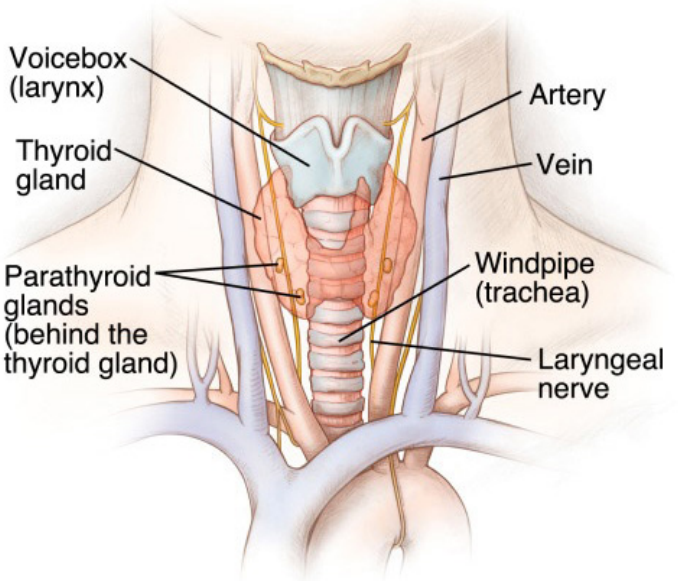
A PATIENT GUIDE TO **PRIMARY HYPERPARATHYROIDISM**

Introduction



The parathyroids are four, very small glands, located in the neck just behind the thyroid gland. They secrete a natural chemical called parathyroid hormone (PTH) into the bloodstream.

PTH helps to control the levels of two salts in the body: calcium and phosphate.

The most common parathyroid disorders are Primary Hyperparathyroidism (PHPT) and Hypoparathyroidism (HypoPT).

How are PHPT and HypoPT different?

PHPT occurs when one or more of the parathyroid glands produce too much PTH. In contrast, HypoPT occurs when the parathyroid glands produce too little PTH.

What happens when PTH is high?

A high level of active PTH causes blood calcium levels to rise (hypercalcemia), and phosphate levels to fall (hypophosphatemia).

Why does the body need calcium and phosphorus?

Calcium and phosphorus combine to make calcium phosphate in the body. Calcium phosphate gives bones and teeth strength. They are also needed for muscles and nerves to work properly.

Calcium and phosphate levels are kept within a narrow range for your well-being. Both high and low values of these salts can be harmful to your health.

What are common causes of PHPT?

PHPT is commonly caused by a non-cancerous tumour (adenoma) on one of the parathyroid glands.

Less commonly, it can occur if 2 or more parathyroid glands become enlarged.

Women are more likely to develop PHPT than men, and most patients are over 60.

What symptoms occur in PHPT?

In many cases, PHPT is diagnosed incidentally during a blood test for other reasons. This is because patients may not show symptoms when calcium levels are only slightly elevated. As calcium levels increase, so too does the likelihood of symptoms, which can include:

- thirst
- increased frequency of urination
- fatigue
- memory impairment
- mood disturbance
- muscle/bone pain
- constipation
- abdominal pain

PHPT increases the risk of bone fractures and kidney stones.

Which tests help diagnose PHPT?

The body's normal response to high calcium levels is to suppress the production of PTH. Therefore, the combination of high calcium levels with high (or inappropriately normal) PTH levels almost always points to the diagnosis of PHPT.

A much rarer inherited (genetic) condition (familial hypocalciuric hypercalcemia or FHH) can cause hypercalcemia. It is screened for by assessing calcium levels in a urine sample.

PHPT is not normally inherited but genetic testing may be recommended in patients under 30 years, or when there is a strong family history of PHPT.

After the initial diagnosis is confirmed, further tests are recommended to identify people at high risk of bone fractures (a DXA scan) and to screen for kidney stones (by scanning the abdomen).

How much calcium should you consume if you suffer from PHPT?

People with PHPT do not need to restrict their dietary calcium intake, but should not exceed the recommended daily limits.

To help estimate your dietary calcium intake, you may be asked to complete an online questionnaire.

Do you need to take vitamin D if you suffer from PHPT?

