Craniopharyngioma

This factsheet provides information about craniopharyngioma and its treatment. If you would like further information, please ask a nurse, doctor, or other healthcare professional involved in your care.

What is a craniopharyngioma?
Craniopharyngioma is a type of brain tumour that is benign. This means it is not cancerous and will not spread to other parts of the body. It often consists of a solid part and a cystic part made up of small balloon-like pockets containing fluid. Parts of the craniopharyngioma may have been present for a long time. They may enlarge very slowly, so symptoms can appear at any time in childhood or in adult life.

Craniopharyngiomas are more common in childhood but can appear at any age. Very slow-growing tumours may not become symptomatic until adulthood. The cause of craniopharyngioma is not known.

What are the signs and symptoms?
The enlarging craniopharyngioma may put pressure on neighbouring parts of the brain which may cause the following symptoms:

- **Headaches** can be caused by pressure on the brain or from blockage of the flow of fluid around the brain (cerebro-spinal fluid or CSF); this is known as hydrocephalus.
- **Alteration in vision** can be caused by pressure on the optic nerves (nerves to the eyes).
- **Hormone deficiencies** can be caused by increased pressure on the hypothalamus and pituitary gland. This can disrupt the production of hormones normally made by these glands. Deficiency of one or more of the hormones may cause the following symptoms:

    - Tiredness
    - Weight gain
    - General slowing-down
    - Thirst
    - Passing large quantities of urine
    - Infertility or loss of sex drive (libido)
    - Skin changes (such as dry skin or thinning of the skin).

Symptoms caused by hormone deficiencies may take a long time to become apparent. They can be treated effectively by taking replacement hormones, usually in tablet form. Most people will need to be under the care of an endocrinologist (a doctor who specialises in managing hormone-related conditions) who will monitor the hormone levels regularly.

Revised: August 2020 Planned review: August 2023

What tests and investigations will I need?

- A CT (computerised tomography) scan and /or an MRI (magnetic resonance imaging) scan to find out the position and size of the craniopharyngioma. This will help with the planning of the treatment.
- Baseline blood tests to check the hormone status.
- An eye examination to check for pressure on the optic nerve and simple tests to check visual fields and visual acuity (how well you see).
- In children, it is important to check height and weight.

What is the treatment for craniopharyngioma?
Craniopharyngiomas can be surgically removed, treated by radiotherapy, or a combination of both. The aim of treatment is to stop the craniopharyngioma from growing or causing further problems.

Surgery
The pressure caused by an enlarging craniopharyngioma is best relieved by surgery. Craniopharyngiomas can be approached through the nose or by a small opening under the lip using keyhole surgery. This is called a transphenoidal resection. It can also be approached through the front of the head by a craniotomy (opening of the skull bone).

The aim of surgery is to remove part or all of the craniopharyngioma and relieve the pressure on the surrounding areas. Complete removal might seem the ideal treatment but this can carry considerable side effects depending on the size and position of the craniopharyngioma. Side effects include possible damage to the optic nerves and worsening of sight, damage to the pituitary gland which can cause hormone problems or damage to the hypothalamus causing excessive hunger and obesity. Removal of **some** rather than all of the craniopharyngioma may be a safer procedure to reduce the risk of damage. Removal may improve eyesight, but usually does not result in recovery of pituitary gland hormone production.

If the cystic part of the craniopharyngioma expands causing rapidly worsening symptoms of pressure, a small operation to drain the cyst will relieve the pressure, preventing further damage.

Occasionally there may be a build-up of CSF (cerebrospinal fluid) causing the ventricles (fluid-filled spaces inside the brain) to enlarge, known as hydrocephalus. This may require a shunt (a tube) to be inserted to drain the fluid.

Radiotherapy
Radiotherapy is delivered using high-energy x-ray beams (photons) which are specifically focused on the tumour. Radiation is commonly given in the form of fractionated stereotactic radiotherapy.

