Classification of female's genital tract malformations

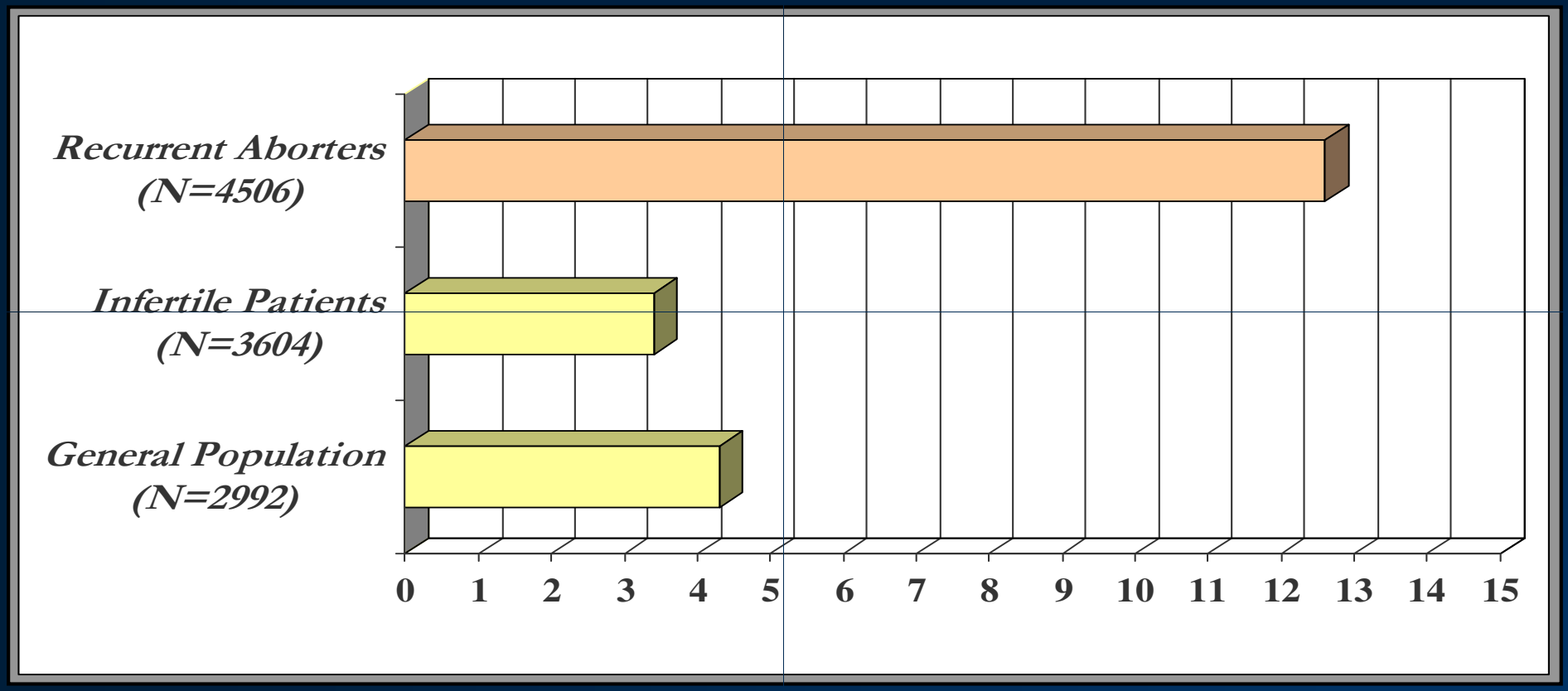
Grigoris F. Grimbizis
Ass. Professor

1st Dept of Obstetrics & Gynecology
Aristotle University of Thessaloniki

Congenital Malformations - Comments

- *Miscellaneous deviations from normal anatomy resulting from embryologic maldevelopment of Müllerian or paramesonephric ducts*
- *High prevalence in the general population (although not absolutely known) and even higher in women with pregnancy losses and implantation failures*

