ACTA MEDICA IRANICA
Vol. XIX. 1976, P. 120-124

Cyclic Hormonal Changes in Patients With Congenital Abscence of the Uterus

H. Naficy, M.D.* H. Parsa, M.D.** R. Barakat, M.D.***

H. Nasr, M.D.**** P. Hall, PhD.*****

The scarcity of data in world literature about measurements of gonadotropins and sex steroids in patients with congenital abscence of the uterus and the frequency of this syndrome in Iranian women has stimulated this study.

This preliminary report proves that the cyclic hormonal changes in these patients is comparable with normal menstruating women.

Clinical Data

Five patients with various degrees of Mullerian duct agenesis were selected for this research. Their ages ranged from 18 to 25 years and all were married at the time of their first visit to the out-patient clinic of the Jahanshah Saleh Maternity Hospital. The chief complaint of all patients was primary amenorrhea and infertility. Three husbands of these patients were complaining of difficulities with intercourse because of the short vagina. Two of the five patients had vague cyclic lower

* From the department of obstetrics and gynecology and reproductive biology unit, university of Tehran
The institute for resarch in human reproduction, ministry of health

abdominal pain and fullness which lasted 2-3 days and followed a monthly interval pattern. All of the patients had developed scondary sex characteristics by the age of 14 years and were normal in that respect. The family history in most cases was pertinent in that there was more infertility in their aunts and cousins.

The general physical examinations revealed a well developed group of females with normal breasts and hair distribution for females of their ages.

Pelvic examinations showed normal external genitalia with abscence of the upper two-thirds of the vagina and also abscence of the uterus in all cases.

Buccal smears of all patients were positive and all has a normal XX female karyotype. Intravenous pyelogram in all patients were normal except for one that showed bilateral uteropelvic obstruction. Arteriography was carried out on three of these patients and the results showed a short blunted end uterine artery. Laparascopy was also done on all of the patients and the rudimentry uteruses were removed in the two patients that had cyclic lower abdominal pain. The three patients that had difficulties with intercourse underwent surgery for construction of longer and larger vaginas.

Material and Method

Blood samples and 24 hour urine samples were collected every other day from each patient for at least 28 days. Quantitiative determination of serum levels of follicle stimulating hormone (F.S.H.), luteinizing hormone (L.H.), and progesterone, and also urinary estrogenes were measured. In one case 17 betaestradioal was measured in serum. All serum measurements were done by radioimmuno-assay procedures and the urinary estrogenes were measured by the Brown's technic.

Results

The levels of 4 hormones measured and their cyclic changes in our patients with abscence of functioning uteruses were identical to the levels and patterns characteristic of subject with intact uteri and normal menstrual cycles. Serum F.S.H. and L.H. exhibited a normal ovulatory surge and serum progesterone levels rose normally 2 days after L.H. peak and remained elevated for at least 12 days. Urinary estrogenes and plasma 17 beta-estradiol had two peaks in each cycle. The second one seamed especially significantly higher in patients that had no cyclic lower abdominal pain. Only one patient had a basal bady temperature taken regularly enough to be fo any value and it showed a biphasic pattern corresponding to a normal 12-14 day luteal phase.

Discussion

By reviewing the literature available, we found that cyclic hormone patterns of serum gonadotropines and sex steroids had been measured only in one case of congenital abscence of the uterus.(3)

Our series of studies showed that hormonal patterns observed in patients with no functioning uterus are very similar to those observed in normally menstruating women who have a definitely functioning uterus. The higher level of urinary estrogenes in the second half of the cycle could be insignificant but also it could be due to lack of its utilization by the uterus and endometrium.

The role of the intact uterus in the regulation of the menstrual cycle is polemical. Luteolytic effects of the intact uterus have been demonstrated in some experimental animals where, upon removal of the uterus, the life span of the corpus luterum is prolonged. (1,2)

Corpus luteum function in women after hysterectomy is prolonged according to some investigations and unaffected, according to others, (2, 4, 5) In our series, all patients had normally functioning corpus luteum for at least 12 days. This proves that a life-long abscence of a functioning uterus appears to have had no effect on the functional

life-span of the corpus luterum, or other endocrine features of these patients. Rudimentary uteruses which were removed had no uterine cavity or endomential tissue.

The frequency of this syndrome in Iran, and especially its geographical distribution, would warrant further studies; but it suggests the autosomal recessive genetic pattern of transmission is most likely to be ture. (6)

In our series, congenital urinary tract anomalies were less than it is reported in literature. (6)

We feel that pelvic arteriography, which was, to our knowledge, done for the first time in the world, in this type of patients is helpful enough to be a significant diagnostic tool.

Acknowledgment

This study was supported by a grant from W.H.O. and the United Nations development fund.

The authors are grateful for the help of doctors Shahrzad, Mirdamadi, and Sattarian and Miss Akbarzadeh and their kind assistance in this project.

Summary

Our studies have shown normal cyclical patterns of ovarian steroids and pituitary gonadotropines in patients with congenital abscence of the uterus.

It appears that the life-long abscence of a functioning uterus has no effect on the functional life span of the corpus luterum, but may have some effect in the level of estrogenes in the second half of the cycle.

References

1. Anderson, L. L., K. P. Bland, and R. M. Melampy, Recent Progress in Hormonal Research 25:57, 1969.
2. Beling, C. G., S. L. Marcus, and S. M. Markham; Journal of Clincial Endocrinol Metabolism 30:30-39, 1970.
3. Coyotupa, Juan and et al; "Normal cyclical patterns of serum gonadotropines and ovarian steroids despite congenital absence of the uterus" Journal of Clincial Endocrinology Metabolism 36:395, 1973.
4. Kistner, R. W.; Gynecology principle and practice, Yearbook Medical Publishers, Chicago, p. 75, 1971.
5. Niswender, G. D., and et al; Fertility and Sterility 23:432, 1972.
6. Novak, E. R., G. S. Jones and H. N. Jones, Jr.; Textbook of Gynecology, Williams and Williasn Company, Baltimore p. 132-133, 1970.