The Royal College of Pathologists
Pathology: the science behind the cure

Standards and datasets for reporting cancers

Dataset for histopathological reporting of renal tumours in childhood

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Contents

Foreword

The cancer datasets published by the Royal College of Pathologists (RCPath) are a combination of textual guidance, educational information and reporting proformas. The datasets enable pathologists to grade and stage cancers in an accurate, consistent manner in compliance with international standards and provide prognostic information, thereby allowing clinicians to provide a high standard of care for patients and appropriate management for specific clinical circumstances. This guideline has been developed to cover most common circumstances. However, we recognise that guidelines cannot anticipate every pathological specimen type and clinical scenario. Occasional variation from the practice recommended in this guideline may therefore be required to report a specimen in a way that maximises benefit to the patient. Clinicians should be able to explain the reasoning behind any variation from recommended practice.

Each dataset contains core data items (see Appendices E and F) that are mandated for inclusion in the Cancer Outcomes and Services Dataset (COSD – previously the National Cancer Data Set) in England. Core data items are items that are supported by robust published evidence and are required for cancer staging, optimal patient management and prognosis. Core data items meet the requirements of professional standards (as defined by the Information Standards Board for Health and Social Care [ISB]), and it is recommended that at least 95% of reports on cancer resections should record a full set of core data items. Other non-core data items are described. These may be included to provide a comprehensive report or, with appropriate patient consent, to meet local clinical or research requirements. All data items should be clearly defined to allow the unambiguous recording of data.

The following stakeholder was contacted to consult on this document:

- Children's Cancer and Leukaemia Group (CCLG).

The information used to develop this dataset was obtained by undertaking a systematic search derived from the IMPORT (Improving Population Outcomes for Renal Tumours of childhood) protocols followed in the UK. Most of the evidence for this dataset was taken from the International Society of Paediatric Oncology (SIOP) trials.[1–10] Key terms searched included Wilms' tumour, renal tumours, children and pathology, and dates searched were between January 2000 and December 2022. A number of studies met the selection criteria and were considered for review. Published evidence was evaluated using modified SIGN guidance (see Appendix G). Consensus of evidence in the guideline was achieved by expert review. Gaps in the evidence were identified by College members via feedback received during consultation.

No major organisational changes or cost implications have been identified that would hinder the implementation of the dataset.

A formal revision cycle for all guidelines takes place on a three-year cycle. The College will ask the authors of the guideline to consider whether or not the guideline needs to be revised. A full consultation process will be undertaken if major revisions are required. If minor revisions or changes are required, a short note of the proposed changes will be placed on the College website for two weeks for members' attention. If members do not object to the changes, the short notice of change will be incorporated into the guideline and the full revised version (incorporating the changes) will replace the existing version on the College website.

The dataset has been reviewed by the Professional Guidelines team, Working Group on Cancer Services and Lay Advisory Group and was placed on the College website for consultation with the membership from 27 February to 27 March 2023. All comments received from the membership were addressed by the author to the satisfaction of the Chair of the Working Group and the Clinical Lead for Guideline Review.

This dataset was developed without external funding to the writing group. The College requires the authors of guidelines to provide a list of potential conflicts of interest; these are monitored by the

Professional Guidelines team and are available on request. The authors of this document have declared that there are no conflicts of interest.

1 Introduction

Renal tumours comprise 7–8% of all tumours in children under 15 years of age. The most common paediatric renal tumours include nephroblastoma (Wilms' tumour; 85%), mesoblastic nephroma (5%), clear cell sarcoma of the kidney (4%), rhabdoid tumour of the kidney (2%) and miscellaneous rare tumours (4%).[11] Their treatment and prognosis are very different and depend on accurate histological diagnosis and their stage.

Renal tumours in children in the UK are treated and reported[12] according to the protocols of SIOP.[13] A pre-chemotherapy biopsy, previously performed routinely to establish tumour type and subsequently determine the preoperative chemotherapy regimen, A pre-chemotherapy biopsy, previously performed routinely to establish tumour type and subsequently determine the preoperative chemotherapy regimen, is now recommended only for patients aged 10 years and older, or patients aged between 7 and 10 years with smaller tumour volumes, and in some rare indicating cases as defined in the SIOP-Renal Tumour Study Group/CCLG's recommendations for the use of paediatric renal tumour biopsy.[14,18] Chemotherapy is typically followed by surgery and then further chemotherapy and/or radiotherapy, if necessary, depending on the tumour's histological subtype and stage.[15]

The pathologist has an essential role in:

- diagnosis
- identifying the histological subtype and risk group
- making a precise evaluation of the abdominal stage of the tumour. Even in children with stage IV disease, local staging is critical to determine the utilisation of radiotherapy. Based on the correlation between the histological features and survival, three prognostic groups of typical renal tumours of childhood were discerned in the SIOP trials and studies (Appendix C).[1–10]

