Mitral Valve Prostheses
Aortic Stenosis, Spectrum of Disease, Low Flow, Low Gradient, and Different Variants
Aortic Stenosis, Bicuspid Valve, and Aortic Root Dialation
Hypertrophic Cardiomyopathy or Phenocopy
Systemic Disease

Participate in Board Questions

1. Select Agenda
3a. Ask Questions
3b. Select Practice Board
4. VOTE
Echocardiographic Evaluation of Mitral Valve Prostheses

Dennis A. Tighe, MD, FASE

Case 1
History

• A 54 year-old woman with hypothyroidism presents to her PCP with worsening of shortness of breath.
  – Systolic and diastolic murmurs are auscultated.
  – Transthoracic echocardiography is requested for further evaluation.
Q1. Echocardiography confirms the presence of aortic stenosis (orifice area 0.6 cm²) and identifies the presence of moderate aortic regurgitation. Mitral valve thickening is also observed. Which of the following constitutes the most likely etiology for the valvular abnormalities:

• A. Age-related degenerative valve disease
• B. Rheumatic heart disease
• C. Annular calcific disease
• D. Carcinoid heart disease
• E. Radiation-associated valve disease
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Q2. Which of the following conditions would be an expected complication resulting from the disease process causing these left-sided valvular abnormalities?

- A. Flushing
- B. Constrictive pericarditis
- C. Coronary artery spasm
- D. Hypertrophic cardiomyopathy
- E. Cardioembolic stroke
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Radiation-Associated Valve Disease

• Frequent complication
• Regurgitant lesions > Stenotic lesions
  – Left sided > right sided
• Risk greater with ≥30 Gy
• Women > men
• Suggestive echocardiographic appearance
  – Calcification and thickening of aortic-mitral curtain
  – Anterior changes more profound than posterior (vs MAC)
  – No leaflet doming/commissural involvement (vs RHD)
  – Aortic root calcification increases the likelihood
• Progressive
• Periodic screening required
Radiation Therapy

- Cardiovascular complications
  - Coronary artery disease
  - Cardiomyopathy
    - Restrictive or dilated
  - Pericardial effusion
  - Constrictive pericarditis
  - Conduction system/arrhythmias
  - Valvular heart disease
  - Carotid artery disease

Case 2
Based on this M-mode tracing, which of the following findings is *unlikely* to be present?

- A. Restrictive mitral inflow pattern
- B. Soft S1
- C. Diastolic mitral regurgitation
- D. Premature closure of the aortic valve
- E. Brief diastolic murmur
Choice Explanations

• D. Premature closure of the aortic valve. *This is the correct answer.*
  – This M-mode tracing displays premature closure of the mitral valve along with high frequency diastolic fluttering of the anterior mitral leaflet (and the interventricular septum). This constellation of findings occurs when acute, severe aortic regurgitation is present.
  – This answer is false because the aortic valve is incompetent. With the rapid rise in LV diastolic pressure characteristic of this lesion, premature opening of the aortic valve may be observed.

• A. Restrictive mitral inflow pattern. This answer is true due to the rapid increase in LV diastolic pressure characteristic of acute severe AR.

• B. Soft S1. This answer is true because the rapidly rising LV diastolic pressure leads to premature closure of the mitral valve.

• C. Diastolic mitral regurgitation. This answer is true. Rapid increases in LV diastolic pressure can lead to transient reversal of the LA-LV pressure gradient in diastole and the occurrence of (low velocity) diastolic mitral regurgitation.

• E. Brief diastolic murmur. This answer is true. The regurgitant murmur is brief in duration because the aortic diastolic pressure rapidly equilibrates with that of the LV.

Case 3
History

• A 56-year old woman with shortness of breath and the finding of right heart volume overload on outside echocardiography due an ostium secundum ASD is referred for percutaneous closure. RHC revealed PASP 29 mm Hg and PVR <2 WU. Non-obstructive CAD was found.
• TEE performed at the time of the procedure confirms the presence of the ASD and documents adequate surrounding rims and appropriate PV drainage. However an incidental finding is made.
Q1. Based on this finding which of the following courses of action should you recommend?

