

Primary Hyperparathyroidism

Information and advice on your condition.

This information sheet is for people who have primary hyperparathyroidism (PHPT). It provides some background information and advice on PHPT. It also discusses when treatment is needed and the types of treatment available.



hypopara.org.uk

What is primary hyperparathyroidism?

Primary hyperparathyroidism (often called PHPT or hyperpara) is a disorder of the parathyroid glands. One or more of the parathyroid glands becomes overactive and produces excessive parathyroid hormone (PTH). As a result, the calcium concentration in the blood may be increased.

What are the parathyroid glands and what do they do?

Parathyroid glands are endocrine (hormone-producing) glands that produce parathyroid hormone (PTH). Most people have four parathyroid glands which are each about the size of a grain of rice. These are in the neck behind the thyroid gland. The parathyroid glands control the concentration of calcium in the blood which is vital for the body to work properly.

When the parathyroid glands sense that the calcium in the blood is low, they release PTH. PTH increases circulating calcium through direct and indirect actions on the kidneys, the intestines and the bones, bringing the blood calcium back to normal. When the blood calcium is normal, the parathyroid glands stop releasing PTH. In PHPT, one or more parathyroids keep producing PTH irrespective of the blood calcium level, as the control mechanisms do not work effectively. This results in calcium being removed from the bones and causes high blood calcium (hypercalcaemia) which might cause problems and make you feel unwell.

What causes PHPT?

The most common cause is a benign (non-cancerous) growth (sometimes called an adenoma) in a single parathyroid gland. In other cases, the condition may affect more than one gland and occasionally, all four glands may be affected (which is sometimes called hyperplasia). Exceedingly rarely, the growth may be cancerous. There are other causes of high blood calcium; in these cases the PTH is usually low.

Who gets PHPT?

PHPT is the third most common endocrine condition. It is more common in women than men and commonly occurs in women after menopause. It does not usually run in families. However, there are rare inherited conditions such as multiple endocrine neoplasia (MEN) types 1 and 2A, familial isolated hyperparathyroidism (FIH) and hyperparathyroidism-jaw tumour syndrome (HPT-JT). There is some evidence of an increased risk of PHPT in people who have had radiotherapy to the head or neck but most often the cause of PHPT is not known.

What are the symptoms of PHPT?

You may not feel unwell at all even if your blood calcium is high, or you may feel some of the following symptoms, especially if your calcium increases.

The symptoms may include:

- Loss of energy
- Tiredness
- Feeling thirsty
- Frequent urination
- Passing an abnormally high amount of urine
- Stomach pain
- Constipation
- Bone pain

Some patients also report having:

- Mood changes
- Confusion
- Memory issues
- Depression

You may also suffer from more severe symptoms:

- Nausea and vomiting
- Heart rhythm disturbances
- High blood pressure
- Pancreatitis
- Kidney stones
- Osteoporosis (low bone density)
- Bone fractures

In exceptional cases of severe untreated high blood calcium the level of consciousness can be affected.

Symptoms of hypercalcaemia can also be associated with other disorders so it is important to see your doctor to get the right diagnosis.

How is PHPT diagnosed?

Doctors usually diagnose PHPT after finding a high level of calcium on a blood test along with a high or 'inappropriate' level of PTH. Phosphate levels may be normal or low. Vitamin D may also be low and should be corrected. Some people may already have symptoms suggesting PHPT but it is often found with a routine blood test.

Your GP will refer you to a specialist endocrinologist who will arrange for further tests to confirm the diagnosis, rule out other causes, and assess you for possible adverse consequences of the disease. If surgery is required you will then be referred to a surgeon who specialises in parathyroid surgery – usually an endocrine or head and neck surgeon.

Some people may feel symptoms of high calcium even though their blood calcium is within the reference range (usually the upper end of normal). The term 'insidious' or normocalcaemic PHPT has sometimes been used to describe this but the diagnosis is difficult to demonstrate. It is important to exclude vitamin D deficiency and other causes.

What other tests may be performed for PHPT?

Various blood tests and scans may be done to help with diagnosis. The most common are:

- *Blood testing* for kidney function, other possible causes of hypercalcaemia, and vitamin D status (measurement of 25(OH)D and 1,25 dihydroxy vitamin D)
- *DXA scan* – a special type of X-ray that measures bone mineral density (BMD)
- *Morning fractional calcium excretion test* or a *24 hour urine collection* – to measure calcium in urine
- *Kidney ultrasound* or *CT scan* – to check for kidney stones

If you are recommended to have surgery you may be referred for the following tests to localise the enlarged parathyroid(s):

- *Ultrasound neck scan*
- *Sestamibi parathyroid scan*
- *CT neck scan* (less common)
- *Parathyroid venous sampling* (this should be

performed only after previous failed surgery at centres with experience in this technique, i.e., when the parathyroids are difficult to localise)

If the tests locate one or more enlarged glands, a minimally invasive or 'key-hole' surgery may be possible. However, even in the best of hands, imaging scans are negative in about 20-30% of cases. Your endocrinologist and surgeon will discuss with you the option of an exploratory operation to find the enlarged/overactive gland(s). Enlarged glands not seen on scans can often be seen by experienced surgeons.

