

A STUDY ON RECONSTRUCTIVE OPTIONS FOR VAGINAL AGENESIS

*Dissertation submitted in partial fulfillment of the requirements
for the degree of*

M.Ch. (Plastic Surgery) - Branch III



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CHENNAI**

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CERTIFICATE

This is to certify that, this dissertation titled “A STUDY ON RECONSTRUCTIVE OPTIONS FOR VAGINAL AGENESIS”, submitted by Dr. PRABHAKAR. U. Appearing for M.Ch, (plastic surgery) degree examination in August 2007 is a bonafide record of work done by him under my guidance and supervision.

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INTRODUCTION

Congenital absence of vagina occurs in 0.001 to 0.025 percent of population. [1] Incidence is 1:5000 to 1:10000 live births. [2] It is the main noticeable manifestation of Rokitansky sequence. [3] The primary defect is the disturbance in the development of the caudal end of paramesonephric ducts. [4]

With normal development of all secondary sexual characteristics, these females with congenital utero- vaginal agenesis must face the cruel fact of being unable to bear children and enjoy sexual satisfaction, affecting both their mates and themselves. Considering these functional and psychosocial impacts to womanhood, there is no reason for not making any endeavor in pursuing excellent results in neovaginal reconstruction.

The scientific literature indicates that there have been more than 100 methods for vaginal reconstruction and still we are looking for a more effective method. [5] Surgeons often say that if there is more than one operation for a condition then the ideal procedure does not exist. In our study we have tried to compare various techniques and arrive at better understanding of this rare but complicated disease.

AIM OF THE STUDY

This study on reconstructive options for vaginal agenesis was done with following aims.

- 1) To study various clinical features of vaginal agenesis.
- 2) To study the effectiveness of VCUAM classification in the diagnosis and management of vaginal agenesis.
- 3) To study the various reconstructive options and treatment methods for vaginal agenesis.
- 4) To compare the effectiveness and complications of various treatment methods for vaginal agenesis.

REVIEW OF LITERATURE

CLASSIFICATION OF UTEROVAGINAL ANOMALIES

The 1988 American fertility society (AFS) classification of mullerian anomalies is based on the degree of failure of normal uterine development.(Table no 1) vaginal agenesis forms a part of syndrome in the following two categories.[6]

CLASS- I. DYSGENESIS OF THE MULLERIAN DUCTS

This includes agenesis of the uterus and vagina.

CLASS- II. DISORDERS OF VERTICAL FUSION

These are considered to represent faults in the junction between the down-growing mullerian ducts and the up growing derivative of the urogenital sinus. These are characterized by an atretic portion of vagina.

They should be regarded as a transverse vaginal septum and classified as either obstructed or unobstructed. The so-called partial vaginal agenesis with uterus and cervix present is probably a misnomer for a large segment of atretic vagina. Cervical agenesis or Dysgenesis is also included in the group of disorders of vertical fusion.

EMBRYOLOGY

Even in the 3.5 – to 4 mm embryo, it is possible to recognize the bilateral thickenings of the coelomic epithelium known as the gonadal ridges

TABLE-I

AMERICAN FERTILITY SOCIETY CLASSIFICATION OF UTEROVAGINAL ANOMALIES

CLASS I. DYSGENESIS OF THE MULLERIAN DUCTS

CLASS II. DISORDERS OF VERTICAL FUSION OF THE MULLERIAN DUCTS.

A. Transverse vaginal septum

1. Obstructed
2. unobstructed

B. Cervical Agenesis or Dysgenesis

CLASS III. DISORDERS OF LATERAL FUSION OF THE MULLERIAN DUCTS

A. Asymmetric-obstructed disorder of uterus or vagina usually associated with ipsilateral renal agenesis.

1. Unicornuate uterus with a non-communicating rudimentary anlage or horn.
2. Unilateral obstruction of a cavity of a double uterus.
3. Unilateral vaginal obstruction associated with double uterus.

B. Symmetric-unobstructed

1. Didelphic uterus
 - a. Complete longitudinal vaginal septum.
 - b. Partial longitudinal vaginal septum.
 - c. No longitudinal vaginal septum.
2. Septate uterus
 - a. Complete
 1. Complete longitudinal vaginal septum.
 2. Partial longitudinal vaginal septum.
 3. No longitudinal vaginal septum.
 - b. Partial
 1. Complete longitudinal vaginal septum.
 2. Partial longitudinal vaginal septum.
 3. No longitudinal vaginal septum.
3. Bicornuate uterus
 - a. Complete
 1. Complete longitudinal vaginal septum.
 2. Partial longitudinal vaginal septum.
 3. No longitudinal vaginal septum.
 - b. Partial
 1. Complete longitudinal vaginal septum.
 2. Partial longitudinal vaginal septum.
 3. No longitudinal vaginal septum.
4. T-shaped uterine cavity (diethylstilbestrol related)
5. Unicornuate uterus
 - A. With a rudimentary horn
 1. with endometrial cavity.
 - a) Communicating
 - b) Noncommunicating
 2. without endometrial cavity.
 - B. Without a rudimentary horn.