Vitamin D deficiency is very common in European populations and measurement of blood vitamin D levels is recommended in people with PHPT. Vitamin D replacement is known to be safe in the context of PHPT although it is not recommended in people with very high calcium levels.

How is PHPT treated?

The only cure for PHPT is to surgically remove the affected gland(s). Not everyone with PHPT will require surgery but the following factors tend to support surgical intervention:

- Presence of symptoms likely to be due to high calcium levels
- Markedly elevated calcium levels
- Younger age
- Bone-thinning (osteoporosis) or bone fractures
- Kidney stones

Where none of these features are present, monitoring of calcium levels and, in some cases, medication may be considered as an alternative to surgery. Where surgery is being considered, neck imaging (e.g. ultrasound) is typically performed to identify the enlarged parathyroid gland(s) prior to the operation.

More than one type of scan may be required to identify the abnormal gland(s).

What are the benefits of surgery?

Curative surgery is associated with improvements in bone density and reduces the risk of kidney stones. It is much more difficult to predict whether surgery will improve symptoms in people with PHPT.

Parathyroid surgery is associated with improved quality of life but it is often difficult to know whether an individual's symptoms are directly linked to high calcium levels.

What risks are associated with surgery?

Parathyroid surgery is typically a safe procedure when performed by experienced surgeons.

Most parathyroid surgery takes place under general anaesthesia (fully asleep) and involves a small incision at the front of the neck. Potential complications of surgery include:

- Wound infection
- Bleeding
- Scar
- Neck stiffness (typically transient)
- Hoarseness, if voice box nerve injury occurs
- Low calcium levels (typically transient and treated with calcium and vitamin D tablets)
- A 2nd operation, if the initial surgery is not curative

What happens if you do not have surgery?

In PHPT patients with mildly elevated calcium levels, it is quite common for their blood test to remain stable over several years. Regular monitoring of calcium levels is recommended, where surgery is not pursued, to identify individuals with rising calcium.

Even if calcium levels remain stable, PHPT may cause symptoms and complications that can affect the bone and kidney. During follow up, careful consideration should always be given to whether surgery is indicated.

Can you treat PHPT with drugs?

The only cure for PHPT is surgery. Medications can be prescribed to help improve bone density but these typically have little effect on calcium levels.

A medication called cinacalcet can be prescribed to lower calcium levels but has no significant effect on bone density and kidney stone risk.

Is it safe to become pregnant when you have PHPT?

PHPT with hypercalcemia can be harmful for the unborn and newborn, so it is recommended that pregnancy be deferred until surgery has been performed.

Please refer to the ESE Parathyroid Disorders during Preconception, Pregnancy and Lactation guide.

Does a high level of PTH always mean you have PHPT?

PTH levels are strongly influenced by dietary calcium intake (and absorption from the gut) and vitamin D status (partly dietary and partly through exposure to sunlight).

Therefore, PTH levels can be raised in people with insufficient calcium intake and in people with vitamin D deficiency. In these situations, calcium levels will typically be normal. PTH will normalize following a period of calcium and/or vitamin D supplementation.

People with significant kidney disease frequently have high levels of PTH but this is also typically associated with normal calcium levels and is known as secondary hyperparathyroidism.

Other causes of raised PTH include increased urinary calcium, malabsorption, drugs and other less common causes.

European Society of Endocrinology

Patients: The concise information in this guide reflects the latest expert recommendations on managing this condition. Please consult your own family doctor or patient advisory group for further advice and support.

Clinicians: This introductory patient guide reflects the latest expert consensus recommendations of PARAT - the ESE educational programme on parathyroid disorders. The guide should not replace clinical consultation with individual patients.

Please view "European Expert Consensus on Practical Management of Specific Aspects of Parathyroid Disorders in Adults and in Pregnancy". *European Journal of Endocrinology* 186 (2) February 2022, for recommendations in full.

Faculty members Fraser Gibb (UK), Claudio Marcocci (Italy), Luis Cardosa (Portugal), Elena Tsourdi (Germany) and Nik Screen (ESE/Versatility.org.uk) prepared this patient information. Further patient information guides for hypoparathyroidism and preconception, pregnancy and lactation, plus all other PARAT educational materials are available at **www.ese-hormones.org** or by searching; **bit.ly/paratlz**