

Dysplasia epiphysealis hemimelica (DEH), also known as Trevor's disease, is a developmental bone disease of childhood. It is rare and clinical experience with this condition is limited. Most cases are diagnosed before 8 years of age. It is characterized by an abnormal growth of cartilage arising from the cartilage of the terminal ends (epiphysis) of the long bones, particularly of the lower limbs. The bones of the knee and ankle joints are most commonly affected, as well as part of the foot (tarsal bones). The upper limbs and spine are rarely involved. The abnormal cartilage produces an irregular nodular mass located either in the medial or lateral part of the bone (hemimelic), usually medial. DEH may affect a single bone (localized form), multiple bones in a single limb (classical form) or an entire limb (generalized) usually involving a leg from the pelvis to the foot. Approximately two-thirds of affected children have multiple lesions. DEH was first described in the medical literature in 1926. Trevor recognized this condition in 1950. The name, dysplasia epiphysealis hemimelica first appeared in the medical literature in 1956. The cause of DEH is unknown. There is no evidence that hereditary factors play a role in the development of this disease. More research is necessary to determine the exact underlying cause(s) of this disorder. DEH is benign and there are no reports of malignant transformation of the cartilage abnormality. DEH usually affects children between the ages of 1 and 15. Males are affected more often than females. The incidence of DEH has been estimated at 1 in 1,000,000 individuals in the general population. However, some authors consider that the incidence is probably higher because some patients may be misdiagnosed with other conditions.