Microangiopathy evolution score was 5.19 (2.04), VAS for DU was 75.52 (16.17), VAS for Raynaud was 67.43 (14.16), HAQ was 1.62 (0.55). 5 patients received Bosentan less than 6 months, so they were excluded from the statistical analysis. 6 month month evaluation revealed significant decrease in the number of DU (p<0.01), the VAS for DU (p<0.01), the VAS for Raynaud (p=0.03) and the HAQ (p=0.04), but not of the microangiopathy evolution score. No significant difference was noticed of the above mentioned parameters at the next follow-up evaluations. Regarding Bosentan safety: 6 patients died during the follow up (3 cases of severe pulmonary arterial hypertension, 1 scleroderma renal crisis, 1 heart failure, 1 post vascular surgery). Bosentan was stopped due to lack of efficacy in 2 case and due to side effects in 3 cases: 2 elevated liver enzymes, 1 severe trombocytopenia and 1 dyspepsia agravation.

12 patients had a follow up after a 6 months Bosentan stop. We did not notice any significant increase in the number of DU, the VAS for DU or Raynaud, the capillaroscopic semi-quantitative scoring of the skin or the HAQ.

Conclusions: We noted a significant decrease in the number of DU, patients perception of Raynaud and of DU after 6 months of treatment and the effect was maintained in the 3 years follow-up, even 6 months after Bosentan was stopped. In this long-term follow-up no new unidentified adverse reactions were found, except for the unexpected severe trombocytopenia. The present study is limited due to the small sample size, to the observational nature and should be viewed as descriptive. Questions rise about drug costs (6 months or long term treatment), but it also has to be emphasised that most of these lesions were chronic and non-responsive to previous treatments.

REFERENCES:

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AB0780
PROLONGED PROTON PUMP INHIBITOR EXPOSURE IS ASSOCIATED WITH DEVELOPMENT OF CALCINOSIS IN SYSTEMIC SCLEROSIS
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Background: Long-term use of proton pump inhibitors (PPI) has been associated with some safety concerns, including potential vascular calcification. In a previous retrospective analysis, we noted a possible association between PPI use and calcinosis in scleroderma (SSc).

Objectives: To investigate the association between PPI use and the presence and extent of calcinosis in SSc patients.

Methods: Data from prospectively recruited patients were collected by patient survey, physician assessment and medical records. Calcinosis was graded; size (3), III=2, body site, II=1 cm, +++=3 cm) and number of sites involved (NSL-I = number of body sites, II-2, III-3, IV-4). A total daily PPI equivalent dose (TDED) was calculated for each patient. We calculated PPI exposure score (PPE) by multiplying the total duration of use by TDED. For analysis, PPE was categorised into four groups: 0=no exposure, 1-up to 5 years, 2=6–10 years, 3=10 years. Fisher’s exact test was used to assess categorical variables. Logistic regression assessed association between calcinosis and independent variables.

Results: 216 patients were recruited, 81.5% females, mean age 57.46 (SD 13.5) years. 56.5% had limited, 31.5% diffuse SSc, 9.7% had overlap features and 2.3% other CTD. Mean disease duration was 10 years (SD 9), ANA subtypes were defined: ACA positive (31.5%), ATA (25.5%), ARA (12.0%), ANA (+ENA- (11.6%), USRNP (5.1%), ANA- (4.2%), PmScl (3.7%) and 6.5% other antibodies. Gastroesophageal reflux symptoms occurred in 83.3% of patients, most were on PPI (81%) and 14.8% had previously been on PPI. Current calcinosis (CC) was present in 30.1% patients, 9.7% reported past calcinosis. 39.8% had calcinosis at any time (CAT). 60.2% of patients never had calcinosis. Of those with CC, 47.7% had >1 site involved. The most frequent sites affected were: finger (70.8%), elbow (35.4%) and knee (18.5%).

Univariable analysis found an association between disease duration and calcinosis, with odds of CAT increased by 7% per year (OR 1.07, CI 1.04–1.11, p=0.001). Similarly, every year of PPI exposure increased odds of CAT by 3% (OR 1.03, CI 1.01–1.05, p=0.05). Increasing age associated with CAT odds increasing by 2% per annum, p=0.004. Exposure to a standard dose of PPI for over 10 years increased the odds of calcinosis by 4 times (OR 4.07, CI 1.68–9.85, p=0.002) compared to no exposure. PPI category associated with NSL (p=0.04). 73.3% of patients with large volume calcinosis (>3 cm) had a PPE for >10 years and all with calcinosis >3 cm had exposure to PPI.

