Pregnancy Outcomes and Management in Patients with Classic Congenital Adrenal Hyperplasia

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Materials and Methods

- In a Natural History Study of CAH at the National Institutes of Health Clinical Center in Bethesda, MD data was collected for 351 patients. Within this patient cohort there were 251 patients with Classic CAH of which 110 were female. Of these women, 51 were above the age of 18 and 8 women had one or more pregnancies.
- Subjects were recruited through listings on www.Clinicaltrials.gov (identifier no. NCT00290159), the national support group of CAH (Congenital Adrenal Hyperplasia Research Education & Support Foundation) or self-referral. The study was approved by the Eunice Kennedy Shriver National Institute of Child Health and Human Development Institutional Review Board and all patients were provided with informed consent.

Results

Table 1. Genetic and Descriptive Characteristics of Patient Cohort

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<th>Genotype</th>
<th>Age at Menarche</th>
<th>Menstrum</th>
<th>Infertility</th>
<th>Menses</th>
<th>Pregnancy</th>
<th>Length of Gestation</th>
<th>Outcome</th>
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Figure 1. Graph of Glucocorticoid Dose Equivalent in Patients Pre, During and Post Pregnancy

- *indicates patients that had an elective early termination of their pregnancy
- Shaded region = normal range

Results Continued

- During pre-pregnancy management, our patients had an average free testosterone of 0.45 ± 0.3 ng/dL (normal 0.3 – 1.9 ng/dL) and average total testosterone is 23.4 ± 11.9 ng/dL (normal range 8 – 60 ng/dL).

Conclusion

- In our large cohort of patients with CAH, 8 (15.7%) adult women with classic CAH became pregnant, despite 2 (25%) having irregular menses in the year prior and mildly elevated adrenal steroids.

- Although infertility has been commonly reported amongst patients with classic CAH, we report here 3 unwanted pregnancies in women with classic CAH who were sexually active.

- Approximately half of the women had uneventful pregnancies with no change in glucocorticoid dose, while half had an increase in glucocorticoid due to symptoms of adrenal insufficiency.

- No change occurred in mineralocorticoid replacement during pregnancy.

- Patients with classic CAH, can become pregnant, experience uneventful pregnancies and have healthy full-term infants with little intervention. However, close monitoring is of use in signs and symptoms of adrenal insufficiency throughout pregnancy and stress doses of glucocorticoid during labor and delivery are essential.

- One limitation of our study is that we had a small patient cohort and not all patients got pregnancy management and care at NIH. Thus, data collection is ongoing.

References