



Multiple Pituitary Hormone Deficiency (MPHD)



SERIES 1



SERIES 2



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SERIES 4



SERIES 5



SERIES 6



SERIES 7



SERIES 8



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GROWTH AND GROWTH DISORDERS – SERIES NO: 11

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CGF INFORMATION BOOKLETS

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No. Title

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10. Constitutional Delay of Growth & Puberty
11. Multiple Pituitary Hormone Deficiency
12. Diabetes Insipidus
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INTRODUCTION

The aim of this booklet is to tie together many aspects of pituitary hormone insufficiencies and their management. There are other booklets in the series which give more information about particular subjects such as growth hormone insufficiency, delayed or absent puberty etc, and your specialist will give these to you if they may be helpful.

HORMONES

Hormones are the chemicals that carry messages from one cell to another via the blood stream. Part of the brain, called the hypothalamus, controls the levels of hormones in the blood by triggering the pituitary gland into producing the required hormones. Any low level, or insufficiency, of a hormone may be the result of problems in the hypothalamus, or with the link with the pituitary, or with the pituitary gland itself

An insufficiency of a hormone may occur alone or it may be associated with other pituitary hormone insufficiencies, as occurs in Multiple Pituitary Hormone Deficiencies (MPHD). Frequently, growth hormone, the gonadotrophins (the hormones which control sexual development) and thyroid stimulating hormone, or TSH, (the hormone that controls the thyroid gland) are affected. Much less frequently, adrenocorticotrophic hormone, or ACTE, (the hormone which stimulates the adrenal gland), is involved. All these hormones can be replaced by regular medication.

This multiple loss of hormones mainly affects the anterior (front) part of the pituitary. If there is loss of hormones from the posterior (back) part of the pituitary, such as antidiuretic hormone (ADH — also called vasopressin), it indicates a more serious problem which requires further investigation.

The study of the glands, and the hormones they produce and release, is called *endocrinology*.

WHERE ARE HORMONES PRODUCED IN THE BODY?

Hormones are produced in an area of the brain called the hypothalamus, as well as from glands including the pituitary gland, the gonads, the thyroid and the adrenals. *These glands, and the hormones they produce, are described overleaf and are shown in the diagram (Figure 1) on page 7.*

Hypothalamus

The hormones produced by the hypothalamus stimulate the pituitary gland and so control the release of the pituitary hormones. Some of these hormones are referred to as “releasing hormones” and include growth hormone releasing hormone (GHRH) and gonadotrophin releasing hormone (GnRH). The role of the hypothalamus is like that of the director – it has an overall controlling activity and its link with the pituitary gland is very important.

Pituitary gland

The pituitary is a pea sized gland which is found in the centre of the head at the base of the brain, just below the hypothalamus. The pituitary is divided into two areas (lobes) and these are called the anterior (front) and posterior (rear) lobes. The hormones released from the anterior part of the pituitary are:

- growth hormone (GH),
- the gonadotrophins (luteinising hormone (LH) and follicle stimulating hormone (FSH))
- thyroid stimulating hormone (TSH) and
- adrenocorticotrophic hormone (ACTH).

The hormones released from the posterior lobe are antidiuretic hormone (ADH; also called vasopressin) and oxytocin.

The pituitary hormones in turn stimulate the other glands to release their hormones.

The gonads (ovaries and testes)

The **ovaries** are the female reproductive organs that produce eggs and sex steroid hormones (oestrogen and progestogen) in a regular cycle in response to gonadotrophin release from the pituitary gland. There are two ovaries, each the size of a large olive, situated in the lower abdomen, either side of the womb (uterus). Each ovary contains follicles (small secretory glands), within which the eggs develop. The follicles secrete oestrogen and a small quantity of androgens. After an egg has been released from a follicle, the site which remains secretes progesterone.

