Respondents were asked about use of a digital tool to support lifestyle behaviour change. Over 3/4 were open to using digital tools. Regarding the respondents' preferences: the top three preferences were for reminders, contact with peers and information on the importance of changing the behaviour in question.

Conclusions: The survey suggests that SRA members have digital access despite 1/3 being older than 65. The focus for current and future behaviour change is physical activity and to a lesser extent, healthy eating. Smoking and risky alcohol use behaviours are low in this group. Willingness to engage in a digital tool is high, and preferences are clear, with interest in human contact with professionals and peers through the digital tool, as well as automated functions.

Disclosure of Interest: None declared

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Advances in biologic therapy of small vessel vasculitis

OP0332 PAEDIATRIC OPEN-LABEL CLINICAL STUDY OF RITUXIMAB FOR THE TREATMENT OF GRANULOMATOSIS WITH POLYANGIITIS (GPA) AND MICROSCOPIC POLYANGIITIS (MPA)

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Background: Rituximab in combination with glucocorticoids (GC) is approved to treat adult patients (pts) with GPA or MPA; however, limited data exist on the safety and efficacy of rituximab in paediatric pts with these potentially life- and organ-threatening diseases.

Objectives: To report the interim safety, pharmacokinetics (PK) and exploratory efficacy data from the 6 month remission induction phase of a Phase IIa international, open-label, 18 month clinical study of rituximab in paediatric pts with GPA or MPA.

Methods: Pts aged ≥ 2 to ≤ 18 years with newly diagnosed or relapsing GPA/MPA received 4 intravenous (IV) rituximab infusions of 375 mg/m² body surface area (BSA) on Days 1, 8, 15 and 22 with concomitant GC 1 mg/kg/day (max 60 mg/day) tapered to 0.2 mg/kg/day minimum (max 10 mg/day) by Month 6. All pts received 3 doses of pulse IV methylprednisolone (30 mg/kg/day, max 1 g/day) prior to first rituximab infusion and mandatory prophylaxis for *Pneumocystis jirovecii* infection. Pts were also pre-medicated with acetaminophen and an antihistamine, 1 hour before each rituximab infusion. Adverse events (AEs) and laboratory data were measured at each study visit (1, 2, 4 and 6 months). Plasma samples for PK analysis were collected throughout the study; clearance and area under the curve (AUC) were calculated using population PK modelling from the RAVE study of rituximab in adult pts with GPA/MPA.¹ For exploratory efficacy assessment, the Paediatric Vasculitis Activity Score (PVAS) was measured at each study visit.

Results: Of the 25 pts enrolled, 19 (76%) had GPA and 6 (25%) had MPA (median [range] age 14⁶⁻¹⁷ years; 80% female). Median (range) disease duration was 0.5 (0.2-0.72) months; 2 pts had received prior cyclophosphamide therapy. All received 4/4 rituximab infusions and completed the 6 month induction phase. By Month 6, all pts had experienced ≥ 1 AE. The most common AEs by system organ class were infections and infestations in 16 pts (64%). AE terms reported in ≥ 3 pts are listed in the table 1. Eleven serious AEs occurred in 7 pts (28%), including 3 serious infections (viral gastroenteritis, one lower and one upper respiratory tract infection). 32% of pts had ≥ 1 infusion related reaction (IRR). No serious IRRs or deaths were reported. The relationship between AUC and BSA was flat and comparable to adult pts. A total of 13 pts (52%) achieved remission by 6 months, defined as PVAS of 0 and GC dose 0.2 mg/kg/day (max 10 mg/day) or PVAS of 0 on 2 consecutive readings ≥ 4 weeks apart irrespective of GC dose.

Abstract OP0332 – Table 1 Adverse events reported in ≥ 3 patients

Adverse event	Number of events (% of pts with ≥ 1 AE)
Headache	6 (24%)
Upper respiratory tract infection	5 (20%)
Abdominal pain (upper)	3 (12%)
Back pain	3 (12%)
Blood IgG decreased	3 (12%)
Chest pain	3 (12%)
Cough	3 (12%)
Epistaxis	3 (12%)
Hypertension	3 (12%)
Nausea	3 (12%)
Pyrexia	3 (12%)
Vomiting	3 (12%)

AE, adverse event; IgG, immunoglobulin G.

Conclusions: In the initial 6 months of this first global clinical trial of rituximab in paediatric pts with GPA/MPA, rituximab was generally safe and well tolerated. The overall safety profile and PK parameters were comparable to adults with GPA/MPA. No new safety signals were observed. However, the study size and interim nature of the analysis limit firm conclusions. The clinical trial and additional efficacy, PK and safety analyses are ongoing.

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OP0333 SURVIVAL IN ANCA ASSOCIATED VASCULITIDES: A RETROSPECTIVE MULTICENTRIC ANALYSIS IN NORTHERN ITALY

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Background: Patients affected by ANCA associated vasculitides (AAV) show lower survival than general population, even if the mortality decreased significantly in the last decade.

Objectives: Aim of our study is to analyse the early mortality (within 6 months) and the long-term survival in a multicentric Italian cohort of AAV patients.

Methods: we identified all patients affected by AAV, diagnosed from 1995 until 2017, followed routinely in four vasculitis referral centres in Northern Italy. We enrolled patients with diagnosis of granulomatosis with polyangiitis (GPA) and micro polyangiitis (MPA), fulfilling EMA algorithm or ^{CHCC} definitions, with complete survival data at last follow up. The analysis focused on early mortality, long-term survival and their predictors.

Results: We enrolled 200 AAV patients (F/M 110/90, Caucasian 98%) with a median age at diagnosis of 54.6 $\pm$ 15.2 years. 157 (78.5%) were affected by GPA and 43 (21.5%) by MPA. Data about ANCA antibodies were available in 181 patients and 157 (87%) resulted ANCA positive: 100 c-ANCA/PR3, 56 p-ANCA/MPO and one with double specificity PR3-MPO-ANCA.

During the follow up period [53²⁶⁻¹⁰⁰ months], we registered 21 (10.5%) deaths, 6 (28.5% of all mortality) within 6 months after diagnosis: 9 patients died due to infectious complications, 1 due to hepatic cancer, 1 due to end stage heart failure, 1 due to massive cholestasis and 9 due to unknown causes.

Early mortality was significantly associated with a higher frequency of alveolar haemorrhage (p=0.01; OR 11.1 IC95% 2.1-60.1) and respiratory failure (p<0.001, OR 28.3 95% CI: 4.7 to 170.6).

The long-term survival, analysed with Kaplan-Maier method, did not show significant differences between GPA and MPA patients, while a significant poorer