



Canadian Cystic Fibrosis Conference

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Maternal-Infant CF Pathways

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w/o limits

Disclosures

I have the following disclosures:

Advisory Board: Vertex



Index Cases

- Recent review of 3 cases has prompted us to re-evaluate the management of:
 - The fetus of a CF pregnant person
 - Fetuses who are antenatally diagnosed
 - Fetuses with a positive family hx or echogenic bowel on ultrasound
 - CF positive neonates and their management across our hospital

Case 1

- 2nd pregnancy of a known CF carrier with first born homozygous dF508
- CVS completed, fetus positive for CF
- Query echogenic bowel identified on 18 week U/S
- Monitoring completed by local physician
- Delivered at community hospital
- Diagnosed with meconium ileus with volvulus
- Significant bowel resection required (short gut physiology) and TPN dependent for almost 2 years

Case 2

- 4th pregnancy
- Known family history
- Parental screening declined
- Echogenic bowel on ultrasound
- Referral made to FDC, repeat imaging but no further follow up
- ? resolution
- Born at regional hospital
- Delayed passage of meconium
- Transferred to tertiary centre for laparotomy and evacuation of bowel
- No resection required

Case 3

- Known CF pregnant person on Trikafta
- Referral to genetics for partner carrier testing but no-showed
- At Risk infant alert sent to NSO after birth
- NBS negative
- Offered precautionary sweat test but no follow up from mom
- Unsure if modulator monitoring completed in infant

Maternal Pathways

After discussion around these cases, we worked with key stakeholders across the organization to create 3 specific maternal pathways

Stakeholders included:

Paeds CF team

Adult CF team

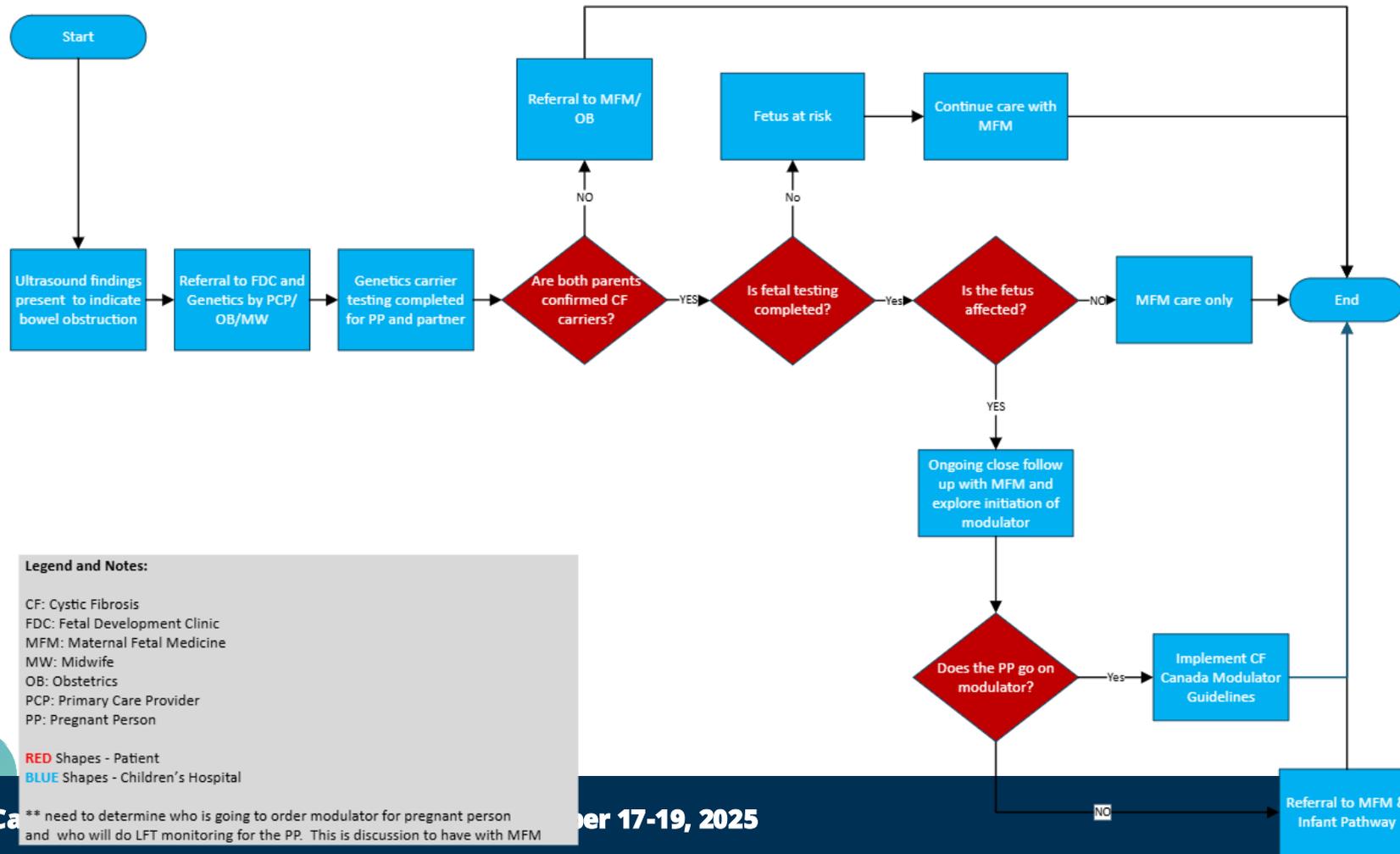
Genetic counsellor

OB/MFM

Parent liaison



Ideal State Process Map – Non-CF affected pregnant person with abnormal ultrasound



