Cardiac Manifestation of Behcet's disease

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M/43

- Chief Complaint: DOE Fc III to IV (3 month ago)

- Present Illness
  - **2005. 5.** Prosthetic valve replacement d/t severe aortic regurgitation
  - **2006. 4.** Aortic root replacement d/t prosthetic valve dysfunction & paravalvular leakage

    CABG d/t Lt. main-LAD injury

    Pacemaker insertion d/t complete AV block

    -> Diagnosed Behcet's disease

  - **2013. 10.** Exertional dyspnea, chest discomfort

OPERATION RECORD

등록번호 200502559 성명 양영환	Op Date 05-03 31	Op No 19710
Age/Sex 34 / M (Birthdate 1970 12 11)	Surgeon 나찬영 / 김재현 / 김윤정	
체중 56 kg 체표 1 6 m2	Physician 유길웅	

Dx Severe AR
moderate MR
LV dysfunction(Efx-45%)
Jehovah s witness
R/O Bechet s disease

Clinical

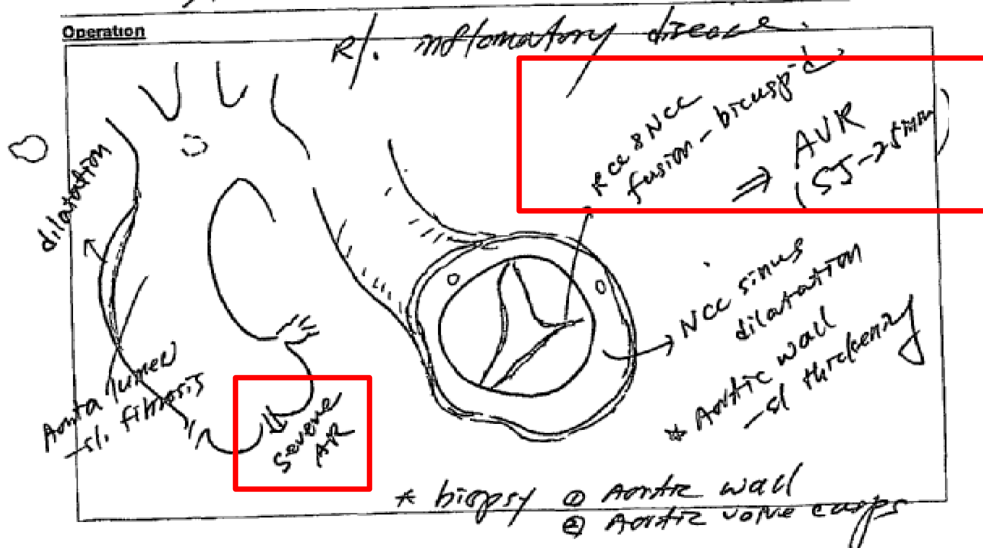
Past Hx 1 Frequent oral and genital ulcer history negative
2 Preop LVD(71/51mm) Efx-45% severe AR aortic valve annulus-21 8mm Sinus portion 36 4mm STJ 3 02mm moderate MR

Remark 1 Preop Hb 12 1gm%
2 Postop Hb 12 1gm%(immediate posrop)
3 Ascending aorta wall and calve cusps-

Op Name
AVR(SJ-25mm)

Op finding
1 Ascending aorta- greater curvature- mild aneurysmal dilatation aortic wall mild thickening R/O inflamation-- send to biosy(aortic wall and aortic valve)
2 Aortic valve- bicuspid RCC 7 NCC fusion- normal cusps- morphology
3 NCC 7 RCC commissural area의 sinus portion이 downward diplaced(due to R/O imflmation)

Operation



양면출력용

이 서식은 사본입니다(원본 대조필)

OPERATION RECORD

Age/Sex 34/M (Birthdate 1970-12 11)
체중 56 kg 체표 1 6 m2

Op Procedure

GEA
Full median sternotomy
No thymectomy
Pericardium tenting
Approach Aorta

CPB 70 min ACC 45 min TCA 0 min [Fibrillation time 0 min

Arterial cannulation Aorta Venous cannulation RA

Vent RUPV LAA Cardioplegics retrograde

Under the general anesthesia median sternotomy incision was done Pericardium was opened Aortic and single RA venous cannualtions were followed after systemic heparinization CPB was started and hypothermia was induced LV vent cannulation was done through RUPV ACC and retrograde cold blood cardioplegia delivery were followed Aorta was opened and aortic valve and aorta morphology were inspeced(same as op finding) Aortic cusps were resected and Aortic valve replacement sutures were done with 2 0 Ticron pledgeted interrupted sutures(16#) Valve was selected with SJ 25mm and sewing ring sutures and tie were followed Rewarming was started and retrograde warm blood reperfusion was started Aortic wall biopsy was done Aorta was closed with 4-0 prolene mattressed sutures ACC release and deairing were followed CPB weaning and modified hemofilteration were followed Decannualtion and heparin reverse were followed Pacign wires and ches ttubes were palced Pericardium was closed Chest wound was closed after meticulous bleeding control
AV replacement with SJ 25 mm

Deairing Aorta declamping CPB weaning
Pericardial closure with Autopericardium
Pleural opening none
C tube 2
Pacing wire atrium(2) ventricle(2)
Postop hemodynamics - stable

Described by [signature]

혜원의료재단 세종 병원 Sejong General Hospital

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OPERATION RECORD

등록번호: 200502559 성명: 양영환
Age/Sex: 35 / M (Birthdate:1970-12-11)
체중: 60 kg 체표: 1.64 m2

Op. Date 06-04-20 Op No: 21205
Surgeon 나찬영 / 김재현 / 권혁봉
Physician 유철웅

Dx Behcet disease
S/P AVR(SJ-25mm)
Prosthetic AV dysfunction
Prosthetic aortic valve paravalvualr leakage
Severe AR(IV)
R/O Prosthetic aortic valve enodocarditis
Jehovah's witness
Complete AV block
S/P temporary pacing
AR gr 4,
Etiology - Degenerative
Functional class 3, Rhythm - Others

Clinical

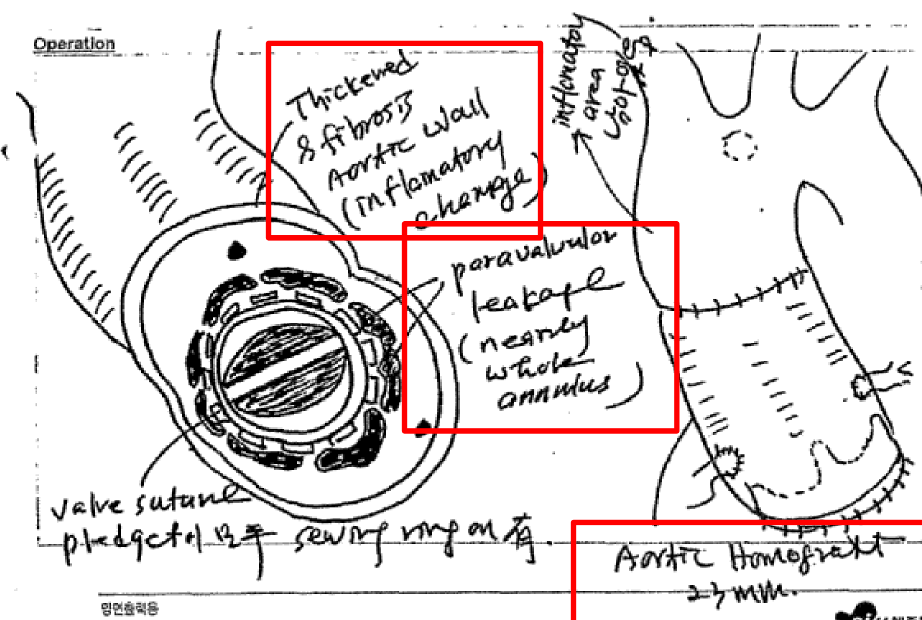
Past Hx 1. 2005-03-31 - AVR(SJ-25mm)- Biopsy- inflmation cell(+).
2. 2005-08-02 echo : LVD(45/30mm), no paravalvualr leakage
3. 2006-04-06 echo : aortic valve dehiscence, AR(III-IV), aortic sinsu-57mm

Remark 1. Preop. Hb- 13.7gm%, postop. Hb- 12.4gm%
2. Homograft- 삼성, donor- 62/F- traffic patient
3. Holmograft- not good quality- aortic

Op. Name
Redo- surgery
Aortic root replacement(Aortic homograft 23mm
Conventional CABG(SV-mLAD)

Op. finding
1.severe adhesion of substernal space.
2. Aortic root and ascending aorta- dilation and inflamatory change(thickened wall and fibrosis)
3. Aortic sinus -dilated.
4.prosthetic AV- annulus dehiscence(all pledgeteds- sewing ring 에 위치하여 scissors나 blade를 사용하지 않고 removed by forcep).
5. Aortic valve sewing ring & 일부 sinus portion에 vegetation-like materal(+)- send to culture.

Operation



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흉부외과 – 2006 OpNo 21205 세종병원

OPERATION RECORD

등록번호 2005 02559 성명 양영환
Age/Sex 35/M (Birthdate 1970 12 11)
체중 60 kg 체표 1 64 m2

Op Procedure

GEA
Full median sternotomy
No thymectomy
Pericardium tenting
Approach Aorta

CPB 235 min ACC 182 min TCA 0 min {Fibrillation time 0 min
Arterial cannulation Asc Aorta Venous cannulation SVC IVC
Vent RUPV Cardioplegics antegrade Ao root retrograde direct coronary artery

Under the general anesthesia median resternotomy incision was done Substernal space was carefully dessected Aortic and bicaval cannualtions were followed after systemic heparinization CPB was started and hypothermia was induced LV vent cannualtion was doen through RSPV ACC and retrograde cold blood cardiopleiga delivery were followed Aorta was opened Aortic valve(prosthetic valve) was easily removed due to severe valve dehiscence without use of blafe ro scissors) Both coroanry buttons were made During left coroanry button dissection left main or Cx coroanry was injured so repaired with 6 0 prolene interrupted sutures(3#) Valve repalcemetn sutures were done with 4 0 prolene interrupted sutures(29#) Homograft was trimmed Homograft annulus suturesd were done and tie were aslo followed Both coronary buttons were reattached using 5 0 & 6 0 prolene continuous suture(aortic wall thick so 5 0 prolene used) During distal anastomosis SV was harvested due to left coronary injury Rewarming and retrograde warm blood reperfusionw ere followed SV was bypassed to aorta to mlAD using 7 0 prolene ACC release and deairing were followed CPB weanign and modified hemofilteraiton were followed Decannulation and heparin reverse were followed Bleeding control was done Pericardium was clsoed with Goretex membrane and self pericardium Pacign wires and chest tubes were placed Chest wound was closed with usual manner
AV replacement with Autograft (AV) 23 mm

Deairing Aorta declamping CPB weaning
Pleural opening both
C tube 3
Pacing wire atrium(2) ventricle(2)
Postop hemodynamics stable

* Lt coronary button dissection 中
coronary artery injury (Lt main or Lt Cx)
⇒ CABG (SV – LAD 시행)
postop MI battery – Normal
EKG – block – pacing
Echo : good LV function

