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Original Research Article

Congenital Mullerian anomalies may lead to acute life threatening emergency in reproductive age group- An experience in a tertiary care centre in Eastern India

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ABSTRACT

Background: Congenital Mullerian anomalies are associated with higher incidence of reproductive failure and adverse obstetrical outcomes. The new ESHRE classification is a precise way to describe a genital anomaly in simple way. Many genital tract anomalies lead to devastating life-threatening conditions but under-reported and poorly explained. This study aims at finding out those anomalies which more frequently lead to specific diseases needing emergency intervention to save life of the patient.

Materials and Methods: This longitudinal observational study evaluates the diagnosis and treatment of 12 patients with congenital Mullerian abnormalities which led to life threatening emergency with case discussion and pictorial documentation.

Results: Among 12 women with uterine anomalies 4 abortions, 7 delivered preterm, 1 term delivery occurred. Majority of complications associated with U3 types (58.3%). Other types with complications are U4 (25%) and U2 (16.7%). Presenting features at emergency varied like mimicking ectopic, labour complications, ruptured corpus, placenta previa and accreta as well as iatrogenic perforation.

Conclusions: Presence of congenital uterine anomalies have adverse effect on obstetrical outcome. Majority of complications are associated with U2, U3 & U4. Probably U1 doesn't interfere with ongoing pregnancy and U5 fails to continue pregnancy. This knowledge warrants the need for a larger case control study to extrapolate these findings to the general population and also to recommend the need for universal Prenatal Screening for uterine anomalies so as to improve the obstetrical outcome in patients with uterine anomalies.

1. Introduction

Congenital uterine anomalies could be defined in simpler terms as malformation in uterus occurring during the development of embryo. Congenital uterine malformations are generally and most common misdiagnosed disease we encountered. Adequate knowledge concerning their prevalence and varieties and early detection is important in recognizing and managing the obstetric and gynaecologic complications.[1]

Congenital uterine anomalies result from failure of complete or incomplete development, fusion or canalisation of one or both Mullerian ducts during foetal life[2] and has been associated with an increased rate of miscarriage, preterm delivery and other adverse foetal outcomes.[3,4] They represent a rather common benign condition with a wide ranging symptoms from dysmenorrhea, infertility, miscarriage, preterm labour, obstructed labour 3^{rd} stage complications soon.[5–7] In 1988 American Fertility Society came out with the first classification based on work of Buttram and Gibbons.[8] They are classified as The American Fertility Society(currently American Society of Reproductive Medicine system,

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ASRM),[9] The embryological-clinical classification system of genito-urinary malformations[10] and the Vagina, Cervix, Uterus, Adnexa and associated Malformations system based on the Tumour, Nodes, Metastases principle in Oncology.[11] Normal pregnancies can occur in patients with Mullerian duct anomalies, but obstetric complications are frequent.[12,13] Adequate assessment is essential in these patients which could further improve their prenatal outcome.[14] But in our set up, where most people belong to very low socio-economic striate of the society whom often lack proper education and awareness, fail to attend proper ANC check-ups and ante-natal diagnosis of any uterine anomalies becomes difficult. In the present study, we evaluated the pregnancy outcome of patients with uterine malformations, who have severe obstetric complications due to Mullerian anomaly.

2. Materials and Methods

A longitudinal observational study was planned and conducted at the Department of Obstetrics at College of Medicine & JNM hospital, Kalyani between the time period from 1st September 2017 to 31st August 2022 . Before starting the study, clearance was taken from ethical committee of the hospital.

2.1. Case selection

A total of 48 women were selected for the study with mullerean anomalies in the Department of Gynaecology & Obstetrics.

2.2. Inclusion criteria

Reproductive age group patients with mullerian anomalies who got admitted leading to emergency life threatening conditions requiring immediate interventions and expertise to save maternal or fetal life. Cases may be already known to have Mullerian anomalies or may be incidentally found during operative procedures.

2.3. Exclusion criteria

Admitted patients with known Mullerian anomaly but not presenting with life-threatening obstetric emergencies.