The criteria for subclassifying the tumours are detailed elsewhere.[13,16] Since the tumours are treated with preoperative chemotherapy, it is important to assess the percentage of non-viable and viable tumour, followed by the percentage of different histological components of the viable tumour.[16]

1.1 Target users of these guidelines

The target primary users of the dataset are trainee and consultant (paediatric) pathologists who are dealing with and reporting these tumours and, on their behalf, the suppliers of IT products to laboratories. The secondary users are surgeons and oncologists, cancer registries and the National Cancer Registration and Analysis Service. Standardised cancer reporting and multidisciplinary team (MDT) working reduce the risk of histological misdiagnosis and help ensure that clinicians have all the relevant pathological information required for tumour staging, management and prognosis. Collection of standardised cancer-specific data also provides information for healthcare providers and epidemiologists and facilitates international benchmarking and research.

These are rare tumours, and it is recommended that they are handled by pathologists with a special interest in paediatric oncology or renal tumour pathology. There should be ready access to an expert opinion. However, the document emphasises the need for meaningful communication between pathologists and treating clinicians.

2 Clinical information required on the specimen request form

Clinical information provided must include details of the patient's age, surgical procedure, tumour side, preoperative biopsy (if performed), preoperative chemotherapy (if given) and information regarding distant metastases. Clinical details should also include information regarding pre- or intraoperative rupture. Ideally, the surgeon should mark the site of preoperative rupture. All these elements are required for accurate assignment of the tumour stage.

3 Preparation of specimens before dissection

The intact surgical specimen should be presented to the pathologist without being opened by the surgeon. The specimen should be received fresh and unfixed in the laboratory. Specimens must be transferred promptly to the laboratory to enable snap-freezing of fresh tissue, which should be done within 60 minutes of excision.

4 Specimen handling and block selection

To obtain accurate information about the stage of the tumour, the nephrectomy specimen should be dealt with as described below.

4.1 Description

The whole specimen should be weighed, measured and photographed. Photography allows difficult cases to be discussed with the MDT and facilitates central pathological review with regard to sample site interpretation. Any areas of rupture or fissuring should be identified and any suspicious areas should be inked in different colours from the rest of the specimen. The specimen should not be decapsulated, as this makes determination of growth beyond the capsule impossible.

[Level of evidence D – Tumour volume, with other parameters, may be a significant prognostic factor.]

Any perirenal and perihilar lymph nodes (which are rare) should be blocked separately and the site recorded.

[Level of evidence A – Lymph node involvement affects SIOP staging.]

The renal vein, artery and ureter should be identified and a transverse section block of each taken near the resection margin.

[Level of evidence A – Margin involvement affects SIOP staging.]

The surface of the whole specimen (or at least areas in which excision margins are possibly involved) and renal sinus should be inked and allowed to dry before opening the specimen. This is a critical step as without inking it might be impossible to stage the tumour correctly; for example, it may be difficult to assess whether resection margins are clear or not.

The specimen should be opened with a longitudinal incision to bivalve and reveal the tumour and its relation to the kidney, capsule and renal sinus. The cut surface should be photographed to demonstrate the tumour, the extent of tumour necrosis and multicystic cut surface (if present).

The report must include the size of the tumour in three dimensions and the percentage of non-viable tumour. The latter is of critical importance in the classification of tumours treated with preoperative chemotherapy.[9]

[Level of evidence A – percentage of necrotic tumour affects SIOP risk group classification.]

The following samples are required for molecular biology studies (these are prospective studies performed to try to identify biological markers of prognosis):

- tumour. At least two pieces (0.5–1 cm^3 each) of morphologically different parts of the tumour should be sampled and snap-frozen in liquid nitrogen or at –70°C (freeze more aliquots if available). If a biopsy is performed prior to commencing preoperative chemotherapy, then a sample of this should also be frozen, if adequate tissue is available.
- a 'mirror' sample of tumour adjacent to the frozen sample should be fixed in formalin and studied for histology. This paraffin block or corresponding slides should accompany the frozen tissue, when requested for additional studies.
- adjacent normal kidney. Two pieces (0.5–1 cm^3) should be snap-frozen in liquid nitrogen or at –70 °C.
- a sample of nephrogenic rest tissue, if identified.
- 10 ml peripheral blood in ethylenediaminetetraacetic acid (EDTA) (if national procedure for storage is available).

Samples should be stored at –70 °C or under liquid nitrogen until transported to the appropriate national research laboratory on dry ice for cases consenting to research studies.

The time interval between removal of the tumour and the freezing of the samples should be as short as possible and certainly not exceed a period of 30–60 minutes.

