

Fontan Circulation and Gynecologic Care Across the Life Span

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with
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Gynecologic Care Across the Life Span:

- Pubertal development and onset of menstruation
 - Onset may be later
- Menstrual cycle concerns, contraception and fertility planning
 - Development of regular cycles – can be affected by body weight, stress, illness
 - Contraception – options available affected by menstrual pattern, clot risk
 - Fertility – studies suggest lower ovarian reserve, higher need for assisted conception
- Pregnancy, delivery and postpartum care
 - Prepregnancy cardiac assessment and team development
 - Monitoring of cardiac function, rhythm, fluid balance; fetal growth and placental function
- Perimenopause and menopausal transitions
 - Earlier onset, heavy bleeding secondary to venous pressures
 - Therapeutic options limited by special considerations for Fontan circulation

Special Considerations for Puberty:

- Delayed onset of growth spurt, pubertal development in some studies
 - May be related to being born smaller for dates/premature
 - May also be related to decrease ovarian reserve
 - Investigations with hormone and genetic testing if more than 3 years behind peers
- Menstrual cycle onset may be later than peers
 - Heavier flow common – often treated by GPs, peds gynecology

Menstrual Cycles:

- Cycle pattern
 - Irregularity of periods more common
 - Ovulation cadence less regular
 - Longer cycles tend to be heavier
 - Intermenstrual spotting
- Heavier menstrual bleeding
 - More common in higher hemoglobin levels, worse ventricular function
 - Need to rule out other common causes
 - Fibroids, polyps, uterine anomalies
- Management options for management depend upon
 - Underlying thrombosis risk/use of anticoagulation
 - Desire for future fertility

Contraceptive Concerns:

- Condoms will not address menstrual concerns but only one with STI protection
- Menstrual flow may limit utility of copper IUD
 - Often increase blood loss with cycles
- Estrogen containing contraceptives carry a risk of clotting
 - Superior control of heavy bleeding for some patients on anticoagulation for short term
- Progesterone only contraceptive generally an excellent option
 - May be oral, implantable, IUD, injection
 - Good control of heavy bleeding but may have increase in spotting
 - May be used as emergency contraception
 - Common side effects include breast tenderness, mood changes

Fertility Planning and Concerns:

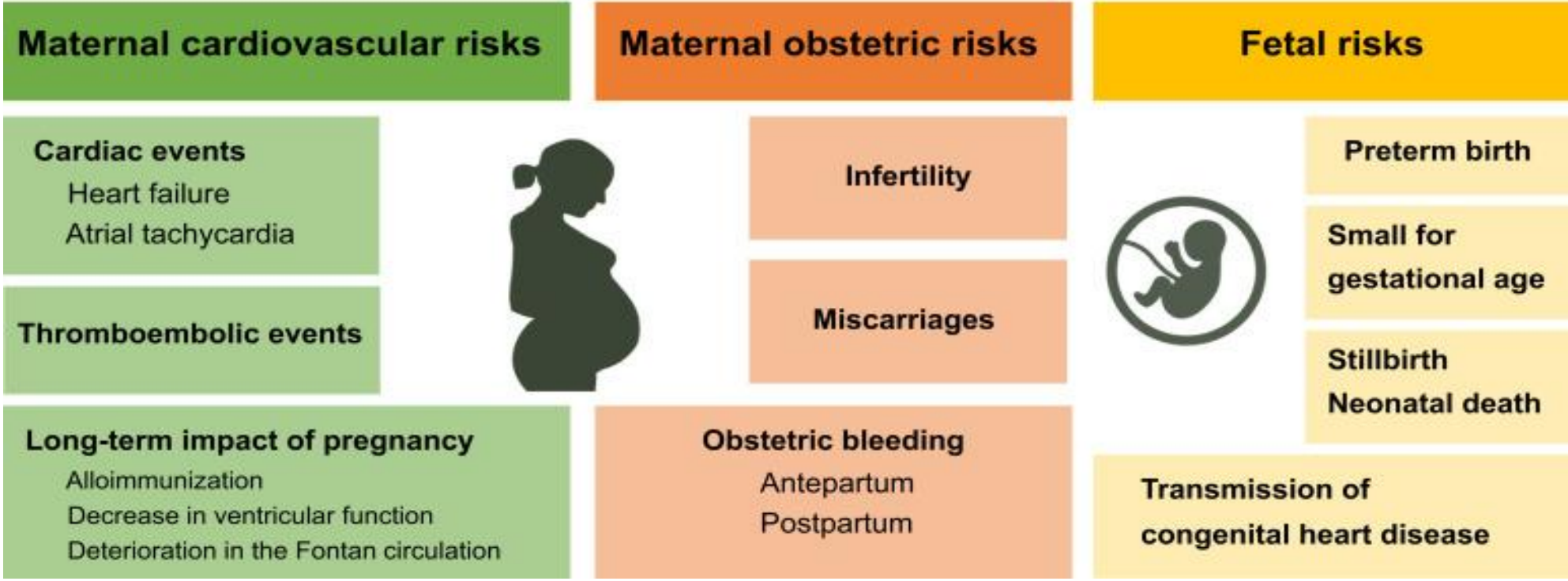
- Spontaneous conception is possible if having cycles with ovulation
- Delayed conception common due to multiple reasons
 - Impaired ovulation /ovarian reserve
 - Impaired uterine blood flow/oxygenation
- Fertility assistance options carry own risks and benefits
 - Ovulation assistance – may cause higher estrogen and clot risk
 - IVF increases the risk of blood clotting / bleeding around hormone stimulation
 - Fluid overload risks can be decreased by using single frozen embryo