Uterine anomalies: Incidence



Grimbizis et al, Hum Reprod Update, 7: 1-14, 2001

Prevalence: Methodological bias

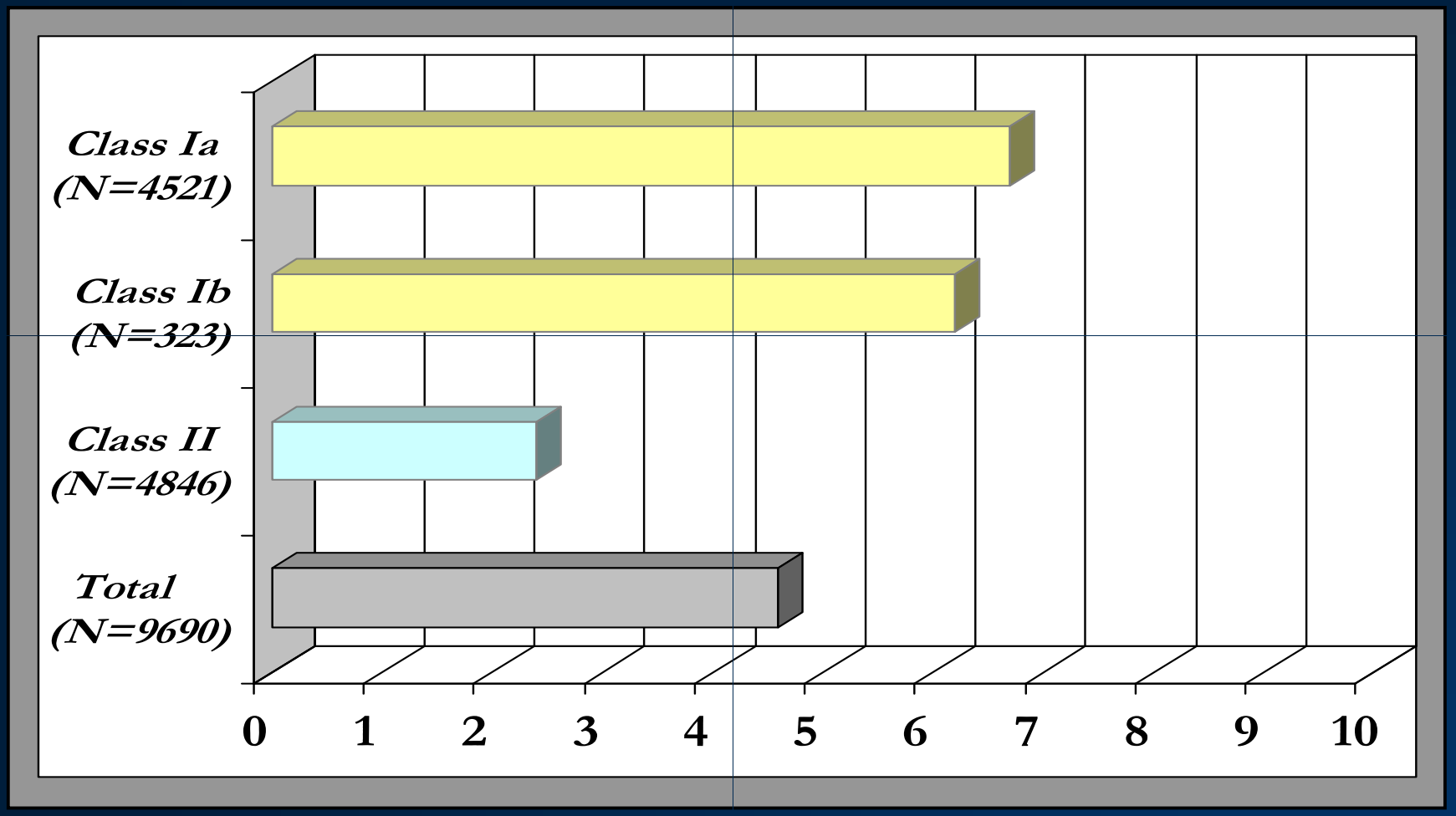
- *different diagnostic methods with different diagnostic accuracy*
- *subjectivity of the criteria used and classify anomalies*
- *non-standarized classification systems (unclear definitions)*
- *inconsistent interpretation of the classification of congenital malformations*

Classification of diagnostic procedures
according to their diagnostic accuracy

Class Ia	**Hyst and Lap SHG 3D US**	***Identification accuracy >90% Classification accuracy into subtypes>90%***
Class Ib	**Hyst (alone)**	***Identification accuracy >90% Non-effective classification into subtypes***
Class II	**HSG 2D US**	***Identification and classification accuracy <90%***
Class III	**MRI Gynecol exam**	***Uncertain accuracy (insufficient data)***

Saravelos et al, Hum Reprod Update, 14: 415-429, 2008

Congenital anomalies: Prevalence in the general population from selected series



Saravelos et al, Hum Reprod Update, 14: 415-429, 2008

Classification Systems: Comments

- Useful and necessary in organizing knowledge
- *Systematic categorization of the patients into groups with similar characteristics*
- *The basic characteristics selected and how they are used create the differences between the systems*
- *The acceptance of a system indicates its ability to effectively corresponds to the needs of the users*

Classification System - Characteristics

1. *Clear and accurate for diagnosis and differential diagnosis*
2. *Comprehensive, incorporating all possible variations*
3. *Correlated with the clinical presentation and the prognosis of the patients*
4. *Correlated with the treatment of the patients*
5. *As simple as possible*

Classification of Female Genital Anomalies
Current Proposals

The American Fertility Society classifications of adnexal adhesions, distal tubal occlusion, tubal occlusion secondary to tubal ligation, tubal pregnancies, Müllerian anomalies and intrauterine adhesions

The American Fertility Society
Müllerian anomalies – Editorial: William Gibbons, M.D.
Committee: Veasy C. Buttram, Jr., M.D., Jan Behrman, M.D., William Gibbons, M.D., Howard Jones, M.D. and John Rock, M.D.

Fertil Steril, 49: 944-955, 1988

Classification of Female Genital Anomalies
Current Proposals

Complex malformations of the female genital tract. New types and revision of classification

Pedro Acién[1,2]*, Maribel Acién[2] and Marisa Sánchez-Ferrer[1]

Acien et al, Hum Reprod, 10: 2377-2384, 2004

Classification of Female Genital Anomalies
Current Proposals

The VCUAM (Vagina Cervix Uterus Adnex–associated Malformation) Classification: a new classification for genital malformations

Peter Oppelt, M.D.,[a] Stefan P. Renner, M.D.,[a] Sara Brucker, M.D.,[b] Pamela L. Strissel, Ph.D.,[a] Reiner Strick, Ph.D.,[a] Patricia G. Oppelt, M.D.,[a] Hellmuth G. Doerr, M.D.,[c] Guenther E. Schott, M.D.,[d] Juergen Hucke, M.D.,[e] Diethelm Wallwiener, M.D.,[b] and Matthias W. Beckmann, M.D.[a]

