

The case number 506 of Mostyn Embrey Syndrome: Imaging Studies and Ethics of Naming Syndromes

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Abstract

Background: Mostyn Embrey syndrome is a rare disorder associated with unilateral renal agenesis and malformations of the female reproductive tract. Delayed diagnosis is associated with serious diagnostic difficulties that may lead to inappropriate management including harmful surgery. The aim of this paper is to present imaging studies of case number 506 of the syndrome which was the first case of this rare syndrome in Iraq, and was originally reported in 2016. Ethics of naming syndromes relevant to this syndrome is discussed.

Patients and methods: An 18-year-old female presented with acute abdominal pain and poor urine output associated with retention of urine; about three years after the onset of menses. The pain was radiating to the back and pelvis. Her menstrual cycles started at the age of fifteen, and were lasting seven days. Cycles were associated with normal flow. The first physician the patient consulted considered the diagnosis of uterine tumor and the need for hysterectomy, but the family of the patient consulted another physician. Vaginal exam was performed and showed bulging left vaginal wall. The diagnosis of hematocolpos was made. A clotted blood was drained by trans- vaginal approach.

Results: It was difficult to identify the two separate uteri and two separate cervixes and to make a diagnosis of didelphys uterus on hysterosalpingography. Abdominal MRI showed left renal agenesis, and two uterine bodies, cervixes, and vaginas with normal myometrium and endometrium (uterine didelphys), and normal ovaries.

Conclusions: It is recommended that pediatricians, and pediatric nephrologists need to be aware of this syndrome to avoid unexpected presentations which may lead to delayed diagnosis and sometimes to inappropriate management. The imaging modalities that can be used to diagnose Mostyn Embrey syndrome include ultrasonography, hysterosalpingography and MRI. In this case, hysterosalpingography was not of much help. Ultrasound is affordable, non-invasive, widely available imaging modality that contributes to the accurate diagnosis of Mostyn Embrey syndrome. However, there can be difficulties in visualizing the vaginal septum on ultrasound which best shown on MRI. MRI can facilitate early diagnosis and thus help in the prevention of further complications. Syndromes in medicine are often named after the physician or group of physicians that discovered them or initially provided the full clinical picture or the best description of the syndrome. Unfortunately, Mostyn Embrey syndromes has been attributed unfairly and inappropriately to physicians other than those first described them in almost all previous papers.

Keywords: mostyn embrey syndromes; Iraq; imaging studies; hysterosalpingography; ultrasound; MRI

Introduction

Mostyn Embrey syndrome is a very rare syndrome essentially consisting of the triad of:

1-Duplication abnormalities of the female reproductive tract (uterus, cervix, and vagina). Uterine abnormalities commonly didelphys and bicornuate uterus or unicornuate.

Duplication malformations of uterus and vagina are caused either by absence or incomplete fusion of the paired Müller's ducts. Uterus didelphys which is the most common uterine abnormalities associated

with this syndrome. Uterus didelphys is associated with total duplication of the uterus with two independent horns, two cervixes, and a double vagina.

The classic Mostyn Embrey syndrome is associated with uterus didelphys. Patients who have uterine abnormalities other than didelphys uterus are generally considered to have a variant of the syndrome.

2-Hemi-vaginal obstruction resulting in unilateral hematocolpos and usually resulting in some degree of hematometra and hemato-salpinx

caused by the back flow of retained menses in the site of the hematocolpos. However, variants of the classic syndrome may be associated with cervical obstruction or atresia.

3-Ipsilateral renal agenesis. Atypical variants can be associated with contra-lateral renal agenesis or other renal anomalies. The uterus, fallopian tubes, cervix and upper two thirds of the vagina develop from the paired müllerian ducts while the lower third of the vagina develops separately from the urogenital sinus.

Duplication of the female reproductive tract results from a lack of fusion of the paired müllerian ducts which appear at the sixth week of embryonic development. The müllerian ducts develop just lateral to the mesonephric (urinary) ducts, which have been hypothesized to act as a guide to the growth of the müllerian ducts. As they move caudally, the müllerian ducts cross over the mesonephric ducts ventrally and fusion of the lowermost portion occurs. The cranial non-fused portions form the paired fallopian tubes; the caudal fused ducts form the uterus, cervix, and the upper three-quarters of the vagina. The distal vagina is formed by invagination of the urogenital sinus whose endoderm then replaces the entire original vaginal epithelium. The association of duplication abnormalities of the uterus with obstructed hemi-vagina and renal agenesis ipsilateral to the side of obstruction was explained by embryologic arrest at the eighth week of gestation which simultaneously affects the müllerian and metanephric ducts. Other renal anomalies may also be associated particularly renal dysplasia, double collecting system and ectopic ureter [1, 2, 3, 4].

The aim of this paper is to present imaging studies of case number 506 of the syndrome which was the first case of this rare syndrome in Iraq, and was originally reported in 2016 [2]. Ethics of naming syndromes relevant to this syndrome is discussed.

Patients and Methods

Thirty four year old female who was born in 1981 presented initially at the age of 18 years (January, 31, 1999), about three years after the onset of menses with acute abdominal pain and poor urine output associated

with retention of urine. The pain was radiating to the back and pelvis.

The first gynecologist the patient consulted considered the diagnosis of uterine tumor and the need for hysterectomy, but the family of the patient consulted another gynecologist. Later, vaginal exam was performed and showed bulging left vaginal wall. The diagnosis of hematocolpos was made. A clotted blood was drained by trans-vaginal approach.