CLASS IV. UNUSUAL CONFIGURATIONS OF VERTICAL-LATERAL FUSION DEFECTS.

TABLE-II

CLASSIFICATION OF MULLERIAN DEFECTS

(Tarry WF 1986 modification of Monie and Sigurdson 1950)

MULLERIAN DEFECT (M)

M0- unilateral system normally formed but unfused or septum retained

M1- vaginal agenesis alone

M2- vaginal uterine agenesis

M3- total mullerian agenesis

M4- mullerian and ovarian agenesis

Each side is graded individually

CLASSIFICATION OF MRKH SYNDROME

[According to Schmid- Tankwald and Hauser (1977) and Duncan et.al. (1979)]

MRKH SYNDROME

Typical

Atypical

MURCS

MURCS- mullerian aplasia, renal aplasia, and cervico thoracic somite dysplasia (association)

ASSOCIATED MALFORMATIONS

Tubes, ovaries, renal system generated and developed

Malformation in the ovary or renal system

Malformation in the skeleton and/ or heart, muscular weakness, renal malformation

VCUAM CLASSIFICATION

(Vagina, cervix, uterus & adnexa associated malformation) (Peter Oppelt et.al.)

VAGINA-(V)

0- normal

1a- partial hymenal atresia

1b- complete hymenal atresia

2a- incomplete septate vagina <50%

2b- complete septate vagina

3- Stenosis of introitus

4- Hypoplasia

5a- unilateral atresia

5b- complete atresia

S1- sinus urogenitalis (deep confluence)

S2- sinus urogenitalis (middle confluence)

S3- sinus urogenitalis (high confluence)

C- Cloacal

+ - other

- unknown

CERVIX (C)

0- normal

1- duplex vagina

2a- unilateral atresia / aplasia

2b - bilateral atresia / aplasia

+ - other

- unknown

UTERUS (U)

- 0- normal
- 1a- arcuate
- 1b- septate <50% of the uterine cavity
- 1c- septate >50% of the uterine cavity
- 2- bicornuate
- 3- hypo plastic uterus
- 4a- unilaterally rudimentary or aplastic
- 4b- bilaterally rudimentary or aplastic
- + - other
- # - unknown

ADNEXA (A)

- 0- normal
- 1a- unilateral tubal malformation, ovaries normal
- 1b- bilateral tubal malformation, ovaries normal
- 2a- unilateral hypoplasia/ gonadal streak (including tubal malformation if appropriate)
- 2b- bilateral hypoplasia/ gonadal streak (including tubal malformation if appropriate)
- 3a- unilateral aplasia
- 3b- bilateral aplasia
- + - other
- # - unknown

ASSOCIATED MALFORMATION (A)

