

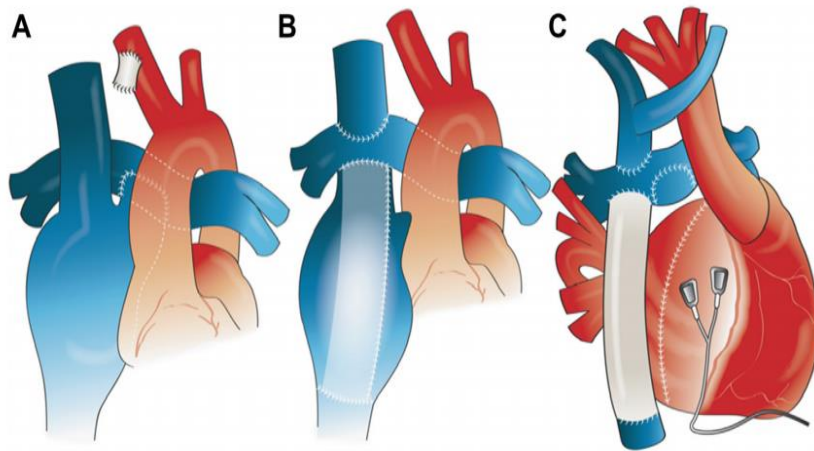


# Pregnancy in patients after Fontan operation

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# The Fontan procedure: palliative surgery for univentricular heart



Modified  
classic  
Fontan

Intracardiac  
lateral tunnel  
Fontan

extracardi  
ac Fontan

- 2% of life births with congenital heart disease (French registry EPICARD source)
- Great anatomic variety
- Significant physiologic alteration

# Expected complications

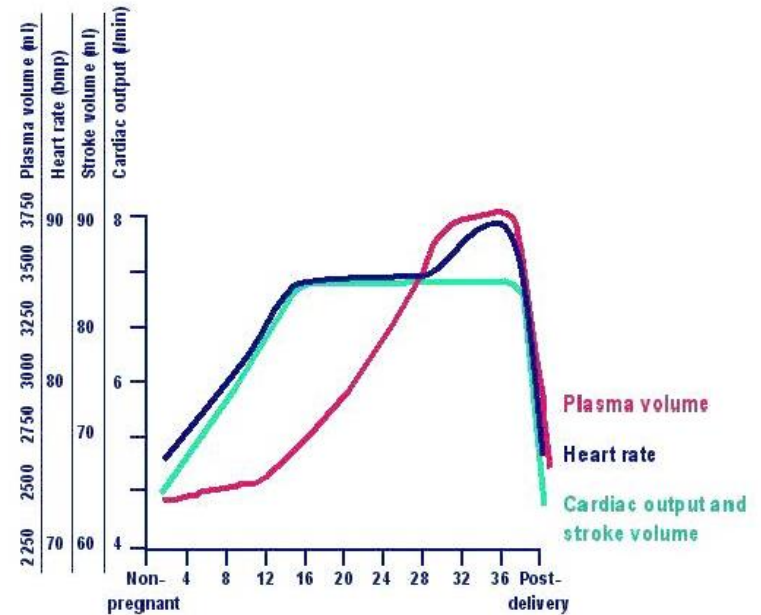
## During long-term FU:

### High prevalence of complications

- Atrial arrhythmia: 60%
- Heart failure-Failing Fontan
- Thromboembolism: 20%
- Systemic complications:
  - Worsening hypoxemia
  - Hepatic fibrosis (cirrhosis, Hepato-cellular carcinoma)
  - Protein losing enteropathy

## During pregnancy

### Expected complications



- + hypercoagulable state
- + Decrease systemic arterial resistance

# Observed cardiac complications

Series	n patients	n pregnancies	CV complications during pregnancy and PP		
			Atrial arrhythmia	Heart failure	Thromboembolism
Siu et al; 2001	5	5	2	0	0
Drenthen et al,2006	6	10	1	0	0
Gouton et al, 2015	37	59	3	3	2
Pundi et al, 2015	35	70	7	7	0
Cauldwell et al, 2016	14	42	3	0	1
Total	97	186	<b>16 (8.6%)</b>	<b>10 (5.3%)</b>	<b>3 (1.6%)</b>

Arrhythmia: 64% prior history of atrial arrhythmia

# Obstetrical outcomes

Studies	patients/ pregnancies	Obstetrical complications				
		Miscarriages	PRM	Premature labor	PE	PPH
Drenthen et al 2006	6/10	5*	1	0	0	1
Gouton et al 2015	37/59	16*	7	4	1	3
Pundi et al 2015	35/70	35*	?	?	?	?
Cauldwell et al 2016	14/42	28	0	0	0	6
Total	92/181	<b>84 (49%)*</b>	<b>8 (9.3%)</b>	<b>4 (4.7%)</b>	<b>1 (1.2%)</b>	<b>10 (11.6%)</b>

