Gh. Shahrzad MD[1]
F. Zafarani BSc[2]
A. Vosough MD[2]
F. Ahmadi MD[2]

Developmental Defects of Uterine Cavity: Presentation of Seven Patients

The diagnosis of intrauterine conditions that may cause infertility is generally made by hysterosalpingography (HSG). Herein, we are presenting seven hysterosalpingographically-proven patients of Developmental Uterine Cavity Defects. We believed that some of developmental defects of these uterine cavity have not been addressed in preceding medical texts. Accurate diagnosis and reports of such cases are important not only for the benefit of treatment, but also to reflect the true incidence of these anomalies and to consolidate embryologic concept.

The presented anomalies included "flying bird" uterus, "wine-glass-shaped" uterus, "buffalo head" uterus, "heart-shaped" uterus, "phantom-shaped" uterus, "candle light" uterus, and "jackal-shaped" uterus.

The presented cases belonged to many years before, thus they were not evaluated by recently-developed advanced diagnostic modalities.

KeyWords: congenital anomalies, uterine developmental defect, hysterosalpingography

Introduction

Mullerian duct anomalies, though uncommon, are often treatable causes of infertility. The true incidence and prevalence of congenital Mullerian duct anomalies are difficult to assess because some of these cases remain asymptomatic throughout their lives.[1,2] The prevalence of these anomalies was reported from 0.16%–10% by different researchers. Several reasons can be counted for this wide range, such as studies on different populations, non-standardized classification systems and differences in the diagnostic data.[3-8]

Uterine malformations are usually detected during manual inspection of the post-partum uterus, routine ultrasound examination, hysterosalpingography (HSG) and magnetic resonance imaging (MRI).

HSG is a simple and one of the sensitive and accurate methods that enables visualization of the uterine cavity contour and lesions.

Congenital uterine abnormalities are associated with an increased incidence of spontaneous abortion and other obstetrics complications such as premature labor, abnormal fetal lie and dystocia at delivery.[9-10]

Case Presentation

Case 1: Flying bird Uterus

A 32-year-old woman presented with a three-year history of primary infertility. Her menstrual cycles were regular with moderate bleeding and without intermenstrual bleeding and dysmenorrhea. On speculum examination, a normal vagina and a normal exocervix were observed. On HSG, the uterus was characterized by a canal-shaped body. The corneas were like wings of a bird and the

1. Department of Radiology, Tehran University of Medical Sciences, Tehran, Iran.
2. Department of Endocrinology and Female Infertility, Royan Institute, Tehran, Iran.

Corresponding Author:
Ahmad Vosough
Address: Royan Institute, No 36, Simin Alley, Zaferanieh, Tehran, Iran.
P.O.Box: 19395-4644
Tel: +9821-22413790
Fax: +9821-22409314
Email: vosough@royaninstitute.org

Received January 2, 2006;
Accepted after revision April 29, 2006.

