



**PHSANZ**

**Australian and New Zealand PHT Registry**

**3<sup>rd</sup> annual report**

**Prof Anne Keogh for the Registry Steering Committee**

A/Prof Geoff Strange, Prof Trevor Williams

A/Prof Susanna Proudman, Carolyn Corrigan

Friday 23<sup>rd</sup> October 2015, Pier 1, Walsh Bay

# PHSANZ Registry

- Funded by unrestricted grants (Actelion, GSK, Bayer and Pfizer)
- Steering committee
  - Anne Keogh - Medical director
  - Trevor Williams - President
  - Fiona Kermeen
  - Susanna Proudman

## 2014 report

- Final entry of data and cleaning of all prevalent and incident cases by “travelling PH nurses” - 2004 to Jan 2014 final cut
- Allowed detailed survival analyses (available on website)
- Identified increasing age at diagnosis as a likely limitation to improvement in survival
- CTEPH cases being identified at an increasing rate
- Underutilisation of epoprostenol in FC III and IV
- Extremely low numbers of transplants for PH across ANZ (under referral, late referral)

# 2015

- Centres are now autonomous and are entering incident (new) cases and completing followup of prevalent cases as per PBS visits
- Large and important database
- Any user is welcome to request binational data to solve hypotheses
- Project requests on entire data set are available by application to committee (use of own data at your discretion)
- Publication policy first 2 authors are those who propose and write up project and other authors at discretion ([www.phsanz.com.au](http://www.phsanz.com.au))
- *please use it as it was designed – to be a clinically useful tool*



Patient Location:

No: 000006 Mortality Status: **Alive** Date of Death:  
Treating Physician:

Patient & Diagnosis Snapshot Diagnostics RHC ECHO SHRT PFT Medications Social Economic Status

### Patient Details

### Diagnosis

### Mortality

Title:   
First Name:   
Middle Name:   
Last Name:   
Former Names:   
Gender:   
Date of Birth:  Age Years: **34**  
Address L1:   
Address L2:   
Suburb:   
State:   
Country:   
Home Ph:   
Work Ph:   
Mobile Ph:   
Email:   
Ethnicity:   
Origin:  Aboriginal  Torres Strait Is.  
Country of Birth:   
Lang. spk. at Home:   
Marital Status:

Primary PHT Diagnosis:   
Secondary Diagnosis:   
Tertiary Diagnosis:   
Date of Diagnosis:  -> Age: **68**  
Treating Physician:   
Diagnosis Confirmed Date:   
Symptom Onset Date:   
Functional Class:   
Hospital MRN / FNZ:

Mortality Status:   
Mortality Status:   
Date of Death:   
Location of Death:   
Primary Cause of Death:   
Autopsy Performed:

### Print Options

These reports will be e-mailed instead of printing.

### Hospitalizations

Year	Date	Event	Institutions

### Summary Form Comments

Very user friendly



# Six Minute Walk Test

Record 3 of 12 (Total 9,459)

Delete Menu

Age Group:   
Location:

Patient No: 000006  
Owner:

## 6MWT Test Details

Test Date: 7 Aug 2012

Baseline:

Age at Test: 72

Functional Class: III

Weight: 103.7 kg

Height: 161.8 cm

O2 Usage:  L/Min

Walking Aid:  Notes:

### Comments

	Start	Finish
SaO2 %:	95	94
HR:	88	90
Systolic BP:	111	110
Diastolic BP:	78	80

\*Distance: 158 m

\*Predicted Distance: 355 m

% Predicted: 53%

Post Borg: 3 /10

Sequential SMWD with individual patients results against predicted results adjusted for height, weight and gender

Add Another

Done



# Right Heart Catheter Test

Record 1 of 1 (Total 3,000) Delete Menu

Age Name:   
Location:

Patient No: 000006  
Owner:

## RHC Test Details

Test Date: 18 Sep 2008

Baseline:

Age at Test: 69

Weight: 303.0  kg  
Height: 163.0  cm  
BSA: 2.07  
BMI: 38.77

Challenge:  +   
Dose:

### Post Challenge

HPAP:   
CI:   
FVR:

### Comments

Item with \* are used on the snapshot chart - >

	Reading	Normal Values
Rhythmic	<input type="checkbox"/>	*
HR:	<input type="checkbox"/>	60 - 100 bpm
SAP:	<input type="checkbox"/>	85 - 140 mmHg
DAP:	<input type="checkbox"/>	40 - 90 mmHg
*MAP:	<input type="checkbox"/>	70 - 90 mmHg
RA:	10 <input type="checkbox"/>	1 - 7
SPAP:	96 <input type="checkbox"/>	20 - 35 mmHg
DPAP:	40 <input type="checkbox"/>	4 - 12 mmHg
MPAP:	58 <input type="checkbox"/>	10 - 20 mmHg
MPVWP:	10 <input type="checkbox"/>	6 - 12 mmHg
LVDDP:	<input type="checkbox"/>	4 - 12 mmHg
TPG:	48 <input type="checkbox"/>	0 - 12 mmHg
CO:	3.00 <input type="checkbox"/>	4 - 8 l/min
*CI:	1.4 <input type="checkbox"/>	2.0 - 4.2 l/min/m <sup>2</sup>
*FVR:	1,280 <input type="checkbox"/>	100 - 240 dynes/cm <sup>2</sup>
SVR:	<input type="checkbox"/>	770 - 1500 dynes/cm <sup>2</sup>

RHC includes an area for challenge results. Comments eg on dobutamine at the time.

