

Supplemental File S4. Articles excluded and the reason of exclusion

Articles excluded and the reason of exclusion	
Reference	Exclusion criteria
Badesch DB, Tapson VF, McGoon MD, Brundage BH, Rubin LJ, Wigley FM, Rich S, Barst RJ, Barrett PS, Kral KM, Jöbsis MM, Loyd JE, Murali S, Frost A, Girgis R, Bourge RC, Ralph DD, Elliott CG, Hill NS, Langleben D, Schilz RJ, McLaughlin VV, Robbins IM, Groves BM, Shapiro S, Medsger TA Jr. Continuous intravenous epoprostenol for pulmonary hypertension due to the scleroderma spectrum of disease. A randomized, controlled trial. <i>Ann Intern Med</i> . 2000 Mar 21;132(6):425-34. doi: 10.7326/0003-4819-132-6-200003210-00002. PMID: 10733441.	No subgroup analyses reported
Givertz MM, Colucci WS, LeJemtel TH, Gottlieb SS, Hare JM, Slawsky MT, Leier CV, Loh E, Nicklas JM, Lewis BE. Acute endothelin A receptor blockade causes selective pulmonary vasodilation in patients with chronic heart failure. <i>Circulation</i> . 2000 Jun 27;101(25):2922-7. doi: 10.1161/01.cir.101.25.2922. PMID: 10869264.	No subgroup analyses reported
Channick RN, Simonneau G, Sitbon O, Robbins IM, Frost A, Tapson VF, Badesch DB, Roux S, Rainisio M, Bodin F, Rubin LJ. Effects of the dual endothelin-receptor antagonist bosentan in patients with pulmonary hypertension: a randomised placebo-controlled study. <i>Lancet</i> . 2001 Oct 6;358(9288):1119-23. doi: 10.1016/S0140-6736(01)06250-X. PMID: 11597664.	No subgroup analyses reported
Gessler T, Schmehl T, Hoeper MM, Rose F, Ghofrani HA, Olschewski H, Grimminger F, Seeger W. Ultrasonic versus jet nebulization of iloprost in severe pulmonary hypertension. <i>Eur Respir J</i> . 2001 Jan;17(1):14-9. doi: 10.1183/09031936.01.17100140. PMID: 11307743.	No subgroup analyses reported
Ghofrani HA, Wiedemann R, Rose F, Schermuly RT, Olschewski H, Weissmann N, Gunther A, Walmrath D, Seeger W, Grimminger F. Sildenafil for treatment of lung fibrosis and pulmonary hypertension: a randomised controlled trial. <i>Lancet</i> . 2002 Sep 21;360(9337):895-900. doi: 10.1016/S0140-6736(02)11024-5. PMID: 12354470.	No subgroup analyses reported
Simonneau G, Barst RJ, Galie N, Naeije R, Rich S, Bourge RC, Keogh A, Oudiz R, Frost A, Blackburn SD, Crow JW, Rubin LJ; Treprostinil Study Group. Continuous subcutaneous infusion of treprostinil, a prostacyclin analogue, in patients with pulmonary arterial hypertension: a double-blind, randomized, placebo-controlled trial. <i>Am J Respir Crit Care Med</i> . 2002 Mar 15;165(6):800-4. doi: 10.1164/ajrccm.165.6.2106079. PMID: 11897647.	No subgroup analyses reported
Barst RJ, McGoon M, McLaughlin V, Tapson V, Rich S, Rubin L, Wasserman K, Oudiz R, Shapiro S, Robbins IM, Channick R, Badesch D, Rayburn BK, Flinchbaugh R, Sigman J, Arneson C, Jeffs R; Beraprost Study Group. Beraprost therapy for pulmonary arterial hypertension. <i>J Am Coll Cardiol</i> . 2003 Jun 18;41(12):2119-25. doi: 10.1016/s0735-1097(03)00463-7. Erratum in: <i>J Am Coll Cardiol</i> . 2003 Aug 6;42(3):591. PMID: 12821234.	No subgroup analyses reported

Galiè N, Hinderliter AL, Torbicki A, Fourme T, Simonneau G, Pulido T, Espinola-Zavaleta N, Rocchi G, Manes A, Frantz R, Kurzyna M, Nagueh SF, Barst R, Channick R, Dujardin K, Kronenberg A, Leconte I, Rainisio M, Rubin L. Effects of the oral endothelin-receptor antagonist bosentan on echocardiographic and doppler measures in patients with pulmonary arterial hypertension. <i>J Am Coll Cardiol</i> . 2003 Apr 16;41(8):1380-6. doi: 10.1016/s0735-1097(03)00121-9. PMID: 12706935.	Post-hoc analysis of a previously published RCT
Haché M, Denault A, Bélisle S, Robitaille D, Couture P, Sheridan P, Pellerin M, Babin D, Noël N, Guertin MC, Martineau R, Dupuis J. Inhaled epoprostenol (prostacyclin) and pulmonary hypertension before cardiac surgery. <i>J Thorac Cardiovasc Surg</i> . 2003 Mar;125(3):642-9. doi: 10.1067/mtc.2003.107. PMID: 12658208.	No subgroup analyses reported
Kanazawa H, Hirata K, Yoshikawa J. Effects of captopril administration on pulmonary haemodynamics and tissue oxygenation during exercise in ACE gene subtypes in patients with COPD: a preliminary study. <i>Thorax</i> . 2003 Jul;58(7):629-31. doi: 10.1136/thorax.58.7.629. PMID: 12832683; PMCID: PMC1746721.	No subgroup analyses reported
