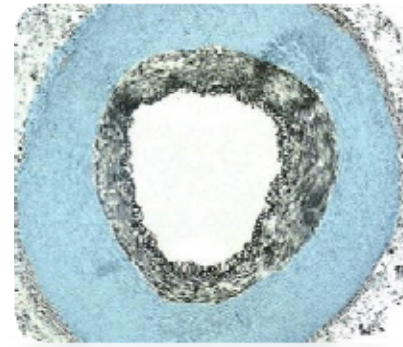
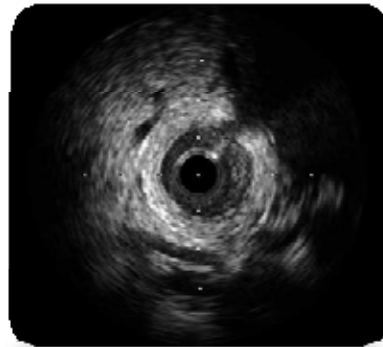


IVUS imaging of renal artery fibromuscular dysplasia in a pediatric patients



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Disclosure Information

Supported by research grants from

- None

CASE 1 : the older of the two sisters



Childhood Idyll
William-Adolphe Bouguereau

Chief complaint

Dyspnea : 2 month ago, exertional

Present illness

A 11-year-old girl was admitted to our hospital because of exertional dyspnea. She had no coronary risk factors.

PMHx, FHx, SHx

- Past medical history : Bronchial asthma
- Familial history : none
- Social history : none
- Herb medication (+) 2month ago

ROS and P / E

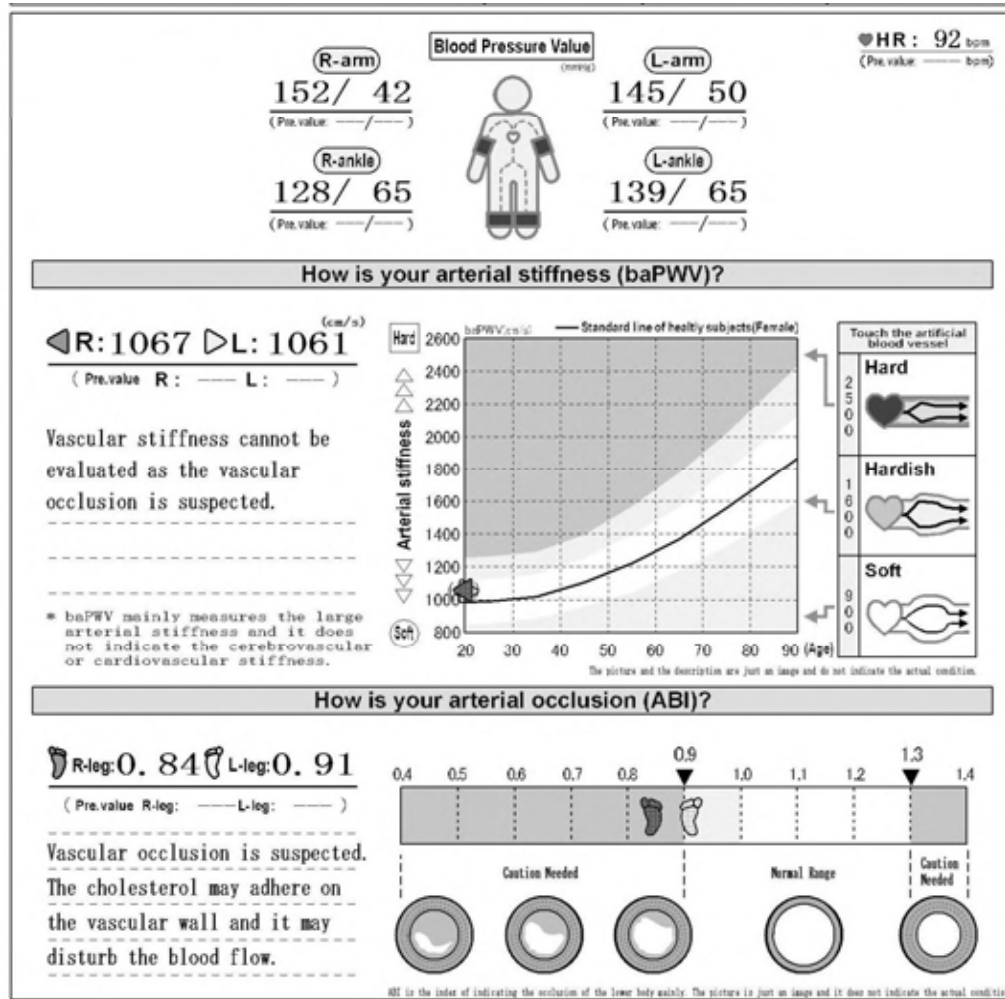
ROS

- URI Sx.(-), fever(-), anorexia (+)

P/E

- both mild anemic sclera (+/+)
- not palpable cervical LNs
- Coarse BS with both lung wheezing
- RHB without murmur
- not palpable abdominal mass / not palpable liver or spleen
- pitting edema(-)
- Weight loss 4kg (2 month)

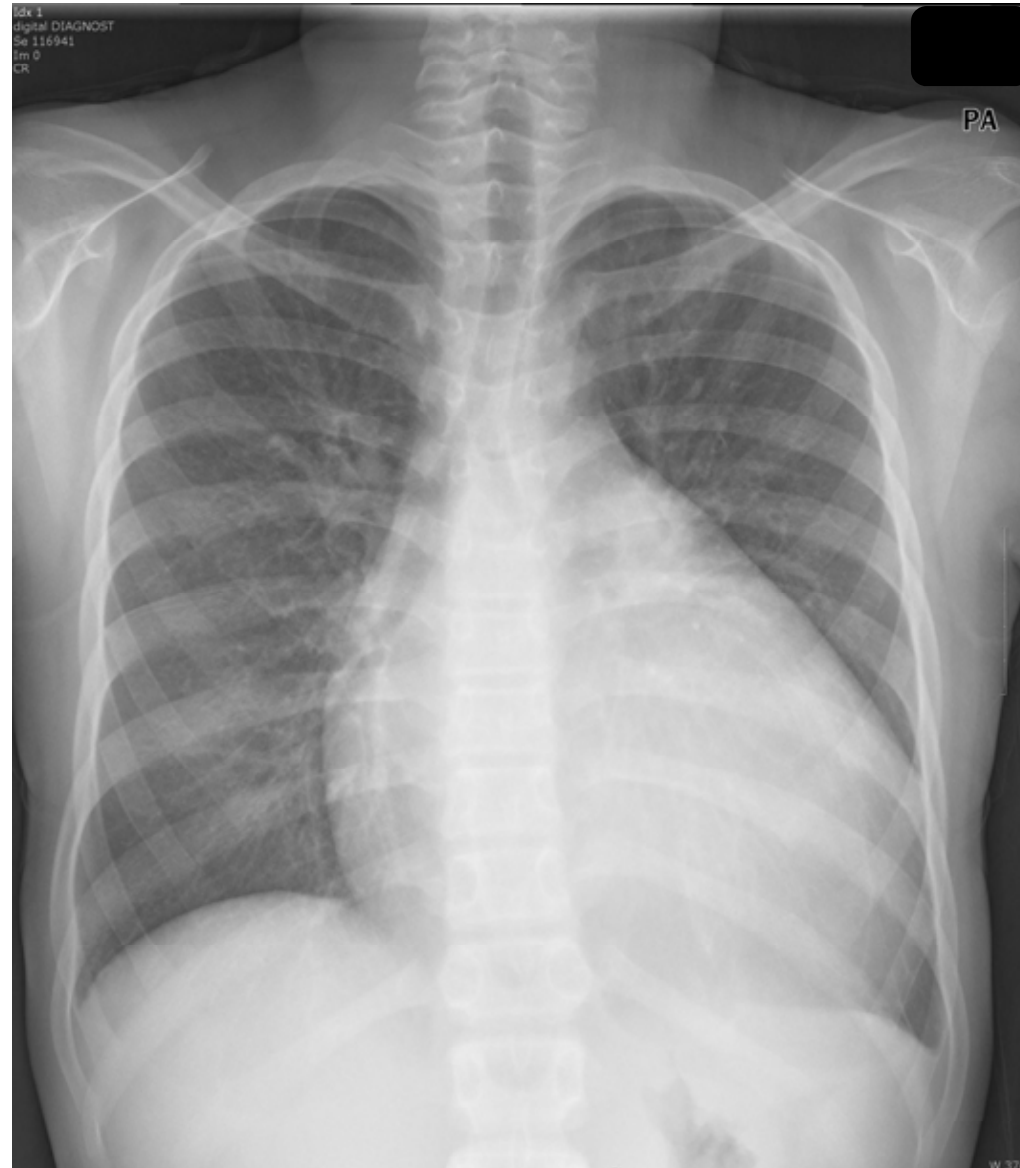
High blood pressure, abnormal ABI



Antihypertensive medication

- Nifedipine 60mg qd
- Furosemide 40mg tid
- Spironolactone 25 mg qd
- Carvedilol 6.25 mg bid

Chest X-ray :



no active lesion in lung / cardiomegaly - CT ratio 0.70

LVE 66mm, EF 30 %, LA, RA size : normal



echoCG

1) Dilated LV cavity size with moderate to severe LV systolic dysfunction

: calculated EF = 25 %, EF by modified simpson's method = 24 %

Visually estimated **EF = 30 %**

2) Global hypokinesia

Normal LV wall thicknes

3) Trivial MR (dP = 34 mmHg

4) Trivial TR with estimated RV systolic pressure of 34 mmHg

Normal IVC size without IVC plethora : assumed RAp = 10 mmH

5) Indetermined diastolic function due to E, A wave summatio

6) LAVI (by Simpson) = 24.4 ml/m²

7) EPSS = 2.61 cm

8) SEC in LV

[Conclusion]

LVE with moderate to severe LV systolic dysfunction

LVIDs	58	mm(25~41)	IVS motion		
LVIDd	66	mm(35~55)	thickness	10	mm(7~11)
EF 24	%(>50)		LVPW motion		
(Visually	30	%)	thickness	10	mm(7~11)
Aorta	27	mm(20~37)	E/A ratio		
LA 22	mm(19~38)	DT	msec출		

Blood lab

Pro-BNP : **11338**pg/ml
Sodium 138
Potassium ▼3.3
Chloride 99
Glucose ▲144
Creatinine 0.96
BUN ▲25.9
Uric acid 6.2
Cholesterol 138
Protein 6.8
Albumin 4.1
AST(SGOT) 21
ALT(SGPT) 13
Alk. phosphatase(ALP) ▲266
Bilirubin total 0.61
eGFR
Digoxin ▼0.40(ng/mL)

2ndary HTN work up

ACTH E.B. ▲71.5 10-60 pg/ml
Cortisol (Serum) S.B. 20.7 morning serum:9.4-26.1ug/dl ug/dl
2013-07-19 2013-07-18 13:39:31
evening serum:1.8-12.7ug/dl
Renin(Basal) E.B. 73.05 supine:0.15-2.33 ng/ml/hr 2013-07-22
2013-07-18 13:39:30
upright:1.31-3.95
Aldosteron/Renin ratio E.B. 5.91
Aldosteron(Basal) S.B. 432 adult supine:1.0 - 10.5 ng/dl 2013-
07-18 2013-07-18 13:39:30
upright:3.4 - 27.3
child at birth :30 - 190
1 month- 2years:2.0 - 110
3 - 16 years:1.2 - 34.0

