Pheochromocytoma and Paraganglioma

The adrenal glands are just above your kidneys and create hormones involved in processes like responding to stressful events and controlling blood pressure.

There are three types of neuroendocrine tumour that can start in or near these glands: adrenocortical carcinoma, pheochromocytomas and paragangliomas.

Phaeochromocytomas (phaeos) and paragangliomas Phaeochromocytomas, often known as ‘phaeos' (‘fee-ohs’), are rare tumours of the adrenal gland arising from the inner section of the gland called the medulla. Similar tumours can arise from sites outside of the adrenal gland and these are called (extra-adrenal) paragangliomas.

For more information, please read:

THE NET HANDBOOK - Your Guide to Life with a Neuroendocrine Tumour

This is a comprehensive 158 page guide produced just for you. We would really appreciate it if you were able to purchase a copy but understand that this is not possible for everyone...We want everyone to benefit from this handbook.

GET YOUR COPY TODAY!
You can buy from our online shop for £7.50 (P&P included) OR email or call us for a free copy. You can also read it online on our website.

Registered charity number 1092386
# Diagnosis for Pheochromocytoma and Paraganglioma

## Blood / Urine Tests

<table>
<thead>
<tr>
<th>Test</th>
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<tbody>
<tr>
<td><strong>Full blood count</strong></td>
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<td><strong>Liver and kidney function</strong></td>
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<td><strong>Hormonal work-up -</strong></td>
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<td>Glucocorticoid excess (minimum 3 of 4 tests):</td>
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<tr>
<td>- Dexamethasone suppression test (1 mg, 23:00 h)</td>
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<td>- Excretion of free urinary cortisol (24 h urine) Basal cortisol (serum)</td>
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<td>Basal ACTH (plasma)</td>
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<td>Sexual steroids and steroid precursors:</td>
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<tr>
<td>- DHEA-S (serum) 17-OH-progesterone (serum) Androstenedione (serum) Testosterone (serum) 17-beta-estradiol (serum, only in men and postmenopausal women) 24-h urine steroid metabolite examination</td>
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<tr>
<td>Mineralocorticoid excess:</td>
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<tr>
<td>- Potassium (serum) Aldosterone/renin ratio (only in patients with arterial hypertension and/or hypokalemia)</td>
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<tr>
<td>- Catecholamine excess:</td>
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<tr>
<td>- Normetanephrine, metanephrine, and 3-methoxytyramine (3-MT) (serum or urine)</td>
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In patients with a clearly established diagnosis of an ACC, one can omit the workup on catecholamine excess and conversely for established pheochromocytoma, one can omit the steroid analysis.
- Consider ALL patients for genetic testing
- Chromogranin-A (especially in NM and 3-MT negative patients) (serum)

## Scans

- CT or MRI of abdomen and CT thorax
- Bone scintigraphy (when suspecting skeletal metastases)
- FDG-PET
- MIBG scintigraphy, DOTA-TATE-PET, Dopa/Dopamine PET
- Differentiation and cellular morphology (Weiss Score)
- Steroidogenesis Factor-1 (if available)
- Ki67

## Pathology (tissue examined in the laboratory)

- Differentiation and cellular morphology
- Synaptophysin
- Chromogranin
- Ki67
**Treatment**

For all NET Patients, treatments need to be completely personalised to the type of NET that you may have. Even if you have a diagnosis that sounds the same as another patient, your treatment plan maybe different, because you are unique. Your care team will discuss your treatment options with you so that together you can agree on the most appropriate treatment plan.

For information on all treatments currently utilised in the care of NET patients, please read the Treatments section in our NET Handbook - Your Guide to Life with a Neuroendocrine Tumour.

**Follow-up**

Patients who underwent successful surgery for nonmetastatic disease are at risk of malignant recurrence and require long-term clinical (adrenergic symptoms and blood pressure levels) and biochemical follow-up.

Biochemical testing (MN, 3-MT and Chromogranin-A) is repeated ~14 days following surgery, thereafter every 3 – 4 months for 2 – 3 years. This should subsequently be repeated every 6 months.

Follow up for a minimum 10 years – except for high-risk patients (young, with genetic disease, large tumour and / or paraganglioma) who should receive lifelong annual follow up.

Imaging to be repeated at least every 6 months during the first year and yearly afterward, irrespective of the negative results of biochemical tests.

In patients with (pre-operative) biochemically inactive disease, imaging every 1 - 2 years.

Patients with new events or pathological endocrine tests and/or elevated circulating chromogranin A should undergo imaging that includes thorax and abdomen CT and best functioning imaging (PET FDG in most cases).

**REFERENCES**


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

European Journal of Endocrinology (2016) 174 : G1-10

doi: 10.153/oveje/16-0033

Plouin et al : European Society of Endocrinology Clinical Practice Guideline for the long-term follow up of patients operated on for a phaeochromocytoma or a paraganglioma

For more information and videos on NETs, diet and nutrition, tests, scans, treatments and side effects, please visit our website: www.netpatientfoundation.org and download ‘The NET Handbook’ and view our NET TV online channel.
A diagnosis of cancer is one of the greatest challenges you can face. It is normal to feel anxious, frightened and worried about the future. But when you’re dealing with a NET, there’s no need to ‘go it alone’. From diagnosis and throughout treatment and beyond, our team and our services are here to offer guidance, support, education and information to anyone affected by NETs every step of the way - whether it’s over the phone, on our website or a chat in person.

The NET Patient Foundation is the only charity in the UK dedicated to providing support and information to people affected by NETs. Here are some of the services we offer:

**Helpline – 0800 434 6476**

Our free, confidential helpline is for anyone who has questions about NETs. Your call will be answered by one of our nurses or trained staff members with experience of NETs.

**Discussion Forums**

The discussion forums are easy to use. If you’re feeling anxious or just need to hear from someone else who has been there they offer a way to gain support and reassurance from others in a similar situation to you.

**Patient Education Meetings**

We run information and support sessions for people living with NETs. These meetings include talks from some of the country’s top NET specialists.

**NET Natter Groups**

NET Natter meetings are informal support groups which offer an opportunity to meet with others in your area who are affected by NETs – patients, supporters, friends and family.