Genetic defects in pediatric renal tumors and their associated syndromes

Renal Tumor	Associated Genetic Defect	Any Known Syndromes
Wilms' tumor	11p13/WT1; also PAX6 gene	WAGR
	11p13/WT1	Denys-Drash, Frasier
	11p15	Beckwith-Wiedemann
	Xp26/GPC3	Simpson-Golabi-Behmel
	5q35/NSD1	Sotos
	17q12-21	Familial WT1
	19q13.4	Familial WT2
	Unknown	Perlman
Cellular CMN	t(12;15)(p13;q25)–ETV6-NTRK3 fusion	
	Trisomy 8, 11, 17	
Malignant rhabdoid tumor	22q deletion, mutation or loss of hSNFS/INI1 gene	Rhabdoid predisposition syndrome
Renal PNET	t(11;22)(q24;q12) (EWS-FLI1)	
	t(21;22)(q22;q12) (EWS-ERG)	
	t(7;22)(p22;q11.2) (EWS-ETV1)	
	t(17;22)(q12;q12) (EWS-E1AF)	
	t(2;22)(q33;q12) (EWS-FEV)	
	t(X;22)(q27;q11) (EWS-?)	
Synovial sarcoma	t(X;18)(p11;q11) (SYT-SSX)	
RCCs		
Xp11.2 translocation	t(X;17)(p11.2;q25) (ASPL-TFE3)	
	t(X;1)(p11.2;q21) (PRCC-TFE3)	
	t(X;1)(p11.2;p34) (PSF;TFE3)	
	t(X;17)(p11.2;q23) (CLTC-TFE3)	
t(6;11) carcinomas	t(6;11)(p21;q12) (Alpha-TFEB)	
PRCC	Chromosome 7, 17 gains	
Adult-type RCC	VHL locus