**Atlas of Genetics and Cytogenetics in Oncology and Haematology**

OPEN ACCESS JOURNAL AT INIST-CNRS

# Cancer Prone Disease Section

## Mini Review

# Familial nervous system tumour syndromes

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Published in Atlas Database: January 2001

Online updated version : http://AtlasGeneticsOncology.org/Kprones/FamilNervousCanID10067.html
DOI: 10.4267/2042/37739



## Identity

**Inheritance**
Eight genetic syndromes are associated with nervous system tumours; these are:
Neufibromatosis 1 (NF1),
Neufibromatosis 2 (NF2),
Tuberous sclerosis,
Turcot syndrome,
Von Hippel-Lindau syndrome,
Li-Fraumeni syndrome,
Gorlin syndrome,
Cowden syndrome,

## Clinics

### *Neoplastic risk*

Neufibromatosis 1 (NF1):
**Nervous System Tumors** Neurofibromas, Astrocytomas, Optic nerve gliomas
**Other tumors** Pheochromocytoma , Osseous lesions, Iris hamartomas
**Genes** NF1 located in 17q11
Neufibromatosis 2 (NF2):
**Nervous System Tumors** Schwanomas, Meningiomas, Spinal ependymomas, Astrocytomas
**Other tumors** Retinal hamartoma
**Genes** NF2 located in 22q12
Tuberous sclerosis:
**Nervous System Tumors** Astrocytomas, Subependymal giant cell tumors
**Other tumors** Cutaneous angio-fibroma, Cardiac rhabdomyomas, Adenomatous polyps of duodenum, Renal hamartomatous tumors, Cysts of the lung and kidney.
**Genes** TSC1 and TSC2 located in 9q34 and 16p13 respectively
Turcot syndrome:
**Nervous System Tumors** Medulloblastomas, Glioblastomas
**Other tumors** Colorectal polyps
**Genes** APC, hMLh1 and hPMS2 located in 5q21, 3p21, and 7p22 respectively
Von Hippel-Lindau syndrome:
Nervous System Tumors Hemangioblastomas
**Other tumors** Retinal hemangioblastomas, Renal cell carcinoma, Pheochromocytoma
**Genes** VHL located in 3p25
Li-Fraumeni syndrome:
Nervous System Tumors Astrocytomas, PNET
**Other tumors** Breast carcinoma, Bone and soft tissues sarcomas, Adenocortical carcinoma, leukaemia
**Genes** TP53 located in 17p13
Gorlin syndrome:
Nervous System Tumors Medulloblastomas
**Other tumors** Multiple basal cell carcinomas, Ovarian fibromas.
**Genes** PTCH located in 9q31
Cowden syndrome:
**Nervous System Tumors** Dysplastic gangliocytoma of the cerebellum
**Other tumors** Hamartomatous polyps of the colon, Thyroid neoplasms, Breast carcinoma
**Genes** PTEN located in 10q23

*This article should be referenced as such:*

Capodano AM. Familial nervous system tumour syndromes. Atlas Genet Cytogenet Oncol Haematol. 2001; 5(2):141.