

# Pulmonary Hypertension Society of Australia and New Zealand 2016 Scientific Abstracts

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## Making Australia the benchmark in echocardiography databases: The National Echo Database Australia (NEDA)

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Epidemiological research from population-based cohort studies have shaped public health strategies. Echocardiography (echo) is one of the most commonly performed cardiac investigations in Australia; however, there are limited epidemiological data quantifying cardiovascular risk for various echo measurements. From Medicare Australia data, 919,309 echos were processed in 2015, excluding State Government hospital echo data. The aim of this paper is to develop the National Echocardiography Database Australia (NEDA), capturing measurement data from digital echo labs across Australia, and to link this data with national death index (NDI). We seek to obtain mortality risk statistics for each cardiac abnormality studied. We have developed an architectural prototype and a “scraper” tool to retrieve every variable from each echo lab, including retrospective data. We identified 650 unique measurements obtained from a comprehensive echo exam. We wrote a unique data dictionary to account for differing variable names from different labs. Text was converted into variables using a parsing algorithm. Two complete echo databases from different software vendors, have been scraped and combined totaling 307,656 echocardiograms collected between 2001 and 2015. Conversion of variable names and measurement units was performed to unify data formats. A total of 5,477,019 valid data points were collected (mean age  $62.9 \pm 16.95$  years). Using novel database engineering, we combined two echo databases from different echo software manufacturers into one database

containing over 300,000 individual echocardiograms. Phased roll-out of NEDA to multiple sites is now planned along with linkage to the NDI, allowing large scale epidemiological research.

## Establishing a pulmonary hypertension exercise program at Fiona Stanley Hospital

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The aim of this paper is to review a newly established pulmonary arterial hypertension (PAH) exercise program describing: (1) referral numbers and program uptake, (2) clinical outcomes, and (3) adverse events. Data were prospectively collected as routine clinical practice from March 1, 2015 to June 30, 2016. Patients with a confirmed diagnosis of PAH were referred from the pulmonary hypertension clinic by specialist physicians. A senior physiotherapist assessed all patients, including six-minute walk distance (6MWD) and muscle strength before and after the program. The 12-week program comprised supervised, twice-weekly, 60-min classes. Exercise prescription and progression were based on patient symptoms (perceived dyspnea using the BORG scale and workload using the RPE scale). Baseline CPET was not used to determine exercise prescription. Patients were monitored symptomatically and with pulse oximetry throughout training. Twenty-four patients were referred to the program ( $n = 24$ ), 17 (70%) were enrolled and seven (30%) declined. Of the 17 enrolled patients, 13 completed training. 6MWD improved at 12 weeks (mean  $\pm$  SD;  $426 \pm 126$  m pre versus  $503 \pm 112$  m post). There were four adverse events (chest pain and dizziness), none requiring hospitalization or discontinuation from the program. Two patients have died from bleeding complications unrelated to the program, one many weeks after completion of training. Preliminary data from our service suggest supervised group PAH exercise training in a hospital outpatient environment is safe and feasible. Exercise training guided by symptoms, BORG dyspnea and RPE scales can improve exercise capacity in patients with PAH.



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## Relationship between pulmonary artery wedge pressure, age, co-morbidities, and treatment response in idiopathic pulmonary arterial hypertension

