THREE-DIMENSIONAL ULTRASOUND IN GYNECOLOGY: UTERINE MALFORMATIONS*

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Abstract **Uterine malformations are unusual findings in the clinical practice of gynecology. Statistics regarding this abnormality are flawed, and there is a lack of standardization of the pertinent terminology, besides difficulty in the diagnosis. Starting in the 1980's, ultrasound became a procedure indispensable to the clinical practice in the field of tocogynecology, modifying concepts and procedures, and bringing an important contribution to this specialty. The advent of endocavitary transducers, amplitude and spectral color Doppler, as well as the increasing improvement in the imaging quality, has contributed to this progress. Over the last ten years, much has been studied, published and discussed about the role of the three-dimensional ultrasonography. The authors review the topic and emphasize the importance of this method as a diagnostic modality.**
Keywords: Uterine malformations; Müllerian malformations; Three-dimensional ultrasonography.

Resumo *Ultra-sonografia tridimensional em ginecologia: malformações uterinas.*
As malformações uterinas são achados pouco comuns na clínica ginecológica. As estatísticas nesta área são muito falhas. Corrobora, ainda, a falta de uniformização na terminologia empregada e as dificuldades diagnósticas. A partir da década de oitenta, a ultra-sonografia tornou-se um procedimento indispensável à prática toco-ginecológica, contribuindo e modificando conceitos e procedimentos dentro dessa especialidade. O advento dos transdutores endocavitários, a análise com Doppler colorido de amplitude e espectral, assim como a melhoria crescente da qualidade de imagem contribuíram para isso. Nos últimos dez anos muito se tem pesquisado, publicado e discutido sobre o papel da ultra-sonografia tridimensional. Os autores fazem uma revisão do tema e ressaltam a importância dessa metodologia como modalidade diagnóstica.
Unitermos: Malformações uterinas; Malformações müllerianas; Ultra-sonografia tridimensional.

INTRODUCTION

Uterine malformations are secondary to failure in development, reabsorption or fusion of Müllerian ducts. Around the sixth week of the embryogenesis, an invagination of the coelomic lining epithelium forms a depression creating a sulcus, whose borders fuse to form the lateral Müllerian ducts (or paramesonephric ducts) (Figure 1). The Müllerian ducts initially are formed in the upper dorsal wall of the coelomic cavity and progress caudally to enter the pelvis where they incline towards the center, fusing medially. Farther on, the caudal progress results in a contact of these fused ducts with the urogenital sinus. The proximal segments of the uterovaginal canal originated from coelomic epithelium remain unfused and open into the peritoneal cavity to form the Fallopian tubes. The upper portion of the vagina is, therefore, considered to have Müllerian origin, and the lower portion as originating from the urogenital sinus. He whole lining epithelium (uterus and tubes) originates from the coelomic epithelium. This is the reason for uterine malformations being denominated Müllerian malformations or anomalies[1].

In the past, the uterus only could be clinically evaluated by means of a physical examination. Several methods have been introduced for gynecological evaluation. As an example, we can mention; radiological examinations[2] by means of hysterosalpingography[3–5] (Figure 2), surgical procedures — laparotomy, laparoscopy, and most recently, videolaparoscopy (Figure 3) — and hysteroscopy[6] (Figure 4). Undoubtedly, ultrasound is the method that

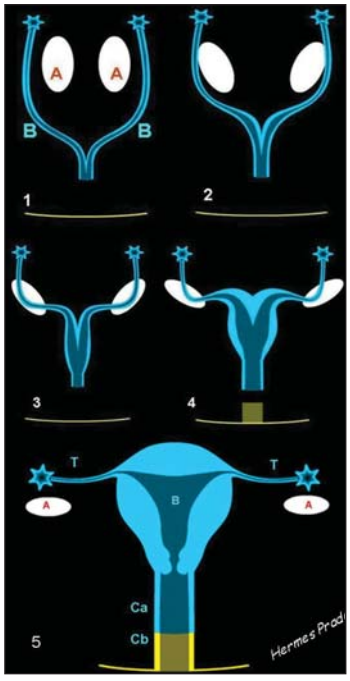

Figure 1. Scheme showing the embryological development and sequence (1 to 4) of Müllerian ducts fusion (B). (A) indicates the ovaries. Number 4 demonstrates the formation of the uterine body after fusion. 5B, uterine body; T, Fallopian tubs; Ca, proximal third of the vagina; Cb, distal third.