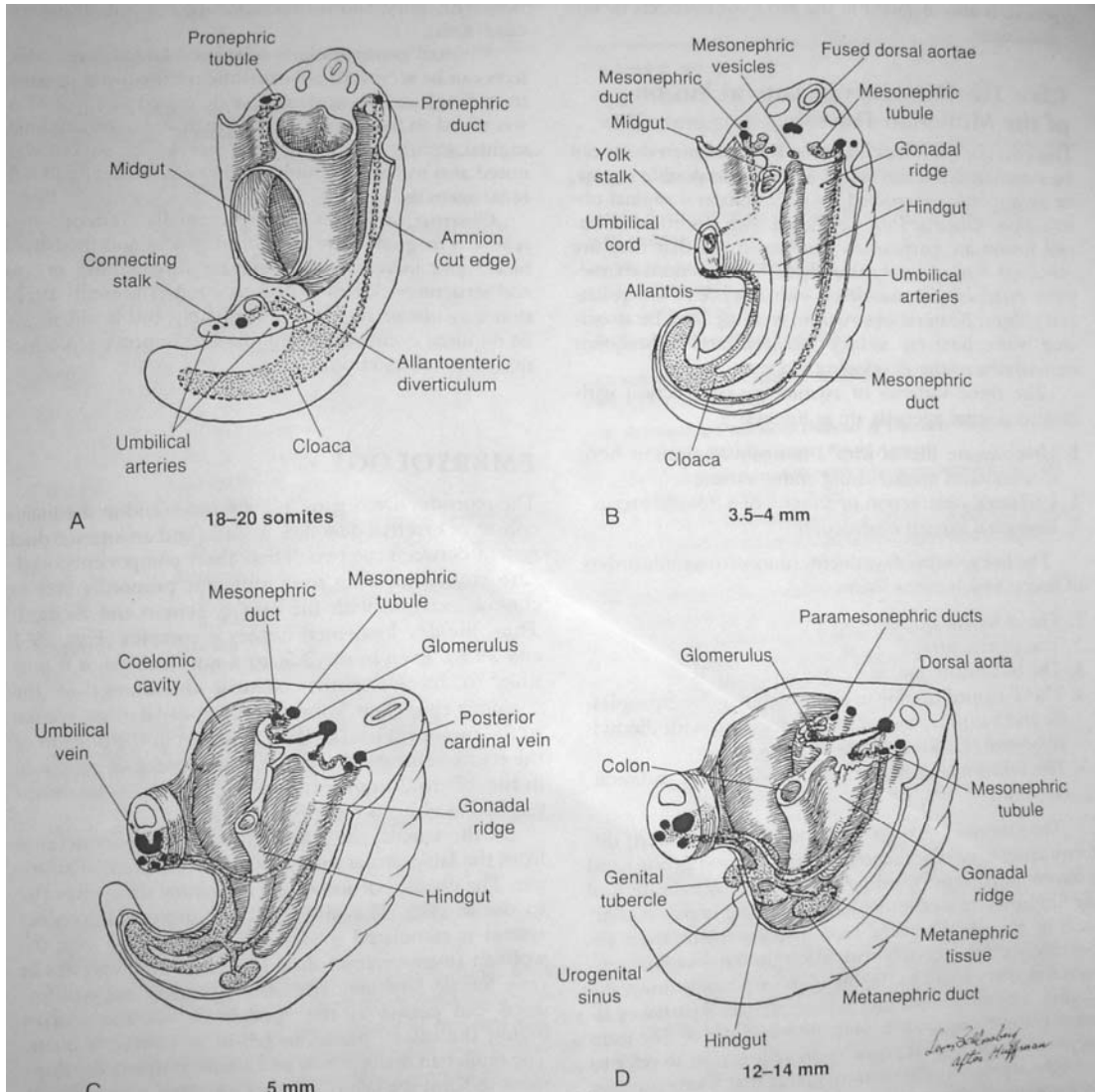
- 0- normal
- R- Renal system
- S- Skeleton
- C- Cardiac
- N- Neurological
- + - other
- # - unknown

medial to the mesonephros (primitive kidney) in the dorsum of the coelomic cavity. At about the 6th week of gestation, in the 17 to 20 mm embryo, the gonad can be distinguished as either a testis or an ovary.

In the female, the labia minora and majora develop from the labioscrotal folds, which are ectodermal in origin. The phallic portion of the urogenital sinus gives rise to the urethra. The mullerian (paramesonephric) duct system is stimulated to develop preferentially over the wolffian (mesonephric) duct system, which regresses in early female fetal life. The cranial parts of the wolffian ducts can persist as the epoophoron of the ovarian hilum; the caudal parts can persist as Gartner's ducts. [7]

About 37 days after fertilization, the mullerian ducts first appear lateral to each Wolffian duct as invagination of the dorsal coelomic epithelium. The site of origin of the invaginations remains open and ultimately forms the fimbriated ends of the fallopian tubes. At their point of origin, each of the mullerian ducts forms a solid bud. Each bud penetrates the mesenchyme lateral and parallel to each of Wolffian duct. As the solid buds elongate, a lumen appears in the cranial part, beginning at each coelomic opening. The lumina extend gradually to the caudal growing tips of the ducts. The paired mullerian ducts continue to grow in a medial and caudal direction until they eventually meet in the midline and become fused

