ESPR
European Society of Paediatric Radiology

56th Annual Meeting &
42nd Post Graduate Course

SOCIÉTÉ INTERNATIONALE D'ONCOLOGIE PÉDIATRIQUE
INTERNATIONAL SOCIETY OF PAEDIATRIC ONCOLOGY

SIOP Renal Tumour Study Group
RTSG

From Wilms to kidney tumors: Which ones require a biopsy ?

institut Curie

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Background - Kidney tumors / Gamuts

Benign

- **Congenital mesoblastic nephroma (CMN)**

Nephroblastomatosis (nephrogenic rests)

- Cystic nephroma
- Stromal metanephric tumors
- Angiomyolipoma (Tuberous sclerosis)
- Urothelial Cyst

Malignant

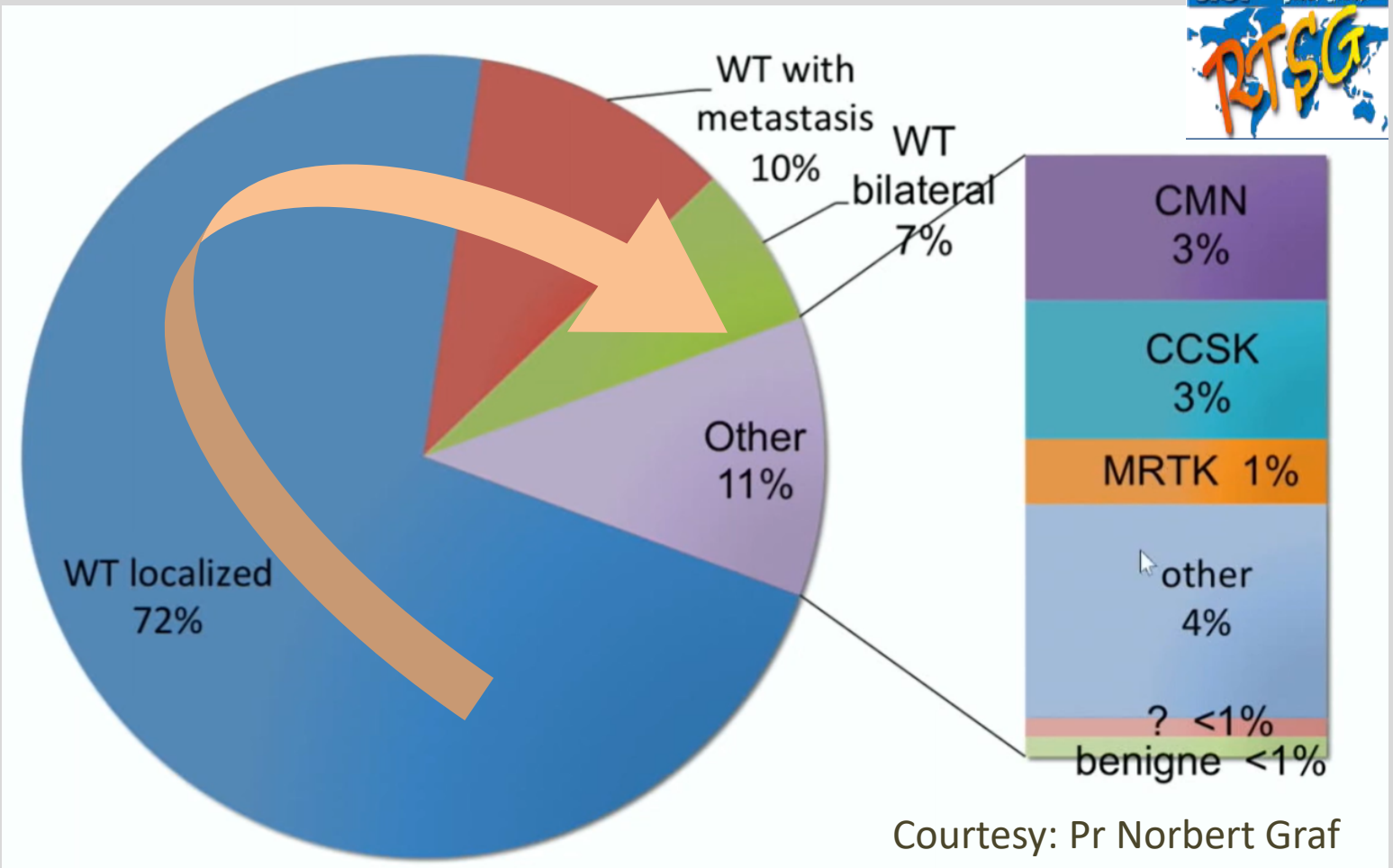
- **Nephroblastoma (Wilms' tumor)**
- **Carcinomas**
  - Juvenile Renal Cell Carcinoma (RCC)
  - Renal Medullary Carcinoma (RMC)
  - Others
- **Sarcomas**
  - Clear Cell Sarcoma of the Kidney (CCSK)
  - Malignant Rhabdoid Tumor (MRTK)
  - Cellular CMN / Fibrosarcoma
  - Others : Ewing, SNVS…
- **Hematologic malignancies**

Pseudotumors

- **Infectious diseases**
  - Acute PN
  - Abscess
  - Chronic PN (XGPN)
- **Malformations**
  - Segmental cystic dysplasia
  - Lymphangioma
- **Miscellaneous**
  - Urinoma
  - Sarcoïdosis….

Background - Kidney tumors / relative incidence

Wilms ≅ 90% !

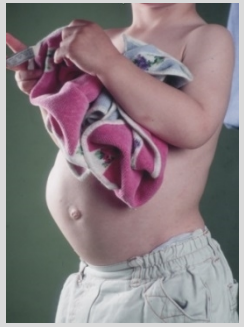


Courtesy: Pr Norbert Graf

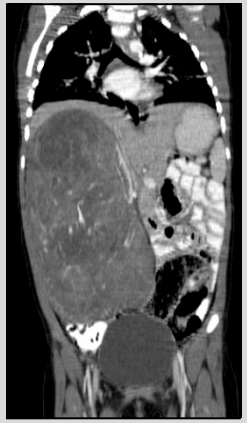
https://www.youtube.com/watch?v=bs0kAHElrg0

Background - unilateral localized tumors: current approaches

COG-North-Am. *approach*

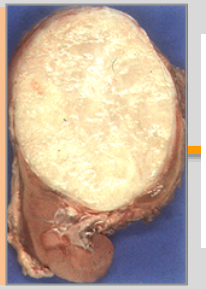


Diagnosis



Primary Surgery

Pathology
- Histotype
- Genetics
- Staging



Risk-adapted postoperative Tt chemo +/- RT

Typical Wilms ?

Yes 2/3

Presumptive neoadjuvant chemo

No 1/3

Surgery ? Biopsy ?

