Clinical study

PAPILLON-LEFEVRE SYNDROME (PLS)

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ABSTRACT

Background. The syndrome was first described by Papillon and Lefevre in 1924, it is characterized by palmoplantar hyperkeratosis and severe periodontal breakdown, resulting in early loss of teeth.

Objective. The authors were engaged in obtaining data on Papillon-Lefevre cases in the population of Slovenia.

Methods. Clinical records were studied and patients' families were investigated at their homes. Among the 2 million Slovenian population 13 persons were detected in 7 families. Consanguinity could not be proven. 11 affected persons were monitored by the investigators.

Results. The patients who cooperated in the study, showed in most cases psoriasis-like skin lesion and palmoplantar hyperkeratosis. A severe periodontitis appeared soon after the teeth eruption. Some of them were treated with antibiotics and retinoids, nonetheless they lost all the teeth up to the end of puberty.

Conclusion. The genetic defect remains unknown, The therapy is symptomatic: antibiotics and good dental cleaning are important. Retinoids influence primarily the skin lesions. Cooperation between dermatologist and stomatologist is needed and an early treatment is indicated.

KEY WORDS

Papillon-Lefevre syndrome, hereditary hyperkeratosis with periodontosis, patients, Slovenia

INTRODUCTION

The combination of a transgredient palmoplantar hyperkeratosis which is usually mild with a severe periodontal breakdown, resulting in early loss of both, the deciduous and permanent teeth was first described by Papillon and Lefevre in 1924 (1). Until now about 200 cases of the Papillon-Lefevre syndrome (PLS) have been described. During the 1994-95 period, 17 cases were reported (2,3,4,5,6,7,8). However, exact numbers cannot be given while some cases are mentioned two times or more, and certain cases are not reported. Though most case reports deal with Caucasians there are also PLS reports from Mongoloid and Negroid races. It is interesting to note that in Slovenia with its 2 million inhabitants 11 PLS patients were diagnosed (9). The syndrome may be supposed to be more frequent than believed, since dentists may overlook the mild palmar and plantar hyperkeratosis and dermatologists may not

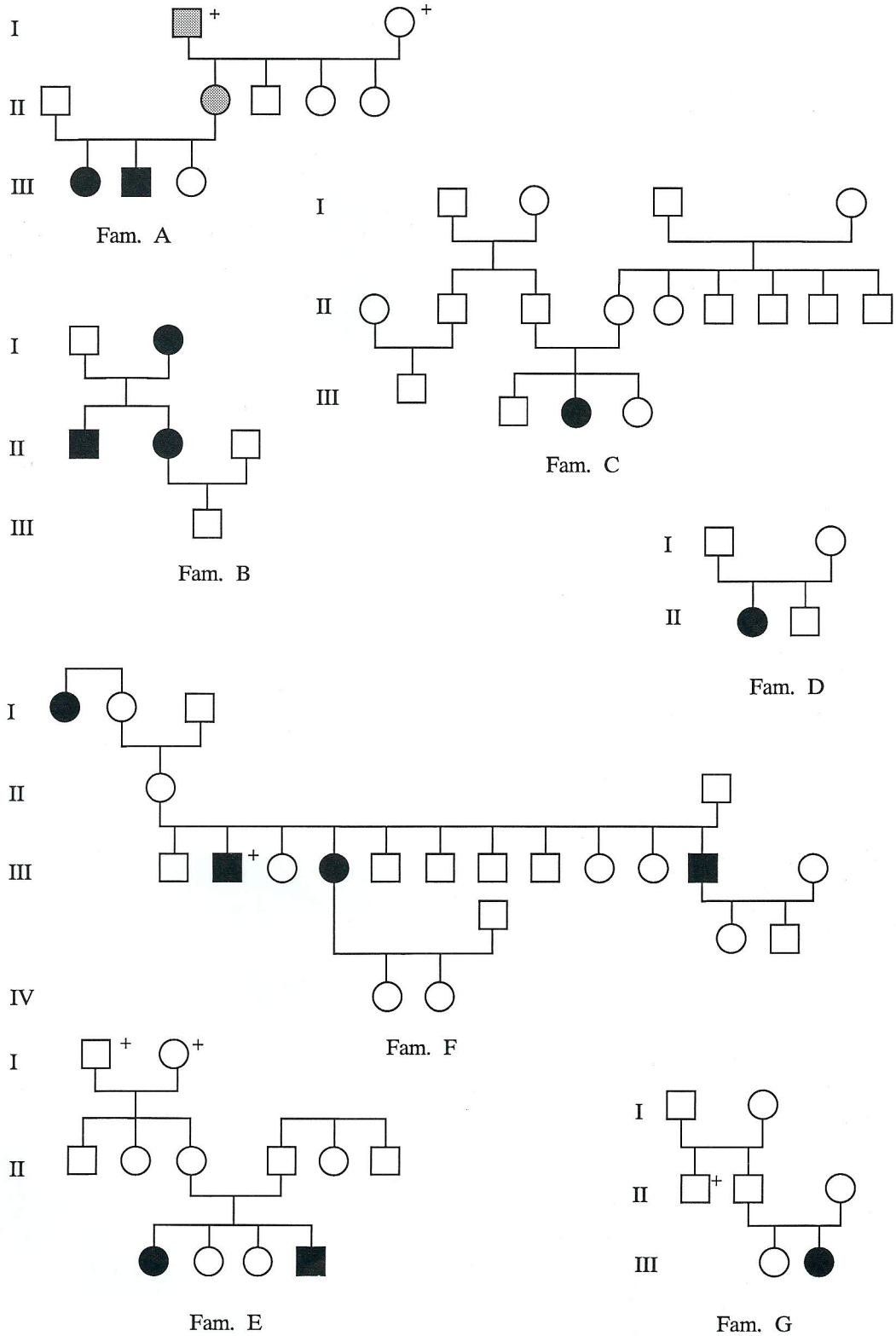

Fig. 1. Pedigrees of 7 families in Slovenia in which 13 patients affected with Papillon-Lefevre syndrome are presented