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Figure 2. Radiological study by hysterosalpingography in a bicornuate uterus. Details of the Fallopian tubes are observed.

Figure 3. Videolaparoscopy in the suspicion of bicornuate uterus. The normal uterine fundus visualization excludes this diagnostic hypothesis. The final diagnosis was of partial septate uterus.

Figure 4. Videohysteroscopy in the suspicion of bicornuate uterus. Visualization of a partial septum (see Figure 3).

has brought the greatest contribution as non-invasive method for evaluation of the uterus and its attachments[7], initially as a two-dimensional (2D) modality with an abdominal approach (Figure 5), and later with a transvaginal approach (Figure 6). Most recently, ultrasonography has been added of the three-dimensional (3D) processing[8–11] in both modalities — the multiplanar (Figure 7) and the volumetric ones (Figures 8, 9 and 10). Magnetic resonance imaging also has been utilized in many services[12].

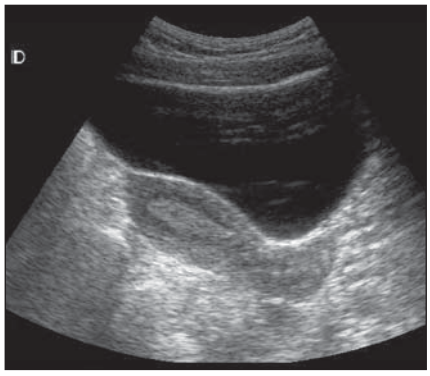

Figure 5. Longitudinal view of a normal uterus on 2D transabdominal ultrasound. Observe the adequately filled bladder, and the secretory pattern of the endometrium.

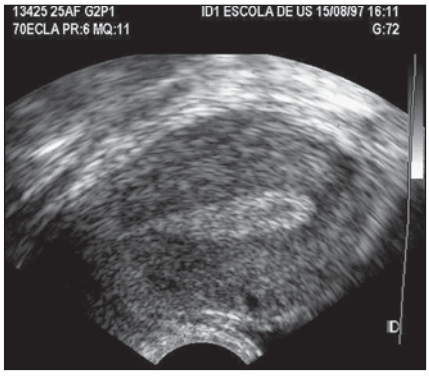

Figure 6. Longitudinal view of a normal uterus on 2D transvaginal ultrasound Observe the secretory pattern of the endometrium, the transvaginal way and the approach of choice for the endometrial evaluation.

DISCUSSION

Uterine malformations are unusual findings in the gynecological clinical practice. This abnormality is a reflection of an array of presentations, associated with the fact that the diagnosis of the majority of malformations does not occur before a gestation or are diagnosed only after manifestation of an obstetric problem[13–15]. Statistics in this area are flawed, and there is a lack of standardization of the pertinent terminology, besides the difficulty in the diagnosis[16]. Patients with these alterations frequently are oligosymptomatic or even asymptomatic, with preserved menstrual, sexual and even reproductive functions. So, frequently presented case reports and casuistics reflect particularities of certain groups of women with obstetric complications, patients of infertility and sterility services, or cases of medical urgency resulting from menstrual flow obstruction[17].

Presently, 2D ultrasound, and especially 3D ultrasound are indispensable diagnostic tools for evaluation of uterine malfor-