Oestrogen stimulates the development of female sexual features including breast development and the androgen (in combination with androgens from the adrenal gland) stimulates the development of pubic and underarm (axillary) hair. Oestrogen also builds up the lining of the uterus while progesterone stimulates the shedding which is the monthly bleed of the menstrual cycle. This is important for keeping the uterus healthy.

The **testes** are the male sex organs that produce sperm and secrete the sex steroid hormone testosterone (an androgen) in response to gonadotrophins released from the pituitary gland. Before a boy is born, the testes develop within the abdomen but then descend into the scrotum (the sac that holds the testes). Testosterone stimulates male sexual development including facial and body hair, deepening of the voice and muscle development.

Thyroid

The thyroid gland is found at the front of the neck. It has two lobes which are connected. This gland is involved in the regulation of metabolic rate through the secretion of thyroid hormone (thyroxine) in response to thyroid stimulating hormone (TSH or thyrotrophin) released from the pituitary gland. *Metabolism* can be described as the balance of all the chemical and physical changes that take place in the body which enable it to continue growing and working. This includes the breakdown of food to release energy as well as the development of tissues in the body such as muscle and bone. *Metabolic rate* is the speed at which the changes happen. This varies from person to person and is affected by factors including age, size, diet and exercise, as well as by various hormone levels.

Adrenals

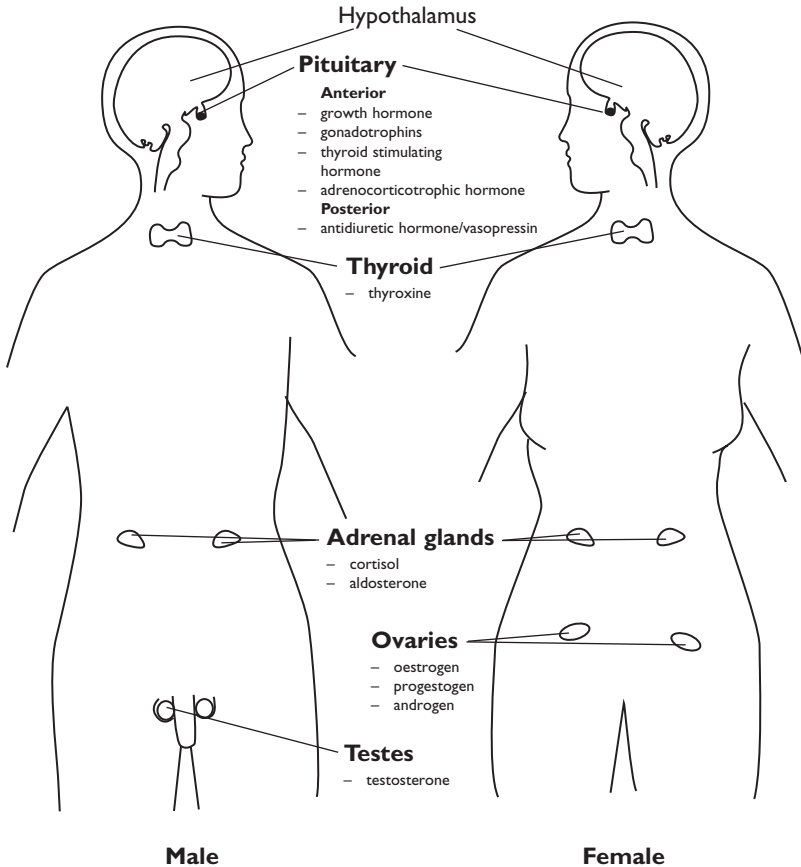
The adrenal glands are two triangular glands, each about the size of a broad bean, which lie close to the kidneys. The adrenal glands have two parts the medulla (the inside part) and the cortex (the outer part). It is the cortex which is stimulated by adrenocorticotrophic hormone (ACTH) from the pituitary to release cortisol and aldosterone. Cortisol is important for the metabolism of sugar in the body and so the control of level of sugar in the blood. It is also very important for the maintenance of blood pressure and the body's response to "stress". Aldosterone controls salt concentrations in the body. However, aldosterone secretion is not usually affected in MPHD.

