

The BSPED is one of the affiliated speciality groups of the Royal College of Paediatrics and Child Health.

The society aims to improve the care of children and young people with endocrine disorders or diabetes mellitus, by bringing together professionals from a range of disciplines.

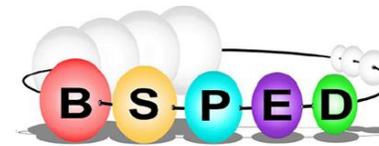
This leaflet has been written by members of the BSPED & reviewed by the Clinical Committee. It is designed to give you some general information about your child's condition and treatment. Your child's doctor or specialist nurse will be able to answer any further questions you have about your child.

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For more information, contact:

BSPED Office
Bioscientifica
Starling House
1600 Bristol Parkway North
Bristol
BS34 8YU
T: + 44 (0) 1454 642258
E: bsped@endocrinology.org



**British Society for
Paediatric Endocrinology
and Diabetes**

Cushing's Syndrome

Information for patients, parents and carers

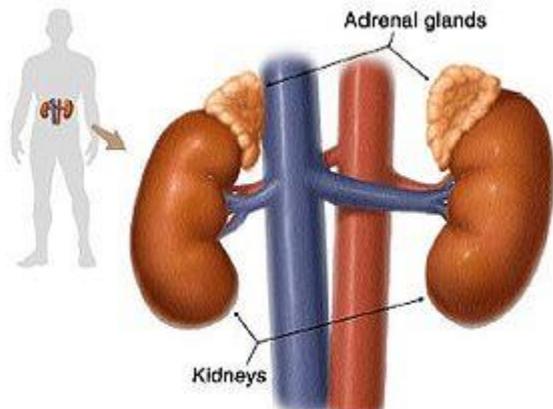


What is a hormone?

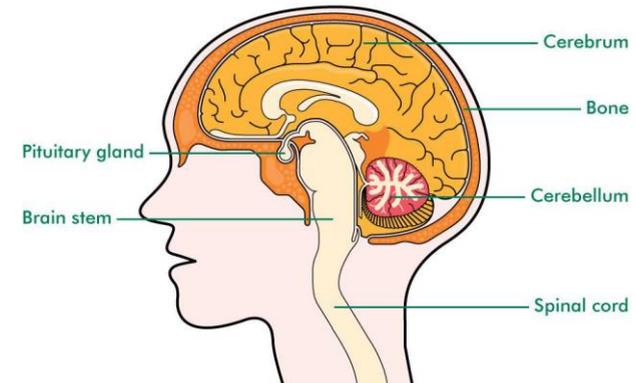
Hormones are chemical messengers. They are made in glands and travel round the body in the bloodstream. Hormones affect how other organs in the body work.

What is Cushing's syndrome?

Cushing's syndrome is a very rare condition (especially in children) that is caused when there is too much of a hormone called cortisol circulating around the body. Cortisol is made by the adrenal gland (small glands that are found on top of each kidney).



To produce cortisol, the adrenal gland is told to make cortisol by a hormone called adrenocorticotrophic hormone (ACTH) which is produced by the pituitary gland (a pea-sized gland found in the head



just below the front part of the brain). The ACTH is released by the pituitary gland into the blood stream, and then it travels down to the adrenal glands which make cortisol.

Why does Cushing's syndrome happen?

There are a number of reasons why a child may have high amounts of cortisol in their body:

1. The cortisol may be from a child's medicine; medicines containing cortisol are often known as steroids. Steroid medicines are used to treat many medical conditions such as asthma, inflammatory bowel disease and leukaemia; it is important to remember they are safe if used as prescribed by your doctor or health care professional. When steroid treatment is used for a long time and in high doses symptoms of Cushing's syndrome may occur.
2. The body is making too much cortisol. The most common reason for

this is a small growth (often called an adenoma or benign tumour) on the pituitary gland. This is a specific type of Cushing's syndrome called Cushing's disease. About 4 out of 5 children with Cushing's syndrome have Cushing's disease. The growth on the pituitary gland makes too much of the hormone ACTH which in turn stimulates the adrenal gland to make large amounts of cortisol.

3. Small growths may form in the adrenal glands that produce large amounts of cortisol.
4. The least common reason why a child may have Cushing's syndrome is if there is a small growth somewhere else in the body (for example the lungs), which makes large amounts of cortisol. This is called ectopic Cushing's syndrome

How does it affect a child?

Cortisol is essential for life – it is needed to maintain normal energy levels, blood pressure and the salt and sugar balance in the body. However when levels of cortisol in the body are too high this can cause changes in the body. In children and teenagers this can include:

- Weight gain (particularly around the middle of the body)
- Puffiness of the face
- Redness or flushing of the cheeks
- Poor growth
- Stretch marks
- High blood pressure

- Emotional changes (e.g. depression, anger, anxiety, tearfulness)
- Tiredness
- Disruption of pubertal development

How is it confirmed?

It can be very difficult to make a diagnosis of Cushing's syndrome. It can be months to years after the condition started to reach the diagnosis.

Cortisol levels change a lot throughout the day and night so a single blood or urine test is often not very helpful. To find out whether a child has Cushing's syndrome they will need blood tests at different times of the day and night, over several days. They will also need other specialist tests and urine collections. These tests should be carried out under the supervision of a specialist Children's Endocrine doctor.

When a child has been diagnosed with Cushing's syndrome, they will need more tests to find out where the extra cortisol is being made. These tests will include specialist endocrine blood tests and MRI scans.

How is it treated?

When the diagnosis has been made, the child may be given a medicine

that will lower the levels of cortisol. However this medicine is not a long-term solution but is often used to help with symptoms leading up to an operation.

The best way to treat Cushing's syndrome is to remove the source of the high cortisol levels. For example in Cushing's disease (i.e. a growth on the pituitary gland) an operation would be carried out by an experienced surgeon to remove the growth. On the other hand if one of the adrenal glands themselves were the problem, then an operation would need to be carried out to remove the growth from the gland.

In about one third of children the pituitary surgery for Cushing's disease, is not always completely successful due to the difficulty of the operation. Radiotherapy to the pituitary gland may then be needed.

Are there any long-term problems from the surgery?

If the surgery is to the pituitary gland because of Cushing's disease, then the other hormones produced by the pituitary gland may be affected; the leaflet Hypopituitarism (Multiple Pituitary Hormone Deficiency) has more details on this. The pituitary hormones affected vary from child to child, but hormone production recovers in many children in the months after surgery.

If Cushing's syndrome is being caused by a growth in the adrenal gland, the surgery can involve either removal of part or all of the adrenal gland.

If only part or all of one adrenal gland is removed then there is no need for long-term hormone treatment. However if the problem is with both adrenal glands and they both need to be removed, life-long replacement of the adrenal hormones would be required. There are two adrenal hormones that would need to be replaced:

- cortisol: which is replaced by a medicine called hydrocortisone and given 3-4 times each day
- aldosterone: which is replaced by a medicine called fludrocortisone and this is given once or twice a day

Suggested sites for further information

www.pituitary.org.uk

www.pituitary.org

<https://patient.info/health/the-pituitary-gland>

www.yourhormones.info