Nevoid Basal Cell Carcinoma Syndrome

National Cancer Institute

Source

National Cancer Institute. Nevoid Basal Cell Carcinoma Syndrome. NCI Thesaurus. Code C2892.

An autosomal dominant genetic syndrome caused by abnormalities in the PTCH gene. It is characterized by multiple basal cell carcinomas at a young age, odontogenic keratocysts, and skeletal defects (bifurcated and splayed ribs, fusion of vertebrae, spinal bifida). Patients with this syndrome may also develop medulloblastomas and ovarian fibromas.