

Canadian Pulmonary Hypertension Registry Annual Report

Version: 1.0, 23 September 2021

Reporting Timeframe: 01 July 2020 – 30 June 2021

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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 centers 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 8 active sites that are entering patient data at their centers. Additional 5 centers are in various stages of start-up, and 2 pediatric centers also in start-up stages.

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. Jason Weatherald	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 (SBGH)	Dr. David Christiansen	ongoing data entry since 01Sep2020
Québec City	Dr. Steeve Provencher	ongoing data entry since 01Jul2021
Toronto	Dr. John Granton	finalizing internal processes
Edmonton	Dr. Rhea Varughese	finalizing internal processes
St. John's	Dr. George Fox	finalizing internal processes
London	Dr. Sanjay Mehta	obtaining ethics approvals
Kingston	Dr. Christine D'Arsigny	preparing ethics application
BC Children's (pediatric)	Dr. Martin Hosking	obtaining ethics approvals
SickKids – Toronto (pediatric)	Dr. Luc Martens	obtaining ethics approvals

3. DATA COLLECTED

Data below is cumulative data entered into the registry database across all participating sites from inception **01Jan2017** to **30June2021**.

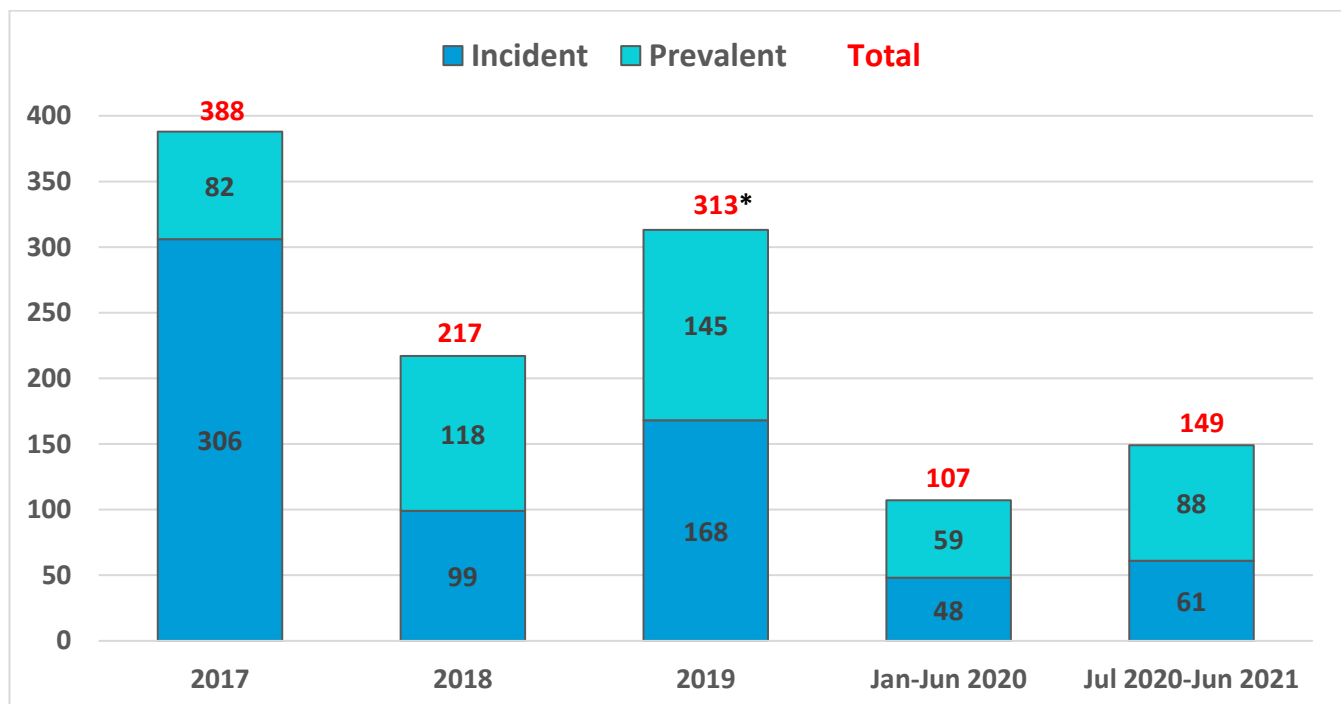
3.1. Patients in the Registry with Confirmed Diagnosis

Site	Patients Entered	PAH	CTEPH
Vancouver	939*	429	132
Hamilton	171	127	24
Calgary	207	148	54
Ottawa	140	118	21
Halifax	15	13	1
Moncton	46	31	6
Winnipeg (SBGH)	35**	-	-
TOTAL	1553	866	238

* Vancouver site enters all WHO groups into the database

** Data has not yet been entered on consented patients.

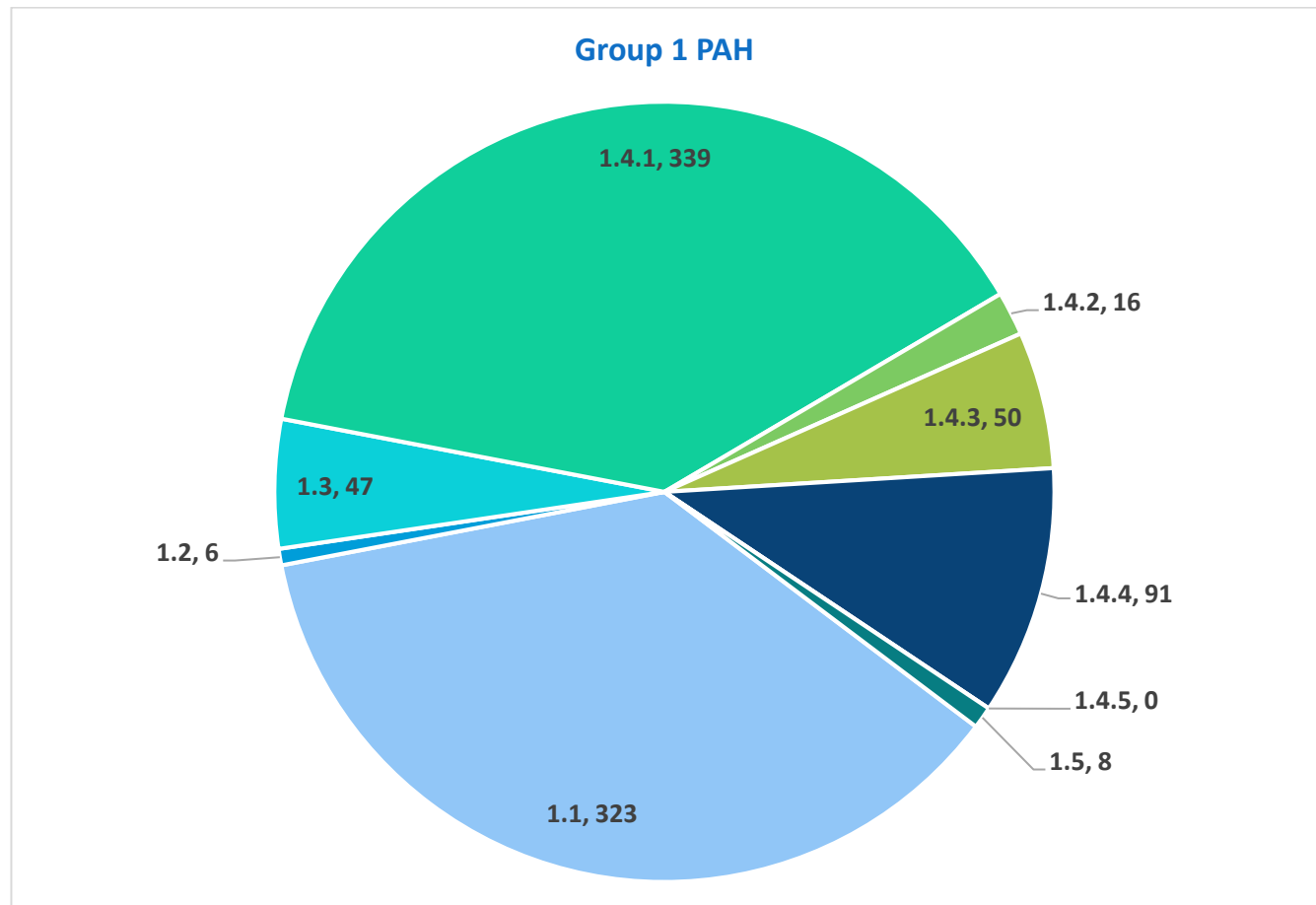
Figure below represents total number of incident and prevalent patients that have been entered into the registry database.



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

3.2. Group 1 Specifics

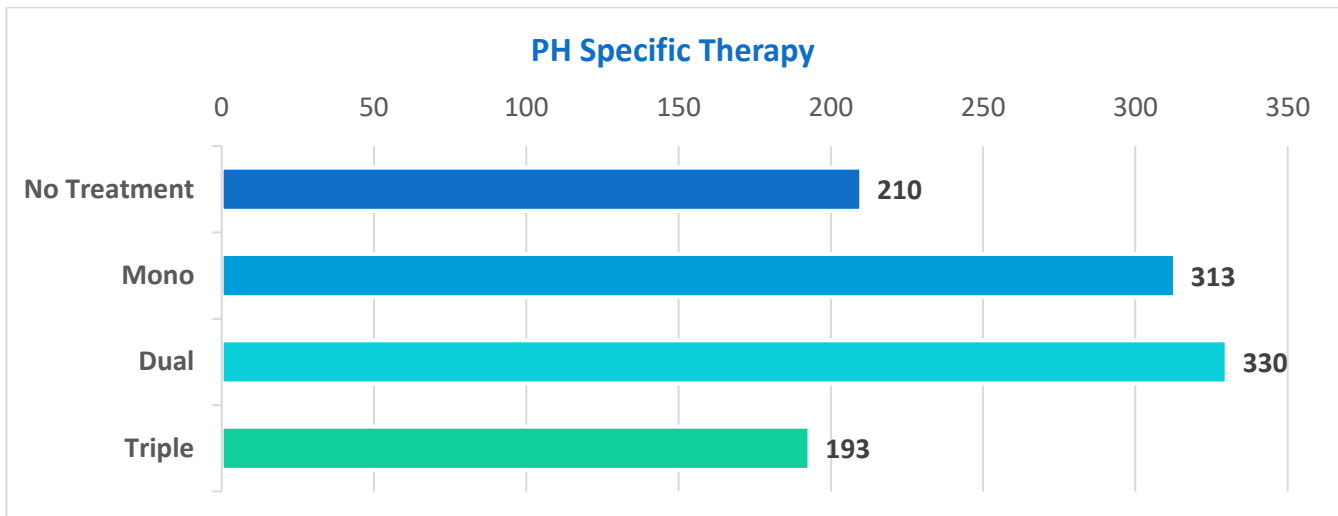
Breakdown of clinical classification of Group 1 PH.



