

Recommendations for cross-sectional imaging in cancer management Paediatric neoplasms



Contents

1	Background	3
2	Wilms' tumour	4
3	Neuroblastoma	6
4	Rhabdomyosarcoma	9
References		

1 Background

While it is acknowledged that most children will have their initial imaging at a local non-specialist hospital, children's tumours are most appropriately imaged thereafter in centres where their treatment will be given.

Techniques and protocols should be according to patterns of tumour spread. An approach that obtains as much of the essential information as possible at a single investigation is required, particularly if general anaesthesia or sedation are to be used. Where feasible, this should also include non-imaging investigations, such as bone marrow biopsy. In paediatric oncology, the TNM classification is not used. Individual staging systems of common paediatric neoplasms (Wilms' tumour, neuroblastoma and rhabdomyosarcoma) are not included in this document, but it is noteworthy that the international neuroblastoma risk grouping (INRG) is incorporated into new international collaborative studies.^{1,7}

2 Wilms' tumour

Clinical background

Wilms' tumour is the most frequent renal tumour of childhood and typically presents in the first six years of life with a painless abdominal mass.^{2,3,8} Initial imaging is usually with ultrasound, and the differential diagnosis for a renal mass includes nephroblastomatosis, clear cell sarcoma, renal cell carcinoma, renal lymphoma, congenital mesoblastic nephroma and malignant rhabdoid tumour. Tumours may be sufficiently large that the organ of origin is not always clear, and computed tomography (CT) or magnetic resonance imaging (MRI) may be needed to discriminate between renal and adrenal mass lesions.

Who should be imaged?

All children presenting with an abdominal mass should be imaged initially with ultrasound. After ultrasound has confirmed the abdominal mass is a tumour, CT or ideally MRI is then performed, which can also aid biopsy.

Staging objectives

- To confirm the organ of origin.
- To assess tumour extent.
- To assess tumour extension into vessels.
- To detect local and regional lymph node involvement.
- To detect ipsilateral or contralateral renal tumour(s).
- To detect ipsilateral or contralateral nephroblastomatosis.
- To detect distant metastatic disease (for example, liver and lungs).

Staging

CT or MRI are used for assessment of abdominal disease. Ultrasound is the optimum method of assessing for tumour thrombus in the renal vein or inferior vena cava (IVC).

Use of sedation or general anaesthesia for CT and MRI depends on individual patient requirements and local circumstances.

With CT and MRI, intravenous (IV) contrast medium administration is mandatory to assess the primary tumour and contralateral kidney. MRI is preferred where possible to CT as it has higher sensitivity for nephroblastomatosis and no radiation burden.

СТ

Dosage of IV contrast medium at CT is 1 mI/0.5 kg patient body weight, with scanning at 65–70 seconds post-injection to allow opacification of the portal vein, renal veins and IVC.

Routine chest CT should also be undertaken to detect or exclude metastases.

Values of CTDI_{vol} should normally be below the relevant national reference dose for the region of scan and patient group (see chapter on radiation protection for the patient).

MRI

MRI technique is based on T2W and T1W spin-echo sequences in axial and coronal planes using a surface coil. Vascular supply and venous drainage may be assessed using angiographic sequences with or without IV contrast agents. Diffusion-weighted imaging can aid in the detection of multifocal tumours, lymph node and metastatic disease evaluation.⁸

A suggested protocol is as follows.

Protocol for imaging of Wilms' tumours

Coil	Sequence	Plane	Slice thickness	Field of view
To fit patient	T1W	Axial	4 ± 1 mm	To fit patient
To fit patient	T2W	Axial	4 ± 1 mm	To fit patient
To fit patient	T1W	Coronal	4 ± 1 mm	To fit patient
To fit patient	T2W	Coronal	4 ± 1 mm	To fit patient
To fit patient	MR angiography or venography 3D isovolumetric T2 sequence	3D coronal		To fit patient
To fit patient	T1W + IV contrast medium, if required	Axial/ coronal	4 ± 1 mm	To fit patient
To fit patient	Diffusion-weighted sequences (Multiple b values)	Axial	4 ± 1 mm	To fit patient

Partial nephrectomy and other forms of nephron-sparing surgery are sometimes considered, particularly in the presence of bilateral tumours. Under these circumstances, angiographic studies may be of benefit to the surgeon.

