



# Craniopharyngioma

## A Guide for Parents and Patients



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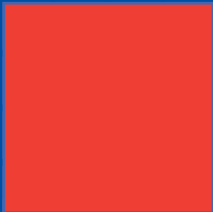
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## ***INTRODUCTION***

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The aim of this pamphlet is to provide general information about a type of brain tumour called a craniopharyngioma, how it is treated, and some of the problems that may be encountered both in the short and long term following treatment. It has been written in general terms and, therefore, not all of the information provided will apply to, or be relevant for, you or your child. How the condition is diagnosed, its treatment, and the outcome will vary from person to person but the information in this booklet will hopefully help you understand the condition better, and give you a basis for discussions with your GP and your specialists.

A craniopharyngioma is a type of brain tumour. It is a benign tumour, which means it is NOT cancerous and therefore will not spread to other parts of the body.

## ***WHAT IS A CRANIOPHARYNGIOMA?***

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It is a congenital tumour which means that it is present from birth. It is thought to form and grow from some misplaced cells collecting in an area of the brain close to the pituitary gland and its stalk (the pituitary stalk), early on in the development of a baby in the womb. There are no known reasons for this to happen, such as taking medications or being ill during pregnancy, and the condition is not hereditary (i.e. passed on from parents or grandparents).

Although present from birth, craniopharyngiomas vary in the timing of when the symptoms first appear. The symptoms indicate that there has been a change in the size of the craniopharyngioma and the effect it is having on other areas of the brain. The impact of a craniopharyngioma may happen at any age, from birth to old age, because some grow faster and cause symptoms much earlier, than others. In children, most are diagnosed between the ages of 5 and 10 years, the tumour being slightly more common in boys than girls. Even though craniopharyngiomas are fairly uncommon, in childhood they are the most common tumours in the area of the pituitary gland and represent about 9% of all childhood brain tumours.

The tumour itself is made up of solid parts, which usually contain pieces of calcium and cysts which are filled with a thick fluid. Occasionally, a craniopharyngioma may be entirely fluid-filled or, more rarely, entirely solid. These tumours are often very “sticky” and they adhere to the surrounding tissues which means it can be difficult to remove them surgically without damaging some of the tissues, particularly an area called the hypothalamus.

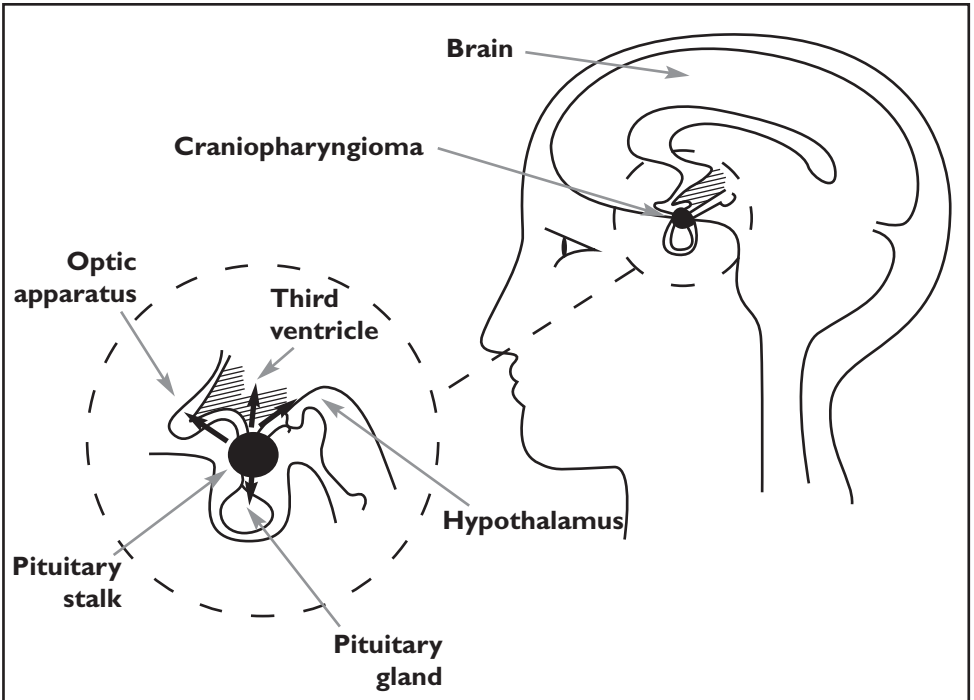
Even if it is removed surgically, a craniopharyngioma may re-grow at the place where it was first found and so will need further treatment.

A craniopharyngioma is a congenital tumour (congenital meaning that a person is born with it), and so if the symptoms appear in early childhood it means the tumour is growing faster than if the symptoms do not appear until adulthood. This means that in childhood it is a more serious condition and is more difficult to treat.

