

Cystic Fibrosis in Pregnancy

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Hospital**

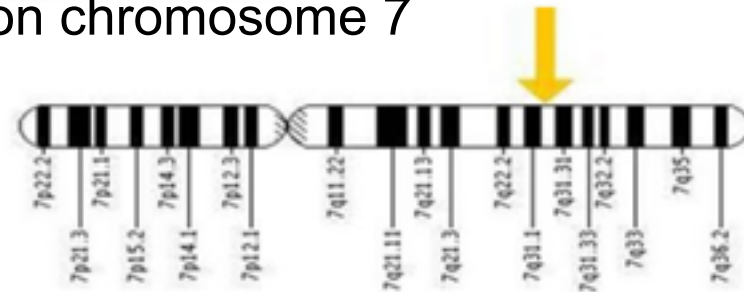
Sinai Health System
Joseph & Wolf Lebovic
Health Complex

Objectives

- Review features of cystic fibrosis
- Effects of CF on pregnancy and vice versa
- Management of women with CF during pregnancy

Cystic Fibrosis

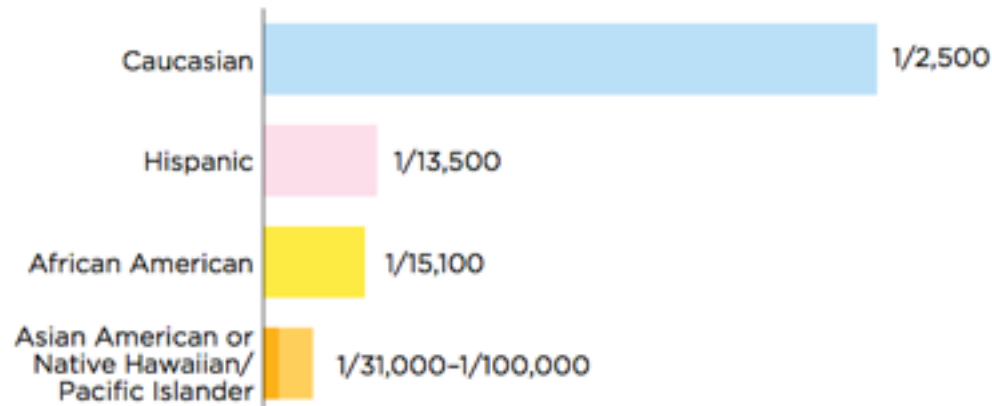
- Autosomal recessive condition: mutation of the gene for CFTR protein on chromosome 7



Cystic Fibrosis

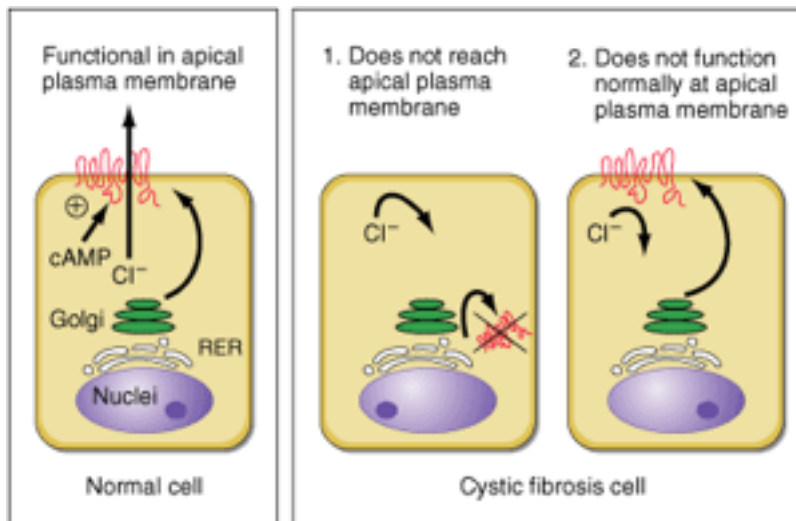
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Estimated Cystic Fibrosis Prevalence Rates by Race and Ethnicity



