

**Table S1.** Search terms

<b>Embase</b>	(bilateral:ab,ti OR (stage AND v:ab,ti) OR (stage AND 5:ab,ti)) AND (wilm*:ab,ti OR nephroblastom*:ab,ti) (bilateral;ab,ti OR (stage AND v:ab,ti) OR (stage AND 5:ab,ti)) AND (CCSK:ab,ti OR clear cell sarcoma of the kidney:ab,ti) (bilateral:ab,ti OR (stage AND v:ab,ti) OR (stage AND 5:ab,ti)) AND (MRTK:ab,ti OR malignant rhabdoid tumor of the kidney:ab,ti) (bilateral:ab,ti OR (stage AND v:ab,ti) OR (stage AND 5:ab,ti)) AND (RCC:ab,ti OR renal cell carcinoma:ab,ti) (bilateral:ab,ti OR (stage AND v:ab,ti) OR (stage AND 5:ab,ti)) AND (CN:ab,ti OR cystic nephroma:ab,ti) (bilateral:ab,ti OR (stage AND v:ab,ti) OR (stage AND 5:ab,ti)) AND (CPDN:ab,ti OR cystic partially differentiated nephroblastoma:ab,ti) (bilateral:ab,ti OR (stage AND v:ab,ti) OR (stage AND 5:ab,ti)) AND (CMN:ab,ti OR MN:ab,ti) OR (mesoblastic nephrom*:ab,ti)
<b>PubMed</b>	("bilateral"[Title/Abstract] OR "stage v"[Title/Abstract] OR "stage 5"[Title/Abstract]) AND ("wilm*" [Title/Abstract] OR "nephroblastom*" [Title/Abstract]) ("bilateral"[Title/Abstract] OR "stage v"[Title/Abstract] OR "stage 5"[Title/Abstract]) AND ("CCSK"[Title/Abstract] OR "clear cell sarcoma of the kidney" [Title/Abstract]) ("bilateral"[Title/Abstract] OR "stage v"[Title/Abstract] OR "stage 5"[Title/Abstract]) AND ("MRTK"[Title/Abstract] OR "malignant rhabdoid tumor of the kidney" [Title/Abstract]) ("bilateral"[Title/Abstract] OR "stage v"[Title/Abstract] OR "stage 5"[Title/Abstract]) AND ("RCC"[Title/Abstract] OR "renal cell" [Title/Abstract]) AND ("child*" [Title/Abstract] OR "pediatr*" [Title/Abstract]) ("bilateral"[Title/Abstract] OR "stage v"[Title/Abstract] OR "stage 5"[Title/Abstract]) AND ("CN"[Title/Abstract] OR "cystic nephrom*" [Title/Abstract]) AND ("child*" [Title/Abstract] OR "pediatr*" [Title/Abstract]) ("bilateral"[Title/Abstract] OR "stage v"[Title/Abstract] OR "stage 5"[Title/Abstract]) AND ("CPDN"[Title/Abstract] OR "cystic partially differentiated nephroblastom*" [Title/Abstract]) ("bilateral"[Title/Abstract] OR "stage v"[Title/Abstract] OR "stage 5"[Title/Abstract]) AND ("CMN"[Title/Abstract] OR "MN"[Title/Abstract] OR "mesoblastic nephrom*")

**Table S2.** Detailed information of 24 patients with bilateral renal tumors.

Pt No.	Sex	Age (mths)	Metastasi zed disease	HTN at Dx	Preop CT	Tumor volume response (%)		Surgery type		Local stage		Tumor histology		Postop therapy		Outcome		eGFR last FU (ml/min/ 1.73m²)	FU (mths)
						L	R	L	R	L	R	L	R	CT	RTx	Relap se	Death		
1	F	44	No	No	VA	-91	-93	NSS	NSS	I	III	RT-WT	RT-WT	Yes	Yes	No	No	>90	74
						NA	NA					NB (PLNR)	NB (PLNR)						
2	M	46	No	Yes	VA-D	-93	-73	NSS	TN	I	I	RT-WT	RT-WT	Yes	No	No	No	>90	66
3	M	52	No	Yes	VA	-87	-66	NSS	TN	III	I	BT-WT	ST-WT	Yes	Yes	No	Yes	<15	20 (died)
						-100	NA					RT-WT	NB (PLNR)						
4	F	11	No	Yes	VA-CE	+43	-30.5	TN	NSS (3y later)	I	III^	ST-WT	ST-WT (2nd tumor)	Yes	No	No <sup>#</sup>	No	>90	58
							-6.5						NB (ILNR)						
5	F	56	No	No	VA	-90	-95	B	TN	I	II	CNT-WT	DA-WT	Yes	No	No	No	>90	51
							-33						RT-WT						
						NA	-15					NB (PLNR)	TLNR*						
							NA					NB (PLNR)							
6	M	57	Yes (osseous Th10)	Yes	VAD	-78	-77	NSS	NSS	III	I	RT-WT	CNT-WT	Yes	Yes	No	No	>90	33
						Stable	-80					NB (PLNR)	NB (PLNR)						
7	F	59	Yes (pulmonar y)	Yes	VAD	-79	-98	TN	NSS	III	I	DA-WT	RT-WT	Yes	Yes	No	No	89	27
						-67	NA					DA-WT	NB (PLNR)						
						NA						NB (PLNR)							
8	M	49	Yes (pulmonar y)	Yes	VA-D	+29	-59	TN	None	III	NA	DA-WT	DA-WT	Yes	Yes	No	Yes	<15	12 (died)
9	F	15	No	Yes	VA-D	-77	-91	NSS	NSS	NA	I	PLNR*	BT-WT	Yes	No	No	No	>90	34
							NA						NB (PLNR)						

10	F	21	No	Yes	VA	-100	-91	None	TN	NA	III	PLNR <sup>1</sup>	RT-WT	Yes	Yes	No	No	>90	63
							-97						RT-WT						
							-94						NB (PLNR)						
							-100						PLNR						
11	F	54	No	No	VA	-78	↓ <sup>3</sup>	TN	B	I	NA	RT-WT	NR*	Yes	No	No	No	>90	49
						NA						PLNR							
12	F	51	No	Yes	VA	-75	>-10 <sup>2</sup>	TN	NSS	II	NA	RT-WT	PLNR*	Yes	No	No	No	>90	48
						NA						RT-WT							
						NA						NB (PLNR)							
13	F	38	No	Yes	VA	Stable	-71	B	TN	NA	I	NB	RT-WT	Yes	No	No	No	>90	60
							NA						NB (PLNR+ILNR)						
14	M	9	No	Yes	VA-D	-1.4	-60	TN	NSS	I	NA	ST-WT	NB (ILNR)*	Yes	No	No	No	>90	33
15	F	12	No	Yes	VA-CE	↓ <sup>3</sup>	+164	None	TN	II	NA	NR <sup>1</sup>	ST-WT	Yes	No	No	No	>90	29
							+48.2						ST-WT						
16	M	19	No	No	VA	-90	↓ <sup>3</sup>	NSS	None	NA	I	MT-WT	NB (PLNR) <sup>1</sup>	Yes	No	No <sup>#</sup>	No	>90	28
						↓ <sup>3</sup>						NB (PLNR)							
17	F	24	No	No	VA	-77	Stable	TN	None	II	NA	ST-WT	NB (ILNR) <sup>1</sup>	Yes	No	No	No	>90	21
						NA						ILNR							
18	M	7	No	Yes	VA	Stable	+59	None	TN	NA	I	NB <sup>1</sup>	ST-WT	Yes	No	No <sup>#</sup>	No	>90	19
							NA						ILNR						
19	F	51	Yes (m. psoas + pulmonary + osseous Th12)	Yes	VAD	Stable	-91	None	TN	NA	III	NB <sup>1</sup>	RT-WT	Yes	Yes	No	No	>90	36
20	M	17	No	Yes	VA	-34.5	+145	None	TN	NA	III	NB <sup>1</sup>	ST-WT	Yes	Yes	No	No	>90	32

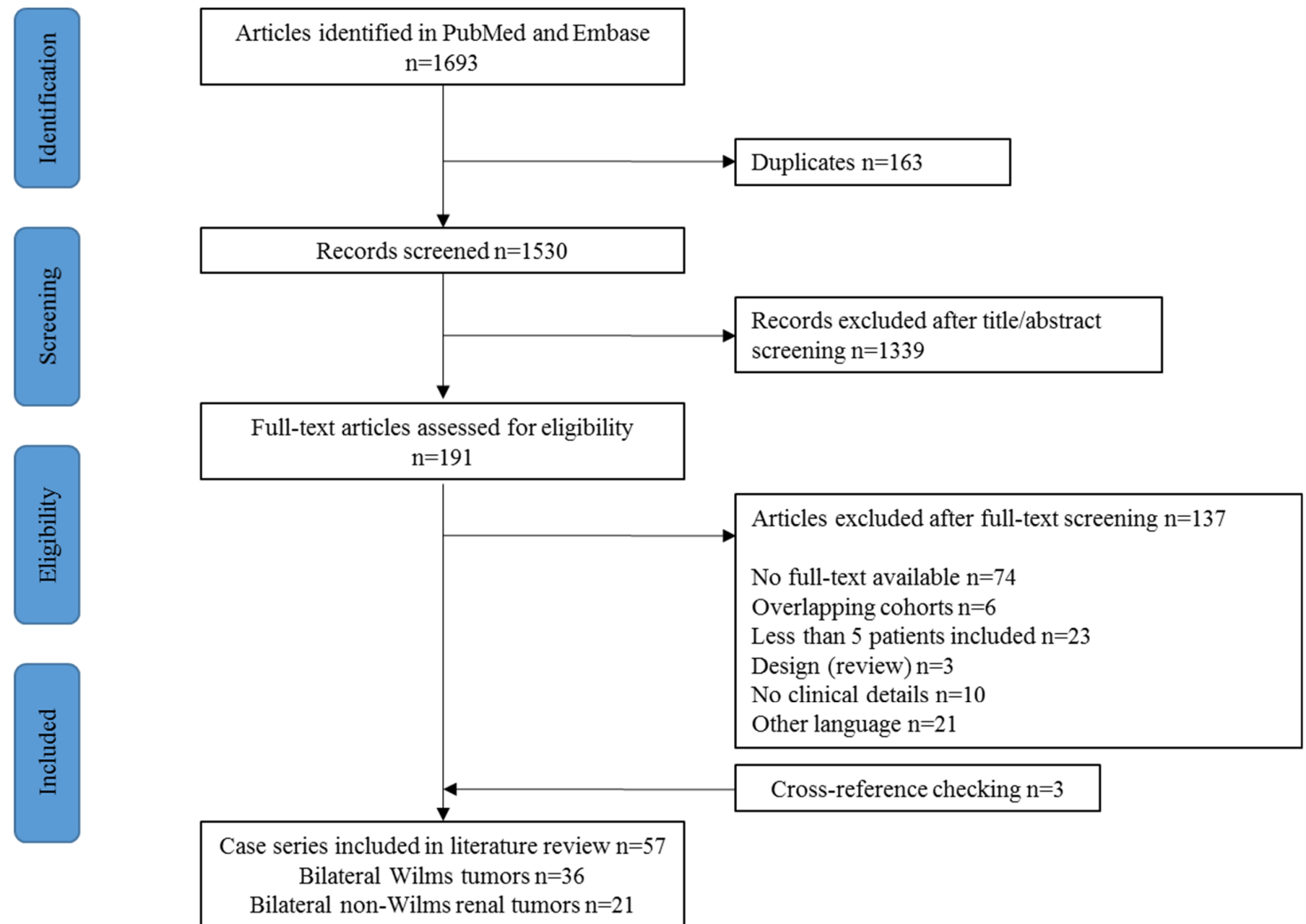
							Stable						NB (PLNR+ILNR)						
21	M	12	No	Yes	VA	↓ <sup>3</sup>	↓ <sup>3</sup>	None	B	NA		NB <sup>1</sup>	NB <sup>1</sup>	NA	No	No	No	>90	36
22	M	24	No	No	VA	>-10 <sup>2</sup>	-97	NSS	NSS	NA		NB (PLNR)	NB (PLNR)	Yes	No	No	No	>90	23
23	M	25	No	Yes	VA	Stable	Stable	None	None	NA		NB <sup>1</sup> (PLNR+ILNR)	NB <sup>1</sup> (PLNR+ILNR)	NA	No	No	No	>90	62
24	F	184	No	No	No	NA	NA	None	TN	NA	I	NA	Papillary RCC type 2	No	No	No	No	>90	60