Pre pregnancy assessment:

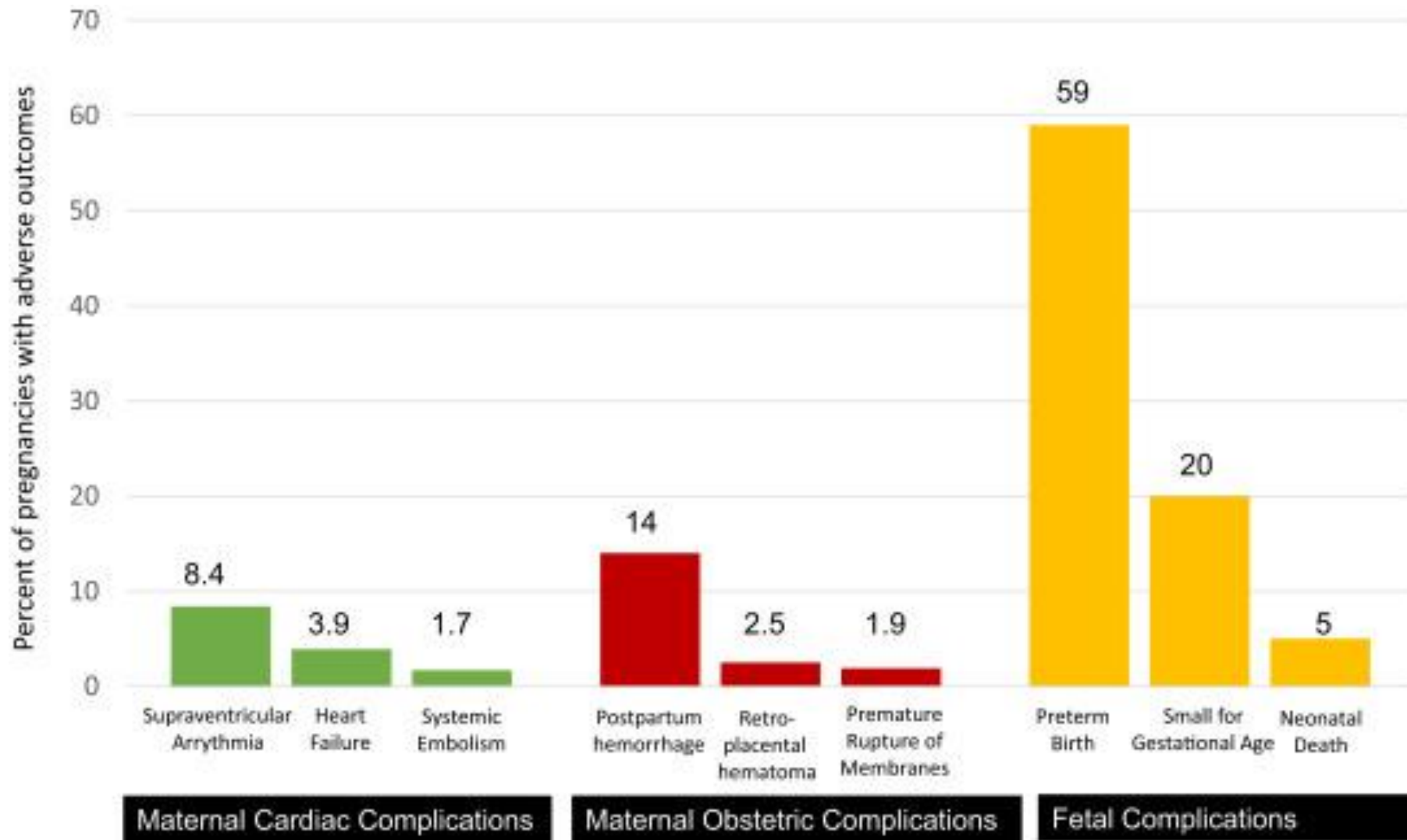
- Pregnancy ought to be a planned event in individuals with a Fontan circulation.
- Pre pregnancy appointment in cardiac obstetrics (COB) clinic:
 - Status and history of any Fontan complications (Heart Failure, arrhythmias, blood clots)
 - Physical assessment (weight, cyanosis, oxygen saturations)
 - 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