2.4. Statical methods

Total of 48 cases we got with mullerean anomalies in the emergency of Gynaecology & Obstetrics department. Among them 32 cases were in the reproductive age group. Most of the cases were with subfertility related issues. 16 cases were in the adolescence period with some minor problems. Amongst the 32 patients, 12 cases presented with acute emergency life threatening conditions due to congenital Mullerian defects. The study was multiparametric in design. It focused on detecting the type of anomaly which led to life threatening conditions like hemorrhagic shock due to ruptured uterus, or ectopic, obstructed labour, iatrogenic uterine perforation etc. There are multiple parameters of an individual case. The study focused on the chief complaint of the cases at presentation to Emergency Room , relevant investigations, ant treatment approach specially the OT notes and findings. In this study we tried to estimate the relationship between uterine anomalies and their outcomes on women of reproductive age group. We followed the ESHRE classification of female gental tract anomalies in this study.

2.5. Evaluation criteria

To determine and evaluate the life threatening condition we followed the gravity of the situation which made us to treat the patient promptly to save the life. The conditions in which patient was in shock, with hemoperitoneum obviating urgent intervention, retained placenta, PPH etc. All these cases almost led to tachycardia>120/min., BP<100/70, severe pallor, increased respiratory rate, need of HDU for post-operative recovery even need of mechanical ventilation, and lastly most of the cases needed more than 4 PRBC transfusion. Case discussions with pictorial presentation is used to elaborate the cases as mullerean anomaly is uncommon but often missed in our day to day practice. We kept all the data and photograph of patients presented with life threatening obstetric complications and mullerian anomalies in the in-patient ward and High Dependency Unit in that time period as much as possible.

	Uterine anomaly		Cervical / Vaginal anomaly	
	Main class	*Sub-class*	*Co-existent class*	
U0	Normal uterus		C0	*Normal cervix*
U1	Dysmorphic uterus	a. T-shaped b. Infantilis c. Others	C1	*Septate cervix*
U2	Septate uterus	a. Partial b. Complete	C2	*Double "normal" cervix*
U3	Bicorporeal uterus	a. Partial b. Complete c. Bicorporeal septate	C3	*Unilateral cervical aplasia*
U4	Hemi-uterus	a. With rudimentary cavity (communicating or not horn) b. Without rudimentary cavity (horn without cavity / no horn)	C4	*Cervical Aplasia*
U5	Aplastic	a. With rudimentary cavity (bi- or unilateral horn) b. Without rudimentary cavity (bi- or unilateral uterine remnants / Aplasia)	V0	*Normal vagina*
			V1	*Longitudinal non-obstructing vaginal septum*
			V2	*Longitudinal obstructing vaginal septum*
			V3	*Transverse vaginal septum and/or imperforate hymen*
			V4	*Vaginal aplasia*
U6	Unclassified Malformations			
U			*C*	*V*

Figure 1: The following ESHRE Classification for diagnosing the congenital Mullerian defects were followed.
Source: https://gynecolsurg.springeropen.com/articles/10.1007/s10397-013-0800-x

Table 1: Brief case discussions and classification according to ESHRE

Case No.	Presenting Features	Provisional diagnosis at presentation	Intervention	Intraoperative findings and management	ESHRE Classificatio	Final diagnosis
Case 1	Newly married, 18 years, nullipara, 6 wks amenorrhoea f/b bleeding P/V. Acute pain at left iliac fossa Cyclical pain of similar type UPT negative Outside USG- A COMPLEX SOL at left adnexa-exclude ectopic	Suspected chronic ectopic pregnancy	Laparotomy followed by resection of rudimentary horn. 5 units PRBC transfusion HDU monitoring	Hemiuterus with non-communicating rudimentary horn with menstrual blood in cavity mimicking chronic ectopic (Figure 2)	U4aC0V0	Ruptured rudimentary horn with functional endometrial cavity in a hemiuterus. Mimics ectopic
Case 2	20 years, primi para, 12 wks amenorrhoea with features of hemorrhagic shock, UPT-positive,	Ruptured ectopic	Laparotomy followed by resection of horn 6 units PRBC transfusion HDU monitoring	pregnancy in a communicating rudimentary horn that ruptured – (Figure 3)	U4aC0V0	Ruptured horn with pregnancy in communicating rudimentary hornin a hemiuterus.
CASE 3	26 years, P0A0, 19wks amenorrhoea presented to Emergency Room (ER) with pain abdomen and abdominal distension for 2 days. on peritoneal tap hemoperitoneum confirmed, departmental USG proved hemoperitoneum	Ruptured uterus	Resuscitation and emergency laparotomy performed for suspected ruptured ectopic.	After opening abdomen massive hemoperitoneum found with badly ruptured uterine left horn found with around 16-18 wks foetus protruding out of the ruptured horn. it was communicating with the hemiuterus which was normal. the horn resected out and stump repaired. she needed relaparotomy on 2^{nd} post-operative day as she had oozing from the stump. 11 units of PRBC, 6 Units of FFPand 4 units of platelet were neede to make the patient recovered. (Figure 4)	U4aC0V0	Ruptured horn with pregnancy in communicating rudimentary hornin a hemiuterus.

