

## PRIMARY HYPERPARATHYROIDISM

Primary Hyperparathyroidism is a hormonal problem due to one or more of the parathyroid glands producing too much parathyroid hormone. When this happens, the blood calcium becomes higher than normal. Bones may lose calcium, and too much calcium may be excreted by the kidneys.

### 1Q. *What are the parathyroid glands?*

1A. They are four small glands located in the neck, close to the thyroid gland. Rarely there are more than four, and they may be located in other parts of the neck, or even in the chest.

### 2Q. *What is their purpose?*

2A. The glands produce parathyroid hormone which plays a critical role in maintaining normal blood calcium levels.

### 3Q. *How do these small glands maintain normal calcium levels in the blood?*

3A. Parathyroid hormone, released by these glands, keeps the amount of calcium in the blood from falling below normal in three ways:

1. By conserving calcium at the kidneys.
2. By releasing calcium from the bones.
3. By increasing absorption of calcium from food.

### 4Q. *What goes wrong in primary hyperparathyroidism?*

4A. One or more of the glands becomes enlarged and overactive, producing too much parathyroid hormone. This leads to a rise in the blood calcium. In most patients (80-85%), a single parathyroid gland becomes enlarged and develops into a benign tumor, known as an adenoma. In nearly all other patients (15-20%), two or more glands enlarge, again in a benign fashion, a condition called hyperplasia. Parathyroid cancer is an extremely rare cause of primary hyperparathyroidism.

### 5Q. *What are the harmful effects of primary hyperparathyroidism?*

5A. When the blood calcium is very high the most common symptoms are loss of appetite, thirst, frequent urination, lethargy, fatigue, muscle weakness, joint pains, and constipation. Symptoms can also include nausea, vomiting, abdominal pain, memory loss and depression. These problems are not usually present unless the blood calcium is very high. Most patients with primary hyperparathyroidism in the United States do not have blood calcium levels in the range where these signs typically occur. In fact, many patients with primary hyperparathyroidism have no symptoms at all. They are said to be asymptomatic. Even so, patients with primary hyperparathyroidism who are asymptomatic can lose bone density and be at risk for fractures because of a loss of calcium from the skeleton. Primary hyperparathyroidism can also lead to kidney stones and calcification of the kidney.

### 6Q. *Do all patients with primary hyperparathyroidism have symptoms?*

6A. No. Most patients are discovered to have primary hyperparathyroidism incidentally, in the course of a routine blood test.

### 7Q. *Do all patients with hyperparathyroidism develop complications including bone loss, kidney stones, weakness, fatigability, etc?*

7A. No. many patients do not develop obvious complications, but bone loss is not uncommon.

**8Q. *Does the physician know who will develop complications?***

8A. At the time of diagnosis, the physician cannot predict who will develop complications. However, complete biochemical and radiological evaluation over time will identify individuals who may exhibit the first signs of these complications before they progress to a more serious stage.

**9Q. *What is a complete evaluation?***

9A. Blood is tested for calcium and parathyroid hormone to establish the diagnosis and assess severity. Blood is also tested for 25-hydroxyvitamin D and creatinine to ensure that the disease is not being made worse by abnormalities in these factors. Urine is tested for calcium and creatinine to determine if there is an increased risk for kidney stones. Measuring "bone markers" in blood, such as alkaline phosphates and in urine, such as collagen breakdown products, can assess the effect of the disease on bone. Kidney x-rays or ultrasound tests are obtained to assess for the presence of kidney stones. Bone mineral density (spine, hips, forearm) is measured to detect bone loss. It is important to measure the forearm as well as the lumbar spine and hip because the forearm can be the first site where bone is lost in people who have primary hyperparathyroidism.

**10Q. *What causes primary hyperparathyroidism?***

10A. In most cases the cause is unknown. Previous x-ray treatments of the face or neck years earlier may be a cause in some patients. Treatment with drugs such as lithium and thiazide diuretics may lead to a higher risk for developing primary hyperparathyroidism. In some patients there is a family history of primary hyperparathyroidism. However, the great majority of patients do not have any relatives with this disorder.

**11Q. *How common is primary hyperparathyroidism?***

11A. Estimates are difficult to obtain. Between 1983 and 1992, a study at the Mayo Clinic in Rochester, Minnesota found that the annual number of newly diagnosed patients was 20.8 per 100,000. IN 1999 approximately 12,000 patients had parathyroid surgery in the United States. Women patients outnumber men by three to one. Primary hyperparathyroidism can occur at any age, but is more common with aging, and its highest occurrence is in people over the age of 50.

**12Q. *Is there a cure for primary hyperparathyroidism?***

12A. Yes. Surgery to remove the affected gland(s) cures the condition. When performed by an experienced parathyroid surgeon, the operation is successful in over 95% of cases. Serious surgical complications are uncommon. The surgery usually leaves a thin, faint, horizontal scar about three inches long in the lower neck.

**13Q. *Should all patients with primary hyperparathyroidism undergo surgery?***

13A. Not necessarily. Although surgery may be considered an appropriate treatment even in patients without signs or symptoms, such patients are not always operated upon because the disease is mild. In these patients, the disease may not get worse. If surgery is performed, these patients should be monitored regularly with blood testing every six months, and bone mineral density testing yearly. This will allow the physician to identify those patients who show signs of progression and who require active treatment.

**14Q. *Who should have surgery?***

14A. Patients who have symptoms of primary hyperparathyroidism are generally recommended for surgery unless they have other medical problems which might make the surgery too risky. Patients who have no symptoms may also have surgery suggested if their physician, after careful analysis of their condition, feels that surgery would likely prevent future complications.

