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

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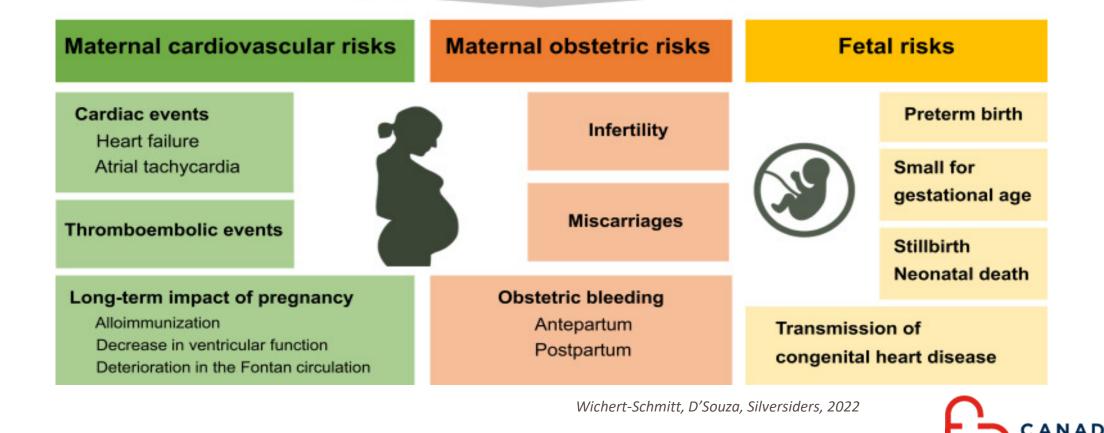


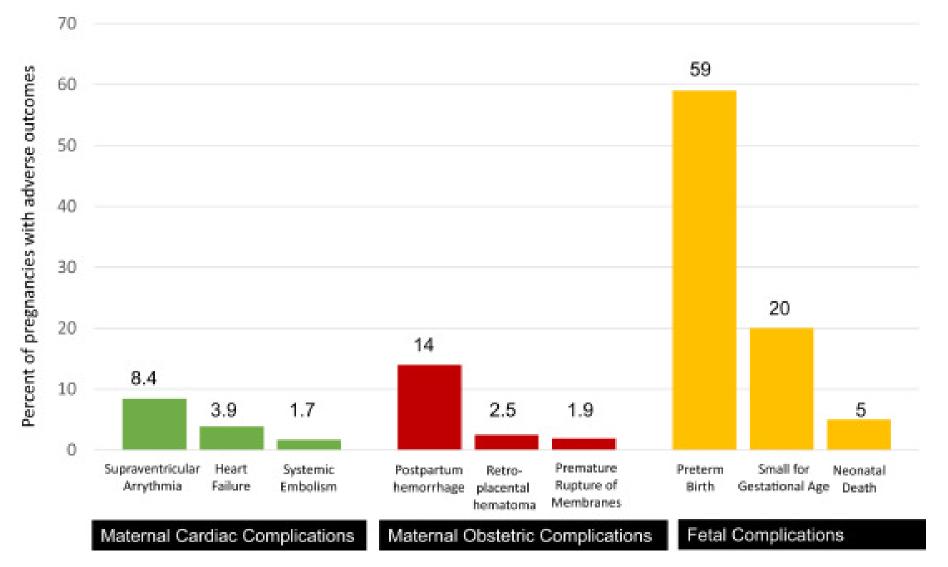
#### Hemodynamic changes of pregnancy

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



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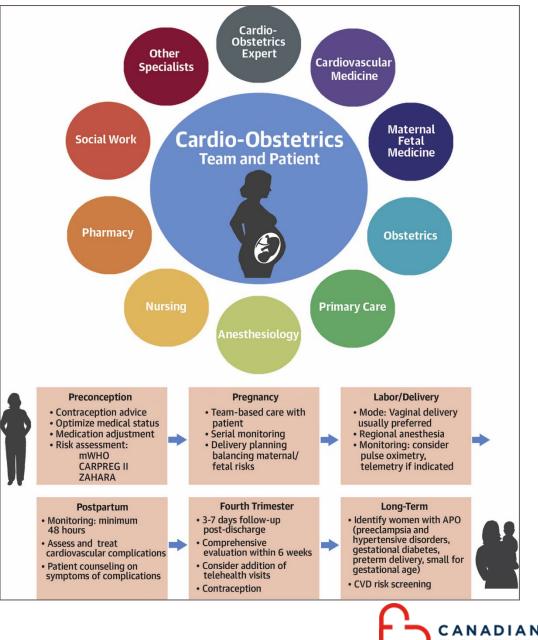
## What to expect before and during pregnancy

- 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