Continued on next page

Table 1 continued

Case 4	24 yrs, P0A1, delivered at 35 wks, vaginally uneventfully Retained placenta- severe PPH	Retained placenta	Emergency laparotomy as manual evacuation unsuccessful Ended up with removal of the uterine corpus with placenta 3 units PRBC transfusion HDU monitoring	Bicorporeal uterus- right uterine corpus enlarged with placenta in situ- resection of right corpus done- longitudinal cut section as in figure showed placenta acreta- left uterine corpus kept intact. (Figure 5)	U3bC0V0	Placenta accreta in a bicorporeal uterus.
Case 5	22 years, P0A1, 37 wks on Labor table with tachycardia, dehydration OS- 6 cm, Partograph- crossed alert line	Obstructed labor Foetal bradycardia	Emergency LSCS done on the pregnant uterine corpus. NICU admission of baby for 1 month	During LSCS, complete bicorporal uterus with septate cervix found (Figure 6)	U3aC1V0	Obstructed labour in a bicorporeal uterus due to cervical anomaly
Case 6	27 years, P0A1, 27 wks, PPROM, on Labor table with tachycardia, hypotension, dehydration Previous H/O of D&E - P/V:- OS- 5 cm, Partograph- poor progress of labour hemoperitonium (departmental USG confirmed the diagnosis)	Rupture uterus	Emergency laparotomy- ICU with ventilatory support needed	Hemoperitonium found. Bicorporeal uterus upto level of cervix found. One uterine corpus found to be ruptured with partial expulsion of foetus. Ruptured corpus 8 units PRBC transfusion was done. (Figure 7)	U3cC0V0	Ruptured corpus in a bicorporeal septate uterus.

Continued on next page

Table 1 continued

Case 7	22 years, P0A1, 36 wks presented to ER in Labour. On per vaginal examination it was found that she had longitudinal vaginal septum but head can be felt by fingers.	May have Mullerian anomaly.	Emergency LSCS due to Mullerian anomaly	After opening abdomen it was found to be a bicorporeal uterus with two cervix. baby delivered from the enlarged uterine corpus (Figure 8)	U3bC2V1	Obstructed labour in a bicorporeal uterus due to double cervix and longitudinal non obstructive vaginal septum.
Case 8	22 years, P0A1, 31wks presented to ER in shock. per vaginal examination there was longitudinal vaginal septum which was obstructing the foetal head.	obstructed labour and Suspected rupture uterus due to severe Mullerian anomaly.	Resuscitation and Emergency hysterectomy of the pregnant uterine corpus	After opening abdomen massive hemoperitoneum found with badly ruptured uterus found with stillborn baby in the pregnant corpus of the uterus as was found to be a bicorporeal uterus with two cervix. vaginal septum was longitudinal but obstructing type (Figure 9)	U3bC2V2	Obstructed labour leading to ruptured corpus of bicorporeal uterus due to double cervix and longitudinal obstructive vaginal septum.
Case 9	35 years, P0A3, 35 wks presented with severe APH and IUFD. Previous H/O Recurrent Pregnancy Loss due to bicorporeal uterus for which laparoscopic metroploasty was done-She had placenta previa in present pregnancy.	Complete placenta previa with APH with haemorrhagic shock	Initial resuscitation followed emergency LSCS	LSCS done with delivery of a 2.4 kg stillborn baby. HDU admission was needed. 6 units of PRBC, 4 units FFP, 2 units of platelet were transfused. (figure absent due to non-capture)	U3bC0V0	Placenta previa in a case of metroplasty done on a bicorporeal uterus.

