**Table F-2. Study characteristics table for KQ 2**

| StudyLocation | PopulationTotal NFemale : Male | Index Test(s) | Comparator(s) | Result | Quality |
| --- | --- | --- | --- | --- | --- |
| Andreassen, 200662Europe | Adults with suspected chronic precapillary PHN=6142:19 | BNPBNP | Cardiac indexFunctional classRHC-mPAPRHC-PVRRHC-sPAPRAPMortality | CorrelationOdds ratio | Good |
| Badesch, 201263Badesch 201164US/Canada | Adults with PAHN=224156:68 | BNP |  | Mean change from baseline in response to treatment (ambrisentan) | Good |
| Barst, 199665US/Canada | Adults with PPH N=81 59:22 | mPAP |  | Change in mean from baseline in response to therapy (epoprostenol) | Good |
| Bendayan, 200266Asia | Adults and children with PAHN=2925:4 | Uric acid | 6MWD (absolute)Functional classMortalityRHC-CORHC-mPAP | Correlation | Good |
| Benza, 201067US | Adults with PAH N=2716 NR  | BNP >180BNP <50Pericardial effusion | Mortality | HR | Good |
| Bernus, 200968US | Children with PAHN=7842:26 | BNP | Cardiac index Peak TRVPCWPRHC-mPAPRHC-PVRRight atrial pressure RV sizeTRV | Correlation | Good |
| Bharani, 200769Asia | Adults and children with suspected or symptomatic PAH N=84:4 | sPAP |  | Change in mean in response to therapy (tadalafil) | Fair |
| Borges, 200670Europe | Chronic PAHN=3724:13  | RIMP/MPI/Tei indexTAPSETAPSERV size | 6MWD (absolute)RHC-PVR | CorrelationChange in mean in response to therapy (bosentan ± beraprost or iloprost) | Good |
| Brierre, 201071Europe | Adults with PAHN=7936:43 | mPAPmPAP ≥ 49Pericardial effusionRIMP/MPI/Tei indexRIMP/MPI/Tei index ≥0.98TAPSE | Mortality | HR | Good |
| Bustamante-Labarta, 200272South America | Adults with PPHN=2519:6  | RA size | Survival free from lung transplant | HR | Good |
| Campana, 200473Europe | Adults with pre-capillary PHN=2214:8  | BNPCardiac index FACBNPRV sizeTAPSE | mPAPRight atrial pressure RVEFTAPSE | CorrelationChanges in means in response to therapy (epoprostenol) | Good |
| Cella, 200974Europe | Adults with PAH associated with CTDN=1813:5 | RVSPNitric oxide | 6MWD (change) | CorrelationChange in mean over time in response to therapy (bosentan) | Good |
| Channick, 200175Badesch 200276US/Europe | Adults with PPH or PAH associated with sclerodermaN=3228:4 | mPAP |  | Change in mean from baseline in response to therapy (bosentan) | Good |
| Chin, 200777US | Epoprostenol-treated patients with pulmonary hypertensionN=2719:8 | BNP | 6MWD (absolute)PCWPRHC-CORHC-mPAPRHC-PVRRight atrial pressure  | Correlation | Good |
| Dimitroulas, 200778Europe | Adults with PAH associated with sclerodermaN=109:1 | BNP |  | Change in median over time in response to therapy (Bosentan) | Good |
| D’Alto, 201079Europe | Adults with PAH due to CHDN=3218:14 | BNP |  | Change in mean in response to therapy (bosentan + sildenafil) | Fair |
| Dyer, 200680US | Children with IPAHN=12NR | RIMP/MPI/Tei index | RHC-mPAP | Correlation | Fair |
| Elstein, 200481Asia | Adults and children with Gaucher diseaseN=4727:20 | BNP | Tricuspid insufficiency  | Correlation with stratified TI values | Good |
