

VALVULAR HEART DISEASE - PULMONARY INCOMPETENCE



Mamluk Flask, Glass, gold, and enamel c. 1287, Syrian, (Height 43.5 cm), Mamluk Dynasty, Metropolitan Museum of Art, New York.

This stunning glass enamelled flask, dates from the last quarter of the Thirteenth Century. It is a superb example of the fabulously skilled glassware that was produced in both Egypt and Syria during the Mamluk dynasty of the Thirteenth and Fourteenth centuries. This work was free blown from greenish tinted clear glass and then gilded in gold leaf. The decoration was applied with an extensive palette of oil suspended enamels including red, blue, green, yellow, purple, brown, pink, white, grey-blue and black. The piece is in stunningly good condition, considering it is well over 700 years old.

The figures and decoration are exquisitely rendered in the finest of detail. There are no fewer than fourteen horsemen arranged around the base who carry various weapons and implements of war. A simurgh, or Iranian mythical creature, that had the head of a lion and the body of a bird, gracefully entwines itself around the neck of the flask. Above the horsemen are three intricate medallions of brilliant blue floral motif, each radiating outwards from a central blue star. The work has both Iranian and Mediterranean influences. The overall style closely resembles Iranian enamelled ceramics of the period, but the type of glass used was that seen in the eastern Mediterranean littoral.

Scholars and Artists even today do not understand exactly how the work was constructed, in particular how the glassmaker was able to prevent the enamels from running together during the glazing process. The technical skill of its creator was astonishing, as the gold leaf and various enamels required different temperatures to affix them to the surface of the glass., without the aid of modern electronics and machinery. It is clear that many aspects of its creation were produced by the most consummate arts, now long lost.

In the days before cardiac echocardiography, physicians could assess any manner of heart valve lesions, by means of a device known as a “stethoscope”; now used principally as a symbolic or ceremonial accessory, rather than a useful diagnostic instrument. Indeed these learned physicians of ages past could, by dint of assessing the sounds, clicks, snaps, rumbles, pitch and tone of various “murmurs”, diagnose the exact nature of valvular lesions, either isolated or mixed, as well as give some prognostication on the severity of the patient’s condition. In the 21st century, heart valve lesions are now assessed by means of a host of modern marvels of medical imaging technology, and sadly, the ancient art of stethoscopic auscultation, is, now as lost to us as the exquisite arts of Thirteenth century Iranian and Syrian glass making!

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Introduction

Pulmonary incompetence (or regurgitation) is uncommon.

It is usually remarkably well tolerated when it does occur.

Natural History

Pulmonary incompetence of itself is often well tolerated and the overall natural history and progress will more correlate with the underlying causative pathology.

Causes

1. Chronic pulmonary hypertension from any cause.

These may include:

- Primary pulmonary hypertension.
- Cor pulmonale
- Recurrent pulmonary embolism

2. Post Operative:

- Pulmonary incompetence it is an almost unavoidable result of either surgical or balloon valvuloplasty of valvular pulmonic stenosis or surgical repair of tetralogy of Fallot.

3. **Mild** pulmonary regurgitation may be a normal finding on Doppler echocardiography.

Other causes are rare:

4. Endocarditis.
5. Rheumatic heart disease.
6. Congenital lesions.

Complications

1. Infective endocarditis.

- Note that pulmonary incompetence may also be **caused by** endocarditis, as well as predisposing to it. But overall pulmonary valve endocarditis is rare.
2. Right heart failure:
 - Patients with *longstanding* pulmonary regurgitation may develop significant RV failure.

Clinical Features

The signs and symptoms will depend largely on the underlying pathology and its extent.

Symptoms

The symptoms of severe pulmonary incompetence/ pulmonary hypertension will include:

- Dyspnea
- Fatigue
- Syncope

Signs

1. Pulse:

- May be reduced in severe disease.

2. Blood pressure:

- May be reduced in severe disease.

3. Palpation:

Apex beat:

- Not typically displaced.
- There may be a right ventricular parasternal heave in cases of significant right ventricular strain.

Thrill:

- Not typically detected.

4. Heart sounds:

- An S4 may be heard, (right ventricular strain)
5. Murmur:
- High pitched blowing diastolic murmur best heard at the second left intercostal space.
 - There may be an associated mid-systolic ejection flow murmur.

Investigations

CXR

Check for:

- Cardiomegaly
- Calcified pulmonary valve.

ECG

- Check for signs of RHV and strain.

Echocardiography

- This will confirm the diagnosis/ assess severity/ help determine the presence pulmonary hypertension.

Cardiac MRI

Cardiac magnetic resonance has proven to be a useful tool for evaluating pulmonary regurgitant fraction, RV end-diastolic and end-systolic volumes, and RV ejection fraction.³

Coronary angiography:

Coronary angiography is indicated in selected cases to detect associated coronary artery disease when surgery is planned. Knowledge of coronary anatomy improves risk-stratification and determines whether coronary revascularization is indicated in association with valvular surgery.

Management

1. Medical Therapy:
 - Diuretics may alleviate the symptoms of right heart failure.
2. Surgical Therapy:

- Isolated pulmonary incompetence is well tolerated and surgical intervention will only be necessary in severe cases.

In general terms pulmonary valve replacement is done in patients with NYHA functional class II or III symptoms and severe pulmonary regurgitation.³

For asymptomatic patients, the indications based on regurgitant fraction, RV end-diastolic or end-systolic volume, and RV ejection fraction remain poorly defined.³

3. Antibiotic prophylaxis for surgical procedures, (see latest edition of Antibiotic guidelines)

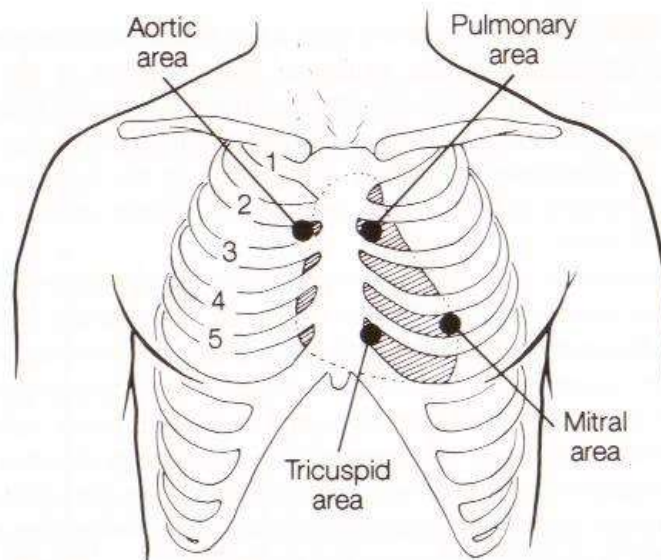
Appendix 1

Grading of the loudness of murmurs:

- Grade 1 *Very soft, requires an experienced listener.*
- Grade 2 *Soft.*
- Grade 3 *Moderate and without a thrill*
- Grade 4 *Loud with thrill just palpable.*
- Grade 5 *Very loud and thrill easily palpable.*
- Grade 6 *Very loud, may be heard without the aid of a stethoscope.*

Appendix 2

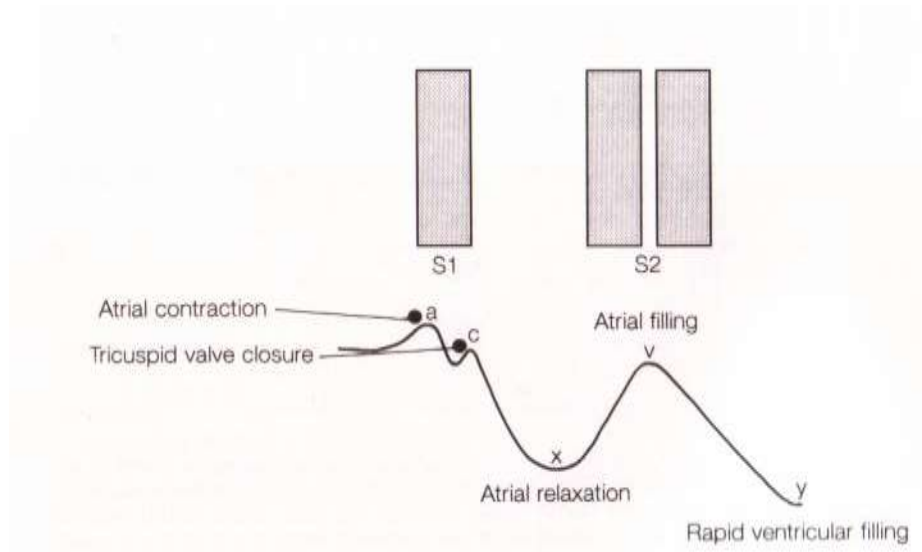
Auscultatory regions of the heart:



Note, these regions show the optimal areas for listening to the heart valve indicated, they do not exactly correlate with surface anatomy of the anatomical location of the valve.

Appendix 3

The JVP wave form:



Components of the jugular venous pressure wave with relationships to the first and second heart sounds.

References

1. Guidelines on the Management of Valvular Heart Disease. The Task Force on the Management of Valvular Heart Disease of the European Society of Cardiology. European Heart Journal Version 2012. European Heart Journal (2012) 33, 2451-2496. doi:10.1093/eurheartj/ehs109
2. Talley N.J, Clinical Examination 3rd ed 1996.
3. 2008 Focused Update Incorporated Into the ACC/AHA 2006 Guidelines for the Management of Patients With Valvular Heart Disease. Journal of the American College of Cardiology. Vol. 52, No. 13, 2008. doi:10.1016/j.jacc.2008.05.007.

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Reviewed July 2016.