Continued on next page

Table 1 continued

Case 10	24 years, P0A3 presented with severe PPH on day 10 post-partum H/O uneventful vaginal delivery at 35 wks at our hospital. USG revealed retained placental bits with hematometra- failed attempted repeated ERPOC.	Secondary PPH after VD on day 10 postpartum-	Laparotomy done with consent of hysterectomy Hysterotomy done Septum resection done with removal of retained bits of placental tissue	A complete septum found, which was obstructing during ERPOC Retained placental bits removed. (Figure 10)	U2bC0V0	Complete septate uterus leading to failed ERPOC and Secondary PPH.
Case 11	Newly married, P0A0, 19 years with incomplete abortion at 8 wks. D& E by resident. On 2^{nd} post-operative day, patient had pain and abdominal distension. USG revealed haemoperitoneum and suspseptate uterus .	Uterine perforation during D&E	Emergency laparoscopy and proceed	laparoscopy guided ERPOC and repair of rent by intracorporeal suturing hemostasis secured. (Figure 11) Iatrogenic perforation	U2bC0V0	Complete septate uterus leading to iatrogenic perforation.
Case 12	28 years, P1A0L1, came for removal of PPIUCD(post-partum IUCD) after 12 months of Preterm(34 wks)vaginal delivery for missing thread of the IUCD and Abnormal uterine bleeding. Underwent D & C procedure but even after repeated try, IUCD couldn't be removed.	After a failed IUCD removal, in immediate postoperative period abdominal distension & USG revealed hemoperitoneum along with bicorporeal uterus with IUCD in left horn. provisional diagnosis – Iatrogenic perforation.	Immediate laparotomy	CU-T removed by hysterotomy. During laparotomy it was found to be a case of bicorporeal uterus. IUCD found in left corpus a which was removed and repaired. (figure- absent)	U3bC0V0	Complete bicorporeal uterus leading to iatrogenic perforation during IUCD removal.

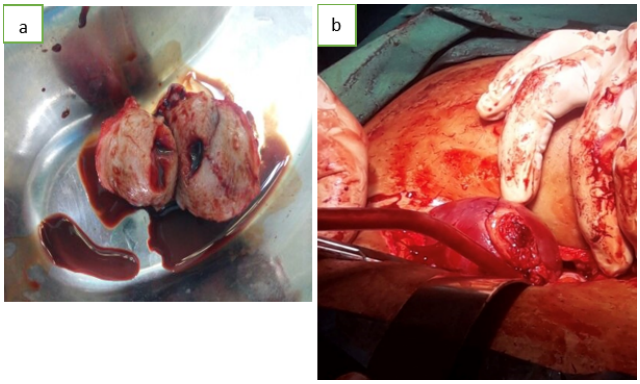

Figure 2: (**a,b**) Cut section of resected out right rudimentary hemiuterus which was non-communicating and menstrual blood seen in it mimicking chronic ectopic; (**a**). Left hemiuterus kept intact (**b**)

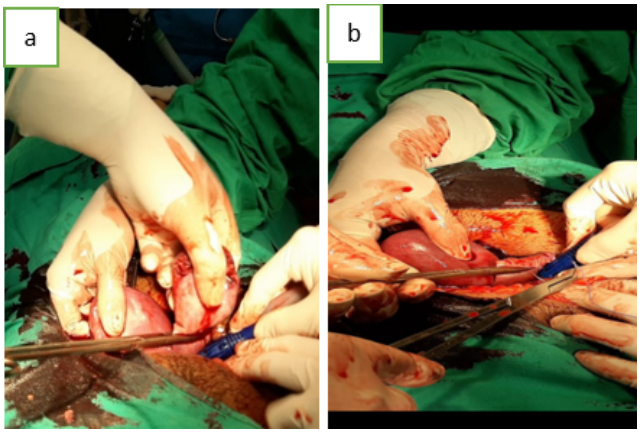

Figure 3: (**a,b**) pregnancy in the communicating rudimentary horn which had ruptured,clamped to cut (**a**) The resected portion being sutured(**b**)