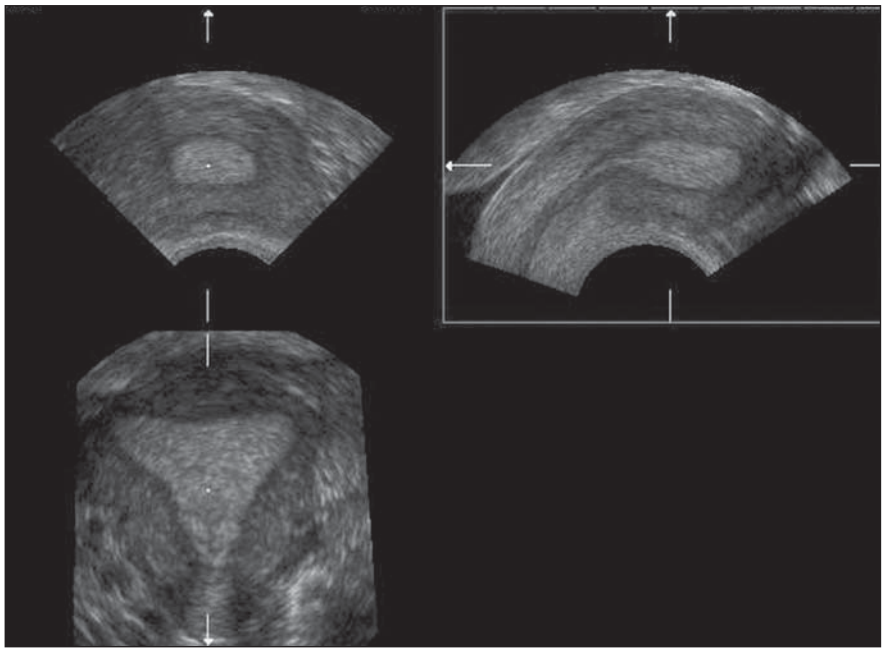

Figure 7. Normal uterus on 3D multiplanar ultrasound. Observe the secretory pattern of the endometrium on the three views.

Figure 8. Coronal view of a septate uterus on 3D volumetric ultrasound. Observe the secretory pattern of the endometrium.

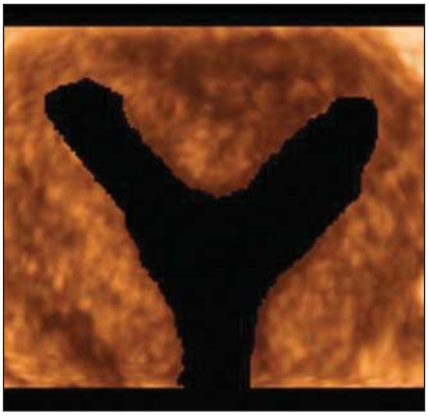

Figure 9. Coronal view of a septate uterus on 3D volumetric ultrasound with digital subtraction of the endometrium.

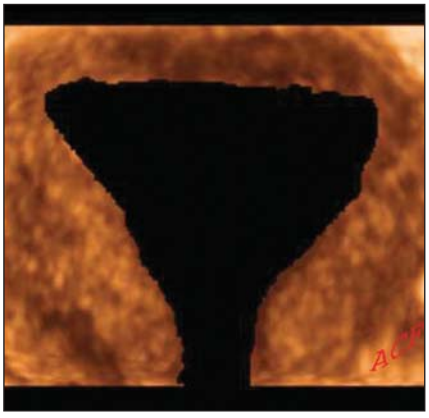

Figure 10. Coronal view of a septate uterus on 3D volumetric ultrasound with digital subtraction of the septum.

mations, allowing accurate diagnoses, most of times more specific than a simple description of a septate uterus, characterizing the abnormality and providing information to assist in the definition of therapeutic regimen and reproductive prognosis[18–21].

The appropriate classification of Müllerian anomalies is important, the most relevant one being proposed by Jarcho in 1946, and later adapted by Zanetti et al., based on the embryonal development. Later on, this classification was modified by Butram and Gibbons, in 1975, and is currently adopted by the American Society of Fertility.

A better evaluation with 2D ultrasound is achieved by the association between transabdominal and transvaginal approaches. The first one allows a better visualization of the uterine fundus, and analysis of the bladder and ureteral jets. The second allows a more detailed analysis of the cervix (cervices) and endometrial cavity (cavities).

The main view for correctly diagnosing the type of the malformation is the coronal view, sometimes difficult to be obtained with the 2D technique, but feasible, provided some technical prerequisites are met: the bladder must be almost emptied; the transducer must be placed transversely to the patient's abdomen with slight, cranial movements.

Several anomalies may originate from an incomplete fusion the Müllerian ducts: clefts in the uterine funds, but not in the cervix, a complete division of the uterus by a septum into two endometrial cavities, forming the called septate uterus. The septum may be partial or complete, with variable extent from the uterine fundus towards the cervix (Figures 11, 12, 13, 14 and 15). Sometimes, the differential diagnosis with bicornuate uteri is difficult, especially if the sonographic evaluation is restricted to the transvaginal approach. Also, it may be associated with a longitudinal or oblique vaginal septum.

