	Study population	**Cases**	**SDH negative immunohistochemistry**	**Loss of heterozygosity (LOH)**
Morris MR et al., 2004[1]	51 RCC and 4 oncocytomas	29 RCC	0/0	0/29
Vanharanta S et al., 2004[2]	352 Pheo/PGL patients	3 RCC in *SDHB* patients (2/3 with PGL) 60 sporadic RCC 35 kidney cancers with clear cell histology	0/0	3 *SDHB* mutations/3 0/60 0/35
Benn DE et al., 2006[3]	116 patients (83 affected and 33 clinically unaffected) from 62 families with PCC/PGL syndromes and *SDHB* or *SDHD* mutations	1 GIST	1 SDHB-/1	0/0
McWhinney SR et al., 2007[4]	9 *SDHx* GIST	9 GIST	0/0	5 *SDHB* mutations, 2 *SDHC* mutations, 1 *SDHD* mutation/8
Solis DC et al., 2009[5]	41 *SDHB* patients	1 papillary thyroid carcinoma 1 uterine leiomyoma 1 HNPGL + hybrid renal tumor 1 HNPGL + papillary bladder carcinoma 1 HNPGL + basocellular carcinoma precordial 1 lung cancer	0/0	1 *SDHB* mutation in hybrid renal tumor /1 RCC + 0/1 PTC
Gill AJ et al., 2010[6]	121 GIST (104 sporadic, 5 Carney Triad, 3 NF1, 2 cKIT mutated, 1 multifocal GIST, 1 pediatric, 5 in young people)	121 GIST	10 SDHB- (3 sporadic, 5 Carney Triad, 1 pediatric, 1 multifocal)/121	0/0
Housley SL et al., 2010[7]	1 *SDHB* patient affected by RCC	1 RCC	0/0	1 *SDHB* mutation/1
Gaal J et al., 2011[8]	57 GIST (4 Carney Stratakis, 6 Carney Triad, 5 cKIT or PDGFRA mutated, 42	1 medullary thyroid carcinoma in patient with sporadic GIST 57 GIST	0/1 11 SDHB-/57	1 *RET* mutation/1 0/0

	sporadic), 1 medullary thyroid carcinoma		(4/4 Carney Stratakis, 6/6 Carney Triad, 0/5 cKIT or PDGFRA mu, 1/42 sporadic)	
Gill AJ et al., 2011[9]*	4 *SDHB* patients affected by 5 RCC	5 RCC	4 SDHB-/4	0/0
Gill AJ et al., 2011[10]	3 RCC in SDHB patients	3 RCC	3 SDHB-/3	0/0
Janeway KA et al., 2011[11]	34 GIST (30 wt patients, 1 *SDHC*, 3 *SDHB*)	34 GIST	2 SDHB- (2 *SDHB* patients)/3 (2 *SDHB* patients, 1 *SDHC* patient)	0/0
Miettinen M et al., 2011[12]	1134 GIST	1134 GIST	66 SDHB-/1134	0/0
Alvarenga CA et al., 2012[13]	1 patient affected by chromophobe RCC + PGL	1 chromophobe RCC	0/0	0/1
Doyle LA et al. 2012[14]	264 GIST (179 cKIT mutated, 32 PDGFRA mutated, 53 wt)	264 GIST	22 SDHB-/264	0/0
Italiano A et al. 2012[15]	28 GIST in young adults	28 GIST	4 SDHB- (3/4 SDHB-/SDHA-)/7	3 *SDHA* mutations/28
Malinoc A et al., 2012[16]	35 *SDHC* patients + 1 proband's mother	2 RCC (1 clear cell cancer, a papillary carcinoma,	1 SDHB- (clear cell cancer)/2	2 *SDHC* mutations/2
Xekouki P et al., 2012[17]	1 PA (GH) in *SDHD* patient	1 PA	0/0	1 *SDHD* mutation/1
Xekouki P et al., 2012[18]	1 PA (macroGH) + PGLs + PCCs in *SDHD* patient	1 PA	1 SDHB-/1	1 *SDHD* mutation/1
Belinsky MG et al., 2013[19]	24 GIST (12 wt, 12 KIT/PDGFRA/BRAF mutated)	24 GIST	11 SDHB- (3/11 wt GIST in Carney Triad)/24	4 *SDHA* mutations/6
Dwight T et al., 2013[20]	Colonic polyps + HNPGL 1 (NS) in *SDHA* patient	Colonic polyps 1 PA	0/1 1 SDHB- and A-/1	0/0 0/1
Dwight T et al., 2013[21]	10 patients with SDH-deficient GIST	10 GIST	10 SDHB- (3/10 SDHB- and A-)/10	3 *SDHA* mutations/10
Gill AJ et al., 2013[22]	GIST + leiomyosarcoma + unclassified renal tumor + PTC in *SDHC* patient	1 leiomyosarcoma 1 PTC 1 GIST 1 RCC	1 SDHB-/3 (1 PTC, 1 GIST, 1 RCC)	0/0
Kim S et al., 2013[23]	721 breast cancers	721 breast cancers	41 SDHA-/319 24 SDHB-/245	0/0

