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
- Medtronic
- *Gilead
- Lilly
- Reata
- *United Therapeutics
- Co Therix
- Actileon
- *Bayer
- Elgier
- Pfizer
- Ikaria
- Novartis
- GENO
- Univ of SD-CTEPH

* Appointed to Advisory Boards
Evolution of PAH and Pregnancy Data

- 2003
  Observe & Counsel
  CHEST

- 2011
  Multidisciplinary Team
  PHA / ERS

- 2016
  Pre-Pregnancy Counseling
  CPP / CHEST / PHPN

- 2008
  Pharmacology & Team -
  CHEST / CPP / ACC
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%)
- Anorexie 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)
<table>
<thead>
<tr>
<th>Pharmacologic agents used in pregnant patients with PAH at our center</th>
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<tbody>
<tr>
<td><strong>Inhaled vasodilators</strong></td>
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<tr>
<td>Nitric oxide</td>
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<tr>
<td>Epoprostenol (Flolan, Veletri)</td>
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<tr>
<td>Treprostinil (Tyvaso)</td>
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<tr>
<td><strong>Intravenous prostacyclins</strong></td>
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<tr>
<td>IV Epoprostenol</td>
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<tr>
<td>Flolan (early years)</td>
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<tr>
<td>Veletri (more recently)</td>
</tr>
<tr>
<td>IV Treprostinil</td>
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<tr>
<td>Remodulin</td>
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<tr>
<td><strong>Oral prostacyclins</strong></td>
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<tr>
<td>Orenitram (Treprostinil Oral)</td>
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<tr>
<td>Selexipec (Upravi)</td>
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<tr>
<td><strong>Calcium channel blocker</strong></td>
</tr>
<tr>
<td>Nifedipine (Procardia)</td>
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<tr>
<td>Amiodipine (Norvasc)</td>
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<tr>
<td><strong>Diuretics</strong></td>
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<tr>
<td>Furosemide (Lasix)</td>
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<td>Torsemide (Demadex)</td>
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<td>Bumetanide (Bumex)</td>
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</table>
Medications not recommended in pregnancy secondary to teratogenicity

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

- Severe PAH – age 25, 1\textsuperscript{st} 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