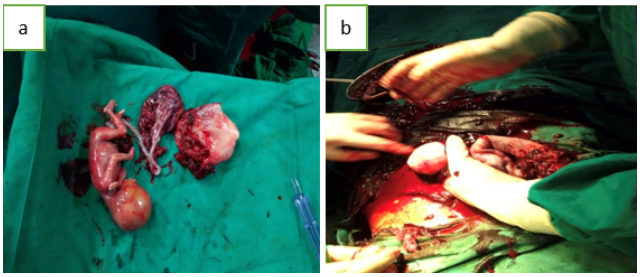

Figure 4: a,b Expulsed foetus; (**a**): Ruptured right uterine corpus communicating with the left hemiuterus (**b**)

3. Results

Out of the total sample of 12 cases, no cases of ESHRE- U1(dysmorphic uterus) , U5 (aplastc uterus) and

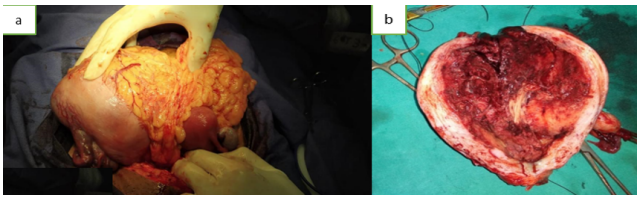

Figure 5: a,b: Bicorporeal uterus(a).cut opened right uterine corpus showing placenta increta

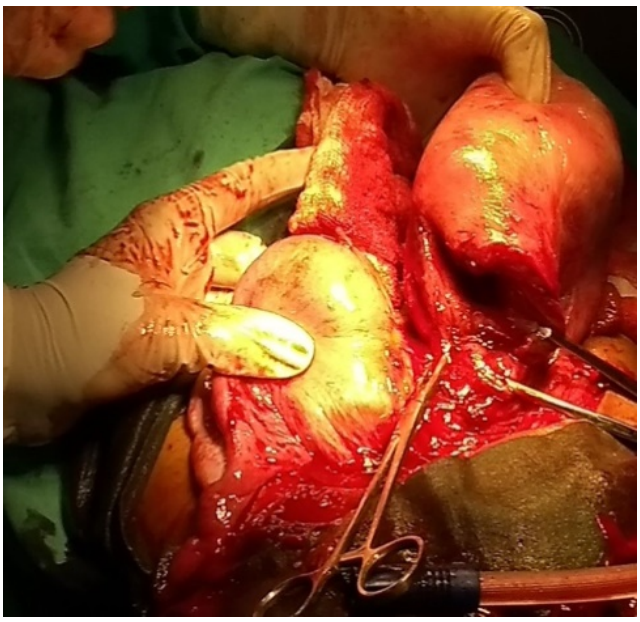

Figure 6: (a,b) : During LSCS, complete bicorporal uterus with septate cervix found

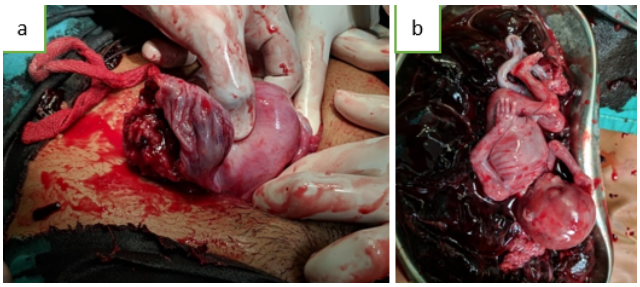

Figure 7: (a,b) Ruptured left uterine corpus (**a**): Expulsed foetus from the ruptured corpus (**b**)

