

Renovascular hypertension in children

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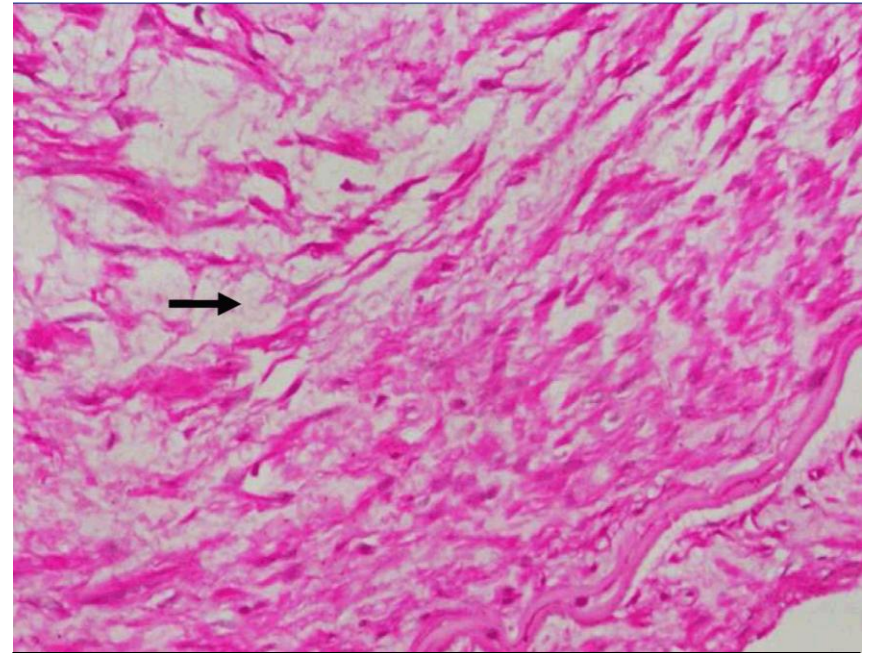
Nottingham Children's Hospital

Perspective

- May account for up to 10% of childhood hypertension
- Most commonly caused by fibromuscular dysplasia
- Frequently bilateral
- Some syndromic associations – particularly neurofibromatosis
- May be associated with extra-renal vascular disease
- Potentially curable!

Fibromuscular dysplasia

- Generally regarded as medial disease
- Aetiology uncertain



dense fibrous connective tissue in the outer media (black arrow), distorted inner medial smooth muscles and normal intimal tissues. (H & E, x200)

Presentation

Shroff et al, 2006; Pediatrics 118: 268-75.

Mode of Presentation	No.
Incidental	9
Cardiac (congestive cardiac failure, palpitations, murmur)	7
Headache with or without vomiting or lethargy	6
Acute hypertensive encephalopathy	3
Cerebrovascular accident	2
Facial palsy	2
Poor feeding and failure to thrive	2
Screening for NF1	2

How useful are non-angiographic imaging techniques?

Table 2 Diagnostic accuracy of ultrasound and pre- and post-captopril isotope studies

Technique	Sensitivity (%)	Specificity (%)
Ultrasound	73–85	71–92
Captopril renography	52–93	63–92
CTA	64–94	62–97
MRA	64–93	72–97

Long-term follow-up

Shroff, 2006. Angioplasty for renovascular hypertension in children: 20-year experience

- 33 children underwent PTH and/or stenting at 1.9 – 17.9 years
- Median follow-up 12 years
- 2/27 re-stenosed from PTA
- 7/19 re-stenosed after stenting
- 11/13 normotensive after main RAS

Long-term follow-up: angioplasty

- Alfonso, 2006
- 11 children (2 in combination with surgery)
- f/u as surgery
- 9/11 normalised bp

Long-term follow-up: surgery

- Kimura, 2010
- 25 children/adolescents over 40 years
- 17 due to FMD
- Surgery included:
 - Aortorenal prosthetic bypass graft
 - Autologous vein/iliac artery graft
 - Nephrectomy
 - Auto-transplant
- Median follow-up 24 years
- Hypertension cured/improved in 21

