Parathyroid problems in children

Patient and family information, brought to you by the Education Committee of APSA

Overview - "What is it?"

Parathyroid glands are small organs of the endocrine system that are located in the neck behind the thyroid. Parathyroid glands (we all generally have four of them) are normally the size of a pea (or even smaller). These glands control the calcium in our bodies by making a hormone called parathyroid hormone or "PTH". PTH controls the level of calcium and phosphorus—minerals that are in bones but also circulate in the blood. PTH controls how much calcium is lost in the urine (by its effects on the kidneys) and works with vitamin D to control how much calcium we absorb from our food.

Our bodies very carefully regulate calcium because it is important in many of the body's functions. If the parathyroid glands make too much or too little hormone, it disrupts this delicate balance. If the glands do not make enough PTH, then you have hypoparathyroidism. More commonly, diseases of the parathyroid gland result in hyperparathyroidism—where your body is making too much PTH and the blood calcium rises. Too much calcium in the blood is called hypercalcemia. Although the thyroid and parathyroid glands are neighbors and are part of the endocrine system, the thyroid and parathyroid glands are unrelated—they just have similar names. Parathyroid problems in children are less common in children compared to adults.

Hypoparathyroidism: Not enough PTH and low blood calcium levels is most commonly caused by injury during an operation of the parathyroid glands—but it only happens in 1-2% of those patients. Some children are born without parathyroid glands, or glands that cannot make enough PTH such as DiGeorge Syndrome. Although DiGeorge Syndrome may affect one baby out of every 2,000 births, not all of those children will suffer from hypoparathyroidism.

Hyperparathyroidism: Too much PTH and high body calcium can be caused by a number of problems, but there are two general types.

- **Primary Hyperparathyroidism**: an enlargement of one or more of the parathyroid glands causing too much PTH to be made and therefore high calcium in the blood. The chance of primary hyperparathyroidism in a child is less than 5 in 100,000. The most

common cause of primary hyperparathyroidism is a benign (non-cancerous growth) tumor on a single parathyroid gland that makes it overactive. Sometimes, extra hormone is made from all four parathyroid glands being bigger than normal. Cancer of the parathyroid glands is very uncommon, but usually causes symptoms that are also due to high blood calcium levels.

- **Secondary hyperparathyroidism:** occurs as a result of another disease that initially causes low levels of calcium in the body which then causes the parathyroid glands to make lots of PTH in an effort to increase blood calcium levels. Over time, this may cause hypercalcemia. Chronic kidney failure is the most common cause of secondary hyperparathyroidism as the kidneys have lost the ability to retain enough calcium in the body.

Signs and Symptoms - "What symptoms will my child have?"

Hypoparathyroidism: In mild cases, there may be no symptoms at all. When symptoms are seen, they are usually vague. Health care professionals call these symptoms "nonspecific". These include feeling tired, irritable, depressed, or anxious. When calcium levels drop very low, the patient may experience muscle or abdominal pain, tingling of the fingers, toes or face, numbness around the mouth, twitching of the face muscles, headaches, brittle nails, dry skin and hair, and uncontrolled spasms that cause muscle cramps. When calcium levels fall very fast or are extremely low, patients may experience seizures.

Hyperparathyroidism: Like hypoparathyroidism, mild cases of hyperparathyroidism may cause no symptoms. When symptoms are seen, they are vague, and many are very similar to having low calcium levels. In mild cases these include muscle weakness, feeling tired, weak, depressed, or muscle pain. In severe cases the patient may experience back or flank pain from kidney stones, belly pain, troubles concentrating, personality changes, memory problems, constipation, or broken bones.

Diagnosis - "What tests are done to find out what my child has?"

Blood Test: Doctors usually diagnose a parathyroid problem after finding an abnormal level of calcium on a blood test when a child has some of the symptoms noted previously. To narrow down why there is too much calcium in the blood, your doctor may also obtain other blood tests to look at the level of other elements in your body such as phosphorous, alkaline phosphate and the PTH level.

Family History: It is especially important to tell the doctor about other family members that may have had similar calcium or parathyroid problems in the past. If the child was healthy

before the onset of symptoms, then high calcium is almost always due to primary hyperparathyroidism.

The doctor may also order a test of the urine to see if the high blood calcium levels are due to a kidney problem.

X-ray called "bone densitometry" is more commonly done in adults to see if the parathyroid problem has caused damage to the bones. This test is not performed commonly in children because a child's bones are still growing and adapting, and the results of that X-ray test are unreliable in kids.

The most common reason why the parathyroid gland works too hard is overgrowth of a single parathyroid gland called an "adenoma". An adenoma is a benign (non-cancerous) growth that only causes a problem because it makes too much parathyroid hormone. Less commonly, one or more glands just get too large, which is called parathyroid hyperplasia. To see if there is evidence of an adenoma, the doctor may order a Sestamibi scan. Sestamibi is a very safe liquid radioactive compound that is absorbed by the overactive parathyroid but not by the healthy ones. The compound is injected through a small IV and then a special X-ray machine called a gamma camera is used to try and identify the abnormal parathyroid gland(s). This test is performed to locate which gland or glands may be abnormal to plan for treatment.

Treatment - "What will be done to make my child better?"

Patients with low blood calcium (hypoparathyroidism) are treated with medicine to replace calcium to keep the blood levels normal. These medicines include calcium and vitamin D (Calcitrol®), both of which can be given by mouth to maintain a normal level of calcium circulating in the body. Currently, giving a patient a drug that mimics PTH is not practical or effective. There is no surgical treatment for hypocalcemia.

Hyperparathyroidism

- **Medical Management:** Although researchers continue to try and find medicines for the treatment of primary hyperparathyroidism, there are no medicines currently available that can block the overproduction of PTH. In patients with secondary hyperparathyroidism that occurs with chronic kidney problems, medicine is available to try and treat the problem, and surgery is typically used as a last resort. For the rare patient who has severe hypercalcemia that has resulted in a seizure or other life-threatening problems, immediate admission to the hospital for intensive therapy with IV fluids and other medicines to bring the calcium level down to a lower level is required prior to considering any type of surgery.
- **Surgical Treatment:** The standard and most effective treatment for primary hyperparathyroidism is to remove the parathyroid tissue that is overproducing PTH—

typically a single adenoma. The Sestamibi scan helps the surgeon to determine which gland(s) is abnormal and allows him or her to focus the operation on removal of the overactive gland(s). This operation is done under general anesthesia though a small incision (1-2 inches) in the neck. The surgeon identifies the abnormal gland and removes it. Many surgeons confirm that the gland in question is the only problem gland prior to closing the incision by using a blood test, while the patient is still under anesthesia, to make sure the PTH level has dropped to a more normal level after the gland has been removed. Since PTH doesn't last for very long in the body, once the surgeon takes the gland out, the PTH level should drop to a normal level within 20-30 minutes.

Risks: from parathyroid surgery include temporary low blood calcium levels while the other normal parathyroid glands left behind regain their ability to make PTH. In modern surgery where the surgeon removes the adenoma and doesn't "explore" the other glands, this may last a few days, and is generally treated with oral calcium supplements until it gets better. Risk of damage to a nerve that is close to the parathyroid glands (recurrent laryngeal nerve) is low (1%). If the nerve is injured, it typically regains function, but if the nerve is permanently damaged it can cause permanent hoarseness. This is very rare in the hands of a surgeon with considerable experience in parathyroid surgery.

Benefits: Removing the overactive gland resolves the symptoms of high calcium. Bones are stronger. The results of surgery are generally excellent, with more than 99% of patients being "cured" of the disease, and in the vast majority of cases there is little risk of the high calcium levels returning because of disease of another parathyroid.

Informed consent: A consent form is a legal document that states the tests, treatments or procedures that your child may need and the doctor or practitioner that will perform them. Before surgery, your doctor should tell you what the operation is, the goal of the surgery and other possible treatment options that are available. Your doctor should explain the risks and benefits of the surgery. You give your permission when you sign the consent form. You can have someone sign this form for you if you are not able to sign it. You have the right to understand your child's medical care in words you know. Before you sign the consent form, make sure all of your questions are answered. It is important to know that during surgery, there are things that can happen that your doctor may have not predicted before going in. He or she will explain these to you after the surgery.

Postoperative care: The operation may require an overnight stay just to ensure that the patient doesn't exhibit any symptoms that would require more aggressive therapy for the temporary low calcium levels.

Home Care - "What do I need to do once my child goes home?"

Diet: Usually the child can be on a regular diet appropriate for age when they go home.

Activity: Regular activity can resume slowly a few days after surgery.

Wound care: Specific wound care issues should be addressed with your child's surgeon. Usually, wounds are kept dry for about three days, then the child may shower. Soaking the wound (such as baths or swimming) should wait until a week after surgery.

Medicines

- Medicines for pain such as acetaminophen (Tylenol®) or ibuprofen (Motrin® or Advil®) or something stronger like a narcotic may be needed to help with pain for a few days after surgery. Stool softeners and laxatives are needed to help regular stooling after surgery, especially if narcotics are still needed for pain.
- If your child has persistent low calcium levels, he or she will need calcium supplements.

What to call the doctor for: Call to the surgeon if there is worry about infection (unexplained fevers, redness and drainage of the wound). If your child experiences numbness and/or tingling around the fingers and the face, you should call the surgeon or endocrinologist as well.

Follow-up care: A wound check is often performed 2-3 weeks after surgery. Often, the child's endocrinologist can also provide follow up of the wound as well.

Long Term Outcomes - "Are there future conditions to worry about?"

Future outcomes for hypercalcemia secondary to hyperparathyroidism is excellent.

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