The comprehensiveness of the ESHRE/ESGE classification of female genital tract congenital anomalies: a systematic review of cases not classified by the AFS system

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Submitted on August 3, 2014; resubmitted on January 30, 2015; accepted on February 11, 2015

**STUDY QUESTION:** How comprehensive is the recently published European Society of Human Reproduction and Embryology (ESHRE)/European Society for Gynaecological Endoscopy (ESGE) classification system of female genital anomalies?

**SUMMARY ANSWER:** The ESHRE/ESGE classification provides a comprehensive description and categorization of almost all of the currently known anomalies that could not be classified properly with the American Fertility Society (AFS) system.

**WHAT IS KNOWN ALREADY:** Until now, the more accepted classification system, namely that of the AFS, is associated with serious limitations in effective categorization of female genital anomalies. Many cases published in the literature could not be properly classified using the AFS system, yet a clear and accurate classification is a prerequisite for treatment.

**STUDY DESIGN, SIZE AND DURATION:** The CONUTA (CONgenital UTerine Anomalies) ESHRE/ESGE group conducted a systematic review of the literature to examine if those types of anomalies that could not be properly classified with the AFS system could be effectively classified with the use of the new ESHRE/ESGE system. An electronic literature search through Medline, Embase and Cochrane library was carried out from January 1988 to January 2014. Three participants independently screened, selected articles of potential interest and finally extracted data from all the included studies. Any disagreement was discussed and resolved after consultation with a fourth reviewer and the results were assessed independently and approved by all members of the CONUTA group.

**PARTICIPANTS/MATERIALS, SETTING, METHODS:** Among the 143 articles assessed in detail, 120 were finally selected reporting 140 cases that could not properly fit into a specific class of the AFS system. Those 140 cases were clustered in 39 different types of anomalies.

**MAIN RESULTS AND THE ROLE OF CHANCE:** The congenital anomaly involved a single organ in 12 (30.8%) out of the 39 types of anomalies, while multiple organs and/or segments of Müllerian ducts (complex anomaly) were involved in 27 (69.2%) types. Uterus was the organ most frequently involved (30/39: 76.9%), followed by cervix (26/39: 66.7%) and vagina (23/39: 59%). In all 39 types, the ESHRE/ESGE classification system provided a comprehensive description of each single or complex anomaly. A precise categorization was reached in 38 out of 39 types studied. Only one case of a bizarre uterine anomaly, with no clear embryological defect, could not be categorized and thus was placed in Class 6 (un-classified) of the ESHRE/ESGE system.

**LIMITATIONS, REASONS FOR CAUTION:** The review of the literature was thorough but we cannot rule out the possibility that other defects exist which will also require testing in the new ESHRE/ESGE system. These anomalies, however, must be rare.
Introduction

Congenital malformations of the female genital tract consist of a group of miscellaneous deviations from normal anatomy. Although certain types of congenital malformation are the result of a clear disturbance in one stage of embryologic development, others are the result of disturbances in more than one stage of normal formation. The combination of malformations, which occur at different stages of development, seems to be the reason for the extremely wide anatomical variations and the large number of combinations of congenital malformation of the female genital tract observed (Grimbizis and Campo, 2010).

At time of writing, three systems have been proposed for the classification of female genital tract anomalies: that of the American Fertility Society (AFS), now the American Society of Reproductive Medicine (AFS, 1988); the embryological–clinical classification system of genito-urinary malformations (Acién et al., 2004a; Acién and Acién, 2011); and the Vagina, Cervix, Uterus, Adnexa and associated Malformations (VCUAM) system, based on the Tumor, Nodes, Metastases principle in oncology (Oppelt et al., 2005).

The AFS classification system has been successfully adopted as the main classification system for almost two decades as it is simple, user-friendly and clear enough. However this system has several limitations in terms of effective categorization of the anomalies; many congenital anomalies could not be classified in the main categories and subcategories of the AFS system, the borders of arcuate and septate uterus are not clear, AFS class I groups an excessive number of anomalies with totally different clinical presentation, and obstructive anomalies are not adequately represented. A systematic re-evaluation of the all the existing proposals (i.e. the AFS, the embryological clinical and the VCUAM systems) has been already published, underlying the need for a new and updated clinical classification system (Grimbizis and Campo, 2010).

