Definitive and Conservative Surgical Management of Congenital Cervical Atresia: Report of 2 Cases

Allen Gideon R. Tan, MD and Madonna Victoria C. Domingo, MD, FPSREI

Two cases of congenital cervical atresia are presented in this paper. The first is a 16-year old who presented with a 2-year history of amenorrhea with associated abdominal pain. Upon laparoscopy, it was confirmed that she had cervical agenesis, and subsequently underwent total hysterectomy. No cervical tissue was identified on histopathologic examination. The second case is a 15-year old who likewise presented with amenorrhea and cyclic abdominal pain. She underwent uterovaginal canalization by a combined transvaginal and transabdominal surgery, after a failed attempt at canalization from a different hospital. Twelve months after the surgery, however, there was distal stenosis of the created uterovaginal canal, prompting a re-admission. She underwent vaginoplasty and total abdominal hysterectomy. After histopathologic examination, a diagnosis of cervical dysgenesis was confirmed. The early recognition of congenital anomalies of the uterine cervix is imperative, but its accurate diagnosis poses a great challenge. Numerous treatment options are available, but to date, its surgical management remains controversial. Nevertheless, individualized treatment is paramount, as described in the cases presented.

Key words: Mullerian anomaly, cervical atresia, cervical agenesis, cervical dysgenesis, conservative surgical management, uterovaginal canalization

Introduction

Mullerian duct anomalies consist of a spectrum of structural malformations from the failure or abnormal development of the paramesonephric duct system. The true prevalence of these malformations is not absolutely known, but have been said to range from 0.001% to 10% in the general population.[1] They may result from 1) failure of development of one or more Mullerian ducts, 2) failure of, or abnormalities in fusion of the ducts, and 3) total or partial failure of septum absorption. Because disturbances may occur at any number of stages of embryologic development, these anomalies encompass a wide variety of manifestations.[2]

Congenital atresia of the uterine cervix is an uncommon Mullerian anomaly that occurs in only 1 in 80,000 to 100,000 births.[3] According to the American Society of Reproductive Medicine (ASRM) classification system,[4] this group of anomalies belongs to Class I – hypoplasia and agenesis. Cervical atresia is further categorized under Class Ib (Table 1). The most common clinical manifestations of this group of malformations involve symptoms of obstructed menstrual flow. Patients present with primary amenorrhea, and cyclic abdominal pain related to hematometra or endometriosis as a consequence of retrograde menstruation.[5] The early and accurate diagnosis of Mullerian abnormalities at the time of its presentation is imperative, in order to prevent the emergence or progression of such complications that may damage structures and impact fertility. More importantly, it allows for a prompt preoperative evaluation that aids in providing the best treatment options for patients. In cases of anomalies of the uterine cervix, however, there have been no carefully designed cohort or randomized trial to support a best surgical approach.[6]

Many authors advocate total hysterectomy for cervical agenesis with a functioning endometrium. However, with advances in reconstructive surgery, and the advent of assisted reproductive technologies, conservative (uterus-sparing) modalities of treatment have been adopted by some.[7] Nonetheless, conservative surgical management of such cases remains controversial, owing to its considerable risks.[8]

Table 1. American Society of Reproductive Medicine classification of Mullerian anomalies[4]

Class I	Hypoplasia / agenesis	a) Vaginal b) Cervical c) Fundal d) Tubal e) Combined
Class II	Unicornuate	a) Communicating b) Noncommunicating c) No cavity d) No horn
Class III	Didelphys	
Class IV	Bicornuate	a) Partial b) Complete
Class V	Septate	a) Partial b) Complete
Class VI	Arcuate	
Class VII	DES drug-related	

Two cases of congenital cervical atresia are presented. While the first was managed with an outright total laparoscopic hysterectomy, the second case initially underwent uterovaginal canalization before she subsequently had an abdominal hysterectomy for stenosis of the reconstructed uterovaginal canal.

Case 1

A 16-year old female was referred to the out-patient clinic of the Section of Reproductive Endocrinology and Infertility (REI) of a tertiary training hospital for amenorrhea. The patient and her immediate family lived in Bohol. She was the youngest of 6 siblings, born to a then 43-year old homemaker and a 37-year old construction worker. At the time of consultation, the patient was a 4th year high school student, and denied any history of smoking or illicit drug use. She had no sexual intercourse.

Upon review of her developmental history, she had thelarche at 11, and pubarche at 13 years of age. The patient had experienced intermittent hypogastric pain, radiating to the back, at 2 to 3-month intervals since 14 years old. She took mefenamic acid as needed, which afforded little to no relief of her symptoms. A year since the onset of her abdominal pains, the patient sought consultation at a provincial hospital where a holoabdominal ultrasound showed hematocolpometra, with note of a normal-sized uterus, and distended endometrial and endocervical cavities. Both kidneys were visualized with no disparity in size or configuration. Magnetic resonance imaging (MRI) showed hematometra with a hypoplastic cervix. The cervical os was not delineated, but the vagina appeared intact. Both ovaries were visualized, and a 3.1 cm right adnexal cyst was seen anterior to the right ovary and superior to the urinary bladder (Figure 1). The patient was eventually advised to seek further evaluation and management at the tertiary training hospital – 2 years since the onset of her symptoms.

On physical examination, the patient stood 145 cm tall and weighed 41 kg with a body mass index (BMI) of 19.5 kg/m^2. She appeared phenotypically female, with a breast Tanner stage 4 and pubic hair Tanner stage 3 (Figure 2). Upon examination of the perineum, she had normal external genitalia with an annular hymen. On speculum examination, she had pink vaginal mucosa with no identifiable cervix. Her vagina admitted one finger with ease, measured 6 cm in length, and ended blindly with no palpable cervix. There was a 6cm x 4 cm movable, slightly tender mass palpated superior to the vagina, left of midline, and a 4cm x 3 cm cystic, movable, non-tender mass at the right adnexal area. Recto- vaginal examination revealed

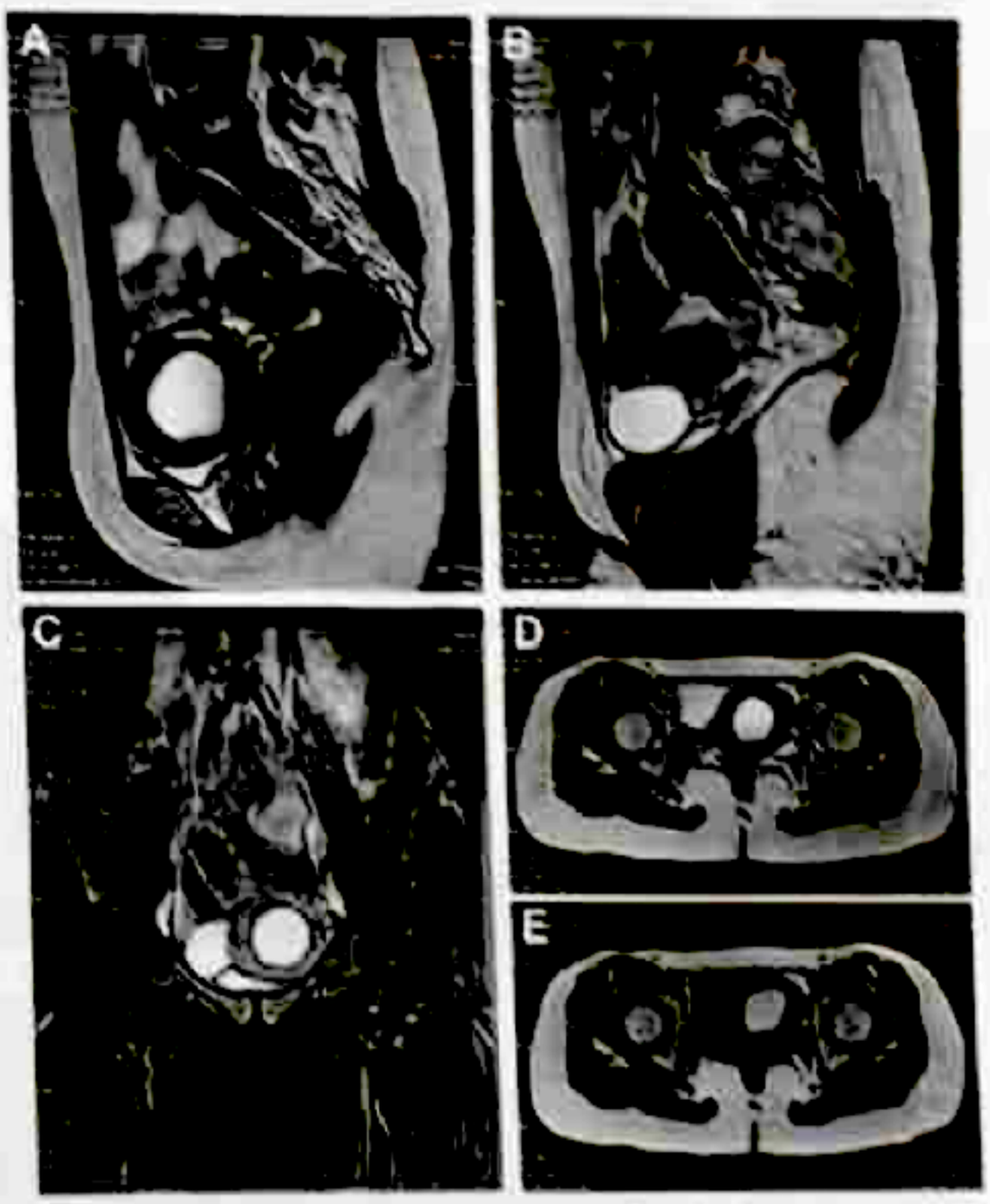


Figure 1. Magnetic resonance imaging of first index patient. A) T2-weight sagittal view showing the hyperintense hematometra. B) T2-weighted sagittal view showing the thin-walled right adnexal cyst. C) T2-weighted coronal view showing the uterus and right adnexal cyst. D-E) T2 and T1-weight transverse view showing the uterus and right adnexal cyst.

Table 1. American Society of Reproductive Medicine classification of Mullerian anomalies[4]

Class I	Hypoplasia / agenesis	a) Vaginal b) Cervical c) Fundal d) Tubal e) Combined
Class II	Unicornuate	a) Communicating b) Noncommunicating c) No cavity d) No horn
Class III	Didelphys	
Class IV	Bicornuate	a) Partial b) Complete
Class V	Septate	a) Partial b) Complete
Class VI	Arcuate	
Class VII	DES drug-related	

Two cases of congenital cervical atresia are presented. While the first was managed with an outright total laparoscopic hysterectomy, the second case initially underwent uterovaginal canalization before she subsequently had an abdominal hysterectomy for stenosis of the reconstructed uterovaginal canal.

Case 1

A 16-year old female was referred to the out-patient clinic of the Section of Reproductive Endocrinology and Infertility (REI) of a tertiary training hospital for amenorrhea. The patient and her immediate family lived in Bohol. She was the youngest of 6 siblings, born to a then 43-year old homemaker and a 37-year old construction worker. At the time of consultation, the patient was a 4th year high school student, and denied any history of smoking or illicit drug use. She had no sexual intercourse.

Upon review of her developmental history, she had thelarche at 11, and pubarche at 13 years of age. The patient had experienced intermittent hypogastric pain, radiating to the back, at 2 to 3-month intervals since 14 years old. She took mefenamic acid as needed, which afforded little to no relief of her symptoms. A year since the onset of her abdominal pains, the patient sought consultation at a provincial hospital where a holoabdominal ultrasound showed hematocolpometra, with note of a normal-sized uterus, and distended endometrial and endocervical cavities. Both kidneys were visualized with no disparity in size or configuration. Magnetic resonance imaging (MRI) showed hematometra with a hypoplastic cervix. The cervical os was not delineated, but the vagina appeared intact. Both ovaries were visualized, and a 3.1 cm right adnexal cyst was seen anterior to the right ovary and superior to the urinary bladder (Figure 1). The patient was eventually advised to seek further evaluation and management at the tertiary training hospital—2 years since the onset of her symptoms.

On physical examination, the patient stood 145 cm tall and weighed 41 kg with a body mass index (BMI) of 19.5 kg/m^2. She appeared phenotypically female, with a breast Tanner stage 4 and pubic hair Tanner stage 3 (Figure 2). Upon examination of the perineum, she had normal external genitalia with an annular hymen. On speculum examination, she had pink vaginal mucosa with no identifiable cervix. Her vagina admitted one finger with ease, measured 6 cm in length, and ended blindly with no palpable cervix. There was a 6cm x 4 cm movable, slightly tender mass palpated superior to the vagina, left of midline, and a 4cm x 3 cm cystic, movable, non-tender mass at the right adnexal area. Recto- vaginal examination revealed

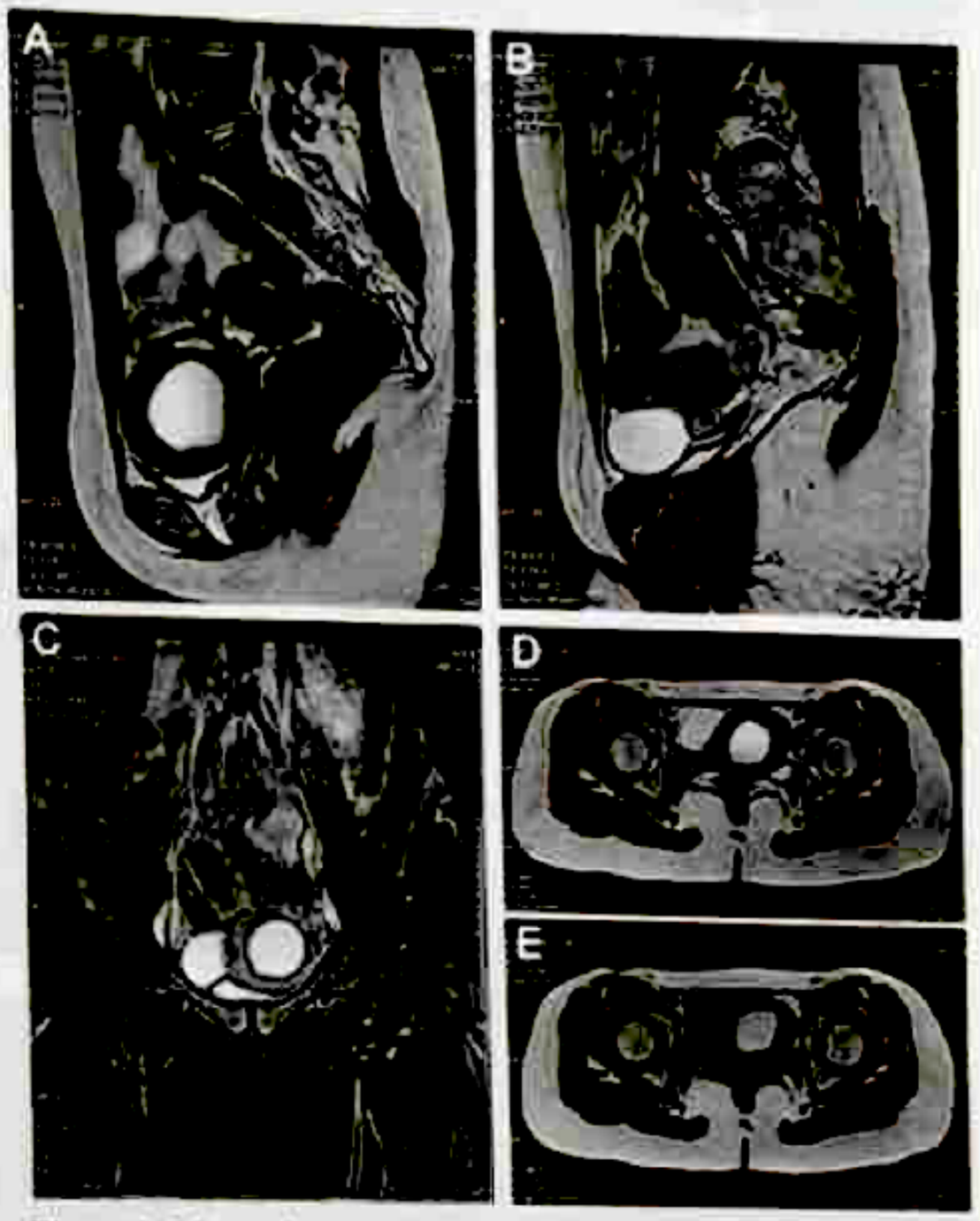


Figure 1. Magnetic resonance imaging of first index patient. A) T2-weight sagittal view showing the hyperintense hematometra. B) T2-weighted sagittal view showing the thin- walled right adnexal cyst. C) T2-weighted coronal view showing the uterus and right adnexal cyst. D-E) T2 and T1-weight transverse view showing the uterus and right adnexal cyst.

good anal sphincter tone, and an intact rectal vault with smooth mucosa. The uterosacral ligaments converged below the previously described left pelvic mass, which was palpated 6 cm from the anal verge.