Follow-up

Neoadjuvant chemotherapy is used initially for Wilms' tumour. Serial measurement with ultrasound is undertaken, often with a CT scan or MRI prior to surgical excision. Over 80% of Wilms' tumour relapses occur within two years after surgery.⁴ Following removal of the tumour, three-monthly chest X-rays and ultrasound examinations are employed initially; MRI is not routinely employed in follow-up care except with bilateral tumours or known nephroblastomatosis.

Tips

- Post-contrast scans may demonstrate the normal renal cortex as a 'claw' around the tumour.
- In patients with large tumour masses, nodal disease may be difficult to distinguish from the primary tumour.

Clinical background

Sympathetic nervous system (SNS) tumours arise from cells of the embryonal neural crest, and there is a spectrum of disease that ranges from malignant undifferentiated neuroblastoma to well-differentiated ganglioneuroma. These tumours arise along the sympathetic neural axis, with the most frequent site being the adrenal glands. However, pelvic, thoracic and cervical neuroblastomas are also encountered. Peak age incidence is around two years.¹ Surgery alone is adequate treatment for localised neuroblastoma, but the majority of patients present with more widespread tumour. Children over the age of one with an abdominal primary tumour often have metastatic disease at presentation (approximately 75%).¹ Neuroblastoma diagnosed antenatally or in the first year of life behaves differently from tumours presenting later in childhood and has a good long-term prognosis.

Who should be imaged?

All patients with an abdominal mass or symptoms and signs suggestive of neuroblastoma should be imaged initially with abdominal ultrasound.^{1,7} Once the diagnosis of neuroblastoma is strongly suspected or confirmed, radionuclide imaging with metaiodobenzylguanidine (MIBG) scintigraphy including single-photon emission tomography-CT (SPECT-CT) where suitable should be undertaken in all patients. MR or CT is used to stage the primary tumour (which may be extra-abdominal).

Staging objectives

- To characterise primary tumour and define extent.
- To identify encasement of vessels.
- To identify imaging-defined risk factors such as encasement of the vessel.
- To identify extension of tumour into spinal canal ('dumbbell' tumour).
- To identify bone erosion by metastatic disease.
- To identify regional lymph node enlargement.
- To identify metastatic marrow or liver disease.

Staging

Ultrasound is frequently used as a first diagnostic investigation. CT or ideally MRI can be used for staging of neuroblastoma at diagnosis. Preoperative CT or MRI is likely to be needed for surgical planning. Preoperative CT may be preferred by the surgeon as a shrunken calcified retroperitoneal mass may be more easily visible at CT than at follow-up MRI. Radionuclide scanning with MIBG scintigraphy is performed routinely in all patients at diagnosis, and during follow-up in those with metastatic disease. A formal diagnosticquality CT scan with contrast may be performed in the nuclear medicine department at the same time as MIBG scintigraphy, in place of the low-dose non-contrast CT, especially at the post-induction chemotherapy, pre-surgical time point. This will be valuable for surgical and radiotherapy planning and reduces the need for two separate general anaesthesia procedures in young children.

MRI

Both MR and CT can be used but MR should be used where possible. This is usually governed by availability of equipment. MRI is superior at diagnosing intraspinal extension and metastatic disease in the bone marrow.

Protoco	l for	imaging	of neuro	blastoma
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Coil	Sequence	Plane	Slice thickness	Field of view
To fit patient	T1W	Axial	4 ± 1 mm	To fit patient
To fit patient	T2W	Axial	4 ± 1 mm	To fit patient
To fit patient	T1W	Coronal	4 ± 1 mm	To fit patient
To fit patient	T2W	Coronal	4±1mm	To fit patient
To fit patient	T1W + IV contrast medium, if required	Axial/ coronal	4 ± 1 mm	To fit patient
To fit patient	MR angiography or venography	3D acquisition		To fit patient or body part if clinically indicated
To fit patient	Diffusion-weighted sequences (Multiple b values)	Axial	4 ± 1 mm	To fit patient

СТ

Multidetector CT (MDCT) is preferred, as reformatted images provide additional information useful for surgical planning.

