



## DAFTAR PUSTAKA

- Aldred, M.A., Vijayakrishnan, J., James, V., Soubrier, F., Gomez-Sanchez, M.A., Martensson, G., Galie, N., Manes, A., Corris, P., Simonneau, G., Humbert, M., Morrel, N.W., Trembath, R.C. 2006. BMPR2 gene rearrangements account for a significant proportion of mutations in familial and idiopathic pulmonary arterial hypertension. *Hum Mutat*;27:212-3.
- Archer, S. and Rich, S. 2000. Primary pulmonary hypertension: a vascular biology and translation research “Work in progress”. *Circulation*; 102: 2781-91. [SEP]
- Badesch, D.B., Raskob, G.E., Elliott, C.G., Krichman, A.M., Farber, H.W., Frost, A.E., Barst, R.J., Benza, R.L., Liou, T.G., Turner, M. 2010. Pulmonary arterial hypertension: Baseline characteristics from the reveal registry. *Chest*;137, 376–387.
- Barst, R.J., Chung, L., Zamanian, R.T., Turner, M., McGoon, M.D. 2013. Functional class improvement and 3-year survival outcomes in patients with pulmonary arterial hypertension in the REVEAL Registry. *Chest*;144:160–168.
- Baumgartner, H., Backer, J.D., Babu-Narayan, S.V., Budts, W., Chessa, M., Diller, G.P., Iung, B., Kluin, J., Lang, I.M., Meijboom, F., Moons, P., Mulder, B.J.M., Oechslin, E., Roos-Hesselink, J.W., Schwerzmann, M., Sondergraad, L., Zeppenfeld, K. 2020. 2020 ESC Guidelines for the management of adult congenital heart disease. *European Heart Journal*;00:1-83.
- Benza, R.L., Gomberg-Maitland, M., Naeiji, R., Arneson, C.P., Lang, I.M. 2011. Prognostic factors associated with increased survival in patients with pulmonary arterial hypertension treated with subcutaneous treprostinil in randomized, placebo-controlled trials. *J Heart Lung Transplant*; 30: 982–9.
- Benza, R.L., Miller, D.P., Barst, R.J., Badesch, D.B., Frost, A.E., McGoon, M.D. 2012. An evaluation of long-term survival from time of diagnosis in pulmonary arterial hypertension from the REVEAL Registry. *Chest*;142:448–456.
- Benza, R.L., Miller, D.P., Foreman, A.J., Frost, A.E., Badesch, D.B., Benton, W.W., McGoon, M.D. 2015. Prognostic implications of serial risk score assessments in patients with pulmonary arterial hypertension: a Registry to Evaluate Early and Long-Term Pulmonary Arterial Hypertension Disease Management (REVEAL) analysis. *J Heart Lung Transplant*; 34: 356–361.
- Benza, R.L., Miller, D.P., Gomberg-Maitland, M., Frantz, R.P., Foreman, A.J., Coffey, C.S., Frost, A., Barst, R.J., Badesch, D.B., Elliot, G., Liou, T.G., McGoon, M.D. 2015. Predicting survival in Pulmonary Arterial Hypertension. *Circulation*;122:164-172.
- Bogaard, H.J., Abe, K., Noordegraaf, A.V., Voelkel, N.F. 2009. The right ventricle under pressure: cellular and molecular mechanisms of right-heart failure in



