Canadian Pulmonary Hypertension Registry Annual Report

Version: 1.0

Reporting Timeframe: 01 July 2022 – 30 June 2023

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1. OVERVIEW

Canadian Pulmonary Hypertension Registry (CPHR) is a multicenter, prospective registry of incident and prevalent patients with pulmonary arterial hypertension (PAH) and chronic thromboembolic pulmonary hypertension (CTEPH) who are evaluated and treated at expert centres across Canada in adult populations. The main goal of the CPHR is to collect real-world epidemiological information, to facilitate monitoring of outcomes in the Canadian PH community, and to be a resource to answer focused research questions and quality improvement questions.

To date there are 11 adult and 2 pediatric active centres that are entering patient data at their centres. Additional 1 adult and 1 pediatric centres are in various stages of start-up.

2. PARTICIPATING CENTERS STATUS

Center Name	PI Name	Status
Vancouver	Dr. John Swiston	ongoing data entry since 01Jan2017
Hamilton	Dr. Nathan Hambly	ongoing data entry since 01Mar2017
Calgary	Dr. Doug Helmersen	ongoing data entry since 01Oct2017
Ottawa	Dr. Lisa Mielniczuk	ongoing data entry since 01Apr2018
Halifax	Dr. Paul Hernandez	ongoing data entry since 01Aug2019
Moncton	Dr. Krista Kemp	ongoing data entry since 01Feb2020
Winnipeg	Dr. David Christiansen	ongoing data entry since 01Sep2020
Québec City	Dr. Steeve Provencher	ongoing data entry since 01Jul2021
Edmonton	Dr. Rhea Varughese Dr. Jason Weatherald	ongoing data entry since 15Oct2021
St. John's	Dr. George Fox	ongoing data entry since 15Oct2021 (start date is 01July2020)
BC Children's - Vancouver (pediatric)	Dr. Martin Hosking	ongoing data entry since 01Jul2022 (start date is 01May2021)
Toronto	Dr. John Granton	ongoing data entry beginning of 2023
London	Dr. Sanjay Mehta	obtaining ethics approvals
SickKids – Toronto (pediatric)	Dr. Luc Martens	have ethics approval
Sainte-Justine – Montreal (pediatric)	Dr. Anne Fournier	obtaining ethics approvals

3. DATA COLLECTED

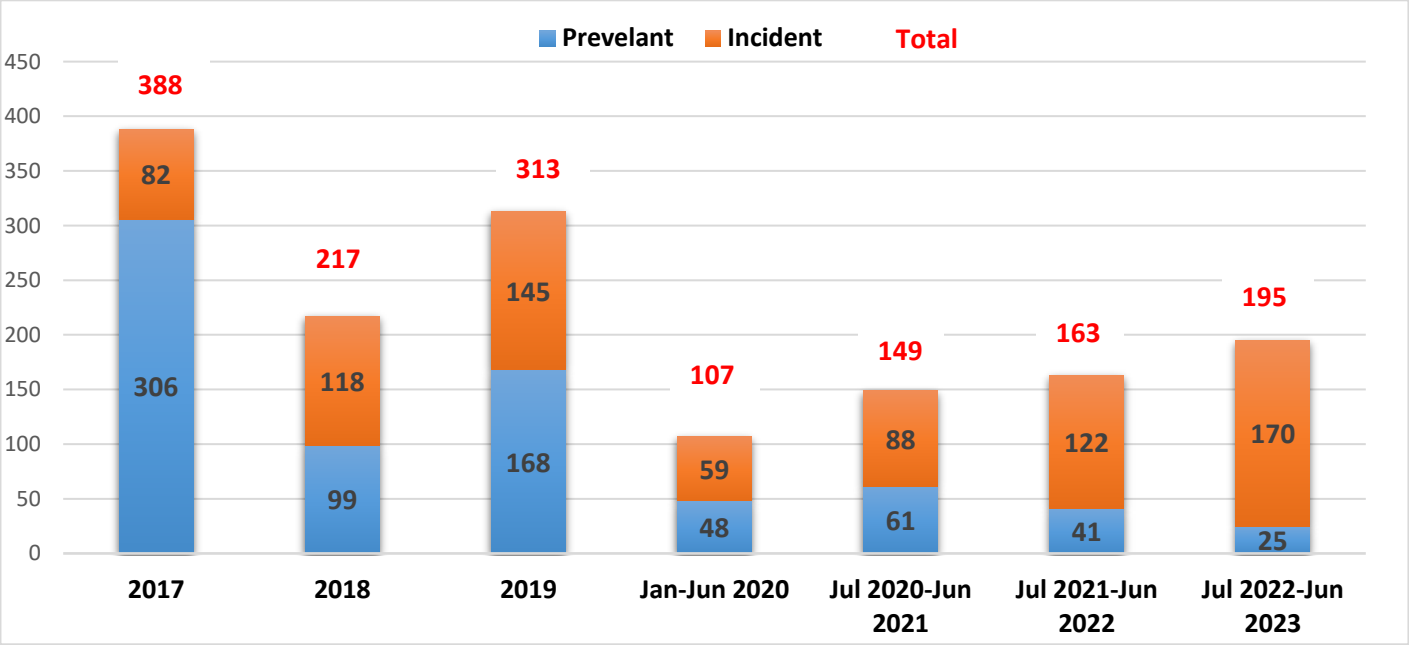
Data below is cumulative data entered into the registry database across all participating sites from inception **01Jan2017** to **30June2023.**