EMBRYOGENESIS OF FEMALE GENITAL TRACT



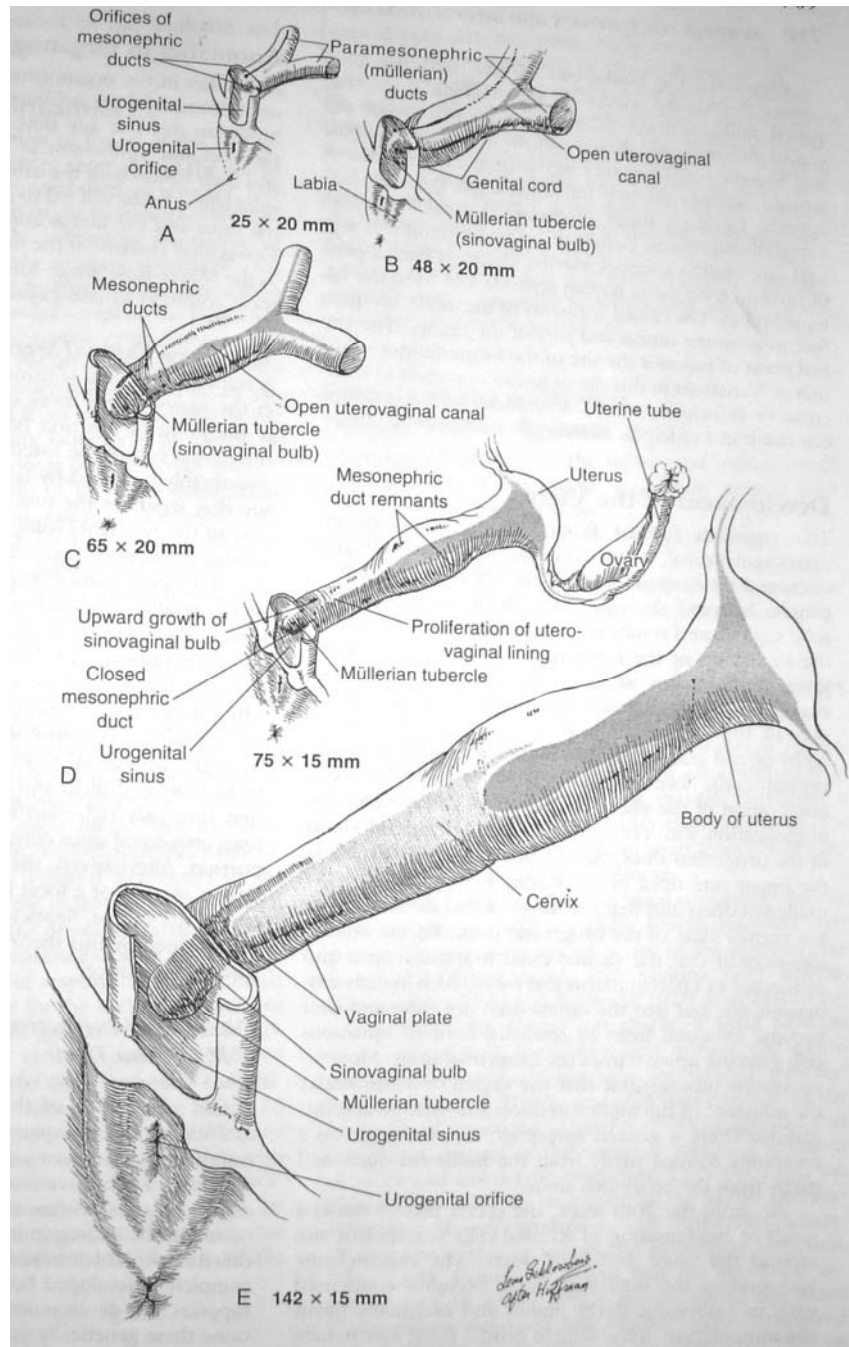
A: 18 TO 20 SOMITE STAGE

B: 3.5 TO 4 MM STAGE

C: 5MM STAGE

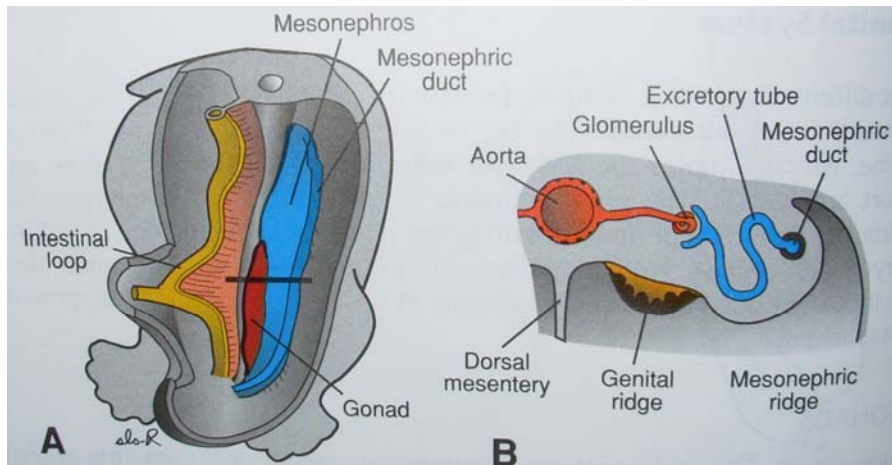
D: 112- 14 MM STAGE

EMBRYOGENESIS OF FEMALE GENITAL TRACT

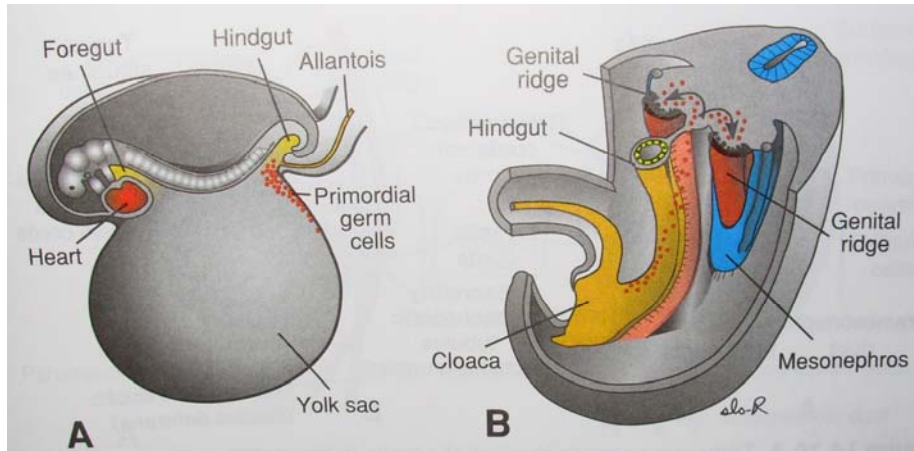


- A: 25 X20 MM STAGE
- B: 48 X20 MM STAGE
- C: 65 X20 MM STAGE