- Increase in rate of miscarriage to 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
- Increased rates of bleeding in first and second trimester
 - May settle spontaneously or be ongoing



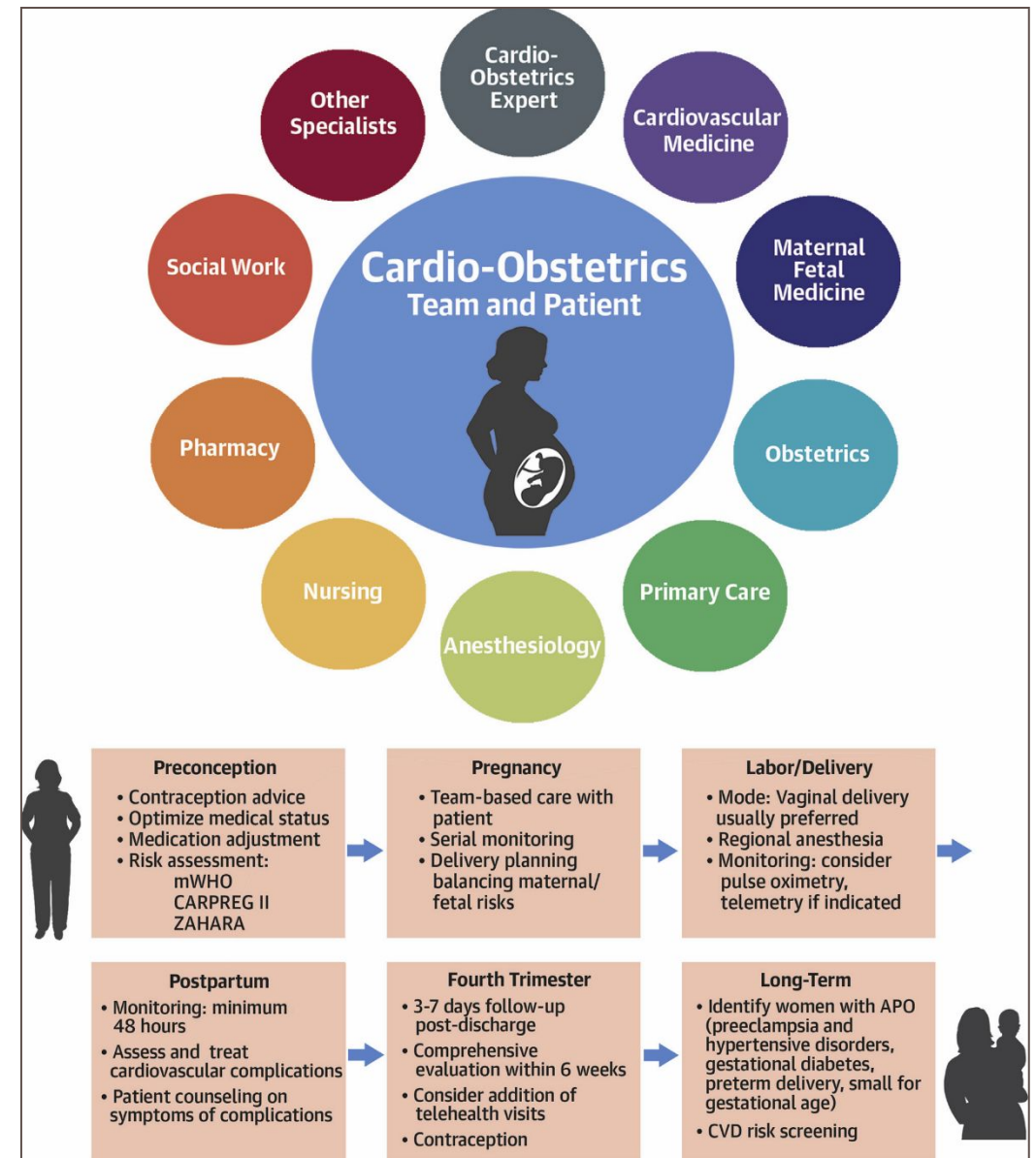
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Pregnancy Management

