

# Canadian Pulmonary Hypertension Registry Annual Report

**Version:** 1.0, 14 October 2020

**Reporting Timeframe:** 01 January 2017 – 30 June 2020

**Table of Contents**

1. OVERVIEW ..... 3

2. PARTICIPATING CENTERS STATUS ..... 3

3. DATA COLLECTED..... 4

    3.1. Patients in the Registry with Confirmed Diagnosis..... 4

    3.2. Group 1 Specifics..... 5

    3.3. PH Specific Therapies ..... 6

    3.4. Transplants, PEA and BPA ..... 6

    3.5. Cause of Death ..... 7

3. DATA USE AND RESEARCH..... 8

4. FUTURE PLANS..... 10

## 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 7 active sites that are entering patient data at their centers. Additional 5 centers 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
Calgary	Dr. Jason Weatherald	ongoing data entry since 01Oct2017
Hamilton	Dr. Nathan Hambly	ongoing data entry since 01Jan2018
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	finalizing internal processes
Toronto	Dr. John Granton	finalizing legal/ethics approvals
Edmonton	Dr. Ali Kapasi	obtaining ethics approvals
St. John's	Dr. George Fox	obtaining ethics approvals
London	Dr. Sanjay Mehta	obtaining ethics approvals

### 3. DATA COLLECTED

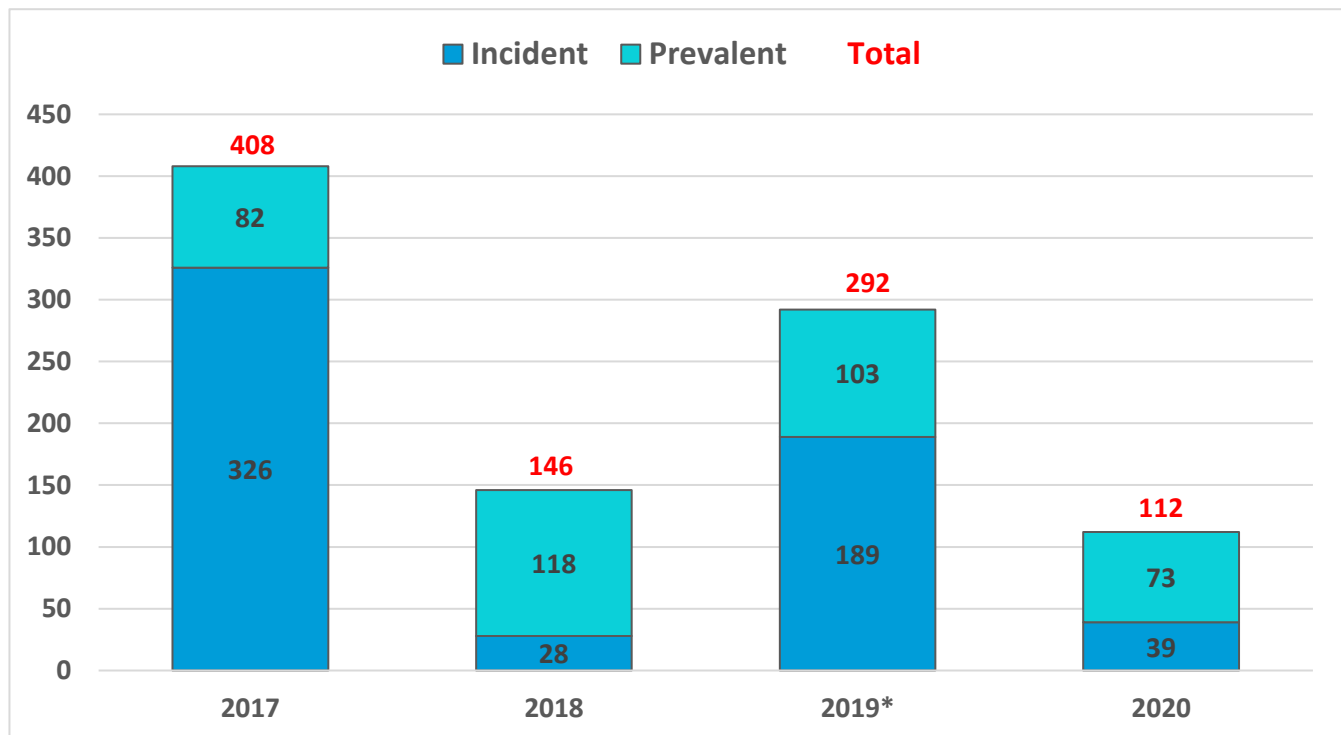
Data below is cumulative data entered into the registry database across all participating sites from inception **01Jan2017 up until 30June2020**.

#### 3.1. Patients in the Registry with Confirmed Diagnosis

Site	Patients Entered	PAH	CTEPH
Vancouver	811*	364	119
Calgary	166	121	42
Hamilton	148	106	21
Ottawa	134	113	20
Halifax	9	9	0
Moncton	24	12	6
<b>TOTAL</b>	<b>1292</b>	<b>725</b>	<b>208</b>

\* Vancouver site enters all WHO groups into the database

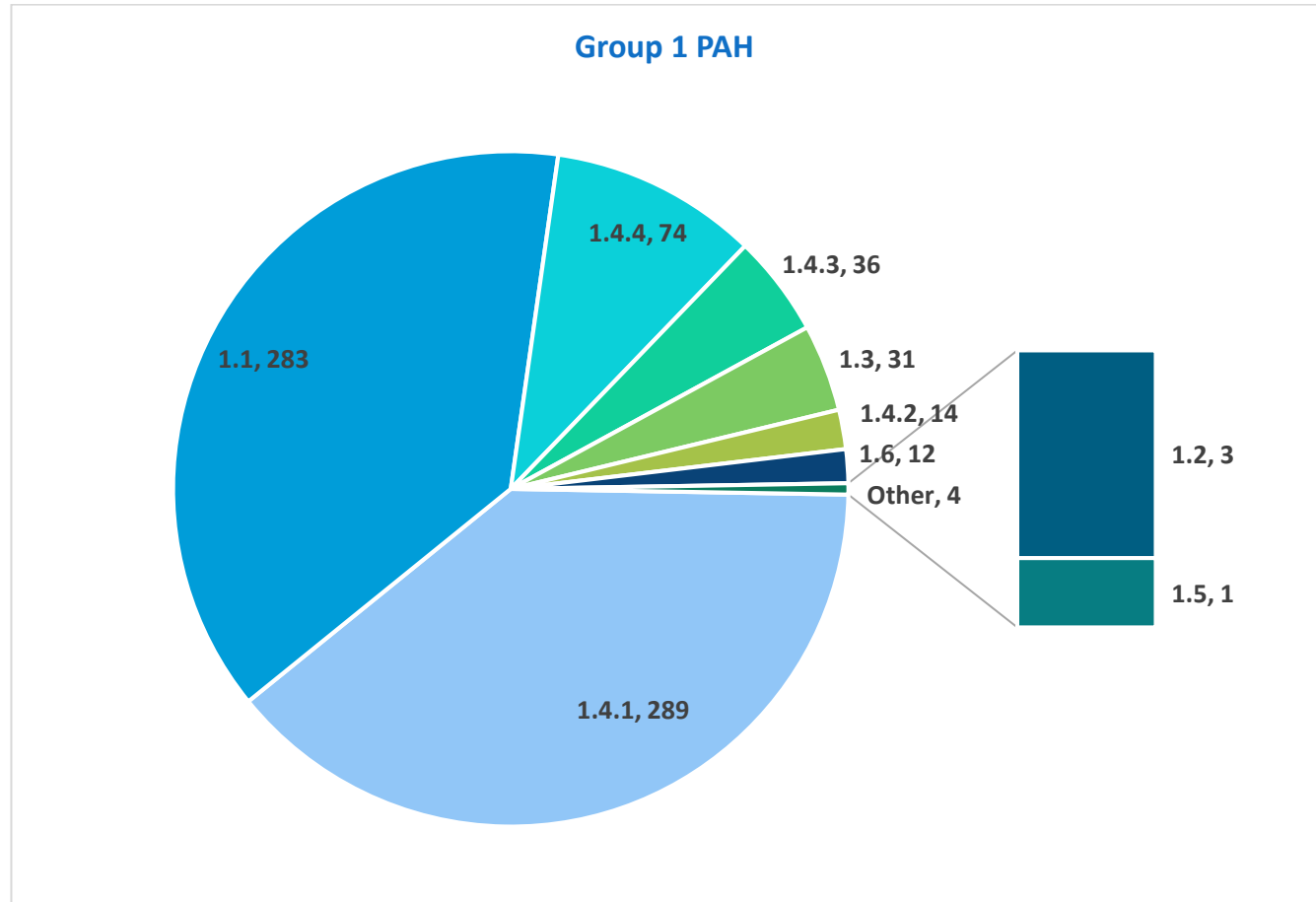
Figure below represents total number of incident and prevalent patients that have been entered into the registry database.



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

### 3.2. Group 1 Specifics

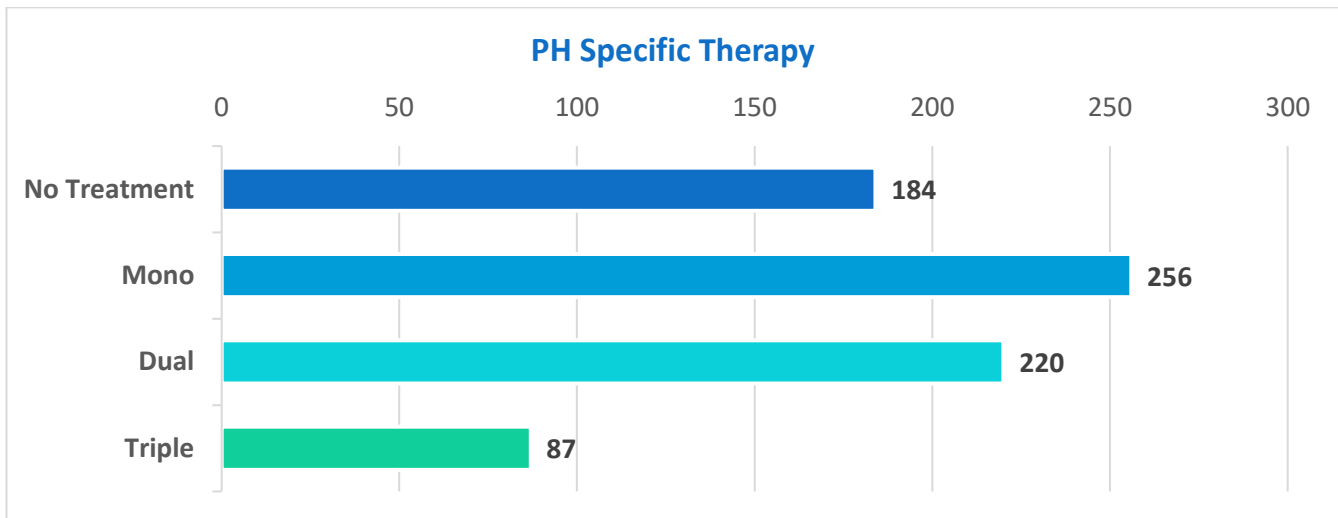
Breakdown of clinical classification of Group 1 PH.



- 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

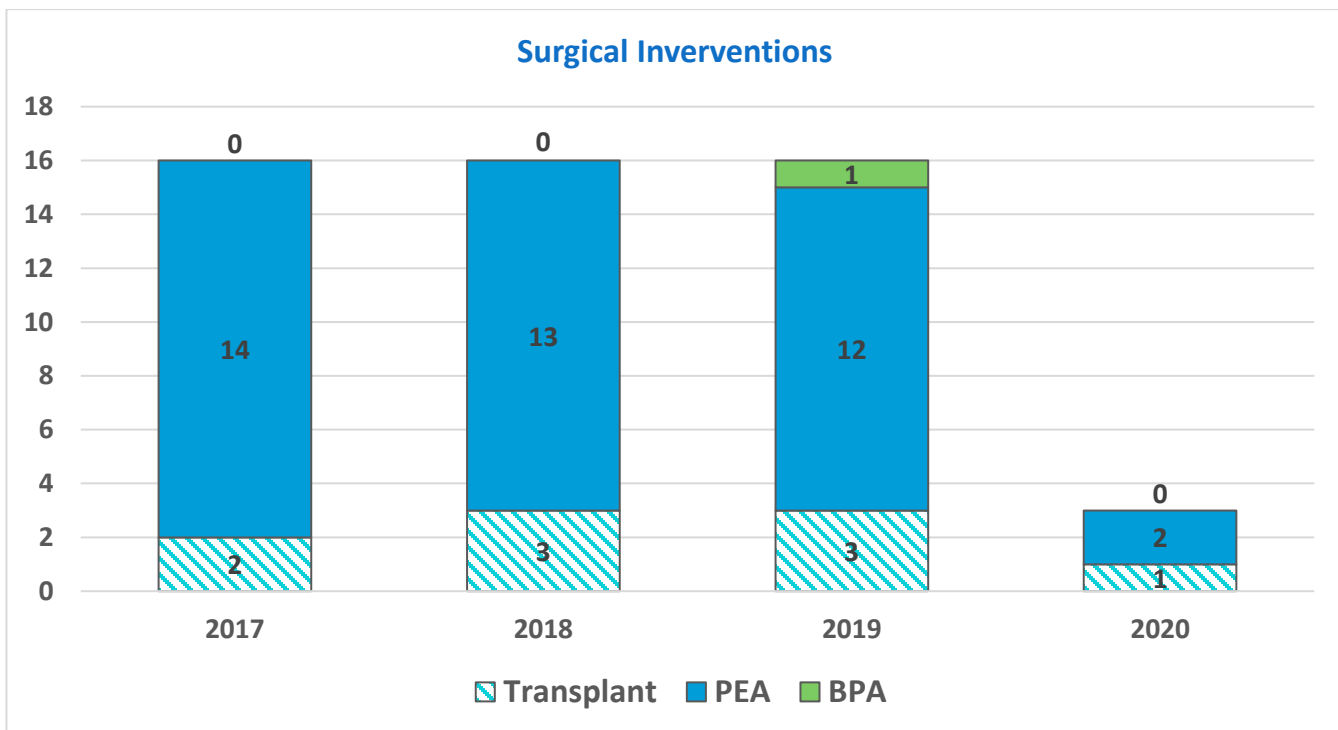
### 3.3. PH Specific Therapies

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.



