Liver tumours

Approximately 10 children in the UK develop primary liver tumours each year. Boys are affected more commonly than girls. Liver tumours can be non-cancerous (benign) or cancerous (malignant). This factsheet is about malignant liver tumours.

There are two main types of primary liver cancer:

**Hepatoblastoma**
This usually occurs in children under five years of age. About eight children in the UK are diagnosed with hepatoblastoma each year.

**Hepatocellular carcinoma (HCC)**
This is rarer and usually occurs in older children. About two children in the UK are diagnosed with hepatocellular carcinoma each year.

More children than ever are surviving childhood cancer. There are new and better drugs and treatments, and we can now also work to reduce the after-effects of having had cancer in the past.

It is devastating to hear that your child has cancer, and at times it can feel overwhelming but there are many healthcare professionals and support organisations to help you through this difficult time.

Understanding more about the cancer your child has and the treatments that may be used can often help parents to cope. We hope you find the information here helpful. Your child's specialist will give you more detailed information and if you have any questions it is important to ask the specialist doctor or nurse who knows your child's individual situation.

This factsheet has been published in conjunction with CCLG's booklet entitled: ‘Children and Young People with Cancer: A Parent’s Guide’.

The liver
The liver is the largest organ in the body. It is surrounded by a fibrous capsule and is divided into sections called lobes. It lies across the upper part of the abdomen and is larger on the right hand side of the body than on the left. It’s surrounded, and protected from injury, by the lower ribs.

The liver is an extremely important organ that has many functions. One of these functions is to produce proteins that circulate in the blood. Some of the proteins help the blood to clot and prevent excessive bleeding. Others are essential for maintaining the balance of fluid in the body.

The liver also destroys harmful substances. It breaks down waste products not used by the body so that they can be passed out in the urine or stools (bowel motions).

The liver is responsible for breaking down food containing carbohydrates (sugars) and fats, so they can be used by the body for energy. It stores substances such as glucose and vitamins so that they can be used by the body when needed.

The liver also produces bile, a substance that breaks down the fats in food so that they can be absorbed from the bowel (intestine).

The liver is connected to the small intestine (duodenum) by a tube called the bile duct. This duct takes the bile produced by the liver to the intestine.
The liver has an amazing ability to repair itself. It can function normally even if only a small part of it is working.

**Causes**

The cause of most primary liver tumours in the western world is unknown. However, in other parts of the world, hepatocellular cancer (HCC) is commonly related to the presence of liver infection. This happens in countries where mothers commonly have hepatitis B and vaccination is not readily available at birth for their children.

Children who are infected with hepatitis B have a higher risk of developing HCC in late childhood than uninfected children.

**Signs and symptoms**

The most common symptom is a lump or swelling in the abdomen, which can be painful. Other possible symptoms include weight loss, loss of appetite, feeling sick (nausea) and being sick (vomiting).

**How liver tumours are diagnosed**

A variety of tests and investigations may be needed to diagnose a liver tumour.

An ultrasound scan and x-rays will be taken, which can show if there is a tumour in the liver. Further tests, including CT (computerised tomography) scans or MRI (magnetic resonance imaging) scans, will be done to find out the extent of the disease, both inside and outside the liver. Blood tests will also be carried out.

Most hepatoblastomas and hepatocellular carcinomas produce a protein that is released into the bloodstream. This protein is known as alpha-fetoprotein (AFP). It’s possible to measure levels of AFP in the blood, which can be a useful indicator of whether the liver tumour is responding to treatment or whether it may have come back after treatment. AFP is also known as a tumour marker.

Any tests and investigations that your child needs will be explained to you.

**Grouping**

To assess the extent of the cancer, a grouping system called PRETEXT (pre-treatment extent of disease) is used. It uses an MRI scan to measure the amount of cancer in the liver at diagnosis. It’s also used to help establish the best treatment for each child.

This grouping system is unique to liver tumours and is increasingly used all over the world.

This grouping process is essential because liver tumours need to be removed surgically. Grouping divides the liver into four surgical areas (sectors) and gives an indication of the kind of surgery that is needed to remove the tumour. Grouping also helps doctors decide whether a liver transplant surgeon needs to be involved from the start or not.