A. Inform your interventional colleague to cease the procedure immediately and discuss the finding with the patient and her family.
B. Obtain an urgent CT surgery consult.
C. Provide additional antibiotic coverage for oral cavity organisms given a high-risk of infective endocarditis.
D. Continue with the planned procedure and discuss the incidental finding and its implications with the referring cardiologist.

Q1. Based on this finding which of the following courses of action should you recommend?

A. Inform your interventional colleague to cease the procedure immediately and discuss the finding with the patient and her family.
B. Obtain an urgent CT surgery consult.
C. Provide additional antibiotic coverage for oral cavity organisms given a high-risk of infective endocarditis.
D. Continue with the planned procedure and discuss the incidental finding and its implications with the referring cardiologist.
Q2. Which of the following statements concerning this valvular abnormality is correct?

A. Aortic stenosis is the predominant hemodynamic abnormality.
B. Aortic regurgitation is the predominant hemodynamic abnormality.
C. Long-term survival with this condition is poor.
D. Aortic dilatation is not commonly found in association with this lesion.
E. Aortic dissection is strongly associated with this valvular abnormality.
Quadricuspid Aortic Valve

- Rare congenital abnormality (<0.05% frequency)
  - Recent single-center review showed frequency of 0.006% of all echocardiograms
- Hurwitz and Roberts classification
  - Types A and B most frequent

![Aortic Valve Findings Table]

* Aortic regurgitation is the predominant hemodynamic abnormality
  Aortic stenosis present in only 8%

Associated Findings

• Aorta
  – Dilatation 29%
    • Usually mild
• Other cardiac lesions
  – MVP/bowing
  – TVP
  – Pulmonary valve stenosis
  – ASD
  – VSD
  – Coronary anomaly

Survival

16% required surgery
Case 4

History

• Echo number 25 of a long day.
• A 22 year-old man previously cared for in pediatric cardiology clinics is referred by his new cardiologist for echocardiography to evaluate mitral regurgitation and LV function.
• He has seen multiple subspecialists over the years. The patient has ESRD and was recently initiated on HD. He has poorly-controlled HTN and his CBC/diff is distinctly abnormal.
LAVI = 39 mL/m²

GLS = -12%

RV-RA grad

Pulm vein

4 years prior
Q1. Based on the available data, how would you best characterize his left heart function?

A. Normal LVEF, elevated mean LAP, elevated LVeDP
B. Normal LVEF, elevated mean LAP, normal LVeDP
C. Normal systolic function with normal filling pressures.
D. Normal systolic function with elevated filling pressures.
Q2. Which of the following conditions is the most likely etiology for the observed echo findings?

- A. Amyloidosis
- B. Apical hypertrophic cardiomyopathy
- C. Hypertensive heart disease
- D. Hypereosinophilic syndrome
- E. Rheumatic heart disease

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- E. Rheumatic heart disease
Lab Studies

• BMP
  – Na 139; K 3.7; Cl 100; Co2 29; BUN 15; Cr 6.4; eGFR 11.

• CBC/diff
  – WBC 12.7; Hgb 8.5; Hct 25.7.
  – PMN 18%; Lymph 13%; Eos 64%; Baso 2%.

Hypereosinophilic Syndrome

<table>
<thead>
<tr>
<th>Stages of cardiac involvement in HES</th>
<th>Characterisation of stage</th>
</tr>
</thead>
<tbody>
<tr>
<td>Acute necrosis</td>
<td>Eosinophilic myocarditis with eosinophilic and lymphocytic infiltration. Myocardial necrosis and apoptosis with rare microembolic phenomena. Typically no other cardiac symptoms. Rarely can exhibit a fulminant course.</td>
</tr>
<tr>
<td>Thrombotic stage</td>
<td>Thrombus along damaged endocardium. Thrombi within apices of one of both ventricles, and can encroach on base of the heart into the subvalvular regions. Embolic phenomena can occur.</td>
</tr>
<tr>
<td>Fibrotic stage</td>
<td>Thrombi are replaced by fibrosis. Scarring occurs. Restrictive cardiomyopathy with signs and symptoms of left or right sided heart failure. Fibrosis that occurs at the base of the heart can lead to valvular regurgitation.</td>
</tr>
</tbody>
</table>

HES, hypereosinophilic syndrome.