3.1. Patients in the Registry with Confirmed Diagnosis

Site	Patients Entered	PAH	CTEPH
Vancouver	1162*	514	167
Hamilton	207	159	26
Calgary (data from last year)	292	205	79
Ottawa	178	143	32
Halifax	25	20	2
Moncton	77	45	10
Winnipeg	73**	59	12
Quebec	190	97	11
Edmonton (data from last year)	2	2	0
St. John's	6	3	2
TOTAL	**2212**	**1247**	**341**

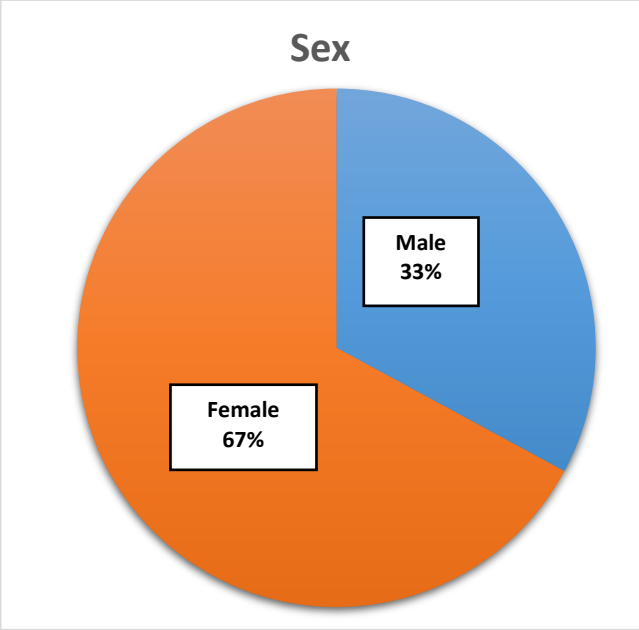
* Vancouver site enters all WHO groups into the database
** Data has not yet been entered on consented patients.

Figure below represents total number of incident and prevalent patients that have been entered into the registry database per year.



* Higher numbers are attributed to addition of new sites to the registry.