The complete non-fusion of Müllerian ducts originates an anomaly previously called complete double uterus with double cervix, currently denominated didelphys uterus (Figures 16 and 17), with each endometrial cavity ending in a solitary fallopian tube. Such anomalies are perfectly compatible both with normal fertility and menstrual cycles, but sometimes they may result in significant clinical problems[20].

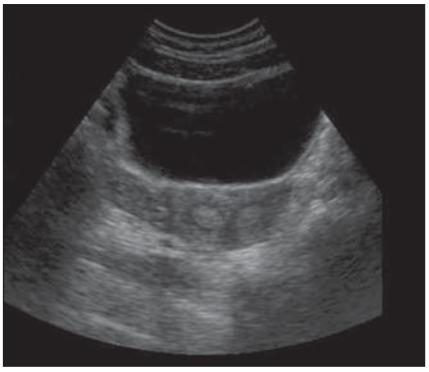

Figure 11. Double cavity uterus. Final diagnosis: septate uterus. Evaluation by 2D transabdominal ultrasound.

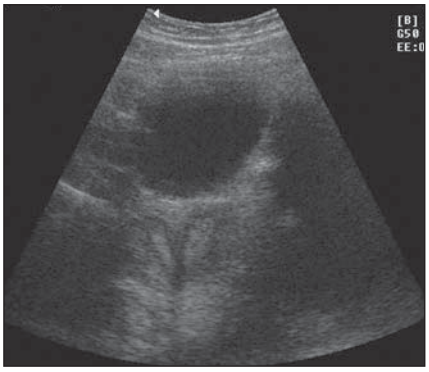

Figure 12. Double cavity uterus. Final diagnosis: septate uterus. Evaluation by 2D transabdominal ultrasound. Observe the difference in relation to Figure 11. In this image, the transducer was slightly more cranially inclined.

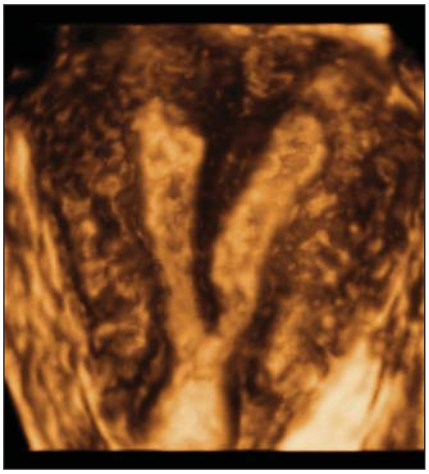

Figure 13. Double cavity uterus. Final diagnosis: septate uterus. Evaluation by transvaginal 3D volumetric ultrasound. Observe the symmetrical endometrial cavities and the septum proximal to the internal ostium.

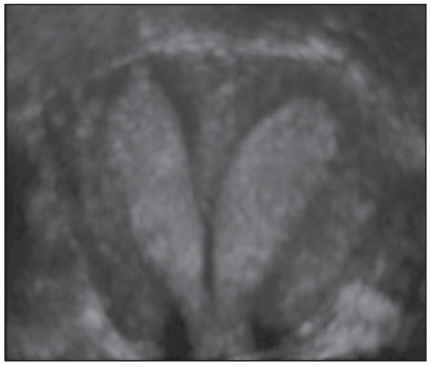

Figure 14. Double cavity uterus. Final diagnosis: septate uterus. Evaluation by multiplanar, transvaginal 3D ultrasound. Observe the symmetrical endometrial cavities.

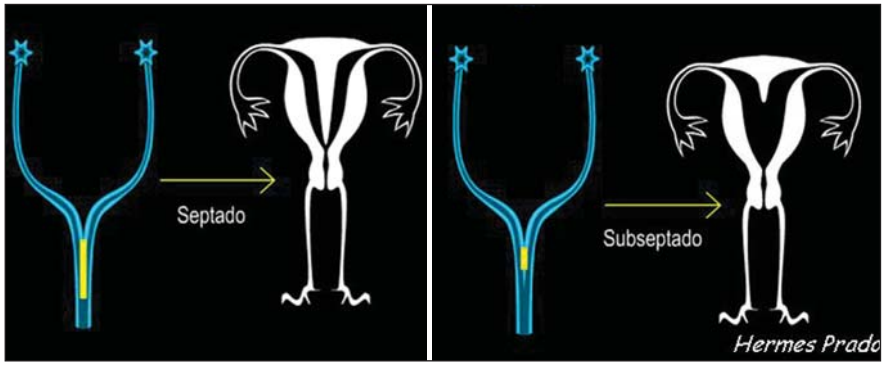

Figure 15. Schemes demonstrating failure both in complete and partial absorption of Müllerian ducts.

