**Appendix F. Study Characteristics Tables (KQ 1 and KQ 2)**

**Table F-1. Study characteristics table for KQ 1**

| **Study** | **Population**  **Total N** | **Test Measures** | **Study Objectives** | **Quality** |
| --- | --- | --- | --- | --- |
| Ajami, 20111 | Children and young adults with congenital heart disease referred for RHC  N=20 | TRV/VTIRVOT | Accuracy of TRV/VTIRVOT for diagnosing elevated PVR | Good |
| Allanore, 20082 | SSc patients with echocardiography sPAP<40 mmHg and no NYHA III/IV symptoms  N=101 | NT-proBNP, plasma  sPAP | Screening for prospective development of PAH (predicting development of PAH in at-risk population) | Good |
| Arcasoy 20033 | Patients with advanced lung disease undergoing evaluation for lung transplantation  N=374 | sPAP  RAP | Estimate performance characteristics of echocardiography compared with RHC in determining sPAP and diagnosing PH | Good |
| Bogdan, 19984 | PAH patients and controls  N=19 | cGMP, urine | Test association of PAH with urine cGMP | Poor |
| Bonderman, 20115 | Referred for evaluation of suspected PAH;  more than half had NYHA III/IV symptoms  N=372 | NT-proBNP  sPAP  RA size  RV size  TAPSE | Diagnostic accuracy for distinguishing PAH from secondary PH  Precision/calibration of echocardiographic measures | Good |
| Cavagna, 20106 | SSc patients; symptoms not described  N=135 | BNP  NT-proBNP | Screening for PAH  Discrimination between PAH or not Reference standard based on echocardiography sPAP screening with RHC verification of positives | Good |
| Cevik, 20127 | Children with pulmonary hypertension, with and without congenital heart disease  N=70 | RIMP/MPI/Tei index  mPAP  S’  TAPSE  TRV/VTIRVOT | Evaluation of RV function using transthoracic echocardiography | Fair |
| Ciurzynski, 20118 | SSc patients. Patients with signs or symptoms of heart or lung disease excluded  N=71 | Transtricuspid gradient rest/exercise | Association with diagnosis of PAH | Good |
| Colle, 20039 | Liver transplant candidates  N=165 | sPAP | Screening for portopulmonary hypertension | Good |
| Condliffe, 201110 | SSc patients with suspected PAH; symptoms not described  N=89 | Tricuspid gradient | Discrimination between PAH or not  Reference standard=RHC | Fair |
| Dahiya, 201011 | Referred for evaluation of suspected PH; all patients had dyspnea  N=114 | TRV/VTIRVOT | Diagnostic accuracy, calibration, and precision of echocardiography estimation of elevated PVR | Good |
| Denton, 199712 | SSc patients suspected of PAH, most due to reduced DLCO  N=93 | RV size  sPAP | Diagnostic accuracy, discrimination of echocardiography for diagnosing PAH | Fair |
| Farber, 201113 | Patients with PAH  N=1883 | sPAP  RAP | Accuracy of echocardiography for sPAP and RAP | Fair |
| Fisher, 200914 | Patients undergoing RHC for known or suspected PAH; symptoms not described  N=65 | sPAP  Transtricuspid gradient | Precision/calibration of echocardiography for mPAP, sPAP compared with RHC | Good |
| Fitzgerald, 201215 | Sickle cell disease with TRV ≥2.5 m/s and RHC  N=75 | TRV  mPAP | Comparison of TRV measurement to RHC for diagnosing PH | Poor |
| Fonseca, 201216 | Sickle cell disease; symptoms not described  N=80 | TRV  Uric acid | Screening for PAH  Echocardiography screening of TRV with RHC verification of positives | Fair |
| Frea, 201117 | SSc patients with no signs or symptoms of PAH  N=76 | NT-proBNP  FAC  RIMP/MPI/Tei index  RV size  TRV/VTIRVOT TAPSE | Screening for prospective development of PAH  (Predicting development of PAH in at-risk population) | Fair |
| Fukuda, 201118 | Patients with known PH  N=67 | FAC  TAPSE  RIMP/MPI/Tei index  sPAP | Correlation between echocardiography and RHC hemodynamics in patient with elevated mPAP | Fair |
| Ghio, 200419 | HIV and confirmed PAH. Controls with HIV and no known cardiac or pulmonary disease  N=93 | NT-proBNP | Diagnostic accuracy for NT-proBNP for discriminating HIV-positive PAH patients from HIV-positive controls | Fair |
| Gialafos, 200820 | SSc patients. Some were symptomatic  N=106 | NT-proBNP  RIMP/MPI/Tei index | Association with diagnosis of PAH | Fair |