As recommended by the 2014 AHA/ACC Valvular Heart Disease Guideline, which of the following statements regarding follow-up of prosthetic heart valves by echocardiography is true?

- A. Annual TTE is reasonable starting at 5 years following mechanical valve replacement.
- B. An initial TEE should be performed routinely to assess valve hemodynamics within 2 months of implantation.
- C. Change in clinical status should prompt early echocardiography.
- D. Annual TTE is reasonable starting at 5 years following bioprosthetic valve replacement.
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Which of the following statements regarding the obstructed/thrombosed prosthetic heart valve is correct?

- 1. A PHT > 130 msec is the single best indicator of prosthetic mitral obstruction.
- 2. Taking into account heart rate is not necessary when assessing trans-mitral gradients.
- 3. Pannus in-growth is more common in the aortic position than with mitral PHVs.
- 4. A peak velocity ≥2.5 m/sec suggests significant aortic PHV stenosis.
- 5. Randomized, controlled trials have demonstrated that bolus infusion of rt-PA is the fibrinolytic regimen of choice.
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Which of the following statements concerning prosthetic heart valve regurgitation is correct?

• A. Pseudo-regurgitation is an issue most often encountered during performance of TEE.
• B. Any degree of regurgitation indicates dysfunction of a mechanical valve.
• C. Structural valve deterioration is an uncommon cause of pathological regurgitation.
• D. Mitral bioprostheses are less prone to suffer structural valve deterioration than are aortic bioprostheses.
• E. Annular dehiscence most often is a consequence of infective endocarditis.
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What can lead to **underestimation** of the aortic valve peak gradient on echo as compared with invasive hemodynamics?

1. Pressure Recovery
2. Equating Peak Instantaneous Gradient To “Peak-to-peak” Gradient
3. A Large Incident Angle To The Aortic Outflow
4. Failure To Account For High Subvalvular Flow
5. Low Stroke Volume
Echo AoV PG vs. Cath/Hemo AoV PG

1. Pressure Recovery – **Echo PG HIGHER**
2. Equating Peak Instantaneous Gradient (Echo) To “Peak-to-peak” (Hemodynamic) Gradient - **Echo PG HIGHER**
3. A Large Incident Angle To The Aortic Outflow – **Echo PG Mis-measured LOWER**
4. Failure To Account For High Subvalvular Flow – **BOTH Echo And Hemo Pg’s Elevated**
5. Low Stroke Volume – **BOTH Echo And Hemo Pg’s Lower**

Review Question #2:
*Reflect upon the image below*

**Transesophageal (TEE):**
Which Of The Following Statements Best Describes This Aortic Valve:

1. Unicuspid - Single Commissure
2. Bicuspid - Fusion Of Left & Right Cor. Cusps
3. Bicuspid - Fusion Of Left & Noncoronary Cusps
4. Functionally Bicuspid Aortic Valve (Trileaflet)
5. Cannot Be Determined
A patient presents with the following echo findings:

- **LVOT diameter** = 2.0 cm
- **LVOT velocity** = 130 cm/s
- **Aortic velocity** = 4.1 m/s

2D: Moderately calcified AV, Normal LVEF (70%)

The aortic valve area is most likely:

1. Normal
2. Mildly Reduced
3. Moderately Reduced
4. Severely Reduced
5. Cannot Be Calculated (Incongruent Units)
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\[ \text{DI} = \frac{130}{410} \]
\[ \text{DI} = 0.32 \]
• 85 Y/O Woman With H/O “Mild” Aortic Stenosis Presents With Severe Dyspnea
  • Hypoxic On Room Air, 2+ Pitting Edema
  • 3/6 Mid-late Peaking SEM Radiating To Neck