3.2. Sex and Age



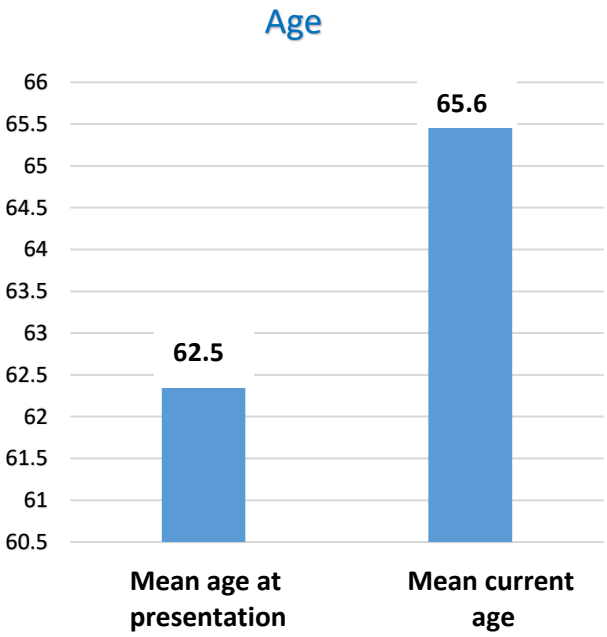
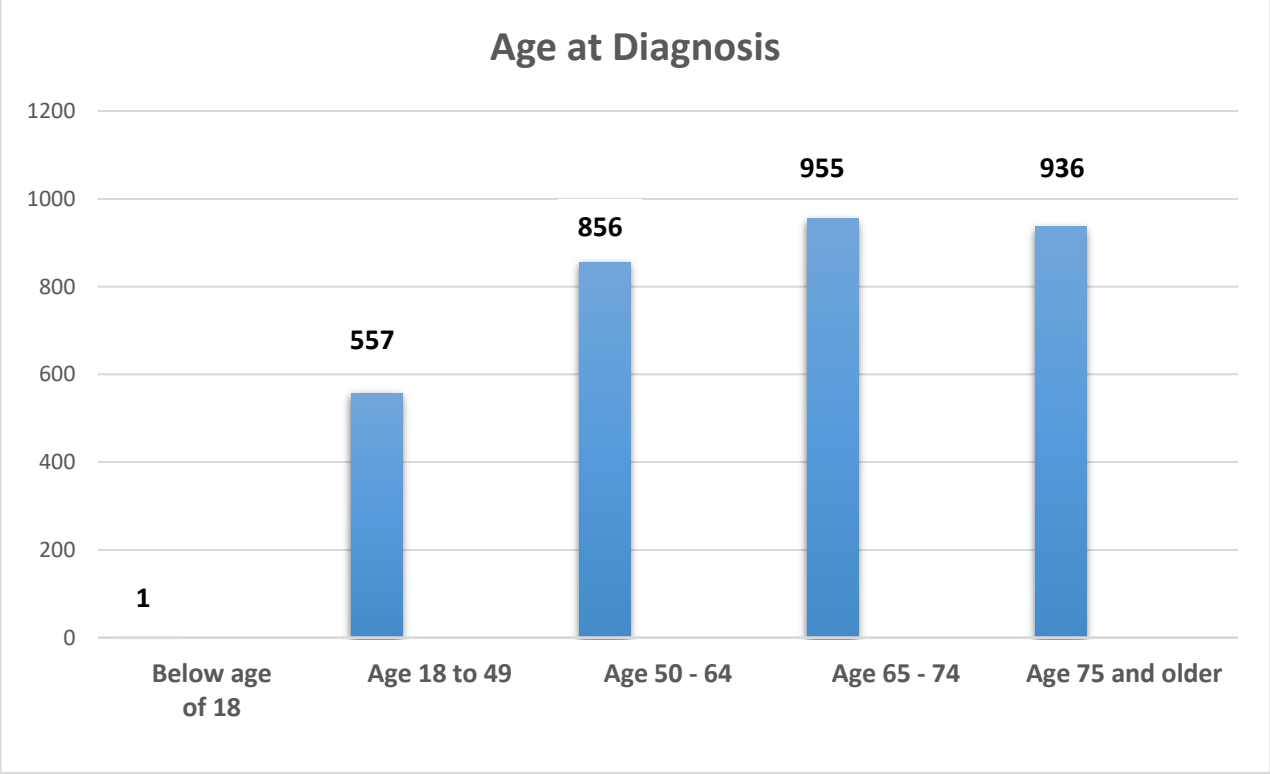
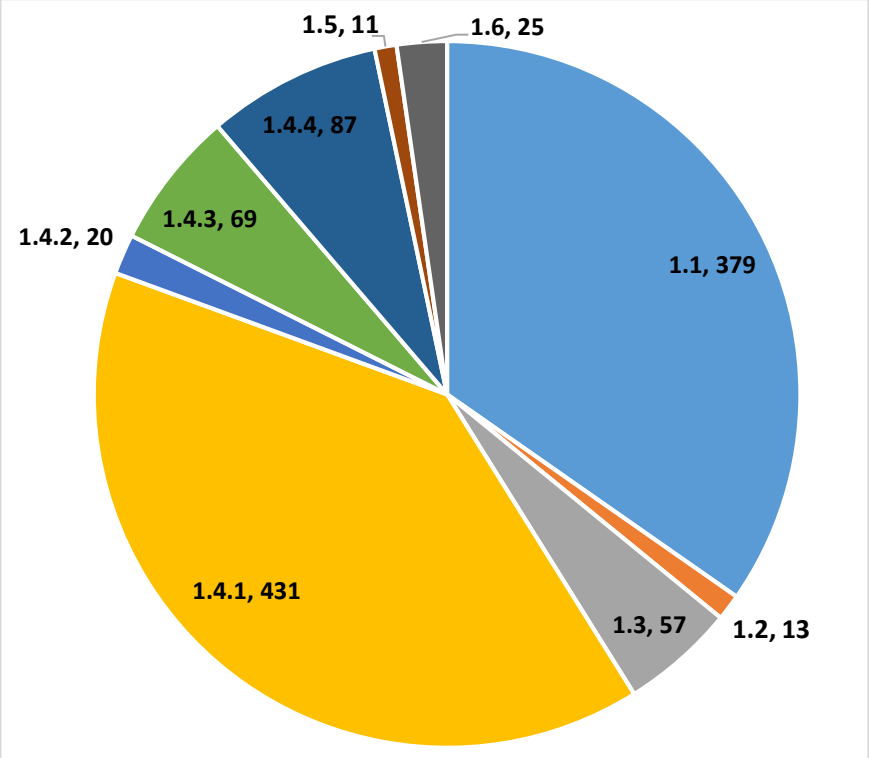


Figure below is the breakdown of the number of patients at the age of diagnosis.