Results

It was difficult to identify the two separate uteri and two separate cervixes and to make a diagnosis of didelphys uterus on hysterosalpingography (Figure-1).

As early as 1977, Brezina emphasized the possibility of not diagnosing Mostyn Embrey syndrome for many years after menarche even by hysterosalpingography. Abdominal MRI showed double uterus and left renal agenesis. MRI (Figure-2) performed at the 25th of September, 2005 showed two uterine bodies, cervixes, and vaginas with normal myometrium and endometrium (uterine didelphys), and normal ovaries.

Her menstrual cycles started at the age of fifteen, and were lasting seven days. Cycles were associated with normal flow. However, she was frequently experiencing dysmenorrhea. During school she was less tolerant to sport activities and was frequently developing palpitation, shortness of breath and hypotensive episodes. Echocardiography showed mild mitral prolapse. However, her performance in school was good and she graduated from the college of dentistry at the age of 23 years during the year 2004.

She had also myopia (-3) and also dental abnormalities including spacing of teeth, and absence of the upper third molars, and malposition of the lower third molars. She developed dry socket infections of the lower third molars and their positions were corrected thereafter. During the years 2009 and 2010 she was complaining frequently from generalized aches and joint pain. Radiographs taken during the 17th of October showed minimal degenerative changes at the knees and cervical spine radiographs showed spondylotic changes at multiple disc space.



Figure 1A: *It was difficult to identify the two separate uteri and two separate cervixes and to make a diagnosis of didelphys uterus on hysterosalpingography*



Figure 1B: *It was difficult to identify the two separate uteri and two separate cervixes and to make a diagnosis of didelphys uterus on hysterosalpingography*



Figure 1C: *It was difficult to identify the two separate uteri and two separate cervixes and to make a diagnosis of didelphys uterus on hysterosalpingography*

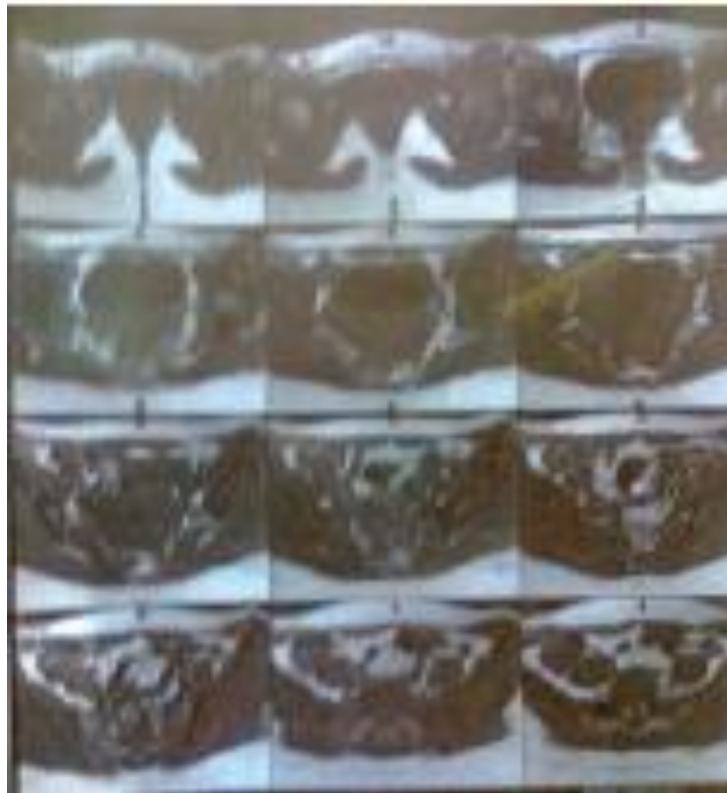


Figure 2A: MRI showed two uterine bodies, cervices, and vaginas with normal myometrium and endometrium (uterine didelphys), and normal ovaries

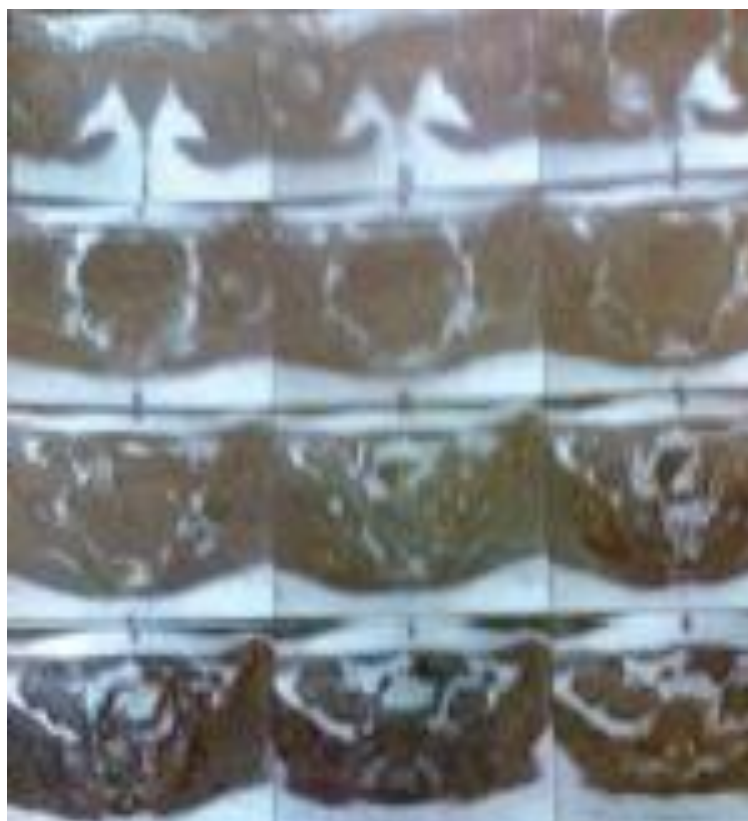


Figure 2B: MRI showed two uterine bodies, cervices, and vaginas with normal myometrium and endometrium (uterine didelphys), and normal ovaries