| Fahmy Elnoamany, 200782Africa | Adults with arterial PH with different cardiac pathologiesN=538:45  | Endothelin-1sPAP | RHC-sPAPRIMP/MPI/Tei IndexRVEFsPAPRHC-sPAP | Correlation | Fair |
| Feliciano, 200583Europe | Adults with severe PAHN=119:2 | RIMP/MPI/Tei index |  | Change in mean in response to therapy (bosentan or Iloprost) | Good |
| Fijalkowska, 200684Europe | Adults with PHN=5543:12 | BNPFACBNPBNPPericardial effusionRA sizeRIMP/MPI/Tei indexRV sizeTroponin T | 6MWD (absolute)Cardiac index Functional classPeak TRVPericardial effusionRHC-mPAPRHC-PVRRight atrial pressure RIMP/MPI/Tei IndexRV sizeTroponin Mortality | CorrelationHR | Good |
| Filusch, 201085Europe | Adults with PAHN=5533:22 | cTroponin ThsTroponin TBNP | MortalityWHO class | SensitivitySpecificityNPVPPV | Good |
| Forfia, 200686US | Adults with PHN=6352:11 | TAPSETAPSE | RHC-PVRMortality | CorrelationHR | Good |
| Friedberg, 200687US | Adults and children who had undergone RHCN=11248:64 | mPAPBNPsPAP | RHC-mPAPRHC-sPAP | Correlation | Good |
| Galie, 200888Oudiz 200989Shapiro 201290US/Europe/Mexico/ South America/ Australia/NZ | Adults with PAHN=201168:33  | BNP |  | Change in mean in response to therapy (ambrisentan) | Good |
| Gan, 200691Europe | Adults with PHN=3022:8 | BNP | 6MWD (absolute)Cardiac index RHC-mPAPRHC-PVRRight atrial pressure RVEF | Correlation | Good |
| Ghio, 201092Europe | Adults with IPAHN=5937:22 | FACPericardial effusionRIMP/MPI/Tei indexsPAPTAPSE | Mortality | HR | Good |
| Ghofrani, 200293Europe | Adults with severe precapillary PH. N=20 (36 tests)NR | BNPcGMP | RHC-PVRCyclic guanosine monophosphateRHC-PVR | Correlation | Fair |
| Goto, 201094Asia | Adults with PAHN=4634:12 | BNPsPAP | RHC-mPAPRHC-sPAP | Correlation | Good |
| Grapsa, 200795UK | Adults with PHN=9350:43  | RIMP/MPI/Tei index | Pericardial effusionRA sizeTRV | Correlation | Good |
| Grubstein, 200896Asia | Adults with PHN=3827:11 | sPAP | RHC-sPAP | Correlation | Fair |
| Haddad, 200997US | Adults with PAHN=5135:16 | mPAPsPAP | RHC-mPAPRHC-sPAP | Correlation | Good |
| Halank, 201198Europe | Adults with portopulmonary hypertensionN=149:5 | BNP | Median | Change in median over time in response to therapy (ambrisentan) | Fair |
| Hampole, 200999US | Adults with PHN=162126:36 | BNP | Mortality | HR | Good |
| Heresi, 2012100US | Adults with PAHN=6862:6 | cTnI (detectable vs. nondetectable) | BNPNYHA classRA size6MWD | Correlation | Good |
| Heresi, 2010101US | Adults with PPHN=4037:3 | BNP | 6MWD (absolute)Cardiac indexRHC-mPAPRHC-PVRRight atrial pressure Mortality | CorrelationHR | Good |
| Hinderliter, 1997102Other | Adults with PPHN=8159:22 | FACPericardial effusionRV sizesPAPRV sizeFACTRV | 6MWD (absolute)Cardiac index RHC-mPAPRight atrial pressure 6MWD (absolute)Cardiac index (CI)RHC-mPAPRight atrial pressure 6MWD (absolute)Cardiac index RHC-mPAPRight atrial pressureRHC-sPAP | CorrelationCorrelationCorrelationCorrelationChange in mean from baseline in response to therapy (epoprostenol) | Fair |
| Hiramoto, 2009103Asia | Adults with PAHN=1611:5 | BNP | Endothelin-1 | Changes in mean stratified by % change in ET-1 | Fair |
| Ho, 2009104Asia | Adults with PAHN=64:2 | RIMP/MPI/Tei IndexFACBNPsPAPRVEF |  | Changes in median in response to therapy (bosentan) | Good |