Pt. no: patient number; mths: months; Dx: diagnosis; M: male; F: female; HTN: hypertension; NSS: nephron sparing surgery; TN: tumornephrectomy; B: biopsy; RT: regressive type; ST: stromal type; BT: blastemal type; DA: diffuse anaplastic; CNT: completely necrotic type; MT: mixed type; WT: Wilms tumor; NB: nephroblastomatosis; NR: nephrogenic rest; PLNR: perilobar nephrogenic rest; ILNR: intralobar nephrogenic rest; TLNR: tumor like nephrogenic rest; preop: preoperative; postop: postoperative; CT: chemotherapy; RTx: radiotherapy; eGFR: estimated glomerular filtration rate; VA: vincristine/actinomycin-D; VAD: vincristine/actinomycin-D/doxorubicin; VA-D: vincristine/actinomycin-D/addition of doxorubicin for stable disease; VA-CE: vincristine/actinomycin-D/addition of carboplatin/etoposide for stable disease; <sup>1</sup>: no histological confirmation of diagnosis; Th: thoracic vertebra; ^: only nephrogenic rest in resection margins, no Wilms tumor cells; †: development of Wilms tumor during follow-up in a previously completely treated (suspected) nephroblastomatosis lesion; \*: diagnosis of nephroblastoma cannot be excluded, but nephrogenic rest is preferred diagnosis; <sup>2</sup>: Volume after chemotherapy was too small for reliable volume assessment; <sup>3</sup>: decrease in size of the nephrogenic rest, dimensions were not specified; FU: follow-up; NA: not applicable; RCC: renal cell carcinoma

**Table S3.** Presenting symptoms

<b>Presenting symptom</b>	<b>Frequency (%)</b>
Abdominal mass	14 (58%)
Abdominal pain	6 (25%)
Asymptomatic	6 (25%)
Surveillance for underlying syndrome	5 (21%)
Incidental finding upon traumatic workup	1 (4%)
Fever	2 (8%)
Loss of appetite	2 (8%)
Hematuria	1 (4%)
Fatigue	1 (4%)
Constipation	1 (4%)
Headache	1 (4%)

**Figure S1.** PRISMA flow diagram of article selection



**Table S4.** Stage V renal tumor patients with metastatic disease

<b>Author</b>	<b>No. of patients with metastasis</b>	<b>Histology</b>	<b>Outcome (Histology)</b>
Aydin 2019 [1]	4	NA	NA
Ehrlich 2017 [2]	27	3 anaplasia n.o.s.	NA (at least 3 anaplasia died)
Davidoff 2015 [3]	7	At least 1 DA At least 1 FH n.o.s.	2/7 died (DA and FH)
Hubertus 2015 [4]	4	2 blastemal, 2 NA	2/4 died (blastemal)
Agarwala 2014 [5]	3	NA	NA
Furtwängler 2014 [6]	19	NA	NA
Oue 2014 [7]	2	NA	NA
Hadley 2013 [8]	4	NA	NA
Indolfi 2013 [9]	11	NA (No DA)	NA
Sulkowski [10]	1	NA	NA
Sudour 2012 [11]	5	NA	NA
Halim 2012 [12]	5	3 UH n.o.s. 2 FH n.o.s.	5 (3 UH, 2 FH)
Millar 2011 [13]	4	NA	4/4 died
Hamilton 2011 [14]	16	NA	NA
Weirich 2004 [15]	3	NA	NA
Kubiak 2004 [16]	4	NA	NA
Tomlinson 1999 [17]	1	NA (No DA)	1/1 died (NA)
Kumar 1998 [18]	9	NA	NA
Shearer 1993 [19]	4	NA	NA
Montgomery 1991 [20]	35	NA	NA
Coppes 1989 [21]	5	NA	NA
Cohen 1986 [22]	4	NA	NA
Asch 1985 [23]	3	NA (No DA)	2/3 died (NA)
Jones 1982 [24]	4	NA (No DA)	4/4 died (NA)
This cohort, 2021	4	1 Regressive bilateral	1/4 died (DA)

		1 Blastemal + stromal	
		2 bilateral DA	
<b>Total</b>	188	162 NA, 16 specified	24/35 died

No.: number; NA: not available; DA: diffuse anaplasia; FH: favorable histology; n.o.s.: not otherwise specified; UH: unfavorable histology.



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