**15Q. *Is there a way to locate the enlarged parathyroid gland(s) before surgery is performed?***

15A. An experienced surgeon who performs many parathyroid operations annually, generally does not require a preoperative localization test to do a successful operation. However, preoperative localization technology has advanced to the point where many expert parathyroid surgeons now are favorably inclined to having an imaging study performed prior to surgery. There are certain specific situations where a localization study prior to surgery is necessary: (1) for patients who have had previous neck surgery; (2) for patients who elect to have minimally invasive surgery (see question 18). It should be noted that the abnormal gland may not always be found by localization studies, and that some localization studies may indicate that a parathyroid gland is present when it is not, a 'false positive test'.

**16Q. *What preoperative localization tests are available?***

16A. There are a number of non-invasive imaging tests to locate abnormal parathyroid glands. They include ultrasound, computerized tomography (CT), magnetic resonance imaging (MRI), positron emission tomography (PET), and the sestamibi scan. Sestamibi is an imaging agent that localizes in the abnormal parathyroid gland. The sestamibi scan, especially when it is performed with computerized tomography (SPECT), is the most sensitive and specific test available. In unusual situations, the quest to find the parathyroid gland advances to invasive tests (arteriography and blood sampling of veins in the neck for parathyroid hormone) that require great expertise and are more difficult. Patients who are considering minimally invasive parathyroid surgery (see question 18), require successful preoperative imaging to localize the abnormal parathyroid glands.

**17Q. *What is the standard surgical approach for primary hyperparathyroidism, and are there alternative approaches?***

17A. The traditional approach is to perform the operation under general or local anesthesia. All four glands are identified. Any enlarged glands, usually one, are removed. If all glands are enlarged at least three are usually removed.

Recently, three newer techniques have been developed. They go under the term "minimally invasive parathyroidectomy". These approaches require successful preoperative localization with a sestamibi scan.

1. A smaller incision is made in the neck in the area where the gland was identified and the gland is removed. This approach should be combined with the use of parathyroid hormone measurements done while the patient is still in the operating room. If the blood parathyroid hormone, which can be measured within 15 minutes, falls to below 50% of the level before surgery, the surgery is nearly always successful, and the other glands do not have to be examined.

2. A second approach is to do the sestamibi scan a few hours before surgery and use a radioactivity detector to more rapidly identify the abnormal gland(s). Once more, an appropriate 50% decrease in blood parathyroid hormone confirms the success of the procedure.

3. A third approach is to use an endoscopy tube through a small incision to locate the gland(s) that have been identified by the sestamibi test.

These three approaches should be undertaken only by surgeons with great experience in parathyroid surgery.

**18Q. *Can the standard parathyroid operation be performed under local anesthesia?***

18A. Yes. Parathyroid surgery can be performed under general, regional (spinal nerve block), or local anesthesia. In many centers, local anesthesia has become routine. In all cases, an experienced surgeon is strongly recommended, regardless of the kind of operation that is performed.

**19Q. *What is parathyroid autotransplantation?***

19A. Parathyroid autotransplantation may be used in patients when all four parathyroid glands are abnormal. It is also of benefit in patients who have previously had an unsuccessful operation to treat hyperparathyroidism since these patients are more likely to develop hypoparathyroidism, a condition in which the blood calcium is low because of inadequate secretion of parathyroid hormone. After all four glands are removed; a small piece of one gland is transplanted into the muscle of the forearm. If a high blood calcium develops because the transplanted parathyroid tissue enlarges over time, this can be corrected by removing some of the tissue in an outpatient procedure under local anesthesia. Parathyroid autotransplantation should only be performed by surgeons with experience in this technique.

**20Q. *Is surgery always successful?***

20A. Not always. The abnormal gland may not be found at surgery. There are several reasons for this:

1. Many abnormal glands are small.
2. The gland may not be in its normal location in the neck, but instead may be in the chest or another unusual location.
3. The surgeon is not highly experienced in the operation.
4. In patients with four abnormal glands, the patient may not be cured because not enough parathyroid tissue was removed; or after apparent cure, the growth of the residual overactive parathyroid tissue produces a recurrence of high blood calcium.
5. The diagnosis is not correct.

**21Q. *Are there alternatives to surgery?***

21A. There are no other cures for primary hyperparathyroidism besides surgery. Estrogen therapy in post menopausal women reduces the blood calcium to some extent, and increases bone mineral density, but does not control the increased secretion of the parathyroid gland. Experimental drugs are being developed to reduce parathyroid hormone secretion. One such drug, Sensipar® (cinacalcet), is approved by the U.S. Food and Drug Administration (FDA) for reducing parathyroid secretion in rare patients with parathyroid cancer and in patients with chronic renal failure who are on dialysis. Although cinacalcet has been studied in primary hyperparathyroidism, the FDA has not yet approved it for primary hyperparathyroidism. Alendronate, one of the bisphosphonates, a group of drugs used to treat osteoporosis and Paget's disease, increases the bone mineral density in patients with primary hyperparathyroidism but does not affect the level of calcium or parathyroid hormone.

**22Q. *What general measures should patients with primary hyperparathyroidism follow?***

22A. Patients should always maintain good hydration by drinking enough fluids. (Dehydration will lead to an increase in the blood calcium.) Immobilization, which can also cause increased blood calcium, should be avoided. A low level of calcium intake should be avoided since this may stimulate the parathyroid glands further. On the other hand, it is not advisable for patients to receive too much calcium. A daily intake of about 1000 mg daily from food and/or supplements is recommended.

**23Q. *Which types of physicians are specialists in treating primary hyperparathyroidism?***

23A. Specialists include endocrinologists, physicians who specialize in hormonal disorders ie. the staff at AAEDA, and surgeons who specialize in endocrine surgery.

**Be sure to contact your AAEDA physician if you have any questions regarding primary hyperparathyroidism, or if you would like to schedule a consultation.**