So you have primary hyperparathyroidism

You may feel fine—but you could need surgery.

If you’ve been told that you have primary hyperparathyroidism, chances are the diagnosis came as a complete surprise. It’s typically discovered in the course of blood testing for other reasons, and there are usually no symptoms—or only nonspecific ones, such as fatigue, constipation, aches and pains, trouble concentrating, or low mood. Primary hyperparathyroidism occurs mainly in postmenopausal women, who may blame such vague symptoms on other health problems or just on normal aging. In the United States, about one in 500 women ages 60 and over develops the disorder each year.

At one time, the condition was recognized only when it had progressed to the point of causing serious problems such as bone disease, kidney stones, gastrointestinal disorders, and cognitive difficulties—classic signs and symptoms that clinicians have dubbed “the bones, stones, abdominal moans, and groans.” These days, some people with primary hyperparathyroidism do have such symptoms. But since the introduction of automated blood chemistry screening in the 1970s, clinicians have been able to detect the disease, which causes elevated levels of calcium in the blood (hypercalcemia), before symptoms appear. This raises the question of what to do about asymptomatic (non-symptomatic) primary hyperparathyroidism, which now accounts for 80% of cases. Most people with asymptomatic primary hyperparathyroidism can have a fairly benign course for years, even a lifetime. But the condition does progress in some people, and they should undergo surgery, which is the only complete cure, as soon as possible. The question is, who?

Expert panels have met three times since 1990 to review the evidence and update guidelines for managing the disorder in people without symptoms, in an effort to identify those most likely to benefit from surgery and to make recommendations for monitoring the rest. It’s important to know your options and understand all the risks and benefits. If you’ve been diagnosed with primary hyperparathyroidism, your first step is to find an endocrine specialist with extensive experience in treating the condition.

Anatomy of the parathyroid glands

The parathyroid glands are tiny glands located at the back of the thyroid gland in the neck. They produce parathyroid hormone (PTH), increasing output when blood levels of calcium are low and decreasing output when levels return to normal. Primary hyperparathyroidism occurs when one or more of these glands become enlarged and produce too much PTH, raising the level of calcium in the blood above the normal range. The usual cause is an adenoma—a noncancerous tumor.
**What causes it?**

Primary hyperparathyroidism is a disorder of the parathyroid glands, four tiny pea-sized glands located behind the thyroid gland in the neck. Rarely, a parathyroid will be located elsewhere in the neck or upper chest region, and it’s possible, though not common, to have more than four glands.

These glands produce parathyroid hormone (PTH), a hormone that helps regulate the balance of calcium and phosphorus in the body. Normally, the parathyroids pump out just enough PTH to counteract a drop in blood levels of calcium. PTH raises calcium levels by triggering the release of calcium from the bones and increasing calcium absorption in the kidneys and intestines. When calcium levels come back into line, PTH production drops off. Because calcium levels fluctuate throughout the day, the parathyroids are continually tweaking their PTH output to keep calcium at a normal level.

In primary hyperparathyroidism, one or more parathyroid glands produce more PTH than needed, raising calcium levels above the normal range. Usually, the cause is a benign (noncancerous) tumor, or adenoma, in a single parathyroid gland. Occasionally, adenomas grow on more than one parathyroid gland. In very rare instances (less than 0.5% of cases), a parathyroid tumor is cancerous.

Parathyroid glands may become overactive because of lithium therapy, past radiation to the neck, or certain gene defects. Roughly 3% to 5% of cases are linked to inherited syndromes. In some cases, the cause is simply unknown.

**How is it diagnosed?**

Some experts consider the term “asymptomatic” something of a misnomer because of the many nonspecific complaints associated with the disease. However, these aren’t clear heralds of the condition, and people often discount them and don’t seek medical attention for them.

Sometimes the diagnosis of hyperparathyroidism is made when doctors seek an explanation for low bone density noted on testing. Most often, it’s made when routine blood screening detects hypercalcemia. The clinician will follow up with a repeat calcium measurement to confirm the elevated reading. PTH will also be tested. Inappropriately high levels of calcium and PTH on follow-up indicate primary hyperparathyroidism.

Thiazide diuretics and lithium can also cause hypercalcemia, so if PTH is high in people taking either drug, they should temporarily halt drug therapy, if possible, and have repeat PTH and calcium tests two or three months later.

Other diseases, including cancer, may raise calcium levels, but few are also associated with excessive PTH. One exception is familial hypercalciuric hypercalcemia (FHH), a genetic disorder that resembles primary hyperparathyroidism but doesn’t need to be treated. FHH is ruled out with urine tests in order to avoid unnecessary parathyroid surgery.

To confirm the diagnosis, an endocrinologist may order additional tests for blood levels of phosphorus, vitamin D, creatinine, and biomarkers of bone turnover, as well as bone density testing using standard dual energy x-ray absorptiometry scanning. Bone loss in primary hyperparathyroidism is usually more pronounced at the forearm than at the spine or hip, but all three sites should be evaluated.

