A GUIDE TO CRANIOPHARYNGIOMAS

Australian Pituitary Foundation

WHAT ARE CRANIOPHARYNGIOMAS?

A craniopharyngioma is a rare, benign (non-cancerous) brain tumour. They can be solid or cystic (fluid-filled sacs). These tumours begin near the pituitary gland, the small pea-sized gland that sits near the base of the brain. The pituitary gland releases hormones that control many body functions.

Craniopharyngiomas can occur at any age but are most common in children aged five to 15 and older adults over 50.

These tumours can affect the function of the pituitary gland and other nearby structures in the brain. Symptoms include gradual changes in vision, tiredness, excessive urination and headaches. Children with craniopharyngiomas may grow slowly and be smaller than expected.

HOW COMMON ARE CRANIOPHARYNGIOMAS?

Around 450 Australians are living with craniopharyngiomas. Each year, about 30 people are diagnosed.[1]

WHAT ARE THE SIGNS AND SYMPTOMS?

COMMON SIGNS

- Headaches (sometimes accompanied by vomiting)
- Diabetes insipidus (excessive thirst and urination)
- Vision disturbance
- Disturbed sleep patterns
- Slow growth
- Behavioural problems
- Early or delayed puberty
- Increased sensitivity to cold or heat
- Tiredness
- Frequent infections
- Hypopituitarism (in children, this causes impaired growth, delayed puberty and other symptoms related to the other hormone deficiencies that may be present).

If the tumour affects the hypothalamus, signs can include:

- Increased or decreased appetite (resulting in weight problems)
- Increased thirst
- Mood swings
- Sleep disturbance
- Reduced concentration
- Short-term memory loss

EARLY-CHILDHOOD SYMPTOMS

In early childhood, tumours can be fast-growing and more aggressive

Symptoms are often present for a number of years before a diagnosis is made.

Because these tumours may be associated with raised intracranial pressure, symptoms can also include headaches, nausea and vision problems.

DIAGNOSIS

Standard tests to diagnose a craniopharyngioma include:

- **Clinical assessment** – a medical history and neurological exam
- **Blood tests** – these can identify changes in hormone levels that suggest a tumour is affecting the pituitary gland
- **Scans** – a magnetic resonance imaging (MRI) or computerised tomography (CT) scan can find the tumour's size and position

After Diagnosis

After diagnosis, it is essential to see:

- **An endocrinologist** with experience in managing pituitary diseases, and
- **A neurosurgeon** with pituitary expertise.

TREATMENT

Surgery - Surgery is the preferred treatment. The type of surgery depends on the size, location and position of the tumour. Surgery options include:

- Minimally invasive (transsphenoidal) – the surgeon reaches the tumour through the nose
- Open surgery (craniotomy) – this involves opening the skull, most commonly in the centre of your scalp, to gain access to the tumour.

Surgery may not be able to remove the tumour altogether because the tumour sticks to surrounding structures. If some tumour cells remain, you may need radiotherapy or chemotherapy.

Sometimes, you may need a cerebrospinal fluid shunt to drain the fluid before surgery or if the tumour regrows. Your neurosurgeon can talk to you more about this.

Radiotherapy - Radiotherapy is often needed after surgery – especially if there is residual tumour after surgery. Radiotherapy uses X-rays and protons to kill tumour cells.

Chemotherapy - Chemotherapy is a drug treatment that destroys tumour cells.

Medication - Surgery may cause new hormonal imbalances or hypopituitarism. After surgery, you may need medication replacing cortisol, growth hormone, thyroid hormone and sex hormones. It is important to have regular checks with your endocrinologist on hormone levels following surgery.

ONGOING MANAGEMENT

After the surgery, you will need long-term follow-up to:

- **Monitor for tumour regrowth** – especially important in the first three years after surgery
- **Treat side effects** – related to pituitary function, such as pituitary hormone deficiencies and Arginine Vasopressin Deficiency (AVP-D)
- **Monitor emotional health** – surgery can affect the person's emotions and cause depression
- **Monitor steroid therapy** – and how to adjust the dose when stressed or ill.

It is common for the tumour to reoccur; however, it is less likely if the person has surgery and radiotherapy.

When to go to the hospital

Seek urgent medical care if:

- **Your vision gets worse or becomes impaired** – this can indicate that cysts are enlarging rapidly
- **You notice clear fluid dripping down the back of the throat or through the nose soon after surgery** – this may indicate a cerebrospinal fluid leak.

COMMON QUESTIONS

Is it cancer?

No. A craniopharyngioma is a rare benign (not cancerous) brain tumour. These tumours rarely spread to other parts of the body.

What causes craniopharyngiomas?

These tumours likely grow from leftover pieces of tissue that remain in early pregnancy when the baby's head, face and brain are forming.

Who gets craniopharyngiomas?

They can affect people of any age. Cases occur most commonly in children aged 5 to 14 but also adults between 50-70.

Are there any new treatment options?

You or your child may be a candidate for a clinical trial. Talk to your doctor about the options.

Are they inherited?

We still don't know. Research to find a genetic cause is ongoing.

MORE INFORMATION

The Australian Pituitary Foundation provides social support for patients and carers, and has published a range of patient resources on pituitary conditions and treatments.

For more information, please visit our website: www.pituitary.asn.au

Email: support@pituitary.asn.au

Phone: 1300 331 807

REFERENCES

1. Nielsen EH, Feldt-Rasmussen U, Poulsgaard L, et al. Incidence of craniopharyngioma in Denmark (n = 189) and estimated world incidence of craniopharyngioma in children and adults. J Neurooncol 2011; 104(3):755–763.
2. Garnett MR, Puget S, Grill J, Sainte- Rose C. Craniopharyngioma. Orphanet J Rare Dis 2007;2:18.

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