Add Another

Done

Medications Physicians Registry Reports Menu

PFT Registry Patient No: 000006

Record 2 of 1,665 (Total 1,665)

Admin Portal New Record Delete Print Find Find All List Menu

Patient No: 000006 Mortality Status: **Alive** Date of Death:

### Patient PFT Snapshot

This is summary information. Please go the appropriate tab to edit any details. Charts show last 12 months

Diagnosis		Current Medications	
Primary PFT Diagnosis:	<b>PAC Idiopathic</b>	Medication	Start Date
Secondary Diagnosis:		Furosemide 40mg TABLETS (PLAIN)	
Date of Diagnosis:	8 Sep 2008	Warfarin 2mg TABLETS (PLAIN)	20 Jun 2008
Treating Physician:		Bosentan 125mg TABLETS	5 Jan 2011
Diag. Confirmed Date:	18 Sep 2008	Sildenafil 25mg TABLETS/Sildenafil 100mg TABLETS	24 Jan 2012
Functional Class:	III		

GMVT Tests		RHC Tests		ECHO Tests	
Distance	Predicted Distance	HPAP	CI	PVR	PAP

Registry ID: 000006    Owner: RANG (24)    Hospital Provider No.:    Entered: 13 Jan 2012 by Peter King    Modified: 30 Aug 2013 by Peter King

Snapshot sheet is useful in electronic or paper records

Can send to referring physicians as the summary sheet



2,980 patients  
entered registry

If diagnosed < 2004  
removed from analysis

297 patients ineligible due  
to small numbers in each  
diagnostic group / coded as  
out of proportion PAH

2,672 cleaned  
patients in current  
analysis

**Up from 2,104  
in Jan 2014!**

# Registries for PHT

	number of patients
Israel (2001)	44
Chinese (2007)	72
French (2010)	674
COMPERA	785
VOLT Amb (2011)	1,003
REVEAL (2012)	3,515.....from 270 mill pop and 55 centres
PHS ANZ PHT (2015)	2,980.....from 25 mill pop and 20 centres!

- Largest registry per capita worldwide!

Users	Patients	Medis	EMAT	RHC	EOHO	PFT	Hospital Events	Surveys	Physicians
81	2,973	14,013	17,353	5,154	16,657	6,093	4,090	833	65
7	200	481	1,624	439	751	230	67	13	5
1	61	616	124	68	106	136	116	0	0
1	0	0	0	0	0	0	0	0	0
1	0	0	0	0	0	0	0	0	0
7	365	779	620	508	635	307	146	22	5
5	8	30	16	7	20	7	17	0	3
5	23	107	36	17	53	8	56	1	4
9	524	3,694	4,164	735	3,645	889	730	558	2
2	115	278	808	118	849	71	62	2	3
2	303	486	299	121	334	151	102	76	2
3	56	171	155	76	260	11	69	1	13
4	315	1,261	2,080	426	1,907	3,470	77	1	1
1	1	2	2	2	3	2	1	0	2
5	301	426	254	125	283	83	160	0	3
4	135	483	595	142	627	149	251	23	9
5	712	3,701	3,783	1,989	4,090	816	1,763	67	5
5	19	152	70	20	96	87	51	2	1
7	275	861	2,075	574	2,236	338	310	16	5
4	93	382	304	112	390	100	76	1	1
3	67	103	342	75	372	258	36	50	1

16 nurse users  
doing a lot of work!!

# Patients included in analysis

(all pts diagnosed before 2004 were excluded)

Aetiology	N=2,672	%
iPAH	732	28
PAH / SSC	511	20
PAH / MCTD incl SLE	165	6
PAH / CHD	202	7.5
PAH Left Heart (WHO group 2)	176	6.5
PAH Lung (WHO group 3)	300	11.5
CTEPH	284	10.5