• Cardiac Catheterization
  • Ra = 16, Rv = 97/19, Pa 93/27 (53)
  • Pcwp = 22, Co 3.2 L/Min, CI 1.5 L/Min/M²
  • Arteriography – Minimal Luminal Irregularities
BSA = 2.14

Sv_i = 28 ml
Mean gradient <40 mmHg means:

1. This Is Definitely NOT Severe AS
2. The Gradient Is Low Because There Is Depressed LV Ejection Fraction
3. The Gradient Is Low Because The CW Doppler Was Mis-measured
4. The Gradient Is Low Because Of Low Stroke Volume
5. As Long As Calculated AVA Is 0.8 Cm², The Mean Gradient Doesn’t Matter.
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Low Gradient AS:
There’s a LOT to think about!!

LOW STROKE VOLUME (SV₁)

- DEPRESSED EF
- PRESERVED EF –
- TINY VENTRICLE
- IMPAIRED FILLING
- SEVERE MR
- IMPAIRED RV OUTPUT

INCREASED VASCULAR AFTERLOAD
Marty –
There's an opening on the TAVR schedule next Tuesday, and this lady has TAVR written all over.
YOU know this is severe AS, I know this is severe AS.... MAKE THIS HAPPEN!!!

Marty –
You know that I don't agree with Howard (or anyone) all that often. Despite the PVR of 6.3 WU, I firmly believe that this is predominant pulmonary venous hypertension.
Her PHTN will improve post AVR!!!

The best “next step” for diagnosis is

1. No Further Workup Is Needed.
2. Treadmill Stress Testing With Echo Assessment Of AV At Peak Stress.
3. Dobutamine Stress Echocardiography With Staged Assessment Of SV, Ava And Gradients
4. TEE Assessment Of Aortic Valve Morphology And Planimetry
5. Pulmonary Function Testing To Evaluate Lung Disease As An Etiology For Symptoms
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How to Evaluate Aortic Stenosis, Bicuspid Valve, and Aortic Root Dilation

Amr E. Abbas, MD, FASE

The most common form of bicuspid aortic valve is:

1. Fusion of the LCC/RCC
2. Fusion of the LCC/NCC
3. Fusion of the RCC/NCC
4. Equal distribution of cusp fusion
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The difference between Doppler maximum instantaneous gradient and Catheter peak to peak gradient is due to:

1. Pressure recovery
2. Is due to difference in the timing of the aortic pressure measurement between cath and echo
3. Is due to difference in the timing of the LV pressure measurement between cath and echo
4. Is related to the severity of aortic stenosis
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Discordance between echo and invasive measurements of mean transvalvular gradient is due to:

1. Pressure recovery
2. Eccentric jets
3. Very severe aortic stenosis
4. HOCM
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3. Very severe aortic stenosis
4. HOCM

Hypertrophic Cardiomyopathy or Phenocopy?

Steven J. Lester, MD, FASE
7. 33 year old male with a history of hypertrophic cardiomyopathy presents with increasing symptoms of shortness of breath (NYHA III). There was no continuous wave Doppler interrogation of the left ventricular outflow tract (LVOT) obtained, however, the peak mitral regurgitation velocity was obtained at 720 cm/sec. The systolic blood pressure was 110mmHg. Assuming a left atrial pressure of 15mmHg, what is the LVOT peak instantaneous systolic gradient?

What is the LVOT Gradient?

MR Velocity = 720 cm/sec

Left Atrial Pressure = 15 mmHg

Systolic BP = 110 mmHg

1. 222 mmHg
2. 97 mmHg
3. 112 mmHg
4. 207 mmHg
Estimating LVOT Gradient Using MR Peak Velocity

MR Velocity = 7.2 m/sec  Systolic BP = 110 mmHg

\[ \Delta P \approx 4 \times (7.2)^2 = 207 \]

LVOT gradient \( \approx 112 \)

LV Pressure \( \approx 222 \)

Aortic \( \approx \) Systolic BP = 110

LAP \( \approx 15 \)

Gradient: Amyl Nitrite

BP 119/62 MMHG

112 mmHg

5.3 m/sec
8. You are called to the ICU to perform an echocardiogram on a 72 year old male with HCM being evaluated for hypotension. The arterial line is recording a systolic blood pressure of 89 mmHg. Your echocardiogram records the following continuous wave Doppler profiles from the left ventricle. Assuming a left atrial pressure of 15mmHg you conclude that the arterial line is providing an inaccurate reading and you report an estimated systolic blood pressure of xx mmHg?