CAUSES OF PITUITARY HORMONE INSUFFICIENCY

In many cases of pituitary hormone insufficiency there is no clear cause and the term that is used to describe this is **idiopathic**. In other cases the cause is known and the hormone insufficiency is then described as **secondary**. Secondary causes include — an abnormality in the brain; cranial irradiation given as treatment for different types of cancer; damage caused by a cyst, a tumour, and/or surgery in the same area of the brain as the hypothalamus or the pituitary.

Although pituitary hormone insufficiency may at first only affect the secretion of one hormone, the most commonly affected being growth hormone, it may develop into a insufficiency of two or more hormones. *The pattern of the developing loss of hormones*

Figure 1
The endocrine glands and their hormones



may be referred to as an *evolving endocrinopathy*. In patients with growth hormone insufficiency (idiopathic or secondary), further hormone insufficiencies are not always seen and the growth hormone insufficiency therefore remains “isolated”. However, if a child has been treated with high doses of irradiation to the head the pituitary gland will be damaged. This will almost always result in an evolving loss of all the pituitary hormones over a period of time.

When a single pituitary hormone is insufficient, this is described as “isolated”, eg isolated growth hormone insufficiency. When more than one hormone is insufficient, this is called multiple pituitary hormone deficiencies (MPHD) or panhypopituitarism.

The timing of when these further hormone insufficiencies occur varies between individuals and is often related to the original problem. Some pituitary tumours may cause a rapid loss of all the hormones, whereas following some irradiation treatments it may take ten years or more for all the insufficiencies to appear.

The development of this evolving loss of hormones always occurs according to a fixed pattern. The first hormone to become insufficient is growth hormone followed by, in this order:

- the gonadotrophins (LU and FSH)
- thyroid stimulating hormone (TSH)
- adrenocorticotrophic hormone (ACTH)

Even though the order in which these hormones become insufficient always stays the same, the time period over which it happens is very variable. This evolving loss occurs to the hormones of the anterior lobe of the pituitary.

HORMONE REPLACEMENT TREATMENT

All the hormones which are produced by the anterior lobe of the pituitary gland are released into the bloodstream in bursts (pulsatile). In addition, there is a characteristic daily (diurnal) pattern of release. For example, growth hormone is usually released in bigger bursts at night-time with smaller bursts being released during the day. Although growth hormone is usually released in bursts every 3 hours, it is not necessary to copy this pattern when growth hormone is being given as a treatment. Thus, having one injection every evening, just before bedtime, seems to be enough to produce the required response in terms of growth.

However, with other hormones, such as those released by the hypothalamus (eg gonadotrophin releasing hormone) it is necessary to give the treatment in a pulsatile way so as to mimic the normal release of the hormone. This is needed in order to stimulate the pituitary to release the gonadotrophins (LH & FSH).

The pattern of release of cortisol from the adrenal glands means that more is released in the morning and low levels occur in the evening. Thus, when replacement treatment is being given, most of the daily dose is given first thing in the morning (before getting out of bed) with a smaller dose being given in the evening. This, however, can vary between individuals and may be changed in discussion with your specialist.

All hormone replacement treatments are usually given as a standard dose related to the size of the patient (child, adolescent or adult). The dose may be altered every 6 months depending on any change in growth and/or body size. This is especially the case for

growth hormone and for thyroxine. Too big a dose of thyroxine may, over time, increase the rate of growth of a child and this is why it is very important that the correct dose is given. Sometimes, depending on how well a child is growing, it is necessary to do some blood tests to check the levels of hormones and to make sure that the replacement dose that is being given is correct.

