**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 RHCN=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, plasmasPAP | 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 | sPAPRAP | 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 symptomsN=372 | NT-proBNPsPAPRA sizeRV sizeTAPSE | Diagnostic accuracy for distinguishing PAH from secondary PHPrecision/calibration of echocardiographic measures | Good |
| Cavagna, 20106 | SSc patients; symptoms not describedN=135 | BNPNT-proBNP | Screening for PAHDiscrimination 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 diseaseN=70 | RIMP/MPI/Tei indexmPAPS’TAPSETRV/VTIRVOT | Evaluation of RV function using transthoracic echocardiography | Fair |
| Ciurzynski, 20118 | SSc patients. Patients with signs or symptoms of heart or lung disease excludedN=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 notReference 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 sizesPAP | Diagnostic accuracy, discrimination of echocardiography for diagnosing PAH | Fair |
| Farber, 201113 | Patients with PAH N=1883 | sPAPRAP | Accuracy of echocardiography for sPAP and RAP | Fair |
| Fisher, 200914 | Patients undergoing RHC for known or suspected PAH; symptoms not describedN=65 | sPAPTranstricuspid gradient | Precision/calibration of echocardiography for mPAP, sPAP compared with RHC | Good |
| Fitzgerald, 201215 | Sickle cell disease with TRV ≥2.5 m/s and RHCN=75 | TRVmPAP | Comparison of TRV measurement to RHC for diagnosing PH | Poor |
| Fonseca, 201216 | Sickle cell disease; symptoms not describedN=80 | TRVUric acid | Screening for PAHEchocardiography screening of TRV with RHC verification of positives | Fair |
| Frea, 201117 | SSc patients with no signs or symptoms of PAH N=76 | NT-proBNPFACRIMP/MPI/Tei indexRV sizeTRV/VTIRVOT TAPSE | Screening for prospective development of PAH(Predicting development of PAH in at-risk population) | Fair |
| Fukuda, 201118 | Patients with known PHN=67 | FACTAPSERIMP/MPI/Tei indexsPAP | 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 diseaseN=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 symptomaticN=106 | NT-proBNPRIMP/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 echocardiographyN=155 | sPAPmPAP | Diagnosis of PH and differentiating between pre- and postcapillary PH | Fair |
| Hsu, 200823 | SSc patients with dyspnea or other clinical features suggestive of PAHN=49 | sPAP | Diagnostic accuracy for diagnosing PAH | Good |
| Hua, 200924 | Liver transplant candidatesN=105 | sPAP | Diagnostic accuracy for portopulmonary hypertension | Good |
| Jansa, 201225 | SSc patients some with dyspneaN=203 | TRV | Screening for PAH in at-risk population | Fair |
| Kovacs, 201026 | Patients with CVD some with symptomsN=52 | sPAP rest and exercise | Screening for PAH in at-risk population | Good |
| Lindqvist, 201127 | Patients with PH undergoing RHCN=30 | TRV/VTIRVOT | Accuracy for diagnosis of elevated PVRPrecision/calibration of echocardiography estimate of PVR | Fair |
| Low, 201128 | Referred for evaluation of suspected or definite PAH, most with symptomsN=200 | Transtricuspid gradient | Diagnostic accuracy for diagnosing PAH | Poor |
| Machado, 200629 | Sickle cell diseaseN=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 symptomsN=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 diseaseN=25 | RA sizeRV sizeTranstricuspid 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 DLCON=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 DLCON=135 | sPAP | Precision/calibration of echocardiography for estimating invasive pulmonary hemodynamics | Fair |
| Nakayama, 199834 | Patients with known, symptomatic CTEPH or PPHN=35 | sPAPmPAP | Accuracy of echocardiography for discrimination between CTEPH and PPH | Fair |