1. 137 mmHg
2. 89 mmHg
3. 95 mmHg
4. 122 mmHg
5. 211 mmHg

What Is The Systolic BP?

1. 137 mmHg
2. 89 mmHg
3. 95 mmHg
4. 122 mmHg
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3. 95 mmHg
4. 122 mmHg
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LVOT Obstruction vs Mitral Regurgitation

MR  LVOT

4.3 m/sec 74 mmHg
9. A 71-year-old woman has had a murmur since childhood and was diagnosed with hypertrophic cardiomyopathy. She is increasingly symptomatic with shortness of breath with minimal exertion (NYHA III). She is referred for consideration of surgical myectomy. You perform a transthoracic echocardiogram and from the findings you make the following conclusion?
A. Confirm the diagnosis of hypertrophic cardiomyopathy and proceed with myectomy.
B. The Doppler signal is mitral regurgitation and the patient may not be a candidate for a myectomy.
C. There is valvular aortic stenosis.
D. There is subvalvular aortic stenosis.
9. An 18 year old male had a syncopal episode after a blood donation. Because of this event he presented for a pre-participation medical evaluation prior to playing ice hockey at a division 1 college program. Which echocardiographic finding is most consistent with “athletes heart”?

a. Left ventricular wall thickness to be ≥ 13mm.

b. Left ventricular end-diastolic diameter to be reduced.

c. Left ventricular global longitudinal peak systolic strain to be more negative than (<) -16%.

d. Abnormal left ventricular filling.
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10. Mitral regurgitation that is secondary to systolic anterior motion of the mitral valve has a jet generally travelling in which direction?

a. Directed anteriorly.
b. Directed infero-laterally.
c. Is centrally directed.
d. Often produces two distinct mitral regurgitation jets that flow in opposite directions.
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A 68-year-old man presents with fatigue and abdominal bloating. On cardiac exam, the jugular venous pressure revealed “CV” waves to angle of the jaw. An RV lift is present. There is a grade 2/6 pansystolic murmur at the lower sternal border that gets louder with inspiration. There is a soft systolic ejection murmur and diastolic murmur at the second left interspace. In addition, there is an enlarged and pulsatile liver. Images obtained from his TTE are shown.

Which of the following is the most likely diagnosis?

1. Rheumatic heart disease
2. Carcinoid heart disease
3. Ebstein’s anomaly
4. Endocarditis
What is the most likely etiology of the mixed valve disease in this 56 year old patient with a hx of Hodgkins Lymphoma?
What is the most likely etiology of the mixed valve disease in this 56 year old patient with a hx of Hodgkins Lymphoma?

1. Chemotherapy induced valve disease
2. Radiation induce valve disease
3. Ergotamine induced valve disease
4. Degenerative calcific valve disease

Radiation Induced Cardiac Disease

- Pancarditis: pericardial, myocardial, endocardial/valvular (fibroelastosis)
- Acute pericarditis during therapy
- Delayed pericarditis: constriction, pericardial effusion
- Cardiomyopathy: diastolic/systolic dysfunction
- CAD: intimal proliferation, endothelial dysfunction
- Conduction system defects

<table>
<thead>
<tr>
<th>Condition</th>
<th>Incidence</th>
<th>Median Time After Therapy</th>
</tr>
</thead>
<tbody>
<tr>
<td>Coronary Artery Disease</td>
<td>10.4%</td>
<td>9 Yrs</td>
</tr>
<tr>
<td>Carotid ± Subclavian Disease</td>
<td>7.4%</td>
<td>17 Yrs</td>
</tr>
<tr>
<td>Significant Valvular Disease</td>
<td>6.2%</td>
<td>22 Yrs</td>
</tr>
</tbody>
</table>

