



# POSNA

The Pediatric Orthopaedic Society of North America

## Ollier's Disease

### Objectives

1. Describe the radiographic features of solitary enchondroma
2. Describe the clinical and radiographic features of Ollier's disease
3. Describe orthopaedic problems secondary to Ollier's disease
4. Discuss risk of chondrosarcoma in relation to Ollier's disease

### Discussion point

1. What is Maffucci's syndrome?

### Discussion

Solitary enchondroma is relatively common. The most common perception of its genesis is that a segment of growth plate does not ossify and persists as an ovoid or linear radiolucent extension of cartilage cells into the metaphysis and sometimes the diaphysis. In children, they can cause pathologic fracture, but are more often incidental findings on radiographs taken for some other reason. Metaphyseal bone affected by large enchondromas does not funnelize normally and may appear expanded due to the absence of remodeling. Forty percent of solitary enchondromas involve the hands or feet, usually the phalanges.

Ollier's disease is an uncommon condition of multiple enchondromatosis. Most patients have bilateral involvement, but more severe on one side. Unilateral involvement also occurs. Genu varum is very common, other angular deformity can result depending on the location of the lesions. Leg length discrepancy is common; but bone affected with

an enchondromatous lesion can be lengthened, sometimes dramatically. The incidence of malignant differentiation of multiple enchondromatosis is high, 30% in one report from the Mayo clinic. The rare Maffucci's syndrome consists of multiple enchondromatosis accompanied by soft tissue hemangiomas, and, in an historical quirk, was described even before Ollier's disease. The potential for malignant degeneration in Maffucci's syndrome is also very high. Cytogenetic study of a low grade chondrosarcoma in a patient with Ollier's disease revealed a deletion at the same site as was found in patients with chondrosarcoma. Undoubtedly, there will be reports of further investigation into the cytogenetics of these neoplasms in the near future.

## References

1. Cannon SR, Sweetnam DR. Multiple chondrosarcomas in dyschondroplasia (Ollier's disease). *Cancer* 1985; 55( 4): 836-40.
2. Chew DK, Menelaus MB, Richardson MD. Ollier's disease: varus angulation at the lower femur and its management. *Journal of Pediatric Orthopedics* 1998; 18( 2): 202-8.
3. Collins PS, Han W, Williams LR, Rich N, Lee JF, Villavicencio JL. Maffucci's syndrome (hemangiomas osteolytica): a report of four cases. *Journal of Vascular Surgery* 1992; 16( 3): 364-71.
4. Gabos PG, Bowen JR. Epiphyseal-metaphyseal enchondromatosis. A new clinical entity. *Journal of Bone & Joint Surgery -American Volume* 1998; 80( 6): 782-92.
5. Liu J, Hudkins PG, Swee RG, Unni KK. Bone sarcomas associated with Ollier's disease. *Cancer* 1987; 59( 7): 1376-85.
6. Ozisik YY, Meloni AM, Spanier SS, Bush CH, Kingsley KL, Sandberg AA. Deletion 1p in a low-grade chondrosarcoma in a patient with Ollier disease. *Cancer Genetics & Cytogenetics* 1998; 105( 2): 128-33.
7. Pandey R, White SH, Kenwright J. Callus distraction in Ollier's disease. *Acta Orthopaedica Scandinavica* 1995; 66( 5): 479-80.
8. Raupp P, Kemperdick H. Neonatal radiological aspect of enchondromatosis (Ollier's disease). *Pediatric Radiology* 1990; 20( 5): 337-8.
9. Shapiro F. Ollier's disease. An assessment of angular deformity, shortening, and pathologic fracture in twenty-one patients. *J Bone Joint Surg (Am)* 1981; 64: 95-103.
10. Springfield DS. Bone and soft tissue tumors. In: Morrissy RT, Weinstein SL, editors. *Pediatric Orthopaedics*. Philadelphia: Lippincott-Raven; 1996. p. 423-67.
11. Sun TC, Swee RG, Shives TC, Unni KK. Chondrosarcoma in Maffucci's syndrome. *Journal of Bone & Joint Surgery -American Volume* 1985; 67( 8): 1214-9.

12. Urist MR. A 37-year follow-up evaluation of multiple-stage femur and tibia lengthening in dyschondroplasia (enchondromatosis) with a net gain of 23.3 centimeters. *Clinical Orthopaedics & Related Research* 1989( 242): 137-57.