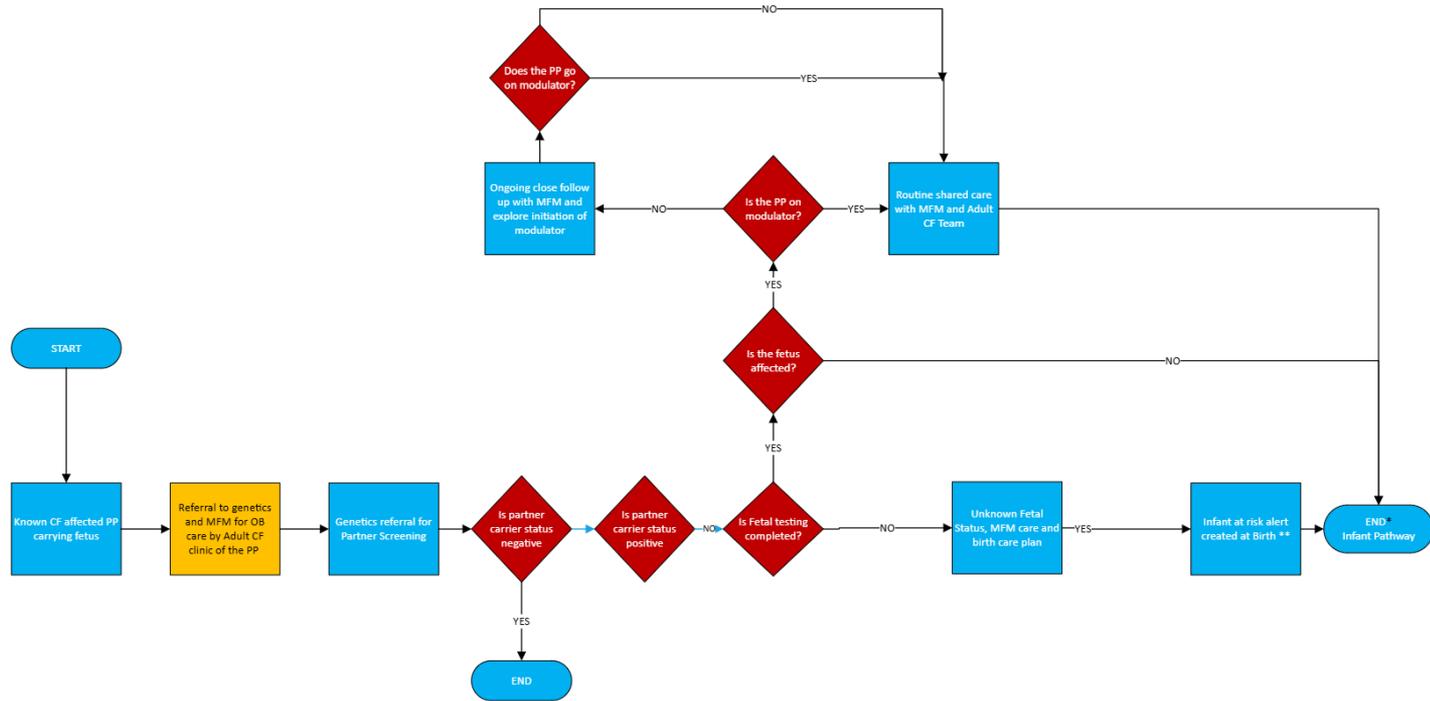
Legend and Notes:

- CF: Cystic Fibrosis
- FDC: Fetal Development Clinic
- MFM: Maternal Fetal Medicine
- MW: Midwife
- OB: Obstetrics
- PCP: Primary Care Provider
- PP: Pregnant Person

RED Shapes - Patient
 BLUE Shapes - Children's Hospital

**** need to determine who is going to order modulator for pregnant person and who will do LFT monitoring for the PP. This is discussion to have with MFM**

Ideal State Process Map - Known CF Affected Pregnant Person



Legend and Notes:

- PP: Pregnant Person
- CF: Cystic Fibrosis
- MFM: Maternal Fetal Medicine
- OB: Obstetrics
- LFT: Liver Function Test

Color Coding:

- RED Shapes - Patient
- BLUE Shapes - Children's Hospital
- YELLOW Shapes - Adult Hospital

Notes:

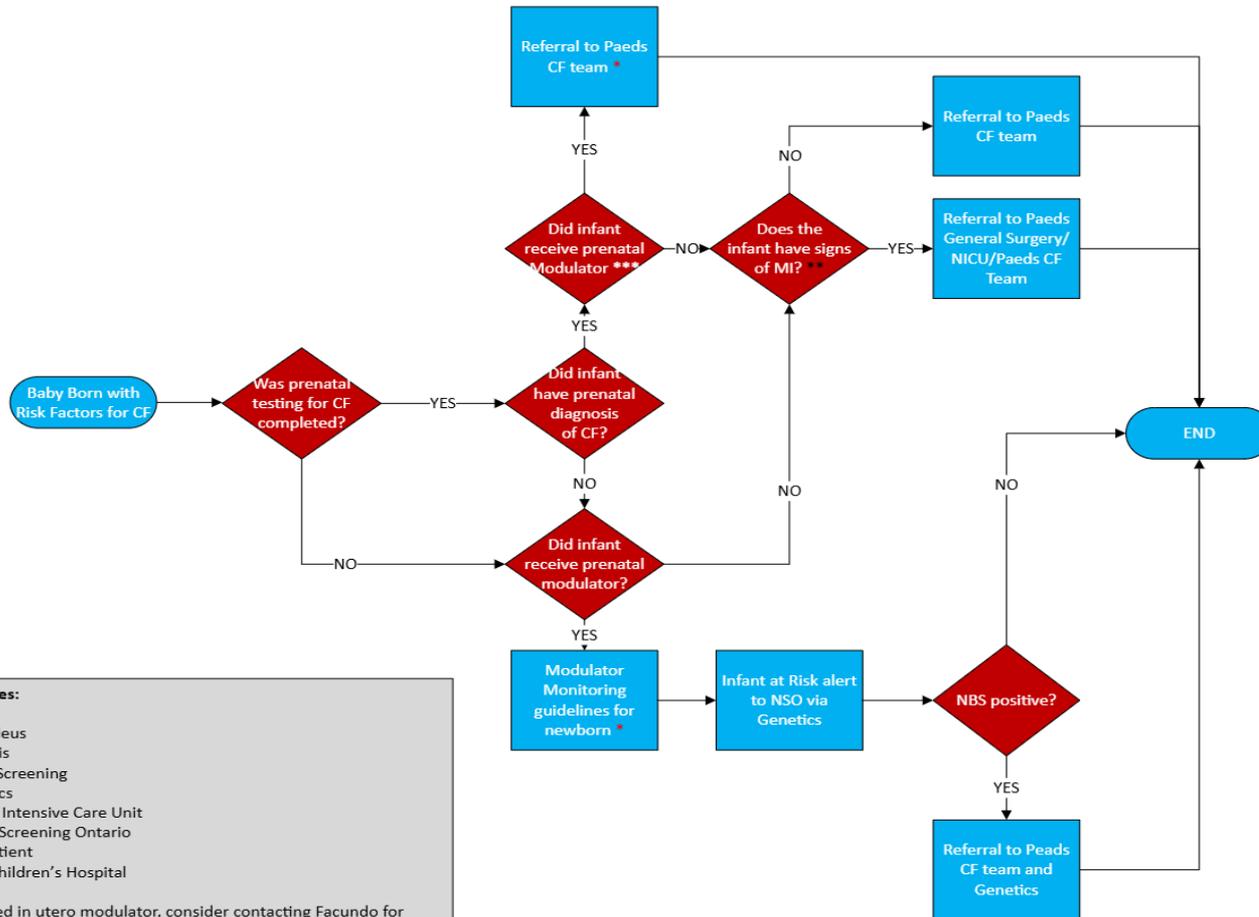
- **END please see infant pathway
- Refer to Infant Monitoring Modulator Guidelines



Infant Pathway

- Care of the infant at birth
 - Key stakeholders: addition of NICU and Paeds General surgery and Paediatricians
- Takes into consideration both known CF diagnosis and unknown
- Management plan for infants exposed to Trikafta
 - Alerts to NSO for at risk infant
 - Management plan for suspected Meconium ileus
 - Paediatric Respiriology to be involved

Infant At Risk Pathway



Legend and Notes:

MI: Meconium Ileus
 CF: Cystic Fibrosis
 NBS: Newborn Screening
 Paeds: Paediatrics
 NICU: Neonatal Intensive Care Unit
 NSO - Newborn Screening Ontario
RED Shapes - Patient
BLUE Shapes - Children's Hospital

*if infant received in utero modulator, consider contacting Facundo for clinical pharmacy study. Please refer to neonatal modulator monitoring guidelines

**if any child who presents to hospital with Meconium Ileus, Paediatric Respiriology should be consulted

*** please see prenatal birth plan from MFM

Implementation

Present pathways to OB/MFM and Paediatrician group

Narratives to go with each pathway

Creating condensed versions of each pathway for easier understanding

Have pathways available in OB/MFM clinics, PPCU for paediatricians

Next steps would be regional education/implementation through partnership with our OB/MFM group and regional MNCYN educators

Questions



Thank You.

A decorative graphic consisting of two overlapping circles, one light blue and one teal, with a small orange dot positioned to the right of the teal circle.