Oppelt et al, Fertil Steril, 84: 1493-1497, 2005

Congenital Anomalies: A.F.S. Classification

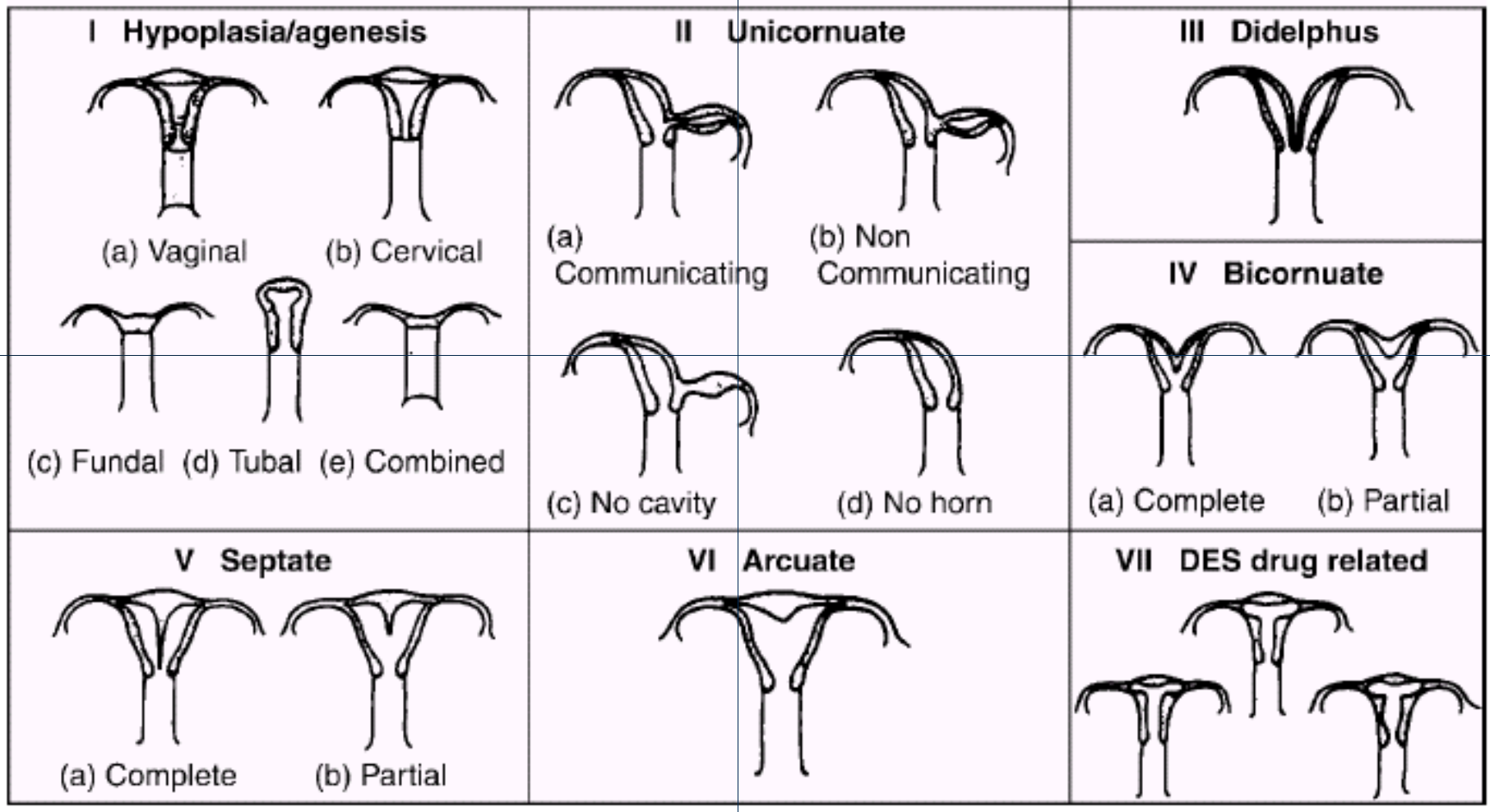


Figure 2. Classification system of müllerian duct anomalies developed by the American Fertility Society (43).

Congenital Anomalies: A.F.S. Classification

Class I	***Hypoplasia & Agenesis***	a. Vaginal b. Cervical	c. Fundal d. Tubal
Class II	***Unicornuate***	a. Communicating b. Non-communicating	c. No cavity d. No horn
Class III	***Didelphys***		
Class IV	***Bicornuate***	a. Partial b. Complete	
Class V	***Septate***	a. Partial b. Complete	
Class VI	***Arcuate***		
Class VII	***DES drug-related***		

A.F.S. Classification: Comments

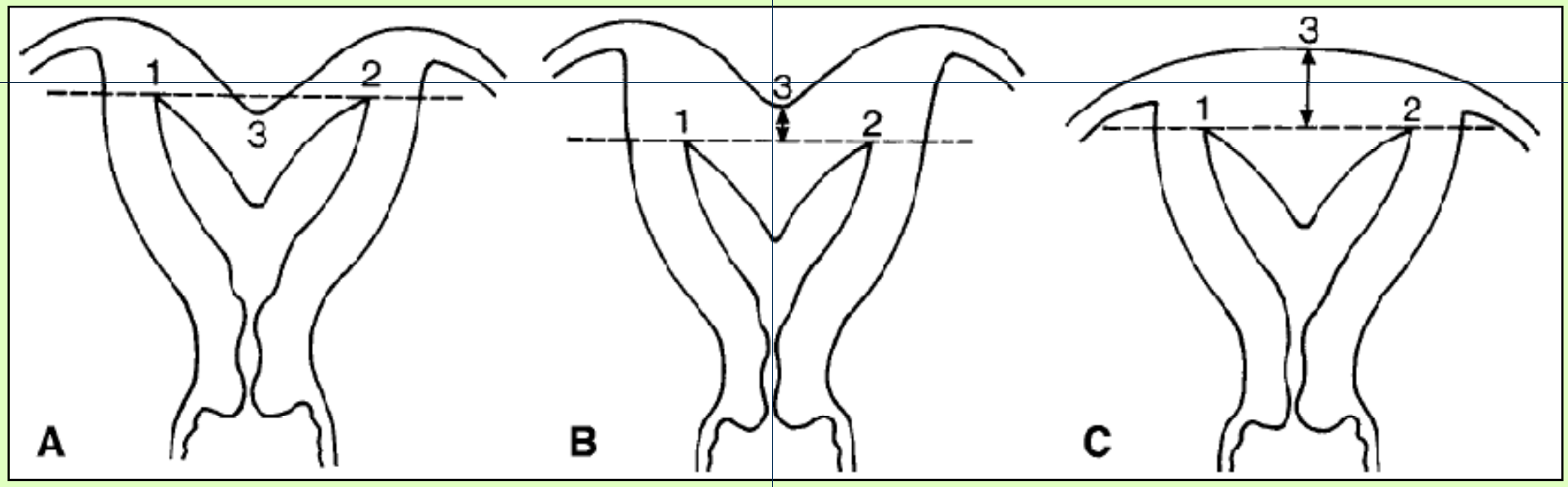
1. *The basis for A.F.S classification system is the anatomy of the female genital tract and especially uterine anatomy*
2. *It is simple, users friendly and clear enough*
3. *It is adopted as the main classification system*
4. *The wide acceptability of this system is explained by the facts*
    1. *the vast majority of congenital malformations are uterine ones and,*
    2. *the classification of congenital anomalies according to the degree of uterine deformity seems to correlate well with patients prognosis*