Dosage of IV contrast medium is 1 ml/0.5 kg patient body weight with scanning at 65–70 seconds post-injection to demonstrate arterial and venous anatomy.

Values of CTDI_{vol} should normally be below the relevant national reference dose for the region of scan and patient group (see chapter on radiation protection for the patient).

MIBG

MIBG scanning is routinely part of all neuroblastoma protocols regardless of the site of disease.^{7,10} MIBG imaging should be restricted to nuclear medicine departments with appropriate facilities and staff experienced in imaging children with SPECT-CT as not all centres will have access to this.

FDG PET-CT

Occasionally patients may have MIBG-negative disease, and in this setting, individual patients may benefit from evaluation with 18f-fluorodeoxyglucose (FDG) PET-CT where local expertise and facilities supporting scans on young children are available.^{7,10,11}

Follow-up

Neoadjuvant chemotherapy is used to reduce tumour bulk, and repeat imaging before an attempt at surgical resection should ideally use the same technique as at diagnosis. Post-surgical imaging may be undertaken to provide a baseline for follow-up. Subsequent imaging is directed by clinical suspicion of recurrence.

Tip

 CT has the advantage of detecting calcification within the tumour, and this is usually visible on the post-contrast-enhanced CT images, obviating the need for non-contrast CT images (to keep the radiation burden to a minimum).

4 Rhabdomyosarcoma

Clinical background

Rhabdomyosarcoma is the third most common soft-tissue tumour of childhood following Wilms' tumour and neuroblastoma.⁵ Rhabdomyosarcomas can occur at almost any site but most are seen in the head and neck region and genitourinary tract.⁹ Rhabdomyosarcoma is usually chemo-sensitive, and sometimes there is no or a minimal residual mass on cross-sectional imaging following treatment.

Who should be imaged?

On discovery of a mass likely to be rhabdomyosarcoma, the primary tumour should be imaged using MRI of the appropriate body part in all patients. In addition, evaluation of the locoregional lymph nodes is crucial for accurate staging.

Staging objectives

- To define extent of local disease. Bulky masses may be present and metastasise to regional lymph nodes (cervical for head and neck rhabdomyosarcoma, and pelvic and retroperitoneal for pelvic rhabdomyosarcoma).
- Parameningeal spread through the neural foramina and skull base should be actively sought in head and neck rhabdomyosarcoma.
- Diagnosis for distant metastatic disease, which is seen in up to 10–18% of patients at the time of diagnosis, most frequently to lung, bone and liver.⁵

Staging

MRI

MRI is the preferred technique for imaging head and neck rhabdomyosarcoma owing to its greater sensitivity for detection of parameningeal spread and greater accuracy in discriminating tumour from retained secretions in paranasal sinuses. CT better defines the degree of skull base bony erosion but the degree of skull base destruction is not critical for staging. Sagittal sequences are very useful for midline pelvic masses such as bladder, prostate or vaginal primary tumours.

Coil	Sequence	Plane	Slice	Field of view
			thickness	
Head or neck	T1W	Coronal	4±1 mm	To fit the body part
Head or neck	T2W	Coronal	4±1 mm	To fit the body part
Head or neck	T1W	Axial	4±1 mm	To fit the body part
Head or neck	T2W	Axial	4±1 mm	To fit the body part
Head or neck	STIR	Coronal – whole neck for detection of nodes		To fit the body part
Head or neck	T1W + IV contrast medium, if required	Axial/coronal	4 ± 1 mm	To fit patient
Head or neck	Diffusion- weighted sequences (Multiple b values)	Axial	4 ± 1 mm	To fit patient

Protocol for imaging of head and neck rhabdomyosarcoma

MRI is also the preferred technique for imaging rhabdomyosarcomas elsewhere, particularly for pelvic, paraspinal and extremity tumours. For pelvic and abdominal rhabdomyosarcoma, MRI or CT may be adequate.