Iran. J. Radiol. Spring 2007;4(3):185-189

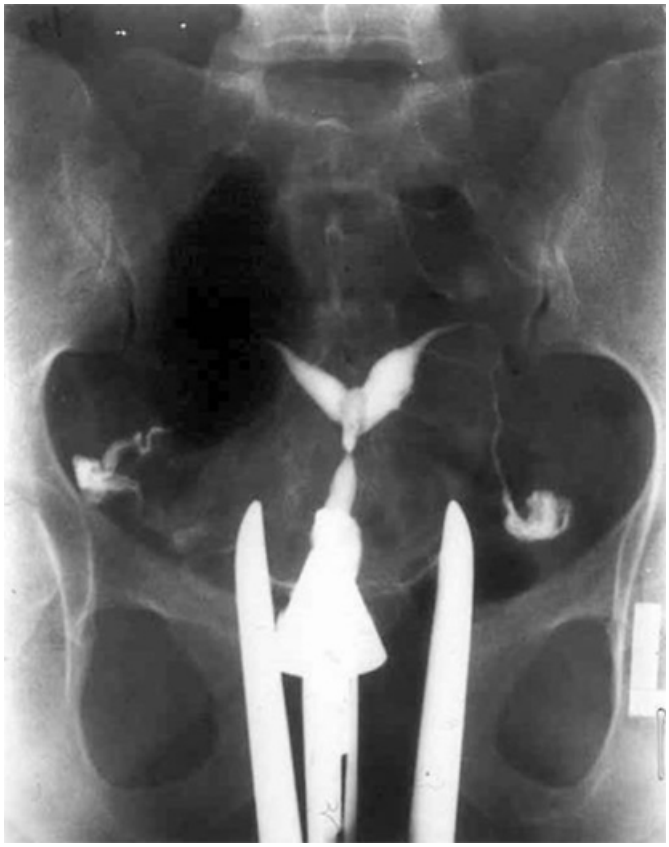

Fig. 1. Flying bird uterus; Canal-shaped body with wing-shaped corneas.

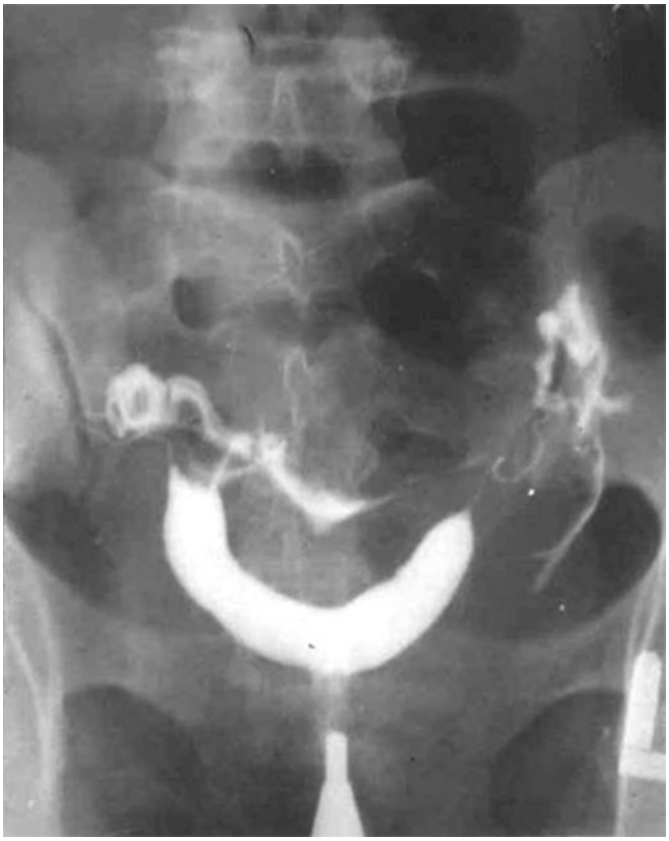

Fig. 2. Wine glass uterus with broad concavity in the fundal area.

uterine cavity presented like a flying bird. This anomaly may be classified as an anatomical variation of bicornuate uterus (Fig. 1).

Case 2: Wine glass uterus

The 35-year-old woman with a five-year history of primary infertility and no history of myoma referred to our center. On speculum examination, a normal vagina and a normal exocervix were observed. On HSG performed on the 9th day of her menstrual cycle, the uterus was presented by a broad concavity in fundus, convexity in lateral margins, thread-like isthmus and conical cervix (Fig. 2).

Case 3: Buffalo head-shaped uterus

A 30-year-old woman presented with three years of primary infertility. Her menstrual cycles were regular with normal bleeding. On speculum evaluation, a normal vagina and a normal exocervix were seen. On HSG the cavity was specious and the corneas were extended. There was convexity in lateral margins. The body was funnel-shaped and uterus looked like buffalo head (Fig. 3).