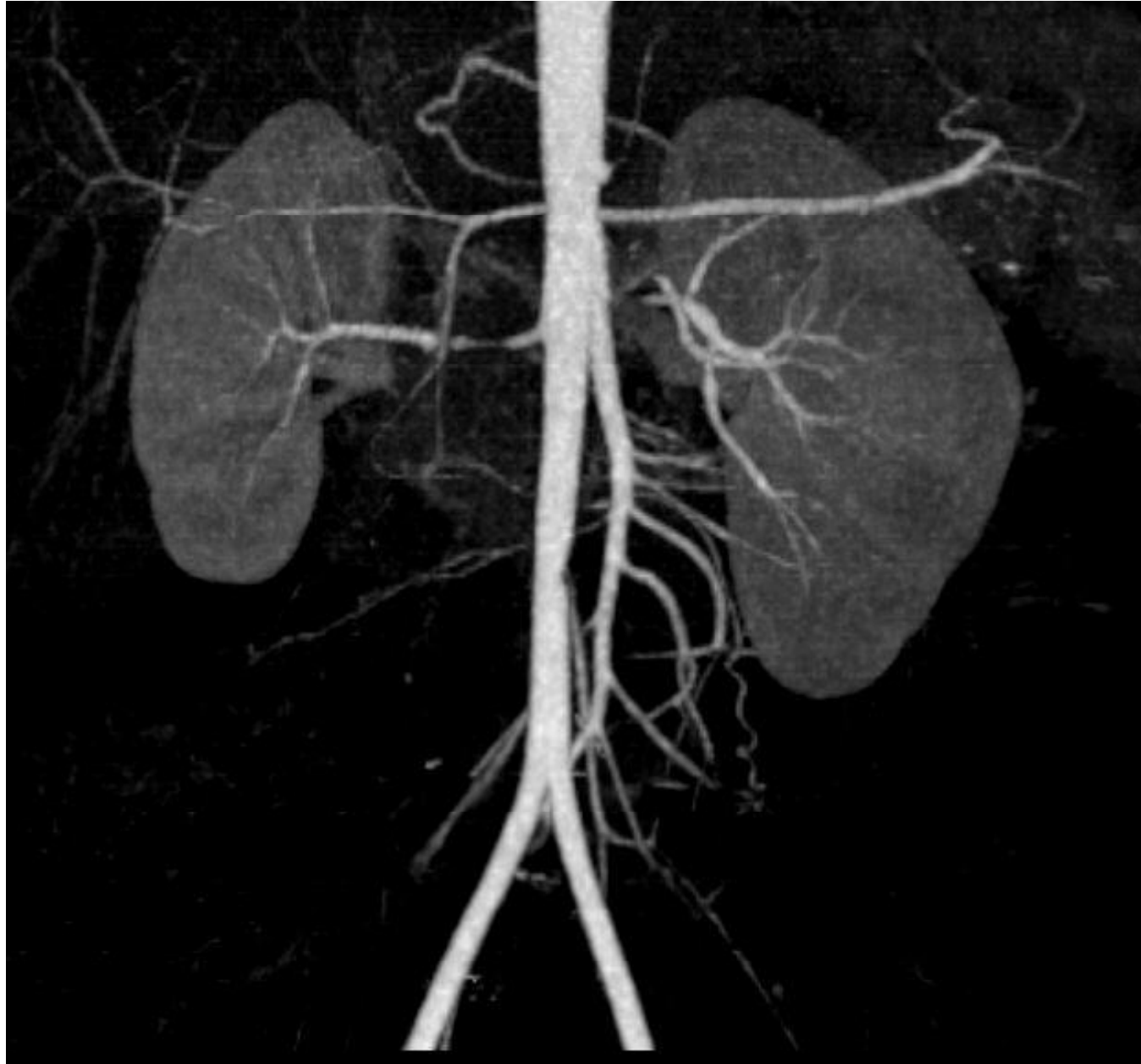
CT angiography

1. multifocal stenosis in both renal arteries
2. suspicious variceal vessel at Lt. perirenal area connected with Lt. renal segmental artery.
--> R/O AV fistula.
3. small size of Rt. kidney more than Lt. kidney but normal enhancement
4. otherwise no remarkable findings.

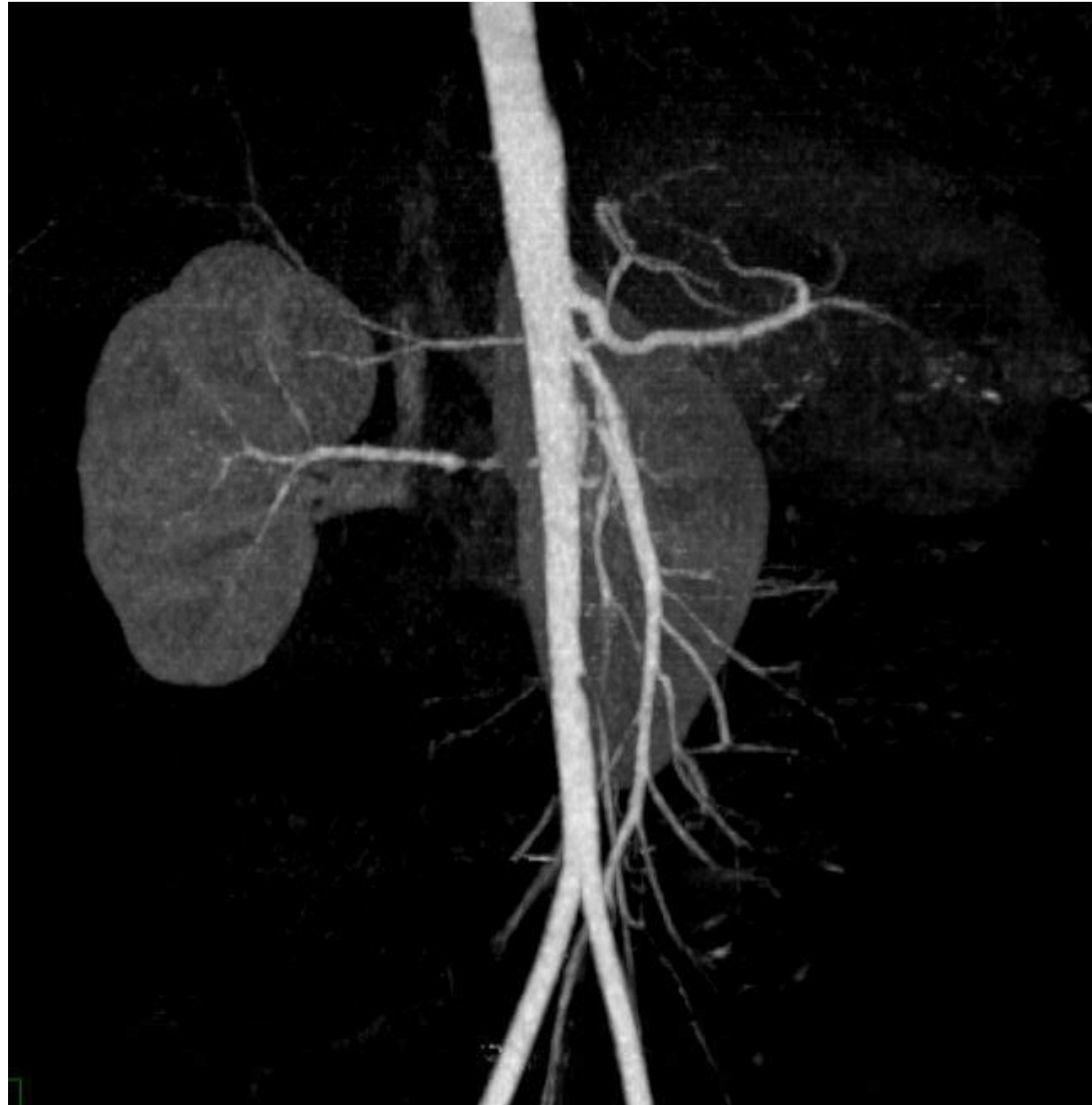
CTA : multifocal stenosis in both renal arteries



