**INTRODUCTION**

- Life expectancy for individuals with cystic fibrosis (CF) has significantly increased and subsequently, more women with CF are considering pregnancy.
- Cystic fibrosis is a chronic and life-threatening disease characterized by a defective CF transmembrane regulator protein that affects multiple organ systems.
- Primary manifestations of CF in adults include progressive lung disease associated with infection and bronchiectasis, pancreatic insufficiency, CF-related diabetes, gastroesophageal reflux, distal intestinal obstruction syndrome, nutritional deficiencies, and possible liver impairment (Geake et al., 2014).
- As the disease state varies depending on several factors, including genetics and the pre-pregnancy clinical course, coordination of the complex care during pregnancy for women with CF involves an interprofessional team of health care providers.
- A detailed understanding of the management of pregnancy, specific pulmonary exacerbations, and necessary medications is essential to provide specialized physical and psychosocial care for pregnant women with CF.

**CARE OF WOMEN WITH CF DURING PREGNANCY**

- **Pulmonary Care**
  - Close monitoring of lung function, pulse oximetry values, and spum cultures is essential throughout pregnancy and the postpartum period.
  - Pregnant women with CF may experience more frequent pulmonary exacerbations for many reasons, including the immunosuppressive effects of pregnancy, severity of lung disease, adherence to treatment, discontinuation of certain routine medications not recommended during pregnancy, and colonization with certain organisms such as B. cepacia.
  - Nebulized Pulmozyme, nebulized selective short-acting β₂-agonists, and nebulized hypertonic saline are routinely used to maintain lung function and reduce pulmonary exacerbations. These medications are unlikely to cause any risk to the woman or fetus (Panchaud et al., 2016).
  - Airway clearance therapy (ACT) or high-frequency chest compression vests help remove pulmonary secretions. Nebulized treatments should be administered before ACT or in combination with ACT.
  - Antibiotics are a mainstay in CF treatment plans.

**Common Intraoperative and Inhaled Antibiotics Used for Women With Cystic Fibrosis**

<table>
<thead>
<tr>
<th>Drug</th>
<th>Route</th>
<th>Recommended Use</th>
</tr>
</thead>
<tbody>
<tr>
<td>Penicillin</td>
<td>Intra</td>
<td>Drug of choice for Staphylococcus aureus or Haemophilus influenzae</td>
</tr>
<tr>
<td>Amoxicillin</td>
<td>Intra</td>
<td>Drug of choice for Pseudomomas aeruginosa</td>
</tr>
<tr>
<td>Cephalosporins</td>
<td>Intra</td>
<td>Drug of choice for S. aureus or H. influenza</td>
</tr>
<tr>
<td>Macrolides</td>
<td>Intra</td>
<td>Drug of choice for S. aureus or H. influenza</td>
</tr>
<tr>
<td>Carbapenem</td>
<td>Intra</td>
<td>Second-line treatment for Pseudomonas aeruginosa</td>
</tr>
<tr>
<td>Quinolones</td>
<td>Intra</td>
<td>Avoid in pregnancy, if needed, oprofloxacin is the drug of choice</td>
</tr>
<tr>
<td>Aminoglycosides</td>
<td>Intra</td>
<td>First-line treatment, low systemic absorption</td>
</tr>
<tr>
<td>Monobactams</td>
<td>Intra</td>
<td>Minimal systemic absorption second-line treatment</td>
</tr>
</tbody>
</table>

*From Kisan et al. (2010) and Panchaud et al. (2016)*

**Nutrition and GI Care**

- With estimated increased caloric need of approximately 300 kcal/day throughout pregnancy, pregnant women with CF may find it difficult to gain adequate weight (Michel and Mueller, 2015). Adequate weight gain may be achieved through oral supplementation, enteral feeds, or parenteral nutrition.
- Pancreatic enzyme replacement therapy is a necessary treatment for women with CF and is not considered a risk for the fetus.
- Insulin is the drug of choice to maintain stable serum glucose levels throughout pregnancy.
- Vitamin A supplementation is necessary for pregnant women with CF. Serum levels of vitamins D, E, and K are low and should be assessed and supplemented as needed.

**Labor and Delivery Considerations**

- Induced preterm births are more frequent for women with CF, with declining lung function as a common indicator.
- Most women can give birth vaginally with the use of epidural anesthesia.

**Psychosocial Care**

- Social isolation during hospital admission for pulmonary exacerbation.
- Financial difficulties as result of CF-related health issues.
- Exhaustion from the physical and emotional demands of caring for a newborn while caring for self, concern about possibly leaving the child motherless.
- Risk of postpartum depression.

**References Upon Request**

**DISCUSSION**

- Induction of labor occurred at 35 weeks with a cervical balloon catheter for cervical ripening, no CF related contraindications to the use of misoprostol, dinoprostone, or oxytocin.
- Approximately 36 hours later she experienced a spontaneous vaginal birth after abdominal epistaxis of a healthy female newborn weighing 2,256 g with Apgar scores of 9 and 9 at 1 and 5 minutes, respectively.
- She and her newborn were in stable condition and doing well immediately after birth.

**CASE REPORT**

- Key points:
  - Pregnancy progression from approximately 10 weeks to 35 weeks gestation of a 29-year-old White, primigravid woman with a genotype delta F508/R76X of CF.
  - She presented for her first prenatal visit with a planned pregnancy in mid-December and experienced three exacerbations requiring hospitalization during this pregnancy.

**Exacerbation 1\textsuperscript{Approximately 14 Weeks Gestation**

- Diagnosis of Influenza A
- Exacerbation followed by eight day home IV antibiotic treatment
- Intensified ACT with maintenance intravenous therapies
- IV cerftaroline & meropenum
- Sputum culture for acid-fast bacillus
- Monitoring of glucose levels on admission and throughout pregnancy
- FEV, predicted = 30% severe obstruction (14 weeks gestation)
- Mid-Fetal ultrasound = normal fetal anatomy

**Exacerbation 2\textsuperscript{Approximately 27 Weeks Gestation**

- Admission with exacerbation, shortness of breath, & hypoxia
- Hospitalized until delivery
- PE ruled out at tertiary hospital
- IV and intravenous antibiotics, steroids, normal home med routine continued
- Continued electronic fetal monitoring
- Oxygen 1 – 2 liters at right to maintain adequate saturation levels
- FEV, predicted = 31% severe obstruction (27 weeks gestation)
- Continued with ultrasounds, mid-March no uroperitoneal insufficiency noted

**Exacerbation 3\textsuperscript{Approximately 32 Weeks Gestation**

- Admitted with MRSA sepsis, worsening symptoms & hypoxia
- Hospitalized until delivery
- High flow nasal cannula oxygen & noninvasive positive pressure ventilation
- Zosyn & Zyvox along with antibiotics, home med routine continued
- Continued electronic fetal monitoring
- Betamethasone injections for fetal lung maturity
- Deep vein thrombosis prophylaxis
- FEV, predicted = 29% severe obstruction at delivery (36 weeks gestation)

**Cystic Fibrosis Foundation (CF) Annual Patient Data Registry Report (August, 2019)**