## ***HOW DOES A CRANIOPHARYNGIOMA FIRST APPEAR?***

As shown in the diagram below, many of the symptoms and potential problems of having a craniopharyngioma result from its position and size in relation to the surrounding parts of the brain. As the tumour grows, it starts to exert pressure against other delicate areas such as the pituitary gland, the hypothalamus, and the connections between them, producing effects which are described below.

The most commonly occurring symptoms are headaches and disturbed vision, both of which result from pressure as the craniopharyngioma grows and presses on other parts of the brain.



## ***Hypothalamus***

This is an extremely important area of the brain. It influences a number of essential behavioural and functional aspects of the body including temperature regulation, food intake, thirst (and, therefore, water intake), sleep-wake patterns, emotional behaviour and memory. The hypothalamus also serves as the main ‘communication centre’ for the pituitary gland by sending messages or signals to the pituitary in the form of hormones which travel via the bloodstream and nerves, down the pituitary stalk. These signals, in turn, control the production and release of further hormones from the pituitary gland which signal to other glands and organs in the body. The role of the hypothalamus is like that of a director – it has an overall controlling activity and its link with the pituitary gland is very important.

## ***Pituitary gland***

The pituitary gland is linked to the hypothalamus by the pituitary stalk. The pituitary releases hormones which trigger other glands in the body to release hormones. The pituitary gland is about the size of a pea and is divided into two lobes – these are called the anterior (front) and posterior (rear) lobes. The hormones produced by the anterior lobe include those responsible for growth (**growth hormone or GH**), development in puberty (**gonadotrophins**), as well as hormones which stimulate the thyroid (**thyroid stimulating hormone or TSH**) and the adrenal glands (**adrenocorticotrophic hormone or ACTH**). The posterior lobe stores and releases **vasopressin**, also called **antidiuretic hormone (ADH)**, which regulates the levels of fluid in the body and so the amount of water passing out through the kidneys and into the urine.

## ***Optic nerves***

The nerves from both eyes, the **optic nerves**, meet and cross at what is called the **optic chiasm** which is found just above the pituitary gland. From here they carry the visual information to the back of the brain. If there is pressure on the optic nerves from the growing craniopharyngioma, vision can be affected, the most serious outcome being complete loss of sight.

## ***Ventricles of the brain***

In the brain there are areas called **ventricles** which contain the fluid known as cerebrospinal fluid (CSF). The CSF flows through the ventricles via channels and openings, circulates over the surface of the brain, and eventually drains into the bloodstream. There is a continuous production and circulation of CSF and so, if the ventricular channels become blocked, preventing escape of fluid from this area, there will be an increase in pressure as the fluid builds up. This causes the ventricles to enlarge resulting in a condition known as **hydrocephalus** (sometimes called ‘water on the brain’ and the symptoms of this are headaches and vomiting.

## **WHAT ARE THE SYMPTOMS OF A CRANIOPHARYNGIOMA?**

With the help of the previous diagram and definitions, you can see that a craniopharyngioma may affect many parts of the brain. It can, therefore, cause some or all of the following symptoms due to the increased pressure caused by the tumour squashing these surrounding areas, disturbing the communication system between the hypothalamus and the pituitary, and preventing both the normal functioning of the pituitary gland and the production and release of hormones.

The most common symptoms are headaches, vomiting and disturbed vision.

Your child may experience some or all of the following symptoms. They usually appear gradually and will require investigation:

- ***Headaches with or without nausea and vomiting***

It is particularly significant if these occur first thing in the morning or at night. These are due to increased pressure within the brain caused by the increased size of the tumour. If the affected area extends far enough upwards, this can cause blockage of the CSF circulation in the ventricles which leads to hydrocephalus.

- ***Disturbance of vision, occasionally leading to loss of sight***

This is due to the closeness of the craniopharyngioma to the optic nerves and optic chiasm. As the craniopharyngioma grows, it starts to press against the nerves and this stops the signals from the eyes reaching the brain and so sight can be impaired or lost altogether.

- ***Effects on hormone secretion***

Because the pituitary gland, the hypothalamus and/or the connection between them can be affected by the growing craniopharyngioma, the production and release of the pituitary hormones may be impaired and so the following problems may result:

***Poor growth.*** One of the causes of poor growth in children with a craniopharyngioma is the effect on the pituitary gland which means that there may be growth hormone insufficiency. In these cases normal growth is affected. However, it is not fully understood why children with a craniopharyngioma, but who do not have a hormone deficiency, have poor growth.

***Delayed onset of puberty.*** Pubertal development includes breast and pubic hair growth in girls and growth of the penis, facial and body hair and muscular development in boys. These events will not occur if the hormones (the gonadotrophins) which are needed to trigger them are deficient as a result of damage to the pituitary gland.