3.3. Group 1 Specifics

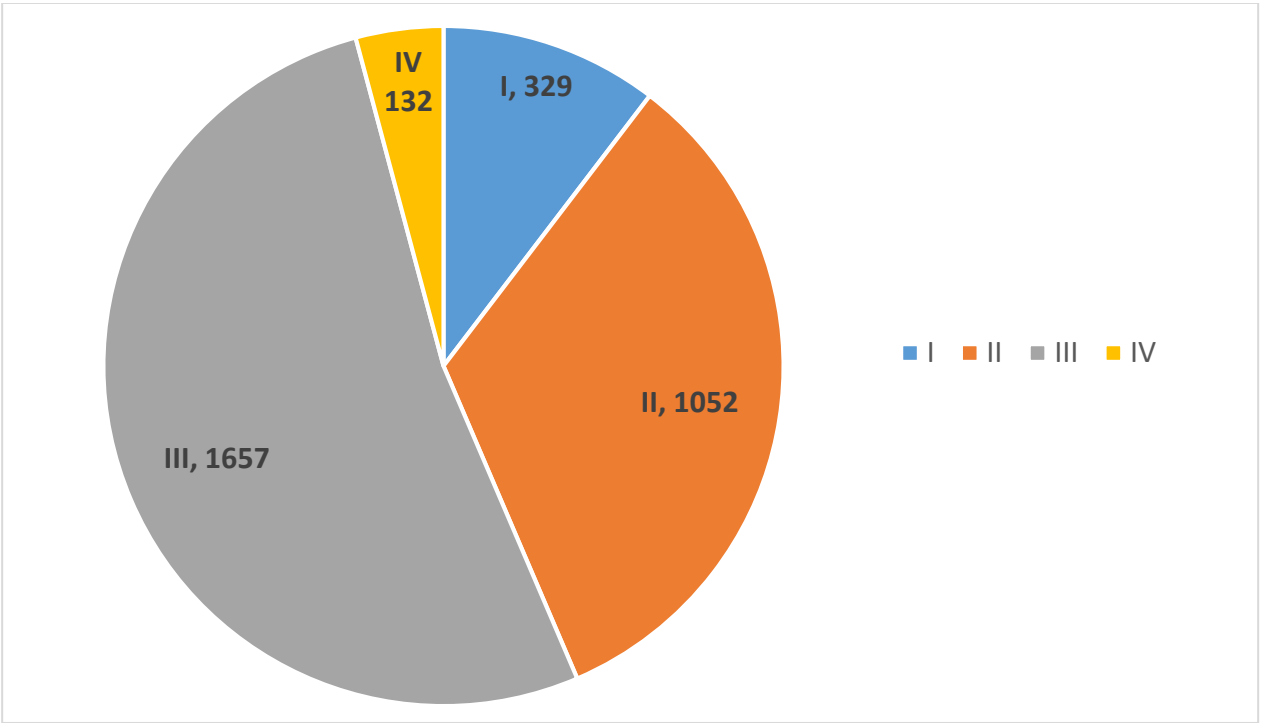
Breakdown of clinical classification of Group 1 PH.



1.1 Idiopathic
1.2 Heritable
1.3 Drug and toxins
1.4.1 Connective Tissue Disease
1.4.2 HIV
1.4.3 Portal Hypertension
1.4.4 Congenital Heart Disease
1.5 PAH long-term responders to calcium channel blockers
1.6 PVOD/PCH