Vonbank K, Ziesche R, Higenbottam TW, Stiebellehner L, Petkov V, Schenk P, Germann P, Block LH. Controlled prospective randomised trial on the effects on pulmonary haemodynamics of the ambulatory long term use of nitric oxide and oxygen in patients with severe COPD. <i>Thorax</i> . 2003 Apr;58(4):289-93. doi: 10.1136/thorax.58.4.289. PMID: 12668787; PMCID: PMC1746623.	No subgroup analyses reported
Barst RJ, Langleben D, Frost A, Horn EM, Oudiz R, Shapiro S, McLaughlin V, Hill N, Tapson VF, Robbins IM, Zwicke D, Duncan B, Dixon RA, Frumkin LR; STRIDE-1 Study Group. Sitaxsentan therapy for pulmonary arterial hypertension. <i>Am J Respir Crit Care Med</i> . 2004 Feb 15;169(4):441-7. doi: 10.1164/rccm.200307-957OC. Epub 2003 Nov 20. PMID: 14630619.	No subgroup analyses reported
Ghofrani HA, Voswinckel R, Reichenberger F, Olschewski H, Haredza P, Karadaş B, Schermuly RT, Weissmann N, Seeger W, Grimminger F. Differences in hemodynamic and oxygenation responses to three different phosphodiesterase-5 inhibitors in patients with pulmonary arterial hypertension: a randomized prospective study. <i>J Am Coll Cardiol</i> . 2004 Oct 6;44(7):1488-96. doi: 10.1016/j.jacc.2004.06.060. PMID: 15464333.	No subgroup analyses reported
Goldsmith DR, Wagstaff AJ. Inhaled iloprost: in primary pulmonary hypertension. <i>Drugs</i> . 2004;64(7):763-73; discussion 774-5. doi: 10.2165/00003495-200464070-00009. PMID: 15025551.	The article did not report results from a controlled randomized trial
Humbert M, Barst RJ, Robbins IM, Channick RN, Galiè N, Boonstra A, Rubin LJ, Horn EM, Manes A, Simonneau G. Combination of bosentan with epoprostenol in pulmonary arterial hypertension: BREATHE-2. <i>Eur Respir J</i> . 2004 Sep;24(3):353-9. doi: 10.1183/09031936.04.00028404. PMID: 15358690.	No subgroup analyses reported

Langleben D, Brock T, Dixon R, Barst R; STRIDE-1 study group. STRIDE 1: effects of the selective ET(A) receptor antagonist, sitaxsentan sodium, in a patient population with pulmonary arterial hypertension that meets traditional inclusion criteria of previous pulmonary arterial hypertension trials. <i>J Cardiovasc Pharmacol</i> . 2004 Nov;44 Suppl 1:S80-4. doi: 10.1097/01.fjc.0000166207.74178.d0. PMID: 15838366.	Post-hoc analysis of a previously published RCT
Oudiz RJ, Schilz RJ, Barst RJ, Galié N, Rich S, Rubin LJ, Simonneau G; Treprostinil Study Group. Treprostinil, a prostacyclin analogue, in pulmonary arterial hypertension associated with connective tissue disease. <i>Chest</i> . 2004 Aug;126(2):420-7. doi: 10.1378/chest.126.2.420. PMID: 15302727.	The article did not report results from a controlled randomized trial
Sastry BK, Narasimhan C, Reddy NK, Raju BS. Clinical efficacy of sildenafil in primary pulmonary hypertension: a randomized, placebo-controlled, double-blind, crossover study. <i>J Am Coll Cardiol</i> . 2004 Apr 7;43(7):1149-53. doi: 10.1016/j.jacc.2003.10.056. PMID: 15063421.	No subgroup analyses reported
Aldashev AA, Kojonazarov BK, Amatov TA, Sooronbaev TM, Mirrakhimov MM, Morrell NW, Wharton J, Wilkins MR. Phosphodiesterase type 5 and high altitude pulmonary hypertension. <i>Thorax</i> . 2005 Aug;60(8):683-7. doi: 10.1136/thx.2005.041954. PMID: 16061711; PMCID: PMC1747498.	No subgroup analyses reported
Fattouch K, Sbraga F, Bianco G, Speziale G, Gucciardo M, Sampognaro R, Ruvolo G. Inhaled prostacyclin, nitric oxide, and nitroprusside in pulmonary hypertension after mitral valve replacement. <i>J Card Surg</i> . 2005 Mar-Apr;20(2):171-6. doi: 10.1111/j.0886-0440.2005.200383w.x. PMID: 15725144.	No subgroup analyses reported
Genovese MC, Chakravarty EF, Boyle DL, Tutuncu Z, Thorburn CM, Halilhodzic M, Kroll S, Baughman J, Stewart S, Kavanaugh A. A randomized, blinded, parallel group, placebo controlled pilot study evaluating the effect of PVAC treatment in patients with diffuse systemic sclerosis. <i>J Rheumatol</i> . 2005 Dec;32(12):2345-50. PMID: 16331761.	The article was not available
