



Pregnancy outcomes in Eisenmenger syndrome: a French multicentric cohort study.

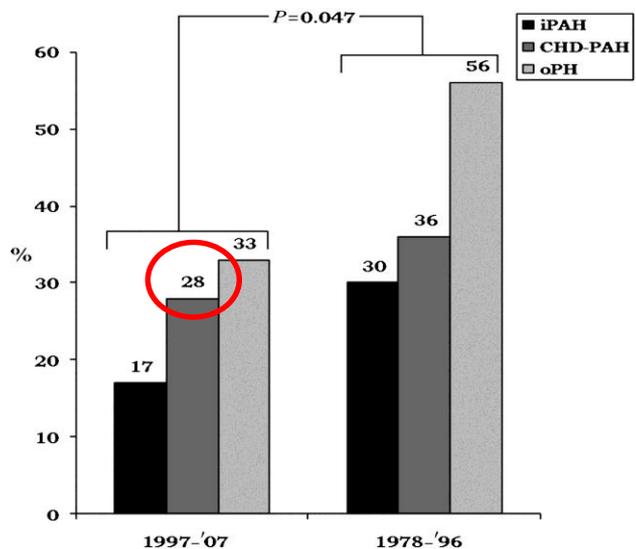
Magalie Ladouceur, M.D.

Centre de référence des Malformations Cardiaques
Congénitales Complexes-M3C, Necker-Hôpital Européen
Georges Pompidou.



Pregnancy and PAH-CHD

- Physiological changes during pregnancy and the peripartum are poorly tolerated : high mortality rate



- Fourfold higher risk of death compared to healthy women (NIS data)
- Reduction of maternal mortality (review from 1997 to 2007)
- but **remains prohibitively** high
- Pregnancy in this condition is still strongly discouraged and termination should be indicated

Opotowski AR et al. Heart 2012

Bédard E et al. Eur Heart J 2009

Baumgartner H et al. Eur Heart J 2010

Hypothesis and aims

- **Hypothesis** : changes in PAH treatment and high-risk pregnancies management could improve outcomes of pregnancy in Eisenmenger syndrome
- **Aims** : To determine management and outcomes of pregnancy in Eisenmenger Syndrome
 - Cohort of patients followed in tertiary centers
 - With the new approaches (>1997)

Methods

- Pregnant patients with ES followed up in 7 French tertiary centers from 1997 to 2015 (median of inclusions =2006)
- All pregnancies were counted :complete (> 20WG), miscarriages, first trimester elective abortions, and terminations
- Information covered ante-, per- and post partum (6 weeks after delivery), and last FU
- outcomes :
 - maternal cardiac and obstetrical complications:
 - Fetal/neonatal complications

Study population : baseline

Variables (n=20)	
age at first pregnancy , mean±SD	26±6
Simple/ complex CHD, n	9/11
BMI, mean±SD	19,2±2,7
NYHA functional class, n (%)	
I	4 (20%)
II	13 (65%)
III	3(15%)
IV	0 (0%)
Saturation (%), mean±SD	87±6
Hemoglobin (g/dl), mean±SD	17.0±1.7
Hematocrit (%), mean±SD	49.8±6.5
Pulmonary arterial pressure (S/D/M), mmHg, mean±SD	101±16/43±13/62±1 3

Maternal complications : cardiac

- Mortality : 5% (one patient)
- Cardiac complications: 33% (n=6)
 - worsening of hypoxemia (n=5),
 - **Severe** heart failure (n=4).
 - All heart failures except one occurred during the **early postpartum** period.

Predictive factors

	no complication (n=12)	cardiac complications (n=6)	p value
Basal characteristics			
Age, median [IQ]	25 IQ[22-27]	30.5 IQ[23-32]	0.3
Hemoglobin, mean±SD	16,1±0,8	19,4±0,8	0.01
Saturation, mean±SD	89,3±3,8	79,6±4.1	<0.01
NYHA ≥3	1 (7.7%)	1 (16.7%)	—
Complex CHD, n (%)	8 (66%)	4 (66%)	0.3
Obstetrical management			
Obstetrical complications, n (%)	4 (33%)	2 (33%)	—
mode of delivery (CS /vaginal)	6/5	6/0	—
anesthesia (general/epidural/spinal)	2/4/4	1/2/3	—
Treatment during pregnancy			
Anticoagulation therapy	5	2	—
Advanced PAH therapy*	1	4	—

* Sildenafil (2), Tadalafil (1), NO (2), Prostacyclin (1), Bosentan (0)

Obstetrical outcomes

- 8 elective abortion (mean term of 8.5 WG, 5 to 15WG) : no complication
- 2 miscarriages
- Eighteen (64%) pregnancies were completed
 - mean duration of 33 ± 3 weeks, (28 to 38 weeks).

Obstetrical management and complications

- Mode of delivery : 67% caesarean section (n=12)
 - 66% planned CS (n=8): cardiac reasons (n=7)
 - Planned vaginal deliveries in 1 patient for SGA
- General anesthesia : 18%;
 - 35% epidural and 45% spinal anesthesia
- Preventive antibiotic for IE : 7 patients; no maternal infectious complication
- Obstetrical complications occurred in 25% (n=7)

Obstetrical complications	n
Preeclampsia	1
Premature rupture of membranes	1
Preterm labor	3
Abruptio placentae	2
Postpartum hemorrhage	4
Diabetes mellitus or infection or gravid hypertension	0

Fetal/neonatal complications

Fetal/Neonatal outcomes and complications	n (%)
Live births, n (%)	18 (64%)
SGA (<10 percentile)	7 (39%)
Birth weight (g), mean \pm SD	1717 \pm 487
Intrauterine fetal death or stillbirth	0
Neonatal death	0
Prematurity (birth<37 WG)	14 (78%)
33-36+6 WG	8
28-32+6 WG	6
22-27+6 WG	0
Recurrence of CHD:	2 (11%)



- patients SGA+ had a significantly lower baseline saturation (85% vs. 90% p=0.04)
- Term was significantly shorter in women with cardiac complications: 30,2 \pm 1,5 vs. 35, 9 \pm 1,6 (<0.01)

Follow-up

- median follow-up of 8.8 years 95%CI[3.7-12.7],
- 2 patients died (37 and 45 years old), 4 and 16 years after delivery.
- NYHA functional class impaired in 7 patients
 - PAH therapy was started or strengthen at a median delay of 4.5 years,
- Moreover, 3 patients experienced arrhythmia and 2 haemoptysis during follow-up.

Conclusion

- Outcomes of pregnancy in women with ES may be improved in the modern management era
- However the severity of maternal complications and the high rate of prematurity and SGA remain substantial.
- Pregnancy must be avoided or early terminated in females with ES.
- If such females refuse termination or want to become pregnant,
 - risk assessment in an experienced centre : severe PAH
 - Patient (and her partner) must be fully counselled.
- Management in an experienced multi-disciplinary team.