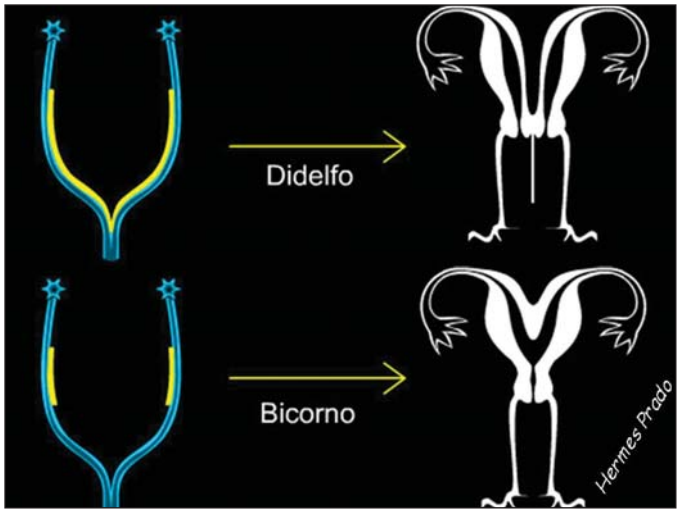

Figure 16. Schemes showing failures in complete (didelphys uterus) and partial (bicornuate uterus) fusion of the Müllerian ducts.

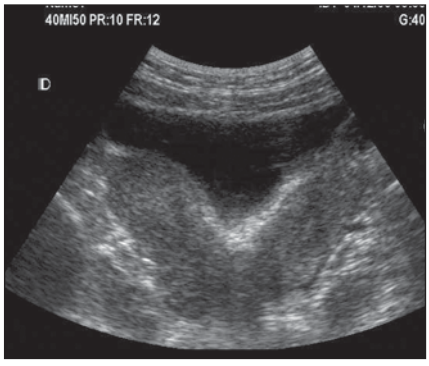

Figure 17. Double cavity uterus. Final diagnosis: didelphys uterus. Transabdominal 2D ultrasound is the method of choice for the correct diagnosis of this anomaly. Observe the complete separation between uterine bodies.

Figure 18. Bicornuate uterus with a ten-week gestation. Transabdominal evaluation. Observe decidua in the non-pregnant uterine cavity.

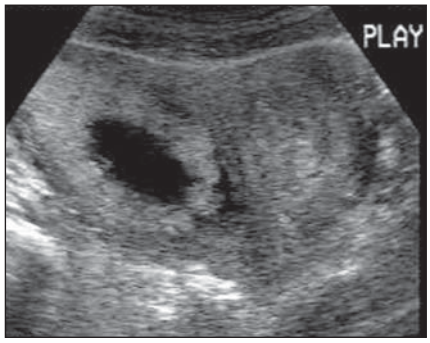

Figure 19. Bicornuate uterus with gestational sac. Transabdominal evaluation.

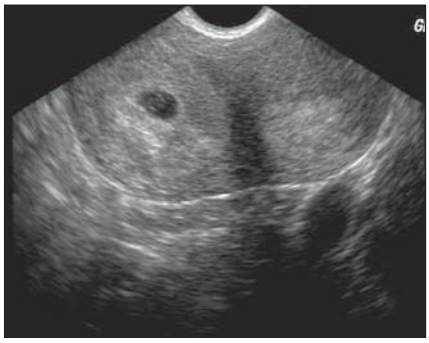

Figure 20. Bicornuate uterus with a five-week gestation. Transvaginal evaluation.

Pregnancy in a half of a septate, bicornuate (Figures 18, 19 and 20) or didelphys uterus may be associated with bleeding of the non-pregnant half of the uterus. In rare cases, the pregnancy may occur in a half of the uterus after the other is already pregnant; this is called superfetation. The 2D ultrasound is sufficient for an adequate evaluation and diagnosis.