A.F.S. Classification: limitations

1. *Arcuate uterus should be placed as a separate class?*
2. *Definitions of the different categories are not clear enough for the needs of differential diagnosis between them*
   - *As a result several authors tried to "describe" the differences*
   - *Some "transitional" cases have been reported*

Proposed differential diagnosis between bicornuate & septate uterus

Müllerian Duct Anomalies: Imaging and Clinical Issues



Troiano & McCarthy, Radiology, 233: 19-34, 2004

A.F.S. Classification: limitations

3. *A lot of congenital anomalies are not included in the categories of the system*
   - *bicervical septate uterus, didelphys with obstructing vaginal septum, bicornuate with vaginal/cervical aplasia etc*
4. *"Obstructive" anomalies (vaginal and/or cervical aplasia/dysplasia with functioning uterus) are not representing clearly in the classification system*
5. *General and non-functional place of all aplasias/dysplasias as the first class of the system*
   - *It seems as an effort to avoid a problem than to solve it*

A.F.S. Classification: limitations

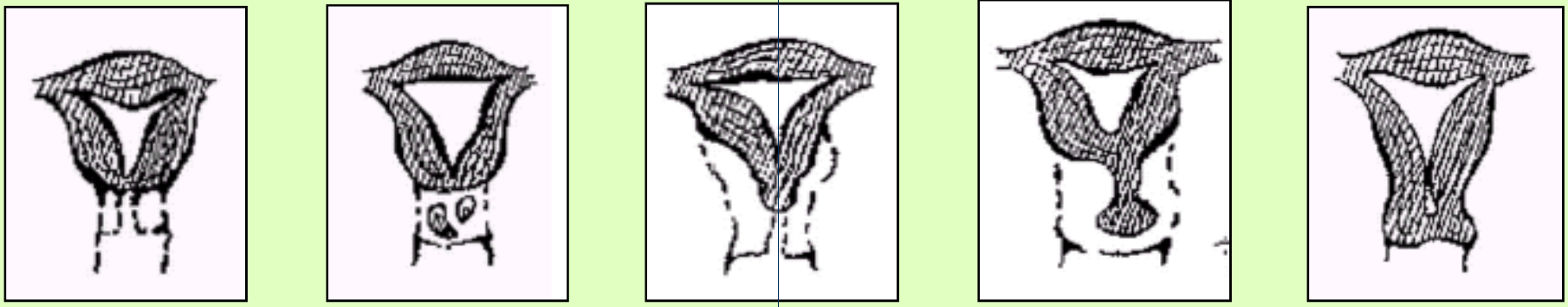
The inability of the AFS classification system to effectively classify "complex" anomalies has as a result

- *two other proposals for a different classification system*
- *subdivisions proposed for certain categories of genital malformations*

Cervical hypoplasia/agenesis (AFS Class Ib) Rock's subclassification

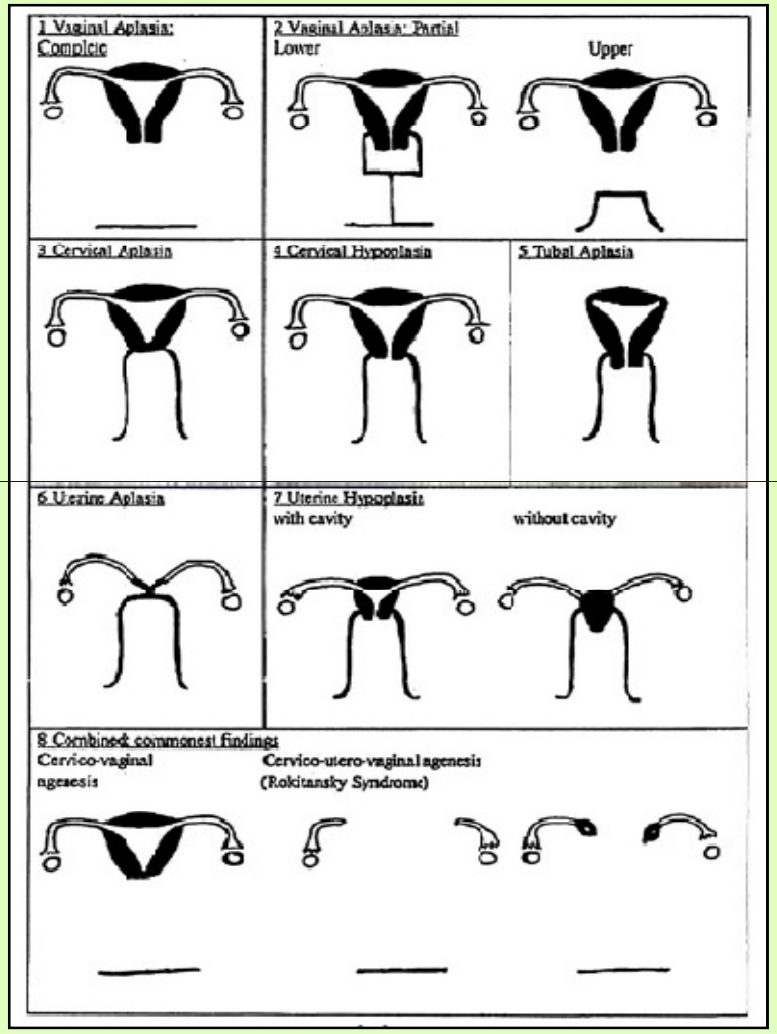
(a) Cervical agenesis (b) Cervical fragmentation (c) Cervical fibrous cord and (d) Cervical obstuction -with or without vaginal aplasia (~50%) (Class I c)