Her karyotype was 46 XX. Transrectal ultrasound showed a rudimentary cervix measuring 0.9cm x 1.7cm x 1.6cm (with no endocervical canal noted), and a corpus with 2 endometrial cavities separated by a septum 0.4 cm thick. The left endometrial cavity measured 5.7cm x 5.9cm x 3.9cm (volume = 68.6 cc), and the right rudimentary cavity measured 2.9cm x 3.3cm x 1.7cm (volume = 8.5 cc). Both ovaries were visualized, and a right para-ovarian cyst, which measured 3.0cm x 3.7cm x 2.1cm was seen medial to the right ovary (Figure 3).

The preoperative diagnosis was primary amenorrhea secondary to cervical dysgenesis, rule out cervical agenesis; hematometra; right para-ovarian cyst. After preoperative counseling, the patient underwent operative laparoscopy. On examination under anesthesia, the pelvic findings were consistent with the initial assessment. Intraoperatively, there was no ascites and no note of endometriotic implants. The uterus was firm and globular, with a blunt inferior pole and with no connection to the proximal vagina. No cervix was identified. Both ovaries were grossly normal. The para-ovarian cyst seen on ultrasonography and MRI was a right paratubal cyst with a smooth capsule and no adhesions (Figure 4). Total laparoscopic hysterectomy with excision of right paratubal cyst was done. Upon reconstruction of the morcellated uterus, it measured 6.5cm x 4.5cm x 4 cm. The cervix was grossly absent. The myometrium was asymmetrically thick measuring 1.5 cm anteriorly and 2.3cm posteriorly. There was only one endometrial cavity, which measured 2cm in length and contained chocolate-like fluid. The endometrium measured 0.1cm. The rudimentary endometrial cavity identified on ultrasonography was a blood lake (Figure 5). The right paratubal cyst measured 4cm x 3cm x 3cm, which was found to be consistent with a paratubal cyst on histologic examination. Histopathologic examination of the hysterectomy specimen showed fragments of myometrial tissues with adenomyosis, and basal endometrium. No cervical tissue was identified.

The final diagnosis was primary amenorrhea secondary to cervical agenesis; hematometra, adenomyosis; right paratubal cyst; status post total laparoscopic hysterectomy, excision of right paratubal cyst (July 2013). The postoperative course of the patient was uneventful. She was sent home 3 days after the surgery. On subsequent clinic visits, she remained symptom-free.

Case 2

A 15-year old female was admitted twice, twelve months apart, under the care of the Section of REI of the same tertiary training hospital. She underwent uterovaginal canalization during the first admission, and was re-admitted for distal stenosis of her reconstructed uterovaginal canal, twelve months after. The patient and her family resided in Quezon Province. She was the fifth of 6 siblings, born to a then 32-year old homemaker and a 37-year old driver. She had been retained in her 3rd year

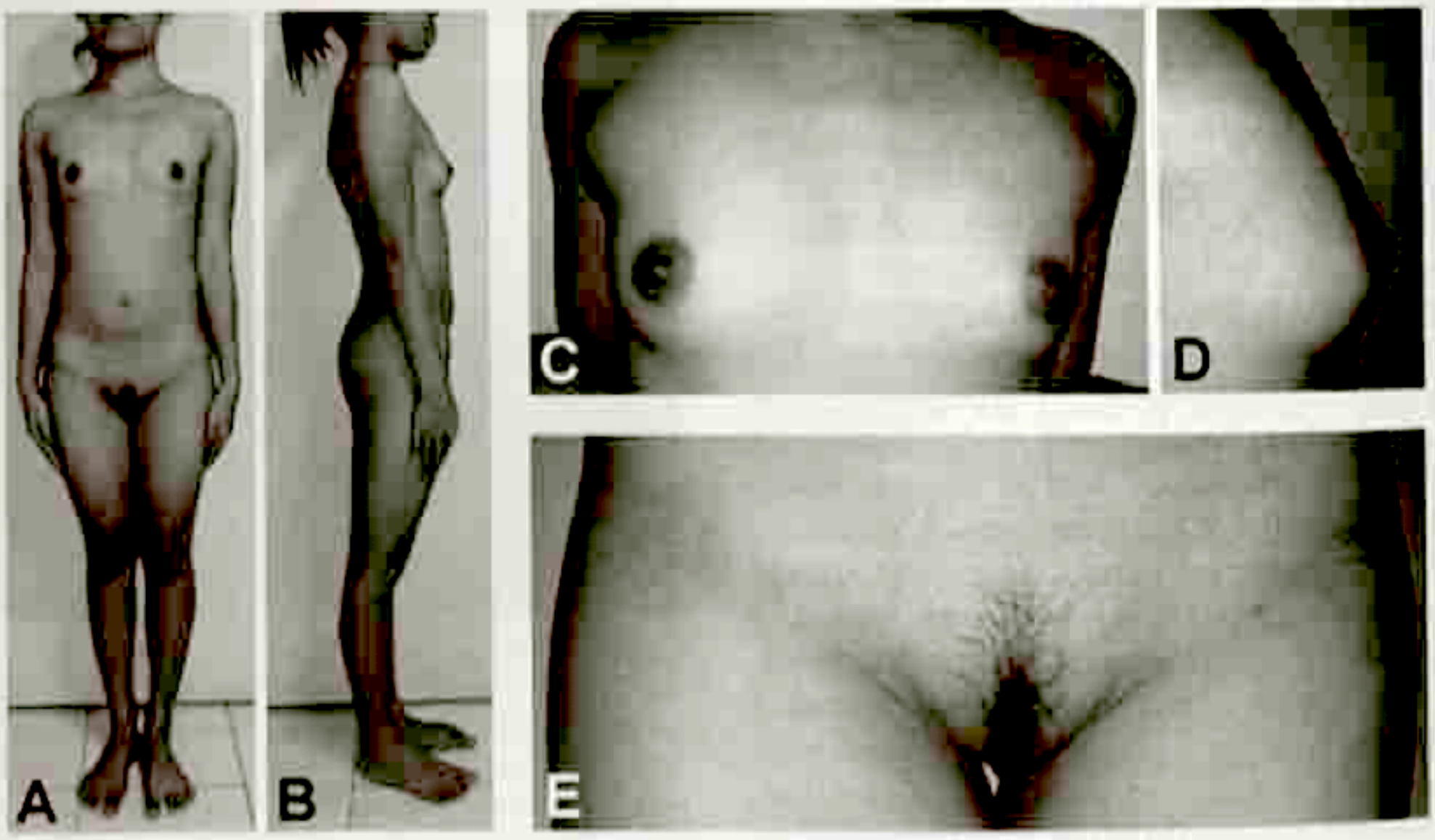
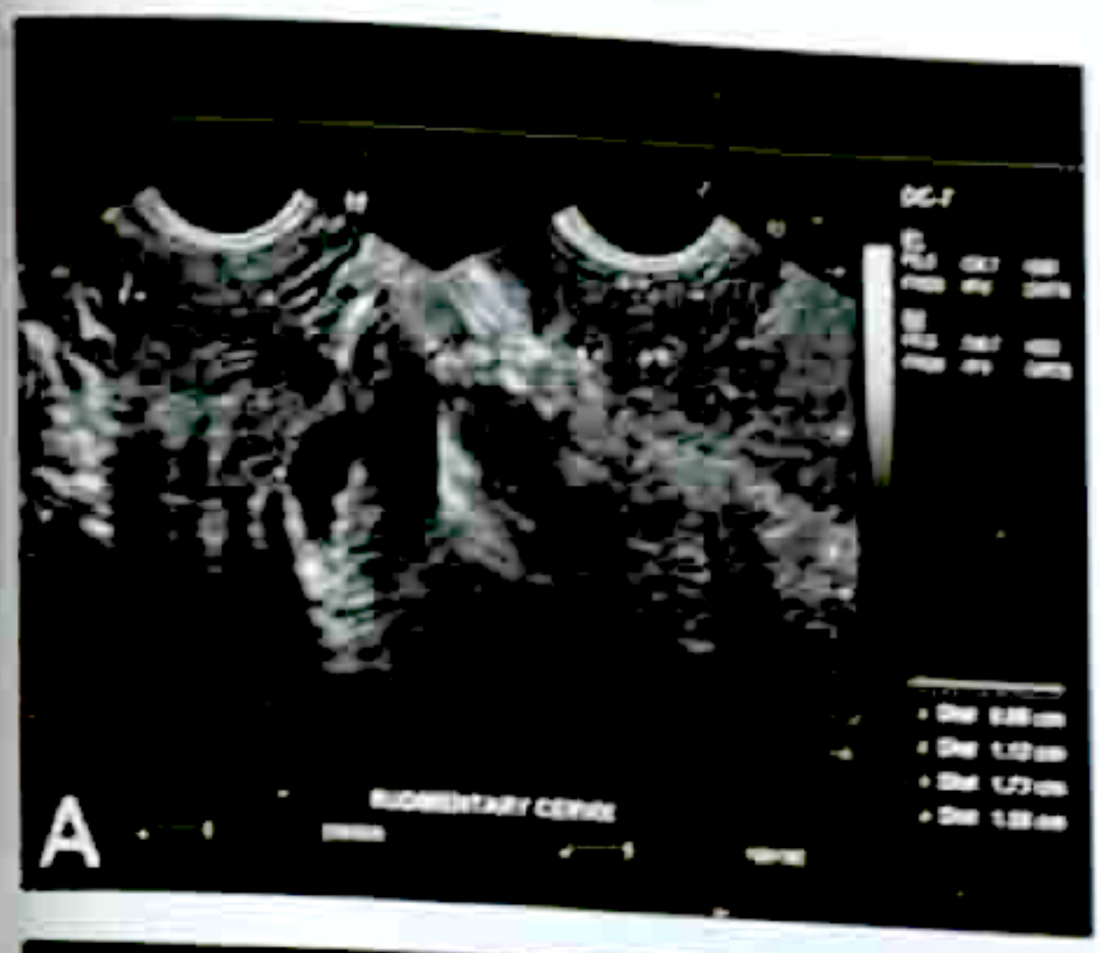
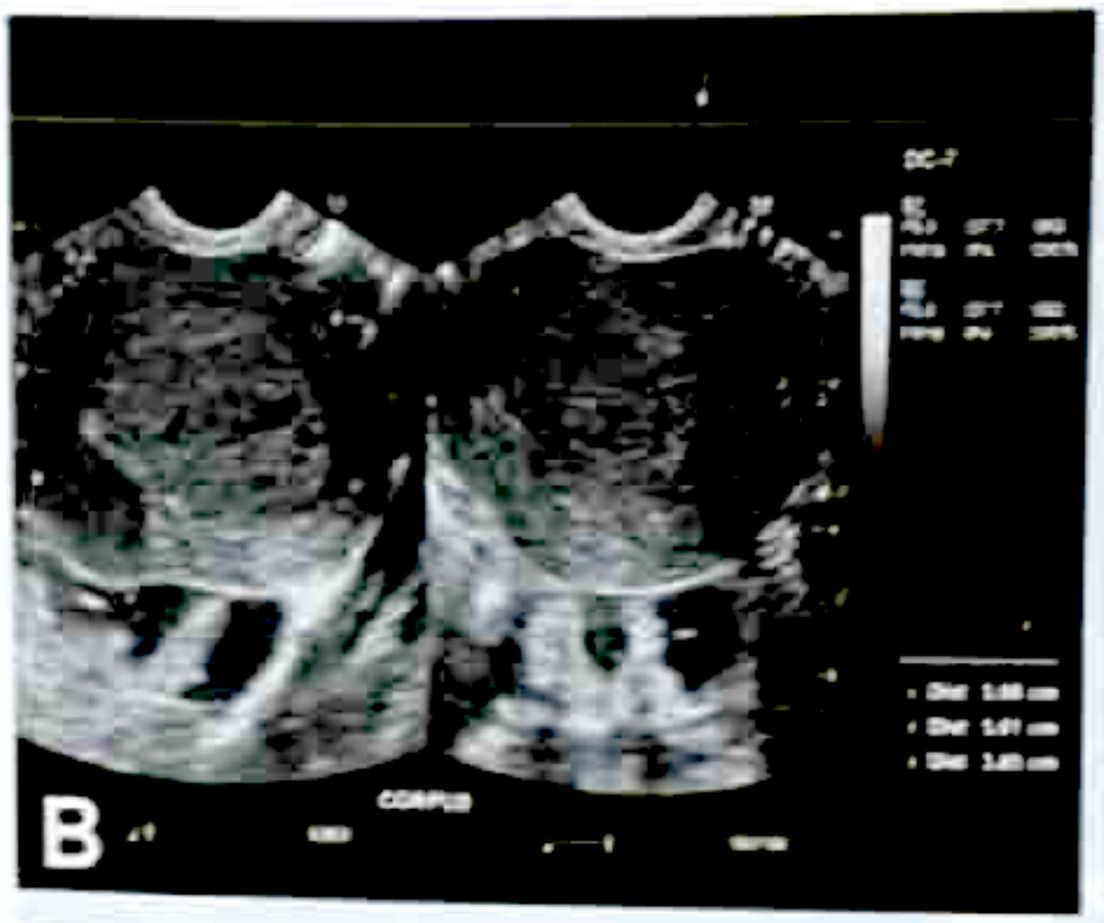
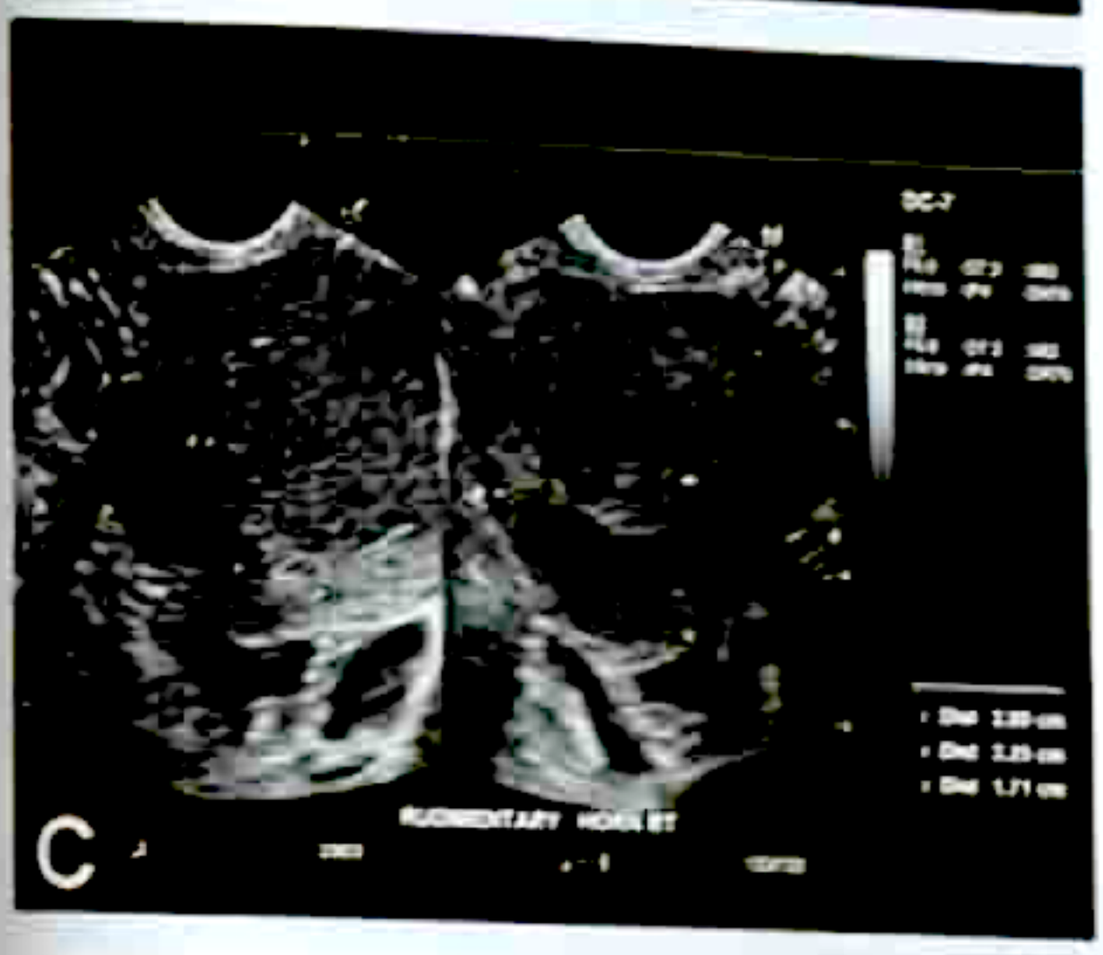
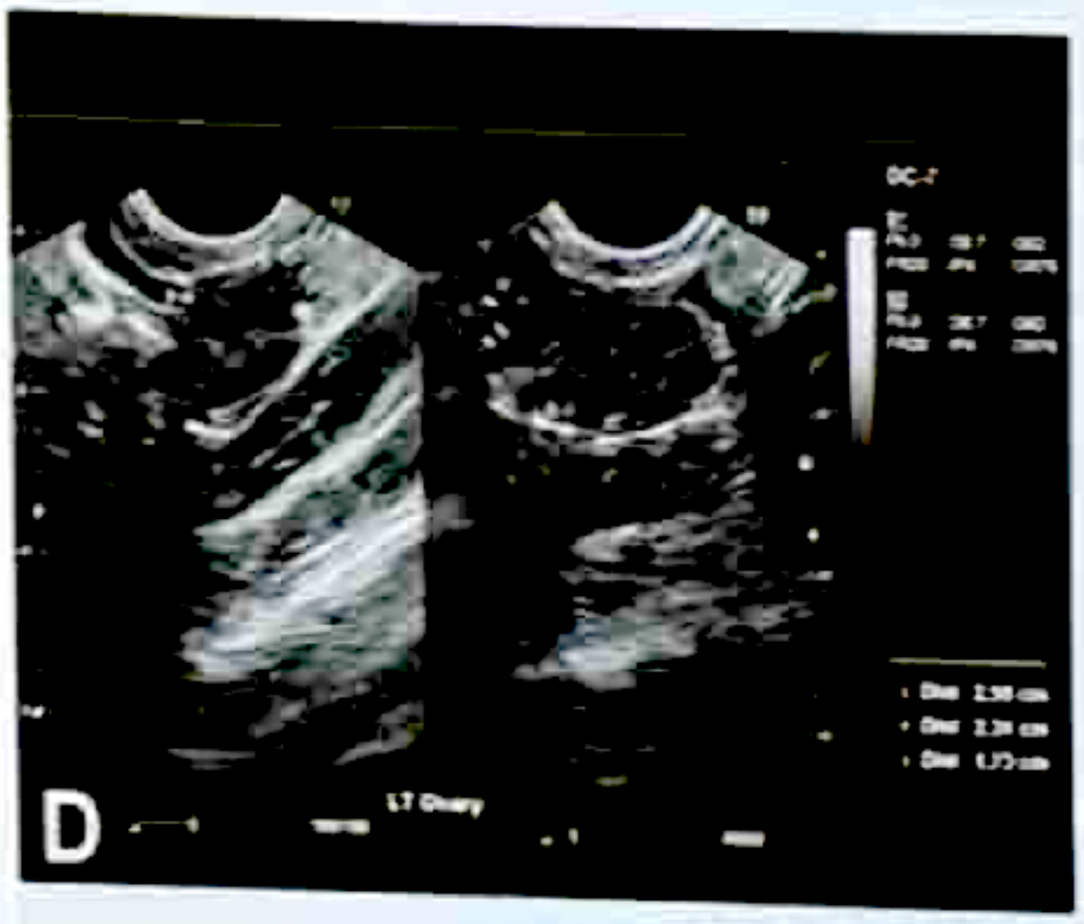