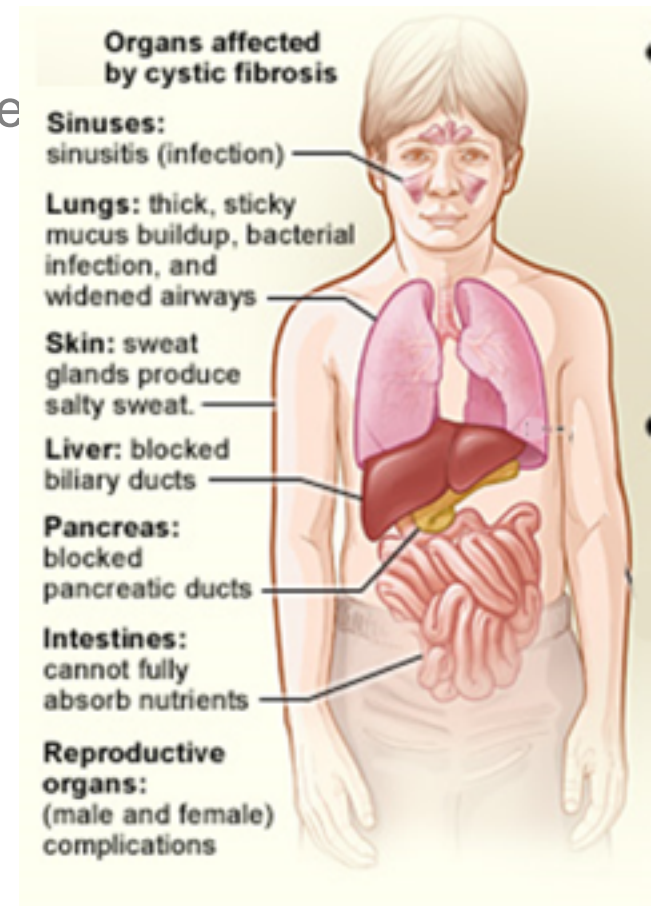
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- CFTR protein is a Cl^- ion channel: sweat, digestive juices, mucus



Cystic Fibrosis

- Autosomal recessive condition: mutation of the gene for CFTR protein on chromosome 7
- Common genetic abnormality: 1/25 carry gene
- CFTR protein is a Cl⁻ ion channel: sweat dige
- Disease affects: lungs (bronchiectasis)
GI tract (malabsorption)
pancreas (diabetes)



CF - bronchiectasis

Effects:

- Hypoxemia
- Pulmonary hypertension
- Respiratory failure
- Barotrauma
- Recurrent infections
 - Pseudomonas*
 - Staphylococcus*
 - Burkholderia. cepacia*



Physiological changes in pregnancy

Anatomic effects

Functional effects

airway edema,
friability

widened AP and
transverse diam.