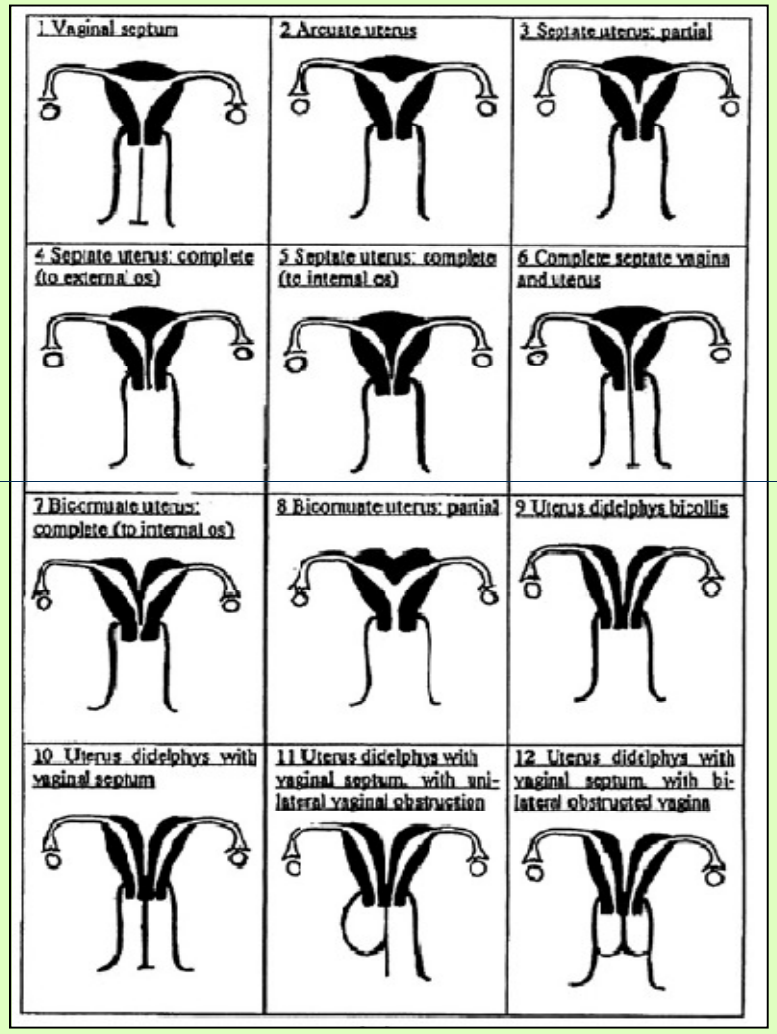
(a) *(b)* *(b)* *(c)* *(d)*



Rock et al, J Pelvic Surg, 1: 129-133, 1995

Subclassification of obstructing Müllerian anomalies





Minto et al, BJOG, 108: 791, 2001

Strawbridge et al, J Pediatr Adolesc Gynecol, 20:195-200, 2007

A.F.S. Classification: Concluding remark

AFS classification system could function as a framework for the description of anomalies rather than an exhaustive list of all possible anomaly types

Clinical & Embryological Classification

Table I. Clinical and embryological classification of the malformations of the female genital tract (modified from Acién, 1992).

1. Agenesis or hypoplasia of a whole urogenital ridge: Unicornuate uterus with uterine, tubal, ovarian and renal agenesis on the contralateral side.
2. **Mesonephric anomalies** with absence of the Wolffian duct opening to the urogenital sinus and of the ureteral bud sprouting (and therefore, renal agenesis). The 'inductor' function of the Wolffian duct on the Müllerian duct is also failing and there is usually:
Utero-vaginal duplicity plus blind hemivagina ipsilateral with the renal agenesis, clinically presented as:
a) Large unilateral hematocolpos*
b) Gartner's pseudocyst on the anterolateral wall of the vagina*
c) Partial reabsorption of intervaginal septum, seen as a 'buttonhole' on the anterolateral wall of the normal vagina which allows access to the genital organs on the renal agenesis side.
d) Vaginal or complete cervico-vaginal unilateral agenesis, ipsilateral with the renal agenesis, and (1) with no communication, or (2) with communication between both hemiuteri (communicating uteri).
3. Isolated **Müllerian anomalies** affecting:
a) Müllerian ducts: they are the common uterine malformations as unicornuate (generally, with uterine rudimentary horn), bicornuate, septate and didelphys uterus.
b) Müllerian tubercle: cervico-vaginal atresia and segmentary anomalies such as transverse vaginal septum.
c) Both, Müllerian tubercle and ducts: (uni- or bilateral) Mayer-Rokitansky-Kuster-Hauser syndrome.
4. Anomalies of the urogenital sinus: cloacal anomalies and others.
5. Malformative combinations: Wolfian, Müllerian and cloacal anomalies.

*These types can associate a vaginal ectopic ureter and interseptal or interuterine communication.

