FAMILIAL PULMONARY FIBROSIS

[1]Krzysztof Wytrychowski, [2]Anna Hans-Wytrychowska, [3]Beata Nowakowska

[1]Department and Clinic of Internal Diseases, Geriatry and Allergology and [2]Department of Family Medicine, Wroclaw Medical University, Wroclaw; [3]Laboratory of Tissue Immunology, Institute of Immunology and Experimental Therapy, Polish Academy of Sciences, Wroclaw, Poland

Introduction: Iidiopathic pulmonary fibrosis (IPF) is a progressive interstitial lung disease of unknown etiology. IPF is a chronic fibrotic interstitial pneumonia (histopathologically known as usual interstitial pneumonia (UIP). To date, about 100 cases of families with at least 2 members diagnosed with IPF have been described. Familial pulmonary fibrosis is hereditary most probably as a feature which is autosomal, dominant with variable penetration. Its incidence is equally often in both sexes. A positive correlation has been observed between IPF and alleles of gene of a1-antitrypsin inhibitor and HLA genes such as B15, B8, B12, DR2 and Dw6. **Material and methods:** Two families with IPF have been treated since 2002 at the Clinic of Internal Diseases, Geriatry and Allergology, Wroclaw Medical University. The patients with IPF in family A included brother (1A), sister (2A), and sister's daughter (3A). The other members of the family – three children of patient 3A (4A, 5A, 6A), sister of patient 3A (7A), and her two children (8A, 9A) – are healthy. The patients with IPF in family B included father (patient 1B) and his two children: daughter (4B - sick), son (5B - sick). The other son (6B) is healthy. Additionally, we examined patients: 2B (healthy sister of patient 1B), 3B (wife of patient 1B.) Patient 4B is wife of patient 7B (healthy) with whom she has two children: patient 8B (daughter – healthy), patient 9B (son – healthy). We examined HLA genes in both families, including antigens class I (locus A, B, and C) and class II (locus DR). Tables 1 and 2 show the results. **Results:**

Table 1.

Male patient 1A (IPF)	**Female patient 2A (IPF)**	**Female patient 3A (IPF)**
A2;B18;Cw7;DR11 A1;B7;Cw1;DR14	A2;B18;Cw7;DR11 A2;B56;Cw1;DR4	A2;B56;Cw1;DR4 A3;B35;Cw4;DR1
Male patient 4A	**Female patient 5A**	**Male patient 6A**
A3;B12;Cw4;DR11 A28;B35;Cw1;DR1	A3;B12;Cw4;DR11 A28;B35;Cw1;DR1	A3;B12;Cw4;DR11 A28;B35;Cw1;DR1
Female patient 7	**Male patient 8A**	**Male patient 9A**
A2,3;B56;Cw1,4;DR1,4,w53 A28;B35;Cw1;DR1	A3;B12;Cw4;DR11 A28;B35;Cw1;DR1	A3;B12;Cw4;DR11 A28;B35;Cw1;DR1

Table 2

Male patient 1B (IPF)	**Female patient 2B**
A*02;B*40;Cw*03;DRD1*13;DQB1*06	A*02;B*40;Cw*03;DRD1*13;DQB1*06 A2;B56;Cw1;DR4
Female patient 3B	**Female patient 4B (IPF)**
A*25;B*18;Cw*12;DRD1*04, DQB1*03 A*68;B*38;Cw*12;DRD1*11;DQB1*03	A*25;B*18;Cw*12;DRD1*04;DQB1*03 A*02;B*40;Cw*03;DRD1*13, DQB1*06
Male patient 5B (IPF)	**Male patient 6B**
A*25;B*18;Cw*12,4;DRD1*04;DQB1*03 A*02;B*40;Cw*03;DR1*13;DQB1*06	A*68;B*38;Cw*12;DR1*11;DQB1*03 A*02;B*40;Cw*03;DR1*13;DQB1*06

Male patient 7B	**Female patient 8B**
A*02;B*07;Cw*07;DR1*14;DQB1*05 A*30;B*13;Cw*06;DRD1*07;DQB1*02	A*02;B*07;Cw*07;DR1*14;DQB1*05 A*25;B*18;Cw*12,4;DRD1*04;DQB1*03
Male patient 9B	
A*02;B*40;Cw*03;DR1*13;DQB1*06 A*30;B*13;Cw*06;DRD1*07;DQB1*02	

Conclusions: It is impossible to determine the relation between HLA antigens and the incidence of the disease on the basis of the results.