

Conn's syndrome with a focus on a unilateral adrenal gland

HOSP #	Mrs DW	WARD	Endocrine Department – CathLab – UCT private Hospital
CONSULTANT	Dr Jody Rusch	DOB/AGE	49y Female

Abnormal Result

49yr old female

Presenting Complaint

Medical complaint: Suspected Conn's disease – right adrenal lesion/ irregular left adrenal gland

History

Past Medical History: Resistant Hypertension, primary hyperaldosteronism (confirmed previously with saline infusion test), hypokalaemia, hypercholesterolaemia, newly diagnosed DM.

Family History: Hypertension – Mother.

Past Surgical History: TAH – 7 years ago.

Allergies: Nil known

Smoker

Meds: Amlodipine/Valsartan 10/320 daily, Doxazosin 8mg daily, Furosemide 40mg daily, Spironolactone 25mg daily, Carvedilol 25mg daily, Metformin 1g nocte, Simvastatin 20mg nocte,

Zolpidem 10mg nocte.

Examination

Not available

Laboratory Investigations

	A	B	C	D	E	F	G	H	I	J
		Time	Aldosterone Episode	Aldosterone pmol/L	Cortisol nmol/L	Selectivity Index Cortisol AV/PV	ACR	Lateralisation Index Dom A/C : nonDom A/C	Mean Aldo/Cort RAV	Aldo/Cort LAV
3	RAV 1	10:10	SA04663221	1310	659	0.2	2.0			
4	RAV 2	10:36	SA04663224	1490	681	0.2	2.2			
5	RAV 3	10:36	SA04663229	INS	712	0.2	#VALUE!			
6	RAV 4	10:39	SA04663232	771	340	0.1	2.3			
7	RAV 5	10:49	SA04663235	1470	692	0.2	2.1			
8	Mean RAV			1260.25	616.8	0.2			2.0	
9	LAV 1	10:01	SA04663256	2160	10790	3.0	0.2	0.1		0.2
10	LAV 2	10:02	SA04663250	3210	14540	4.0	0.2	0.1		0.2
11	LAV 3	10:03	SA04663242	5260	2621	0.7	2.0	1.0		2.0
12	LAV 4	10:59	SA04663238	2760	11870	3.3	0.2	0.1		0.2
13	LAV 5	10:03	SA04663246	3590	10770	3.0	0.3	0.2		0.3
14	Mean LAV			3396	10118.2	2.8				0.3
15	PIVC 1	11:00	SA04663213	2540	3609					
16	PFEM 1	9:43	SA04663217	803	301					
17	Arm	10:12	SA04663208	1330	724					
18										
19	Key:									
20	RAV	Right Adrenal Vein				Peripheral				
21	LAV	Left Adrenal Vein				Aldosterone		2540		
22	PIVC	Peripheral Inferior Vena Cava				Cortisol		3609		
23	PFEM	Peripheral Femoral Vein								
24	UTC	Unable to calculate								
25	*	Not assayed in dilution								
26	AV/PV	Adrenal Vein to Peripheral Vein Ratio								
27	ACR	Adrenal to Cortisol Ratio								

Other Investigations

Not available for this patient.

Ideally one would need a CT with contrast beforehand to adequately visualize the positions of the adrenal veins, as this may aid in the canulation, especially of the right adrenal vein.

One needs to diagnose hyperaldosteronism (by an appropriate salt loading test) before proceeding to bilateral adrenal vein sampling.

Final Diagnosis

Interpretation

Definition	Formula	Clinical significance
Selectivity index	$\text{PCC}(\text{side}) / \text{PCC}(\text{ivc})$	>cutoff confirms canulation of adrenal vein >3 stimulated >2 unstimulated
Lateralization index	$\text{PAC}/\text{PCC}(\text{dom}) : \text{PAC}/\text{PCC}(\text{non-dom})$	>cutoff confirms laterilization of hyperaldo secretion >4 stimulated >2 unstimulated
Contralateral suppression index	$\text{PAC}/\text{PCC}(\text{non-dom}) : \text{PAC}/\text{PCC}(\text{ivc})$	<cutoff indicate ipsilateral suppression and suggest contralateral aldosterone overproduction.

Table 1 – Interpretation of bilateral adrenal vein sampling. PCC: plasma cortisol concentration, PAC: plasma aldosterone concentration, ivc: inferior vena cava or peripheral vein, dom: dominant side, non-dom: non-dominant side.

Selectivity index

Right: 0.2 (mean)

Left: 2.8 (mean)

These two results indicate that the left adrenal has likely been canulated adequately, but the right vein inadequately.

Lateralization index

Unable to comment because of the inadequate canulation of the

right adrenal vein. If determined, it would very likely provide a false result.

Contralateral suppression index

616.8 /1260.25 : 2540/3609

= 0.70

This falls in between some of the referenced cutoffs (<1 and <0.5)

All of the other samples also fall somewhere in this range. Biochemically, these results suggests inadequate right sided venous sampling (a commonly described problem)

Take Home Message

- Procedure is done in the Cath Lab
- The patient received continuous synacthen infusion
- Done under imaging with contrast used for the localisation of the adrenal gland and adrenal vein
- Sequential sampling technique used, generally > 20 mins infusion
- Multi-disciplinary: nurses, anaesthetist, radiographer, intervention radiologists, students, chemical pathologists
- Difficulty with sampling right side for both patients
- Difficulty with interpreting results – most likely due to inadequate cannulation of the right adrenal vein

Some important learning points

1. Adrenal vein sampling may be a valuable tool that is underutilised
2. Careful selection of patients essential – also patient should consent to surgical removal of the affected adrenal before this invasive procedure is initiated

3. Inter-disciplinary approach is necessary
4. Obtaining cosyntroponin (aka synacthen) can be difficult (Section 21), but recommended
5. Right adrenal access difficult: may require specific imaging. Recommended to start on the right or do simultaneous sampling
6. Adrenalectomy may be curative or help achieve better control of BP thus decrease associated morbidity and mortality in those with unilateral adenoma