The new ASRM müllerian anomaly classification: a picture is worth one thousand words

In this issue of Fertility and Sterility, the Müllerian Anomaly Task Force of the American Society for Reproductive Medicine (ASRM) (previously known as the American Fertility Society [AFS]) introduces us to a long-awaited new classification system for müllerian anomalies (1). The most commonly used previous classification, the AFS system, was published in 1988 (2). It was limited to uterine anomalies. Other classifications have been developed, but they are relatively complex (3, 4). Here is the challenge: the müllerian anomalies each represent defects of embryonic development and have been shown to occur at any site in the process. The possible separate defects are numerous, and many of them have yet to be identified.

Let us put this in perspective by pausing and thinking about reproductive development. The very early embryo has the primordia for both the Wolfian and Müllerian systems. The Müllerian system continues to develop to completion if its development is not arrested by Müllerian inhibiting substance (also known as antimüllerian hormone). In contrast, the Wolfian system will only develop to a certain point and will continue after that only if androgens (i.e., testosterone) are present. The Müllerian system develops first as 2 uterine anlagen (buds) that come together in the midline, fuse, and then canalize. Defects are largely fusion failures or canalization failures or both. One can only imagine the almost limitless possible defects that can occur during this process.

THE ASRM MÜLLERIAN ANOMALY CLASSIFICATION

Like the AFS 1988 classification, the new Müllerian Anomaly Classification (MAC) is based on known defects and uses as its foundation 9 descriptive terms (the first 6 taken from the 1988 classification): müllerian agenesis; cervical agenesis; unicorneate uterus; uterus didelphys; bicornuate uterus; septate uterus; longitudinal vaginal septum; transverse vaginal septum; and complex anomalies. Since there are often several different names given to the same anomaly, it was decided to use the most common and best descriptive terms for each of them. In addition, the new system describes common variations to each of the main categories. It acknowledges the fact that because variations depend on the temporal relationship of the insult to fusion and canalization during the embryologic process, the resulting physical findings will vary, and the number of specific defects is limitless. Rather than present an exhaustive list of defects, only common variations are presented in this new system, and the classification leaves room for adding variations as they are identified. In other words, the new MAC is iterative. Using these common descriptive terms allows the clinician encountering a müllerian anomaly to describe their findings at physical examination and radiologic assessment. In a way, the new MAC is a physical and radiologic examination-based system.

The new MAC goes further than any previous system by including diagnostic studies that are most helpful depending on the defect and adding common treatments. Moreover, it incorporates associated anomalies into the system.

Here is an example of how the new MAC could work. In 1992, we were referred a young adolescent with delayed menarche and monthly cyclic severe pelvic pain (5). On examination, she was found to have an absent vagina with a solid structure in the midline of the pelvis. On ultrasound, she had seemingly a noncanalized uterus and vaginal agenesis. The uterus was removed, and it was confirmed that this was a canalization failure of the mullerian tract. In the uterus, there were very small islands of endometrium along the midline. Using the new MAC, one would proceed from examination to the category of müllerian agenesis and then to the variants. Supplemental information for diagnosis and treatment would be present to help guide management. At future MAC updates, new variants identified could be added to this iterative system if it was felt that by doing so, it would enhance the MAC.

THE MAC INTERACTIVE TOOL

The goals for the development of this new MAC were lofty but not impossible to achieve. You will find in this article that a robust interactive-format educational tool was developed to lead the clinician through the management of the patient presenting with an anomaly. This computer tool is slick and most impressive. One can start with the common anomaly that best fits what is encountered on examination and move through the similar anomalies as well as variants using the diagnostic modalities suggested. While this educational tool is impressive, it, by no means, suggests that it will allow any clinician to care completely for these patients. Historically, the correct diagnoses have often been missed, gone undiagnosed, or incorrectly treated surgically for periods of time. The new tool allows for enhanced awareness of these anomalies and expedient diagnoses. However, the treatment of these anomalies clearly should be performed by the most knowledgeable clinician. Often, the best result occurs with the first treatment. This is especially true if the first treatment is surgical.

The tool associated with this manuscript has a real wow effect. It is cool and slick and covers all bases. It is complete and includes such an enormous amount of information about müllerian anomalies that it could be overwhelming. We believe that it is such a valuable tool that the ASRM needs to develop programs specifically around the ASRM MAC to guide users on how to navigate through its intricacies. For example, it would be ideal for the ASRM to develop a grand rounds presentation for obstetrics and gynecology departments showing specific clinical examples on how to use the tool. Presentations with audience participation would best facilitate retention of the material.

As pointed out in the manuscript, it is also critical that the tool be introduced to physicians who evaluate and/or treat anomalies outside of gynecology. This, for example, would include pediatric and urologic surgeons along with adolescent
medicine specialists. We encourage the ASRM to facilitate the introduction of this tool also to these specialties outside of gynecology by developing and sharing a case-based curriculum that applies the tool to various presentations of congenital anomalies that clinicians may encounter. Short videos would also be helpful.

For any curriculum developed, the drawings/diagrams of this tool will immediately hook the learner who will find themselves moving from common defect to variants and making diagnoses more expediently than ever. Dr. Samantha Pfeifer, Chair, and the MAC Task Force should be congratulated for their outstanding work in this difficult area. By developing this descriptive MAC and the associated tool, they have truly shown that a picture is worth one thousand words!

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