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Clinical Radiology
The Royal College of Radiologists

Recommendations for cross-sectional imaging in cancer management, Second edition

Paediatric neoplasms

Faculty of Clinical Radiology

Contents

Background

Children's tumours are most appropriately imaged 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 newly formulated international neuroblastoma risk grouping (INRG) is now incorporated into new international collaborative studies.[1]

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] Initial imaging is usually with ultrasound, and the differential diagnosis for a renal mass includes nephroblastomatosis, clear cell sarcoma, renal cell carcinoma, renal lymphoma and congenital mesoblastic nephroma. Tumours may be sufficiently large that the organ of origin is not always clear, and CT and 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 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.

CT
Dosage of intravenous contrast medium at CT is 1 ml/0.5 kg patient body weight, with scanning at 65–70 seconds post-injection to allow opacification of the portal vein, renal veins and the 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 Appendix and section on Radiation protection for the patient in CT in Section 2).

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/without IV contrast agents. A suggested protocol is as follows.

Protocol for imaging of Wilms' tumours

Sequence	Plane	Slice thickness	Field of view
T1W	Axial	6 ± 2 mm	To fit patient
T2W	Axial	6 ± 2 mm	To fit patient
T1W	Coronal	6 ± 2 mm	To fit patient
T2W	Coronal	6 ± 2 mm	To fit patient
MR angiography or venography 3D isovolumetric T2 sequence	3D Coronal		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.

Neuroblastoma

Clinical background

Neuroblastoma arises 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.[5] Once the diagnosis of neuroblastoma is strongly suspected or confirmed, radionuclide imaging with metaiodobenzylguanidine (MIBG) scintigraphy should be undertaken in all patients. CT or MRI 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 extension of tumour into spinal canal (‘dumbbell’ tumour).
- To identify bone erosion by primary tumour.
- 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 may 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.

MRI

Both CT and MRI may be used and the choice of MRI is usually governed by availability of equipment. MRI is superior at diagnosing intraspinal extension and metastatic disease in the bone marrow.

Protocol for imaging of neuroblastoma

Coil	Sequence	Plane	Slice thickness	Field of view
To fit patient	T1W	Axial	6 ± 2 mm	To fit patient
To fit patient	T2W	Axial	6 ± 2 mm	To fit patient
To fit patient	T1W	Coronal	6 ± 2 mm	To fit patient
To fit patient	T2W	Coronal	6 ± 2 mm	To fit patient
To fit patient	T1W + IV contrast medium	Axial/coronal if required	6 ± 2 mm	To fit patient
To fit patient	MR angiography or venography	3D acquisition		To fit patient or body part if clinically indicated

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

Dosage of intravenous 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 Appendix and section on Radiation protection for the patient in CT in Section 2).

MIBG
MIBG scanning is routinely part of all neuroblastoma protocols regardless of the site of disease. MIBG imaging should be restricted to nuclear medicine departments with appropriate facilities and staff experienced in imaging children.

FDG PET-CT
Occasionally patients may have MIBG-negative disease and, in this setting, individual patients may benefit from evaluation with FDG PET-CT where local expertise and facilities supporting scans on young children are available.[6]

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).

Rhabdomyosarcoma

Clinical background

Rhabdomyosarcoma is the third most common soft tissue tumour of childhood following Wilms' tumour and neuroblastoma.[7] Most rhabdomyosarcoma occurs in the head and neck region and in the pelvis. 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 loco-regional 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.[7]

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.

Protocol for imaging of head and neck rhabdomyosarcoma

Coil	Sequence	Plane	Slice thickness	Field of view
Head or neck	T1W	Coronal	6 ± 2 mm	To fit the body part
Head or neck	T2W	Coronal	6 ± 2 mm	To fit the body part
Head or neck	T1W	Axial	6 ± 2 mm	To fit the body part
Head or neck	T2W	Axial	6 ± 2 mm	To fit the body part
Head or neck	STIR	Coronal – whole neck for detection of nodes		To fit the body part

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

Protocol for imaging of pelvic/abdominal rhabdomyosarcoma

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

For detection of pulmonary, nodal and hepatic disease, a combination of US and CT is adequate. Radionuclide bone scanning is performed routinely in all patients at diagnosis, although in the future bone scans may not be recommended for low-risk patients.[8] MRI may be more sensitive in demonstrating bone metastases if there is a clinical history suggestive of bone involvement.

CT

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 intravenous contrast medium.

Dosage of intravenous 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 Appendix and section on Radiation protection for the patient in CT in Section 2).

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 loco-regional lymph nodes, with some form of biopsy for suspicious lymph nodes, is crucial for appropriate risk grouping and accurate staging.

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