Abstract FRI0481 – Figure 1. Interactions between GCA, IBD and covariates

Conclusions: The probability that GCA patients may also suffer from IBD is increased in comparison with age- and gender-matched controls. Our findings indicate that this association is most prominent in younger patients (<70). Screening for IBD amongst GCA patients in this age group may be warranted.

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FRI0482

CAUSES OF DEATH IN CONNECTIVE TISSUE DISEASE (CTD'S) AND VASCULITIDES; DATA FROM THE NORWEGIAN CONNECTIVE TISSUE DISEASES AND VASCULITIS REGISTRY (NOSVAR)

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Background: Mortality as an outcome of disease's severity and causes of death can give extended insight into the nature of each specific diagnose and highlight distinct needs for monitoring. In connective tissue diseases (CTD) and systemic vasculitides, mortality and causes of death have been studied within most single diseases, but with heterogeneity of the studies and widely varying results. Studies applying similar methods across the diagnoses are lacking. Consequently, it is difficult to compare causes of death between the different diagnoses.¹

Objectives: To identify the causes of death within different CTDs and systemic vasculitides.

Methods: We performed a prospective, observational, controlled study between 1999 and 2017 of adult patients (at least 18 years of age) with diagnosis of CTD or vasculitides. All patients were included in the Norwegian connective tissue disease and vasculitis Registry (NOSVAR). In total, 2140 patients were diagnosed and followed up until death or study end by April 31st 2017. To avoid bias by selection, we included only incident cases, excluding patients with diagnoses set prior to 1999. Moreover, cases with a disease course not consistent with the initial diagnoses were excluded. Causes of death were identified by linking NOSVAR to the Norwegian Causes of Death Registry and by reviewing hospital charts. We divided the causes of death into the main groups of cardiovascular diseases (CVD), neoplasms, chronic respiratory disease (CRD), infections and other (gastrointestinal, renal insufficiency and trauma). To compare causes of death to the general population we used data from WHO Mortality Database, Causes of death.²

Results: During a mean (SD) follow-up time of 9.2 years (4.7), 279 patients (13%) deceased. The major causes of death were, in descending order of frequency; CVD (27%), neoplasms (25%), CRD (16%), infections (11%), gastrointestinal manifestations (4%), renal insufficiency (2%). Data from the general population, adjusted for age and gender, showed that deaths by CVD, CRD and infections were more prevalent among the patients.² The leading causes of death are shown in figure 1. In Takayasu arteritis and ISSc, CVD was the most frequent cause of death; (56%) and (41%), respectively. More than half of the patients (53%) with antisynthetase syndrome died of CRD. Those with dermatomyositis died most frequently of neoplasms (50%).

Conclusions: Compared to general population, patients with CTD and vasculitis died more often of CVD, CRD and infections. CVD as a cause of death was most prevalent in patients with Takayasu arteritis, giant cell arteritis and systemic sclerosis, while neoplasm was the major cause of death in dermatomyositis. In antisynthetase syndrome, both CRD was the major causes of death. The study gives the clinician valuable information on how to monitoring the different CTDs and vasculitides regarding serious outcome.

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FRI0483

SULFASALAZINE AS A POTENTIAL TREATMENT FOR IGA-VASCULITIS (HENOCH-SCHÖNLEIN PURPURA) IN ADULTS

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Background: Primary IgA-vasculitis and inflammatory bowel diseases (IBD) share many clinical, endoscopic, and radiological signs. It may suggest a common pathogenetic background for both pathological conditions. While sulfasalazine (SASP) is one of the well-known potential agents able to improve symptoms in IBD, the usefulness of SASP in treatment of the primary IgA-vasculitis remains unclear.

Objectives: Retrospective study to assess a therapeutic value of the SASP in primary IgA-vasculitis.

Methods: Totally 78 adult patients with primary IgA-vasculitis were enrolled in this study. Diagnosis was made on the basis of EULAR/PRES criteria¹ after thorough screening to exclude a secondary nature of the disease, including colonoscopy. Purpura/petechia was present in all patients being mandatory diagnostic criterion. Arthritis/arthralgia was seen in 48 patients (61.5%). There were no patients with abdominal syndrome just before enrollment, although 46 (59%) patients had transient abdominal pains in the history. Mild to moderate signs of renal involvement (hematuria and/or proteinuria) was seen in 35 (45%) patients without renal impairment. There were 20 (25%) drug-naïve patients, 40 (51%) patients after unsuccessful immunosuppressive treatment and 18 (23%) patients failed to respond to anticoagulants or antiplatelet agents. Initially SASP was prescribed in a daily dose 1 g followed with gradual titrating up to the 2 g/day depending on tolerability and clinical response. Most patients (96%) have been taking SASP longer than 6 month and about a half of the patients (56%) – longer than year. The longest treatment was 5 years.

Results: Complete clinical remission of the skin rash was achieved in 48 patients (58.9%). In 27 patients (35%), there was partial improvement of the skin eruptions, characterised with less quantity of the skin purpura or longer periods free of