Canadian Pulmonary Hypertension Registry Annual Report

Version: 1.0

Reporting Timeframe: 01 July 2025 – 30 June 2026

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1. OVERVIEW

Canadian Pulmonary Hypertension Registry (CPHR) is a multicentre, 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 and pediatric 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 12 adult and 3 pediatric active centres that are entering patient data at their centres.

2. PARTICIPATING CENTRES STATUS

Centre 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. George Chandy	ongoing data entry since 01Apr2018
Halifax	Dr. Paul Hernandez	ongoing data entry since 01Aug2019
Moncton/Saint John	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. Danny Wadden	ongoing data entry since 15Oct2021 (start date is 01July2020)
BC Children's - Vancouver (pediatric)	Dr. Iman Naimi	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 Mertens	ongoing data entry since 01Feb2026
Sainte-Justine – Montreal (pediatric)	Dr. Anne Fournier	ongoing data entry since 01Feb2025
Kingston	Dr. Stephen Archer	ethics approval 16May2025

3. DATA COLLECTED

Data below is cumulative data entered into the registry database across all participating sites from inception **01Jan2017** to **30June2026.**

***Some sites are still in the process of completing their data entry completeness checks. As a result, the figures in this annual report may not fully represent data collected up to June 30, 2026.

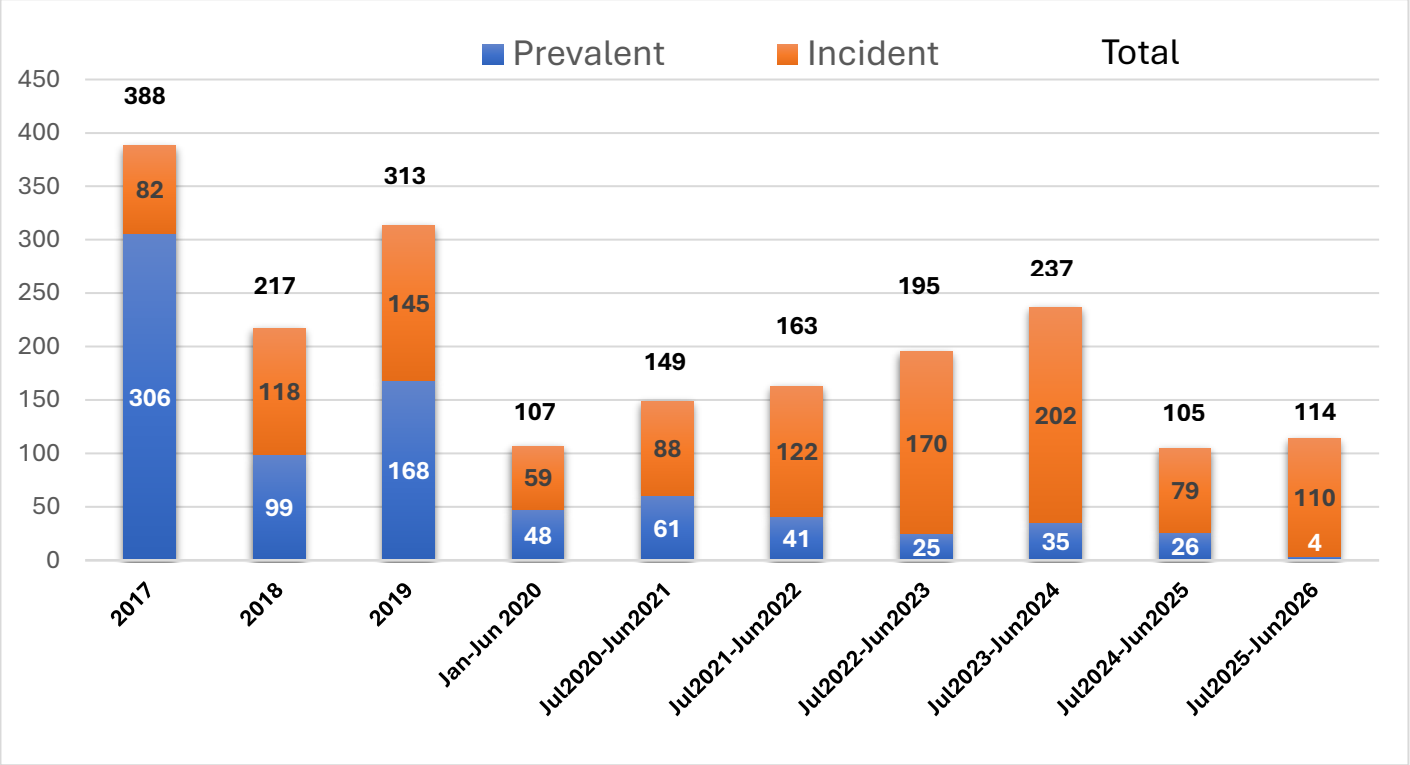
3.1. Patients entered in the Registry

Site	Patients Entered	PAH	CTEPH
Vancouver	*1712	459	162
Hamilton	248	182	35
Calgary	521	246	125
Ottawa	184	146	32
Halifax	34	19	1
Moncton/Saint John	140	48	6
Winnipeg	131	104	24
Quebec	720	280	55
Edmonton	43	17	5
Toronto	22	18	1
St. John's	15	8	4
BC Children's Hospital; Vancouver	**	**	**
Sainte-Justine Hospital; Montreal	**	**	**
The Hospital for Sick Children; Toronto	**	**	**
TOTAL (adult sites)	**3770**	**1527**	**450**

* Vancouver site entered all WHO groups into the database from 01Jan2017 to 31Dec2024.
** Pediatric site numbers are currently withheld due to small population size.

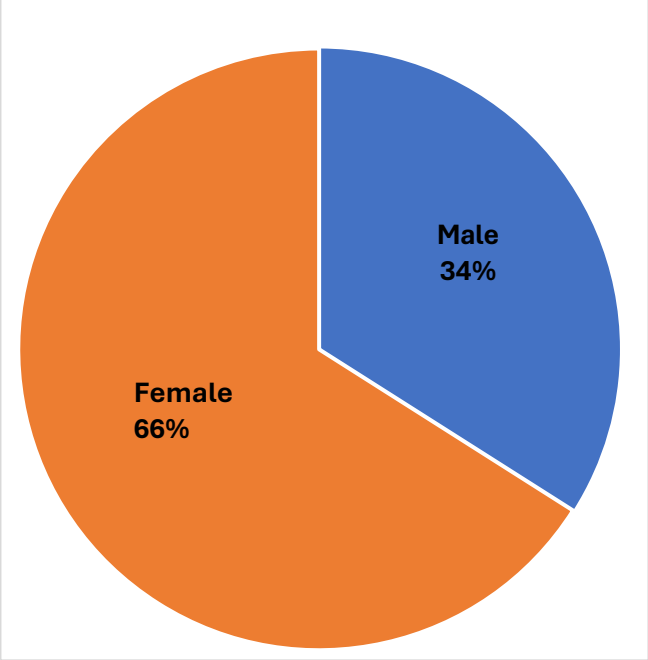
3.2. Incident and Prevalent

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.