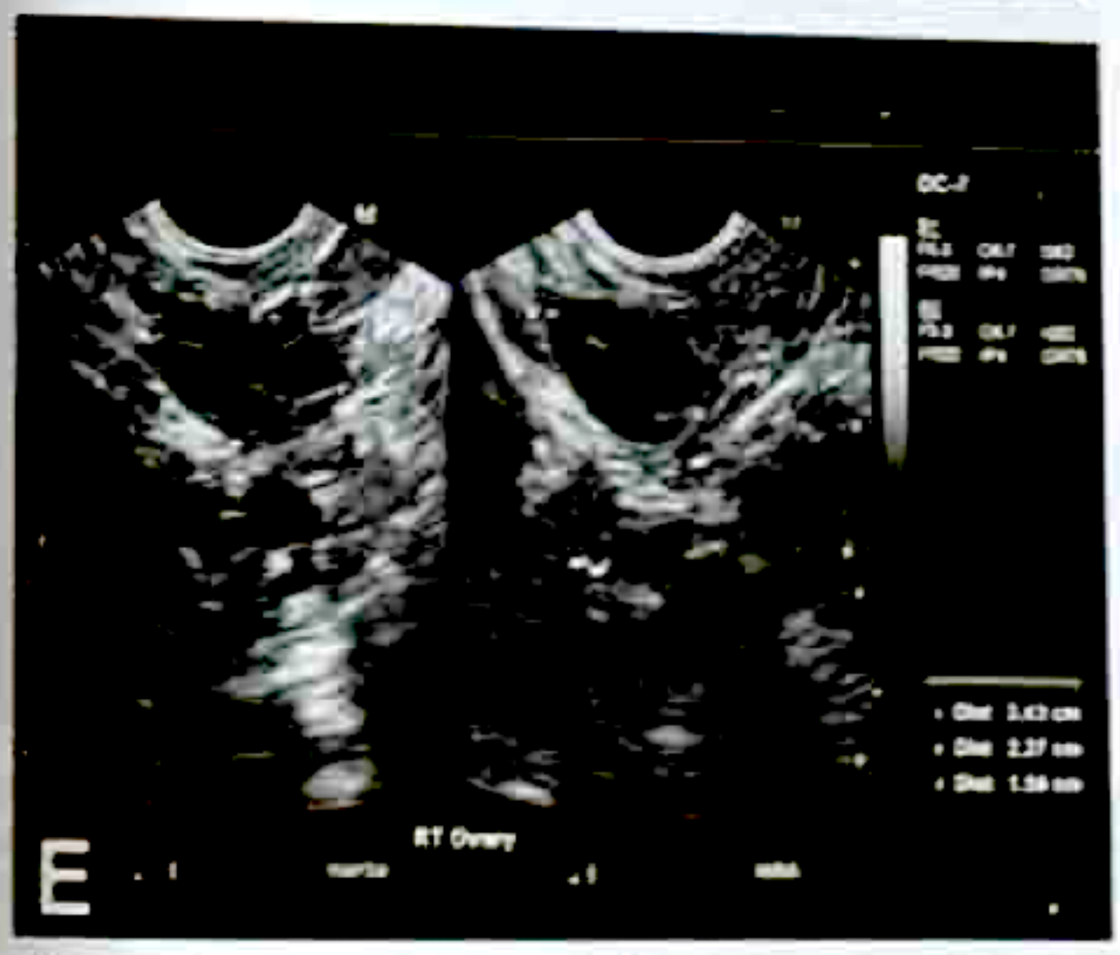
Figure 2. Images of first index patient. A-B) Full body. C-D) Breasts. E) Pelvis.











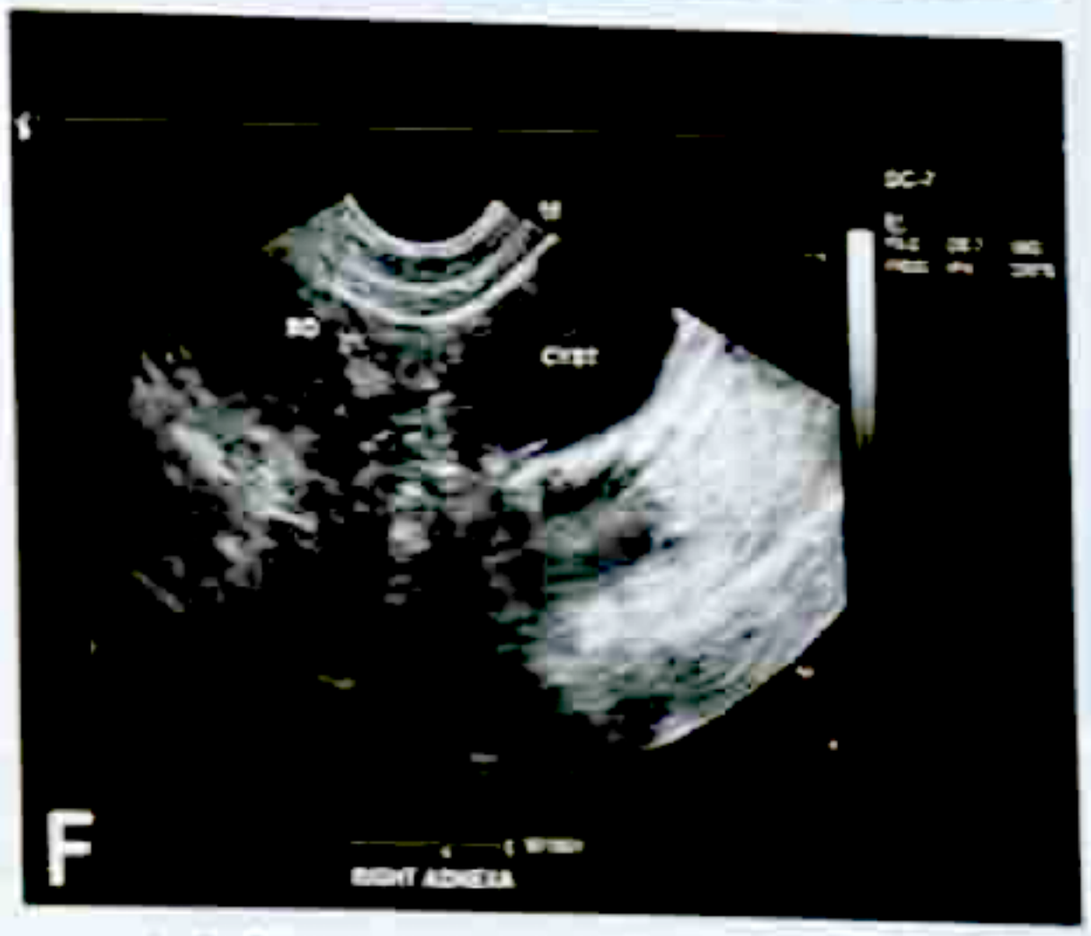


Figure 3. Transrectal ultrasound of first index patient. A) Rudimentary cervix. B) Corpus, endometrial cavity. C) Corpus, rudimentary endometrial cavity. D) Left ovary. E) Right ovary. F) Right para-ovarian cyst in relation to right ovary.

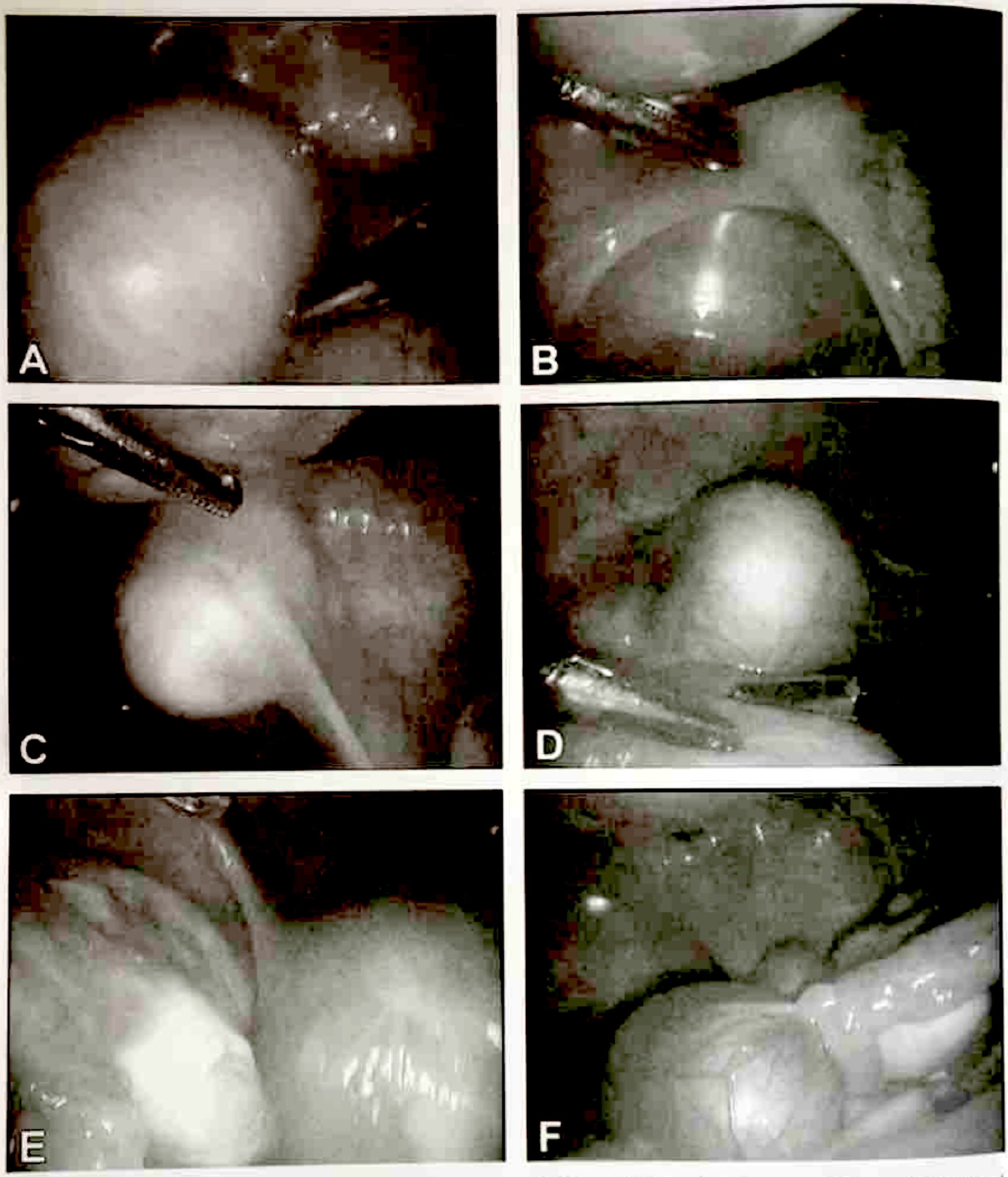


Figure 4. Intraoperative findings of first index patient on laparoscopy. A) Uterus. B) Postero-inferior aspect of the uterus in relation to the uterosacral ligaments. C) Postero- inferior aspect of the uterus in relation to the proximal end of the vagina, vaginal probe in place. D) Antero-inferior aspect of the uterus in relation to the proximal end of the vagina, vaginal probe in place. E) Left ovary and fallopian tube. F) Right paratubal cyst in relation to right ovary and fallopian tube.

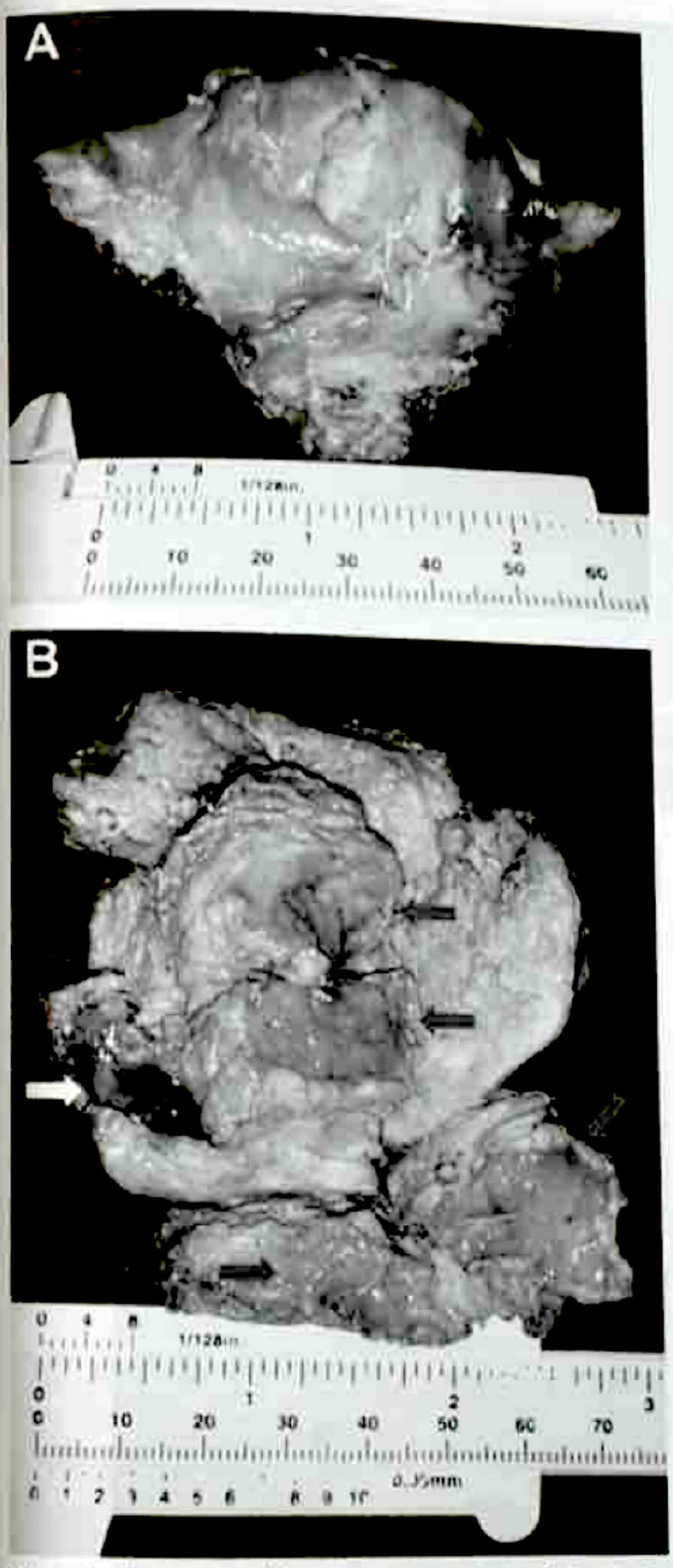


Figure 5. Reconstructed hysterectomy specimen of first index patient. A) Uterus with no cervix. B) Cut section of uterus showing myometrium with blood lake (encircled arrow) and endometrium (arrows).

of high school due to multiple absences. She denied having vices, and had no sexual contact.