CTA : multifocal stenosis in both renal arteries



CTA : multifocal stenosis in both renal arteries



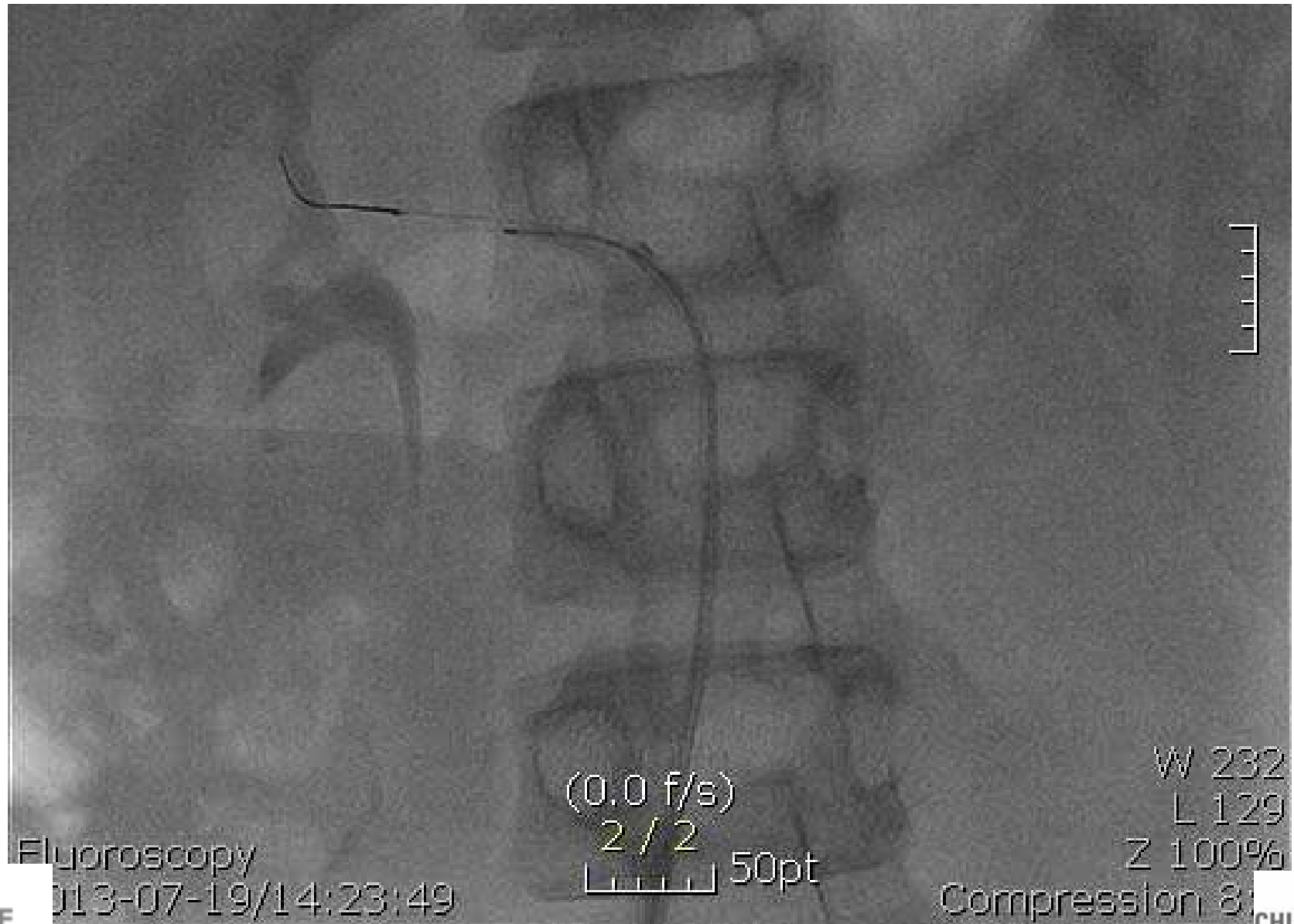
Renal angiography



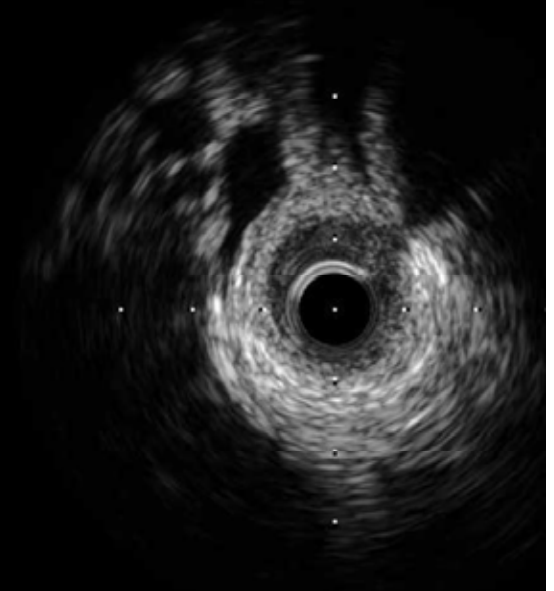
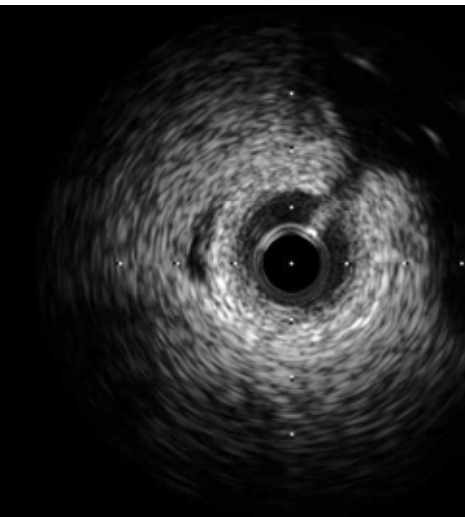
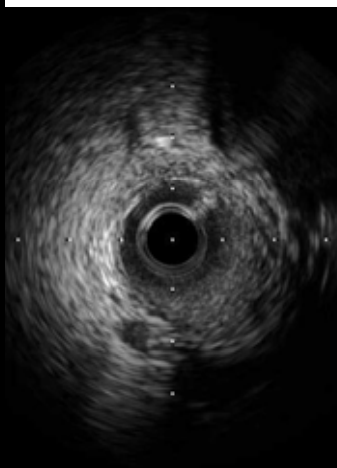
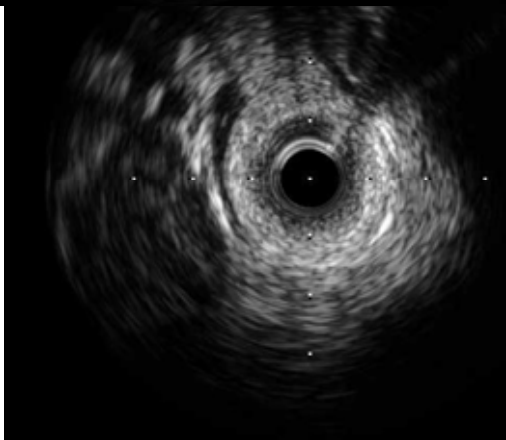
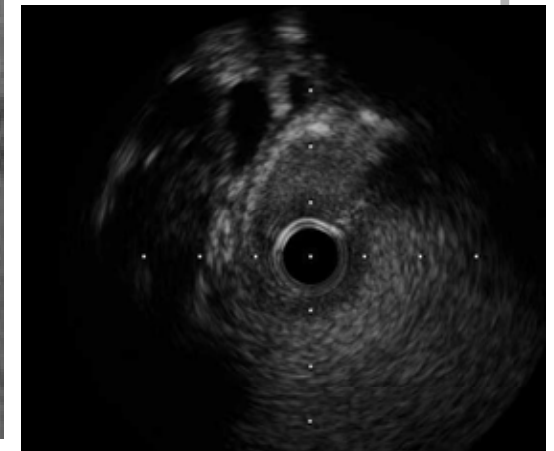
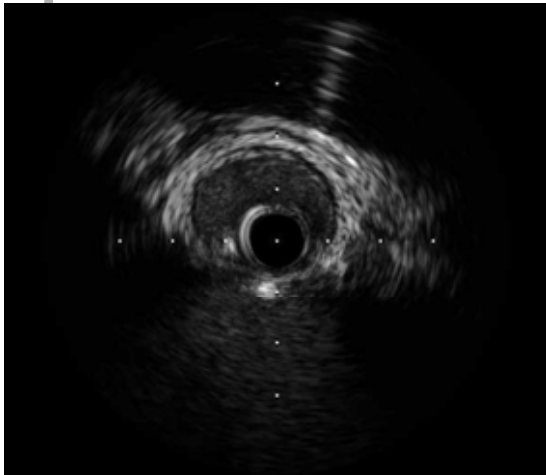
.014 wire (coronary)