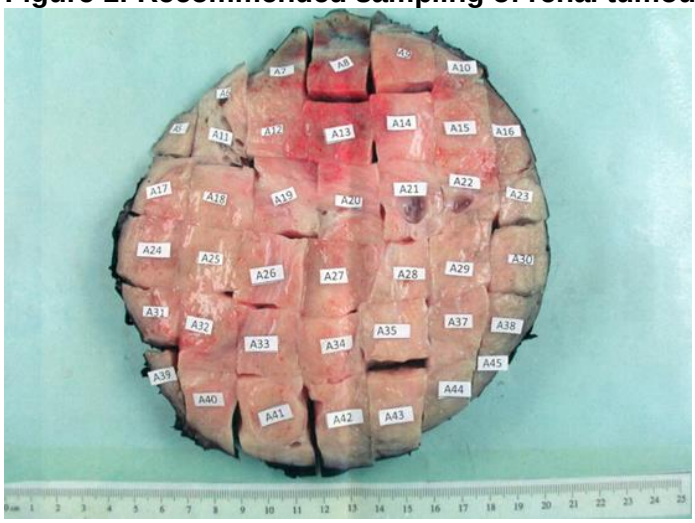
[Level of evidence GPP – for preservation of samples the time interval should not exceed 30–60 minutes.]

The specimen should be fixed in 10% buffered formalin for 24–48 hours according to the usual procedure of the laboratory. Several additional cuts can be made parallel to the initial cut to divide the specimen into 'slabs' for better fixation.

4.2 Block selection

A photograph or a pre-prepared diagram in the SIOP Institutional Pathology Form should preferably be used (Appendix A). The samples for histological examination should include at least one longitudinal slice of tumour and kidney surface, completely sampled (see Figure 1; mega-blocks make histological assessment much easier and they are less time consuming for both pathologists and their labs).

Figure 1: Recommended sampling of renal tumours.



In addition, the following should be sampled:

- the macroscopically different areas of the tumour
- areas suspected of being incompletely resected or surgically adherent, which are marked by the surgeon (with appropriate ink or dye) for the special attention of the pathologist
- sinus lymph nodes when present
- other lymph nodes
- renal pelvis and pelvic fat, ureter and sinus vessels (the renal vein should be inspected for evidence of tumour thrombus in particular – if present, it is critical to assess whether it is completely resected)
- each nodule away from the main mass (in multifocal tumours)
- tumour–kidney interface
- tumour–kidney capsule
- areas of the capsule that are suspected of being invaded by the tumour
- areas of perirenal fat where tumour infiltration is suspected (this is important in assessing whether or not the tumour is completely resected)
- areas of adhesions of the tumour to surrounding tissues
- at least two blocks of the normal kidney and blocks from abnormal looking areas in the remaining renal tissue.

A ‘block guide’ (as in Figure 1) is essential to allow for central review, i.e. all the samples should be numbered and their sites recorded as well as all other samples taken at the time of operation (e.g. adrenals, lymph nodes and various biopsies).

In the histopathology report, all relevant findings should refer to the block/slide number (e.g. ‘There is renal sinus invasion in block A7’), as this assists central pathology review.

5 Core data items

Core data items include:

- total weight of kidney with tumour (in grams)

- size of specimen in all 3 dimensions (in millimetres)
- size of the tumour (in all three dimensions)
- location of tumour
- tumour focality

  [Level of evidence D – tumour volume, with other parameters, may be a significant prognostic factor.]
- specimen integrity, i.e. was it received intact from the operating theatre?
- renal capsule integrity i.e. was it grossly intact?

  [Level of evidence A – renal capsule status is important prognostic information used in SIOP staging.]
- was the surface inked?

  [Level of evidence GPP – inking of resection margins affects certainty of margin status and therefore staging.]
- block key identification
- percentage of necrosis/regressive changes on gross examination
- percentage of necrosis/regressive changes on microscopic examination

  [Level of evidence A – percentage of necrosis/regressive change provides prognostic information.]
- percentage of blastema as a proportion of viable tumour

  [Level of evidence A – relative percentage of tumour components provides prognostic information.]
- presence of anaplasia and whether it is focal or diffuse

  [Level of evidence A – the presence of anaplasia affects risk group stratification.]
- presence of perirenal fat invasion

  [Level of evidence A – perirenal fat invasion is important prognostic information used in SIOP staging.]
- presence of renal sinus invasion

  [Level of evidence A – renal sinus invasion is important prognostic information used in SIOP staging.]
- presence of renal vein tumour

  [Level of evidence A – renal vein invasion by tumour is important prognostic information used in SIOP staging.]
- resection margin involvement and if involved, whether by viable or non-viable tumour

  [Level of evidence A – resection margin status is important prognostic information used in SIOP staging.]
- lymph node examination, i.e. whether or not lymph nodes have been examined. (For each lymph node group, state the number of nodes identified, the number of nodes positive, negative or uncertain, and whether the tumour involvement is viable or non-viable for each node.)

  [Level of evidence A – lymph node involvement is important prognostic information used in SIOP staging.]
- histological diagnosis and subtype

[Level of evidence A – histological type determines SIOP tumour risk group.]