- pulmonary hypertension. *Chest*; 135: 794–804.
- Campo, A., Mathai, S.C., Pevec, J.L., Zaiman, A.L., Hummers, L.K., Boyce, D., Houston, T., Lechtzin, N., Chami,H., Girgis, R.E., Hassoun, P.M. 2011. Outcomes of hospitalisation for right heart failure in pulmonary arterial hypertension. *Eur Respir J*; 38: 359–367.
- Champion, H.C., Michelakis, E.D., Hassoun, P.M. 2009. Comprehensive invasive and noninvasive approach to the right ventricle-pulmonary circulation unit: state of the art and clinical and research implications. *Circulation*; 120: 992–1007.
- Chaouat, A., Savale, L., Chouaid, C., Tu, L., Sztrymf, B., Canuet, M., Maitre, B., Housset, B., Brandt, C., Corvoisier, P.L., Weitzenblum, E., Eddahibi, S., Adnot, S. 2009. Role for interleukin-6 in COPD-related pulmonary hypertension. *Chest*;136:678-87.
- Christmas, B.W., McPherson, C.D., Newman, J.H., King, G.A., Bernard, G.R., Groves, B.M., Loyd, J.E. 1992. An imbalance<sup>[1]</sup>between the excretion of thromboxane and prostacyclin metabolites in pulmonary hypertension. *N Engl J Med*; 327: 70-5.
- Diller, G.P., Dimopoulos, K., Broberg, C.S., Kaya, M.G., Naghotra, U.S., Uebing, A., Harries, C., Goktekin, O., Gibbs, J.S.R., Gatzoulis, M.A. 2006. Presentation, survival prospects, and predictors of death in Eisenmenger syndrome: a combined retrospective and case-control study. *Eur Heart J*; 27(14):1737–42.
- Dimopoulos, K., Diller, G.P., Opotowsky, A.R., D'Alto, M., Gu, H., Giannakoulas, G., Budts, W., Broberg, C.S., Veldtman, G., Swan, L., Beghetti, M., Gatzoulis, M.A. 2018. Definition and Management of segmental pulmonary hypertension. *J Am Heart Assoc*;7:e008587.
- Engelfriet, P.M., Duffels, M.G., Moller, T., Boersma, E., Tijssen, J.G., Thaulow, E., Gatzoulis, M.A., Mulder, B.J. 2007. Pulmonary arterial hypertension in adults born with a heart septal defect: the Euro Heart Survey on adult congenital heart disease. *Heart*;93:682 687.
- Fang, Y.H., Piao, L., Hong, Z., Toth, P.T., Marsboom, G., Bache-Wiig, P., Rehman, J., Archer, S.L. 2012. Therapeutic inhibition of fatty acid oxidation in right ventricular hypertrophy: exploiting Randle's cycle. *J Mol Med (Berl)*;90:31-43.
- Fernandes, S.M., Lui, G.K., Long, J., Lin, A., Rogers, I.S., Sillman, C., Romfh, A., Dade, T.C., Dong, E., Haeffele, C., Scribner, C., Major, M., McElhinney, D.B. 2021. Predicting 10-year mortality in adults with congenital heart disease. *Int J Cardiol Congenital Heart Disease*; 2: 100057.
- Fijalkowska, A., Kuryzuna, M., Torbicki, A., Szewczyk, G., Florczyk, M., Pruzsczyk, P., Szturmowicz, M. 2006. Serum N-terminal brain natriuretic peptide as a prognostic parameter in patients with pulmonary hypertension. *Chest*; 129(5):1313-1321.
- Fine, N.M., Chen, L., Bastiansen, P.M., Frantz, R.P., Pellikka, P.A., Oh, J.K., Kane, G.C. 2013. Outcome prediction by quantitative right ventricular function assessment in 575 subjects evaluated for pulmonary hypertension. *Circ*



*Cardiovasc Imaging*;6: 711 – 721.