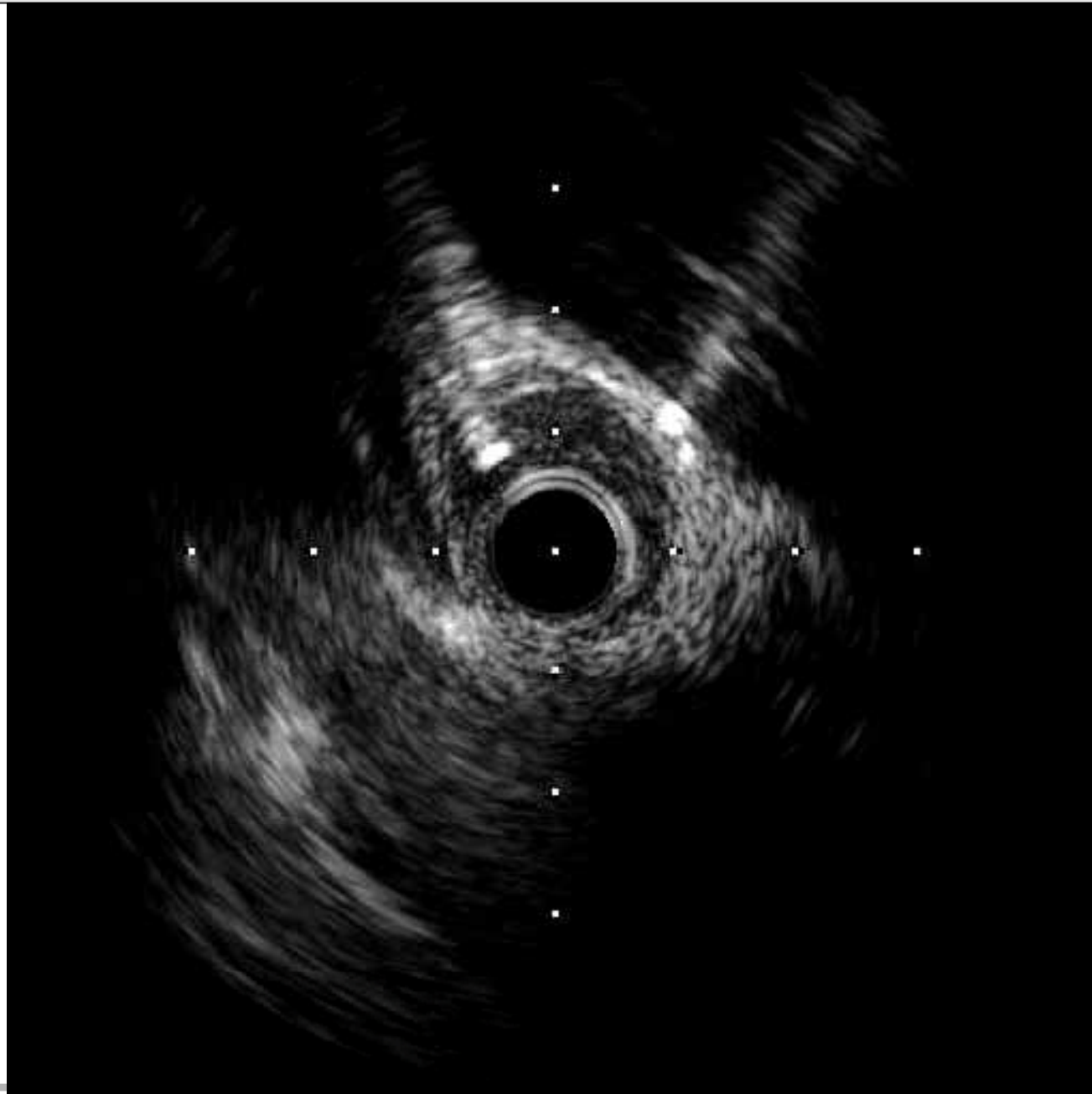
IVUS exam



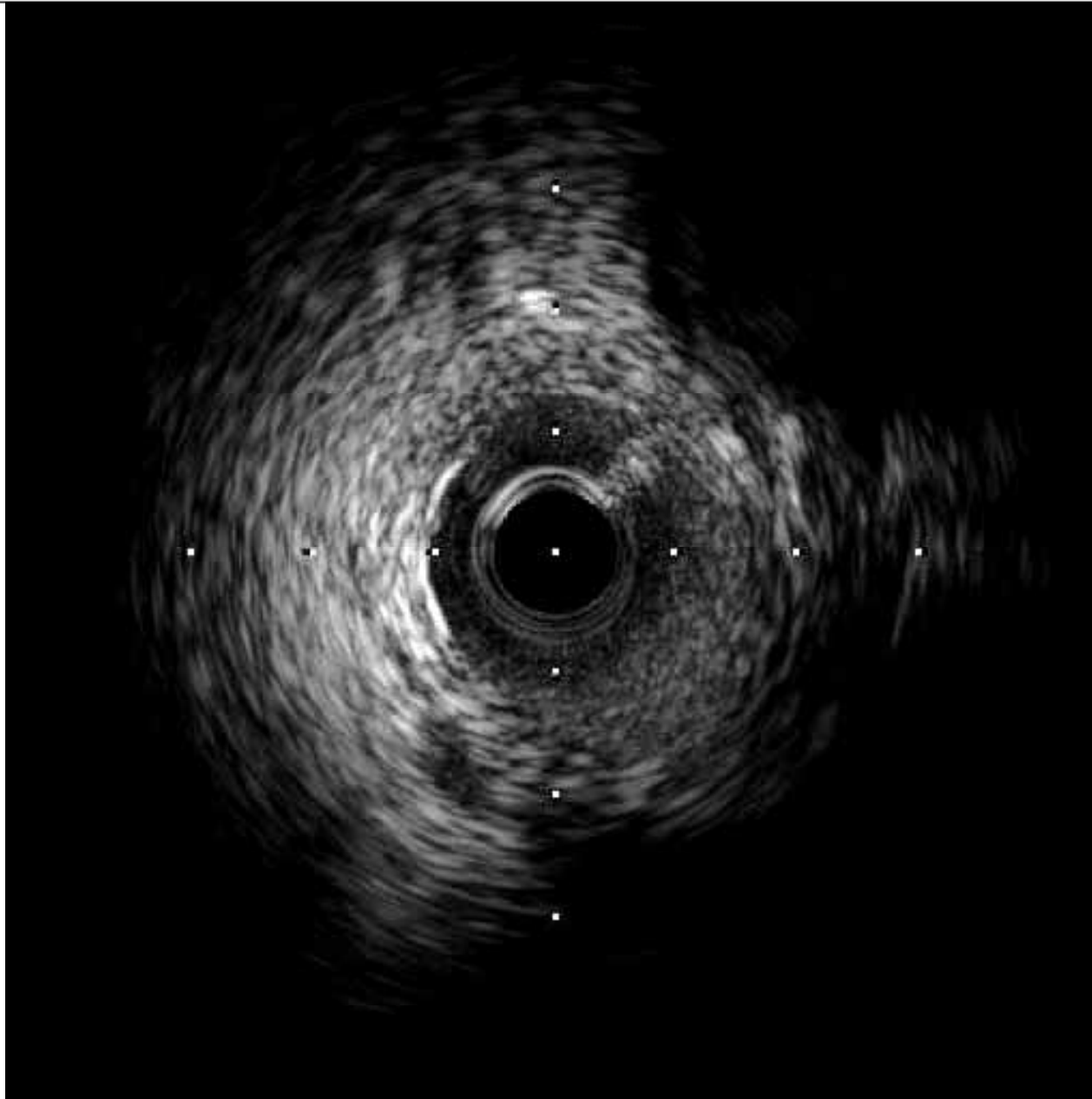
IVUS finding

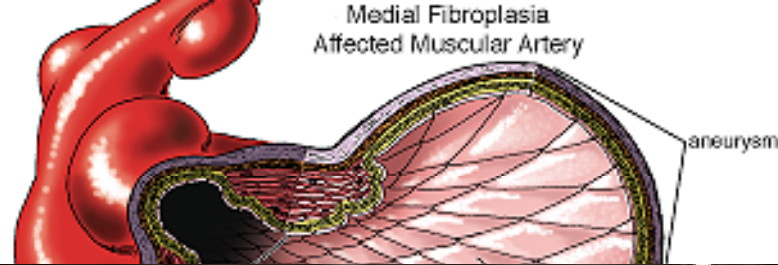
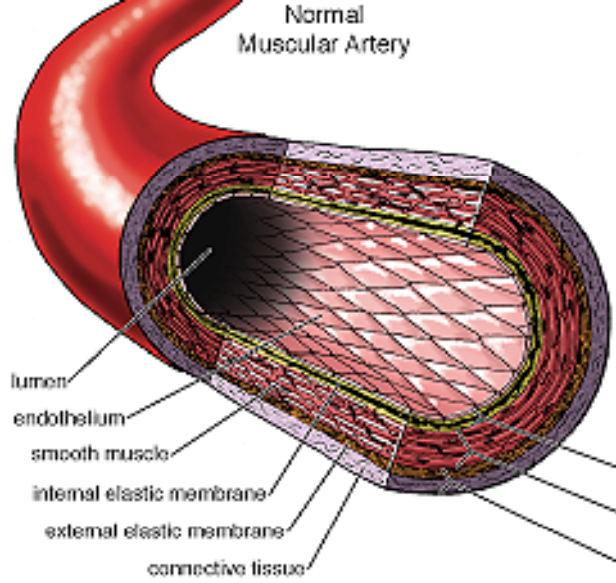