Before her first admission in August 2012, the patient was noted to have had normal development until she was referred to the tertiary training hospital for amenorrhea. She had thelarche at 12 years old, and pubarche at 13 years old. Prior to the first admission, she had a 7-month history of episodic hypogastric pain, for which she took oral Hyocine-N-butyl bromide, which afforded only transient relief. Five months from the onset of her symptoms, due to persistent severe abdominal pain, she was admitted at a private provincial hospital, diagnosed as a case of transverse vaginal septum. Excision of the septum was attempted, but allegedly due to technical difficulties encountered intraoperatively, the surgery was abandoned. The patient did not experience any menstrual bleeding post-operatively. Two months from the procedure, she was admitted at the tertiary training hospital for the same complaint.

Upon examination during her 2012 admission, her abdomen was flat, with direct tenderness on the right hypogastric area on deep palpation. She had grossly normal external genitalia, with an annular hymen. Upon digital rectal examination, she had good anal sphincter tone and an intact rectal vault. No bulging anterior rectal wall mass was appreciated. There was a 4cm x 4cm cystic, movable, right adnexal mass palpated on bimanual examination.

Abdominal ultrasound showed a single, enlarged kidney on the left, with no structure noted at the right renal fossa. Transperineal and transabdominal ultrasound revealed findings suggestive of a transverse vaginal septum, hematocolpometra and a right hematosalpinx. The uterus and cervix were dilated by a collection of low-level echo fluid with a volume of 20 cc. The upper two-thirds of the vagina was likewise dilated to 5.1cm x 3.0cm x 3.5cm (volume = 28 cc). A band measuring 0.4 cm thick was noted 2.8 cm from the introitus, separating the identified fluid collection from the distal third of the vagina. Both ovaries were visualized. Lateral to the right ovary was a tubulocystic structure measuring 4.7cm x 6.0cm x 2.7cm, with incomplete septations and low-level echo fluid within (Figure 6).

The pre-operative diagnosis was primary amenorrhea secondary to transverse vaginal septum; hematocolpometra with right hematosalpinx; Status post failed excision of transverse vaginal septum (June 2012, Lucena); Congenitally absent right kidney. On pelvic examination under anesthesia, the vagina had pink mucosa. It admitted 1 finger with ease, and was found to be 5 cm long, ending blindly with no cervix identified. It was compressed by a fluctuant mass measuring 4cm x 3cm x 3.5cm, located at the right lateral vaginal wall, with its

cm, shortened by the contracted median episiotomy scar. Hymenal tags were noted (Figure 8). The vagina was pink and measured 2 cm in length. It ended blindly, and tapered towards its proximal end. On digital rectal examination, there was good anal sphincter tone and intact rectal vault. Six centimeters from the anal verge, there was a 3cm x 3cm cystic mass palpated, which was noted to be continuous with the enlarged right unicornuate uterus. At the right adnexal area was a fixed, cystic mass measuring 5cm x 4cm.

Transabdominal ultrasound revealed hematocolpometra, a right hematosalpinx and an endometriotic cyst on the right. The uterus measured 5.5cm x 5.1cm x 4.0cm, while the proximal vagina measured 5.0cm x 5.2cm x 4.6cm. Both structures were dilated by low-level echo fluid. The left ovary was normal, while the right ovary was converted to a 2.4cm x 2.9cm x 1.4cm unilocular, cystic mass with low to medium-level echo fluid within. Superior to the right ovary was a tubulocystic mass measuring 3.9cm x 3.8cm x 2.5cm, with incomplete septations and low-level echoes within (Figure 9).

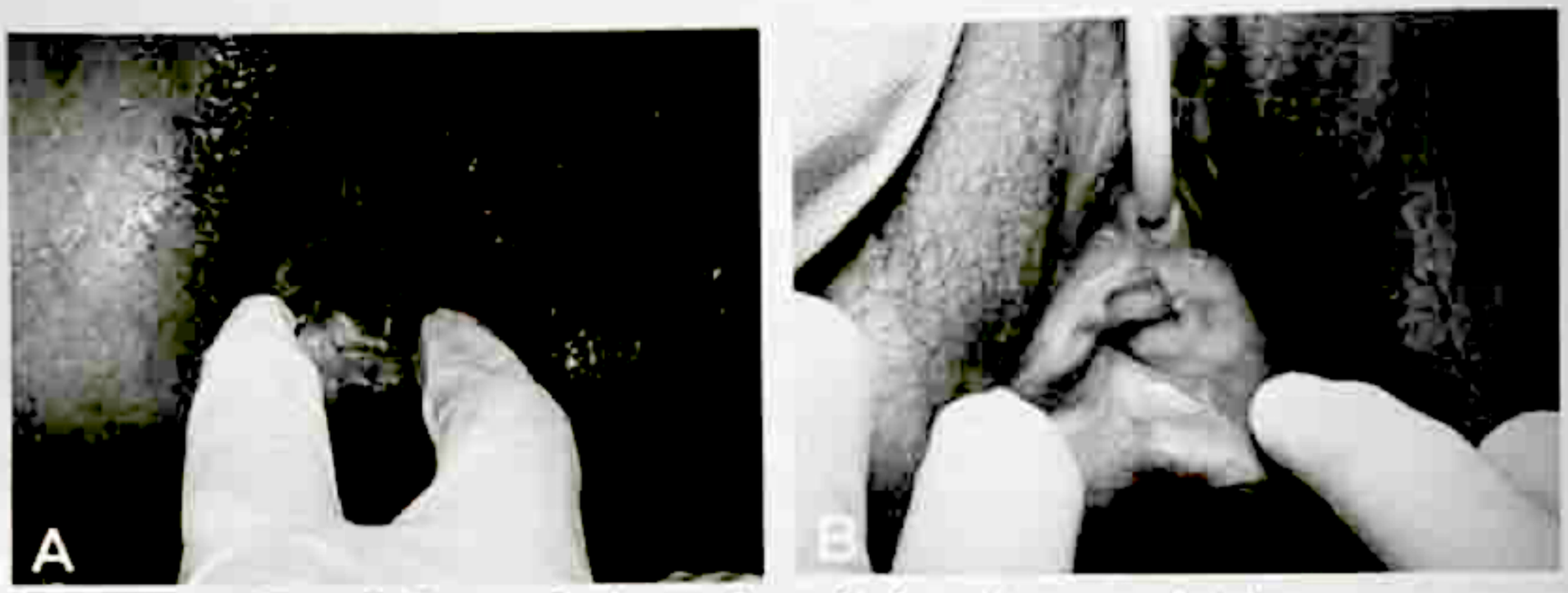


Figure 8. Images of perineum of second index patient on re-admission.

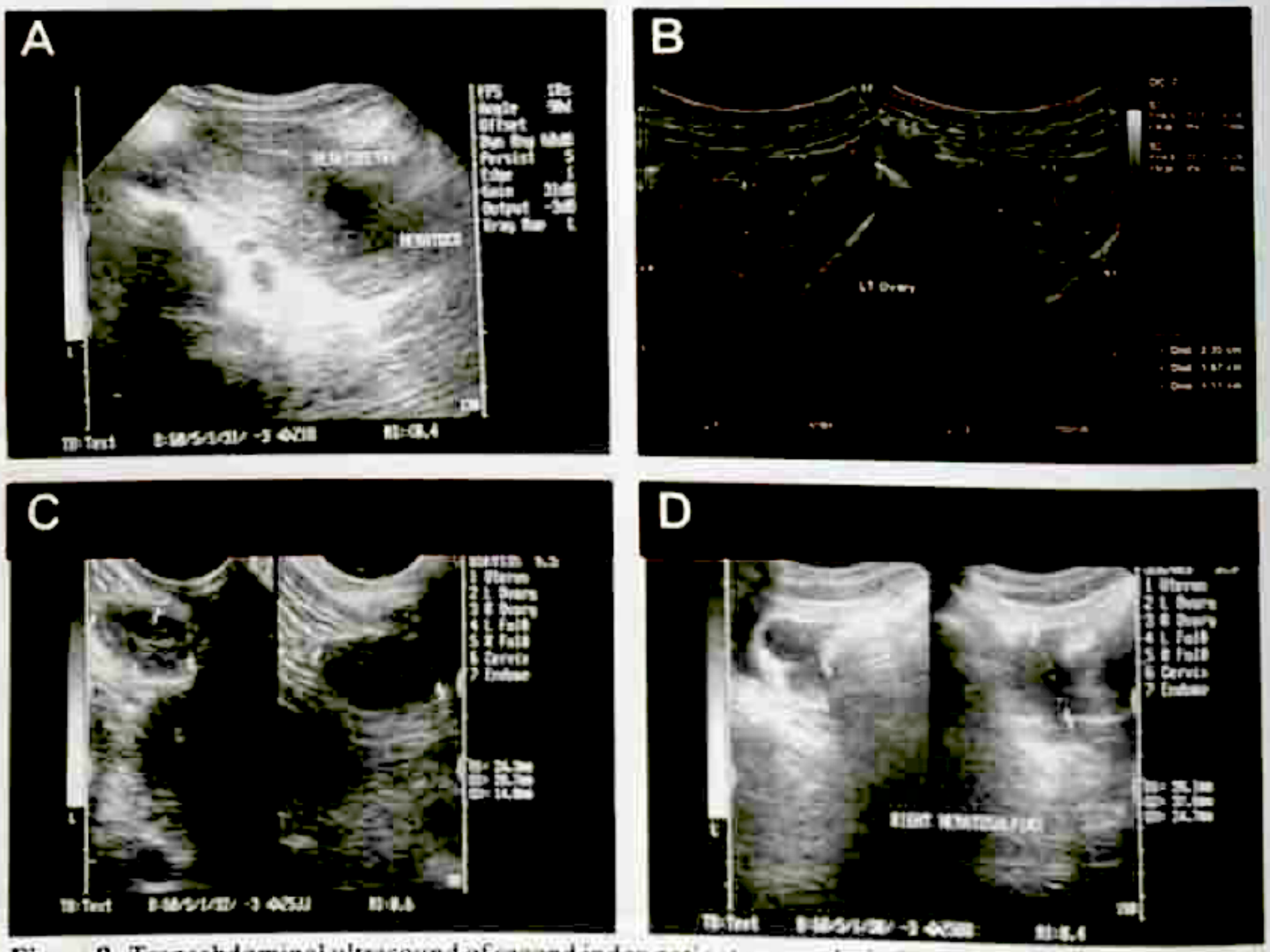


Figure 9. Transabdominal ultrasound of second index patient on re-admission. A) Hematocolpometra. B) Left ovary. C) Right endometriotic cyst. D) Right hematosalpinx.

The preoperative diagnosis was distal stenosis of uterovaginal canal; pelvic endometriosis with right endometriotic cyst; hematocolpometra with right hematosalpinx; status post uterovaginal canalization for right unicornuate uterus with cervical hypoplasia and transverse vaginal septum (August 2012); status post failed excision of transverse vaginal septum (June 2012, Lucena); congenitally absent right kidney. After thorough pre- operative counseling, the patient underwent vaginoplasty and total abdominal hysterectomy with right salpingectomy. The pelvic examination findings under anesthesia were consistent with the initial assessment. An attempt to re-canalize the stenotic uterovaginal canal by the transvaginal approach under ultrasound guidance was performed, but was unsuccessful due to technical difficulty.

A neovagina was created by blunt dissection of the dense fibrotic tissue proximal to the blind end of the vagina, and was kept patent by insertion of a vaginal mold. On laparotomy, there were dense adhesions noted between the omentum, right adnexum and posterior aspect of the unicornuate uterus. There were multiple blebs containing serous fluid on the right pelvic structures, with note of pseudocysts at the posterior cul-de-sac. The left ovary was grossly normal (Figure 10). The uterus measured 5cm x 4.5cm x 3.5cm, with no grossly identified cervical tissue. The myometrium measured 1.3cm, while the endometrium, 0.2cm. The vaginal cuff of the specimen measured 4cm in length, with dense tissue circumferentially (Figure 11). The right hematosalpinx measured 5cm x 3cm x 3cm, and contained chocolate-like fluid (Figure 12).

Histopathologic examination of the hysterectomy specimen showed proliferative phase endometrium and chronic endocervicitis with squamous metaplasia. The structure identified as the vaginal cuff on gross inspection (the obstructed vagina on ultrasonography) showed only endocervical glands and stroma along its entire length. Neither ectocervical nor vaginal tissue was identified.

The revised diagnosis was primary amenorrhea secondary to right unicornuate uterus with cervical dysgenesis; hematometra with right hematosalpinx; pelvic endometriosis; status post failed excision of transverse vaginal septum (June 2012); status post uterovaginal canalization (August 2012); status post vaginoplasty, total abdominal hysterectomy, right salpingectomy (August 2013); congenitally absent right kidney.

The patient had an unremarkable post-operative course. The plan was to maintain the vaginal mold in place, until adequate epithelialization and dilatation of the neovagina.

