



The tumour takes its name from German surgeon Max Wilms who was active around the turn of the 20th century. Back then, surgery was the only available treatment, but surgery alone cured only a small number of children. It was soon realised that this kind of tumour was sensitive to radiation, and it subsequently proved to be highly sensitive to chemotherapy. Combinations of these types of treatment have led to a dramatic increase in survival. From being able to cure at best one child in ten through surgery alone, today we expect almost nine out of ten to recover.

#### SYMPTOMS

Wilms' tumour rarely has any very obvious symptoms. Often the parents themselves notice that their child has a painless lump or swelling on one side of the abdomen which prompts them to contact a doctor. In the absence of other symptoms, these tumours are therefore often very large by the time they are detected. Sometimes discovery of the tumour may be preceded by stomach pain with or without a tendency towards constipation. Less frequently, blood in the urine may lead to diagnosis.

#### DIAGNOSIS

Wilms' tumour is imaged most easily with an ultrasound scan of the abdomen. Sometimes there may be reason for a more detailed scan, in which case a CT scan may be performed. It

is important to examine both kidneys because the tumour sometimes appears on both sides. In around one in five children with Wilms' tumour, there will be detectable secondary tumours, or metastases, in the lungs at the time of diagnosis. It is therefore important to X-ray the lungs as well. Metastases in other organs, such as the liver and bones, are possible but very rare. Biopsies for microscopic tissue diagnostics are rarely warranted in the first instance.

#### TREATMENT

Only in exceptional cases will the treatment of a child with Wilms' tumour begin with an operation. The tumour is almost always pre-treated with chemotherapy for four to six weeks to shrink it and make surgery easier and less risky. The operation involves removing the kidney affected together with the tumour. If both kidneys are affected, there are alternative solutions. Lymph nodes around the kidney also need to be removed for analysis. The tumour is then examined under a microscope to assess the stage of the disease and the histological risk group. This information is used to plan the child's subsequent treatment with a view to maximising the chances of a cure while minimising the risk of lasting side-effects. This can involve anything from no further treatment at all to up to 34 weeks of chemotherapy. These days only a minority of children need radiotherapy, but this may sometimes still be required for a child to sur-

vive. In this case, local radiotherapy is performed in the area from which the affected kidney was removed.

#### FOLLOW-UP

Once treatment is complete, many years of follow-up begin. Initially these check-ups concentrate on picking up any recurrence of the disease, whereas subsequently the focus switches to detecting any late side-effects of the treatment given. These check-ups, which gradually become less frequent, normally consist of a general medical examination, an abdominal ultrasound and lung X-ray, and tests of kidney function. The treatment given to the majority of children with Wilms' tumour is relatively gentle, and so the risk of late side-effects is very small. Sometimes the treatment given can also warrant tests of heart function.

#### PROGNOSIS

These days almost nine children out of ten treated for the tumour recover. If the tumour has spread to the lungs, stronger treatment is required, but the prognosis is not notably different. What has more of an effect on the prognosis is the histological tumour type. Of children with Wilms' tumour with high-risk histology, only two out of three can be cured despite much more intensive treatment.

With few exceptions, children cured of Wilms' tumour can lead normal lives. Having only one kidney does not generally affect everyday life – some people are born with just one kidney and never know it.

*Factual accuracy verified by specialist Niklas Pal from the Childhood Cancer Unit at the Astrid Lindgren Children's Hospital in Stockholm*