IVUS : Lt renal artery

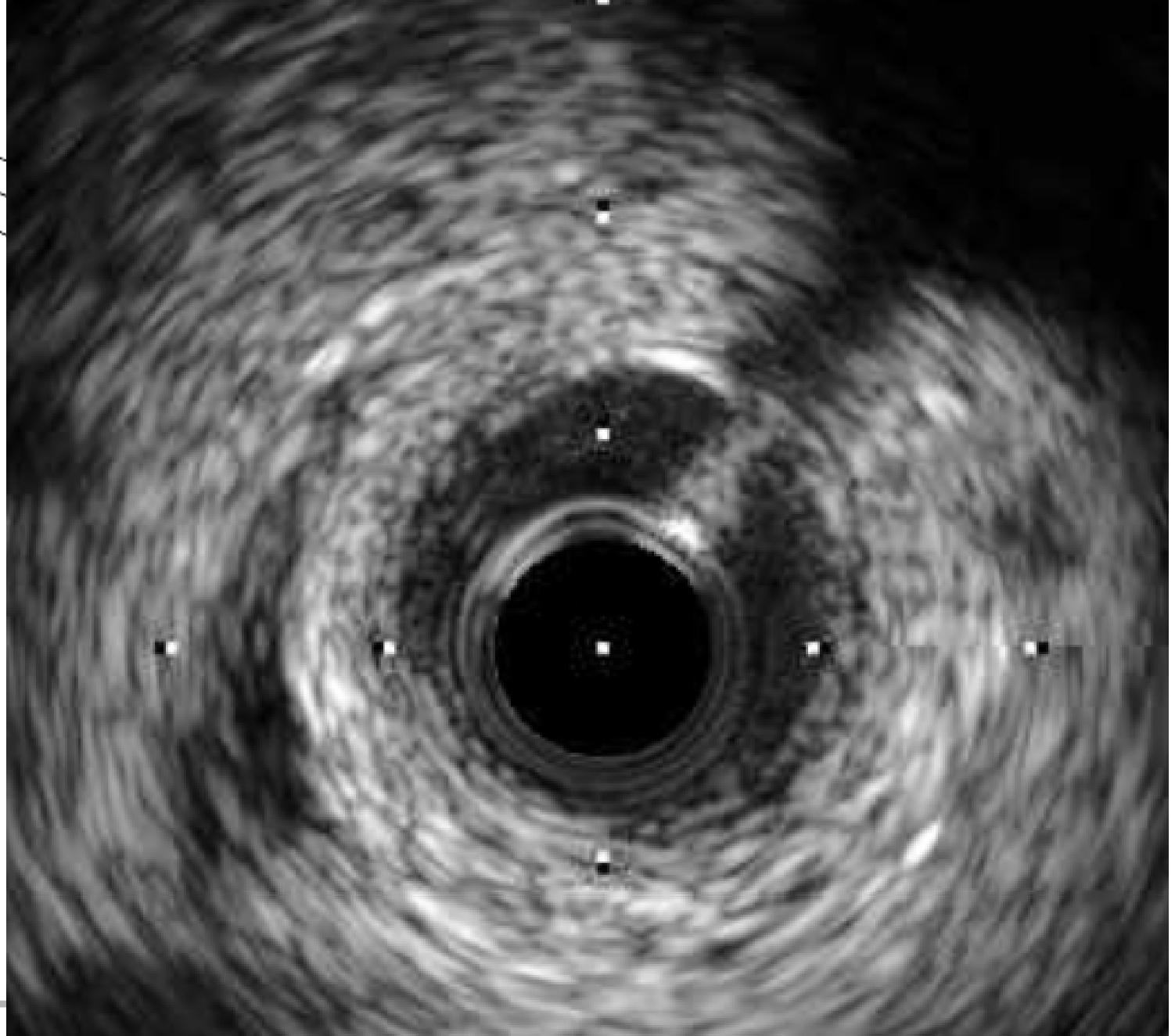


IVUS : Rt renal artery

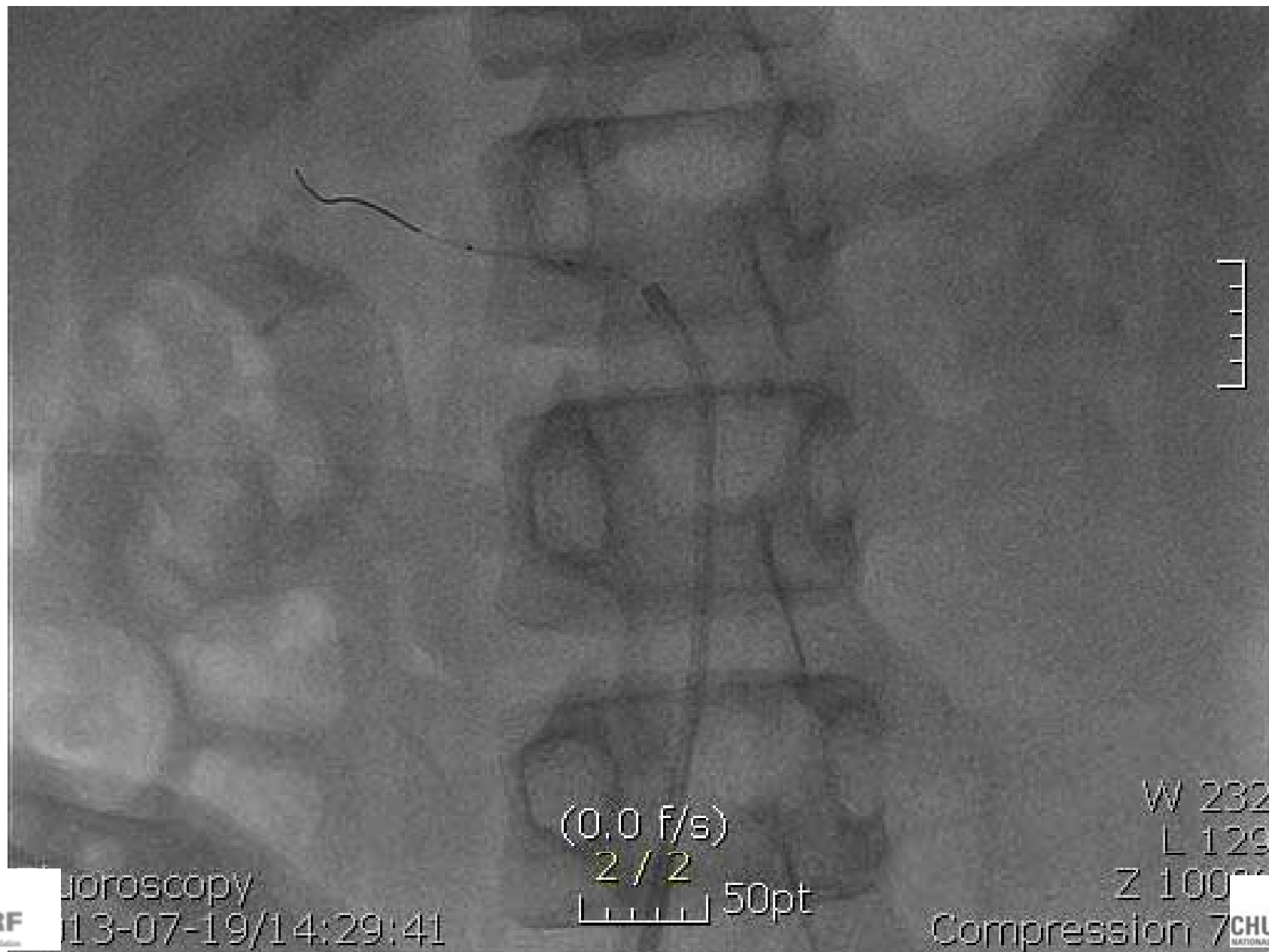




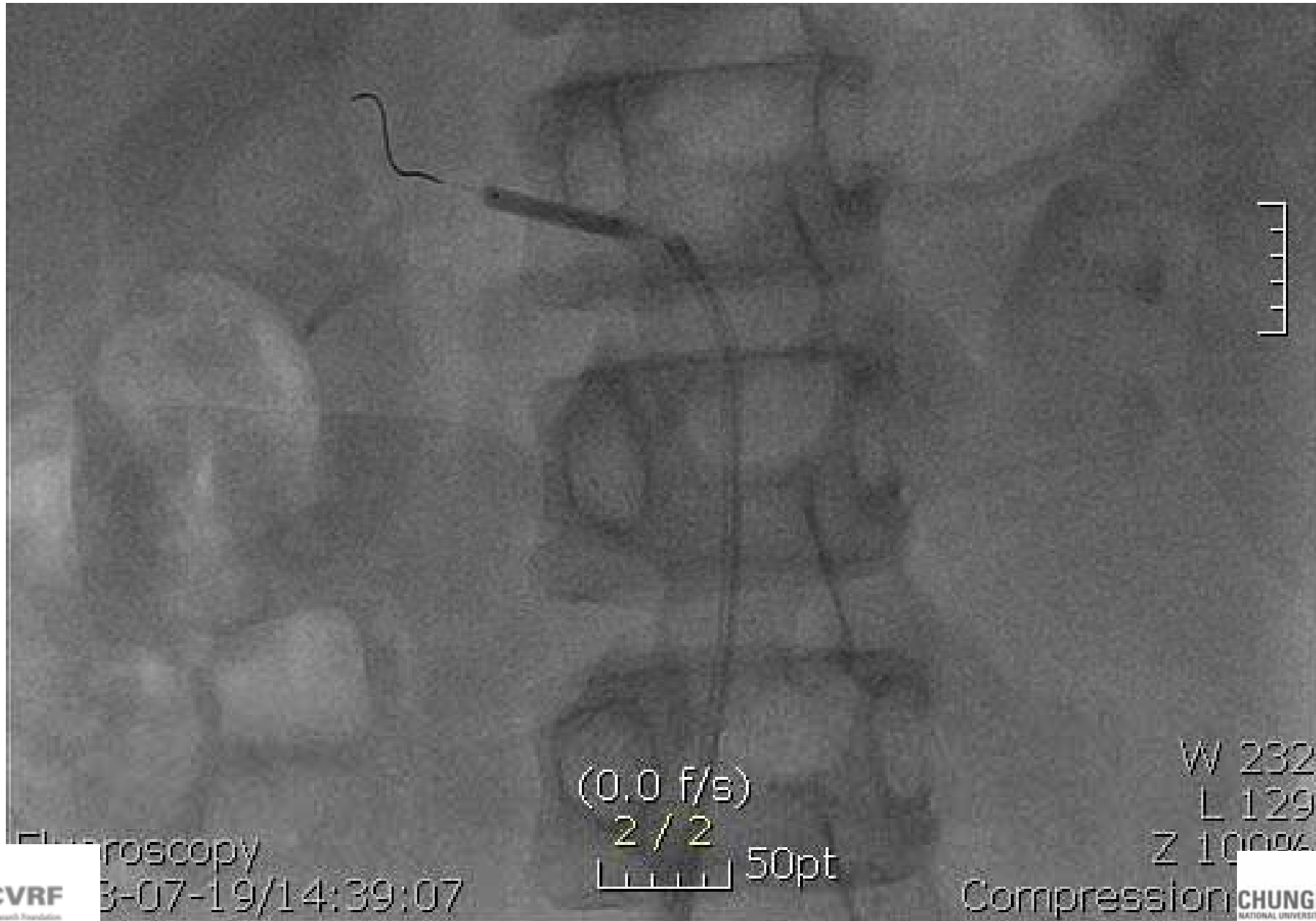
al thickness



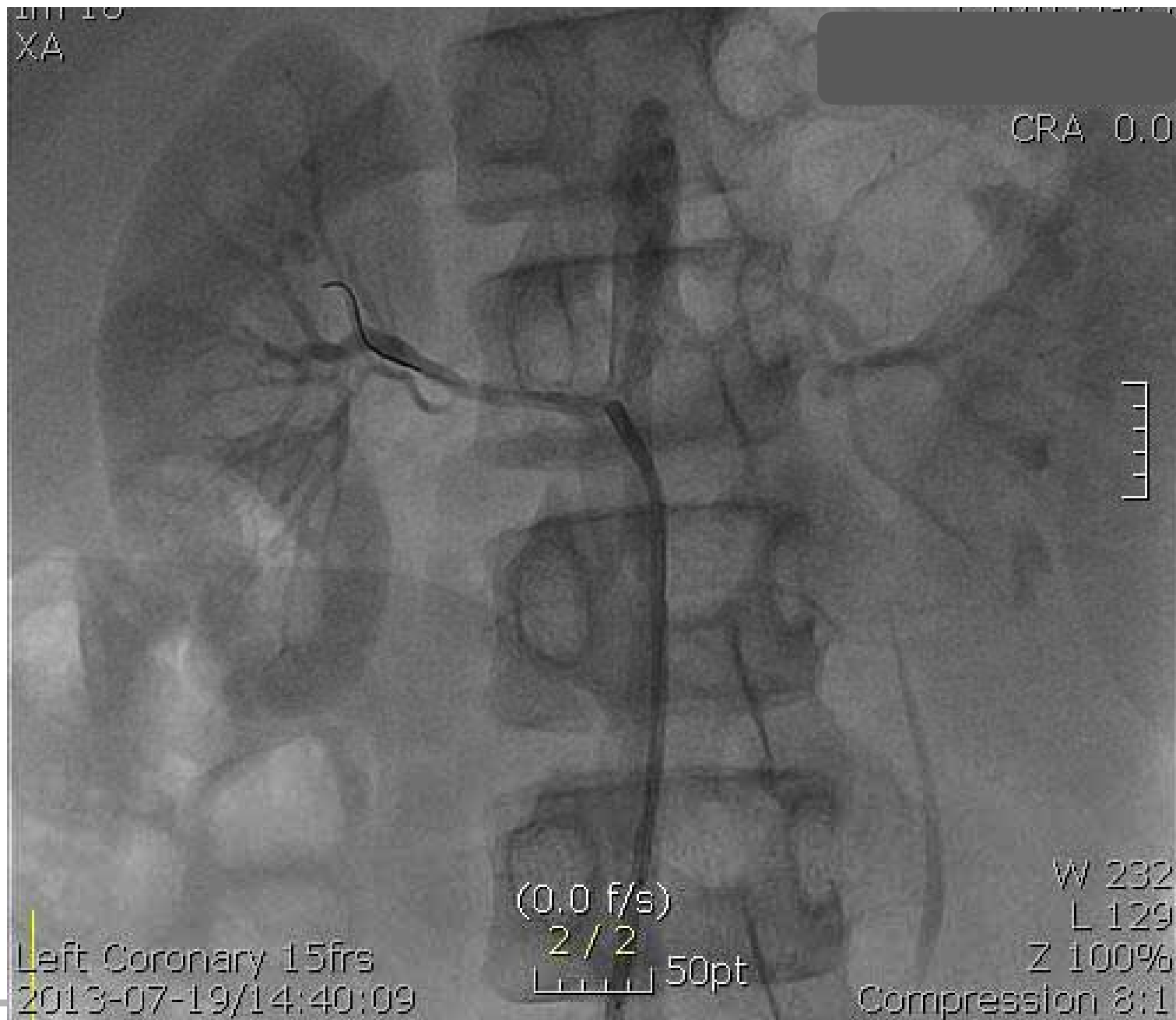
Coronary noncompliant balloon 2.5 X 15 mm



