The pituitary neuroendocrine tumor classification is based upon the size, invasion of adjacent structures, sporadic pituitary neuroendocrine tumor or familial cases, biochemical activity, clinical manifestations, morphological characteristics, response to treatment, and recurrence [1].

Although most pituitary neuroendocrine tumors occur sporadically, these common tumors can present in a familial setting in approximately 5% of cases. Germline mutations in several genes with autosomal dominant (AIP, MEN1, CDKN1B, PRKAR1A, SDHx) or X-linked dominant (GPR101) inheritance are causative of familial pituitary neuroendocrine tumors.

1)
Syro LV, Rotondo F, Ramirez A, Di Ieva A, Sav MA, Restrepo LM, Serna CA, Kovacs K. Progress in the Diagnosis and Classification of pituitary neuroendocrine tumors. Front Endocrinol (Lausanne). 2015 Jun 12;6:97. doi: 10.3389/fendo.2015.00097. eCollection 2015. Review. PubMed PMID: 26124750; PubMed Central PMCID: PMC4464221.