Discussion

The functional role of the uterine cervix is to provide a conduit for menstrual flow, to serve as a barrier to ascending infection, to supply cervical mucus for sperm transport, and to maintain a pregnancy in utero.[4] Its differentiation is a complex process involving tissues derived from both mesoderm and endoderm derivatives. Formation of the uterine cervix begins at around the

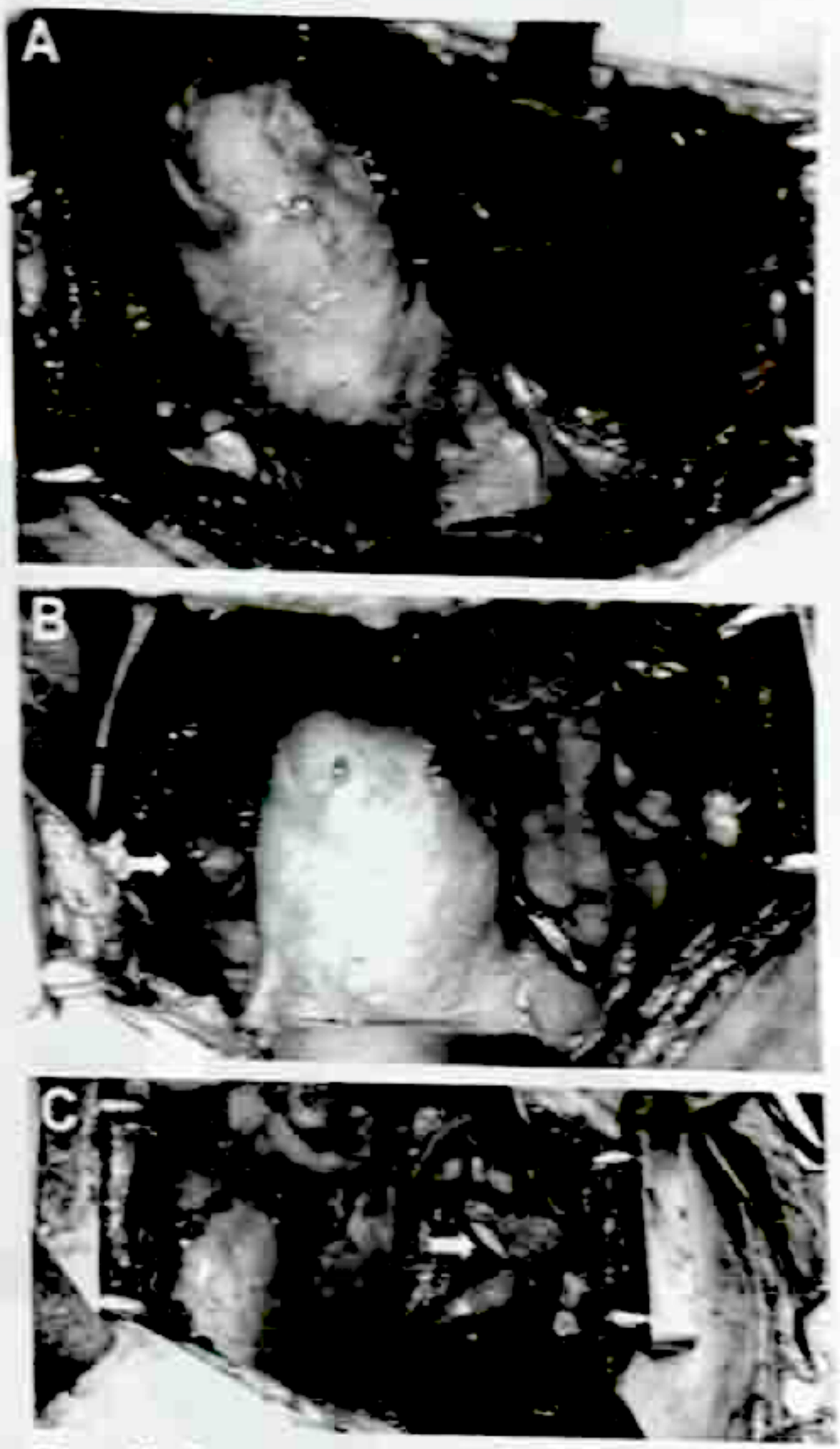


Figure 10. Intraoperative findings of second index patient on laparotomy on re-admission. A) Right unicornuate uterus with adhesions and endometriotic implants. B) Right unicornuate uterus in relation to right ovary (white arrow) and fallopian tube (black arrow). C) Left ovary (white arrow) and fallopian tube (black arrow) on left pelvic sidewall.

The preoperative diagnosis was distal stenosis of uterovaginal canal; pelvic endometriosis with right endometriotic cyst; hematocolpometra with right hematosalpinx; status post uterovaginal canalization for right unicornuate uterus with cervical hypoplasia and transverse vaginal septum (August 2012); status post failed excision of transverse vaginal septum (June 2012, Lucena); congenitally absent right kidney. After thorough pre- operative counseling, the patient underwent vaginoplasty and total abdominal hysterectomy with right salpingectomy. The pelvic examination findings under anesthesia were consistent with the initial assessment. An attempt to re-canalize the stenotic uterovaginal canal by the transvaginal approach under ultrasound guidance was performed, but was unsuccessful due to technical difficulty.

A neovagina was created by blunt dissection of the dense fibrotic tissue proximal to the blind end of the vagina, and was kept patent by insertion of a vaginal mold. On laparotomy, there were dense adhesions noted between the omentum, right adnexum and posterior aspect of the unicornuate uterus. There were multiple blebs containing serous fluid on the right pelvic structures, with note of pseudocysts at the posterior cul-de-sac. The left ovary was grossly normal (Figure 10). The uterus measured 5cm x 4.5cm x 3.5cm, with no grossly identified cervical tissue. The myometrium measured 1.3cm, while the endometrium, 0.2cm. The vaginal cuff of the specimen measured 4cm in length, with dense tissue circumferentially (Figure 11). The right hematosalpinx measured 5cm x 3cm x 3cm, and contained chocolate-like fluid (Figure 12).

Histopathologic examination of the hysterectomy specimen showed proliferative phase endometrium and chronic endocervicitis with squamous metaplasia. The structure identified as the vaginal cuff on gross inspection (the obstructed vagina on ultrasonography) showed only endocervical glands and stroma along its entire length. Neither ectocervical nor vaginal tissue was identified.

The revised diagnosis was primary amenorrhea secondary to right unicornuate uterus with cervical dysgenesis; hematometra with right hematosalpinx; pelvic endometriosis; status post failed excision of transverse vaginal septum (June 2012); status post uterovaginal canalization (August 2012); status post vaginoplasty, total abdominal hysterectomy, right salpingectomy (August 2013); congenitally absent right kidney.

The patient had an unremarkable post-operative course. The plan was to maintain the vaginal mold in place, until adequate epithelialization and dilatation of the neovagina.

Discussion

The functional role of the uterine cervix is to provide a conduit for menstrual flow, to serve as a barrier to ascending infection, to supply cervical mucus for sperm transport, and to maintain a pregnancy in utero.[8] Its differentiation is a complex process involving tissues derived from both mesoderm and endoderm derivatives. Formation of the uterine cervix begins at around the

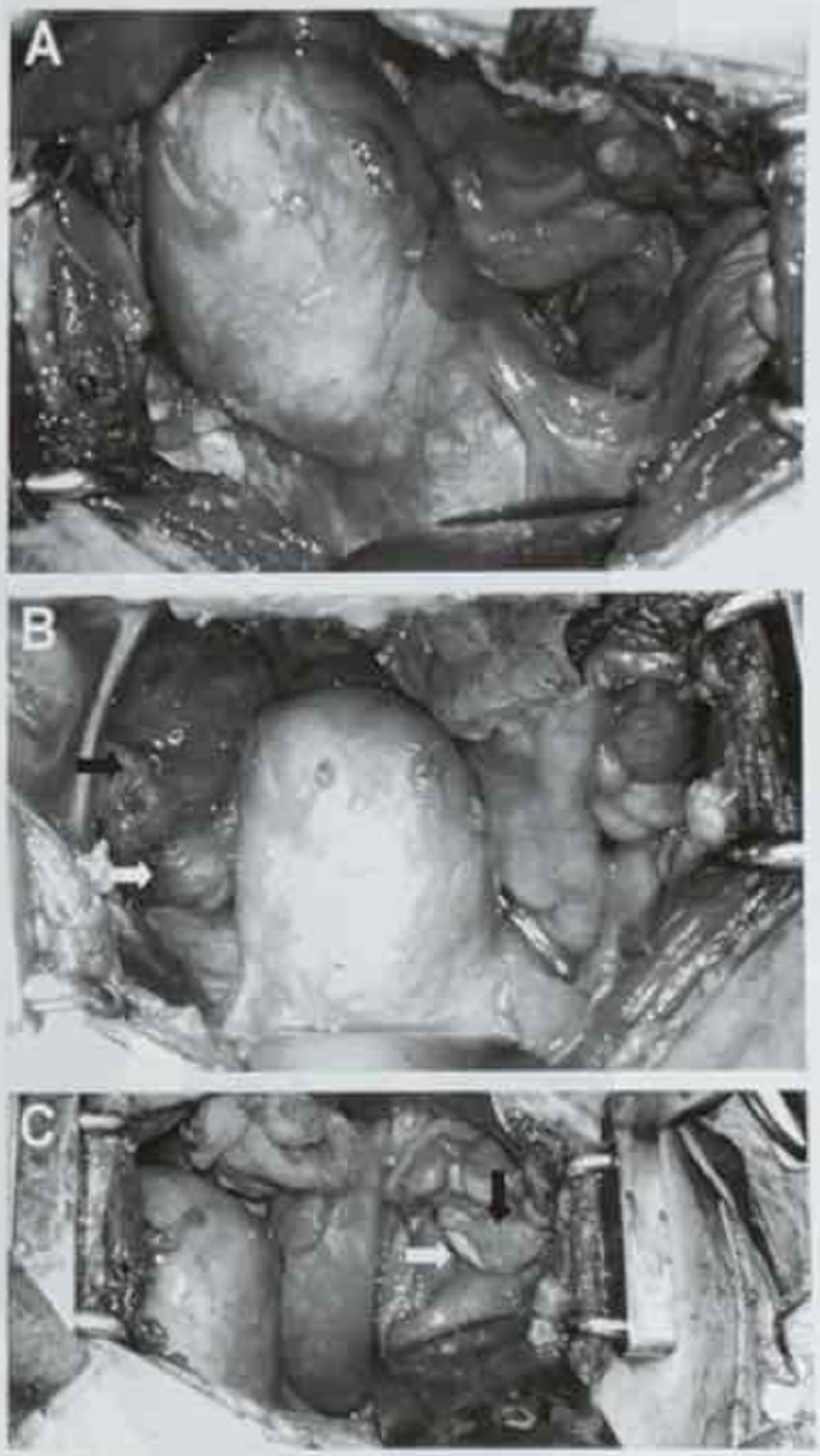


Figure 10. Intraoperative findings of second index patient on laparotomy on re-admission. A) Right unicornuate uterus with adhesions and endometriotic implants. B) Right unicornuate uterus in relation to right ovary (white arrow) and fallopian tube (black arrow). C) Left ovary (white arrow) and fallopian tube (black arrow) on left pelvic sidewall.

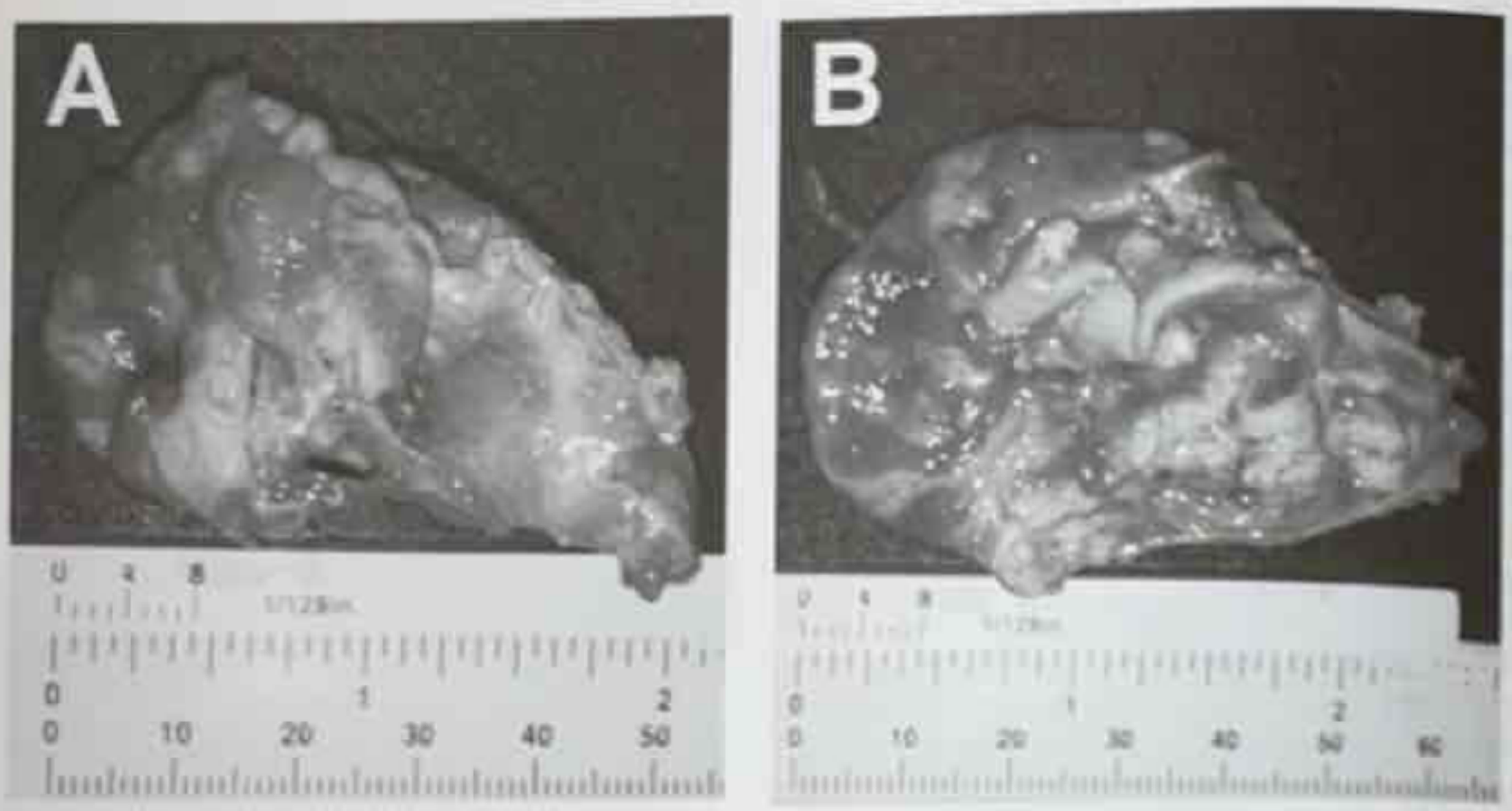


Figure 11. Right fallopian tube specimen of second index patient.

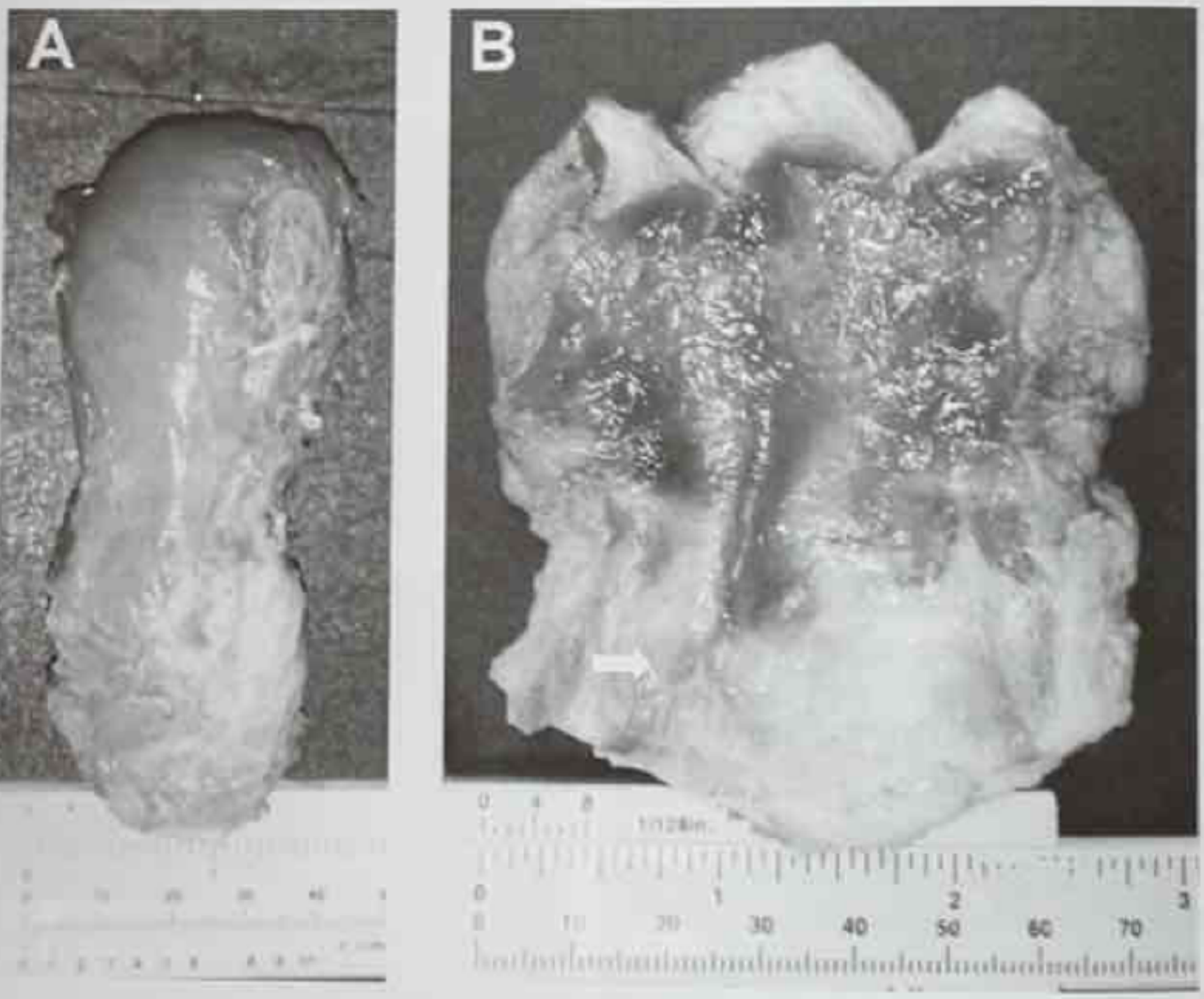


Figure 12. Hysterectomy specimen of second index patient. A) Gross specimen. B) Cut section of specimen with "vagina" (arrow).

fifteenth week of gestation, with the thickening of the caudal aspect of the Mullerian duct tissue in contact with the urogenital sinus and portions of the lateral Wolffian ducts. Abnormalities of the cervix may involve both early and late stages of differentiation, from elongation to canalization.[9]

The incidence of congenital abnormalities of the uterine cervix is very low. In 2011, Roberts and Rock[6] reported that a review of literature lists less than 200 cases since 1942. No local study has described its incidence in the Philippines. Recently, however, the Philippine Journal of Reproductive Endocrinology and Infertility published two case reports[10,11] of cervical atresia seen at tertiary hospitals in Metro Manila.

