A 3-year-old girl presented with leg deformities diagnosed as enchondromatosis (Ollier disease), which is a rare, sporadic, skeletal disorder characterized by hamartomatous growth of cartilage cells within metaphyses of long bones. In 2008, fluid-attenuated inversion recovery signal hyperintensity in her left frontal lobe (Figure, A) and skull-base enchondroma (Figure, B) were discovered. Her enchondromatosis required amputation of digits that were heavy and unusable (Figure, C and D). In 2017, surveillance imaging revealed enlargement of the frontal tumor, leading to craniotomy for a WHO grade 2 astrocytoma. Ollier disease is associated with IDH mutations\(^1\) and patients must be monitored for sarcomatous transformation of enchondromas and extraosseous malignancies including gliomas.\(^2\)

**Study Funding**
The authors report no targeted funding.

**Disclosure**
The authors report no disclosures relevant to the manuscript. Go to Neurology.org/N for full disclosures.
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## References


Teaching NeuroImage: Histopathologically Confirmed Intracranial Enchondroma/Low-Grade Chondrosarcoma and IDH1-Mutated Diffuse Glioma in Ollier Disease

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Neurology 2021;97;e1747-e1748 Published Online before print May 26, 2021
DOI 10.1212/WNL.0000000000012269