- tumour risk group for tumours treated with preoperative chemotherapy – this is the risk grouping based on the SIOP classification (see Appendix C)
- local tumour stage for tumours treated with preoperative chemotherapy using the SIOP staging system
- tumour risk group for tumours treated with immediate surgery – this is the risk grouping bases on the SIOP classification (see Appendix C)
- local tumour stage for tumours treated with immediate surgery using the SIOP staging system
- reason for staging – the reason for the stated SIOP stage
- SNOMED CT codes or SNOMED T and M codes (Appendix B).

6 Non-core data items

Non-core data items include:

- presence of associated anomalies and/or relevant syndromes (clinical information)
- microscopic assessment of the percentage of epithelial and stromal components as proportions of the viable tumour
- presence or absence of nephrogenic rests
- coexisting pathological changes
- ancillary studies

7 Diagnostic staging and coding

The tumours are staged according to the SIOP staging system (see Appendix D).

Tumour stage is one of the most important therapeutic and prognostic criteria for renal tumours. It has been shown in all multicentre trials that accuracy of staging still represents a major problem.[17] This is partly because renal tumours are usually very large at nephrectomy and it is often very difficult to assess their relationship with normal renal anatomical structures such as the renal capsule and the renal sinus. The renal sinus is best recognised by the presence of nerves, blood vessels and lymphatics; nerves are of particular significance as they are never present within tumours.

The local (abdominal) staging of primary tumour is carried out following pre-nephrectomy chemotherapy and is very important even in stage IV cases. The presence or absence of metastases is evaluated at presentation, on the basis of imaging studies.

Separate proformas should be completed for bilateral tumours and the local stage stated for each. For multifocal tumours, each nodule should be assessed individually and then a tumour as a whole.

The tumour should be coded according to the SNOMED system using appropriate body structure and morphologic abnormality codes for SNOMED CT (see Appendix B).

SNOMED procedure codes should be recorded for the procedure. Procedure codes vary according to the SNOMED system in use in different organisations, therefore local procedure codes should be recorded and used for audit purposes. SNOMED ceased to be licensed by the International Health Terminology Standards Development Organisation from 26 April 2017.

A list of typically applicable SNOMED CT codes is provided in Appendix B. Mapping from SNOMED to SNOMED CT terminology is provided.

8 Reporting of biopsy specimens

The main purpose of a biopsy is to establish whether the tumour is a Wilms tumour or another renal tumour that may require different preoperative treatment.[13,14,18]

9 Reporting of frozen sections

Frozen section diagnosis is not appropriate for paediatric tumours since many entities share a common morphological phenotype (‘small round blue cell’) and cannot be distinguished on morphological grounds alone. Frozen sections are not recommended for renal tumours of childhood.

10 Criteria for audit

The following are recommended by the RCPath as key assurance indicators (see Key Assurance Indicators for Pathology Services, November 2019) and key performance indicators (see Key Performance Indicators – Proposals for implementation, July 2013):

- cancer resections should be reported using a template or proforma, including items listed in the English COSD, which are, by definition, core data items in RCPath cancer datasets. English trusts were required to implement the structured recording of core pathology data in the COSD.
  - standard: 95% of reports must contain structured data
- histopathology cases must be reported, confirmed and authorised within seven and ten calendar days of the procedure.
  - standard: 80% of cases must be reported within seven calendar days and 90% within ten calendar days.

All paediatric pathologists should participate in the national external quality assessment scheme.

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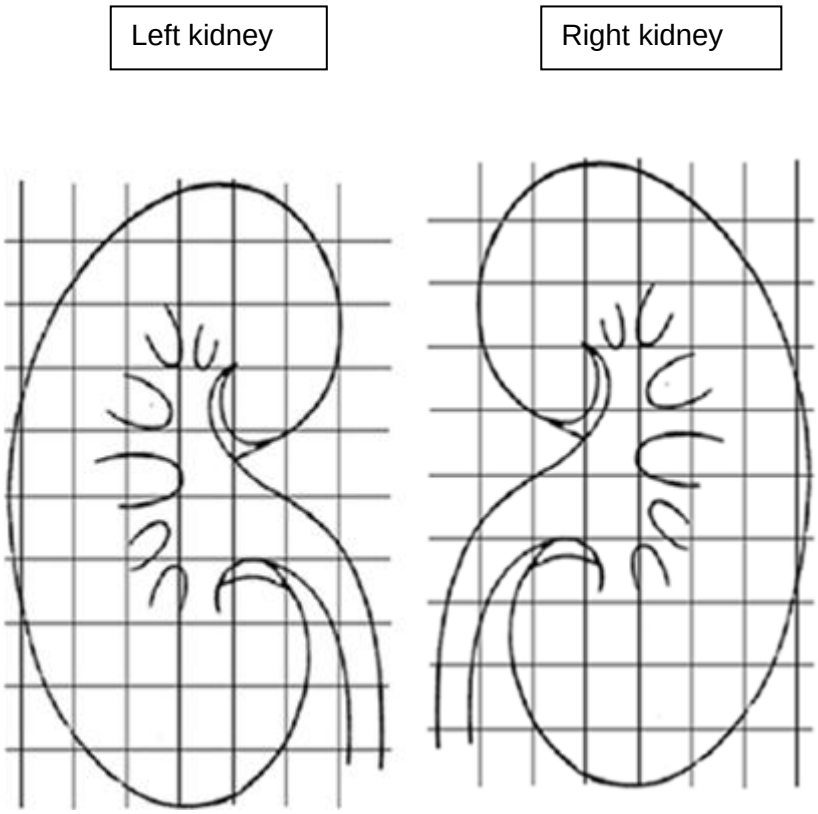
Appendix A SIOP diagram for renal tumours



Please draw or photograph the tumour and document the exact site (by using numbers or letters) of each section taken.

