

Resources for Pheochromocytoma and Paraganglioma

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Pheo-para alliance (<https://pheopara.org/>)

VHL Alliance (<https://www.vhl.org/>)

Mutation databases:

LOVD (<https://www.lovd.nl/>) (individual gene pages)

ClinVar <https://www.ncbi.nlm.nih.gov/clinvar/>

Clinical Trials

<https://clinicaltrials.gov/>

our repository: NCT <https://clinicaltrials.gov/ct2/show/NCT03160274?term=dahia&draw=2&rank=1>)

PPGL trials:

https://clinicaltrials.gov/ct2/results?cond=pheochromocytoma%2C+paraganglioma&Search=Apply&recrs=b&recrs=a&recrs=f&recrs=d&age_v=&gndr=&type=&rslt=

Guideline papers and other relevant research

Next Generation Sequencing in PPGLs(1)

SDH carrier Guidelines(2)

TMEM127 genotype-phenotype correlations (3)

Metastatic PPGLs(4)

New target therapy (5,6)

1. Group NGSiPS, Toledo RA, Burnichon N, Cascon A, Benn DE, Bayley JP, Welander J, Tops CM, Firth H, Dwight T, Ercolino T, Mannelli M, Opocher G, Clifton-Bligh R, Gimm O, Maher ER, Robledo M, Gimenez-Roqueplo AP, Dahia PL. Consensus Statement on next-generation-sequencing-based diagnostic testing of hereditary pheochromocytomas and paragangliomas. *Nat Rev Endocrinol.* 2017;13(4):233-247.
2. Amar L, Pacak K, Steichen O, Akker SA, Aylwin SJB, Baudin E, Buffet A, Burnichon N, Clifton-Bligh RJ, Dahia PLM, Fassnacht M, Grossman AB, Herman P, Hicks RJ, Januszewicz A, Jimenez C, Kunst HPM, Lewis D, Mannelli M, Naruse M, Robledo M, Taieb D, Taylor DR, Timmers H, Treglia G, Tufton N, Young WF, Lenders JWM, Gimenez-Roqueplo AP, Lusse-Lepoutre C. International consensus on initial screening and follow-up of asymptomatic SDHx mutation carriers. *Nat Rev Endocrinol.* 2021;17(7):435-444.
3. Armaiz-Pena G, Flores SK, Cheng ZM, Zhang X, Esquivel E, Poullard N, Vaidyanathan A, Liu Q, Michalek J, Santillan-Gomez AA, Liss M, Ahmadi S, Katselnik D, Maldonado E, Salgado SA, Jimenez C, Fishbein L, Hamidi O, Else T, Lechan R, Tischler AS, Benn DE, Dwight T, Clifton-Bligh R, Sanso G, Barontini M, Vincent D, Aronin N, Biondi B, Koops M, Bowhay-Carnes E, Gimenez-Roqueplo AP, Alvarez-Eslava A, Bruder JM, Kitano M, Burnichon

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4. Fishbein L, Del Rivero J, Else T, Howe JR, Asa SL, Cohen DL, Dahia PLM, Fraker DL, Goodman KA, Hope TA, Kunz PL, Perez K, Perrier ND, Pryma DA, Ryder M, Sasson AR, Soulen MC, Jimenez C. The North American Neuroendocrine Tumor Society Consensus Guidelines for Surveillance and Management of Metastatic and/or Unresectable Pheochromocytoma and Paraganglioma. *Pancreas.* 2021;50(4):469-493.
 5. Kamihara J, Hamilton KV, Pollard JA, Clinton CM, Madden JA, Lin J, Imamovic A, Wall CB, Wassner AJ, Weil BR, Heeney MM, Vargas SO, Kaelin WG, Jr., Janeway KA, Perini RF, Zojwalla NJ, Voss SD, DuBois SG. Belzutifan, a Potent HIF2alpha Inhibitor, in the Pacak-Zhuang Syndrome. *N Engl J Med.* 2021;385(22):2059-2065.
 6. Jonasch E, Donskov F, Iliopoulos O, Rathmell WK, Narayan VK, Maughan BL, Oudard S, Else T, Maranchie JK, Welsh SJ, Thamake S, Park EK, Perini RF, Linehan WM, Srinivasan R, Investigators MK. Belzutifan for Renal Cell Carcinoma in von Hippel-Lindau Disease. *N Engl J Med.* 2021;385(22):2036-2046.