Described by 나찬영

Sejong General Hospital

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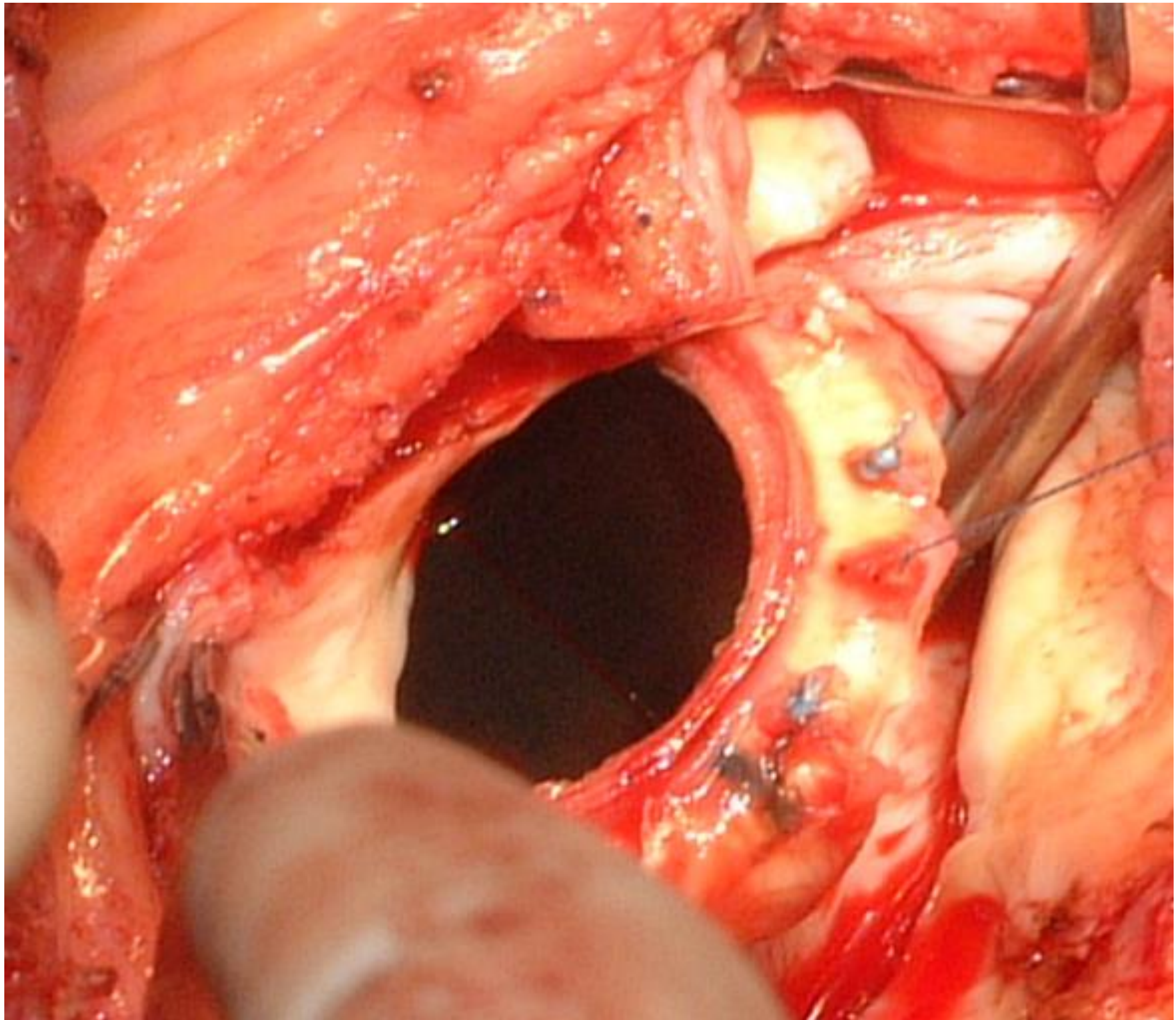


In summary

- HTN / DM / Tb / Hepatitis (-/-/-/-)
  - **Behcet's disase (+)**: diagnosed in 2005.
- **Adm / Op history (+/+)**
  - **Prosthetic aortic valve replacement (2005/05)**
  - **Aortic root replacement with homograft (2006/04)**
    - **Coronary artery bypass graft op (SVG-LAD) (2006/04)**
    - **Pacemaker insertion (2006/04)**
- Social history
  - Alcohol / **Current smoking** (-/**+**): 20 PY (1 PPD x 20 yrs)
- Medication (+)
  - **Cardiology**: Aspirin, Clopidogrel, Lasix 20mg, Spironolactone 12.5mg, Bisoprolol 1.25mg.
  - **Rheumatology**: Azathioprine 100mg, Prednisolone 10mg

ROS, P/Ex, Lab

- **Review of system**

Chest discomfort / **Dyspnea** (+/+): **NYHA III-IV**.

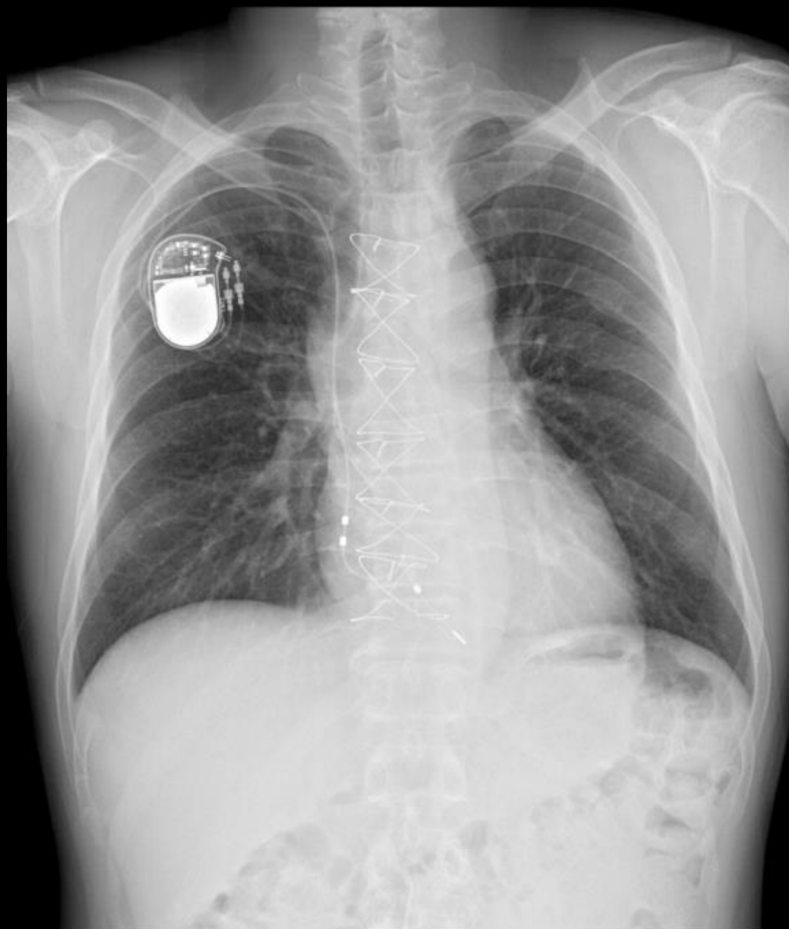
- **Physical examination**

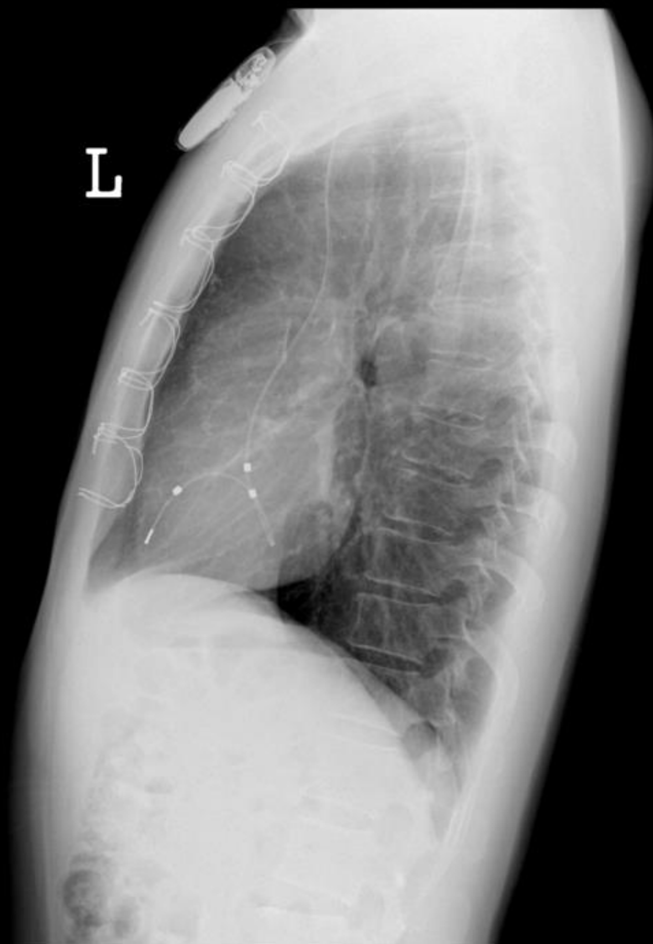
Clear breathing sound without crackle, wheezing.

Systolic murmur, Gr3, left sternal border.

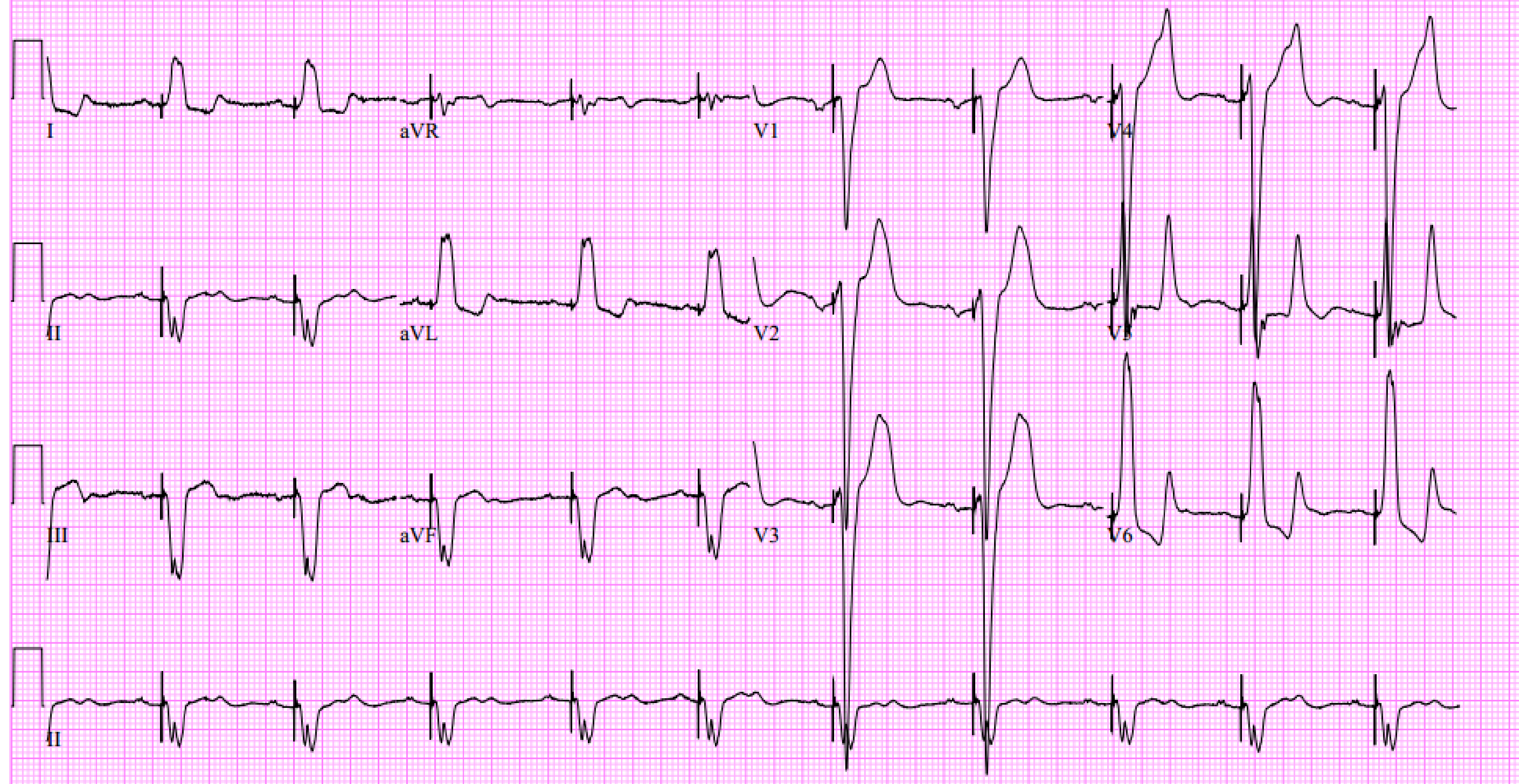
- Lab
  - NT Pro BNP **360.5** pg/mL, BUN / Cr 14.0 / 0.97 mg/dL
  - Hb 15.1 g/dL - WBC 9,200 /uL – PLT 227,000 /uL