% Female = 68%

WHO FC I = 1%

WHO FC II = 13%

WHO FC III = 69%

WHO FC IV = 14%

# Demographics

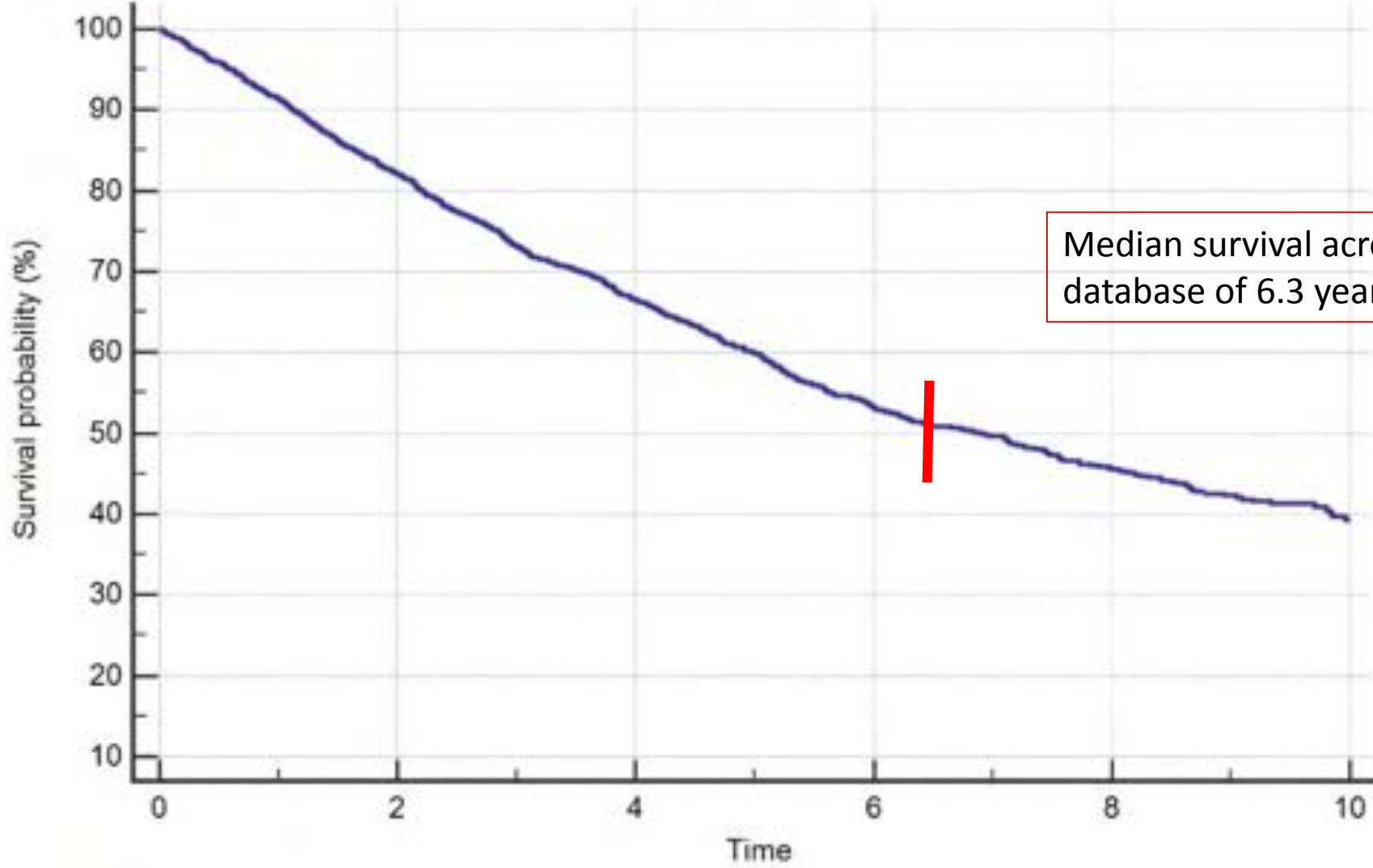
	iPAH	PAH / SS <sub>c</sub>	PAH / CHD	PHT / LHD	PHT / RESP	CTEPH
Mean age at diagnosis	57±19	62±12	36±19	70±11	67±13	57±16
Delay to diagnosis (yrs)	2.1±3.5	2.2±2.7	5.0±8.7	3.0±2.7	3.7±4.1	2.4±3.1
SMWD m	302±136	309±130	361±139	271±120	249±118	353±144
RA mmHg	9.5±4.9	8.3±4.7	8.5±4.7	12.2±5.9	8.7±5.6	10.2±5.8
PVR dyn/s/cm-5	681±466	513±359	791±579	381±212	590±363	671±352
mPAP mmHg	45±15.1	36±13.3	55±16.6	39±11.5	42±11	46±13
CI L/min/m <sup>2</sup>	2.5	2.7	3.1	2.4	2.5	2.3

STATE \* Diag\_Groups Crosstabulation

		Diag_Groups						Total	
		iPAH	PAH/SSc	PAH/MCTD	PAH/CHD	PHT/LHD	PHT/Respir		CTEPH
STATE	Count	152	192	72	74	121	189	87	887
	% within STATE	17.1%	21.6%	8.1%	8.3%	13.6%	21.3%	9.8%	100.0%
	Count	159	91	42	61	14	39	108	514
	% within STATE	30.9%	17.7%	8.2%	11.9%	2.7%	7.6%	21.0%	100.0%
	Count	79	5	1	14	2	0	2	103
	% within STATE	76.7%	4.9%	1.0%	13.6%	1.9%	0.0%	1.9%	100.0%
	Count	98	97	19	8	12	25	9	268
	% within STATE	36.6%	36.2%	7.1%	3.0%	4.5%	9.3%	3.4%	100.0%
	Count	145	59	10	28	17	37	29	325
	% within STATE	44.6%	18.2%	3.1%	8.6%	5.2%	11.4%	8.9%	100.0%
	Count	19	24	5	7	2	4	11	72
	% within STATE	26.4%	33.3%	6.9%	9.7%	2.8%	5.6%	15.3%	100.0%
	Count	85	43	16	10	8	6	38	206
	% within STATE	41.3%	20.9%	7.8%	4.9%	3.9%	2.9%	18.4%	100.0%
Total	Count	737	511	165	202	176	300	284	2375
	% within STATE	31.0%	21.5%	6.9%	8.5%	7.4%	12.6%	12.0%	100.0%

		functionalClass				Total
		1	2	3	4	
STATE	Count	4	145	687	104	940
	% within STATE	0.4%	15.4%	73.1%	11.1%	100.0%
	Count	10	69	387	124	590
	% within STATE	1.7%	11.7%	65.6%	21.0%	100.0%
	Count	0	0	104	2	106
	% within STATE	0.0%	0.0%	98.1%	1.9%	100.0%
	Count	1	32	204	44	281
	% within STATE	0.4%	11.4%	72.6%	15.7%	100.0%
	Count	6	25	249	59	339
	% within STATE	1.8%	7.4%	73.5%	17.4%	100.0%
	Count	1	12	56	10	79
	% within STATE	1.3%	15.2%	70.9%	12.7%	100.0%
	Count	7	59	135	26	227
	% within STATE	3.1%	26.0%	59.5%	11.5%	100.0%
Total	Count	29	342	1822	369	2562
	% within STATE	1.1%	13.3%	71.1%	14.4%	100.0%