- D: 75 X15 MM STAGE
- E: 142 X15 MM STAGE

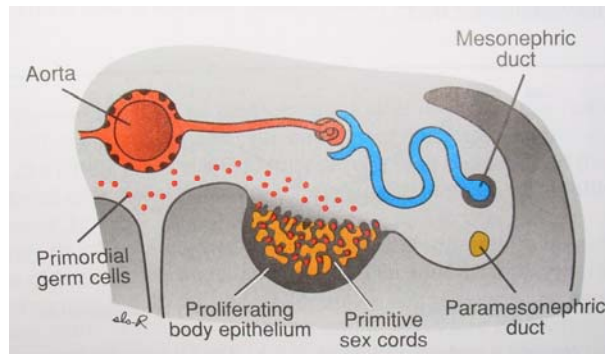
EMBRYOGENESIS OF FEMALE GENITAL TRACT



RELATIONSHIP OF THE GENITAL RIDGE AND THE MESONEPHROS

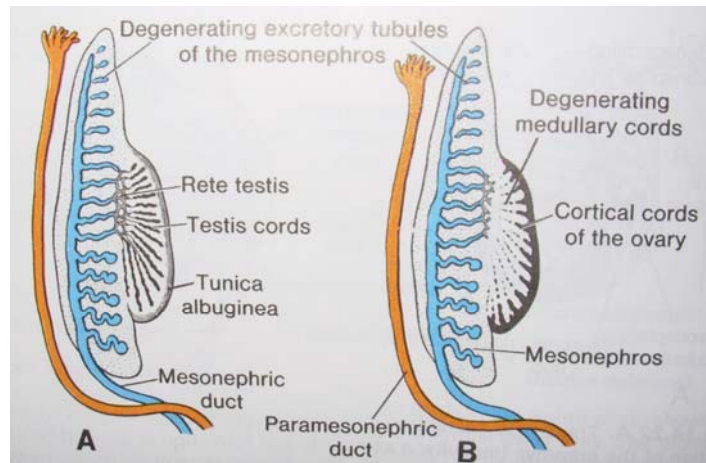


MIGRATIONAL PATH OF THE PRIMORDIAL GERM CELLS ALONG THE WALL OF THE HINDGUT AND THE DORSAL MESENTRY INTO THE GENITAL RIDGE