Initial radiologic findings



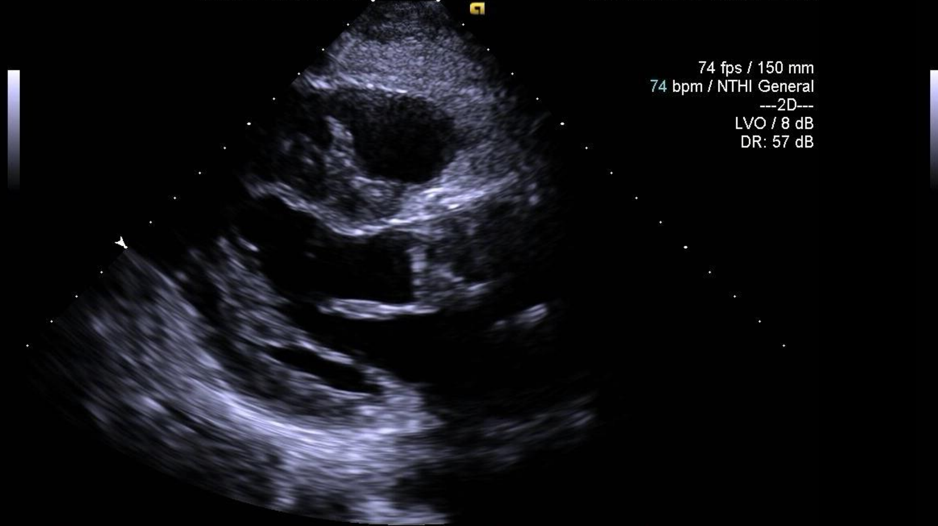


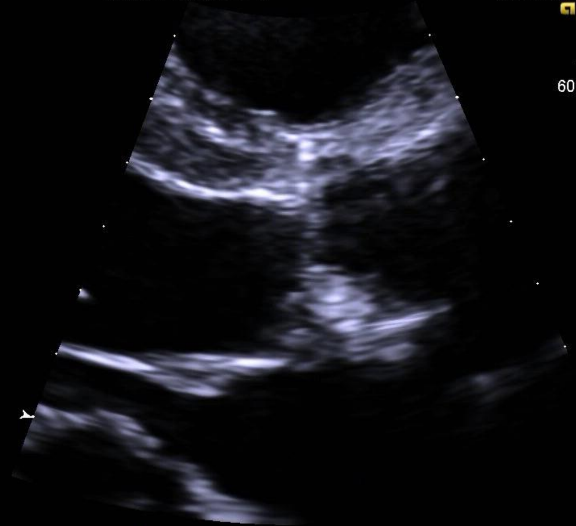
EKG findings

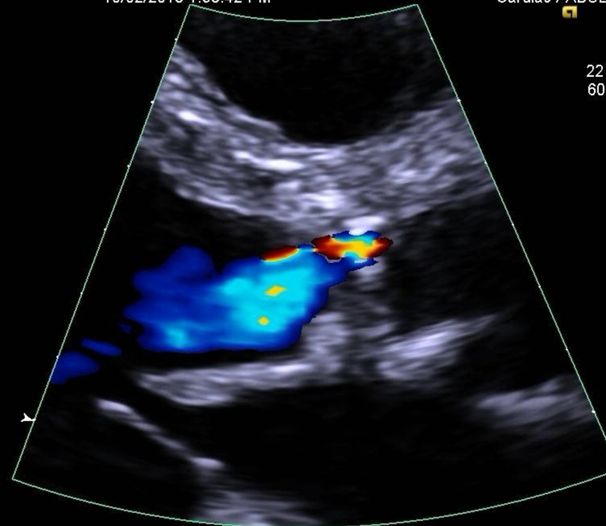


Electrical pacing rhythm. HR 63 bpm

2-D ECHO (TTE)







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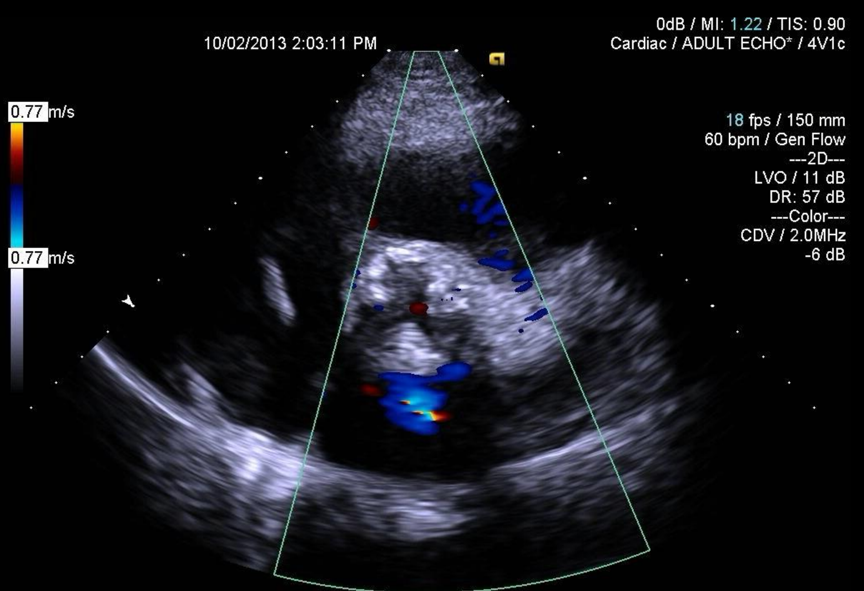
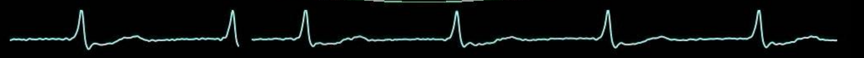
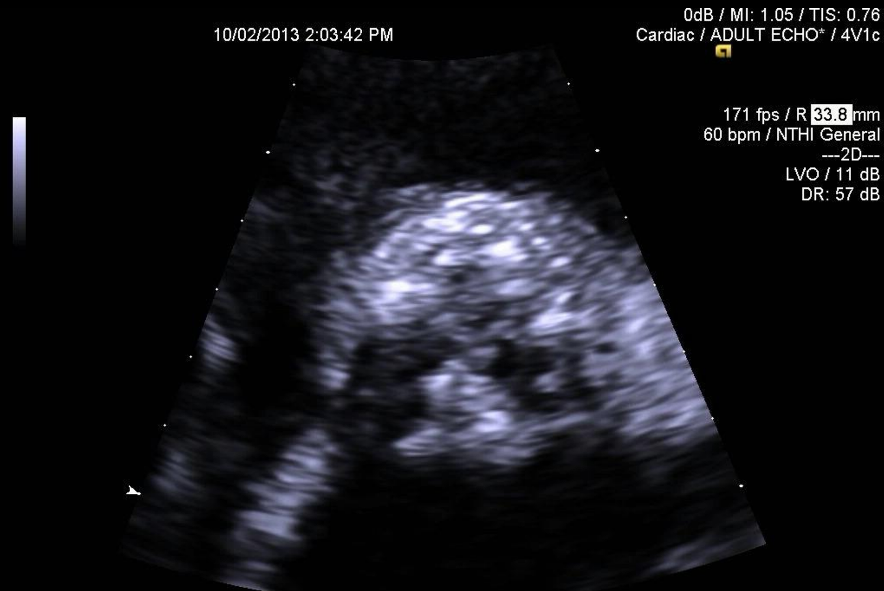
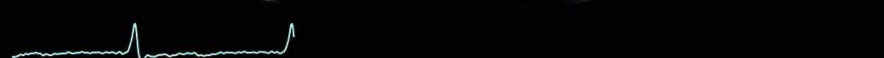
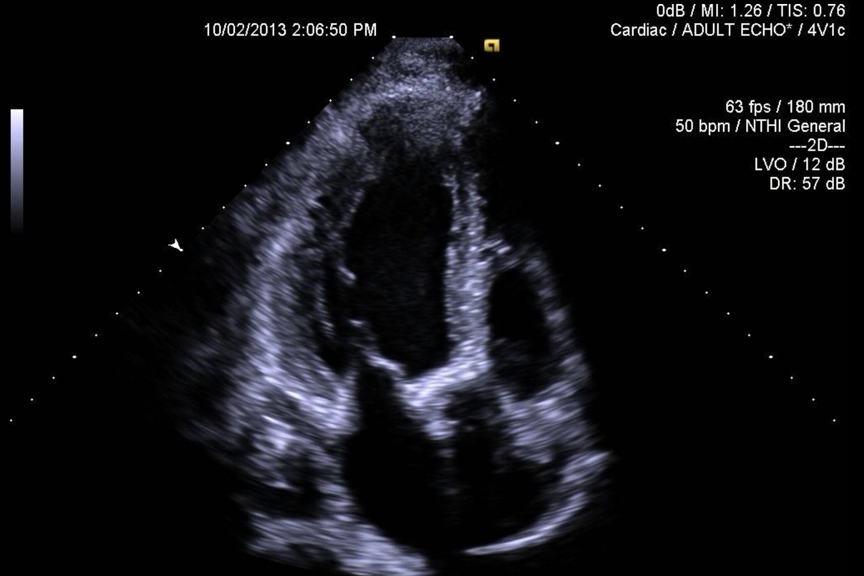
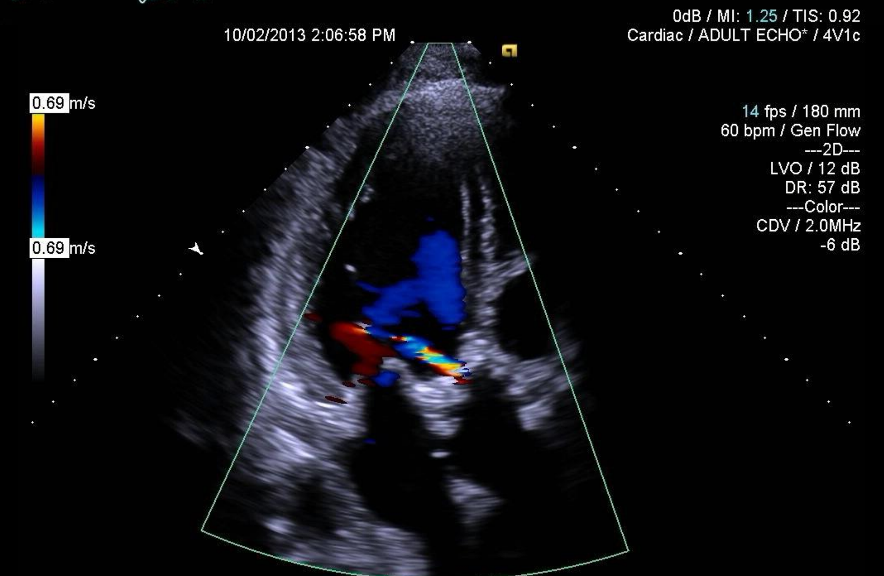




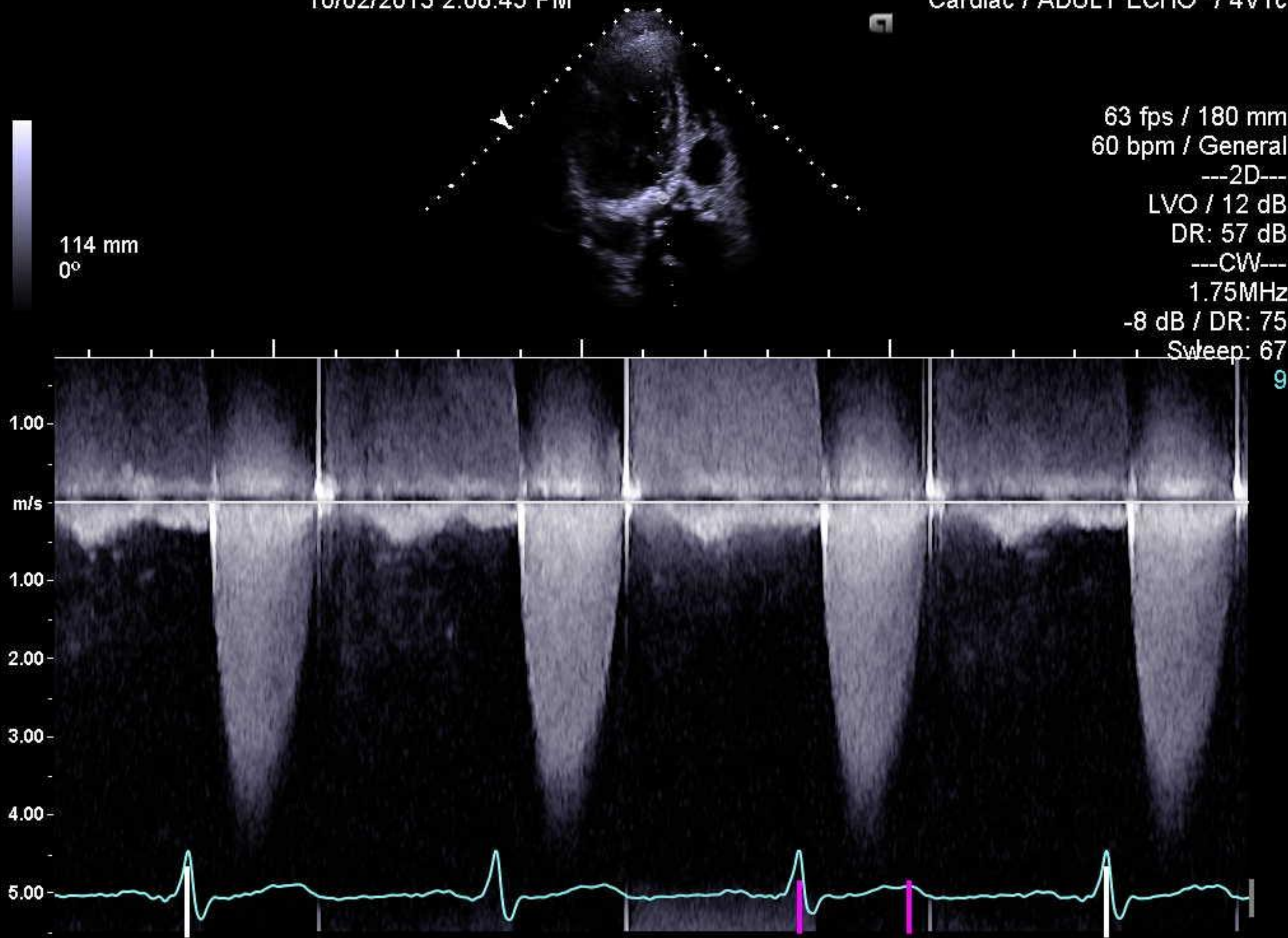






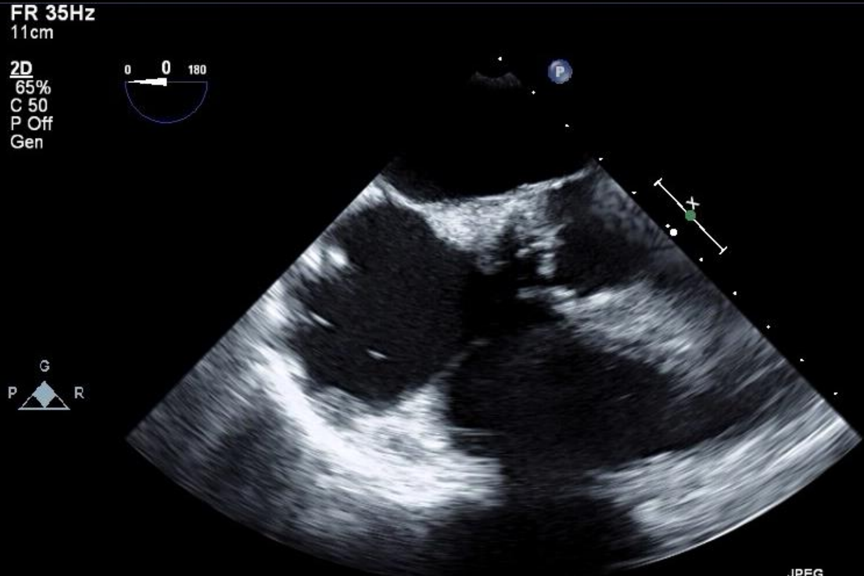


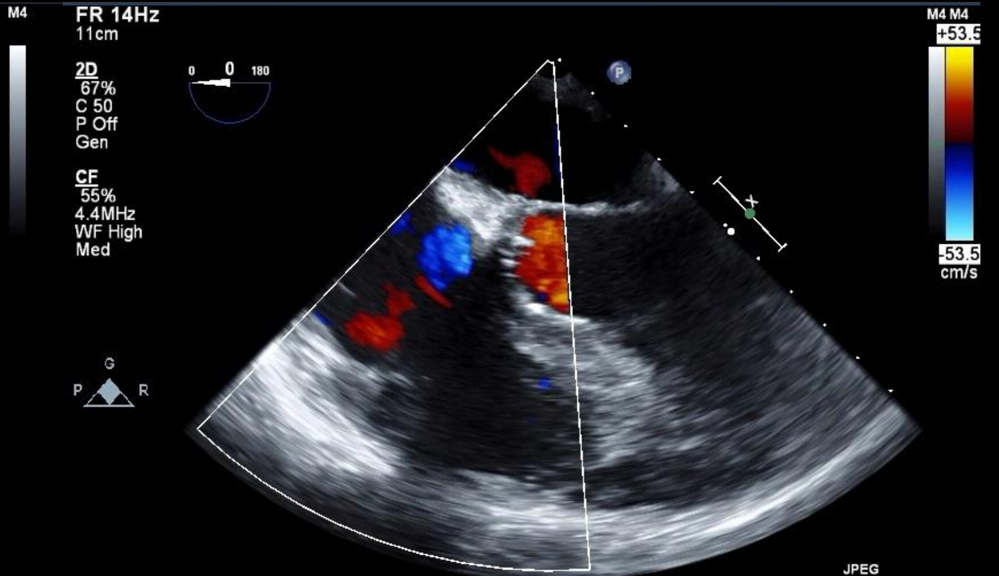
2-D ECHO (TTE)

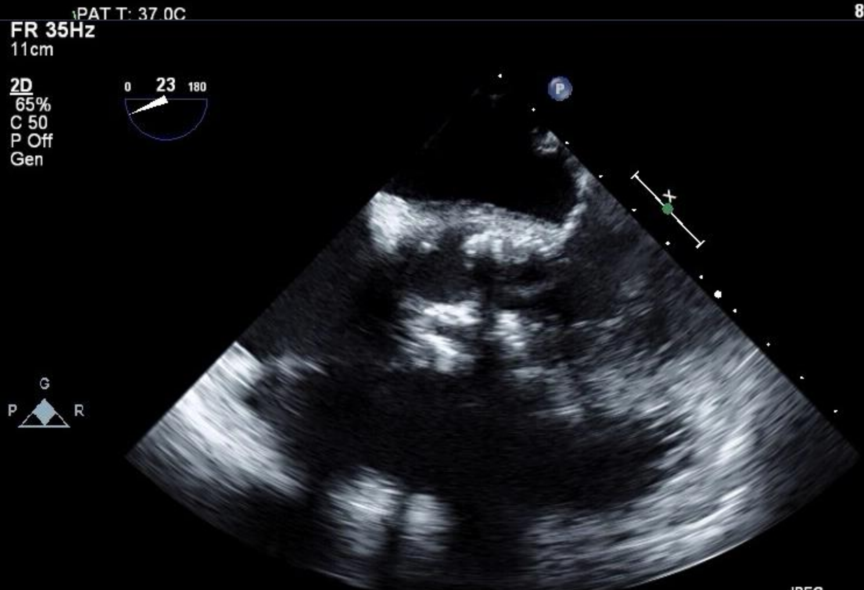


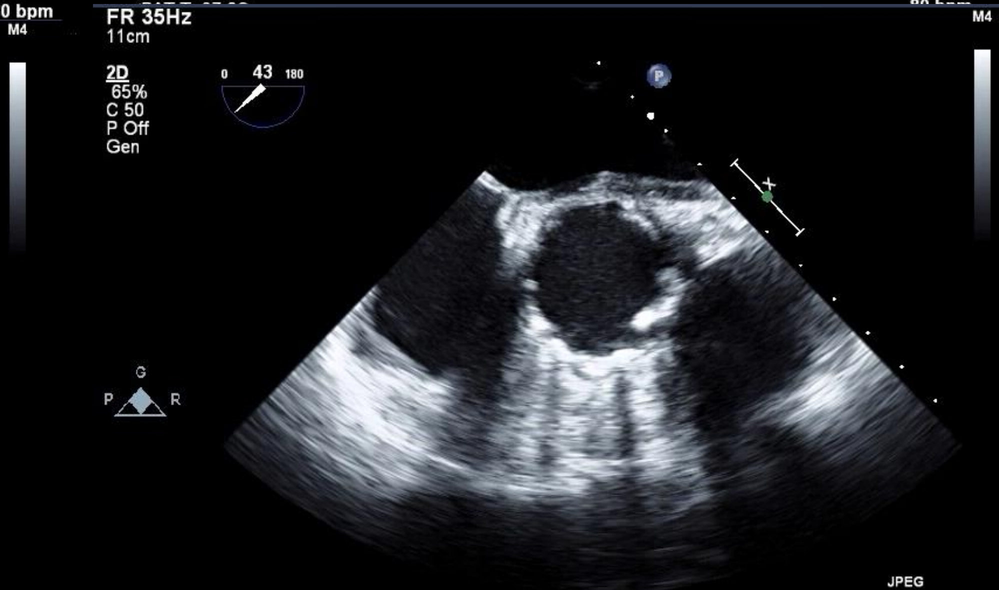
PEAK V=4.3M/SEC

2-D ECHO (TEE)









2-D ECHO

- **Comment**

  EF 32-37%, Mild diffuse hypokinesia.

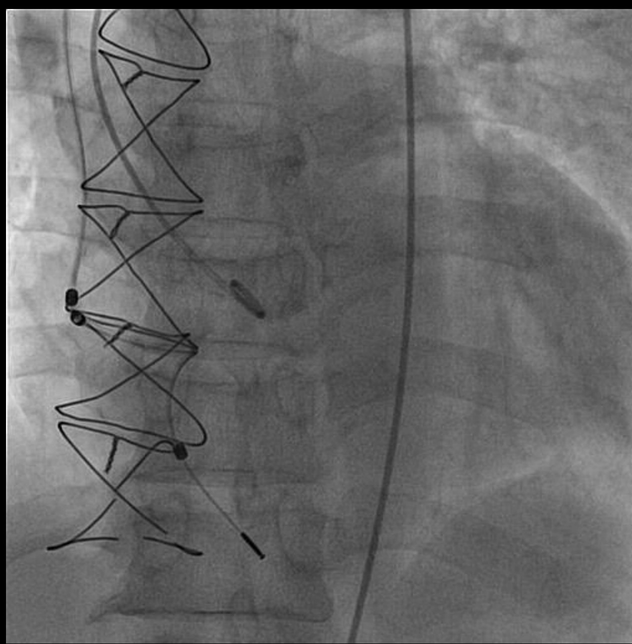
  Aortic valve: calcified and stenotic.

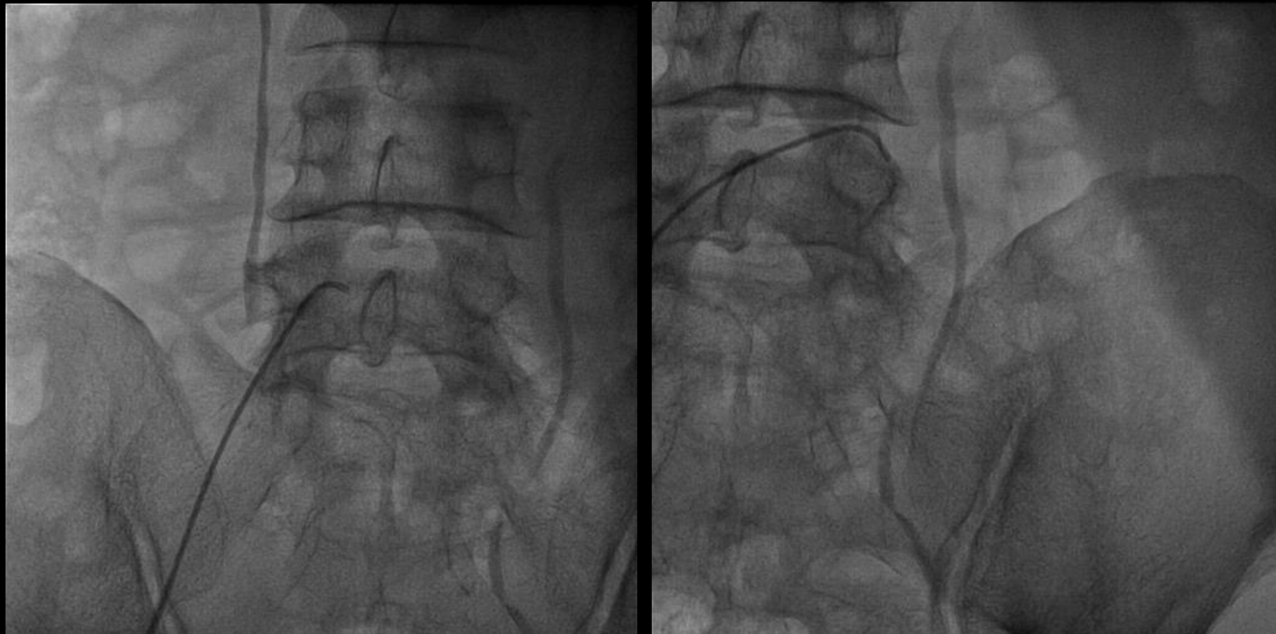
  - Aortic velocity = 4.3 m/s (▲).
  - Mean pressure gradient = 75.0 mmHg (▲).
  - Aortic area = 0.90 cm^2 (▼).
  - Mild aortic regurgitation.

- **Conclusion**

  Aortic steno-insufficiency. Mild LV systolic dysfunction.

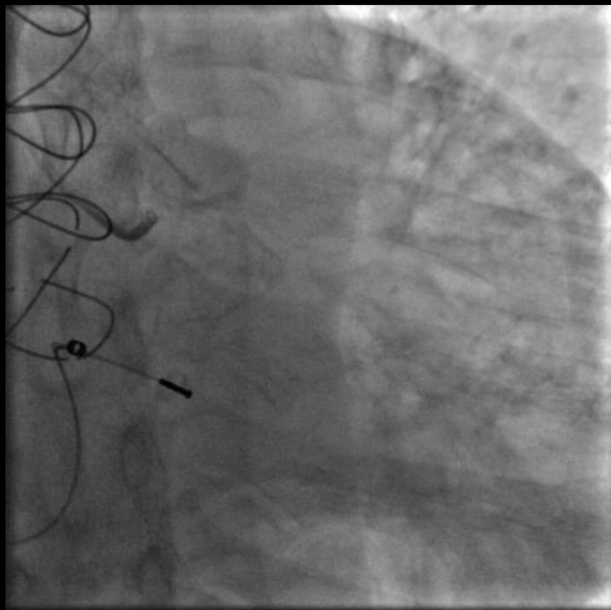
Angiography

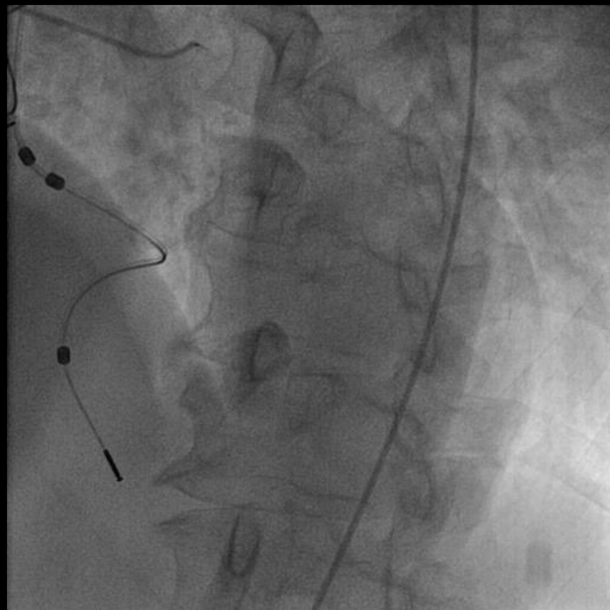


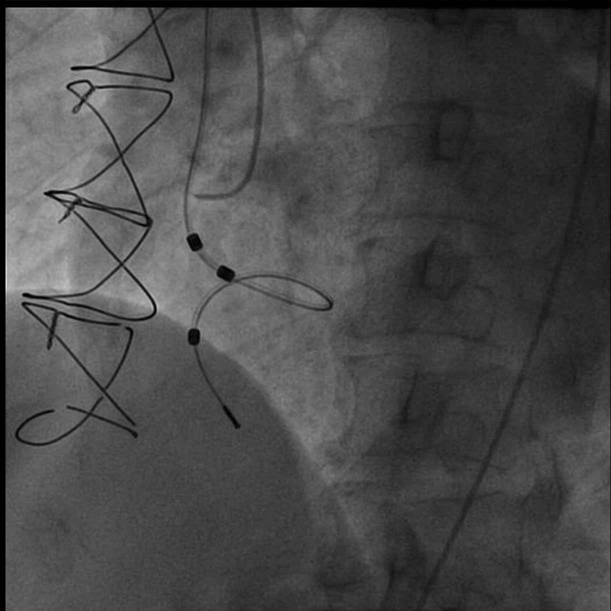


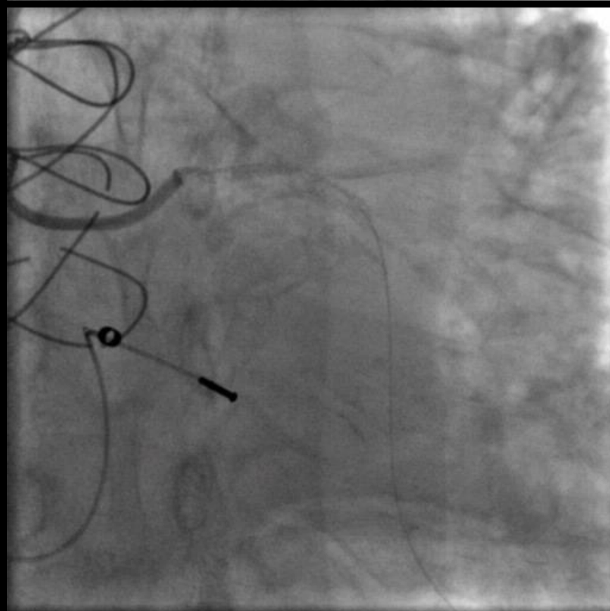
Severe calcification at aortic valve area
and limited motion of aortic valve leaflets.

CAG/PCI









LCX: os , focal, critical stenosis up to 90%.
Direct stenting : Xient prime =3.0mmx18mm

Diagnosis

- Severe aortic stenosis.
  - Mild aortic regurgitation.
  - s/p AVR(SJ 25mm)
  - s/p Homograft aortic valve replacement (2006/04)
  - s/p Coronary bypass graft op(SVG-LAD) (2006/04)

- Behcet's disease.

- Coronary arterial disesase
  - s/p PCI on Lt. main to LCXos

STS risk score

- **Perioperative mortality**
  - **Mortality: 2.58%.**
  - Low STS risk score.

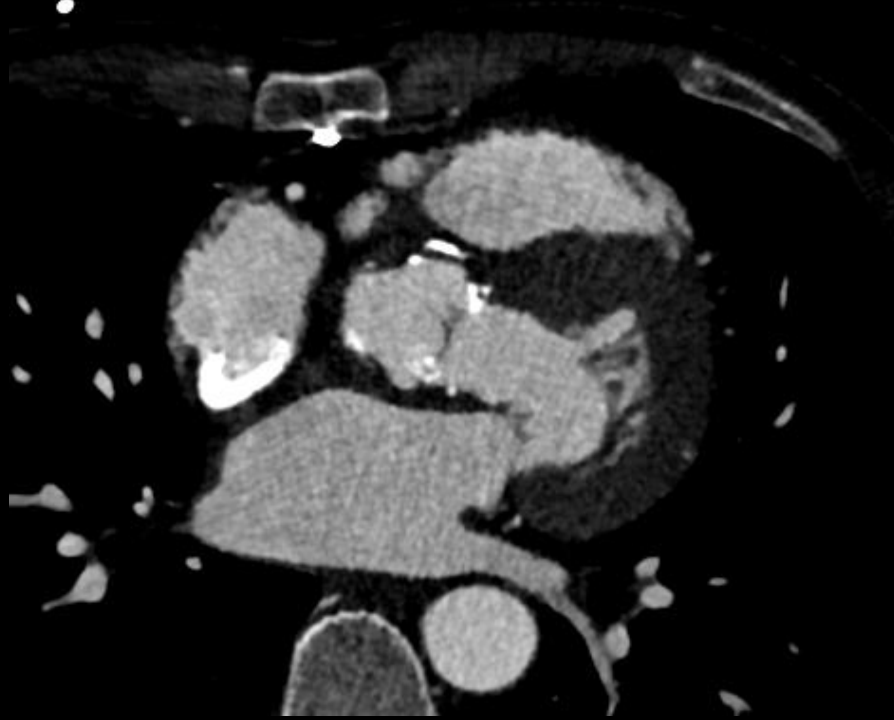
- **Patient's medical history**
  - History of open heart surgery for twice.
  - Underlying autoimmune inflammatory disease.
  - Not an appropriate candidate for **conventional aortic valve replacement op**.

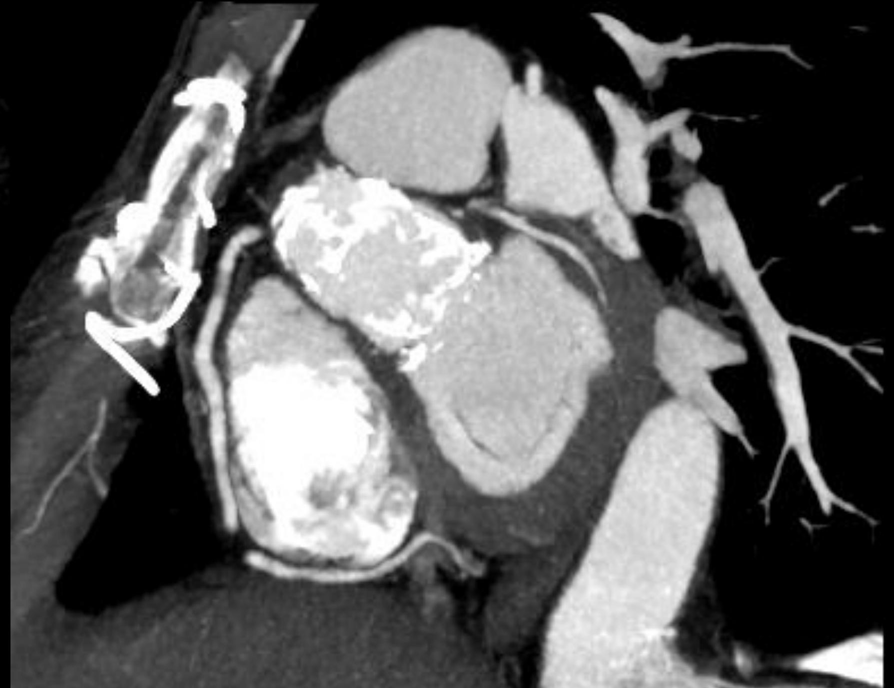
Treatment option

Non-surgical aortic valve replacement.

-> **TAVR (Transcatheter Aortic Heart Valve Replacement)**.