GROWTH HORMONE INSUFFICIENCY

One of the main functions of the hormone called growth hormone is to stimulate growth and this is why it has this name. However, in adult life, as well as in childhood, growth hormone has other equally important functions which are not directly related to growth such as the control of the level of sugar in the blood. An alternative name for growth hormone is somatotropin.

Growth hormone is the hormone which most often becomes insufficient when there is any type of pituitary insufficiency (idiopathic or secondary causes). Idiopathic growth hormone insufficiency occurs in approximately one in five thousand children and affects boys more than girls. Further information about growth hormone insufficiency is available in two other booklets, No.2 (Growth Hormone Deficiency) and No.7 (Adult GHID).

Approximately 50% of children with growth hormone insufficiency have isolated GH insufficiency, ie no further hormone insufficiencies occur. Growth failure in a child born with growth hormone insufficiency usually becomes obvious after the age of one to 1½ years. Before this time, ie from birth to 18 months, growth is more dependent on nutrition than on growth hormone secretion.

However, because of some of the metabolic actions of growth hormone, a baby with growth hormone insufficiency may show signs of low levels of sugar in the blood (hypoglycaemia) which will require growth hormone treatment. This metabolic use is independent of the use of growth hormone to stimulate growth. Despite growth hormone treatment, episodes of hypoglycaemia may well occur during infections. However, after the age of three to four years, this problem usually goes away.

If the hypoglycaemia continues, even when the child is having growth hormone treatment, this may indicate that there is also an insufficiency of another hormone, ACTH. As a result of the ACTH insufficiency, the adrenal gland is not stimulated and so there is insufficient cortisol being released to control the level of sugar in the blood. When an individual has hypoglycaemia associated with GH insufficiency, it is very important that they do not miss any of their growth hormone injections. Advice about hypoglycaemia and its treatment, especially in emergencies, is given in booklet No.5.

The only way to give growth hormone treatment is by injection. Because growth hormone is a protein, if it was taken by mouth it would be digested. These injections are given each evening and the needle goes just under the skin (subcutaneous). The dose of growth hormone needed is calculated according to the size (either body weight or body surface area) of the individual. However, even when the dose increases, the volume of the injection remains very small.

In children, the response to treatment is assessed by monitoring the rate of growth. In adolescents and young adults, where growth hormone may be appropriate even though growth has stopped, other methods may be used to assess response to treatment and these could include measures of muscle mass, bone density and overall well-being.

GONADOTROPHIN AND SEX STEROID INSUFFICIENCY

Gonadotrophins are released from the pituitary gland following stimulation by gonadotrophin releasing hormone (GnRH) which is released from the hypothalamus. The gonadotrophins send messages to the gonads (ovaries in females, testes in males) to trigger them to release oestrogen (females) or testosterone (males). The gonadotrophins, therefore, are necessary for the physical signs of puberty to occur and for the development of fertility (the production of eggs in females and of sperm in males).

In individuals with gonadotrophin insufficiency, the cause is often an insufficiency of GnRH release from the hypothalamus. As a result, the pituitary gland is not stimulated to release the gonadotrophins. More details about these hormones are given in booklet No.3 (Puberty and the Growth Hormone Deficient Child).

In children with GnRH insufficiency which started before birth, the diagnosis can often be made from signs such as a very small penis (micropenis) and both testes having not descended (undescended testes). MPHJ should always be suspected in a boy if these signs are present, even more so if hypoglycaemia (low level of sugar in the blood) is also present. However, it is much more difficult to diagnose GnRH insufficiency in the middle childhood years. At this age (2-9 years) gonadotrophin levels are usually low, and so it is difficult to tell whether the levels are low but normal, or whether there is GnRH insufficiency.

About 50% of children who are thought to have isolated GH insufficiency also have gonadotrophin insufficiency. Often, however, the diagnosis cannot be confirmed until the pubertal years when it can be seen whether or not the child is showing the expected physical signs of puberty at the appropriate age (triggered by the actions of the gonadotrophins and the sex steroids).