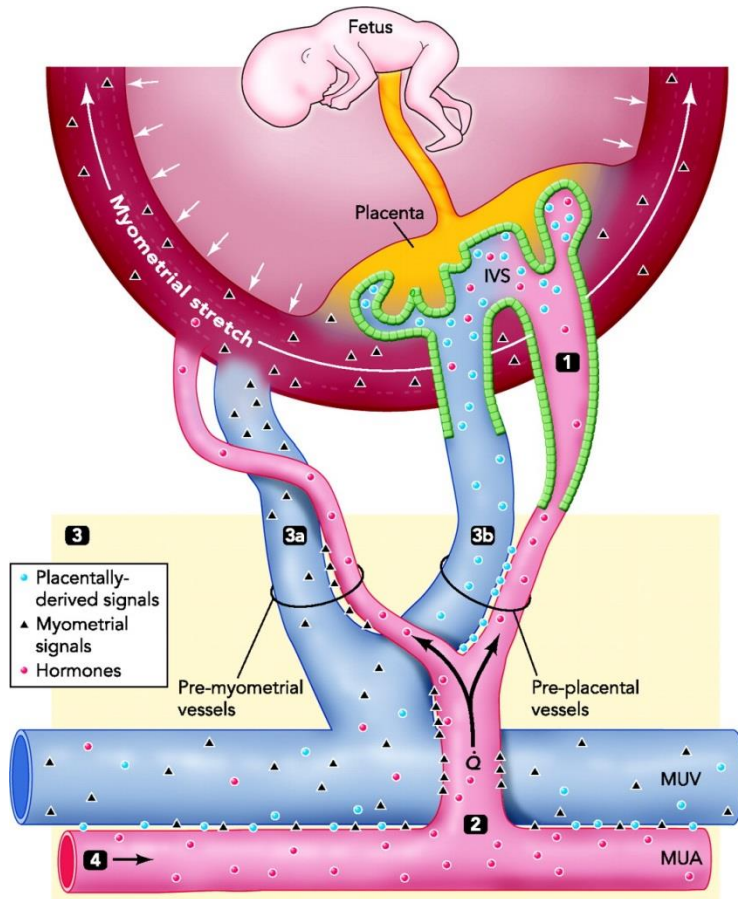
Miscarriages and unsuccessful conception are related to maternal cyanosis and CARPREG score

# Fetal/neonatal outcomes

Outcomes	Rates
Live births	49%
Mean term	33WG
Mean birth weight	1912g (10th percentile)
intra-uterine fetal death	1.2%
Still birth	1.2%
Prematurity	79%
SGA	16.7%
Neonatal death	4.8%
Recurrence of CHD	4.7%

- Viable pregnancies are possible in 54 to 78% of patients
- But:
  - High rate of **prematurity**
  - High rate of SGA
- Neonatal death related to prematurity

# Utero-placental flow



George Osol, and Maurizio Mandala *Physiology*  
2009;24:58-71

Table 1. Processes, pathways, signals, and events implicated in maternal uterine vascular remodeling during pregnancy

## Processes

- Implantation/placentation
- Endovascular trophoblast invasion
- Myometrial stretch
- Altered endocrine regulatory mechanisms

## Pathways

- Local
  - Physical (e.g., increased shear stress on endothelium)
  - Molecular (e.g., venoarterial exchange)
- Systemic (humoral/endocrine)

## Signals

- Sex steroids
- Nitric oxide
- Prostaglandins
- Growth factors and angiogenic molecules
- Angiotensin
- Other (?)

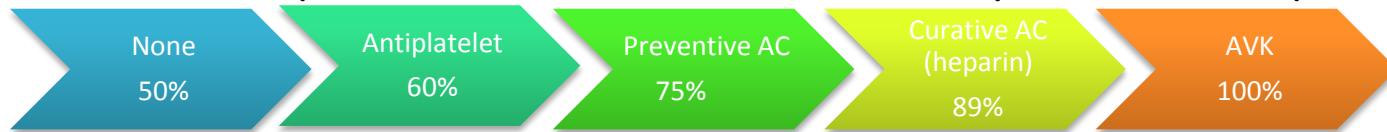
## Events occurring within the vascular wall

- Vasodilation
- Cellular hypertrophy
- Cellular hyperplasia
- Matrix remodeling
- Recruitment of periadventitial cells (?)

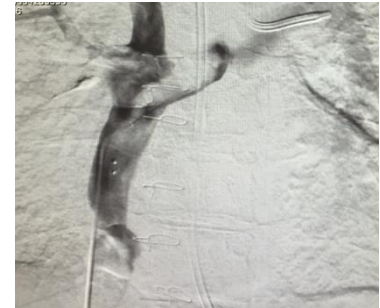
*Physiology*

# Management: Anticoagulation or not?

- Relationship with miscarriages is debated
  - Most miscarriages were under antiplatelet or preventive AC therapy
  - But, 63% of miscarriages were under curative AC
  - AC may be an additional risk of prematurity



- Thromboembolic risk : 3 cases
  - 2/3 Pulmonary embolism
  - 2/3 without anticoagulation
  - Could be fatal
- Hemorrhagic complications are not related with the use of AC





# Fertility

- Menstrual cycle disorders : delayed menarche
- High incidence of amenorrhea
- High infertility rate : 21%
- Therapeutic may also carry risks: ovarian stimulation
  - estrogen stimulation : thromboembolic risk
  - ovarian hyperstimulation syndrome.



# Follow-up

- No evidence of an accelerated deterioration,
- The impact of pregnancy on the natural history of a Fontan circulation is still uncertain



# Conclusion

- Pregnancy in women with Fontan palliation is possible, without maternal mortality. **However:**
- **High risk for the mother** with arrhythmia, heart failure and TE
- **Very high risk for the fetus** : miscarriages, prematurity , SGA.
- Anticoagulation therapies during pregnancy are still debated:
  - thromboembolisms can threaten Fontan circulation
  - We suggest **at least prophylactic anticoagulation**
- **Preconception counseling** is highly recommended
- strict **multidisciplinary monitoring** by obstetrician, cardiologist and anesthetist is essential.