MeetingsPremium
MP Journal of Medical Research
Journal of poster communications · ISSN 3144-8402 (Online)
Vol. 1 · 2026-0044
OPEN ACCESS

TYPE
Poster communication

SPECIALTY
Oncology

PRESENTED AT
13th Middle East Pediatric Orthopedic Conference and 5th Khareef Dhofar Military Orthopedic Conference

PUBLISHED
26 October 2026

DOI
10.5281/zenodo.23675457

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KEYWORDS
Chondrosarcoma · Multiple Hereditary Exostosis · malignent transformation

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POSTER COMMUNICATION · ONCOLOGY

Risk of Chondrosarcoma Transformation from Multiple Hereditary Exostosis Under the Age of 18, and Suggestion for Screening Protocol. Literature Review.

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KEY MESSAGES

1. Malignant transformation of MHE into chondrosarcoma is extremely rare in patients under 18 years.
2. Routine screening for malignant transformation in pediatric MHE patients is not currently supported by available data.
3. Surveillance by skeletal survey every 2-3 years is recommended for asymptomatic children with MHE.

ABSTRACT

Introduction. Multiple hereditary exostosis (MHE) is a rare autosomal dominant condition with a prevalence of 1 in 50,000. MHE usually occurs in long bones such as the femur, tibia, and humerus. Malignant transformation into chondrosarcoma is rare and occurs in about 3-5%, usually at the 4th and 7th decade of life.

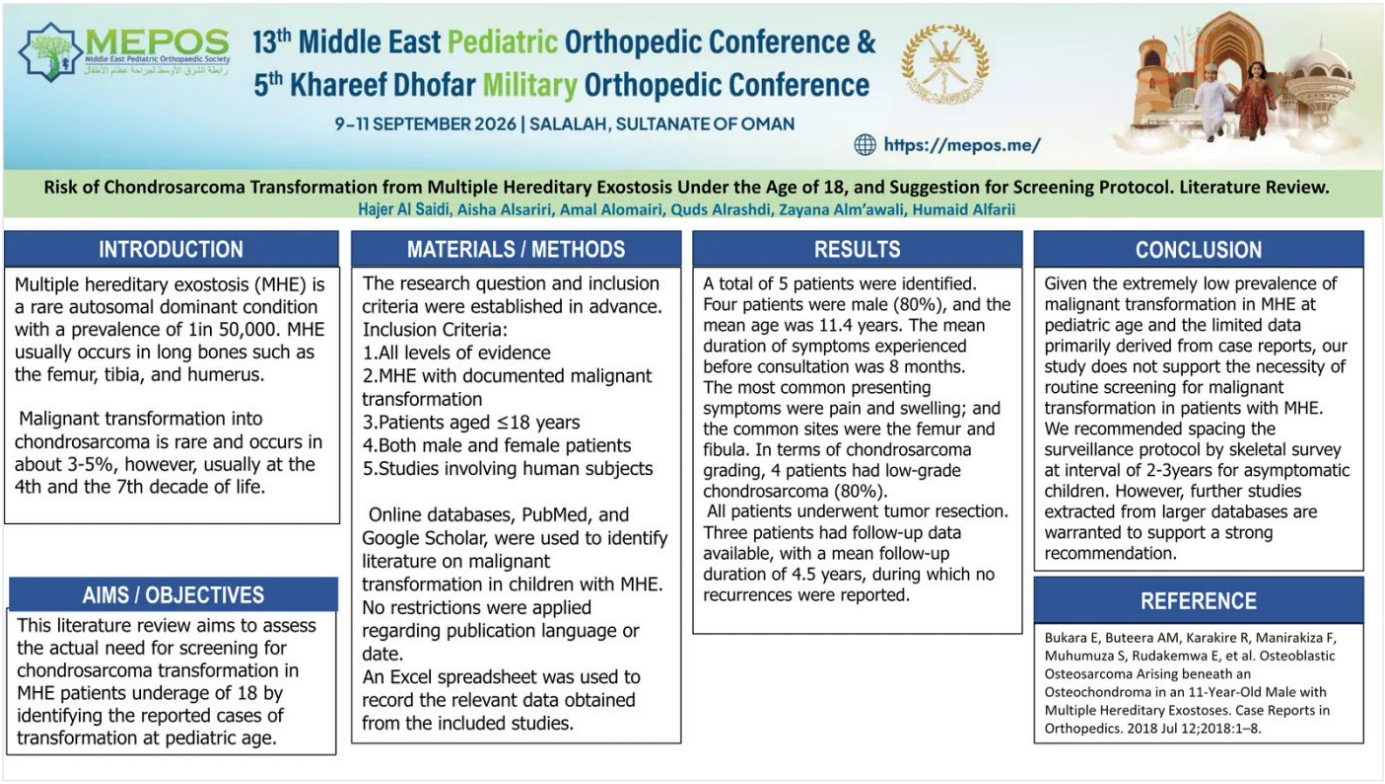
Materials and methods. The research question and inclusion criteria were established in advance. Inclusion criteria were: all levels of evidence, MHE with documented malignant transformation, patients aged ≤18 years, both male and female patients, and studies involving human subjects. Online databases PubMed and Google Scholar were used to identify literature on malignant transformation in children with MHE. No restrictions were applied regarding publication language or date. An Excel spreadsheet was used to record the relevant data obtained from the included studies.

Results. A total of 5 patients were identified. Four patients were male (80%), and the mean age was 11.4 years. The mean duration of symptoms before consultation was 8 months. The most common presenting symptoms were pain and swelling; the common sites were the femur and fibula. Four patients had low-grade chondrosarcoma (80%). All patients underwent tumor resection. Three patients had follow-up data available, with a mean follow-up duration of 4.5 years, during which no recurrences were reported.

Conclusion. Given the extremely low prevalence of malignant transformation in MHE at pediatric age and the limited data primarily derived from case reports, this study does not support the necessity of routine screening for malignant transformation in patients with MHE. It is recommended to space the surveillance protocol by skeletal survey at intervals of 2-3 years for asymptomatic children. Further studies extracted from larger databases are warranted to support a strong recommendation.

Online record, HD poster
journal.meetingspremium.com/p/2026-0044

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Al Saidi H, Al Sariri A, Al Omairi A, Al Rashdi Q, Al M'Awali Z, Al Farii H. Risk of Chondrosarcoma Transformation from Multiple Hereditary Exostosis Under the Age of 18, and Suggestion for Screening Protocol. Literature Review [Poster]. MP J Med Res. 2026;1:2026-0044. doi:10.5281/zenodo.23675457

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PROCESS
Record checked by the editorial team, approved by the corresponding author, then reviewed by the journal's editorial board.

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