elevated diaphragm

widened subcostal
angle

enlarging uterus

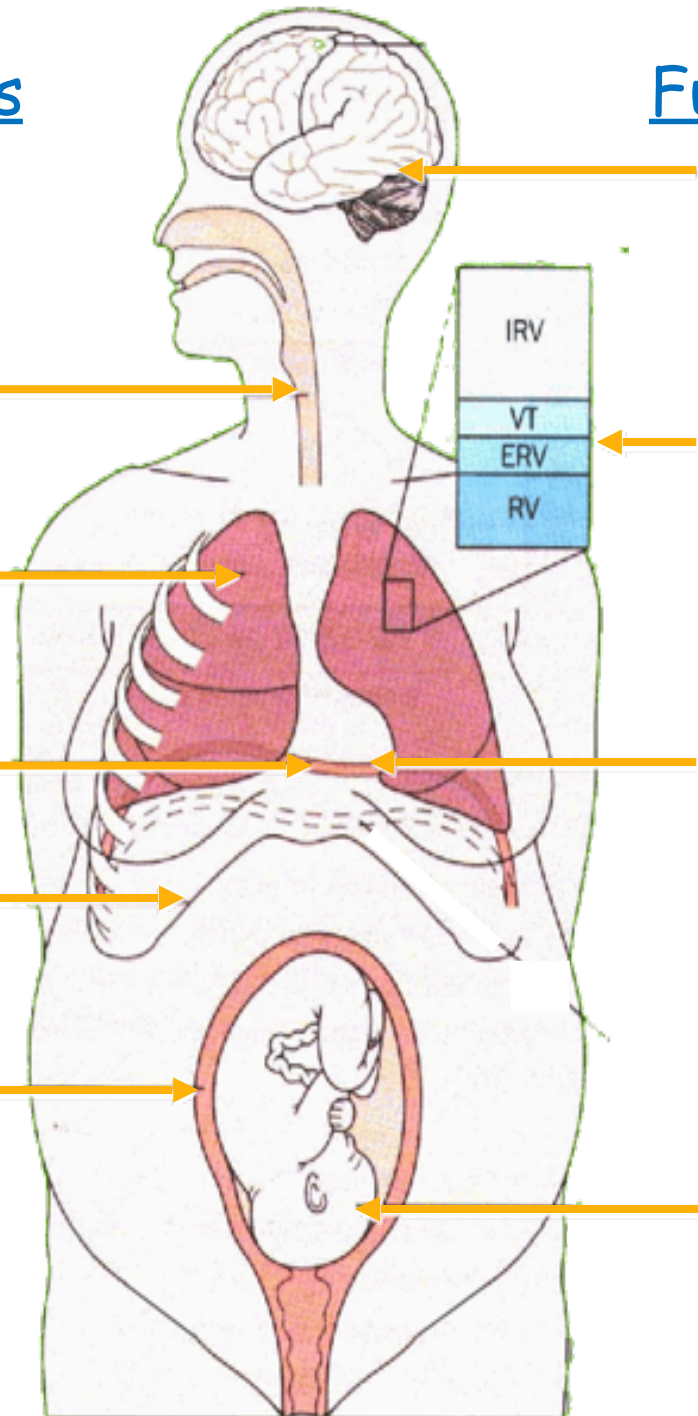
increased respiratory
drive

minimal change in TLC

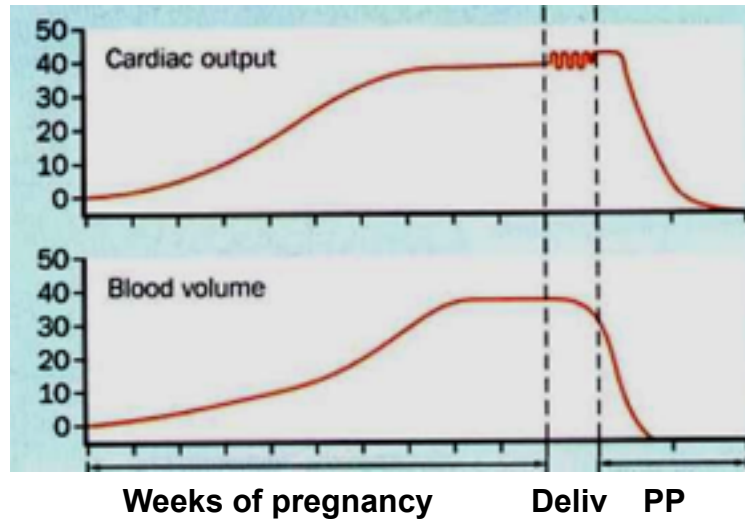
increased V_t
reduced FRC

normal diaphragmatic
function

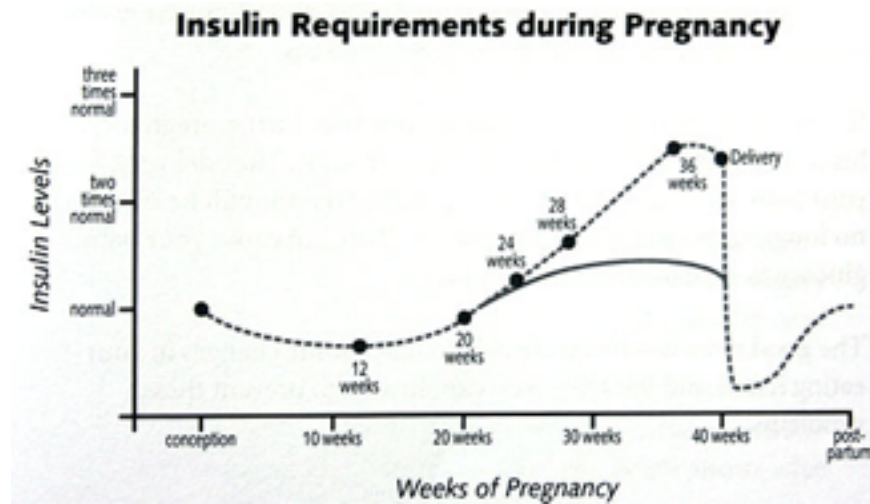
increased O_2
consumption and CO_2
production