Secondary Surgery

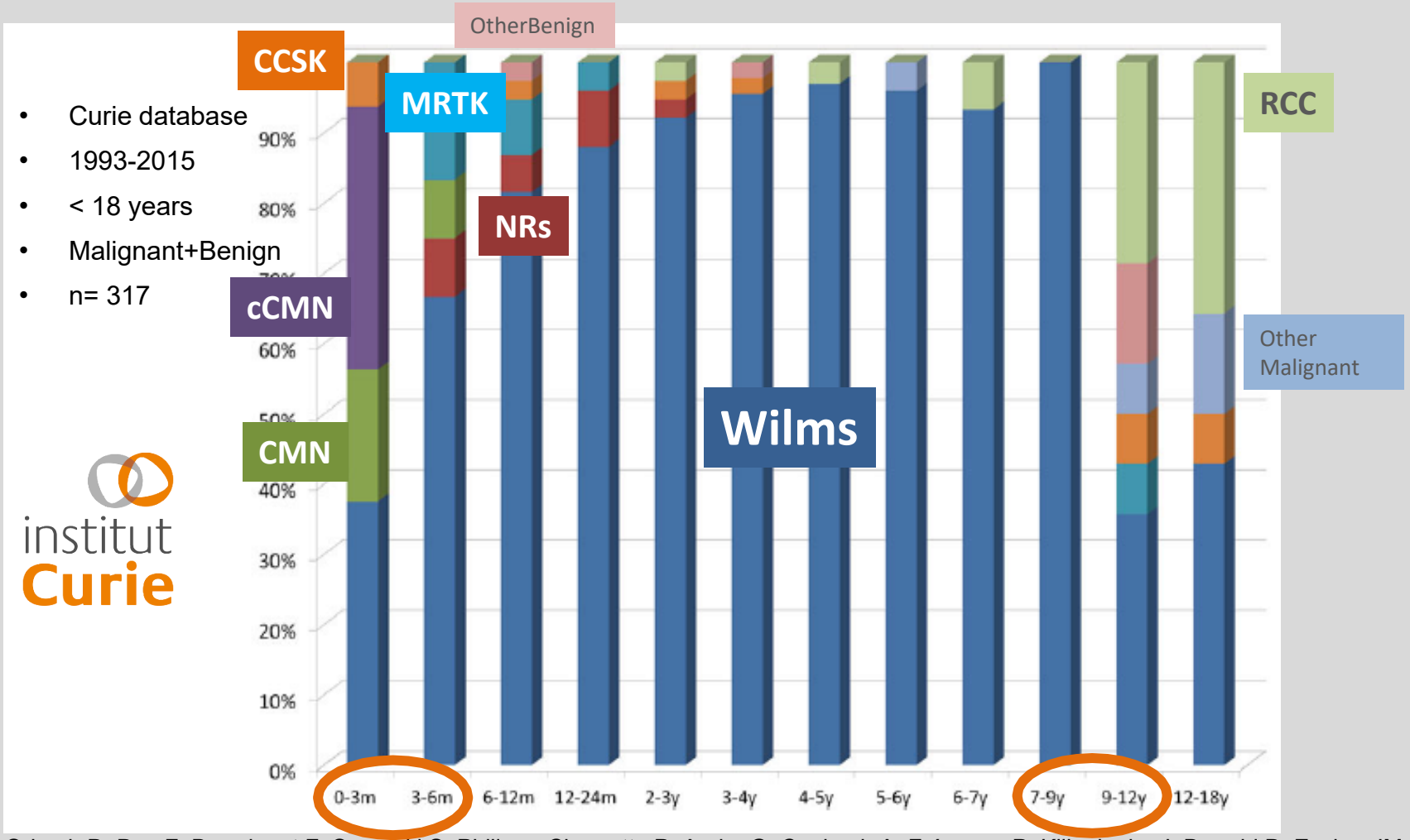
Rational
- ↘ tumor volume ↘ perop rupture
- ↘ local stage ↘ RT
- ↗ Information / chemosensitivity
- ↘ need for biopsy

SIOP-RTSG *approach*

Diagnostic criteria: Wilms vs Non-Wilms

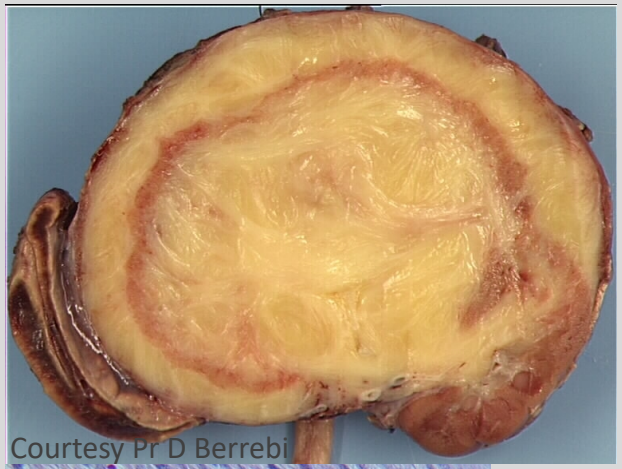
Best criterion:

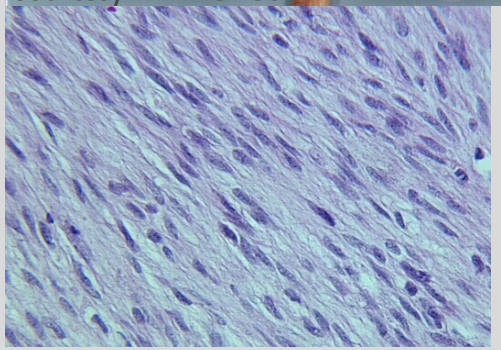
Patient's **age** !

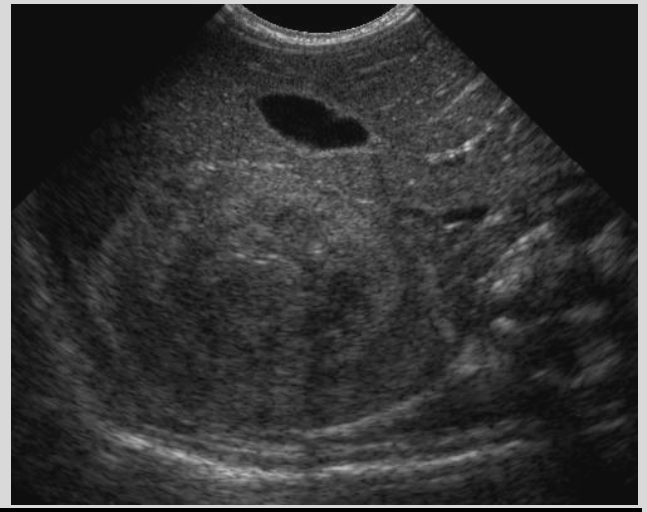


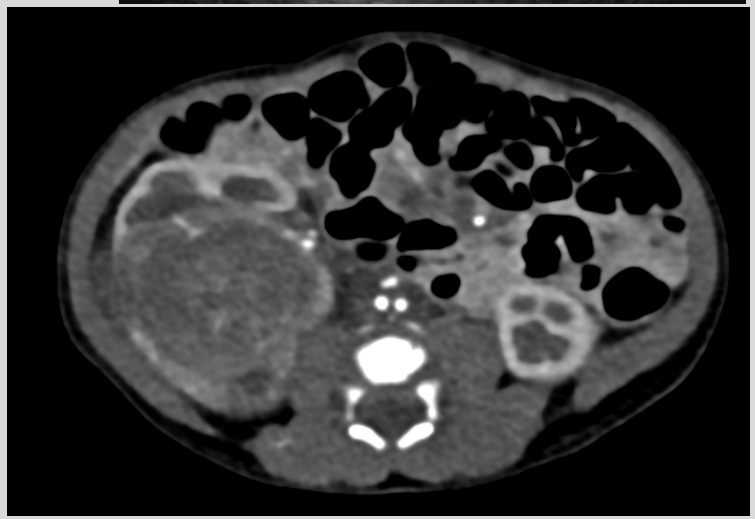
de la Monneraye Y, Michon J, Pacquement H, Aerts I, Orbach D, Doz F, Bourdeaut F, Sarnacki S, Philippe-Chomette P, Audry G, Coulomb A, Fréneaux P, Klijanienko J, Berrebi D, Zucker JM, Schleiermacher G, Brisse HJ. Indications and results of diagnostic biopsy in pediatric renal tumors: A retrospective analysis of 317 patients with critical review of SIOP guidelines. **Pediatr Blood Cancer. 2019**