Figure 2C: MRI showed two uterine bodies, cervices, and vaginas with normal myometrium and endometrium (uterine didelphys), and normal ovaries



Figure 2D: MRI showed two uterine bodies, cervices, and vaginas with normal myometrium and endometrium (uterine didelphys), and normal ovaries



Figure 2E: MRI showed two uterine bodies, cervixes, and vaginas with normal myometrium and endometrium (uterine didelphys), and normal ovaries



Figure 2F: MRI showed two uterine bodies, cervixes, and vaginas with normal myometrium and endometrium (uterine didelphys), and normal ovaries



Figure 4A: The patient was evaluated twice with ultrasound early during the year 2010 more than two years before the second pregnancy



Figure 4B: The patient was evaluated twice with ultrasound early during the year 2010 more than two years before the second pregnancy



Figure 5A: An ultrasound image taken at the 27th of February, 2010

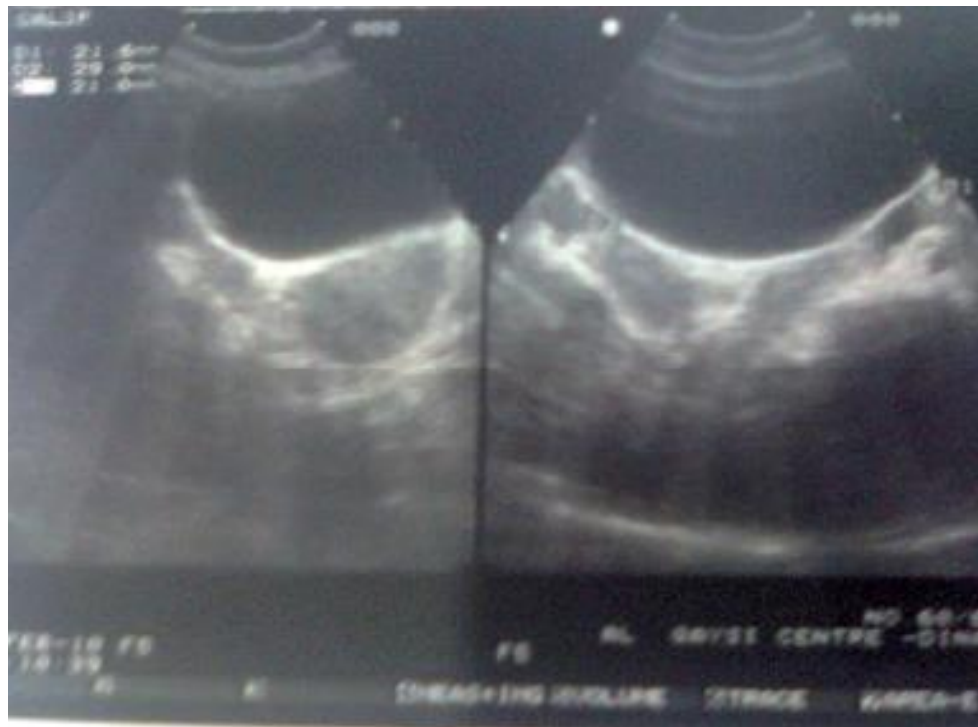


Figure 5B: An ultrasound image taken at the 27th of February, 2010

daughter was considered normal and imaging studies excluded Mostyn Embrey syndrome.

Her parents were unrelated. She has two brothers; one brother has two healthy male children and the other brother has a daughter with

cricopharyngeal achalasia. Her mother and two brothers had history of renal stone. Her cousin who is four years older was known to have bicornuate uterus and single kidney, but she didn't have hematocolpos, and she gave birth to two males. Table-1 summarizes the features and management of the first case of Mostyn Embrey syndrome.