Long-term follow-up: surgery

- Alfonso, 2006
- 6 children, aged 2-18 y
- Mean follow-up 11.5 y
- All cured

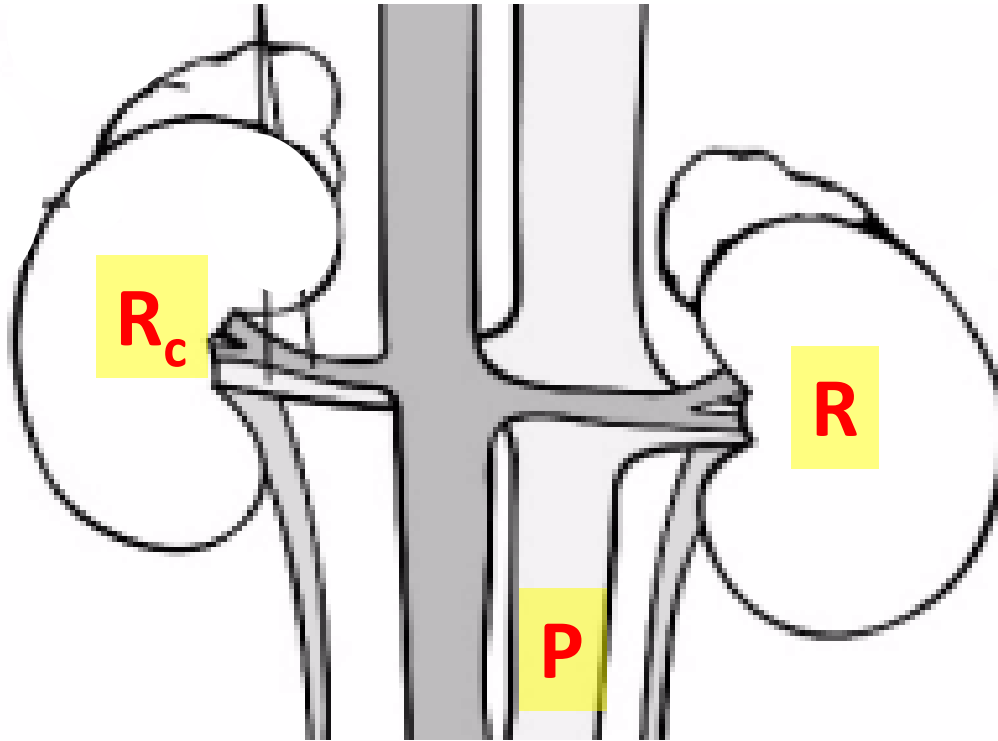
Unilateral nephrectomy follow-up

- 4 cases presenting in early infancy and childhood
- Age at surgery: 0.1 – 2.5 years
- % Function 14 – 27%
- Follow-up: 5-16 years
- All normotensive off treatment with normal GFR

Table 2 Clinical and laboratory signs that indicate an increased chance of renovascular disease [3] (*BP* blood pressure)

Clinical or laboratory sign

1. Very high BP
 2. Secondary symptoms of high BP
 - including cerebral symptoms, cardiac failure and facial palsy
 3. Hypertension not controlled on >2 antihypertensive drugs
 4. Diagnosis of a syndrome with a higher risk of vascular disease, such as neurofibromatosis, tuberous sclerosis and Williams' syndrome
 5. Signs of vasculitis, in particular Takayasu disease
 6. Known or suspected previous vascular insult, such as renal artery thrombosis or umbilical artery catheterisation
 7. Bruit heard over the renal artery or arteries
 8. Elevated peripheral plasma renin or moderate hypokalaemia
-



Lateralisation: $R/R_c > 1.5$

Suppression of contralateral kidney: $R_c/P < 1.3$

25 year review of renal vein renin sampling. Goonasekara, 2002.