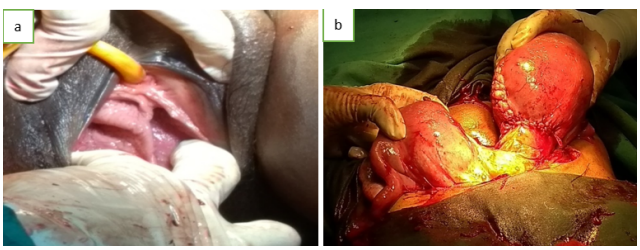

Figure 8: (a,b) Fingers introduced intwo vagina cervix; (**a**): LSCS wound repaired in right corpus(b)

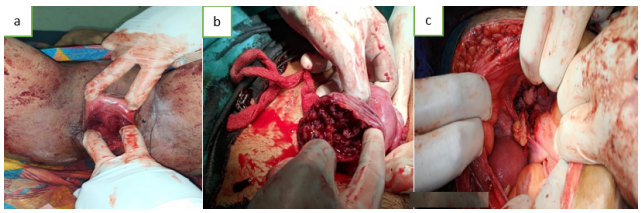

Figure 9: (a,b,c) Vaginal septum with two cervix; (**a**): **R**uptured corpus of a bicorporeal uterus; (**b**): Two corpus separated by a longitudinal omental tag , right corpus ruptured but left corpus intact (**c**)

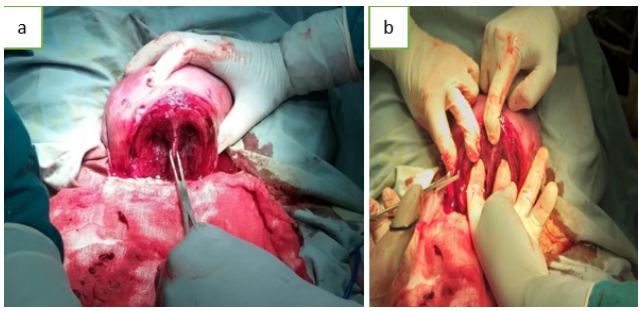

Figure 10: (a,b) Complete septum

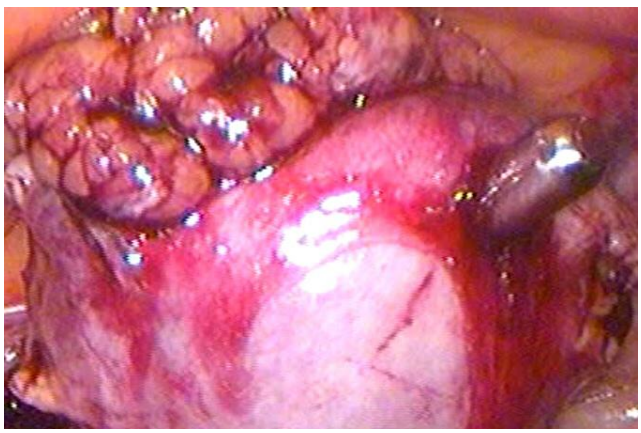

Figure 11: (a) laparoscopic view of uterine perforation done during ERPOC

U6 (otherwise unclassified) contributed in a complicated pregnancy outcome. Among 12 women with uterine anomalies 4 abortions, 7 delivered preterm, 1 term delivery occurred.

Majority of complications associated with U3 types (58.3%). No cases in U1 and U6 found to cause life threatening emergencies. Majority of cases (50%) continued pregnancy in between 32 - 36+6 wks pregnancy. Only one case of U3a continued pregnancy till term. Major cases of type 3 (6 out of 7) continued pregnancy beyond 32 wks. Other types with complications are U4 (25%) and U2 (16.7%). Presenting features at emergency varied like mimicking ectopic, labour complications, ruptured corpus, placenta previa and accreta as well as iatrogenic perforation. 4 out of 12 cases presented with abortion. The major contributor to cause acute life-threatening conditions, in this study, is the U3 which is the bicorporeal type of uterus (58.3%).