Clinical & Embryological Classification

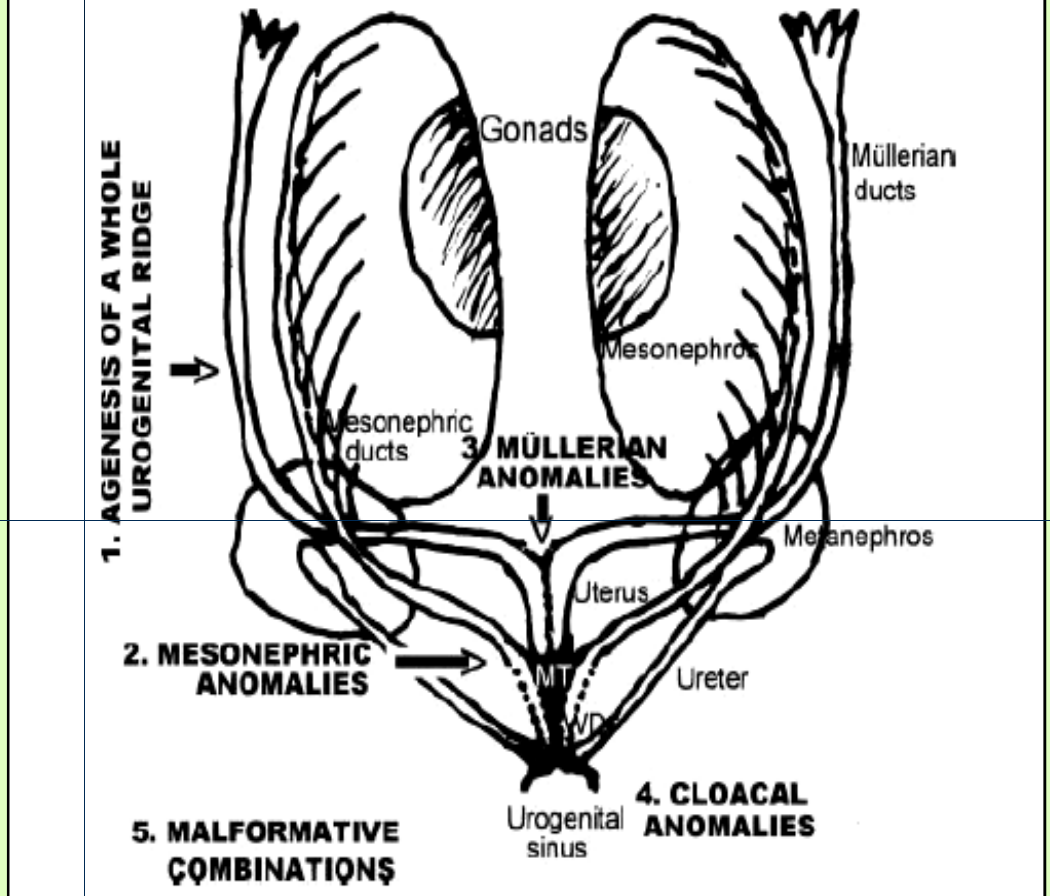


Figure 6. Diagram showing the embryology of the female genital tract and the places for the origin of the five groups of malformations pointed out in Table 1 (WD, Wolffian duct; MT, Müllerian tubercle).

Embryological Classification: Comments

1. *The basis for clinical and embryological classification system is the embryological origin of the different elements of the genitourinary tract*
2. *This system could probably lead to a better understanding of the pathogenesis of the anomalies*
3. *It could be probably more effective than the AFS system in the classification of complex anomalies (a hypothesis that should be tested) as it is based on their pathogenesis*

Emryological Classification: Limitations

1. *It is not simple and users friendly*
2. *There is a radical change on the basis of the classification system from the anatomy to embryogenesis reducing the chances for acceptance*
   - *Anatomy is the basis of the widely accepted AFS System*
   - *Congenital malformations by definition are miscellaneous deviations from normal anatomy*
3. *Patient's clinical manifestations and prognosis seem to be related with anatomical deviations from normality*
4. *Treatment of the patients tend to restore normal anatomy*

Emryological Classification: Concluding remark

Clinical & Emryological classification system could, probably, better explain pathogenesis of congenital malformation but it could not act as a functional framework for the description and treatment of the anomalies

Description of the individual malformation relative to the organ
V C U A M Classification System

Vagina (V)	0	Normal
	1a	Partial hymenal atresia
	1b	Complete hymenal atresia
	2a	Incomplete septate vagina <50%
	2b	Complete septate vagina
	3	Stenosis of the introitus
	4	Hypoplasia
	5a	Unilateral atresia
	5b	Complete atresia
	S1	Sinus urogenitalis (deep confluence)
	S2	Sinus urogenitalis (middle confluence)
	S3	Sinus urogenitalis (high confluence)
	C	Cloacae
	+	Other
	#	Unknown

Cervix (C)	0	Normal
	1	Duplex cervix
	2a	Unilateral atresia/aplasia
	2b	Bilateral atresia/aplasia
	+	Other
	#	Unknown
Uterus (U)	0	Normal
	1a	Arcuate
	1b	Septate <50% of the uterine cavity
	1c	Septate >50% of the uterine cavity
	2	Bicornate
	3	Hypoplastic uterus
	4a	Unilaterally rudimentary or aplastic
	4b	Bilaterally rudimentary or aplastic
	+	Other
	#	Unknown

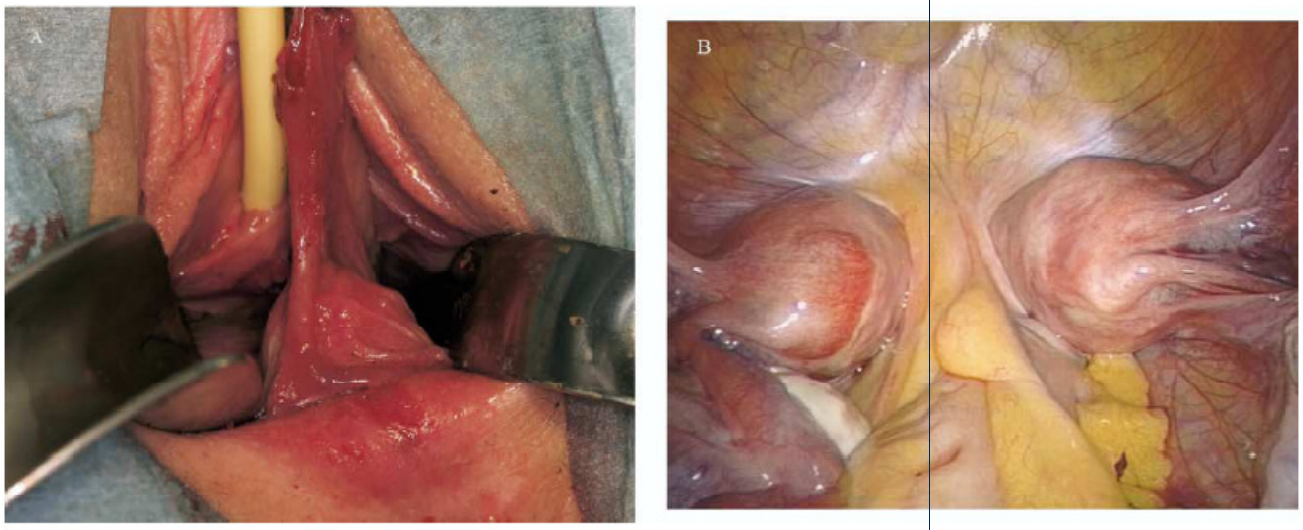
Adnexa (A)	0	Normal
	1a	Unilateral tubal malformation, ovaries normal
	1b	Bilateral tubal malformation, ovaries normal
	2a	Unilateral hypoplasia/gonadal streak (including tubal malformation if appropriate)
	2b	Bilateral hypoplasia/gonadal streak (including tubal malformation if appropriate)
	3a	Unilateral aplasia
	3b	Bilateral aplasia
	+	Other
	#	Unknown
Associated malformation (M)	0	None
	R	Renal system
	S	Skeleton
	C	Cardiac
	N	Neurologic
	+	Other
	#	Unknown