Cardiac/hemodynamic



Endocrine



Implications of physiological changes:

- Maintaining oxygen requirements of pregnancy
- Achieving the normal increased ventilation
- Nasal obstruction
- Pulmonary hypertension
- Managing diabetes

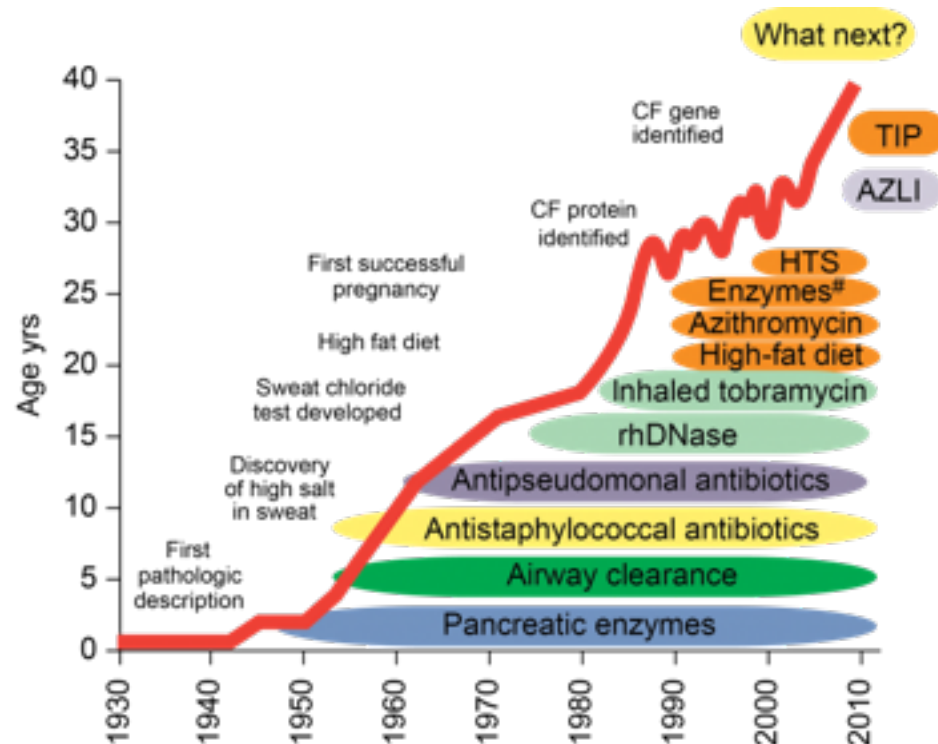
Fertility and pregnancy in women with CF

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 - decreased body mass index
 - thick mucus:
 - cervical mucus (cyclic), uterine fluid
 - ovarian dysfunction