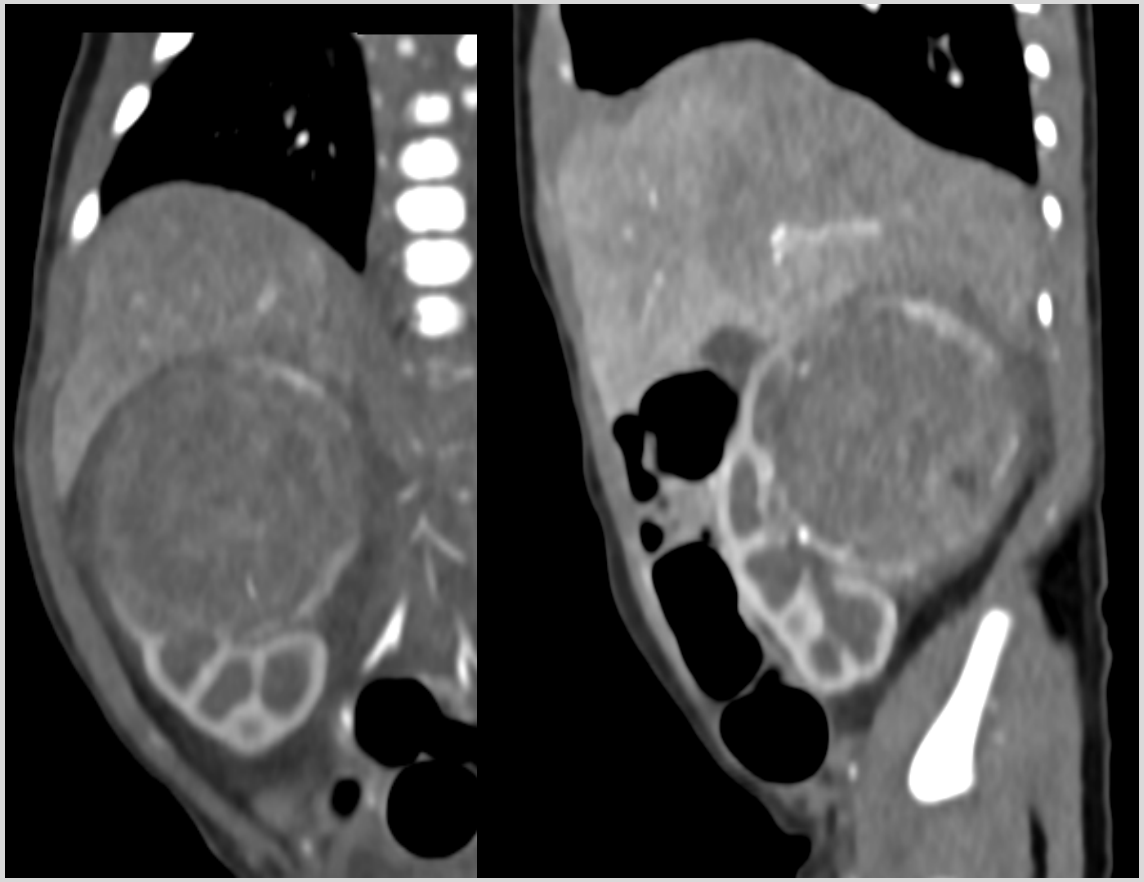
Diagnostic criteria: Wilms vs Non-Wilms











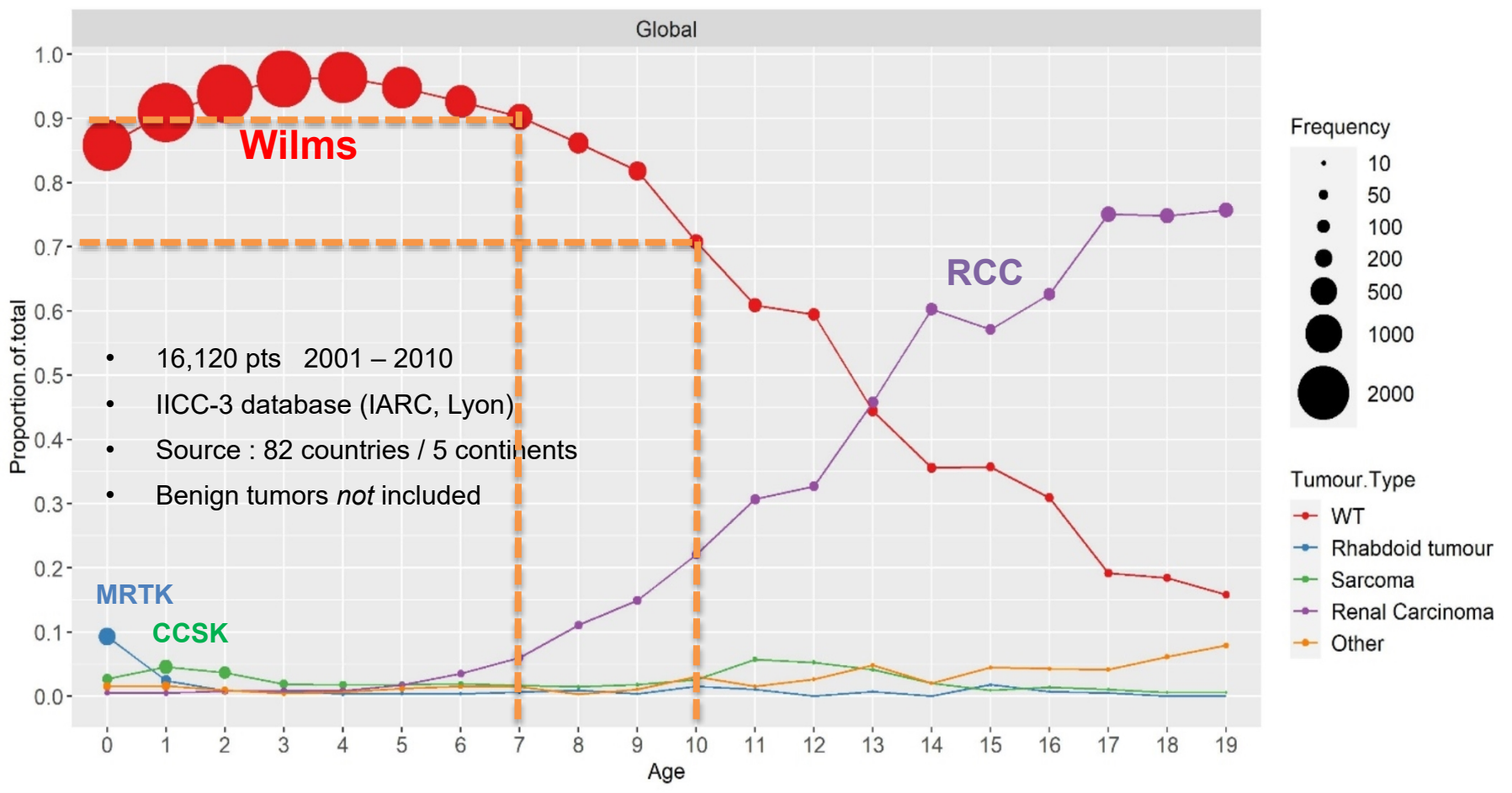
CMN: infiltrative pattern

Infants < 3 months: upfront surgery

Diagnostic criteria: Wilms vs Non-Wilms

Best criterion:

Patient's **age** !