**Early onset of puberty (very rare).** An unusually early onset of the pubertal changes can occur in some children, again resulting from the impact of the craniopharyngioma on the hypothalamus and the pituitary.

**Increased thirst and more frequently passing urine.** The effect on the hypothalamus and the posterior lobe of the pituitary may mean that vasopressin insufficiency develops leading to the condition called diabetes insipidus. The symptoms of this are excessive urine output and thirst with individuals drinking up to 6 – 10 litres of fluid per day.

● **Other symptoms that may occur include:**

- Tiredness and frequent infections which are difficult to 'shake off'
- Intolerance to cold temperatures
- Disturbance of normal sleep patterns with either excessive sleepiness during the daytime. or increased waking at night
- Weight loss or weight gain with a normal, decreased or increased appetite
- Behavioural problems such as becoming withdrawn, introverted or aggressive or having poor concentration
- Poor balance or weakness in the arms or legs
- Rarely, fits may occur

## ***HOW IS A CRANIOPHARYNGIOMA DIAGNOSED?***

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The following investigations will be needed to confirm the diagnosis:

**Skull X-ray:** This may be helpful in showing up flecks of calcium which are almost always present in childhood craniopharyngiomas. However, this investigation is not always performed because brain scans give more information and are now much more widely available.

**Brain scan (either an X-ray (CT) scan or magnetic (MRI / NMR) scan or both):**

This will enable the tumour, as well as the surrounding areas of the brain, to be seen in great detail so that further treatment, especially surgery, can be planned very carefully.

**Additional tests that form part of the overall assessment may include:**

**Visual assessment:** This will be done by the endocrinologist, or the eye specialist, to check if there has been any effect on your child's sight, particularly to their field of vision.

**Measurements of height and weight:** This will be done by your endocrinologist and the measurements will be plotted to assess your child's rate of growth. Your child's stage of development in puberty will also be checked when appropriate.



**Blood tests:** These will be done to check the levels of hormones to see if there are any which are insufficient.

**Water balance:** This is a test which is performed to establish the level of fluid in the body. It is done by comparing the concentration of the blood with the concentration of the urine, as there is a relationship between the two which should be maintained. If there is a difference between the two concentrations, it may indicate that your child has diabetes insipidus resulting from vasopressin insufficiency.

**Psychological assessment:** Depending on the severity of the craniopharyngioma, and what treatment is planned, it may be important to assess your child's stage of learning, memory and behaviour, as all these may be affected by the development of the tumour and/or its treatment.

## **WHAT ARE THE TREATMENTS FOR A CRANIOPHARYNGIOMA?**

Because of the problems a craniopharyngioma causes, particularly if it is growing rapidly, it is usually best to try and remove it surgically. There are different ways this can be done, although usually, in children, it will require a major operation.

Normally the surgeon will try to remove as much of the tumour as possible, but this should not be at the expense of serious damage to the nearby areas of the brain. As mentioned previously, these tumours can be very 'sticky' and they attach themselves to the surrounding tissues, so removing them can be a very difficult and delicate procedure.

It may also be necessary to follow the surgery with radiotherapy, to help prevent any re-growth of the tumour, but this will also depend on the age of your child.

Some of the treatments will now be discussed in more detail.

### **Surgery**

#### **Craniotomy:**

In most hospitals where treatment for children with a craniopharyngioma is available, the primary treatment is surgery to remove all, or part, of the tumour. This involves a major operation known as a craniotomy which requires opening the skull at the front, or front and side, of the head, near the hair-line, usually on the right side. The hair around the area of the incision will need to be shaved, but usually no more than this is needed.

By looking carefully at the scans which were taken before the operation, the surgeon will decide whether it is best to try and completely remove the craniopharyngioma. This is usually the aim but if, during the operation, it can be seen that there are solid or cystic parts of the tumour stuck very firmly to the optic nerves, the hypothalamus, or a major blood vessel, then it will be far too hazardous to try and remove all of the craniopharyngioma without causing severe damage to these sensitive parts of the brain.

Often, the tumour can be only partially removed, leaving a significant portion behind. Other treatments, such as radiotherapy or cyst drainage, may then be required and these will be discussed with you by your specialist.

### ***Trans-nasal operation (operation through the nose):***

Very rarely, if the tumour is small and confined to the area where the pituitary gland lies, the operation can be done through the nose. However, it is **very much the exception** for this operation to be performed in children.

### ***Cyst aspiration:***

The cystic part of a craniopharyngioma may be a single cyst or may be formed by several cysts joining with one another. Before attempting to remove the tumour, the surgeon may decide to drain or **aspirate** the contents of the cyst, particularly if there is one major cyst which is pressing against a critical structure and causing symptoms of increased pressure within the brain. This involves a small operation whereby a hole (**burr hole**) is made in the skull and a tube is passed into the cyst to drain its contents. The surgeon will then go on to perform a craniotomy at a later date. Cyst aspiration may also be done if the cystic part of a tumour re-grows.