3.3. Sex and Age

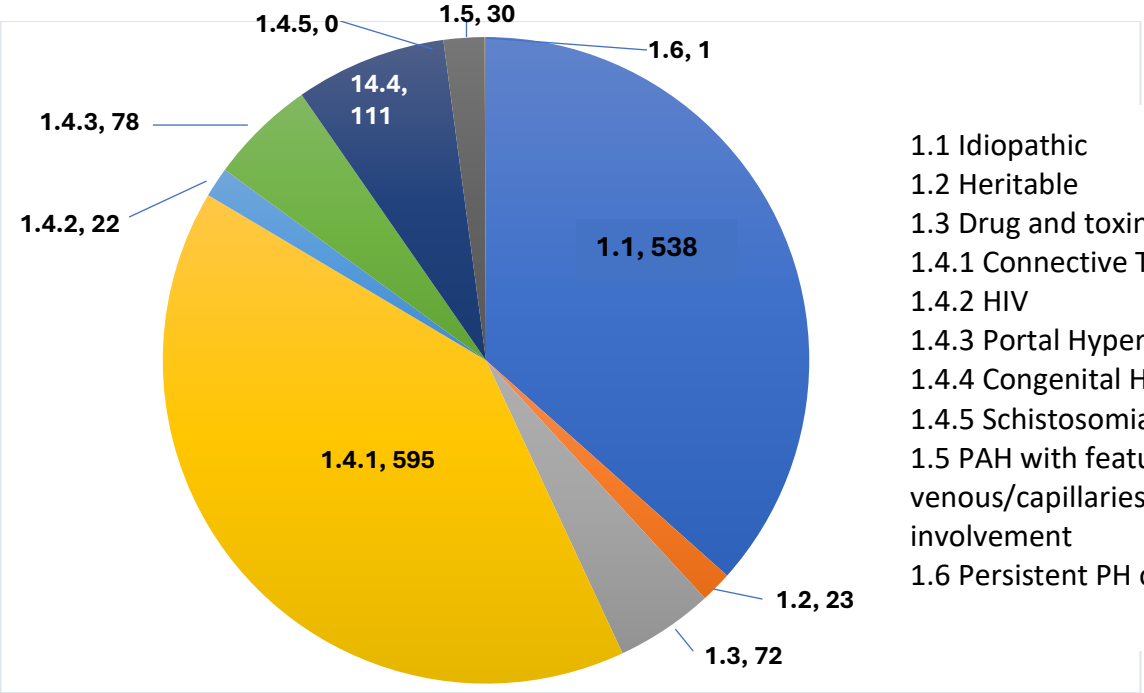


59.29
Age (Diagnosis)

69.70
Age (Current)

3.4. Group 1 Specifics

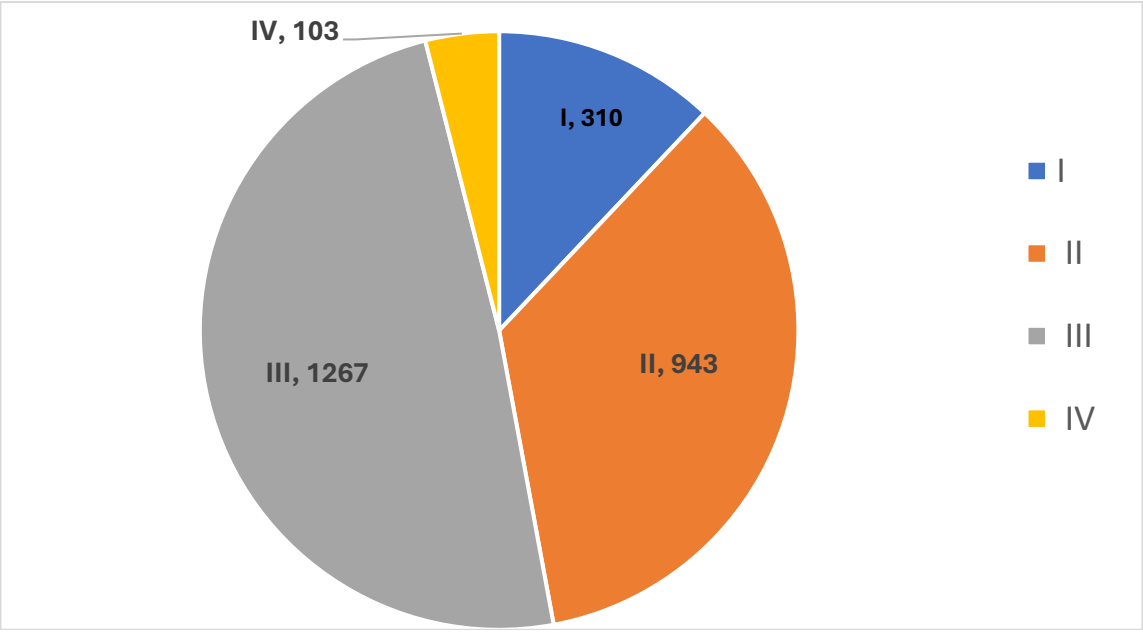
Breakdown of clinical classification of Group 1 PH.



* Some participants have multiple Group 1 specific classifications marked; therefore, the total number of Group 1 specifics does not equal the total number of PAH.