- 137 procedures reviewed
- 30% false positive rate in diagnosing RVD
- Useful in predicting response to intervention?
 - High sensitivities but low specificities

Renal vein renin sampling – practical considerations

- Easy to undertake at the same time as angiogram +/- angioplasty
- Review results retrospectively
- Indication for imaging review if hypertension does not improve

DISCUSSION OF CASES

3 year old boy

- Developed right Bell's palsy during intercurrent viral illness
- Blood pressure: 200 systolic (by doppler)
- Renal function normal
- LVH on echo; fundoscopy normal
- Controlled reduction on PICU with labetalol

Imaging

- USS and doppler – normal
- MAG3: 43% function on left
- Angiogram

Follow-up

- Good angioplasty result
- 2 antihypertensives at discharge
- Slow/near complete resolution of Bell's palsy
- Atenolol finally stopped after 10 months

Learning points

- Importance of checking BP in the context of Bell's palsy
- When to proceed to invasive hypertension investigations?
- Angioplasty effective but long-term follow-up needed
- BP can take months to normalise through vascular remodelling

13 month old girl

- Presented with left Bell's palsy on a background of growth faltering
- Initial systolic blood pressure 240 mmHg
- Labetolol infusion to stabilise
- Required 5 oral agents to control, including captopril after definitive management decision