**PRETEXT 1**

One liver sector is affected and the tumour can be removed by straightforward surgery.

**PRETEXT 2**

Two adjoining sectors are affected and the tumour can be removed with more extensive surgery.

**PRETEXT 3**

Two or three sectors are affected with no two adjoining sectors free of disease, and the tumour can be removed with major surgery but sometimes may require a liver transplant.

**PRETEXT 4**

All four sectors of the liver are affected, and the tumour cannot usually be removed without replacing the liver with a donor liver transplant.

Doctors also look at the extent of the spread of cancer beyond the liver:

**In the blood vessels**

Sometimes the tumour gets into the blood vessels that are entering or leaving the liver. This may affect the type of surgery needed to remove the tumour.

**In the abdomen**

Sometimes the tumour spreads outside the liver and into the abdomen (tummy). The tumour cannot be removed completely.

**In the lungs or other organs**

If the tumour spreads outside the liver through the bloodstream (metastatic disease), it usually goes to the lungs. About 1 in 5 children are found to have affected lungs when they are diagnosed. Doctors use x-rays and CT or MRI scans to assess whether the lungs are affected.

**Treatment**

The diagnosis is made by taking a sample of cells from the tumour (biopsy). This is done under a general anaesthetic. Once the diagnosis and staging have been confirmed, plans for treatment will be made.
Hepatoblastoma

For hepatoblastoma, the type of treatment will depend on the PRETEXT staging and whether the cancer has spread to other parts of the body (metastatic disease).

<table>
<thead>
<tr>
<th>PRETEXT 1, 2 and 3 tumours are called ‘standard risk’.</th>
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<tr>
<td>PRETEXT 4 is called ‘high risk’ which also includes tumours that are pressing on the blood vessels in the liver.</td>
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<tr>
<td>METASTATIC tumours are those that have spread outside the liver, normally to the lungs.</td>
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Treatment is broadly similar for all PRETEXT stages. Chemotherapy (anti-cancer drugs) is given first. The aim of chemotherapy is to shrink the tumour in the liver and hopefully get rid of any metastatic tumours. Surgery to remove any remaining tumour will take place after a few weeks of chemotherapy. Further chemotherapy is usually given after surgery. Your child’s doctor will give you more detailed information about the chemotherapy drugs and their side effects.

Hepatocellular carcinoma

For hepatocellular carcinoma, treatment is a little different. The role of chemotherapy is less certain and surgery is the main treatment. Doctors usually recommend removing the tumour (if it’s small enough) at diagnosis. Chemotherapy may be given afterwards. However, in many situations the tumour is too large to remove at first and so chemotherapy is given to shrink the tumour so that surgery can be done later.

Hepatocellular tumours don’t always respond to chemotherapy as well as hepatoblastomas. For this reason, other treatments, such as chemoembolisation and targeted treatments (see below), may be used.

Chemoboembolisation

This refers to the giving of drugs directly into the artery going into the liver. Very occasionally, this may be used for hepatocellular cancer.

Targeted drugs

Targeted drugs are a group of treatments that work in a different way to conventional chemotherapy. Some drugs may cause cancer cells to die directly. Others act to cut off the blood supply to the tumour cells (antiangiogenesis drugs). Some drugs, called multi-targeted agents, work in both ways.

Research is looking into how useful targeted treatments are on their own and in combination with chemotherapy. Your child’s doctor may talk to you more about this research.

Surgery

All children, if they are able to, will have surgery following chemotherapy. If there’s cancer in the lungs, which hasn’t completely disappeared with the chemotherapy, the lungs will usually be operated on first. If the liver tumour can be surgically removed, the operation to do so will follow, usually a week or two later.

If the tumour involves all four sectors of the liver (PRETEXT 4), a liver transplant will probably be necessary. This is recommended for hepatoblastoma but only in particular circumstances for hepatocellular carcinoma. In a transplant, the whole liver is removed and replaced with a liver from another person.

This will be discussed with you from the beginning, and you will be given the opportunity to think about donating half of your liver, or for your child to have a liver from a donor. The transplant team will be there to answer all of your questions. A liver transplant is only possible if all the cancer outside the liver has gone.