Hoeper MM, Kramm T, Wilkens H, Schulze C, Schäfers HJ, Welte T, Mayer E. Bosentan therapy for inoperable chronic thromboembolic pulmonary hypertension. <i>Chest</i> . 2005 Oct;128(4):2363-7. doi: 10.1378/chest.128.4.2363. PMID: 16236895.	No subgroup analyses reported
Kramm T, Eberle B, Guth S, Mayer E. Inhaled iloprost to control residual pulmonary hypertension following pulmonary endarterectomy. <i>Eur J Cardiothorac Surg</i> . 2005 Dec;28(6):882-8. doi: 10.1016/j.ejcts.2005.09.007. Epub 2005 Oct 19. PMID: 16242948.	No subgroup analyses reported
McLaughlin VV, Sitbon O, Badesch DB, Barst RJ, Black C, Galié N, Rainisio M, Simonneau G, Rubin LJ. Survival with first-line bosentan in patients with primary pulmonary hypertension. <i>Eur Respir J</i> . 2005 Feb;25(2):244-9. doi: 10.1183/09031936.05.00054804. Erratum in: <i>Eur Respir J</i> . 2005 May;25(5):942. PMID: 15684287.	No subgroup analyses reported

Morrell NW, Higham MA, Phillips PG, Shakur BH, Robinson PJ, Beddoes RJ. Pilot study of losartan for pulmonary hypertension in chronic obstructive pulmonary disease. <i>Respir Res.</i> 2005 Aug 1;6(1):88. doi: 10.1186/1465-9921-6-88. PMID: 16060962; PMCID: PMC1198258.	No subgroup analyses reported
Ocal A, Kiriş I, Erdiñç M, Peker O, Yavuz T, İbrişim E. Efficiency of prostacyclin in the treatment of protamine-mediated right ventricular failure and acute pulmonary hypertension. <i>Tohoku J Exp Med.</i> 2005 Sep;207(1):51-8. doi: 10.1620/tjem.207.51. PMID: 16082155.	No subgroup analyses reported
Radovancevic B, Vrtovec B, Thomas CD, Croitoru M, Myers TJ, Radovancevic R, Khan T, Massin EK, Frazier OH. Nitric oxide versus prostaglandin E1 for reduction of pulmonary hypertension in heart transplant candidates. <i>J Heart Lung Transplant.</i> 2005 Jun;24(6):690-5. doi: 10.1016/j.healun.2004.04.016. PMID: 15949728.	No subgroup analyses reported
Seyfarth HJ, Pankau H, Hammerschmidt S, Schauer J, Wirtz H, Winkler J. Bosentan improves exercise tolerance and Tei index in patients with pulmonary hypertension and prostanoid therapy. <i>Chest.</i> 2005 Aug;128(2):709-13. doi: 10.1378/chest.128.2.709. PMID: 16100158.	No subgroup analyses reported
Shen J, He B, Wang B. Effects of lipo-prostaglandin E1 on pulmonary hemodynamics and clinical outcomes in patients with pulmonary arterial hypertension. <i>Chest.</i> 2005 Aug;128(2):714-9. doi: 10.1378/chest.128.2.714. PMID: 16100159.	No subgroup analyses reported
Wilkins MR, Paul GA, Strange JW, Tunariu N, Gin-Sing W, Banya WA, Westwood MA, Stefanidis A, Ng LL, Pennell DJ, Mohiaddin RH, Nihoyannopoulos P, Gibbs JS. Sildenafil versus Endothelin Receptor Antagonist for Pulmonary Hypertension (SERAPH) study. <i>Am J Respir Crit Care Med.</i> 2005 Jun 1;171(11):1292-7. doi: 10.1164/rccm.200410-1411OC. Epub 2005 Mar 4. PMID: 15750042.	No subgroup analyses reported
Fattouch K, Sbraga F, Sampognaro R, Bianco G, Gucciardo M, Lavalle C, Vizza CD, Fedele F, Ruvolo G. Treatment of pulmonary hypertension in patients undergoing cardiac surgery with cardiopulmonary bypass: a randomized, prospective, double-blind study. <i>J Cardiovasc Med (Hagerstown).</i> 2006 Feb;7(2):119-23. doi: 10.2459/01.JCM.0000203850.97890.fe. PMID: 16645371.	No subgroup analyses reported
Galiè N, Beghetti M, Gatzoulis MA, Granton J, Berger RM, Lauer A, Chiossi E, Landzberg M; Bosentan Randomized Trial of Endothelin Antagonist Therapy-5 (BREATHE-5) Investigators. Bosentan therapy in patients with Eisenmenger syndrome: a multicenter, double-blind, randomized, placebo-controlled study. <i>Circulation.</i> 2006 Jul 4;114(1):48-54. doi: 10.1161/CIRCULATIONAHA.106.630715. Epub 2006 Jun 26. PMID: 16801459.	No subgroup analyses reported
McLaughlin VV, Oudiz RJ, Frost A, Tapson VF, Murali S, Channick RN, Badesch DB, Barst RJ, Hsu HH, Rubin LJ. Randomized study of adding inhaled iloprost to existing bosentan in pulmonary arterial hypertension.	No subgroup analyses reported

Am J Respir Crit Care Med. 2006 Dec 1;174(11):1257-63. doi: 10.1164/rccm.200603-358OC. Epub 2006 Aug 31. PMID: 16946127.	