- Freed, B.H., Gomberg-Maitland, M., Chandra, S., Mor-Avi, V., Rich, S., Archer, S.L., Jamison, E.B., Lang, R.M., Patel, A.R. 2012. Late gadolinium enhancement cardiovascular magnetic resonance predicts clinical worsening in patients with pulmonary hypertension. *J Cardiovasc Magn Reson*;14:11.
- Fritz, J.S., Blair, C., Oudiz, R.J., Dufton, C., Olschewski, H., Despain, D., Gillies, H., Kawut, S.M. 2013. Baseline and follow-up 6-min walk distance and brain natriuretic peptide predict 2-year mortality in pulmonary arterial hypertension. *Chest*;143: 315 – 323.
- Galie, N., Rubin, L.J., Hoeper, M.M., Jansa, P., Al-Hiti, A., Meyer, G.M.B., Chiossi, E., Kusic-Pajic, A., Simonneau, G. 2008. Treatment of patients with mildly symptomatic pulmonary arterial hypertension with bosentan (EARLY study): a double-blind, randomised controlled trial. *Lancet*; 371: 2093–2100.
- Galie, N., Hoeper, M.M., Humbert, M., Torbicki, A., Vachiery, J.L., Barbera, J.A., Beghetti, M., Corris, P., Gaine, S., Gibss, J.S., Gomez-Sanches, M.A., Jondeau, G., Klepetko, W., Opitz, C., Peacock, A., Rubin, L., Zellweger, M., Simonneau, G. 2009. Guidelines for the diagnosis and treatment of pulmonary hypertension: the Task Force for the Diagnosis and Treatment of Pulmonary Hypertension of the European Society of Cardiology (ESC) and the European Respiratory Society (ERS), endorsed by the International Society of Heart and Lung Transplantation (ISHLT). *Eur Heart J*; 30: 2493–2537.
- Galie, N., Humbert, M., Vachiery, J.L., Gibss, S., Lang, I., Torbicki, A., Simonneau, G., Peacock, A., Noordegraaf, A.V., Beghetti, M., Ghofrani, A., Gomez-Sanches, M.A., Hansmann, G., Klepetko, W., Lancellotti, P., Matucci, M., McDonagh, T., Pierard, L.A., Trindade, P.T., Zompatori, M., Hoeper, M. 2015. 2015 ESC/ERS Guidelines for the diagnosis and treatment of pulmonary hypertension: The Joint Task Force for the Diagnosis and Treatment of Pulmonary Hypertension of the European Society of Cardiology (ESC) and the European Respiratory Society (ERS). *Eur Respir J*; 46: 903–975.
- Garson, A., Nihill, M.R., McNamara, D.G., Cooley, D.A. 1979. Status of the adult and adolescent after repair of tetralogy of fallot. *Circulation*; 59:6.
- Ghio, S., Klfersy, C., Magrini, G., D'Armini, A.M., Scelsi, L., Rainieri, C., Pasotti, M., Serio, A., Campana, C., Vigano, M. 2010. Prognostic relevance of the echocardiographic assessment of right ventricular function in patients with idiopathic pulmonary arterial hypertension. *Int J Cardiol*; 140: 272–278.
- Grunig, E., Tiede, H., Enyimayew, E.O., Ehlken, N., Seyfarth, H.J., Bossone, E., D'Andrea, A., Naeije, R., Olschewski, H., Ulrich, S., Nagel, C., Halank, M., Fischer, C. 2013. Assessment and prognostic relevance of right ventricular contractile reserve in patients with severe pulmonary hypertension. *Circulation*;128:2005 – 2015.
- Hjalmarsson, C., Radegran, G., Kylhammar D., Rundqvist, B., Multing, J., Nisell,



