The Adrenal Glands

The adrenal glands are hormone producing organs that sit on top of the kidneys.

The outer part is called the cortex - responsible for producing glucocorticoids and mineralocorticoids – (cortisol and aldosterone) as well as small amounts of male and female sex hormones.

- Glucocorticoids play a role in converting fats, carbohydrates and proteins into energy, whilst also helping to regulate blood pressure and heart function. They also help regulate the immune system
- Mineralocorticoids help control blood pressure by regulating our salt / water balance.

The inner part is called the medulla – responsible for producing adrenaline and noradrenaline. These are the fight or flight hormones, which are released when the sympathetic nervous system recognises physical or emotional stress.

ACC is found in the outer layer – the cortex.

Signs and symptoms of an ACC are related to tumour position/size (1) and /or excess hormone production (2):

1. Discomfort / pain, occasionally nausea/vomiting
2. Cushings or mixed Cushings/virilising syndrome

Further information including support: www.acccsupport.org.uk
Diagnosis for ACC

Blood / Urine Tests

- Full blood count
- Liver and kidney function
- **Hormonal work-up:**
  - Glucocorticoid excess (minimum 3 of 4 tests)
  - Dexamethasone suppression test (1mg 23:00)
  - Excretion of free urinary cortisol (24 hr urine)
  - Basal cortisol (serum) Basal ACTH (plasma)
- **Sexual steroids and steroid precursors**
  - Serum DHEA-S, 17-OH-progesterone, Androstenedione & Testosterone
  - 17-beta-estradiol (serum, men & postmenopausal women only), 24-h urine steroid metabolite examination
- **Mineralocorticoid excess**
  - Potassium (serum) Aldosterone/renin ratio (only in those with arterial hypertension +/- hypokalaemia)
  - Catecholamine excess
    - Normetanephrine, metanephrine & methoxytyramine (plasma)
    - Alternatively - fractionated metanephrine excretion (24 hr urine)

In those with a clearly established diagnosis of ACC, catecholamine excess work-up is not required

Scans

- CT chest & abdomen and / or CT chest &MRI abdomen
- Bone scintigraphy/brain imaging - where there is clinical suspicion of metastases
- FDG-PET (optional)
- mIBG scintigraphy, DOTA_TATE_PET, Dopa/Dopamine PET or FDG-PET if pheochromocytoma is proven

Pathology

- Differentiation and cellular morphology (Weiss score)
- Steroidogenesis Factor-1 (if available)
- Ki67

nb a biopsy is only indicated where surgery is precluded and histology is required for oncological management
Treatment for ACC

For all cancer patients, medical care including treatments need to be personalised not only to the specific type of cancer, but also the specific background medical history and current health status of the individual affected. So even if you have a diagnosis that sounds the same as someone else’s, your care and treatment may be different. Your medical team will discuss your care and treatment options with you, so that, together, you can make an informed choice about your ongoing plan of care.

For further information about treatments visit: www.accsupport.org.uk

Follow-up for ACC

As per national and international guidelines nb local policy may vary

For patients following complete resection:

- CT/MRI* 3 monthly for 2 years, then 3 - 6 monthly for further 3 years
- Ongoing surveillance beyond 5 years is suggested but can be adapted according to clinical indication
- Regular hormone screen.

Advanced ACC (incomplete resection, metastatic and inoperable disease):

Imaging and hormone monitoring – recommendation is this should be guided by prognosis, expected treatment efficacy and treatment related toxicity (performance status and clinical indication for active intervention).

*Cross sectional imaging: chest, abdomen and pelvis is recommended.
A big part of meeting with your doctors, or specialist nurse, is to make sure you get the information you need to understand what’s happening, so that you can make an informed choice about your care. Asking questions can be difficult, especially if you’re feeling nervous, confused, frightened or struggling to understand what you are being told. You might want to know as much as possible straight away or prefer to take things in small amounts at your own pace.

**Suggestions that may help:**
- Prepare a list of questions that are important to you
- Ask for simple explanations - do not be worried about asking your nurse or doctor to repeat what they have said
- Take someone with you or ask if you can record the conversation. Many mobile phones have a record function or an app you can download
- Ask for a copy of any letters sent to your GP and/or other care team(s)
- If you have a nurse specialist - keep in touch. They can be a great source of information and support for you.

**Example questions:**
- Who can I call if I have any questions? Who is my main point of contact?
- Who will be involved in my care?
- What are the treatment options for me? How might they affect me?
- How often will I need to have scans and tests?
- Are there any flags or warning signs I need to look out for?

Further information about making the most of your consultations can be found in our handbook: www.netpatientfoundation.org

**REFERENCES**
Berruti et al: Adrenal cancer: ESMO Clinical Practice Guidelines for diagnosis, treatment and follow-up

Fassnacht et al: European Society of Endocrinology Clinical Practice Guidelines on the management of adrenocortical carcinoma in adults, in collaboration with the European Network for the Study of Adrenal Tumors