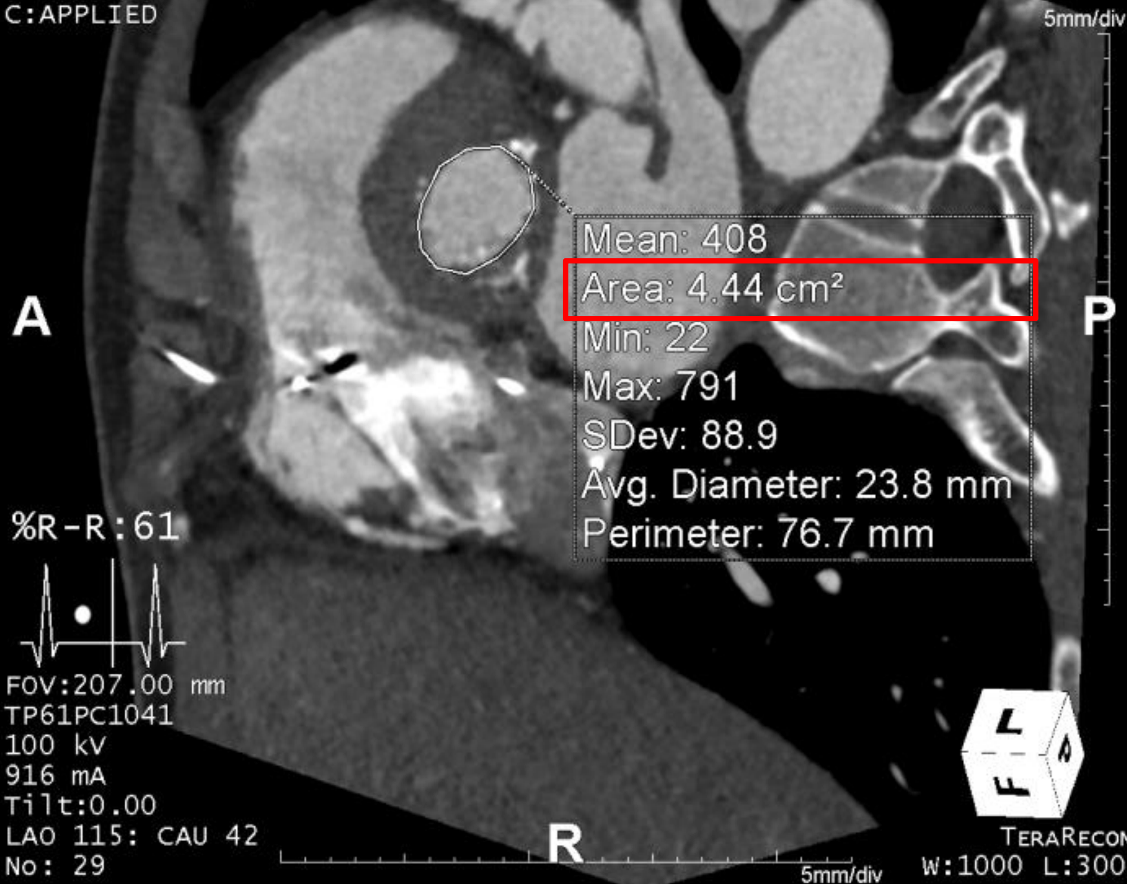
Coronary CT



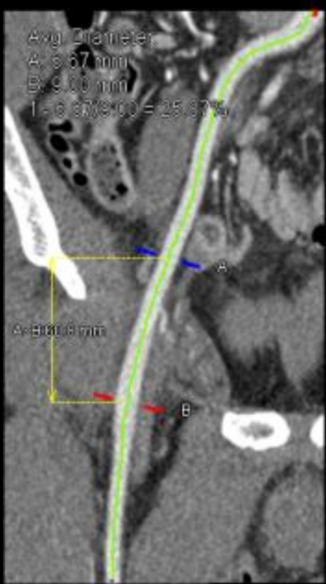


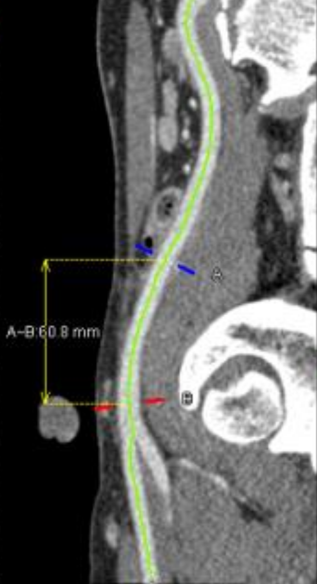
Multiple aortic valve leaflet dense calcifications.