Coronary noncompliant balloon 2.5 X 15 mm



POST PTA



Lt renal angiography



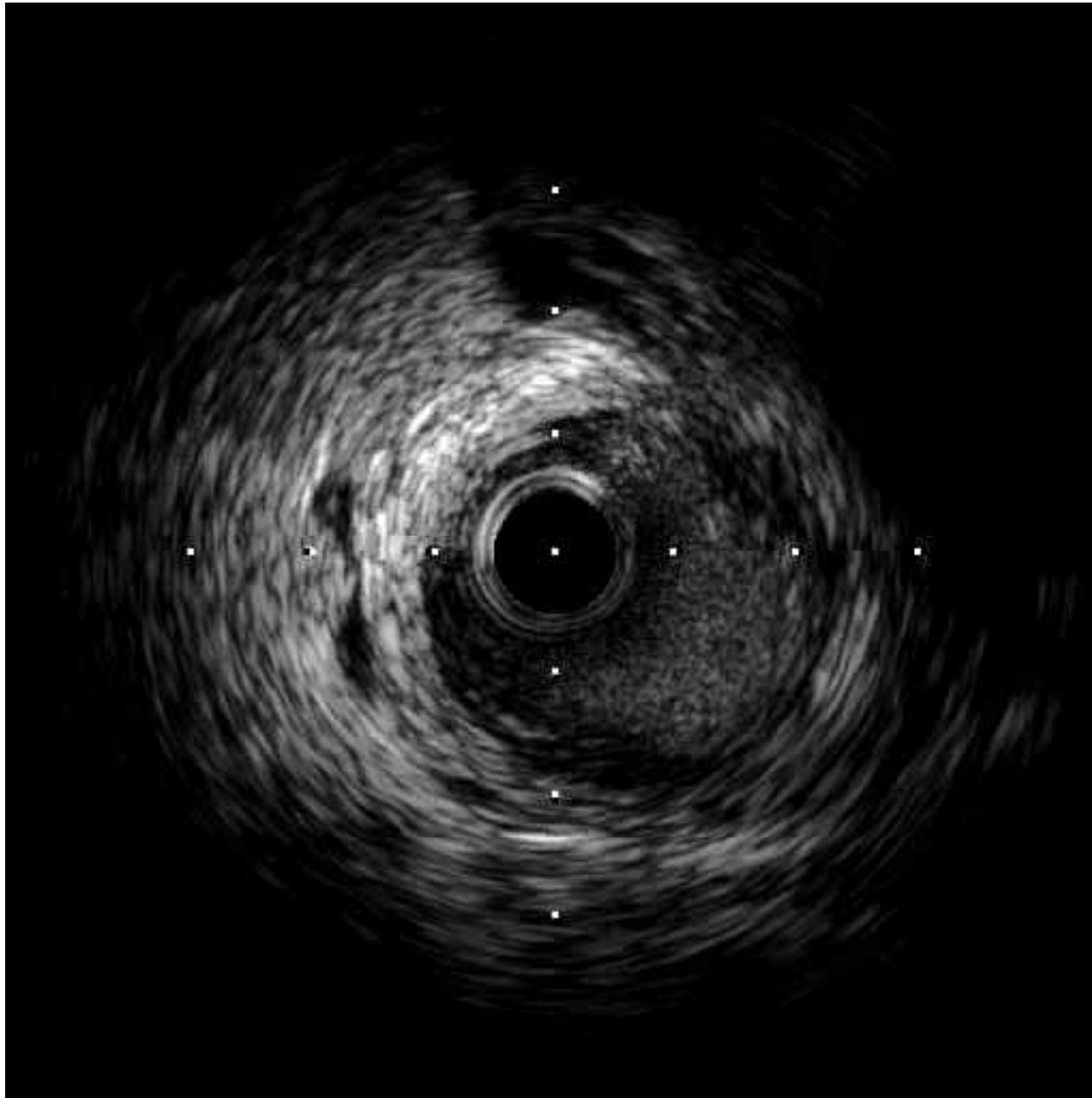
Wiring & PTA



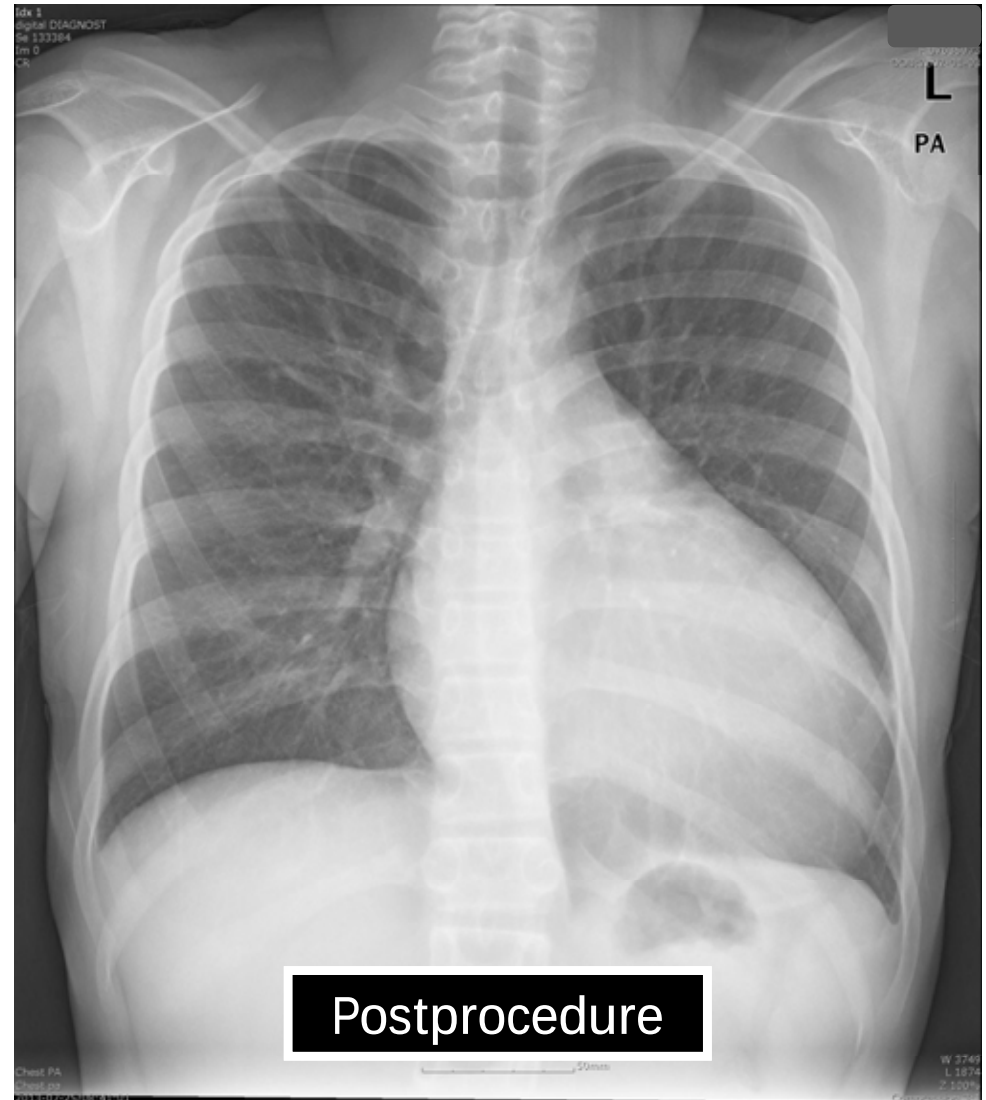
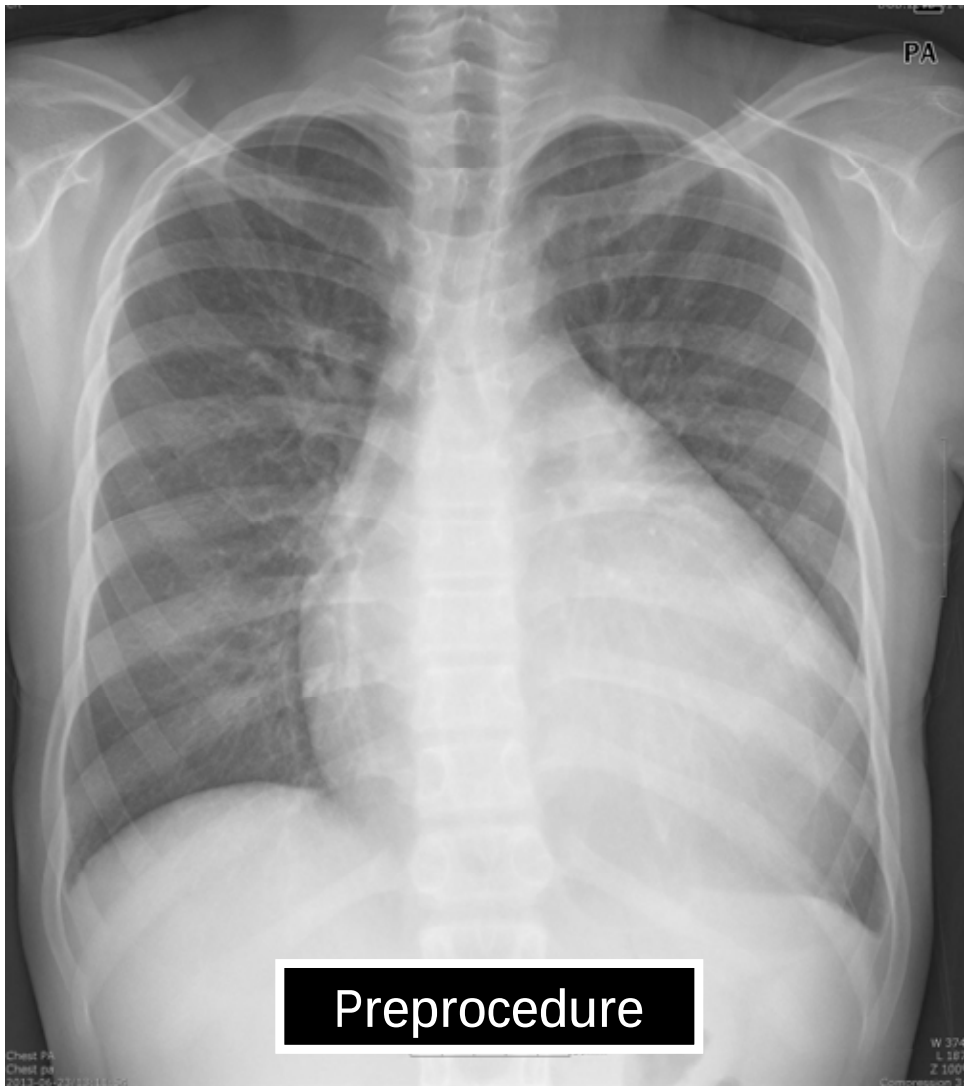
Final result



IVUS : Post PTA



Chest PA : pre and post procedure # 4



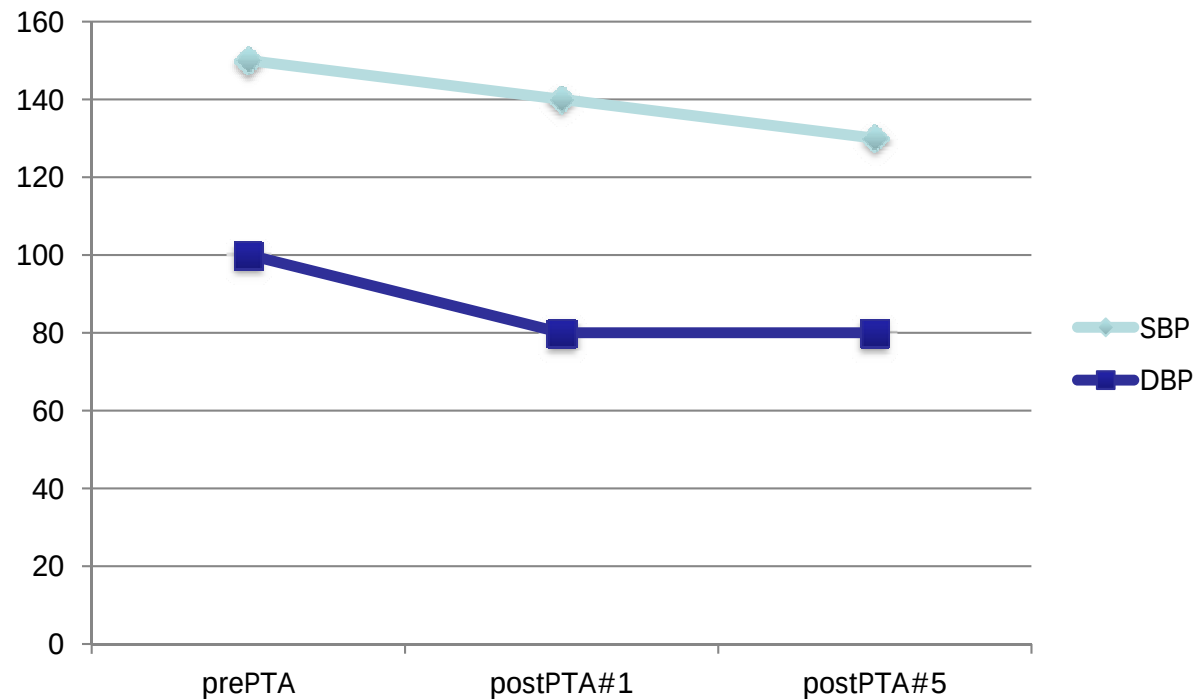
BP F / U

medication(pre PTA)

- Nifedipine 60mg qd
- Furosemide 40mg tid
- Spironolactone 25 mg bid
- Carvedilol 6.25 mg bid

medication (post PTA)

- Amlodipine 2.5 mg qd
- Furosemide 20mg qd
- Spironolactone 12.5 mg qd



CASE 2 : the younger of the two sisters



Childhood Idyll
William-Adolphe Bouguereau

Chief complaint

High blood pressure : 1 month ago

Present illness

A 9-year-old girl was admitted to our hospital because of high blood pressure. She had no coronary risk factors.

PMHx, FHx, SHx

- Past medical history : none
- Familial history : Fibromuscular dysplasia with both renal artery stenosis – her older sister
- Social history : none

ROS and P / E

ROS

- URI Sx.(-), fever(-), anorexia (-)

P/E

- both mild anemic sclera (-/-)
- not palpable cervical LNs
- Clear BS without rale
- RHB without murmur
- not palpable abdominal mass / not palpable liver or spleen
- pitting edema (-)

High blood pressure

BP : 170/100 mmHg

Antihypertensive medication

Nifedipine 60mg qd

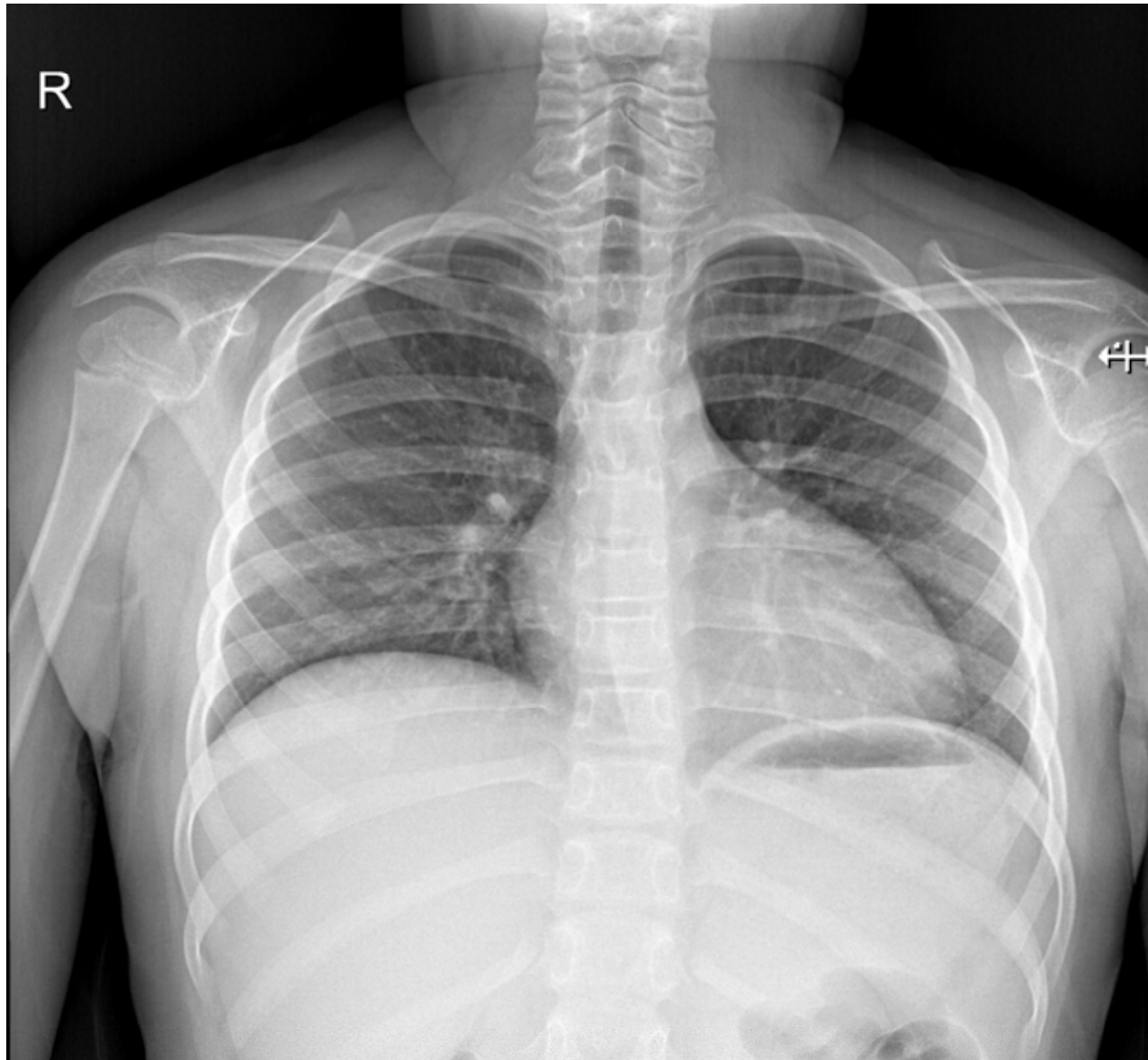
Norvasc 5mg tab(Amlodipine) 1tab [P.O]bid q12h

Dilatrend 25mg tab(Carvedilol) 25mg [P.O]bid q12h

Tenormin 50mg tab(Atenolol) 1tab [P.O]bid q12h

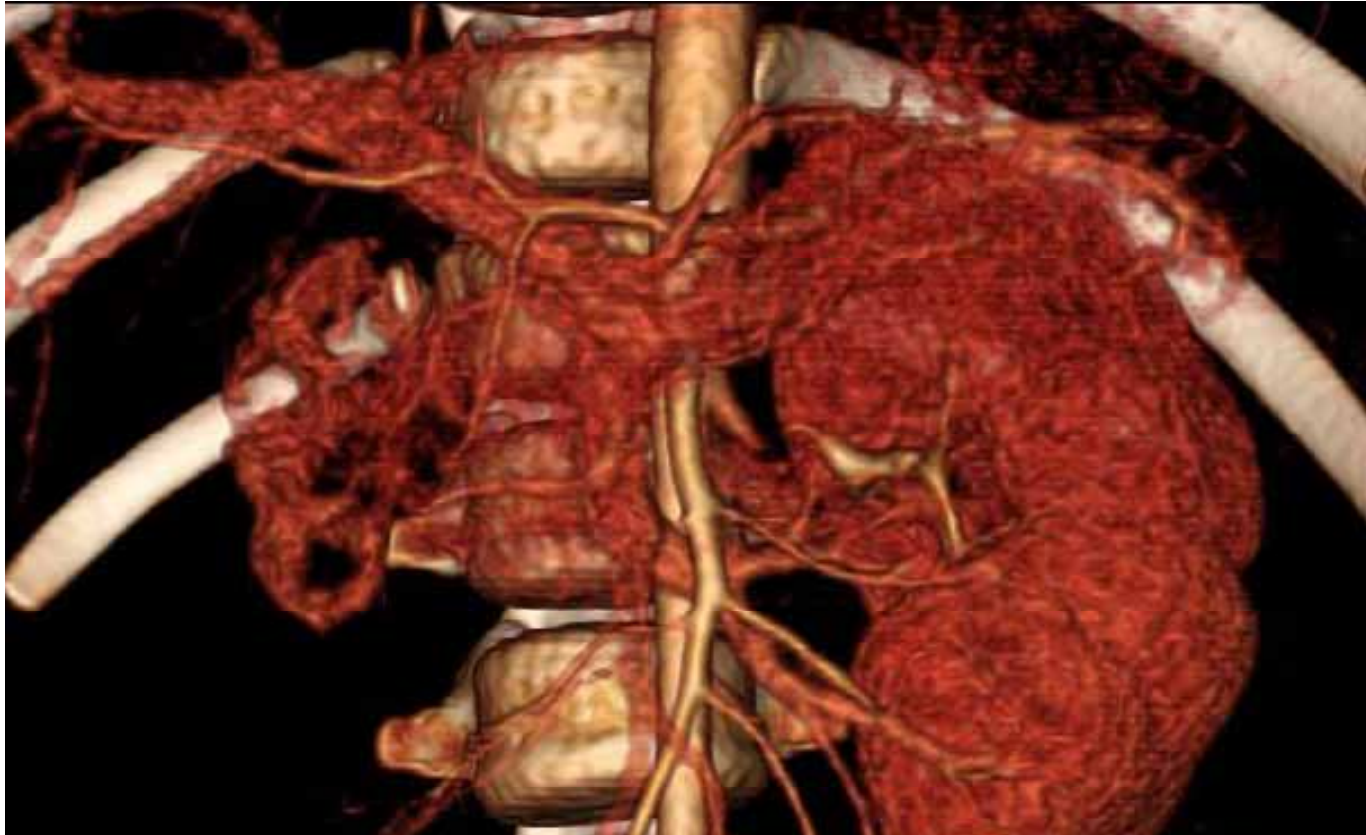
Dichlozid 25mg tab(Hydrochlorothiazide) 0.5tab

Chest X-ray :



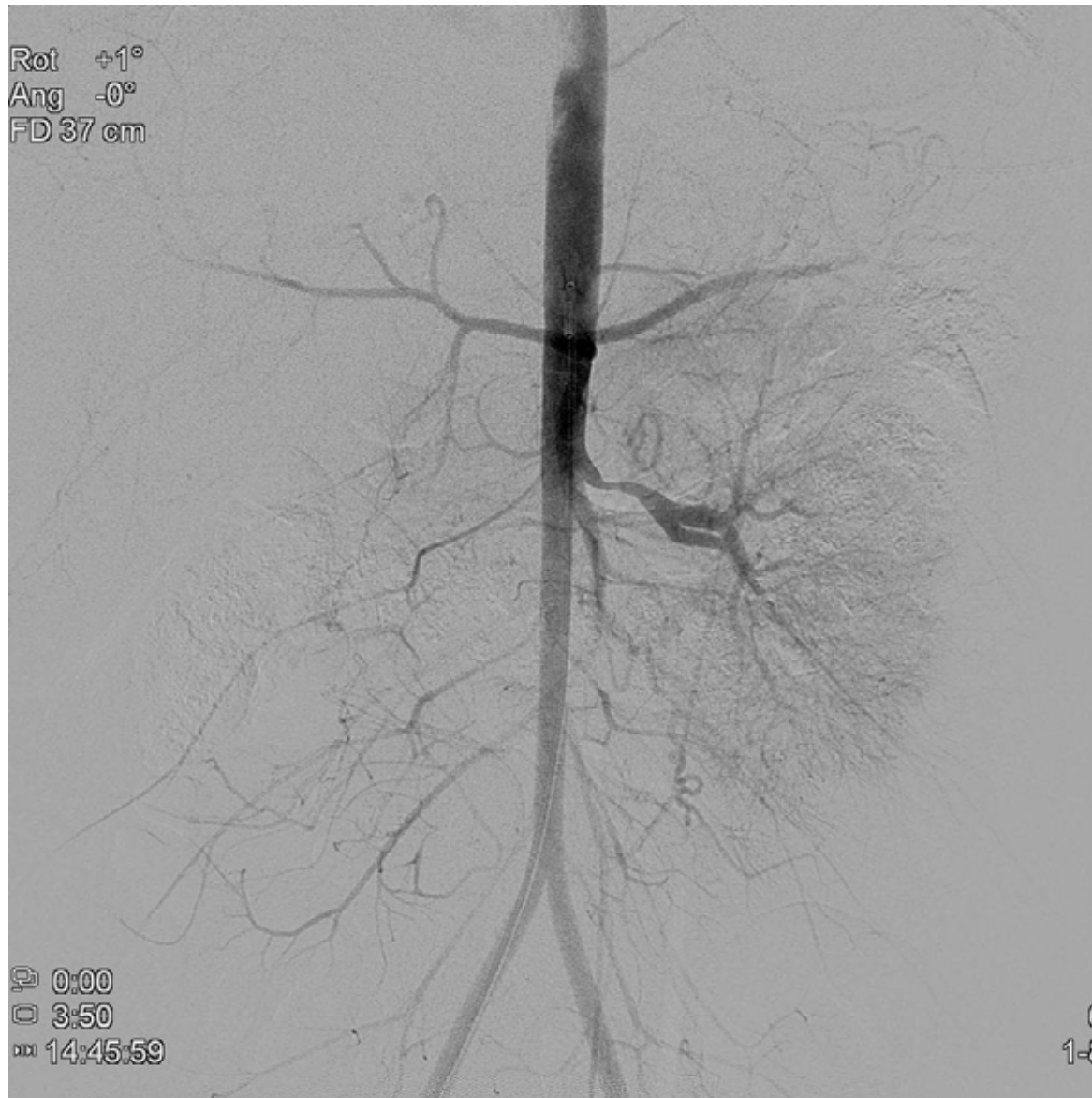
Imaging information from SNUH pediatric nephrology

CT angiography



- Atrophic change of right kidney without visualization of right renal artery
--> probable total obliteration of right renal artery
- Severe stenosis in mid and distal portion of left renal artery showing beaded appearance, but relatively intact the os of left renal artery.
--> r/o FMD

Angiography : right kidney without visualization of right renal artery --> probable total obliteration of right renal artery



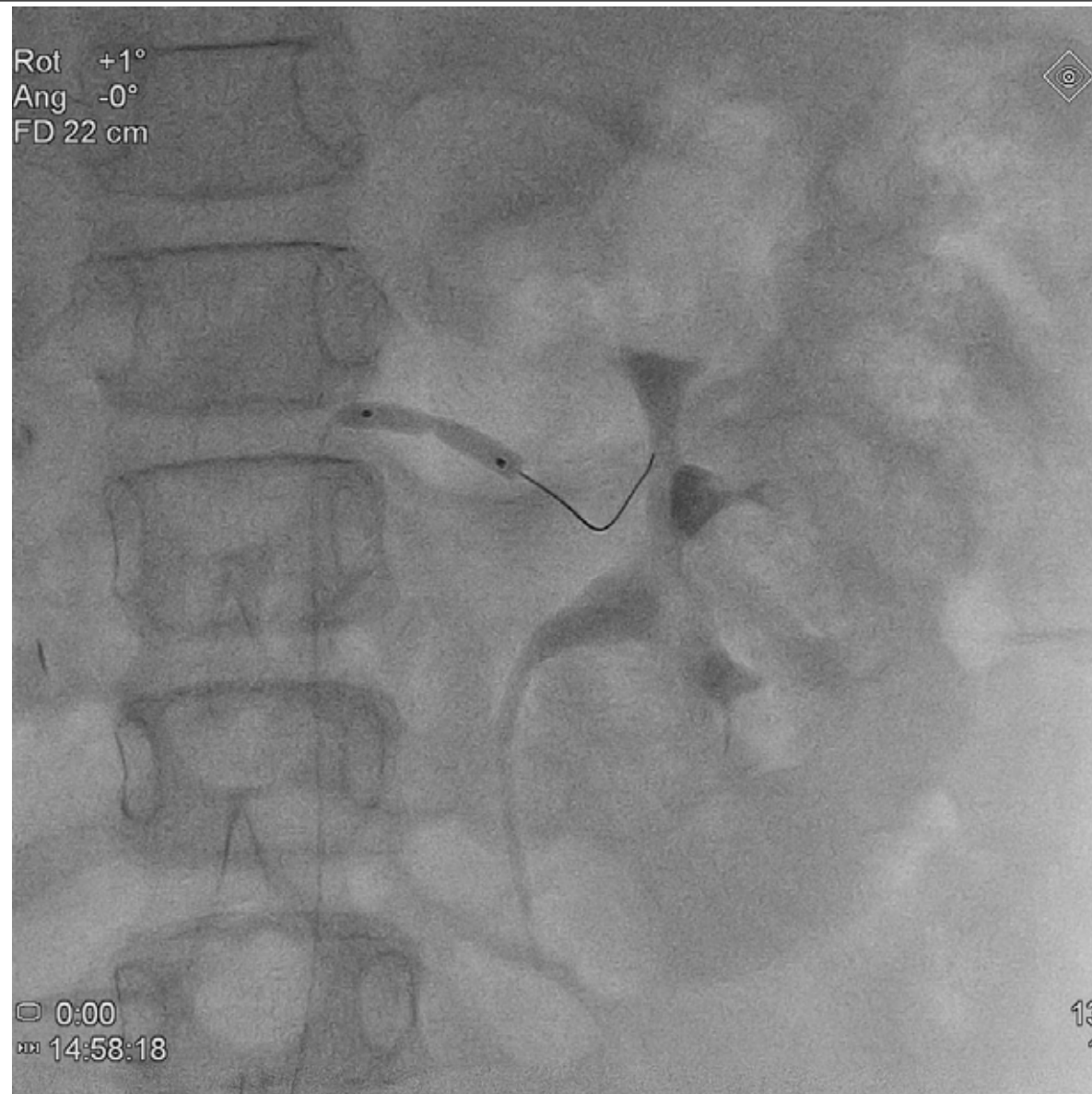
Angiography :