The new European Society of Human Reproduction and Embryology (ESHRE)/European Society for Gynaecological Endoscopy (ESGE) classification system of female genital anomalies is designed mainly for clinical orientation and is based on the anatomy of the female genital tract (Grimbizis et al., 2013a,b). This classification system seems to overcome the limits of the previous attempts; however its clinical value still needs to be proved (Grimbizis et al., 2013a,b).

The aim of the present study is to evaluate the effectiveness and the comprehensiveness of the ESHRE/ESGE classification system focusing on those cases reported in the literature that could not be properly classified by the AFS system.

Materials and Methods

Literature search methodology, study selection and data extraction

We conducted an electronic literature search through Medline, EMBASE and Cochrane library from January 1988 (i.e. date of publication of AFS classification system) to January 2014 using MESH combinations of the following key words: ‘female genital tract anomaly’, ‘müllerian anomaly’, ‘müllerian duct anomaly’, ‘uterine anomaly’ ‘cervical anomaly’, ‘vaginal anomaly’ AND ‘categorization’, ‘classification’, ‘diagnosis’, ‘case report’, ‘exceptional case’, ‘rare anomaly’, ‘rare case’. Only scientific publications in English, Italian, French and Spanish were included. The study was designed and approved by the Scientific Committee (SC) of the CONUTA Working Group.

Three participants in the systematic review (A.D.S.S., C.C., M.S.) screened independently titles and abstracts of studies obtained by the search strategy. All cross-references were hand-searched, as were relevant conference abstracts. All types of studies were selected and each potentially relevant study was obtained in full text and assessed for inclusion independently by the three authors.

The three authors independently extracted data from all included studies. The results were compared and any disagreement was discussed and resolved by consensus after consultation with a fourth reviewer (G.G.). Finally, the results were assessed independently and approved by all the members of the SC of the CONUTA Group.

Outcome measure

The primary outcome measure was the classifiability of all the identified anomalies reported as exceptional and ‘not classifiable’ according to the AFS classification system, into a specific class and/or subclass of the novel ESHRE/ESGE classification system.

Results

Search results

Of the 10 514 related papers, 874 were removed as duplicated articles. A total of 9497 were excluded after reading the abstracts or screening titles (Fig. 1).

Among the 143 remaining articles retrieved for detailed evaluation, 23 were excluded: 15 articles were excluded because the anomaly was described as ‘exceptional’ but could be easily classified with AFS classification system (i.e. unicorurate uterus with a non-communicating rudimentary horn) and 8 articles were excluded because the anomalies were not clearly described or they represented review articles or non-homogenous case series of anomalies already considered in our analysis (Fig. 1).

Classifiability of the included cases

The 120 papers included in the study reported on 140 cases, which could not properly fit into a specific class of the AFS system.

These 140 cases were clustered in 39 different types of anomalies (Table I); the uterus was the organ most frequently involved (30/39: 76.9%), followed by cervix (26/39: 66.7%) and vagina (23/39: 59%). The congenital anomaly involved a single organ in 12 types of anomalies (12/39: 30.8%), while multiple organs and/or segments of Müllerian ducts in more than one stages of embryologic development (complex
anomalies) were simultaneously affected in 27 types of anomalies (27/39: 69.2%).

In all 39 types of anomalies, the ESHRE/ESGE classification system provided a comprehensive description of each single or complex anomaly (Table I). A precise categorization of single or complex anomalies was reached in 38 out of the 39 different types studied. In the following cases a consultation with the fourth reviewer was necessary before classification: Capito and Sarnacki (2009), Gupta et al. (2007) and Singhal et al. (2003) (Robert’s uterus or Complete septate uterus/unilateral cervical aplasia/normal vagina ESHRE/ESGE Class U2b C3 V0), Nezhat and Smith (1999) (Hemi-uterus with rudimentary cavities/ESHRE/ESGE Class U4a C0 V0), Wright et al. (2011) (Aplastic uterus with bilateral rudimentary cavities ESHRE/ESGE Class U5a C0 V0), Sadik et al. (2002) (Aplastic uterus/cervical aplasia/normal vagina/ESHRE/ESGE Class U3b C4 V0), Medema et al. (2008) (Normal uterus with rudimentary horns/ESHRE/ESGE Class U6). The case reported by Nezhat and Smith (1999) having a hemi-uterus with ipsilateral rudimentary horns with cavity could be classified as Hemi-uterus with rudimentary cavity (ESHRE/ESGE Class U4a C0 V0) since the presence of the cavity and not the number is the clinically important parameter for the classification. The only anomaly that could not be perfectly categorized with the ESHRE/ESGE system was that reported by Medema et al. (2008), in which a ‘tricavitated’ uterus was described (Table I).