Oppelt et al, Fertil Steril, 84: 1493-1497, 2005

VCUAM Classification: Comments

1. *The basis for VCUAM classification system is the anatomy of the female genital tract*
2. *Each organ is classified separately as is done for breast tumors in the TNM classification*
3. *This gives the opportunity to have a precise, detailed and extremely representative way of classification*
4. *Each type of anomaly, even the more complex, could theoretically and practically be described with this system*

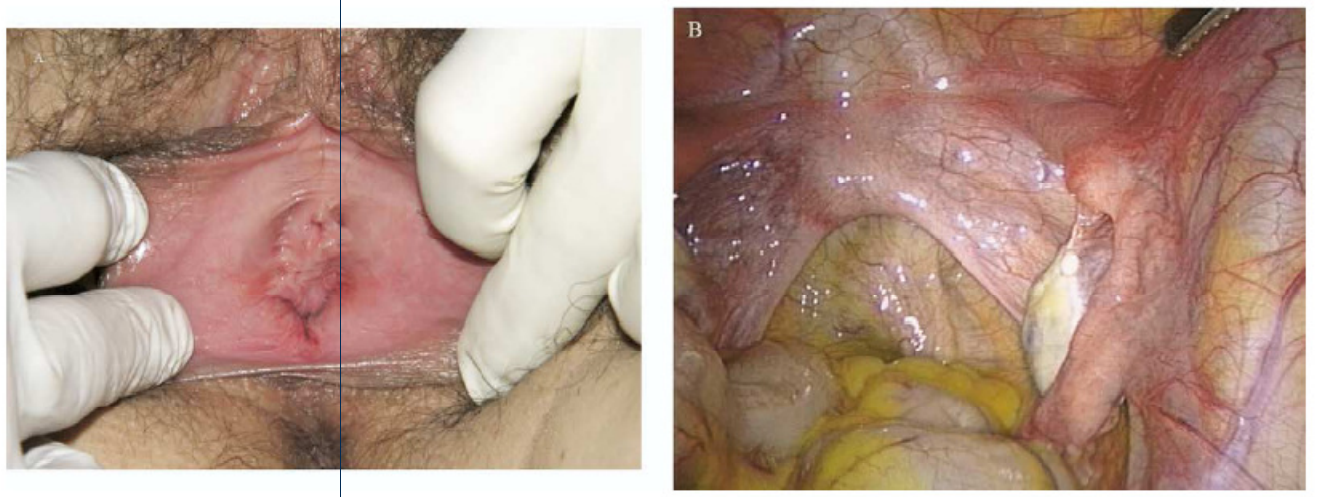
VCUAM classification of uterus didelphys: V2b, C1, U2, A0, M0. (**A**) Vagina. (**B**) Intraoperative view.



V2b, C1, U2, A0, M0

Didelphys Uterus with complete vaginal septum

VCUAM classification of atypical Mayer-Rokitansky-Küster-Hauser syndrome: V5b, C2b, U4b, A0, MR. (**A**) Vagina. (**B**) Intraoperative view.



V5b, C2b, U4b, A0, MR

MKRH Syndrome

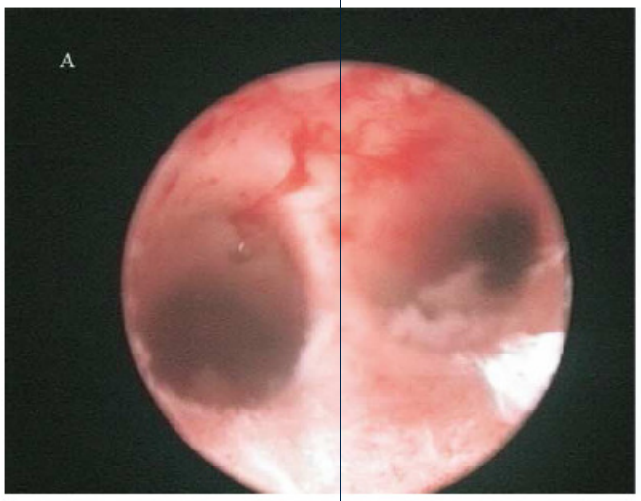
VCUAM Classification: Limitations

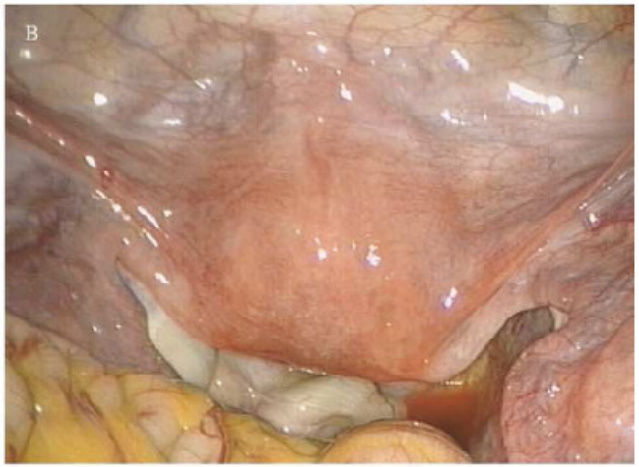
1. *It is not simple and users friendly*
2. *The classification of each patient could only be done with the help of the classification system's tables*
3. *The "translation" of each classified type could also be done only with the use of the classification system's tables*
4. *The anomalies of each organ separately have exactly the same independent importance in the classification system*
   - *Frequency is not taken into account*
   - *Overestimation of the anatomy*
5. *These explain the low acceptability of this proposal (non-functional for everyday use)*