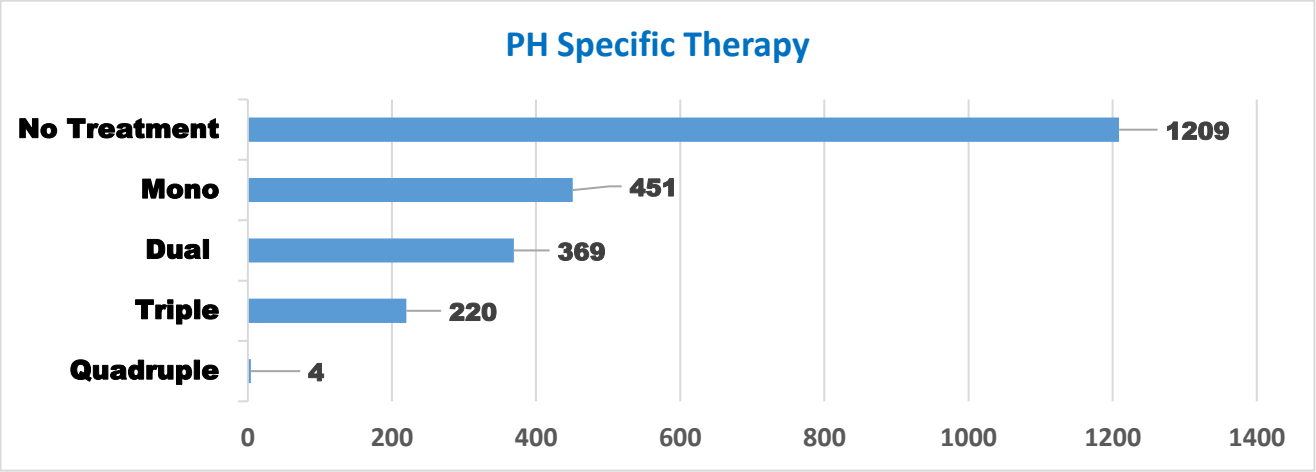
3.4 WHO FC Groups

Breakdown of the WHO FC at the time of first visit.



3.5 PH Specific Therapies

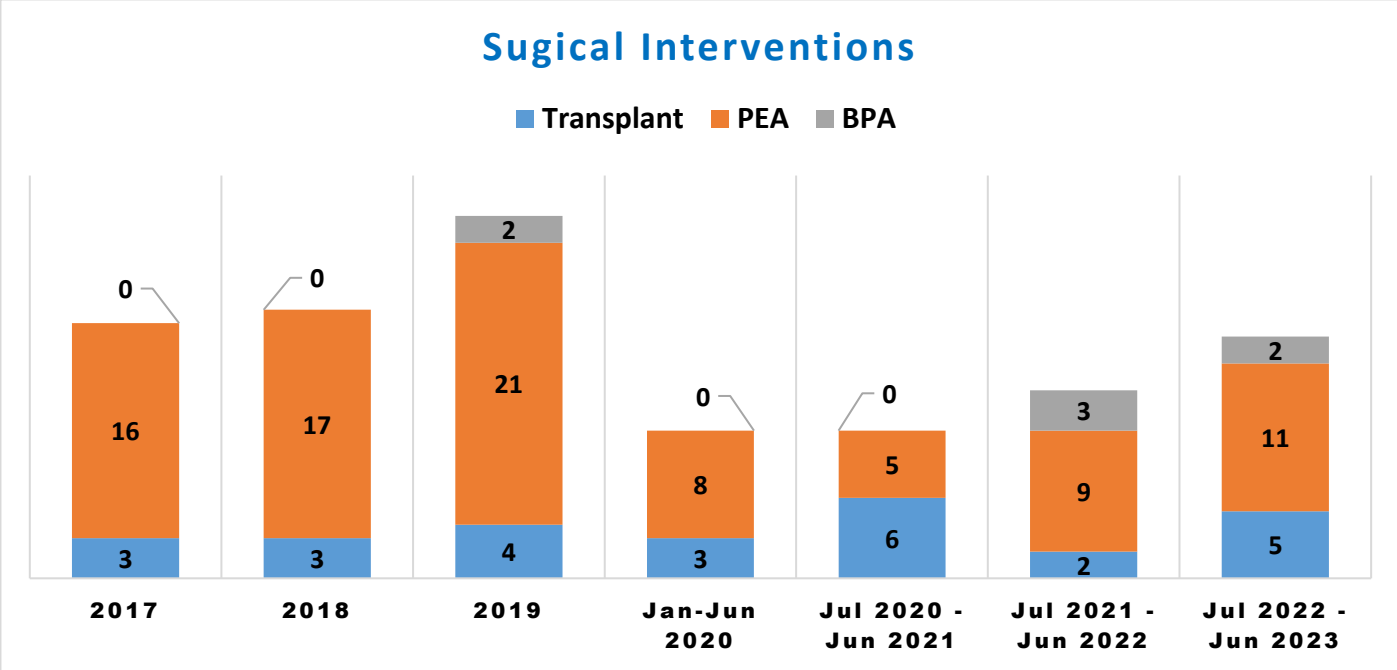
PH specific therapies approved in Canada: Ambrisentan, Bosentan, Macitentan, Sidenafil, Tadalafil, Riociguat, Selexipag, Epoprostenol, Treprostinil. Figure below depicts treatment combination distribution of PH specific therapy.