### All Mortality



Number at risk  
2594

1727

1045

572

278

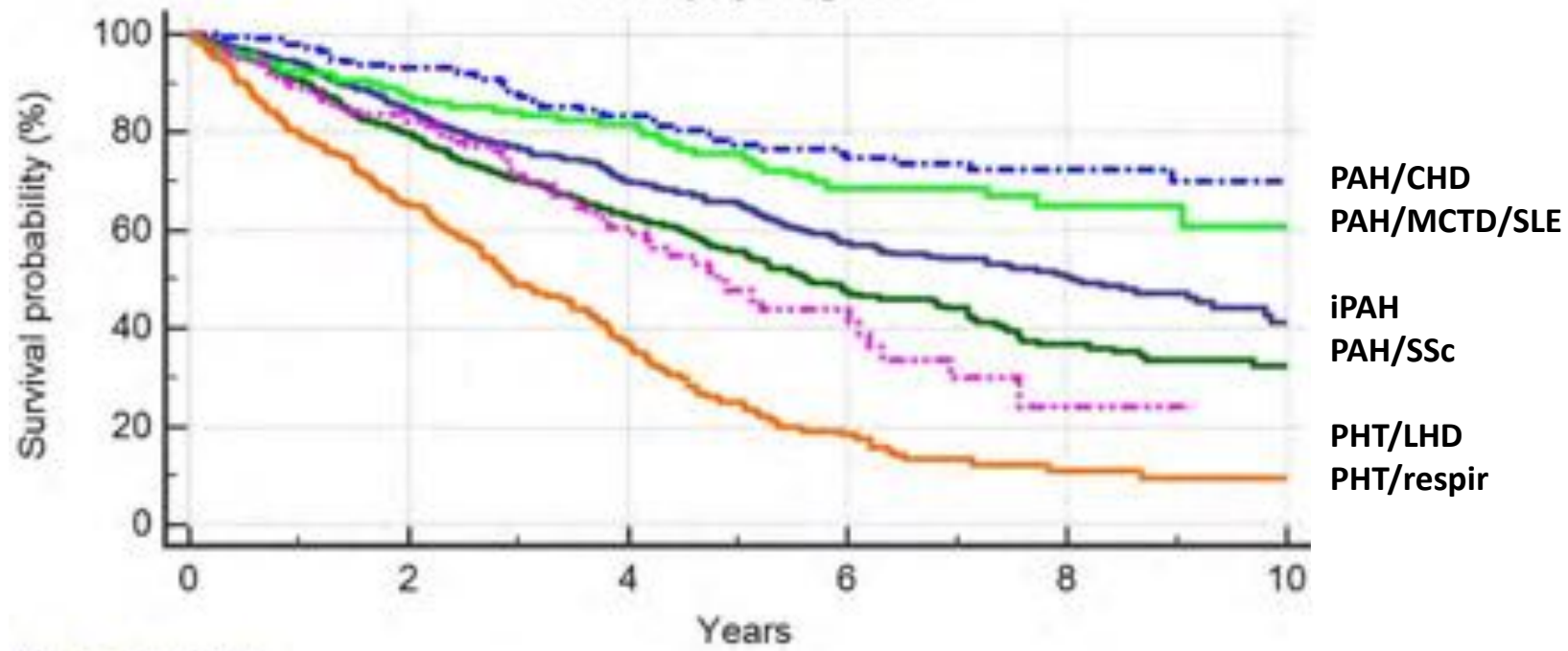
87



Pulmonary  
Hypertension  
Society  
of Australia and New Zealand



Mortality by Diagnosis

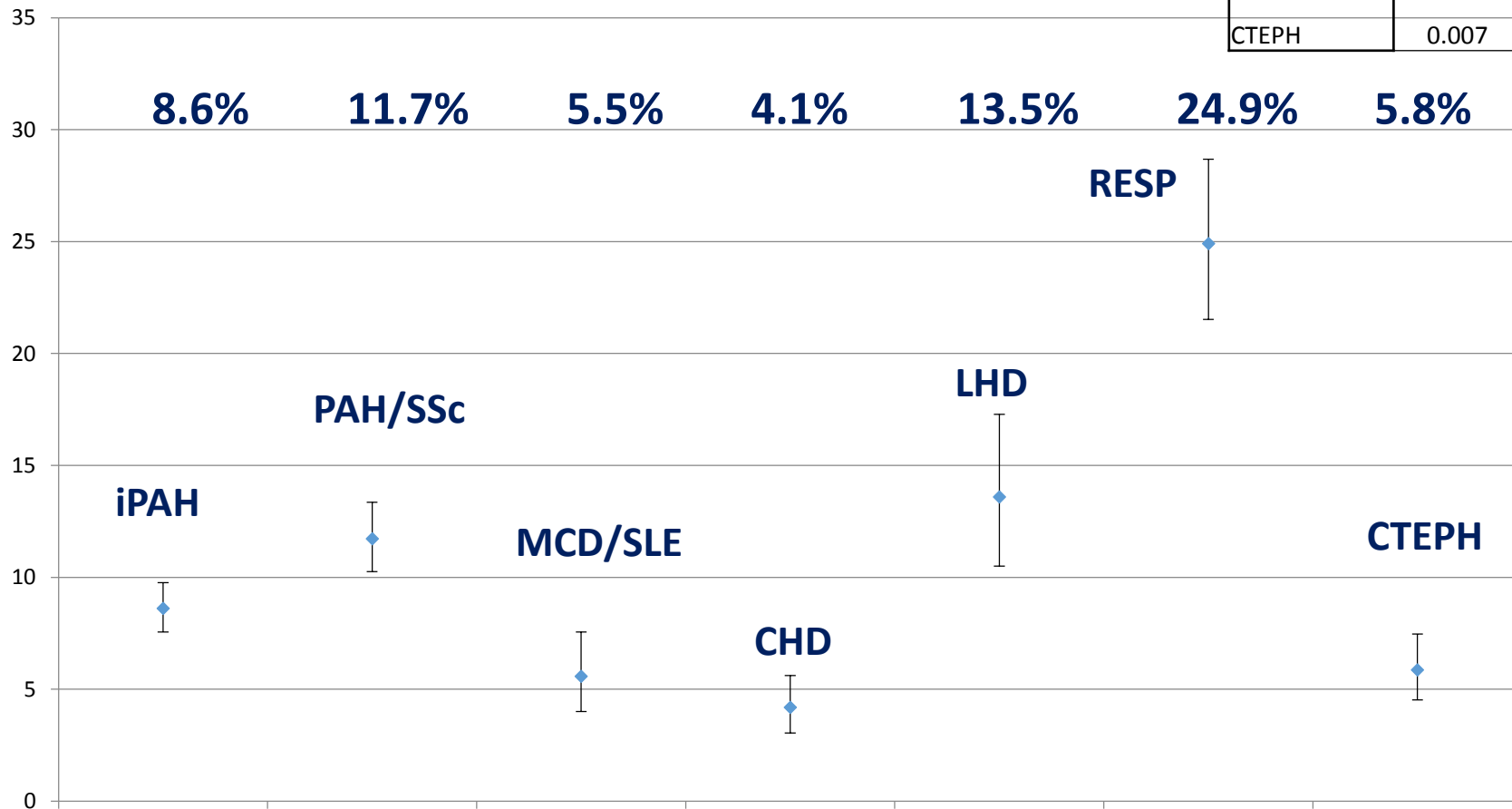


Number at risk

Diagnosis	0	2	4	6	8	10
iPAH	708	487	304	151	88	25
PAH/SSc	499	334	209	122	51	22
PAH/MCTD	163	113	85	54	29	7
PAH/CHD	200	164	121	79	43	17
PHT/LHD	167	103	41	18	2	0
PHT/Respir	298	156	63	23	9	2

