What are Phaeochromocytoma & Paraganglioma Sydromes?

PPGL syndromes are conditions that may cause multiple tumours called paraganglioma or phaeochromocytoma. These syndromes are caused by gene changes (mutations) that can be passed down in families. Sometimes the tumours may make greater than normal amounts of hormones, the body's chemical messengers, which in turn may cause a range of different symptoms. These tumours grow in particular cells of the body that mean that they are classed as neuroendocrine tumours or neoplasms (NET or NEN). All the tumours in PPGL syndromes can occur alone (sporadic) and independently from an inherited syndrome.

How are they diagnosed?

A person may be diagnosed with an inherited PPGL syndrome after being offered a gene test. Genetic testing may be offered either after someone is diagnosed with a paraganglioma or phaeochromocytoma, or because there is a family history of a PPGL syndrome.

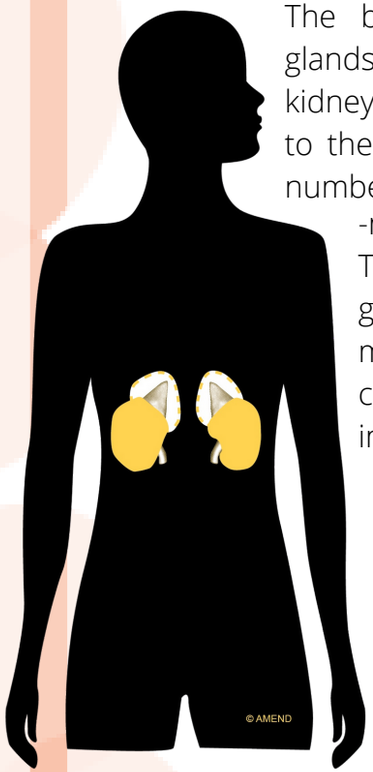
What are the syndromes called?

There are several different Phaeo Para syndromes that are named after the gene changes that cause them: SDHA, SDHB, SDHC, SDHD, SDHAF2, TMEM127 and MAX.

Each different syndrome has a different risk of developing different types of tumour and at different ages.

About Phaeochromocytomas

Phaeochromocytomas, often known as 'phaeos' ('fee-ohs'), are rare tumours that grow in the inner part of the adrenal gland.



The body has two adrenal glands, one on top of each kidney ('ad-renal' means 'next to the kidney'). They make a number of important hormo--nes including adrenaline. Tumours in the adrenal glands often make too much of these hormones causing symptoms including:

- headaches
- excess sweating
- anxiety
- dizziness
- heart palpitations
- nausea
- fatigue
- high blood pressure

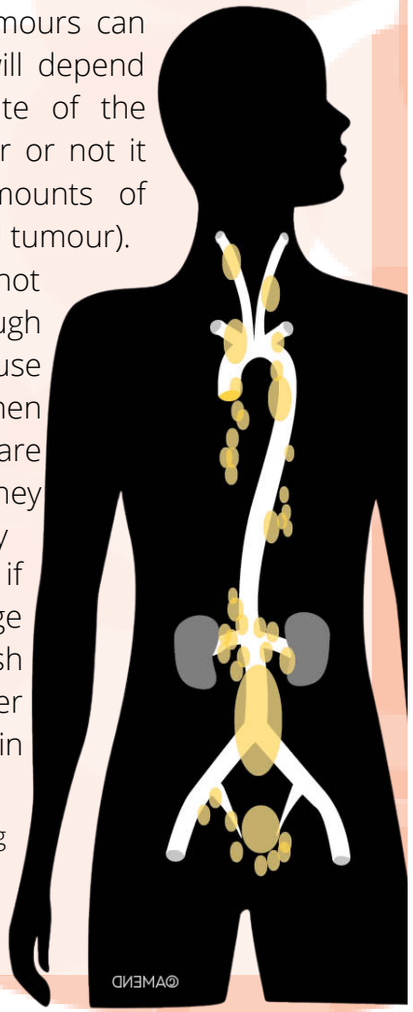
Treatment usually consists of surgery to remove the tumour and the gland, but only after the blood pressure has been controlled with special medicine called alpha-blockers.

About Paragangliomas

Paragangliomas are similar to phaeos but are more rare and grow outside of the adrenal glands. Paragangliomas arise from the extra-adrenal paraganglia. These are cells that form part of the nervous system that runs between the base of the skull and the pelvis. Therefore, these tumours can grow anywhere between these two points.

The type of symptoms that occur with these tumours can vary widely. They will depend on the size and site of the tumour, and whether or not it is making large amounts of hormones (functional tumour). Paragangliomas do not often make enough hormones to cause symptoms, but when they do, symptoms are similar to phaeos if they do. Instead, they may cause symptoms if they grow large enough to push against other structures or organs in the body. E.g.

- near the ear = hearing problems;
- near the wind-pipe = breathing difficulties



Patient Care

Anyone with these tumours and syndromes should be cared for in a specialist centre by an experienced multi-disciplinary team (MDT)

You will need regular tests and scans to make sure that you remain as healthy as possible and that any tumours that grow are found early on before they can cause severe symptoms.

About Phaeo Para Support UK

Phaeo Para Support UK is here to help you and your family by providing free patient-friendly information resources and support services. Joining Phaeo Para Support UK gives you access to the following:

- information resources
- information events
- monthly E-news and biannual newsletter
- our professional Counselling Service
- peer support meetings
- social media groups

All our information resources and support services are provided free to our registered members - membership is also free!

Information resources are developed with the help of our Medical Advisory Team to ensure accuracy.

Visit our website now to join for free!
www.phaeoparasupport.org.uk

Contact Us

Phaeo Para Support UK is a part of the Association for Multiple Endocrine Neoplasia Disorders (AMEND) which is a charity registered in England & Wales (no. 1153890)

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Information and Support for Families Affected by Inherited Phaeochromocytomas and Paragangliomas

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