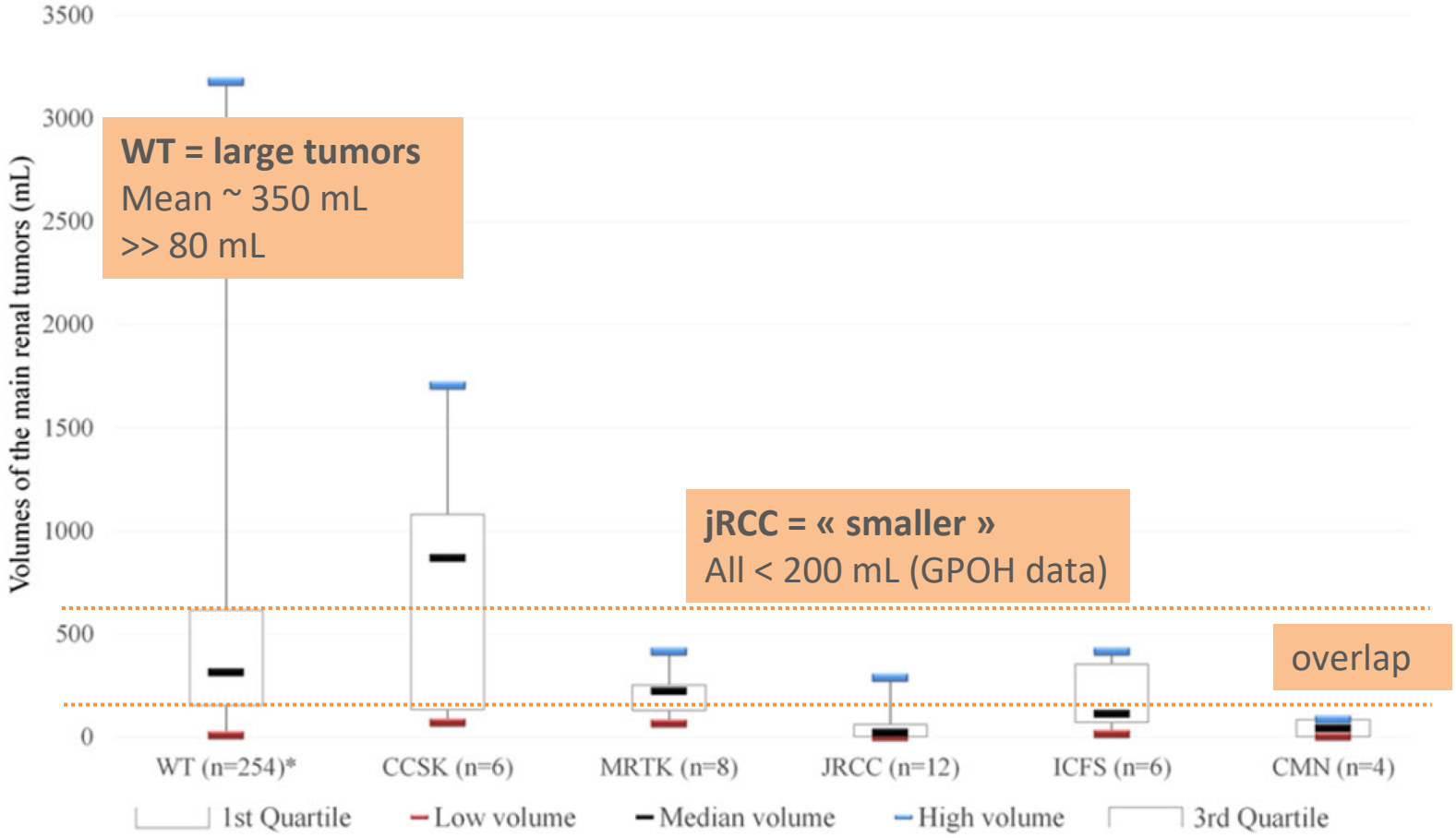


- Nakata K, Colombet M, Stiller CA, Pritchard-Jones K, Steliarova-Foucher E. Incidence of childhood renal tumours: An international population-based study. **Int J Cancer. 2020**
- Jackson TJ, Brisse HJ, Pritchard-Jones K, Nakata K, Morosi C, Oue T, Irtan S, Vujanic G, van den Heuvel-Eibrink MM, Graf N, Chowdhury T; SIOP RTSG Biopsy Working Group. How we approach paediatric renal tumour core needle biopsy in the setting of preoperative chemotherapy. **Pediatr Blood Cancer. 2022**

Diagnostic criteria: Wilms vs Non-Wilms

Tumor Volume

Useful criterion
depsite overlaps



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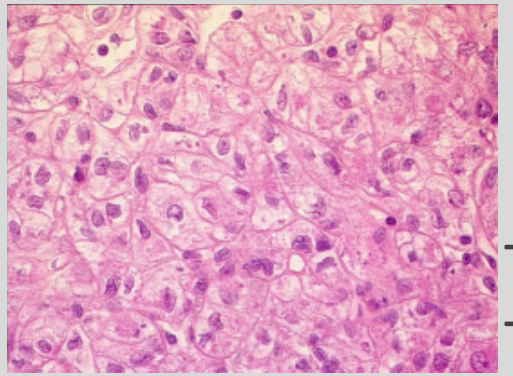
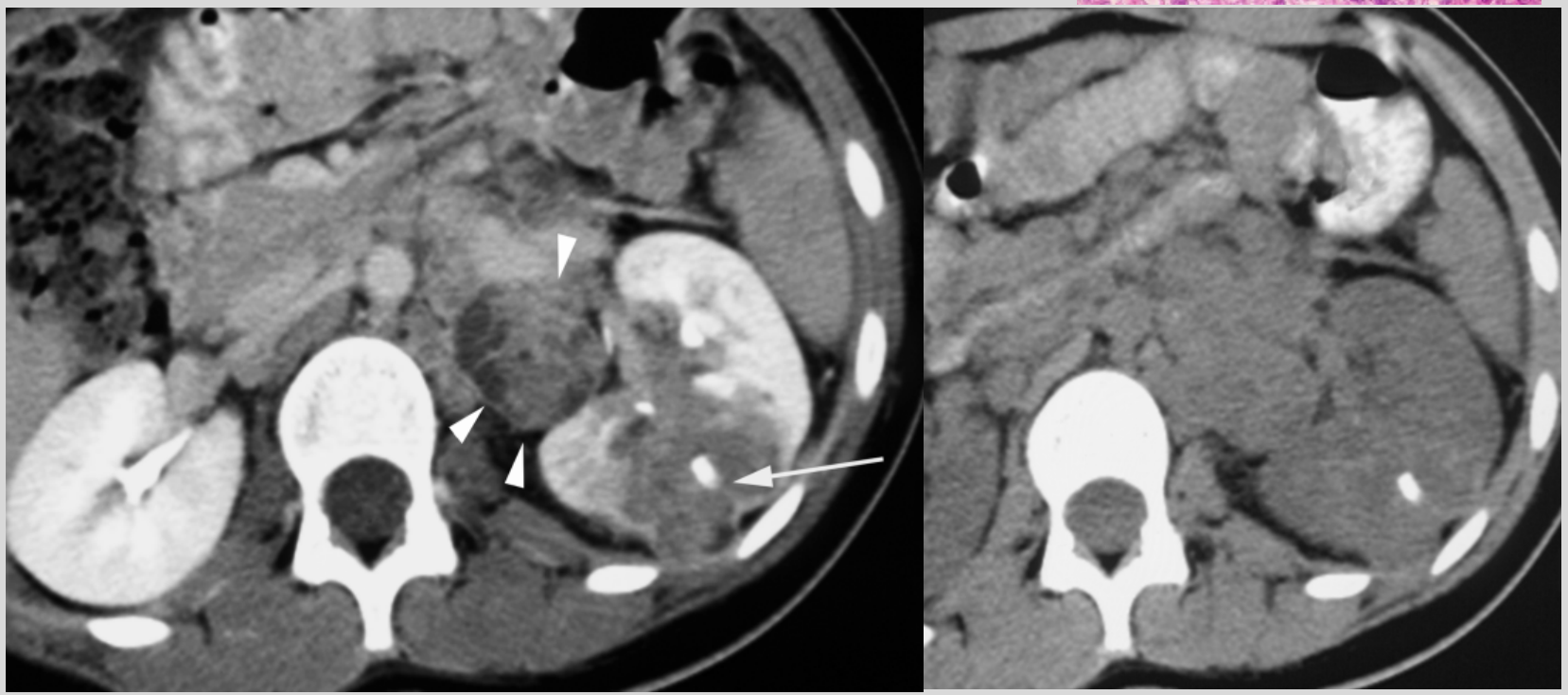
Diagnostic criteria: Wilms vs Non-Wilms