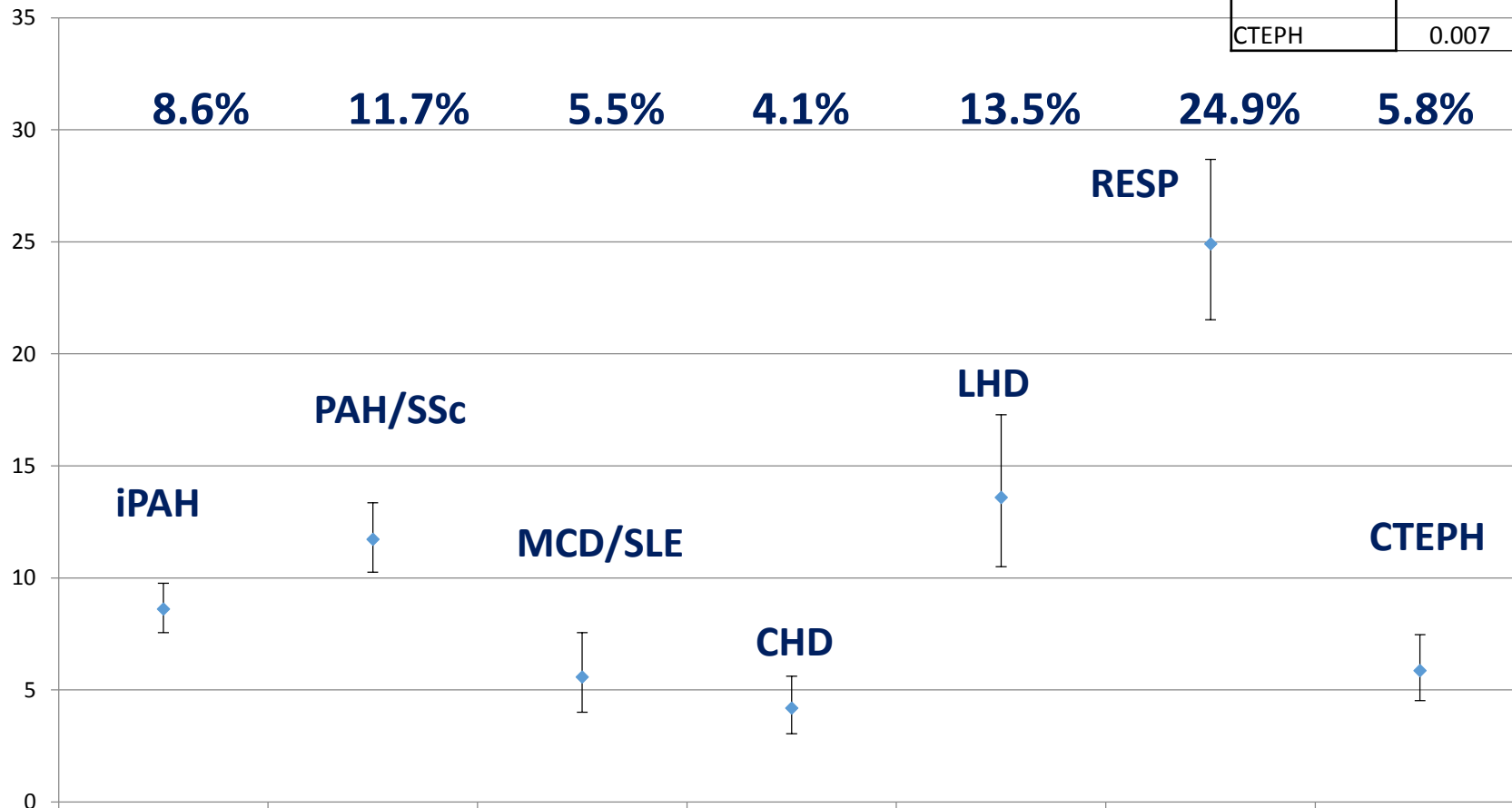
# Annualised mortality

	iPAH	PAH/SSc	PAH/MCTD	PAH/CHD	PHT/LHD	PHT/Respir
PAH/SSc	0.001					
PAH/MCTD	0.011	<0.001				
PAH/CHD	<0.001	<0.001	0.165			
PHT/LHD	0.001	0.254	<0.001	<0.001		
PHT/Respir	<0.001	<0.001	<0.001	<0.001	<0.001	
CTEPH	0.007	<0.001	0.813	0.056	<0.001	<0.001



