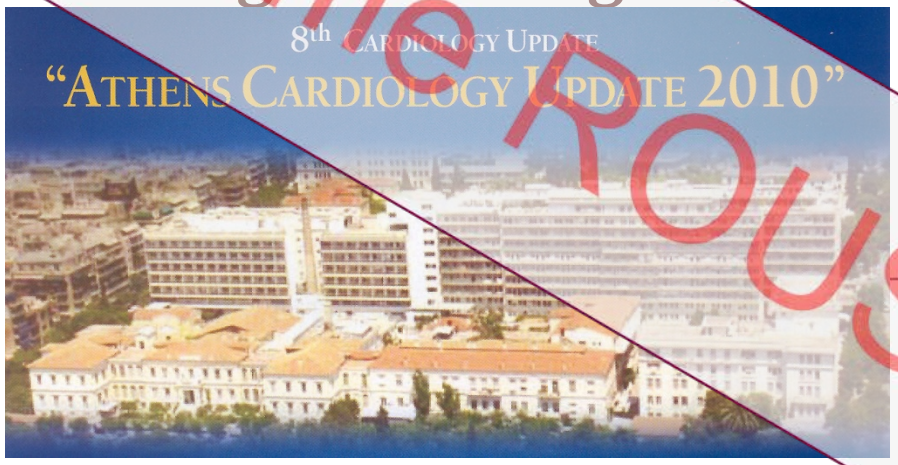
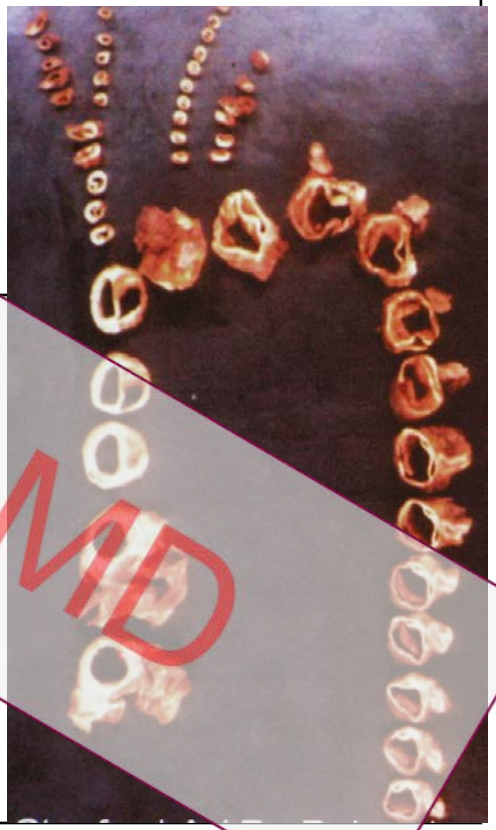
Late Reoperation for Proximal Aortic Complication in a Marfan Patient following Ascending Aortic Grafting





Efthymia G.Rouska[1],
Anastasios D.Spoulos[1], Charalampos F.Ntavelas[1],
Christos Charitos[2], Michalis Argyriou[2],
Efstathia J.Prappa[1], Antonios Sideris[1],

[1] Second Department of Cardiology, **Evangelismos General Hospital, Athens**
[2] Second Department of Cardiac Surgery, **Evangelismos General Hospital ,Athens**

Case

- 51-year-old Marfan female patient
- with a history of surgery for type A aortic dissection

On admission

- presented with uncontrolled hypertension
- known hypertensive on medical treatment
  - B-Blocker
  - ACE-inhibitor
- medical treatment became not efficient recently
- admitted neglect in her appointments over the last 3 yrs

PCH

Marfan patient - Ghent Criteria

System	Major criterion	Involvement
Skeletal	At least 4 of the following features:	2 of the major features, or 1 major feature and 2 of the following:
	● Pectus carinatum	● Pectus excavatum
	● Pectus excavatum requiring surgery	● Joint hypermobility
	● ULSP <0.86 or span:height >1.05	● High palate with dental
	● Wrist and thumb signs	● Crowding
	● Scoliosis >20° or spondylolisthesis	● Characteristic face
	● Reduced elbow extension (<170°)	
	● Pes plenus	
	● Protrusio acetabulae	
Ocular	Lens dislocation (ectopia lentis)	Flat cornea
		Increased axial length of globe (causing myopia)
		Hypoplastic iris or ciliary muscle (causing decreased miosis)
Cardiovascular	Dilatation of the aortic root	Mitral valve prolapse
	Dissection of the ascending aorta	Dilatation of the pulmonary artery, below age 40
		Calcified mitral annulus, below age 40
		Other dilatation or dissection of the aorta
Pulmonary	None	Spontaneous pneumothorax
		Apical blebs
Skin/Integument	None	Striae atrophicae
		Recurrent or incisional hernia
Dura	Lumbosacral dural ectasia	None
Genetic findings	Parent, child or sibling meets these criteria independently	None
	Fibrillin 1 mutation known to cause Marfan syndrome	
	Inheritance of DNA marker haplotype linked to Marfan syndrome in the family	

PCH

- Type –A aortic dissection 11 years ago

- confusional state
- bruises on both carotid arteries
- no palpable pulses in the right upper and the left lower extremity

CT angiography *demonstrated Type –A aortic dissection extended from aortic root to iliac arteries*

PCH

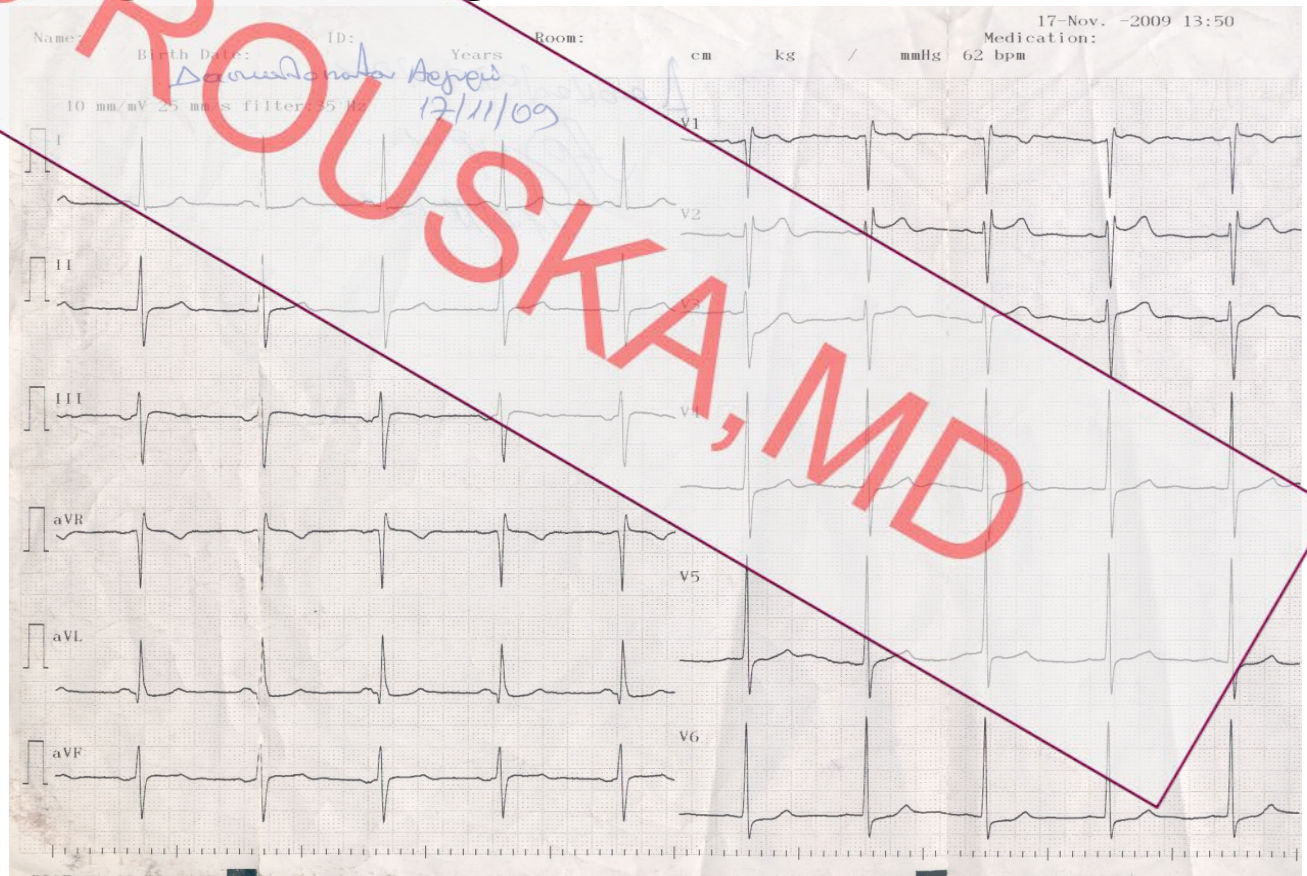
- **Operation:**
  - Supracoronary ascending aorta replacement with a tubular graft (Vascutec, 25mm) and resuspension of the incompetent aortic valve
- **Recovery:**
  - Very good
- **Histopathological examination:**
  - media of the aortic wall with a profound decrease in the amount of elastin and loss of the highly aligned and ordered lamellar arrangement
  - extensive deposits of mucopolysaccharides

