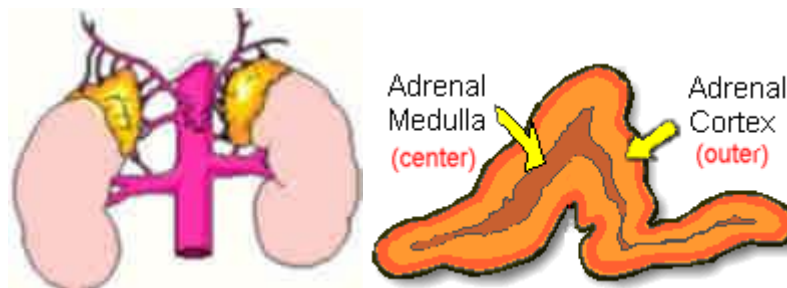


Your Adrenal Glands

► **The adrenal glands are orange-colored endocrine glands which are located on the top of both kidneys.** The adrenal glands are triangular shaped and measure about one-half inch in height and 3 inches in length. Each gland consists of a **medulla** (the center of the gland) which is surrounded by the **cortex**. The medulla is responsible for producing epinephrine and norepinephrine (adrenaline). The adrenal cortex produces other hormones necessary for fluid and electrolyte (salt) balance in the body such as cortisone and aldosterone. The adrenal cortex also makes sex hormones but this only becomes important if overproduction is present.



Indications for Surgical Removal of Adrenal Glands

✓ **The indications for surgical removal of the adrenal gland are relatively straightforward and include, but are not limited to the following:**

- **Tumors of the adrenal cortex which overproduce hormones.** One such tumor is shown in the picture...it is the round yellow mass located within the triangular adrenal gland. This picture is blown up about 2-fold to make it easier to see. The normal adrenal gland is NOT this big. Endocrine glands are very important despite their small size since they make important hormones.
- **Tumors of the adrenal medulla which overproduce adrenaline (pheochromocytoma).**
- **Most solid tumors of the adrenal gland which are greater than 4 cm (~2 inches) regardless of whether they produce hormone or not (to rule out malignancy).**
- **Primary cancers of the adrenal.**



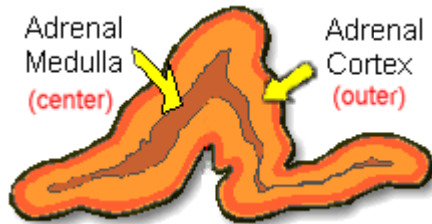
- **Any size adrenal mass which causes symptoms** such as flank pain or tenderness.

Note: the above conditions which indicate the surgical removal of an adrenal gland all exclude the situation in which cancer has spread from another organ to the adrenal (metastatic spread)...with *very few* exceptions.

Diseases of the Adrenal Cortex

~ Cushing's Syndrome ~

Note: this page contains information which assumes you have read our [introduction to the adrenal gland page](#). If not, go there first. Remember, the adrenal gland has a center gland (medulla) which produces [adrenaline](#), and an outer gland (the cortex) which produces several hormones such as cortisol and aldosterone. **This page is about cortisol overproduction by the adrenal cortex.**



✓ In 1932, a physician by the name of Harvey Cushing described eight patients with **central body obesity, glucose intolerance, hypertension, excess hair growth, osteoporosis, kidney stones, menstrual irregularity, and emotional lability**. It is now known that these symptoms are the result of excess production of cortisol by the adrenal glands. Cortisol is a powerful steroid hormone, and excess cortisol has detrimental effects on many cells throughout the body. Although some of these symptoms are common by themselves, the combination of these suggests that a workup for this disease may be in order. Keep in mind that Cushing's syndrome is rare, occurring in only about 10 patients per one million population. On the other hand, simple obesity can be associated with some of these symptoms in the absence of an adrenal tumor--this is related to the slightly different mechanism by which normally produced steroids are metabolized by individuals who are obese. *Note: The most common cause of excess steroids in the blood and its side effects, however, is long-term use of steroid medications for other disorders.*

▶ **Since cortisol production by the adrenal glands is normally under the control of the pituitary** (like the thyroid gland), **overproduction can be caused by a tumor in the pituitary or within the adrenal glands themselves**. When a pituitary tumor secretes too much ACTH (Adrenal Cortical Tropic Hormone), it simply causes the otherwise normal adrenal glands to produce too much cortisol. This type of Cushing's syndrome is termed "Cushing's Disease" and it is diagnosed like other endocrine disorders by measuring the appropriateness of hormone production. In this case, serum cortisol will be elevated, and, serum ACTH will be elevated at the same time.

▶ When the adrenal glands develop a tumor, like any other endocrine gland, they usually produce excess amounts of the hormone normally produced by these cells. If the adrenal

tumor is composed of cortisol producing cells, excess cortisol will be produced which can be measured in the blood. Under these conditions, the normal pituitary will sense the excess cortisol and will stop making ACTH in an attempt to slow the adrenal down. In this manner, physicians can readily distinguish whether excess cortisol is the result of a pituitary tumor, or an adrenal tumor.

▶ Even more rare (but placed here for completion sake) is when excess ACTH is produced somewhere other than the pituitary. This is extremely uncommon, but certain lung cancers can make ACTH (we don't know why) and the patients develop Cushing's Syndrome in the same way they do as if the ACTH was coming from the pituitary.