| Homma, 2001105US | Adults with PHN=85:3 | sPAP | RHC-sPAP | Correlation | Good |
| Jacobs, 2009106Europe | Adults with idiopathic PAHN=1613:3 | BNP |  | Change in mean in response to therapy | Fair |
| Kaya, 2012107NR | Adults and children with Eisenmenger syndromeN=2313:10 | RV sizeRA sizesPAPs-prime |  | Change in mean over time in response to therapy (bosentan) | Good |
| Keogh, 2011108Australia/NZ | Adults with PAHN=11289:23 | sPAP |  | Change in mean in response to therapy (monotherapy vs. combination therapy) | Fair |
| Knirsch, 2011109Europe | Children with heart diseaseN=103NR | BNP |  | Changes in mean in response to therapy (standardized protocol) | Good |
| Kopec, 2012110Europe | Adults with Eisenmenger syndromeN=74:3 | BNPET-1 |  | Change in median over time in response to therapy (bosentan) | Fair |
| Lammers, 2009111UK | Children with PHN=5018:32 | BNP | 6MWD (absolute)Functional class | Correlation | Good |
| Langleben, 1999112US/Canada | Patients with PPHN=18NR | Endothelin-1 |  | Change in mean in response to therapy | Good |
| Leuchte, 2005113Europe | Adults with PAHN=3018:12 | Change in BNPBNP | 6MWD (absolute)Cardiac index RHC-CORHC-mPAPRHC-PVRRight atrial pressure  | CorrelationChanges in mean levels over time (no specific therapy) | Good |
| Lorenzen, 2011114Europe | Adults with PAHN=7048:22 | BNPUric acid | Mortality | HR | Good |
| Machado, 200629US | Patients with sickle cell diseaseN=230138:92  | BNPBNP≥160, unadjusted≥160, adjustedlog10, adjustedlog10, unadjusted | 6MWD (absolute)mPAPPCWPRA sizeRHC-CORHC-dPAPRHC-PVRRHC-sPAPRV sizeTRVMortality | CorrelationHR | Poor |
| Machado, 2004115US | Patients with PAHN=1717:0 | Nitric oxidesPAP | mPAPRHC-sPAP | Correlation | Fair |
| Mahapatra, 2006116US | Adults with PHN=5441:13 | RIMP/MPI/Tei IndexRVSP | Mortality | HR | Fair |
| Mathai, 2011117US | Adults with known or suspected PAHN=5049:1 | FACPeak TRVPericardial effusionRA sizeTAPSE | Mortality | HR | Fair |
| Mauritz, 2011118Europe | Adults with PAHN=198149:49 | BNP |  | Baseline means only | Good |
| McLaughlin, 2010119Frantz, 2012120US/Europe | Adults with PHN=235191:44 | BNP |  | Median change from baseline in response to treatment (treprostinil) | Good |
| Michelakis, 2002121Canada | Adults with PHN=139:4 | cGMP |  | Acute change in mean levels after dose of various vasodilators (iNO, sildenafil, iNO + sildenafil) | Fair |
| Minniti, 2009122US | Adults with SCD and PHN=1410:4 | BNPTRV |  | Change in mean in response to therapy (ambrisentan) | Poor |
| Montani, 2007123Europe | Adults with PAHN=3321:12 | Endothelin-1 | Cardiac index RHC-PVRRight atrial pressure  | Correlation | Fair |
| Morishita, 2009124Asia | Adults and children with PAHN=76:1 | Pericardial effusionRA sizeRA sizeBNP | Functional classBNP | CorrelationChanges in mean in response to therapy (epoprostenol) | Good |
| Mukerjee, 2003125Europe | Adults with systemic sclerosisN=2321:2 | BNP | RHC-mPAPRHC-PVR | Correlation | Good |
| Nagaya, 2000126Asia | Patients with PPHN=6042:18 | ANPANPBNPBNP | PCWPRHC-CORHC-mPAPRight atrial pressure MortalityPCWPRHC-CORHC-mPAPRHC-mPAPRHC-PVRRight atrial pressure RV sizeMortality | CorrelationHRCorrelationHR | Good |
