

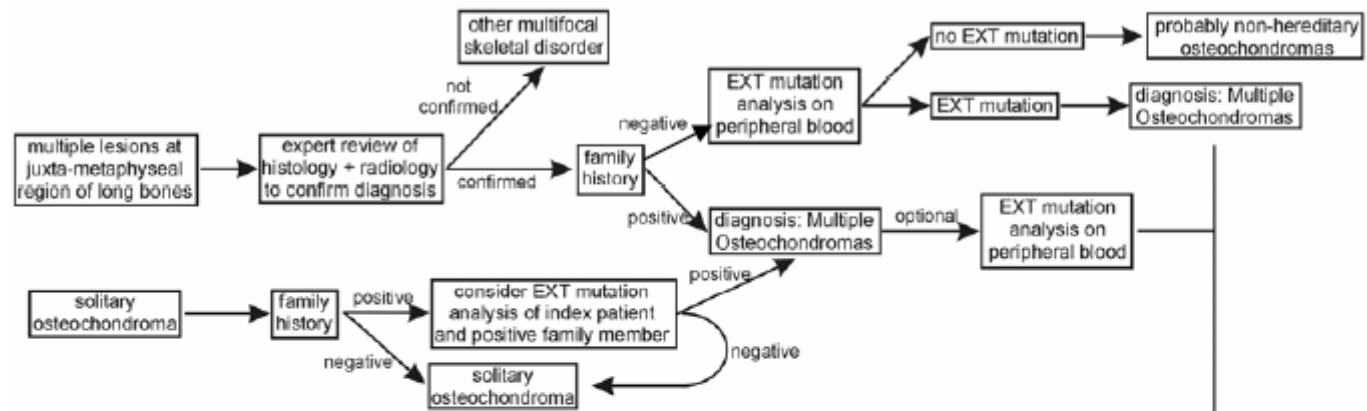
Pancras C.W. Hogendoorn, MD, PhD Principle Researcher MHERF research registry Professor of Pathology, Department of Pathology, Leiden University Medical Center, Molecular tumor pathology and tumor genetics, Netherlands. Head of EuroBoneT consortium, a European Commission granted Network of Excellence for studying the pathology and genetics of bone tumors. In 2002 the World Health Organization (WHO) has redefined the definition of Multiple Hereditary Exostoses (MHE) into Multiple Osteochondromas (MO)(Bovee and Hogendoorn, 2002).

This was done because of the very wide and a-specific use both inside the medical community as well as by patients of the term "exostosis". It ranges in use from osteochondroma(Khurana et al., 2002), the benign cartilage tumor involved in MHE, to a perversion in the direction of growth (eg Nora's lesion or Trevor disease) and even to a reactive process (eg subungual exostosis). Therefore the WHO decided to redefine the terminology for the bony outgrowths into different terms reflecting the different nature of the diseases involved aiming at a better defined diagnosis. "Exostoses" involved in MHE are now specified as osteochondromas and defined as benign cartilaginous tumors.

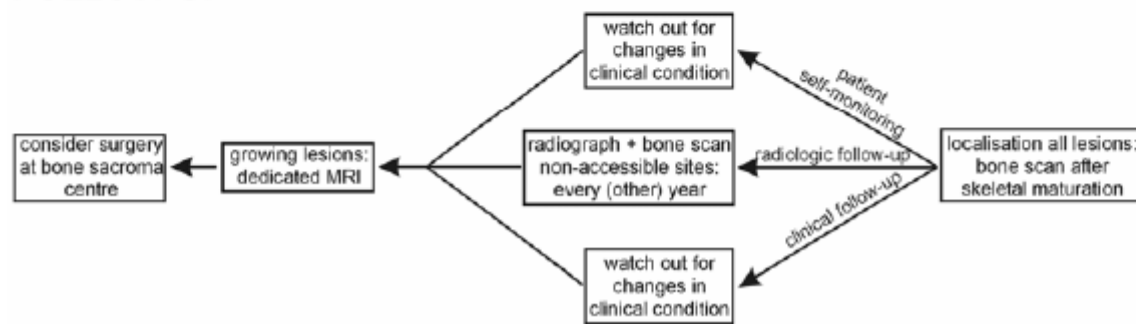
The current WHO definition of an osteochondroma is "a cartilage capped bony projection arising on the external surface of bone containing a marrow cavity that is continuous with that of the underlying bone" (WHO 2002)(Khurana et al., 2002). With this change in terminology, Multiple Hereditary Exostoses is no longer applicable as name for this disorder . That is why the WHO also changed the name of MHE to Multiple Osteochondromas (MO) and stated that: "A diagnosis of multiple exostoses can be made when radiologically at least two osteochondromas of the juxta-epiphyseal region of long bones are observed. MO is diagnosed in case of a positive family history and/or a proven germline mutation in one of the EXT genes" (WHO 2002) (Bovee and Hogendoorn, 2002).

It is very important to correctly diagnosis Multiple Osteochondromas. Especially good monitoring of the patient is important after Multiple Osteochondromas is established. This scheme was taken from "Multiple Osteochondromas: clinicopathological and genetic spectrum and suggestions for clinical management." by L. Hameetman, J.V.M.G. Bovee, A.H.M. Taminiau, H.M. Kroon and P.C.W. Hogendoorn published Hereditary Cancer in Clinical Practice (Volume 2(4) pp. 161-173)(Hameetman et al., 2004).

DIAGNOSIS



FOLLOW-UP



Literature:

References

1. Bovee, J.V.M.G. and P.C.W.Hogendoorn. 2002. Multiple osteochondromas. In World Health Organization classification of tumours. Pathology and genetics of tumours of soft tissue and bone. C.D.M.Fletcher, K.K.Unni, and F.Mertens, editors. IARC Press, Lyon. 360-362.
2. Hameetman, L., J.V.M.G.Bovee, A.H.M.Taminiau, H.M.Kroon, and P.C.W.Hogendoorn. 2004. Multiple Osteochondromas: Clinicopathological and Genetic Spectrum and Suggestions for Clinical Management. Hereditary Cancer in Clinical Practice 2:161-173.
3. Khurana, J., F.Abdul-Karim, and J.V.M.G.Bovee. 2002. Osteochondroma. In World Health Organization classification of tumours. Pathology and genetics of tumours of soft tissue and bone. C.D.M.Fletcher, K.K.Unni, and F.Mertens, editors. IARC Press, Lyon. 234-236.