My child’s Wilms’ tumour has come back
Treatment for relapsed Wilms’ tumour: information for parents

It can be devastating to hear your child’s cancer has returned, but there are treatments available for them if this does happen.

What is relapse?
Most children diagnosed with Wilms’ tumour are treated successfully with surgery, chemotherapy and sometimes radiotherapy but in a small number of children, their cancer comes back\(^1\). Usually this happens after a period of time during which the cancer could not be detected. This is known as relapse or tumour recurrence. Sometimes the tumour starts to grow again before it has been removed. This is known as tumour progression. Most relapses (about 85%) occur within two years of diagnosis of the original Wilms’ tumour, but for some children it can come back later than this. For children who relapse, their treatment will depend on what treatment they have already had and what their cancer looks like under the microscope (histology)\(^1\).

How many children with Wilms’ tumour relapse?
In the UK only about 5 to 10 children with Wilms’ tumour relapse each year\(^2\). The chance of relapse is greater in children whose original Wilms’ tumour had already spread (stage 4) or had ‘high risk’ histology.

Out of all childhood cancers, kidney cancers account for around 5% approximately 90% of childhood kidney cancers are Wilms’ tumours, being the most common kidney cancer in children.

As with most childhood cancers, the causes of Wilms’ tumour are unknown.
My child’s Wilms’ tumour has come back

Why and where does the tumour come back?

Wilms’ tumours consist of billions of cancer cells. The first operation your child has removes most of the cancer cells, then treatment with chemotherapy and radiotherapy kill any remaining cancer cells.

However, sometimes there are cancer cells that are not killed by the treatment. These are known as resistant cancer cells and even though the Wilms’ tumour appears to be gone, undetectable resistant cells can remain and start to grow again.

The original Wilms’ tumour can spread to other parts of the body through a process known as metastasis. This is where undetectable cancer cells of the tumour travel and spread to other parts of the body through the blood system.

The most common site of metastatic relapse is in the lungs (80%), followed by the liver (15%), and then bone or brain (less than 5%).

Local relapse is when the tumour grows back in the abdomen, usually near the original tumour site.

Metastatic and local relapse can occur at the same time.

Metachronous relapse is when Wilms’ tumour appears in the other kidney. It is thought that this is a new Wilms’ tumour rather than the original tumour returning.

Hearing the news Alice’s cancer was back was devastating and in many ways was worse than the initial diagnosis. Whilst we understood the treatment process better, we also knew what she had to face. Alice has almost finished her treatment and then hopefully this battle will be over once and for all.

Jane, mum to Alice, diagnosed with relapsed Wilms’ tumour in 2015
What influences the success of treatment for relapsed Wilms’ tumour?

How your child’s relapsed Wilms’ tumour responds to treatment can depend on a number of different factors:

1. The original **tumour stage** and how it looked under the microscope (**histology**)
   Children with ‘high risk’ histology (anaplastic or blastemal type Wilms’ tumour) or more advanced tumour stage (stages 3 and 4) are more difficult to treat at relapse[1]

2. How many chemotherapy drugs were used to treat your child’s original Wilms’ tumour
   Children treated initially with two drugs do better than children treated initially with three or more drugs (including doxorubicin)[1]

3. If radiotherapy was used to treat your child’s original tumour
   If relapse occurs in an area where radiotherapy was used to treat your child’s tumour the relapse may be more difficult to eradicate[3]

The factors listed above are used to decide your child’s ‘risk group’ and help doctors to decide the best treatment for your child. Studies show that the time from initial diagnosis to relapse, and the site of recurrence, do not influence the success of treatment for relapse.

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**What are the risk groups and what does this mean?**

There are three risk groups that are used to help doctors decide the best treatment for your child and predict the chance of successful treatment.

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<th>Risk Group</th>
<th>Reason</th>
<th>Proportion of children predicted to be successfully treated</th>
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| **Standard** | • Initially received vincristine with or without actinomycin D only. No radiotherapy.  
• Usually the original tumour was low stage and favourable histology | 70%                                                       |
| **High**    | • Doxorubicin already given and/or radiotherapy already given  
• Usually the original tumour was stage 1 with high risk histology, or stage 2/3 with intermediate risk histology. Some stage 4 tumours are also in this group. | 50%                                                       |
| **Very High**| • Four or more chemotherapy drugs already used  
• The original tumour had high risk histology | 10%                                                       |
What treatment is my child likely to have?

The treatment your child receives will depend on which risk category your child is in (see previous page) and how their original tumour was treated.

