

Supplementary Materials

Table S3. Description of studies included in the systematic review by region, sampling method, method of diagnosis, mean onset age of von Hippel-Lindau disease (vHL), number of participants with vHL, mean cohort age and number of participants with pheochromocytoma and/or paraganglioma (PPGL).

Study	Region	Sampling	Diagnosis, (mean onset age, years)	No. vHL participants, (% males), [mean cohort age, years]	No. of participants with PPGL
Feletti et al, 2016(6)	Europe	Consecutive	Included genetic testing, (-)	128, (-), [-]	39
Ciotti et al, 2009(12)	Europe	Not specified	Included genetic testing, (-)	27, (-), [-]	4
Cybulski et al, 2002(13)	Europe	Consecutive	Included genetic testing, (-)	82, (-), [-]	3
Dollfus et al, 2002(14)	Europe	Consecutive	Included genetic testing, (28.0)	211, (-), [-]	44
Erlic et al, 2010(15)	Europe	Consecutive	Included genetic testing, (-)	469, (-), [-]	177
FJ OB et al, 2014(16)	Europe	Consecutive	Included genetic testing, (-)	36, (41.7%), [-]	9
Frantzen et al, 2012(17)	Europe	Convenience	Included genetic testing, (-)	29, (0.0%), [-]	2
Gergics et al, 2009(18)	Europe	Convenience	Included genetic testing, (-)	14, (-), [-]	4
Graziani et al, 2014(19)	Europe	Convenience	Included genetic testing, (-)	23, (43.5%), [45.0]	3
Losonczy et al, 2013(20)	Europe	Not specified	Included genetic testing, (-)	7, (-), [-]	0
Maher et al, 1990(21)	Europe	Consecutive	Clinical criteria only, (26.3)	152, (52.0%), [-]	11
Stanojevic et al, 2007(22)	Europe	Convenience	Included genetic testing, (-)	13, (61.5%), [-]	6
Wittstrom et al, 2014(23)	Europe	Not specified	Included genetic testing, (-)	15, (53.3%), [39.3]	2
Binderup et al, 2018(24)	Europe	Consecutive	Included genetic testing, (-)	165, (-), [32.3]	24

Glushkova et al, 2017(25)	Europe	Not specified	Included genetic testing, (-)	10, (-), [-]	2
Hes et al, 2007(26)	Europe	Consecutive	Included genetic testing, (-)	110, (-), [-]	17
Ruiz-Llorente et al, 2004(27)	Europe	Not specified	Included genetic testing, (29.0)	35, (51.4%), [-]	10
Ong et al, 2007(28)	Europe	Consecutive	Included genetic testing, (24.7)	573, (-), [-]	115
Prasad et al, 2016(29)	Europe	Consecutive	Included genetic testing, (-)	20, (40.0%), [44.7]	8
Vanbinst et al, 2019(30)	Europe	Not specified	Clinical criteria only, (-)	18, (-), [-]	3
Wiklund et al, 1997(31)	Europe	Convenience	Included genetic testing, (-)	11, (27.3%), [-]	0
Chen et al, 2013(32)	Eastern Asia	Convenience	Included genetic testing, (-)	21, (66.7%), [-]	5
Hattori et al, 2006(33)	Eastern Asia	Not specified	Included genetic testing, (-)	25, (-), [-]	3
Hwang et al, 2014(34)	Eastern Asia	Consecutive	Included genetic testing, (-)	25, (44.0%), [-]	6
Igarashi et al, 2014(35)	Eastern Asia	Consecutive	Not specified, (-)	377, (-), [-]	64
Kwon et al, 2014(36)	Eastern Asia	Consecutive	Included genetic testing, (37.9)	72, (58.3%), [37.9]	18
Siu et al, 2011(37)	Eastern Asia	Not specified	Included genetic testing, (-)	14, (-), [-]	3
Sriphrapadang et al, 2017(38)	Eastern Asia	Not specified	Included genetic testing, (40.3)	16, (-), [-]	2
Huang et al, 2007(39)	Eastern Asia	Not specified	Included genetic testing, (-)	13, (46.2%), [-]	0
Kim et al, 2008(40)	Eastern Asia	Consecutive	Included genetic testing, (39.2)	24, (66.7%), [-]	5
Lee et al, 2016(41)	Eastern Asia	Not specified	Included genetic testing, (-)	26, (65.4%), [-]	4
Liu et al, 2018(42)	Eastern Asia	Convenience	Included genetic testing, (-)	21, (42.9%), [-]	11

Wang et al, 2018(43)	Eastern Asia	Consecutive	Included genetic testing, (31.2)	340, (52.6%), [-]	46
Wang et al, 2018(44)	Eastern Asia	Convenience	Included genetic testing, (-)	7, (85.7%), [-]	1
Zhang et al, 2008(45)	Eastern Asia	Not specified	Included genetic testing, (-)	97, (-), [-]	4
Aufforth et al, 2015(46)	North America	Consecutive	Included genetic testing, (30.5)	273, (46.2%), [-]	84
Bradley et al, 2009(47)	North America	Consecutive	Included genetic testing, (22.4)	19, (-), [-]	2
Chen et al, 1995(48)	North America	Consecutive	Included genetic testing, (-)	522, (-), [-]	92
Salama et al, 2019(49)	North America	Consecutive	Included genetic testing, (24.0)	86, (38.4%), [35.0]	32
Chacon-Camacho et al, 2010(50)	North America	Convenience	Included genetic testing, (-)	20, (35.0%), [29.5]	1
Rasmussen et al, 2010(51)	North America	Not specified	Included genetic testing, (-)	36, (36.1%), [21.8]	3
Fagundes et al, 2019(52)	South America	Not specified	Included genetic testing, (28.3)	31, (-), [-]	17
Gomy et al, 2010(53)	South America	Convenience	Included genetic testing, (-)	12, (-), [-]	1
Ganesh et al, 2010(54)	Southern Asia	Convenience	Clinical criteria only, (-)	7, (42.9%), [33.3]	6
Vikkath et al, 2015(55)	Southern Asia	Not specified	Included genetic testing, (35.6)	31, (54.8%), [-]	3
