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.