Anesthesia Considerations for a patient with Marfan's Syndrome

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Background: Marfan's Syndrome

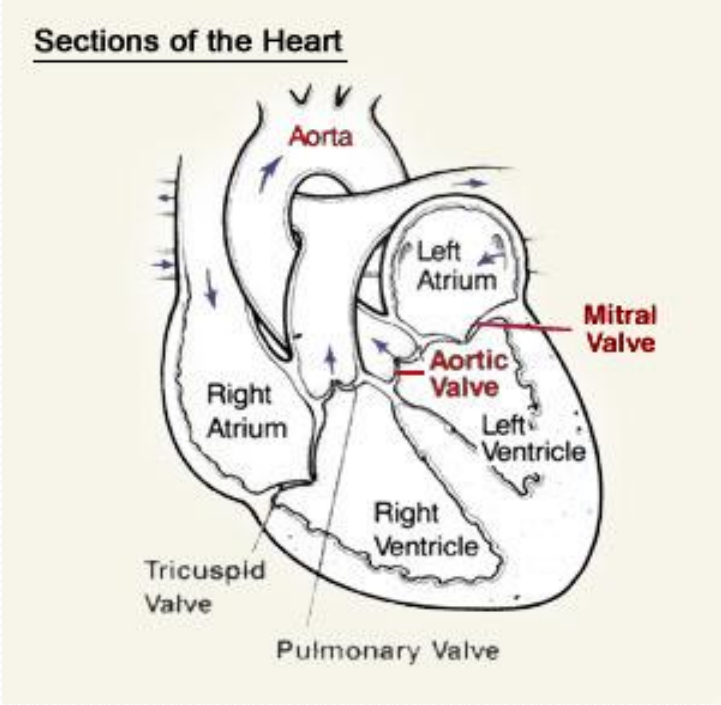
- Inherited autosomal dominant trait
- Incidence 1 in 5,000
- Connective tissue disorder with mutation of the FBN1 gene that encodes for fibrillin-1
- Fibrillin-1 is important in the structural support of tissues
- Average age of survival is 45 years; with treatment extending up to 70 years
- Premature death due to cardiovascular abnormalities

Characteristics of Marfan's

- Skeletal System
  - Tall, long slender arms, legs, fingers, and toes
  - Scoliosis, pectus excavatum or carinatum
  - Abnormal joint flexibility
  - High palate, small jaw
- Ocular Findings:
  - Myopia
  - Dislocated lens
  - Detachment of the retina
  - Glaucoma
  - Cataracts

