2017 EULAR Recommendations for the Treatment of Systemic Sclerosis (SSc)

The European League for Rheumatism (EULAR) periodically reviews the most research literature on the treatment of systemic sclerosis (SSc) and publishes updated treatment recommendations. The most recent guidelines were published in April 2017. Here is a direct link to the full published paper:

http://ard.bmj.com/content/early/2017/04/25/annrheumdis-2016-209909?papetoc.

The paper is targeted at clinicians and may be difficult to understand unless you have a medical background.

The paper includes a table which summarizes their recommendations in a concise format. I have "translated" the summary table into a somewhat more understandable format for those of you who are not comfortable reading medical papers. The revised table is below:

Symptom	Recommendation	Grade*
Raynaud's Phenomenon (RP)	Calcium channel blockers, usually oral nifedipine, should be considered for first-line therapy for scleroderma related RP. Phosphodiesterase Type 5 (PDE5) inhibitors (e.g., Viagra) should also be considered in treatment of RP.	A
	Intravenous iloprost (e.g., Ventavis) should be considered for severe RP. Experts recommend that intravenous iloprost should be used for treatment of RP attacks after first trying oral therapy such as nifedipine.	A
	Fluoxetine (Prozac) might be considered in treatment of RP attacks.	C
Digital Ulcers (DU)	Intravenous iloprost (e.g., Ventavis) should be considered in the treatment of digital ulcers in patients with SSc.	A
	PDE-5 inhibitors (e.g., Viagra) should be considered in the treatment of digital ulcers in patients with SSc.	A
	Endothelin receptor antagonists (e.g., Tracleer) should be considered for reduction of the number of new digital ulcers in SSc, especially in patients with multiple digital ulcers despite use of calcium channel blockers, PDE-5 inhibitors or iloprost therapy.	A

* strength of recommendation

Symptom	Recommendation	Grade*
Pulmonary Artery Hypertension (PAH)	Endothelin receptor antagonists (e.g., Tracleer), PDE-5 inhibitors (e.g., Viagra) or riociguat (Adempas) should be considered to treat PAH.	B
	Intravenous epoprostenol (e.g., Flolan) should be considered for the treatment of patients with severe PAH.	A
	Prostacyclin analogues (e.g., Ventavis) should be considered for the treatment of patients with PAH.	B
Skin and lung disease	Methotrexate may be considered for treatment of skin manifestations of early diffuse SSc.	A
	In view of the results from two high-quality randomized controlled trials and despite its known toxicity, cyclophosphamide (Cytoxan) should be considered for treatment of interstitial lung disease (ILD), in particular for patients with progressive ILD.	A
	Autologous stem cell transplants (HSCT) should be considered for treatment of selected patients with rapidly progressive SSc at risk of organ failure. In view of the high risk of treatment-related side effects and of early treatment-related mortality, careful selection of patients with SSc for this kind of treatment and the experience of the medical team are of key importance.	A
Scleroderma Renal Crisis (SRC)	Experts recommend immediate use of ACE inhibitors in the treatment of SRC.	C
	Blood pressure and renal function should be carefully monitored in patients with SSc treated with glucocorticoids (e.g., prednisone).	C
Gastrointestinal symptoms	Proton pump inhibitors (PPIs) (e.g., omeprazole) should be used for the treatment of gastro-oesophageal reflux disease (GERD) and prevention of oesophageal ulcers and strictures.	C
	Prokinetic drugs should be used for the management of SSc-related symptomatic motility disturbances (difficulty swallowing, GERD, early satiety, bloating, pseudo-obstruction, etc).	C
	Intermittent or rotating antibiotics should be used to treat symptomatic small intestine bacterial overgrowth in patients with SSc.	D

* strength of recommendation

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