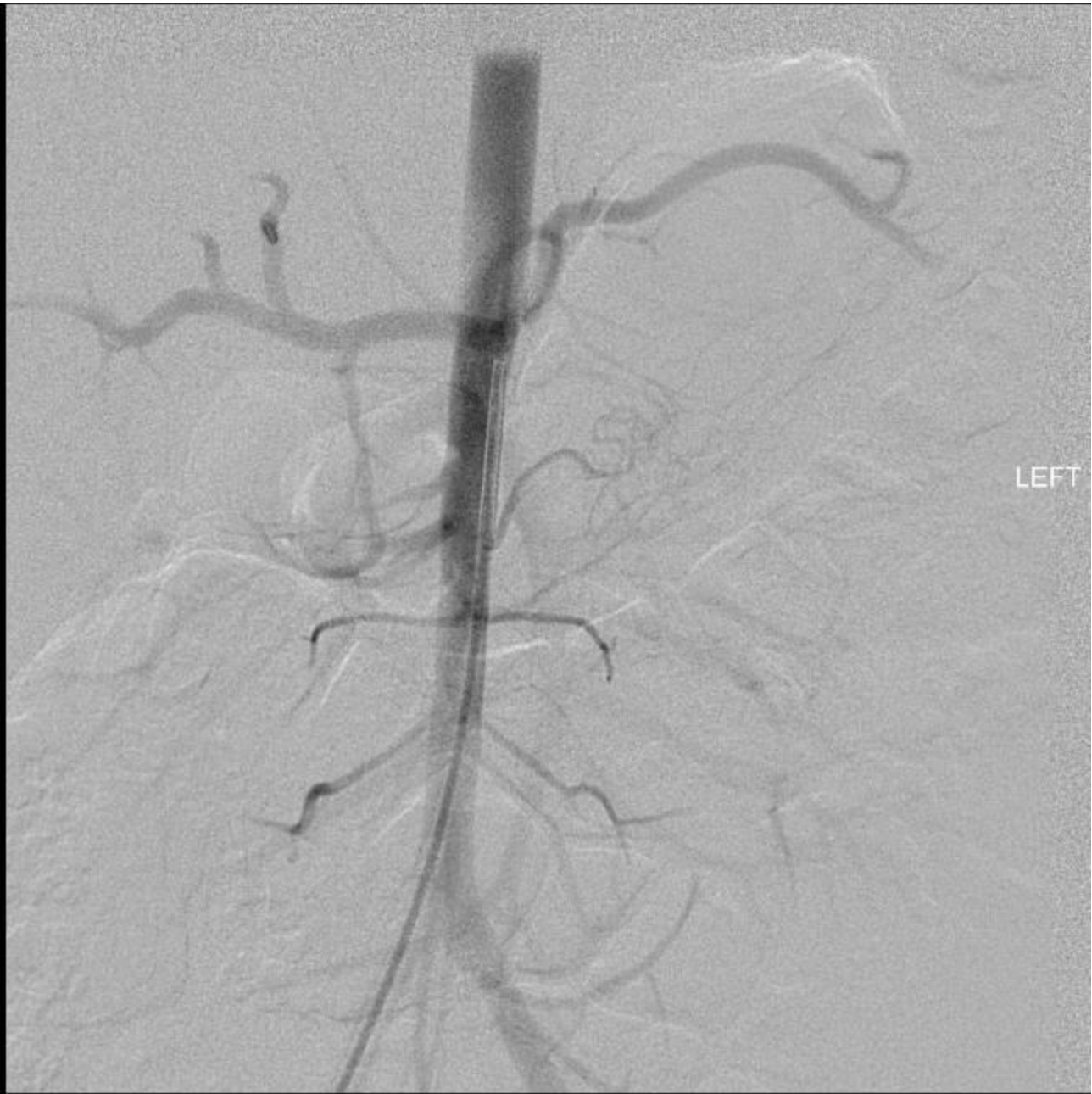
Imaging

- Ultrasound:
 - Small left kidney with smooth outline and possible delayed doppler upstroke
 - Increased echogenicity on right – c/w hypertensive change
- Angiogram – left renal artery stenosis
- 19% function on left on DMSA

RIGHT

LEFT

RIGHT



- Decision to proceed to left nephrectomy in view of patient size and poor function
- Open procedure
- Discharged on 3 agents
- Drugs weaned and finally stopped after 3 months
- Facial palsy improved slowly
- Remains off treatment after 21 months

