PL2. FISH - ODOUR SYNDROME : SOUTHEAST ASIA EXPERIENCE

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ABSTRACT

A study of the metabolism of trimethylamine was carried out in 103 healthy Thai volunteers (70 men and 33 women) and it was found that under normal dietary conditions 84-100% of trimethylamine was excreted in the urine in its N-oxide form. Five propositi living in different parts of the country were identified as having deficiency in the N-oxidation of this tertiaryamine, because they excreted only 8-35% of this chemical as trimethylamine N-oxide. This metabolic defect was also confirmed by the results of an oral trimethylamine (600 mg) challenge experiment in which all five propositi were found to excrete an even smaller percentage of trimethylamine as trimethylamine N-oxide in their urine. The results of a study of the families of the two proband individuals, as well as those members of their preceding generations under normal dietary conditions, are consistent with the view that the disorder or metabolic defect is inherited in a Mendelian fashion as an autosomal recessive trait, similar to that reported for white Caucasian subjects.