| Hachulla, 200521 | SSc patients; some symptomatic  N=599 | TRV | Screening for PAH in at-risk population | Poor |
| Hammerstingl, 201222 | Patients with PH undergoing RHC and transthoracic echocardiography  N=155 | sPAP  mPAP | Diagnosis of PH and differentiating between pre- and postcapillary PH | Fair |
| Hsu, 200823 | SSc patients with dyspnea or other clinical features suggestive of PAH  N=49 | sPAP | Diagnostic accuracy for diagnosing PAH | Good |
| Hua, 200924 | Liver transplant candidates  N=105 | sPAP | Diagnostic accuracy for portopulmonary hypertension | Good |
| Jansa, 201225 | SSc patients some with dyspnea  N=203 | TRV | Screening for PAH in at-risk population | Fair |
| Kovacs, 201026 | Patients with CVD some with symptoms  N=52 | sPAP rest and exercise | Screening for PAH in at-risk population | Good |
| Lindqvist, 201127 | Patients with PH undergoing RHC  N=30 | TRV/VTIRVOT | Accuracy for diagnosis of elevated PVR  Precision/calibration of echocardiography estimate of PVR | Fair |
| Low, 201128 | Referred for evaluation of suspected or definite PAH, most with symptoms  N=200 | Transtricuspid gradient | Diagnostic accuracy for diagnosing PAH | Poor |
| Machado, 200629 | Sickle cell disease  N=416 | NT-proBNP | Association between biomarker and hemodynamic measures. Diagnosis based on echocardiography screen with partial verification by RHC of some test positives. | Poor |
| McLean, 200730 | Referred for echocardiography with adequate TR jet on Doppler, nearly all with symptoms  N=108 | RV end-diastolic diameter (RVD)  time to peak (RV tricuspid annular motion by TDI, time from beginning of IC to first systolic myocardial peak) | Correlation between echocardiography RVD/time to peak and RHC PASP | Poor |
| Mourani, 200831 | Children under 2 years of age undergoing RHC for chronic lung disease  N=25 | RA size  RV size  Transtricuspid gradient | Asses echocardiography feasibility, calibration for estimating hemodynamics, and accuracy for diagnosis of PAH | Fair |
| Mukerjee, 200432 | SSc patients with suspected PAH, symptoms of exercise limitation and reduced DLCO  N=137 | sPAP | Accuracy of echocardiography sPAP at different thresholds for diagnosis of PAH | Fair |
| Murata, 199733 | SSc patients. Symptoms not described, but most had reduced DLCO  N=135 | sPAP | Precision/calibration of echocardiography for estimating invasive pulmonary hemodynamics | Fair |
| Nakayama, 199834 | Patients with known, symptomatic CTEPH or PPH  N=35 | sPAP  mPAP | Accuracy of echocardiography for discrimination between CTEPH and PPH | Fair |
| Nogami, 200935 | Suspected pulmonary hypertension; all patients symptomatic  N=29 | sPAP | Precision/calibration of echocardiography for estimating invasive pulmonary hemodynamics | Good |
| Phung, 200936 | SSc patient referred with or without suspicion of PAH; 10% had NYHA III/IV symptoms  N=184 | sPAP | Accuracy of echocardiography sPAP for diagnosing PAH | Good |
| Pilatis, 200037 | Liver transplant candidates  N=55 | RV size  sPAP | Accuracy of echocardiography for diagnosing portopulmonary hypertension | Fair |
| Rajagopalan, 200838  Rajagopalan, 200739 | Known pulmonary hypertension  N=52 | TRV/VTIRVOT  sPAP  S’ | Accuracy of echocardiography for estimating PVR in PH patients  Calibration/precision of echocardiography for estimating RHC hemodynamics | Fair |
| Rajaram, 201240 | Connective tissue disease patients with suspected PH  N=81 | sPAP  mPAP  Pericardial effusion | Comparison of magnetic resonance imaging, computed tomography, and echocardiography for diagnosing PAH | Fair |
| Rich, 201141 | Patients with both RHC and Doppler echo  N=183 | sPAP | Calibration/precision of echocardiography for estimating RHC hemodynamics | Good |
| Roeleveld, 200542 | Known PH  N=47 | sPAP | Calibration/precision of echocardiography for estimating RHC hemodynamics | Fair |
| Roule, 201043 | Known PH  N=37 | TRV  TRV/VTIRVOT | Calibration/precision for estimating RHC hemodynamics at elevated PA pressures  Accuracy for diagnosing elevated PVR in PH patients | Good |