Learning points

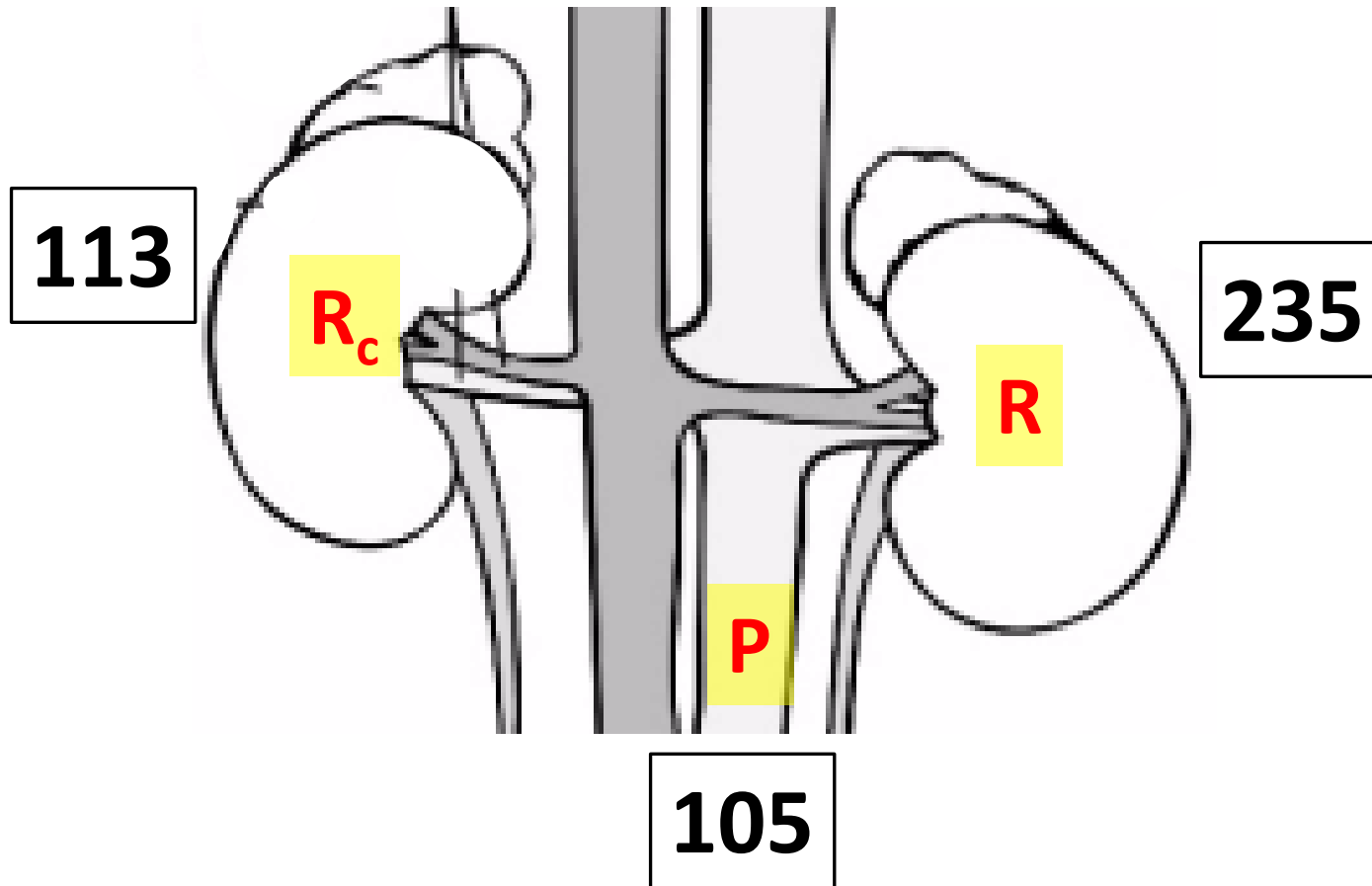
- Nephrectomy may be indicated rather than angioplasty in some cases
- Judicious use of ACEIs has a role in management of difficult cases

15 year old girl

- Neurofibromatosis type 1
- Mild hypertension
- BP 135/95 measured in clinic
- BP systolic load 70% on ABPM
- Peripheral renin level: high normal
- BP unchanged with two agents

- Discharged on 2 drugs
- Atenolol stopped after 3 months
- Amlodipine reduced after further 2 months but BP rebounded
- Amlodipine stopped again 6 months later
- Seen every 6 months
- BP currently around 90th centile off treatment





Lateralisation: $R/R_c > 1.5$

1.7 = lateralised

Suppression of contralateral kidney: $R_c/P < 1.3$

1.1 = suppressed

- Low threshold for angiogram when syndromic association
- Lesions in NF-1 are often ostial and can be more difficult to treat
- RV renin sampling may be helpful in complex cases

14 year old girl

- Incidental finding of hypertension
- BP uncontrolled in single agent
- Initial USS and DMSA normal
- Peripheral renin/aldosterone normal
- BP remained poorly controlled on two agents
- Decision to proceed to angiogram after 12 m

