2022-3

The Unseen Burdens of Pediatric Pulmonary Hypertension: Treatment Side Effects and Access to Care

Rebecca Brown
Brigham Young University - Provo, beccauburn@gmail.com

Megan Pierce
Brigham Young University - Provo

Samara Nelson
Utah State University

Ashley Bangerter Seelos
Brigham Young University - Provo

Ella Cook
Brigham Young University

See next page for additional authors

Authors

Rebecca Brown, Megan Pierce, Samara Nelson, Ashley Bangerter Seelos, Ella Cook, Heather Stickle, Michael Johansen, and Erik J. Nelson

The Unseen Burdens of Pediatric Pulmonary Hypertension: Treatment Side Effects and Access to Care

Rebecca Brown,[1] Megan Pierce,[1] Samara Nelson,[2] Ashley Bangerter Seelos,[1] Ella Cook,[1] Heather Stickle,[1] Michael Johansen, DO[3] and Erik J. Nelson, PhD, MPH[1]

[1]Department of Public Health, Brigham Young University, Provo, Utah; [2]Emma Eccles Jones College of Education & Human Services, Utah State University, Logan, Utah; [3]Indiana University School of Medicine, Indianapolis, Indiana

INTRODUCTION

- Pulmonary hypertension (PH) is a serious and life-threatening disease characterized by an elevation in mean pulmonary artery pressure and pulmonary vascular resistance, leading to right heart failure and death.[1,2]
- PH has primarily been studied among adults and therefore clinical therapies have also aimed at treating this population.[3-5]
- Several pharmaceutical treatments are available and approved for use in adults, however, the effectiveness among children has yet to be sufficiently determined though they are considered the standard treatment regimen in children.
- Accessing specialized pediatric PH care providers who are versant in these therapies is difficult.

Want to learn more about how PH affects children?

Watch this 3-minute Video:

OBJECTIVE

- The purpose of this research is to better understand the quality of life issues associated with PH treatment in children.

METHODS

Study Participants

- Parents/Caregivers of children living with PH were recruited via Facebook from December 2021 – January 2022

Eligibility Criteria

- Members of the Facebook Group
- Self-identified as parents, guardians, or primary caregivers of a child living with PH.
- Aged 18 years or older

Facebook Recruitment Strategy

- Participants were recruited through discussion board posts and direct messages within the "Families of Children with Pulmonary Hypertension" Facebook group.

Table 1. Characteristics of children living with PH in the study sample

Child's Characteristics	n (%)
Child's race/ethnicity	
White/Caucasian	114 (82.0%)
Other	25 (18.0%)
Gestational age at birth	
Less than 24 weeks	6 (4.6%)
more than 36 weeks	87 (66.9%)
24 to 28 weeks	12 (9.2%)
29 to 32 weeks	4 (3.1%)
33 to 36 weeks	21 (16.2%)
Age at PH diagnosis	
First 30 days	32 (23.0%)
1-12 months	37 (26.6%)
1-4 years	40 (28.8%)
5-10 years	23 (16.5%)
11-15 years	7 (5.0%)
Current age	
<1 year	13 (9.4%)
1-5 years	45 (32.4%)
6-10 years	36 (25.9%)
11-15 years	28 (20.1%)
16-17 years	15 (10.8%)
>=18 years	2 (1.4%)
Down Syndrome	
Yes	12 (8.7%)
No	126 (91.3%)
Congenital heart defect	
Yes	67 (48.2%)
No	72 (51.8%)

RESULTS

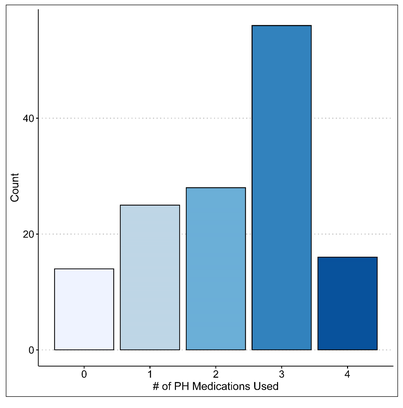
PH Therapy Side Effects

- More than 70% of Children with PH take ≥2 medications to combat symptoms of PH, with 51.7% taking ≥3 medications.
- The medicines with the highest average number of side effects reported were Remodulin (4.4), Uptravi (2.8) and Adcirca (2.6).
- Across all PH medicines, the most commonly reported side effects were flushing of the skin (18.6%), headache/dizziness (11.4%), joint/muscle pain (8.1%), and nausea (7.9%).