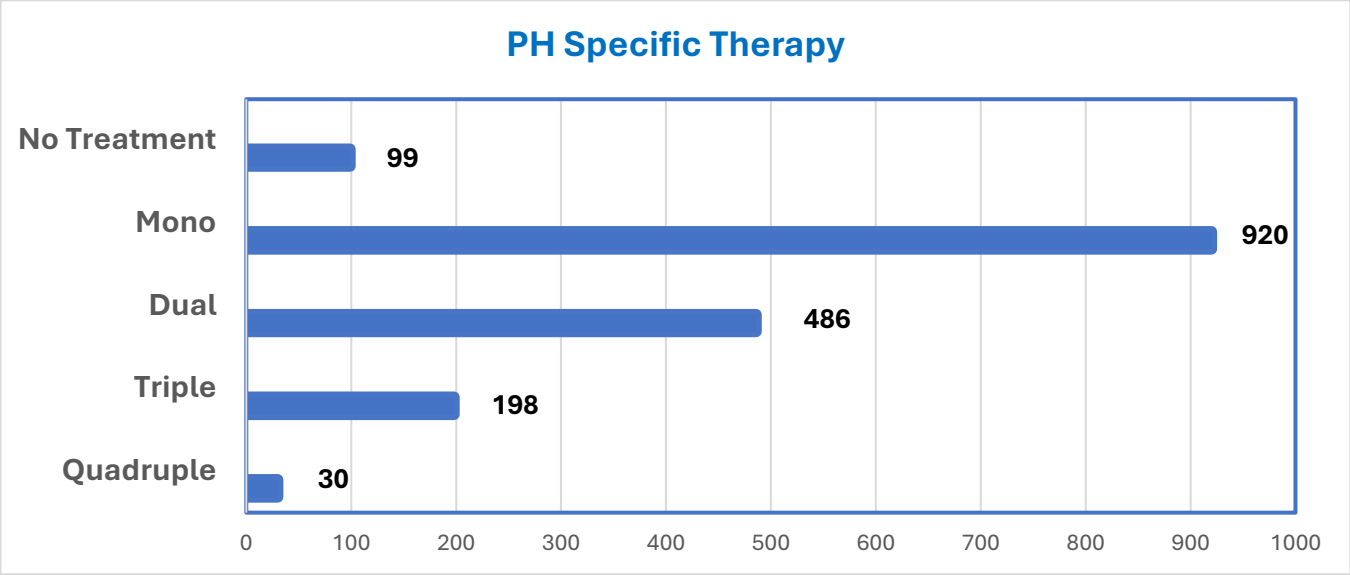
3.5. WHO FC Groups

Breakdown of the WHO FC for all patients.



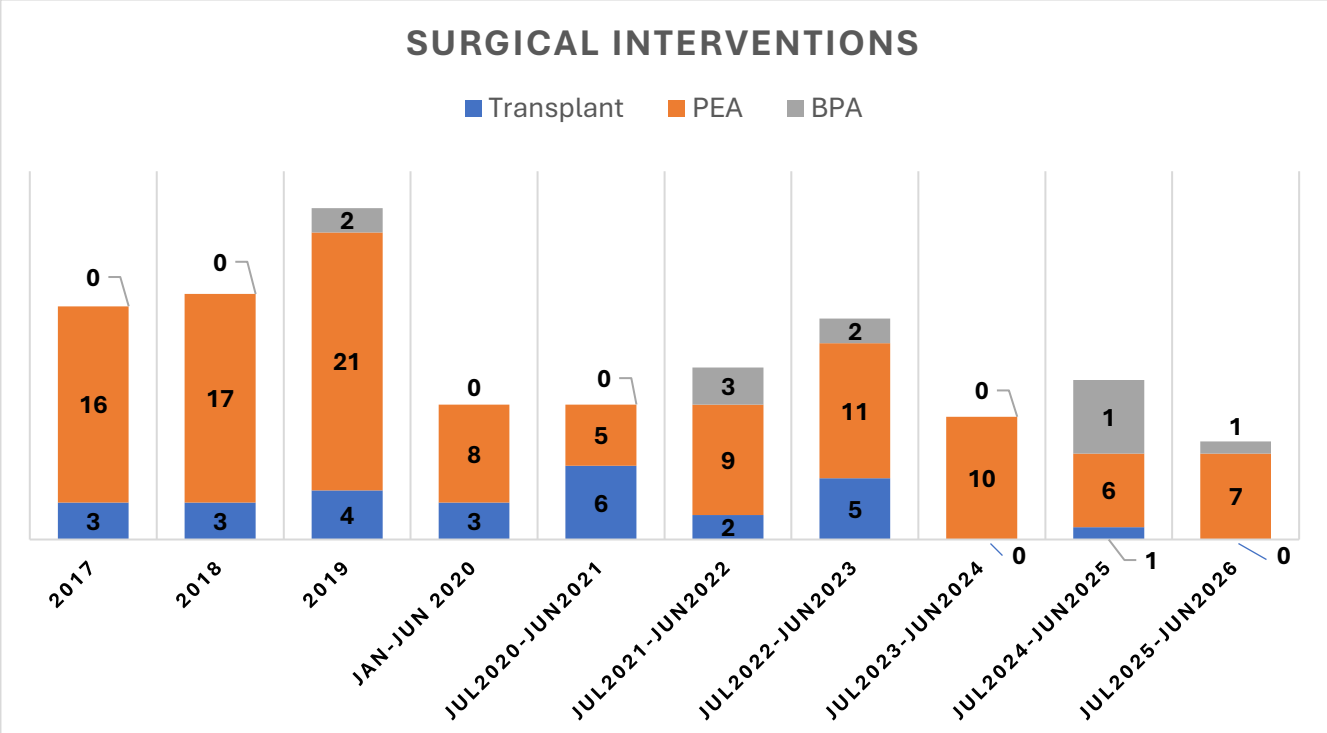
3.6. PH Specific Therapies

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



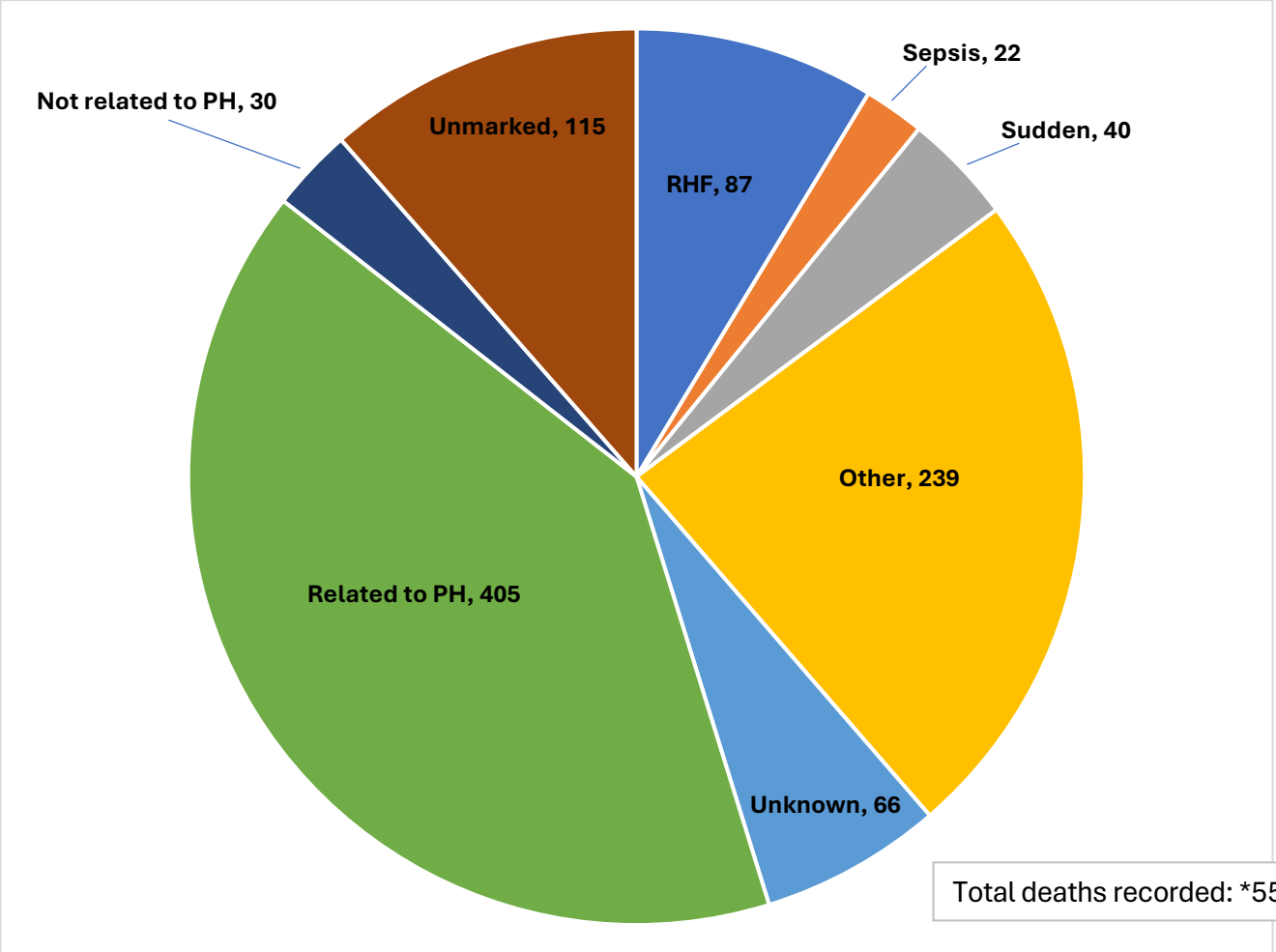
3.7. Transplants, PEA and BPA

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



3.8. Cause of Death

Breakdown of the cause of death of deceased patients.