Children with growth hormone insufficiency should have a hormone (endocrine) re-evaluation at around the age of 9 to 10 years to check whether GnRH insufficiency is present. In girls, a pelvic ultrasound examination will show the size, appearance and maturity of the ovaries which may be helpful in assessing the levels of circulating gonadotrophins (the gonadotrophins will stimulate the growth and maturity of the ovaries).

Delayed puberty is common in boys with GH insufficiency. It is therefore difficult to tell whether there is also gonadotrophin insufficiency, or whether the release of gonadotrophins from the pituitary is simply delayed. Even if there is doubt about the actual diagnosis, puberty should be started at the appropriate age (11 to 12 years). The investigation of gonadotrophin levels can be made during the late teenage years, after growth has stopped. It is, of course, important that the proper diagnosis of GnRH insufficiency is made as this will be needed in order to assess the opportunity for future fertility.

In boys and girls, the outward signs of GnRH insufficiency, and so gonadotrophin insufficiency, is a lack of pubertal development. This is treated by giving sex steroid treatment. In boys this will be as testosterone (injections or tablets) and in girls as oestrogen and progestogen (tablets or skin patches).

One of the main difficulties associated with testosterone injections is that they cause a very sudden, rapid rise in the level of testosterone in the blood. This is associated with increased libido (sex drive) and frequency of erections. These increases in testosterone are followed later by decreases to levels well below normal. Such swings in testosterone levels can be very difficult to cope with, both emotionally and sexually. If you are having real problems with this, do discuss it with your specialist as he/she may be able to modify the dose to help prevent this from happening. In the near future, skin patches containing testosterone (similar to patches of oestrogen, also called hormone replacement treatment (HRT), used in women) will become available. The main advantage of the patches should be that a more steady level of testosterone is released into the blood which ought to have considerable advantages for the patient.

In adulthood, when an individual wants to start a family, treatment to bring about fertility will be required. This is a more complex treatment which is achieved by injections of GnRH or gonadotrophins according to a fixed plan. Your specialist will be able to arrange a referral to a doctor who specialises in fertility treatment.

Treatment of Undescended Testes

When an unborn baby boy is growing in the womb, the testes start to develop at the back of the abdomen, close to the developing kidneys. While he is still in the womb, the testes descend down into the back of his abdomen, into his groin, and then into his scrotal sacs.

By the time the boy is 2 years of age, both testes should have descended into the scrotum. If one testes does not descend, the cause is usually a local problem and is not hormonal. It will therefore require an operation to bring the testis down and this operation is called an orchidopexy. If both testes do not descend then this *may* be due to gonadotrophin insufficiency. This will require hormone investigations. It may be that giving gonadotrophins for 3 to 6 weeks will trigger the testes to descend. If this does not work, surgery will be required.

It is very important that the testes are not left in the abdomen during adult life as this may increase the risk of the development of testicular cancer. In addition to the undescended testes, the boy may have a very small penis (micropenis).

Treatment of Micropenis

Once a boy has started school, he may be very worried if he has a very small penis (micropenis). It may therefore be helpful to give a short course of low dose testosterone treatment which will increase the size of his penis (both length and width). However, the best time to give the treatment may be before he gets to the age of starting school, probably when he is 2 to 3 years of age. Once the boy is older, treatment with testosterone to trigger his puberty will also produce an increase in the size of the penis.

TRH / TSH INSUFFICIENCY

An insufficiency of either thyroxine releasing hormone (TRH) from the hypothalamus, or thyroid stimulating hormone (TSH) from the pituitary, leads to the loss of stimulation of the thyroid gland. As a result there will be low levels of thyroid hormone (thyroxine) in the blood. If the thyroid gland is 'underactive', this will affect growth as well as slowing many other mental and metabolic functions in the body.