Access to PH Specialty Care

- Almost all PH care is received at a Specialty Children's Hospital
- Over 85% of families travel more than 20 miles to visit their primary PH care provider
- More than 68% of families travel more than 60 minutes to receive routine PH care
- In our sample, 12 families (8.6%) had to relocate in order to be closer to primary PH care.

Figure 1. Number of Medications Used by Children Living with PH



For additional Study Results, open this QR Code link:

Table 2. Frequency of Side Effects Reported by Children Using PH Therapies

Medication	# of Children Taking Medication	% of children reporting Side Effects	Total # of Reported Side Effects	Average # of Side Effects Per Child
Remodulin	43	97.7%	190	4.4
Adcirca (Tadalafil)	70	88.6%	183	2.6
Revatio (Sildenafil)	49	73.5%	74	1.5
Letairis (Ambrisentan)	56	33.9%	30	0.5
Tracleer (Bosentan)	19	21.1%	5	0.3
Opsumit (Macitentan)	12	25.0%	8	0.7
Uptravi (Selexipag)	26	80.8%	74	2.8
Tyvaso	2	50.0%	2	1.0
Adempas (Riociguat)	2	50.0%	3	1.5

Table 3. Care provider characteristics and access reported by parents/caregivers of children living with PH (n=139).

Care Provider Characteristics	n (%)
Where PH care is received	
Local pediatrician's office	2 (1.4%)
Specialty Children's Hospital	133 (95.7%)
There is no PH care where we live	4 (2.9%)
Type of PH provider	
Pediatric cardiologist & pediatric pulmonologist	61 (43.9%)
Pediatric Cardiologist	56 (40.3%)
Pediatric Pulmonologist	10 (7.2%)
PH Specialist	10 (7.2%)
Primary care physician (Local Pediatrician)	2 (1.4%)
Distance to primary PH provider	
Less than 5 miles	4 (2.9%)
6-10 miles	8 (5.8%)
11-20 miles	13 (9.4%)
More than 20 miles	113 (86.9%)
Travel time to PH provider	
Less than 10 minutes	3 (2.2%)
10-30 minutes	16 (11.5%)
30-45 minutes	15 (10.8%)
46-60 minutes	9 (6.5%)
61-90 minutes	21 (15.1%)
More than 90 minutes	74 (53.2%)
Had to move residence to be closer to PH care provider	
Yes	12 (8.6%)
No	127 (91.4%)

DISCUSSION AND CONCLUSIONS

- This study found that the unseen burdens of PH care are manifest in the number of treatment side effects and the distance traveled to access specialty care.
- Our results show that over half of children with PH take more than 3 medications to mitigate symptoms of PH, sometimes experiencing as many as 4 side effects.
- Additionally, most families travel more than 60 miles to receive PH care at a Specialty Children's hospital.
- These findings fill a gap in the literature about the unseen burdens of PH care.
- More research is needed to fully understand the side effects and burdens of PH among pediatric populations

References

1. Gaine SP, Rubin LJ. Primary pulmonary hypertension. *Lancet.* 1998;352(9129):719-725.
2. Berger RM, Beghetti M, Humpl T, et al. Clinical features of paediatric pulmonary hypertension: a registry study. *Lancet.* 2012;379(9815):537-546.
3. van Albada ME, Berger RM. Pulmonary arterial hypertension in congenital cardiac disease--the need for refinement of the Evian-Venice classification. *Cardiol Young.* 2008;18(1):10-17.
4. van der Linde D, Konings EE, Slager MA, et al. Birth prevalence of congenital heart disease worldwide: a systematic review and meta-analysis. *J Am Coll Cardiol.* 2011;58(21):2241-2247.
5. Zijlstra WM, Douwes JM, Rosenzweig EB, et al. Survival differences in pediatric pulmonary arterial hypertension: clues to a better understanding of outcome and optimal treatment strategies. *J Am Coll Cardiol.* 2014;63(20):2159-2169