• polyostotic fibrous dysplasia
• café au lait spots
• endocrine dysfunction

Albright syndrome *(also called* **McCune-Albright** *syndrome)* is the most severe form of **polyostotic fibrous dysplasia.** It affects young people *(males and females equally)*. It is characterized by Café au lait spots on the skin, and endocrine abnormalities *(the most common of which is precocious sexual development in females)*. The extent to which each of these problems exists in those with the syndrome is quite variable. The hallmark of Albright syndrome is **premature puberty in the female.** Early sexual development in the male is less common than in the female.

Clinical features:

- Early childhood
- Multiple, slow-growing, painless expansile bone lesions confined to the craniofacial area or throughout the skeleton
- endocrine manifestations; in females often **sexual precocity**
- Irregular shaped Café au lait spots on the torso and sometimes intraorally
- Disfigurement
- Increased level of serum alkaline phosphatase

*** **Pathologic fractures** are frequently associated with this syndrome.

There is no specific treatment for this syndrome. Drugs that inhibit estrogen production, such as testolactone, have been tried with some success.

Important: An additional complication is the **malignant transformation potential** of both the monostotic *(mainly)* and polyostotic fibrous dysplasia into **osteosarcomas.**