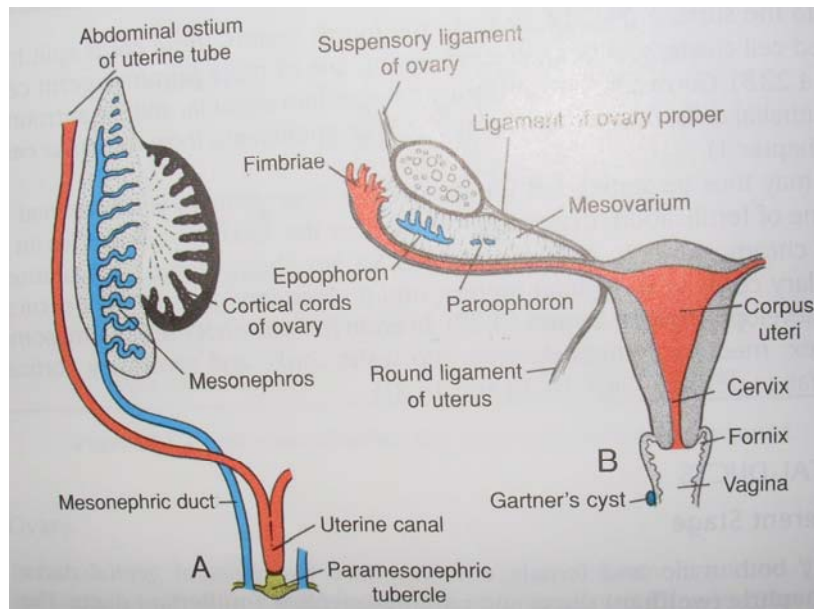


6- WEEK EMBRYO PRIMITIVE GERM CELLS ARE SURROUNDED BY CELLS OF THE PRIMITIVE SEX CORDS

EMBRYOGENESIS OF FEMALE GENITAL TRACT

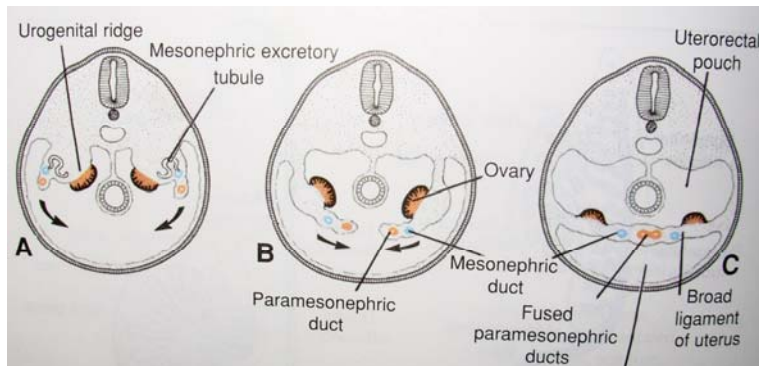


GENITAL DUCTS -6-WEEK EMBRYO A: MALE B: FEMALE

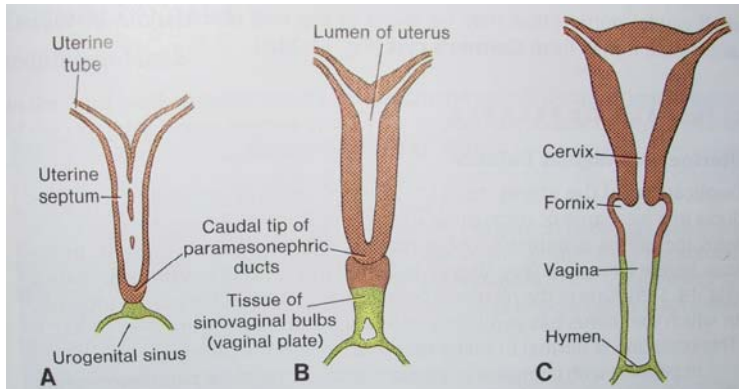


A: GENITAL DUCTS- 2 MONTHS

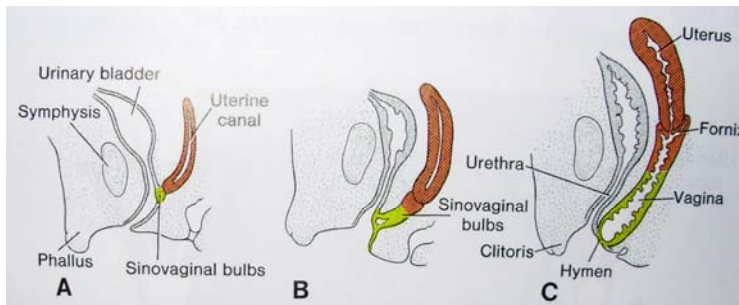