Replacement treatment for thyroid hormone insufficiency is as a daily tablet. The daily dose will be based on body size. To check whether the correct replacement dose is being given, occasional blood tests may be needed although in children. Some specialists just use growth rate and bone development as a way of assessing whether the dose is correct.

In children with isolated growth hormone insufficiency, starting growth hormone treatment may result in an apparent lowering of the thyroid hormone levels in the blood and this can temporarily confuse the assessment. The levels will usually return to normal within six months and long term thyroxine treatment may not be needed.

ACTH INSUFFICIENCY

Adrenocorticotrophic hormone stimulates the adrenal gland which produces a group of hormones including hydrocortisone. These hormones are very important for general well-being and for maintaining the correct blood pressure and sugar balance in the body. In stressful situations, such as following accidents, or during surgery or illness, they become even more important.

Hydrocortisone replacement treatment is usually given in the form of a tablet, normally three times, but sometimes twice, a day. The normal release of cortisol from the adrenal gland is higher in the morning, and so when replacement treatment is given, most of the dose (up to two thirds of the daily dose) may be given first thing in the morning. The remainder is given in the late afternoon or early evening. In children, a normal growth rate will show that the correct dose is being given. If too much is given, the growth rate will be affected and will slow down.

Occasionally, in some individuals, it may be necessary to perform blood tests to check that the dose being given is enough but not too much (this is done while continuing on the regular dose of hydrocortisone). Such tests are performed over 24 hours with blood samples being taken every 2 to 3 hours. For this test, an overnight stay in hospital is needed.

A few children gain weight very quickly when they are receiving cortisol treatment. If this does occur, it is very important to check the level of thyroid hormone, and of cortisol, in the blood (as described above). If the level of thyroid hormone is too low, an individual will gain weight and thus their dose of thyroid hormone replacement treatment may need to be increased. With cortisol, even if the level in the blood is correct, a child may still gain weight, although the reason for this is unknown. It appears that they may be very sensitive to levels of cortisol in their body. Such children may benefit from the advice of a dietician to help with their weight control. Your specialist should be able to advise you about this.

During minor illnesses it may be necessary to increase the dose of hydrocortisone to two or three times what is normally given. In addition, during severe stress or illness, especially when vomiting occurs, the hydrocortisone may need to be given by intramuscular injection rather than by tablets. As well as causing low blood pressure, cortisol insufficiency may result in low levels of sugar in the blood (hypoglycaemia). This is even more likely when growth hormone insufficiency is additionally present. More information about this, and the treatment of hypoglycaemia, is given in booklet No.5.

ANTIDIURETIC HORMONE INSUFFICIENCY

Antidiuretic hormone (ADH), which is also called vasopressin, is released from the posterior part of the pituitary gland. This hormone controls levels of fluid in the body and thus the concentration of the blood, making sure it is not too dilute or too concentrated. It does this by signalling to the kidneys to adjust the amount of water that is retained in the body or excreted in the urine. If the blood becomes too concentrated (too little water in the body) the “thirst centre” in the hypothalamus detects this and it stimulates the secretion of ADH from the posterior pituitary. This then acts on the kidneys causing them to prevent further loss of water in the urine. The urine therefore becomes more concentrated and the blood more dilute. Once the water concentration in the blood is back to normal, the pituitary stops releasing ADU.

ADH insufficiency causes a condition known as *diabetes insipidus* in which large volumes of dilute urine are passed. This happens because there is no signal to the kidneys to retain the water. Following the increased loss of fluid, the blood becomes more concentrated and the thirst centre in the hypothalamus detects this. As a result the sense of thirst is stimulated and the individual has to drink very large quantities of liquids to make up for the water lost in the urine.

Diabetes insipidus is usually caused by an abnormality at the base of the brain, by a cyst, or by a tumour. In rare cases, diabetes insipidus can be due to a genetic defect inherited from the parents. Whatever the suspected cause, any child with diabetes insipidus, especially when there are also insufficiencies of the anterior pituitary hormones, should have a specialised x-ray or magnetic scan of the brain to get a clear image of the pituitary gland.