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Normal pulmonary artery wedge pressure (PAWP) is  $\leq 12$  mmHg but current guidelines include a PAWP up to  $\leq 15$  mmHg for the diagnosis of pulmonary arterial hypertension (PAH). Using registry data, our aims were to determine whether idiopathic PAH (iPAH) patients with a "borderline" PAWP of 13–15 mmHg (iPAH<sub>13–15</sub>) were more likely to have co-morbid risk factors for heart failure with preserved ejection fraction (HFpEF) or to exhibit a differential treatment response, compared to those with strictly normal PAWP (iPAH <sub>$\leq 12$</sub> ). From the PHSANZ Registry, we analyzed 303 treatment-naïve patients (85 had iPAH<sub>13–15</sub>, 218 had iPAH <sub>$\leq 12$</sub> ) who received PAH therapy and had follow-up data at 12 months. Risk factors for HFpEF included hypertension, coronary artery disease (CAD), diabetes, and BMI  $> 30$  kg/m<sup>2</sup>. Compared with iPAH <sub>$\leq 12$</sub> , patients with iPAH<sub>13–15</sub> were older ( $63 \pm 17$  versus  $54 \pm 18$  years,  $P < 0.001$ ) but had similar BMI ( $30.5 \pm 7.2$  versus  $29.2 \pm 6.4$ ,  $P = 0.19$ ). Prevalence of CAD and hypertension but not diabetes were higher in the iPAH<sub>13–15</sub> group (23.5% versus 13.8% had  $\geq 3$  HFpEF risk factors,  $P = 0.05$ ). Although baseline hemodynamics were milder in the iPAH<sub>13–15</sub> group (mPAP:  $43 \pm 16$  versus  $47 \pm 13$  mmHg and PVR:  $7.4 \pm 6.6$  versus  $10.4 \pm 5.9$  WU, both  $P < 0.05$ ), 6MWD was lower ( $270 \pm 130$  versus  $305 \pm 125$  m,  $P = 0.031$ ), compared with iPAH <sub>$\leq 12$</sub> . At 12 months, both iPAH<sub>13–15</sub> and iPAH <sub>$\leq 12$</sub>  groups responded to PAH therapy with a similar increase in 6MWD ( $61 \pm 95$  versus  $64 \pm 88$  m,  $P = 0.78$ ) and proportion displaying functional class improvement (26.7% versus 31.1%,  $P = 0.48$ ). iPAH patients with "borderline" PAWP (13–15 mmHg) are older and have more HFpEF risk factors. Despite this, treatment response to PAH drugs appears similar in both groups.

## Pulmonary arterial hypertension: effects of ERA switch to macitentan

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Macitentan, is a relatively new endothelin receptor antagonist (ERA) with theoretical advantages over the existing ERAs, Bosentan (Bos) and Ambrisentan (Amb), in the treatment of pulmonary arterial hypertension (PAH). The hypothesis of this paper is that switching sub-optimally controlled patients from Bos or Amb would be associated with an improved six-minute walk test (6MWT) and WHO functional class. A retrospective cohort analysis of 37 patients from a single center controlled for age, gender, and PAH type single was conducted. Patients were separated into three treatment groups and followed for 18 months: Group 1 ( $n = 14$ ), patients switched to macitentan from Bos/Amb due to clinician determined sub-optimally controlled PAH; Group 2 ( $n = 11$ ), patients commenced on macitentan as de-novo therapy ( $n = 5$ ) or as add-on to Sildenafil ( $n = 6$ ); and Group 3, patients deemed stable by treating clinician and therapy with Sildenafil and/or ERA (Bos/Amb) ( $n = 12$ ) unchanged. Eleven patients had at least one hospital admission. Mortality was 2/37 (group 1,  $n = 1$ ; group 2,  $n = 1$ ) with an annualized mortality of 3.6%. Patients in group 3 remained stable with one hospital admission and 100% survival. Functional class showed a statistically significant improvement ( $P = 0.001$ ) in groups 1 and 2 and was unchanged in group 3. By six months, there was a 37 m improvement in 6MWT in Group 1 ( $P = 0.17$ ), a 98 m improvement in group 2 ( $P = 0.01$ ), and no change in group 3. Of the patients, 92% continued macitentan throughout the study. One patient stopped due to anemia and one patient stopped all therapy, was palliated, and died shortly thereafter. Annualized mortality in this center's PAH population, all on targeted therapy, is low. Clinical determination of stability appears accurate as none of the patients clinically assessed as stable had significant deterioration over the subsequent 18 months. Macitentan is well tolerated. Switching sub-optimally controlled patients from Bos/Amb to macitentan may be associated with improved functional class.

### Keywords

echocardiography, nutrition, pulmonary arterial hypertension, scleroderma

### Targeting the prostacyclin pathway in the treatment of connective tissue disease associated pulmonary arterial hypertension (PAH): insights from the randomized controlled GRIPHON trial with selexipag