Singh TP, Rohit M, Grover A, Malhotra S, Vijayvergiya R. A randomized, placebo-controlled, double-blind, crossover study to evaluate the efficacy of oral sildenafil therapy in severe pulmonary artery hypertension. Am Heart J. 2006 Apr;151(4):851.e1-5. doi: 10.1016/j.ahj.2005.09.006. PMID: 16569546.	Subgroup analyses missing data
Voswinckel R, Enke B, Reichenberger F, Kohstall M, Kreckel A, Krick S, Gall H, Gessler T, Schmehl T, Ghofrani HA, Schermuly RT, Grimminger F, Rubin LJ, Seeger W, Olschewski H. Favorable effects of inhaled treprostinil in severe pulmonary hypertension: results from randomized controlled pilot studies. J Am Coll Cardiol. 2006 Oct 17;48(8):1672-81. doi: 10.1016/j.jacc.2006.06.062. Epub 2006 Sep 26. PMID: 17045906.	No subgroup analyses reported
Barst RJ, Langleben D, Badesch D, Frost A, Lawrence EC, Shapiro S, Naeije R, Galie N; STRIDE-2 Study Group. Treatment of pulmonary arterial hypertension with the selective endothelin-A receptor antagonist sitaxsentan. J Am Coll Cardiol. 2006 May 16;47(10):2049-56. doi: 10.1016/j.jacc.2006.01.057. Epub 2006 Apr 24. PMID: 16697324.	No subgroup analyses reported
Hoepfer MM, Leuchte H, Halank M, Wilkens H, Meyer FJ, Seyfarth HJ, Wensel R, Ripken F, Bremer H, Kluge S, Hoeffken G, Behr J. Combining inhaled iloprost with bosentan in patients with idiopathic pulmonary arterial hypertension. Eur Respir J. 2006 Oct;28(4):691-4. doi: 10.1183/09031936.06.00057906. PMID: 17012628.	No subgroup analyses reported
Barst RJ, Galie N, Naeije R, Simonneau G, Jeffs R, Arneson C, Rubin LJ. Long-term outcome in pulmonary arterial hypertension patients treated with subcutaneous treprostinil. Eur Respir J. 2006 Dec;28(6):1195-203. doi: 10.1183/09031936.06.00044406. Epub 2006 Aug 9. PMID: 16899485.	No subgroup analyses reported
Ludwik B, Lewczuk J, Piszko P, Nowak M, Wrabec K. Roczna, kontrolowana ocena stosowania doksazosyny u chorych na nadciśnienie płucne na tle przewlekłej obturacyjnej choroby płuc [One-year controlled therapy with doxazosin in patients with pulmonary hypertension due to chronic obstructive pulmonary disease]. Pol Merkuriusz Lekarski. 2006 May;20(119):527-30. Polish. PMID: 16875154.	The article was not available
McLaughlin VV. Survival in patients with pulmonary arterial hypertension treated with first-line bosentan. Eur J Clin Invest. 2006 Sep;36 Suppl 3:10-5. doi: 10.1111/j.1365-2362.2006.01688.x. PMID: 16919005.	No subgroup analyses reported
But AK, Ozgul U, Erdil F, Gulhas N, Toprak HI, Durmus M, Ersoy MO. The effects of pre-operative dexmedetomidine infusion on hemodynamics in patients with pulmonary hypertension undergoing mitral valve replacement surgery. Acta Anaesthesiol Scand. 2006 Nov;50(10):1207-12. doi: 10.1111/j.1399-6576.2006.01136.x. Epub 2006 Sep 15. PMID: 16978159.	No subgroup analyses reported
Shim JK, Choi YS, Oh YJ, Kim DH, Hong YW, Kwak YL. Effect of oral sildenafil	No subgroup analyses

citrate on intraoperative hemodynamics in patients with pulmonary hypertension undergoing valvular heart surgery. <i>J Thorac Cardiovasc Surg.</i> 2006 Dec;132(6):1420-5. doi: 10.1016/j.jtcvs.2006.08.035. PMID: 17140970.	reported
Yurtseven N, Karaca P, Uysal G, Ozkul V, Cimen S, Tuygun AK, Yuksek A, Canik S. A comparison of the acute hemodynamic effects of inhaled nitroglycerin and iloprost in patients with pulmonary hypertension undergoing mitral valve surgery. <i>Ann Thorac Cardiovasc Surg.</i> 2006 Oct;12(5):319-23. PMID: 17095973.	The article was not available
Yi S, Ran L, Gu XH. [Effects of pulmonary arterial perfusion with shenqi fuzheng injection on lung injury during cardiopulmonary bypass]. <i>Zhongguo Zhong Xi Yi Jie He Za Zhi.</i> 2006 Oct;26(10):938-41. Chinese. PMID: 17121051.	The article was written in a language other than english/spanish/french
Badesch DB, Hill NS, Burgess G, Rubin LJ, Barst RJ, Galiè N, Simonneau G; SUPER Study Group. Sildenafil for pulmonary arterial hypertension associated with connective tissue disease. <i>J Rheumatol.</i> 2007 Dec;34(12):2417-22. Epub 2007 Nov 1. PMID: 17985403.	Post-hoc analysis of a previously published RCT
Benza RL, Mehta S, Keogh A, Lawrence EC, Oudiz RJ, Barst RJ. Sitaxsentan treatment for patients with pulmonary arterial hypertension discontinuing bosentan. <i>J Heart Lung Transplant.</i> 2007 Jan;26(1):63-9. doi: 10.1016/j.healun.2006.10.019. PMID: 17234519.	No subgroup analyses reported