Treatment for diabetes insipidus is given as an “analogue” of the natural ADH. An analogue is a product which has a slight change made to it. It has the same actions as the original hormone but lasts longer, therefore it often can be given less often. Also, it can often be given in a different way, eg by tablets or intranasal drops, rather than by injection. Different forms of ADH/vasopressin replacement therapy are available and your specialist will assess which one is most suitable for you or your child. Treatment may be given by intranasal drops or spray. The spray tends to be more suitable for adults or older children as the dose of each puff of spray is quite high. In younger children it is often easier to use the intranasal drops. Vasopressin tablets are also available and can replace the use of intranasal forms.

Over-treatment of diabetes insipidus, through giving too much vasopressin, may result in a build up of fluid in the body and this could result in convulsions (fits). For this reason it is very important not to exceed the dose of vasopressin recommended by your specialist. Under-treatment is less dangerous and just causes the individual to pass more urine and so become extremely thirsty.

Further details regarding the care, and treatment, of individuals with diabetes insipidus are given in Booklet No. 12.

DIAGNOSING AN EVOLVING PITUITARY HORMONE INSUFFICIENCY

Predicting which children with an initial diagnosis of isolated growth hormone insufficiency will develop further hormone insufficiencies can be difficult. Help may be obtained by doing a special x-ray (CT scan) or a magnetic (MRI) scan which will show the size and appearance of the pituitary gland and this may help in deciding the diagnosis. Some children will have additional signs or symptoms such as:

- a slowing down of their rate of growth due to thyroid hormone insufficiency
- hypoglycaemia due to the development of cortisol insufficiency
- a failure to go into puberty due to gonadotrophin insufficiency

When your specialist does the first tests at the hospital, these are to assess the pituitary gland in order to see how many of the hormones are insufficient. However, as already described, the loss of hormones can occur over a number of years and so the picture may change over the coming months and years. It may, therefore, be necessary to repeat these tests every few years.

It is particularly important not to miss the diagnosis of ACTH insufficiency because of the importance of cortisol during stress and illness. In addition, a check of hormone levels will be needed just before puberty, at approximately ten or eleven years of age, in order to see if additional treatment should be given. A further hormone check should be done at the end of growth hormone treatment, when growth has stopped, in order to confirm which hormones are insufficient before care is transferred to an adult endocrinologist.

SUMMARY

Not all the hormone insufficiencies described in this booklet will apply to you/your child. Those hormone insufficiencies that are appropriate for you will be discussed with you by your specialist. However, it is important that you understand that the loss of pituitary hormones can occur over time, and that the growth hormone insufficiency which is initially diagnosed may develop into multiple pituitary hormone insufficiencies.

ADDITIONAL INFORMATION

1. It is important that you/your child carries notification on them at all times that they have MPHD. This is particularly important for those children who have cortisol insufficiency in addition to GH insufficiency, as they commonly develop hypoglycaemia. One way is to wear an engraved bracelet or necklet and these can be purchased from the following companies (if you are on a low income, let them know):
 - MedicAlert Foundation – 12 Bridge Wharf
156 Caledonian Road
London N1 9UU
Tel: 0171 833 3034
 - SOS Talisman
2. During trips abroad it is wise to have a “To whom it may concern...” letter from your specialist giving details of any treatment requirements for you/your child. This may include the necessary drugs as well as explaining why you are carrying needles and syringes.
3. If you/your child has episodes of hypoglycaemia associated with MPHD, or has cortisol insufficiency, you should always have a vial of hydrocortisone in the fridge at home to be given in emergency situations. You should also know how to give it (by intramuscular injection). Booklet 5 will help with this but you should also check with your specialist.
4. There is no reason why you/your child should not be treated normally and undertake normal age-related activities.
5. Children and adults with MPHD may have learning or behavioural difficulties associated with their condition. This often depends on many factors including the underlying cause of the MPHD. This will therefore vary from individual to individual.

