GENERAL GYNECOLOGY

Double cervix: clarifying a diagnostic dilemma

Benjamin C. Smith, MD; Douglas L. Brown, MD; Rickey E. Carter, PhD; Abimbola O. Famuyide, MBBS

OBJECTIVES: Although double cervix is often considered to indicate uterus didelphys, it may be challenging to determine the true associated anomaly as double cervix occurs with other müllerian anomalies. Our purpose is to report the frequency of uterine müllerian anomalies that occur with a double cervix and identify imaging or clinical criteria to help classify the associated anomaly.

STUDY DESIGN: After institutional review board approval, an electronic search was performed to identify patients with double cervix and pelvic magnetic resonance imaging (MRI) between 1976-2010 (using *International Statistical Classification of Diseases and Related Health Problems, Edition 9* and Hospital International Classification of Diseases Adapted codes). MRIs were reviewed to classify the müllerian anomaly. Clinical characteristics including cervical canal thickness, intercervical distance and orientation were recorded. Kruskal-Wallis and χ^2 tests were used to test for differences in patient and imaging characteristics across müllerian anomalies.

RESULTS: There were 64 patients who met inclusion criteria: 32 (50%) septate uterus, 27 (42%) uterus didelphys and 5 (8%) bicornuate uterus. Cervical canal divergence was present in 34% (11/32) with septate uterus, 26% (7/27) with uterus didelphys, and none with bicornuate uterus. Mean intercervical distance was significantly greater ($P < .001$) in uterus didelphys (12.05 mm) compared with septate (5.43 mm) or bicornuate uterus (5.40 mm).

CONCLUSION: Septate uterus is as common as uterus didelphys, and considerably more common than bicornuate uterus, in women with a double cervix. Although a widened intercervical distance may support the MRI diagnosis of uterus didelphys, careful evaluation of the uterine fundus is required for correct diagnosis when encountering a double cervix.

Key words: cervical duplication, double cervix, duplicated cervix

Cite this article as: Smith BC, Brown DL, Carter RE, et al. Double cervix: clarifying a diagnostic dilemma. Am J Obstet Gynecol 2014;210:x-ex-x-ex.

The incidence of müllerian duct anomalies (MDAs) of the uterus is difficult to determine and although incidence rates up to 10% have been reported, the incidence is probably closer to 1% in the general population and up to 3% in women with recurrent pregnancy loss or poor reproductive outcomes.[1,2] These abnormalities occur secondary to errors in embryogenesis and include uterine, cervical, and vaginal anomalies. Müllerian anomalies are usually classified in terms of uterine configuration according to the classification proposed by the American Fertility Society in 1988.[3] This classification is not all encompassing and does not include specific classification for cervical or vaginal anomalies.[4] Because of this, there may be confusion regarding classification of patients who also have abnormalities of the cervix or vagina.

Most MDAs are associated with a single cervix. In the minority of patients with MDAs who have a double cervix,[2] confusion may arise when attempting to diagnose and classify the associated uterine anatomy. Classic teaching under the unidirectional theory of müllerian development suggests that a double cervix should occur only with uterus didelphys.[5,6] Uterus didelphys has been reported as the most common anomaly to occur with a double cervix.[7] However, septate uterus with a double cervix may be misdiagnosed as uterus didelphys.[8,9] Further experience has shown that double cervix may occur with septate uterus, bicornuate uterus, uterus didelphys, and even a normal uterus, although the frequency of these occurrences is unknown due to the low numbers of reported cases.[4,10] Because septate uterus is the most common MDA, accounting for about 55% of all MDAs,[2] there is the potential to misdiagnose these patients when they also have a double cervix.

Reproductive and surgical implications are dependent on correct diagnosis and classification of müllerian anomalies. Some anomalies carry less obstetric implications than others. Patients with septate uteri (particularly if they have had recurrent pregnancy losses) are generally treated with hysteroscopic resection of the septum, whereas bicornuate uterus and uterus didelphys are not generally treated with surgery.[11] Misdiagnosis between septate uterus, bicornuate uterus, and uterus didelphys carries significant clinical implications. If a patient with a septate uterus is mistakenly diagnosed with a bicornuate or didelphys uterus, she may be denied a useful surgical treatment. If a patient with bicornuate or didelphys uterus is mistakenly diagnosed with a septate uterus, surgical treatment

From the Department of Radiology (Drs Smith and Brown), Department of Biomedical Statistics and Informatics (Dr Carter), and Department of Obstetrics and Gynecology (Dr Famuyide), Mayo Clinic-Rochester, Mayo Clinic College of Medicine, Rochester, MN.

Received Dec. 22, 2013; revised Feb. 1, 2014; accepted March 6, 2014.

The authors report no conflict of interest.

Presented at the 97th Scientific Assembly and Annual Meeting of the Radiological Society of North America, Chicago, IL, Nov. 27-Dec. 2, 2011.

Reprints: Benjamin C. Smith, MD, 200 1st St. SW, Mayo E2, Rochester, MN 55905. Smith.benjamin@mayo.edu.

0002-9378/$36.00

http://dx.doi.org/10.1016/j.ajog.2014.03.014

may result in complications such as uterine perforation.[9,12]

Multiple imaging techniques are used for diagnosis of uterine and cervical anomalies with varying degrees of accuracy. These include hysterosalpingography, ultrasound, and magnetic resonance imaging (MRI). Accuracy for hysterosalpingography and 2-dimensional (2D) ultrasound are less than that reported for MRI. MRI accuracy in diagnosis of müllerian anomalies is reported from 96-100%.[2,7,13] Three-dimensional (3D) ultrasound is playing an increasing role in the evaluation of müllerian anomalies of the uterus.[14]

The primary purpose of our study is to report the frequency of the different müllerian anomalies of the uterus that occur in patients with a double cervix as well as identify imaging or clinical diagnostic criteria that may help more easily classify the type of uterine anomaly associated with double cervix. Secondarily, we report the frequency of vaginal septa in the same patient population. We anticipate the results will help decrease the confusion and misdiagnosis that may occur when encountering patients with a double cervix.

Materials and Methods

This retrospective review was approved by our institutional review board. Patients were excluded if they had denied consent for use of their medical information for research purposes. Female patients with double cervix were retrospectively identified from our institution's patient database by searching for specific *International Statistical Classification of Diseases and Related Health Problems, Edition 9* and Hospital International Classification of Diseases Adapted codes, which included *International Statistical Classification of Diseases and Related Health Problems, Edition 9* code 752.2 (double uterus) from 1995-2010 and Hospital International Classification of Diseases Adapted codes 07525-34-0, 07525-36-0, 07525-35-0 (double cervix, including anomaly cervix or uterus) from 1976-2010. Thus, double cervix was defined by the appearance of the cervix clinically as documented by the evaluating gynecologist. Patients identified from this search were then further evaluated to determine who had a pelvic MRI available for review. Patients without an MRI were excluded. Pelvic MRI was the standard imaging method in our department for evaluating patients with MDAs during most of this time period.

Chart review was conducted to identify age at presentation and presenting symptoms, as well as physical examination findings, including the presence of a vaginal septum. MRIs were reviewed by a board certified radiologist with experience in gynecologic imaging. The MRI findings were used as the gold standard to confirm the presence of a double cervix. T2-weighted images were available in all cases and used to evaluate the cervix. Double cervix was defined as 2 cervical canals, from the internal to the external os. Care was taken not to mistake plicae palmatae for a cervical septum. Plicae palmatae are distinguished by recognizing that they do not extend completely across the endocervical canal and that they do not extend from more superiorly in the uterus.[15-17]

Categorization of uterine anomalies was based on the American Fertility Society classification scheme,[3] however, that scheme does not include imaging criteria. T2-weighted images obtained in the coronal plane of the uterus were available in all cases and were used to determine the type of MDA. A cleft in the fundal myometrium greater than 1 cm was the criterion used to diagnose a bicornuate uterus (Figure, A); septate uterus (Figure, B) was diagnosed when there was no cleft or a cleft of less than 1 cm.[14,18] Uterus didelphys (Figure, C) was diagnosed when there were 2 separated uterine horns without communication.[2,18] If separated uterine horns merged above the level of the internal cervical os, we classified the MDA as a bicornuate uterus. If separated uterine horns merged below the level of the internal cervical os, we classified the MDA as uterus didelphys.[2,19] If the distinction between uterus didelphys and bicornuate uterus was equivocal, the uterine anomaly was classified as didelphys.

Additional features related to the cervix were also assessed on the MRI examinations. Orientation of the cervical canals at the external os,[20] cervical canal thickness, and distance between cervical canals was recorded. If the long axis of each cervical canal angled away from each other at the external os, the canals were considered to diverge; if they angled toward each other at the external os, the canals were considered to converge. Cervical canals that neither converged nor diverged were classified as parallel. Distance between cervical canals was measured from inner margin to inner margin in the transverse plane. Cervical canal thickness was measured at its widest point in the transverse plane. The presence of a longitudinal or transverse vaginal septum was recorded if identified on MRI or reported in clinical notes (as some patients had their septum resected before the MRI was performed).

The predominate presenting symptom for each patient was determined by review of clinical notes and was classified into one of the following 8 categories: difficulty obtaining Papanicolaou smear; infertility or reproductive performance concerns; history of recurrent miscarriages; history of second-trimester pregnancy loss; abnormal vaginal bleeding; Papanicolaou smear abnormality; incidental finding/asymptomatic; and dysmenorrhea. These symptom categories were also correlated with the type of MDA.

Statistical analyses were conducted with SAS (version 9.2; SAS Institute, Cary, NC). To test for differences in continuous measures between cervical defects, the Kruskal-Wallis test was used. Tests for differences in the distributions of categorical variables among groups was accomplished with a Pearson χ^2 test. *P* values less than .05 were taken as statistically significant. No adjustment for multiple comparisons has been applied to reported *P* values.

Results

Initial search results revealed 519 patients with a double cervix. Sixty-four of the patients identified with a double cervix had undergone a pelvic MRI with confirmation of a double cervix and constitute our study group.

Of those 64 with double cervix, 32 patients were classified with a septate