*Quadruple therapy - transitioning between two therapies.

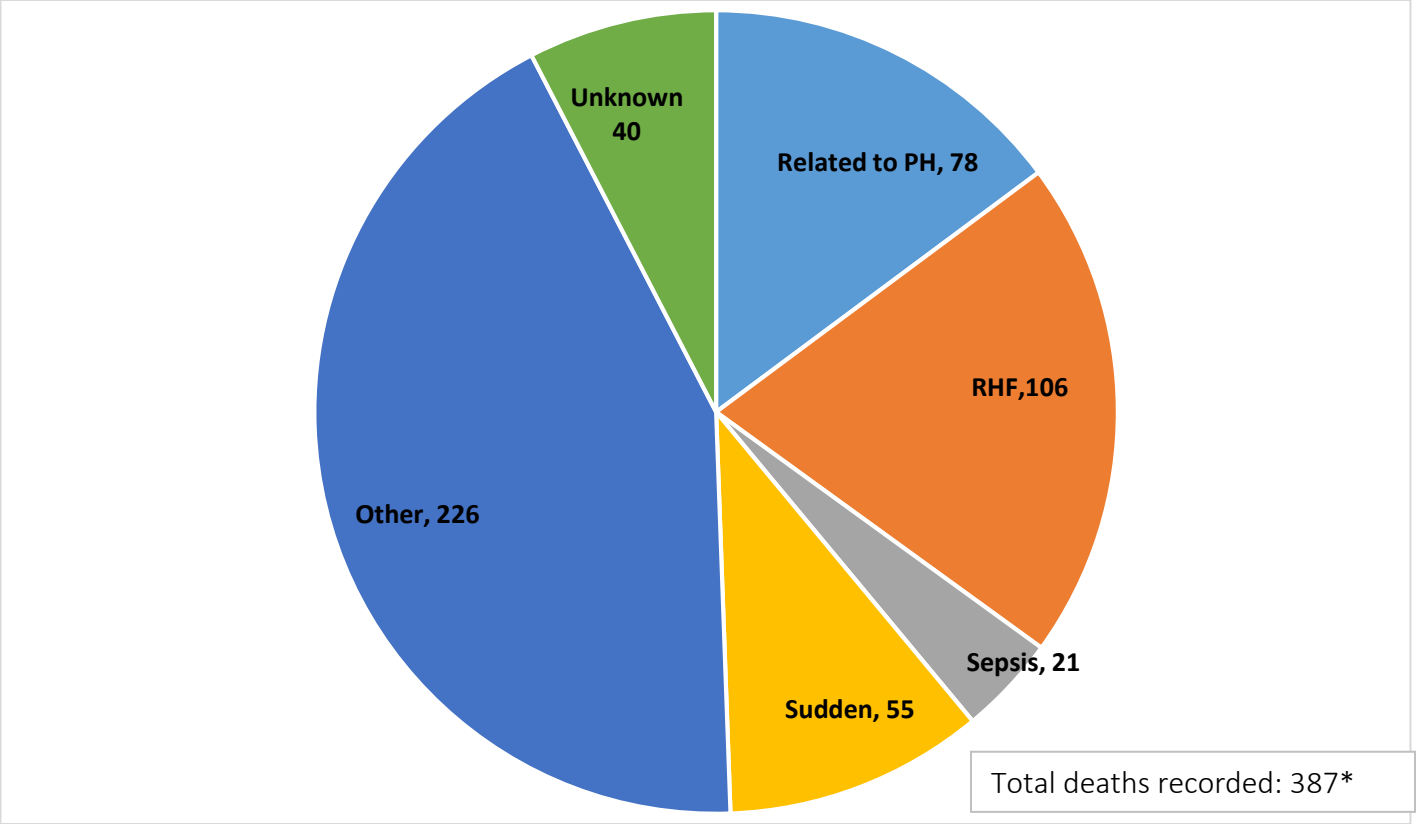
3.6 Transplants, PEA and BPA

Number of transplants, pulmonary endartectomy (PEA) and balloon pulmonary angioplasty (BPA) surgeries performed on patients across all sites.



3.7 Cause of Death

Breakdown of the cause of death of deceased patients.