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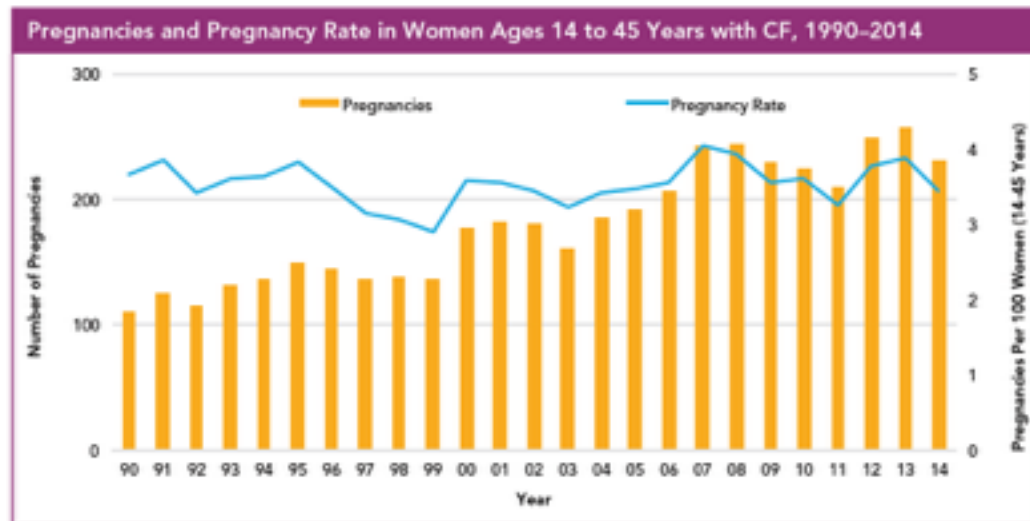
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 - artificial reproductive techniques

Have made pregnancy increasingly common

Fertility and pregnancy in women with CF

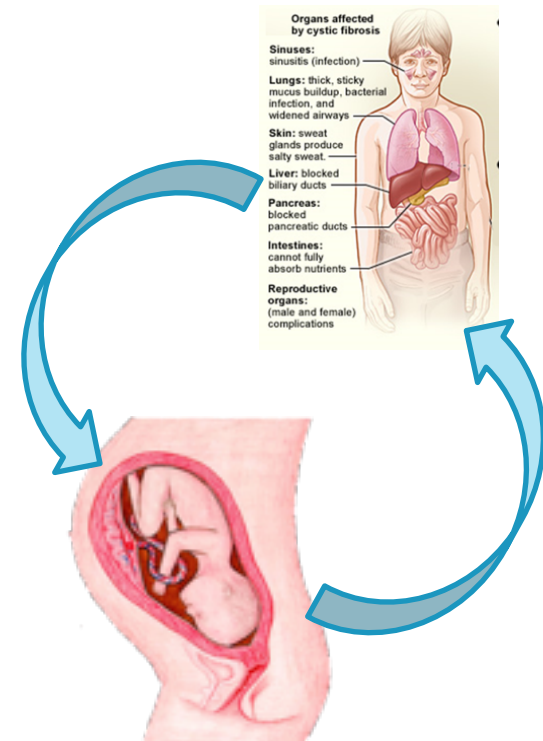
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Pregnancy in the woman with CF

- Does CF adversely affect the pregnancy?
- Does pregnancy adversely affect maternal outcome?
- Genetic counselling
- Management during pregnancy:
 - Pulmonary
 - Nutrition
 - Diabetes management
 - Labor and delivery



CF effects on pregnancy

- Data derived from case series of expert centres (mostly series from 1970's to 2000)

- No increased risk of fetal demise or birth defects
- Preterm delivery common – 25%
 - associated with poor lung function and low maternal weight

Barak et al, Isr Med Assoc J 2005; 7:95-98

Ødegaard et al. Acta Obstet Gynecol Scand 2002; 81:698-705

- Toronto data (1963-1998): 8% preterm delivery
 - Not associated with poor lung function

Gilljam et al, Chest. 2000;118:85-91

Pregnancy effects on CF patients

- **No negative effect on maternal long-term outcome**
 - pregnant women usually less severe lung disease
 - no adverse effect even when corrected for PFT
 - including women with FeV1 < 40%
- **Increased antibiotic use (v. non-pregnant):**
increased infections versus lower threshold?
- **Comparison of pregnant v. non-pregnant CF patients**
 - no difference in decline of FEV1 over time
 - increased diabetes during pregnancy, but not long-term

