-ch view (**Fig 3C, yellow arrow**) and in a 3-ch view, where it can be observed that this structure is being pulled into the left ventricular outflow tract during systole (**Fig 3D, yellow arrow**). Images obtained during the surgical resection of the mass, show high correlation with the echocardiographic findings, in terms of anatomic features and location (**Fig 3E**). Pathology allowed the establishment of the diagnosis of MV papillary fibroelastoma (PFE).

Teaching points: PFE is the third most frequent tumor of the heart after atrial myxoma and lipoma, and is the most frequent tumor of the cardiac valves (8). PFE are single in 90% of cases and more than 95% are located in the left heart (8). Although histologically benign, PFE is a dangerous condition due to the potential for cerebral and coronary embolization. (9) Two thirds of cases are fortuitously diagnosed by routine TTE, and one third are diagnosed after an embolic event that triggers exploration. (9) Surgical resection is recommended to avoid embolic complications; this constitutes definitive treatment as recurrence is very rare. (8)

CASE 4: Bioprosthetic Thrombosis

70 year-old woman with PMH of hypertension, chronic kidney disease, and MV replacement three years ago with a bioprosthetic St. Jude Epic 29 mm valve presents with progressive dyspnea on exertion, chest tightness and palpitations. NSTEMI was ruled out according to guidelines. (9) Due to bad acoustic window, a TEE was performed.

Mid-esophageal 4ch view shows restricted opening of the MV bioprosthesis and the presence of a hyperechogenic structure attached to the ventricular side of the MV bioprosthetic leaflets (**Fig 4A**, **yellow arrow**). Color Doppler showed the presence of acceleration flow towards the MV inflow (**Fig 4B**, **white arrow**). En-face 3D Zoom image of the MV bioprosthesis allows visualization of the restricted motion of two of the leaflets (**Fig 4C**, **white asterisks**), while only one was fully opening (**Fig 4C**, **dashed white lines**). PW Doppler mean transmitral gradient of 12 mmHg, suggests the presence of severe MS (**Fig 4D**). Once the bioprosthesis was removed, the restricted motion of two of the leaflets is tested by the surgeon (**Fig 4E**, **white asterisks**), showing that only one of the leaflets has preserved excursion (**Fig 4E**, **dashed white lines**). When the excised bioprosthesis is seen from its ventricular side, organized thrombi involving both immobile leaflets is seen (**Fig 4F**, **yellow arrows**). **Teaching points:** Bioprosthetic MV thrombosis should be suspected when there is a restricted motion of the prosthetic leaflets. In cases presenting with heart failure, large clot burden (>0.8 cm2) or coronary artery disease, particularly when fibrinolysis is contraindicated, cardiac surgery is preferred. (11) 3DE analysis of the MV from the surgeon's perspective (as seen from the LA) provides additional information for the diagnosis. 3DE multi-planar reconstruction can also be used to better visualize the pathology in multiple anatomic planes.





Case.4

CASE 5: Paravalvular Leak

62-year-old man, with PMH of chronic obstructive pulmonary disease, long-standing atrial fibrillation and bioprosthetic MV replacement 3 years ago presents with fever. One month ago, he was hospitalized with a diagnosis of sepsis due to a multiresistant E.coli and was treated with Ertapenem. He now presents with progressive shortness of breath and dyspnea on exertion. Hemoglobin at admission was 6.9 gr/dL, requiring blood transfusions to maintain a hemoglobin > 7.0 gr/dL and signs of hemolysis were noted on a blood smear. A TEE was performed.

In a mid-esophageal 4-ch view, there is a large posterior dehiscence of the bioprosthetic annulus (**Fig5A**, **yellow arrow**). Color Doppler corroborated the presence of a severe regurgitant jet across this gap, which extends along the posterior LA wall, creating the Coanda effect (**Fig 5B**, **white arrow**). These findings are consistent with a large paravalvular leak (PVL). En-face 3DE Zoom acquisition allows a better depiction of the bioprosthetic valve (**Fig 5C**), and the use of 3D Color rendering, provides additional information regarding the extension of the paravalvular defect (**Fig 5D**, **white arrow**). During surgery, the surgeon observes a complete posterior dehiscence (**Fig 5E**, **yellow arrow**. White dashed lines correspond to the virtual extension of the Lahey clamp underneath the prosthesis). After the surgeon lifts up the bioprosthesis, the real dimensions of the defect can be better observed (**Fig 5F**, dashed white lines).

Teaching points: PVL, which occurs in nearly 3% of surgical MV replacements (12), should be suspected in patients with prosthetic valves who present with heart failure symptoms, anemia and/or evidence of hemolysis on their blood smear. Valvular dehiscence and excessive prosthetic rocking are usually associated with active or prior endocarditis or other systemic inflammatory disorders. (12) Imaging after MV interventions includes careful evaluation of device integrity, position, stability, and interaction with adjacent structures. (13) Color Doppler assessment of MR severity with TTE is challenging due to interaction of the device or adjacent structures with the MR jets, and the frequent presence of multiple MR jets, often crisscrossing in different planes (13). Although 3DE is critical for identifying the origin and extent of the defect, a comprehensive approach that integrates several parameters of valve morphology and function is key to appropriately detect and quantitate prosthetic dysfunction. The treatment of patients with severe PVL and NYHA III/IV heart failure requires surgical or percutaneous intervention. (11) The type of prosthesis, patient's surgical risk, and feasibility of transcatheter approach better, should determine whether surgery of percutaneous treatment is more appropriate.



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