| Ruan, 200744 | Known PAH and healthy controls  N=180 | FAC  RV size  sPAP | Diagnostic accuracy of echocardiography for discriminating PAH and control patients | Fair |
| Ruiz-Irastorza, 201245 | Systemic lupus erythematosus patients with or without suspicion of PAH  N=245 | sPAP  mPAP | Prevalence of and strategy for diagnosing PH in patients with lupus | Fair |
| Sanli, 201246 | Congenital heart disease with and without known PAH  N=70 | RV size  mPAP  Nitric oxide  RIMP/MPI/Tei index  TAPSE | Relationship between biomarkers and hemodynamic measurements | Fair |
| Selby, 201247 | Patients with HIV infection with or without suspicion of PAH  N=129 | sPAP | Comparison of sPAP measured by echocardiography versus RHC | Fair |
| Selimovic, 200748 | Patients with suspected pulmonary vascular disease. 37/42 NYHA III/IV  N=42 | sPAP  mPAP | Calibration/precision of echocardiography for estimating RHC hemodynamics | Good |
| Simeoni, 200849 | Known SSc-associated PAH and controls with SSc but no PAH  N=20 | NT-proBNP | Diagnostic accuracy of NT-proBNP for discriminating PAH and control patients | Poor |
| Steen, 200850 | SSc patients with suspected PAH based on symptoms or signs  N=54 | sPAP rest/exercise | Accuracy of rest/exercise echocardiography to diagnose PAH | Fair |
| Takatsuki, 201251 | Children with idiopathic PAH  N=102 | S’  mPAP | Assessing disease severity and prognostic value with tissue Doppler imaging | Fair |
| Tei, 199652 | Known PPH and health controls  N=53 | RIMP/MPI/Tei index | Association of Tei index with PPH versus normal controls | Poor |
| Thakkar, 201253 | SSc patients with known PAH, high risk for PAH, interstitial lung disease, or no cardiopulmonary disease  N=94 | sPAP  NT-proBNP | NT-proBNP as a replacement for transthoracic echocardiography in screening for SSc-related PAH | Fair |
| Tian, 201154 | Suspected PH based on symptoms  N=42 | sPAP  mPAP | Calibration/precision of echocardiography for estimating RHC hemodynamics | Fair |
| Torregrosa, 200155 | Liver transplant candidates  N=94 | sPAP | Accuracy for diagnosing portopulmonary hypertension | Fair |
| Toyono, 200856 | Children with VSD and severe PH  N=24 | BNP | Correlation between BNP levels and invasive PVR | Good |
| Tutar, 199957 | Children with left-to-right shunt and health controls  N=23 | Endothelin-1, plasma | Association of endothelin-1 levels and pulmonary hypertension | Fair |
| Vlahos, 200758 | Known or suspected pulmonary hypertension  N=12 | TRV/VTIRVOT | Accuracy of echocardiography for diagnosing elevated PVR | Poor |
| Vonk, 200759 | Connective tissue diseases. One-third NYHA III/IV  N=98 | RIMP/MPI/Tei index  sPAP | Accuracy for diagnosis of PAH or not | Fair |
| Willens, 200860 | Patients with known PH and elevated sPAP and controls with CHF and elevated sPAP  N=47 | sPAP | Association of sPAP with PH versus CHF | Fair |
| Williams, 200661 | SSc patients with PAH and controls with SSc but without PAH  N=109 | NT-proBNP | Accuracy for diagnosis of PAH | Fair |

Abbreviations: BNP=brain natriuretic peptide; CHF=congestive heart failure; CTEPH=chronic thromboembolic pulmonary hypertension; CVD=collagen vascular disease; DLCO=diffusion capacity of the lung for carbon monoxide; FAC=fractional area change; mPAP=mean pulmonary artery pressure; MPI=myocardial performance index; NT-proBNP=N-terminal pro-B-type natriuretic peptide; NYHA=New York Heart Association; PAH=pulmonary arterial hypertension; PH=pulmonary hypertension; PPH=primary pulmonary hypertension; PVR=pulmonary vascular resistance; RA=right atrium; RHC=right heart catheterization; RIMP=right index of myocardial performance; RV=right ventricle; S’=tricuspid lateral annular systolic velocity; sPAP=systolic pulmonary artery pressure; SSc=systemic sclerosis; TAPSE=tricuspid annular plane systolic excursion; TDI=tissue Doppler imaging; TRV=tricuspid regurgitant jet velocity; VSD=ventricular septal defect; VTIRVOT=velocity-time integral of right ventricular outflow tract