

2. ABSTRACT

Agensis or isolated hypodontia of maxillary permanent canines is a rare dental anomaly. We report p.Gly213Ser heterozygous missense mutation in *WNT10A* in nine Thai patients, three of them had congenital absence of maxillary permanent canines. One of those with agensis of maxillary permanent canines had peg-shaped maxillary lateral incisors and dens invaginatus of the left maxillary lateral incisor. Six patients had various types of isolated hypodontia which appeared to be quite severe. One female patient was affected with Tricho-Odonto-Onycho-Dermal dysplasia had a compound heterozygous mutation (p.Gly213Ser and p.Glu347x).

KEY WORDS: Ectodermal dysplasia, missing teeth, Odonto-Onycho-Dermal dysplasia, Tricho-Odonto-Onycho-Dermal dysplasia, nail dysplasia, hypotrichosis, sparse hair, hypodontia