



Liver tumours in children

An information guide for parents, carers and families

The purpose of this guide is to give information about liver tumours to help you understand more about the type of cancer your child has.

Information in this guide should be used to support professional advice specific to your child's diagnosis. If you have any questions, it is important to ask your child's medical team.

What are liver tumours?

Approximately 20 children in the UK develop a liver tumour each year. Liver tumours affect more boys than girls. Liver tumours can be non-cancerous (benign) or cancerous (malignant). This factsheet is about cancerous (malignant) liver tumours.

There are two main types of primary liver cancer:

- **Hepatoblastoma** - usually occurs in children under five years of age. About eight children in the UK are diagnosed with hepatoblastoma each year.
- **Hepatocellular carcinoma (HCC)** - usually occurs in older children. About two children in the UK are diagnosed with hepatocellular carcinoma each year.

The liver

The liver is the largest solid organ in the body and is divided into sections called lobes. It is in the upper part of the tummy and is larger on the right-hand side of the body than on the left. It is surrounded, and protected, by the lower ribs.

The liver is good at repairing itself. It can still do its job even if only a small part of it is working. The liver is a vital organ that has lots of important jobs:

- makes proteins to help prevent too much bleeding
- helps maintain the balance of fluid in the body
- breaks down harmful substances so that they can be passed out in wee (urine) or poo (stools)
- breaks down food and converts it to energy
- stores sugars and fats for when they are needed
- produces bile which breaks down the fats in food so that they can be absorbed by the body

Causes of liver tumours

We don't know what causes most liver tumours. In some parts of the world, hepatocellular carcinoma (HCC) is related to liver infection. This happens in countries where hepatitis B is common in pregnant mothers and vaccination is not readily available for children at birth. Children who are infected with hepatitis B have a higher risk of developing HCC than uninfected children. There is a link to increased rates of hepatoblastoma in premature and low birth weight babies. It is important to remember that nothing you have done has caused your child's cancer.

Signs and symptoms

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

How it is diagnosed

A variety of tests and investigations may be needed to diagnose a liver tumour. An ultrasound scan will be taken to show if there is a tumour in the liver. Further tests, including CT or MRI scans, of the tummy and chest, will be done to find out the extent of the disease. Blood tests will also be carried out. Most liver tumours produce a protein called alphafetoprotein (AFP). The levels of AFP in the blood can be measured and this 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 by your child's medical team.

Grouping

All liver tumours are assessed by using a grouping system called PRETEXT (pre-treatment extent of disease). It helps your child's doctors to decide which treatment is best for your child.

The grouping system is essential because liver tumours need to be removed surgically. The grouping divides the liver into four areas and helps decide the kind of surgery that is needed. Grouping also helps doctors decide whether a liver transplant surgeon needs to be involved from the start or not.

The grouping system

- **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 whether the cancer has spread beyond the liver:

- **In the blood vessels** – sometimes the tumour gets into the blood vessels which enter or leave the liver. This may affect the type of surgery needed.
- **In the tummy (abdomen)** – sometimes the tumour spreads outside the liver and into the tummy.

- **In the lungs or other organs** – if the tumour spreads outside the liver (metastatic disease), it usually goes to the lungs. About 1 in 5 children have tumour in the lungs when they are diagnosed. X-rays and CT scans are used to see whether the lungs are affected.

Treatment

Treatment depends on the type of liver cancer and the grouping of the tumour. Your child's doctor will discuss the treatment options with you.

Hepatoblastoma

Treatment will usually be a combination of chemotherapy and surgery. The exact timing of surgery and the intensity of chemotherapy is decided using a combination of 'risk' factors which includes grouping, whether the tumour has spread, AFP levels and the age of the child.

- **Very low risk** tumours are usually small and only in the liver. It may be safe to remove the tumour at diagnosis. Most children will need some chemotherapy after surgery.
- **Low risk** tumours are only in the liver but cannot be removed at diagnosis. Chemotherapy will be given to shrink the tumour before it is removed. More chemotherapy is usually given after surgery.
- **Intermediate risk** tumours have other risk factors but are usually only in the liver. Intense chemotherapy is given at the start. Many children will be able to have their tumour removed with standard surgery, some children will need a liver transplant to safely remove all of the tumour. More chemotherapy is usually given after surgery.
- **High risk** tumours have usually spread outside of the liver (metastases). Intense chemotherapy is given at the start to try and clear the disease that has spread outside the liver before having surgery or a transplant. If the cancer outside of the liver is not cleared after initial chemotherapy, treatment may be more challenging and more courses of chemotherapy may be needed.

Hepatocellular carcinoma

For hepatocellular carcinoma, surgery is the main treatment. If the tumour is small enough it will usually be removed at diagnosis. Chemotherapy may be given afterwards. If the tumour is too large to remove at first, or has spread, chemotherapy is given to shrink it so that surgery can be done later. Hepatocellular tumours don't always respond well to chemotherapy, so other treatments, such as chemoembolisation and targeted drugs, may be used.

Chemotherapy

Chemotherapy is the use of anti-cancer drugs to destroy cancer cells. It is usually given directly into a vein (intravenously).

