Brainstem Gliomas

This information sheet is designed to be used with the accompanying booklet *Brain and Spinal Tumours in Children and Young People – A Parent's Guide*. You will find some references to the Guide in this factsheet. You may also find it helpful to read the CCLG/Cancerbackup booklet *A Parent's Guide to Children's Cancers* which provides more information about cancers in children, their diagnosis and treatment and the support services available. You may also want to discuss this information with a nurse or doctor involved in the treatment of your child.

What is a brainstem glioma?

Brainstem gliomas account for 10 to 15% of all brain tumours in children, with 30 to 40 new cases seen in the UK each year. These tumours can develop at any time but the most common ages are between 5 and 8 years of age.

The brainstem is located at the base of the brain and plays an essential role within the central nervous system (see diagram page 37 and glossary page 39). This area is the main communication centre for sending information from the brain to the body and vice versa. Every movement and sensation of the body is transmitted through the brainstem. The brainstem also contains the central part of many of the cranial nerves controlling the movement and sensitivity of the eyes, the face, hearing, steadiness, and taste, for example. The brainstem is also responsible for basic life functions such as blood pressure and breathing. All these functions are concentrated in a very small area, and even a small amount of damage to the structure of the brainstem may lead to severe problems.

Most of the tumours arising in the brainstem are GLIOMAS. This is why doctors use either the term brainstem tumour or brainstem glioma. The commonest type of brainstem tumours are "infiltrative/diffuse" (most often called diffuse pontine glioma), meaning that abnormal cells are growing in between normal fibres and normal cells. This often means surgery is impossible and why sometimes even a small biopsy may be too difficult.

Some tumours, however, grow on the edge of the brainstem. These tumours are called exophytic and are less common than the infiltrative/diffuse form. They can be relatively shallow and therefore it is possible to surgically remove all or part of them with minimal problems.

What are the symptoms?

Brainstem gliomas commonly cause the following symptoms:

- squint
- problems with eye movements
- uncoordinated or clumsy walking
- one sided weakness
- speech difficulties
- swallowing difficulties
- headaches
- vomiting

How is it diagnosed?

When a child arrives at a hospital with clinical symptoms suggestive of a brainstem glioma, the MRI *(see glossary page 42)* is the most appropriate scan to give a diagnosis. If the brainstem tumour is diffuse usually no other tests will be needed before treatment since the diagnosis is almost certain.

What is the treatment for 'Exophytic' tumours?

Surgery is the main type of treatment. Radiotherapy may be given after surgery depending on what the individual tumour cells look like under the microscope (histology) at diagnosis, the position of the tumour and how much of the tumour it was possible to remove by surgery.

Radiotherapy may be particularly beneficial for low grade (slower growing) tumours. All treatment decisions will be discussed with you in detail by your doctor and your liaison nurse.

What is the treatment for 'Diffuse brainstem gliomas' ?

Radiotherapy is the only effective treatment in these particular tumours. This treatment is given just to the area of the tumour over a 6 to 7 week period. New radiotherapy techniques such as hyperfractionated radiotherapy (delivering a higher dose in 2 treatments per day) have not shown a benefit over classical radiotherapy (one treatment per day). All treatment decisions will be taken only after discussion with you and possibly your child.

Steroids Due to the pressure on the vital cells in the brainstem, children with brainstem tumours are often very ill at the time of presentation. It is common to prescribe steroids to help to relieve some of their symptoms, as they reduce any extra swelling around the tumour and so help to relieve pressure. Despite high dose steroids, some children can experience worsening of their symptoms during the first weeks of radiotherapy. This may be related to some extra brainstem swelling that can occur as tumour cells are destroyed by radiotherapy. Generally, the child's condition will improve as the course of radiotherapy continues and sometimes all symptoms disappeared by the end, or soon after, the end of treatment. As soon as this improvement occurs, doctors will be keen to reduce the dose of steroids as they too have side effects that might add to the child's problems if taken for too long. This is possible in most cases but some children can become "steroid-dependent" which means that their symptoms come back when the dose of steroids is reduced below a certain level. These children need careful management by their consultant and community nursing team.

Chemotherapy For diffuse brainstem glioma, studies have shown no convincing evidence that chemotherapy will be helpful. However, there are ongoing trials on selected chemotherapies and some new drugs that have been designed specifically to attack these tumour cells.

In exophytic tumours some improvement with chemotherapy has been reported, mainly in young children. Chemotherapy might be considered in these cases.

What is the outlook for diffuse brainstem gliomas?

Regrettably, even after all of the original symptoms have disappeared, there is very little chance of cure for children with diffuse brainstem gliomas. In most cases, the symptoms come back or get worse (progress) after a symptom free time usually measured in months not years. The MRI will be of limited use to show tumour progression. Monitoring of this is now mainly based on how the child is feeling and behaving.

When progression occurs steroids may once again be used to control new symptoms. This use of steroids is one of the most complex problems in caring for children with this disease. Daily doses are likely to cause side effects including increased appetite, weight gain, sleeping disorders, irritability and skin and bone problems. To reduce these side effects doctors use high doses of steroids given in pulses (short bursts) for a few days at a time only.

What is the outlook for exophytic tumours?

This varies considerably from child to child. The outlook very much depends on firstly the amount of tumour that it was possible to remove and the response to radiotherapy, if it was given. Secondly, it can depend on what grade the tumour was or rather what the cells looked like down the microscope at diagnosis. In general, for low grade tumours with surgery and possible radiotherapy, the prognosis is hard to predict but with any high grade tumour the prognosis is as poor as for the diffuse form.

Who has produced this information?

This factsheet has been produced by the Children's Cancer and Leukaemia Group. The factsheets and accompanying booklet *Brain and Spinal Tumours in Children and Young People, A Parent's Guide* have been sponsored by a donation from the Samantha Dickson Brain Tumour Trust

Children's Cancer and Leukaemia Group (CCLG)

An organisation for professionals treating children with cancer, including coordination of clinical trials. The CCLG provides a range of information for patients and families affected by childhood cancer.

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Samantha Dickson Brain Tumour Trust

The Samantha Dickson Brain Tumour Trust raises funds for research and offers a vital support link for patients diagnosed with a brain tumour and their families.

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