Case 4: Heart-shaped uterus

A 28-year-old woman presented with a six-year history of primary infertility. She had regular menses with normal bleeding and no history of uterine sur-

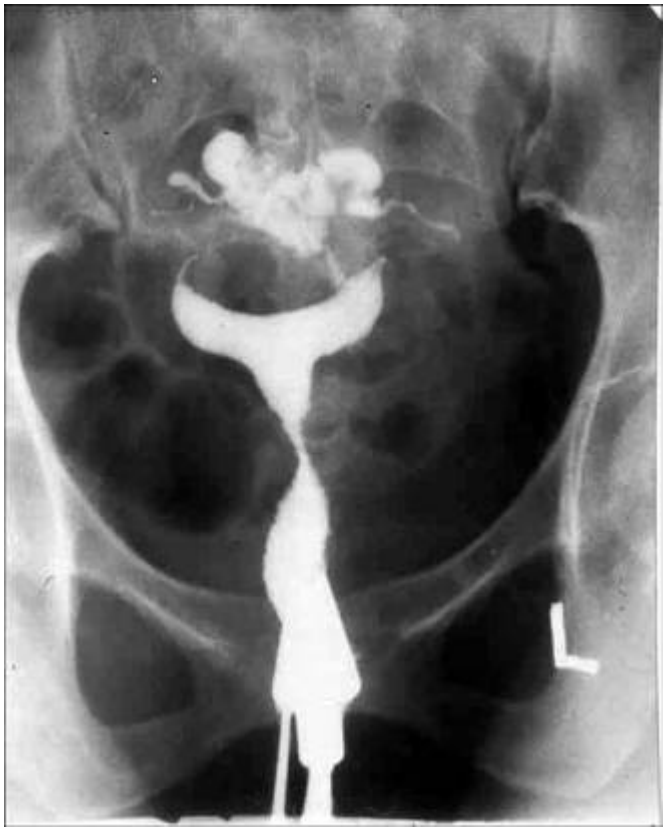

Fig. 3. Buffalo head-shaped uterus; the cavity is funnel-shaped with a concavity in fundal area.

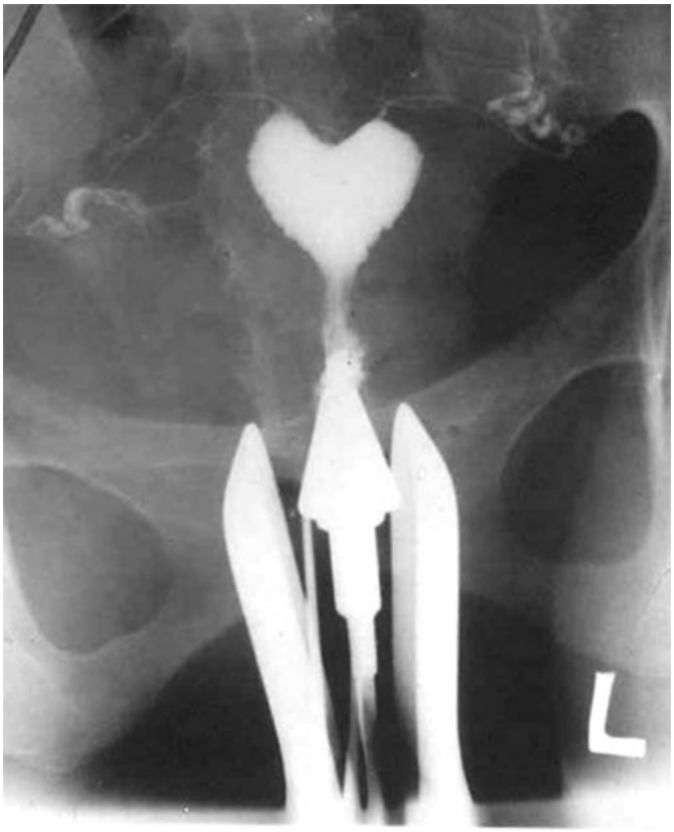

Fig. 4. Heart-shaped uterus; Deep indentation in fundal area and convexity in lateral margin.

gery. On speculum examination, a normal vagina and a normal exocervix were observed. On HSG the cavity was characterized by a deep indentation in the fundus, and a convexity in lateral margin, giving the uterus a typical heart shape (Fig. 4).

