Wilms Tumour

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Renal neoplasms in childhood are usually malignant, the most common being nephroblastoma Wilms tumour (WT). The incidence varies from 10.9 per million in the United States to 2.5 per million in China. A genetic predisposition exists, and nephrogenic cell clusters, (which are premalignant lesions), are found in one third of patients with WT. Some syndromes are associated with WT:

- WAGR (WT, Aniridia, Genitourinary anomalies, mental Retardation)
- Denys-Drash (WT, disorders of sexual development, nephropathy)
- Beckwith-Wiedemann (exomphalos, macroglossia, visceromegaly)
- Hemihypertrophy is associated with an increased incidence of WT

WT1 and *WT2* genes are associated with WT, especially *WT2* with Beckwith-Wiedemann syndrome.

53.1 Presentation

WT presents as a palpable, asymptomatic abdominal mass in a toddler. It is usually discovered when the parents are bathing or dressing the child. Weight loss, malaise, abdominal pain, hypertension, and haematuria may be present. Rarely, the patient may present with a varicocoele, where the left renal vein is occluded by tumour thrombus. On examination, the smooth, rounded tumour occupies most of the abdomen, and in about 10% of patients, the tumour thrombus from the nephroblastoma may invade the inferior vena cava. At times, it may extend to the right atrium, causing cardiac dysfunction or even a pulmonary embolus. Obstruction of the hepatic veins may rarely cause an acute hepatic encephalopathy and produce a Budd-Chiari syndrome. Patients with syndromes such as WAGR, Denys-Drash, Beckwith-Weidemann, or hemihypertrophy should receive regular ultrasound screening because they have an increased risk of developing WT.

53.2 Diagnosis

The mainstay of investigation is imaging. A plain x-ray of the abdomen usually shows a soft tissue mass, and calcification may be seen in about 10% of patients. A plain chest radiograph may show pulmonary metastases. Abdominal ultrasound confirms that the tumour is renal in origin and can demonstrate a normal contralateral kidney. It also evaluates the inferior vena cava for blood flow and for the presence of tumour thrombus. A CT scan will outline the tumour and may show a lesion in the contralateral kidney; it is more sensitive than the plain chest radiograph for the identification of pulmonary metastases. MRI can add a further dimension to renal evaluation, with visualisation of blood vessels. Echocardiography may be necessary to exclude the presence of an intra-atrial extension of the tumour thrombus.

The histology of WT mimics the development of the normal kidney with the proportion of the three components (blastema, tubules, and stroma) varying greatly in different tumours. Histologic differentiation allows good clinical correlation between a "low risk" group of patients who can be cured and a "high risk" group who need more intensive treatment and do less well.

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53.3 Treatment and Staging

The surgical treatment of Wilms tumour involves three stages: (1) making a diagnosis by biopsy; (2) operative excision of the tumour; and (3) staging of the patient.

53.3.1 Staging Systems

The staging system of the National Wilms' Tumor Study (NWTS) uses five stages:

- *Stage I*: Tumour limited to the kidney and completely excised. Surface of the renal capsule intact; no tumour rupture; no residual tumour apparent beyond margin of excision
- *Stage II:* Tumour extends beyond kidney but is completely excised; regional extension of tumour; vessel infiltration; tumour biopsy or local spillage or tumour confined to flank; no residual tumour apparent at or beyond margins of excision.
- *Stage III*: Residual non-hematogenous tumour confined to the abdomen, lymph node involvement of the hilum, periaortic chains or beyond; diffuse peritoneal contamination by tumour spillage; peritoneal implants; tumour extends beyond resection margins, either microscopically or macroscopically; tumour not completely resectable because of local infiltration into vital structures.
- *Stage IV*: Deposits beyond stage III in lung, liver, bone, or brain.
- *Stage V:* Bilateral renal involvement at diagnosis.

The SIOP (International Society of Pediatric Oncology) group subdivides Stage II into IIa, in which the lymph nodes are tumour-free, and IIb, with tumour-positive regional (hilar) lymph nodes.