Hull MC, et al. JAMA 2003; 290:2831

Radiation Therapy for Hodgkin’s Lymphoma Clinically Significant Valvular Disease

- Aortic Stenosis (48%)
- Mitral Regurgitation (28%)
- Mitral Stenosis (10%)
- Tricuspid Regurgitation (10%)
- Aortic Regurgitation (4%)

Hull MC, et al. JAMA 2003; 290:2831
What is the most likely diagnosis in this 58 yo man with pulmonary infiltrates and syncope?
What is the most likely diagnosis in this 58 yo man with pulmonary infiltrates and syncope?

A. Ischemic Cardiomyopathy  
B. Arrhythmogenic RV Cardiomyopathy (ARVC)  
C. Cardiac Sarcoidosis  
D. Systemic Lupus Erythematosus

Cardiac Sarcoidosis

- Noncaseating granuloma  
- Regional wall motion abnormalities in unusual distribution  
- Heart block  
- Sudden death

Courtesy William Edwards, MD
**Sarcoidosis – Granulomas**

11% right atrium

7% left atrium

73% interventricular septum

96% left ventricular free wall

46% right ventricular wall

Bargout R: Int J Cardio, 2004

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**Sarcoidosis – Echo features**

Echo abnormalities are rare

LV dilatation

LV aneurysms

LV scar RWMAs

Mitral valve regurgitation

Pulmonary hypertension

Cardiac Sarcoïdosis (Echo)

Question 4

33 Year Old Female with Lupus Erythematosus, antiphospholipid antibody positive, negative blood cultures, and multiple strokes

What is the most likely diagnosis?
1. Staph aureus endocarditis
2. Libman-Sacks endocarditis
3. Thickened bicuspid aortic valve
4. Eosinophilic valvulitis
Systemic Lupus Erythematosus  
- Cardiac Involvement

- Pericarditis (fluid ANA+)
- Lupus anticoagulant
- Anticardiolipin antibodies
- Myocarditis
- Coronary arteritis
- Libman-sacks (marantic) vegetations

Question 5
What is the most likely diagnosis in this 35 year old female with a history of migraine headaches and no history of rheumatic fever?

1. Ergot associated valvular disease
2. Aortic and mitral valve endocarditis
3. Parachute mitral with coexistent aortic regurgitation
4. Left sided carcinoid valve disease

Drug-Induced Valvular Disease
Echocardiographic Findings

- Thickening and retraction of valve leaflets or cusps
  - No commissural fusion
  - Reduced mobility, restricted closure coaptation
- Thickened, fused, shortened MV/TV chordal support apparatus
- Variable regurgitation, rarely significant stenosis
MDMA (3,4-Methylenedioxymethamphetamine)  
Echo Findings with “Ecstasy” Abuse

<table>
<thead>
<tr>
<th></th>
<th>MDMA Users (n=33)</th>
<th>Controls (n=29)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Duration of use</td>
<td>6.1 ± 3.4 yrs</td>
<td>0</td>
</tr>
<tr>
<td>Age (yrs)</td>
<td>24.3 ± 3.1</td>
<td>25.6 ± 3.1</td>
</tr>
<tr>
<td>MR ≥ Grade 2/4</td>
<td>4 (14%)</td>
<td>0</td>
</tr>
<tr>
<td>Restricted MV motion</td>
<td>7 (24%)</td>
<td>0</td>
</tr>
<tr>
<td>TR ≥ Grade 2/4</td>
<td>13 (45%)</td>
<td>0</td>
</tr>
<tr>
<td>Restricted TV motion</td>
<td>7 (24%)</td>
<td>0</td>
</tr>
<tr>
<td>AR ≥ Grade 1/4</td>
<td>4 (14%)</td>
<td>0</td>
</tr>
</tbody>
</table>

• PREVALENCE OF MDMA ABUSE: 0.4 – 6% WORLDWIDE

Droogmans S, et al. Am J Cardiol 2007; 100: 1442