Lewis GD, Shah R, Shahzad K, Camuso JM, Pappagianopoulos PP, Hung J, Tawakol A, Gerszten RE, Systrom DM, Bloch KD, Semigran MJ. Sildenafil improves exercise capacity and quality of life in patients with systolic heart failure and secondary pulmonary hypertension. <i>Circulation.</i> 2007 Oct 2;116(14):1555-62. doi: 10.1161/CIRCULATIONAHA.107.716373. Epub 2007 Sep 4. PMID: 17785618.	No subgroup analyses reported
Bharani A, Patel A, Saraf J, Jain A, Mehrotra S, Lunia B. Efficacy and safety of PDE-5 inhibitor tadalafil in pulmonary arterial hypertension. <i>Indian Heart J.</i> 2007 Jul-Aug;59(4):323-8. PMID: 19126937.	The article was not available
Rubenfibre M, McLaughlin VV, Allen RP, Elliott G, Park MH, Wade M, Schilz R. Transition from IV epoprostenol to subcutaneous treprostinil in pulmonary arterial hypertension: a controlled trial. <i>Chest.</i> 2007 Sep;132(3):757-63. doi: 10.1378/chest.06-2118. Epub 2007 Mar 30. Erratum in: <i>Chest.</i> 2007 Nov;132(5):1721. PMID: 17400684.	No subgroup analyses reported
Sfikakis PP, Papamichael C, Stamatelopoulos KS, Tousoulis D, Fragiadaki KG, Katsichti P, Stefanadis C, Mavrikakis M. Improvement of vascular endothelial function using the oral endothelin receptor antagonist bosentan in patients with systemic sclerosis. <i>Arthritis Rheum.</i> 2007 Jun;56(6):1985-93. doi: 10.1002/art.22634. PMID: 17530638.	The trial was not design to include pulmonary hypertension patients
Oztekin I, Yazici S, Oztekin DS, Goksel O, Issever H, Canik S. Effects of low-dose milrinone on weaning from cardiopulmonary bypass and after in	No subgroup analyses reported

patients with mitral stenosis and pulmonary hypertension. <i>Yakugaku Zasshi</i> . 2007 Feb;127(2):375-83. doi: 10.1248/yakushi.127.375. PMID: 17268158.	
Kaluski E, Cotter G, Leitman M, Milo-Cotter O, Krakover R, Kobrin I, Moriconi T, Rainisio M, Caspi A, Reizin L, Zimlichman R, Vered Z. Clinical and hemodynamic effects of bosentan dose optimization in symptomatic heart failure patients with severe systolic dysfunction, associated with secondary pulmonary hypertension--a multi-center randomized study. <i>Cardiology</i> . 2008;109(4):273-80. doi: 10.1159/000107791. Epub 2007 Sep 17. PMID: 17873492.	No subgroup analyses reported
Skoro-Sajer N, Lang IM, Harja E, Kneussl MP, Sing WG, Gibbs SJ. A clinical comparison of slow- and rapid-escalation treprostinil dosing regimens in patients with pulmonary hypertension. <i>Clin Pharmacokinet</i> . 2008;47(9):611-8. doi: 10.2165/00003088-200847090-00004. PMID: 18698881.	No subgroup analyses reported
Suntharalingam J, Treacy CM, Doughty NJ, Goldsmith K, Soon E, Toshner MR, Sheares KK, Hughes R, Morrell NW, Pepke-Zaba J. Long-term use of sildenafil in inoperable chronic thromboembolic pulmonary hypertension. <i>Chest</i> . 2008 Aug;134(2):229-236. doi: 10.1378/chest.07-2681. Epub 2008 Feb 8. PMID: 18263674.	No subgroup analyses reported
von Dossow V, Spies C, Schenk H, Schlesinger S, von Heymann C. Secondary pulmonary hypertension: haemodynamic effects of torasemide versus furosemide. <i>Clin Drug Investig</i> . 2008;28(1):17-26. doi: 10.2165/00044011-200828010-00003. PMID: 18081357.	No subgroup analyses reported
Behling A, Rohde LE, Colombo FC, Goldraich LA, Stein R, Clausell N. Effects of 5'-phosphodiesterase four-week long inhibition with sildenafil in patients with chronic heart failure: a double-blind, placebo-controlled clinical trial. <i>J Card Fail</i> . 2008 Apr;14(3):189-97. doi: 10.1016/j.cardfail.2007.11.006. PMID: 18381181.	No subgroup analyses reported
Stolz D, Rasch H, Linka A, Di Valentino M, Meyer A, Brutsche M, Tamm M. A randomised, controlled trial of bosentan in severe COPD. <i>Eur Respir J</i> . 2008 Sep;32(3):619-28. doi: 10.1183/09031936.00011308. Epub 2008 Apr 30. PMID: 18448495.	No subgroup analyses reported
Pepke-Zaba J, Gilbert C, Collings L, Brown MC. Sildenafil improves health-related quality of life in patients with pulmonary arterial hypertension. <i>Chest</i> . 2008 Jan;133(1):183-9. doi: 10.1378/chest.07-0592. PMID: 18187744.	No subgroup analyses reported