Miettinen M et al., 2013[24]	944 GIST	944 GIST	127 SDHB- (36/127 SDHB-/A-)/944	6 *SDHA* mutations/7
Oudijk L et al. 2013[25]	33 GIST	33 GIST	16 SDHB- (4/16 SDHB-/A-, one associated with metastatic medullary thyroid carcinoma)/33	4 *SDHA* mutations/4
Papathomas TG et al., 2014[26]	348 tumors	5 RCC 2 PA	4 SDHB-/4 1 SDHB-/2	4/4 1/2
Wagner AJ et al. 2013[27]	33 GIST	33 GIST	33 SDHB- (9/33 SDHB- and A-)/33	9 *SDHA* mutations/9
Choi J et al., 2014[28]	206 filloid breast tumors	206 filloid breast tumors	36 SDHB- (5/36 SDHB- and A-)/206	0/0
Fleming S et al., 2014[29]	3 RCC in *SDHB* patients (one previously described[§])	2 RCC	2 SDHB-/2	0/0
Gill AJ et al., 2014[30]	36 RCC[‡] in 27 *SDHx* patients	36 RCC	36 SDHB-/36	0/0
Gill AJ et al., 2014[31]	309 PA	309 PA	1 SDHB-/A- (macro PRL)/309	1 *SDHA* mutation/1
Miettinen M et al., 2014[32]	2258 epithelial tumors	1547 other tumors 711 RCC	3 SDHB- (1 prostate adenocarcinoma, 1 stomach adenocarcinoma, 1 testis seminoma)/1547 4 SDHB- (1 clear cell carcinoma 1 papillary carcinoma 2 unclassified carcinomas)/711	0/0 0/0
Paik JY et al., 2014[33]	1 RCC in *SDHB* patient	1 RCC	1 SDHB-/1	0/0
Pantaleo MA et al., 2014[34]	358 GIST (34 KIT/PDGFRA wt)	34 wt KIT/PDGFRA GIST	6 SDHB– (4 SDHB- and A-)/20	6 *SDHA* mutations, 1 *SDHB* mutation, 1 *SDHC* mutation/34
Renella R et al., 2014[35]	1 *SDHB* patient affected by lymphoma + PGL	1 lymphoma	1 markedly reduced immunoreactivity/1	0/0
Tirumani SH et al., 2014[36]*	34 *SDHx* GIST	34 GIST	34 SDHB-/34	0/0
Cornejo KM et al. 2015[37]	420 renal epithelial tumors	420 renal epithelial tumors	0/420	0/0
Dénes J et al., 2015[38]	39 patients (19 sporadic, 20 familial)	23 PA: 12 PRL, 6 GH, 5 NFPA (5 *SDHB*, 1 *SDHC*, 1	1 SDHB-/7	3 SDHB mutations/5