Case 5: Phantom-shaped uterus

A 27-year-old woman presented with nine years of primary infertility. She had regular menses with normal bleeding and no history of uterine surgery. On speculum examination, a normal vagina and a normal exocervix were observed. On HSG, the uterus was presented by a prominent convexity in the fundal area, diminished intercornual distance, and narrow distal portion of the body. The lateral outline was rigid and straight. The body was cylindrical and the uterus was seen like a phantom (Fig. 5).

Case 6: Candle light-shaped uterus

A 35-year-old woman with a six-year history of primary infertility referred to our center with primary amenorrhea and with no history of tuberculosis. On speculum examination, a normal vagina and a normal exocervix were observed. On HSG, the uterus was characterized by an elongated and normal cervix, narrow and cylindrical body with no left tube. The right tube was short and just interstitial portion of right tube was seen. After spillaging of contrast in the right side, the appearance of uterus looks like a burning candle (Fig. 6).

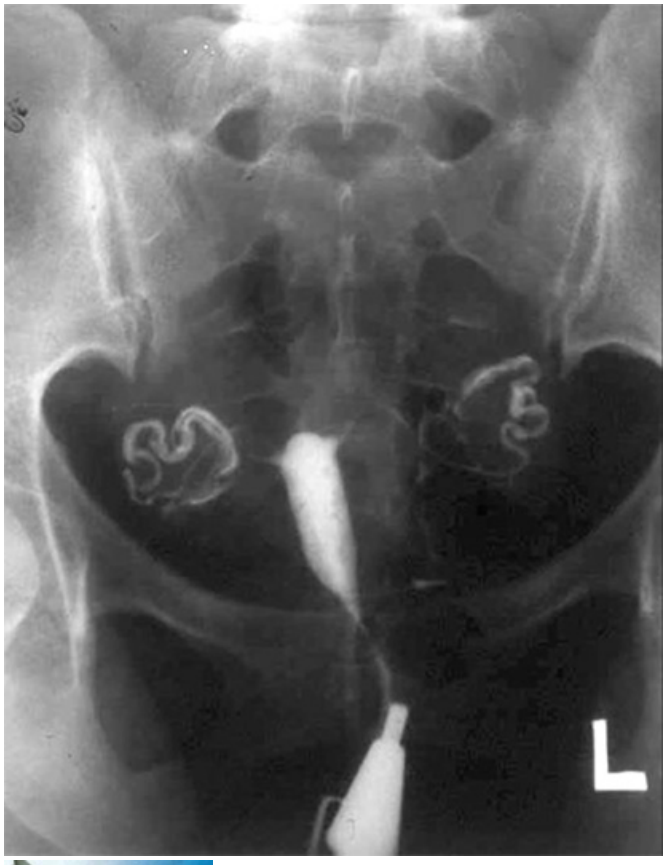

Fig. 5. Phantom-shaped uterus; Cylindrical cavity with diminished intercornual distance and narrow distal portion of the body.

