

Nevoid basal cell carcinoma syndrome:

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Abstract:

Nevoid basal cell carcinoma syndrome (NBCCS) is characterized by the development of multiple jaw keratocysts, frequently beginning in the second decade of life, and/or basal cell carcinomas (BCCs) usually from the third decade onward. Approximately 60% of individuals have a recognizable appearance with macrocephaly, bossing of the forehead, coarse facial features, and facial milia. Most individuals have skeletal anomalies (e.g., bifid ribs, wedge-shaped vertebrae). Ectopic calcification, particularly in the falx, is present in more than 90% of affected individuals by age 20 years. Cardiac and ovarian fibromas occur in approximately 2% and 20% of individuals respectively. Approximately 5% of children with NBCCS develop medulloblastoma (primitive neuroectodermal tumor [PNET]), generally the desmoplastic subtype. Peak incidence is at age two years. Life expectancy in NBCCS is not significantly different from average