		SDHA, 1 *SDHAF2*, 15 no mut)		
Jiang Q et al. 2015[39]	1 GIST + chromophobe renal carcinoma + kidney cist in a *SDHA* patient	1 GIST 1 chromophobe renal carcinoma	1 SDHB-/1 1 SDHB-/1	1 *SDHA* mutation/1 1 *SDHA* mutation/1
Ni Y et al., 2015[40]	802 patients affected by differentiated thyroid carcinomas or breast carcinomas + data from TGCA thyroid cancer (THCA) dataset (476 patients)	754 patients	0/0	5 (3 *SDHB* and 2 *SDHD* mutations)/37 apparently sporadic epithelial thyroid carcinomas (7 follicular, 4 follicular variant of papillary histology, 26 PTC)
Niemeijer ND et al., 2015[41]	26 *SDHx* patients affected by 35 non paragangliomic tumors	4 *SDHx* patients with 5 different tumors 2 GIST 3 RCC 2 PA	2 SDHB- (ganglioneuroma and neuroendocrine pancreas tumor)/5 2 SDHB-/2 3 SDHB-/3 2 SDHB- (1/2 SDHB-/A-)/2	1 *SDHD* mutation (neuroendocrine pancreas tumor)/2 1 *SDHD*, 1 *SDHB* mutation/2 3 *SDHB* mutations /3 1 *SDHD* mutation (+ 1 somatic mutation)/2
O'Toole SM et al., 2015[42]	72 literature cases	60 new cases [#]	0/0	2 *SDHB* mutations/ number of cases not known
Ozluk Y et al., 2015[43]	1 metastatic RCC	1 RCC	1 SDHB- and A-/1	1 *SDHA* mutation/1
Pantaleo MA et al., 2015[44]	69 metastatic GIST	11 KIT/PDGFRA wild-type GIST	6 SDHB-/11	6 *SDHA* mutations/6
Williamson SR et al., 2015[45]	37 RCC with histology in line with SDH	37 RCC	11 SDHB- (1 SDHB- and A-/11)/37	5 *SDHB* mutations/6
Yakirevich E et al. 2015[46]	1 RCC	1 RCC	1 SDHB- and A-/1	1 *SDHA* mutation/1
Falzarano SM et al., 2016[47]	7 RCC	7 RCC	0/7	0/0
Kim E et al., 2017[48]	11 RCC, 10 GIST	11 RCC, 10 GIST	11 SDHB-/11 RCC 10 SDHB-/10 GIST	0/0
Mason EF et al., 2016[49][$]	705 GIST	76 GIST	76 SDHB- (28/76 SDHB-/A-)/76	0/0
Belinsky MG et al., 2017[50]	A GIST	1 GIST	1 SDHB-/1	1 *SDHB* mutation/1
Caliò A et al., 2017[51]	4 RCC (3/4 *SDHB* patients)	4 RCC	4 SDHB-/4	0/0

Carter CS et al., 2017[52]	96 cutaneous leiomyomas from 87 patients (12/96 from 7 patients with documented HLRCC)	96 cutaneous leiomyomas from 87 patients	9 FH- (7/9 from HLRCC patients) + 2 equivocal 1 equivocal SDHB /96	0/0
Casey R et al., 2017[53]	12 patients affected by PCC/PGL/HNPGL + RCC (1/12 + breast carcinoma)	1 oncocytoma, 11 RCC	0/0	1 *MAX* mutation/1
De Sousa SMC et al., 2017[54]	44 patients affected by PA	2 parathyroid tumors 1 NFPA 1 microPRL 1 PRL 1 GH + pituitary gangliocytoma 1 PA	0/2 parathyroid tumors 0/1 PA	0/0
Else T et al., 2017[55]	4 *SDHx* cortisol secreting ACC	4 *SDHx* cortisol secreting ACC	0/3	1 possible LOH/4
Elston MS et al., 2017[56]	1 GIST in *SDHB* patient	1 GIST in *SDHB* patient	1 SDHB- and SDHA+/1	1 *SDHB* mutation/1
Gupta S et al., 2017[57]	14 RCC + PGL (3 affected by VHL)	11 RCC	0/11	0/0
Gupta S et al., 2017[58]	1 composite PGL-ganglioneuroma, 7 composite PCC-ganglioneuromas	1 composite PGL-ganglioneuroma, 7 composite PCC-ganglioneuromas	0/8	0/0
Iwashita H et al., 2017[59]	1 RCC	1 RCC	1 SDHB-/1	1 *SDHB* mutation/1
Liu W et al., 2017[60]	355 GIST	355 GIST	12 SDHB- /355	0/0
Schaefer IM et al., 2017[61]	76 GIST	76 GIST	14 SDHB-/22	17 MAX mutations/76
Tufton N et al., 2017[62]	1 pituitary carcinoma (NF) + HNPGL in *SDHB* patient	1 PA	0/0	2 somatic *SDHA* inactivating mutations/1
Kumar R et al., 2018[63]	1 papillary RCC	1 RCC	1 SDHB-/1	0/0
Li Y et al., 2018[64]	33 unclassified RCC	33 RCC	8 SDHB- and A+, 4 FH-/33	0/0
Maher M et al., 2018[65]	1 PA (macroPRL)	1 PA	0/1	Partial retention of the normal allele at the mutation site of *SDHB* gene/1
McEvoy CR et al., 2018[66]	1 RCC	1 RCC	1 SDHB- and A -/1	1 *SDHA* likely pathogenic mutation/1
Michalova K et al., 2018[67]	10 papillary RCC	10 papillary RCC	0/10	0/0
Williamson SR et al., 2018[68]	12 RCC† + 16 clear cell papillary renal cell	12 RCC† + 16 clear cell papillary renal cell	28 SDHB-/34 Equivocal in 1 unclassified	0/0