### ***Drainage of the ventricles (a 'shunt'):***

In about a third to a half of children with a craniopharyngioma, the ventricles of the brain become enlarged due to the build-up of CSF and it may be necessary to re-establish the flow and drainage of the CSF before attempting removal of the tumour itself. This involves inserting a form of drainage system into the ventricles that can be either a temporary or permanent measure. Most commonly, the drainage system is a tube, called a shunt, that connects a ventricle to another body cavity, usually the abdomen (this is then called a **ventriculo-peritoneal shunt**). There is an interconnecting valve so that the speed and pressure at which the fluid drains can be controlled.

Occasionally, after the main operation to remove a craniopharyngioma, it is necessary to insert a shunt. Less commonly a shunt may be needed if the tumour re-grows and causes obstruction of the CSF flow.

## ***Radiotherapy***

Radiotherapy is a way of targeting radiation at a tumour in order to kill the tumour cells with minimal damage to the surrounding normal tissues. This form of treatment is more commonly used for malignant or cancerous types of tumour but has been found to be effective in preventing re-growth of a craniopharyngioma.

Radiotherapy is mainly used in children who have had surgery to remove their craniopharyngioma, especially if it was decided to remove only part of the tumour. It may be that a course of radiotherapy is given as soon as the child has recovered from their

surgery. Alternatively, a ‘watch and see’ policy may be adopted, with the option of giving radiotherapy at a later date if scans show that the craniopharyngioma is regrowing. If the neurosurgeon is confident that all of the tumour has been removed at surgery, and this is confirmed on further brain scans, then it is *unlikely* that radiotherapy will be needed.

The age of the child is also important when deciding whether radiotherapy is given. It is generally **not** used for children under two years of age, and rarely under five years, because of the concerns of the long-term effects of radiation on the developing brain. However, newer, more refined techniques of giving radiotherapy are being developed all the time so that this form of treatment may play a more important role in the future.

## **Chemotherapy**

Like radiotherapy, chemotherapy is used only for certain cancers and is not used for the **treatment** of craniopharyngiomas. However, a particular antibiotic-like drug called **bleomycin** is being used in some centres in Europe prior to surgical removal of the cystic portion of a craniopharyngioma. A month or so before the operation, bleomycin is injected into the cyst to thicken or shrink the wall of the cyst, and this can make surgical removal easier.

## ***WHAT HAPPENS DURING THE INITIAL TREATMENT?***

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Your child may be in hospital for a few weeks, although this will depend upon the individual patient and what treatment he/she receives. Usually, the first few days before the operation are spent performing the investigations already described and assessing the results of the brain scans.

If the insertion of a shunt, or the aspiration of a cyst, is needed first, there may be a wait of a few days, or occasionally a few weeks, before the main operation to remove the tumour is carried out. This time may be spent in or out of hospital depending on the individual child.

After the operation to remove the tumour (a craniotomy) your child is likely to be in hospital for two to three weeks. Again, much will depend upon the progress of the individual child.

If radiotherapy is additionally required, then arrangements for this will be made once your child has recovered from surgery. The course of treatment takes approximately six weeks, as small doses of radiation are given every day in order to reduce the side effects of the total dose. Your child may not have to stay in hospital for this course of radiotherapy; however, this will depend on the age, co-operation and well-being of the individual child.

## ***WHAT HAPPENS IMMEDIATELY AFTER THE OPERATION?***

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As with any major operation, your child will be very closely monitored for the first 24 to 48 hours following surgery. This period of observation may need to take place in the Intensive Care Unit depending on whether your child requires the support of a breathing machine. However, this is usually only the case if the operation has taken several hours and it is therefore felt better to wake up your child more gradually, giving a period of complete rest and stability for the first few hours after surgery.

A careful record of fluid balance, i.e. liquid intake and urine output, will need to be kept during, as well as after, the operation (see below). All urine produced is measured and checked for its concentration and a urinary catheter may be used initially to make this easier and more comfortable for your child. Likewise, all fluid taken in, through a drip at first, and then by mouth, will be carefully calculated and recorded. Blood and urine samples will be taken at least daily for the first few days to make sure this balance of intake and output is correctly maintained.