53.3.2 Preoperative Chemotherapy

A variety of protocols have been devised that show benefit from preoperative chemotherapy once the diagnosis has been established. Thus, on suspicion of a Wilms tumour, preoperative chemotherapy may shrink the tumour, downgrade the staging, and reduce the incidence of intraoperative tumour rupture. Preoperative chemotherapy has been very effective in Europe, where the SIOP trials have demonstrated a reduced incidence of operative tumour rupture in patients who had received chemotherapy. The chemotherapy shrinks the tumour considerably in 80% of cases, and therefore makes the surgery safer (particularly for intravascular tumour thrombus). In the UK, patients currently may have percutaneous biopsies to obtain histologic confirmation of the tumour prior to starting preoperative chemotherapy. (This is an attempt to identify those histologic types that are likely to progress on chemotherapy and may benefit from expedited surgery.) Surgery is normally delayed for several weeks whilst neoadjuvant chemotherapy is administered according to the protocol.

53.3.3 Surgical Procedure

The operation performed for Wilms tumour is a transabdominal nephrectomy. It is essential to have all the necessary prerequisites done before surgery can start, such as a central venous catheter with central venous pressure monitoring, arterial line with continuous arterial pressure monitoring, urinary catheter, etc. Epidural block is part of the routine procedure, and cross-matched blood should be available if needed.

The abdomen and chest are prepared and draped. It is best to have the flank raised with a roll underneath the patient. The operative procedure begins with a large transverse incision (Fig. 53.1). The incision is carried out well into the flank on the involved side and across to the flank on the opposite