Surgery

All children, if they are able to, will have surgery, either upfront (very low risk group), or after chemotherapy. If there's still cancer in the lungs, this will usually be operated on first. If the liver tumour can be surgically removed, this operation usually follows shortly after.

If the tumour involves all four sectors of the liver (pretext 4), a liver transplant will probably be needed. This is recommended for hepatoblastoma but only in some 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 to your child, or for your child to have a liver from a donor. A liver transplant is only possible if all the cancer outside the liver has gone. The transplant team will be able to answer all of your questions.

Chemoembolisation

Chemoembolisation is giving drugs directly into the artery going into the liver. This may be used for hepatocellular cancer.

Targeted drugs

Targeted drugs work differently to conventional chemotherapy. Some drugs may cause cancer cells to die directly. Others cut off the blood supply to the tumour cells (antiangiogenic drugs). Some drugs, called multi-targeted agents, work in both ways.

Side effects during treatment

Treatment can cause side effects, and your child's doctor will discuss these with you before the treatment starts. Any side effects will depend on the part of the body that's being treated and what treatment is being used.

Most side effects are short-term and gradually disappear once treatment stops. Side effects can include:

- feeling sick (nausea) and being sick (vomiting)
- temporary hair loss
- tiredness
- low blood count leading to an increased risk of infection and bruising and bleeding (sometimes blood and/or platelet transfusions are needed)
- sore mouth and tummy
- diarrhoea

Relapse

If the cancer comes back after initial treatment, 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 happen in the weeks after surgery, as the liver repairs itself. It can come back in the liver or in other parts of the body. Your child's doctor will discuss all the treatment options with you.

Clinical trials

Many children have their treatment as part of a clinical research trial or study. Trials and studies are carried out to try to improve our understanding of the best way to treat an illness. There are better results for curing children's cancers compared with just a few years ago because of clinical trials. Your child's medical team will talk to you about taking part in a clinical trial and will answer any questions you have. Taking part is completely voluntary, and you'll be given plenty of time to decide if it's right for your child. You may decide not to take part, or you can withdraw from a trial at any stage. Your child will still receive the best treatment available.

National treatment guidelines

Sometimes, clinical trials are not available for your child's tumour. In this case, your doctors will still offer the most appropriate treatment, using guidelines which have been agreed by experts across the UK. Children's Cancer and Leukaemia Group (CCLG) is an important organisation which helps to produce these guidelines.

Donating to a tissue bank

Liver tumours are rare and more research is needed to help doctors develop better treatment for the future. Your child's hospital team will offer you the opportunity to anonymously donate tissue left over from tests carried out, for example, a biopsy or bone marrow test, to the tissue bank. This sample of tissue can then be used by scientists to learn more about liver tumours and how best to treat them. This is voluntary, and you will have plenty of time to decide if you wish to take part.

Late side effects

Months or years later some children may develop late side effects from the treatment they have had. These may include a reduction in bone growth, hearing problems, a change in the way the heart, lungs and kidneys work, and a small increase in the risk of developing another cancer in later life. For more information please visit www.cclg.org.uk/life-after-childhood-cancer. Your child's doctor or nurse will talk to you about any possible late side effects and will keep a close eye on possible long-term side effects in follow-up clinics.

Follow-up care

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.

Once treatment has finished, the doctors will monitor your child closely with regular appointments to be sure that the cancer has not come back and there are no complications. 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. After a while you will not need to visit the clinic so often.

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

Your feelings

It's devastating to hear that your child has cancer and you may feel overwhelmed but there are many professionals and organisations to help you through this difficult time. You may have many feelings, such as fear, guilt, sadness, anger, and uncertainty. These are all normal and are part of the process that many parents go through.

It's not possible to address in this guide all of the feelings you may have. However, the CCLG booklet 'A parent's guide to children and young people with cancer', 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.



The CCLG booklet 'A parent's guide to children and young people with cancer' is available FREE of charge from your child's hospital

All CCLG booklets and leaflets can be downloaded or ordered from our website:

www.cclg.org.uk/publications

USEFUL ORGANISATIONS

Children's Cancer and Leukaemia Group (CCLG) publishes a variety of free resources to order or download.

www.cclg.org.uk

Young Lives vs Cancer offers practical support to children and young people with cancer and to their families

www.younglivesvscancer.org.uk

Macmillan Cancer Support offers support and advice to those affected by cancer.

www.macmillan.org.uk

Childhood Liver Tumours Strategy Group - SIOPEL

International group of medical specialists who promote research into childhood liver cancer.

www.siopep.org



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Children's Cancer and Leukaemia Group (CCLG) is a leading national charity and expert voice for all childhood cancers.

Each week in the UK and Ireland, more than 30 children are diagnosed with cancer. Our network of dedicated professional members work together in treatment, care and research to help shape a future where all children with cancer survive and live happy, healthy and independent lives.

We fund and support innovative world-class research and collaborate, both nationally and internationally, to drive forward improvements in childhood cancer. Our award-winning information resources help lessen the anxiety, stress and loneliness commonly felt by families, giving support throughout the cancer journey.

Our work is funded by donations. If you would like to help, text 'CCLG' to 70300 to donate £3. This will cost £3 plus a standard rate message.

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