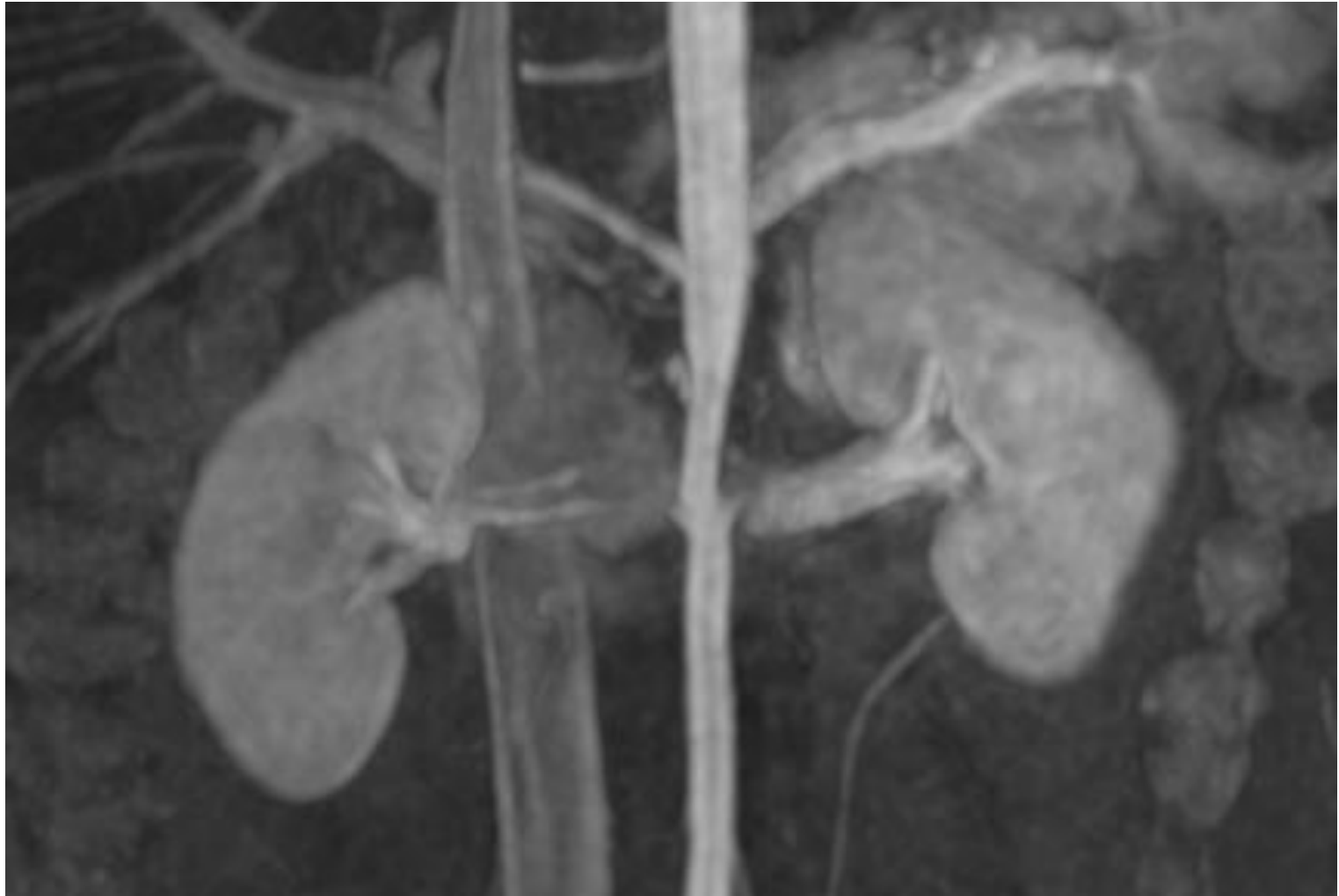
RIGHT

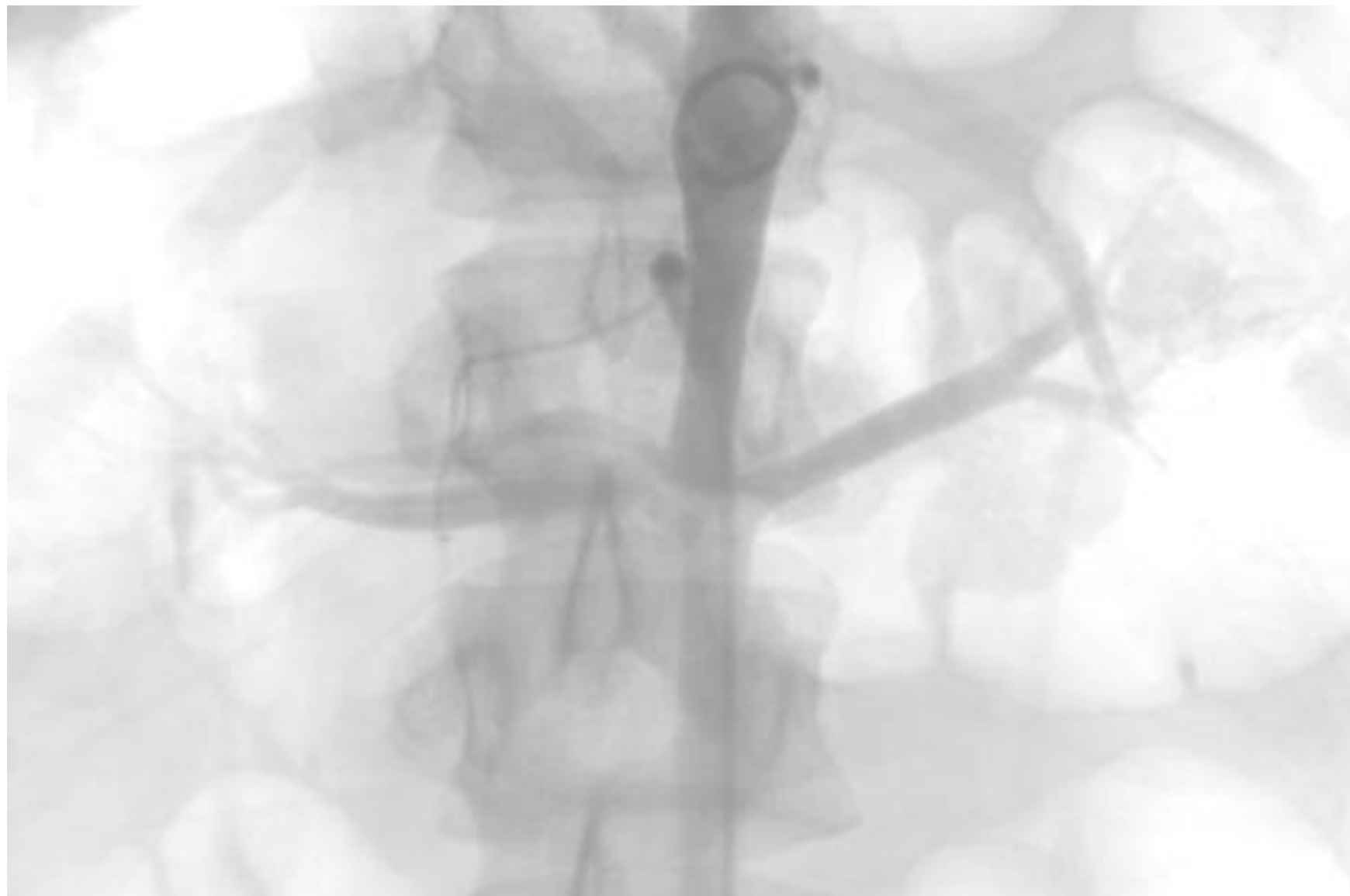
LEFT



- Mid-aortic syndrome and left lower pole stenosis
- Decision to plan angioplasty / canvas opinion
- Angioplasty 4 months later with safety wire across aorta
- Both aorta and right lower pole artery angioplastied
- BP remained high on 2 agents