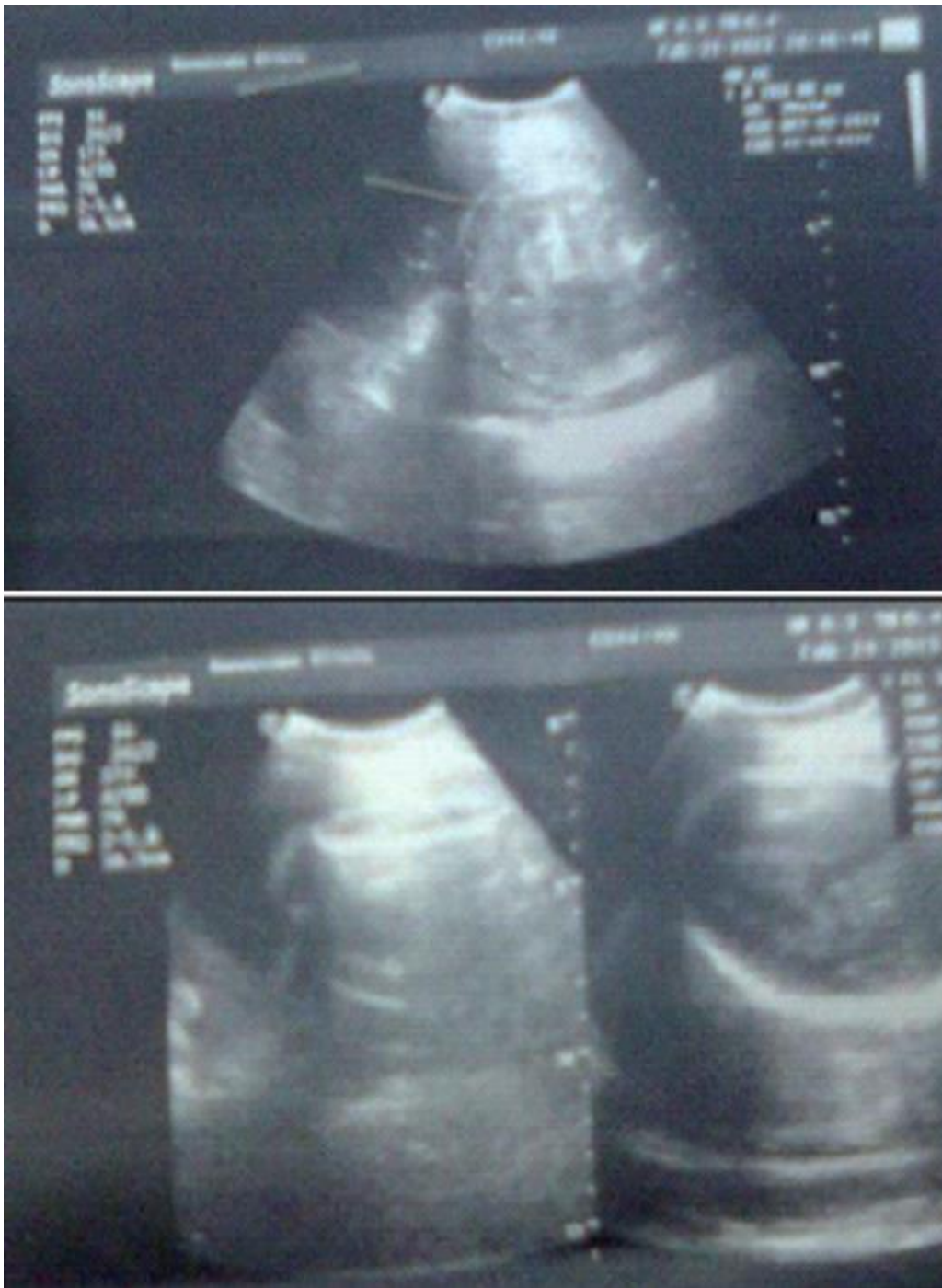


Figure 6A: Ultrasound at the 6th of September 2012 showed bulky gravid bicornuate uterus with well-defined gestational sac with in the right horn (19 X 13 mm). A tiny fetal pole was seen within the yolk sac



Figure 6B: Ultrasound at the 6th of September 2012 showed bulky gravid bicornuate uterus with well-defined gestational sac with in the right horn (19 X 13 mm). A tiny fetal pole was seen within the yolk sac

Age presentation	18 years , unmarried
Presentation	Acute abdominal pain and poor urine output associated with retention of urine about three years after the onset of menses. The pain was radiating to the back and pelvis
Family history	Her cousin who is four years older was known to have bicornuate uterus and single kidney, but she didn't have hematocolpos, and she gave birth to two males. The daughter of the patient didn't have Mostyn Embrey syndrome.
General exam	Appeared well, normal development
Abdominal exam	A mass originating in the pelvis
Vaginal exam	Bulging left vaginal wall
MRI findings	Double uterus and left renal agenesis: two uterine bodies, cervixes, and vaginas with normal myometrium and endometrium (uterine didelphys), and normal ovaries
Early management	Clotted blood was drained by trans-vaginal approach
Other abnormalities	1-Cardiovascular abnormalities: Poor tolerant to school sport, palpitation, shortness of breath and hypotensive episodes. Echocardiography: mild mitral prolapse. 2-Myopia (-3).

	3-Dental abnormalities: spacing of teeth, and absence of the upper third molars, and malposition of the lower third molars. She developed dry socket infections of the lower third molars. 4-Minimal degenerative changes at the knees and spondylotic changes at multiple disc space in the cervical spine.
Obstetric history	Became pregnant twice, one pregnancy ended with

Table-1: *the features and management of the first case of Mostyn Embrey syndrome in Iraq*

Discussion

The rare association of complete duplication of the genital tract-uterus didelphys; complicated by unilateral atresia with menstrual retention and a hematocolpo-metrosalpinx was probably first reported in the literature during 1922. Hadden [5] and Purslow [6] were probably the first to report this rare association. Purslow in 1922 reported a 16-year-old girl who had double uterus with right hematocolpos, hematometra and hematosalpinx [6].

Purslow treated patient by removal of the affected uterus and appendages (right hemi-hysterectomy) and vaginal septum incision with vaginal drainage [6].

Wilson reported in 1925 the treatment of a 14 years old girl who had double uterus and vagina with unilateral hematocolpos and hematometra who presented with pelvic mass and treated by total hysterectomy and salpingoophorectomy [7].