Appendix B SNOMED T and M codes and SNOMED CT codes for paediatric renal tumours

SNOMED T codes

Topographical codes	SNOMED	SNOMED CT terminology	SNOMED CT code
Kidney	T71000	Kidney structure (body structure)	64033007

M codes

Morphological codes	SNOMED	SNOMED CT terminology	SNOMED CT code
Cystic nephroma	M89590	Benign cystic nephroma (morphologic abnormality)	128757006
Cystic partially differentiated nephroblastoma	M89591	Cystic partially differentiated nephroblastoma (morphologic abnormality)	128758001
Mesoblastic nephroma	M89601	Mesoblastic nephroma (morphologic abnormality)	11793003
Nephroblastoma (Wilms tumour)	M89603	Nephroblastoma (morphologic abnormality)	25081006
Rhabdoid tumour of the kidney	M89633	Malignant rhabdoid tumour (morphologic abnormality)	83118000
Clear cell sarcoma of the kidney	M89643	Clear cell sarcoma of kidney (morphologic abnormality)	24007003

Appendix C The revised SIOP working classification of renal tumours of childhood (2016)[15]

For pre-treated cases

- Low-risk tumours
  - mesoblastic nephroma[1,2]
  - cystic partially differentiated nephroblastoma
  - completely necrotic nephroblastoma[3]
- Intermediate-risk tumours
  - nephroblastoma: epithelial type[4]
  - nephroblastoma: stromal type[4]
  - nephroblastoma: mixed type
  - nephroblastoma: regressive type
  - nephroblastoma: focal anaplasia[5,6]
- High-risk tumours
  - nephroblastoma: blastemal type[14]
  - nephroblastoma: diffuse anaplasia[5,6]
  - clear cell sarcoma of the kidney[7,8]
  - rhabdoid tumour of the kidney[9,10]

For primary nephrectomy cases

- Low-risk tumours
  - mesoblastic nephroma
  - cystic partially differentiated nephroblastoma
- Intermediate-risk tumours
  - non-anaplastic nephroblastoma and its variants
  - nephroblastoma: focal anaplasia
- High-risk tumours
  - nephroblastoma: diffuse anaplasia
  - clear cell sarcoma of the kidney
  - rhabdoid tumour of the kidney

Appendix D SIOP staging criteria for paediatric renal tumours (2016)[15]

Stage I

a) The tumour is limited to the kidney.

b) Tumour is present in the perirenal fat but is surrounded by a fibrous (pseudo)capsule. The (pseudo)capsule may be infiltrated by viable tumour, which does not reach the outer surface.

c) Tumour may show botryoid/protruding growth into the renal pelvis or the ureter, but it does not infiltrate their walls.

d) The vessels or the soft tissues of the renal sinus are not involved by tumour.

e) Intrarenal vessel involvement may be present.

Notes:

- *Fine needle aspiration or percutaneous cutting needle ('tru-cut') biopsy does not upstage the tumour.*
- *The presence of necrotic tumour or chemotherapy-induced change in the renal sinus, renal veins and/or within the perirenal fat should not be regarded as a reason for upstaging the tumour.*
- *Viable tumour infiltration of fat between the kidney and the adrenal gland, or of the adrenal gland itself, does not upstage the tumour if the tumour is contained within the (pseudo)capsule. However, the presence of viable tumour in the lymphatic or blood vessels in this area is regarded as stage II.*
- *Liver: tumour might be attached to the liver capsule and this should not be regarded as infiltration of the adjacent organ; only if clear infiltration of the liver parenchyma is present, the tumour should be regarded as stage II (if completely resected) or stage III (if incompletely resected).*

Stage II

a) Viable tumour is present in the perirenal fat and is not covered by a (pseudo)capsule, but is completely resected (resection margins 'clear').[19]

b) Viable tumour infiltrates the soft tissues of the renal sinus.

c) Viable tumour infiltrates blood and/or lymphatic vessels of the renal sinus or of the perirenal tissue, but it is completely resected.

d) Viable tumour infiltrates the wall of the renal pelvis or of the ureter.

e) Viable tumour infiltrates the vena cava or adjacent organs (except the adrenal gland, see above) but is completely resected in one piece.