| Nogami, 200935 | Suspected pulmonary hypertension; all patients symptomaticN=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 symptomsN=184 | sPAP | Accuracy of echocardiography sPAP for diagnosing PAH | Good |
| Pilatis, 200037 | Liver transplant candidatesN=55 | RV sizesPAP | Accuracy of echocardiography for diagnosing portopulmonary hypertension | Fair |
| Rajagopalan, 200838Rajagopalan, 200739 | Known pulmonary hypertensionN=52 | TRV/VTIRVOTsPAPS’ | Accuracy of echocardiography for estimating PVR in PH patientsCalibration/precision of echocardiography for estimating RHC hemodynamics | Fair |
| Rajaram, 201240 | Connective tissue disease patients with suspected PHN=81 | sPAPmPAPPericardial effusion | Comparison of magnetic resonance imaging, computed tomography, and echocardiography for diagnosing PAH | Fair |
| Rich, 201141 | Patients with both RHC and Doppler echoN=183 | sPAP | Calibration/precision of echocardiography for estimating RHC hemodynamics | Good |
| Roeleveld, 200542 | Known PHN=47 | sPAP | Calibration/precision of echocardiography for estimating RHC hemodynamics | Fair |
| Roule, 201043 | Known PHN=37 | TRVTRV/VTIRVOT | Calibration/precision for estimating RHC hemodynamics at elevated PA pressuresAccuracy for diagnosing elevated PVR in PH patients | Good |
| Ruan, 200744 | Known PAH and healthy controlsN=180 | FACRV sizesPAP | Diagnostic accuracy of echocardiography for discriminating PAH and control patients | Fair |
| Ruiz-Irastorza, 201245 | Systemic lupus erythematosus patients with or without suspicion of PAHN=245 | sPAPmPAP | Prevalence of and strategy for diagnosing PH in patients with lupus | Fair |
| Sanli, 201246 | Congenital heart disease with and without known PAHN=70 | RV sizemPAPNitric oxideRIMP/MPI/Tei indexTAPSE | Relationship between biomarkers and hemodynamic measurements | Fair |
| Selby, 201247 | Patients with HIV infection with or without suspicion of PAHN=129 | sPAP | Comparison of sPAP measured by echocardiography versus RHC | Fair |
| Selimovic, 200748 | Patients with suspected pulmonary vascular disease. 37/42 NYHA III/IVN=42 | sPAPmPAP | Calibration/precision of echocardiography for estimating RHC hemodynamics | Good |
| Simeoni, 200849 | Known SSc-associated PAH and controls with SSc but no PAHN=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 signsN=54 | sPAP rest/exercise | Accuracy of rest/exercise echocardiography to diagnose PAH | Fair |
| Takatsuki, 201251 | Children with idiopathic PAHN=102 | S’mPAP | Assessing disease severity and prognostic value with tissue Doppler imaging | Fair |
| Tei, 199652 | Known PPH and health controlsN=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 diseaseN=94 | sPAPNT-proBNP | NT-proBNP as a replacement for transthoracic echocardiography in screening for SSc-related PAH | Fair |
| Tian, 201154 | Suspected PH based on symptomsN=42 | sPAPmPAP | Calibration/precision of echocardiography for estimating RHC hemodynamics | Fair |
| Torregrosa, 200155 | Liver transplant candidatesN=94 | sPAP | Accuracy for diagnosing portopulmonary hypertension | Fair |
| Toyono, 200856 | Children with VSD and severe PHN=24 | BNP | Correlation between BNP levels and invasive PVR | Good |
| Tutar, 199957 | Children with left-to-right shunt and health controlsN=23 | Endothelin-1, plasma | Association of endothelin-1 levels and pulmonary hypertension | Fair |
| Vlahos, 200758 | Known or suspected pulmonary hypertensionN=12 | TRV/VTIRVOT | Accuracy of echocardiography for diagnosing elevated PVR | Poor |
| Vonk, 200759 | Connective tissue diseases. One-third NYHA III/IVN=98 | RIMP/MPI/Tei indexsPAP | Accuracy for diagnosis of PAH or not | Fair |
| Willens, 200860 | Patients with known PH and elevated sPAP and controls with CHF and elevated sPAPN=47 | sPAP | Association of sPAP with PH versus CHF | Fair |
| Williams, 200661 | SSc patients with PAH and controls with SSc but without PAHN=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