Recent admission – ***O/E***

- HR 76bpm, SR
- BP 175/95mmHg
- <u>On auscultation</u>
  - parasternal diastolic murmur
    (third left intercostal space
    radiating widely along the left sternal border)

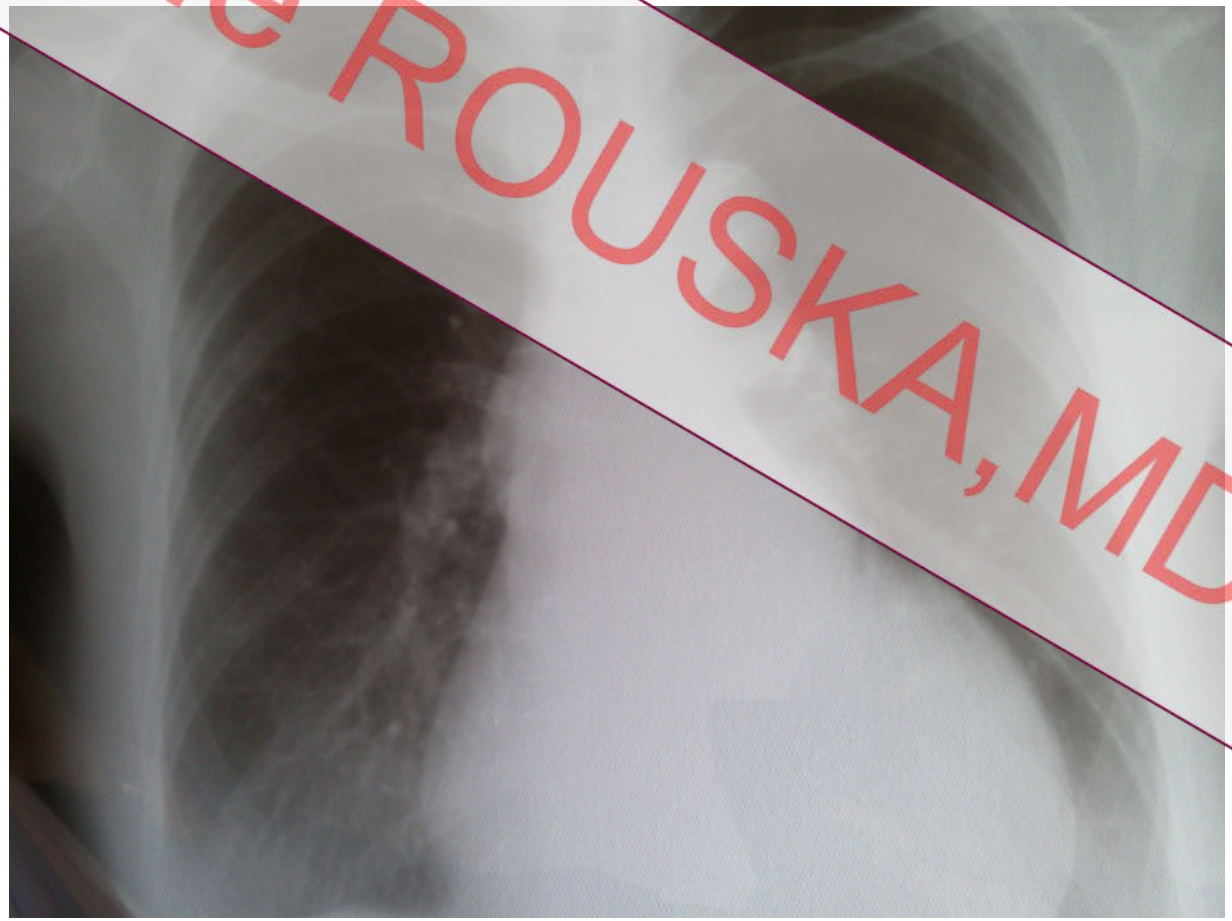
ECG

- *SR,*
- *nonspecific ST segment changes*
- *RBBB*
- *LVH*



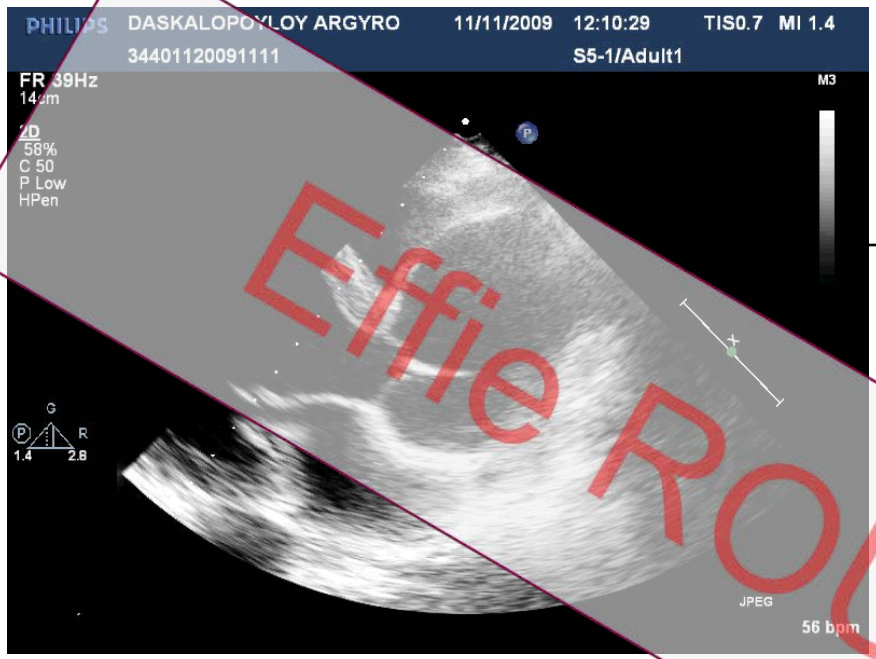
CXR

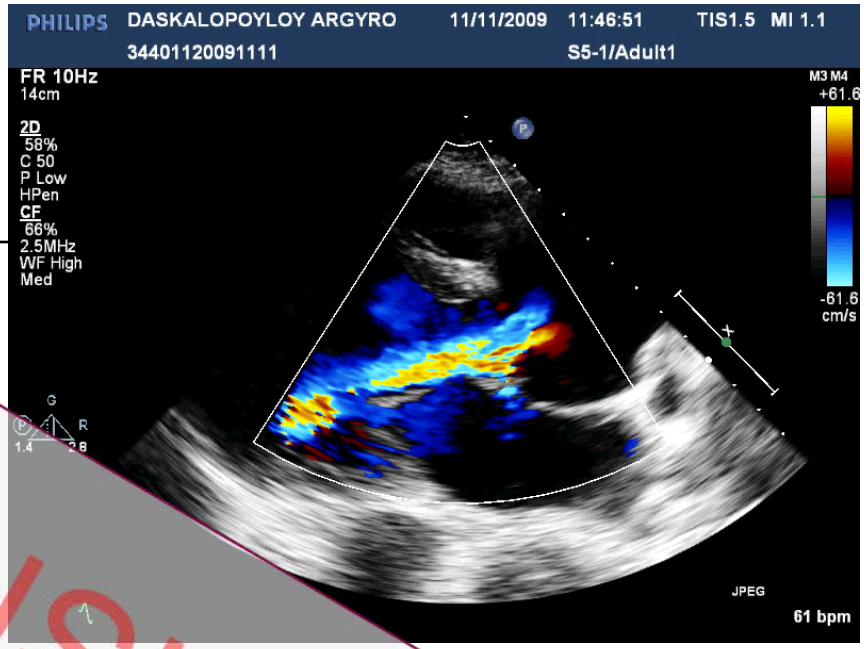
- cardiomegaly

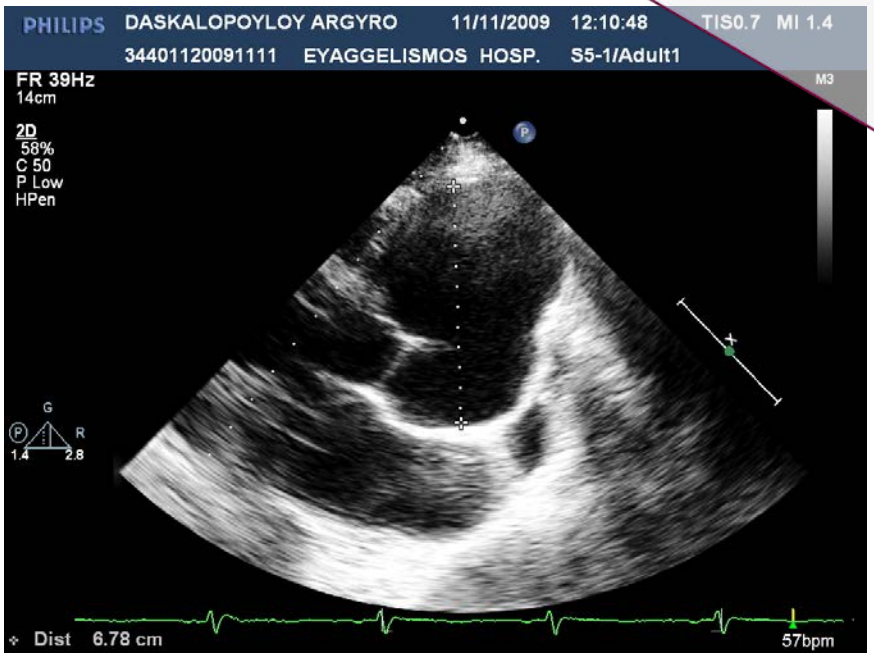


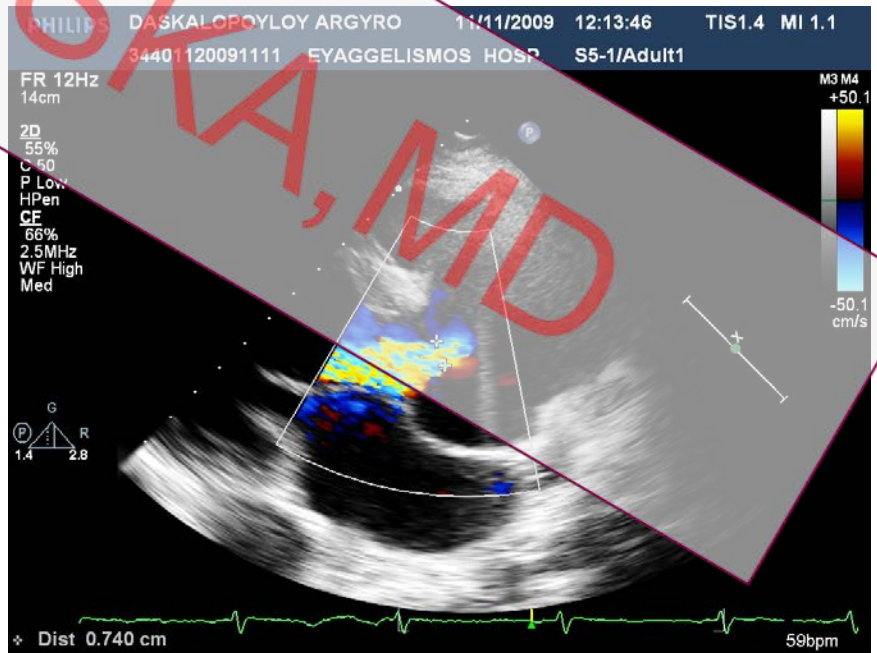
TTE findings !!!

