11.08 Incidence and outcomes of pulmonary arterial hypertension in the Republic of Ireland

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Pulmonary hypertension (PH) is a progressive disease of the pulmonary vasculature, which is characterised by premature morbidity and mortality. The aim of this study is to define the characteristics of PH in the national PH unit in Ireland.

This study received institutional ethical approval(IRB:1/378/2176TMR). Cases of PH which were referred to the NPHU between 2010 and 2020 were included. PH was defined as a mean pulmonary artery pressure $\geq$ 25mmHg at right heart catheterisation.

Four hundred and fifteen cases of PH were identified. Group 1 pulmonary arterial hypertension (PAH) accounted for 39% (n=163) of cases, with a calculated annual incidence of 3.11 per million population (95% CI, 1.53-4.70). Connective tissue disease associated PAH (CTD-PAH) was the predominant subgroup, responsible for 49% of cases (**Table 1**). The mean age at PAH diagnosis was 56 ±15 years and 86% (n=111) received double- or triple-combination therapy within the first twelve months of diagnosis. The 1-, 3- and 5-year transplant-free survival for PAH was 89%, 75% and 65%. This was significantly lower for individuals with CTD-PAH ($p<0.05$).

This study describes the incidence and outcomes of PAH in Ireland. While the outcomes are comparable to other centres, the incidence appears low, suggesting that improved disease awareness and case recognition are required.

Table 1:

	CTD-PAH	IPAH	CHD-PAH	PoPH	PVOD	DPAH	CCB	HPAH	HIV-PAH	HHT-PAH
Subjects, n	80 (49)	33 (20)	22 (13)	11 (7)	5 (3)	3 (2)	3 (2)	2 (1)	2 (1)	2 (1)
Sex: female n (%)	72 (90)	26 (79)	13 (59)	2 (18)	2 (40)	3 (100)	3 (100)	2 (100)	2 (100)	1 (50)
Age (years): mean ± SD	64 ± 11	56 ± 17	55 ± 21	47 ± 9	69 ± 9	58 ± 15	34 ± 13	46 ± 14	45 ± 1	47 ± 29
WHO functional class: % I/II/III/IV	0/22/60/18	3/24/55/18	0/50/50/0	0/36/64/0	0/20/40/40	0/33/33/33	0/67/33/0	0/0/100/0	0/50/50/0	0/50/50/0
BNP (ng/L): mean ± SD	611 ± 795	268 ± 295	271 ± 348	63 ± 40	417 ± 675	1330 ± 1800	53 ± 41	298 ± 87	---	187 ± 127
Risk stratification: % Low/intermediate/High risk	8/61/31	27/45/28	27/55/18	9/91/0	20/60/20	33/33/33	33/67/0	0/50/50	50/50/0	0/100/0
Right heart catheterization (mean, ±SD)										
mPAP (mmHg)	43 ± 11	50 ± 12	51 ± 25	51 ± 17	38 ± 14	43 ± 25	38 ± 1	49 ± 0	44 ± 6	46 ± 0
PAWP (mmHg)	11 ± 4	8 ± 3	13 ± 5	12 ± 7	12 ± 10	7 ± 0	7 ± 2	8 ± 0	12 ± 0	---
CO (L/min)	4 ± 1	4 ± 2	4 ± 1	5 ± 1	4 ± 0	3 ± 0	5 ± 1	2 ± 0	4 ± 0	9 ± 0
PVR (WU)	9 ± 4	12 ± 7	12 ± 13	6 ± 3	8 ± 0	13 ± 0	6 ± 1	18 ± 0	15 ± 0	---
DLCO, % predicted	35 ± 14	69 ± 20	73 ± 21	50 ± 6	43 ± 13	19 ± 0	88 ± 0	---	---	---
PH therapy: n (%)	79 (99)	32 (97)	14 (64)	9 (82)	5 (100)	3 (100)	1 (33)	2 (100)	2 (100)	1 (50)
Monotherapy	20 (25)	4 (12)	4 (29)	4 (44)	2 (40)	1 (33)	1 (100)	0	1 (50)	0
PD5 inhib.	10 (50)	4 (100)	2 (50)	3 (75)	2 (100)	1	1 (100)	0	0	0
ERA	10 (50)	0	2 (50)	1 (25)	0	0	0	0	1 (100)	0
Double combination therapy	45 (56)	22 (67)	9 (64)	5 (56)	2 (40)	1 (33)	0	2 (100)	1 (50)	1 (100)
PD5 inhib. & ERA	44 (98)	21 (95)	9 (100)	5 (100)	2 (100)	1 (100)	0	2 (100)	1 (100)	1 (100)
sGCS + ERA	0	1 (5)	0	0	0	0	0	0	0	0
ERA + PGI2	1 (2)	0	0	0	0	0	0	0	0	0
Triple combination therapy	14 (18)	6 (18)	1 (7)	0	1 (20)	1 (33)	0	0	0	0
PD5 inhib. + ERA + neb PGI2	12 (86)	4 (67)	1 (100)	0	1 (100)	1 (100)	0	0	0	0
PD5 inhib. + ERA + oral PGI2	2 (14)	2 (33)	0	0	0	0	0	0	0	0

Table 2 describes the characteristics of pulmonary arterial hypertension subgroups at the time of diagnosis, with a specific focus on demographics and treatment patterns. Data was incomplete for the following characteristics and parameters at the time of diagnosis: mPAP was missing for 22 (13%) subjects, PAWP for 22 (13%), CO for 85 (52%), PVR for 95 (58%) and DLCO was missing in 87 (53%) cases. Of note: While mPAP was unavailable at diagnosis for 13% of PAH subjects, a subsequent RHC with evidence of a mPAP≥25mmHg was available for all subjects, but not included in this analysis. *Abbreviations: CTD-PAH: Connective tissue disease associated pulmonary arterial hypertension; IPAH: Idiopathic pulmonary arterial hypertension; CHD-PAH: Congenital heart disease associated PAH; PoPH: Portopulmonary hypertension; PVOD: Pulmonary veno occlusive disease; DPAH: Drug-associated PAH; CCB: Long-term responsive to calcium channel blockers PAH; HPAH: Hereditary PAH; HIV-PAH: HIV associated PAH; Hereditary haemorrhagic telangiectasia associated PAH; SD: Standard deviation; WHO: world health organisation; BNP: B-type natriuretic peptide; mPAP: mean pulmonary artery pressure; PAWP: pulmonary artery wedge pressure; CO: cardiac output; PVR: pulmonary vascular resistance; DLCO: diffusion capacity for carbon monoxide; PH: pulmonary hypertension; PD5 inhib: phosphodiesterase type-5 inhibitor; ERA: Endothelin receptor antagonist; sGCS: Soluble guanylate cyclase stimulator; PGI2: prostacyclin; neb: nebulised. Inadequate data was omitted and replaced by "---".*