The ROYAL MARSDEN
NHS Foundation Trust

# Intracranial germ cell tumours

## Children's Unit

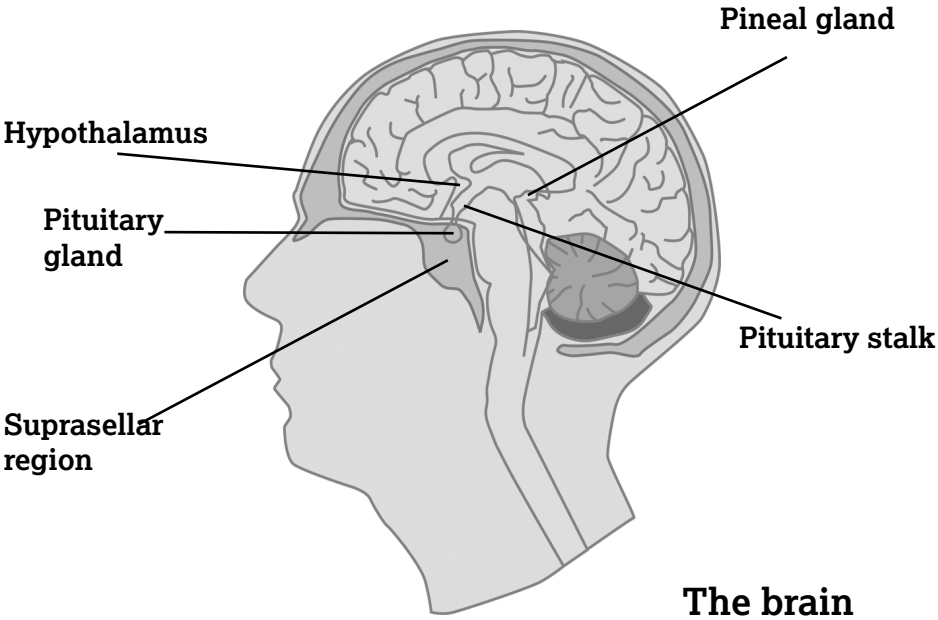
## Patient Information

NHS

## What is an intracranial germ cell tumour?

An intracranial germ cell tumour is a malignant (cancerous) tumour formed from early (primitive) germ cells (the reproductive cells of the body). These tumours mostly develop in one of two areas; around the pituitary gland and the pineal gland. They often secrete (release) chemical substances into the blood, called alpha-fetoprotein (AFP) and beta HCG (B-HCG). These can be used as tumour markers and may help with monitoring progress during your child's treatment.

There are two main types of intracranial germ cell tumour; non-secreting germinomas and secreting germ cell tumours. They can spread to the cerebro-spinal fluid (CSF) surrounding the brain and spine. Another type of germ cell tumour which can be either benign or malignant is known as a teratoma.




The brain

## Who gets an intracranial germ cell tumour?

These are very rare tumours. Fewer than 10 children a year develop an intracranial germ cell tumour in the UK. Young adults can also develop intracranial germ cell tumours but they are again very rare. These types of tumours occur more commonly in South East Asia (especially Japan) but the reason for this is not fully understood. Like most brain tumours, the cause of intracranial germ cell tumours is not known.

## What are the signs and symptoms?

Symptoms may be caused by the tumour blocking the flow of fluid surrounding the brain. This increases the pressure inside the brain. Symptoms may also happen as a result of direct pressure on or damage to the pituitary gland or nerves from the eyes. It also possible to develop symptoms (such as early puberty) related to the secretion of the tumour markers. Sometimes hormone deficiency can happen before the obvious development of a tumour. Symptoms may include:

- hormone disturbances
- headaches with nausea or vomiting
- disturbed sleep patterns
- visual disturbance
- behavioural changes
- slow growth
- increased sensitivity to cold or heat
- early or delayed puberty
- change in appetite and weight gain/loss.

If the tumour spreads to the spine:

- difficulty walking
- problems with bladder and bowel control.

