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## THE BASELINE CHARACTERISTICS AND SURVIVAL OF CHINESE PATIENTS WITH CONNECTIVE TISSUE DISEASE ASSOCIATED PULMONARY ARTERIAL HYPERTENSION

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**Objectives** Pulmonary arterial hypertension (PAH) is a severe complication of connective tissue disease (CTD) with a poor prognosis. There have been sporadic reports with respect to the clinical features and survival of CTD associated PAH (CPAH), however, those in Chinese with CPAH are unknown yet. The purpose of this study is to investigate the baseline characteristics, survival and risk factors of mortality in Chinese with CPAH.

**Methods** All consecutive adult patients who visited the three medical centres with confirmed diagnosis of CPAH between July 2006 and May 2011 were enrolled into the study. For all these patients, PAH was confirmed by right heart catheterisation.

**Results** A total of 144 patients (40.6±12.6 years old) were included in the study and 44% of them were associated with SLE. The other underlying CTDs, in the descending rank order, are pSS (15%), Takayasu arteritis (12%), MCTD (10%), SSc (8%) and some others (RA 3%, PM/DM 2%, adult onset Still'disease 2%, UCTD 2%, primary APS 1%, and ANCA associated vasculitis 1%).

The median duration between symptom onset and diagnostic catheterisation was 16.5 months. At diagnosis, 57.6% of patients were in WHO functional class III/IV. The 6-min walk distance was 377.0±99.7 m. Mean pulmonary artery pressure was 49.7 ±14.4 mm Hg. Eighty-five percent of patients received vascular-targeted therapy.

One hundred and twenty-nine patients were followed up with a median duration of 15.8 months (ranged 1.1–55.1 months). The survival rates of these patients at 1 and 3 years were 87.8% and 53.8%. The survival rates of patients with SLE associated PAH at 1 and 3 years were 90.0% and 57.1%. K-M survival analysis showed there were no significant differences in the survivals among different connective tissue diseases.

Univariate Cox analysis showed shorter 6-min walk distance, lower cardiac output, cardiac index and mixed venous oxygen saturation, higher pulmonary vascular resistance (PVR), alkaline phosphatase (ALP), total bilirubin and direct bilirubin, lower total cholesterol and low-density lipoprotein were associated with high risk of death (all $p<0.05$). Multivariate Cox analysis showed higher PVR and ALP were independent predictors of mortality (HR were 1.32 (1.03–1.68) and 1.70 (1.01–2.87) respectively, both $p<0.05$). K-M analysis demonstrated the survival rate in PVR<15 wood unit group was higher significantly than that in ≥15 wood unit group ($p=0.009$), and the survival rate in ALp<150 U/l group higher than that in ≥150 U/l group ($p=0.012$).

**Conclusions** SLE was the most common underlying disease of CPAH in China; however, SSc-associated PAH was fewer in Chinese patients, which were much different from Caucasians. The survival of Chinese patients with CPAH at 1 and 3 years were 87.8% and 53.8%, which were similar with the data of Western countries. Furthermore, elevated PVR and ALP were independent risk factors of bad outcomes.