**Discussion**

Although several classification systems for female genital tract anomalies have been proposed (AFS, 1988; Acien et al., 2004a, 2011; Oppelt et al., 2005), the AFS classification is still the most widely used for categorizing such abnormalities. The AFS system provides a description and classification of the main uterine anomalies appropriate for the vast majority of the patients. However, it is not comprehensive, which hampers precise description of each anomaly and prediction of feasibility and safety of surgical correction (Mazouni et al., 2008; Saravelos et al., 2008; Grimbizis and Campo, 2010).

Thus, ‘obstructive’ anomalies, as a result of cervical and/or vaginal aplasias and/or dysplasias in the presence of either a normal or deformed but functional uterus, are not represented in the AFS classification system. Malformations with anatomical characteristics included in more than one category cannot be classified individually and precisely. AFS class I, including cases with hypoplasia and/or dysgenesis of the vagina, cervix, uterus and/or adnexae, incorporates severe and complex types of congenital anomalies with serious clinical manifestations usually needing complex surgical treatments. A clear and accurate classification is a prerequisite for their treatment, which is not the case with the AFS system. It is noteworthy to mention that these limitations also gave rise to further subdivisions for these categories of anomaly (Rock et al., 1995, 2010; Joki-Erkkila and Heinonen, 2003; Troiano and McCarthy, 2004; Strawbrigde et al., 2007).

In 38 out of 39 types of anomalies included in this study as previously un-classified, the ESHRE/ESGE classification provided a comprehensive description and categorization of each single or complex anomaly in all cases.

All these 39 types of anomalies could not be described and categorized previously with the AFS system; as a consequence the terminology used by the authors to describe them is often ‘liberal’ and mostly subjective. The ESHRE/ESGE classification system gives the opportunity to replace inappropriate descriptions within the AFS system (i.e. ‘didelphys’
Table I  Classification of the previously un-classified cases using the European Society of Human Reproduction and Embryology/European Society for Gynaecological Endoscopy (ESHRE/ESGE) Classification system of female genital anomalies: the terminology provided by the authors for the description of the anomaly is ‘translated’ to that of the new system before classification.