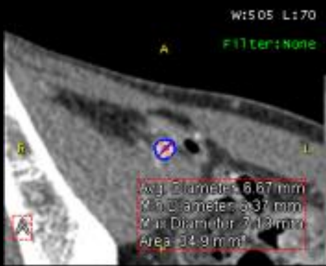
Coronary CT

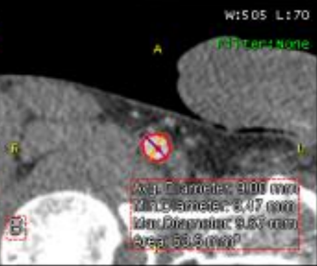


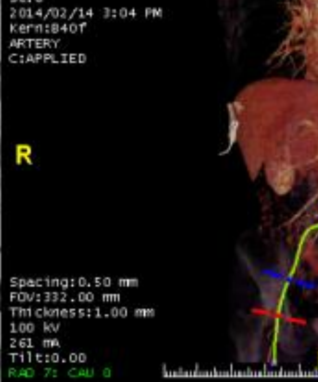
CT scan

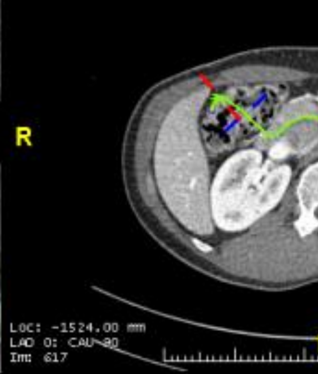


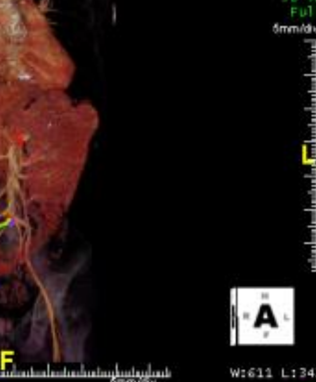


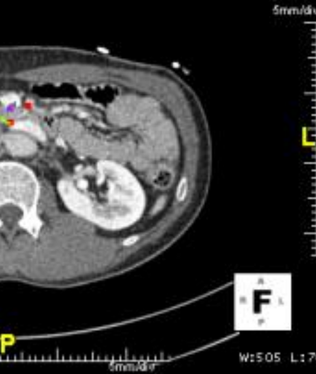


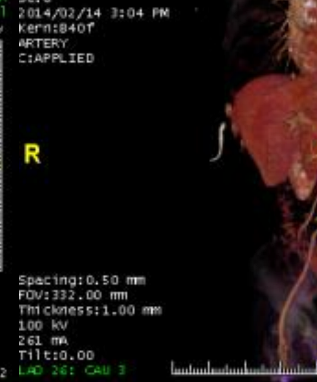


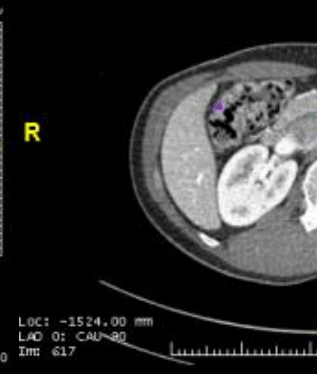


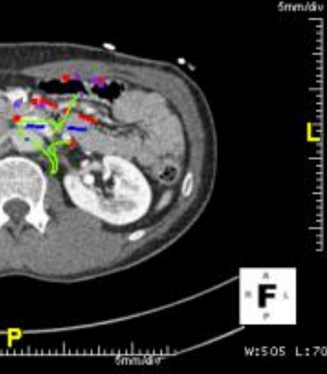




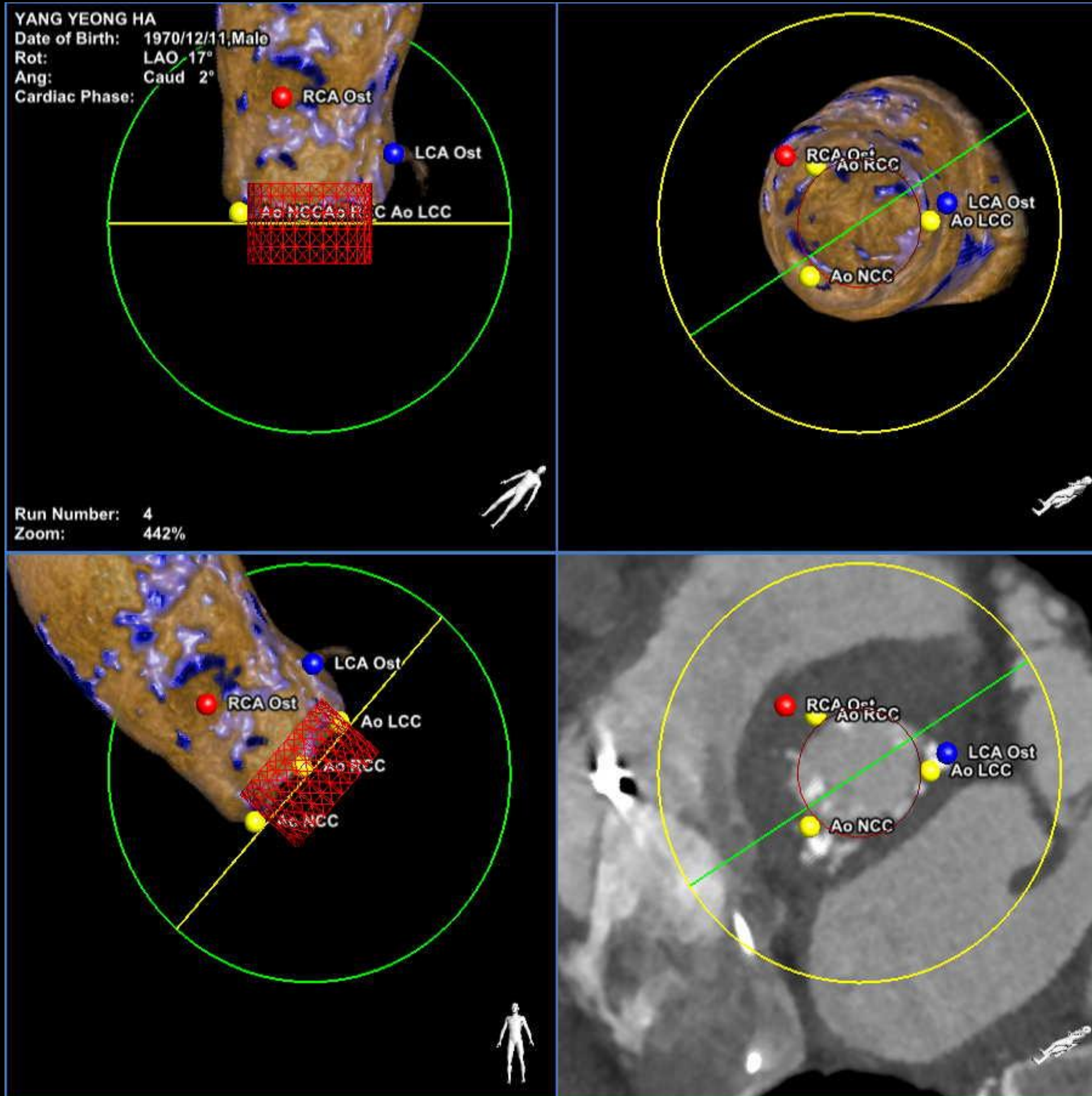






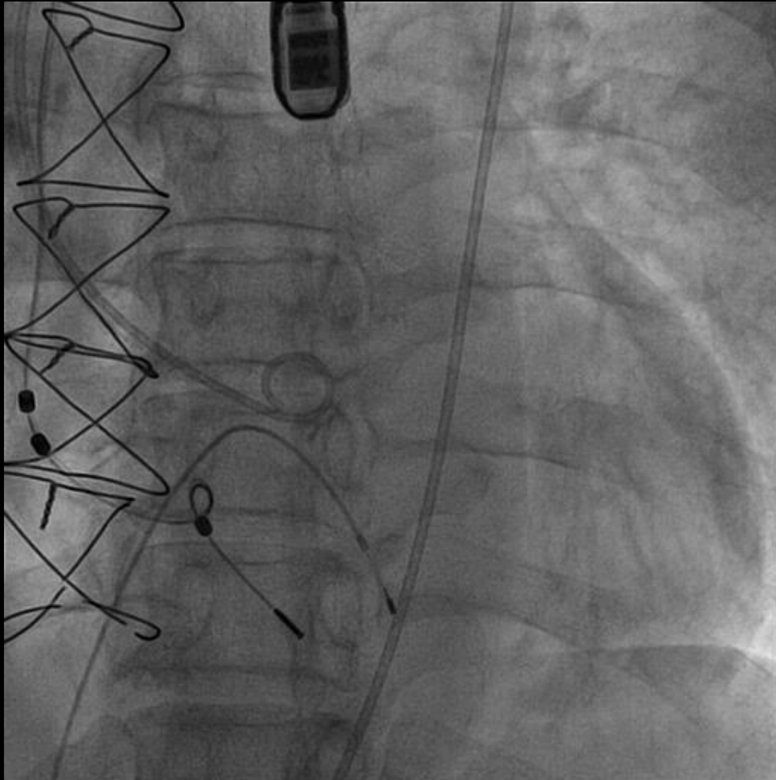


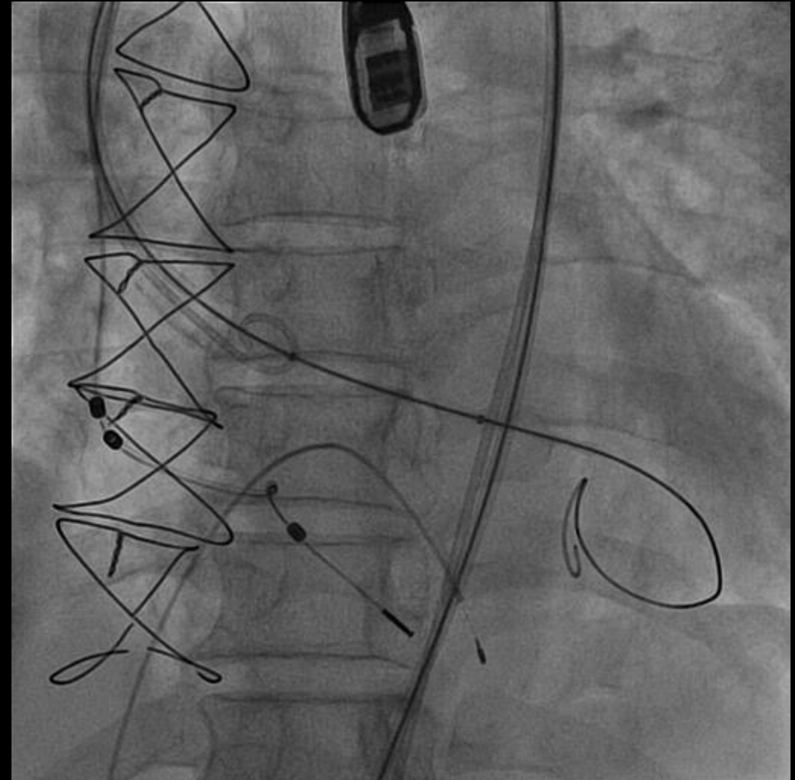
Coronary CT simulation for TAVI



25mm
Aortic valve insertion.

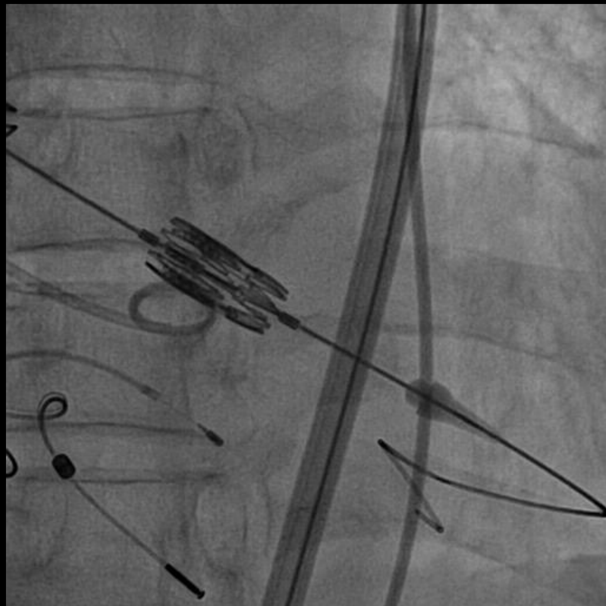
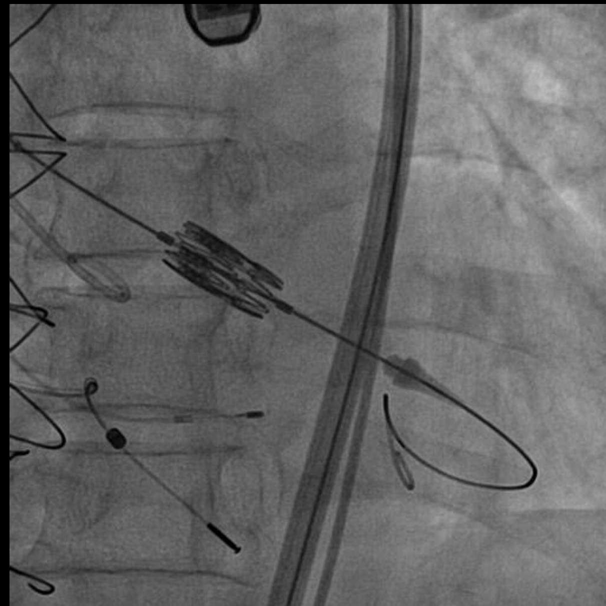
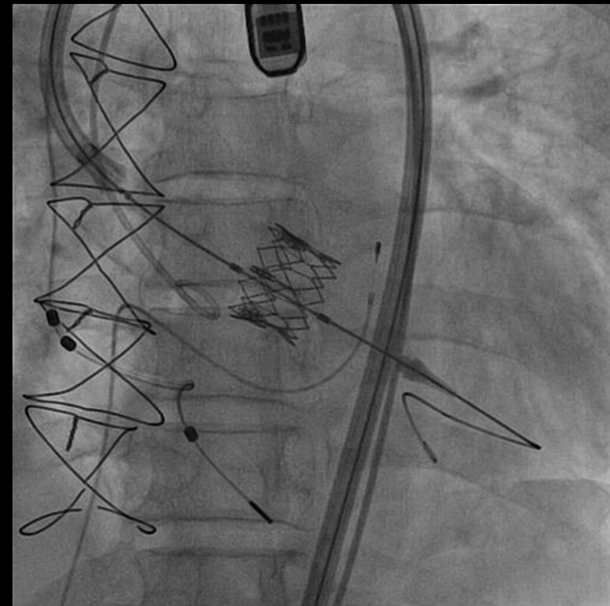
TAVR (Ballooning)





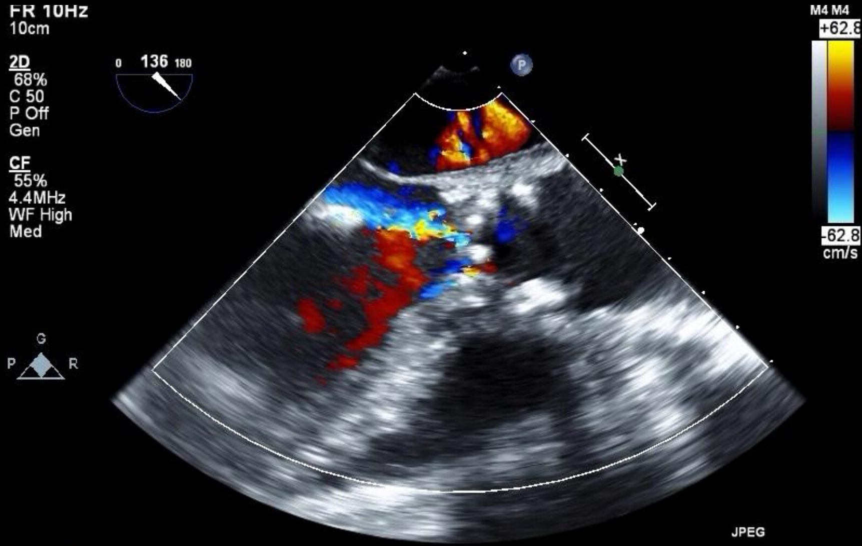
BAV performed by 24mm x 40mm balloon

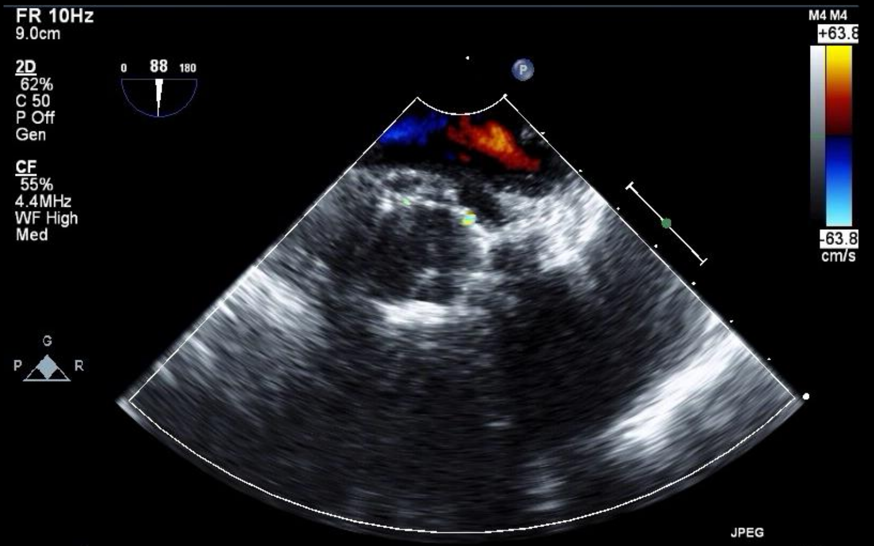
TAVR (THV deployment)

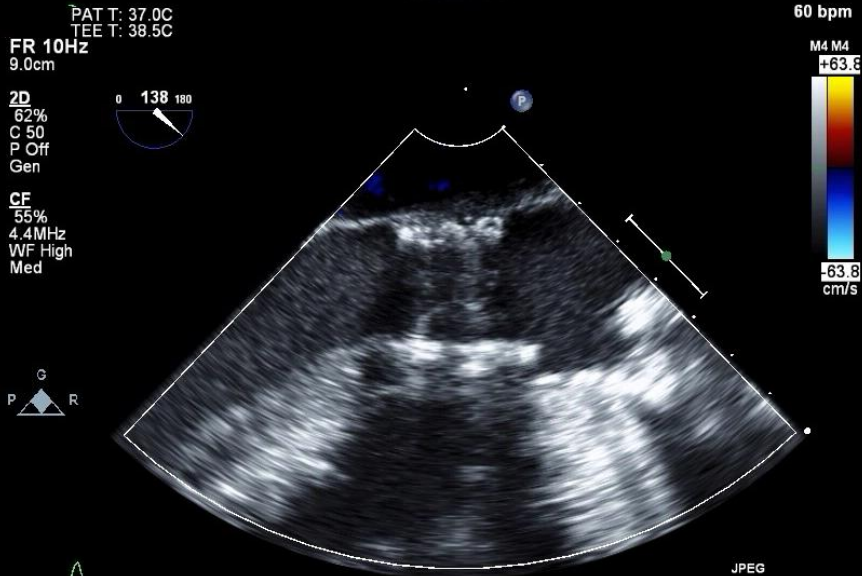




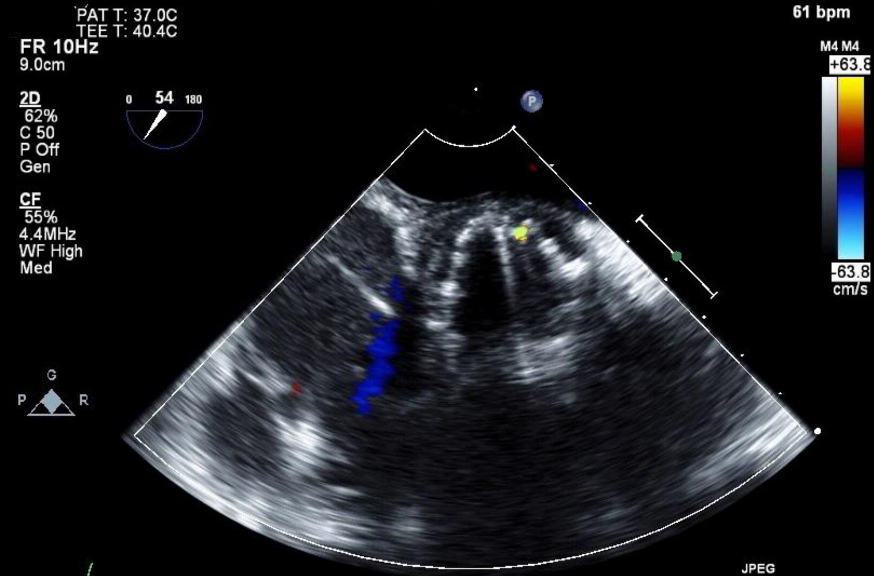
SapienXT valve (26mm) deployment.