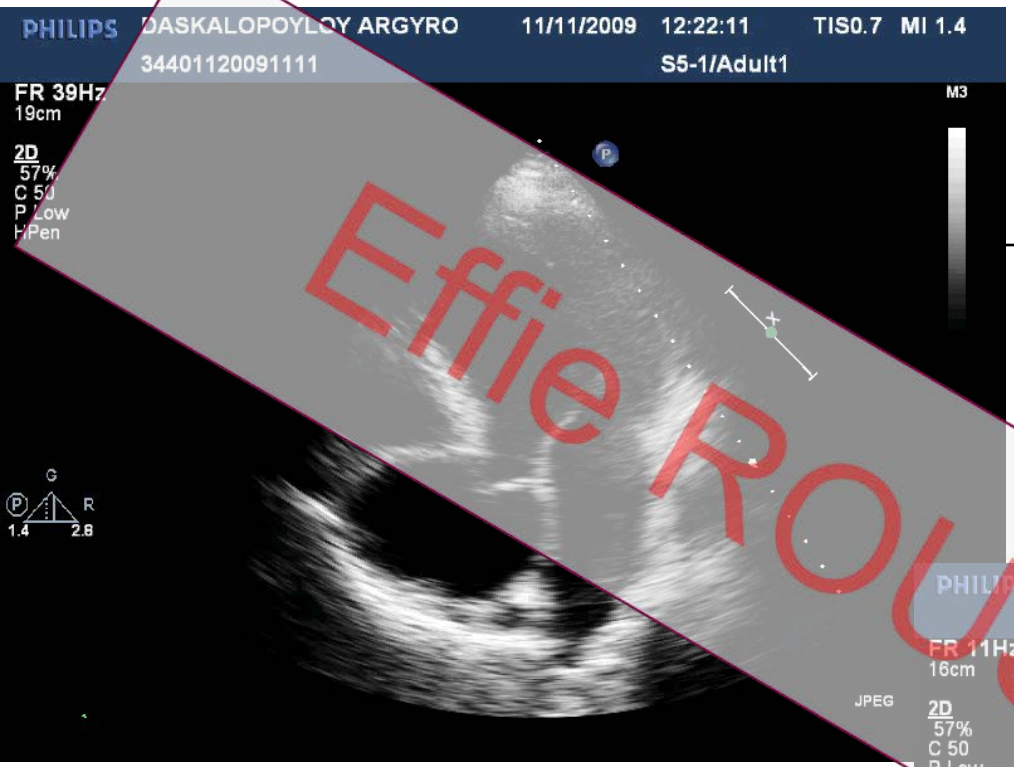
- severe dilatation of the Aortic root (d : 68mm)
- severe AR.
- Right Sinus of Valsalva aneurysm
- LV mildly hypertrophic and dilated.
- LV function mildly impaired.
- mild MR (functional due to AMVL prolapse)