Cervical anomalies can be classified into 2 basic categories. The first type, cervical aplasia or agenesis, is the congenital absence of the uterine cervix, where the lower uterine segment narrows into a peritoneal sleeve above the normal site of communication with the vaginal apex (Figure 13).[6] The second type, cervical dysgenesis, can be divided into four subtypes: 1) Cervical body consisting of a fibrous band of variable length and diameter, where endocervical glands may be noted on histologic examination, 2) Intact cervical body with obstruction of the cervical os, 3) Stricture of the midportion of the cervix, with a bulbous tip and no identifiable canal, and 4) Fragmentation of the cervix (Figure 14).[6]

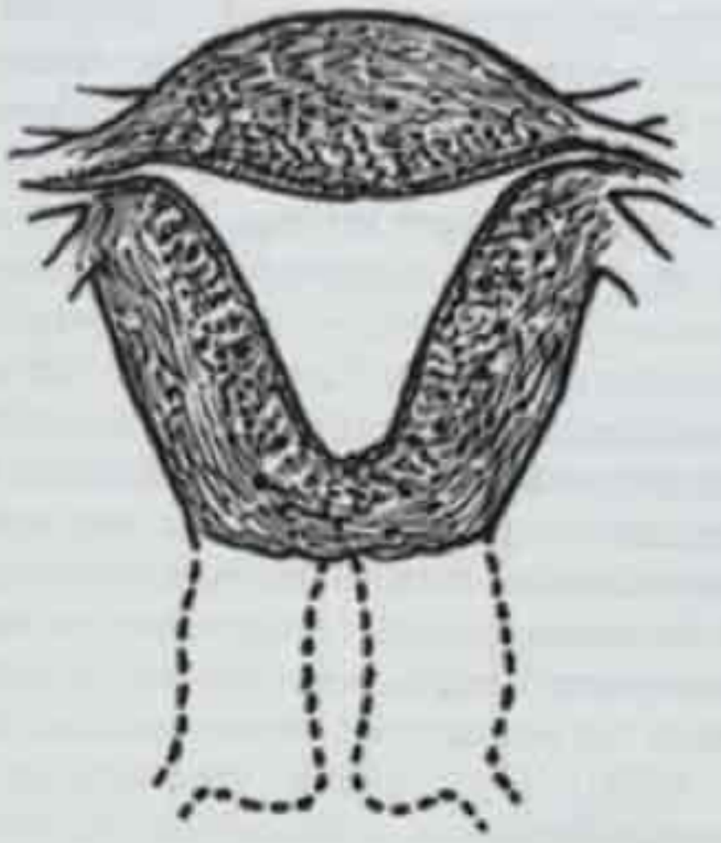


Figure 13. Cervical agenesis[6]

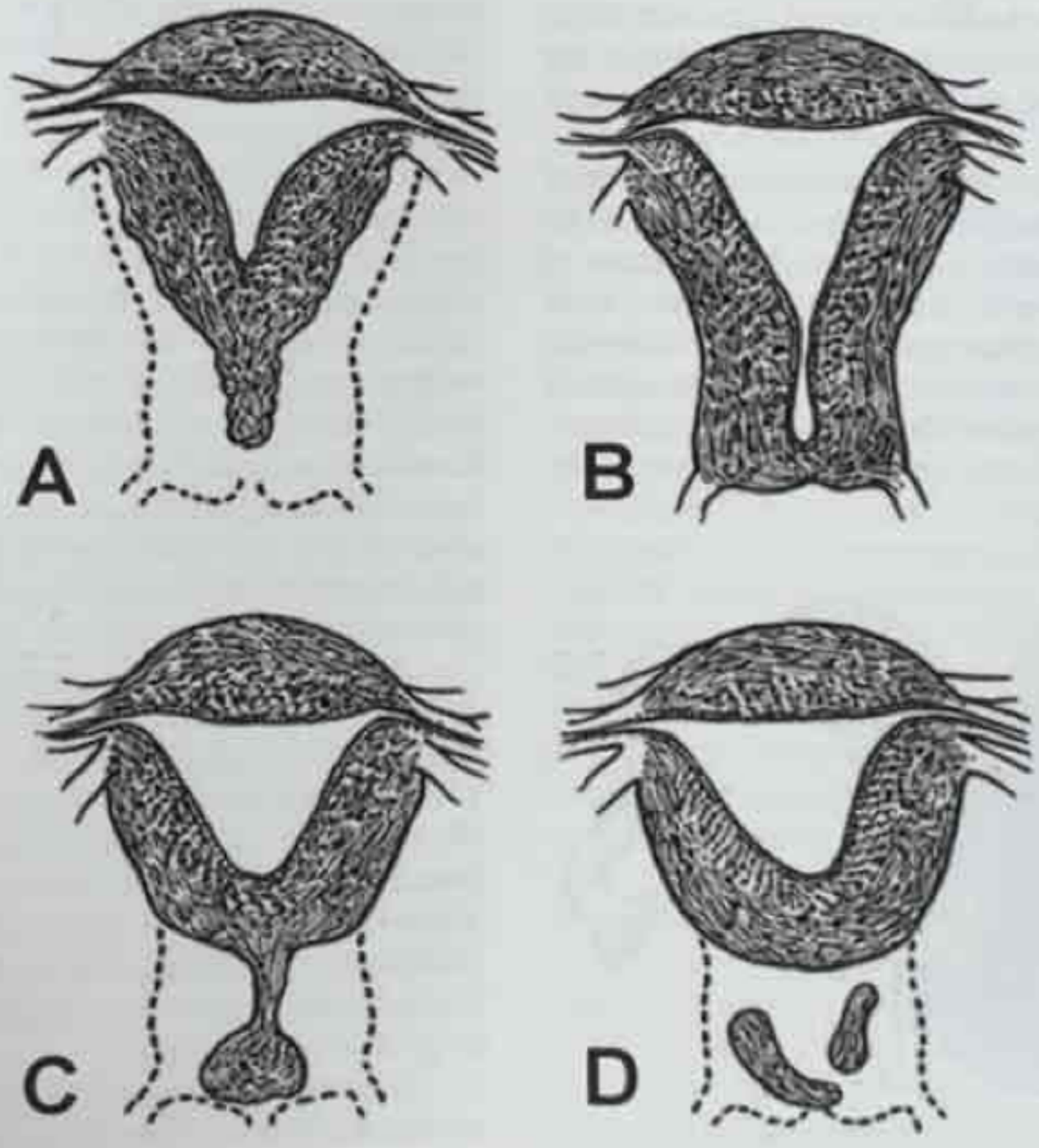


Figure 14. Cervical dysgenesis subtypes[6]

The first case presented is that of cervical agenesis. Although preoperative imaging revealed a hypoplastic cervix by ultrasonography and MRI, intraoperatively, no cervical tissue was identified. On laparoscopy, the inferior border of the uterus was blunt and rounded. The uterosacral ligaments, which normally attach to the upper portion of the cervix posteriorly, did not connect to any structure (Figure 14). Histopathologic examination confirmed the absence of any cervical tissue.

The second patient belongs to the second subtype enumerated under cervical dysplasia. Histopathologic examination of the supposed obstructed vagina revealed only endocervical tissue. Reviewing the first admission, what was identified as a transverse vaginal septum, was in fact the obliterated end of the dysplastic cervix of the right unicornuate uterus. The hematometra was so severe, that it extended caudad, compressing the normal vagina, which led to the assumption that the obstruction was due to a vaginal septum. Simpson reported a similar malformation in 1999, when he described incomplete Mullerian fusion with a blindly ending hemiuterus causing hydrometrocolpos or hydrocolpos (Figure 15).[12] In contrast, the second index patient had a unicornuate uterus with an obstructed cervical os. From both her admissions, the finding of a distended and obstructed vagina on pelvic examination and ultrasonography was actually the expanded endocervical cavity of a distally obstructed dysplastic cervix.

Atresia of the uterine cervix is associated with 52% of cases of partial or complete vaginal agenesis.[9] The incidence of concomitant renal anomalies in cases of congenital cervical atresia is rare, but do occur.[6] Both index patients had normal vaginal anatomy. During her re-admission, however, the second patient was noted to have had vaginal stenosis due to her first surgery, necessitating a vaginoplasty. Abdominal ultrasonography revealed normal kidneys in the first, while an absent right kidney in the second case.

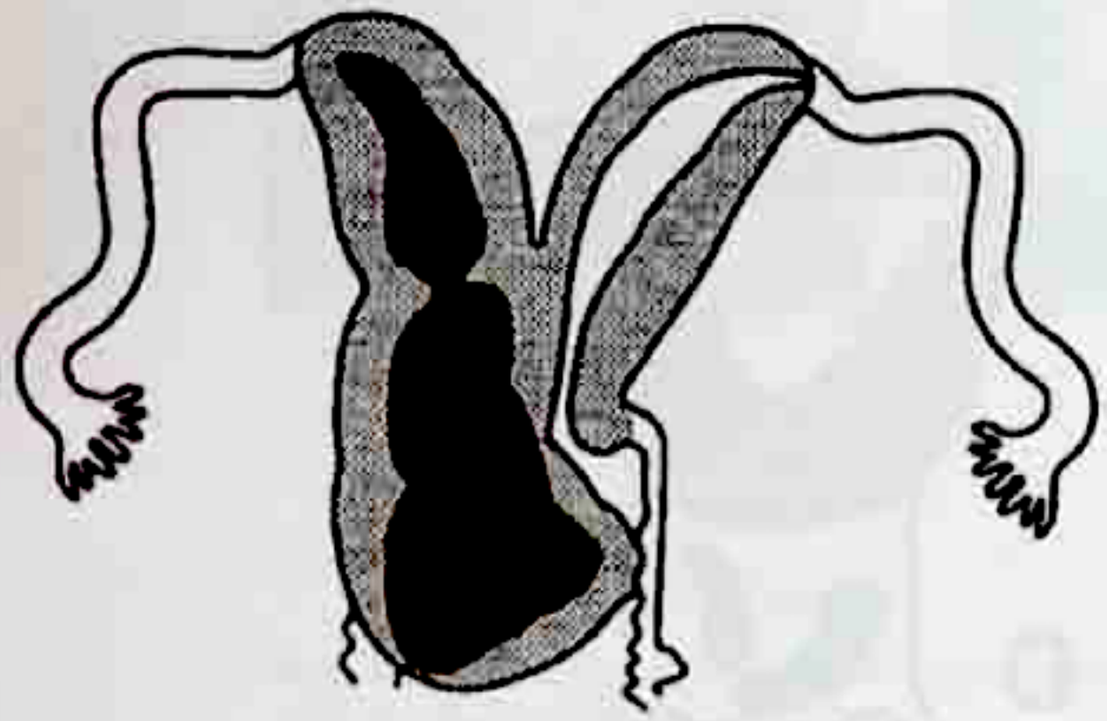


Figure 15. Incomplete Mullerian fusion with obstruction due to blindly ending hemiuterus[12]

Interestingly, the second patient had renal agenesis ipsilateral to her unicornuate uterus. Since a unicornuate uterus is due to the complete agenesis of the organs derived from the contralateral urogenital ridge, typically, it is associated with contralateral renal agenesis or hypoplasia, as well. Ipsilateral renal agenesis with a unicornuate uterus therefore, is a deviation from the expected, and as such, is an extremely rare condition. It may be explained by the abnormal development of organs derived from the unilateral urogenital ridge. Haydardedeoglu[13] published the first case report of unicornuate uterus with ipsilateral renal agenesis in Fertility and Sterility in 2006. The second index patient may very well be the second reported case.

Clinical symptoms of congenital cervical atresia manifest at the time of expected menarche. Any acute or chronic abdominal or genital pain in a pubescent girl must then evoke a high index of suspicion. In a review of 18 cases of uterine cervix atresia, Deffarges, et al.[14] found that 61% complained of cryptomenorrhea, with 2 of the 18 patients presenting as acute abdomen. They reported that 44% had associated hematometra, 22% had severe endometriosis, while only 3 of the 18 patients had no concomitant upper genital tract lesion. Such complications associated with abnormalities of the cervix often necessitate surgical treatment.[8] This was true for both index patients. Both presented with primary amenorrhea with cyclic abdominal pain. For the second case, her re-admission was due to stenosis of the previous operation, presenting as amenorrhea with associated pain. The first index patient did not have any evidence of pelvic endometriosis on laparoscopy, but had adenomyosis on both gross and histopathologic examination. This may have been due to defects in, or obstruction of the fallopian tubes, hindering retrograde menstruation and peritoneal seeding. On the other hand, the second case had a hematosalpinx and severe pelvic endometriosis and pelvic adhesions on laparotomy.

These concomitant lesions are evidence of the chronicity of both patients' conditions. There was a definite delay in the management of these cases. To avoid complications that require aggressive surgery, the diagnosis of uterine cervix anomalies should be made as early as possible.[15] Furthermore, early diagnosis offers significant advantages in patient care, especially with regard to preoperative planning. Establishing congenital cervical malformations, however, presents a diagnostic challenge to gynecologists.[6]

Hysterosalpingography (HSG) is the traditional method used to evaluate the cervical canal, uterine cavity and fallopian tubes. However, its efficacy in diagnosing

anomalies is debatable, and varies greatly with specific types of malformation.[1] HSG has no role in the evaluation of congenital cervical atresia, given the inherent inability to catheterize the cervix. Ultrasonography has been proposed as the initial diagnostic modality for patients with a suspicion of a Mullerian abnormality.[16] It allows a more detailed analysis of the endometrium, uterine cavity and cervix, with a sensitivity of ~44%, depending on the type of malformation, among other factors. Its specificity ranges from 85 to 92%.[1] On evaluation by ultrasonography, blood collection in the uterus and vagina appears as a cystic mass with diffuse low-level echoes. The uterus is differentiated by the thicker myometrial wall and milder distention of the uterine cavity, as compared to the thin and often imperceptible wall of the vagina. The presence or absence of a patent cervix, however, is difficult to document.[17]

On ultrasonography, the first index case was found to have a hypoplastic cervix but with no distinct endocervical canal. Upon laparoscopy, the measured structure was confirmed to be the peritoneum between the proximal end of the vagina and the distal end of the uterine body (Figure 3). On ultrasonography of the second case during both her admissions, the distended endocervical canal was mistaken for an obstructed vagina, owing to the presence of a notch assumed to be the cervix, and the thin wall and caudad extension of the presumed obstructed vagina (Figures 6 & 9). Intraoperatively, however, a normal, albeit compressed vagina was found during her first admission. On histopathologic examination, only endocervical glands and stroma were seen from the presumed vagina. These findings confirm the diagnosis of cervical dysgenesis, of the obstructed type.

Three-dimensional (3-D) ultrasonography has been proposed to be the first line imaging technique for the diagnosis of congenital uterine anomalies. In a review of 8 publications, Alcazar, et al.[18] found that 3-D ultrasonography has a sensitivity of 93% to 100% and a specificity of 85 to 100% in this regard. In retrospect, this could have been offered to both index patients. It is difficult, however, to imagine 3-D ultrasonography producing a significantly different sonologic picture, compared to the two-dimensional ultrasound findings of the patients.

Magnetic resonance imaging (MRI) is said to be the study of choice, as it is the most accurate in diagnosing Mullerian malformations, with a specificity between 96% and 100%.[1] Hematometra is well delineated in MRI, given the preserved zonal anatomy of the uterus, as well as the cervix and endocervical canal, when present. Its multiplanar capability confers a helpful advantage in delineating complex anomalies with marked distortions of the uterovaginal anatomy. In addition, MRI can better characterize secondary processes such as endometriosis and hematosalpinges, compared to ultrasonography.[17]

MRI confirmed the sonologic findings of the first index case. Undoubtedly, the second patient would have benefitted from magnetic resonance imaging, given the complexity of her Mullerian abnormality. Unfortunately, this was not done owing to its high cost, which is the major disadvantage of this imaging modality, and a tangible problem for many patients. When readily available, however, it is ideally done prior to any surgical intervention, as it has limited value in cases of previous surgeries.[19]

It cannot be over-emphasized that both ultrasonography and MRI are more helpful when correlated with findings of a careful and thorough pelvic examination under anesthesia.[8] Despite all efforts, however, in most cases, the nature of uterine outflow obstruction in congenital cervical atresia is confirmed intraoperatively.[8] In that regard, does frozen section have a role in the management? Unfortunately, no report has been published to elucidate this issue.