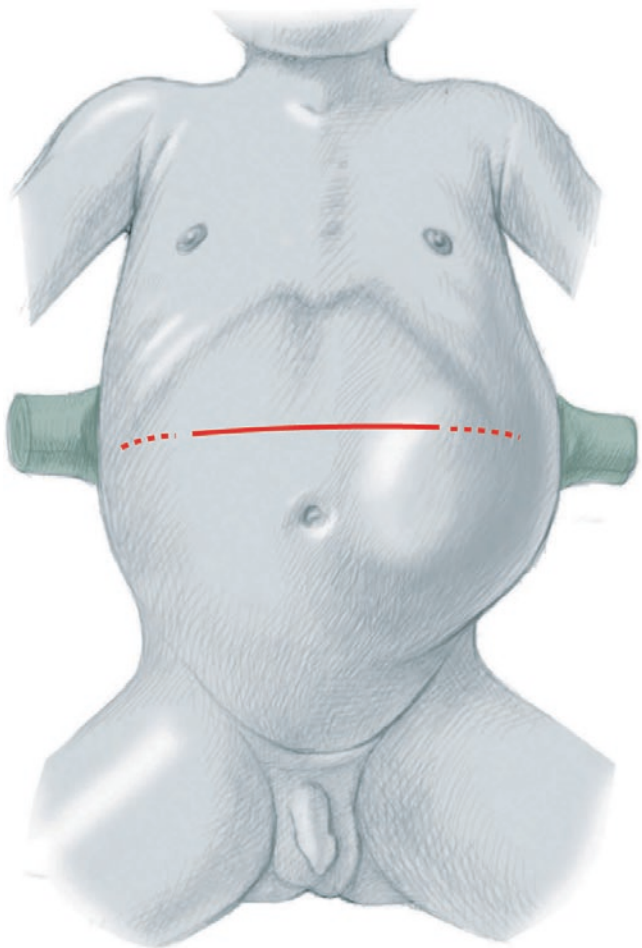


Fig. 53.1 Transverse incision for transabdominal nephrectomy

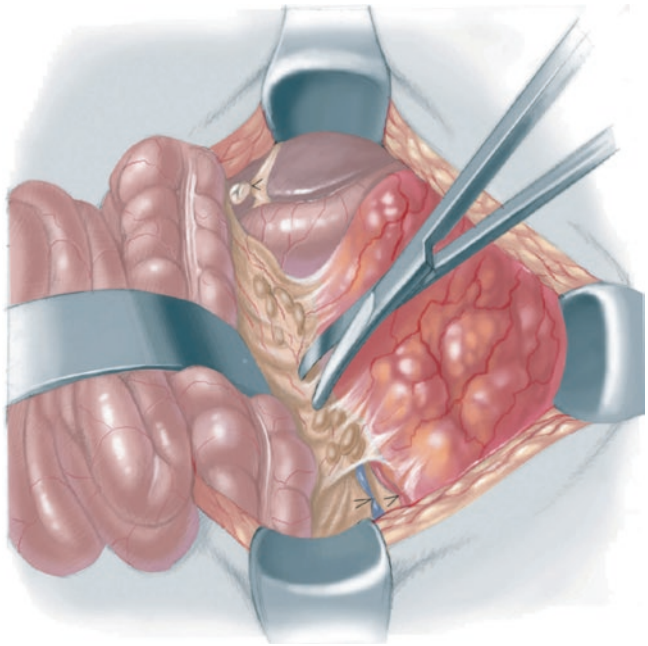


Fig. 53.2 Mobilizing the colon off the tumour

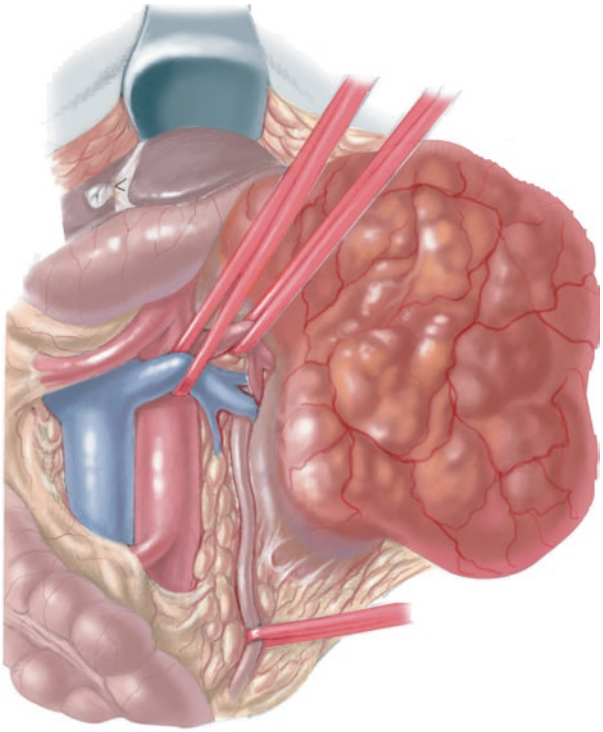


Fig. 53.3 Slings preventing embolization

side. The incision must be large enough to allow large tumours to be mobilized without risking intraoperative rupture. The incision is deepened through the subcutaneous fat and the rectus and oblique muscles.

The peritoneum is then opened and the ligamentum teres is divided between ligatures. Any free fluid (particularly if it is blood-stained) should be sent for cytology, and any peritoneal deposits should be excised and sent for histopathology. The small intestine is then delivered out of the abdomen and protected by warm, moist packs in order to assess the tumour. A full assessment should be made by palpating the tumour, the liver, lymph nodes, and the contralateral kidney (although preoperative imaging may be sufficient).

The retroperitoneal space is opened by an incision made lateral to the reflection of the peritoneum of the ascending colon for a right-sided tumour, or lateral to the descending colon for a left-sided tumour. The colon is mobilized off the tumour and reflected medially (Fig. 53.2). Occasionally the tumour invades the mesentery and the vessels; these need to be ligated and divided, preserving the marginal artery to avoid unnecessary resection of bowel.

A sling is passed around the renal vein or veins, and the contralateral renal vein is visualized. (Tumour may distort the normal anatomy.) A sling around these structures will prevent any risk of embolization when mobilizing the tumour. Sling the renal artery or arteries and the ureter (Fig. 53.3). Dissection of the hilum of the kidney before mobilization is not always possible, especially when the tumour crosses the midline or if large lymph nodes are in the way.

The renal vein and inferior vena cava should be palpated early for an assessment of whether an intravascular thrombus with tumour extension is present. Careful palpation of the contralateral vein is also indicated at this step of the surgical procedure. If a thrombus is detected in the renal vein, or the thrombus is extending into the vena cava, the appropriate vessels can be opened by a transverse incision between slings or after placement of vascular clamps (Fig. 53.4), and the thrombus can be removed with an open-ended suction cannula or a Fogarty balloon catheter. The vein is then closed with a 5/0 monofilament non-absorbable continuous running suture.

In cases without intravascular tumour involvement, the renal artery or arteries are transfixed and the ends are ligated. Double-ligate the renal veins with nonabsorbable suture material (Fig. 53.5). The renal artery (or arteries) should be ligated and divided before the renal vein (veins) to avoid excess congestion of the tumour. Lymph nodes from para-caval, para-aortic, supra-hilar, infra-hilar, mesenteric and bilateral iliac regions should be sampled (and carefully labeled) for staging. A sling is passed around the ureter at the pelvic brim and the gonadal vessels are usually divided (although may be preserved if tumour anatomy allows). The ureter should be isolated off the pelvic brim and transfixed as low down as possible using absorbable suture material.

