** objetivos:** el estudio objetivo fue el de hacer que el sujeto impuesto que impone la caracterización de un nuevo concepto. Las participantes fueron seleccionadas para que se registraran en el Registro de PID. El diagnóstico de varios casos de PID fue llevado a cabo de acuerdo con los criterios establecidos por la IUIS [1].

**resultados:** durante el período de 1993 a 2020, 49 casos de PID fueron registrados en el adulto población de Chuvashia. Aunque la frecuencia de PID, común variable inmunodeficiencia (CID) es la más común en Chuvashia (26 personas). En el segundo lugar se seleccionó la IgG yi (10 personas); en el caso de los niños se seleccionó la IgM y la enfermedad autoinmune (4 personas). Los resultados de los casos de PID se presentan en el cuadro y en el texto 4. Se presentan los resultados de varios casos de PID, los cuales fueron registrados de acuerdo con los criterios establecidos por la IUIS [1].

**referencias:**

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**Disclosure de intereses:** No declarado.

**DOI:** 10.1136/annrheumdis-2021-eular.3526

**POS1375**

**ENfekte der M694V Homozygotie auf die CAROTIS INTRA-Media-THICKNESS UND FLOW MEDIATED DILATATION IN PATIENTS WITH FMF RELATED AMYLOIDOSIS**

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**Background:** There are limited data in FMF associated AA amyloidosis patients regarding the vascular abnormalities including atherosclerosis and endothelial dysfunction, all of which are contributors of cardiovascular disease (CVD) risk. And, this risk assessment for future CV events have not yet been evaluated between FMF amyloidosis patients with different genotypes.

**Objectives:** We aimed to compare early markers of endothelial dysfunction and atherosclerosis, mortality and other disease characteristics in FMF-related amyloidosis patients with a homzygous M694V genotype and in patients with other genotypes with this cross-sectional comparative study.

**Methods:** For this purpose, patients with FMF-related amyloidosis were assigned according to the MEFV gene mutation to one of the two groups. Group 1: Patients homozygous for M694V (which is the most common genotype associated with the most severe clinical phenotype in FMF) Group 2: Patients homzygous (other than M694V) or compound heterozygous (including M694V) for other MEFV variants. Potential confounders were controlled by excluding the patients with untreated hypertension, diabetes mellitus, obesity, smoking, previous history of CVD low glomerular filtration rate (eGFR <70mL/min). Flow-mediated dilatation (FMD), pentraxin-3 (PTX3) as early markers of endothelial dysfunction and carotid intima-media thickness (cIMT), Fibroblast Growth Factor 23 (FGF23) as an indicator of atherosclerotic vascular disease, all of which are non-invasive tests that are also used to identify subjects at increased risk for future CV events were measured in these 2 groups.

**Results:** We analyzed demographic, clinical, genetic, survival data and these non-invasive markers of endothelial dysfunction (FMD and PTX3) and atherosclerosis (cIMT, fibroblast Growth Factor 23 (FGF23)) in 76 FMF amyloidosis patients with homzygous M694V mutations (Group 1) and 93 FMF amyloidosis patients with two pathogenic mutations apart from M694V homozygosity (Group 2) (Table 1). Brachial artery FMD was significantly lower in Group 1 when compared with Group 2 subjects (0.6 ± 0.5%, p<0.001). cIMT, FGF23 and PTX3 levels were higher in Group 1 when compared with Group 2 (cIMT: 0.84 ± 0.67 mm, p<0.001; FGF23: 52.4 vs. 38.0 pg/dL, p<0.001; PTX3: 14.0 vs. 3.2 ng/mL, p<0.001). Time to develop amyloidosis was similar in patients homozygous for M694V (median 10.0 years, 95% CI: 8.99-11.0) and in Group 2 (median 10.0 years, 95% CI: 9.87-11.0) (p=0.05). Mortality rate was significantly higher in Group 1 compared to Group 2 (18.4% vs. 11.1%, p<0.001) over a median of 7.8 years of follow-up. FMF amyloidosis patients homozygous for M694V had reduced survival (mean survival: 57 months, 95% CI: 49.9 to 101.0 months) as compared to Group 2 (mean survival: 100.7 months, 95% CI: 100.0 to 101.3 months) (p<0.001) (Figure 1). The proportion of the clinical manifestations including fever, serositis, cryoglobulinaemia-like...