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<th>Publication</th>
<th>Uterus Authors’ description</th>
<th>ESHRE/ESGE terminology</th>
<th>Cervix Authors’ description</th>
<th>ESHRE/ESGE terminology</th>
<th>Vagina Authors’ description</th>
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<td>Rajamaheswari et al. (2009), Di Spiezio Sardo et al. (2007) and Demirci et al. (1995)</td>
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<td>Normal</td>
<td>Normal</td>
<td>Normal</td>
<td>Longitudinal vaginal septum</td>
<td>Longitudinal non-obstructing vaginal septum</td>
<td>U₀ C₀ V₁</td>
<td>Non-obstructive transverse vaginal septum (partial failure of canalization/vertical fusion defect with incomplete unification of urogenital sinus and paramesonephric duct)</td>
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<tr>
<td>Cetinkaya et al. (2011) and Levy et al. (1997)</td>
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<td>Normal</td>
<td>Normal</td>
<td>Normal</td>
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</tr>
<tr>
<td>Acien et al. (2009) (case 6)</td>
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<td>Normal</td>
<td>Bicervical</td>
<td>Double normal</td>
<td>Double or septate vagina</td>
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<td>*One cervix is blind (not communicating with uterine cavity)</td>
</tr>
<tr>
<td>Morales-Rosello and Peralta Llorens (2011)</td>
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<td>Normal</td>
<td>Double*</td>
<td>Double normal</td>
<td>Double vagina</td>
<td>Longitudinal non-obstructing vaginal septum</td>
<td>U₀ C₂ V₁</td>
<td>*Double cervix communicating bilaterally with uterine cavity § The use of term ‘double vagina’ is not correct as in such case the vagina is single and divided by a septum</td>
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<td>Candiani et al. (1996)<em>, Dunn and Hantes (2004)</em>, Goldberg and Falcone (1996)* and Keltz et al. (1994)*</td>
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<td>Normal</td>
<td>Double*</td>
<td>Double</td>
<td>Longitudinal non-obstructing vaginal septum</td>
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<td></td>
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<tr>
<td>Shirota et al. (2009) and Varras et al. (2007)*§</td>
<td>Normal</td>
<td>Normal</td>
<td>Double*</td>
<td>Double normal</td>
<td>Double vagina</td>
<td>Longitudinal non-obstructing vaginal septum</td>
<td>U₀ C₂ V₁</td>
<td>*Double cervix communicating bilaterally with uterine cavity § The use of term ‘double vagina’ is not correct as in such case the vagina is single and divided by a septum</td>
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<th>Publication</th>
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<th>Authors' description</th>
<th>ESHRE/ESGE terminology</th>
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<td></td>
<td>The use of term ‘two cervices’ is not correct as in such case the cervix is single and divided by a septum</td>
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<td>Fedele et al. (2012)</td>
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<td>Complete septate uterus</td>
<td>Septate</td>
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<td>Acien et al. (2009) (case 5) and Lev-Toaff et al. (1992)</td>
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<td>Complete septate uterus</td>
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<td>Hur et al. (2007), Fedele et al. (2013) (10/87 cases) and Acien et al. (2004c)</td>
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<td>Complete septate uterus</td>
<td>Double cervices</td>
<td>Double normal</td>
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<td>Longitudinal obstructing vaginal septum</td>
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<td>Ziebarth et al. (2007) (case 2)</td>
<td>Bicornuate</td>
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<td>Single</td>
<td>Normal</td>
<td>Obstructed hemivagina</td>
<td>Longitudinal obstructing vaginal septum</td>
<td>U3b C0 V2</td>
<td></td>
<td></td>
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<tr>
<td>Fedele et al. (2013) (1/87 case)</td>
<td>Bicornuate</td>
<td>Partial Bicorporeal</td>
<td>Septate</td>
<td>Septate</td>
<td>Unilateral obstructed hemivagina</td>
<td>Longitudinal obstructive vaginal septum</td>
<td>U3b C1 V2</td>
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<td>Fedele et al. (2013) (9/87 cases)</td>
<td>Bicornuate</td>
<td>Partial Bicorporeal</td>
<td>Bicollis</td>
<td>Double normal</td>
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<td>Longitudinal obstructive vaginal septum</td>
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*In the case of Acien et al. (2004c) associated malformation: an ectopic ureter joined to ipsilateral hemi-cervix

* The described ‘obstructing’ uterine septum is the result of unilateral cervical aplasia
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<td>Single</td>
<td>Normal</td>
<td>Single</td>
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<td>Adair et al. (2011) and Altchek et al. (2009)</td>
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<td>Complete</td>
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<td>Single</td>
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<td>Septate</td>
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<tr>
<td>Acien et al. (2009) (case 2) and Acien et al. (2010a,b)</td>
<td>Didelphys</td>
<td>Complete</td>
<td>Bicorporeal</td>
<td>Septate</td>
<td>Septate</td>
<td>Septate</td>
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<tr>
<td>Coskun et al. (2008)</td>
<td>Didelphys</td>
<td>Complete</td>
<td>Bicorporeal</td>
<td>Double</td>
<td>Double normal</td>
<td>Obstructed unilateral vagina by a transverse vaginal septum</td>
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</table>