## ***WHAT CAN BE THE AFTER-EFFECTS OF A CRANIOPHARYNGIOMA AND/OR ITS TREATMENTS?***

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<p>A craniopharyngioma occurs in a part of the brain which is very close to the hypothalamus and the pituitary gland and so most of the effects are due to damage which has occurred to these very important areas.</p>
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### ***Diabetes insipidus***

The hypothalamus normally monitors the state of water balance in the body. In conditions where the body might become dehydrated, for example in very hot weather, the 'thirst centre' in the hypothalamus is stimulated so that we feel thirsty and drink more water. The hypothalamus also signals to the posterior lobe of the pituitary gland to release the hormone **vasopressin** in larger amounts. This hormone acts on the kidneys in order to prevent water loss, i.e. urine is passed **less frequently** and appears **more concentrated**.

Without **vasopressin** the urine would continue to be dilute and passed out frequently and we would feel very thirsty and want to drink almost constantly so as to try and compensate for this lack of control of water loss from the kidneys. Absent or inadequate production and release of **vasopressin** disturbs the ability of the body to maintain a normal fluid balance and leads to a condition called diabetes insipidus. This is quite different from diabetes mellitus, which is also called 'sugar diabetes'. A very young child with **diabetes insipidus** is not able to say how thirsty he/she feels, and so is dependent

on carers for the provision of food and drink. In these young children it can be seen that there is a real risk of the child becoming rapidly dehydrated without the correct treatment.

As mentioned earlier, the location of a craniopharyngioma is usually very close to the pituitary gland and stalk. During surgery, it is often necessary to cut the pituitary stalk in order to reach the tumour. This disrupts communication between the hypothalamus and the pituitary gland, and the pituitary hormones will no longer be produced and released as needed. **Vasopressin** is one of these hormones and it is therefore extremely common for diabetes insipidus to develop after the operation. This explains why fluid intake and output are so closely observed following surgery.

## ***How is diabetes insipidus treated?***

Signs of diabetes insipidus are quickly noticed because of the strict fluid balance records. Diabetes insipidus is treated by replacing the deficient hormone, vasopressin, with a synthetic form of the hormone called an 'analogue'. 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 and therefore can be given less often. Also, it may be given in a different way, e.g. by tablets or intranasal drops, rather than by injection.

During the first few days after surgery, the vasopressin analogue is given, as necessary, by injection into the muscle. As soon as your child is well enough, the treatment can be given using a nasal preparation, either as a spray which is sniffed up the nose or as drops blown or sniffed from a fine, transparent, plastic tube. It is usually required once or twice a day. Alternatively, vasopressin may be given as tablets and in order to achieve the very much smaller doses needed in children, your specialist will advise you how to break the tablets to give the correct dose.

In the first few days following surgery, the correct fluid balance for your child may be quite difficult to sort out and your child may have periods of feeling very thirsty and wanting to drink frequently. These problems soon settle down once a smoother control has been achieved with the right dose and timing of vasopressin.

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.

Some children may have damage to their thirst centre as well as having diabetes insipidus. In such children, although they lose a lot of fluid in large volumes of dilute urine, they do not get a feeling of thirst. This is called **adipsia**. These children may rapidly dehydrate if

not **made** to drink fixed quantities of fluid. Your doctor will provide you with an idea of how much fluid your child needs each day – this may be called a fluid ‘prescription’.

## ***Will my child always need to take vasopressin?***

Diabetes insipidus, when it occurs, may not be permanent. It will become apparent in the first few weeks after surgery whether vasopressin will be needed in the long term. This depends on the degree of damage to the hypothalamus, the pituitary stalk and the posterior pituitary. Obviously, if the stalk has been cut, recovery cannot take place and vasopressin treatment will be required for life.

## ***Other treatments***

### ***Anticonvulsants (drugs to prevent fits):***

Rarely, a child with a craniopharyngioma has a convulsion as the first sign that there is anything wrong. Medication will then almost certainly be given to cover the period of the operation so as to prevent any further fits occurring. It may be that the medication can be gradually withdrawn a few months after the operation if no further problems arise.

Even if your child has not had any previous fits, any interference with the brain, such as the trauma of an operation, may potentially cause a fit. Therefore, some neurosurgeons like to treat the child with anticonvulsants before and after the operation. The treatment can be gradually withdrawn if there are no problems.

A proportion of children do continue to have problems with fits, or else develop them later on, in which case they may need medication for a longer time to keep these under control.

### ***Steroids:***

Steroids are always given before and after surgery for a craniopharyngioma. There are two main reasons for this:

- To help reduce the swelling and build-up of pressure within the brain, high doses (**dexamethasone** or **hydrocortisone**) are given.
- **Adrenocorticotrophin (ACTH)**, one of the hormones produced by the anterior lobe of the pituitary gland, triggers the adrenal glands to make and release a steroid called cortisol. Cortisol is produced in varying amounts during the day and night but, more importantly, it enables us to cope with stress such as illness and trauma. At these times, greatly increased amounts of cortisol are released into the blood-stream.