What is fractionated stereotactic radiotherapy (FSRT)?
Fractionated **s**tereotactic radiotherapy is a high precision treatment delivered on a machine called a linear accelerator (Linac). The Linac focuses radiation on the tumour and gives as little radiation as possible to the rest of the brain. This is usually delivered with 30 daily treatments sessions over a period of six weeks, Monday to Friday. It requires a specially fitted face mask to be worn which keeps the head in the same position for each session. This allows the treatment to be targeted very accurately and minimises damage to healthy cells.

What are the side effects of fractionated stereotactic radiotherapy?

Short term side effects

Having daily treatment often makes people feel tired. Towards the end of treatment there may be small patches of hair loss. After treatment the hair grows back normally. (Please also see the separate factsheet *Fractionated brain radiotherapy*).

Before, during and immediately after radiotherapy, the fluid filled part of the craniopharyngioma (the cyst) may swell or increase in size. We do not know what causes it but it does not mean that the treatment is ineffective.

If the cyst gets bigger, which happens in 10–20% of cases (10-20 people in 100), it may put further pressure on the optic nerves and therefore eyesight may worsen. Occasionally, it may block the flow of fluid from the brain causing hydrocephalus. This can result in headaches and sickness. During radiotherapy, a doctor or clinical nurse specialist (CNS) will review you once a week to check your symptoms and your field of vision. It is important that you report any headache or worsening in eyesight. If the cyst gets bigger, the fluid may need to be drained by a simple operation. You will be monitored carefully throughout treatment.

Long term side effects

- **Vision** - There is a small risk that the nerves to the eyes may be damaged by radiation. In 1-2% of cases (one to two people in 100), this may lead to worsening of vision months to years after treatment.
- **Hormones -** Radiation may cause damage to the production of pituitary hormones. Your hormone levels will be monitored once or twice a year. The risk of deficiency in people who have normal hormones before radiotherapy, is 30-50% at 10 years and increases with time. It can be easily treated by taking the necessary hormones by mouth or by injection (these need to be continued indefinitely).
- **Other tumours** - There is a small risk of developing another tumour in the brain - approximately 1% (one in 100) at 10 years and 2% (two in 100) at 20 years.
- **Other possible side effects -** Based on research in other conditions where similar radiotherapy is given, it is estimated that there may be a slightly higher risk of having a stroke. There is concern that radiation may cause damage to parts of the brain surrounding the tumour but overall, this risk is low.

How effective is radiotherapy and what are the benefits?

The aim of radiotherapy is to stop the tumour growing. The success of radiotherapy is described in terms of tumour control. A tumour is controlled if it remains the **same size** and does not need further treatment.

After conventional radiotherapy, tumour control is in the region of 90% (90 in100) in 10 years and 80% (80 in 100) in 20 years. This means there is approximately a 10% chance (10 in 100) of needing further treatment over 10 years and a 20% (20 in 100) chance over 20 years.

These results are based on studies performed over many years. With new radiotherapy techniques the present results are even better. However, the new techniques have not been in use for as long and

it is therefore not known whether in 10-20 years the results would be the same or better than achieved in the past.

How do I decide what to do?
The advice to have radiotherapy is based on the balance of risks and benefits. Your doctor will be happy to discuss this with you in further detail. Radiotherapy is considered to be relatively safe compared to the risks of not having the treatment. Overall, partial surgical removal of the craniopharyngioma followed by radiotherapy results in a very good chance of never needing further treatment.

To date there are no clinical trials which have directly compared the different treatment options for craniopharyngioma. A review of the literature suggests that limited surgery combined with fractionated radiotherapy achieves the best results, in terms of long term control of the tumour with the least side effects.

Contact details

Neuro-oncology Clinical Nurse Specialist (CNS)	020 8915 6011
The Royal Marsden Macmillan Hotline (available 24 hours a day, 7 days a week)	020 8915 6899