Background: Scleroderma renal crisis (SRC) represents a rare but life-threatening manifestation in systemic sclerosis (SSc). Survival remains poor despite therapeutic use of ACE inhibitors (ACEi). Factors influencing the risk of SRC are not well characterised. In particular, ACEi are discussed as promoting but also as pro-therapeutic use of ACE inhibitors (ACEi).

Methods: EUSTAR database analysis with focus on arterial hypertension, antihypertensive medication and glucocorticoids. Subgroup analysis of a dataset with defined documentation of medication from January 2009 until November 2017.

Results: Out of 14,924 patients in the database we identified 7,648 patients with at least one follow-up after 2009. 102 patients developed SRC in 27,450 person-years (py), representing an incidence of 3.72 (3.06–4.51) per 1,000 py. In a multi-variable time-to-event analysis adjusted for age, sex, disease severity and onset, arterial hypertension, tendion friction rubs, SCL70 and ACA positivity, 78 of 6083 patients developed SRC. Herein, use of ACEi displayed an increased risk for the development of SRC with a hazard ratio (HR) of 2.07 (95% confidence interval (CI):1.28–3.36). Calcium channel blockers (CCB), angiotensin receptor blockers, endothelin receptor antagonists and glucocorticoids did not influence SRC incidence. Medication strategies were not altered after renal crisis. Cumulative mortality 5 years after renal crisis was 18.6% (95% CI:13.0%–26.3%).

Conclusions: This EUSTAR analysis supports the notion that ACEi should be avoided in arterial hypertension in SSC patients. Alternative antihypertensive drugs as CCB might be preferred.

REFERENCES:

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