If the pituitary gland and stalk are damaged, either by the craniopharyngioma itself or the surgery, then the ability to produce cortisol is lost and the body will be unable to cope with the stress of an operation unless replacement steroids are given. The initial high doses are gradually reduced, following the operation to much smaller doses which are then continued in the form of **hydrocortisone** tablets until further tests of pituitary function have been carried out (see below).

**Thyroid hormone or thyroxine:**

Following the operation, if there has been damage to the hypothalamus, the pituitary or the connection between, the pituitary will be unable to release the hormone necessary to stimulate the thyroid gland. Replacement thyroid hormone treatment will be needed and this is given in the form of tablets.

**Growth hormone:**

As mentioned previously, damage to the hypothalamus and/or the pituitary will mean that the anterior lobe of the pituitary is unable to release growth hormone in sufficient amounts and replacement treatment will be needed. The only way to give growth hormone treatment is by injection because growth hormone is a protein and 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.

**Sex hormones:**

If there is an insufficiency of the gonadotrophins, there will not be the stimulus to the gonads (ovaries in girls, testes in boys) to produce the sex hormones (oestrogen and testosterone respectively) which are needed to trigger the physical changes which occur during puberty. At the appropriate age, replacement treatment will be needed.

More information about some of these conditions is available in other CGF booklets as follows:	
Growth hormone deficiency	Booklet 2
Delayed puberty	Booklet 3
Premature puberty	Booklet 4
Emergency treatment for cortisol deficiency	Booklet 5
Multiple pituitary hormone deficiencies:	Booklet 11
Diabetes insipidus:	Booklet 12

Approximately two to three months after the operation, some blood tests, which take a morning to perform, will usually be done to assess how well the pituitary gland is working. From the results of these, it should be clear which hormones will need to be replaced. Treatment, once started, will almost certainly be **for life**.

When all the pituitary hormones are deficient, the term **panhypopituitarism** or **multiple pituitary hormone deficiencies (MPHD)** is used. Unfortunately, most children will become panhypopituitary after surgery for a craniopharyngioma. Even if the pituitary is still functioning shortly after the operation, it is likely, with time, that it will gradually stop producing hormones in sufficient amounts, particularly if radiotherapy has been given as well. Careful follow-up is therefore **always** required by a hormone specialist (**endocrinologist**), and this will continue throughout adult life.

These few additional points are very important to remember:

- Medication **MUST** always be given regularly as prescribed. Do **NOT** run out of supplies.
- Hydrocortisone is **VITAL** in helping the body cope with stress. At times of illness, the prescribed dose will need to be **INCREASED** and if vomiting occurs, the hydrocortisone will need to be given by injection (see emergency pamphlet).
- Children who are deficient in pituitary hormones and/or who suffer with fits, for whatever reason, should wear an identity necklet or bracelet and carry a medical card.

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## **WHAT OTHER FOLLOW-UP WILL MY CHILD NEED?**

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### **Eyes**

Problems with sight can improve after surgery but may be permanent. Eye tests will therefore be needed at regular intervals and special help organised at school if your child's sight is very restricted in one or both eyes. Some children may need to be registered as partially-sighted or blind.

### **Growth**

Measurements of height, weight and pubertal development will need to be recorded carefully and regularly. This is best done in a growth clinic by a paediatrician/paediatric endocrinologist. Children who have a craniopharyngioma can grow normally following surgery, even though the pituitary gland is no longer producing **growth hormone**. Normal growth may continue for many months or years but usually slows down after one to two years, at which point growth hormone injections can be started to re-establish normal growth once more.

However, those children who do grow without growth hormone are usually those who gain a lot of weight because they eat very large quantities of food. This is thought to happen as a result of injury to the hypothalamus when the craniopharyngioma is removed during surgery which damages the eating control centre (see below). It is therefore very



important to monitor your child's weight. Other children grow very poorly before and after surgery due to their growth hormone deficiency and therefore need treatment at a much earlier stage than the above individuals.

## ***Brain scans***

The timing of when further X-ray (CT) or magnetic (MRI) scans are needed will vary from child to child. As a guideline, a scan will be performed shortly after surgery, either within the first 24-48 hours or before your child is discharged from hospital, to make sure all is well. A repeat scan will probably be arranged for six months after the operation and then, depending on the results of this, six-monthly to yearly for the next two or three years, and then as recommended by your specialist.

## ***School progress***

It will be very important to monitor your child's progress at school as, for various reasons, up to two-thirds of children may experience learning problems following treatment of a craniopharyngioma. Problems may range from:

- difficulty in catching up on missed work because of the amount of time taken off school
- difficulties keeping up due to poor sight
- difficulties with concentration because the child is distracted by thoughts of food (see below)
- difficulties due to true learning or memory problems

Learning difficulties may result from injury to the hypothalamus, injury to other areas of the brain during the operation, or from the effects of radiation therapy which may build up over a period of years. Ideally, a psychologist should be involved in the care of your child from the beginning so that further tests of learning and memory can be done at intervals after the operation and/or radiotherapy. In this way, any special needs can be picked up early and prompt referral can be made to an Educational Psychologist so that appropriate support and supervision can be organised at school as soon as possible. Some children's needs may be better met in schools geared to special educational aspects and it is crucial not to let these individuals struggle for several years before finding this out.