B: GENITAL DUCTS -DESCENT OF THE OVARY



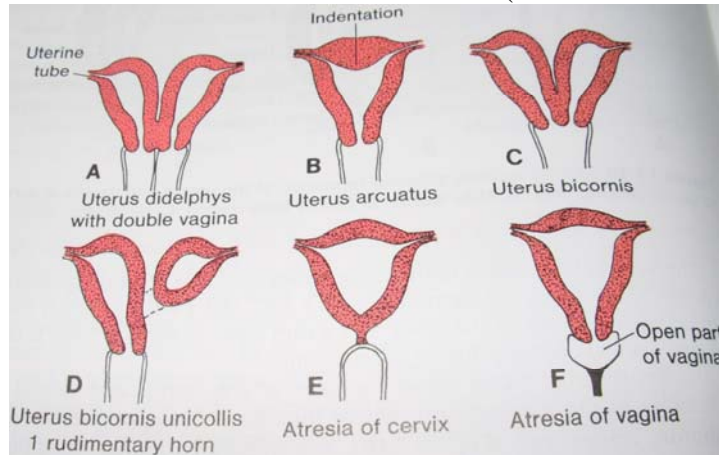
EMBRYOGENESIS OF FEMALE GENITAL TRACT



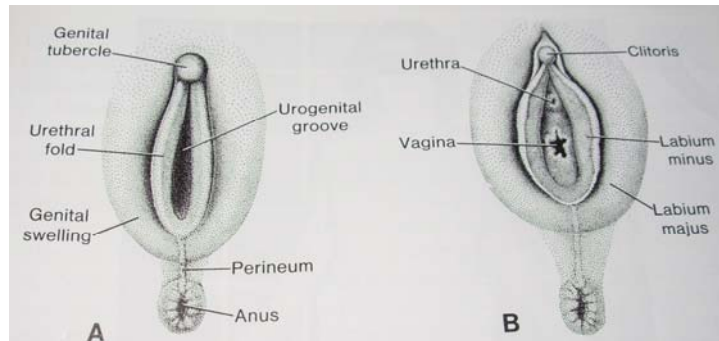
A: 9 WEEKS B: 3 MONTHS C: NEW BORN



FORMATION OF UTERUS & VAGINA (SAGGITAL VIEW)

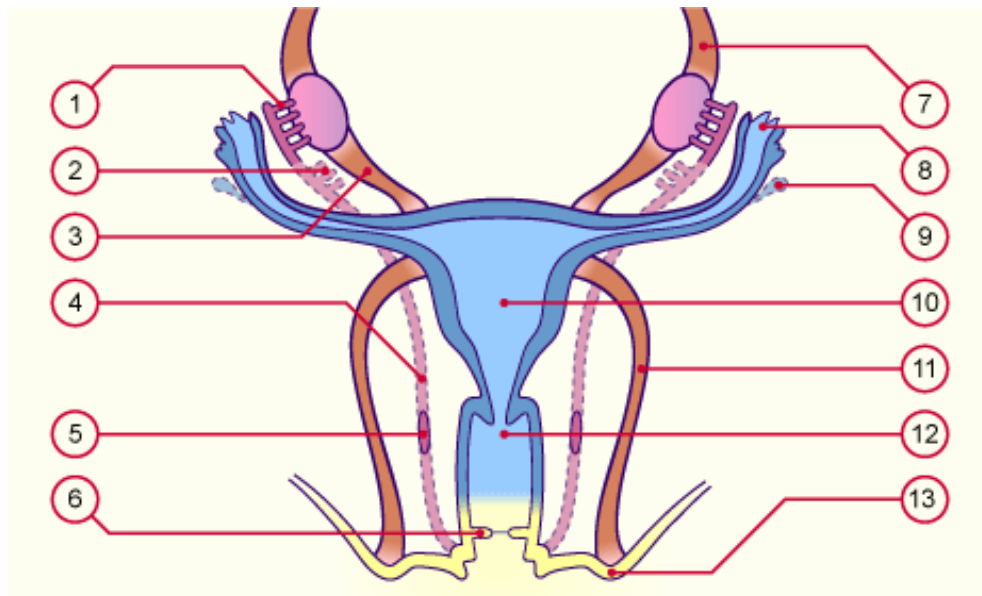
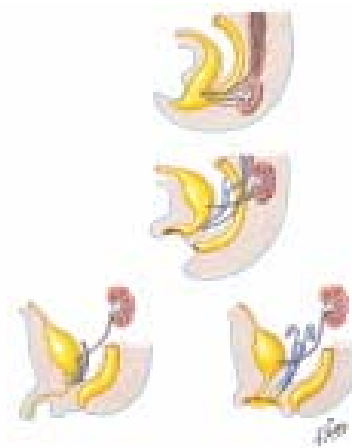
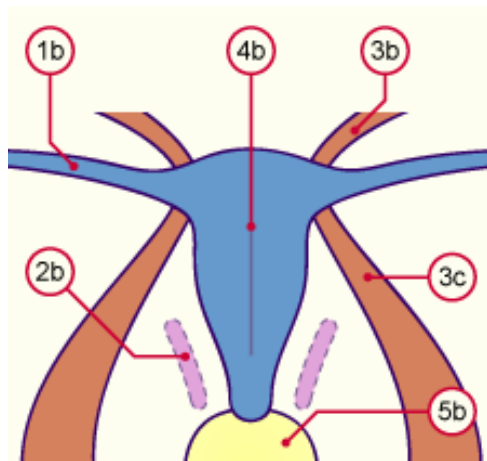
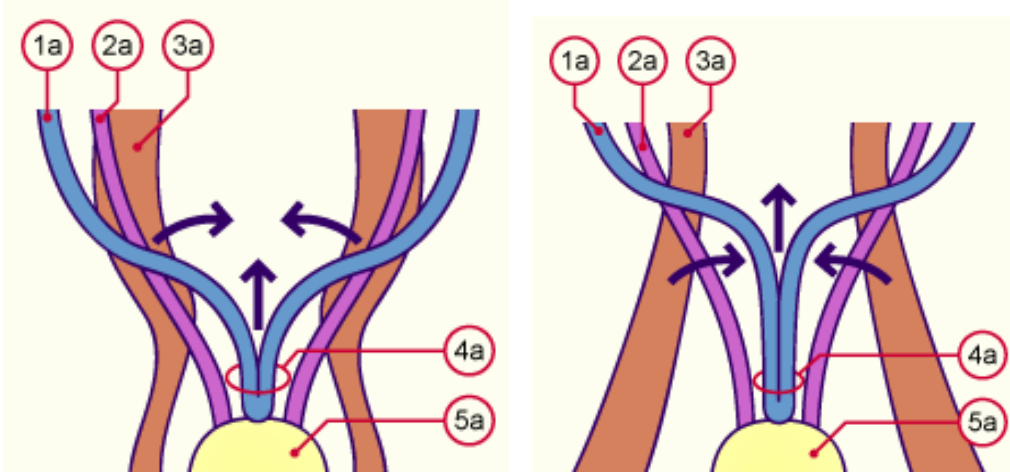