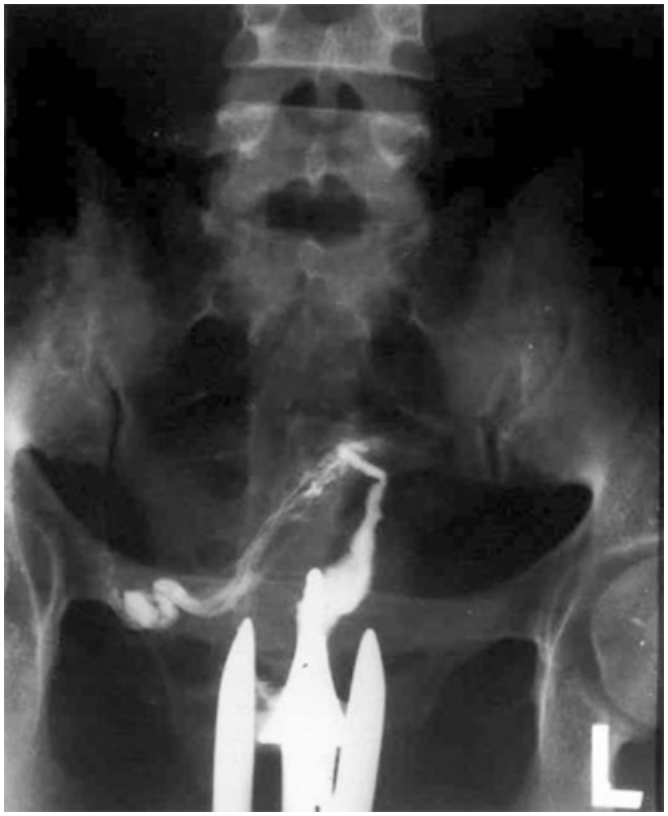

Fig. 6. Candle light-shaped uterus with a narrow and cylindrical body and a short right tube.

Case 7: Jackal-shaped uterus

A 27-year-old woman with a nine-year history of primary infertility referred to our center. Her menstrual cycles were irregular with normal bleeding. On speculum examination, a normal vagina and a normal exocervix were seen. On HSG, the cavity is funnel-shaped and the corneas are elongated. The fundal and lateral margin are straight. The isthmus is cylindrical and cervical canal is spindle shaped (Fig. 7).

Discussion

A brief review of embryology is useful for better understanding of congenital anomalies. Both the tube and the fallopian tubes develop from the mullerian duct. Normal development of the uterus and fallopian tubes requires craniocaudal growth of the paired mullerian ducts, fusion of the caudal segments of the ducts (unfused cranial ends form the paired fallopian tubes), resorption of the median septum diving the fused segments, and finally, maternal or placental hormone stimulation of the normally-formed uterus and fallopian tubes.[11]

Malformations occurring at each of these phases have been shown by HSG.

Several attempts have been made over the past years to establish a comprehensive classification of mullerian duct anomalies. Such a classification should be compatible with the hypothesis concerning the normal embryologic development of mullerian system as well as the development of mullerian anomalies. [12-14]

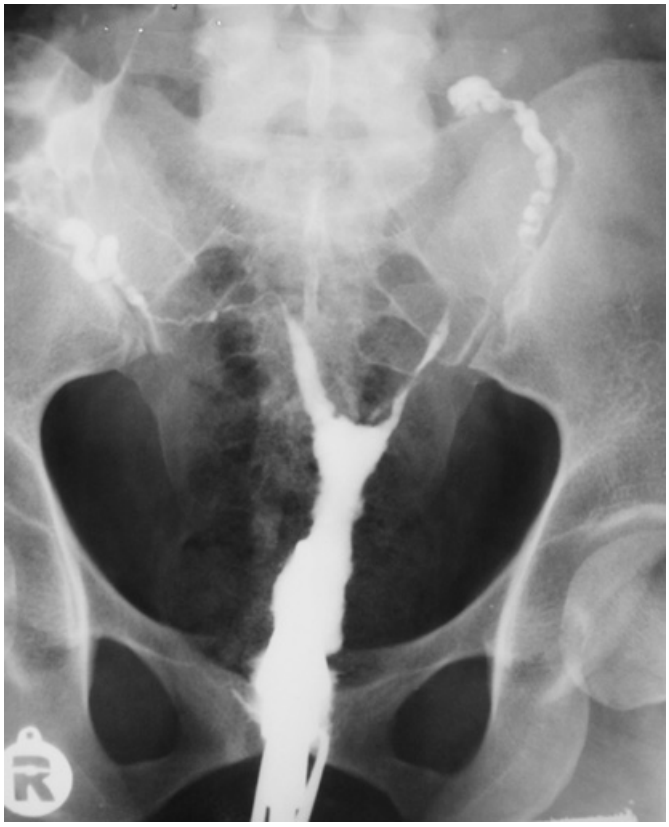

Fig. 7. Jackal-shaped uterus with a funnel-shaped body and elongated corneas.