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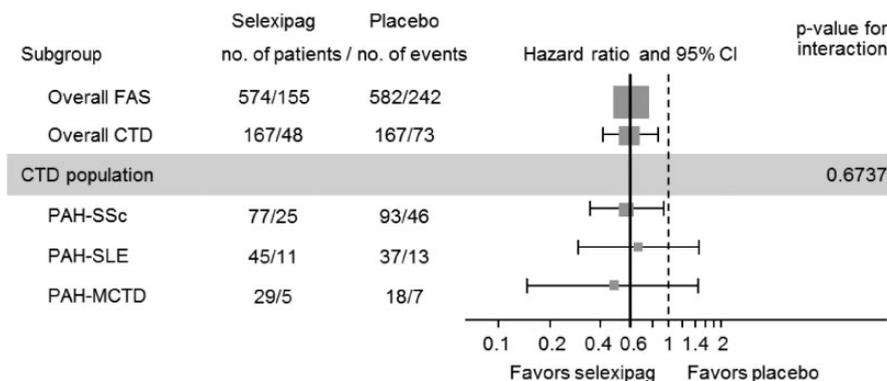
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The phase III GRIPHON study (NCT01106014) enrolled 1156 patients including 334 with pulmonary arterial hypertension–connective tissue disease (PAH-CTD). We examined the

effect of selexipag versus placebo in PAH-CTD subgroups: PAH associated with systemic sclerosis (PAH-SSc), systemic lupus erythematosus (PAH-SLE), and mixed CTD (PAH-MCTD). Patients were randomized 1:1 to placebo or selexipag. Hazard ratios (HRs) (95% CI) were calculated using Cox regression models. Of the 334 PAH-CTD patients, 170 had PAH-SSc, 82 PAH-SLE, and 47 PAH-MCTD; CTD sub-classification was not reported in 35 patients. The majority were receiving an endothelin receptor antagonist, a phosphodiesterase type-5 inhibitor, or both at baseline (73–83%). In the PAH-SSc, PAH-SLE, and PAH-MCTD subgroups, 65%, 33%, and 45% were in WHO functional class III, respectively. Selexipag reduced the risk of morbidity/mortality events by 44% (HR, 0.56; 95% CI, 0.34–0.91) in PAH-SSc, 34% (HR, 0.66; 95% CI, 0.30–1.48) in PAH-SLE, and 53% (HR, 0.47; 95% CI, 0.15–1.48) in PAH-MCTD. Treatment effect was consistent across subgroups (interaction test indicated no heterogeneity;  $P=0.6737$ ) (Fig. 1). By study end, 22 PAH-SSc, seven PAH-SLE, and three PAH-MCTD placebo patients, and 17 PAH-SSc, four PAH-SLE, and eight PAH-MCTD selexipag-group patients had died. Common prostacyclin-associated side effects observed with selexipag in PAH-CTD patients generally occurred at a similar incidence to PAH-non-CTD patients and within the PAH-CTD subgroups. GRIPHON included the largest randomized cohort of PAH-CTD patients to date. The treatment effect of selexipag on time to first morbidity/mortality event was consistent across the subgroups, suggesting that selexipag is an effective therapeutic option in these difficult-to-treat patients.



<sup>1</sup>Other' CTD (n=15) not shown.

CI, confidence interval; CTD, connective tissue disease; FAS, full analysis set; MCTD, mixed connective tissue disease; PAH, pulmonary arterial hypertension; SLE, systemic lupus erythematosus; SSc, systemic sclerosis.

Fig. 1.

## Survival and health-related quality of life in incident systemic sclerosis related pulmonary arterial hypertension: a multicenter Australian cohort study

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Pulmonary arterial hypertension (PAH) is the leading cause of systemic sclerosis (SSc)-related mortality. We sought to determine survival; predictors of mortality, and health-related quality of life (HRQoL) of PAH in a large SSc cohort. Patients enrolled in a SSc longitudinal cohort between 2009 and 2015 were included. Group I PAH was diagnosed on right heart catheterization (RHC) (mPAP ≥ 25 and PAWP < 15 mmHg). Other types of pulmonary hypertension were excluded based on TTE, HRCT scan, PFTs, and V/Q scan. Summary statistics, chi-square tests and survival methods were used to determine survival rates and identify predictors of mortality. HRQoL was calculated using the Medical Outcomes Study Short Form 36 (SF-36). Among 132 SSc-PAH patients, 84.9% were female and 68.9% had limited disease subtype. The mean age at diagnosis of