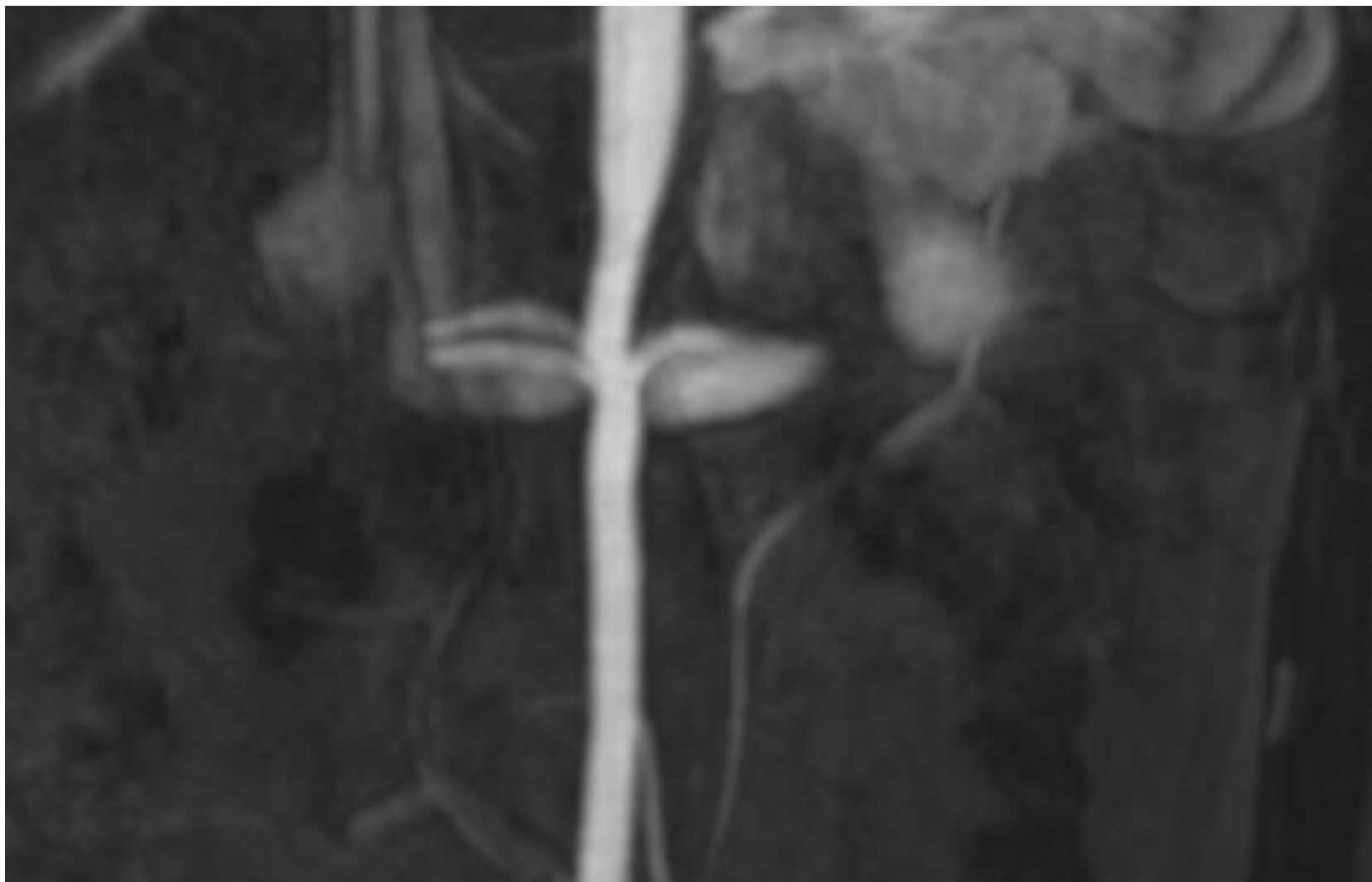
- Follow-up CT angiogram showed possible stenosed right upper pole vessel
- Repeat MR angiogram 6 m later – appearances unchanged
- Repeat angiogram/angioplasty







- BP again creeping up
- Further MR angiogram



- Now transitioned to adult care
- Remains on single agent with controlled BP
- Likely need for on-going imaging and possible repeat angiography/surgery

Learning points

- Indication for angiography when uncontrolled BP on two agents
- CT/MR angiography useful in follow-up imaging

Questions?