

## Supplementary Material

### The detailed search strategy:

((“randomized controlled trial”[Publication Type] OR “randomized controlled trials as topic”[MeSH Terms] OR “randomized controlled trial”[All Fields] OR “randomised controlled trial”[All Fields]) AND (((((“lupus erythematosus, systemic”[MeSH Terms] OR (“lupus”[All Fields] AND “erythematosus”[All Fields] AND “systemic”[All Fields]) OR “systemic lupus erythematosus”[All Fields] OR (“systemic”[All Fields] AND “lupus”[All Fields] AND “erythematosus”[All Fields])) OR ((“scleroderma, systemic”[MeSH Terms] OR (“scleroderma”[All Fields] AND “systemic”[All Fields]) OR “systemic scleroderma”[All Fields] OR (“systemic”[All Fields] AND “sclerosis”[All Fields]) OR “systemic sclerosis”[All Fields] OR (“scleroderma, systemic”[MeSH Terms] OR (“scleroderma”[All Fields] AND “systemic”[All Fields]) OR “systemic scleroderma”[All Fields] OR “scleroderma”[All Fields] OR “scleroderma, localized”[MeSH Terms] OR (“scleroderma”[All Fields] AND “localized”[All Fields]) OR “localized scleroderma”[All Fields]))) OR (“sjogren’s syndrome”[MeSH Terms] OR (“sjogren’s”[All Fields] AND “syndrome”[All Fields]) OR “sjogren’s syndrome”[All Fields] OR (“sjogren”[All Fields] AND “syndrome”[All Fields]) OR “sjogren syndrome”[All Fields])) OR (“polymyositis”[MeSH Terms] OR “polymyositis”[All Fields])) OR (“dermatomyositis”[MeSH Terms] OR “dermatomyositis”[All Fields])) OR (“connective tissue diseases”[MeSH Terms] OR (“connective”[All Fields] AND “tissue”[All Fields] AND “diseases”[All Fields]) OR “connective tissue diseases”[All Fields])) AND (“familial primary pulmonary hypertension”[MeSH Terms] OR (“familial”[All Fields] AND “primary”[All Fields] AND “pulmonary”[All Fields] AND “hypertension”[All Fields]) OR “familial primary pulmonary hypertension”[All Fields] OR (“pulmonary”[All Fields] AND “arterial”[All Fields] AND “hypertension”[All Fields]) OR “pulmonary arterial hypertension”[All Fields])) OR (“familial primary pulmonary hypertension”[MeSH Terms] OR (“familial”[All Fields] AND “primary”[All Fields] AND “pulmonary”[All Fields] AND “hypertension”[All Fields]) OR “familial primary pulmonary hypertension”[All Fields] OR (“pulmonary”[All Fields] AND “arterial”[All Fields] AND “hypertension”[All Fields]) OR “pulmonary arterial hypertension”[All Fields])) AND ((((((((((“prostaglandins”[MeSH Terms] OR “prostaglandins”[All Fields] OR (“epoprostenol”[MeSH Terms] OR “epoprostenol”[All Fields])) OR (“treprostinil”[Supplementary Concept] OR “treprostinil”[All Fields])) OR (“iloprost”[MeSH Terms] OR “iloprost”[All Fields])) AND (((“endothelin receptor antagonists”[Pharmacological Action] OR “endothelin receptor antagonists”[MeSH Terms] OR (“endothelin”[All Fields] AND “receptor”[All Fields] AND “antagonists”[All Fields]) OR (“ambrisentan”[Supplementary Concept] OR “ambrisentan”[All Fields])) OR (“bosentan”[Supplementary Concept] OR “bosentan”[All Fields])) OR (“macitentan”[Supplementary Concept] OR “macitentan”[All Fields])) AND (((“phosphodiesterase 5 inhibitors”[Pharmacological Action] OR “phosphodiesterase 5 inhibitors”[MeSH Terms] OR “sildenafil citrate”[MeSH Terms] OR (“sildenafil”[All Fields])) OR (“vardenafil dihydrochloride”[MeSH Terms] OR (“vardenafil”[All Fields] AND “dihydrochloride”[All Fields]) OR “vardenafil dihydrochloride”[All Fields] OR (“vardenafil”[All Fields])))) OR (((“endothelin receptor antagonists”[Pharmacological Action] OR (“endothelin”[All Fields] AND “receptor”[All Fields] AND “antagonists”[All Fields]) OR “endothelin receptor antagonists”[All Fields] AND “ambrisentan”[Supplementary Concept] OR “ambrisentan”[All Fields])) OR (“bosentan”[Supplementary Concept] OR “bosentan”[All Fields])) OR (“macitentan”[Supplementary Concept] OR “macitentan”[All Fields])) AND (((“soluble guanylyl cyclase”[MeSH Terms] OR (“soluble”[All Fields] AND “guanylyl”[All Fields] AND “cyclase”[All Fields]) OR “soluble guanylyl cyclase”[All Fields] OR (“soluble”[All Fields] AND “guanylate”[All Fields] AND “cyclase”[All Fields]) OR “soluble guanylate cyclase”[All Fields] AND “guanylyl”[All Fields] AND “cyclase”[All Fields]) AND stimulators[All Fields])) OR (“riociguat”[Supplementary Concept] OR “riociguat”[All Fields])))) OR (((((“prostaglandins”[MeSH Terms] OR (“prostaglandins”[All Fields] OR (“epoprostenol”[MeSH Terms] OR “epoprostenol”[All Fields])) OR (“treprostinil”[Supplementary Concept] OR “treprostinil”[All Fields])) OR (“iloprost”[MeSH Terms] OR “iloprost”[All Fields])) AND (((selective[All Fields] AND (“receptors,epoprostenol”[MeSH Terms] OR (“receptors”[All Fields] AND “epoprostenol”[All Fields]) OR “epoprostenol receptors”[All Fields] OR (“prostacyclin”[All Fields] AND “receptor”[All Fields]) OR “prostacyclin receptor”[All Fields]) AND agonist[All Fields]) OR (“selexipag”[Supplementary Concept] OR “selexipag”[All Fields])))) OR (((((“endothelin receptor antagonists ”[Pharmacological Action] OR “endothelin receptor antagonists”[MeSH Terms] OR (“endothelin”[All Fields] AND “receptor”[All Fields] AND “antagonists”[All Fields]) OR “endothelin receptor antagonists”[All Fields] AND “ambrisentan”[Supplementary Concept] OR “ambrisentan”[All Fields])) OR (“bosentan”[Supplementary Concept] OR “bosentan”[All Fields])) OR (“macitentan” [Supplementary Concept] OR “macitentan”[All Fields])) AND (((“phosphodiesterase 5 inhibitors”[Pharmacological Action] OR “phosphodiesterase 5 inhibitors”[MeSH Terms] OR “sildenafil citrate”[MeSH Terms] OR (“sildenafil”[All Fields])) OR (“vardenafil dihydrochloride”[MeSH Terms] OR (“vardenafil”[All Fields] AND “dihydrochloride”[All Fields]) OR “vardenafil dihydrochloride”[All Fields] OR (“vardenafil”[All Fields])))) OR (((“endothelin receptor antagonists”[Pharmacological Action] OR (“endothelin”[All Fields] AND “receptor”[All Fields] AND “antagonists”[All Fields]) OR “endothelin receptor antagonists”[All Fields] AND “ambrisentan”[Supplementary Concept] OR “ambrisentan”[All Fields])) OR (“bosentan”[Supplementary Concept] OR “bosentan”[All Fields])) OR (“macitentan”[Supplementary Concept] OR “macitentan”[All Fields])) AND (((“soluble guanylyl cyclase”[MeSH Terms] OR (“soluble”[All Fields] AND “guanylyl”[All Fields] AND “cyclase”[All Fields]) OR “soluble guanylyl cyclase”[All Fields] OR (“soluble”[All Fields] AND “guanylate”[All Fields] AND “cyclase”[All Fields]) OR “soluble guanylate cyclase”[All Fields]) AND stimulators[All Fields])) OR (“riociguat”[Supplementary Concept] OR “riociguat”[All Fields])))) OR (((((“endothelin receptor antagonists”[Pharmacological Action] OR (“endothelin”[All Fields] AND “receptor”[All Fields] AND “antagonists”[All Fields]) OR “endothelin receptor antagonists”[All Fields] AND “en-