The American Fertility Society (AFS) classification system functions as a framework for the description of anomalies, for communication between clinicians, and for comparison among various therapeutic modalities. It combines embryologic aspects of these anomalies with some consideration of their clinical and surgical significance.

The cases presented in this article failed to fit in AFS classification. Also we believe that some of developmental defects of the uterine cavity have not been addressed in the preceding medical texts.

As the most of these cases refer to many years ago, we could not evaluate the patients by laparascopy, hysteroscopy or MRI.

The majority of mullerian duct anomalies are considered to be sporadic or multi-factorial in nature. However, polygenic and genetic patterns of inheritance have been described in the expression of these anomalies.

Extrauterine and intrauterine environmental factors, such as exposure to ionizing radiation, intrauterine infections, and drugs with teratogenic effects such as talidomide and diethylstilbestrol (DES), can also cause defects of the developing fetal genital tracts.

In conclusion, accurate diagnosis and reports of such cases are important not only for their treatment, but also for deriving the true incidence of these anomalies and for consolidating the embryologic concepts.

References

1. Patton GW. The uterus in infertility evaluation. In: Behrman SJ, Kistner RW, Patton GW, editors. Progress in Infertility. 3rd ed. Boston: Little & Brown Co., 1988.
2. Troiano RN, McCarthy SM. Mullerian duct anomalies: imaging and clinical issues. Radiology 2004;233(1):19-34.
3. Byrne J, Nussbaum-Blask A, Taylor WS, Rubin A, Hill M, O'Donnell R, et al. Prevalence of Mullerian duct anomalies detected at ultrasound. Am J Med Genet 2000;94(1):9-12.
4. Maneschi F, Zupi E, Marconi D, Valli E, Romanini C, Mancuso S. Hysteroscopically detected asymptomatic mullerian anomalies prevalence and reproductive implications. J Reprod Med 1995;40(10):684-8.
5. Simon C, Martinez L, Pardo F, Tortajada M, Pellicer A. Mullerian defects in women with normal reproductive outcome. Fertil Steril 1991;56(6):1192-3.
6. Ashton D, Amin HK, Richart RM, Neuwirth RS. The incidence of asymptomatic uterine anomalies in women undergoing transcervical tubal sterilization. Obstet Gynecol 1988;72(1):28-30.
7. Stampe Sorensen S. Estimated prevalence of mullerian anomalies. Acta Obstet Gynecol Scand 1988;67(5):441-5.
8. Rock JA, Schlaff WD. The obstetric consequences of uterovaginal anomalies. Fertil Steril 1985;43(5):681-92.
9. Pennes DR, Bowerman RA, Silver TM. Congenital uterine anomalies and associated pregnancies: findings and pitfalls of sonographic diagnosis. J Ultrasound Med 1985;4(10):531-8.
10. Golan A, Langer R, Bukovsky I, Caspi E. Congenital anomalies of the mullerian system. Fertil Steril 1989;51(5):747-55.
11. Longman J. Urogenital system. In: Longman J, editor. Medical Embryology. 4th ed. Baltimore: Williams&Wilkins; 1981.
12. Crosby WM, Hill EC. Embryology of the Mullerian duct system. Review of present-day theory. Obstet Gynecol 1962;20:507-15.
13. Buttram VC, Gibbons WE. Mullerian anomalies: a proposed classification (an analysis of 144 cases). Fertil Steril 1979;32(1):40-6.
14. The American Fertility Society. The AFS classifications of adnexal adhesions, distal tubal occlusion secondary to tubal ligation, tubal pregnancies, mullerian anomalies and intrauterine adhesions. Fertil Steril 1988;49(6):944-55.