department of Peking University First Hospital from January 2007 to May 2016.The primary causes of death were identified, the standardised mortality ratio (SMR) and years of life lost (YLL) were calculated based on the National Bureau of Statistics of China for the general population, the survival in the first decade was performed using Kaplan-Meier analysis, and the predictors of mortality were evaluated by multivariable cox regression.

Results: A total of 226 DM and 54 PM cases were included and the mean age of onset was 49.9±14.8 years for DM and 48.1±17.1 years for PM. The median follow-up duration was 40.6 (11.6–77.6) months. Among 267 patients who were successfully traced, 66 patients died. Infection (50.0%) was the leading cause of death followed by malignancy (19.7%), and interstitial lung disease (ILD) (9.1%). The overall age and sex adjusted SMR was 9.0 (95% CI 6.8–11.2) for DM, and 5.0 (95% CI 2.4–7.5) for PM. The overall age and sex adjusted SMR of DM/PM patients with ILD was 8.4 (95% CI 5.8–11.0), and the SMR of the patients with malignancy was 14.9 (95% CI 8.5–21.2). The YLL of women and men were 37.5 and 28.4 years respectively for DM, and 24.3 and 12.0 years respectively for PM (Table1). The 10-year survival of patients with ILD or malignancy was significantly worse than those without ILD or malignancy respectively (Figure 1 and 2). The independent predictors of mortality for DM were age of disease onset, respiratory muscle involvement and malignancy; and the independent predictor of mortality for PM was age at disease onset (Table2).

Table 1. The standardized mortality ratio (SMR), life expectancy (LE) and years of life lost (YLL)

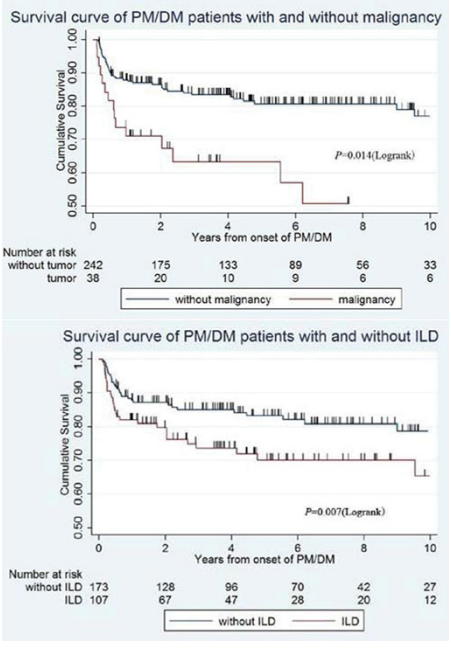
	Overall (n=280)	Females (n=201)	Males (n=79)
Dermatomyositis	226	163	63
Death number	55	37	18
SMR (95% CI)	9.0 (6.8–11.2)	12.0 (6.8–11.2)	6.0 (3.2–8.7)
LE of general population (years)	–	80.8	75.8
LE of DM patients (years)	–	43.3	47.4
YLL (years)	–	37.5 8	28.4
Polymyositis	54	38	16
Death number	11	8	3
SMR (95% CI)	5.0 (2.4–7.5)	4.2 (1.3–7.2)	9.3 (-1.2–19.8)
LE of general population (years)	–	80.8	75.8
LE of PM patients (years)	–	56.5	63.8
YLL (years)	–	24.3	12.0

Abbreviations: DM: dermatomyositis; SMR: standardised mortality ratio; CI: confidence interval; LE: life expectancy; YLL: years of life lost; PM: polymyositis.

Table 2. Multivariable cox regression analyses of risk factors in the DM/PM patients

Variables	HR	95% CI	*P* value
Dermatomyositis			
Age*	1.04	1.01–1.06	<0.001
ILD	1.35	0.74–2.48	0.319
Respiratory muscle involvement	2.58	1.19–5.58	0.016
Malignancy	3.12	1.49–6.58	0.003
Polymyositis			
Age*	1.08	1.00–1.16	0.044
ILD	2.47	0.18–34.00	0.500
ESR	1.02	0.99–1.04	0.174

*Age: Age at disease onset. Abbreviations: HR: Hazard Ratio; CI: confidence interval; ILD: interstitial lung disease; ESR: erythrocyte sedimentation rate.



Conclusions: Mortality of DM/PM patients in China is substantial, especially in females, and those with ILD or malignancy. Infection was the leading cause of death. Patients with older age at onset, respiratory muscle involvement, ILD, and malignancy need to be paid more attention.

Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2017-eular.2873

OP0126 A PHASE 2 STUDY OF SAFETY AND EFFICACY OF ANABASUM (JBT-101) IN SYSTEMIC SCLEROSIS

R. Spiera[1], L. Hummers[2], L. Chung[3], T. Frech[4], R. Domsic[5], D. Furst[6], J. Gordon[1], M. Mayes[7], R. Simms[8], S. Constantine[9], B. White[9]. *[1]Hospital for Special Surgery, New York City; [2]Johns Hopkins, Baltimore; [3]Stanford, Palo Alto; [4]University of Utah, Salt Lake City; [5]University of Pittsburgh, Pittsburgh; [6]Arthritis Association of Southern California, Los Angeles; [7]University of Texas, Houston; [8]Boston University, Boston; [9]Corbus Pharmaceuticals, Inc., Norwood, United States*

Background: Anabasum (JBT-101) is a synthetic, oral, non-immunosuppressive, preferential CB2 agonist. It inhibits onset and activates resolution of innate immune responses in animal models of systemic sclerosis (SSc).