- M.D., Kjellstrom, B. 2018. Impact of Age and Comorbidity on Risk Stratification in Idiopathic Pulmonary Arterial Hypertension. *Eur Respir J*; 51(5): 1702310.
- Hoeper, M.M., Pletz, M.W., Golpon, H., Welte, T. 2007. Prognostic value of blood gas analyses in patients with idiopathic pulmonary arterial hypertension. *Eur Respir J*; 29:944 – 950.
- Howard, L.S. 2011. Prognostic factors in pulmonary arterial hypertension: assessing the course of the disease. *Eur Respir Rev*; 20: 122, 236–242.
- Jacobs, W., van de Veerdonk, M.C., Trip, P., de Man, F., Heymans, M.W., Marcus, J.T., Kawut, S.M., Bogaard, H-J., Boonstra, A., Noordegraaf, A.V. 2014. The right ventricle explains sex differences in survival in idiopathic pulmonary arterial hypertension. *Chest*; 145:1230-6.
- Kanemoto, N. 1987. Natural history of pulmonary hemodynamics in primary pulmonary hypertension. *Am Heart J*;114(2): 407e13.
- Khirfan, G., Naal, T., Abuhalimeh, B., Newman, J., Heresi, G.A., Dweik, R.A., Tonelli, A.R. 2018. Hypoxemia in patients with idiopathic or heritable pulmonary arterial hypertension. *PLoS ONE*; 13(1): e0191869.
- Krasuski, R.A., Hart, S.A., Smith, B., Wang, A., Harrison, J.K., Bashore, T.M. 2011. Association of anemia and long-term survival in patients with pulmonary hypertension. *Int J Cardiol*; 150:291-295.
- Lau, E.M.T., Giannoulatou, E., Celermajer, D.S, Humbert, M. 2017. Epidemiology and treatment of pulmonary arterial hypertension. *Nature Reviews Cardiology*,14: p. 603.
- Le, R.J., Fenstad, E.R., Maradit-Kremers, H., McCully, R.B., Frantz, R.P., McGoon, M.D., Kane, G.C. 2011. Syncope in adults with pulmonary arterial hypertension. *J Am Coll Cardiol*; 58: 863–867.
- Leuchte, H.H., El, N.M., Tuerpe, J.C., Hartmann, B., Baumgartner, R.A., Vogeser, M., Muehling, O., Behr, J. 2007. N-terminal pro-brain natriuretic peptide and renal insufficiency as predictors of mortality in pulmonary hypertension. *Chest*;131: 402 – 409.
- Li, X., Sun, X., Huang, Y., Wang, Y., Yang, X., Wang, J., Zhang, N., Gu, L., Zhang, M., Wang, Q. 2019. Simplified risk stratification for pulmonary arterial hypertension associated with connective tissue disease. *Clin Rheumatol*;38(12):3619-3626
- McGoon, M., Guetterman, D., Steen, V., Barst, R., McCrory, D.C., Fortin, T.A., Loyd, J.E. 2004. Screening, early detection, and diagnosis of pulmonary arterial hypertension: ACCP evidence-based clinical practice guidelines. *Chest*; 126:14S-34S
- McLaughlin, V.V., Archer, S.L., Badesch, D.B., Barst, R.J., Farber, H.W., Lindner, J.R., Mathier, M.A., McGoon, M.D., Park, M.H., Rosenson, R.S., Rubin, L.J., Tapson, V.F., Varga, J., Harrington, R.A., Anderson, J.L., Bates, E.R., Bridges, C.R., Eisenberg, M.J., Ferrari, V.A., Grines, C.L., Hlatky, M.A., Jacobs, A.K., Kaul, S., Lichtenberg, R.C., Moliterno, D.J., Mukherjee, D., Pohost, G.M., Schofield, R.S., Shubrooks, S.J., Stein, J.H., Tracy, C.M., Wesley, D.J. 2009. ACCF/AHA 2009 expert consensus document on pulmonary hypertension a report of the American College of Cardiology



Foundation Task Force on Expert Consensus Documents and the American Heart Association developed in collaboration with the American College of Chest Physicians; American Thoracic Society, Inc.; and the Pulmonary Hypertension Association. *J Am Coll Cardiol*;53:1573-619.