# Annualised mortality

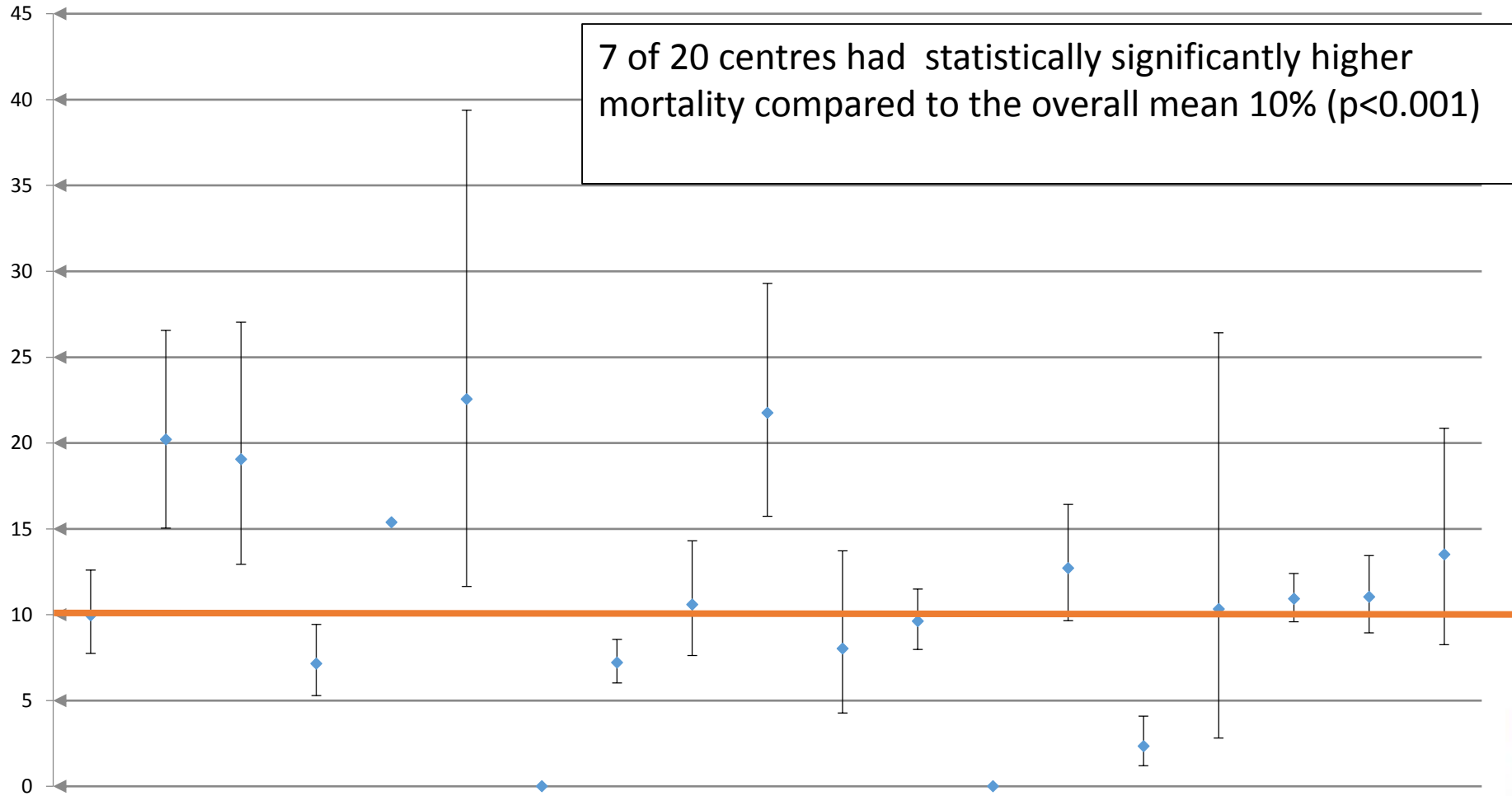
	iPAH	PAH/SSc	PAH/MCTD	PAH/CHD	PHT/LHD	PHT/Respir
PAH/SSc	0.001					
PAH/MCTD	0.011	<0.001				
PAH/CHD	<0.001	<0.001	0.165			
PHT/LHD	0.001	0.254	<0.001	<0.001		
PHT/Respir	<0.001	<0.001	<0.001	<0.001	<0.001	
CTEPH	0.007	<0.001	0.813	0.056	<0.001	<0.001



