Objective: Pulmonary hypertension (PH) is the most severe complication with Systemic Scleroderma (SSc). In this study, we analyzed the prognostic factors related to PH using a database registered as an intractable disease by the Ministry of Health, Labor, and Welfare (MHLW).

Methods: The anonymized data were made available to us for analysis (MHLW; No.0708-1; 2010). We used data of patients with SSc registered as a specific disease in 2003-2008 by MHLW. Data cleansing was carried out following the SSc Treatment Guidelines (2012) applied by the MHLW research group. We performed logistic regression for the association between autoantibodies and visceral lesions and prognostic factors analysis for the onset of PH.

Results: With analyzing 22,524 cases registered at 2003-2008, anti-Scl-70 antibody was related with PH (odds ratio 1.54 [95% CI 1.35-1.76]), renal crisis (1.30 [1.00-1.69]), and cardiac conduction disorder (1.15 [1.01-1.31]). Anti-U1-RNP antibody was related to PH (1.46 [1.24-1.73]). We focused on primary and secondary PH and found anti-centromere antibody was related to primary PH (2.01 [1.55-2.62]) while anti-U1-RNP and anti-Scl-70 antibody was related to secondary PH (1.74 [1.43-2.12], 2.11 [1.81-2.45]). As for prognostic factors analysis for the onset of PH after three years with 4,135 cases, the prostaglandin (PG) was identified as a prognostic factor (1.95 [1.17-3.25]), while steroids, immunosuppressants (anti-rheumatic agents), and angiotensin-converting enzyme inhibitors were not identified.

Conclusion: The anti-centromere antibody is associated with primary PH, and the anti-Scl-70 antibody is related to secondary PH related to pulmonary fibrosis. Although PG has been considered to suppress PH's onset, PG is suggested as a prognostic factor for PH.