

Endochondromatosis

The term **enchondromatosis** refers to multiple **enchondromas** (islands of cartilaginous tissue in the metaphyses of tubular bones) that retard growth and deform the bone.

Ollier's disease (Louis Léopold Ollier (1830-1900), French surgeon) - affecting ½ of the body

Maffucci syndrome (Angelo Maffuccini (1847-1903), Italian pathologist) - enchondromatosis + multiple hemangiomas (i.e. hemangiomatosis)

Clinical picture

- affected long bones shortened and bent, enlarged metaphyses
- fingers enlarged and deformed in the hands, pseudomadelung's deformity on the forearm (see Madelung's deformity), genu valgum

X-ray image

- puffy lucencies, calcified stripes running from the metaphysis to the diaphysis

Therapy

- removal of symptomatic enchondromas, spongioplasty
- corrective osteotomy and bone lengthening
- dispensation (risk of malignancy)

Links

Related articles

- Achondroplasia
- Thanatophoric dwarfism
- Diastrophic dysplasia
- Spondyloepiphyseal dysplasia
- Larsen's syndrome
- Congenital multiple exostoses
- Fibrous bone dysplasia
- Neurofibromatosis
- Dysostosis cleidocranialis
- Apert syndrome
- Osteogenesis imperfecta
- Morbus Albers-Schönberg
- Osteopoikilosis
- Arthrogryposis multiplex congenita
- Marfan syndrome
- Mucopolysaccharidosis
- Hereditary osteoonychodysplasia (nail-patella syndrome)

Used literature

- DUNGL, P., et al. *Ortopedie*. 1. vydání. Praha : Grada Publishing, 2005. ISBN 80-247-0550-8.
- Wikipedia. *Ollier Disease* [online]. ©2006. Poslední revize 2009, [cit. 2009]. <https://en.wikipedia.org/wiki/Ollier_disease>.



Radiograph showing distal radius enchondromas in a 7-year-old girl with Ollier's disease