Coil	Sequence	Plane	Slice thickness	Field of view
Pelvic/ abdominal	T1W	Coronal	6 ± 2 mm	To fit patient
Pelvic/ abdominal	T2W	Coronal	6 ± 2 mm	To fit patient
Pelvic/ abdominal	T1W	Axial	6 ± 2 mm	To fit patient
Pelvic/ abdominal	T2W	Axial	6 ± 2 mm	To fit patient
Pelvic/ abdominal	Short tau inversion recovery (STIR)	Coronal – whole pelvis for detection of nodes		To fit patient
Pelvic/ abdominal	T1W + IV contrast medium, if required	Axial/coronal	4 ± 1 mm	To fit patient
Pelvic/ abdominal	Diffusion- weighted sequences (Multiple b values)	Axial	4 ± 1 mm	To fit patient

Protocol for imaging of pelvic/abdominal rhabdomyosarcoma

For detection of pulmonary, nodal and hepatic disease, a combination of ultrasound and CT is adequate. Radionuclide bone scanning is increasingly being replaced by whole-body 18F-FDG PET/CT or when not available by whole-body MRI if there is the clinical suspicion of bone metastases.^{69,13}

СТ

CT scans, if preferred to MRI for the primary site evaluation, should be obtained through the region of the primary tumour and the chest following the injection of IV contrast medium.

Dosage of IV contrast medium is 1 ml/0.5 kg patient body weight with scanning at 65–70 seconds post-injection.

Values of CTDI_{vol} should normally be below the relevant national reference dose for the region of scan and patient group (see chapter on radiation protection for the patient).

FDG PET-CT

FDG PET/CT is being increasingly used in the staging of children with rhabdomyosarcoma where the local expertise and facilities to scan younger children are available.^{9,10,11}

Follow-up

Repeat imaging after neoadjuvant chemotherapy is used to plan further management. Local recurrence after surgery or radiotherapy is not infrequent, and post-treatment baseline imaging is useful for further follow-up, particularly when surgical resection is thought to be incomplete.

Tip

Evaluation of the locoregional lymph nodes, with some form of biopsy for suspicious lymph nodes, is crucial for appropriate risk grouping and accurate staging.

References

- Brisse HJ, McCarville MB, Granata C *et al.* Guidelines for imaging and staging of neuroblastic tumors: consensus report from the International Neuroblastoma Risk Group Project. *Radiology* 2011; 261(1): 243–257.
- 2. Swinson S, McHugh K. Urogenital tumours in childhood. Cancer Imaging 2011; 11: S48–S64.
- 3. Geller E, Kochan P. Renal neoplasms of childhood. Radiol Clin North Am 2011; 49(4): 689–709.
- Campbell AD, Cohn SL, Reynolds M et al. Treatment of relapsed Wilms' tumor with high-dose therapy and autologous hematopoietic stem-cell rescue: the experience at Children's Memorial Hospital. J Clin Onc 2004; 22: 2885–2890.
- 5. Van Rijn RR, Wilde JC, Bras J *et al.* Imaging findings in noncraniofacial childhood rhabdomyosarcoma. *Pediatr Radiol* 2008, **38**(6): 617–634.
- Weiss AR, Lyden ER, Anderson JR *et al.* Histologic and clinical characteristics can guide staging evaluations for children and adolescents with rhabdomyosarcoma: a report from the Children's Oncology Group Soft Tissue Sarcoma Committee. *J Clin Oncol* 2013, **31**(26): 3226–3232.
- 7. Orr K, McHugh K. The new international neuroblastoma response criteria. *Pediatr Radiol* 2019; **49**: 1433–1440.
- 8. Servaes SE, Hoffer FA, Smith EA *et al.* Imaging of Wilms tumor: an update. *Pediatr Radiol* 2019; **49**:1441–1452.
- 9. Jawad N, McHugh K. The clinical and radiological features of paediatric rhabdomyosarcoma. *Pediatr Radiol* 2019; **49**: 1516–1523.
- 10. Voss SD. Functional and anatomical imaging in pediatric oncology: which is best for tumors? *Pediatr Radiol* 2019; **49**: 1534–1544.
- The Royal College of Radiologists. Guidelines for the use of PET-CT in children, 2nd edition. London: The Royal College of Radiologists, 2014.
- 12. The Royal College of Radiologists. *iRefer: Making the best use of clinical radiology*, 8th edition. London: The Royal College of Radiologists, 2017. www.irefer.org.uk
- Smets A, Deurloo E, Slager T *et al.* Whole body MR imaging for detection of skeletal metastases in children and young people with primary solid tumors – systemic review. *Pediatr Radiol* 2018; **48**: 241–252.

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