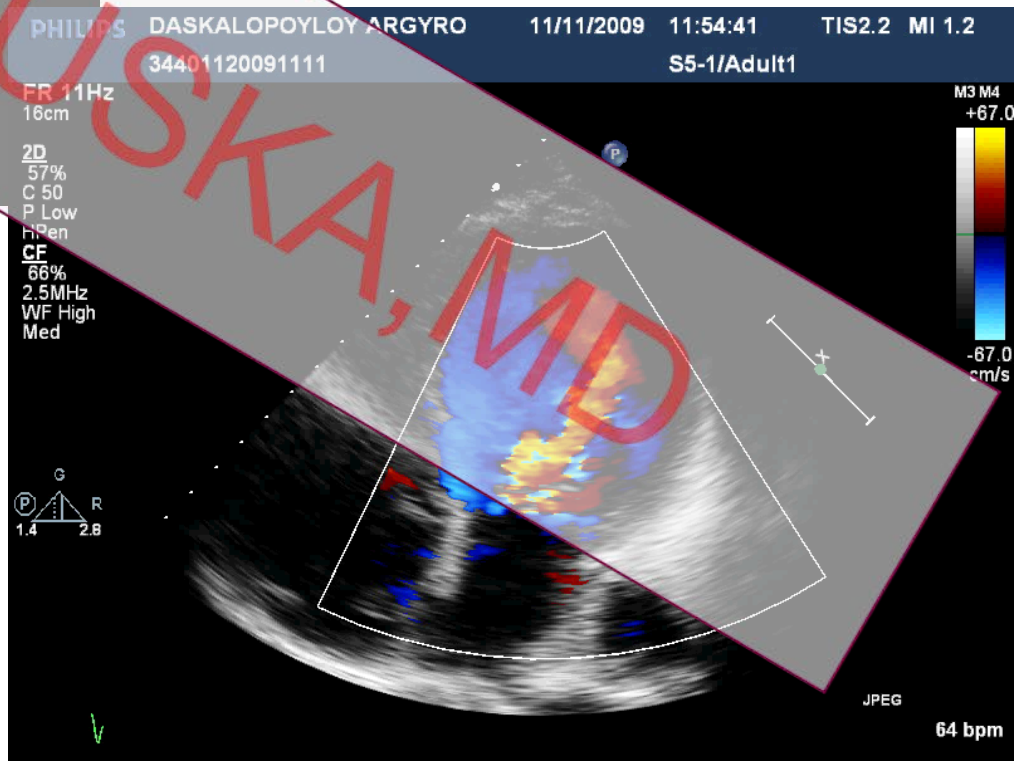




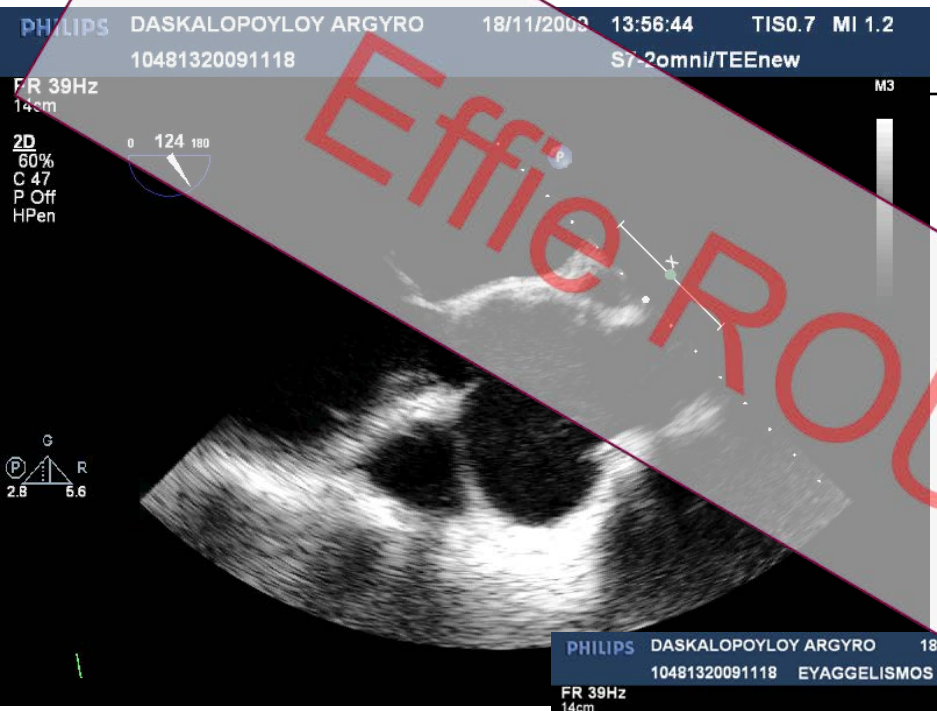


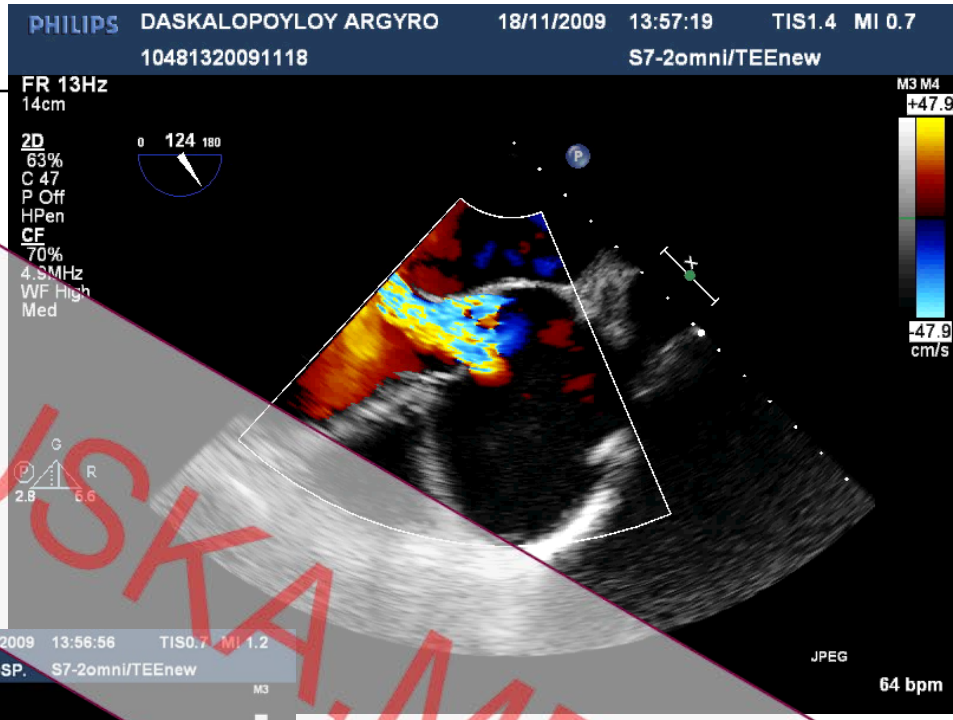


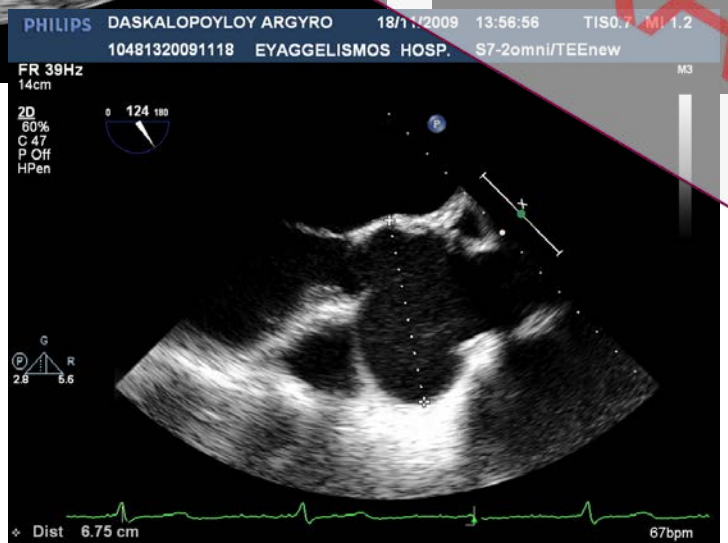


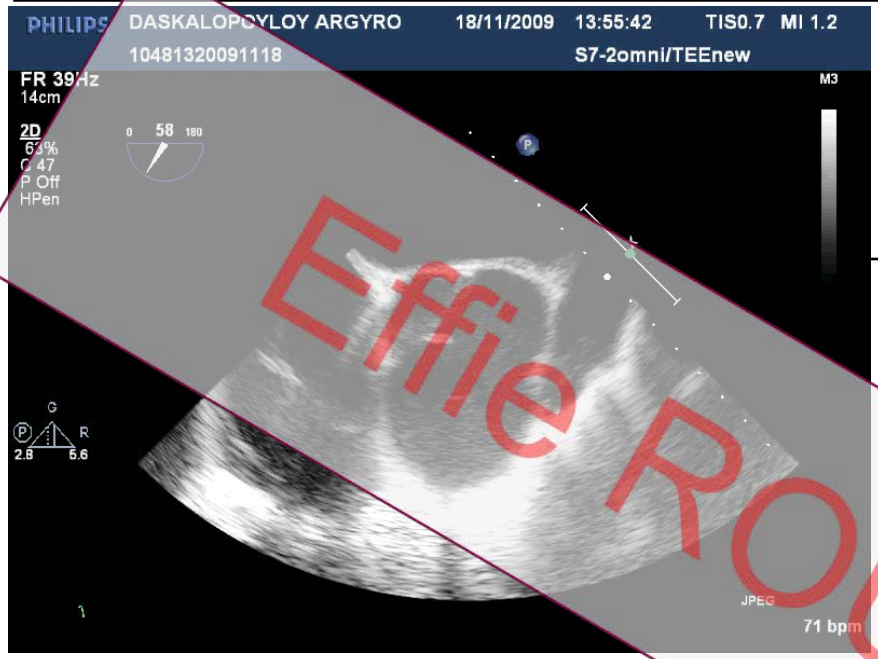


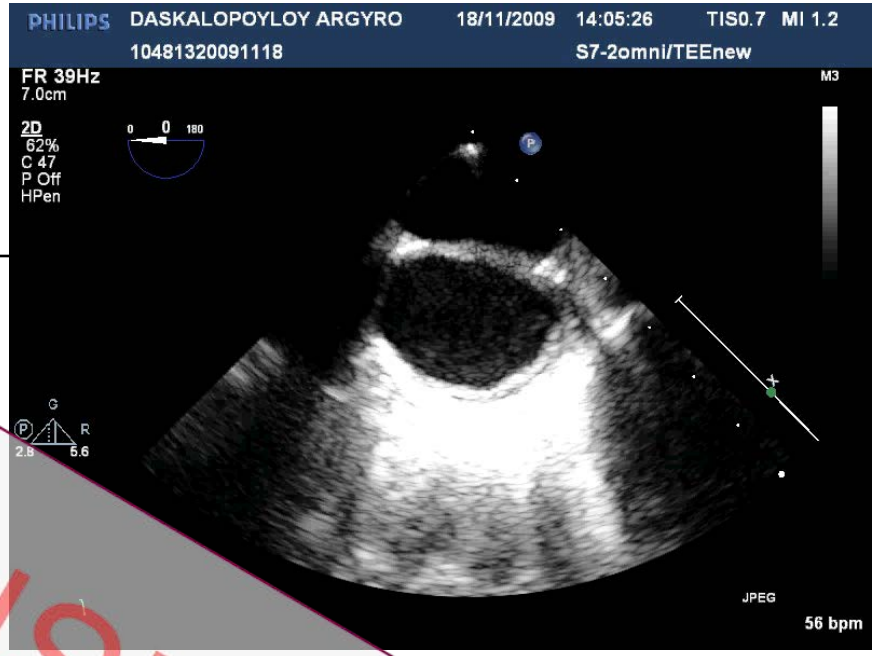
TOE findings confirmed TTE

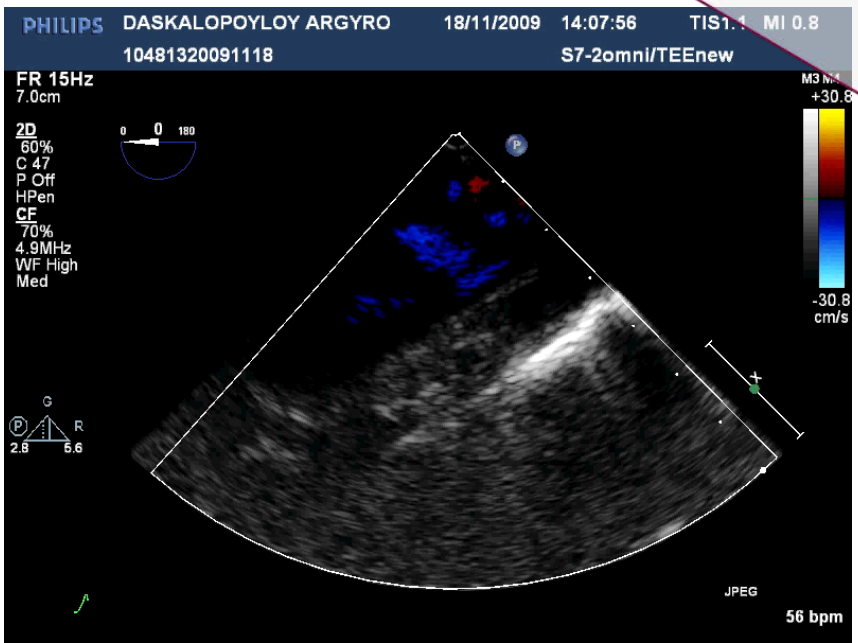


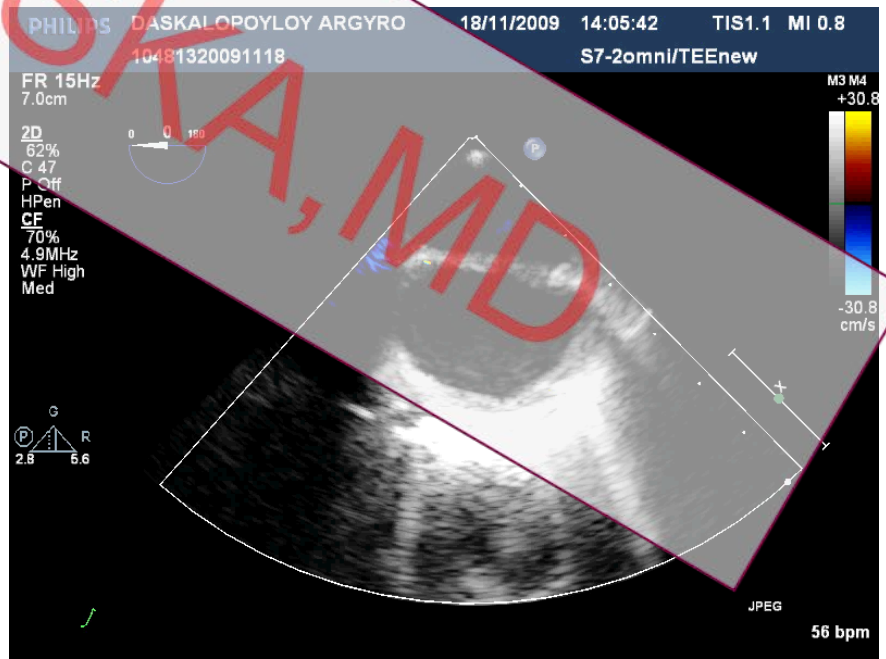