| Nakayama, 2007127Asia | Children with IPAHN=3115:16 | BNP |  | Change in mean in response to therapy (epoprostenol) | Fair |
| Nath, 2005128US | Adults with PPHN=2016:4 | Peak TRVRIMP/MPI/Tei IndexRV sizeRIMP/MPI/Tei IndexRV sizesPAPTRV | Functional class | CorrelationMean changes over time in response to therapy (epoprostenol) | Good |
| Nickel, 2012129NR | Adults with IPAHN=10985:24 | BNPUric acid | Mortality | HR | Fair |
| Nickel, 2008130Europe | Adults with IPAHN=7652:24 | BNPUric acid | Composite outcome (death or lung transplantation) | HR | Fair |
| Njaman, 2007131Asia | Adults with PHN=9077:13 | Uric acid | Mortality | HR stratified by uric acid levels | Good |
| Ogawa, 2012132Asia | Adults with pulmonary veno-occlusive or pulmonary capillary hemangiomatosisN=84:4 | BNP |  | Change in mean over time in response to therapy (epoprostenol) | Fair |
| Park, 2004133US | Adults with PAHN=2016:4 | sPAPBNP | Clinical event | Mean levels at baseline and over time stratified by patients with event vs patients without event | Fair |
| Pyxaras, 2011134Europe | Adults and children with PAHN=6036:24 | sPAPmPAP | RHC-sPAPRHC-mPAP | Correlation | Good |
| Raymond, 2002135Not reported/Unclear | Adults with PPHN=8159:22 | FACPeak TRVPericardial effusionRA size | MortalityComposite outcome (death or transplantation) | HR | Fair |
| Rhodes, 2011136Europe | Adults with IPAHN=13998:41 | BNP | 6MWD (absolute)Cardiac index PCWPRHC-PVRRight atrial pressure Mortality | CorrelationHR | Good |
| Sadushi-Kolici, 2012137NR | Adults with PHN=11177:39 | Pericardial effusion | Mortality | HR | Fair |
| Schumann, 2010138Europe | Adults with PHN=3617:19 | BNP | sPAP | BNP levels stratified by different levels of disease severity | Good |
| Sebbag, 2001139Canada | Adults and children with PPHN=1613:3 | RIMP/MPI/Tei IndexSPAP |  | Changes in mean in response to therapy (epoprostenol) | Good |
| Shimony, 2012140Canada | Adults with PAHN=154129:25 | Pericardial effusion (prevalent v incident) | Mortality | % patients with outcome | Fair |
| Simeoni, 200849Europe | Adults with systemic sclerosis and PHN=2018:2 | BNP |  | Changes in mean in response to therapy (bosentan) | Good |
| Soon, 2011141UK | Adults with PHN=6342:21 | BNP |  | Determination of most accurate log transformed dichotomous BNP variables to predict events | Good |
| Souza, 2007142Not reported/Unclear | Adults with IPAHN=4210:32 | BNP | 6MWD (absolute)Cardiac index Functional classRHC-mPAPRHC-PVRRight atrial pressure | Correlation | Good |
| Taguchi, 2012143Asia | Adults and children with IPAHN=6551:14 | BNP |  | Change in mean over time in response to combination therapy  | Good |
| Takatsuki, 2012144US | Children with PAHN=3322:11 | TRJvRV sizeBNP |  | Change in mean over time in response to therapy (tadalafil) | Good |
| Takatsuki, 2012145US | Children with PAHN=3819:19 | BNP |  | Change in mean over time in response to therapy (transition to or addition of ambrisentan) | Good |
| Takatsuki, 2012146US | Children with PAHN=8846:42 | BNP (BNP and NT-proBNP) | 6MWDRHC-mPAPRHC-RAPRHC-PVRRHC-CITRJv | Correlation | Good |