* In the case of Acien et al. (2010a,b) associated malformations: Gardner’s duct cyst, ipsilateral renal agenesis, blind hemibladder and ectopic ureterocele.
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<td>Double normal</td>
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<td>Longitudinal non-obstructing and obstructive transverse vaginal septum</td>
<td>U36, C2 V1+3</td>
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<td>Growdon and Laufer (2008) and Singhal et al. (2013)</td>
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<td>Complete Bicornoreal</td>
<td>Double</td>
<td>Double normal</td>
<td>Lower vaginal agenesis</td>
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<td>Vaginal aplasia</td>
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<td>Fedele et al. (2013) (4/87 cases), Acién et al. (2004b) and Acién et al. (2008)</td>
<td>Didelphys</td>
<td>Complete Bicornoreal</td>
<td>Unilateral cervical atresia</td>
<td>Unilateral cervical aplasia</td>
<td>Vaginal agenesis</td>
<td>Normal</td>
<td>Vaginal aplasia</td>
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<td>Singh and Sunita (2008) and Bakri et al. (1992)</td>
<td>Didelphys</td>
<td>Complete Bicornoreal</td>
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<td>Jain et al. (2013)</td>
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<td>Nezhat and Smith (1999)</td>
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<td>Normal</td>
<td>Normal</td>
<td>Vaginal aplasia</td>
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<td>Engmann et al. (2004)*</td>
<td>Unicornate uterus with normal external morphology</td>
<td>Hemi-uterus without rudimentary cavity with normal external morphology</td>
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<td>Normal</td>
<td>Normal</td>
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<td>Deligeorgiou et al. (2007), Mais et al. (1994) and Heinonen (1997)</td>
<td>Unicornate uterus without contralateral horn</td>
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<td>Imperforate hymen and transverse vaginal septum</td>
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<td>Wright et al. (2011)</td>
<td>Two hemi-uteri with endometrial cavities (no connection with normal cervix)</td>
<td>Aplastic Uterus with Bilateral rudimentary cavities</td>
<td>Normal</td>
<td>Normal</td>
<td>Normal</td>
<td>Vaginal aplasia</td>
<td>U36/C0/V0</td>
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*Incomplete canalization of one Mullerian duct initiated from two distinct sites resulting in two cavitated horns Failure of lateral fusion.
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<tr>
<td>Christopoulos et al. (2009)</td>
<td>Didelphys uterus with noncanalized horns</td>
<td>Aplastic Uterus with Bilateral rudimentary horns without cavity</td>
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<tr>
<td>Sadik et al. (2002)</td>
<td>'Middle' hypoplastic non-cavitated uterus/Two rudimentary horns—no endometrium</td>
<td>Aplastic uterus*</td>
<td>Cervical aplasia*</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Sole cervix, small in size with non-patent lumen</td>
<td></td>
</tr>
<tr>
<td>Medema et al. (2008)</td>
<td>Tricavitated</td>
<td>Normal uterus with two additional rudimentary functional horns</td>
<td>Not described clearly</td>
</tr>
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AFS, American Fertility Society; OHVIRA, Obstructed Hemivagina and Ipsilateral Renal Anomaly.
in presence of single cervix or ‘two cervices’ in presence of single cervix with a septum) or subjective and mostly ‘liberal’ terms due to the absence of terminology (i.e. ‘obstructive hemivagina’ or ‘hybrid septate and bicornuate uterus’) with the more objective ones used for the classes and subclasses of the system such as ‘complete bicornoreal uterus with single cervix’, ‘septate cervix’, ‘longitudinal obstructing vaginal septum’ and ‘bicornoreal septate’, respectively. This is a ‘proof’ that with the use of the new ESHRE/ESGE system, a common terminology could be adopted for communication among clinicians to convey the exact anatomical status of the female genital tract, which is the primary basic characteristic in the design of the classes and subclasses of the system.

The only anomaly that could not be perfectly categorized with the ESHRE/ESGE system was that of a reported ‘tricavitated uterus’ (Medema et al., 2008). According to the ESHRE/ESGE classification system this ‘bizarre’ uterine anomaly was clearly described as ‘normal uterus with two additional rudimentary functional horns’, but a precise categorization was not possible (i.e. U6C0V0). Two possible explanations of this complex uterine anomaly can be given: (i) Müllerian ‘duplication’ at the level of the uterine body responsible for the presence of the two functional rudimentary horns or (ii) aplasia of the mid part of the uterus combined with a fusion defect of the upper part. Indeed, potential duplication defects of Müllerian ducts like that of Medema et al. (2008), could not be categorized with the use of the new system by its design. In another case of ‘perforated’ vaginal septum, although it could be successfully classified with the ESHRE/ESGE system, the clinical importance of the existing perforation would have been underestimated without adding a proper comment.