Other reasons	#	Other reasons	#
Cancer	41	Cirrhosis	5
Pneumonia	24	Fall	3
Congestive heart failure/disease	21	MAID	5
Liver failure/disease	10	PVOD	4
Palliative	17	Pulmonary fibrosis	4
Other	83	Post-Surgery complications	3
Hypoxemia	7	Aspiration	3
Respiratory Failure	16	Interstitial lung disease	13
COVID	11	GI Bleed	4
Renal failure/disease	21	Influenza	2
AECOPD/COPD	12	Biventricular failure	7
PE	10		

*Not all deaths resulted from a single cause; therefore, total number of causes will not equal total number of deaths.

4. DATA USE AND RESEARCH

The following are published studies that utilized some of the collected registry data.

4.1. Published Journal Articles

Brunner NW, Legkaia L, Al-Ahmadi F, Lee L, Norena M, Lam CSM, Yim JJ, Luong C, Weatherald J, Nador RG, Levy RD, Swiston JR. Does community size or commute time affect severity of illness at diagnosis or quality of care in a centralized care model of pulmonary hypertension?, Int J Cardiol. 2021 Jun 1;332:175-181. https://doi.org/10.1016/j.ijcard.2021.03.035

- lead by Nathan Brunner, Vancouver

Moghaddam N, Swiston JR, Tsang MYC, Levy R, Lee L, Brunner NW. Impact of targeted pulmonary arterial hypertension therapy in patients with combined post-and precapillary pulmonary hypertension. Am Heart J. 2021;235:74-81. https://doi.org/10.1016/j.ahj.2021.01.003

- lead by Nathan Brunner, Vancouver

de Perrot, M., Donahoe, L., McRae, K., Thenganatt, J., Moric, J., Chan, J., McInnis, M., Jumaa, K., Tan, K. T., Mafeld, S., Granton, J., & Canadian CTEPH Working Group. (2022). Outcome after pulmonary endarterectomy for segmental chronic thromboembolic pulmonary hypertension. The Journal of Thoracic and Cardiovascular Surgery. 2022 Feb 28. https://doi.org/10.1016/j.jtcvs.2021.10.078

- lead by Marc DePerrot, Toronto

Zelt JGE, Sugarman J, Weatherald J, Partridge ACR, Liang J, Swiston J, Brunner B, Chandy G, Stewart DJ, Contreras-Dominguez V, Thakrar M, Helmersen D, Varughese R, Hirani N, Umar F, Dunne R, Doyle-Cox C, Foxall J, Mielniczuk L. Mortality trends in pulmonary arterial hypertension in Canada: a temporal analysis of survival per ESC/ERS Guideline Era European Respiratory Journal Jan 2021, 2101552; DOI: https://doi.org/10.1183/13993003.01552-2021

- lead by Lisa Mielniczuk, Ottawa

Jason Weatherald, MD, Hina Iqbal, MD, Lisa Mielniczuk, MD, Abdul Rehman Syed, BHK, Lena Legkaia, BSc, Jennifer Howard, Nicole Dempsey, Tamara Rader, MLIS,John Swiston, MD, and Steeve Provencher, MD. Priorities for pulmonary hypertension research: A James Lind Alliance priority setting partnership Journal of Heart and Lung Transplantation (10.1016/j.healun.2022.09.015) https://www.jhltonline.org/article/S1053-2498(22)02160-X/ppt

- lead by Jason Weatherald, Calgary

Ostad S, Sugarman J, Alkhodair A, Liang J, Mielniczuk LM, Hambly N, Helmersen D, Hirani N, Thakrar M, Varughese R, Norena M, Kularatne M, Swiston JR, Kapasi A, Weatherald J, Brunner NW. Association Between the Pulmonary Artery Pulsatility Index and Prognosis in Pulmonary Arterial Hypertension: A Multicentre Study. CJC Open. 2023 Apr 25;5(7):545-553. doi: 10.1016/j.cjco.2023.04.005. PMID: 37496788; PMCID: PMC10366663. https://doi.org/10.1016/j.cjco.2023.04.005

- lead by Nathan Brunner

4.2. Published Abstracts

Moghaddam N, Swiston JR, Weatherald J, Mielniczuk L, Kapasi A, Hambly N, Langleben D, Brunner NW. Impact of saline loading at cardiac catheterization on the classification and management of patients evaluated for pulmonary hypertension. Int J Cardiol. 2020 May 1;306:181-186. https://doi.org/10.1016/j.ijcard.2019.11.104

- lead by Nathan Brunner, Vancouver

Sugarman J, Weatherald J, ThakrarM, Helmersen D, Hirani N, Varughese R, Liu J. Pulmonary Artery Pulsatility Index as a Predictor of Mortality in Pulmonary Arterial Hypertension. CHEST, Volume 158, Issue 4, A2235 - A2236. https://doi.org/10.1016/j.chest.2020.08.1906