"Unmarked" denotes cases where the "death related to PH" or "death not related to PH" field was left blank in the database.

"Other" causes of death include, but are not limited to, cancer, pneumonia, congestive heart failure/disease, liver failure/disease, palliative, hypoxemia, respiratory failure, covid, renal failure/disease, AECOPD/COPD, PE, cirrhosis, fall, MAID, PVOD, pulmonary fibrosis, post-surgery complications, aspiration, interstitial lung disease, GI bleed, influenza, biventricular failure, and hypoxia.

* Data transfer inaccuracies were identified and corrected following the data completeness check.

Not all deaths were attributed to a single cause; therefore, the total number of reported causes may not equal the total number of deaths.

4. DATA USE AND RESEARCH

The following published studies utilized data from the CPHR registry.

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

Budhram B, Alrasheed SK, Rashad M, Chandy G, Dvorkin-Gheva A, Fox GA, Foxall J, Gardner A, Helmersen D, Hirani N, Kemp K, Kularatne M, Langleben D, Lesenko L, Provencher S, Spence E, Swiston JR, Thakrar M, Wadden D, Weatherald J, Hambly N. Transitioning From Parenteral Prostanoids to Oral Selexipag in Pulmonary Arterial Hypertension: A Multicenter Retrospective Cohort Study. Pulm Circ. 2025 Dec 5;15(4):e70220. doi: 10.1002/pul2.70220

- lead by Nathan Hambly

Renee Reimer, John R. Swiston, Carter McInnis, Freda Tom, Mitesh V. Thakrar, Naushad Hirani, Doug Helmersen, Nathan Hambly, Jason Weatherald, Monica Norena, Nathan W. Brunner. Effect of Community Size and Commute Time on Severity of Pulmonary Arterial Hypertension at Diagnosis and Follow-up in Patients Across 3 Sites. CJC Open. 27 April 2026 https://doi.org/10.1016/j.cjco.2026.04.014

- lead by Nathan Brunner

Ebrahim AlEissa, MD, Lisa Kolkman, NP(F), Ali Kapasi, MD, Defen Peng, PhD, Steeve Provencher, MD, MSc, Nathan Brunner, MD, John Swiston, MD, MPH. Outcomes of Transition from Selexipag to Parenteral Prostacyclin Analogues in Patients with Pulmonary Arterial Hypertension. ERJ Open Research 2026 12(5): 00131-2026; DOI: https://doi.org/10.1183/23120541.00131-2026

- lead by John Swiston

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 several research projects at various stages of completion that utilize registry data.

Participating centres		Title	Notes	Lead Author
Vancouver Calgary	Edmonton Ottawa	*"Severity of Illness and Outcomes in Indigenous patients with Pulmonary Arterial Hypertension in Canada"*	*-other participating sites have/will be submitting to ethics*	Nathan Brunner Jason Weatherald
Vancouver		*"Radiologic Alignment in Chronic Pulmonary Thromboembolism: Retrospective Study from Real-World Data"*	*-data collection completed*	Nathan Brunner
Vancouver		*"Association between echocardiographic markers of increased left ventricular filling pressures at baseline and response to pulmonary vasodilator therapy in patients with pulmonary arterial hypertension"*	*-collecting data*	Nathan Brunner
Quebec Vancouver	Other sites to be determined	*"Ethnicity and immigration status as social determinants of health for patients with pulmonary hypertension (PH): an international comparison study for Canadian PH patients of East and/or South-east Asian ethnicity ('PH-ESASIAN')"*	*-main site received ethics approval*	Steeve Provencher

5. Future Plans

CPHR's future plans include continuing robust data collection at participating sites while expanding participation by recruiting new sites. Efforts will also focus on establishing additional pediatric sites and incorporating pediatric data into research projects. Furthermore, CPHR will continue to leverage existing data to address targeted research and quality improvement questions.