Intraop TEE

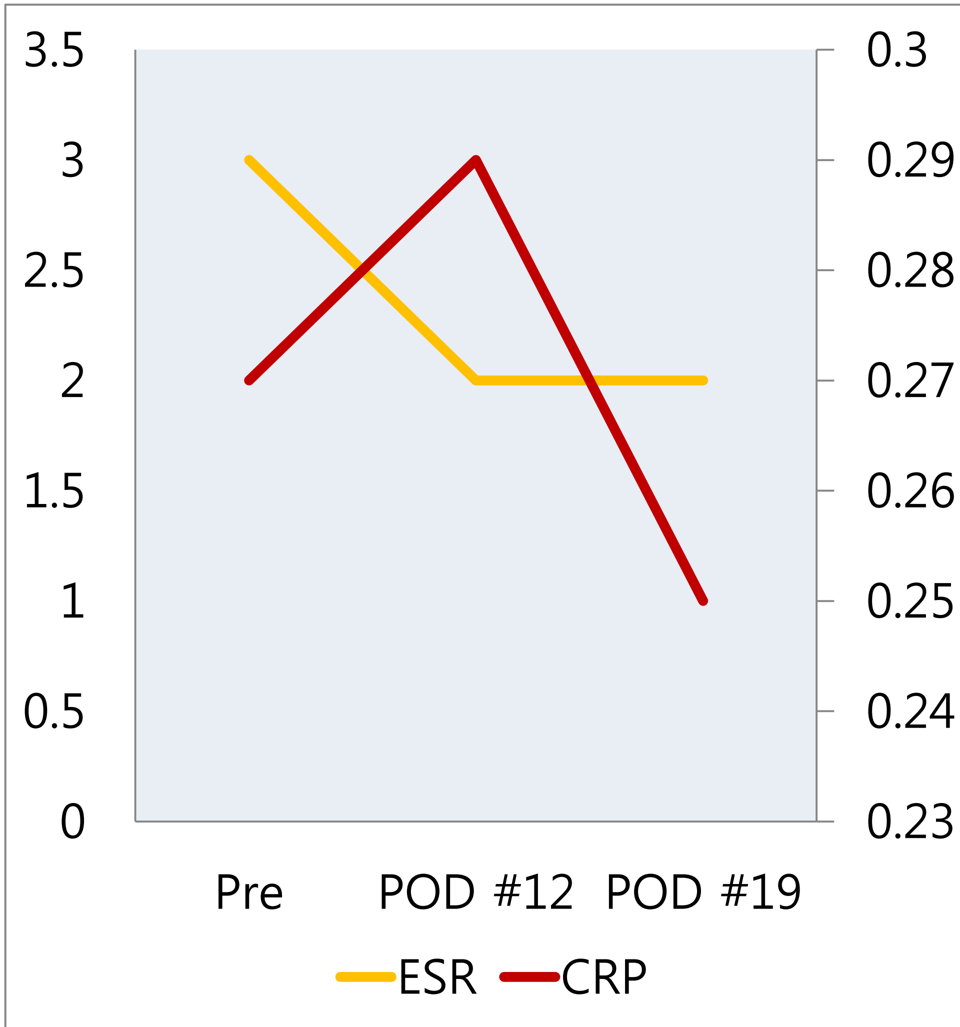




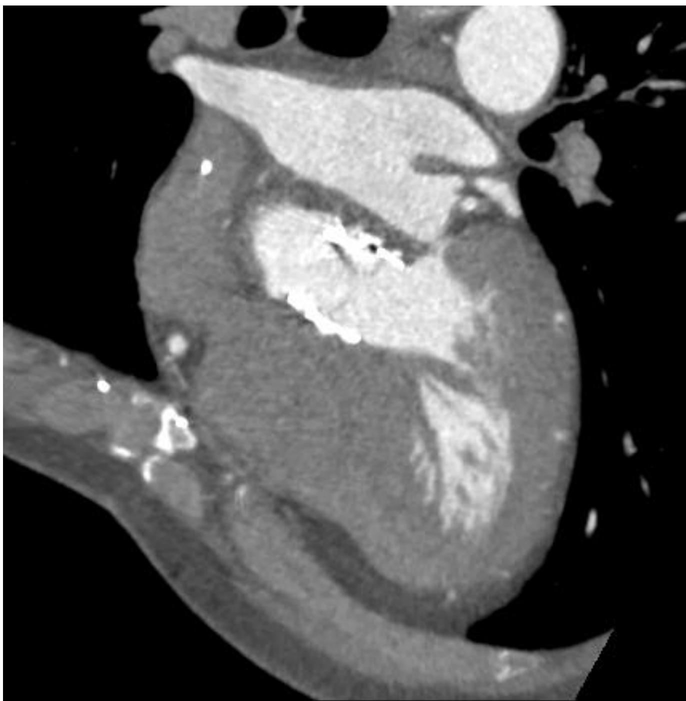
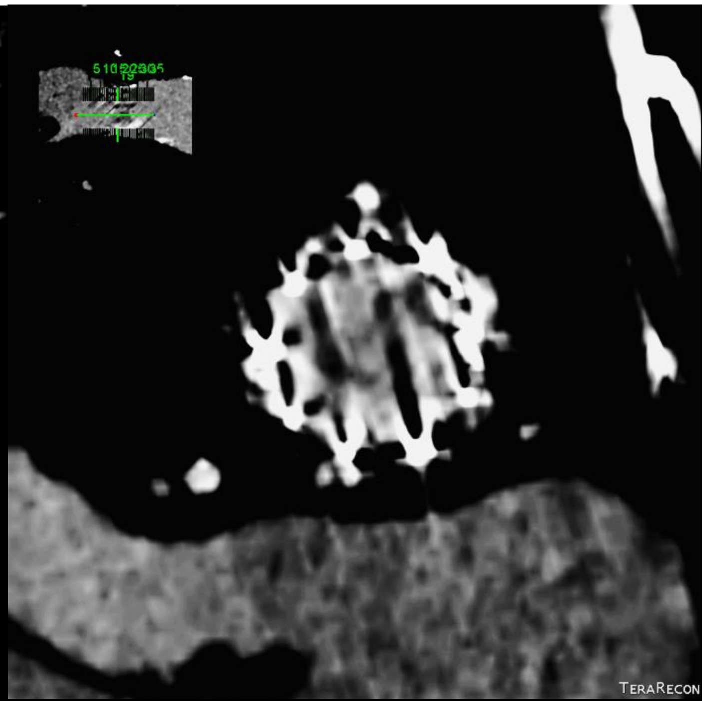
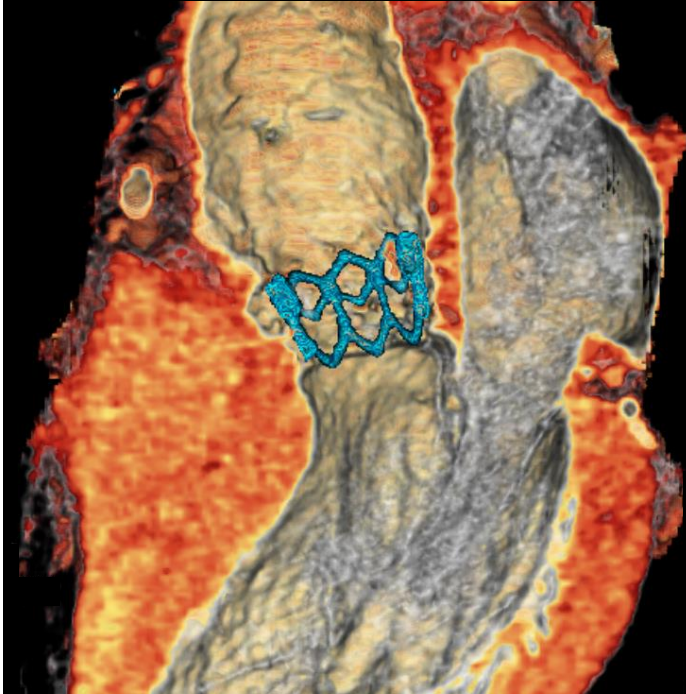
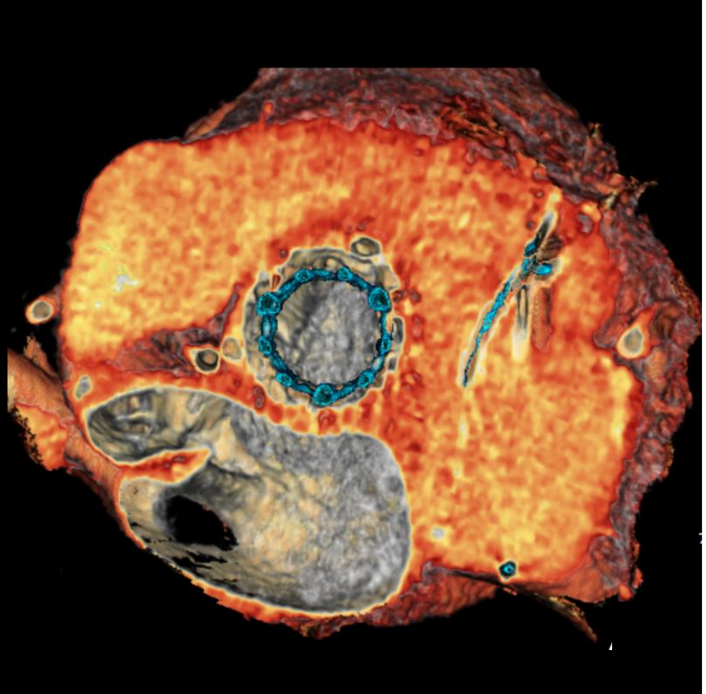




Progress



- **Medication**
  - **Cardiology**: Aspirin, Clopidogrel, Lasix 20mg, Spironolactone 12.5mg, Bisoprolol 1.25mg.
  - **Rheumatology**: Azathioprine 100mg, Prednisolone 10mg



TeraRecon



Take Home Message

1. **Cardiac involvement are rare but often disastrous in Behcet's disease.**
2. **Although many surgical considerations are needed for surgical AVR, diagnosis of Behcet's disease often delayed after the primary AVR.**
3. **Aortic root replacement (ARR) has achieved favorable clinical outcomes for primary/re-operations in Behcet's disease.**
4. **TAVR could be an alternative strategy after the repeated open-heart operation (AVR and ARR).**

Thank you for your attention!