Galiè N, Olschewski H, Oudiz RJ, Torres F, Frost A, Ghofrani HA, Badesch DB, McGoon MD, McLaughlin VV, Roecker EB, Gerber MJ, Dufton C, Wiens BL, Rubin LJ; Ambrisentan in Pulmonary Arterial Hypertension, Randomized, Double-Blind, Placebo-Controlled, Multicenter, Efficacy Studies (ARIES) Group. Ambrisentan for the treatment of pulmonary arterial hypertension: results of the ambrisentan in pulmonary arterial hypertension, randomized, double-blind, placebo-controlled, multicenter, efficacy (ARIES) study 1 and	No subgroup analyses reported

<p>2. Circulation. 2008 Jun 10;117(23):3010-9. doi: 10.1161/CIRCULATIONAHA.107.742510. Epub 2008 May 27. PMID: 18506008.</p>	
<p>Cai J, Su Z, Shi Z, Zhou Y, Xu Z, Liu J, Chen L, Xu Z, Yu X, Ding W, Yang Y. Nitric oxide in conjunction with milrinone better stabilized pulmonary hemodynamics after Fontan procedure. <i>Artif Organs</i>. 2008 Nov;32(11):864-9. doi: 10.1111/j.1525-1594.2008.00643.x. PMID: 18959679.</p>	No subgroup analyses reported
<p>Winterhalter M, Simon A, Fischer S, Rahe-Meyer N, Chamtzidou N, Hecker H, Zuk J, Piepenbrock S, Strüber M. Comparison of inhaled iloprost and nitric oxide in patients with pulmonary hypertension during weaning from cardiopulmonary bypass in cardiac surgery: a prospective randomized trial. <i>J Cardiothorac Vasc Anesth</i>. 2008 Jun;22(3):406-13. doi: 10.1053/j.jvca.2007.10.015. Epub 2008 Jan 22. PMID: 18503929.</p>	No subgroup analyses reported
<p>Kleber FX, Bollmann T, Borst MM, Costard-Jäckle A, Ewert R, Kivikko M, Petterson T, Pohjanjousi P, Sonntag S, Wikström G. Repetitive dosing of intravenous levosimendan improves pulmonary hemodynamics in patients with pulmonary hypertension: results of a pilot study. <i>J Clin Pharmacol</i>. 2009 Jan;49(1):109-15. doi: 10.1177/0091270008325150. Epub 2008 Nov 3. PMID: 18981240.</p>	No subgroup analyses reported
<p>Baughman RP, Judson MA, Lower EE, Highland K, Kwon S, Craft N, Engel PJ. Inhaled iloprost for sarcoidosis associated pulmonary hypertension. <i>Sarcoidosis Vasc Diffuse Lung Dis</i>. 2009 Jul;26(2):110-20. PMID: 20560291.</p>	The article was not available
<p>Khan TA, Schnickel G, Ross D, Bastani S, Laks H, Esmailian F, Marelli D, Beygui R, Shemin R, Watson L, Vartapetian I, Ardehali A. A prospective, randomized, crossover pilot study of inhaled nitric oxide versus inhaled prostacyclin in heart transplant and lung transplant recipients. <i>J Thorac Cardiovasc Surg</i>. 2009 Dec;138(6):1417-24. doi: 10.1016/j.jtcvs.2009.04.063. PMID: 19931670.</p>	No subgroup analyses reported
<p>Seheult RD, Ruh K, Foster GP, Anholm JD. Prophylactic bosentan does not improve exercise capacity or lower pulmonary artery systolic pressure at high altitude. <i>Respir Physiol Neurobiol</i>. 2009 Feb 28;165(2-3):123-30. doi: 10.1016/j.resp.2008.10.005. Epub 2008 Oct 11. PMID: 18977464.</p>	No subgroup analyses reported
<p>Oudiz RJ, Galiè N, Olschewski H, Torres F, Frost A, Ghofrani HA, Badesch DB, McGoon MD, McLaughlin VV, Roecker EB, Harrison BC, Despain D, Dufton C, Rubin LJ; ARIES Study Group. Long-term ambrisentan therapy for the treatment of pulmonary arterial hypertension. <i>J Am Coll Cardiol</i>. 2009 Nov 17;54(21):1971-81. doi: 10.1016/j.jacc.2009.07.033. PMID: 19909879.</p>	No subgroup analyses reported
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Elinoff JM, Rame JE, Forfia PR, Hall MK, Sun J, Gharib AM, Abd-Elmoniem K, Graninger G, Harper B, Danner RL, Solomon MA. A pilot study of the effect of spironolactone therapy on exercise capacity and endothelial dysfunction in pulmonary arterial hypertension: study protocol for a randomized controlled trial. <i>Trials</i> . 2013 Apr 2;14:91. doi: 10.1186/1745-6215-14-91. PMID: 23547564; PMCID: PMC3653687.	Clinical Trial protocol
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Hoeper MM, Barst RJ, Bourge RC, Feldman J, Frost AE, Galié N, Gómez-Sánchez MA, Grimminger F, Grünig E, Hassoun PM, Morrell NW, Peacock AJ, Satoh T, Simonneau G, Tapson VF, Torres F, Lawrence D, Quinn DA, Ghofrani HA. Imatinib mesylate as add-on therapy for pulmonary arterial	The article did not report on pulmonary hypertension specific-therapy

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Schuuring MJ, Vis JC, van Dijk AP, van Melle JP, Vliegen HW, Pieper PG, Sieswerda GT, de Bruin-Bon RH, Mulder BJ, Bouma BJ. Impact of bosentan on exercise capacity in adults after the Fontan procedure: a randomized controlled trial. <i>Eur J Heart Fail</i> . 2013 Jun;15(6):690-8. doi: 10.1093/eurjhf/hft017. Epub 2013 Jan 29. PMID: 23361871.	The trial was not design to include pulmonary hypertension patients