Causes of Cushing's Syndrome

ACTH Dependent (80%)

- Pituitary Tumors (60%)
- Lung Cancers (5%)

ACTH Independent (20%)

- Benign Adrenal Tumors (adenoma) (25%)
- Malignant Adrenal Tumors (adrenal cell carcinoma) (10%) [[new page on this topic](#)]

Testing for Cushing's Syndrome

▶ The most sensitive test to check for the possibility of this disease is to measure the amount of cortisol excreted in the urine during a 24 hour time period. Cortisol is normally secreted in different amounts during the day and night, so this test usually will be repeated once or twice to eliminate the variability which is normally seen. This normal variability is why simply checking the amount of cortisol in the blood is not a very reliable test. A 24 hour free cortisol level greater than 100 ug is diagnostic of Cushing's syndrome. The second test which helps confirm this diagnosis is the suppression test which measures the cortisol secretion following the administration of a powerful synthetic steroid which will shut down steroid production in everybody with a normal adrenal gland. Subsequent tests will distinguish whether the disease is due to an ACTH dependent or independent cause.

▶ Invariably, once the diagnosis is made, patients will undergo a CT scan (or possibly an MRI or Ultrasound) of the adrenal glands to look for tumors in one or both of them ([more information on adrenal x-ray tests on another page](#)). If the laboratory test suggest a pituitary origin, a CT or MRI of the brain (and possibly of the chest as well) will be performed.



Treatment of Cushing's Syndrome

► Obviously, the treatment of this disease depends upon the cause. Pituitary tumors are usually removed surgically and often treated with radiation therapy. Neurosurgeons and some ENT surgeons specialize in these tumors. If the cause is determined to be within a single adrenal gland, this is treated by surgical removal. If the tumor has characteristics of cancer on any of the x-ray tests, then a larger, conventional operation is in order. If a single adrenal gland possesses a small, well defined tumor, it can usually be removed by the new technique of laparoscopic adrenalectomy.

Diseases of the Adrenal Cortex

~ ADRENAL CANCER ~

✓ **Adrenocortical carcinoma is a rare tumor** afflicting only one or two persons per one million population. It usually occurs in adults, and the median age at diagnosis is 44 years. Although potentially curable at early stages, only 30% of these malignancies are confined to the adrenal gland at the time of diagnosis. Because these tumors tend to be found years after they began growing, they have the opportunity to invade nearby organs, spread to distant organs (metastasize) and cause numerous changes in the body because of the excess hormones they produce.

Characteristics of Adrenal Cortical Cancer

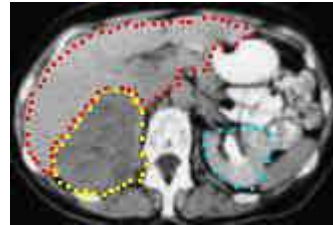
- Typically an aggressive cancer.
- Most (~60%) are found because excess hormone production causes symptoms which prompt patients to seek medical attention.
- Most (60-80%) actually secrete high amounts of one or more adrenal hormones.
- Many will present with pain in the abdomen and flank (nearly all that don't present with symptoms of hormone excess will seek medical attention because of pain).
- Spread to distant organs (metastasis) occurs most commonly to the abdominal cavity, lungs, liver, and bone.

Evaluation of a Suspected Adrenal Cortical Cancer

- **The Initial evaluation should include blood tests to measure the amount of adrenal hormones in the circulation.** Since the vast majority of these cancers make too much hormone (cortisol, testosterone, estrogen, aldosterone, etc) this is an obvious place to start. Keep in mind, however, that most non-cancerous tumors of the adrenal glands (benign adenomas and hyperplasia) will also secrete too much hormones. Therefore, demonstrating overproduction of adrenal hormones helps establish the presence of an adrenal tumor, yet it does not always help distinguish between benign and malignant (cancerous) tumors. Extremely high

levels, however, are more commonly produced by malignant tumors.

- **X-ray tests play a central role in the diagnosis of adrenal cancers**, and undoubtedly will play a central role in determining the type of treatment planned. Computed tomography (CAT scans) and/or magnetic resonance imaging (MRI) are the two central tests in this regard. They give overlapping information, so that all persons do not need both tests, but occasionally the situation will dictate that both are obtained. The CAT scan on the right shows a large right adrenal mass outlined in **yellow** under the normal sized liver (in **red**). This tumor produced large amounts of estrogen and caused the patient (a 66 year old woman) to seek medical attention after she began to have menstrual bleeding 20 years after her menopause.
- **Some adrenal tumors will require special studies of their blood supply** to help define the extent of the tumor, whether it is impinging on the blood supply to other nearby organs, and to help the surgeon decide on which operative approach to use. These tests are referred to as selective angiography and adrenal venography. They also may be helpful in distinguishing tumors of the adrenal gland from tumors of the upper pole of the kidney.



Adrenal Syndromes Caused by Excess Hormone Secretion

✓As noted in our example above, **many patients will seek medical attention with some sort of bodily change which typically comes on quite slowly** (usually over one to three years). When excess female hormones are produced in a female it can be hard to detect, except at extremes of age such as early puberty in a child, or the return of vaginal bleeding in a post-menopausal woman. The same is true for excess testosterone in a male. The opposite, however, will often make the presentation easier such as when a woman begins to develop male characteristics (deeper voice, excess body hair) or when a man begins to develop enlarged breasts. Some of these hormone overproduction problems have specific names and are listed below.

- **hypercortisolism** ([Cushing's syndrome](#)) (excess cortisol produced)
- **adrenogenital syndrome** (excess sex steroids produced)
- **virilization** (acquisition of male traits in a female because of excess testosterone production)
- **feminization** (acquisition of female traits in a male because of excess estrogen production)
- **precocious puberty** (puberty occurring too early because of excess sex steroids produced)
- **hyperaldosteronism** (Conn's syndrome) (excess aldosterone leading to hypertension and low potassium)