The information provided in this factsheet is based on current UK treatment strategies for relapsed Wilms’ tumours. These strategies are based on internationally recognised treatments by the International Society of Paediatric Oncology (SIOP) Renal Tumour Study Group.

Generally, treatment for relapsed Wilms’ tumours will include chemotherapy drugs that have not already been used to treat the original tumour. However, in some cases of metachronous relapse, the same drugs can be used to successfully treat the ‘new’ tumour.

Treatment for standard risk relapsed Wilms’ tumour

The recommendation for children with standard risk relapsed Wilms’ tumour is to be treated with chemotherapy (doxorubicin, etoposide or cyclophosphamide), surgery if appropriate and radiotherapy. This treatment is successful in about 70% of children with standard risk relapsed Wilms’ tumour[1].

Treatment for high risk relapsed Wilms’ tumour

Children with high risk relapsed Wilms’ tumour, are treated with more intensive chemotherapy (etoposide, cyclophosphamide and carboplatin) followed by high-dose chemotherapy (melphalan) with autologous stem cell transplant (see page 5). Surgery may be necessary before the high-dose therapy. Radiotherapy is usually given afterwards[4].

Only a few hundred children have been treated with high dose chemotherapy and autologous stem cell transplant. The current evidence suggests that this treatment can be successful for around 66% of children[3].

The chemotherapy drugs topotecan or irinotecan/vincristine may also be considered as a treatment option for this group of children[1].

In the UK only 5 to 10 children with Wilms’ tumour relapse each year[2].

The chance of relapse is greater in children whose original Wilms’ tumour had already spread (stage 4) or had ‘high risk’ histology.
Treatment for very high risk relapsed Wilms’ tumour

This risk group is very challenging to treat as the tumour is likely to have developed resistance to treatment and no longer responds to available chemotherapy drugs. If your child is in the very high risk group they may have their treatment as part of a clinical trial which investigates new agents or new combinations of treatments to treat relapsed Wilms’ tumours[^1].
New therapies and clinical trials
Many children have their treatment as part of a clinical trial. Clinical trials aim to improve our understanding of the best way to treat an illness. The trial might examine a new treatment or a new combination of existing treatments. 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 will be provided to help explain things. Taking part in a research trial is completely voluntary, and you will be given plenty of time to decide if it’s right for your child.
**Glossary**

**Anaplastic Wilms’ tumour**
About 5-10% of Wilms’ tumours have an appearance called anaplasia, which means the cells look very disorganised under a microscope. Considered to be a ‘higher risk’ type of Wilms’ tumour.

**Blastemal-type Wilms’ tumour**
This group of high-risk tumours cannot be identified by looking at the biopsy because they occur when a particular type of early kidney cell survives the pre-surgery chemotherapy. These cells are known as blastemal cells. Tumours where most of these cells survive chemotherapy are called blastemal tumours.

**Chemotherapy**
Drug treatment that kills cancer cells.

**Histology**
The appearance of tissue under a microscope, which helps to determine the diagnosis.

**Local relapse**
When the tumour grows back in the abdomen, usually near the original tumour site.

**Metachronous relapse**
When Wilms’ tumour appears in the other kidney. It is thought that this is a new Wilms’ tumour rather than the original tumour returning.

**Metastasis**
Tumours that have spread from the first (primary) tumour into another part of the body. Also known as secondary tumours.

**Progression**
Where the tumour continues to grow, even during treatment.

**Radiotherapy**
The use of radiation to treat cancer.

**Relapse/recurrence**
The return of cancer after previous treatment.

**Staging**
Categorising the tumour, based on whether it has spread outside the kidney and whether it can still be removed during surgery, to help decide on a course of treatment.

**Surgery**
An operation.

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**Useful organisations**

**Children’s Cancer and Leukaemia Group (CCLG)**
www.cclg.org.uk
Children’s cancer charity and the UK and Ireland’s professional association for those involved in the treatment and care of children with cancer. Provides a range of information for patients and families affected by childhood cancer.

**Bethany’s Wish - Wilms Tumour Charity UK**
www.bethanyswish.co.uk
The UK’s Wilms’ tumour charity. Campaigns to raise awareness of Wilms’ tumour and childhood cancer, and provides each child in the UK diagnosed with Wilms’ tumour with a ‘B’s Bravery Bear Pack’ with special surprises.

**CLIC Sargent**
www.clicsargent.org.uk
Provides clinical, practical and emotional support for children and young people (and their families) to help them cope with cancer and get the most out of life.

**Macmillan Cancer Support**
www.macmillan.org.uk
A national cancer charity offering support to people with cancer and their families by providing information and practical support.

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**References**


