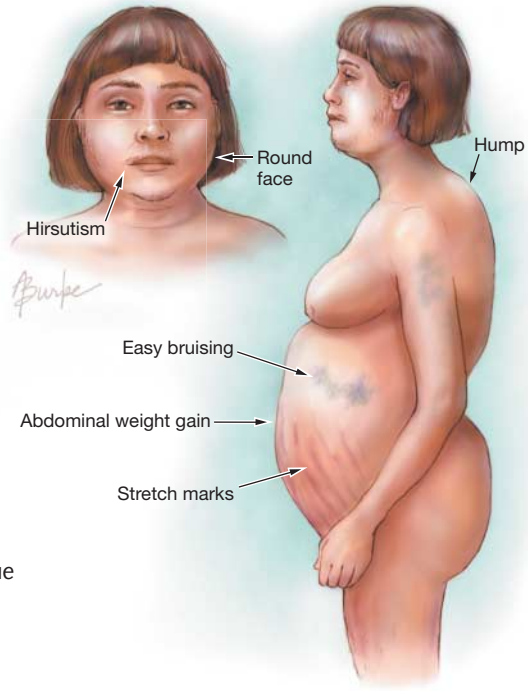
Cushing Syndrome and Cushing Disease

Cortisol is a **hormone**, a chemical that regulates body function. It is produced by the **adrenal** glands, which are located above each kidney. Cortisol helps regulate blood sugar levels and the responses to infection and stress. Cortisol production is increased by **adrenocorticotropic hormone (ACTH)** released from the **pituitary gland** in the brain. **Hypercortisolemia** (cortisol overproduction) causes **Cushing syndrome**, most often when a pituitary **adenoma** (a benign tumor) produces an excess of ACTH, resulting in **Cushing disease**. It also results when an adenoma of the adrenal gland produces too much cortisol. Both Cushing syndrome and Cushing disease affect women more often than men. Rarely, hypercortisolemia is produced by a tumor outside the pituitary or adrenal glands; for example, by a lung cancer that releases ACTH. This is called **ectopic** Cushing syndrome. Cushing syndrome can develop as part of genetic diseases that run in families (such as **multiple endocrine neoplasia type 1**). Prolonged treatment with glucocorticoids, which are cortisol-like drugs, can result in **iatrogenic** Cushing syndrome.

SYMPTOMS

- Weight gain, typically with a round face and a hump on the upper back, but often normal arms and legs
- Stretch marks on thighs and abdomen
- Easy bruising
- **Hirsutism** in women (excessive hair on face, abdomen, and legs)
- Irregular menstrual periods in women; sexual difficulties in men
- Severe fatigue, weak muscles, and easy fractures of bones
- High blood pressure
- Diabetes
- Infections
- Anxiety, irritability, and depression
- Decreased ability to concentrate and reduced memory

Signs and symptoms of Cushing syndrome



DIAGNOSIS

The diagnosis of Cushing syndrome is suggested by characteristic body changes, hypertension, and diabetes. It is confirmed by a high cortisol level in urine or saliva or by blood or urine tests measuring the response to **dexamethasone**, a glucocorticoid. As the next step, the cause of high cortisol is determined by additional blood studies and imaging studies (MRI or CT) of the abdomen or pituitary gland.

TREATMENT

If a pituitary adenoma is the source of Cushing disease, it is removed through an incision inside a nostril or under the lip. This is done using **microneurosurgical** techniques with small instruments and a microscope or using a **fiberoptic** technique with a tiny tube with a camera. These techniques can remove pituitary adenomas smaller than half an inch in diameter. If Cushing disease cannot be controlled by surgery, radiation can be used or the adrenal glands may be removed through the side of the abdomen (**adrenalectomy**).

When an adrenal adenoma produces Cushing syndrome, it is removed by adrenalectomy. However, if surgery is not possible, medications are used to block cortisol production and control hypertension and diabetes. In extremely severe cases of Cushing syndrome, if no source is found, both adrenal glands may be removed and the patient placed on lifelong hormone medications.

FOR MORE INFORMATION

- National Institute of Diabetes and Digestive and Kidney Diseases
  www.endocrine.niddk.nih.gov/pubs/cushings/cushings.htm
- US National Library of Medicine, National Institutes of Health
  www.nlm.nih.gov/medlineplus/ency/article/000410.htm

Source: National Institute of Diabetes and Digestive and Kidney Diseases

INFORM YOURSELF

To find this and previous JAMA Patient Pages, go to the Patient Page link on *JAMA*'s Web site at www.jama.com. Many are available in English and Spanish. A Patient Page on hypothyroidism was published in the December 10, 2003, issue and one on hyperthyroidism was published in the July 6, 2005, issue.

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