Stage III

a) Viable tumour present at a resection margin. Non-viable tumour or chemotherapy-induced changes present at a resection margin is not regarded as stage III.

b) Abdominal lymph node involvement by either viable or non-viable tumour.

c) Pre- or intraoperative tumour rupture, if confirmed by microscopic examination (viable tumour at the surface of the specimen in the area of the rupture).

d) Viable or non-viable tumour thrombus is present at resection margins of ureter, renal vein or vena cava inferior (always discuss resection margins with the surgeon).

e) Viable or non-viable tumour thrombus, which is attached to the inferior vena cava wall, is removed piecemeal by surgeon.

f) Wedge/open tumour biopsy prior to preoperative chemotherapy or surgery.

g) Tumour implants (viable or non-viable) are found anywhere in the abdomen.

h) Tumour (viable or non-viable) has penetrated through the peritoneal surface.

Stage IV

a) Haematogenous metastases (lung, liver, bone, brain, etc.) or lymph node metastases outside the abdomino-pelvic region.

Stage V

a) Bilateral renal tumours at diagnosis. Each side should be substaged according to the above criteria.

Appendix E Reporting proforma for paediatric renal tumours

Surname: …… Forenames: …… Date of birth: …… Sex: ……..
Hospital……..…….. Hospital no: ……..…… NHS no: …….
Date of surgery: ……..….. Date of report authorisation: …… Report no: …….
Date of receipt:…… Pathologist: ……..…….. Surgeon: ……....

Clinical data and specimen type

Prechemotherapy open biopsy		Yes □	No □	Not stated □
Preoperative chemotherapy		Yes □	No □	Not stated □
Pre- or intraoperative tumour rupture		Yes □	No □	Not stated □
Tumour site	Left □	Right □		
	Bilateral	Yes □	No □ (if bilateral, complete separate forms for left and right)	
Nephrectomy	Unilateral □		Total □	Partial □
	Bilateral □	Left:	Total □	Partial □
		Right:	Total □	Partial □

Macroscopic features

Total weight of specimen with tumour ……g Size of specimen …….. x …….. x ……. mm

Tumour size …….. x …….. x ……. mm

Location of tumour: Lower pole □ Upper pole □ Whole kidney □ Multifocal □

Tumour multifocal? Yes □ No □ Uncertain □ If yes, number of foci ……

Specimen received intact from operating theatre? Yes □ No □ Uncertain □

Renal capsule grossly intact? *(before opening specimen)* Yes □ No □ Uncertain □

Surface inked? No □ Before opening specimen □ After opening specimen □

Percentage of necrosis/regressive changes on gross examination…… *(please state)*

Histological features

Percentage of necrosis/regressive changes on histological examination

<65% *(please state)* …… 65–99% *(please state)* …… 100% ……..…

Percentage of: Blastema…….. Epithelium …… Stroma ……..

Anaplastic nephroblastoma:	Focal □	Diffuse □	Uncertain □	No □
Perirenal fat invasion	Yes □	No □	Uncertain □	
Renal sinus invasion	Yes □	No □	Uncertain □	
Perirenal vessels invasion	Yes □	No □	Uncertain □	
Renal vein tumour	Yes □	No □	Uncertain □	
Resection margins involved	Yes □	No □	Uncertain □	
If yes, is tumour:	Viable □	Non-viable □		

Lymph nodes examined Yes □ No □

Site of node	No of nodes identified	Lymph node status			Node involved by viable or non-viable tumour or both
		No of negative nodes	No of positive nodes	No of uncertain nodes	
Hilar					Viable □ Non-viable □ Both □
Para-aortic					Viable □ Non-viable □ Both □
Other					Viable □ Non-viable □ Both □

Total number of positive lymph nodes: ……

Conclusion

Tumour diagnosis and risk group:

Risk group	Diagnosis, for pre-treated cases	Diagnosis, for primary nephrectomy cases
Low risk □	Mesoblastic nephroma □ Cystic partially differentiated nephroblastoma □ Completely necrotic nephroblastoma □	Mesoblastic nephroma □ Cystic partially differentiated nephroblastoma □
Intermediate risk □	Nephroblastoma – epithelial type □ Nephroblastoma – stromal type □ Nephroblastoma – mixed type □ Nephroblastoma – regressive type □ Nephroblastoma – focal anaplasia □	Non-anaplastic nephroblastoma and its variants □ Nephroblastoma – focal anaplasia □
High risk □	Nephroblastoma – blastemal type □ Nephroblastoma – diffuse anaplasia □ Clear cell sarcoma of the kidney □ Rhabdoid tumour of the kidney□	Nephroblastoma – diffuse anaplasia □ Clear cell sarcoma of the kidney □ Rhabdoid tumour of the kidney □

Tumour local SIOP stage (2018): I □ II □ III □

Reason for stage …..……..

SNOMED CODES: T …….. M ……..

Pathologist

Name …… Signature …… Date ……..