**Table 1.** Continued

Characteristic	Mean ± SD or %
Anti-centromere pattern ANA	63 (51.6%)
Antiphospholipid antibodies (>ULN)	33 (30%)
RNA polymerase III positive	8 (11.4%)
Scl 70 positive	9 (7.4%)
WHO functional class at time of PAH diagnosis	
Class I	3 (2.3%)
Class II	23 (17.4%)
Class III	79 (59.9%)
Class IV	12 (9.1%)
Baseline 6MWD (m)	326.13 (±105.53)
Baseline mRAP (mmHg)	8.3 (±4.3)
Baseline mPAP (mmHg)	35.6 (±10.4)
Baseline PAWP (mmHg)	10.5 (±3.4)
Baseline cardiac index (L/min/m <sup>2</sup> )	3.2 (±1.9)
Baseline PVR (Wood Units)	8.7 (±38.8)
Presence of a pericardial effusion	24 (18.2%)
Mean DLCO % predicted	46.6 (±13.5)
Mean DLCO/VA % predicted	56.7 (±20.2)

PAH, pulmonary arterial hypertension; MCTD, mixed connective tissue disease; ANA, antinuclear antibody; ULN, upper limit of normal; WHO, World Health Organization; 6MWD, six-minute walk distance; mRAP, mean right atrial pressure; mPAP, mean pulmonary arterial pressure; PAWP, pulmonary artery wedge pressure; PVR, peripheral vascular resistance; DLCO, diffusing capacity of the lung for carbon monoxide; DLCO/VA, adjusted diffusing.

\*Disease duration from first non-Raynaud manifestation.

**Table 1.** Patient characteristics.

Characteristic	Mean ± SD or %
Total patients (n)	132
Female	112 (84.9%)
Age at PAH diagnosis (years)	62.3 (±10.5)
Disease duration at PAH diagnosis (years)	14.1 (±11.9)
Status at censoring	
Alive	70 (53.0%)
Dead	60 (45.5%)
Withdrawn	1 (0.8%)
Unable to contact	1 (0.8%)
Age at recruitment (years)	62.7 ± 10.3
Race	
Caucasian	112 (84.9%)
Asian	6 (4.6%)
Aboriginal-Islander	1 (0.8%)
Hispanic	1 (0.8%)
Follow-up duration from PAH diagnosis (years) (median + IQR)	3.8 (1.6–5.8)
Survival from PAH diagnosis (years) (median + IQR)	4.0 (2.2–6.1)
Disease duration* at PAH diagnosis (years)	14.4 ± 12.1
Disease subtype	
Limited	91 (68.9%)
Diffuse	30 (22.7%)
MCTD	7 (5.3%)
Autoantibody status	

(continued)

**Table 2.** Independent predictors of mortality in SSc-PAH determined by cox proportional multivariable hazard regression analysis.

Characteristic	Hazard ratio (95% CI)	P value
Age at diagnosis of PAH (years)	1.05 (1.0-1.1)	0.03
ILD (FVC > 60%)	2.83 (1.4-5.6)	0.01
WHO functional class	2.01 (1.1-3.9)	0.03
Pulmonary arterial pressure (mmHg)	1.06 (1.0-1.1)	0.001
Digital ulcers present	3.12 (1.4-7.2)	0.01
Specific PAH therapies and anticoagulation		
Vasodilator monotherapy only	Reference	Reference
Vasodilator monotherapy and anticoagulation	0.39 (0.1–1.2)	0.09
Vasodilator combination therapy only	0.49 (0.2–1.2)	0.10
Vasodilator combination therapy and anticoagulation	0.28 (0.1–0.7)	0.007

PAH, pulmonary arterial hypertension; WHO, World Health Organization; ILD, interstitial lung disease; 6MWD, six-minute walk distance; mRAP, mean right atrial pressure; mPAP, mean pulmonary arterial pressure; HCQ, Hydroxychloroquine.

PAH was 62.3 ( $\pm 10.5$ ) years and disease duration at PAH diagnosis was 14.1 ( $\pm 11.9$ ) years. Over a median follow-up of 3.7 years (range, 1.6–5.8 years), 60 (45.5%) patients died, with a median survival time from PAH diagnosis of 3.7 years (Table 1). The standardized mortality ratio for patients with SSc-PAH compared with the general population was 5.8 (95% CI, 4.3–7.8). The years of life lost with SSc-PAH was 15.22 years (95% CI, 12.3–18.1). Kaplan–Meier survival curves showed a survival advantage with combination PAH therapy and anticoagulation. Older age at PAH diagnosis ( $P=0.03$ ), presence of mild ILD ( $P=0.01$ ), worse WHO functional class ( $P=0.03$ ), higher mean pulmonary arterial pressure at PAH diagnosis ( $P=0.001$ ) and presence of digital ulcers ( $P=0.01$ ) were predictive of PAH mortality (Table 2). Combination therapy together with anticoagulation provided the most significant survival advantage, with a 72% reduction in mortality compared with pulmonary vasodilator monotherapy without anticoagulation (Figure 1). Patients with SSc-PAH had consistently poorer HRQoL scores in all domains of the SF-36 form except mental health than the general population. Despite advanced therapy, the median survival in SSc-PAH is less than four years. The addition of anticoagulation to standard combination therapy was associated with a significant survival advantage, pointing to mechanisms involving endothelial abnormalities and small vessel thrombosis in the pathogenesis of PAH.

### Critical pulmonary arterial hypertension necessitating bilateral lung transplantation

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It is difficult to discern if a diagnosis of pulmonary arterial hypertension (PAH) relates to early asymptomatic pulmonary hypertension unveiled by physiological changes during labor or is a recently postpartum acquired condition.

A 29-year-old woman with presyncope and dyspnea presented 11 days post uncomplicated vaginal delivery. Three previous pregnancies were uneventful. Computed tomographic pulmonary angiography and ventilation perfusion scan demonstrated no thromboembolic disease. Transthoracic echocardiogram revealed a severely dilated right ventricle with severe systolic dysfunction. The left ventricle was of normal size and underfilled. Cardiac catheterization revealed pulmonary systemic equalization. Medical therapy with inhaled nitric oxide, inhaled iloprost, and intravenous epoprostenol was initiated with only a small decrement in pulmonary artery systolic pressure. Within a short time, the patient developed hemoptysis exacerbated by thrombocytopenia. This was further complicated by a rising lactate, suprasystemic pulmonary pressures (135 mmHg), extremely low ScvO<sub>2</sub>, and ineffective pulmonary vasodilator therapy, necessitating emergent central VA ECMO being instituted as a life saving measure and bridge to transplantation. The patient was urgently listed for and received

bilateral sequential lung transplantation with the aid of peripheral VA ECMO to help separate from bypass, later transitioned to peripheral VV ECMO for respiratory support. She was discharged home one month after transplantation.

This case highlights the complex surgical management of a difficult pathology, utilizing mechanical circulatory support as both a bridge to transplantation in a critically ill and deteriorating patient as well as postoperative support to recovery.

### Percentage predicted is not superior to absolute six-minute walk distance as a predictor of mortality in pulmonary hypertension

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Absolute six-minute walk distance (6MWD) and functional class have been validated as predictors of mortality in patients with pulmonary arterial hypertension (PAH) and form the basis of PBS (Pharmaceutical Benefits Scheme) funded prescription of pulmonary vasodilator therapies in Australia. Several equations exist for predicting 6MWD in adults from which percentage predicted 6MWD can be calculated. We sought to determine whether percentage predicted 6MWD was a better predictor of mortality than absolute 6MWD in patients with PAH (all WHO groups). Secondary analysis included the impact of baseline WHO functional class and patient demographics on mortality. Analysis was conducted from registry data of the Pulmonary Hypertension Society of Australia and New Zealand (PHSANZ) which includes patients with pulmonary hypertension from all five WHO groups. Inclusion criteria were age at diagnosis > 18 years, diagnosis from 2004, and baseline 6MWD within three months of diagnostic right heart catheter. Baseline mortality predictors were taken at time of diagnosis. Percentage predicted 6MWD was calculated using Chetta, Jenkins, and Enright predictive equations. Survival analysis was performed using cox-proportional hazards model. De-identified data for 2442 Registry subjects were reviewed, of which 923 subjects met the inclusion criteria. There were 285 deaths with an annual incidence rate of 9.2%. Mean 6MWD was 308  $\pm$  136 m. Mean duration of follow-up from diagnosis was 3.3  $\pm$  2.4 years. Absolute 6MWD, percentage predicted 6MWD (all three equations), functional class, and age at diagnosis were all significant predictors of mortality ( $P < 0.001$ ) on univariate analysis. On multivariate analysis, only absolute 6MWD (hazard ratio [HR], 0.997; 95% CI, 0.995–0.998;  $P < 0.001$ ), functional class (HR, 1.965; 95% CI, 1.509–2.557;  $P < 0.001$ ), and age at diagnosis (HR, 1.023; 95%

CI, 1.014–1.032;  $P < 0.001$ ) remained significant. Percentage predicted is not superior to absolute 6MWD as a predictor of mortality in PAH.

