# 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<br>>3 stimulated<br>>2 unstimulated |
| Lateralization index | PAC/PCC (dom) : PAC/PCC (non-dom) | >cutoff confirms laterilization of hyperaldo secretion<br>>4 stimulated<br>>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

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# Hyperaldosteronism with Hyperreninaemia in a 15 year old

A case of hyperreninemia and hyperaldosteronism in a 15 year old. The cause will surprise you.

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# Hyperaldosteronism

| | | | |
|---|---|---|---|
| **HOSP #** | | **WARD** | Murraysburg Hospital, Female Ward |
| **CONSULTANT** | | **DOB/AGE** | 51 y female |

## Abnormal Result

Aldosterone: 1380 pmol/L

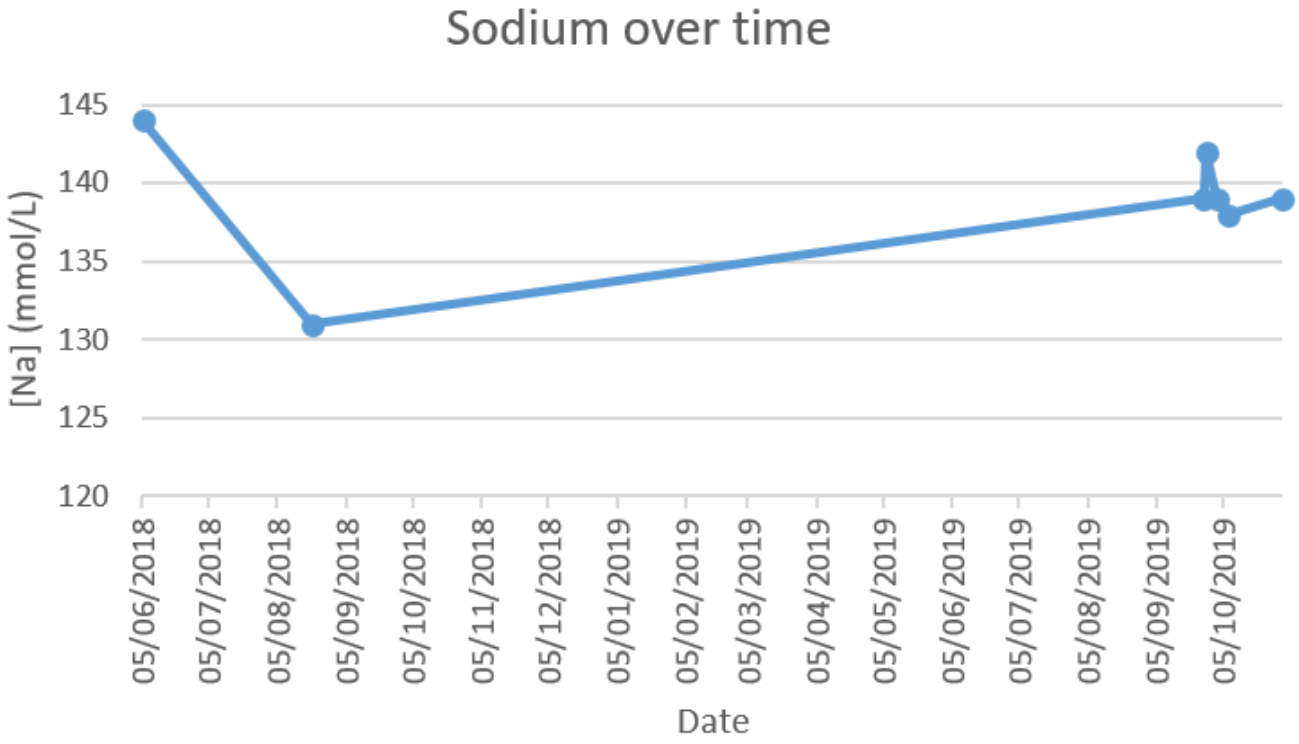
Renin: 2.1 ng/L

Aldosterone: Renin ratio: 657.14 pmol/ng

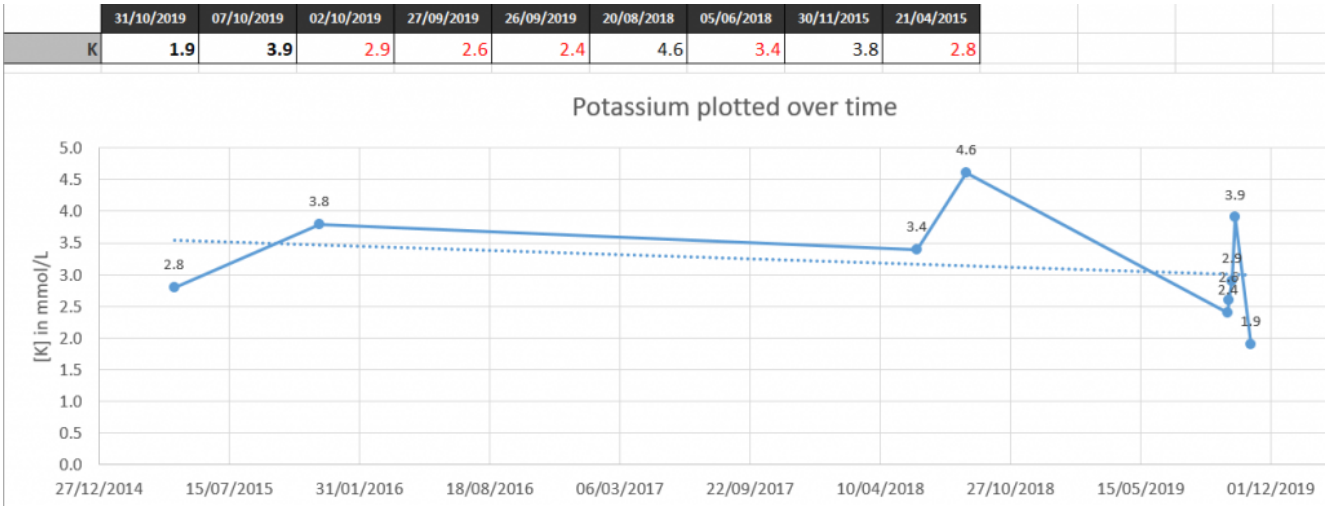
# Presenting Complaint

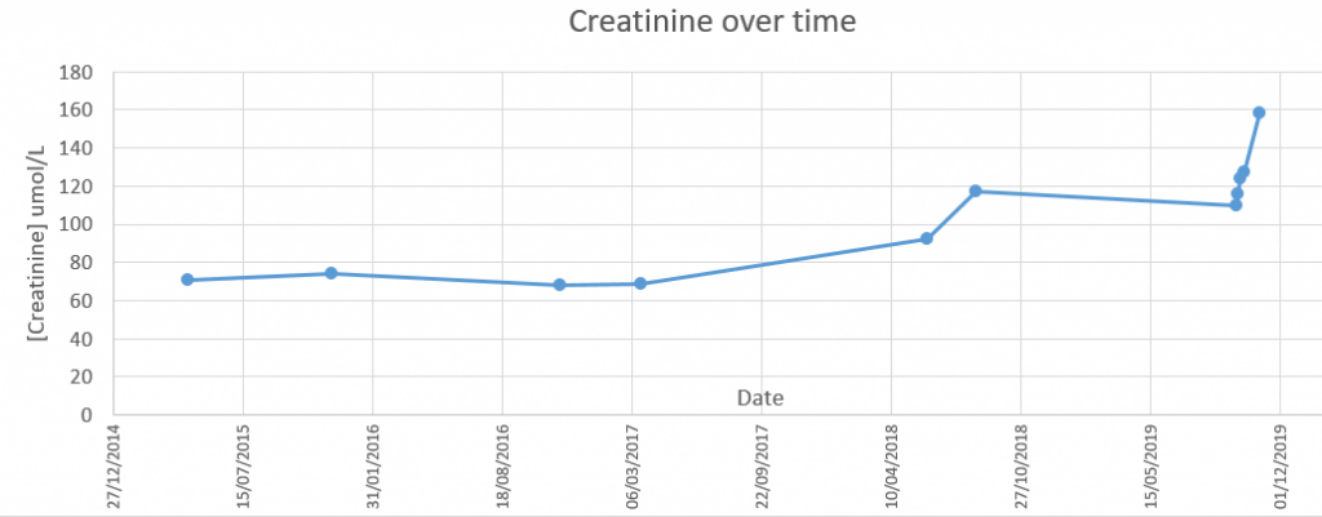
Uncontrolled Hypertension, unresolved on maximum dose of 3 antihypertensives.