Appendix F Reporting proforma for paediatric renal tumours in list format

Please add a box with:

Prechemotherapy biopsy	Yes No Not stated	If yes	FNA tru-cut open

Element name	Values	Implementation notes	COSD v9
Preoperative chemotherapy	Single-selection value list: • Yes • No • Not stated		
Pre- or intraoperative tumour rupture	Single-selection value list: • Yes • No • Not stated		pCT6610
Tumour site	Single-selection value list: • Left • Right		pCR0820
Tumour bilateral	Single-selection value list: • Yes • No	If bilateral, complete separate forms for left and right.	
Nephrectomy unilateral or bilateral	Single-selection value list: • Unilateral • Bilateral		
Unilateral nephrectomy total or partial	Single-selection value list: • Total • Partial	Only applicable if 'Unilateral' is selected for 'Nephrectomy unilateral or bilateral'.	
Left nephrectomy total or partial	Single-selection value list: • Total • Partial	Only applicable if 'Bilateral' is selected for 'Nephrectomy unilateral or bilateral'.	
Right nephrectomy total or partial	Single-selection value list: • Total • Partial	Only applicable if 'Bilateral' is selected for 'Nephrectomy unilateral or bilateral'.	
Total weight of specimen with tumour	Weight in grams		

Size of specimen	Size in mm in three dimensions		
Location of tumour	Single-selection value list: • Lower pole • Upper pole • Whole kidney • Multifocal		
Tumour multifocal	Single-selection value list: • Yes • No • Uncertain		
Number of tumour foci	Integer	Only applicable if 'Tumour multifocal' is selected.	
Specimen received intact from operating theatre?	Single-selection value list: • Yes • No • Uncertain		
Renal capsule grossly intact?	Single-selection value list: • Yes • No • Uncertain		
Surface inked?	Single-selection value list: • Yes • No		
Surface inked timing	Single-selection value list: • Before opening specimen • After opening specimen • Not applicable	Not applicable if 'Surface inked' is 'No'.	
Percentage of necrosis/regressive changes on gross examination, specify	Number between 0 and 100		
Percentage of necrosis/regressive changes on histological examination	Single-selection value list: • <65% • 65–99% • 100%		
Percentage of necrosis/regressive changes on histological examination, specify	Number between 0 and 99.99	Not to be completed if 'Percentage of necrosis/ regressive changes on histological	

		examination' is '100%'.	
Percentage of blastema	Number between 0 and 100		
Percentage of epithelium	Number between 0 and 100		
Percentage of stroma	Number between 0 and 100		
Anaplastic nephroblastoma	Single-selection value list: • Yes • No • Uncertain		
Anaplastic nephroblastoma, subclassify	Single-selection value list: • Focal • Diffuse • Uncertain • Not applicable	Not applicable if 'Anaplastic nephroblastoma' is 'No'.	pCT6620
Perirenal fat invasion	Single-selection value list: • Yes • No • Uncertain		pCT6630
Renal sinus invasion	Single-selection value list: • Yes • No • Uncertain		pCT6640
Perirenal vessels invasion	Single-selection value list: • Yes • No • Uncertain		
Renal vein tumour	Single-selection value list: • Yes • No • Uncertain		pCT6650
Resection margins involved	Single-selection value list: • Yes • No • Uncertain		pCR0880 Yes = (05) Tumour reaches excision margin No = (01) Excision margins are clear (distance from margin not stated) Uncertain = (06) Uncertain
Resection margin tumour viable	Single-selection value list: • Viable • Non-viable • Not applicable	Not applicable if 'Resection margins involved' is 'No' or 'Uncertain'.	pCT6680

Lymph nodes examined	Single-selection value list: • Yes • No		
Hilar, number of nodes identified	Integer		
Hilar, number of negative nodes	Integer		
Hilar, number of positive nodes	Integer		
Hilar, number of uncertain nodes	Integer		
Hilar, type of nodal involvement	Single-selection value list: • Viable • Non-viable • Both • Not applicable	Not applicable if 'Hilar, number of positive nodes' is '0'.	
Para-aortic, number of nodes identified	Integer		
Para-aortic, number of negative nodes	Integer		
Para-aortic, number of positive nodes	Integer		
Para-aortic, number of uncertain nodes	Integer		
Para-aortic, type of nodal involvement	Single-selection value list: • Viable • Non-viable • Both • Not applicable	Not applicable if 'Para-aortic, number of positive nodes' is '0'.	
Other, number of nodes identified	Integer		
Other, number of negative nodes	Integer		
Other, number of positive nodes	Integer		
Other, number of uncertain nodes	Integer		
Other, type of nodal involvement	Single-selection value list: • Viable • Non-viable • Both • Not applicable	Not applicable if 'Other, Number of positive nodes' is '0'.	
Total number of positive lymph nodes	Integer		pCR0900
Risk group	Single-selection value list:		

