



British Society for Paediatric Endocrinology and Diabetes

HYPOPITUITARISM (underactivity of the pituitary gland)

The pituitary gland is a pea-sized endocrine gland (an "endocrine" gland is one that secretes "hormones", chemicals that circulate in the blood and affect other organs) situated in the head below the front part of the brain. It has important functions in the control of other endocrine glands; indeed it has been called "the conductor of the endocrine orchestra". The pituitary has two lobes, anterior and posterior, each of which secretes hormones. The posterior lobe is chiefly involved with the control of water balance through a hormone called ADH (antidiuretic hormone) and is only rarely involved in disease (see sheet on diabetes insipidus). The anterior lobe secretes several hormones of major importance including growth hormone (GH, which, as the name implies, is important in controlling growth and also has other beneficial effects on muscle and bone and is important for maintaining blood glucose levels), thyroid stimulating hormone (TSH, which drives the thyroid gland in the neck to produce thyroxine), adrenocorticotrophic hormone (ACTH, which drives the adrenal glands in the abdomen to produce steroids, especially cortisol), and, at puberty, gonadotrophins (FSH and LH, which control sexual development and function). It also secretes a hormone called prolactin which in women is implicated in lactation, but its role in childhood remains to be determined.

Causes

Underactivity of the pituitary gland may result from several different causes. There may be a "congenital" (present at birth) abnormality in the formation of the gland or a congenital tumour (a "craniopharyngioma") in the region above the gland. Various diseases may damage the gland or the function may be disturbed by a head injury or as a side effect of treatment with radiotherapy to the head which may be needed for the treatment of leukaemia and some related conditions such as brain tumours. Infection and inflammation such as Langerhans cell histiocytosis can also result in hypopituitarism. A number of genetic causes have now been established for hypopituitarism and these can lead to different hormonal deficiencies in children. Many of these genes are important for the way in which the pituitary gland develops in the embryo. Additionally, abnormalities of the midline of the brain and eye abnormalities resulting in blindness may be associated with hypopituitarism, a condition called septo-optic dysplasia. This is because development of the eye, brain and pituitary are closely related and abnormality of one of these can impact on the other structures as well.

Diagnosis

Diagnosis depends on the measurement of pituitary hormones in the blood either in the resting state or after stimulation tests to show more clearly the capacity of the gland. Scanning of the head, usually with an MR (magnetic resonance) scan can show the structure of the gland well. An assessment of the eyes may be indicated.

Treatment

Treatment consists of replacement either of the missing pituitary hormones themselves or of the hormones produced by the target glands. The most important of these hormones are hydrocortisone which replaces cortisol produced from the adrenal glands, which is important in maintaining the blood sugar and protecting against stress and illness such as infection, and thyroxine, from the thyroid gland, which is involved in maintaining the rate of many biochemical processes. Growth hormone may have to be replaced in children before and during puberty to achieve adequate



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growth. Sex hormones, testosterone in boys and oestrogen in girls, may be needed to induce puberty in hypopituitary children and to maintain sexual development in adults. However the pattern of hormone deficiencies is very variable; in some children only GH needs to be replaced whereas in others all the hormones need to be replaced. Puberty is variable: it may occur early in patients with SOD or children who have been treated with radiotherapy for tumours, or it may be normal or absent. Occasionally, children may have problems with fluid balance resulting in diuresis due to absence of vasopressin, a hormone that is required for water reabsorption in the kidney (see note on Diabetes Insipidus). However for vasopressin to act effectively, normal ACTH and cortisol secretion is required. The diabetes insipidus is often only apparent if cortisol concentrations are normal either spontaneously or with hydrocortisone treatment. In patients in whom cortisol deficiency co-exists with diabetes insipidus, it is extremely important that hydrocortisone doses are rapidly doubled at times of illness and that DDAVP is stopped – if not then water will not be adequately excreted in the presence of sub-optimal cortisol concentrations, and hyponatraemia will be exacerbated by the continuing administration of DDAVP.

Once growth is complete, adolescents are usually re-tested to confirm GH deficiency – if this is proven, then GH may be used at lower doses for its beneficial effects on body composition and bone mineral density. Additionally, prednisolone or dexamethasone, both longer-acting glucocorticoids, may be used to substitute hydrocortisone.

Treatment Plan

.....'s current treatment regimen is as follows: Hydrocortisonemg (milligrams) each morning,mg at lunchtime, andmg each evening (this dose has to be increased to cover physical stress, such as an illness or injury, see the sheet on steroid replacement)

Thyroxinemcg (micrograms) in the morning.

Growth hormone.....units.....by subcutaneous injection each evening Testosterone
.....

or

Oestrogen

Outlook

With replacement of the missing hormones, people with hypopituitarism can live a full life with normal health and strength. To achieve fertility, special hormone treatment is usually needed. Learning difficulties may be associated with the hypopituitarism.