Brown and Brews reviewed 50 cases of congenital retention of the menses including a case of a 16 years old girl who had an abdominal mass. Laparotomy and excision of the hymen were performed, but the patient died eight days postoperatively because of peritonitis [8].

Simon HE [Surg Gynec Obstet 1928; 47:356] reviewed 23 cases of hematometra including a patient with an incomplete uterus didelphys." He also quoted Quenu and Le Sourd (1926) as reviewing eight cases of uterus didelphys with hematometra [4].

Masson and Mueller in 1933 reported a patient with uterus duplex bicornis with septate vagina open only on one side associated with right hematocolpos; they performed laparotomy and excision of the vaginal septum [9]. During the same year Carnington and Burlington reported a patient who had incomplete bipartite uterus with left hematocolpos and salpingitis. They treated the patient by partial hysterectomy and salpingoophorectomy [10]. In 1935, Martindale reported a case of uterus didelphys associated with hematocolpos of the right vagina and simulating appendicitis. The patient was treated by excision of the vaginal septum [11].

In 1950, Mostyn P. Embrey provided the first excellent description of the occurrence of uterus didelphys, hemi-vaginal obstruction in association with ipsilateral renal agenesis in an unmarried 17-year old girl referred to hospital because of abdominal swelling (Figure-7). Mostyn P. Embrey (B.Sc., M.D. F.R.C.S., and M.R.C.O.G.) was a lecturer in obstetrics at the University of Bristol and Assistant Obstetrician, Bristol Royal Hospital [1, 2, 3, 4].

It was not clear whether the earlier seven cases of the association of uterine duplication abnormalities commonly didelphys or bicornuate uterus with hemi-vaginal obstruction resulting in unilateral hematocolpos or hematocolpo-metrosalpinx reported during the period from 1922 to 1935 were associated with renal agenesis or not [Hadden (1922), Purslow (1922), Wilson (1925), Brown and Brews (1930), Masson and Mueller (1933), Carnington and Burlington (1933), and Martindale (1935)].

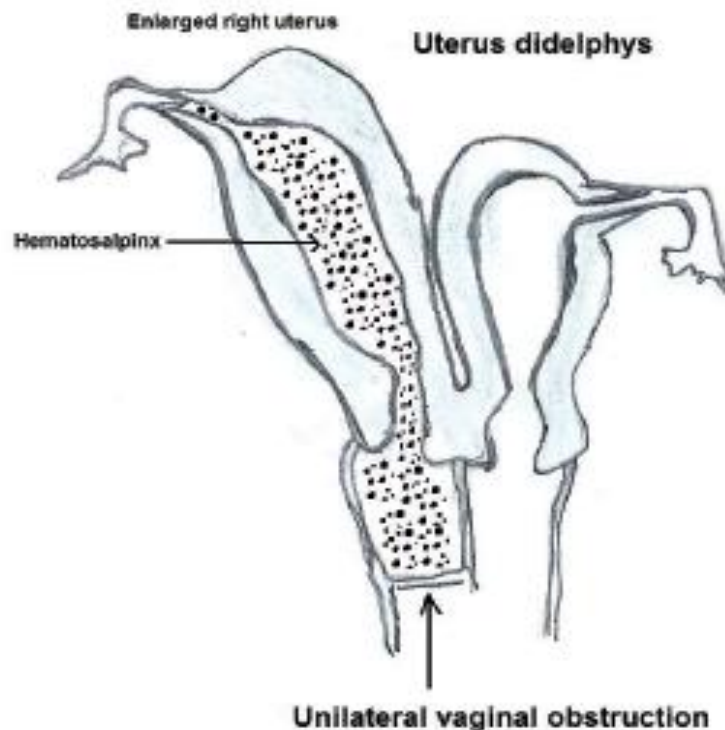


Figure 7: *The anomalies in the reproductive system in Mostyn Embrey syndrome*

The patient described by Embrey was only complaining of indefinite back pain of about one year and she looked to be well and normally developed.

Abdominal examination showed a cystic regular swelling originating in the pelvis and reaching the umbilicus more to the right of the midline. Rectal examination revealed a mass which was palpable anteriorly largely filling the pelvis.

When the patient was examined digitally under anesthesia, the vulva was normal with intact hymen, but the abdominal mass was bulging down the right anterior vaginal wall and displacing the vagina to the left and posteriorly.

Laparotomy revealed a large rounded smooth mass, mostly extra-peritoneal and it was filling most of the pelvic cavity reaching the umbilicus and presenting in the laparotomy wound. The mass was also causing distention of the vagina to a large extent. On the top of the mass there was a uterus on each side. The left uterus was of average size and the left appendages were normal. The right uterus was softer and enlarged to one and half the size of the left uterus and the right tube was 3 times larger because of hematosalpinx.

Right salpingectomy was performed and the right ovary was conserved. Incision of the swelling bulging from the vagina was made and about one liter of blood was drained. Right renal agenesis was confirmed by excretion pyelography which showed only the left renal tract. There was no right uretric orifice on cystoscopy with indigo-carmin excretion study.

After six months, exam under anesthesia showed that the left uterus and

the right uterus were both of normal and similar size. The right vagina opened at the lower end of the left. A second laparotomy showed two separate uteri with normal left tube and ovaries. The absence of the right ureter and kidney was also confirmed. Right hemi-hysterectomy was performed during the second laparotomy. Excision of lower most end of the right vagina was performed from the perineum to eliminate the cavity. Convalescence was uneventful [1, 2, 3, 4]. Table-2 summarizes the features and management of the first case of Mostyn Embrey syndrome.