What are the risks with PHPT?

Patients with PHPT have an increased risk of developing problems in a number of organs, particularly the bones and kidneys. There is a higher chance of developing osteoporosis, kidney stones and gradual deterioration in kidney function. Other risks include neuropsychiatric symptoms (such as depression, anxiety and forgetfulness), falls and fractures, constipation, pancreatitis and (arguably) a slightly higher risk of cardiovascular problems in the long term. Some patients with only a minimal increase in calcium may have significant symptoms.

Does PHPT need treatment?

If there is evidence that PHPT is affecting your health, then treatment will usually be recommended. If your vitamin D is low it should always be corrected before any treatment. If your blood calcium is dangerously high you will be referred immediately to hospital for your calcium concentration to be decreased and may be referred for an urgent operation.

Surgery

Current guidelines for the management of hyperparathyroidism recommended an operation called a parathyroidectomy if you have any of the following i) significant symptoms, (ii) significantly high calcium, (iii) significantly impaired kidney function, (iv) kidney stones, (v) osteoporosis or (vi) you are under the age of 50. This means that

parathyroid surgery is also often recommended in patients over 50 years of age even if they do not have symptoms as it is likely to decrease the long-term consequences of hyperparathyroidism on the bones and kidneys.

A parathyroidectomy involves making an incision in your neck to find and remove the abnormal parathyroid gland(s). This operation does not take very long and has a high chance of curing the condition. Your PTH may be monitored during surgery.

It is important that parathyroid surgery is carried out by a highly skilled parathyroid surgeon. Do not hesitate to ask about your surgeon's experience, the number of parathyroid operations they perform and their complication rate. You will be asked to provide informed and written consent.

After surgery, low concentrations of PTH cause a condition called hypoparathyroidism which may be temporary or permanent. If permanent or if you have most of your parathyroid glands removed (for example, if you have multigland disease) you will need lifelong treatment with vitamin D, magnesium and calcium supplements and regular blood tests.

Medical treatment

If your blood calcium is high but you are unable to have an operation for any reason, you might be prescribed a drug called cinacalcet (Mimpara). This drug can lower the concentration of calcium, reducing symptoms and improving quality of life. It does not however increase bone density or reduce the risk of developing kidney stones. Some patients taking this medication may develop side effects, most commonly, nausea.

If your bone density is reduced, you might be prescribed an anti-osteoporosis drug such as alendronate. Alendronate is a bisphosphonate drug used in treating osteoporosis. Some clinicians advise deferring bisphosphonate treatment if you are planning to have surgery; discuss this with your endocrinologist or surgeon.

If you have no symptoms, or very mild symptoms, then it is possible that no treatment will be needed, although vitamin D levels should be corrected if necessary. You will however be monitored by a specialist or by your GP (with advice from the specialist). You will need to have regular tests to monitor your blood calcium, to check that your kidneys are working normally, and to check your bone density for results which may indicate the need for an operation. You will also be monitored in this way if you decline surgery or if surgery is not an option. You will also be given lifestyle advice such as to drink plenty of fluids, have regular exercise, and to seek medical help if you develop persistent vomiting and diarrhoea. If you have symptoms do not hesitate to contact your doctor.

What is the outlook?

After surgery, your PTH level may take a little time to settle down. If you feel unwell or have tingling, numbness, or cramping symptoms get in touch with a doctor immediately. You may need some medication until the PTH and calcium are within the normal range again.

Following successful parathyroid surgery, you will be able to resume a normal life. Studies have shown that bone density improves, risk of subsequent fractures and kidney stones are reduced, and there may well be improvements in energy, mood, and memory.

Useful organisations

AMEND

Patient support & advice on Multiple Endocrine Neoplasia Disorders (MEN)

Tel: +44 (0)1892 516076

www.amend.org.uk

Society for Endocrinology

Patient information about 'You & Your Hormones'
www.yourhormones.info/Endocrine_conditions/Primary_hyperparathyroidism.aspx

BAETS

Information for patients about surgery
www.baets.org.uk/useful-links/patients/

For further information, support or advice about hyperparathyroidism, please contact:

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East Grinstead, West Sussex RH19 4DF**

Tel: +44 (0)1342 316315 (England & Wales)

Tel: +44 (0)1475 522576 (Scotland)

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About Hypopara UK

Hypopara UK is a national patient organisation for families affected by a parathyroid condition – either hyperparathyroidism or any form of rare hypoparathyroidism. Via its website, groups and helplines it provides support and information to empower patients to become active in the management of their particular condition.

Hypopara UK works closely with its clinical advisory team in the UK and raises awareness about parathyroid conditions internationally to help improve the quality of life for patients.

Written by Judith Taylor & Liz Glenister of Hypopara UK with the Hypopara UK Clinical Advisory Team, and endorsed by the Society for Endocrinology Clinical Committee of the Bone and Calcium Endocrine Network.

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