Juvenile RCC

- > 10 y
- Vol < 200 mL
- +/- Calcifications
- Large LN

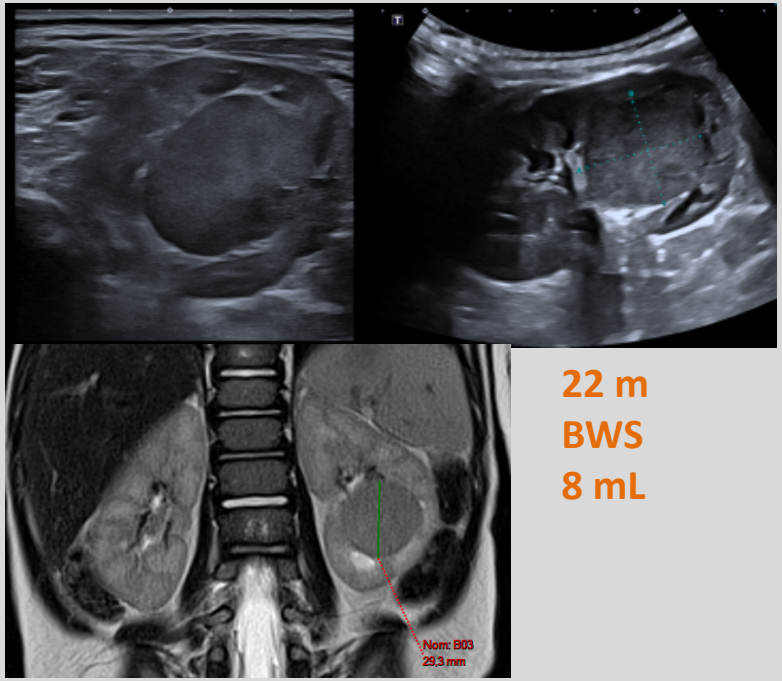


TFE3
TFEB



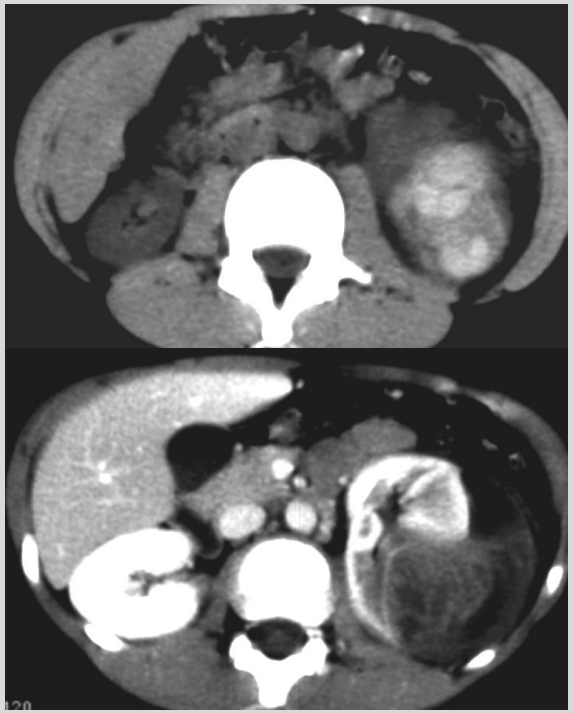
Diagnostic criteria: Wilms vs Non-Wilms

Tumor Volume criterion: Not always relevant (small WT)