- 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

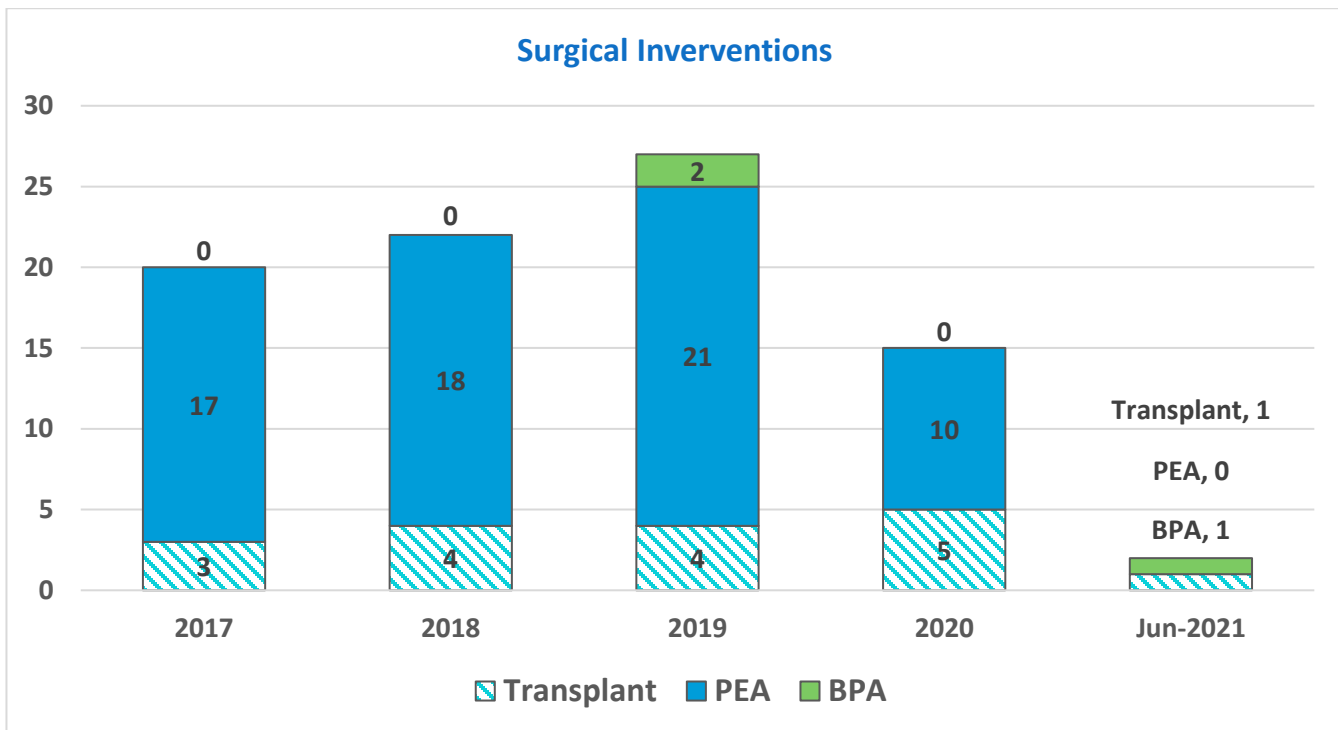
3.3. PH Specific Therapies

PH specific therapies approved in Canada: Flolan, Caripul, Remodulin, Uptravi, Tracleer, Tracleer generic, Volibris, Opsumit, Revatio, Revatio generic, Adcirca, Adempas. Figure below depicts treatment combination distribution of PH specific therapy.



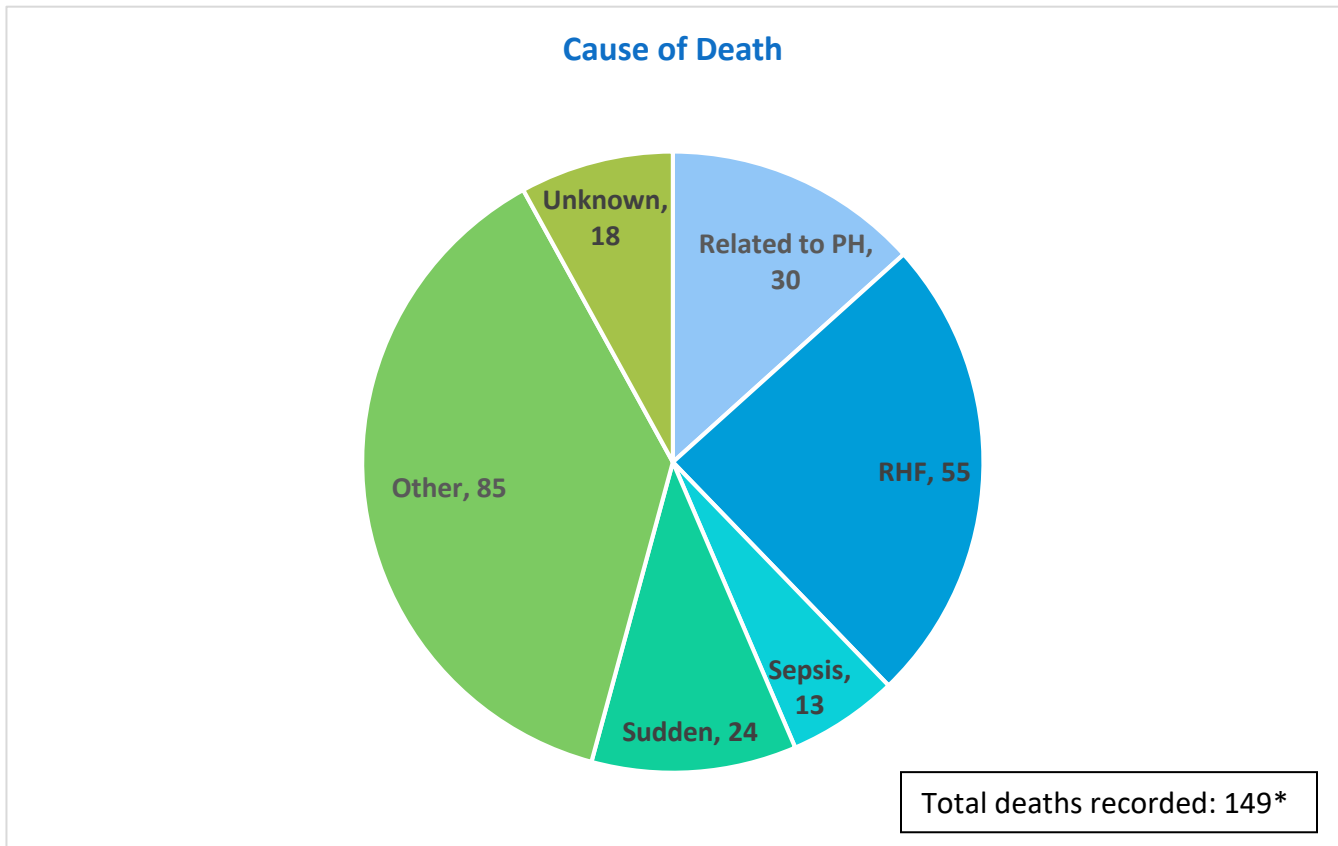
3.4. Transplants, PEA and BPA

Number of Transplants, PEA and BPA surgeries performed on patients across all sites.



3.5. Cause of Death

Breakdown of the cause of death of deceased patients.



“Other” death causes include, but are not limited to, congestive heart failure, atrial fibrillation, biventricular failure, bronchiolitis, cancer, COPD exacerbation, GI bleed, hypoxemia, interstitial lung disease, multisystem organ failure, palliative, pneumonia, PVOD, renal failure, respiratory failure, advanced sarcoidosis, urosepsis.

*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

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

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, Thakrar M, 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

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 centers	Title	Notes	Lead Author
Toronto, et al.	“Outcome of pulmonary endarterectomy according to the level of disease on preoperative CT pulmonary angiogram and at surgery”	- <i>manuscript is under review in the Journal of Thoracic and Cardiovascular Surgery</i>	Marc DePerrot
Calgary Ottawa Vancouver Quebec City Hamilton Halifax Winnipeg	“The EmPHasis-10 (E10) tool in Canadian patients with pulmonary hypertension: Validation and determining the minimal clinically important difference”	- <i>sites undergoing ethics application review</i>	Jason Weatherald
Vancouver Ottawa Calgary Hamilton	“Pulmonary Artery Pulsatility Index as a Predictor of Morbidity and Mortality in Pulmonary Hypertension” – multicenter project	- <i>analyzing data</i>	Jason Weatherald Nathan Brunner
Ottawa Calgary	“Redefining risk prediction in patients with Pulmonary Arterial Hypertension Heart Failure Database Retrospective Data Analysis”	- <i>analyzing data</i>	Lisa Mielniczuk
Hamilton Calgary Halifax Vancouver Ottawa	Quebec City Saskatoon Moncton Montral	“Transitioning from parenteral prostacyclin therapy to oral selexipag in pulmonary arterial hypertension: A multi-center retrospective chart review”	- <i>sites undergoing ethics application review</i> Nathan Hambly
Vancouver Calgary	Hamilton	“Does community size or commute time affect severity of illness at diagnosis or quality of care in a centralized care model of pulmonary hypertension?”	- <i>multicenter project</i> - <i>sites undergoing ethics application review</i> Nathan Brunner

5. FUTURE PLANS

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