



**The 5th International Congress  
on Cardiac Problems in Pregnancy (CPP 2018)**

22-25 February, 2018  
Bologna, Italy

# Pregnancy In Pulmonary Hypertension: An Update

***Dianne L. Zwicke, MD, FACC***

**Adjunct Professor of Medicine**

**University of Wisconsin School of Medicine - Milwaukee Clinical Campus**

**Medical Director**

**Pulmonary Hypertension Clinic / Accredited**

**Aurora St. Luke's Medical Center, Milwaukee, Wisconsin**

# FINANCIAL DISCLOSURE

Dianne L. Zwicke, MD

I have been an investigator and/or received study grants from the following companies:

Glaxo – Welcome

\*United Therapeutics

Pfizer

Medtronic

Co Therix

Ikaria

\*Gilead

Actileon

Novartis

Lilly

\*Bayer

GENO

Reata

Elgier

Univ of SD-CTEPH

\* Appointed to Advisory Boards

# Evolution of PAH and Pregnancy Data

2003  
Observe & Counsel  
CHEST

2016  
Pre-Pregnancy Counseling  
CPP / CHEST / PHPN



2008  
Pharmacology & Team –  
CHEST / CPP / ACC

2011  
Multidisciplinary Team  
PHA / ERS

# 2002 - 2018

## PATIENT POPULATION

### VOLUME

Total Patients (Mothers)	170
Total Infants	186

### Diagnosis of PAH

- Pre Pregnancy	10
- Post Pregnancy	8

### F/U of pre-pregnancy

- Chose Adoption	1
- Lost to F/U	1
- Proceeded with Pregnancy	8

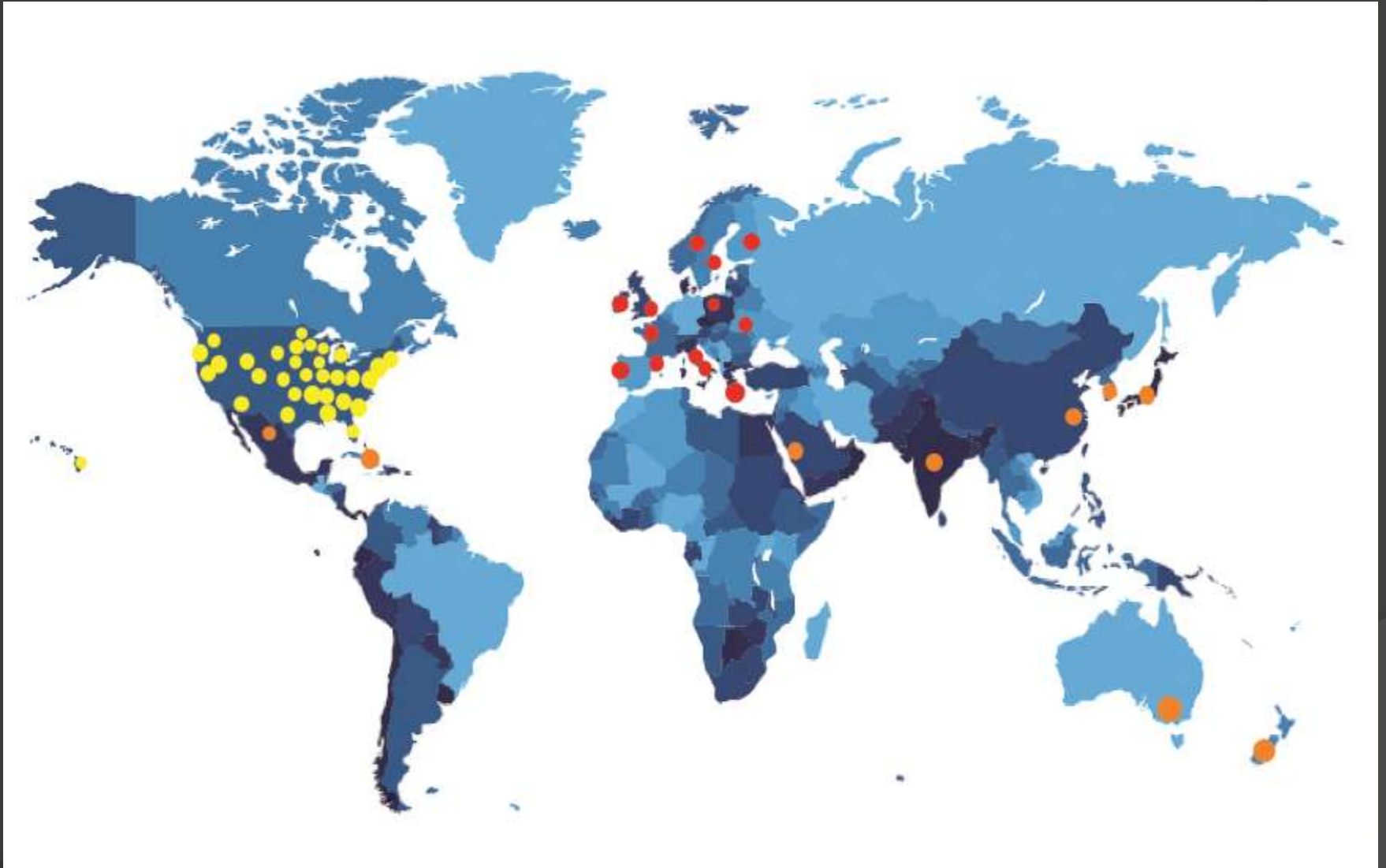
### Infants

- Female	106 (57%)
- Male	80 (43%)