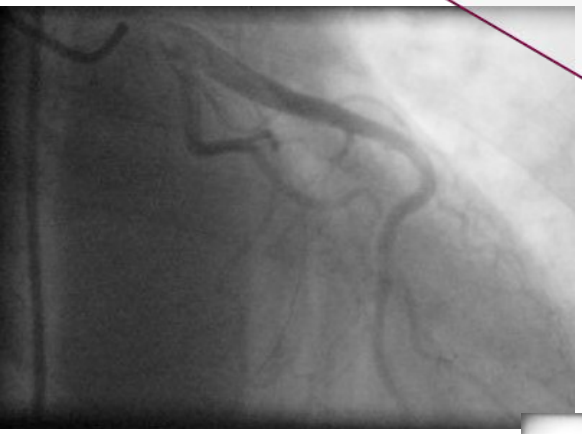


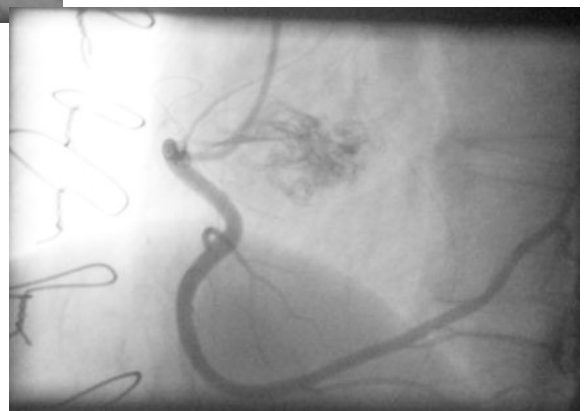


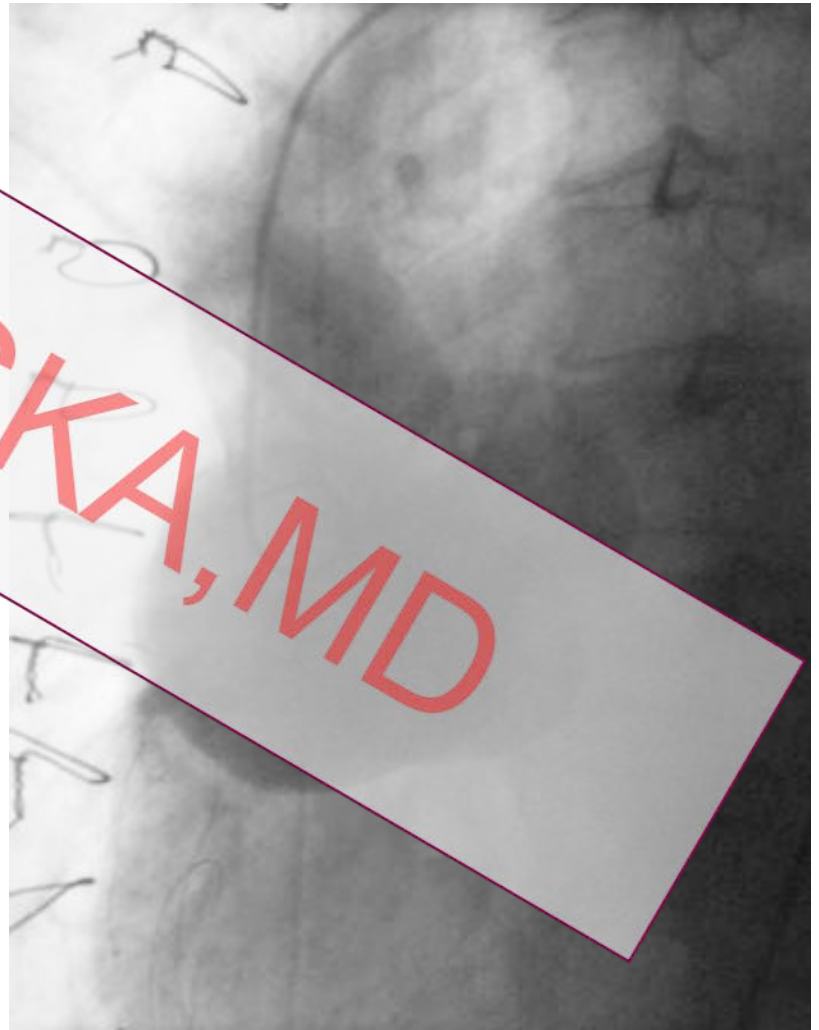


Coronary Angiogram -Aortography

- Normal coronaries
- Aneurysm of Valsalva

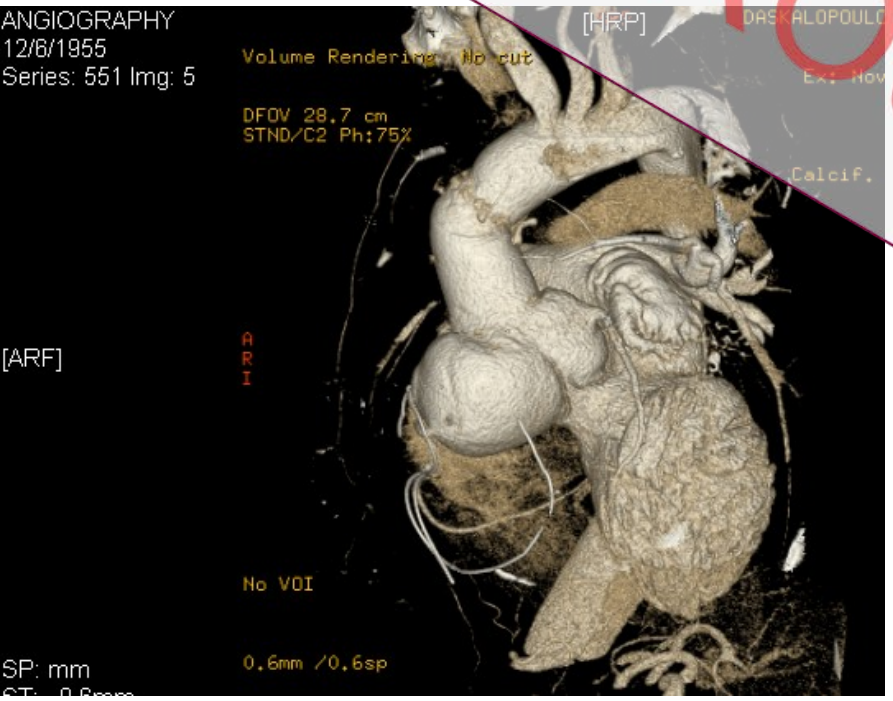


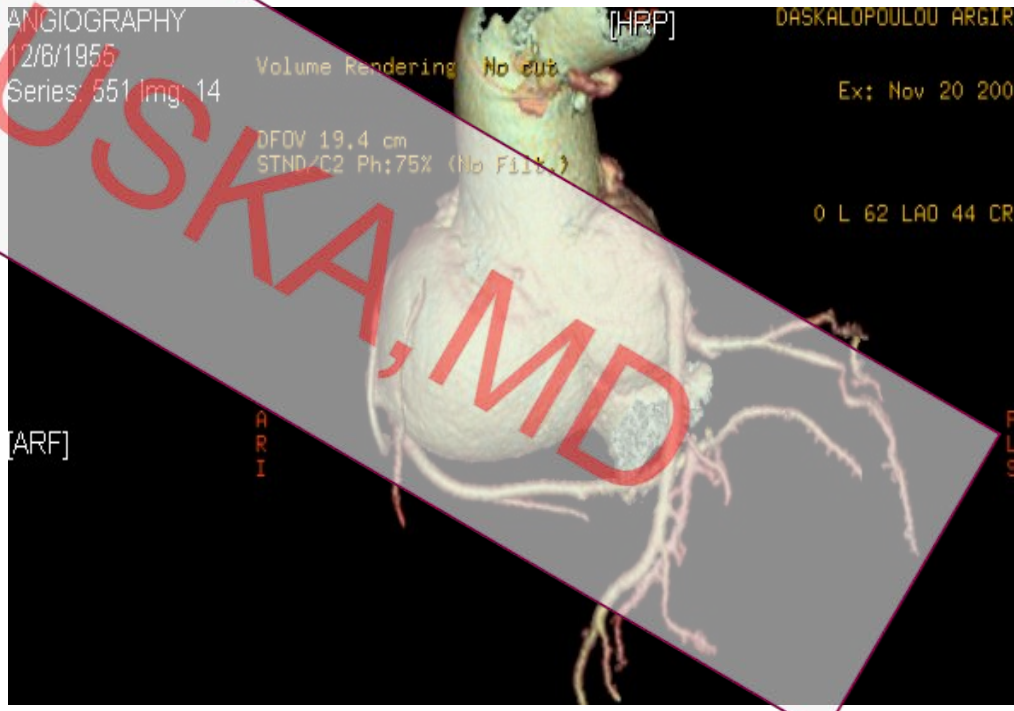




CT angiography

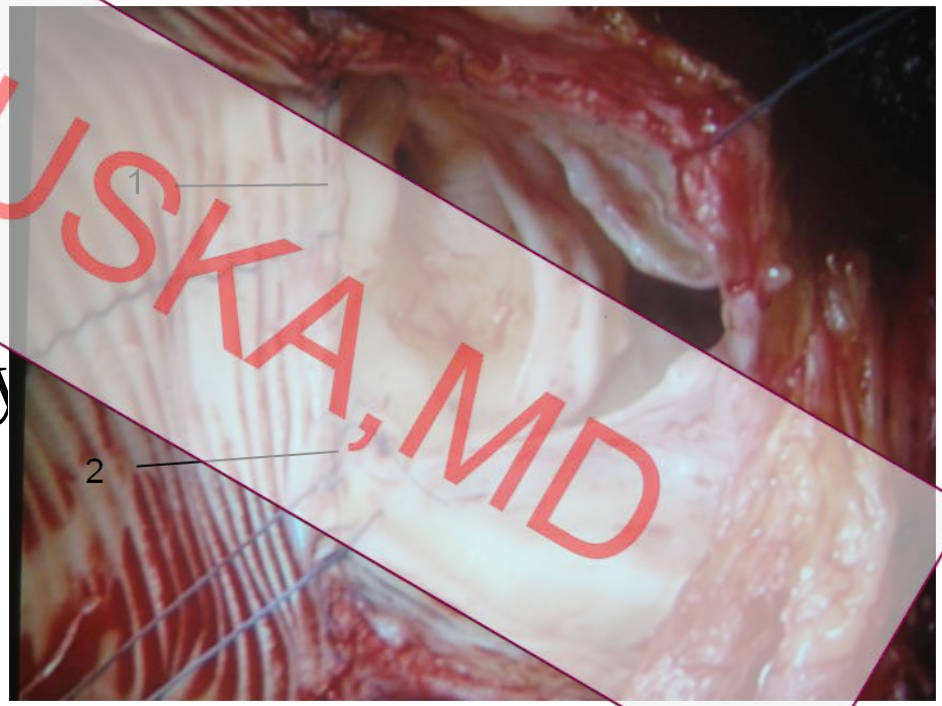
- severe Aortic root dilatation and findings consistent with the previous operation





Urgent Redo-operation

- Bentall procedure
- Post-op complications
- Long hospital stay
- Discharged the 55th day in a good condition



Discussion

- Marfan syndrome (MfS) is an autosomal dominant inherited connective tissue disorder with variable phenotypic expression of cardiovascular, ocular and musculoskeletal manifestations
- Usually associated with mutation in fibrillin-1 (FBN1) gene on chromosome 15, which encodes for the glycoprotein fibrillin.
- Estimated prevalence of MfS is 1 in 10,000
- 26% of cases have no family history (new mutation)

- Dean JC. Heart. 2002 ;88:97
- Westaby S. Ann Thorac Surg. 1999;67:1861
- Detter C, et al . Eur J Cardiothorac Surg. 1998 ;13:416

