Gene Section
Mini Review

SDHB (succinate dehydrogenase complex II, subunit B, iron-sulfur protein or IP)
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Identity
Other names: SDH1 (succinate dehydrogenase 1)
HGNC (Hugo): SDHB
Location: 1p36.1-p35

Function
The complex II (succinate-ubiquinone oxidoreductase) is a key component of the mitochondrial respiratory chain and the tricarboxylic acid cycle. It is involved in the oxidation of succinate (succinate + ubiquinone = fumarate + ubiquinol) and carries electrons from FADH to CoQ. It is composed of four nuclear-encoded subunits. The subunit B protein or iron-sulfur protein, which binds three different iron-sulfur clusters, is directly involved in the catalytic activity of succinate dehydrogenase.

Homology
The complex II includes SDHD (cybS) and SDHC (cybL) which are also implicated in paragangliomas and pheochromocytomas.

Mutations

Germinal
Germline mutations cause hereditary paraganglioma, non-familial paraganglioma, familial and sporadic pheochromocytomas. Different germline mutations have been reported: i) a nonsense mutation (R90X) in a family with cervical paraganglioma and ectopic pheochromocytoma, ii) a missense mutation (P197R) in a family with extraadrenal pheochromocytoma and a 1 bp deletion in a sporadic pheochromocytoma, iii) a missense (P131R) mutation and 1 bp insertion (M71fsX80) in familial paraganglioma and a nonsense mutation (Q59X) in sporadic paraganglioma.

Somatic
Loss of wild type allele in tumor DNA is usually observed.
Implicated in

**Hereditary paraganglioma type 4**

**Note**
Alias: familial non chromaffin paragangliomas 4; familial glomus tumor; familial and sporadic pheochromocytoma.

**Disease**
Hereditary paraganglioma type 4 (PGL4) is a rare autosomal dominant disorder non maternally imprinted. Paragangliomas are slow growing highly vascular tumor, usually benigns, derived from crest-neural cells. They are preferentially located in the neck (carotid body and glomus vagal) and head (glomus jugulare and tympanicum). They may be associated with adrenal or extraadrenal pheochromocytomas which produce catecholamines.

**Prognosis**
It depends on extent of the disease at the time of diagnosis.

**References**


This article should be referenced as such: