subsequently with CNK. At the moment of CNK introduction and at their latest visit while on CNK, patients were asked to fill a questionnaire investigating their experience with either ANK or CNK. The questionnaire included 4 different domains that were assessed through means of Visual Analogue Scales (VAS), ranging from 0 to 10. The following domains were considered: injection-related pain, satisfaction with the frequency of administration, concern for drug-related adverse events including infections, and overall drug tolerability. VAS values for each of the domains were compared between ANK and CNK with Mann-Whitney U test.

Results: Ten AOSD patients treated with both ANK and CNK were included in the analysis. Disease characteristics are shown in Table 1 below. ANK was discontinued due to ineffectiveness in 6 patients and due to adverse reactions in 4. As shown in Figure 1, CNK overall tolerability significantly exceeded ANK one (p=0.021). Only 1 AOSD patient reported a greater tolerability with ANK compared to CNK because she felt more comfortable with pre-filled syringes. The reasons for higher CNK tolerability were the lower pain at injection site (median 1, IQR 1-1 vs median 4.5, IQR 1.25-7; p=0.015) and the greater level of satisfaction with the frequency of administration (median 10, IQR 9.25-10 vs median 3.5, IQR 2.25-4.75, p<0.001). The concern for drug-related adverse events was not significantly different between ANK and CNK (p=0.549) and overall low among AOSD patients. CNK led to a satisfying control of systemic inflammation and steroid-sparing effect in most of patients and it was discontinued in only one case (Patient 5), due to ineffective disease control.

Conclusion: In our cohort of AOSD patients, CNK proved not only to be effective in controlling systemic inflammation in ANK refractory cases, but its use was associated with greater patients' satisfaction and tolerability.

REFERENCES:

Disclosure of Interests: Nicola Farina: None declared, Corrado Campochiaro Speakers bureau: Novartis, Pfizer, Roche, GSK, SOBI, Alessandro Tomelleri: None declared, Giacomo De Luca Speakers bureau: SOBI, Novartis, Celgene, MSD, Pfizer, Giulio Cavalli Speakers bureau: SOBI, Elena Baldissera Speakers bureau: Novartis, Pfizer, Roche, Alpha Sigma, Sanofi, Lorenzo Dagna Speakers bureau: Abbvie, Amgen, Biogen, BMS, Celltrion, Novartis, Pfizer, Roche, SG, SOBI, Celgene Janssen, MSD, MP

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MORTALITY AND ITS RELATED FACTORS IN PATIENTS WITH IGG4-RELATED DISEASE: A JAPANESE SINGLE-CENTER STUDY

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Background: In recent years, IgG4-related disease (IgG4-RD) has become a widely recognized disorder. However, mortality and its related factors in this disease are not well known.

Objectives: This study aimed to clarify mortality and its related factors in patients with IgG4-RD.

Methods: We retrospectively reviewed the medical records of patients with IgG4-RD diagnosed by experts based on fulfillment of the Japanese comprehensive diagnostic criteria and/or the 2019 ACR/EULAR classification criteria for IgG4-RD at a single center in Japan. Using the collected data, we calculated the crude mortality rate and the standardized mortality ratio (SMR) using national Japan mortality statistics and investigated the cause of death. We performed Cox regression analyses to assess mortality-related factors.

Results: A total of 179 patients with IgG4-RD were included: 124 were male (69.3%); the median age was 68 years (interquartile range [IQR] 60-75 years); and the median follow-up from diagnosis was 47 months (IQR 17-84). Ten patients (5.6%) in our cohort died during the follow-up period. Five died of malignancy, one of respiratory failure, two of infectious pneumonia, one of sudden cardiac event, and one of suspected aortic aneurysmal rupture. The crude mortality rate was 11.1 per 1,000 person-years. According to national Japan mortality statistics, 11.6 age- and sex-matched deaths were expected to occur within the follow-up period, resulting in a SMR of 0.86 (95% confidence interval [CI] 0.41-1.59). Univariate Cox regression analyses indicated that the number of affected organs at diagnosis (hazard ratio [HR] 1.45, 95% CI 1.02-2.05), serum creatinine levels at diagnosis (HR 1.82, 95% CI 1.06-3.12), and the presence of malignancy, one of respiratory failure, two of infectious pneumonia, one of sudden cardiac event, and one of suspected aortic aneurysmal rupture. The crude mortality rate was 11.1 per 1,000 person-years. According to national Japan mortality statistics, 11.6 age- and sex-matched deaths were expected to occur within the follow-up period, resulting in a SMR of 0.86 (95% confidence interval [CI] 0.41-1.59). Univariate Cox regression analyses indicated that the number of affected organs at diagnosis (hazard ratio [HR] 1.45, 95% CI 1.02-2.05), serum creatinine levels at diagnosis (HR 1.82, 95% CI 1.06-3.12), and the presence of malignancy during the clinical course (HR 3.93, 95% CI 1.10-14.02) had a significant impact on the time to death, whereas the other factors including age at diagnosis and serum C-reactive protein and IgG4 levels at diagnosis did not.

Conclusion: Our findings suggest that the mortality rate of patients with IgG4-RD does not significantly differ from that of the Japanese general population.

Table 1. Clinical and treatment characteristics of study population.

<table>
<thead>
<tr>
<th>Sex and age (years)</th>
<th>Disease duration (months)</th>
<th>Disease manifestations</th>
<th>Sequential biologic therapies</th>
<th>Steroid dosage varia- tion (mg/day)</th>
<th>ESR variation (mm/ hour)</th>
<th>CRP variation (mg/L)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Patient 1</td>
<td>Male, 70</td>
<td>22</td>
<td>Arthritis, Fever</td>
<td>TCZ, ANK, CNK</td>
<td>25 → 5</td>
<td>120 → 13</td>
</tr>
<tr>
<td>Patient 2</td>
<td>Female, 72</td>
<td>11</td>
<td>Arthritis, Fever</td>
<td>TCZ, ANK, CNK</td>
<td>5 → 0</td>
<td>60 → 24</td>
</tr>
<tr>
<td>Patient 3</td>
<td>Male, 32</td>
<td>48</td>
<td>Arthritis, Fever, Rash</td>
<td>ANK, CNK</td>
<td>15 → 0</td>
<td>29 → 11</td>
</tr>
<tr>
<td>Patient 4</td>
<td>Male, 71</td>
<td>12</td>
<td>Arthritis, Fever</td>
<td>ANK, CNK</td>
<td>20 → 5</td>
<td>120 → 2</td>
</tr>
<tr>
<td>Patient 5</td>
<td>Female, 58</td>
<td>72</td>
<td>Arthritis, Fever</td>
<td>ANK, CNK</td>
<td>0 → 5</td>
<td>27 → 36</td>
</tr>
<tr>
<td>Patient 6</td>
<td>Female, 48</td>
<td>5</td>
<td>Arthritis, Fever</td>
<td>ANK, CNK</td>
<td>10 → 0</td>
<td>8 → 10</td>
</tr>
<tr>
<td>Patient 7</td>
<td>Female, 74</td>
<td>180</td>
<td>Arthritis, Fever, Rash, MAS</td>
<td>ETN, ANK, CNK</td>
<td>5 → 5</td>
<td>33 → 1</td>
</tr>
<tr>
<td>Patient 8</td>
<td>Female, 78</td>
<td>180</td>
<td>Arthritis, Fever</td>
<td>ANK, CNK</td>
<td>40 → 20</td>
<td>46 → 10</td>
</tr>
<tr>
<td>Patient 9</td>
<td>Male, 22</td>
<td>144</td>
<td>Arthritis, Fever</td>
<td>ETN, TCZ, ANK, CNK</td>
<td>20 → 10</td>
<td>86 → 94</td>
</tr>
<tr>
<td>Patient 10</td>
<td>Male, 43</td>
<td>24</td>
<td>Arthritis, Fever</td>
<td>ANK, CNK</td>
<td>15 → 5</td>
<td>15 → 10</td>
</tr>
</tbody>
</table>

1 Before and after canakinumab start.

ANK, anakinra; CNK, canakinumab; CRP, C-reactive protein; ESR, erythrocyte sedimentation rate; ETN, etanercept; MAS, macrophage activation syndrome; MTX, methotrexate; PDN, prednisone; TCZ, tocilizumab.
population. Multi-organ involvement and renal dysfunction at diagnosis as well as malignancy during the clinical course may be associated with higher mortality. An appropriate clinical evaluation for the early detection of these risk factors is required at first diagnosis and during long-term follow-up.

Disclosure of Interests: None declared

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THE JOINT INVOLVEMENT IN ADULT ONSET STILL’S DISEASE IS CHARACTERISED BY A PECULIAR MAGNETIC RESONANCE IMAGING AND A SPECIFIC TRANSCRIPTOMIC PROFILE

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Background: Adult onset Still’s disease (AOSD) is a rare systemic autoinflammatory disease and joint involvement is one of its clinical manifestations [1]. AOSD involves TNF pathway. IL-1β, a proinflammatory cytokine, and its receptor IL-1R1, is a more expressed in AOSD patients than controls, when we explored the iron metabolism, the group using regular medication had significantly fewer and shorter duration, the group using regular medication had significantly fewer and shorter

Disclosure of Interests: None declared

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PREGNANCIES

FAMILIAL MEDITERRANEAN FEVER DURING PREGNANCY: A 26 CASE SERIES WITH 38 PREGNANCIES

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Background: Familial Mediterranean Fever (FMF) is an autoinflammatory disease characterized by recurrent attacks with autosomal recessive inheritance. FMF usually occurs in young age; most patients (90%) begin to suffer from their first attack before 20 years of age. Pregnancy can occur frequently during the course of the disease, as women of reproductive age are affected by the disease.

Objectives: In this study, it was aimed to retrospectively examine the demographic, genetic, and clinical characteristics (frequency of attacks during pregnancy, duration of attacks, treatment of attacks) of 26 cases who were followed up with a diagnosis of FMF in the last five years and experienced pregnancy.

Methods: A total of twenty-six female FMF cases experienced pregnancy and diagnosed or followed up in our rheumatology center between 2015-2020 were included in the study. All patients were diagnosed according to Tel-Heshomer criteria. All data and follow-up visit records of the patients were retrospectively recorded by the rheumatologist. Patients were followed up by an obstetrician working in the same center during pregnancy. The demographic and genetic characteristics of the patients, the treatment used, the duration and characteristics of the attack during pregnancy, and the treatment they received during the attack were recorded retrospectively. Data processing and analysis conducted with SPSS 22 for Windows.

Results: During the follow-up period, a total of 38 pregnancies were observed in 26 female cases. When the genetic mutation tests of all patients were examined, 61% were M694, 15% were V726, 11% were M680I positive and compound mutation was detected in 42% of the patients. The mean age of the patients was 30±2.8, the disease duration was 9.8±5.4 years, the follow-up period was 38±14 months, the attack frequency during pregnancy was 3.6±1.7 and the attack duration was 14±9.8 hours. Considering the clinical features, fever was seen in 92.3%, abdominal pain 96.1%, chest pain 88.4%, arthritis 11.5% and other symptoms seen in 26% during attacks of pregnant FMF patients. All patients used 1 gram of colchicine regularly throughout pregnancy. Steroids were used in 11.5% of patients and non-steroid anti-inflammatory drugs in 53.8% of patients during the attack. Anakinra was used in 11.5% of the cases except for the first trimester following a written consent obtained from the patients. In 10.5% of 38 pregnancies, spontaneous abortion was observed in the early period, 78% of pregnancies resulted in preterm delivery before 32 weeks. In addition, 81.5% of pregnancy completed the planned period and resulted in a healthy birth. Cesarean section was performed in 4 patients and normal delivery procedure in 27 patients. Major malformation-anomaly was not observed in any baby. When patients using colchicine (73%) irregularly and less than 1 gram (26.9%) before pregnancy were compared in terms of attack frequency and duration, the group using regular medication had significantly fewer and shorter attacks (p<0.05).

Three colchicine resistant patients with M694 homozygous mutation became pregnant under anakinra treatment. A total of five pregnancies were followed up in three cases. No medication was used in these patients in the first trimester. As of the second trimester, 100mg/day for 3 days of anakinra was administered in these patients after obtaining an informed consent. In this patient group, no obstetric problem was observed during and after pregnancy, and healthy deliveries were realized.

Conclusion: Pregnancy is common in FMF patients of reproductive age. Disease and relapse treatment during pregnancy is still a problem due to the limited number of medications that can be used for treatment. Further studies required to verify safety of Anakinra in refractory FMF cases. There is a need to develop options for the prevention and treatment of attacks during pregnancy.

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