## What tests (investigations) will be carried out?

We will need to carry out some tests to find out as much as possible about the type, position and size of the tumour. This will help us to decide on the best treatment for your child. These tests include:

- **CT scan** – it is likely a CT scan of your child's brain was the first specific test carried out at your local hospital. Although MRI scans are usually the best way of seeing the tumour and the effects of treatment, sometimes CT scans are also useful. For more information, please see The Royal Marsden leaflet *CT scan*.
- **MRI scan** – this scan allows us to see the brain and spine in great detail and is used regularly to diagnose and follow the effect treatment is having on your child's tumour. For more information, please see The Royal Marsden leaflet *MRI scan*.
- **Lumbar puncture (CSF testing)** – this is carried out under a general anaesthetic about 14 days after surgery. We look for any malignant cells in the CSF. At the same time, we can monitor the levels of the tumour's markers in the CSF.
- **Blood tests for tumour markers** – these are to find out if the tumour is secreting tumour markers.
- **Ophthalmic assessment** – a full assessment of your child's vision will need to be recorded at the time of diagnosis, during and after treatment. This is painless but sometimes difficult for young children.
- **Endocrine assessment** – if there is evidence of the tumour pressing the hypothalamus or pituitary gland, we will need a full test of your child's hormones. This usually involves simple blood tests.

### Staging

Staging is a measure of how far the tumour has spread beyond its original site. It is important to know if any tumour has spread to the spine through the CSF. The level of any secreted tumour

markers can tell us how quickly growing the tumour is and how it may respond to treatment.

## How do you treat an intracranial germ cell tumour?

The treatment depends on the type of tumour and its location and spread. Common treatments include:

### Surgery

Those children with secreting germ cell tumours can be diagnosed using brain scans and tumour marker studies. Children with all other types of intracranial germ cell tumours will have an operation to confirm the diagnosis and to relieve the pressure. If your child has a benign teratoma, surgery may be the main course of treatment. This may be followed by chemotherapy and/or radiotherapy and sometimes further surgery to remove any remaining tumour.

### Radiotherapy

Radiotherapy is commonly used after surgery to destroy any remaining cancer cells. This may be the only treatment needed for a germinoma. Radiotherapy is given to the tumour site. The brain and spinal cord are treated if cancer cells have been found in the CSF and spinal cord.

### Proton beam therapy

Children with intercranial germ cell tumours may benefit from receiving proton beam therapy. Proton beam therapy delivers radiotherapy in a way that may reduce the radiation to healthy tissue outside the area needing treatment, This can mean that children have fewer long-term side effects. In the UK, proton therapy is delivered at University College Hospital, London and The Christie Hospital, Manchester.

### Chemotherapy

Chemotherapy is given together with surgery and or radiotherapy to treat secreting germ cell tumours. Treatment with anti-cancer drugs is used to destroy the cancer cells.

It is usually given by injections and drips into a vein (intravenous infusion). The type of chemotherapy varies in intensity according to whether the tumour has spread, whether radiotherapy will be used and the age of the child. Some children may need high dose chemotherapy with a stem cell transplant. If your child takes part in a clinical trial, the treatment is explained in more detail in the specific trial information sheet.

## What are clinical trials?

Clinical trials are medical research trials involving patients that are carried out to try and find new and better treatments. In cancer, clinical trials are most commonly used to try and improve different forms of treatment such as surgery, radiotherapy or chemotherapy.

The treatment being tested may be aimed at:

- improving the number of people cured (for example, trying new types of surgery or chemotherapy)
- improving survival
- relieving the symptoms of the cancer
- relieving the side effects of treatment
- improving the quality of life or sense of wellbeing for people with cancer.

Clinical trials may also involve research aimed at understanding more about the tumour's biology. You may be asked to allow us to do research on the tumour sample removed at surgery, or on blood samples.

Well-run clinical trials have lead to a significant improvement in the treatment of children with cancer. You can find more general information on clinical trials in a separate information sheet. If you are asked to consider entering your child into a clinical trial, you will be given specific information about the trial

before deciding whether to take part. There is no obligation to participate and your child's care will not be affected should you choose not to take part in a clinical trial.

