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PREGNANCY IN PULMONARY HYPERTENSION: Recommendations for A Successful Outcome

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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

PAH 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

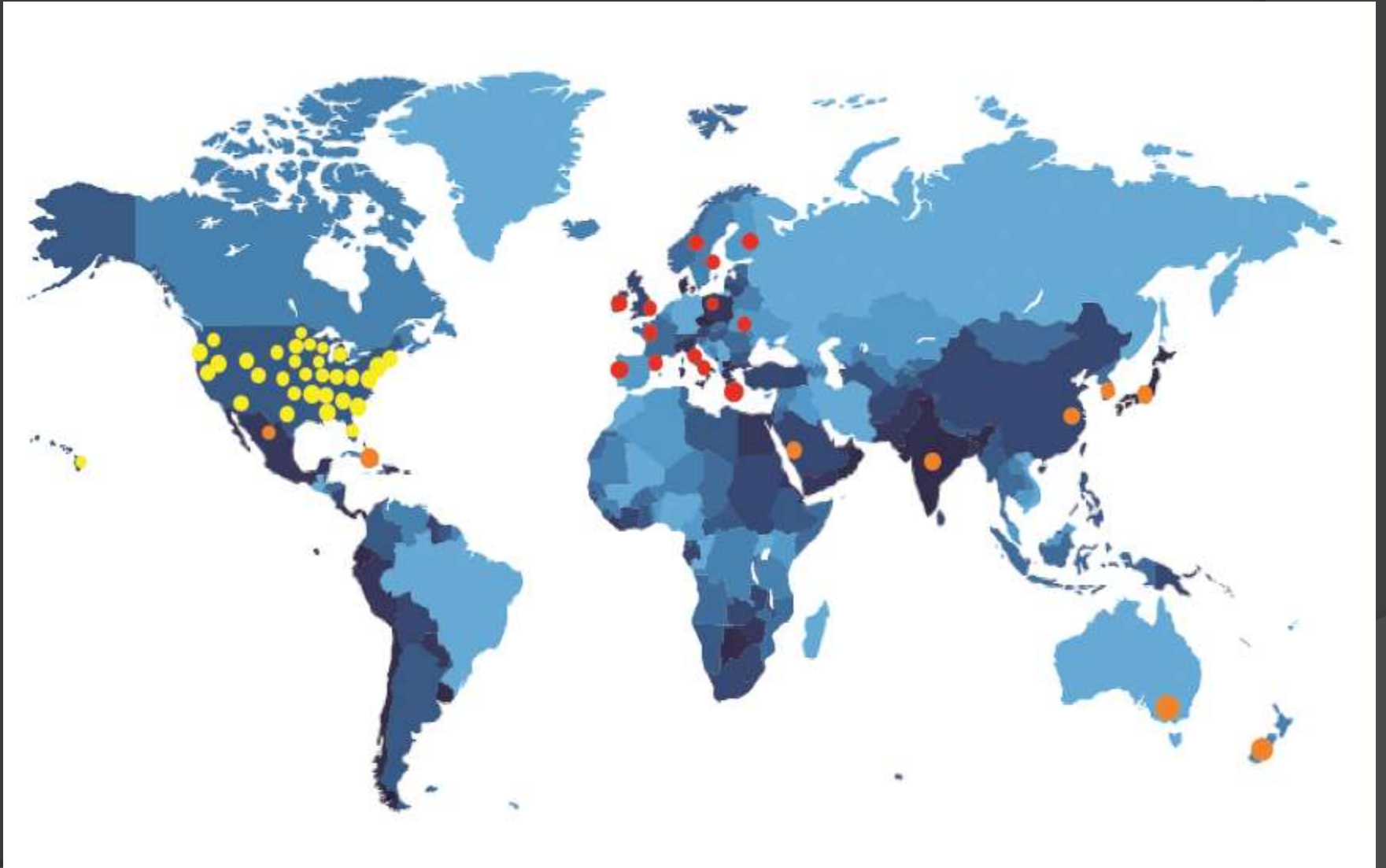
Infants

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

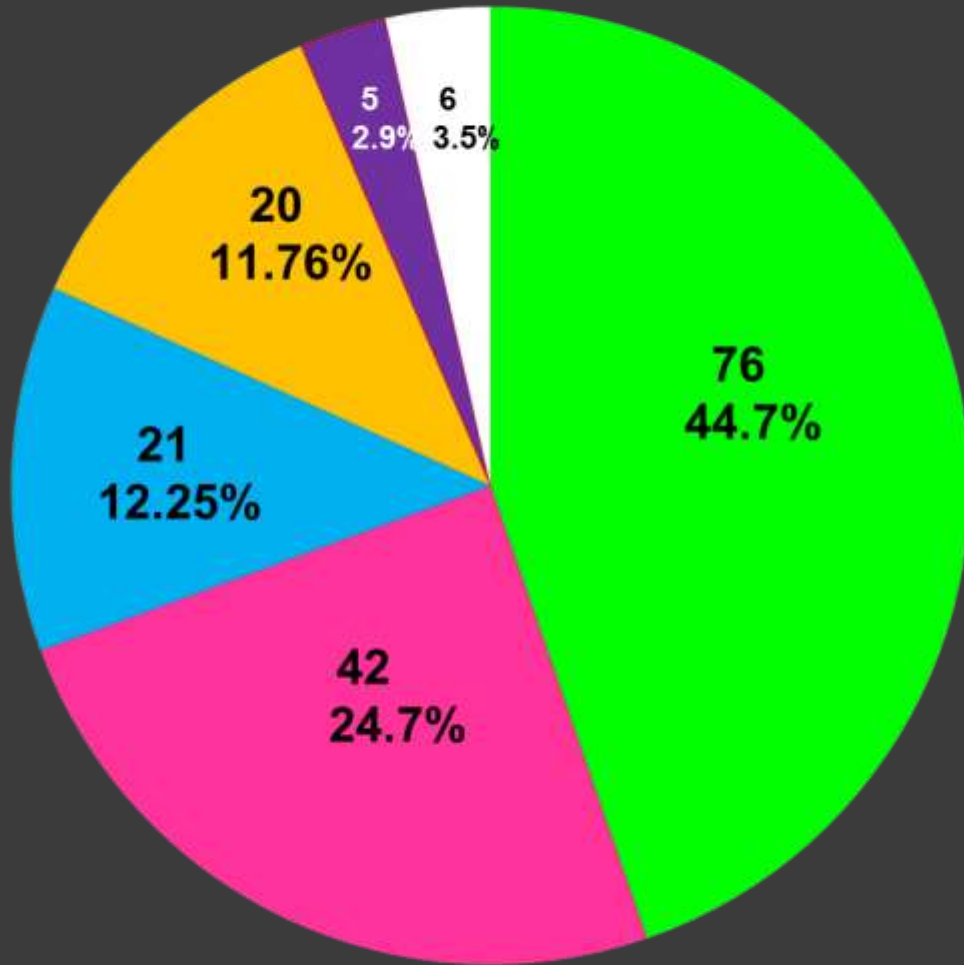
Geography

- 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

OUTCOMES

- Written Care Plan
- Delivery – Cesarean Section - 32 (17.4%)
Vaginal delivery - 138 (80.4%)
Conversion - 4 (2.2%)
- Degree of PAH
 - Severe - 63 patients
 - Moderate - 88 patients
 - Mild -19 patients
- Spinal / Epidural Anesthesia (Mandatory)
- Multidisciplinary team – establish early
- Survival – 170 women – NO DEATHS
186 infants
100% survival

Multidisciplinary Team

Physicians: Cardiologist – PAH, Congenital
 MFM – High Risk OB
 Anesthesia – OB and Cardiac
 Nephrology
 Critical Care Physician
 Pulmonary Medicine
 Psychiatry
 Others: Hematology / CV Surgeon (ECMO)

RN's: ICU / CCU
 Labor and Delivery

Respiratory Therapy

Spiritual Care (if requested)

Pharmacologic agents used in pregnant patients with PAH at our center

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

Medications not recommended in pregnancy secondary to teratogenicity

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

CASES

- Severe PAH – age 25, 1st pregnancy, diagnosed at 4 months of pregnancy.
- PAH diagnosis 5 days prior to delivery. Complicated by a chest tumor.
- Pre-pregnancy counseling.

CONCLUSION OF 16 YEARS OF DATA AND EXPERIENCE

- WE CAN TREAT PAH IN PREGNANCY
- We can do realistic pre-pregnancy counseling
- We have tools to treat PAH in pregnancy