# History




| | 31/10/2019 | 07/10/2019 | 02/10/2019 | 27/09/2019 | 26/09/2019 | 20/08/2018 | 05/06/2018 | 30/11/2015 | 21/04/2015 |
|---|---|---|---|---|---|---|---|---|---|
| K | **1.9** | **3.9** | 2.9 | 2.6 | 2.4 | 4.6 | 3.4 | 3.8 | 2.8 |






# Examination

# Laboratory Investigations

| | | | | | | |
|---|---|---|---|---|---|---|
| Hos | Murraysburg Hospital wc MBH | ☎ | 049 844 0053 | Received | 07/10/2019 | 17:57 |
| Wrd | Female Ward | ☎ | | Registered | 07/10/2019 | 17:58 |
| Doc | DR HUMAN | ☎ | | | ePR | I Deta |

| Test Set | Staff Notes | Test Item | Result | Units | Normal Values | Previous Result 1 |
|---|---|---|---|---|---|---|
| ALDOS | ✓ | Aldosterone | 1,380.0 | pmol/L | | |
| | | Patient condition | | | | |
| | | Aldosterone auto comm | ALDO4 | | | |
| RENIN | | Renin mIU | | mIU/L | | |
| | | Renin ng | 2.1 | ng/L | | |
| | | Aldosterone : renin ratio | 657.14 | pmol/ng | | |

# Other Investigations

## Urine electrolytes

| | 01/10/2019 15:32 |
|---|---|
| UNa | 59 |
| UK | 27,5 |
| Ucreat | 4,1 |
| Uprotein | 0,27 |
| Uprot:creat | 0,066 |

## Serum Results

| Date | Sodium mmol/L | Potassium mmol/L | eGFR ml/min | GGT U/L | Chol mmol/L | TSH mIU/L | T4 pmol/L | FreeT3 pmol/L | Cort nmol/L |
|---|---|---|---|---|---|---|---|---|---|
| 21/04/2015 | | 2,8 | >60 | | 5,07 | | | | |
| 30/11/2015 | | 3,8 | >60 | | 4,53 | | | | |
| 15/11/2016 | | | >60 | | 4,04 | | | | |
| 20/03/2017 | | | >60 | | 4,36 | | | | |
| 05/06/2018 | 144 | 3,4 | 56 | | 4,39 | 1,79 | 11,9 | 5 | 394 |
| 20/08/2018 | 131 | 4,6 | 42 | | | | | | |
| 21/08/2018 | | | | | | | | | |
| 24/08/2018 | | | | | | | | | |
| 26/08/2018 | | | | | | | | | |
| 26/08/2018 | | | | | | | | | |
| 26/09/2019 | 139 | 2,4 | 45 | | | 0.81 | | | |
| 27/09/2019 | 142 | 2,6 | 43 | | | | | | |
| 01/10/2019 | | | | | | | | | |
| 02/10/2019 | 139 | 2,9 | 40 | | | CEGK | | | |
| 03/10/2019 | | | | | | | | | |
| 07/10/2019 | 138 | 3,9 | 38 | | | | | | |
| 31/10/2019 | 139 | 1,9 | 30 | 28 | | | | | |

## Urine metanephrines

| Urine collection period | 24 h | Reference value |
|---|---|---|
| Urine volume | 3080 ml | |
| Ucreat | 2,2 mmol/L | |
| Umetadren | 160 nmol/L | |
| Unormetadren | 870 nmol/L | |
| dUmetadren | 493 nmol/24h | 152-913 |
| dUnormetadren | 2680 nmol/24h | 699-2643 |
| Umetadren:cr | 73 nmol/mmol creat | 17-91 |
| Unormetad:cr | 395 nmol/mmol creat | 75-309 |

# Final Diagnosis

Primary hyperaldosteronism causing secondary hypertension with accompanying renal injury.

# Take Home Messages

## Reference Ranges for Aldosterone:

- Upright 70 – 1066 pmol/L
- Supine 49 – 643 pmol/L

Screening for primary hyperaldosteronism: most sensitive when **>350** pmol/L

## Reference Ranges for Renin:

- Upright: 2.7 – 27.7 ng/L
- Supine: 1.7 – 23.9 ng/L

Beta-blockers suppress renin levels and should be stopped 2 weeks before testing.

## Aldosterone: Renin Ratio:

Most sensitive when the ratio is **>118** pmol/ng.

## Effects of hyperaldosteronism

- One’s expectation is a high serum sodium, but since it normalizes with an increase in fluid volume, hence hypertension as in this case, there is normal sodium.
- Low serum potassium due to loss in urine, although this can also be normal.
- Increased urine potassium concentration (>30 mmol/L) in a random urine specimen suggests increased mineralocorticoid effect.
- The renin:aldosterone ratio is used to compensate for

the increase in aldosterone which is caused by an increase in renin (for instance which is caused by hypovolemia or low blood pressure).

- Some studies recently published are suggesting that the prevalence of hyperaldosteronism are significantly more than was (and is) thought, and hence urinary (24 hour) aldosterone measurement may be more accurate to screen for hyperaldosteronism. The authors of recent estimates of the prevalence of hyperaldosteronism are of opinion that hyperaldosteronism may be the cause of around 10% of unexplained “essential” hypertensives (see attached articles).

Hyperaldo-prevalence-2020Download
Primary-hyperaldo-Editorial-2020Download