

Maladie de Gaucher : ERT

Diminution, espacement, grossesse

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Grossesse et Gaucher

- Les données de l'expérience française n'ont pas été reproduites ici avant publication

Conséquences

- Avortement spontané
- Aggravation de la maladie de Gaucher
(anémie, thrombopénie, crises osseuses)
- Hémorragies du post partum
- Infections du post partum

Traitements de la maladie de Gaucher et grossesse

- Miglustat (Zavesca®) : contre indication
- ERT (Cerezyme®) :
 - pas de consensus
 - Pas d'étude randomisée
 - Études restrospectives et case report

La plus récente Elstein et al (2004) :

revue de la littérature

Comparaison 23 grossesses sous ERT/ 43 sans ERT

| Citations | Patients | Pregnancy | Delivery | Live births | Comments |
|-------------|----------|-----------|--------------------------|-------------|---|
| Eliot | 1 | 1 | Vaginal | 1 | Require transfusion |
| Bromberg | 7 | 13 | Vaginal | 9 | 4 induced abortions « because of Gaucher disease » |
| Decker | 2 | 5 | Vaginal | 4 | 1 therapeutic abortion |
| Teton | 1 | 4 | Vaginal | 4 | Uneventful |
| Greenwald | 3 | 3 | Vaginal | 3 | Uneventful |
| Hoja | 1 | 2 | Vaginal | 2 | Uneventful |
| Addleman | 1 | 3 | Vaginal | 3 | Uneventful |
| Watov | 1 | 4 | Cesarian | 3 | 1 sp.abortion all deliveries anemia, thrombopenia |
| Mandel | 1 | 3 | Vaginal | 2 | 1 sp.abortion |
| Houlton | 2 | 5 | Vaginal | 4 | 1 sp.abortion, 1 pregnancy hemorrhage, 3 N |
| Swinhoe | 1 | 1 | Cesarian | 1 | Infant small for gestational age |
| Young | 1 | 3 | 2 vaginal, 1 cesarian | 3 | Cesarian section was elective |
| Goldblatt | 21 | 11 | 16 vaginal 4 cesarian | 18 | 1 sp abortion, 2 severe anemia, 7 severe bleeding 1 cesarian section for fetal distress, septicemia died |
| Mazor | 1 | 1 | Vaginal | 1 | Portal hypertension |
| Schoenfeld | 1 | 3 | Cesarian | 3 | Spenectomuy during first pregnancy, adhesions of liver in 3rd |
| Zlotogora | 17 | 47 | 33 vaginal 5 cesarian | 38 | 9 sp. Abortions (5 pt), 6 pts required blood infusions (pregnancy), 5 pts required blood infusions (delivery) |
| Torjeman | 53 | 53? | ? | 53 | 3 women required blood transfusion in postpartum |
| Moughabghab | 1 | 1 | Cesarian | 1 | Endometritis |
| Granovski | 53 | 102 | 61 vaginal 9 cesarian | 72 | 25 sp. abortions, 2 intrauterine deaths, 8 pts with vaginal delivery 5 pts with cesarian section required multiple transfusion |
| Ayhan | 1 | 1 | cesarian | 1 | Uneventful |
| Santana | 1 | 1 | Vaginal | 1 | Portal hypertension, anemia |

TOTAL 190 302 264 (87,4 %) 30 sp abortion, 29 hemorrhages

**Elstein 26 43 31 vaginal 34 + 2 (86%) 3 sp abortion, 4 post partum hemorrhage
9 cesarian**

Obstetric characteristics of all pregnancies

| | Enzyme Treated | Untreated | P value |
|---------------------------------|-----------------------------|----------------------|-----------------------|
| No of pregnancies | 23 | 43 | |
| Spontaneous Abortion | 3 in 1 women | 1 in each 3 women | |
| Other outcomes | 1 ectopic pregnancy | 1 termination | |
| Twin pregnancies | 3 | 2 | |
| Infant due | 1 | 2 | |
| Antenatal hemorrhage | 0 | 0 | |
| Pregnancy induced Hemorrhage | 0 | 3 | Trend to significance |
| Postpartum hemorrhage | 49 (1 in a twin pregnancy) | 4 | |
| Preterm delivery | 2 | 5 | |
| Intrauterine growth restriction | 1 | 4 | |
| Vaginal delivery | 9 | 31 | |
| Cesarian section rate | 39% | 11.6% | 0.022 |
| Need for blood | 4% | 3% | |
| Live birth rate | 78.3% | 86% | NS |
| Postpartum febrile morbidity | 7/23 (30.4%) | 1/43 (2.3%) | 0.0019 |
| Total live births | 18 (3 sets of twins) | 37 (2 sets of twins) | |
| N° of previous live births | 4 in 3 women | 29 in 12 women | |
| N° of previous abortions | 15 in 6 women | 1 each in 4 women | |

Postpartum was defined as hemorrhage exceeding 500 mL

Preterm delivery was defined as delivery before 37 completed weeks.

Intrauterine growth restriction was defined using the 10 th percentile cutoff value