Diagnosis

- Ghent Criteria *(sensitivity/specificity 86%)*

System	Major criterion	Involvement
Skeletal	At least 4 of the following features:	2 of the major features, or 1 major feature and 2 of the following:
	• Pectus carinatum	• Pectus excavatum
	• Pectus excavatum requiring surgery	• Joint hypermobility
	• ULSR <0.86 or span:height >1.05	• High palate with dental
	• Wrist and thumb signs	• Crowding
	• Scoliosis >20° or spondylolisthesis	• Characteristic face
	• Reduced elbow extension (<170°)	
	• Pes plenus	
	• Protrusio acetabulae	
Ocular	Lens dislocation (ectopia lentis)	Flat cornea
		Increased axial length of globe (causing myopia)
		Hypoplastic iris or ciliary muscle (causing decreased miosis)
Cardiovascular	Dilatation of the aortic root	Mitral valve prolapse
	Dissection of the ascending aorta	Dilatation of the pulmonary artery, below age 40
		Calcified mitral annulus, below age 40
		Other dilatation or dissection of the aorta
Pulmonary	None	Spontaneous pneumothorax
		Apical blebs
Skin/Integument	None	Striae atrophicae
		Recurrent or incisional hernia
Dura	Lumbosacral dural ectasia	None
Genetic findings	Parent, child or sibling meets these criteria independently	None
	Fibrillin 1 mutation known to cause Marfan syndrome	
	Inheritance of DNA marker haplotype linked to Marfan syndrome in the family	

Natural History

- **Life expectancy is determined by the severity of cardiovascular involvement**
- Without surgical intervention, many patients die in the third decade of their lives from complications of aortic root aneurysm *(rupture, dissection, insufficiency)*
- Life expectancy improved in the past 30years as a result of improved surgical management

•David TE, et al J Thorac Cardiovasc Surg. 2009 ;138:859

Indications for surgery in MfS:

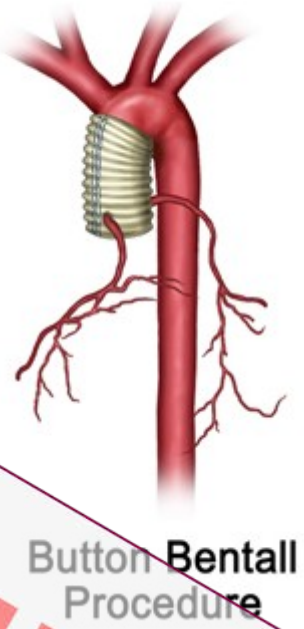
- aortic sinus diameter 5cm or greater or 4.5cm among patients with family history
- ascending aortic dissection
- aneurysm growth more than 1cm/year
- worsening aortic regurgitation in a dilated root when a valve-sparing procedure is desired

• Cameron DE, et al Ann Thorac Surg. 2009;87:1344

Surgery in MfS:

1. Bentall procedure (golden standard)

- Excellent early and late postoperative outcomes
- Complications related to long-term anticoagulation.

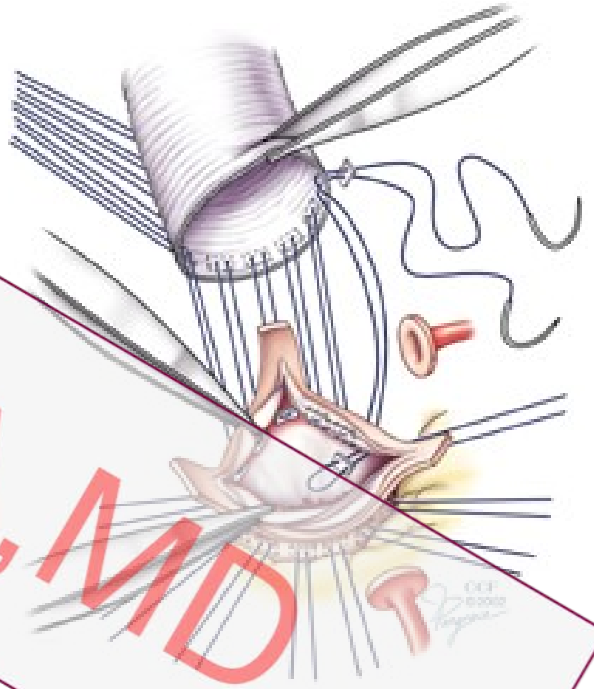


- Karck M, et al J Thorac Cardiovasc Surg. 2004;127:391
- Braverman AC. Curr Opin Cardiol. 2004 ;19:549
- *Elefteriades J .J Thorac Cardiovasc Surg 2002;123:201*

Surgery in MfS:

2. *Valve-sparing root replacement (David / Yacoub)*

- Operative results similar to Bentall procedure
- Time-consuming
- Durability has to be proven



- Cameron DE, et al Ann Thorac Surg. 2009;87:1344
- Patel ND, et al Ann Thorac Surg. 2008;85:2003
- Volguina IV, et al J Thorac Cardiovasc Surg. 2009 ;137:1124
- Kallenbach K, et al Ann Thorac Surg. 2007;83:S764
- *Elefteriades J. J Thorac Cardiovasc Surg 2002;123:201*

Surgery in MfS:

3. Conservative procedures

(replacement of the dissected aorta with tube graft - with/without resuspension of AV)

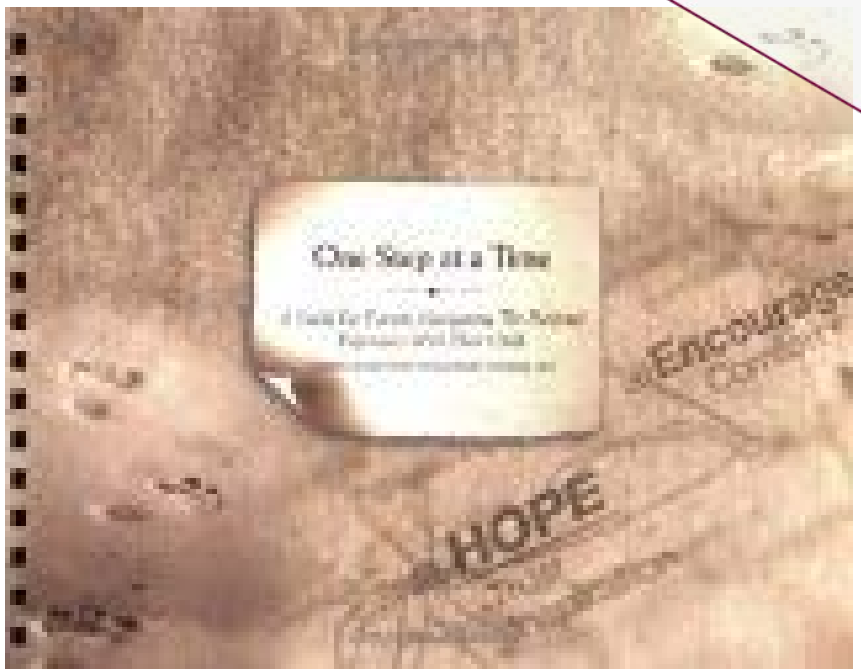
- Saves time
- High risk Reoperation usually needed!
- **Close follow-up of patients is mandatory!**

- Westaby S. Ann Thorac Surg. 1999;67:1861
- De Paulis R, et al Eur J Cardiothorac Surg.2005; 27: 86
- Treasure T. Heart. 2000;84:674

Conclusion

- *In Marfan patients who suffered dissection, **close follow up** is recommended to prevent any other cardiovascular complication, especially if a conservative operation has been performed*

- ***Replacement of the aortic root** is usually required in addition to repair of the dissected aorta, in order to eliminate the re-operation rate which is of high mortality and morbidity*



Thank you