V0, C0, U1b, A0, M0

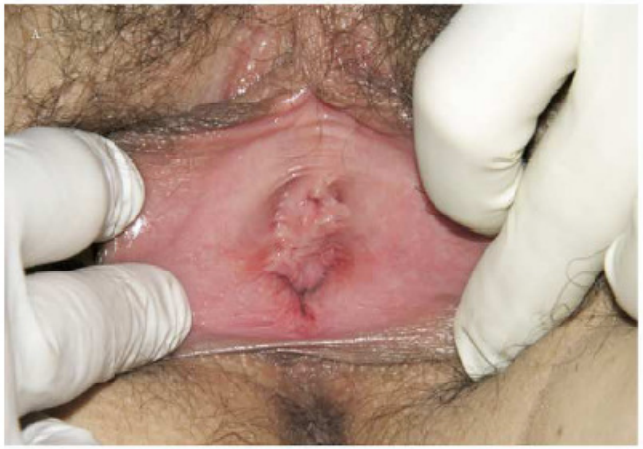
Septate Uterus (partial)

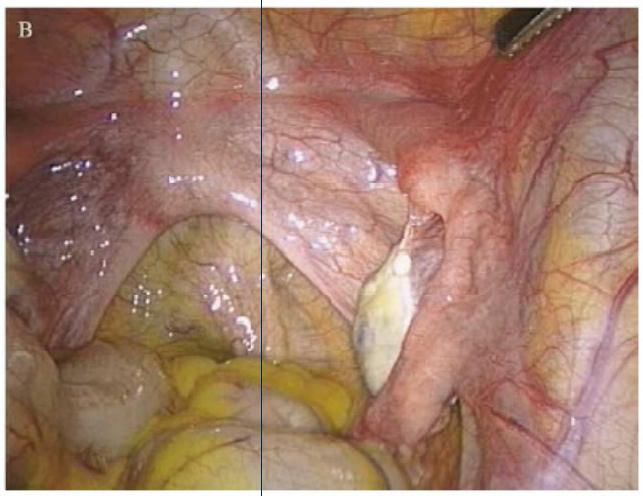
VCUAM classification of uterus septus: V0, C0, U1b, A0, M0. (**A**) Hysteroscopic view. (**B**) Intraoperative view.





VCUAM classification of atypical Mayer-Rokitansky-Küster-Hauser syndrome: V5b, C2b, U4b, A0, MR. (**A**) Vagina. (**B**) Intraoperative view.





V5b, C2b, U4b, A0, MR

MKRH Syndrome

VCUAM Classification: misleading problem

Uterus (U)	0	Normal
	1a	Arcuate
	1b	Septate <50% of the uterine cavity
	1c	Septate >50% of the uterine cavity
	2	Bicornate
	3	Hypoplastic uterus
	4a	Unilaterally rudimentary or aplastic
	4b	Bilaterally rudimentary or aplastic
	+	Other
	#	Unknown

There is a need to discuss the groups of each separate organ from the beginning as is the case for the AFS classification system

VCUAM Classification: Concluding remark

VCUAM classification system could function as a exhaustive list of all possible anomalies types but it could not easily serve as a functional framework for the description of anomalies

Congenital malformations of the female genital tract: the need for a new classification system

Grigoris F. Grimbizis, M.D., Ph.D. and Rudi Campo, M.D.

Fertil Steril, accepted for publication, 2009

The need for a new Classification System

1. *The need for a new classification system is obvious*
2. *The new classification system should critically take into account all the experience gained from the application of the current systems*
3. *The new classification system should fulfill, as much as possible, all the criteria of an ideal classification system*
4. *Although this is a not an easy , it is a necessary task for the patients' management*

The need for a new Classification System
1. Clear and accurate definitions

1. *The classification system should be very clear in the description of its classes and subclasses*
2. *This enables the clinicians to avoid*
   - *subjectivity in the criteria used to recognize and classify each anomaly*
   - *"transitional" cases*
3. *This, also, allows the correct assessment of their prevalence and the prevalence of the different types*
4. *The availability of new methods for the diagnostic work-up make it feasible*

The need for a new Classification System
2. Comprehensive

1. *There are a lot of new variations of undescribed anomalies with unclear classification*
   - *bicervical septate uterus, didelphys with obstructing vaginal septum, bicornuate with vaginal/cervical aplasia etc*
2. *This is the result of our increasing ability to detect them more efficiently and identify their anatomy in details*
   - *due to the use of the new diagnostic methods*
3. *The classification system should be able to incorporate them and be open to new entities*

The need for a new Classification System
3. Correlation with clinical presentation and prognosis

1. *Uterine anomalies are associated with*
   - *poor obstetrical outcome & obstetrical complications dangerous for women's life*
2. *Normal functioning uterus & vaginal and/or cervical aplasia or dysplasia are associated with*
   - *severe health problems urgent for treatment, inability to establish sexual life and reproductive problems*
3. *Uterine & vaginal aplasia are associated with*
   - *inability to establish sexual life and reproductive problems*
4. *The classification system should be correlated in an evidence based basis with clinical presentation and prognosis*

The need for a new Classification System
4. Correlation with patients treatment

1. *In planning our therapeutic strategy it is important to clarify*
   1. *If there is a need of treatment*
   2. *If there is a treatment*
   3. *If the treatment restores the functional problems related to the anomaly*
2. *There is a need for patients' classification according to their therapeutic needs*
3. *The classification system should incorporate treatment related options in their design*

The need for a new Classification System
5. Simple and users' friendly

1. *There should be a direct and obvious association with the anatomy of the female genital system incorporating, if possible, some embryological options*
2. *If possible in one page the scheme of the classification system should be given avoiding the use of complicated tables*
3. *It is not necessary to be analytical and extremely detailed*
4. *Frequency of congenital anomalies should be taken into account*

Concluding Remarks

1. *Congenital malformations represent a common clinical entity*
2. *The most commonly used classification is that of AFS*
3. *There is a need to have a more clear, exact and accurate definition of the different malformations in a new classification system*
4. *This will facilitate researchers and clinicians in evaluating prognosis and planning treatment*

The EAGS in collaboration with ESGE, recognizing the scientific and clinical significance of an updated classification, has established a working group for this issue