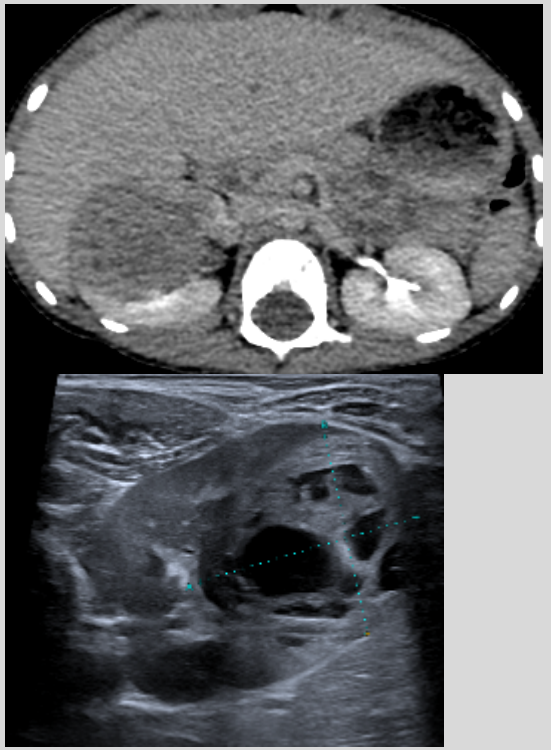


22 m
BWS
8 mL

Diagnosis from **US screening**
Genetical predisposition



Acute pain from retroperitoneal **bleeding**

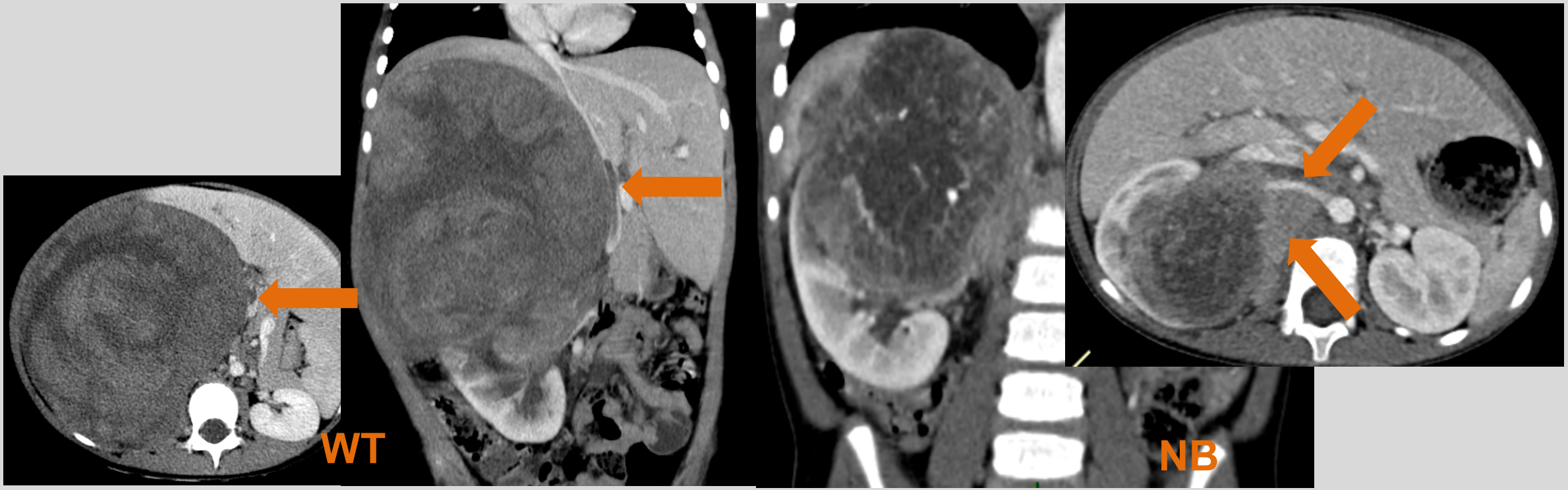


Central tumor revealed by macroscopic **hematuria**

Diagnostic criteria: Wilms vs Non-Wilms

Anatomic location:
Beware the upper-pole

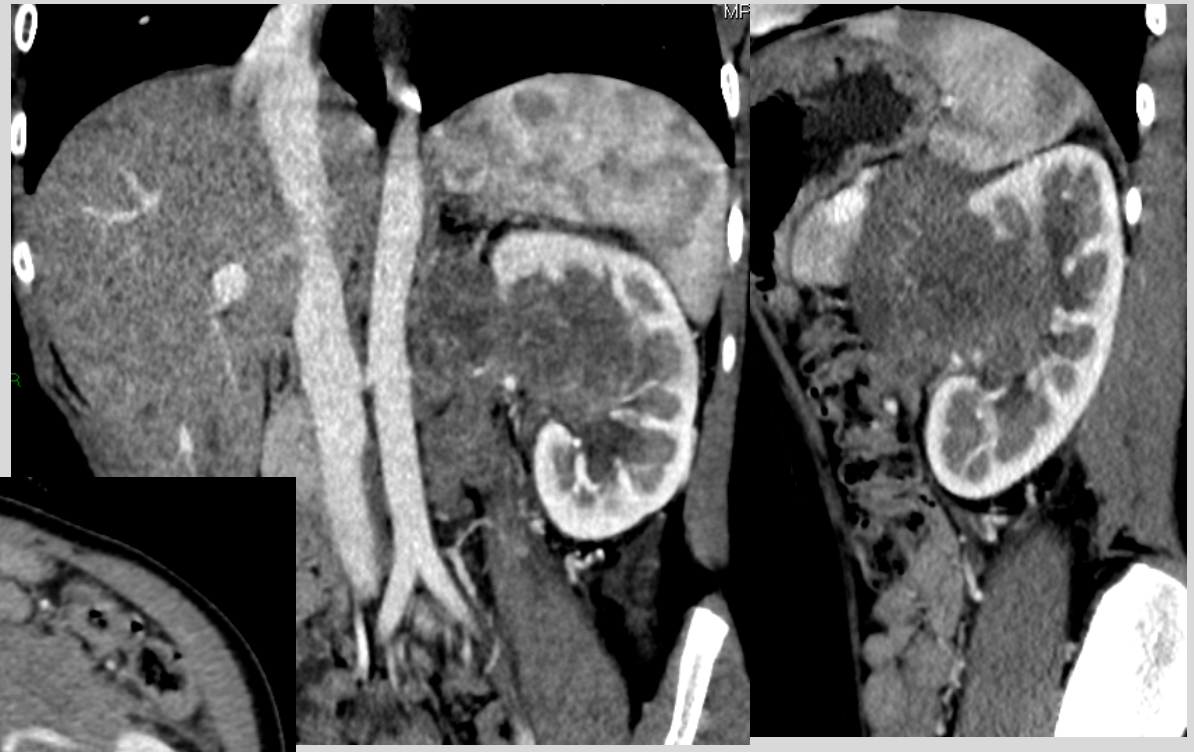
NB and WT occur at the same age !

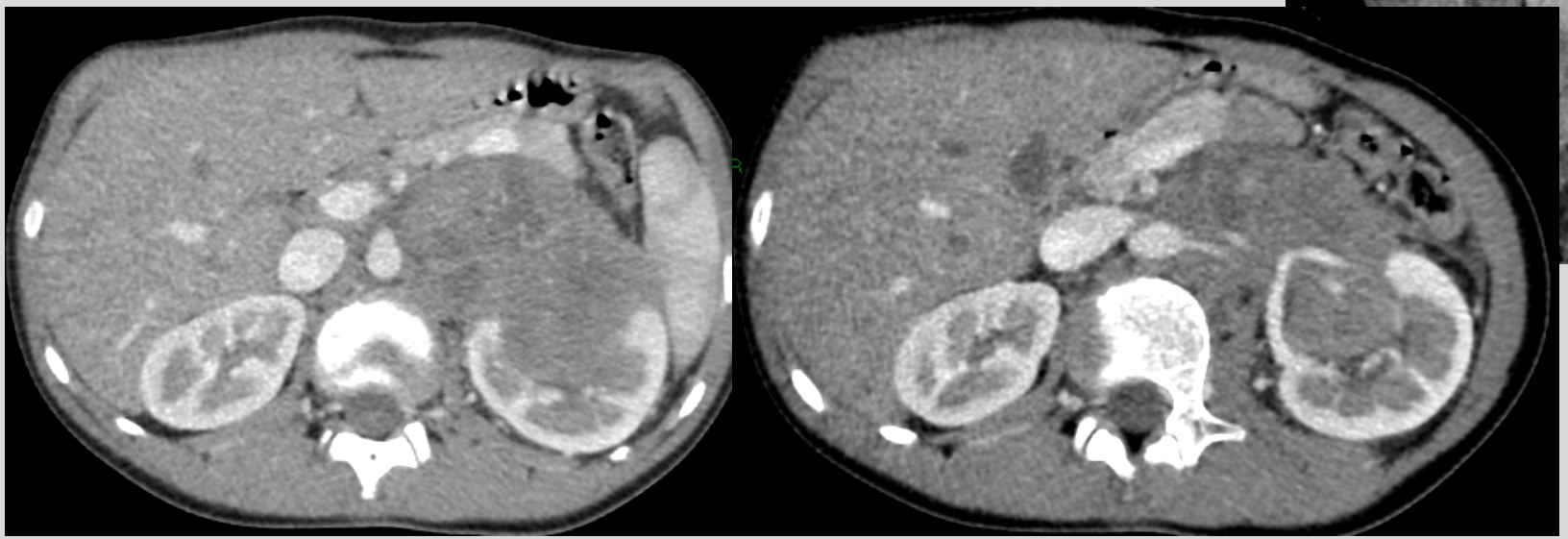


Diagnostic criteria: Wilms vs Non-Wilms

Anatomic location

Beware the renal hilum

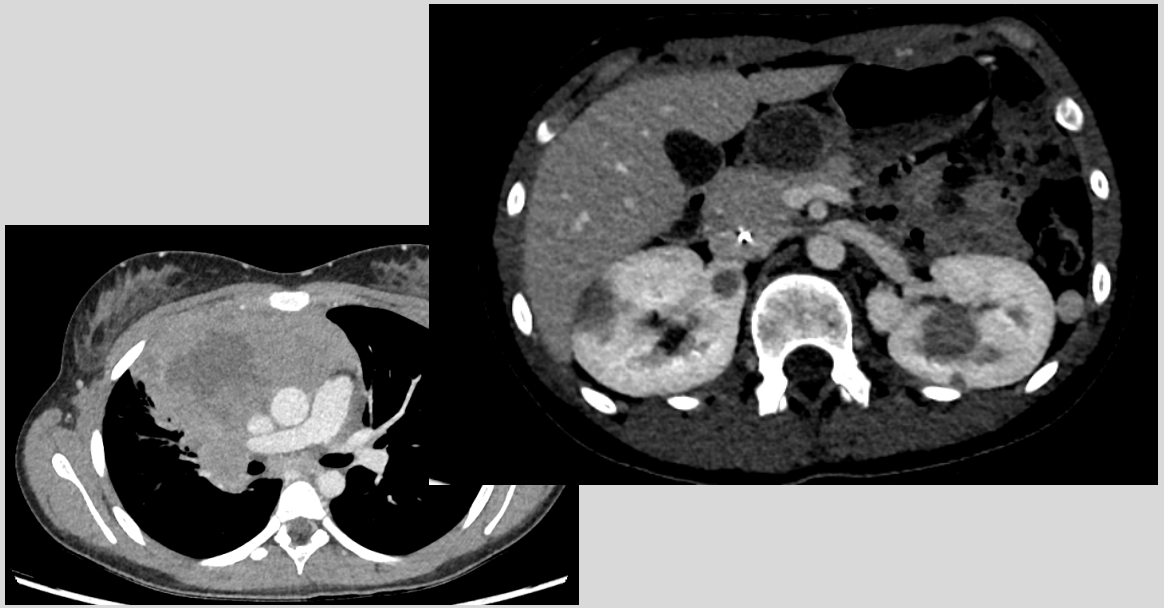


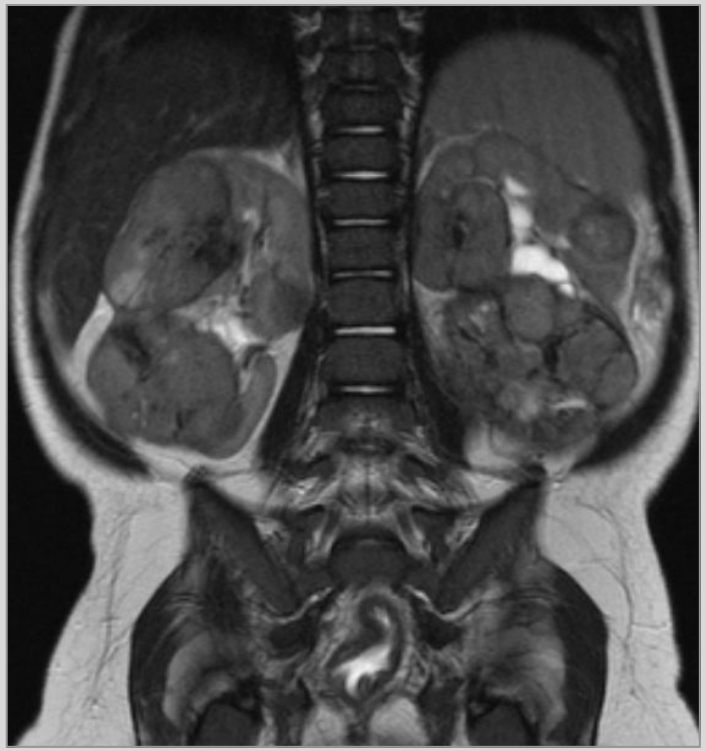


Renal invasion from NB is common

Diagnostic criteria: Wilms vs Non-Wilms

Bilateral/multifocal disease: patients' age ?