The aims of treatment of congenital cervical anomalies are firstly, to relieve symptoms related to obstructed menstrual flow, and secondly, to restore fertility and regular menstruation.[20] To date, there has been a lack of uniformity in literature with regard to the first-line surgical treatment of these cases. Given the substantial occurrence of complications, controversy surround conservative surgical management. Not surprisingly, several authors support performing a total hysterectomy, especially for a patient with a functional endometrium and congenital cervical agenesis. A hysterectomy eliminates the problem of cryptomenorrhea, sepsis, endometriosis and the need for repeated surgeries. If performed early enough, the ovaries are spared and its function preserved.[8]

The first index patient underwent a total hysterectomy, when the cervix was confirmed to be absent intraoperatively. As recommended, she had the procedure done by operative laparoscopy.[1] While there is light in offering a conservative surgical procedure for her, the best, individualized treatment option was definitive. The patient had symptoms for 2 years prior to the operation. She had poor access to health care, requiring considerable efforts to avail quality professional medical attention. She was from a distant, rural, low-income community, and was prone to be lost to follow-up. In addition, a total hysterectomy spared her from the risk of multiple surgeries.

The advantages of a total hysterectomy are clear and numerous, but the problem of sterility in these patients remains. With recent advancements in reproductive medicine and (laparoscopic) surgical techniques, conservative management is a possibility, and perhaps may be the first-line treatment option for patients with

anomalies is debatable, and varies greatly with specific types of malformation.[1] HSG has no role in the evaluation of congenital cervical atresia, given the inherent inability to catheterize the cervix. Ultrasonography has been proposed as the initial diagnostic modality for patients with a suspicion of a Mullerian abnormality.[16] It allows a more detailed analysis of the endometrium, uterine cavity and cervix, with a sensitivity of ~44%, depending on the type of malformation, among other factors. Its specificity ranges from 85 to 92%.[1] On evaluation by ultrasonography, blood collection in the uterus and vagina appears as a cystic mass with diffuse low-level echoes. The uterus is differentiated by the thicker myometrial wall and milder distention of the uterine cavity, as compared to the thin and often imperceptible wall of the vagina. The presence or absence of a patent cervix, however, is difficult to document.[17]

On ultrasonography, the first index case was found to have a hypoplastic cervix but with no distinct endocervical canal. Upon laparoscopy, the measured structure was confirmed to be the peritoneum between the proximal end of the vagina and the distal end of the uterine body (Figure 3). On ultrasonography of the second case during both her admissions, the distended endocervical canal was mistaken for an obstructed vagina, owing to the presence of a notch assumed to be the cervix, and the thin wall and caudad extension of the presumed obstructed vagina (Figures 6 & 9). Intraoperatively, however, a normal, albeit compressed vagina was found during her first admission. On histopathologic examination, only endocervical glands and stroma were seen from the presumed vagina. These findings confirm the diagnosis of cervical dysgenesis, of the obstructed type.

Three-dimensional (3-D) ultrasonography has been proposed to be the first line imaging technique for the diagnosis of congenital uterine anomalies. In a review of 8 publications, Alcazar, et al.[18] found that 3-D ultrasonography has a sensitivity of 93% to 100% and a specificity of 85 to 100% in this regard. In retrospect, this could have been offered to both index patients. It is difficult, however, to imagine 3-D ultrasonography producing a significantly different sonologic picture, compared to the two-dimensional ultrasound findings of the patients.

Magnetic resonance imaging (MRI) is said to be the study of choice, as it is the most accurate in diagnosing Mullerian malformations, with a specificity between 96% and 100%.[1] Hematometra is well delineated in MRI, given the preserved zonal anatomy of the uterus, as well as the cervix and endocervical canal, when present. Its multiplanar capability confers a helpful advantage in delineating complex anomalies with marked distortions of the uterovaginal anatomy. In addition, MRI can better characterize secondary processes such as endometriosis and hematosalpinges, compared to ultrasonography.[17]

MRI confirmed the sonologic findings of the first index case. Undoubtedly, the second patient would have benefitted from magnetic resonance imaging, given the complexity of her Mullerian abnormality. Unfortunately, this was not done owing to its high cost, which is the major disadvantage of this imaging modality, and a tangible problem for many patients. When readily available, however, it is ideally done prior to any surgical intervention, as it has limited value in cases of previous surgeries.[19]

It cannot be over-emphasized that both ultrasonography and MRI are more helpful when correlated with findings of a careful and thorough pelvic examination under anesthesia.[6] Despite all efforts, however, in most cases, the nature of uterine outflow obstruction in congenital cervical atresia is confirmed intraoperatively.[6] In that regard, does frozen section have a role in the management? Unfortunately, no report has been published to elucidate this issue.

The aims of treatment of congenital cervical anomalies are firstly, to relieve symptoms related to obstructed menstrual flow, and secondly, to restore fertility and regular menstruation.[20] To date, there has been a lack of uniformity in literature with regard to the first-line surgical treatment of these cases. Given the substantial occurrence of complications, controversy surround conservative surgical management. Not surprisingly, several authors support performing a total hysterectomy, especially for a patient with a functional endometrium and congenital cervical agenesis. A hysterectomy eliminates the problem of cryptomenorrhea, sepsis, endometriosis and the need for repeated surgeries. If performed early enough, the ovaries are spared and its function preserved.[6]

The first index patient underwent a total hysterectomy, when the cervix was confirmed to be absent intraoperatively. As recommended, she had the procedure done by operative laparoscopy.[1] While there is light in offering a conservative surgical procedure for her, the best, individualized treatment option was definitive. The patient had symptoms for 2 years prior to the operation. She had poor access to health care, requiring considerable efforts to avail quality professional medical attention. She was from a distant, rural, low-income community, and was prone to be lost to follow-up. In addition, a total hysterectomy spared her from the risk of multiple surgeries.

The advantages of a total hysterectomy are clear and numerous, but the problem of sterility in these patients remains. With recent advancements in reproductive medicine and (laparoscopic) surgical techniques, conservative management is a possibility, and perhaps may be the first-line treatment option for patients with

anomalies is debatable, and varies greatly with specific types of malformation.[1] HSG has no role in the evaluation of congenital cervical atresia, given the inherent inability to catheterize the cervix. Ultrasonography has been proposed as the initial diagnostic modality for patients with a suspicion of a Mullerian abnormality.[16] It allows a more detailed analysis of the endometrium, uterine cavity and cervix, with a sensitivity of ~44%, depending on the type of malformation, among other factors. Its specificity ranges from 85 to 92%.[1] On evaluation by ultrasonography, blood collection in the uterus and vagina appears as a cystic mass with diffuse low-level echoes. The uterus is differentiated by the thicker myometrial wall and milder distention of the uterine cavity, as compared to the thin and often imperceptible wall of the vagina. The presence or absence of a patent cervix, however, is difficult to document.[17]

On ultrasonography, the first index case was found to have a hypoplastic cervix but with no distinct endocervical canal. Upon laparoscopy, the measured structure was confirmed to be the peritoneum between the proximal end of the vagina and the distal end of the uterine body (Figure 3). On ultrasonography of the second case during both her admissions, the distended endocervical canal was mistaken for an obstructed vagina, owing to the presence of a notch assumed to be the cervix, and the thin wall and caudad extension of the presumed obstructed vagina (Figures 6 & 9). Intraoperatively, however, a normal, albeit compressed vagina was found during her first admission. On histopathologic examination, only endocervical glands and stroma were seen from the presumed vagina. These findings confirm the diagnosis of cervical dysgenesis, of the obstructed type.

Three-dimensional (3-D) ultrasonography has been proposed to be the first line imaging technique for the diagnosis of congenital uterine anomalies. In a review of 8 publications, Alcazar, et al.[18] found that 3-D ultrasonography has a sensitivity of 93% to 100% and a specificity of 85 to 100% in this regard. In retrospect, this could have been offered to both index patients. It is difficult, however, to imagine 3-D ultrasonography producing a significantly different sonologic picture, compared to the two-dimensional ultrasound findings of the patients.

Magnetic resonance imaging (MRI) is said to be the study of choice, as it is the most accurate in diagnosing Mullerian malformations, with a specificity between 96% and 100%.[1] Hematometra is well delineated in MRI, given the preserved zonal anatomy of the uterus, as well as the cervix and endocervical canal, when present. Its multiplanar capability confers a helpful advantage in delineating complex anomalies with marked distortions of the uterovaginal anatomy. In addition, MRI can better characterize secondary processes such as endometriosis and hematosalpinges, compared to ultrasonography.[17]

MRI confirmed the sonologic findings of the first index case. Undoubtedly, the second patient would have benefitted from magnetic resonance imaging, given the complexity of her Mullerian abnormality. Unfortunately, this was not done owing to its high cost, which is the major disadvantage of this imaging modality, and a tangible problem for many patients. When readily available, however, it is ideally done prior to any surgical intervention, as it has limited value in cases of previous surgeries.[19]

It cannot be over-emphasized that both ultrasonography and MRI are more helpful when correlated with findings of a careful and thorough pelvic examination under anesthesia.[6] Despite all efforts, however, in most cases, the nature of uterine outflow obstruction in congenital cervical atresia is confirmed intraoperatively.[9] In that regard, does frozen section have a role in the management? Unfortunately, no report has been published to elucidate this issue.

The aims of treatment of congenital cervical anomalies are firstly, to relieve symptoms related to obstructed menstrual flow, and secondly, to restore fertility and regular menstruation.[20] To date, there has been a lack of uniformity in literature with regard to the first-line surgical treatment of these cases. Given the substantial occurrence of complications, controversy surround conservative surgical management. Not surprisingly, several authors support performing a total hysterectomy, especially for a patient with a functional endometrium and congenital cervical agenesis. A hysterectomy eliminates the problem of cryptomenorrhea, sepsis, endometriosis and the need for repeated surgeries. If performed early enough, the ovaries are spared and its function preserved.[6]

The first index patient underwent a total hysterectomy, when the cervix was confirmed to be absent intraoperatively. As recommended, she had the procedure done by operative laparoscopy.[1] While there is light in offering a conservative surgical procedure for her, the best, individualized treatment option was definitive. The patient had symptoms for 2 years prior to the operation. She had poor access to health care, requiring considerable efforts to avail quality professional medical attention. She was from a distant, rural, low-income community, and was prone to be lost to follow-up. In addition, a total hysterectomy spared her from the risk of multiple surgeries.

The advantages of a total hysterectomy are clear and numerous, but the problem of sterility in these patients remains. With recent advancements in reproductive medicine and (laparoscopic) surgical techniques, conservative management is a possibility, and perhaps may be the first-line treatment option for patients with

cervical atresia.[7] Whether or not decidedly advantageous, however, a thorough discussion on post-operative complications and treatment outcomes is of utmost importance, and absolutely imperative.

Conservative surgical interventions may be one of the following: uterovaginal canalization, uterovaginal anastomosis, or cervical reconstruction. Canalization has been proposed as the main treatment option in cases of cervical obstruction. It involves the direct drilling and sounding of the cervix.[8] Often, it entails the forced formation of a canal, and as such, is prone to stricture of the reconstructed tract.[21] The lack of normal endocervical glandular function has been implicated as the most important contributing factor to stenosis.[5] In 2010, Rock, et al.[22] reported that canalization is successful only among patients with sufficient rudimentary cervical tissue. Only patients with a well formed cervical body, with at least a palpable cord or only distal obstruction achieved successful surgical outcomes. Despite being complicated by high failure rates and poor functional results, however, canalization remains to be a popular first-line surgical intervention.

Uterovaginal anastomosis restores the genital tract by directly suturing the vagina to the uterine isthmus.[5] The surgical technique entails exposing the vagina by complete dissection of the rectouterine and vesicouterine space, allowing circumferential anastomosis of the vagina to the lower uterine segment. In cases of cervical dysplasia, the atretic cervix is removed completely.[6] In the 2001 publication of Deffarges, et al.[14] on the long-term follow-up of 18 women with uterine cervix atresia who underwent open uterovaginal anastomosis, they recommended that anastomosis be preferred over canalization techniques. In their study, only one patient had secondary stenosis of the anastomosis, and none of the patients underwent a hysterectomy. Ten of their patients had a pregnancy, four of which resulted in a total of six successful spontaneous pregnancies. Recently, laparoscopic uterovaginal anastomosis has been reported successful. In 2006, Creighton, et al.[23] were first to describe their technique, advocating conservative laparoscopic surgery as first-line treatment for cervical agenesis. In 2009, Darai, et al.[24] confirmed the feasibility of this procedure, done without fundal incision, citing decreased risk of intrauterine adhesions and of uterine rupture during pregnancy as significant advantages.

Cervical reconstruction attempts to restore the functional anatomy of the genital tract with the creation of a neocervix. It involves the use of grafts that serve the role of the cervix. Many kinds have been reported, including slim thickness skin grafts, full thickness skin grafts, bladder mucosa graft and saphenous vein graft.[8] These, however, are limited to case reports only.

Our second index patient had 2 failed attempts to remedy her obstructed menstrual flow, before she eventually underwent a total hysterectomy. The surgery done in Lucena may have predisposed the subsequent stenosis of the first operation performed in the tertiary training hospital by contributing to the formation of scarring and fibrosis in the area. However, other causal factors definitely added to the consequent complication that ensued. Despite the failure to recognize the cervical dysgenesis in the case, a successful conduit between the uterine cavity and the vagina was achieved. The canalization allowed for monthly menses reported for 10 months after the operation. In addition, the patient experienced no ailment relating to an ascending infection, during the period between admissions.

As suggested by Bugmann, et al.[25] several other factors could have influenced the eventual poor outcome seen in the patient. These include the size of the created channel, the duration of stenting of the created channel, the presence of residual endocervical glands in the proximity of the created channel, the absence of an added epithelial lining, the presence of normal vaginal tissue adjacent to the distal end of the created channel, and the number of menses allowed to flow through the stented channel. Equally important are the patient's compliance with manual dilatation of the canal, and the observance of prompt and long-term follow-up, both of which the patient failed to do.

Nevertheless, stenosis of the reconstructed canal is an inherent and foreseeable complication of the conservative surgical management of congenital cervical atresia, among many others. In addition, complications of such procedures are not insignificant. They include endometritis, pelvic inflammatory disease, persistent pelvic pain, and bowel or bladder injury.[6] In fact, stenosis usually occurs, and further operations should be anticipated.[26] In 2004, Bedner, et al.[27] reported that of 5 patients with congenital cervicovaginal agenesis who underwent uterovaginal anastomosis, 3 (60%) had stenosis of the canal at the uterovaginal junction, 3 to 8 months after surgery. The patients had to undergo an additional operation to restore canal patency. Rock, et al.[28] reviewed 21 cases of cervical malformation who underwent surgical interventions and found that of the 14 patients with an absent or fragmented cervix, 10 had a primary hysterectomy, while 4 had cervical reconstruction. All 4 patients, however, ultimately had a hysterectomy, due to complications post-operatively. In literature[29-31], three deaths have been reported as a result of severe sepsis following a uterovaginal canalization procedure. The first death was reported in 1973 by Greary and Weed[29], the second in 1980 by Niver, et al.[30], and the third in 1997 by Casey and

Laufer.[11] Thus, it is not surprising that total hysterectomy is still supported by several authors. Especially when maintaining menstrual flow fails or abdominal pain persists after a uterovaginal canalization, hysterectomy is the logical endpoint.[3] Conservative surgical management, therefore, should be reserved for well-selected patients, taking into strict consideration their anatomical findings and treatment preference. If not, young girls may be subjected to multiple surgical procedures with no good evidence of success.

In 2011, Roberts and Rock[6] proposed a guideline for the surgical treatment of congenital cervical atresia, based on cervical anatomical categories (Table 2). According to them, the options for the first index patient, are 1) removal of the uterine fundus, or 2) anastomosis of the lower uterine segment to the vaginal epithelium and stroma. For the second case, they recommended 1) to create a neocervical canal using a drilling or coring technique, with optional grafting of the endocervical canal, or 2) to remove the cervix and perform a uterovaginal anastomosis. Based on their guidelines, therefore, both index patients were managed appropriately. The case of cervical agenesis underwent an outright total hysterectomy, while the case of cervical dysgenesis consented to a total hysterectomy after 2 failed attempts to create a uterovaginal canal.