Side effects of treatment

Treatment often causes side effects, and your child’s doctor will discuss these with you before treatment starts. Side effects can include:

- feeling sick (nausea) and being sick (vomiting)
- hair loss
- an increased risk of infection
- bruising and bleeding
- tiredness
- diarrhoea

Late side effects

The chemotherapy used to treat liver cancer can cause late side effects. These may include hearing problems, kidney problems and possibly heart problems. There will be a slightly increased risk of your child developing another type of cancer later in life. Most children will develop some late effects and need to have some follow-up tests. Your child’s doctor or nurse will tell you more about any possible late side effects.

Recurrence

If the cancer comes back after initial treatment, this is known as a recurrence. It can come back in the liver or in other parts of the body. If the cancer comes back, often (before anything is seen on scans) the levels of alpha-fetoprotein (AFP) in the child’s blood will start to rise again. Small rises in AFP can occur in the weeks after surgery, as the liver regenerates as much as it can.

Clinical trials

Many children have their treatment as part of a clinical research trial. Trials aim to improve our understanding of the best way to treat an illness, usually by comparing the standard treatment with a new or modified version.

Specialist doctors carry out trials for children’s cancer. If appropriate, your child’s medical team will talk to you about taking part in a clinical trial and will answer any questions you have. Written information is often provided to help explain things.
Taking part in a research trial is completely voluntary, and you’ll be given plenty of time to decide if it’s right for your child.

TREATMENT GUIDELINES

Sometimes, clinical trials are not available for your child’s tumour. This may be because a recent trial has just finished, or because the tumour is very rare. In these cases, you can expect your doctors and nurses to offer treatment which is agreed to be the most appropriate, using guidelines which have been prepared by experts across the country. The Children’s Cancer and Leukaemia Group (CCLG) is an important organisation which helps to produce these guidelines.

FOLLOW-UP CARE

When the treatment ends, your child will have regular blood tests to measure the level of AFP in the blood (if appropriate), as well as scans and chest x-rays.

More than three quarters of children with hepatoblastoma are cured, and for children with small tumours that are confined to the liver, the outlook is even better.

The outcome for hepatocellular carcinoma is not quite as good. The staff at the hospital can give you information about the likely outcome for your child.

If you have specific concerns about your child’s condition and treatment, it is best to discuss them with your child’s doctor, who knows your child’s condition in detail.

YOUR FEELINGS

As a parent, the fact that your child has cancer is one of the worst situations you can be faced with. You may have many emotions, such as fear, guilt, sadness, anger and uncertainty. These are all normal reactions and are part of the process that many parents go through at such a difficult time. It’s not possible to address in this factsheet all of the feelings you may have. However, the CCLG booklet ‘Children & Young People’s Cancer; A Parent’s Guide’, talks about the emotional impact of caring for a child with cancer and suggests sources of help and support.

Your child may have a variety of powerful emotions throughout their experience of cancer. The Parent’s Guide discusses these further and talks about how you can support your child.

USEFUL ORGANISATIONS

- **Children’s Cancer and Leukaemia Group (CCLG)**
  www.cclg.org.uk

- **Childhood Liver Tumours Strategy Group - SIOPEL**
  www.siopel.org

SIOPEL is an international group of medical specialists who promote research into childhood liver cancer. The website has a patients and families section.

- **CLIC Sargent**
  www.clicsargent.org.uk

CLIC Sargent offers practical support to children and young people with cancer or leukaemia, and to their families.

- **Macmillan Cancer Support**
  www.macmillan.org.uk

Offers support and advice to those affected by cancer.

With thanks to Dr Bruce Morland, Consultant Paediatric Oncologist at Birmingham Children’s Hospital and Chairman of the International Liver Cancer Group (SIOPEL), who reviewed this factsheet on behalf of the CCLG Publications Committee, comprising multiprofessional experts in the field of children’s cancer.

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If you have any comments on this factsheet, please contact us.

CCLG publications on a variety of topics related to children’s cancer are available to order or download free of charge from our website.

References: