

Canadian Pulmonary Hypertension Registry Annual Report

Version: 1.0, 13 September 2022

Reporting Timeframe: 01 July 2021 – 30 June 2022

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

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. 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
Edmonton	Dr. Rhea Varughese	ongoing data entry since 15Oct2021
St. John's	Dr. George Fox	ongoing data entry since 15Oct2021 (start date is 01July2020)
BC Children's (pediatric)	Dr. Martin Hosking	ongoing data entry since 01Jul2022 (start date is 01May2021)
Toronto	Dr. John Granton	have approvals, finalizing internal processes
London	Dr. Sanjay Mehta	obtaining ethics approvals
SickKids – Toronto (pediatric)	Dr. Luc Martens	obtaining ethics approvals
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 **30June2022**.

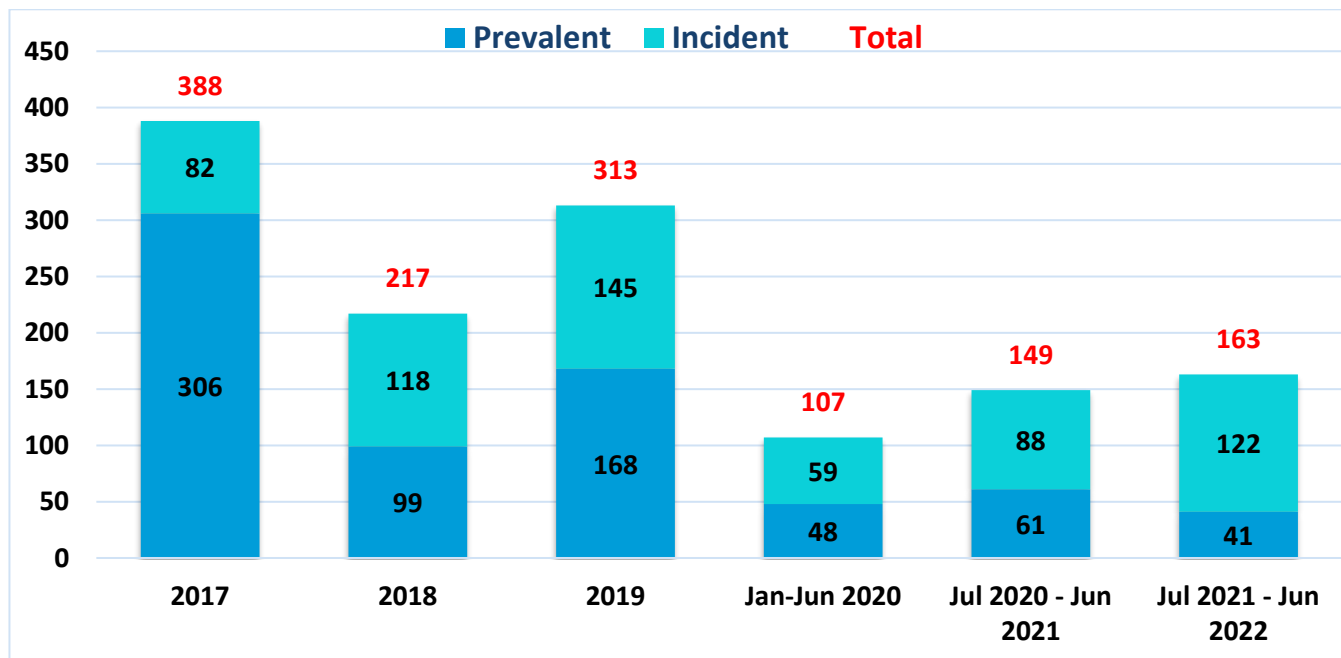
3.1. Patients in the Registry with Confirmed Diagnosis

Site	Patients Entered	PAH	CTEPH
Vancouver	1053*	474	140
Hamilton	189	142	26
Calgary	292	205	79
Ottawa	154	127	26
Halifax	25	20	2
Moncton	63	41	9
Winnipeg (SBGH)	60**	-	-
Quebec	117	65	7
Edmonton	2	2	0
TOTAL	1995	1076	289

* Vancouver site enters all WHO groups into the database

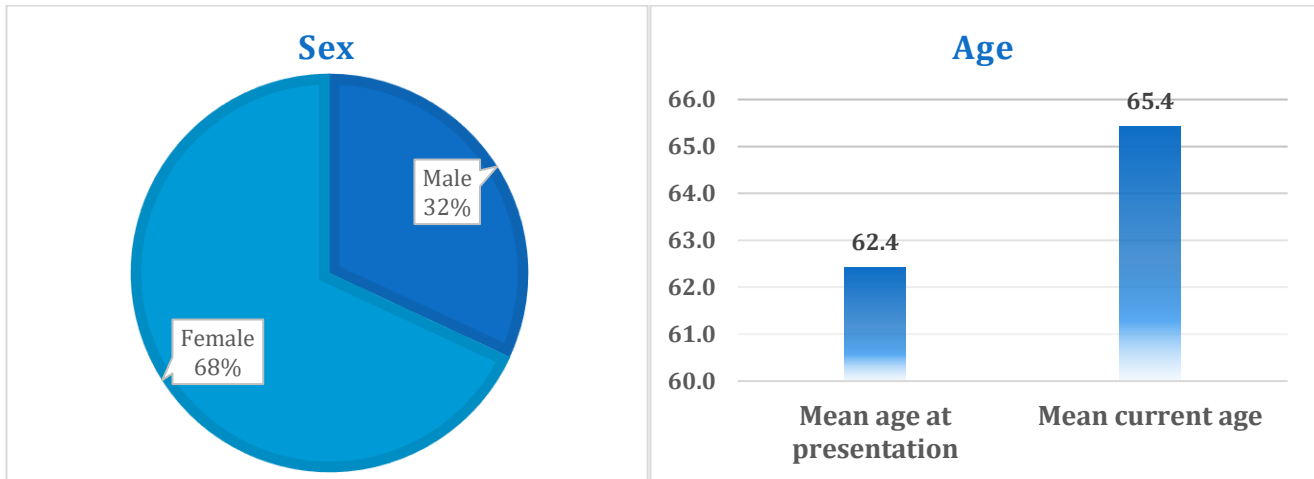
** Data has not yet been entered on consented patients.

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



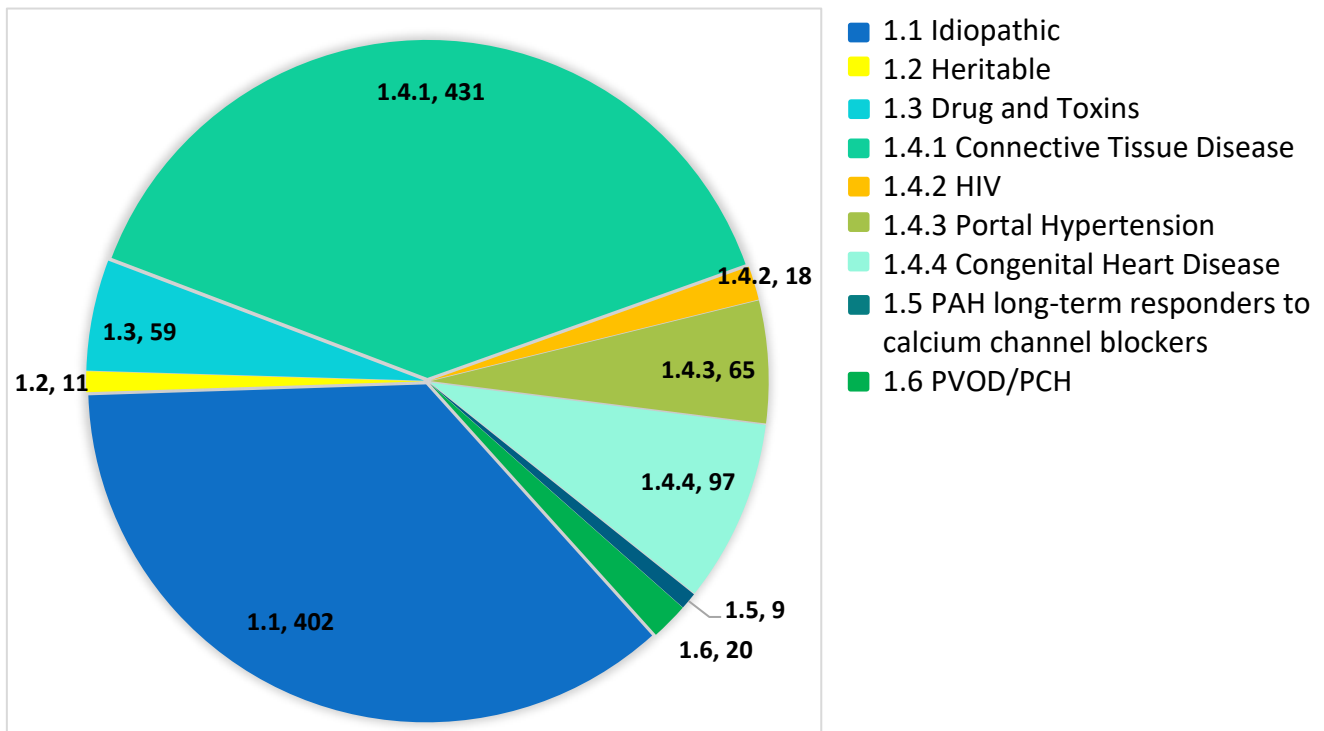
* Higher numbers are attributed to addition of new sites to the registry.

3.2. Sex and Age



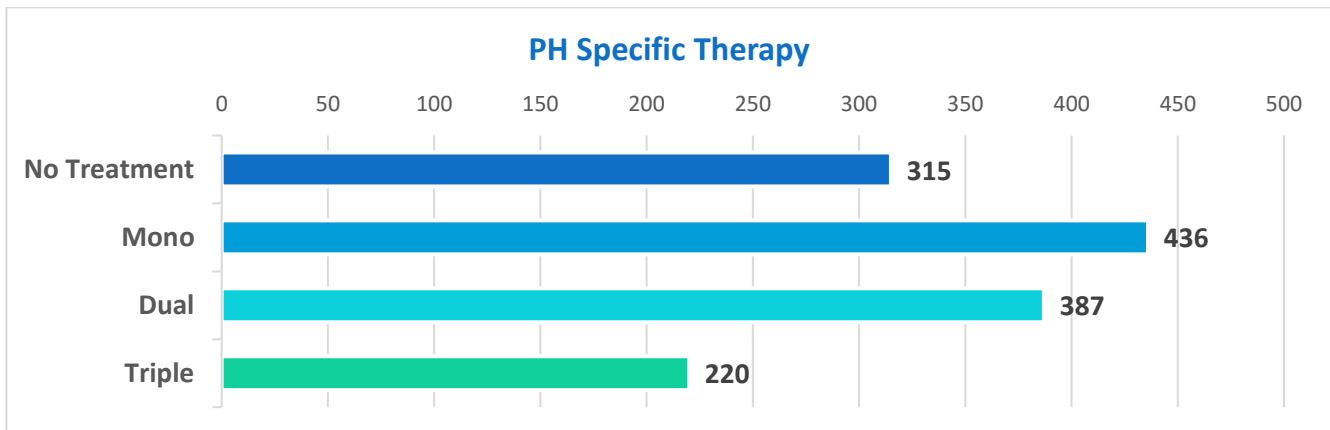
3.3. Group 1 Specifics

Breakdown of clinical classification of Group 1 PH.



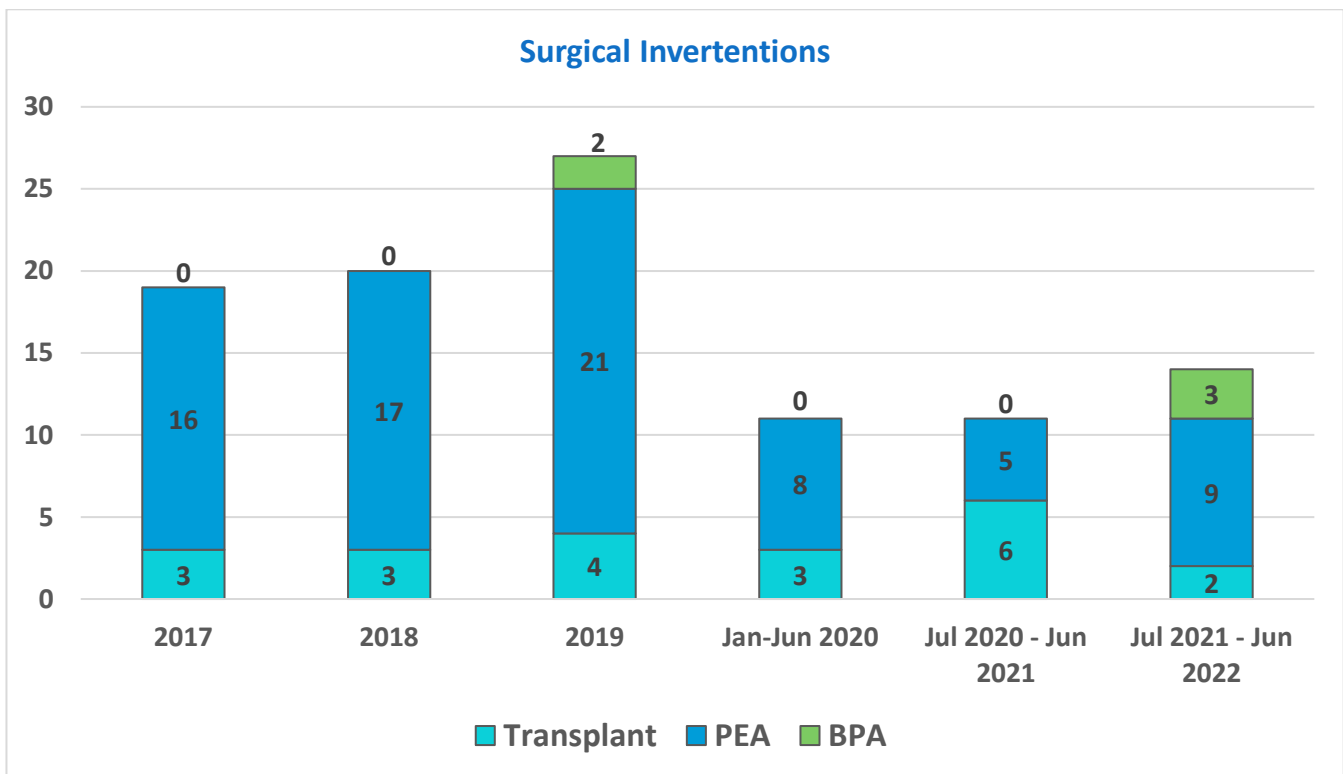
3.4. PH Specific Therapies

PH specific therapies approved in Canada: epoprostenol, treprostinil, selexipag, bosentan, ambrisentan, macitentan, sildenafil, tadalafil, riociguat. Figure below depicts treatment combination distribution of PH specific therapy.



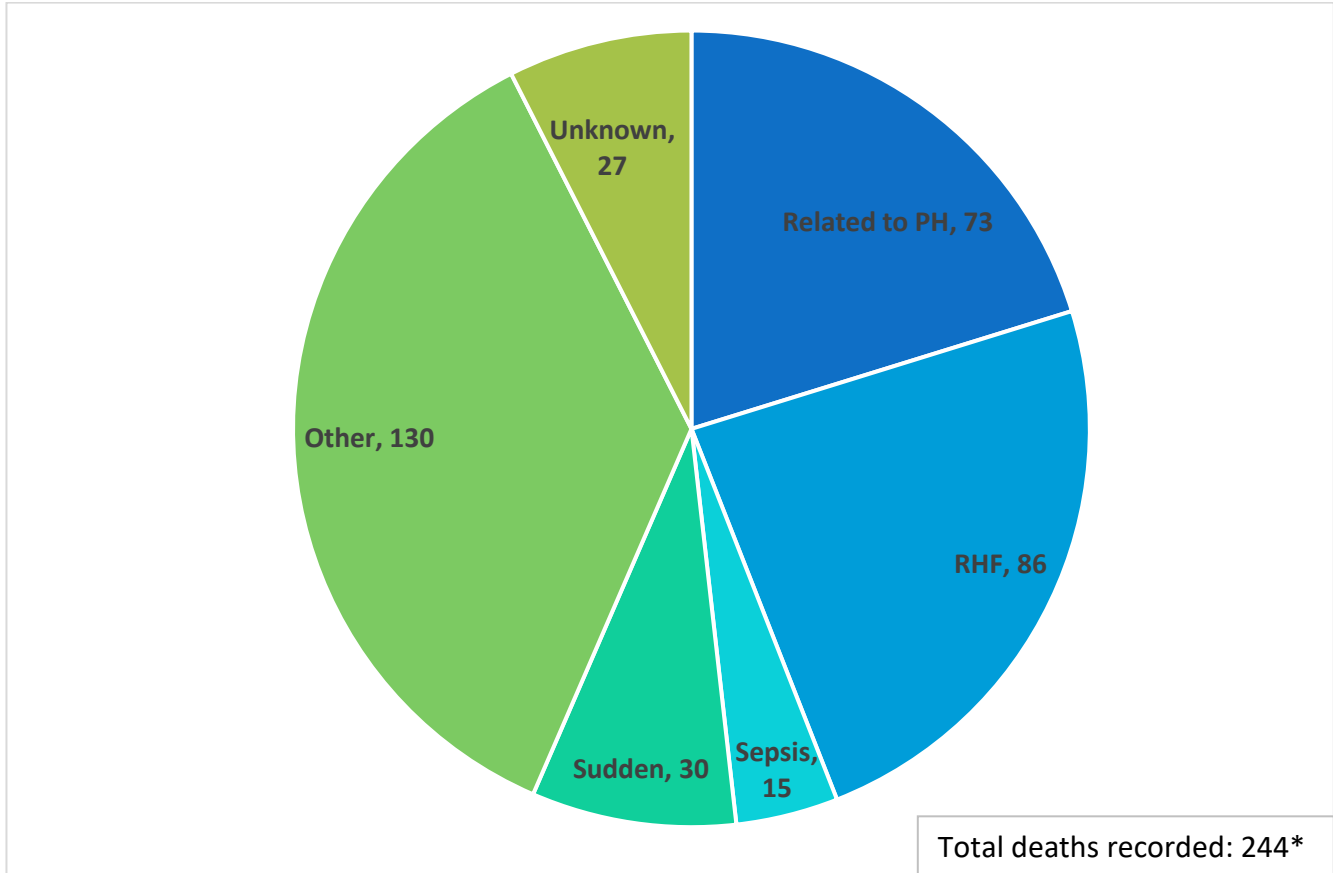
3.5. Transplants, PEA and BPA

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



3.6. Cause of Death

Breakdown of the cause of death of deceased patients.



Other reasons	#	Other reasons	#
Cancer	18	AECOPD	5
Pneumonia	15	PE	5
Congestive heart failure/disease	14	Septis/Septic shock	4
Liver disease	14	Cirrhosis	3
Palliative	13	Fall	3
Other	17	MAID	3
Hypoxemia	8	PVOD	3
Respiratory Failure	7	Pulmonary fibrosis	3
COVID	6	Post-Surgery complications	3
Renal disease	5	Aspiration	2

*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

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

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
Calgary Vancouver Hamilton Winnipeg	Ottawa Quebec City Halifax	“The EmPHasis-10 (E10) tool in Canadian patients with pulmonary hypertension: Validation and determining the minimal clinically important difference”	- sites undergoing ethics application review	Jason Weatherald
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 Montral	"Transitioning from parenteral prostacyclin therapy to oral selexipag in pulmonary arterial hypertension: A multi-centre retrospective chart review"	- sites collecting data	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?”	- multicentre project - sites collecting and cleaning data	Nathan Brunner
Vancouver	Edmonton	“Severity of Illness and Outcomes in Indigenous patients with Pulmonary Arterial Hypertension in Canada”	- sites undergoing ethics application review - analyzing Vancouver Data	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. 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.