



POSTER PRESENTATION

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PReS-FINAL-2171: Consensus: what agent to use when first-line vasodilators fail in Raynaud's phenomenon or digital ulcers secondary to rheumatic diseases in children?

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Introduction

Juvenile Systemic Sclerosis (JSS) is characterized by Raynaud's phenomenon (RP) and digital ulcers (DU). Conventional therapy includes calcium channel blockers (CCB). A growing number of vasodilators is available for treatment of refractory patients but there is no clear evidence of the best option. To aid clinical decision-making, a consensus of experts was undertaken.

Objectives

To identify the best therapeutic options and define the sequence of 2nd line vasodilators for RP and DU.

Methods

Steps in the process of consensus were: a) Identification of expert panel (EP) members, b) Identification of 2nd line vasodilators c) identification of outcome measures to define RP and DU improvement, d) systematic literature review; e) summary report of the latest scientific evidence f) expert consensus meeting; g) rating of the strength of evidence. RAND/UCLA appropriateness method was used for rating the medical decision: items were rated on a 9-point scale on each drug option. There were two scoring rounds: first: anonymous and independent rating of the appropriateness of vasodilators based on scientific evidence and best clinical judgment. Differences in scoring were discussed at a face-to-face meeting, followed by a second rating round. Consensus was reached on appropriate/inappropriateness.

Results

The EP included 10 physicians from a tertiary center who are involved in the care of patients with JSS: 3 pediatric rheumatologists, 2 dermatologists, 1 pediatrician, 1 gastroenterologist, 1 nephrologist, 1 nutritionist, 1 pharmacologist, and a moderator. The EP identified 4 drugs for analysis: bosentan, iloprost, sildenafil, and trepostinil. Outcome measures were selected according to the literature references and EP judgment. RP improvement definition: $\geq 30\%$ improvement according to the physician (in a visual analogue scale, VAS) and $\geq 30\%$ improvement in at least 2 patient-related domains (pain or function). Patient domains were: a) number of episodes, b) pain in a VAS, c) function (impaired activity of daily living, VAS), d) RP episodes average duration (in minutes). DU improvement definition: a favorable change in all physician- and patient-related domains: patient's domains: a) pain (VAS) b) function (VAS); physician's domains: a) ulcer activity (VAS) b) horizontal and transverse DU diameter (in mm). Systematic literature review was performed independently by 5 EP members and guided by the moderator. All articles in English were eligible. Data bases included pubmed and Cochrane. The search strategy included all relevant terms: bosentan, iloprost, sildenafil, trepostinil, RP, DU, combined in different sets of keywords. The summary report of the scientific evidence included 25 articles. Ranking of papers according to the strength of evidence showed: 1a (1 paper), 1b (7), 2b (2), 3b (2), 4(8), 5(5). After second scoring round: 1st appropriate indication Iloprost; 2nd bosentan, 3rd sildenafil; 4th trepostinil.

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Conclusion

The EP reached a consensus on vasodilator drugs, providing direction for common dilemmas in the pharmacologic treatment of RP and DU in refractory patients.

Disclosure of interest

None declared.

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