The decision to proceed with a hysterectomy, or attempt a conservative surgical procedure for cases of congenital cervical atresia is indeed controversial. Despite the reported innovative and advanced surgical techniques that have proven successful and beneficial, the risks and complications of conservative management remain substantial and quite alarming. A comprehensive treatment plan should be devised, after a thorough examination, and in-depth preoperative counseling. Ultimately, the patient's preference, the specific anatomical findings, the availability of a surgeon with expertise in the field of reconstructive surgery, and the assurance of long-term follow-up should be considered.

Once a decision to perform a hysterectomy is made, however, the psychological impact on, and reproductive future of the patients of congenital cervical atresia should not be overlooked. During their adolescent years, when their conditions first manifest, these patients may not fully comprehend the totality of their condition yet. Preoperative psychotherapy is valuable in this respect. With regard to their curtailed childbearing potential, it is important that alternative means of building a family, like adoption, perhaps surrogacy, be made available to them in the future. With adequate counseling, long-term follow-up, and sustained psychosocial support, patients with congenital cervical atresia can live a full and normal life.

Conclusion

Two cases of congenital cervical atresia were presented. The first was a case of cervical agenesis, while the second belonged to a subtype of cervical dysgenesis, with an intact cervical body but obstructed cervical os. While the first underwent an outright total hysterectomy, the second patient initially underwent conservative surgical procedures before subsequently undergoing a hysterectomy.

Congenital cervical atresia is an uncommon Mullerian anomaly, and as such, there are challenges to its accurate and early diagnosis. Its first-line surgical management, whether definitive or conservative, has been the topic of much debate and controversy. Each has its own set of advantages and disadvantage to be considered. In the end, however, treatment is tailored to the individual, as illustrated by the cases presented.

References

1. Ribeiro SC, Tormena RA, Peterson TV, Gonzales MDO, S PG, de Almeida JAM, et al. Mullerian duct anomalies: review of current management. Sao Paulo Med J 2009; 127(2): 92-6.
2. Grimbizis GF, Campo R. Congenital malformations of the female genital tract: the need for a new classification system. Fertil Steril 2010; 94: 401-7.
3. Suganuma N, Furuhashi M, Moriwaki T, Tsukahara S, Ando T, Ishihara Y. Management of missed abortion in a patient with congenital cervical atresia. Fertil Steril 2002; 77(5): 1071-3.
4. American Fertility Society. The AFS classifications of adnexal adhesions, distal tubal occlusion, tubal occlusion secondary to tubal ligation, tubal pregnancies, Mullerian anomalies and intrauterine adhesions. Fertil Steril 1988; 49: 944-55.
5. Lai T-H, Wu M-H, Hung K-H, Cheng Y-C, Chang F-M. Successful pregnancy by transmyometrial and transtubal embryo transfer after IVF in a patient with congenital cervical atresia who underwent uterovaginal canalization during Cesarean section. Human Repro 2001; 16(2): 268-71.
6. Roberts CP, Rock JA. Surgical methods in the treatment of congenital anomalies of the uterine cervix. Current Opinion in Obstet Gynecol 2011; 23: 251-7.
7. Rajaratnam A, D'cunha P, Paul N, Ray A. Cervical aplasia. J Clin Diag Res 2011; 5(8): 1672-3.
8. Grimbizis GF, Tsalikis T, Mikos T, Papadopoulos N, Tarlatzis BC, Bontis JN. Succesful end-to-end cervico-cervical anastomosis in a patient with congenital cervical fragmentation: Case report. Hum Reprod 2004; 19(5): 1204-10.
9. Fujimoto VY, Miller JH, Klein NA, Soules MR. Congenital cervical atresia: Report of seven cases and review of literature. Am J Obstet Gynecol 1997; 177: 1419-25.
10. Estole-Casanova LA, Oblepias EG. What is the essence of a younf hysterectomized woman? A rare case of congenital cervical atresia. Philipp J Repro Endo Inf 2007; 4(2): 33-40.
11. Batto SKT, Gorgonio N. Cervical agenesis: A case report. Philipp J Repro Endo Inf 2012; 9(1): 31-9.
12. Simpson JL. Genetics of the female reproductive ducts. Am J Med Genet (Semin Med Genet) 1999; 89: 224-39.

13. Haydardedeoglu B, Simsek E, Kilicdag EB, Tarim E, Aslan E, Bagis T. A case of unicornuate uterus with unilateral ovarian and renal agenesis. Fertil Steril 2006; 85(3): 750e1-4.
14. Deffarges JV, Haddad B, Musset R, Paniel BJ. Utero-vaginal anastomosis in women with uterine cervix atresia: long-term follow-up and reproductive performance. A study of 18 cases. Human Repro 2001; 16(8): 1722-5.
15. Muddana A, Pawar A, Algotar KM, Shyamsunder, Daver RG. An unusual case of cryptomenorrhea. Bombay Hosp J 2008; 50(1): 113-5.
16. Marten K, Vosshenrish R, Funke M, Obenauer S, Baum F, Grabbe E. MRI in the evaluation of mullerian duct anomalies. J Clin Imaging 2003; 27: 346-50.
17. Troiano RN, McCarthy SM. Mullerian duct anomalies: imaging and clinical issues. Radiology 2004; 233(1): 19-34.
18. Alcazar JL, Auba M, Olartecoechea B. Three-dimensional ultrasound in gynecological clinical practice. Reports in Medical Imaging 2012; 5: 1-13.
19. Lang IM, et al . MR imaging of pediatric utero-vaginal anomalies. Pediatr Radiol 1999; 29: 163-70.
20. Anttila L, Penttila T-A, Suikkari A-M. Successful pregnancy after in-vitro fertilization and transmyometrial embryo transfer in a patient with congenital atresia of cervix. Hum Reprod 1999; 14(6): 1647-9.
21. Prorocic M, Vasiljevic M. Effective uterovaginal anastomosis in patient with congenital atresia of cervix uteri – a case report. Singapore J Obstet Gynecol 2006; 37(2): 112-4.
22. Rock JA, Roberts CP, Jones HWL. Congenital anomalies of the uterine cervix: lessons from 30 cases managed clinically by a common protocol. Fertil Steril 2010; 94: 1858-63.
23. Creighton SM, Davies MC, Cutner A. Laparoscopic management of cervical agenesis. Fertil Steril 2006; 85(5): 1510e13-15.
24. Darai E, Ballester M, Bazot M, Paniel B-J. Laparoscopic-assisted uterovaginal anastomosis for uterine cervix atresia with partial vaginal aplasia. J Minimally Invasive Gynecol 2009; 16: 92-4.
25. Bugmann P, Amaudruz M, Hanquinet S, La Scala G, Birraux J, Le Coultre C. Uterocervicoplasty with bladder mucosa layer for the treatment of complete cervical agenesis. Fertil Steril 2002; 77(4): 831-5.
26. Acien P, Acien MI, Quereda F, Santoyo T. Cervicovaginal agenesis: spontaneous gestation at term after previous reimplantation of the uterine corpus in a neovagina: Case report. Hum Reprod 2008; 23(3): 548-53.
27. Bedner R, Rzepka-Gorska I, Blogowska A, Malecha J, Kosmider M. Effects of a surgical treatment of congenital cervicovaginal agenesis. J Pediatr Adolesc Gynecol 2004; 17: 327-30.
28. Rock JA, Carpenter SE, Wheeless CR, Jones Jr. HW. The clinical management of maldevelopment of the uterine cervix. J Pediatr Surg 1995; 1(3): 129-33.
29. Greary W, Weed J. Congenital atresia of the uterine cervix. Obstet Gynecol 1973; 76: 900-1.
30. Niver D, Barrett G, Jewelewicz R. Congenital atresia of the uterine cervix and vagina: three cases. Fertil Steril 1980; 33: 25-9.
31. Casey AC, Laufer MR. Cervical agenesis: Septic death after surgery. Obstet Gynecol 1997; 90: 706-7.

Laufer.[3] Thus, it is not surprising that total hysterectomy is still supported by several authors. Especially when maintaining menstrual flow fails or abdominal pain persists after a uterovaginal canalization, hysterectomy is the logical endpoint.[2] Conservative surgical management, therefore, should be reserved for well-selected patients, taking into strict consideration their anatomical findings and treatment preference. If not, young girls may be subjected to multiple surgical procedures with no good evidence of success.

In 2011, Roberts and Rock[6] proposed a guideline for the surgical treatment of congenital cervical atresia, based on cervical anatomical categories (Table 2). According to them, the options for the first index patient, are 1) removal of the uterine fundus, or 2) anastomosis of the lower uterine segment to the vaginal epithelium and stroma. For the second case, they recommended 1) to create a neocervical canal using a drilling or coring technique, with optional grafting of the endocervical canal, or 2) to remove the cervix and perform a uterovaginal anastomosis. Based on their guidelines, therefore, both index patients were managed appropriately. The case of cervical agenesis underwent an outright total hysterectomy, while the case of cervical dysgenesis consented to a total hysterectomy after 2 failed attempts to create a uterovaginal canal.

The decision to proceed with a hysterectomy, or attempt a conservative surgical procedure for cases of congenital cervical atresia is indeed controversial. Despite the reported innovative and advanced surgical techniques that have proven successful and beneficial, the risks and complications of conservative management remain substantial and quite alarming. A comprehensive treatment plan should be devised, after a thorough examination, and in-depth preoperative counseling. Ultimately, the patient's preference, the specific anatomical findings, the availability of a surgeon with expertise in the field of reconstructive surgery, and the assurance of long-term follow-up should be considered.

Once a decision to perform a hysterectomy is made, however, the psychological impact on, and reproductive future of the patients of congenital cervical atresia should not be overlooked. During their adolescent years, when their conditions first manifest, these patients may not fully comprehend the totality of their condition yet. Preoperative psychotherapy is valuable in this respect. With regard to their curtailed childbearing potential, it is important that alternative means of building a family, like adoption, perhaps surrogacy, be made available to them in the future. With adequate counseling, long-term follow-up, and sustained psychosocial support, patients with congenital cervical atresia can live a full and normal life.

Conclusion

Two cases of congenital cervical atresia were presented. The first was a case of cervical agenesis, while the second belonged to a subtype of cervical dysgenesis, with an intact cervical body but obstructed cervical os. While the first underwent an outright total hysterectomy, the second patient initially underwent conservative surgical procedures before subsequently undergoing a hysterectomy.

Congenital cervical atresia is an uncommon Mullerian anomaly, and as such, there are challenges to its accurate and early diagnosis. Its first-line surgical management, whether definitive or conservative, has been the topic of much debate and controversy. Each has its own set of advantages and disadvantage to be considered. In the end, however, treatment is tailored to the individual, as illustrated by the cases presented.

References

1. Ribeiro SC, Tormena RA, Peterson TV, Gonzales MDO, S PG, de Almeida JAM, et al. Mullerian duct anomalies: review of current management. Sao Paulo Med J 2009; 127(2): 92-6.
2. Grimbizis GF, Campo R. Congenital malformations of the female genital tract: the need for a new classification system. Fertil Steril 2010; 94: 401-7.
3. Suganuma N, Furuhashi M, Moriwaki T, Tsukahara S, Ando T, Ishihara Y. Management of missed abortion in a patient with congenital cervical atresia. Fertil Steril 2002; 77(5): 1071-3.
4. American Fertility Society. The AFS classifications of adnexal adhesions, distal tubal occlusion, tubal occlusion secondary to tubal ligation, tubal pregnancies, Mullerian anomalies and intrauterine adhesions. Fertil Steril 1988; 49: 944-55.
5. Lai T-H, Wu M-H, Hung K-H, Cheng Y-C, Chang F-M. Successful pregnancy by transmyometrial and transtubal embryo transfer after IVF in a patient with congenital cervical atresia who underwent uterovaginal canalization during Cesarean section. Human Repro 2001; 16(2): 268-71.
6. Roberts CP, Rock JA. Surgical methods in the treatment of congenital anomalies of the uterine cervix. Current Opinion in Obstet Gynecol 2011; 23: 251-7.
7. Rajaratnam A, D'cunha P, Paul N, Ray A. Cervical aplasia. J Clin Diag Res 2011; 5(8): 1672-3.
8. Grimbizis GF, Tsalikis T, Mikos T, Papadopoulos N, Tarlatzis BC, Bontis JN. Succesful end-to-end cervico-cervical anastomosis in a patient with congenital cervical fragmentation. Case report. Hum Reprod 2004; 19(5): 1204-10.
9. Fujimoto VY, Miller JH, Klein NA, Soules MR. Congenital cervical atresia: Report of seven cases and review of literature. Am J Obstet Gynecol 1997; 177: 1419-25.
10. Estole-Casanova LA, Oblepias EG. What is the essence of a younf hysterectomized woman? A rare case of congenital cervical atresia. Philipp J Repro Endo Inf 2007; 4(2): 33-40.
11. Batto SKT, Gorgonio N. Cervical agenesis: A case report. Philipp J Repro Endo Inf 2012; 9(1): 31-9.
12. Simpson JL. Genetics of the female reproductive ducts. Am J Med Genet (Semin Med Genet) 1999; 89: 224-39.

13. Haydardedeoglu B, Simsek E, Kilicdag EB, Tarim E, Aslan E, Bagis T. A case of unicornuate uterus with unilateral ovarian and renal agenesis. Fertil Steril 2006; 85(3): 750e1-4.
14. Deffarges JV, Haddad B, Musset R, Paniel BJ. Utero-vaginal anastomosis in women with uterine cervix atresia: long-term follow-up and reproductive performance. A study of 18 cases. Human Repro 2001; 16(8): 1722-5.
15. Muddana A, Pawar A, Algotar KM, Shyamsunder, Daver RG. An unusual case of cryptomenorrhea. Bombay Hosp J 2008; 50(1): 113-5.
16. Marten K, Vosshenrish R, Funke M, Obenauer S, Baum F, Grabbe E. MRI in the evaluation of mullerian duct anomalies. J Clin Imaging 2003; 27: 346-50.
17. Troiano RN, McCarthy SM. Mullerian duct anomalies: imaging and clinical issues. Radiology 2004; 233(1): 19-34.
18. Alcazar JL, Auba M, Olartecoechea B. Three-dimensional ultrasound in gynecological clinical practice. Reports in Medical Imaging 2012; 5: 1-13.
19. Lang IM, et al . MR imaging of pediatric utero-vaginal anomalies. Pediatr Radiol 1999; 29: 163-70.
20. Anttila L, Penttila T-A, Suikkari A-M. Successful pregnancy after in-vitro fertilization and transmyometrial embryo transfer in a patient with congenital atresia of cervix. Hum Reprod 1999; 14(6): 1647-9.
21. Prorocle M, Vasiljevic M. Effective uterovaginal anastomosis in patient with congenital atresia of cervix uteri – a case report. Singapore J Obstet Gynecol 2006; 37(2): 112-4.
22. Rock JA, Roberts CP, Jones HWL. Congenital anomalies of the uterine cervix: lessons from 30 cases managed clinically by a common protocol. Fertil Steril 2010; 94: 1858-63.
23. Creighton SM, Davies MC, Cutner A. Laparoscopic management of cervical agenesis. Fertil Steril 2006; 85(5): 1510e13-15.
24. Darai E, Ballester M, Bazot M, Paniel B-J. Laparoscopic-assisted uterovaginal anastomosis for uterine cervix atresia with partial vaginal aplasia. J Minimally Invasive Gynecol 2009; 16: 92-4.
25. Bugmann P, Amaudruz M, Hanquinet S, La Scala G, Birraux J, Le Coultre C. Uterocervicoplasty with bladder mucosa layer for the treatment of complete cervical agenesis. Fertil Steril 2002; 77(4): 831-5.
26. Acien P, Acien MI, Quereda F, Santoyo T. Cervicovaginal agenesis: spontaneous gestation at term after previous reimplantation of the uterine corpus in a neovagina: Case report. Hum Reprod 2008; 23(3): 548-53.
27. Bedner R, Rzepka-Gorska I, Blogowska A, Malecha J, Kosmider M. Effects of a surgical treatment of congenital cervicovaginal agenesis. J Pediatr Adolesc Gynecol 2004; 17: 327-30.
28. Rock JA, Carpenter SE, Wheeless CR, Jones Jr. HW. The clinical management of maldevelopment of the uterine cervix. J Pediatr Surg 1995; 1(3): 129-33.
29. Greary W, Weed J. Congenital atresia of the uterine cervix. Obstet Gynecol 1973; 76: 900-1.
30. Niver D, Barrett G, Jewelewicz R. Congenital atresia of the uterine cervix and vagina: three cases. Fertil Steril 1980; 33: 25-9.
31. Casey AC, Laufer MR. Cervical agenesis: Septic death after surgery. Obstet Gynecol 1997; 90: 706-7.