### Climbing Kilimanjaro: time-course changes in pulmonary pressures and gas-exchange during altitude exposure in 27 non-acclimatized climbers

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High altitude exposure results in hypoxic pulmonary vasoconstriction (HPV) and increased pulmonary pressure. This study examined the relationship between acute changes in resting pulmonary pressure and pulmonary gas-exchange measures during exercise in a group of non-acclimatized climbers summiting Mount Kilimanjaro. Twenty-seven climbers (age, 44 + 15 years) completed the study. Exercise testing (4-min step test with gas-exchange) and echocardiographic measurements were completed at four different altitudes: 8801 m (barometric pressure, PB = 690 mmHg); 3500 m (PB = 505 mmHg); 4600 m (PB = 428 mmHg) and on return to 880 m (8802 m). Right ventricular systolic pressure (RVSP, mmHg) progressively increased as PB fell during the ascent (8801 m: 19 + 4; 3500 m: 27 + 8; 4600 m: 33 + 8 mmHg). During exercise, the fall in PB was associated with a decrease ( $P < 0.01$ ) in oxygen saturation (nadir SpO<sub>2</sub>(%): 8801 m: 96 + 2; 3500 m: 82 + 3; 4600 m: 73 + 4). The time-course changes in gas exchange tracked changes in RVSP during ascent with a progressive decrease ( $P < 0.01$ ) in SpO<sub>2</sub> and end tidal carbon dioxide production (end-exercise PETCO<sub>2</sub>, mmHg: 8801 m: 37.1 + 3.6; 3500 m: 28.5 + 2.6; 4600 m: 20.8 + 1.9) and an increase in breathing efficiency (VE/VCO<sub>2</sub>: 8801 m: 28.5 + 2.9; 3500 m: 35.8 + 4.6; 4600 m: 50.7 + 5.8). However, while SpO<sub>2</sub> and RVSP normalized on return to lower altitude (8802 m), gas-exchange measures remained altered (PETCO<sub>2</sub>, mmHg: 31.2 + 3.0; VE/VCO<sub>2</sub>: 32.8 + 3.2,  $P < 0.01$  versus 8801 m). With high altitude exposure, there is an increased ventilatory drive characterized by altered gas exchange and associated changes in SpO<sub>2</sub> and pulmonary pressures. However, following high altitude exposure and once SpO<sub>2</sub> and pulmonary pressures are normalized, gas-exchange measures remain altered suggesting that HPV is no longer a potential stimulus for an increased ventilatory drive.

### The National Echo Database Australia (NEDA) and pulmonary hypertension

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We have previously demonstrated that pulmonary hypertension (PHT), identified using echocardiography (echo) is common and that left heart disease accounts for the majority of PHT. Echo measurements of left heart disease may be helpful in predicting the cause of PHT. The aim of this study is to examine prevalence of PHT within NEDA, and uncover left heart predictors of PHT. NEDA utilizes novel database engineering to combine individual databases into a single database. A total of 307,656 echocardiograms from two laboratories have been included in this analysis. We defined PHT as a right ventricular systolic pressure (RVSP) over 40 mmHg. In total, 180,374 echos (59%) had a measurable tricuspid regurgitation (TR) velocity profile from which an RVSP could be calculated. PHT from any cause was identified in 39,699 (22%) echos. Of those in which PHT was identified, the mean RVSP was 51 ± 11 mmHg, compared with 29.5 ± 5.8 in those without PHT ( $P < 0.0001$ ). These patients were older than the overall average for NEDA (mean age, 74.9 ± 12.1 years versus 62.9 ± 16.6,  $P < 0.0001$ ). The ejection fraction (EF) was similar but significantly different between those with PHT and those without (58.1 ± 13.7 versus 61.9 ± 8.7%,  $P < 0.0001$ ). Measures of diastolic function were markedly different (E:E' ratio 17.1 ± 8.5 versus 11.3 ± 5.5,  $P < 0.0001$ ). PHT is common, representing 22% of those with a measurable RVSP in a large echo cohort (over 300,000 echos). Overall, the EF was similar in PHT compared to those without PHT, whereas surrogate markers of filling pressure such as E:E' ratio were markedly different, underpinning the importance of measuring diastolic function in the evaluation of PHT.