> 7 years

- Beware **hematologic malignancies** (NHL, ALL)
- Check : Other locations & LDH
  - ➢ Myelogram or biopsy

< 7 years

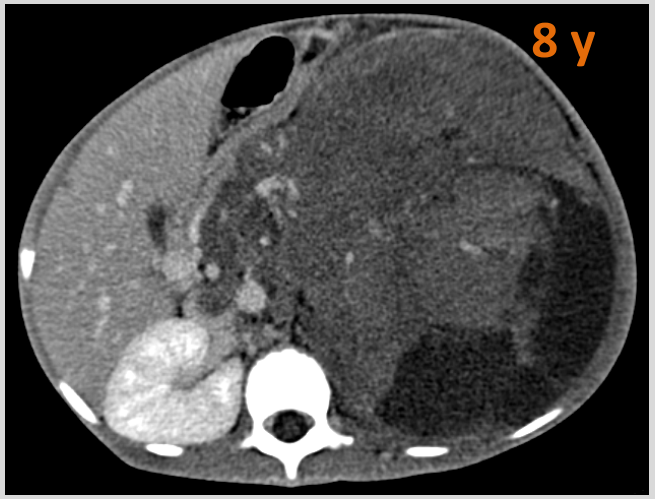
- Probably **Nephroblastomatosis +/- WT**
- Check : predisposing syndrome, proteinuria, cysts
  - ➢ Presumptive chemotherapy