4. Discussion

Mullerian anomalies in reproductive age group has significant adverse effects on pregnancy outcomes from both aspect of maternal and foetal outcome. In this study we have focussed on the life threatening emergency maternal and foetal conditions which were due to congenital reproductive anomalies. The ESHRE classification of Mullerian anomalies are used to categorise different genital anomalies as it is simple and caters a variety of anomalies.[15] After the introduction of ESHRE Classification in 2013 it has been less utilized to determine the effectiveness in calculating the fertility outcome as well as pregnancy outcome. Though it seems ESHRE Classification is comparable tool to measure fertility outcome in respect to the ASRM Classification but it is more effective in measuring the life threatening complications in respect to maternal & foetal outcome. It may be due to the cervical and vaginal sub-group classification along with the main classification makes it easier to define the adverse outcome of pregnancy.

Out of the total sample of 12 cases, no cases of ESHRE U1(dysmorphic uterus), U5 (aplastc uterus) and U6 (otherwise unclassified) contributed in a complicated pregnancy outcome. It may be due to dysmorphic uterus (U1) doesn't affect much in implantation, stretches adequately with the growing foetus and maintains its contractility. On the other hand, uterus with U5 & U6 anomalies are mostly not suitable to hold pregnancy.

4 out of 12 cases presented with abortion. It is quite obvious that mullerian anomalies present with abortion which is 25% in our observation. 3 out of 4 cases are from you U4a group and one case from U2b group. In our study none of hemiuterus with or without communicating horn took pregnancy beyond 20 weeks. One case of U3c continued pregnancy till 27 wks. Majority of cases (50%) continued pregnancy in between 32 - 36+6 wks pregnancy.

Only one case of 3a continued pregnancy till term. Major cases of type 3 (6 out of 7) continued pregnancy beyond 32 wks. So if a mullerian anomaly is properly diagnosed before pregnancy and followed up during pregnancy then viable fetal outcome may raise from present scenario. But life threatening complications always should be kept in mind and closed supervision with early intervention may significantly reduce maternal as well fetal morbidity and mortality.

A total of 2 cases were found in the group U2 (i.e. septate uterus) which lead to obstetric complications. Both

Table 2: Distribution of cases of life saving emergency according to ESHRE Classification (n = 12)

Main Class	Number (n=12)	Percentage (%)	Subclass	Number	Percentage
U1 (Dysmorphic Uterus)	0	0	U1a	0	0%
			U1b	0	0%
			U1c	0	0%
U2 (Septate Uterus)	2	16.7	U2a	0	0%
			U2b	2	16.7%
U3 (Bicorporeal Uterus)	7	58.3	U3a	1	8.3%
			U3b	5	41.6%
			U3c	1	8.3%
U4 (Hemi Uterus)	3	25	U4a	3	25%
			U4b	0	0%
U6 (Unclassified)	0	0	U6a	0	0%
			U6b	0	0%

Table 3: Gestatinal age of **t**presentation of complications due to mullerian anomalies according to ESHRE Main Class

	Total number of cases(n=12)	U2 cases (n=2)		U3 cases (n=7)			U4 cases (n=3)	
		2a	2b	3a	3b	3c	4a	4b
<20 wks	4	0	1(8.3%)	0	0	0	3 (25%)	0
20-27^{+6} wks	1	0	0	0	0	1(8.3%)	0	0
28-31^{+6} wks	0	0	0	0	0	0	0	0
32-36^{+6}wks	6	0	1(8.3%)	0	4 (57%)	1(8.3%)	0	0
37 wks	1	0	0	1(8.3%)	0	0	0	0

Table 4: Different mullerian anomaly and presentations & complications

Presenting complication	U2		U3			U4	
	U2a	U2b	U3a	U3b	U3c	U4a	U4b
Mimicks ectopic	0	0	0	0	0	1(8.33%)	0
Ruptured ectopic (horn)	0	0	0	0	0	2(16.6%)	0
Labour complication	0	0	1	1	0	0	0
Ruptured uterine corpus	0	0	0	1	1	0	0
Retained placenta & Retained product of conception	0	1	0	0	0	0	0
Placental complications (placenta previa, acreta etc)	0	0	0	3	0	0	0
Secondary PPH	0	0	0	0	0	0	0
Iatrogenic uterine perforation	0	1	0	0	0	0	0

Table 5:

Presenting complication	U2		U3		U4	
Mimicks ectopic	0	0%	0	0%	1	8.33%
Ruptured horn (ECTOPIC LIKE)	0	0%	0	0%	2	16.6%
Labour complication	0	0%	2	16.6%	0	0%
Ruptured uterine corpus	0	0%	2	16.6%	0	0%
Retained placenta & Retained product of conception	1	8.33%	0	0%	0	0%
Placental complications (placenta previa, acreta etc)	0	0%	2	16.6%	0	0%
Secondary PPH	1	8.33%	0	0%	0	0%
Iatrogenic uterine perforation	1	8.33%	0	0%	0	0%

were in the sub-group U2b which is complete septate uterus, but no cases of U2a or partial septate were found to cause any adverse effect on pregnancy. 1 case in U2 had been complicated by vaginal procedures like D/E and the other case was complicated due to secondary PPH after a normal vaginal delivery due to retained placental bits. It is evident that vaginal procedures in patients with U2 mullerian anomaly is unpredictable due to its distorted internal anatomy and inadequate diagnosis by radiologists prior to the procedure.

The major contributor to cause acute life-threatening conditions, in this study, is the U3 which is the bicorporeal type of uterus (58.3%). According to ESHRE classification it has three sub-types. The U3a or the partial type contributed in one case, which was a full term pregnancy, landed up in obstructed labor and delivered by emergency LSCS. This patient had C1 anomaly i.e. septate cervix, which may be a contributing factor against her obstruction. There was 2 patient found in the group U3C which is bicorporeal septate type. In this U3c type, there is complete septation till the internal os with almost normal musculature, represents it as two uterus in other term. One case presented with ruptured corpus at 27 wks and other case presented with iatrogenic perforation during removal of CU T 12 months after preterm delivery. It is also evident that removal of CU T without prior USG might lead to a life threatening complication. Type U3b or the complete bicorporeal uterus has contributed maximum in U3. Due to poor stretching effect of any of the cornus or improper implantation has caused rupture of the corpus. Those cases of U3b which were associated with cervical and vaginal anomalies led to obstetric emergencies mainly. Improper placentation also caused placenta previa and retained placenta.

Type U4 or hemi uterus or unicornuate uterus contributed to 3 cases out of total 12 cases which led to complications. Patients with these type uterus mainly suffer for the rudimentary horn. Rest of the cases where rudimentary horn is absent(unicornuate only) can reach upto term delivery as shown by ludmir et al where 60% of cases of unicornuate and didelphys reached upto term.[16] Rudimentary horn is a type of hypoplastic uterine part that has lesser musculature and stretching ability. If it is a connecting type (U4a) then sometimes implantation may occur within rudimentary horn causing rupture of the horn which may be termed as pseudo-ectopic pregnancy. Sometimes in a non-communicating type of hemiuterus i.e. type U4a, mimics ectopic pregnancy, by shedding of the functioning endometrium in rudimentary horn and concealed menstrual bleeding may arise suspicion of ectopic pregnancy. Though dysmenorrhea will be the most common presenting feature but few a time it may mimics ectopic.

Type U5 or aplastic uterus is not suitable for viable pregnancy. Uterine transplantation or surrogacy are options.

5. Conclusion

Congenital Uterine anomalies are common but their effects on reproductive outcome is unclear. Many studies have elucidated the effect of uterine anomalies on fertility. But the effect of these anomalies on later part of pregnancy has been less studied. From present study it is seen that U3 main class of the ESHRE classification is mostly associated with dreaded complications both obstetric and gynaecological aspect like Placenta Acreta, ruptured corpus, obstructed labour. U4 main class leads to life threatening complications like ruptured horn as well as pseudo ectopic pregnancy. U2 main class is relatively associated with miscarriage and retained product. Though we were able to save all the cases bit it needed lots of skill and expertise. Hence we can conclude that mullerian anomalies often leads to grave emergency situation specially in ruptured corpus and ruptured horn cases as because these cases are mostly under diagnosed or undiagnosed before the emergency arises. A good pull of suspicion is to be kept in mind to save mother as well as fetus. Though ESHRE classification is little bit complicated to remember but it's composite structure is very much helpful to diagnose and manage these rare cases.

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7. Conflict of Interest

None.

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