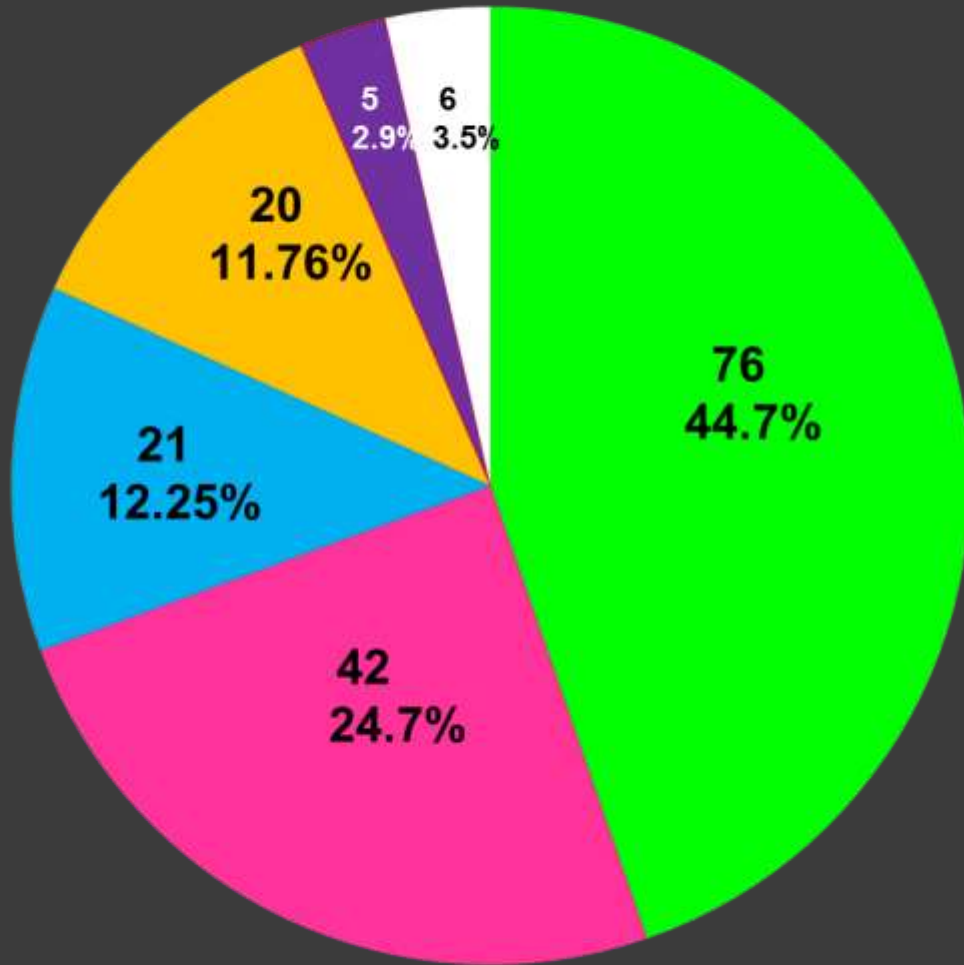
### Geography

- United States	32
- Foreign Countries	22

# OUR NEIGHBORS / WORLD MAP



# ETIOLOGY OF PAH / 170 PATIENTS



- Idiopathic PAH (44.7%)
- Congenital Heart Disease(24.7%)
- Connective Heart Disease (12.25%)
- Familial PAH (11.76%)
- Anorexic Drug Related (2.9%)
- CTEPH (6%)

## **Clinical symptoms at time of first evaluation**

92% Dyspnea on exertion

77% Lower extremity edema

44% Pre-syncope

12% Syncope

82% Increased abdominal girth for gestational age

33% Chest pressure, discomfort, pain with exertion

# WHAT IS CAUSE OF SYMPTOMS

- Serious alterations in normal pregnancy
  - Each trimester
  - Post partum
  - Manifests at 16-28 weeks gestation, especially weeks 24-28
- Systems
  - Cardiovascular System
    - ↑ total blood volume by 20-30%
    - ↑ red cell mass and plasma volume is 30-50%
    - ↑ heart rate by 25 beats per minute
    - ↑ Cardiac output throughout pregnancy, peak at 32 weeks (30-50%)
    - ↑ SVR, PVR, BP decreases
    - ↑ 500 cc of blood wash in/out of uterus with contractions



# PAH SYMPTOMS

- Fixed pulmonary vascular resistance (PVR), with serious increase in blood volume.
- Decreased or fixed cardiac output (supply & demand).
- RV dysfunction → fluid retention.
- Impaired venous return secondary to compression of the IVC, as fetus grows.
- Worsening right to left shunt, if present.
- Evaluate nocturnal O<sub>2</sub>.
- Hypercoagulability.

# Pharmacologic agents used in pregnant patients with PAH at our center

<b>Inhaled vasodilators</b>  Nitric oxide  Epoprostenol (Flolan, Veletri)  Treprostinil (Tyvaso)	<b>Inotropic agents</b>  Dobutamine (Dobutrex)  Digoxin (Lanoxin)  Dopamine