Diagnostic criteria: Wilms vs Non-Wilms

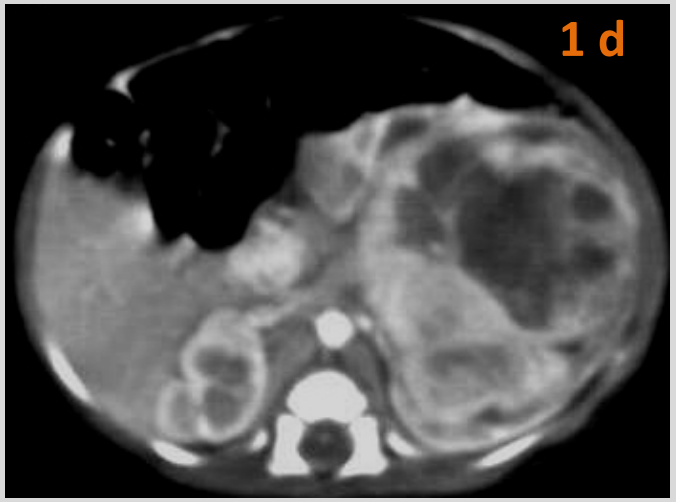
Tumor structure

Solid tumor + necrosis

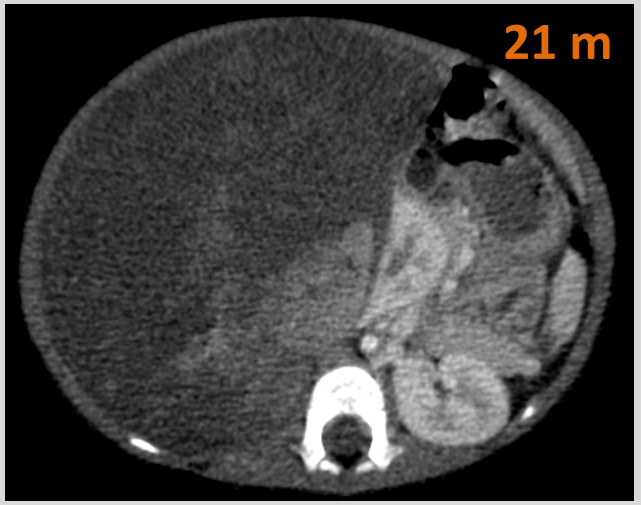
Not discriminatory



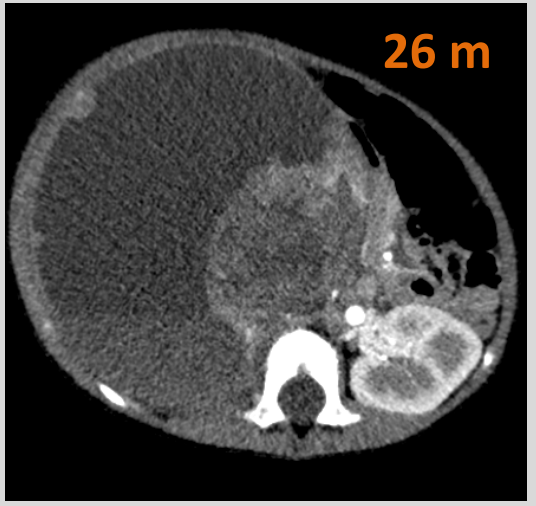
WT



cCMN/IFS



CCSK

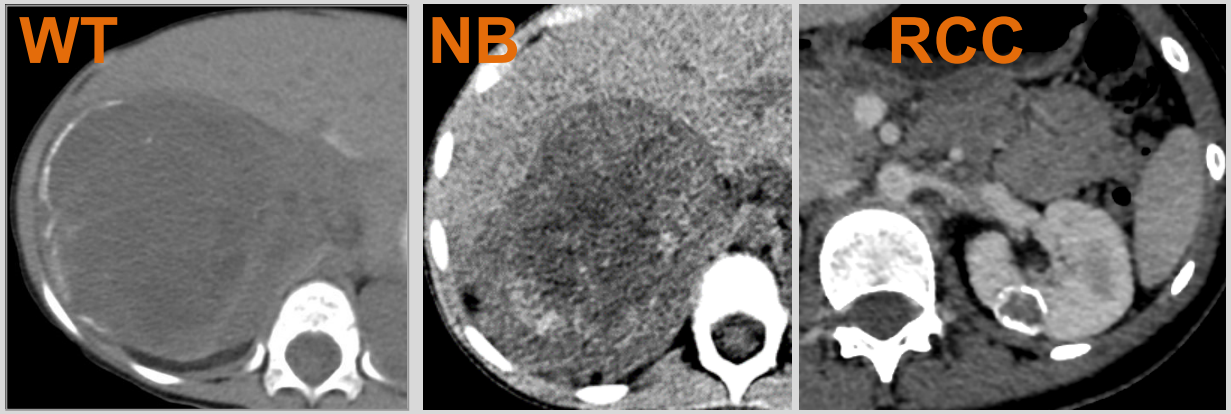


MRTK

Diagnostic criteria: Wilms vs Non-Wilms

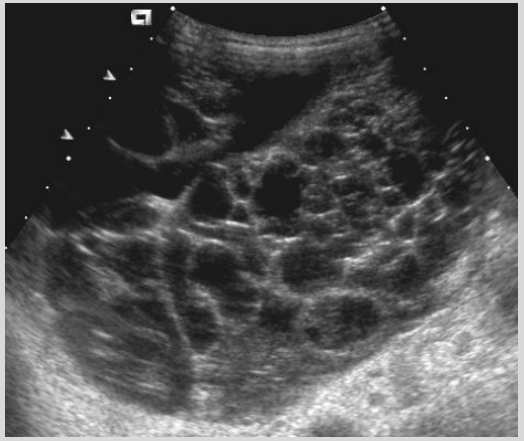
Tumor structure

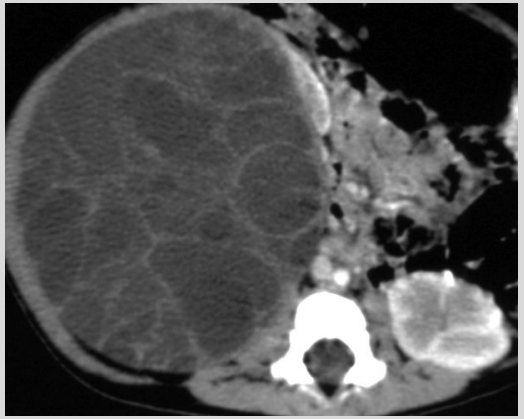
Specific situations

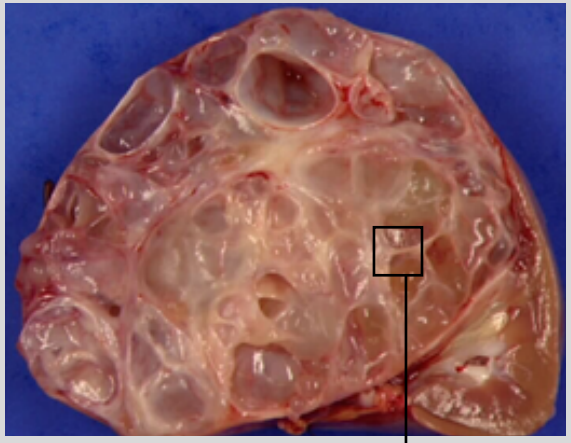


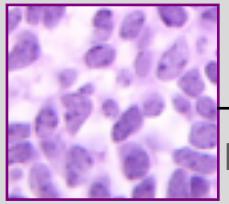
Tumor calcifications

- Analyze number/morphology









Blastemal cells ?

Yes → **WT** / **CPDN**

No → **CN**

Completely Cystic tumors

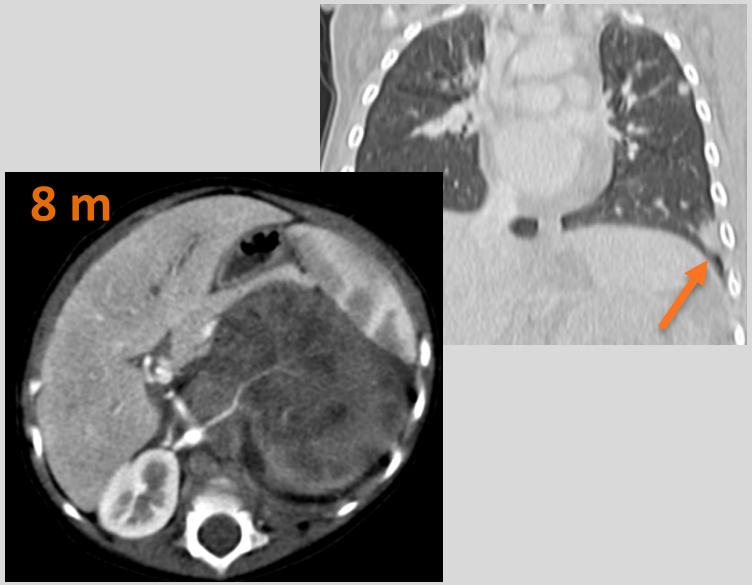
- Upfront surgery

Diagnostic criteria: Wilms vs Non-Wilms

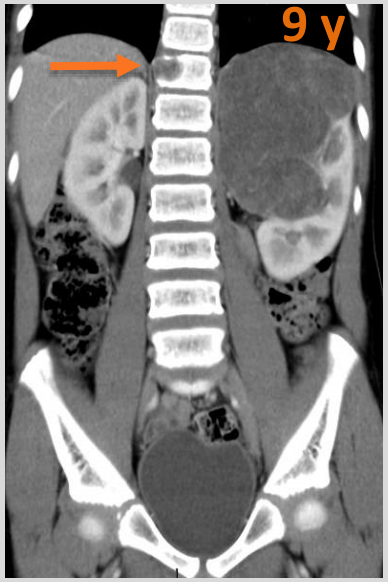
Metastases

Common sites for **WT** : **lung** (15%), liver (2%)

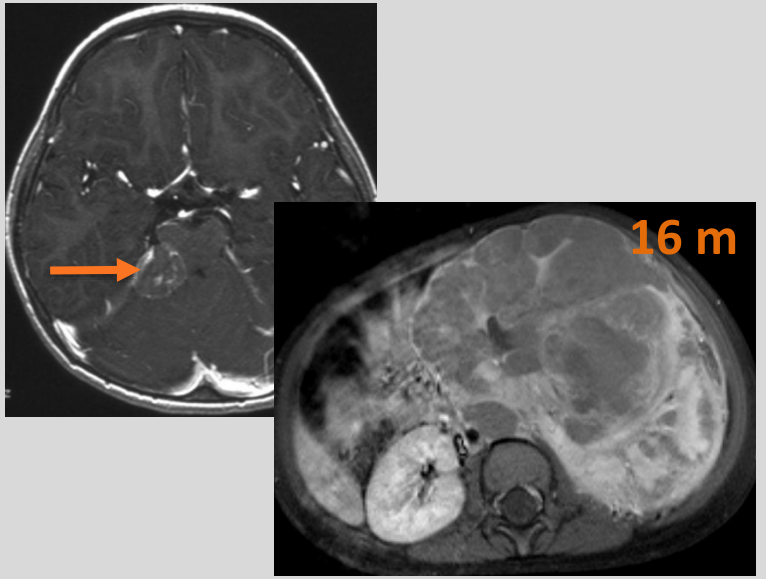
Beware *uncommon* age & sites



Lung < 2 years
MRTK



Bone
CCSK, jRCC



CNS
MRTK, CCSK

SIOP-RTSG Guidelines for CNB of kidney tumors

(genetic predisposition excluded)

	Biopsy NOT recommended **Features typical of WT** ALL criteria required	**Biopsy NOT recommended** if ANY of these criteria met	**Biopsy recommended** **if ANY of these criteria met**	**Indication to be discussed in TBM if ANY of these criteria**
Clinical criteria	**6m ≤ Age < 7y** *No* infectious syndrome	**Age < 3 m** (upfront surgery)	**Age ≥ 10 y** 7 ≤ Age <10 y + *vol* < 200mL	3 m ≤ Age < 6 m Infectious syndrome Urinary tract infection
Radiological criteria	**Obvious renal origin** **Unilateral tumor** **Tumor volume > 80 mL** **Solid or mixed** (solid and cystic) tumor No calcification Metastases absent or lung mets *and* age ≥ 2y	**Totally cystic** tumor (primary surgery) **Bilateral** kidney tumors or Nephroblastomatosis *and* age <7 y (presumptive chemotherapy)	**Uncertain renal origin** **Atypical metastases** for WT: - Bones - CNS - Lungs *and* age < 2y	Intra-tumor calcifications Tumor volume < 80 mL Major necrotic adenopathy Bilateral tumors and age > 7 y
Biological criteria	Normal urinary catecholamines Normal serum calcium LDH < 4 N		Elevated urinary **catecholamines** **Hypercalcemia** *and* age <4 y	**LDH > 4 N**

Primary Surgery

Neoadj. Chemo

Biopsy

Jackson TJ, Brisse HJ, Pritchard-Jones K, Nakata K, Morosi C, Oue T, Irtan S, Vujanic G, van den Heuvel-Eibrink MM, Graf N, Chowdhury T; SIOP RTSG Biopsy Working Group. How we approach paediatric renal tumour core needle biopsy in the setting of preoperative chemotherapy. **Pediatr Blood Cancer. 2022**

Conclusion - take home messages

- **Age** at diagnosis is a basic criterion

- **Essential role of imaging**
  - To first confirm the renal origin
  - To assess tumor structure, local extension, metastases (including skeletum)
  - To measure the tumor volume and check the other kidney

- **Biology** may help: calcium, LDH (+ catecholamines, always !)

- **Biopsy** must be a collegial decision (**TBM)**