During the period from 1950 to 1970, sixteen cases were described in English literature including the first case of Mostyn P. Embrey [1]. Most of the cases were classical Mostyn Embrey syndrome: Woolf and Allen (1953) [four cases], Semmens (1956), Gibberd (1957), Hill (1958), Allan and Cowan (1963) [Three cases], Thompson and Lynn (1966) [Two cases], Lewis and Brent (1966) [2, 3, 4].

In 1971, Herlyn and Werner reported the 17th case of Mostyn Embrey syndrome and the first case in German literature. Unfortunately, they missed the earlier sixteen cases reported in English literature and considered their case inappropriately as a new typical syndrome of anomalies [1, 2, 3, 4].

A total of 41 cases of Mostyn Embrey syndrome were reported by the end of the 1970s. Thirty three cases had the classical syndrome with didelphys uterus and eight cases had atypical variant associated with bicornuate uterus in seven and unicornuate uterus in one patient.

Thirty six cases were reported during the 1980s, in addition to the atypical case reported by Burbige and Hensle who had severe renal dysplasia rather than renal agenesis [2, 4].

Age & marital status	17 years , unmarried
History	Indefinite back pain of about one year duration
General exam	Appeared well, normal development
Abdominal exam	Cystic regular swelling originating in the pelvis and reaching the umbilicus more to the right of the midline
Rectal exam	Palpable mass anteriorly, largely filling the pelvis
Vaginal exam under anesthesia	A swelling bulging down the right anterior vaginal wall and displacing the vagina to the left and posteriorly
Laparotomy finding	Normal left uterus and left appendages. Enlarged right uterus and right tube
Excretion pyelography	Absence of the right renal tract
Cystoscopy with indigo-carmin excretion study	Absence of right uretric orifice
Early management	Right salpingectomy Incision of the hymen and drainage
FOLLOW-UP	AFTER SIX MONTHS
Vaginal exam under anesthesia	Left uterus and the right uterus were both of normal and similar size
Laparotomy	Two separate uteri with normal left tube and ovaries Absence of the right ureter and kidney
Later management	Right hemi-hysterectomy was performed during the second laparotomy. Excision of lower most end of the right vagina was performed from the perineum to eliminate the cavity

Table -2: The features and management of the first case of Mostyn Embrey syndrome

By the end of 1999, Eighty seven cases were reported including the atypical variant associated with hypoplastic cervix reported by Lee et al. The total cases of Mostyn Embrey syndrome reached 164. The 1990s witnessed the report of large series; from USA (15 patients) and from Italy (36 patients). During the 2000s, 225 cases with Mostyn Embrey syndrome were reported (The atypical case of Altchek and colleagues report was not

included). There were reports from Israel, Finland, Tunisia, Italy, India, and USA. The total cases reported by the end of 2010 were 290 cases. After 2010, 171 cases with Mostyn Embrey syndrome were reported. The atypical cases including Garge et al variants and the patients with incomplete syndrome reported by Dorais et al. we're not included in this count. The total cases reported after 2010 were 560 cases [2, 4].

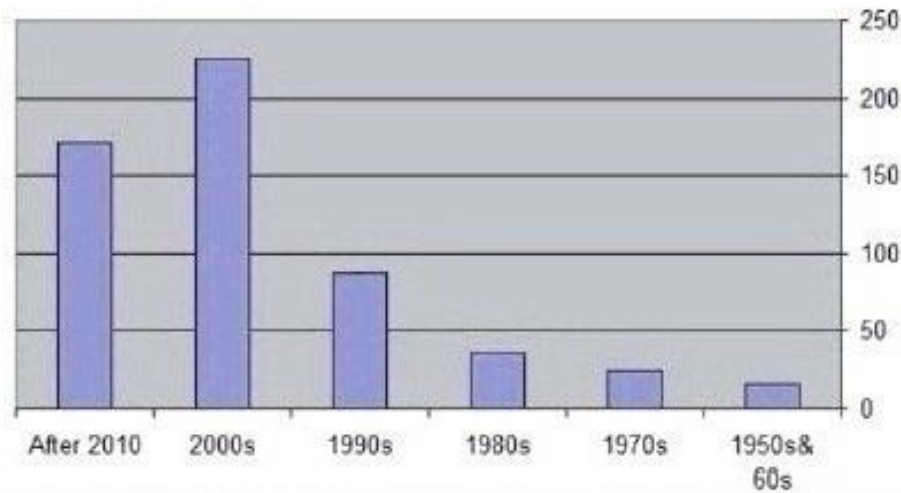


Figure 8: The number of cases reported during the previous 100 years

The vast majority of cases of Mostyn Embrey syndrome have been reported in English literature. However, some cases have been reported in other languages including eight in German, five in Bulgarian, three in French, three cases in Spanish, one in Danish, one in Dutch, and one case published in both English and Turkish.

It was difficult and not convenient to determine the geographic region of many reports. However, there were 139 cases from Italy, 80 from china, 41 cases from the USA, 31 cases from Turkey, 13 from Canada, 9 from Japan, 7 from Taiwan, 7 from India, 6 from France, 5 from Bulgaria. 5

from Tunisia, 4 from UK (Many of the cases reported during the 1950s and 1960s are expected to be from UK), 4 from Australia, 4 from Finland, 4 from Israel, 2 from Spain, 2 from Singapore. There were also cases reported from Korea, Denmark, Poland, Portugal, Austria, Thailand, Netherlands, and Oman [2, 4]. Figure-9 shows the geographic distribution of cases of Mostyn Embrey syndrome reported during the previous 100 years.