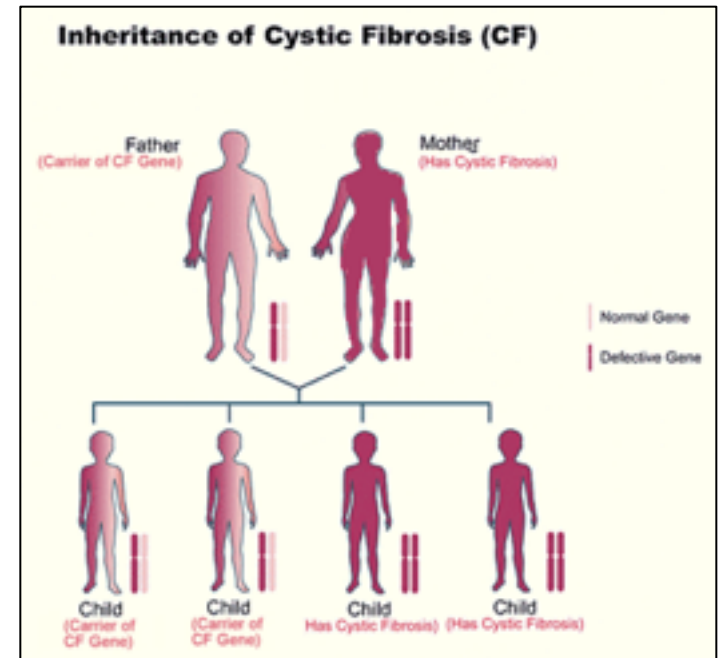
McMullen et al. Chest. 2006;129:706-711

- **No increase in long-term maternal mortality**

Goss et al. Chest. 2003; 124:1460-8

Counselling

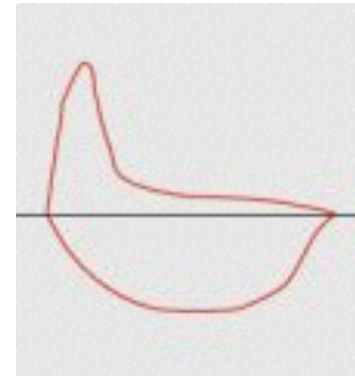
- **Homozygous mother:**
 - increase risk for baby to 1/50
(with unknown father's status)
 - Identify genotype: 1400 defects
 - screening is not 100% sensitive
- **Offer psychosocial counselling**
 - risks of pregnancy
 - frequent medical visits
 - demands of managing baby
 - transplantation/death
- **Contraindications:**
 - pulmonary hypertension?



Managing the pregnant patient with CF

Pulmonary disease

- Monitoring:
 - spirometry
 - pulse oximetry
 - Echo: re pulmonary hypertension
- Anesthesia consultation:
 - epidural/spinal
 - general anesthesia?
- Treatment:
 - supplemental oxygen therapy
 - exercise



Managing the pregnant patient with CF

Treatment of pulmonary disease

- Antibiotic therapy:
 - don't avoid necessary Rx
 - penicillins, cephalo, azithro (short term) – safe in pregnancy
 - ciprofloxacin is used in pregnant CF patients
 - careful with aminoglycosides
 - inhaled antibiotics probably safe (except high dose?)
 - Avoid: TMP-SMX, tetracyclines, carbapenems
- Secretion clearance:
 - physiotherapy
 - inhaled Dornase (DNAase)- used in pregnancy?

Managing the pregnant patient with CF

Pancreatic disease

- **Nutrition:**
 - meet with dietitian
 - monitor weight
 - adequate intake
 - pancreatic enzymes
 - fat soluble vitamins, Fe suppl
- **Diabetes:**
 - if not diabetic, OGTT every trimester
 - monitor sugars, keep HbA1C <6%
 - Rx with insulin
 - sulfonylureas – NB neonatal hypoglycemia
 - newer drugs – no data in pregnancy



Managing the pregnant patient with CF

Labor and delivery

- Analgesia
 - epidural: reduces CV and respiratory work
 - reduces hyperventilation and alkalosis
 - N₂O: barotrauma? (isobaric counterdiffusion)
- Mode of delivery:
 - determined by obstetric factors
- Postpartum:
 - hypoglycemia in diabetics
 - breast feeding acceptable (with good nutrition)