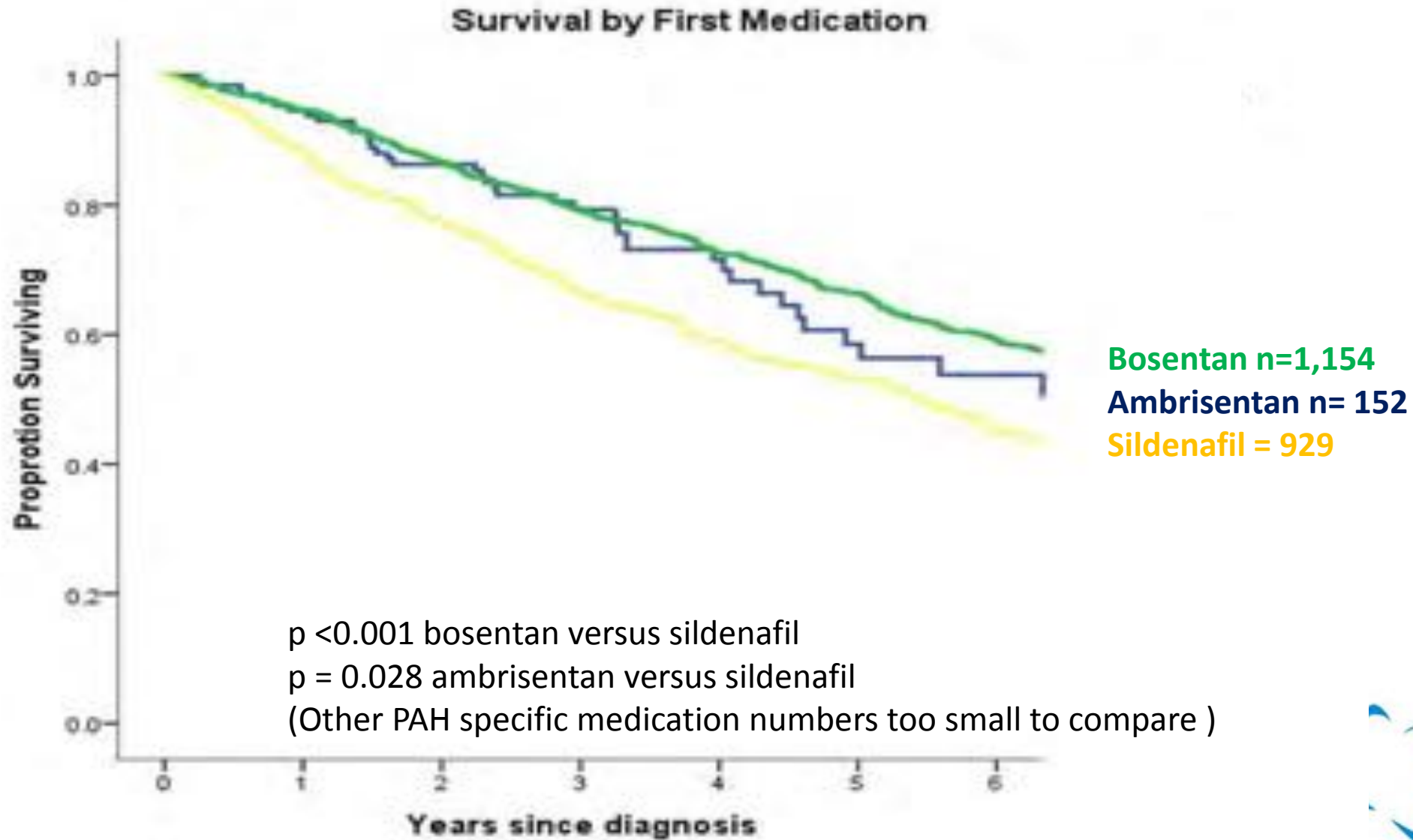
## Summary:

- Resp PH very high annual mortality
- Scleroderma as bad as left heart disease
- Congenital similarly low as SLE
- CTEPH the lowest mortality of all

# All cause PH annualised mortality by centre (% mortality, confidence intervals)



# Survival based on ever initiated monotherapy – 6 year survival



# PHSA ANZ Registry

- n = 2,672 clean patients – largest per capita in the world
- Annualised rate of death acc to etiology ranging from 8.6% for iPAH to 24.9% for PAHResp
- Median survival all cause PHT 6.5yrs - Respiratory related PHT, left heart disease and scleroderma statistically significant highest mortality
- Significant mortality variance by centre which probably reflects the mix of patients being treated
- A survival benefit for first initiation of bosentan and ambrisentan over sildenafil as monotherapy
- 2016
  - More analysis on medications to be done
  - Explore less frequent diagnoses eg portopulmonary
  - Delta change in parameters with treatment



Councillor Carolyn Corrigan  
NSW VIC



Chris Frampton  
Statistician



Dr Geoff Strange  
COO

Thankyou.

Your comments please?