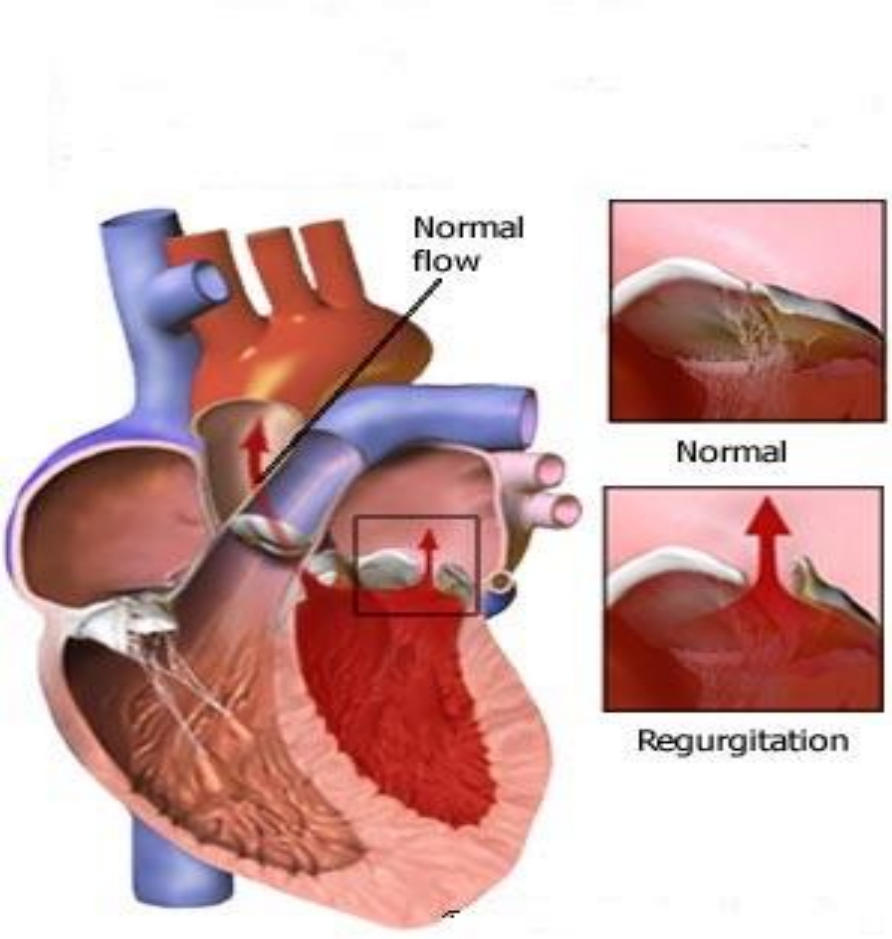
Characteristics of Marfan's

- Cardiovascular System
  - Heart murmur
  - Aortic or mitral regurgitation
  - Aortic root dilation or dissection
  - Pulmonary artery dilatation
  - Congestive heart failure
- Other
  - Spontaneous pneumothorax
  - Decreased muscle mass and subcutaneous fat
  - Obstructive sleep apnea
  - Lumbosacral dural ectasia



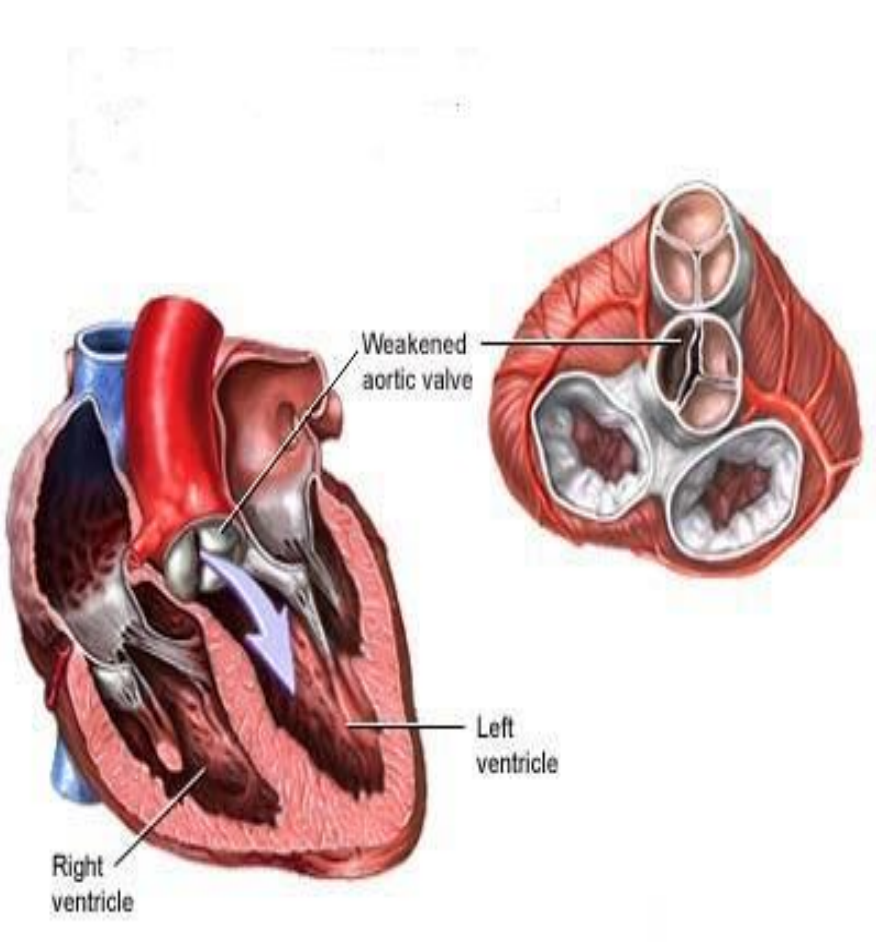
Mitral Regurgitation

- Reduction in forward stroke volume due to backward flow of blood into the left atrium during systole



Aortic Regurgitation

- Failure of the aortic valve to close leads to backflow into the left ventricle resulting in LV volume overload



Flo Hyman

Case Presentation:

- HPI:
- 37 year old Korean male presented to the LAC-USC Emergency Department with shortness of breath for 3 months with worsening symptoms for the past 3 days. On the day of admission, patient stated he felt like he was going to die.
- PMH: None
- Meds: None
- Family Hx: denies
- PSH: ORIF to right lower extremity
- Social Hx: 20 pack/yr smoker, drinks ½ bottle of whiskey every other day, denies illicit drug use

