

Aryl hydrocarbon interacting protein

The [AIP gene](#) provides instructions for making a protein called [aryl hydrocarbon interacting protein](#)

[Familial isolated pituitary neuroendocrine tumor](#) (FIPA) is an autosomal dominant disease, characterized by low penetrance, early-onset disease, more invasive tumor growth, as well as somatotroph and lactotroph adenomas in most cases. It has been indicated that the aryl hydrocarbon receptor interacting protein (AIP) gene is a tumor suppressor gene.

The exact molecular mechanism by which its dysfunction promotes tumorigenesis of pituitary is unclear ¹⁾.

Germline aryl hydrocarbon receptor-interacting protein (AIP) mutations are present in 15-30% of familial isolated pituitary neuroendocrine tumor (FIPA) families, and are responsible for 30% of pituitary [gigantism](#) cases (1). However, pathological accelerated growth and/or tall stature can be unrelated to the [growth hormone](#) (GH) axis, and may occur in isolation or as part of a syndrome, such as in Klinefelter, Marfan or Sotos syndromes (2).

Marques et al. report a five-generation kindred with two brothers with pituitary gigantism due to AIP mutation-positive GH-secreting pituitary neuroendocrine tumors and their first-cousin coincidentally also having gigantism due to Marfan syndrome ²⁾.

¹⁾

Cai F, Zhang YD, Dai CX, Liu XH, Yang YK, Yao Y, Wang RZ. [Aryl hydrocarbon receptor interacting protein gene and familial isolated pituitary neuroendocrine tumors]. Zhongguo Yi Xue Ke Xue Yuan Xue Bao. 2012 Dec;34(6):640-4. doi: 10.3881/j.issn.1000-503X.2012.06.021. Review. Chinese. PubMed PMID: 23286415.

²⁾

Marques P, Collier D, Barkan A, Korbonits M. Coexisting pituitary and non-pituitary gigantism in the same family. Clin Endocrinol (Oxf). 2018 Sep 17. doi: 10.1111/cen.13852. [Epub ahead of print] PubMed PMID: 30223298.

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