An important characteristic of the ESHRE/ESGE classification system is the independent classification of uterine, cervical and vaginal anomalies. Thus, 22 out of the 39 types of anomalies (54.2%) were related to ‘obstructive’ anomalies that could not be described by the AFS classification system. All these cases could be easily and precisely classified into specific subclasses of the ESHRE/ESGE classification system due to this characteristic of the system. Furthermore, 7 out of the 39 (18%) types of anomalies identified referred to cases of hypoplasia and/or dysgenesis of the vagina, cervix and/or the uterus. All these malformations, otherwise nondescript or inappropriately grouped together into the same class I of the AFS classification system, could be properly and correctly classified into subclasses, expressing each one as a specific anatomical deviation. In other words, complex anomalies could be easily classified, due to the possibility to describe independently anomalies of different areas of the genital tract (uterus, cervix and vagina) and combine them case by case. However, it should be noted that, although some of these complex anomalies may be classified equally successfully by using other existing classifications (e.g. U2bC2 of the ESHRE/ESGE classification equates to C1U1c of the VCUAM classification), those systems have other disadvantages (Grimbizis and Campo, 2010), which would limit their use.

The new classification also ‘promotes’ the description of ‘associated anomalies of non-Müllerian origin’, which is so important particularly in the complex anomalies where a significant number will have associated renal tract malformations. This was not accounted for in the AFS classification and is a further advantage of the new classification.

Another important advantage of the ESHRE/ESGE classification system is that embryological origin has been adopted as the secondary basic characteristic in the design of the main classes. In fact, using this classification we could have an image of the embryological defect and, for example, such a rare anomaly as ‘Robert’s uterus’ could be easily categorized as ‘complete septate uterus with unilateral cervical aplasia’ (U2bC3V0).

The system seems to be simple and functional because it has a direct and obvious association with the anatomy of the female genital system, without using complicated tables. As the AFS committee for the classification of congenital anomalies pointed out, the scheme of any classification system should be given in one page and this is fulfilled completely with the ESHRE/ESGE system. Furthermore, for cases where the anatomy is very difficult to be described only with the subclasses of the system (e.g. tricavitated uterus), an additional note could be made within the classification scheme (see ‘Comments’ in Table I). This may improve the skillful combination of simplicity and comprehensiveness.

From our analysis, uterine anomalies defined as ‘arcuate’ uterus with the AFS system are not included because this category no longer exists in the ESHRE/ESGE classification system and the patients should be categorized as normal or septate, depending on the degree of midline indentation; thus all these patients could be classified with the new system, albeit in a different way. Furthermore, accessory and cavitated uterine masses (ACUM) were excluded because their pathogenesis is still controversial. Although some authors consider them as a new type of Müllerian anomaly (probably related to a dysfunction of the female gubernaculum), others support the notion that they are simply adeno-myotic cysts (Acién et al., 2010a, 2011, 2012; Bedaiwy et al., 2013).

Conclusions
An important characteristic of an ‘ideal’ classification system is to be comprehensive, incorporating all possible variations and offering a clear and distinct description and categorization for them. This facilitates enormously diagnosis and differential diagnosis, evaluation of their prognosis and planning their treatment. The new ESHRE/ESGE classification system may overcome the limits of the previous classification systems, providing an effective and comprehensive categorization of almost all the currently known anomalies of the female genital tract.

Suggestions for future research
The comprehensiveness of the ESHRE/ESGE classification adds objective scientific validity to its use. This may, therefore, promote its further dissemination and acceptance, which will have a positive outcome in clinical care and research. Offering a common language between the researchers, in the near future the ESHRE/ESGE classification system of female genital anomalies could be used as a tool for the development of guidelines for their diagnosis and treatment, further facilitating daily clinical practice.

Authors’ roles
Conception and design: G.G., A.D.S.S., R.C. and S.G.; Literature search and data extraction: A.D.S.S., M.S and C.C.; Assessment of the quality of studies: A.D.S.S., M.S and C.C.; Analysis and interpretation of the data: A.D.S.S., M.S., C.C. and G.G.; Drafting the article: A.D.S.S. and G.G.; Critical revision of the article: all the authors (CONUTA Scientific Committee); Approval of results: CONUTA Scientific Committee.

Funding
The authors declare no source of funding or other financial support. Funding to pay the Open Access publication charges for this article was provided by the European Society of Human Reproduction and Embryology (ESHRE).
Conflict of interest

The authors report no conflict of interest. The authors alone are responsible for the content and writing of this paper.

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