[www.phsanz.com.au](http://www.phsanz.com.au)







Survival rates International Registries, incident and prevalent: iPAH

	1 year	2 years	3 years	5 years	7 years
French	83%	67%	58%	na	na
US REVEAL	91%	na	74%	65%	na
Spanish	89%	na	77%	68%	na
ANZ	95%	87%	80%	70%	50%

M McGoon et al. Pulmonary Arterial Hypertension: Epidemiology and Registries. Advances in PH. 2014;13:1:21-6





# Echo Test Results

Record 4 of 12 (Total 9,264) Delete Menu

Age Row:   
Location:

Patient No: 000006  
Owner:

## Echo Test Results

Test Date: 24 Jan 2012  Baseline:  Age at Test: 72

	Readings	Normal Values	Comments
Left Atrium (LA)	48 <input type="checkbox"/>	< 40 mm	<div style="border: 1px solid gray; height: 400px;"></div>
LV Diastole (LVEDD)	40 <input type="checkbox"/>	35 - 50 mm	
LV Systole (LVESD)	29 <input type="checkbox"/>	20 - 40 mm	
TR Volume	4 <input type="checkbox"/>	Normal	
TR Velocity	<input type="checkbox"/>	Normal < 2.5 m/s	
RxSP (Rx-PA gradient)	65 <input type="checkbox"/>	10 - 20 mmHg	
JVP (RA)	15 <input type="checkbox"/>	2 - 3 mmHg	
*PASP	80 <input type="checkbox"/>	---	
Rx Basal Diameter	49 <input type="checkbox"/>	20 - 50 mm	
Rx Distation	<input type="checkbox"/>	Normal	
Rx Function	4 <input type="checkbox"/>	Normal	
Pericardial Effusion	No <input type="checkbox"/>	No	
TAPSE	12 <input type="checkbox"/>	> 16 mm	

Add Another

Done

## STATE \* gender Crosstabulation

		gender		Total	
		Female	Male		
STATE	Count	635	332	967	
	% within STATE	65.7%	34.3%	100.0%	
	Count	394	217	611	
	% within STATE	64.5%	35.5%	100.0%	
	Count	83	24	107	
	% within STATE	77.6%	22.4%	100.0%	
	Count	211	77	288	
	% within STATE	73.3%	26.7%	100.0%	
	Count	257	115	372	
	% within STATE	69.1%	30.9%	100.0%	
	Count	69	22	91	
	% within STATE	75.8%	24.2%	100.0%	
	Count	163	73	236	
	% within STATE	69.1%	30.9%	100.0%	
	Total	Count	1812	860	2672
		% within STATE	67.8%	32.2%	100.0%





# Pulmonary Function Test

Record 1,730 of 1,730 (Total 1,730)

Delete

Menu

Age (yrs):

Location:

Patient No: 000006

Owner:

**Alert! Please enter Test Date.**

## Pulmonary Function Test Details

Test Date:

Baseline:

Age:

Height at Test (cm):

Weight at Test (kg):

### Lung Function (Pre-Bronch)

	Measured	% Predicted	Over-ride % Predicted
FVC - Forced Vital Capacity (L):	<input type="text"/>	<input type="text"/>	<input type="text"/>
FEV1 - Forced Expiry Volume (L):	<input type="text"/>	<input type="text"/>	<input type="text"/>
FEF - (25-75%) (L/s):	<input type="text"/>	<input type="text"/>	<input type="text"/>
Total Lung Capacity (L):	<input type="text"/>	<input type="text"/>	<input type="text"/>
DLCO (ml/min/mmHg):	<input type="text"/>	<input type="text"/>	<input type="text"/>

Add Another

Done

Who does not want to know what role obesity plays in PAH??

patient\_ageAtDiagnosis\_c RA\_PVR\_c MPAP CI\_c BMI\_c distance \* STATE

STATE	patient_ageAtDiagnosis_c	RA	PVR_c	MPAP	CI_c	BMI_c	distance
Mean	60.641	10.060	592.481	43.260	2.615	29.62	300.656
N	967	814	643	637	637	790	895
Std. Deviation	17.1396	5.6887	409.0979	14.1389	.8326	21.041	131.6717
Minimum	1.0	-2.0	0	13.0	.9	12	10.0
Maximum	96.0	37.0	2963.0	108.0	6.2	386	758.0
Mean	57.241	9.290	568.049	43.554	2.526	34.31	319.846
N	610	497	431	514	434	488	552
Std. Deviation	18.2333	5.0983	373.8391	14.1803	.8191	51.537	150.6423
Minimum	.0	.0	-34.0	6.0	.8	14	5.0
Maximum	90.0	34.0	2400.0	122.0	6.7	700	785.0
Mean	59.196	10.338	647.268	47.962	2.515	26.67	309.167
N	107	74	56	78	39	42	90
Std. Deviation	15.6756	4.9664	386.6683	14.7326	.8487	5.913	148.7224
Minimum	25.0	2.0	198.0	21.0	1.4	16	18.0
Maximum	88.0	25.0	1571.0	88.0	4.7	41	810.0
Mean	62.417	9.112	329.564	31.956	3.090	32.77	313.696
N	288	232	172	271	167	189	276
Std. Deviation	13.4396	4.2064	265.8033	13.7716	.8483	40.958	123.6603
Minimum	11.0	.0	14.0	11.0	1.3	16	31.0
Maximum	88.0	24.0	1752.0	96.0	5.9	519	595.0
Mean	51.065	9.484	711.727	46.872	2.494	29.99	282.858
N	370	308	245	313	216	258	309
Std. Deviation	23.1477	5.3286	466.3921	15.3922	.9058	30.030	140.3247
Minimum	.0	.0	-37.0	11.0	1.0	12	.0
Maximum	90.0	26.0	3091.0	102.0	6.5	450	635.0
Mean	50.912	9.560	604.017	41.364	2.582	27.16	339.023
N	91	75	60	79	61	74	87
Std. Deviation	19.4362	5.5489	434.0642	13.7011	.7997	7.661	143.3295
Minimum	4.0	1.0	.0	19.0	1.2	14	10.0
Maximum	62.0	25.0	2133.0	83.0	4.3	49	688.0
Mean	55.051	8.759	812.898	48.446	2.297	28.04	333.775
N	236	174	166	166	162	171	173
Std. Deviation	15.9768	5.3630	405.3018	12.6841	.5477	5.968	133.7061