

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

|                   |                  |                |                                                             |
|-------------------|------------------|----------------|-------------------------------------------------------------|
| <b>HOSP #</b>     | Mrs DW           | <b>WARD</b>    | Endocrine Department –<br>CathLab – UCT private<br>Hospital |
| <b>CONSULTANT</b> | Dr Jody<br>Rusch | <b>DOB/AGE</b> | 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             |
|----|----------|---------------------------------------|-------------|-------------|----------------|-------------------|-----|----------------------|--------------------|---------------|
| 1  |          | Time                                  | Aldosterone | Aldosterone | Cortisol       | Selectivity Index | ACR | Lateralisation Index |                    |               |
| 2  |          | Episode                               | pmol/L      | nmol/L      | Cortisol AV/PV |                   |     | 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                  | $\text{PCC(side)} / \text{PCC (ivc)}$             | >cutoff confirms<br>canulation of adrenal<br>vein<br>>3 stimulated<br>>2 unstimulated                      |
| Lateralization<br>index            | $\text{PAC/PCC (dom)} : \text{PAC/PCC (non-dom)}$ | >cutoff confirms<br>laterilization of<br>hyperaldo secretion<br>>4 stimulated<br>>2 unstimulated           |
| Contralateral<br>suppression index | $\text{PAC/PCC (non-dom)} : \text{PAC/PCC (ivc)}$ | <cutoff indicate<br>ipsilateral suppression<br>and suggest contralateral<br>aldosterone<br>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