	carcinoma tumors + 6 unclassified tumors	carcinoma tumors + 6 unclassified tumors	tumor	
Brcic I et al., 2019[69]	1 GIST	1 GIST	1 SHDB- and A- /1	1 *SDHA* mutation/1
Casey RT et al., 2019[70]	15 GIST	15 GIST	10 SDHB-/15	0/0
Chatzopoulos K et al., 2019[71]	6 pulmonary chondroma, 33 pulmonary hamartoma	6 pulmonary chondroma, 33 pulmonary hamartoma	0/33 5/6 (4 affected by Carney triad)	0/0
Gupta S et al., 2019[72]	1009 RCC (400 papillary, 203 chromophobe, 87 chromophobe, 273 oncocytomas, 46 unclassified)	1009 RCC	3 SDHB- (initially classified ad oncocytomas)/1009	0/0
Kalfusova A et al., 2019[73]	261 GIST	44 GIST (without KIT, PDGFRA, and BRAF mutations)	0/0	1 deletion of the whole *SDHB* gene, deletion of exons 2–4 of the *SDHC* gene, and deletion of exons 2–4 of the *SDHD* gene and 1 deletion of *SDHAF2* exons 1-4 and deletion of *SDHB* exons 1-4/4
Kennedy JM et al., 2019[74]	23 biopsies and 37 resections from 30 patients affected by HRCCS	1 RCC in *SDHD* patient	1 SDHB-/1	0/0
Malik F et al., 2019[75]	1 GIST with 2 different morphological population: one well differentiated and one dedifferentiated	1 GIST	1 SDHB-/1	1 *SDHB* mutation/1
Nicolas E et al., 2019[76]	1 RCC + PTC	1 RCC	1 SDHB -/1	0/1
Ricci R et al., 2019[77]	48 GIST (5 in 3 *NF1* patients were excluded)	43 GIST	9 SDHB- /34	7 *SDHA* mutations, 1 *SDHB* mutation, 1 *SDHC* mutation/9
Roh TH et al., 2019[78]	90 CNS tumors	17 glioblastomas 7 astrocytomas 9 oligodendroglyomas 9 ependymomas 7 central neurocytomas	3 SDHB– (hemangioblastomas)/90	0/0

		3 schwannomas 10 meningiomas 6 hemangiopericytomas 3 hemangioblastomas 5 craniopharyngiomas 12 pituitary adenomas		
	35 hemangioblastomas	35 hemangioblastomas	3 SDHB-/35	0/0
Saavedra A et al., 2019[79]	1 macroPA (GH) + metastatic PGL in *SDHB* patient	1 PA	1 heterogeneus staining pattern for SDHB/1	0/1
Shi SS et al., 2019[80]	366 GIST	26 GIST	26 SDHB-/26	10 missense mutations (6 *SDHD*, 3 *SDHB*, 1 *SDHC*)/26
Stanley K et al., 2019[81]	1 *SDHC* patient affected by GIST + renal and neck cysts	1 GIST	1 SDHB-/1	0/0
Urgate-Camara M et al., 2019[82]	2 RCC in a *SDHB* patient	2 RCC	0 (1 doubt to review)/2	0/0
De Filpo G et al., 2020[83]	1 *SDHB* patient	1 mediastinal germ cell tumor	0/1	0/1
Gokozan HN et al., 2020[84]	1 GIST	1 GIST	1 SDHB-/1	0/0
Heilig CE et al., 2020[85]	1 metastatic high-grade spindle cell sarcoma not otherwise specified	1 metastatic high-grade spindle cell sarcoma not otherwise specified in *SDHB* patient	0/0	1 *SDHB* mutation/1
Wilczek Y et al., 2020[86]	1 RCC	1 RCC	1 SDHB-/1	0/0

Supplementary Table S1: association between non-chromaffin tumors and PCC/PGL and/or mutations in one of the PCC/PGL susceptibly genes.

ACC adrenocortical carcinoma, CNS central nervous system, FH fumarate hydratase, GH growth hormone, HLRCC Hereditary Leiomyomatosis and Renal Cell Carcinoma, HNPGL head and neck paraganglioma, NFPA non-functioning pituitary adenoma, PGL sympathetic paraganglioma, PCC pheochromocytoma, PRL prolactin, PTC papillary thyroid carcinoma, RCC renal cell carcinoma, VUS variant of uncertain significance.

*included RCC reported in Gill AJ et al., 2011[9] and Housley SL et al., 2010[7]

§ Housley SL et al., 2010[7]

‡ 21/36 previously unreported cases

13 PA +PCC + known mutations, 21 PA + PCC + suspect clinical picture, 26 PA + PCC

$ partial temporal overlap between these two studies (Tirumani SH et al., 2014[36] and Mason EF et al., 2016[49]): some patients may have been included in both studies.

† 9/12 RCC previously reported in Williamson SR et al., 2015[45]

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