**What about surgery?**

When it causes kidney or bone disease or severe symptoms, primary hyperparathyroidism almost always calls for parathyroidectomy—removal of the enlarged parathyroid gland (or glands). In experienced hands, this operation has few complications, and it cures the condition 95% to 98% of the time.

Experts are less certain about what to do when the condition is asymptomatic. Asymptomatic primary hyperparathyroidism progresses to a more serious condition only about one-third of the time, so some experts favor simply monitoring most patients and, if necessary, managing the condition with lifestyle measures and medications. Others argue that the long-term cost of such medical management may exceed that of the relatively simple operation, which is increasingly performed as an outpatient procedure. Surgery eliminates the risk of disease progression altogether and may also get rid of many of the nonspecific complaints.

In guidelines published in the February 2009 issue of *The Journal of Clinical Endocrinology and Metabolism*, experts acknowledged that medical management might be appropriate for people who don’t meet the criteria for surgery (see “Decision factors for surgery in asymptomatic primary hyperparathyroidism”) or who can’t or won’t undergo the surgery. But they also determined that surgery might ultimately be recommended even for the most asymptomatic patients. The new evidence comes from recent studies.
indicating that surgery reduces the lifetime risk of fractures. This research, which includes three randomized trials and a 15-year observational study, indicates that without surgery, bone density is likely to start worsening eight to 10 years after the diagnosis. Surgery consistently improves bone density and quality of life.

**Surgery: What’s involved?**

The standard surgery, called bilateral neck exploration, entails making a two- to five-inch incision across the front of the neck, examining all four parathyroid glands, and removing the enlarged ones. It’s usually performed under general anesthesia, although local anesthesia is sometimes an option. A single abnormal parathyroid is usually at fault, but sometimes two or more must be removed.

Today, minimally invasive surgery is increasingly preferred. It involves a smaller, one- to two-inch incision, takes less time, and requires less anesthesia. This technique can be used whenever preoperative imaging indicates that there is a single abnormal parathyroid gland. The most common imaging method is sestamibi scanning, which uses a radioactive isotope called technetium sestamibi to identify the abnormal gland. (See the illustration “What is sestamibi scanning?”) An ultrasound of the neck may be ordered as well.

PTH is tested in the operating room after the abnormal parathyroid gland is removed. The level should drop by more than 50% within a few minutes. If it does, surgery is over. If it doesn’t, the surgeon will abandon the minimally invasive technique and look for other abnormal glands.

When skilled parathyroid surgeons perform these procedures, complication rates are very low (1% to 3%). Possible complications of both traditional and minimally invasive surgery include damage to the laryngeal nerve (which can cause hoarseness) and decreased parathyroid function. Work with your endocrinologist to find a surgeon with a lot of experience—at least 50 parathyroid surgeries a year, according to some experts. To locate an endocrinologist near you, contact the American Association of Clinical Endocrinologists, www.aace.com, 904-353-7878, or the Hormone Foundation, www.hormone.org, 800-467-6663 (toll-free).

**Nonsurgical measures**

If you have asymptomatic primary hyperparathyroidism and aren’t planning on surgery, you should have regular blood tests and bone density scanning (see “Monitoring primary hyperparathyroidism in nonsurgical patients”). You can also take the following steps to reduce the risk of bone loss and kidney problems:

- To lower the chances of kidney stones developing, drink at least six to eight glasses of water each day.
- Get adequate exercise, which helps protect your bones.
- Maintain a moderate intake of calcium (1,000 to 1,200 milligrams per day) and vitamin D (400 to 600 international units per day).
- Avoid thiazide diuretics and lithium therapy, which can contribute to hypercalcemia.

No currently available medication can cure primary hyperparathyroidism, but some may help reduce the complications. Alendronate (Fosamax), a bisphosphonate, decreases bone turnover and increases bone density at the spine and hip region, although it doesn’t correct PTH or calcium levels in the blood. Cinacalcet (Sensipar) is a drug that helps normalize calcium levels by acting on the calcium-sensing receptors in the parathyroid glands to reduce PTH levels. So far, studies have shown no resulting improvement in bone density.

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### Monitoring primary hyperparathyroidism in nonsurgical patients

<table>
<thead>
<tr>
<th>Test</th>
<th>Frequency</th>
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<tbody>
<tr>
<td>Blood calcium</td>
<td>Annually</td>
</tr>
<tr>
<td>Blood creatinine</td>
<td>Annually</td>
</tr>
<tr>
<td>Bone density</td>
<td>Every 1–2 years at the forearm, hip, and spine</td>
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**What is sestamibi scanning?**

Sestamibi scanning is a painless technique often used before surgery to locate abnormal parathyroid glands and determine whether minimally invasive surgery is appropriate. A radiolabeled compound (technetium sestamibi) is injected into a vein and is taken up preferentially by the abnormal parathyroids. The patient lies still on a table while two cameras circle the upper body, recording images from opposite sides. The abnormal glands will show up as areas of increased uptake on the scan.
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