("ambrisentan"[Supplementary Concept] OR "ambrisentan"[All Fields])) OR ("bosentan"[Supplementary Concept] OR "bosentan"[All Fields])) OR ("macitentan"[Supplementary Concept] OR "macitentan"[All Fields])) AND ((selective[All Fields] AND ("receptors,eprostenol"[MeSH Terms] OR ("receptors"[All Fields] AND "eprostenol"[All Fields]) OR "epoprostenol receptors"[All Fields] OR ("prostacyclin"[All Fields] AND "receptor"[All Fields]) OR "prostacyclin receptor"[All Fields]) AND agonist[All Fields]) OR ("selexipag"[Supplementary Concept] OR "selexipag"[All Fields]))) OR (((("phosphodiesterase 5 inhibitors"[Pharmacological Action] OR "phosphodiesterase 5 inhibitors"[MeSH Terms] OR "phosphodiesterase 5 inhibitors"[All Fields]) OR ("sildenafil citrate"[MeSH Terms] OR ("sildenafil"[All Fields] AND "citrate"[All Fields]) OR "sildenafil citrate"[All Fields] OR "sildenafil"[All Fields])) OR ("adalafil"[MeSH Terms] OR "adalafil"[All Fields])) OR ("vardenafil dihydrochloride"[MeSH Terms] OR ("vardenafil"[All Fields] AND "dihydrochloride"[All Fields]) OR "vardenafil dihydrochloride"[All Fields] OR "vardenafil"[All Fields])) AND ((selective[All Fields] AND ("receptors, eprostenol"[MeSH Terms] OR ("receptors"[All Fields] AND "eprostenol"[All Fields]) OR "epoprostenol receptors"[All Fields] OR ("prostacyclin"[All Fields] AND "receptor"[All Fields]) OR "prostacyclin receptor"[All Fields]) AND agonist[All Fields]) OR ("selexipag"[Supplementary Concept] OR "selexipag"[All Fields]))) OR (((("soluble guanylyl cyclase"[MeSH Terms] OR ("soluble"[All Fields] AND "guanylyl"[All Fields] AND "cyclase"[All Fields]) OR "soluble guanylyl cyclase"[All Fields] OR ("soluble"[All Fields] AND "guanylate"[All Fields] AND "cyclase"[All Fields]) OR "soluble guanylate cyclase"[All Fields]) AND stimulators[All Fields]) OR ("riociguat"[Supplementary Concept] OR "riociguat"[All Fields])) AND ((selective[All Fields] AND ("receptors,eprostenol"[MeSH Terms] OR ("receptors"[All Fields] AND "eprostenol"[All Fields]) OR "epoprostenol receptors"[All Fields] OR ("prostacyclin"[All Fields] AND "receptor"[All Fields]) OR "prostacyclin receptor"[All Fields]) AND agonist[All Fields]) OR ("selexipag"[Supplementary Concept] OR "selexipag"[All Fields]))) AND (Clinical Trial[ptyp] AND "humans"[MeSH Terms]))