In addition, for children, there may be issues associated with bullying, delayed puberty, psychological well-being etc. Your specialist will be able to help with these matters. In

addition, some of these issues are covered in other booklets and the Child Growth Foundation will be able to give you further information.

QUESTIONS & ANSWERS

Q1: Are pituitary hormone insufficiencies reversible?

A: No, once they happen it is very rare that they can be reversed. However, they can be treated by giving hormone replacement. There is a rare environmental cause of growth hormone insufficiency, known as psychosocial dwarfism, which can be reversed when the environment (home or school conditions) is improved, or the child is taken out of the adverse environment.

Q2: Can hypothalamic and pituitary hormone insufficiencies develop in adult life?

A: Yes. Some hormone insufficiencies may not appear until adulthood and this is why it is important that care is transferred to an adult endocrinologist who will continue to care for adolescents and adults.

Q3: Does the rapid development of additional hormone insufficiencies suggest that there is a tumour or cyst in the brain?

A: No, not necessarily. An evolving insufficiency can happen in idiopathic (unknown cause) pituitary insufficiency as well as following secondary causes such as a tumour. Your specialist will discuss this with you as well as the need for any special x-rays or magnetic imaging of the pituitary gland.

Q4: Does diabetes insipidus (ADH insufficiency) result from an unknown cause?

A: Diabetes insipidus is a problem associated with the posterior part of the pituitary gland and the cause is usually known. Assessment of the cause of the problem will involve tests of hypothalamic and pituitary function, as well as special x-rays or magnetic imaging. This will enable your specialist to see if there is a cyst or a tumour present. The exception is when diabetes insipidus is due to a genetic defect inherited from the parents.

Q5: Is diabetes insipidus easily treated?

A: The answer, usually, is yes. However, if there has been a lot of damage to the hypothalamus or the pituitary, as can happen during surgery for a tumour such as a craniopharyngioma, there may be additional damage to the thirst centre. If this happens it will affect the individual's ability to feel thirsty. So, if a lot of water is lost through dilute urine, they lose the ability to feel very thirsty and so don't always drink enough fluid to make up for it. This can be very serious and your specialist will advise as to how this condition should be looked after.

Q6: Is it important for my child to have any special x-rays or magnetic scans of the pituitary glands?

A: For children with MPHD, it is usually recommended to have these tests because it is important to check whether there are any abnormalities such as a cyst or a tumour present. Your specialist will discuss this with you.

Q7: Will sexual development and function be normal in individuals with MPHD?

A: Yes, if gonadotrophin insufficiency is present the appropriate sex steroids can be given (oestrogen in girls, testosterone in boys) and this will trigger the development of normal sexual characteristics. Sex steroid treatment will be continued into adulthood to maintain the stage of sexual development as well as libido (sex drive).

Q8: Will my child be able to have children in the future if they have MPHD?

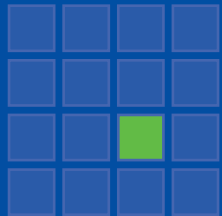
A: Usually yes, although this will depend on the underlying cause of their MPHD. Certainly, it will require some specialised hormone treatment when the time is right. Your specialist will discuss this with you/your child (at an appropriate age) and arrange handover to a specialist with expertise in fertility (reproductive endocrinology).

Q9: If my child has “isolated” growth hormone insufficiency, will he/she need to continue under the care of an endocrinologist during adult life?

A: Almost certainly, yes. It may be that some of the additional hormone insufficiencies do not appear until adult life, even though they have not occurred during childhood. Sometimes it takes many years for additional hormone insufficiencies to develop. As and when they do occur, it is important that you are under the care of an endocrinologist so that you receive the appropriate replacement therapies.



SERIES 11



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