| Takeda, 2010147Asia | Adults with PAHN=3729:8 | BNP | Mortality | HR | Good |
| Torbicki, 2003148Europe | Adults with PAHN=5643:13 | FACBNPPericardial effusionRA sizeTroponin T | Mortality | HR | Good |
| Utsunomiya, 2011149Asia | Adults with PHN=5039:11 | BNPRA sizeRIMP/MPI/Tei Index | Mortality | HR | Fair |
| Utsunomiya, 2009150Asia | Adults with chronic PHN=5039:11 | RA size | Right atrial pressure RIMP/MPI/Tei Index | Correlation | Good |
| Van Albada, 2008151Europe | Children with PAHN=2918:11 | BNPUric acid | 6MWD (absolute) 6MWD (change)Functional classMortalityCardiac indexRHC-mPAPRHC-PVRMortality | Correlation | Good |
| Vizza, 2012152Europe | Adults with IPAHN=4437:7 | ET-1BNP | WHO FCBNPRHC-mPAPRHC-CIRHC-PVRClinical worseningClinical worsening | CorrelationOR | Good |
| Vizza, 2008153Europe | Adults with PAH associated with CTDN=2521:4 | Endothelin-1 | BNP | Correlation | Good |
| Voelkel, 2000154US | Patients with PHN=191NR  | Uric acid | RHC-mPAPRight atrial pressure  | Correlation | Good |
| Williams, 200661UK | Adults with systemic sclerosisN=10988:21 | BNPBNP10-fold increase from baseline levels10-fold increase in baseline levels | Cardiac index RHC-mPAPRHC-PVRRight atrial pressure Mortality | CorrelationHR | Fair |
| Wilkins, 2005155UK | Adults with IPAH or PAH associated with CTDN=2621:5 | RV sizeCardiac index RIMP/MPI/Tei IndexRA sizeBNP |  | Change in mean from baseline in response to therapy (bosentan) | Good |
| Yamada, 2012156Asia | Adults and children with IPAHN=4129:12 | BNPUric acid | MortalityHospitalization | HR | Good |
| Yanagisawa, 2012157Asia | Adults with PAHN=4638:8 | BNP | Mortality | HR | Good |
| Yang, 2012158Asia | Adults and children with Eisenmenger syndromeN=129:3 | RV sizemPAP |  | Change in mean over time in response to therapy (iloprost) | Fair |
| Yoshida, 2012159Asia | Adults with PAHN=2118:3 | BNPmPAP |  | Mean change from baseline in response to treatment (ambrisentan) | Fair |
| Zafrir, 2007160Asia | Adults with PPH +/- collagen vascular diseaseN=2922:7 | RA sizeRIMP/MPI/Tei IndexRVEF | 6MWD (absolute)Functional class6MWD (absolute)Functional class | Correlation | Good |
| Zeng, 2011161Asia | Adults and children with IPAHN=9561:34 | BNP |  | Means stratified by survivor/nonsurvivor | Good |
| Zhao, 2012162Asia | Adults and children with IPAHN=7656:20 | Uric acid | Mortality | HR | Good |

Abbreviations: 6MWD=6-minute walk distance; BNP=brain natriuretic peptide; CHD=congenital heart disease; CHF=congestive heart failure; CI=cardiac index; CTD=connective tissue disease; CTEPH=chronic thromboembolic pulmonary hypertension; CVD=collagen vascular disease; DLCO=diffusion capacity of the lung for carbon monoxide; FAC=fractional area change; HR=hazard ratio; IPAH=idiopathic pulmonary arterial hypertension; mPAP=mean pulmonary artery pressure; MPI=myocardial performance index; NPV=negative predictive value; 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; PPV=positive predictive value; 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; RVEF= right ventricular ejection fraction ; 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