- 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 cardio-obstetric team and circulated well before delivery
- Availability of experienced cardio obstetrics team will determine delivery location



The Fourth Trimester: Postpartum Concerns

- Most patients will be on anticoagulation postpartum even if not during pregnancy
 - Vaginal bleeding may be prolonged or heavier
- Breastfeeding should be unaffected
 - Milk production can be reduced if diuresis is needed for fluid overload
 - Some medications for heart failure may be not recommended for breastfeeding
- Progesterone only contraception safe for both Fontan and for breastfeeding

Diana's Experience

- Modified Fontan in 1989
- Began family planning in early 30's
 - Experienced fertility challenges
 - One spontaneous conception, infertility, several miscarriages
 - underwent fertility treatment
 - IVF (In vitro fertilization) to address fertility issues
 - No cardiac complications
 - Obstetrical Complications
 - obstetrical bleeding/hematoma
 - Premature rupture of membranes
 - 2 healthy children !



Menopause and Perimenopause

- Menopause is diagnosed after 12 months of absent menstruation
 - Any bleeding after this must be investigated (biopsy, ultrasound)
 - Average onset is 51 but is earlier in Fontan circulation
- Symptoms may arise up to 5-10 years prior to cessation of menstruation
 - Menstrual irregularity increases
 - Hot flushing/sleep disturbance/concentration
 - Pelvic floor changes – bladder function/sexual function/vaginal dryness
 - Also reported – joint pain, weight gain, mood changes
- Hormone therapy (primarily estrogen) is first line for symptoms but does carry risk
 - Low dose vaginal estrogen is safe and can help pelvic floor issues
 - Low dose transdermal estrogen has less clot risk than oral tablets
 - Would need to balance with cardiology the risks
 - Progesterone needed for prevention of uterine cancer unless hysterectomy performed

Troublesome Bleeding

- Irregular and heavy bleeding is common in the perimenopause
 - Investigations include: blood testing, ultrasound, Pap testing, endometrial biopsy
- Options for management depend upon
 - Age at onset and fertility desires
 - Presence of other conditions – endometriosis, fibroids, breast pathology
- In general hormonal and surgical options are the mainstay
 - Hormonal options are as per contraception (Mirena, oral progesterone) with possible addition of estrogen for perimenopausal symptoms

Surgical Options for Troublesome Bleeding

- Hysteroscopy
 - Involves looking inside the uterus and possible removing lesions causing bleeding
 - Day care procedure and carries little anaesthetic / cardiac risk
 - Pregnancy possible following completion
- Endometrial ablation
 - The lining of the uterus is burned to prevent periods
 - Has a longer effective period in 40s, does not allow pregnancy
- Uterine artery embolization
 - Blocks blood flow to the uterus and decreases blood flow
 - Radiologic procedure with minimal bleeding risk
 - Not compatible with pregnancy, not helpful in pelvic pain
- Uterine fibroid treatment or hysterectomy
 - Requires general anaesthetic which carries some cardiac risk
 - Laparoscopy may not be well tolerated for longer procedures due to pressures in the abdomen
 - Allows treatment of other problems (endometriosis, pelvic pain, scar tissue)

Reena's Experience

- Fenestrated Fontan in 1995, fenestration closure and pulmonary stent followed
- Heavy menstrual bleeding, debilitating cramps, migraines in teens, 20's, 30's and now
 - Birth control prescribed but problematic - caused increased blood pressure; stopped anticoagulants for a few years but did not help
 - 2013 - endometrial ablation to remove cysts and fibroids followed by 10 years of no periods/complications
 - 2023 - perimenopause symptoms and return of heavy bleeding and debilitating cramps
 - Prescribed HRT, discussed surgical options
 - Scheduled for hysterectomy in May 2024

Take Home Points

- Ovarian function and menstrual cycles are affected by the Fontan circulation
- Pregnancy should be planned and followed by an expert team to get the best outcomes
- Contraception is important for timing of pregnancies and management of heavy bleeding
- Menopause may come earlier and treatment options are altered by cardiovascular risk