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Wilms' Tumour

Synonym: nephroblastoma

Wilms' tumours are the most common intra-abdominal tumours of childhood. A Wilms' tumour is an undifferentiated mesodermal tumour of the intermediate cell mass (primitive renal tubules and mesenchymal cells). It may be sporadic or familial.

Epidemiology

- Wilms' tumour is the most common renal tumour of childhood and affects about 1 in 10,000 children^[1].
- The incidence is estimated at 7 in 1,000,000 children under 16 years of age^[2].
- Wilms' tumour is the second most common intra-abdominal cancer of childhood and the fifth most common paediatric malignancy overall (approximately 6% of all paediatric cancers)^[3].
- Only 3% of Wilms' tumours are reported in adults^[4]. Most adult patients are diagnosed unexpectedly following nephrectomy for presumed renal cell carcinoma^[5].
- In 5-10% of patients, both kidneys are affected at the same time (synchronous bilateral Wilms' tumour) or one after the other (metachronous bilateral Wilms' tumour)^[6].

Associations^[3]

Wilms' tumours usually develop in otherwise healthy children but approximately 10% occur in children with recognised malformations; either:

- 'Overgrowth syndromes' (excessive prenatal and postnatal somatic growth resulting in macroglossia, nephromegaly and hemihypertrophy) - most commonly, [Beckwith-Wiedemann syndrome](#) or isolated hemihypertrophy. Others include Perlman syndrome, Sotos' syndrome, and Simpson-Golabi-Behmel syndrome.
- No 'overgrowth' - associated with [Edwards syndrome \(trisomy 18\)](#), Bloom's syndrome, Denys-Drash syndrome; or WAGR (**W**ilms' with **A**niridia, **G**onadoblastoma (genitourinary malformations), and **R**etardation).

Familial Wilms' tumour^[7]

Hereditary Wilms' tumour (either bilateral tumours or a family history of the neoplasm) is uncommon. Several different families with Wilms' tumours have been identified. All are transmitted in an autosomal dominant manner, caused by mutations in one of at least three genes:

- One related to the WT1 gene on chromosome 11 (11p13) - (includes those patients with WAGR) - encodes a protein which is a transcriptional repressor downregulating IGF-II, an insulin-like growth factor.
- Other families (including those with Beckwith-Wiedemann syndrome) have a different mutation - of the WT2 gene on chromosome 11 (11p15.5).
- Other gene mutations, thought to be on chromosome 16 (WT3-16q) and/or chromosome 1p can also cause the tumour^[8].

Presentation

Usually presents in the first five years of life but 3% of presentations are in adults^[9].

95% are unilateral. Only 1-2% have a positive family history^[10].

Clinical features^[3]

- The most common presentation is an asymptomatic abdominal mass.
- Abdominal pain.
- Haematuria.
- Urinary tract infection.
- Hypertension, gross haematuria and fever may occur but are uncommon.
- Advanced disease may rarely present with respiratory symptoms due to lung metastases.

Screening

A recommendation for surveillance of children at high risk (familial or associated conditions) included^[11]:

- Surveillance should only be offered after review by a clinical geneticist.
- Surveillance should be carried out by renal ultrasound every 3-4 months.

- Surveillance should continue until 5 years of age in all conditions except Beckwith-Wiedemann syndrome, Simpson-Golabi-Behmel syndrome and some familial Wilms' tumour pedigrees, when it should continue until 7 years.

Investigations^[3]

- Useful laboratory tests include FBC, renal function and electrolytes and urinalysis.
- Genetic studies may reveal the chromosomal abnormalities consistent with the condition.
- Ultrasound and/or intravenous pyelogram (IVP) may show distortion of the renal pelvis; hydronephrosis. Dynamic imaging of the renal vein and inferior vena cava may be contributory.
- Renal angiography may help to show a more detailed view of the blood vessels.
- CT and MRI scanning may help to determine the nature of the tumour and may also reveal the degree of involvement of the lymph nodes, whether the other kidney is involved and invasion into blood vessels or the liver.
- Transcutaneous renal biopsy should be avoided, as this may make the condition worse.
- Chest CT to detect lung metastases.

Staging of Wilms' tumour ^[12]	
Stage I	Tumour limited to the kidney and completely excised. Renal capsule is intact. The tumour is not ruptured before or during removal. The vessels of the renal sinus are not involved. There is no residual tumour apparent beyond the margins of excision.
Stage II	Tumour extends beyond the kidney but is completely excised. No residual tumour is apparent at or beyond the margins of excision. There may be: <ul style="list-style-type: none"> • Regional extension of the tumour - ie penetration through the outer surface of the renal capsule into the perirenal soft tissue or more than 1-2 mm of tumour invasion into the renal sinus. • Vessels outside the kidney are infiltrated or contain tumour thrombus. • The tumour was biopsied or there was local spillage of tumour confined to the flank.
Stage III	There is residual tumour confined to the abdomen. There may be one or more of the following: <ul style="list-style-type: none"> • Tumour-positive lymph nodes in the renal hilus, the periaortic chains, or other intra-abdominal sites on biopsy. • There has been diffuse peritoneal contamination by the tumour - eg, spillage of tumour beyond the flank before or during surgery or by tumour growth penetrating through the peritoneal surface. • Implants are found on the peritoneal surfaces. • Tumour extends beyond the surgical margins, either microscopically or grossly. • Tumour is not completely resectable because of local infiltration into vital structures.
Stage IV	Haematogenous metastases - beyond stage III - eg, to the lung, liver, bone, or brain.
Stage V	Bilateral renal involvement at initial diagnosis. Attempt to stage each side according to the above criteria on the basis of extent of disease prior to biopsy. Four-year survival was 94% for those patients whose most advanced lesion was stage I-II; 76% where it was stage III.

Management^[6]

- For most patients, nephrectomy followed by chemotherapy (regimes include vincristine, dactinomycin and doxorubicin, sometimes with additional cyclophosphamide) can be curative.
- Routine postoperative radiotherapy to the flank is beneficial in patients with a stage III tumour.
- Patients with massive, nonresectable unilateral tumours, bilateral tumours, or venacaval tumour thrombus above the hepatic veins should be considered for pre-operative chemotherapy because of the risk of initial surgical resection.

Prognosis^[13]

- With treatment, over 90% of children diagnosed with Wilms' tumour survive long-term^[14].
- Long-term survival is above 75% for those with metastatic disease^[1].
- There is an increased risk of second tumours in survivors of Wilms' tumour. Second tumours include bone and soft tissue sarcomas, breast cancer, lymphoma, gastrointestinal tumours and melanoma. Acute leukaemias may also occur.

Further reading & references

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Author: Dr Colin Tidy	Peer Reviewer: Dr Anjum Gandhi	
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