Adult Congenital Case Report
Complications from an Anomalous Coronary During a Mitral Valve Replacement
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Disclosures
No Disclosures
Overview

- The complexity of today's adult congenital heart cases.
- Increasing numbers of this patient population and an increased likelihood of encountering these patients in your practice.
- Review Shone's syndrome and coronary artery anomalies/aberration and the incidence of these defects.
- Describe the specifics of this case report and the complication arising from the anomalous coronary despite precautionary preoperative studies and intraoperative surgical measures.
- Appreciate the difficulties and potential complications associated with surgical repairs of multifaceted heart lesions in the adult congenital patient population.

Pathophysiology of Congenital Heart Disease in the Adult

by Robert J. Sommer, Ziyad M. Hijazi, and John F. Rhodes

- Congenital heart disease is common, occurring in ≈8 of 1000 live births.
- With the successes in cardiothoracic surgery over the past 3 decades and the ongoing improvements in the diagnostic, interventional, and critical care skills of pediatric cardiologists, ≈90% of children born with heart defects now survive to adulthood.

Today, adults with congenital heart disease outnumber children with the condition.
Case Report

43 year old with PMHx of Shones variant consisting of:

- VSD
- Coarctation of the Aorta
- Mild Sub Aortic Stenosis
- Mild Mitral Valve Dysplasia
- Mitral Stenosis

Case Report

Past Surgical HX

- Coarctation repair with dacron patch and PA banding (neonate).
- VSD closure and PA band take down at ~2 years of age
- No surgery done on Sub AS or Mitral Dysplasia in infancy
- Balloon angioplasty of aorta for re-coarctation
Case Presentation

Patient presents to our center with:
- Worsening fatigue and SOB (CHF)
- Chest pain sharp, brief non radiating
- Increasing gradient across MV
- HTN

Referred for TEE and Cardiac Cath which showed:
1. Shone’s complex with history of coarctation of the Aorta, VSD, Supraventricular Ring, Parachute Mitral Valve, and subvalvular Aortic Stenosis
2. Sp Coarc repair and re-coarc balloon angioplasty
3. Sp VSD repair
4. Interval development of Severe Mitral Stenosis
5. Interval development of Thoracic Pseudoaneurysm associated with coarctation repair
6. Aberrant circumflex coronary artery arising from the Right Sinus

Anomalous circumflex traversing posterior to the aortic root

The LCx has its ostium at the right coronary artery just after its origin at the anterior right coronary sinus
Our patient was referred for two surgeries:

1. Anastomotic Aneurysm of the Thoracic Aorta (coarc)
   1. Left carotid to subclavian transposition (bypass)
   2. Endostent repair of thoracic aortic aneurysm

- 2 months later open heart surgery scheduled...
2. Severe Symptomatic Mitral Stenosis/Shones
   1. Redo sternotomy x 4
   2. MVR with 25 mm OnxM
   3. Partial resection of supramitral ring
   4. ASD closure (Access to MV) (a vessel was found running in the ring)
Case Presentation

- Ht 158 cm
- Wt 63 Kg
- BSA 1.64 M²

Bypass run #1 – 136'
Aortic Cross Clamp #1 – 94’

- Difficulty seating valve so downsized to a 25 mm mechanical valve as there was a vessel running in the supramitral ring described as a vein from the original cath report and it was difficult getting past the ring to the annulus.

- Off CPB ECHO showed:
  - Inferior wall and lateral wall dyskinesis and poor function.
  - The blood vessel running in the supra mitral ring appeared smaller and could not be traced throughout its path.

Procedure:
1. Redo sternotomy x 4
2. MVR with 25 mm OnxM
3. Resection of Supramitral Ring
4. ASD closure

1.3 liters del Nito cardioplegia antegrade 3.8 °C
additional 330 cc slow given at 60° antegrade 3.7 °C

1 liter del Nito cardioplegia antegrade 3.0 °C

Left Ventricle in short axis after first bypass: Inferior wall and lateral wall dyskinesis

Case Presentation

Bypass run #2 – 100’
Aortic Cross Clamp #2 – 51’

- 25mm valve was removed and repositioned in the supra-annular supramitral ring position allowing the supra mitral ring room below the valve so there would not be any impingement on the coronary artery.

- Off CPB ECHO showed:
  - Much better left ventricular function
  - Blood was clearly flowing into the coronary artery traveling within the supramitral ring
Left ventricle in short axis after revision of mitral valve position

Post Operative

- Patient did relatively well post operatively in the ICU
- Over the next 2 days the patient started to decline and required inotropic support.
- Lab order set revealed increased troponin levels indicative of ischemia despite EKG appearing normal
- A trip to the cath lab revealed there was intermittent compression of the anomalous circumflex artery during contraction.
- The decision was made to stent circumflex coronary
Mitral supravalvular ring: a case report

Cineangiography and MRI both failed in reaching this objective, whereas transesophageal echocardiography is the best method to identify this congenital heart disease. Supravalvular ring can occur as an isolated defect, but in nearly 90% of the patients, the supravalvular ring is found in conjunction with other congenital heart defects. In this case, a 36-year-old male was admitted to our Heart department. He experienced progressive dyspnea on effort and at rest. Diagnosis was made by transesophageal echocardiography which showed, on apical 4-chamber section, an anular structure attached to the mitral valve annulus above the valve leaflet.

More Descriptive Title:
Complications from an Anomalous Coronary During a Mitral Valve Replacement in a Patient with Shone’s Syndrome or Shone’s Complex

Shone’s Complex
Shone’s syndrome is a rare congenital cardiac malformation defined by four cardiovascular defects leading to left side heart obstruction at multiple levels.

- Parachute mitral valve (MS/MR)
- Supravalvular mitral ring
- Subaortic membrane
- Coarctation of the aorta

Few cases have been reported since this syndrome was first defined, and very few of these have been adult patients.

Incidence of Shone’s syndrome is <1% of all CHDs
Shone’s Syndrome

Described by Shone in 1963 (also known as Shone’s complex)

Is a rare congenital heart defect consisting of four left sided cardiac anomalies...

Anomalous Origin of the Left Circumflex Coronary Artery

Recognition, Angiographic Demonstration and Clinical Significance

Anomalous origin of the circumflex coronary artery from the proximal right coronary artery or right sinus of Valsalva was recognized in 60 of 2000 patients undergoing selective coronary angiography (0.3%). The relative frequency of this anomaly demands a high level of anticipation during the performance of selective coronary angiography to assure that an adequate study has been obtained. Failure to recognize and appropriately interpret these anomalous origins may result in diagnostic errors.

Two angiographic signs have proved reliable in recognizing the anomalous artery before its selective demonstration. These signs are a profile view of the artery behind the aorta seen during left ventriculography (the “rowboat” sign) and recognition of a short elongated tubular segment of the artery beyond the origin of the anomalous artery. These angiographic signs are described and the clinical implications of proper documentation of this anomalous circumflex coronary artery are discussed.

“Failure to recognize and properly demonstrate the anomaly can be hazardous to patient management.”
Clearly, this aspect of cardiology will not be able to develop fully without extensive collaboration between individual cardiologists and institutions. To further this goal, the Texas Heart Institute has established a Web site designed to promote multicenter collaboration on protocols dedicated to ACAOS patients (http://texasheart.org/Education/Resources/caac.cfm). Only such efforts can give rise to the large-scale studies needed to define the prognosis and optimal treatment of individual forms of CAs.

(Circulation. 2007;115:1296-1305.)
Coronary artery "dominance"

- Most hearts are right dominant where the PDA is supplied by the RCA. However up to 20% of hearts may be left dominant, where the PDA is supplied by the LAD or LCx.

- If both RCA and LCX give rise to the PDCA the system is co-dominant.

- The crux of the heart is usually supplied by the atroventricular node artery from the RCA.

"No mal" Anatomy of the Mitral Valve and Coronary Vessels

Coronary injury complications during mitral valve surgery

"This problem occurs much more frequently than we appreciate. Most people will claim they don’t know it exists but that is because they haven’t looked for it."

Ani C. Anyanwu, MSc, MD, FRCS
Mount Sinai Hospital.
Department of Cardiothoracic Surgery
The RCA and LCx form two half-loops forming the atrioventricular groove (arrowheads). Left circumflex coronary artery giving off two marginal obtuse branches (MO).

Anomalous
The LCx has its ostium at the right coronary artery just after its origin at the anterior right coronary sinus.
Conclusions

- Adult congenital cardiac disease is a fast growing sector of CTS and will likely affect us all

- Shone’s syndrome is a complex set of anomalies often requiring multiple surgical interventions

- Coronary artery anomalies are infrequent but must be considered when the post-op course does not proceed in expected fashion