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Simonneau G, Rubin LJ, Galiè N, Barst RJ, Fleming TR, Frost A, Engel P, Kramer MR, Serdarevic-Pehar M, Layton GR, Sitbon O, Badesch DB; PACES Study Group. Long-term sildenafil added to intravenous epoprostenol in patients with pulmonary arterial hypertension. <i>J Heart Lung Transplant</i> . 2014 Jul;33(7):689-97. doi: 10.1016/j.healun.2014.02.019. Epub 2014 Feb 22. PMID: 24815795.	No subgroup analyses reported
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<p>Sun X, Wang K, Wang W, Li B. [Clinical study on sildenafil in treatment of pregnant women with pulmonary arterial hypertension]. <i>Zhonghua Fu Chan Ke Za Zhi</i>. 2014 Jun;49(6):414-8. Chinese. PMID: 25169631.</p>	<p>The article was written in a language other than english/spanish/french</p>
<p>Baughman RP, Culver DA, Cordova FC, Padilla M, Gibson KF, Lower EE, Engel PJ. Bosentan for sarcoidosis-associated pulmonary hypertension: a double-blind placebo controlled randomized trial. <i>Chest</i>. 2014 Apr;145(4):810-817. doi: 10.1378/chest.13-1766. PMID: 24177203.</p>	<p>No subgroup analyses reported</p>
<p>Simonneau G, Galiè N, Jansa P, Meyer GM, Al-Hiti H, Kusic-Pajic A, Lemarié JC, Hoeper MM, Rubin LJ. Long-term results from the EARLY study of bosentan in WHO functional class II pulmonary arterial hypertension patients. <i>Int J Cardiol</i>. 2014 Mar 15;172(2):332-9. doi: 10.1016/j.ijcard.2013.12.179. Epub 2014 Jan 9. PMID: 24525158.</p>	<p>Post-hoc analysis of a previously published RCT</p>
<p>Schumacher DS, Müller-Mottet S, Hasler ED, Hildenbrand FF, Keusch S, Speich R, Bloch KE, Ulrich S. Effect of oxygen and acetazolamide on nocturnal cardiac conduction, repolarization, and arrhythmias in precapillary pulmonary hypertension and sleep-disturbed breathing. <i>Chest</i>. 2014 Nov;146(5):1226-1236. doi: 10.1378/chest.14-0495. PMID: 24991933.</p>	<p>No subgroup analyses reported</p>
<p>Jiang X, Wang YF, Zhao QH, Jiang R, Wu Y, Peng FH, Xu XQ, Wang L, He J, Jing ZC. Acute hemodynamic response of infused fasudil in patients with pulmonary arterial hypertension: a randomized, controlled, crossover study. <i>Int J Cardiol</i>. 2014 Nov 15;177(1):61-5. doi: 10.1016/j.ijcard.2014.09.101. Epub 2014 Sep 28. PMID: 25499341.</p>	<p>No subgroup analyses reported</p>
<p>Feng EZ, Yang SY, Huang NX, Yin H, Zhang Y, Tian ZX. Plasma endothelin-1 and nitric oxide correlate with ligustrazine alleviation of pulmonary artery hypertension in patients of chronic cor pulmonale from high altitude plateau during acute exacerbation. <i>Zhongguo Ying Yong Sheng Li Xue Za Zhi</i>. 2014 Nov;30(6):532-7. PMID: 26016363.</p>	<p>The article was not available</p>
<p>Chin KM, Badesch DB, Robbins IM, Tapson VF, Palevsky HI, Kim NH, Kawut SM, Frost A, Benton WW, Lemarie JC, Bodin F, Rubin LJ, McLaughlin V. Two formulations of epoprostenol sodium in the treatment of pulmonary arterial hypertension: EPITOME-1 (epoprostenol for injection in pulmonary arterial hypertension), a phase IV, open-label, randomized study. <i>Am Heart J</i>. 2014 Feb;167(2):218-225.e1. doi: 10.1016/j.ahj.2013.08.008. Epub 2013 Oct 16. PMID: 24439983.</p>	<p>No subgroup analyses reported</p>
<p>Jiang G, Li B, Zhang G, Xu E, Liu Y, Xu Z. Effects of sildenafil on prognosis in patients with pulmonary hypertension after left-sided valvular surgery. <i>Heart Lung Circ</i>. 2014 Jul;23(7):680-5. doi: 10.1016/j.hlc.2013.12.014. Epub 2014 Jan 31. PMID: 24613188.</p>	<p>No subgroup analyses reported</p>

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Goudie AR, Lipworth BJ, Hopkinson PJ, Wei L, Struthers AD. Tadalafil in patients with chronic obstructive pulmonary disease: a randomised, double-blind, parallel-group, placebo-controlled trial. <i>Lancet Respir Med</i> . 2014 Apr;2(4):293-300. doi: 10.1016/S2213-2600(14)70013-X. Epub 2014 Mar 5. PMID: 24717626.	No subgroup analyses reported
Lindegger N, Sidharta PN, Reseski K, Dingemanse J. Macitentan, a dual endothelin receptor antagonist for the treatment of pulmonary arterial hypertension, does not affect cardiac repolarization in healthy subjects. <i>Pulm Pharmacol Ther</i> . 2014 Oct;29(1):41-8. doi: 10.1016/j.pupt.2014.04.007. Epub 2014 May 9. PMID: 24813561.	Patients were not treated for pulmonary hypertension