Bicornuate uteri (Figures 21 and 22) is the result of an incomplete fusion of the uterovaginal horns at the level of the fundus; that is to say, a partial fusion has occurred, resulting in two uterine fundi (horns) presenting different fusion degrees, symmetrically or not, most often at the level of the uterine isthmus, and, therefore, frequently presenting a single cervix. This

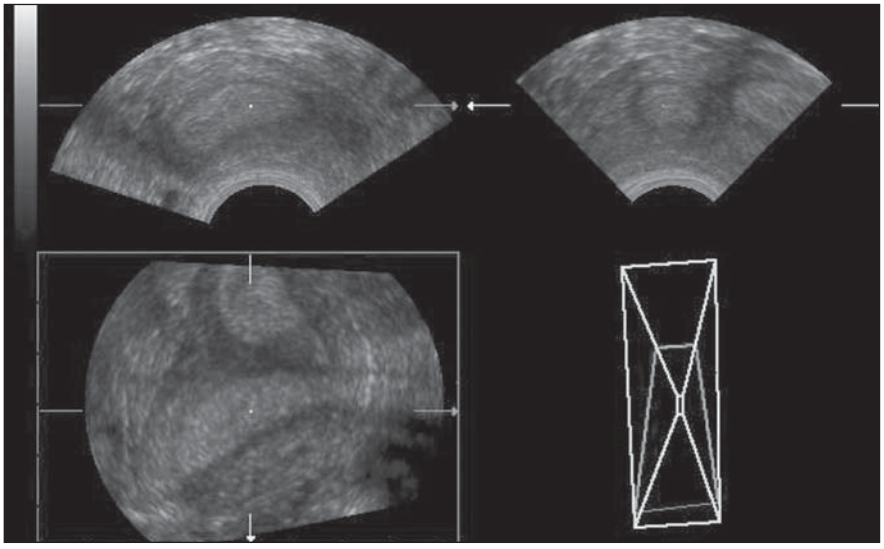

Figure 21. Double cavity uterus. Final diagnosis: bicornuate uterus. Transvaginal 3D multiplanar ultrasound. Observe asymmetrical endometrial cavities.

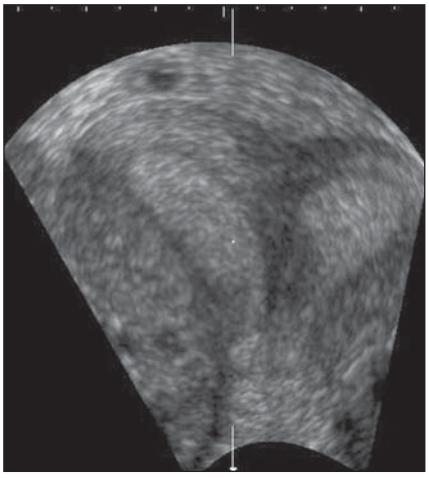

Figure 22. Bicornuate uterus. Coronal, 3D multiplanar evaluation showing asymmetrical cavities.

type of anomaly may be confused with septate uterus so currently 3D ultrasound is a valuable diagnostic method.

The small sized uterus is subdivided into hypoplastic and infantile uterus. The uterine hypoplasia (Figure 23) is found in an array of endocrine disorders, with a 1:1 uterine body/cervix ratio. In the infantile uterus, the uterine body/cervix ratio is 2:1. In the majority of cases, hypoplastic/infantile uterus occurs because of an ovarian or hypophyseal hypofunction.

An aspect that should not be disregarded during an ultrasound examination in a patient with uterine malformation is the evaluation of the renal lodges, considering the frequent association of uterine anomalies with urinary tract malformations such as renal agenesis (Figure 24) and/or ectopia[23]. Li et al. have found renal agenesis in 17 (29.8%) of 57 patients. No other renal anomaly has been found. Renal agenesis was the most frequent association in patients with didelphys uterus (13/16 cases), uterine agenesis (2/5 cases) and unicornuate uterus (2/7 cases). All the 11 cases of obstructed didelphys uterus were associated with homolateral renal agenesis towards the transverse septum of the obstructed hemivagina. So, they have concluded that renal agenesis is more frequently found in didelphys uterus than in other types of uterine malformations[23].

The uterus and vagina agenesis is denominated Mayer-Rokitansky-Kuster-Hauser syndrome and results from dysplasia of the Müllerian ducts, with absence of the normal uterus and part or the whole vagina. The diagnosis is basically clinical, however, the 2D ultrasound may confirm a clinical suspicion. The 3D ultrasound presents no advantage over the 2D ultrasound in these cases.