Physical Exam

Vitals:	BP 102/73 HR 84 bpm RR 16 O2 Sat 100% on N/C 2 L/min
Gen:	Moderate distress
HEENT:	+ 10 cm JVD
Cardiac:	III/VI holosystolic murmur noted at the apex, regular rate and rhythm
Resp:	Bibasilar crackles
Ext:	Long slender limbs, fingers and toes
Airway:	Mall 2, SMS 4 cm, MO 2 cm, high arched palate, Neck FROM

Laboratory and Diagnostic Work-up

- Labs: Unremarkable
- EKG: NSR @ 79 bpm; LAE
- CXR:
  - Severe cardiomegaly
  - Pulmonary edema
- TEE:
  - Trace aortic and pulmonic regurgitation
  - Severe mitral regurgitation
  - Normal LV systolic function
  - EF: 66%
- CT Chest/ABD:
  - Aortic root dilatation 5.8 cm
  - Pulmonary HTN

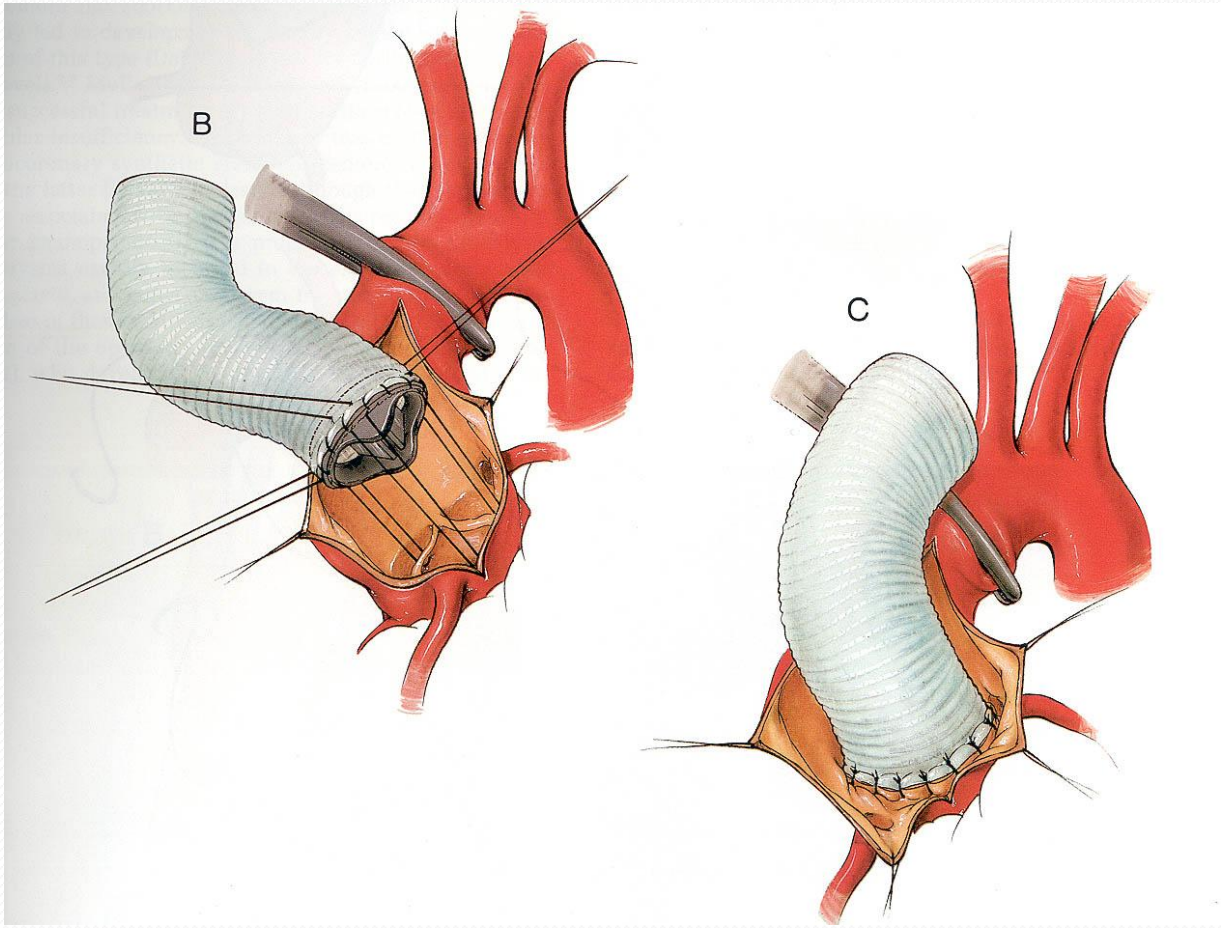
Hospital course:

- Nitroprusside drip
- Furosemide
- Metoprolol
- Lotensin
- Cardiac cath performed:
  - Normal coronary arteries
  - PA 36/21mmHg
  - Trace aortic regurgitation
  - Severe Mitral Regurgitation
  - Global hypokinesis

Diagnosis and Plan

- Diagnosis upon admission:
  - Marfan's Syndrome
  - Severe mitral regurgitation
  - Aortic regurgitation
  - Aortic root aneurysm (5.8cm in diameter)
- Plan:
  - Mitral Valve Replacement
  - Bentall Procedure-replacement of aortic valve and ascending aorta

Bentall Procedure



Anesthesia Considerations

- History and physical exam
- Review cardiac tests
  - EKG, ECHO, cardiology consult
  - Cath data: EF, condition of coronary arteries, valves, wall motion studies
- Review pulmonary tests
  - CXR, CT scan, PFT, ABG, pulmonary consult

Anesthesia Considerations

- Avoid factors that worsens regurgitation and aortic dilatation such as:
- Bradycardia:
  - Keep HR between 80-100 bpm
- Afterload:
  - Controlling afterload such as following endotracheal intubation and surgical stimulation
- Contractility:
  - Avoid sudden increases in contractility to minimize the risk of aortic dissection
- Preload:
  - Avoid excessive volume expansion to prevent further regurgitation and subsequent pulmonary edema

Preoperative Plan

- Arterial line prior to induction
- Right internal jugular Cordis and Swan-Ganz Catheter
- TEE monitoring
- Vasopressors on standby:
  - Dopamine, dobutamine, phenylephrine, ephedrine, epinephrine and levophed
- Vasodilators on standby
  - Nitroglycerin and sodium nitroprusside

Induction and Maintenance

- Induction
  - Fentanyl 200mcg, propofol 100mg, ketamine 50mg, morphine 4mg, cisatracurium 14mg
  - Direct laryngoscopy was done once systolic blood pressure 85-90mmHg
- Maintenance
  - Isoflurane with 100% oxygen
  - Scopolamine 0.4mg
  - Propofol drip 20 mcg/kg/min
  - Ketamine q1h throughout surgery
  - Cisatracurium titrated to TOF

Swan-Ganz values….

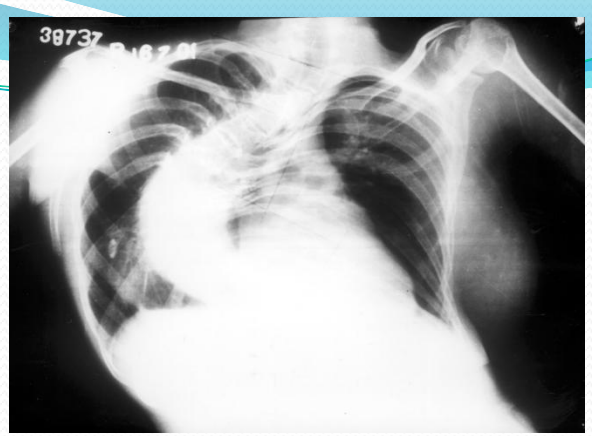
- PAP 49/34mmHg, CVP 13mmHg, CI 2.5 L/min, SVR 1240mmHg
- Management of pulmonary hypertension
  - Avoided systemic hypotension, hypoxemia, hypercarbia, acidosis, hypothermia, hypervolemia.
  - Nitroglycerin
  - Furosemide

Bypass Period

- Preparation for Bypass
  - Nitroglycerin to lower BP before aortic cannulation
  - Monitored for atrial dysrhythmias with venous cannulation
- Termination and weaning from Bypass
  - Prevented awareness with isoflurane and midazolam during termination from bypass
  - Blood pressure limited to systolic 120 mmHg to avoid stress on the aortotomy site
- Post Bypass
  - PRBC (3units), FFP (4units), Platelets (2units)
  - Furosemide IV given to prevent fluid overload

Post-operative

- Patient remained intubated and transferred to CTICU on the following drips:
  - Epinephrine, dopamine, nitroglycerin, amiodarone, insulin, aminocaproic acid, propofol
- POD#2: Extubated; out of bed to chair
- POD#7: Discharged home



Conclusion

- In a patient with Marfan's:
  - Preoperative evaluation should focus on cardiovascular and respiratory abnormalities
  - Aortic dilatation is silent unless aortic regurgitation or dissection is present and can be exacerbated under anesthesia
  - Cautious with positive pressure ventilation
  - Proper positioning to prevent injury and dislocations
- In a patient with mitral/aortic regurgitation:
  - Avoid bradycardia and tachycardia
  - Avoid sudden increases in SVR and contractility
  - Maintain preload

Michael Phelps

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