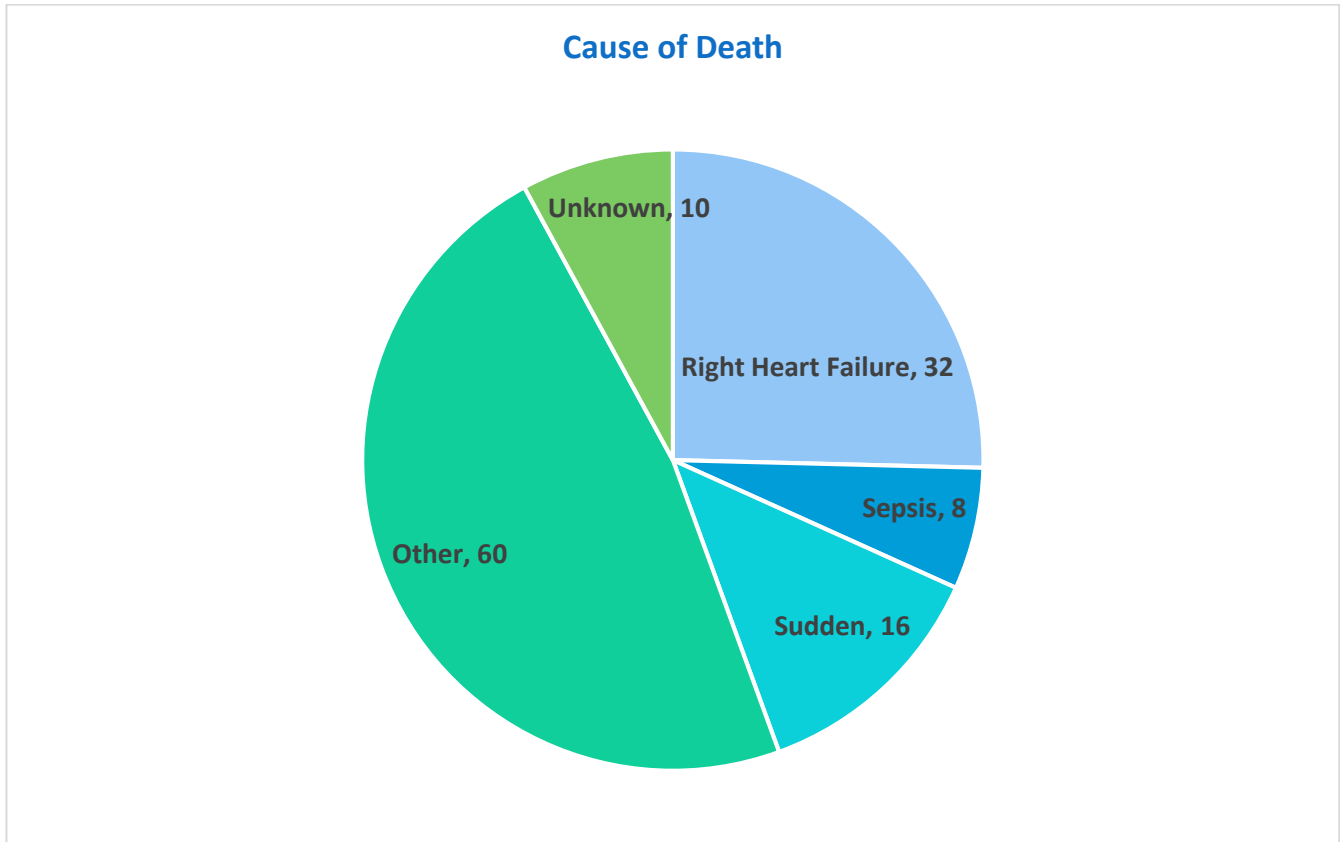
### 3.4. Transplants, PEA and BPA

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, interstitial lung disease, renal failure, PVOD, palliative, pneumonia, COPD exacerbation, severe hypoxemia.

### 3. DATA USE AND RESEARCH

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

Participating centers	Journal and Date of submission/publication	"Title" Notes	Authors/Involved researchers
Vancouver	ISHLT April 22-25, 2020 - <i>cancelled</i>  6-Jun-2019/10-Dec-2019	"Impact of Targeted Pulmonary Arterial Hypertension Therapy in Patients with Combined Pre- and Post-Capillary Pulmonary Hypertension"  <i>Abstract accepted as a moderated poster at the ACC's 69th Annual Scientific Session together with the World Congress of Cardiology, March 28 - 30, 2020, in Chicago, IL.</i>	Nathan Brunner Nima Moghaddam Michael Tsang Lisa Lee Monical Norena John Swiston
Vancouver	International Journal of Cardiology  Published 9-Dec-2019	"Impact of saline loading at cardiac catheterization on the classification and management of patients evaluated for pulmonary hypertension"  <i><a href="https://doi.org/10.1016/j.ijcard.2019.11.104">https://doi.org/10.1016/j.ijcard.2019.11.104</a></i>	Nima Moghaddaman John R. Swiston Jason Weatherald Lisa Mielniczuk Ali Kapasi Nathan Hambly David Langleben Nathan Brunner
Vancouver	ATS Abstract - Presentation 15-20 May, Philadelphia - <i>cancelled</i>  Submitted for publication in International Journal of Cardiology 02-Jul-2020	"Does Community Size or Commute Time Affect Severity of Illness at Diagnosis or Quality of Care in a Centralized Care Model of Pulmonary Hypertension?"	Nathan W Brunner Lisa Lee Lena Legkaia Connie Sherba Kelly Kerwin Charmaine Lam Fayez Al-Ahmadi Jason Weatherald Roland Nador Robert D Levy John R Swiston



Participating centers	Journal and Date of submission/publication	"Title" Notes	Authors/Involved researchers
Calgary Vancouver Hamilton		"Using the EmPHasis-10 tool in Canadian patients with pulmonary hypertension: validation and determining the minimal clinically important difference"  <i>- Protocol is not yet finalized</i>	Jason Weatherald
Vancouver Calgary Ottawa Hamilton Toronto		"Pulmonary artery capacitance index and pulmonary artery pulsatility index as novel hemodynamic parameters for predicting prognosis in patients with pulmonary arterial hypertension"  <i>- obtaining ethics approvals and DTAs</i>	Abdullah Alkhodair John Swiston Robert Levy Jason Weatherald Nathan Brunner
Calgary	Accepted as poster at the CHEST Annual Meeting 2020, Chicago, IL October 18-21	"Pulmonary Artery Pulsatility Index (PAPi) as a Predictor of Mortality in Pulmonary Arterial Hypertension"	Jordan Sugarman Mitesh Thakrar Doug Helmersen Naushad Hirani Jonathan Liu Rhea Varughese Jason Weatherald
Ottawa Calgary Vancouver		"Redefining risk prediction in patients with Pulmonary Arterial Hypertension Heart Failure Database Retrospective Data Analysis"  <i>- obtaining DTAs</i>	Lisa Mielniczuk Jason Weatherald John Swiston

#### 4. 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.