The first case of Mostyn Embrey syndrome is Iraq was the case number 561, and the second in Ethnic Arab [2].

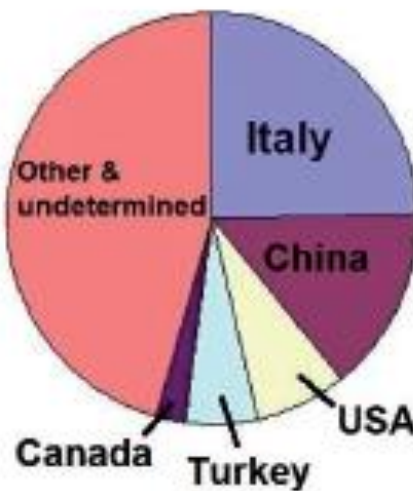


Figure 9: The geographic distribution of cases of Mostyn Embrey syndrome reported during the previous 100 years

Atypical variant of the syndrome associated with bicornuate uterus rather than didelphys uterus were reported by Thompson and Lynn (1966) and Burton (1968). Although it was expected that many of the cases reported before 1970 were from the United Kingdom [2, 3, 4].

Non classical forms of Mostyn Embrey syndrome associated with bicornuate uterus rather than didelphys uterus have already been described before Wunderlich (1976) [13] by Thompson and Lynn (1966)

, Burton (1968), and Amon and colleagues (1972). Therefore these atypical cases can be called Thompson Lynn variant of Mostyn Embrey syndrome [2, 4].

Another five atypical cases (variant) of the Mostyn Embrey syndrome associated with bicornuate uterus rather than didelphys uterus were reported by Brezina in 1977, Gazárek et al in 1979 [two cases], and Shenker and Brickman in 1979 [two cases]. Therefore, the total cases of

Thompson Lynn variant of Mostyn Embrey syndrome were nine by the end of the 1970s [2, 4].

In 1972, Vinstein and Franken reported the first atypical case (variant) of Mostyn Embrey syndrome associated with septate uterus rather than didelphys uterus. Hörr in 1979 described for the first time the occurrence of the syndrome in two sisters. Shenker and Brickman were probably the first to report pregnancy in a patient with Mostyn Embrey syndrome in 1979 [2, 4].

The most common clinical manifestation is dysmenorrhea, pelvic pain, vaginal or pelvic mass after menarche. Early diagnosis is important, to preserve fertility, and adhesion due to retrograde menstruation, hematometra, hematosalpinx and endometriosis which lead to distorted pelvic anatomy. These anomalies could be diagnosed with ultrasonography and magnetic resonance imaging. Resection or incision of vaginal septum is adequate treatments to relieve symptoms and reserve fertility in classic Mostyn Embrey syndrome.

Mostyn Embrey syndrome should be considered in the differential diagnoses in young females with unilateral renal agenesis presenting with pelvic mass, symptoms of acute abdomen, and acute urinary retention [1, 2, 3, 4].

A syndrome is a collection of medical signs and symptoms that are associated with each other and, often, with a specific disease or disorder. Syndromes in medicine are often named after the physician or group of physicians that discovered them or initially provided the full clinical picture or the best description of the syndrome. However, many of the rare syndromes have been described by physicians in many areas of the world before the era of the internet which has been associated with easy access to clinical reports throughout the world. Unfortunately, some syndromes have been attributed unfairly and inappropriately to physicians other than those first described them [2, 4].

Mostyn P. Embrey was the first to report the association of uterus didelphys, hemi-vaginal obstruction in association with ipsilateral renal agenesis. During the period from 1950 to 1970, sixteen cases were described in English literature including the first case of Mostyn P. Embrey. Most of the cases were classical Mostyn Embrey syndrome: Wolf and Allen [four cases], Semmens, Gibberd, Hill, Allan and Cowan [Three cases], Thompson and Lynn [Two cases] and Lewis and Brent. Atypical cases (variants) of the syndrome associated with bicornuate uterus rather than didelphys uterus were reported by Thompson and Lynn in 1966 and Burton in 1968 respectively. In April 1971, Herlyn and Werner reported the 17th case of the syndrome and the first in German literature. Unfortunately, they missed the earlier cases reported in English literature and considered their case as a new typical syndrome of anomalies Wunderlich M reported the thirty first case of the syndrome in 1976; the case was the third in German literature. It is intriguing and disturbing to discover the following facts about the historic reporting of Mostyn Embrey syndrome: Herlyn and Werner reported the 17th case of the syndrome twenty-one years after the first excellent description of the syndrome by Mostyn P. Embrey. Wunderlich M reported the thirty first case of the syndrome more than twenty-five years after the report of Mostyn P. Embrey [1, 2, 3, 4].

Gazárek et al. reported in 1979 four cases of Mostyn Embrey syndrome. They described them as having “duplication of uterus, para-cervically localized cystic resistance and renal aplasia on the same side”. It is very surprising that Gazárek et al. considered their cases to have two different syndromes; the Herlyn-Werner syndrome in two cases and the Wunderlich syndrome in the other two despite all apparent similarity of the four cases.