## Scleroderma as a scapegoat: recognizing the prevalence of left-sided heart disease in patients with pulmonary hypertension and scleroderma in a tertiary referral center

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Pulmonary hypertension (PHT) may result from a myriad of etiologies. Although scleroderma is an obvious suspect in patients with known scleroderma developing PHT, it is important to exclude left-sided cardiac disease. The prevalence of left-sided cardiac disease in scleroderma patients has not previously been well-defined. We conducted a retrospective case-control trial of scleroderma patients at St Vincent's Hospital Melbourne, with patients categorized according to whether or not they had the outcome of PHT. PHT was identified on echocardiography as a right ventricular systolic pressure >35 mmHg. Demographic variables (age, sex, BMI) and the presence of left ventricular systolic dysfunction, diastolic dysfunction, or aortic stenosis were compared between groups. Of 401 patients with scleroderma, 118 (29.4%) had echocardiographic evidence of PHT and 283 (70.6%) did not. Patients with PHT were older ( $66.3 \pm 12.2$  years versus  $56.7 \pm 13.6$  years,  $P < 0.0001$ ) and more likely to be overweight (BMI,  $27.4 \pm 7.5$  kg/m<sup>2</sup> versus  $25.9 \pm 5.0$  kg/m<sup>2</sup>,  $P = 0.02$ ). On echocardiography, patients with PHT more frequently had diastolic dysfunction (54.4% versus 21.8%,  $P < 0.0001$ ) and aortic stenosis (37.6% versus 16.3%,  $P < 0.0001$ ). There were no differences between groups with regards to gender distribution or mean left ventricular ejection function. Our study suggests that diastolic dysfunction is present in up to half of scleroderma patients with PHT, and aortic stenosis in over one-third. In today's aging Australian population, PHT is frequently multifactorial in nature. It remains important to completely delineate the contribution of left-sided cardiac disease prior to commencing pulmonary vasodilator therapy for scleroderma patients.

	Scleroderma + pHTN	Scleroderma w/o pHTN	Significance
Number			
Mild pHTN			
Moderate pHTN			
Severe pHTN	118 (29.4%)		
77			
26			
15	283 (70.6%)		
Age (years)	$66.3 \pm 12.2$	$56.7 \pm 13.6$	$P < 0.0001$
Male gender (%)	16.9	11.7	$P = 0.15$

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	Scleroderma + pHTN	Scleroderma w/o pHTN	Significance
BMI (kg/m <sup>2</sup> )	$27.4 \pm 7.5$	$25.9 \pm 5.0$	$P = 0.02$
Aortic stenosis (%)	37.6	16.3	$P < 0.0001$
LA dilation (%)	53.4	21.9	$P < 0.0001$
Diastolic dysfunction (%)	54.4	21.8	$P < 0.0001$
LVEF (mean %)	$61.0 \pm 6.6$	$60.9 \pm 4.8$	$P = 0.91$

## Nutritional status, dietary intake, and quality of life in older patients with pulmonary arterial hypertension

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The purpose of this pilot study was to investigate the nutritional status, dietary intake, and quality of life (QoL) of patients with pulmonary arterial hypertension (PAH). Patients were approached during their routine attendance at the Wesley Pulmonary Hypertension Unit or if admitted as inpatients at The Wesley Hospital. Nutritional status was assessed using the Patient Generated Subjective Global Assessment (PG-SGA), dietary intake using a multi-pass 24-h recall and QoL using a PAH specific tool, the emPHasis-10. A total of 18 patients participated (4 men, 14 women; mean (SD) age, 71.9 (2.9) years, mean (SD) BMI  $27.2$  (1.7) kg/m<sup>2</sup>). Fourteen patients were assessed as well-nourished and four patients mild-moderately malnourished. Eleven patients showed signs of mild to moderate muscle and/or fat wasting. Nutrition impact symptoms were experienced by 14 patients with the most frequent symptoms of dry mouth, early satiety, no appetite, and problems swallowing. A majority of patients ( $n = 16$ ) met estimated protein requirements and ten patients met estimated energy requirements. The mean (SD) QoL score was 20.8 (2.8). The PGSGA score was positively correlated with emPHasis-10 score ( $P = 0.045$ ), indicating patients with worse nutritional status had poorer QoL. This is the first study to examine nutritional status, dietary intake, and QoL in patients with PAH. The cohort of patients examined was of older average age and showed significant association between nutritional status and QoL. Further research is warranted to determine if nutrition intervention may improve the QoL of these patients.

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