- Miyamoto, S., Nagaya, N., Satoh, T., Kyotani, S., Sakamaki, F., Fujita, M., Nakanishi, N., Miyatake, K. 2000. Clinical correlates and prognostic significance of six-minute walk test in patients with primary pulmonary hypertension. Comparison with cardiopulmonary exercise testing. *Am J Respir Crit Care Med*; 161: 487–492.
- Montani, D., Gunther, S., Dorfmuller, P., Perros, F., Girerd, B., Garcia, G., Jais, X., Savale, L., Artaud-Macari, E., Price, L.C., Humbert, M., Simonneau, G., Sitbon, O. 2013. Pulmonary arterial hypertension. *Orphanet J of Rare Diseases*;8:97.
- Naeije, R., Huez, S. 2007. Right ventricular function in pulmonary hypertension: physiological concepts. *Eur Heart J*; 9: H5-H9.
- Naidu, P., Grigg, L., Zentner, D. 2017. Mortality in adults with congenital heart disease. *Int J Cardiol*;245:125 – 130.
- Nakanishi, T., Baldwin, H.S., Fineman, J.R., Yamagishi, H. 2020. *Molecular mechanism of congenital heart disease and pulmonary hypertension*. Springer Open; Singapore.
- Nickel, N., Golpon, H., Greer, M., Knudsen, L., Olsson, K., Westerkamp, V., Welte, T., Hoeper, M.M. 2012. The prognostic impact of follow-up assessments in patients with idiopathic pulmonary arterial hypertension. *Eur Respir J*;39:589–596.
- Oliver, J.M., Gallego, P., Gonzales, A.E., Garcia-Hamilton, D., Avila, P., Yotti, R., Ferreira, I., Fernandez-Aviles, F. 2017. Risk factor for excess mortality in adults with congenital heart diseases. *Eur Heart J*;38: 1233-1241.
- Olsson, K.M., Nickel, N.P., Tongers, J., Hoeper, M.M. 2013. Atrial flutter and fibrillation in patients with pulmonary hypertension. *Int J Cardiol*;167:2300 – 2305.
- Ramani, G., Park, L.M., Baltimore. 2014. The impact of anemia on survival in pulmonary arterial hypertension. *Jour of Heart and Lung Transplantation*; S229: 620.
- Regitz-Zagrosek, V., Roos-Hesselink, J.W., Bauersachs, J., Blomstrom-Lundqvist, C., Cifkova, R., De Bonis, M., Iung, B., Johnson, M.R., Kintscher, U., Kranke, P., Lang, I.M., Morais, J., Pieper, P.G., Presbitero, P., Price, S., Rosano, G.M.C., Seeland, U., Simoncini, T., Swan, L., Warnes, C.A. 2018. ESC Guidelines for the management of cardiovascular diseases during pregnancy. *Eur Heart J*;39:3165-3241.
- Rexhaj, E., Bloch, J., Jayet, P.Y., Rimoldi, S.F., Dessen, P., Mathieu, C., Tolsa, J-F., Nicod, P., Scherrer, U., Sartori, C. 2011. Fetal programming of pulmonary vascular dysfunction in mice: role of epigenetic mechanisms. *Am J Physiol Heart Circ Physiol*;301:H247-52.
- Rich, J.D., Archer, S.L., Rich, S. 2013. Noninvasive cardiac output measurements in patients with pulmonary hypertension. *Eur Respir J*;42:125–133.
- Shapiro, S., Traiger, G.L., Turner, M., McGoon, M.D., Wason, P., Barst, R.J. 2012.



Sex differences in the diagnosis, treatment, and outcome of patients with pulmonary arterial hypertension enrolled in the registry to evaluate early and long-term pulmonary arterial hypertension disease management. *Chest*;141:363-73.

