Conclusion. high levels of NT-proBNP are associated with increased mortality in SSc patients even in the absence of overt pulmonary arterial hypertension. Our findings lend further support to the inclusion of NT-proBNP dosage in the periodic screening algorithms of SSc patients.

References

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Special Lecture

SL.1.1

DESSCIPHER, A JUMP IN THE FUTURE

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DeSScipher aims at improving clinical practice in the management of SSc for which no orphan drug is available. Over a three-year period, building upon the expertise of a multidisciplinary experienced consortium combining clinicians, physicians, biostatisticians, biologists and chemists, DeSScipher will provide a systemic approach to SSc. This approach isbased on five observational trials including different dimensions of the SSc disease: from early detection to severe complications, from child to adult patients, different organ levels (lung, heart) and integrates all generated outputs at the complexe level of the SSc disease. This project is comparing the outcomes of different preventive measures and treatments in terms of efficacy of off-label drugs and of adverse events from the early phase to severe phases of the SSc-associated diseases. DeSScipher will define appropriate outcome measures (OM) in SSc management. It includes several key experts - experienced in working together - from the EUSTAR consortium with more than 150 expert centres and 9000 well controlled and documented patients. Consequently, it is the aim of the DeSScipher project to improve clinical practice in the management of SScoccuring in adults but also in children by relying on the activities of the EUSTAR consortium. The concept of DeSScipher is using the EUSTAR long-term databank MEDSonline (Minimal Essential Data Set), the EUSTAR biobank and a preliminary diagnostic tool VEDOSS and to analyse and extend these data to shed light on specific molecular, genetic and functional details of SSc pathophysiology ranging from the early inflammatory phase to alterations of the vascular architecture to the clinically overt fibrosis of the connective tissues of the skin and internal organs. To facilitate the approach of DeSScipher, five observational trials are running to cover the disease evolution phases from early diagnosis such as digital ulcers and hand arthritis to the associated morbidity-driving pathologies such as interstitial lung disease, pulmonary hypertension or left heart disease. For this purpose, DeSScipher compares different scenarios: preventative measures and/or treatments to define appropriate primary and secondary outcome measures in order to evaluate the effectiveness as well as the adverse effects of the off-label candidate drugs and orphan drugs. For that purpose, the MEDSonline database as the biggest database in the field of an orphan rheumatic disease and the EUSTAR biobank have also been made available for and implemented by DeSScipher. DeSScipher will also serve as prototype approach for all other orphan diseases in the field of rheumatology and clinical immunology with similar problems in recruiting and documenting patients for testing novel drugs and establishing validated general international recommendations. Interested to contribute ? Visit the booth at the 3rd WSC or our website www.desscipher.eu or mail desscipher@med.uni-giessen.de
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