## ***ARE THERE ANY OTHER EFFECTS OF A CRANIOPHARYNGIOMA OR ITS TREATMENT TO BE MENTIONED?***

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Earlier in this pamphlet, the hypothalamus was mentioned as having a number of important functions and its closeness to a growing craniopharyngioma can be appreciated by the diagram on page 3. At the time of diagnosis, a craniopharyngioma will often have grown so large that it presses against the hypothalamus and this may result in some

changes in behaviour such as disturbance of eating and sleep patterns. Neurosurgeons are increasingly aware of the importance of causing as little damage as possible to this area during the operation, even if this means leaving a small amount of the tumour behind. However, some damage may still occur, and this can cause certain behaviour problems. Sometimes, but not always, these problems will improve with time.

## ***Increased food intake and obesity***

It is **extremely common** for children to gain some weight after surgery for a craniopharyngioma but the amount varies from child to child and is much **less** likely to be a problem in those children whose tumour is not adhering to the hypothalamus at the time of operation. Increase in weight is due to a combination of the high doses of steroids given around the time of the operation, pituitary hormone deficiencies and/or injury to the '**eating control centre**' of the hypothalamus. When this happens, a persistent feeling of hunger results so that your child may feel hungry even if he/she has just eaten. It seems as though they need to eat much more than normal to try and 'fill up'.

In those children where there has been an effort to remove all the tumour, and as a result there has been damage to the eating control centre, up to 60% become obese because of the change in eating behaviour. The increase in appetite can develop very soon after surgery so that the child becomes totally preoccupied with thoughts of food. Understandably, this may cause distress to the child and his/her family, particularly if there is a sudden change in body appearance because weight gain has been so rapid. Problems at school may also occur both from poor concentration and, occasionally, from teasing by other children.

Handling this problem can be very difficult and in a small minority of children total dietary control is necessary, including locking kitchen doors, cupboards and refrigerators. It is important to discuss the issue openly and to encourage the involvement of your child as much as possible in the ideas of how to deal with it. Appreciating that this is a specific medical problem may help you to understand your child's 'need' for food and enable you to plan for when your child becomes more independent. Early referral to a psychologist may be helpful.

There is a booklet available which gives some creative ideas on how to help children with this type of eating disorder. It has been prepared by parents of children with a completely different medical problem to your child but who also have a disturbance of their eating control centre and who become very obese. If you think this booklet would help you and your child, contact the Child Growth Foundation for further details.

## ***Sleep disturbance***

This may mean that your child wakes up several times during the night or falls asleep at odd times during the day. This symptom is related to the effect of the tumour itself

pressing on other areas of the brain and/or due to damage to some of these surrounding areas during surgery. It usually continues even as the child gets older. The main problems associated with this are the impact on the child's education (they may be very tired during the day when they are at school) and the disturbance to other members of the family – parents, brothers and sisters, etc. – particularly if the night-time wakefulness is spent looking for food!

## ***Impaired thirst***

This is fortunately rare because the inability to feel thirsty is a serious problem, particularly when associated with diabetes insipidus (see pages 10–12).

## ***Memory disturbance***

Memory may be affected either by damage to the hypothalamus or by disturbance to other nearby areas of the brain. Short-term rather than long-term memory is usually affected, although this may depend on the individual child.

## ***Impaired temperature regulation***

This is not usually too much of a problem. Parents notice that their child is not sensitive to extremes of temperature—when everyone else is piling on jumpers, their child is walking around in T-shirts or, on a hot day, they are asking for their jumper!

## ***Mood swings***

You may find that your child has a much 'shorter fuse' than before the operation and has temper tantrums which are quite out of character. This may, of course, be due to the fact that your child has been ill although, occasionally, damage to the hypothalamus may play a part. In some cases it may be necessary to seek advice from a child psychiatrist.

## **QUESTIONS AND ANSWERS**

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### **Q1 What are the signs that a craniopharyngioma is re-growing?**

**A** Your child may have no symptoms at all and a re-appearance of the tumour may be found during a routine follow-up scan, or else any of the symptoms that your child first had may return. Poor growth, despite hormone replacement treatment, may be an early sign of the tumour re-growing.