The patient of Wunderlich was described as having bicornuate uterus with simple vagina and isolated hematocervix on right without connection of

the right uterus to the vagina in association with aplasia of the right kidney and ureter. Non classical forms of Mostyn Embrey syndrome associated with bicornuate uterus rather than didelphys uterus have already been described before Wunderlich by Thompson and Lynn, Burton, Amon and colleagues.

Obviously, the occurrence of bicornuate uterus in two of the four patients of Gazárek et al. and their unawareness of all the previously reported cases led them to their inaccurate description of the well-known syndrome [2, 4].

The classical uterine abnormality in Mostyn Embrey syndrome is didelphys uterus which is sometimes described as duplicated or double uterus. However, other uterine abnormalities especially bicornuate uterus and septate have been associated and such cases can be considered a variant of the classical syndrome. Such variants have been reported by many authors including Rock and Jones, Stassart et al. and Candiani et al., Heinonen Gholoum et al., Smith and Laufer and Vercellini et al. [2, 4].

There is only one reports of the familial occurrence of Mostyn Embrey syndrome in two sisters [2, 4]. In this Iraqi case a cousin of the patient have an incomplete variant of the syndrome. Such familial occurrence of the syndrome has not been reported before.

However, incomplete variants of the syndrome exist including the cases reported Fried et al. Their cases had bicornuate uterus or unicornuate uterus and unilateral renal agenesis but without hemi-vaginal obstruction and hematocolpos [2, 4]. Acute retention of urine as the initial clinical presentation has been rarely reported in this syndrome. This case is the seventh case of Mostyn Embrey syndrome presenting with retention of urine.

Strong suspicion and knowledge of this syndrome are mandatory for an accurate diagnosis. In fact, accurate diagnosis and surgical treatment can be delayed for several months or even years.

The treatment of patients with classical Mostyn syndrome is generally simple vaginal excision of obstructive septum. The imaging modalities that can be used to diagnose Mostyn Embrey syndrome include ultrasonography, hysterosalpingography and MRI. In this case, hysterosalpingography was not of much help. In fact, Brezina when reporting a case in 1977 emphasized the possibility of not diagnosing Mostyn Embrey syndrome for many years after the menarche even by hysterosalpingography.

Ultrasound is affordable, non-invasive, widely available imaging modality that contributes to the accurate diagnosis of Mostyn Embrey syndrome. However, there can be difficulties in visualizing the vaginal septum on ultrasound which best shown on MRI. MRI can facilitate early diagnosis and thus help in the prevention of further complications because MRI can show the müllerian duct anomaly complicated by obstructed hemi-vagina in detail and also the ipsilateral renal agenesis. Hollander and colleagues when reporting a case of the syndrome made a recommendation for pre-adolescent screening [2, 3, 4].

Conclusions

It is recommended that pediatricians, and pediatric nephrologists need to be aware of this syndrome to avoid unexpected presentations which may lead to delayed diagnosis and sometimes to inappropriate management. The imaging modalities that can be used to diagnose Mostyn Embrey syndrome include ultrasonography, hysterosalpingography and MRI. In this case, hysterosalpingography was not of much help. Ultrasound is affordable, non-invasive, widely available imaging modality that contributes to the accurate diagnosis of Mostyn Embrey syndrome. However, there can be difficulties in visualizing the vaginal septum on ultrasound which best shown on MRI. MRI can facilitate early diagnosis

and thus help in the prevention of further complications. Syndromes in medicine are often named after the physician or group of physicians that discovered them or initially provided the full clinical picture or the best description of the syndrome. Unfortunately, Mostyn Embrey syndromes has been attributed unfairly and inappropriately to physicians other than those first described them in almost all previous papers.

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References

1. Embrey MP. (1950). A case of uterus didelphys with unilateral gynatresia. Br Med J. 1(4657); 820-821.
2. Al-Mosawi AJ. (2016). Mostyn Embrey syndrome. Lap Lambert Academic Publishing GmbH & Co. KG, Saarbrücken, Germany.
3. Al-Mosawi AJ. (2017). Unilateral Renal Agenesis and the Awareness of Mostyn Embrey Syndrome. Journal of Renal Medicine. 1(1):1-4.
4. Al-Mosawi AJ. (2021). Mostyn Embrey syndrome: Awareness and ethics of naming syndromes. Scholars' Press.
5. Hadden D. (1922). Double uterus and vagina. Am J Obstet Gynecol. 3:526.
6. Purslow CE. (1922). A case of unilateral haematocolpos, hematometra and hematosalpinx. J Obstet Gynaecol Br Emp. 28: 643.
7. Wilson J St G. (1925). A case of double uterus and vagina with unilateral haematocolpos and hematometra. J Obstet Gynaecol Br Emp. 32:127-128.
8. Brown AC, Brews A. (1930). Congenital retention of the menses. J Obstet Gynaecol Br Emp. 37: 233-255.
9. Masson JC, Mueller SC. (1933). Uterus duplex bicornis with septate vagina open only on one side: report of case. Surg Clin North Am. 13:857- 861.
10. Carnington GL, Burlington NC. (1933). Incomplete bipartite uterus with unilateral hematocolpos and salpingitis. Am J Obstet Gynecol. 25: 924-925.
11. Martindale L. (1935). A case of uterus didelphys associated with haematocolpos of the right vagina and simulating appendicitis. Lancet. 1:1389-1390.



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