- Schuuring, M.J., van Riel, A.C.M.J., Vis, J.C., Duffels, M.G., van Dijk, A.P.J., de Bruin-Bon, R.H.A.C.M., Zwinderman, A.H., Mulder, B.J.M. Bouma, B.J. 2014. New predictors of mortality in adults with congenital heart disease and pulmonary hypertension: Midterm outcome of a prospective study. *Int J Cardiol*; 181:270–6.
- Simonneau, G., Montani, D., Celermajer, D.S., Denton, C.P., Gatzoulis, M.A., Krowka, M., Williams, P.G., Souza, R. 2019. Haemodynamic definitions and updated clinical classification of pulmonary hypertension. *Eur Respir J*;53:1801913.
- Sitbon, O., Humbert, M., Nunes, H., Parent, F., Garcia, G., Herve, P., Rainisio, M., Simonneau, G. 2002. Long-term intravenous epoprostenol infusion in primary pulmonary hypertension: prognostic factors and survival. *J Am Coll Cardiol*; 40: 780–788.
- Smith, B.C., Dobson, G., Dawson, D., Charalampopoulos, A., Grapsa, J., Nihoyannopoulos, P. 2014. Three-dimensional speckle tracking of the right ventricle: toward optimal quantification of right ventricular dysfunction in pulmonary hypertension. *J Am Coll Cardiol*;64:41–51.
- Steiner, J., Wu, W.-C., Jankowich, M., Maron, B.A., Sharma, S., Choudhary, G. 2015. Echocardiographic Predictors of Mortality in Patients with Pulmonary Hypertension and Cardiopulmonary Comorbidities. *PLoS ONE*; 10(3):e0119277.
- Swift, A.J., Rajaram, S., Campbell, M.J., Hurdman, J., Thomas, S., Capener, D., Elliot, C., Condliffe, R., Wild, J.M., Kiely, D.G. 2014. Prognostic value of cardiovascular magnetic resonance imaging measurements corrected for age and sex in idiopathic pulmonary arterial hypertension. *Circ Cardiovasc Imaging*;7:100 – 106.
- Taichman, D.B., McGoon, M.D., Harhay, M.O., Archer-Chicko, C., Sager, J.S., Murugappan, M., Chakinali, M.M., Palevsky, H.I., Gallop, R. 2009. Wide variation in clinician's assessment of New York Heart Association/World Health Organization functional class in patients with pulmonary arterial hypertension. *Mayo Clin Proc*;84:586 – 592.
- Thenappan, T., Shah, S.J., Rich, S., Tian, L., Archer, S.L., Gomberg-Maitland, M. 2010. Survival in pulmonary arterial hypertension: a reappraisal of the NIH risk stratification equation. *Eur Respir J*; 35: 1079–1087.
- Thenappan, T., Mark, L.O., John, J.R., Stephen, L.A. 2018. Pulmonary arterial hypertension: pathogenesis and clinical management. *BMJ*;360:j5492.
- Tonelli, A.R., Arelli, V., Minai, O.A., Newman, J., Bair, N., Heresi, G.A., Dweik, R.A. 2013. Causes and circumstances of death in pulmonary arterial hypertension. *Am J Respir Crit Care Med*;188:365–369.
- Van Riel, A.C., Schuuring, M.J., van Hessen, I.D., Zwinderman, A.H., Cozijnsen, L., Reichert, C.L., Hoornje, J.C., Wagenaar, L.J., Post, M.C., van Dijk, A.P., Hoendermis, E.S., Mulder, B.J., Bouma, B.J. 2014. Contemporary



prevalence of pulmonary arterial hypertension in adult congenital heart disease following the updated clinical classification. *Int J Cardiol*;174:299–305.

- Vijarnsorn, C., Durongpisitkul, K., Chungsomprasong, P., Bositthipichet, D., Ketsara, S., Titaram, Y., Chanthong, P., Kanjanauthai, S., Soongswang, J. 2018. Contemporary survival of patients with pulmonary arterial hypertension and congenital systemic to pulmonary shunts. *PLoS ONE*;13(4): e0195092.
- Vonk Noordegraaf, A. and Galie, N. 2011. The role of the right ventricle in pulmonary arterial hypertension. *Eur Respir Rev*;20:243–253.
- Walker, A.M., Langleben, D., Korelitz, J.J., Rich, S., Rubin, L.J., Strom, B.L., Gonin, R., Keast, S., Badesch, D., Barst, R.J., Bourge, R.C., Channick, R., Frost, A., Gaine, S., McGoon, M., McLaughlin, V., Murali, S., Oudiz, R.J., Robbins, I.M., Tapson, V., Abenhaim, L., Constatine, G. 2006. Temporar trends and drug exposures in pulmonary hypertension: an American experience. *American Heart Journal*;152(3):521–526.
- Wang, L.Y., Lee, K.T., Lin, C.P., Hsu, L.A., Wang, C.L., Hsu, T.S., Ho, W.J. 2017. Long-Term Survival of Patients with Pulmonary Arterial Hypertension at a Single Center in Taiwan. *Acta Cardiologica Sinica*, 33(5): p. 498-509.
- Wensel, R., Francis, D.P., Meyer, F.J., Opitz, C.F., Bruch, L., Halank, M., Winkler, J., Seyfarth, H.J., Glaser, S., Blumberg, F., Obst, A., Dandel, M., Hetzer, R., Ewert, R. 2013. Incremental prognostic value of cardiopulmonary exercise testing and resting haemodynamics in pulmonary arterial hypertension. *Int J Cardiol*;167:1193–1198.
- Yong, G., Khairy, P., De Guise, P., Dore, A., Marcotte, F., Mercier, L-A., Noble, S., Ibrahim, R. 2009. Pulmonary arterial hypertension in patients with transcatheter closure of secundum atrial septal defects: a longitudinal study. *Cardiovasc Intervent*; 2: 455-462.