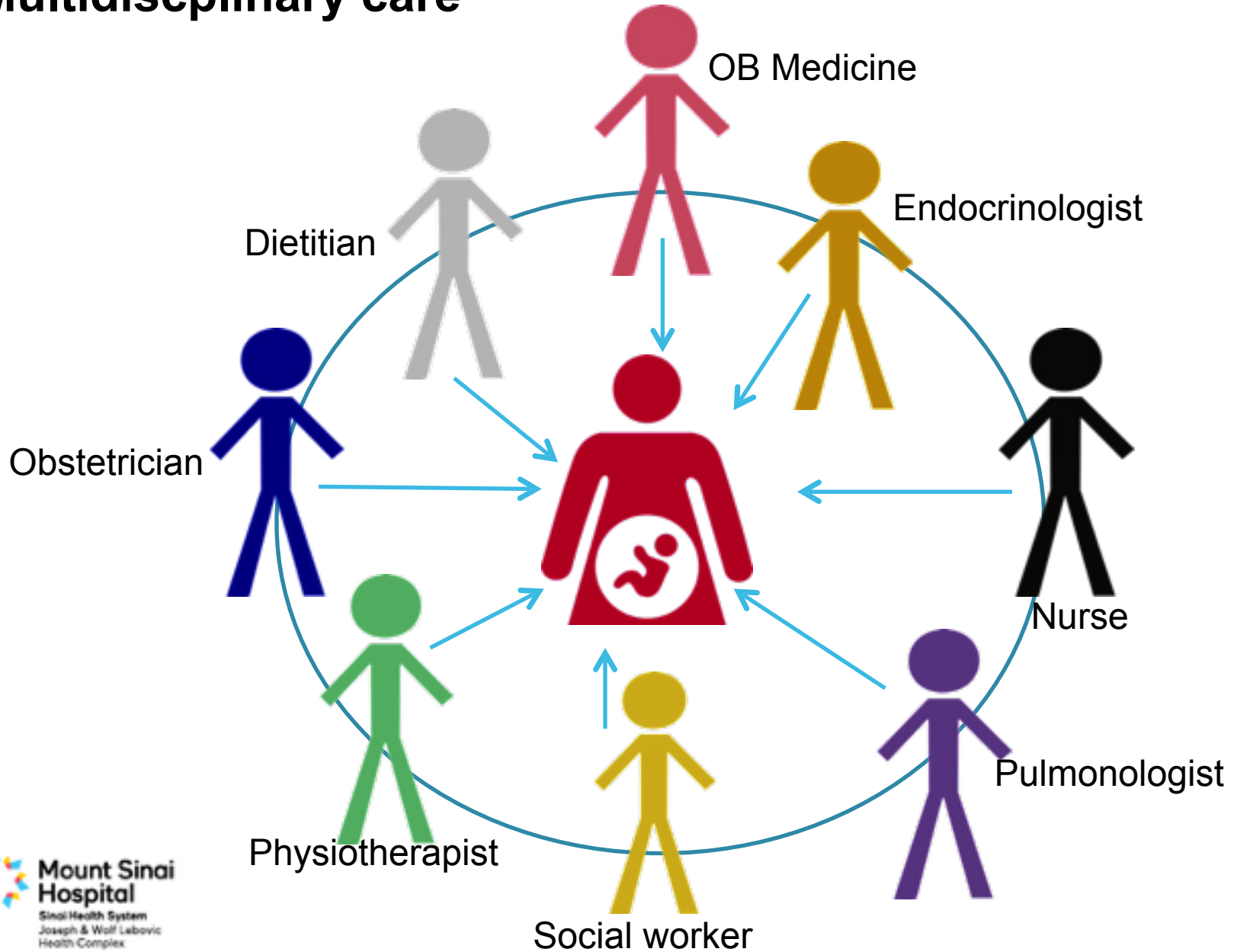
Managing the pregnant patient with CF

Termination of pregnancy

- Incidence
 - unclear: under-reporting
- Indications:
 - C/I to pregnancy: severe pulmonary hypertension
Burkholderia infection?
FEV1 < 50% ??
 - very poor nutritional state (eg. BMI < 18 kg/m²)
- Methods:
 - surgical, under epidural (or GA)
 - medical

Edenborough et al, BJOG 2000; 107:254-61

Multidisciplinary care





Questions?