The Fertility and Pregnancy Journey with a Fontan Circulation – personal experiences and best practices

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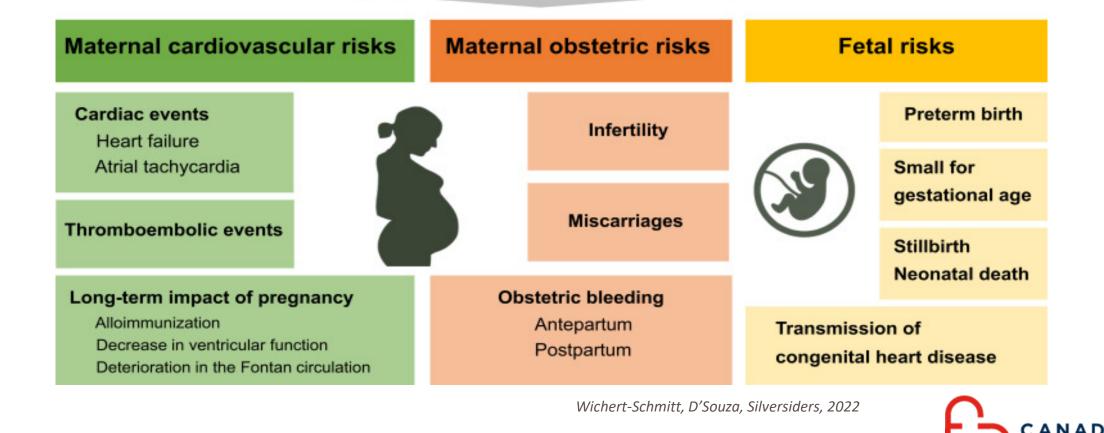


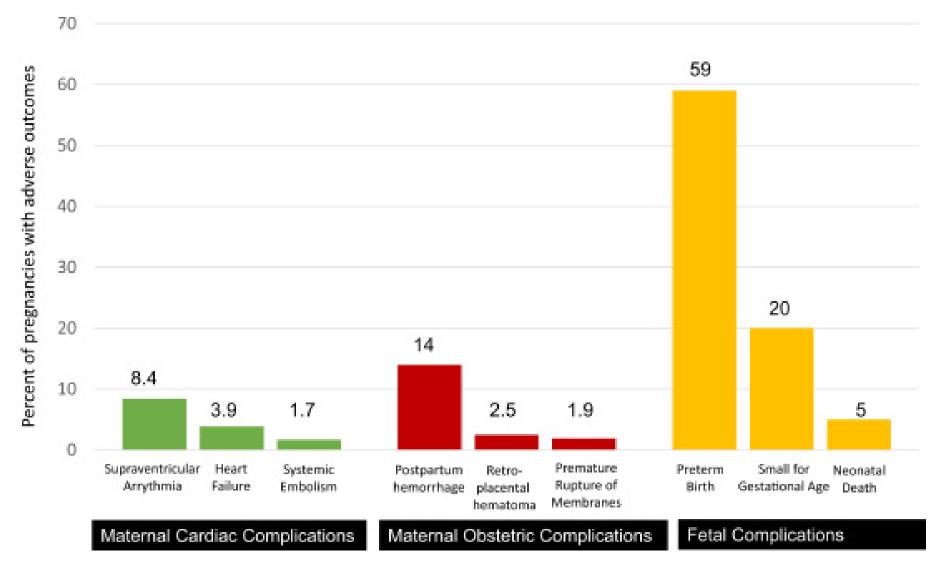
Hemodynamic changes of pregnancy

Increased plasma volume Increased cardiac output Increased heart rate Hypercoagulable state

Fontan physiology

Limited ability to increase cardiac output Hypoxemia Increased central venous pressure Hypercoagulability/antithrombotic therapy





Wichert-Schmitt, D'Souza, Silversiders, 2022



Hemodynamic changes in pregnancy

	Pregnancy	Peripartum (during L&D)	Postpartum
Blood volume	↑ (40 – 50%) and up to 67% for twin pregnancy	1	↓(auto diuresis)
BP	\downarrow	1	↑
Heart rate	↑ (10 – 15 beats/min)	1	Ļ
Cardiac output	↑(30-50%) and an additional 10-20% for twin pregnancy	1	\downarrow

ESC Guidelines. 2011



Pre pregnancy assessment:

- Pregnancy ought to be a planned event in individuals with a Fontan circulation.
- Pre pregnancy appointment in cardiac obstetrics (COB) clinic:
 - Status and history of any Fontan complications (Heart Failure, arrhythmias, blood clots)
 - Physical assessment (weight, cyanosis, oxygen saturations)
 - Heart Rhythm (ECG)
 - Echocardiogram (heart function assessment)
 - Cardiopulmonary exercise test (exercise capacity)
 - Cardiac MRI (to look for any Fontan circulation obstructions)
 - Assessment of liver and kidney function



Fertility Challenges in women with Fontan circulation

- Women with a Fontan circulation have challenges conceiving & maintaining pregnancy related to:
 - menstrual irregularities,
 - Fontan circulation +/- complications,
 - hormonal alterations,
 - morphologic uterine abnormalities
- Overall reported rate of miscarriage ranges between 40 70% (compared to ~ 15% in general population). Potential causes:
 - Lower oxygen levels in the blood decreases the ability of the fetus to survive
 - Fontan complications such as abnormal valve function or heart failure
 - Pregnancy terminations reported to occur ~7 10% -- may also contribute to low live birth rates
 - The patient's cardiac status should be optimized prior to pregnancy (heart valve issues should be repaired, heart rhythm corrections [ablations] should occur prior to pregnancy)



Potential options when fertility is a problem

- In vitro fertilization (IVF)
- Surrogacy



7

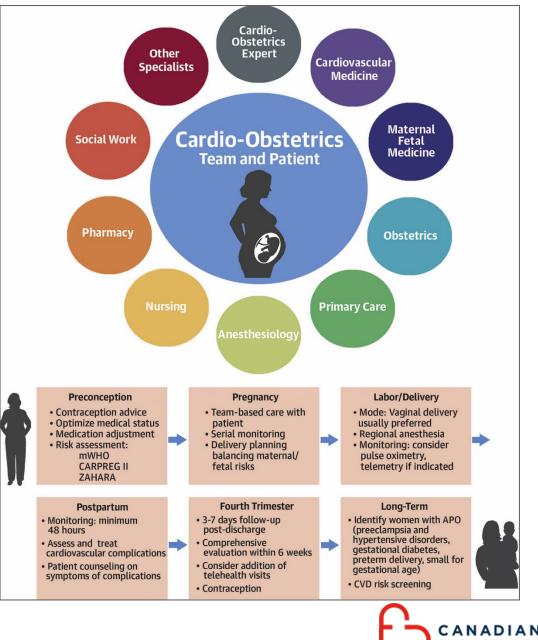
What to expect before and during pregnancy

- Medications:
 - Some medications cannot be used during pregnancy, so alternative medications may be recommended.
- Frequent visits with Obstetrician and Cardiac Obstetrics Team
- Increased surveillance during pregnancy:
 - Blood tests (eg INR)
 - Echocardiograms
 - Fetal echocardiogram
 - Obstetrical ultrasounds



Pregnancy Management

- Pregnancy care should be provided by an experienced cardio-obstetric team
- Monitoring plan should be established early in pregnancy
- A labour and delivery care plan should be prepared by the multidisciplinary cardioobstetric team and circulated well before delivery
- Availability of experienced cardio obstetrics team will determine delivery location



Recommendations

- - good physical condition (including Fontan circuit)
 - healthy lifestyle
 - maintain normal weight
 - avoid risky behaviours -- such as alcohol consumption, smoking



 Fontan deterioration time dependent, complications impact ability to carry viable pregnancy -- ?? benefits of considering pregnancy at younger age (if possible!)



Contraception

- Estrogen containing formulations are <u>contra-indicated</u> because they can cause blood clots to form in the Fontan circuit.
- Safe & reliable to use:
 - Progesterone only contraceptive methods (pills, injectables & implants)
 - Copper and hormone IUDs
 - Emergency contraception → progesterone pill within 72 hours, progesterone receptor modulator within 120 hours, or placement of copper IUD



Diana's Journey

Began family planning a few years in advance:

- Experienced some fertility issues
 - underwent fertility treatment
 - IVF (In vitro fertilisation) to address fertility issues
- Obstetrical Complications
 - obstetrical bleeding
 - preterm delivery
- 2 Successful births and 2 healthy children

