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

	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
RAV 1	10:10	SA04663221	1310	659	0.2	2.0			
RAV 2	10:36	SA04663224	1490	681	0.2	2.2			
RAV 3	10:36	SA04663229	INS	712	0.2	#VALUE!			
RAV 4	10:39	SA04663232	771	340	0.1	2.3			
RAV 5	10:49	SA04663235	1470	692	0.2	2.1			
Mean RAV			1260.25	616.8	0.2			2.0	
LAV 1	10:01	SA04663256	2160	10790	3.0	0.2	0.1		0.2
LAV 2	10:02	SA04663250	3210	14540	4.0	0.2	0.1		0.2
LAV 3	10:03	SA04663242	5260	2621	0.7	2.0	1.0		2.0
LAV 4	10:59	SA04663238	2760	11870	3.3	0.2	0.1		0.2
LAV 5	10:03	SA04663246	3590	10770	3.0	0.3	0.2		0.3
Mean LAV			3396	10118.2	2.8				0.3
PIVC 1	11:00	SA04663213	2540	3609					
PFEM 1	9:43	SA04663217	803	301					
Arm	10:12	SA04663208	1330	724					

Key:	
RAV	Right Adrenal Vein
LAV	Left Adrenal Vein
PIVC	Peripheral Inferior Vena Cava
PFEM	Peripheral Femoral Vein
UTC	Unable to calculate
*	Not assayed in dilution
AV/PV	Adrenal Vein to Peripheral Vein Ratio
ACR	Adrenal to Cortisol Ratio

Peripheral	
Aldosterone	2540
Cortisol	3609

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	PCC(side) / PCC (ivc)	>cutoff confirms canulation of adrenal vein >3 stimulated >2 unstimulated
Lateralization index	PAC/PCC (dom) : PAC/PCC (non-dom)	>cutoff confirms laterilization of hyperaldo secretion >4 stimulated >2 unstimulated
Contralateral suppression index	PAC/PCC (non-dom) : PAC/PCC (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 canulation 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 simulataneous sampling
6. Adrenalectomy may be curative or help achieve better control of BP thus decrease associated morbidity and mortality in those with unilateral adenoma

---

ACTH

HOSP #		**WARD**	G16 Medical Ward
CONSULTANT		**DOB/AGE**	54 y Female

Abnormal Result

21/08/2018 Two ACTH tests (referred to another laboratory) and two
Cortisol levels (at our laboratory) were done.
At first it was thought to be a dexamethasone suppression test, but then
realized the clinicians were suspecting hypopituitarism.

10h05: **ACTH 0.7 pmol/L ↓** (1.6-13.9) Cortisol 8 nmol/L ↓ (Morning: 133- 537; Afternoon 68 – 327)

10h35: ACTH 1.8 pmol/L N (1.6-13.9)
Cortisol 68 nmol/L ↓ (Morning: 133- 537; Afternoon 68 – 327)

Presenting Complaint

? hypopituitarism

History

Known with a pituitary macroadenoma, previously seen at the Radiotherapy clinic in 2016.

Examination

No clinical info available.

For Primary adrenal insufficiency one would expect: Hyperpigmentation
(due to ↑ ACTH), +/- hyperkalemia/hyponatremia (aldosterone effect), +/-
virilization.

For Secondary adrenal insufficiency there is subtle symptoms, electrolytes are not deranged significantly because aldosterone function is preserved. See table on Bishop 7th ed. p. 459.

Laboratory Investigations

Measurement of
plasma ACTH concentration is used to assess Cushing's disease, adrenal tumors,
ectopic ACTH-producing tumors, Addison's disease, Nelson's syndrome, and
hypopituitarism.

The
laboratory diagnosis of hypopituitarism, however is relatively straightforward.
In contrast to the primary failure of an endocrine gland that

is accompanied by
dramatic increases in circulating levels of the corresponding pituitary tropic
hormone, secondary failure (hypopituitarism) is associated with low or normal
levels of tropic hormone. This is the
diagnosis in this case with the history of previous radiotherapy which was
given for a macro-adenoma.

Other Investigations

Free T4 on 19/04/2018 was 7.8 pmol/L (12-22), also suggesting possible hypopituitarism, although a TSH would be helpful.

Final Diagnosis

Hypopituitarism confirmed.

Take Home Messages

Dexamethasone suppression test need only measurement of cortisol, not accompanying ACTH, except in extended work-up however, where a Cosyntropin (CRH) stimulation test can be done to distinguish between pituitary or hypothalamic insufficiency.

Evaluation of pituitary function need the Primary hormone (Cortisol) as well as the tropic hormones from the pituitary (ACTH).