### **Q2 What are the chances of the craniopharyngioma re-growing and what will happen?**

**A** If a craniopharyngioma is going to re-grow, it usually does so within three years from the first treatment, which is why it is important to have regular brain scans during this time. If only a partial removal of the tumour was possible, then there is

a higher chance of the tumour increasing in size again. The treatment options will depend on the individual child. Radiotherapy is commonly used. Occasionally, further surgery is necessary. Whatever the treatment plan decided upon, this will be to allow the best possible Outcome for your child.

**Q3 Will my child die from this condition?**

**A** The chances are no, as approximately 90% of children treated for a craniopharyngioma are alive at 10 years from diagnosis. However, a small proportion of children do not survive their tumour or the related problems.

**Q4 Will other children of ours have this condition?**

**A** No, this is a developmental abnormality that happens by pure chance. It is not familial or hereditary (i.e. passed on from parents or grandparents).

**Q5 Will my child be able to have children?**

**A** Even though your child will almost certainly need hormone replacement (testosterone or oestrogens) in order to go through puberty normally, and will need to stay on this treatment to maintain sexual development, it should still be possible for your child to have children. Additional hormone treatment will be required in order to become fertile, and this is one of the reasons why your child needs to remain under the care of an endocrinologist as an adult.

**Q6 What will happen when my child becomes an adult?**

**A** As an adult, most of the hormone treatments will need to be continued and this may include growth hormone. In addition, because some of the hormone deficiencies may develop at an older age, it is important that your child is referred to an adult endocrinologist at the appropriate time for continuity of their care.

**Q7 Why is this tumour different in adults?**

**A** Although a craniopharyngioma may occur for the first time in adulthood, this tends to be a different type of tumour from the one occurring in childhood. In fact, the cells making up the tumour are usually different. Childhood craniopharyngiomas tend to be more difficult to treat, especially the ones that cause symptoms at a very young age, as these tend to be growing faster. Also, treatments such as radiotherapy may have to be delayed because of the young age of the child.

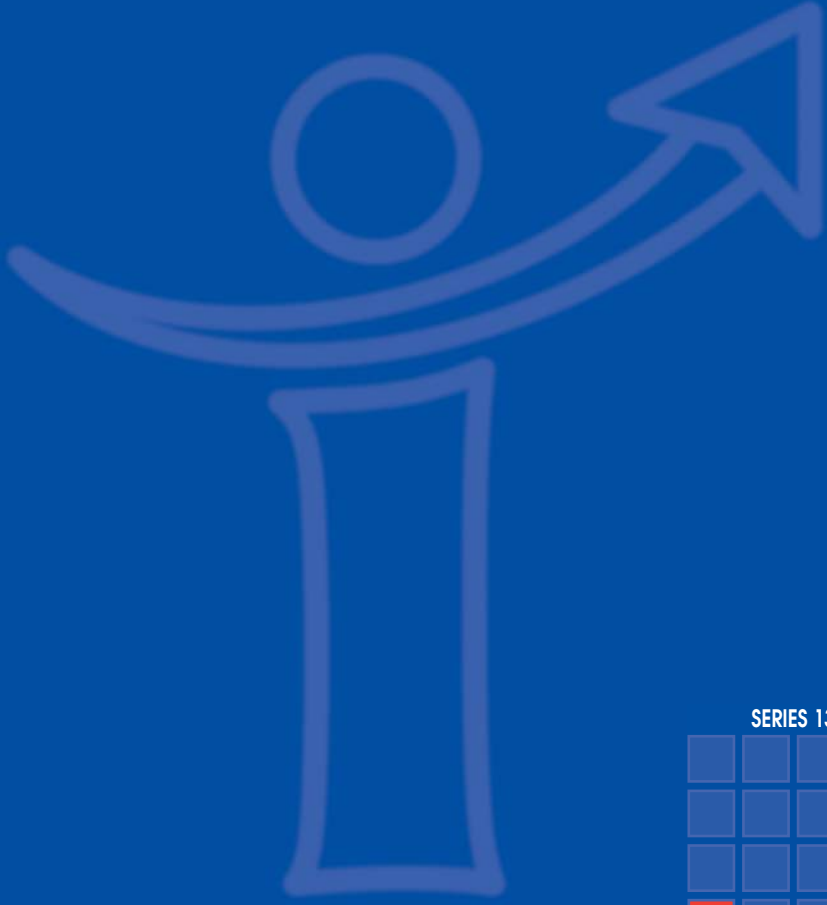
## ***FINAL COMMENTS***

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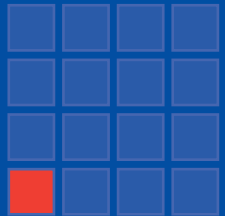
This pamphlet has tried to give a very general overview of the diagnosis, treatment and associated problems of having a craniopharyngioma. As stressed at the beginning, treatment should always be individualised and not everything written will apply to your child. Information given in other pamphlets in the series may also be helpful.

# NOTES

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SERIES 13



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