Multivariable logistic regression found that disease duration (OR 1.07, CI 1.03–1.11, p=0.001) and antibody specificity strongly associated with calcinosis. Presence of ATA (OR 0.32, CI 0.14–0.75 p=0.008), ANA- (OR 0.13, CI 0.02–0.79 p=0.026), and ANA +ENA- (OR 0.17, CI 0.05–0.52, p=0.002) reduced odds of calcinosis. Although the effect of PPIs on calcinosis was attenuated after adjusting for disease duration and antibodies, higher exposure to PPIs remained a significant predictor of calcinosis, with PPE category (>10) increasing risk of CAT (OR 3.34, CI 1.16–9.17, p=0.025).

Conclusions: Our data support a novel association of PPI exposure with calcino- sis and confirm association of disease duration and antibody profile. Given the clinical impact of calcinosis, a potentially modifiable risk factor of PPI exposure warrants further study.

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AB0781
GLUCOCORTICOID DOSE AND CARDIAC INVOLVEMENT MIGHT BE POTENTIAL RISK FACTORS FOR SCLERODERMA RENAL CRISIS
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Background: Scleroderma renal crisis (SRC) is a rare but life-threatening complica-
tion of systemic sclerosis (SSc). SRC remains a major risk factor for mortality in SSc. It is important to identify potential risk factors for SRC, and avoid developing overt SRC.

Objectives: To perform a retrospective case series analysis of the characteristics, management and outcomes of SRC in Chinese SSc patients.

Methods: SSc patients hospitalised at Sun Yat-Sen Memorial Hospital from Jan-
uary 1992 to December 2017 were recruited. Clinical data were collected. SRC was defined as new onset, with blood pressure (BP) >140/90 mmHg or >30 mmHg rise in BP from baseline, rising serum creatinine (Scr) levels and/or oligo-
uria. Data were showed as mean ±standard deviation.

Results: There were 749 SSc patients recruited and 16 patients (2.1%) of them were hospitalised for SRC. Among these 16 patients, 56% were females, age was 54.6±13.6 years, mean duration from SSc onset to SRC occurred was 4 years. SRC developed in 14 patients (87.5%) with diffuse cutaneous SSc (dcSSc), and in 2 patients (12.5%) with limited cutaneous SSc (IcSSc). Eleven patients (68.8%) were under glucocorticoid treatment before SRC onset: 4 patients received >30 mg/d of prednisone, 6 patients received >7.5 mg/d prednisone and 1 patient received <7.5 mg/d prednisone. No patient was treated with angiotensin-converting enzyme (ACE) inhibitors before SRC.

All 16 patients manifested progressive renal failure, with Scr levels increase to 220±256 μmol/L. Ten patients manifested new onset hypertension, with systolic BP 175±21 mmHg and diastolic BP 108±13 mmHg. Five patients who had a his-
tory of well-controlled hypertension manifested accelerated increase in BP 178±117/108±7 mmHg. One patient was normotensive, but manifested rapidly pro-
gressive oliguric renal failure with Scr increase to 969 μmol/L, massive proteinuria and hemolytic anemia.

Twelve patients (75%) had pulmonary fibrosis, 11 patients (88.8%) had cardiac involvement, 6 patients had pulmonary arterial hypertension (P AH) and 6 patients had gastrointestinal dysfunction. Cardiac involvement was common, manifested pericarditis, myocardial damage and heart failure (n=7, 43.8%, respectively). All 5 dead patients were accompanied by cardiac involvement.

Eleven patients had Raynaud’s phenomenon, 8 patients had digital ulcers, 5 patients had arthritis and 2 patients had oliguria. Thirteen patients (81%) mani-
fested anemia, 8 patients (50%) manifested thrombocytopenia, and 8 patients (50%) manifested microangiopathic haemolytic anemia (MAHA).

Eleven patients (68.8%) received ACE inhibitor treatment. Fifteen patients were treated with glucocorticoid and 12 patients with immunosuppressant (Cyclophos-
phamide n=10, Azathioprine n=2). After treatment, renal recovery in 4 patients (25%), kidney function improved and developed to chronic kidney disease (CKD) without dialysis in 5 patients (31%), 2 patients required permanent dialysis (13%). Five patients (31%) died.