- lead by Jason Weatherald, Calgary

Alquraishi H, Swiston J, Lee L, Legkaia L, Norena M, Alobaidellah K, Kapasi K, Levy RD, Brunner NW. The Association Between Median Income and Severity of Pulmonary Hypertension at Diagnosis and Risk at Follow Up in a Public Health Care System. ATS 2022 May 18, 2022 Abstract Presentation. https://doi.org/10.1164/ajrccm-conference.2022.205.1_MeetingAbstracts.A5085

- lead by Nathan Brunner, Vancouver

A McBride, D Helmersen, N Hirani, M Thakrar, M Kularatne, J Liu, L Harper, H Iqbal, A Naser, R Varughese, J Weatherald. Validation of EmPHasis-10 health-related quality of life assessment tool in Canadian patients with pulmonary hypertension. European Respiratory Journal 2022, 60 (suppl 66) 2659; https://doi.org/10.1183/13993003.congress-2022.2659

- lead by Jason Weatherald

Emma E.M. Spence, Brandon Budhram, Doug Helmersen, Mitesh V. Thakrar, Jonathan Liu, Naushad Hirani, Mithum Kularatne, Lea Harper, Jason Weatherald. Evaluating the Transition from Parenteral Prostacyclin Therapy to Oral Selexipag Therapy in Pulmonary Arterial Hypertension: A Single-Center Retrospective Cohort Study. Canadian Respiratory Conference 2023, Montreal, Quebec.

- lead by Jason Weatherald

Brandon Budhram, Emma Spence, Andrea Gardner, Jason Weatherald, John Swiston, Lena Legkaia, Steeve Provencher, Kristina Kemp, George Fox, Julia Foxall, George Chandy, Nathan Hambly. Transitioning Patients with Pulmonary Arterial Hypertension from Parenteral Prostacyclin Therapy to Oral Selexipag: A Multi-center Retrospective Case-Control Study. American Thoracic Society Conference 2023, Washington, D.C. https://www.atsjournals.org/doi/pdf/10.1164/ajrccm-conference.2023.207.1_MeetingAbstracts.A6447

- lead by Nathan Hambly

Amanda Cheung MD, Miles Marchand MD, Lisa Kolkman NP, John Swiston MD, FRCPC, Ali Kapasi MD, FRCPC, Marion Brown, Jason Weatherald MD, MSc, FRCPC, and Nathan W Brunner MD, FRCPC Severity of Illness in Indigenous Patients with Pulmonary Arterial Hypertension in Canada. Abstract Poster, Vascular 2023 Conference, Oct.25-29, 2023, Montreal, Quebec

- lead by Jason Weatherald and Nathan Brunner

4.3 Ongoing projects

There are a number of research projects in various stages of completion that utilize some of the collected registry data.

Participating centres		Title	Notes	Lead Author
Vancouver Calgary	Ottawa Hamilton	“Pulmonary Artery Pulsatility Index as a Predictor of Morbidity and Mortality in Pulmonary Hypertension” – multicentre project	- *analyzing data*	Jason Weatherald Nathan Brunner
Hamilton Calgary Halifax Vancouver Ottawa	Quebec City Saskatoon Moncton Montreal	"Transitioning from parenteral prostacyclin therapy to oral selexipag in pulmonary arterial hypertension: A multi-centre retrospective chart review"	- *data collected* - *analyzing data*	Nathan Hambly
Vancouver Calgary St. John’s	Edmonton Moncton	“Severity of Illness and Outcomes in Indigenous patients with Pulmonary Arterial Hypertension in Canada”	- *sites undergoing ethics application review* - *analyzing Vancouver Data*	Nathan Brunner
Vancouver Hamilton	Calgary	Evaluation of Centralized Care in a Major Canadian Pulmonary Hypertension Centre	- *sites collecting data*	Nathan Brunner
Calgary Hamilton Winnipeg	Vancouver Edmonton Halifax	Access and barriers to exercise rehabilitation programs for pulmonary hypertension in Canada: a patient survey	- *site collecting data*	Jason Weatherald
Vancouver Edmonton London	Calgary Hamilton Quebec	Outcomes in patients with pulmonary arterial hypertension transitioned from selexipag to parental prostacyclin analogues	- *sites undergoing ethics*	John Swiston

5. Future Plans

CPHR plans going forward are to continue robust data collection at participating sites, as well as continually add new interested sites. Establish more pediatric sites and start utilizing their data in research projects. Moreover, continue utilizing existing data in answering specific research and quality improvement questions.