Neurofibromatosis type 2 (NF2), a multiple neoplasia syndrome, is a manifestation of an impaired expression of the merlin protein, exerting inhibitory effects on cell proliferation signals due to abnormalities of the NF2 gene located on chromosome 22. About half of patients inherit a germline mutation from a parent, and nearly 60% of de novo NF2 patients are estimated to have somatic mosaicism.