Objectives: Evaluate safety and efficacy of anabasum in SSc

Methods: A double-blind, randomized, placebo (PBO)-controlled Phase 2 trial dosed 42 diffuse cutaneous SSc subjects with disease duration ≤6 years on stable medication including immunosuppressive drugs. Subjects received anabasum 5 mg QD, 20 mg QD, or 20 mg BID on Days 1–28, then 20 mg BID on Days 29–84, or PBO on Days 1–84. Subjects were followed off study drug on Days 85–113. The primary safety outcome was treatment-emergent adverse events (TEAEs). The primary efficacy outcome was improvement in ACR Combined Response Index in diffuse cutaneous Systemic Sclerosis (CRISS) score, combined anabasum group vs PBO, Days 29–113 (end of Weeks 4–16). The five domains of the ACR CRISS are the modified Rodnan skin score, HAQ-DI, patient and physician global assessments, and FVC % predicted.

Results: Of 42 dosed subjects, 27 (64%) received anabasum and 15 (36%) received PBO. Three anabasum subjects withdrew: 1 (3.7%) for a TEAE of moderate dizziness; 1 withdrew consent; and 1 by physician decision. One PBO subject withdrew consent. Baseline demographic and CRISS domain scores were similar except slightly more anabasum subjects used background immunosuppressive drugs (93% versus 80%, anabasum vs PBO). Seventeen (63%) anabasum subjects had 66 TEAEs, and 9 (60%) PBO subjects had 35 TEAEs. There were no serious, severe, or unexpected TEAEs related to anabasum. Severity and relationship of TEAEs to study drug were similar in both groups. The most frequent TEAEs by MedDRA system (% anabasum vs % PBO) were: nervous system (37% vs 27%); general disorders (30% vs 7%); gastrointestinal (22% vs 20%); infections (22% vs 20%); musculoskeletal (22% vs 13%); and investigations (0% vs 20%). The most frequent TEAEs in anabasum subjects were dizziness (22%) and fatigue (19%) which were usually mild. Anabasum subjects had greater improvement in ACR CRISS scores than PBO subjects (mixed model repeated measures analysis, p=0.044, 1-sided). The median ACR CRISS scores at the end of Weeks 4, 8, 12, and 16 (anabasum vs PBO) were 3.0% vs 1.0%, 19.0% vs 1.0%, 27.5% vs 1.0%, and 33.0% vs 1.0%, respectively. Among anabasum subjects, ∼50% had ACR CRISS ≥20% after 8 weeks of dosing. The individual domains of the ACR CRISS score showed greater improvement, improvement that reached minimal important differences in several domains, and less worsening in anabasum vs PBO groups. Anabasum subjects had greater improvement in SSc skin symptoms and itch. Plasma metabolipidomic profiles showed anabasum, not PBO, shifted lipid mediator production to increase pro-resolving vs pro-inflammatory lipid mediators.

Conclusions: Anabasum provided significant and medically meaningful efficacy in SSc as assessed by the ACR CRISS score and its individual domains and had acceptable safety and tolerability in this Phase 2 trial. These data support continued clinical development of anabasum for the treatment of SSc.

Disclosure of Interest: R. Spiera: None declared, L. Hummers: None declared, L. Chung: None declared, T. Frech: None declared, R. Domsic: None declared, D. Furst: None declared, J. Gordon: None declared, M. Mayes: None declared, R. Simms: None declared, S. Constantine Employee of: Corbus Pharmaceuticals, Inc., B. White Employee of: Corbus Pharmaceuticals, Inc.

DOI: 10.1136/annrheumdis-2017-eular.2712

OP0127 TEN YEARS FOLLOW-UP OF GASTROINTESTINAL INVOLVEMENT BY THE SMALL INTESTINAL CLEARANCE IN PATIENTS WITH SYSTEMIC SCLEROSIS

N. Hashimoto[1], M. Kitano[2], T. Nakazawa[3], T. Iwasaki[4], T. Hashimoto[1]. *[1]Hashimoto Rheumatology Clinic, Osaka; [2]Division of Rheumatology, Department of Internal Medicine, Hyogo College of Medicine, Nishinomiya; [3]Osaka Saiseikai Nakatsu Hospital, Osaka; [4]Department of Pharmacy, Hyogo University of Health Sciences, Kobe, Japan*

Background: Systemic sclerosis (SSc) is an inflammatory autoimmune disease characterized by fibrosis and small vascular involvement in the skin, lungs, heart and gastrointestinal (GI) tract. The esophagus is the most frequently involved GI tract disorder. Although the small intestinal involvement such as malabsorption and pseudo-obstruction is less common, it has been related to morbidity and mortality of SSc patients. We previously reported a close correlation between the