	• Low risk • Intermediate risk • High risk		
Low risk	Single-selection value list: • Pre-treated case: Mesoblastic nephroma • Pre-treated case: Cystic partially differentiated nephroblastoma • Pre-treated case: Completely necrotic nephroblastoma • Primary nephrectomy case: Mesoblastic nephroma • Primary nephrectomy case: Cystic partially differentiated nephroblastoma	Only completed if 'Risk group' is 'Low risk'.	
Intermediate risk	Single-selection value list: • Pre-treated case: Nephroblastoma – epithelial type • Pre-treated case: Nephroblastoma – stromal type • Pre-treated case: Nephroblastoma – mixed type • Pre-treated case: Nephroblastoma – regressive type • Pre-treated case: Nephroblastoma – focal anaplasia • Primary nephrectomy case: Non-anaplastic nephroblastoma and its variants • Primary nephrectomy case: Nephroblastoma – focal anaplasia	Only completed if 'Risk group' is 'Intermediate risk'.	
High risk	Single-selection value list: • Pre-treated case: Nephroblastoma – blastemal type • Pre-treated case: Nephroblastoma – diffuse anaplasia	Only completed if 'Risk group' is 'High risk'.	

	• Pre-treated case: Clear cell sarcoma of the kidney • Pre-treated case: Rhabdoid tumour of the kidney • Primary nephrectomy case: Nephroblastoma – diffuse anaplasia • Primary nephrectomy case: Clear cell sarcoma of the kidney • Primary nephrectomy case: Rhabdoid tumour of the kidney		
Tumour local SIOP stage (2016)	Single-selection value list: • I • II • III		pCT6670
Reason for stage	Free text		
SNOMED Topography code	May have multiple codes. Look up from SNOMED tables.		
SNOMED Morphology code	May have multiple codes. Look up from SNOMED tables.		

Appendix G Summary table – explanation of grades of evidence

(modified from Palmer K *et al. BMJ* 2008;337:1832)

Grade (level) of evidence	Nature of evidence
Grade A	At least one high-quality meta-analysis, systematic review of randomised controlled trials or a randomised controlled trial with a very low risk of bias and directly attributable to the target cancer type or A body of evidence demonstrating consistency of results and comprising mainly well-conducted meta-analyses, systematic reviews of randomised controlled trials or randomised controlled trials with a low risk of bias, directly applicable to the target cancer type.
Grade B	A body of evidence demonstrating consistency of results and comprising mainly high-quality systematic reviews of case-control or cohort studies and high-quality case-control or cohort studies with a very low risk of confounding or bias and a high probability that the relation is causal and which are directly applicable to the target cancer type or Extrapolation evidence from studies described in A.
Grade C	A body of evidence demonstrating consistency of results and including well-conducted case-control or cohort studies and high-quality case-control or cohort studies with a low risk of confounding or bias and a moderate probability that the relation is causal and which are directly applicable to the target cancer type or Extrapolation evidence from studies described in B.
Grade D	Non-analytic studies such as case reports, case series or expert opinion or Extrapolation evidence from studies described in C.
Good practice point (GPP)	Recommended best practice based on the clinical experience of the authors of the writing group.

Appendix H AGREE II compliance monitoring sheet

The datasets of the Royal College of Pathologists comply with the AGREE II standards for good quality clinical guidelines. The sections of this dataset that indicate compliance with each of the AGREE II standards are indicated in the table below.

AGREE standard	Section of guideline
Scope and purpose	
1 The overall objective(s) of the guideline is (are) specifically described	Foreword, 1
2 The health question(s) covered by the guideline is (are) specifically described	Foreword, 1
3 The population (patients, public, etc.) to whom the guideline is meant to apply is specifically described	Foreword
Stakeholder involvement	
4 The guideline development group includes individuals from all the relevant professional groups	Foreword
5 The views and preferences of the target population (patients, public, etc.) have been sought	Foreword
6 The target users of the guideline are clearly defined	1
Rigour of development	
7 Systematic methods were used to search for evidence	Foreword
8 The criteria for selecting the evidence are clearly described	Foreword
9 The strengths and limitations of the body of evidence are clearly described	Foreword
10 The methods for formulating the recommendations are clearly described	Foreword
11 The health benefits, side effects and risks have been considered in formulating the recommendations	Foreword, 1
12 There is an explicit link between the recommendations and the supporting evidence	2–9
13 The guideline has been externally reviewed by experts prior to its publication	Foreword
14 A procedure for updating the guideline is provided	Foreword
Clarity of presentation	
15 The recommendations are specific and unambiguous	2–9
16 The different options for management of the condition or health issue are clearly presented	2–9
17 Key recommendations are easily identifiable	2–9
Applicability	
18 The guideline describes facilitators and barriers to its application	Foreword
19 The guideline provides advice and/or tools on how the recommendations can be put into practice	Appendices A–F
20 The potential resource implications of applying the recommendations have been considered	Foreword
21 The guideline presents monitoring and/or auditing criteria	10
Editorial independence	
22 The views of the funding body have not influenced the content of the guideline	Foreword
23 Competing interest of guideline development group members have been recorded and addressed	Foreword