Although there are several types of uterine malformations, almost all of them can be diagnosed as follows: arcuate uterus, septate uterus (partial or complete), bicornuate uterus, unicornuate uterus, didelphys uterus, agenesis, hypoplasia, and infantile uterus.

With the exception of arcuate uterus, considered as a variant of the normal uterus, the most usual uterine anomalies, frequently resulting in diagnostic difficulty are the septate and bicornuate uterus[9].

The term "arcuate" refers to cases where there is a minimal alteration of the uterine cavity, with a convex or flat uterine fundus. The endometrial cavity may present a minimal, residual septum in the fundal region. There is no need for correction.

Regardless the type of uterine malformation, both 2D and 3D ultrasound should be performed during the second phase of the menstrual cycle for a better visualization of the endometrium, and therefore a better definition of the uterine cavity. During the first phase of the menstrual cycle,

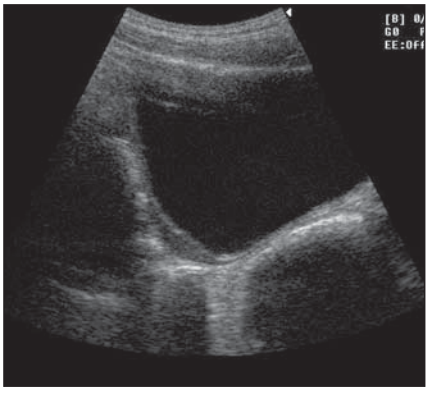

Figure 23. Very reduced volume of the uterus. Final diagnosis: hypoplastic uterus. Observe the almost imperceptible presence of the uterus on the transabdominal study.

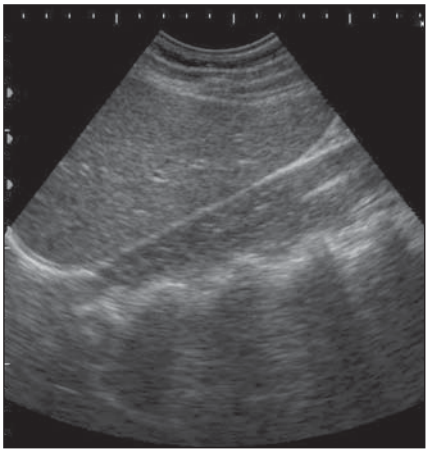

Figure 24. Right renal agenesis in patient diagnosed with didelphys uterus.

the endometrial cavity may not be appropriately visualized, affecting the diagnosis. In the suspicion of uterine malformations, the evaluation should be always performed during the secretory phase (Figure 5). Alternatively, if this is not feasible, sonohysterography is recommended.

Presently, both modalities of 3D ultrasound — multiplanar and volumetric — have been considered as a highly sensitive and specific technique for uterine malformations evaluation[24–26]. Both the transabdominal and transvaginal approaches may be adopted, the latest being preferable. The transrectal modality is an excellent alternative in the unfeasibility of the transvaginal approach. With the multiplanar 3D ultrasound, longitudinal, axial and coronal planes can be concomitantly evaluated, the coronal plane being indispensable to an appropriate diagnosis of Müllerian ducts anomalies.

Although magnetic resonance imaging is considered by some authors[12] as the method of choice for uterine anomalies evaluation, with up to 100% efficacy, this has not been a routine practice because of its high cost.

For the majority of authors, the ultrasound is the primary method for evaluation of Müllerian abnormalities. Fedele et al., studying 43 infertile patients diagnosed with double uterus by hysterosalpingography, have submitted them to subsequent ultrasound and laparoscopy/hysteroscopy to evaluate the ultrasound capacity to correctly demonstrate a malformed uterus. The ultrasound visualization was adequate in 39 cases (90.7%): one of two cases of didelphys uteri, all the 11 bicornuate uteri, all the four complete septate uteri, and all the 22 partial septate uteri with 92.3% sensitivity, and 100% specificity. Therefore, a differential and accurate diagnosis of "double uterus or endometrial cavity" is possible with this technique[21].

New diagnostic methods have been introduced, and ultrasound has consolidated its position as an invaluable tool to complement the clinical rationale. Its role in the evaluation of uterine malformations is already defined and will be more and more consolidated with the utilization of the 3D technique.

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