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Boyko AN, Bosenko LP, Vasilovskiy VV, Volkova LI, Zakharova MN, Kotov SV, Lekomtseva EV, Negrich TI, Parshina EV, Patrusheva OP, Prokopenko SV, Sazonov DV, Timchenko LV, Trinitatskiy YV, Khabirov FA, Khavunka MY, Chichanovskaya LV, Sherman MA, Ivanov RA, Lin'kova YN, Stukalina EY, Zinkina-Orikhan AV, Obukhova IG. Éffektivnost', perenosimost' i bezopasnost' terapii preparatom teberif: rezul'taty dvukhletnego klinicheskogo issledovaniia u patsientov s remittiruiushchim rasseiannym sklerozom, ranee ne poluchavshikh PITRS, i pri perekliuchenii s drugogo interferona β -1a [Efficacy, tolerability and safety of the treatment with teberif: the results of a 2-year randomized clinical trial of treatment naïve patients with remitting multiple sclerosis, who have not received DMT, after switching from other interferon β -1a]. <i>Zh Nevrol Psikhiatr Im S S Korsakova.</i> 2019;119(2. Vyp. 2):73-85. Russian. doi: 10.17116/jnevro20191192273. PMID: 31156245.	The article was written in a language other than english/spanish/french
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Torres F, Farber H, Ristic A, McLaughlin V, Adams J, Zhang J, Klassen P, Shanahan W, Grundy J, Hoffmann I, Cabell C, Escibano Subías P, Sood N, Keogh A, D'Souza G, Rubin L. Efficacy and safety of ralinepag, a novel oral IP agonist, in PAH patients on mono or dual background therapy: results from a phase 2 randomised, parallel group, placebo-controlled trial. <i>Eur Respir J</i> . 2019 Oct 10;54(4):1901030. doi: 10.1183/13993003.01030-2019. PMID: 31391223.	No subgroup analyses reported
White RJ, Vonk-Noordegraaf A, Rosenkranz S, Oudiz RJ, McLaughlin VV, Hoeper MM, Grünig E, Ghofrani HA, Chakinala MM, Barberà JA, Blair C, Langley J, Frost AE. Clinical outcomes stratified by baseline functional class after initial combination therapy for pulmonary arterial hypertension. <i>Respir Res</i> . 2019 Sep 12;20(1):208. doi: 10.1186/s12931-019-1180-1. PMID: 31511080; PMCID: PMC6739949.	Post-hoc analysis of a previously published RCT
Winterhalter M, Rex S, Stoppe C, Kienbaum P, Müller HH, Kaufmann I, Kuppe H, Dongas A, Zwissler B; ILOCARD Investigators. Effect of iloprost inhalation on postoperative outcome in high-risk cardiac surgical patients: a prospective randomized-controlled multicentre trial (ILOCARD). <i>Can J Anaesth</i> . 2019 Aug;66(8):907-920. English. doi: 10.1007/s12630-019-01309-8. Epub 2019 Feb 12. PMID: 30756339.	Patients were not treated for pulmonary hypertension
Ram E, Sternik L, Klempfner R, Eldar M, Goldenberg I, Peled Y, Raanani E, Kogan A. Sildenafil for Pulmonary Hypertension in the Early Postoperative Period After Mitral Valve Surgery. <i>J Cardiothorac Vasc Anesth</i> . 2019 Jun;33(6):1648-1656. doi: 10.1053/j.jvca.2018.12.023. Epub 2018 Dec 25. PMID: 30685151.	No subgroup analyses reported
Sadushi-Kolici R, Jansa P, Kopec G, Torbicki A, Skoro-Sajer N, Campean IA, Halank M, Simkova I, Karlocai K, Steringer-Mascherbauer R, Samarzija M,	No subgroup analyses reported

<p>Salobir B, Klepetko W, Lindner J, Lang IM. Subcutaneous treprostinil for the treatment of severe non-operable chronic thromboembolic pulmonary hypertension (CTREPH): a double-blind, phase 3, randomised controlled trial. <i>Lancet Respir Med</i>. 2019 Mar;7(3):239-248. doi: 10.1016/S2213-2600(18)30367-9. Epub 2018 Nov 23. PMID: 30477763.</p>	
<p>Nathan SD, Behr J, Collard HR, Cottin V, Hoeper MM, Martinez FJ, Corte TJ, Keogh AM, Leuchte H, Mogulkoc N, Ulrich S, Wuyts WA, Yao Z, Boateng F, Wells AU. Riociguat for idiopathic interstitial pneumonia-associated pulmonary hypertension (RISE-IIP): a randomised, placebo-controlled phase 2b study. <i>Lancet Respir Med</i>. 2019 Sep;7(9):780-790. doi: 10.1016/S2213-2600(19)30250-4. Epub 2019 Aug 12. PMID: 31416769.</p>	<p>Subgroup analyses missing data</p>
<p>Pan Z, Marra AM, Benjamin N, Eichstaedt CA, Blank N, Bossone E, Cittadini A, Coghlan G, Denton CP, Distler O, Egenlauf B, Fischer C, Harutyunova S, Xanthouli P, Lorenz HM, Grünig E. Early treatment with ambrisentan of mildly elevated mean pulmonary arterial pressure associated with systemic sclerosis: a randomized, controlled, double-blind, parallel group study (EDITA study). <i>Arthritis Res Ther</i>. 2019 Oct 26;21(1):217. doi: 10.1186/s13075-019-1981-0. PMID: 31655622; PMCID: PMC6815440.</p>	<p>No subgroup analyses reported</p>
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