Severe (70-80%) stenosis in the Lt. proximal renal artery



Angioplasty :

Angioplasty of the Lt. proximal renal artery, using a 3mm x 1.5 cm balloon

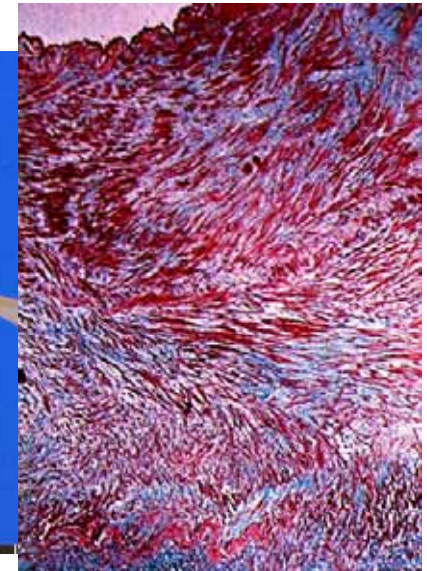
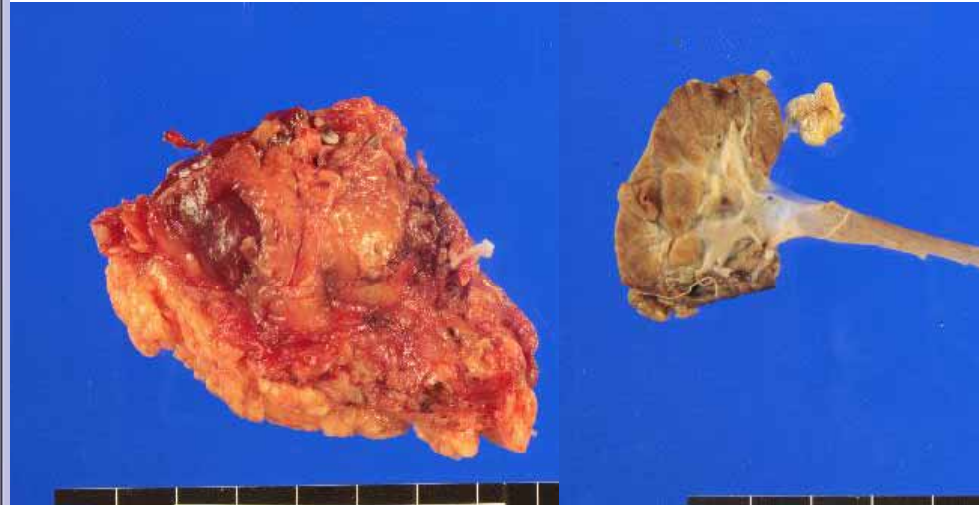
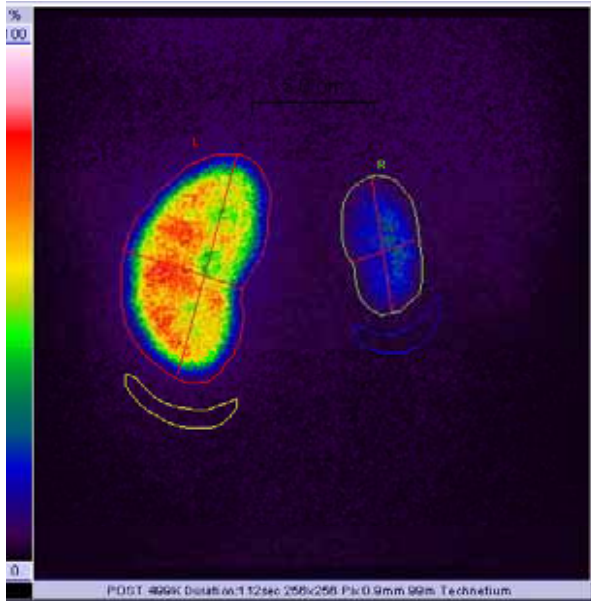


Post-procedural angiography

- Improved renal artery stenosis, with mild (30-40%) residual stenosis.



2 month later, Heminephrectomy, Rt



Kidney, right, radical nephrectomy:

1. **Fibromuscular dysplasia, intimal, medial fibroplasia type**
2. Chronic pyonephritis, focal
3. Congestive renal parenchyme with focal atrophic change

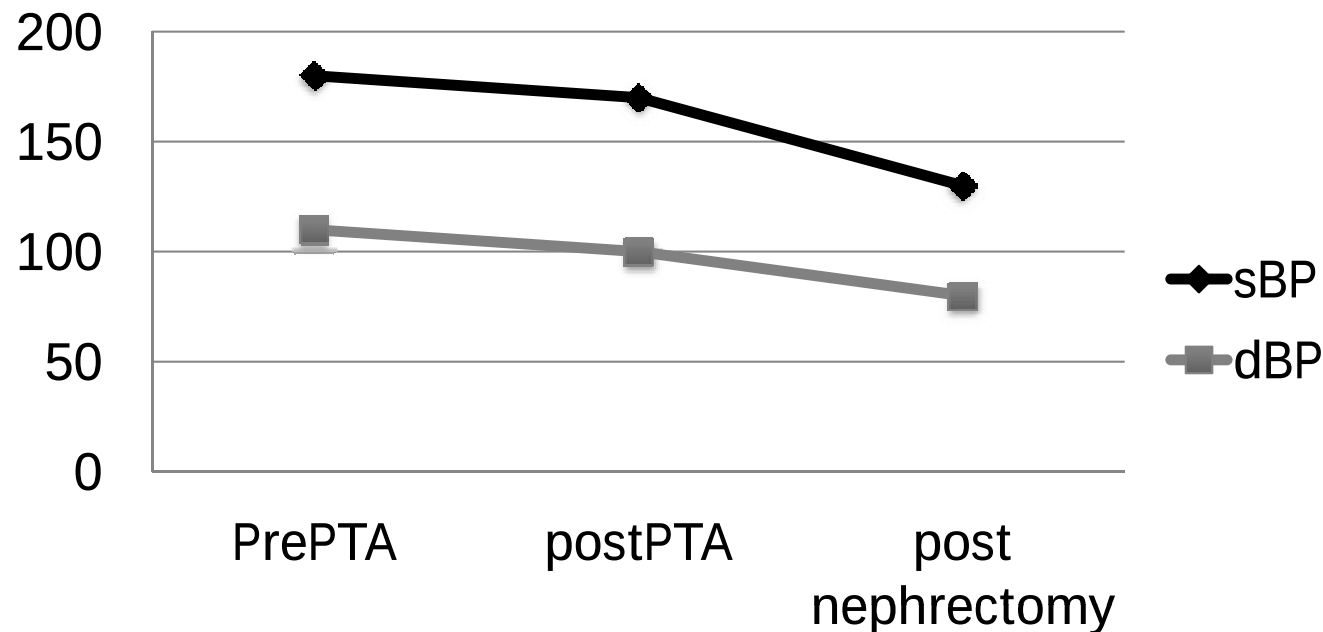
BP F / U

Medication (pre PTA, nephrectomy)

- Nifedipine 60mg qd
- amlodipine 5mg bid
- Carvedilol 25mg bid
- Atenolol 50mg bid
- Dicchlozid 12.5mg

Medication (post PTA)

- Amlodipine 5mg bid
- Carvedilol 25mg bid
- Dichlozid 12.5mg bid
- Atenolol 50mg bid



SUMMARY

- FMD : heterogeneous group of **idiopathic, segmental, noninflammatory, and nonatherosclerotic stenosis of** the musculature of arterial walls in small and medium – sized artery
- IVUS is useful tool in evaluation of renal artery stenosis.
- Familial FMD is found in 10% of FMD.

Thank you



Fibromuscular dysplasia ?

- heterogeneous group of **idiopathic**, segmental, **noninflammatory, and nonatherosclerotic stenosis of** the musculature of arterial walls in small and medium – sized artery
- It is **the second most frequent cause of renovascular hypertension.**
- Its historical classification based on histology is no longer relevant now that **percutaneous revascularization** has replaced surgery in most cases.

Fibromuscular dysplasia

- **Less than 10%** of renal artery stenosis
- mostly affects **women below the age of 40** and more specifically, renal arteries in the **distal two thirds or even segmental** segments; **bilateral** occurrence is quite frequent (60% of cases).

Fibromuscular dysplasia : Pathophysiology ?

Despite various hypotheses linking it to genetic, mechanic or hormonal factors that are being suggested, the pathogenesis of this disease remains **unknown**.

Fibromuscular dysplasia : diagnosis?

In accordance with current definitions

- **nonatherosclerotic stenosing lesions**
- **trunk or branches** of the renal arteries
- **absence of aortic wall thickening or biochemical evidence of inflammation**
- **absence of known syndromic arterial disease** such as type1 neurofibromatosis, pseudoxanthoma elasticum, vascular Ehlers-Danlos syndrome, Williams syndrome, or Marfan syndrome.

Optimal treatment

- Medical treatment is first indicated for the hypertensive patient.
- The current guideline' recommendations for angioplasty refer to
 - ① treatment- resistant hypertension
 - ② drug intolerance
 - ③ signs of ischemic nephropathy (kidney function alteration and kidney size' changes)
 - ④ possible curable hypertension after revascularization.

Good prognosis

Favorable prognostic predictive factors such as

- age under 40
- less than 5 years of hypertension
- maximum BP under 160 mmHg

were considered.

Good prognosis

many controversies regarding the treatment of renal artery stenosis

- balloon angioplasty remains the treatment of choice for FMD
- primary stent placement for the atherosclerotic RAS is

still debatable

→ Two random studies (Astral and Star) **failed to bring enough evidence** in favor of additional stent angioplasty when compared to medical treatment alone, in terms of BP control and renal function

PTA vs surgery

- There are no controlled studies
comparing angioplasty and surgical revascularization.
- Current guidelines recommend
 - ① balloon angioplasty for multifocal or troncular fibro-muscular lesions
 - ② surgery for complex lesions (at the junction or reaching the segmental branches, stenosis associated with micro- aneurysms) or unsuccessful angioplasty

PTA vs surgery

- In 2010, a meta-analysis assessing **2630** FMD patients revascularized surgically and by angioplasty revealed a **36% and respectively 54% success rate (success defined as BP below 140/90 mmHg)**. Periprocedural risks were reported as significant (**12% for balloon angioplasty and 17% for surgery**). However, in terms of major complications, the respective percentages were **6% and 15%**