

Table 1. Hereditary pheochromocytoma (Updated Data from the Freiburg International Paraganglioma Registry^{3,29})

	MEN 2	VHL	NF1	PGL 1	PGL 3	PGL 4
Gene	<i>RET</i>	<i>VHL</i>	<i>NF1</i>	<i>SDHD</i>	<i>SDHC</i>	<i>SDHB</i>
Chromosome	10q11.2	3p25-26	17q11.2	11q23	1q21	1p36
PHEO %	30-60	15-20	3-5	34	<1*	37
Inheritance	AD	AD	AD	AD (maternal imprinting)	AD	AD
Age at Diagnosis	~34	~16	~43	~26	~41	~34
Malignancy rate %	3	4	12	<1	0*	24
A/EA %	97 / 3	92 / 17	94 / 6	86 / 57	50 / 50*	42 / 58
HNP %	<1*	<1*	0	84	~100	47
Multifocality %	65	55	12	48	11	12
Other associated Tumours	MTC A: pHPT B: multiple Neurinoma	A: Retinal, CNS Hemangioblastoma, Endolymphatic Sactumour, Epididymal Cystadenoma B: +Renal cysts and RCC; +Pankreas-cysts and –tumour C: PHEO only	Neurofibroma, Café-au-lait spots, Lisch Nodules	Gastrointestinal Stromal Tumours	Gastrointestinal Stromal Tumours	RCC, Gastrointestinal Stromal Tumours

PHEO: pheochromocytoma, HNP: head and neck paraganglioma, AD: autosomal dominant, A: adrenal, EA: extra-adrenal, MTC: medullary thyroid carcinoma, RCC: renal clear cell carcinoma.

* isolated cases