The kidney is mobilized from the retroperitoneal space (Fig. 53.6). Large veins may need to be divided. Meticulous

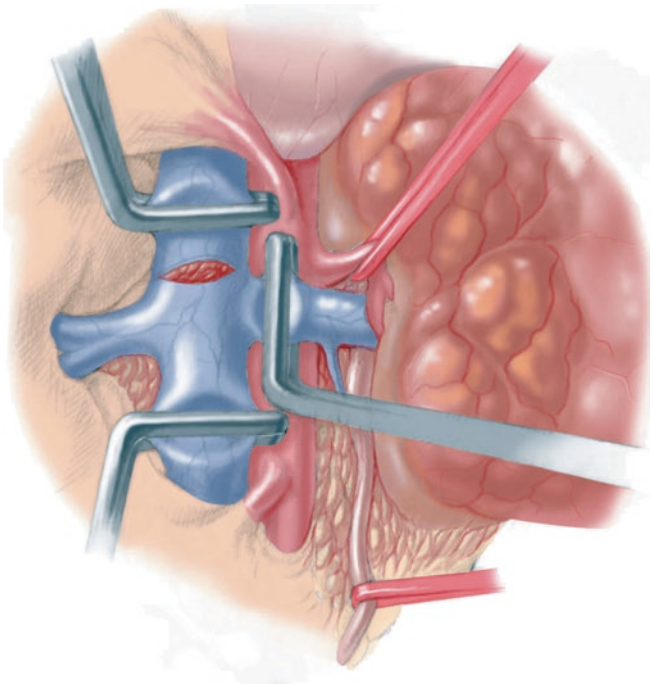


Fig. 53.4 Incision for removal of an intravascular thrombus

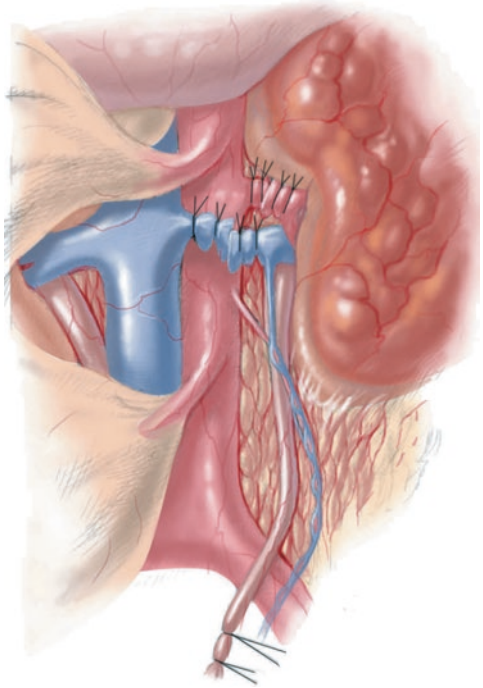
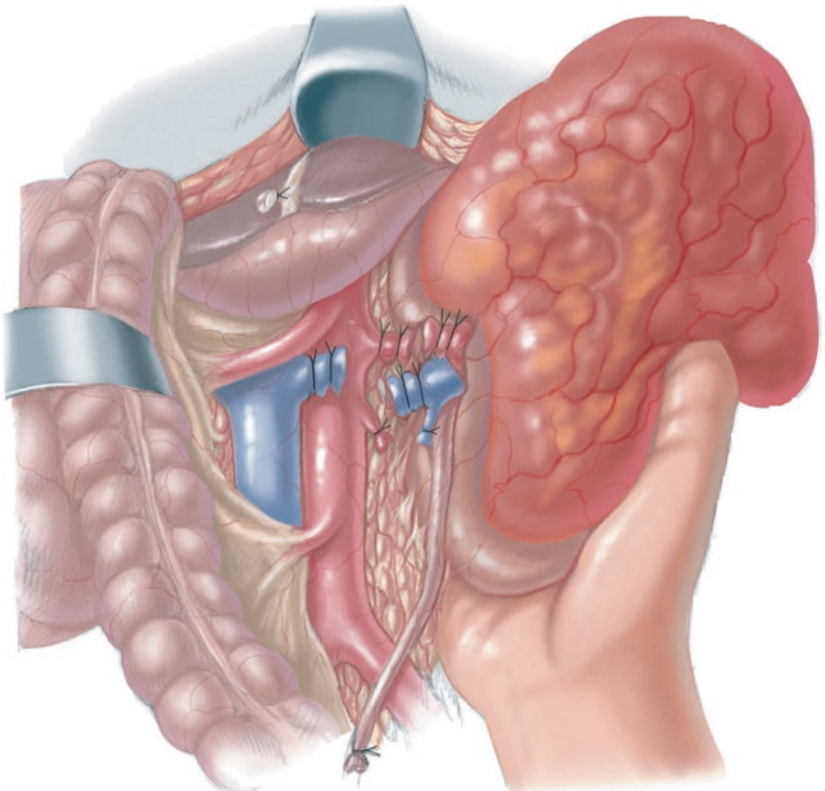


Fig. 53.5 Ligation of renal veins and arteries

Fig. 53.6 Removal of the tumour



dissection is needed. If the tumour is adherent to muscle or diaphragm, this needs to be removed as well. Finger dissection is useful for tissue planes.

The adrenal is removed in most cases (although preservation may be possible in the case of small, lower pole tumours). Any lymph nodes should be included in the mobilization and removed en bloc with the perinephric fat. These should be secured with double sutures and transfixed where appropriate, as in the case of the renal artery. Sometimes large veins in the perinephric fat must be ligated and divided. Direct infiltration of the posterior abdominal wall by the tumour can occur. Similarly, invasion of the diaphragm may require dissection of the muscle together with the tumour. The adjacent liver is usually adherent rather than actually invaded, but invasion of the diaphragm is common and sometimes a portion of the diaphragm must be removed.

It is important to recognize that large tumours may displace the aorta or the vena cava, and the tumour may actually grow behind these large vessels.

Accidental ligation of the aorta or the contralateral renal vein and vena cava as well as injury to the superior mesenteric vessels have been known in the past. To avoid any doubt about which type of blood vessel is being sacrificed, a loop around the vein or artery before ligating is essential. If the adrenal gland is adherent, it may be removed with the tumour. The tumour must be removed en bloc with the hilar lymph nodes and sent fresh to pathology.

The tumour bed should be inspected for haemostasis or residual tumour. Any suspicious area should be removed and sent for biopsy. The contralateral kidney must be carefully inspected for evidence of disease.

Haemostasis needs to be meticulous and the tumour bed should be dry before closure of the wound. Rarely, where there is uncontrolled bleeding, a pack may need to be left in and removed 48 h later. The abdomen is closed in layers, and no drain is left in situ.

53.4 Outcome and Follow-Up

The history of the treatment of patients with WT is one of the most impressive success stories in pediatric surgery. In 1941, W.E. Ladd reported a survival rate of 20%, but today the relapse-free survival is close to 90% in all patients, and even 66% in the histologically high-risk group. Study results of the NWTS (primary surgery) and the SIOP (primary chemotherapy, with or without tumour biopsy) are similar: the rate of complications and tumour rupture is significantly higher with the primary surgery strategy.

Long-term follow-up is important, to allow early detection of a possible metachronous tumour in the contralateral kidney. Synchronous bilateral tumours have an incidence of about 10%; after initial biopsy, chemotherapy should be instituted in these patients, and then partial nephrectomy carried out, as well as lumpectomies. This procedure has been found to be effective in the past. Biopsy only and chemotherapy is indicated in patients with diffuse nephroblastomatosis, but long-term follow-up is also crucial, because late WT occurrence is known.

53.5 Alternative Approaches

Experience with nephron-sparing surgery (NSS) in unilateral WT is increasing. This approach should be considered particularly for children with good response to preoperative chemotherapy who have a single kidney, syndromes predisposing to WT, or other nephrologic conditions. Ideally, for NSS to be feasible, the tumour should be polar, away from renal sinus/vessels and calyces. The aim is to leave two thirds of the kidney in situ. This approach may risk up-staging from stage I to stage III.

Some surgeons have undertaken minimally invasive surgery for WT (particularly for small tumours in the hilum). Careful case selection is vital, and this approach is not currently recommended by SIOP (especially if the alternative would be NSS, which would be preferred). Certainly, if this approach is considered, it is important for the patient to be enrolled in ongoing international studies so that the historical improvements in outcome for this tumour are not compromised by surgical approach.

Suggested Reading

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