Recommendations for the follow-up of pregnancies and newborns of mothers who are receiving cystic fibrosis transmembrane conductance regulator (CFTR) modulators

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- Ivacaftor (Kalydeco®)
- Lumacaftor/Ivacaftor (Orkambi®)
- Tezacaftor/Ivacaftor (Symdeko ®)
- Elexacaftor/Tezacaftor/Ivacaftor (Trikafta®)

The following suggested recommendations are intended for use by obstetricians/midwife, family physicians, adult CF clinics, pediatric CF clinics, CF newborn screening programs and other healthcare providers.

The following suggested recommendations are for follow-up of pregnant patients who are taking any of the above medications for treatment of cystic fibrosis (CF). Preliminary data about the effect of these cystic fibrosis transmembrane conductance regulator (CFTR) modulators on the unborn baby are preliminary but reassuring. There are two elements for CF clinics to consider:

1. **Neonatal screening for cystic fibrosis in newborns born to mothers on a CFTR modulator:** Newborn screening for CF cannot be considered reliable because of the effect of CFTR-modulators on the newborn infants' immunoreactive trypsinogen (IRT) (first stage of newborn screening) and on the subsequent sweat test if necessary. To avoid a false-negative neonatal screening and/or sweat test result for CF, it is recommended to proceed to CFTR analysis irrespective of the IRT value. Please contact your local pediatric CF clinic or newborn screening program on the best way to proceed.

2. **Monitoring the newborn of a mother on a CFTR modulator:**
   As a precaution, it is recommended to perform liver function tests (AST, ALT, GGT, bilirubin) for the infant at birth, then at one month, and three months after birth, **IF** the mother chooses to breastfeed. It is also suggested to carry out an ophthalmological examination within the first 2 months after delivery due to the low risk of developing cataracts.

It is important for everyone to remain vigilant on this expanding population of infants and mothers. Please contact your local pediatric cystic fibrosis clinic team for further questions and advice.

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Reviewed by: Canadian CF Newborn screening group.
Endorsed by: Cystic Fibrosis Canada's Healthcare Advisory Council.