<b>Intravenous prostacyclins</b>  IV Epoprostenol  Flolan (early years)  Veletri (more recently)  IV Treprostinil  Remodulin	<b>Antiarrhythmic (A-Fib)</b>  Diltiazem (Cardizem)  Amiodarone (Cordarone)  Digoxin (Lanoxin)
<b>Oral prostacyclins</b>  Orenitram (Treprostinil Oral)  Selexipeg (Uptravi)	<b>PDE-5</b>  Sildenafil (Revatio)  Tadalafil (Adcirca)
<b>Calcium channel blocker</b>  Nifedipine (Procardia)  Amlodipine (Norvasc)	<b>Anticoagulants</b>  Warfarin (Coumadin)  Enoxaparin (Lovenox)
<b>Diuretics</b>  Furosemide (Lasix)  Torsemide (Demadex)  Bumetanide (Bumex)	<b>Other</b>  Potassium chloride  Magnesium

# Medications not recommended in pregnancy secondary to teratogenicity

- SCG DRUGS (Adempas)
- ERA Drugs (Letaris, Macitentan, Tracleer)

# EVALUATION

- Multidisciplinary teams established early
- Written Delivery / Care plan
- Spinal anesthesia is mandatory
- Delivery –

Cesarean Section	32 (17.4%)
Vaginal delivery	148 (80.4%)
Conversion	4 ( 2.2%)
- Degree of PAH

Severe	- 63 patients
Moderate	- 88 patients
Mild	- 19 patients

# Long Term Follow up of patients

170 Patients – **NO Deaths** related to pregnancy.

Etiology of 8 deaths within observation time of study

- No deaths during or within 18 months after delivery of babies.
- 1 death secondary to progression of CTD (Lupus)
- 3 deaths secondary to progression of congenital heart disease awaiting heart-lung transplant
- 4 deaths from progression of primary disease
  - 1 Anorexic PAH, 1 Sickle Cell disease, 1 Idiopathic PAH, 1 Familial PAH