## General side effects of chemotherapy

- **Bone marrow suppression (myelosuppression)** – chemotherapy drugs decrease the production of blood cells by the bone marrow for a variable period of time. This results in low red blood cells (anaemia), low white blood cells (neutropenia) and low platelets (thrombocytopenia). Your child may need blood or platelet transfusions and will be at increased risk of infections. The doctors and nurses caring for your child will tell you more about these side effects.
- **Nausea and vomiting** – some of the chemotherapy drugs used may make your child feel sick or vomit. We will give anti-sickness drugs at the same time to stop nausea and vomiting which are usually very effective.
- **Sore mouth (mucositis)** – some of the chemotherapy drugs make the lining of the mouth and throat very sore and ulcerated. We will give your child painkillers for this, and explain how to care for your child's mouth during treatment.
- **Hair loss** – temporary hair loss is common. Please see the Macmillan leaflet *Coping with hair loss* for further information.

## Chemotherapy drugs

For details of the side effects of individual drugs, please visit the Macmillan website which provides individual drug information factsheets. You can access this via *www.macmillan.org.uk/*

## What is the outlook (prognosis)?

The prognosis for germinomas is excellent in most cases with radiotherapy or a combination of radiotherapy and chemotherapy. Significant progress has been made in the management of children with secreting germ cell tumours

over the last 20 years. The use of combined chemotherapy and radiotherapy has dramatically increased the survival rate. However, some patients still do not respond or they relapse. The use of surgery to remove residual tumour and possible high dose chemotherapy is being explored in clinical trials. If a teratoma is benign and can be removed, then the outlook is good. Malignant teratomas have a similar outlook to secreting germ cell tumours.

## What are the possible long-term effects?

Germ cell tumours have many potential serious effects on a child and young person's health and development. These include growth, hormonal, behavioural changes and learning difficulties. Visual disturbances can result from both the tumour and the treatment, and may be severe. A specialist hormone doctor (endocrinologist) will need to be involved in the long-term care of your child to replace hormones that are deficient (either in the form of tablets or injections). Your child may need urgent medical attention if they become unwell as they may not be able to control fluid balance and blood pressure normally. This can be very difficult to control and needs the help of more than one health professional. Therefore all children are followed up regularly after treatment is finished, in the 'long-term brain tumour follow up clinic', indefinitely, so that we can help with any long-term effects of the treatment.

## Other sources of information and support

General information on chemotherapy, hair loss and side effects is available from the PALS Help Centre, via *www.royalmarsden.nhs.uk* and in the 'parent held record'.

**Macmillan Cancer Support**

Macmillan Support Line: **0808 808 0000**

Website: *www.macmillan.org.uk*

Macmillan provides a range of free information and support on all aspects of cancer including:

- advice on benefits and other kinds of financial support
- information on local cancer support groups and organisations near you.

**The Children's Cancer and Leukaemia Group (CCLG)**

Website: *www.cclg.org.uk*

CCLG is a national professional body responsible for the organisation of the treatment and management of children with cancer in the UK. They coordinate national and international clinical trials. They also provide information for patients and families and produce a quarterly magazine called Contact.

**The Brain Tumour Charity**

Website: *www.thebraintumourcharity.org*

Support line: **0808 800 0004**

The Brain Tumour Charity is a UK charity, committed to fighting brain tumours. They fund scientific and clinical research into brain tumours and offer support and information to those affected, whilst raising awareness and influencing policy.

**Young Lives vs Cancer**

Website: *www.younglivesvscancer.org.uk*

Support line: **0300 303 5220**

Young Lives vs Cancer is a UK charity for children, young people, and their families, which provides clinical, practical, financial and emotional support to help them cope with cancer.

## Contact details

Children and Young People's Unit

24 hour Helpline: **020 8915 6248**

## References

This booklet is evidence based wherever the appropriate evidence is available, and represents an accumulation of expert opinion and professional interpretation.

Details of the references used in writing this booklet are available on request from:

The Royal Marsden Help Centre

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Sutton 020 8661 3759 / 3951

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Should you require information in an alternative format, please contact The Royal Marsden Help Centre.

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