MAIN ABNORMALITIES OF THE UTERUS AND VAGINA



EXTERNAL GENITALIA DEVELOPMENT A: 5 MONTHS B: NEW BORN

EMBRYOLOGY OF MULLERIAN DUCTS



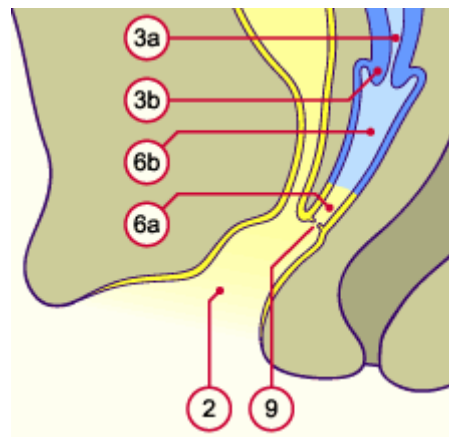
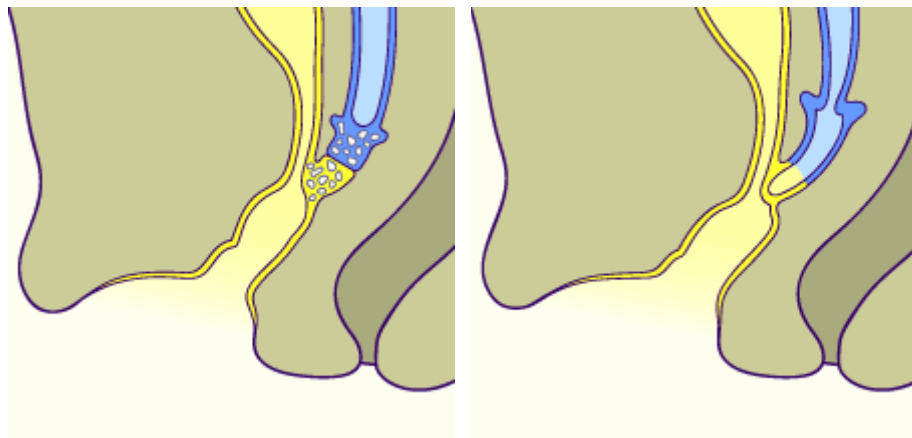
together in the urogenital septum. A septum between the two mullerian ducts gradually disappears, leaving a single uterovaginal canal lined with cuboidal epithelium. The most cranial parts of the mullerian ducts remain separate and form the fallopian tubes. The caudal segments of the mullerian ducts fuse to form the uterus and part of the vagina. [7]

DEVELOPMENT OF THE VAGINA

The vagina is formed from the lower end of the uterovaginal canal, which develops from the mullerian ducts and the urogenital sinus. The point of contact between the two is the mullerian tubercle. A solid vaginal cord results from proliferation of the cells at the caudal tip of the fused mullerian ducts. The cord gradually elongates to meet the bilateral endodermal evaginations (sinovaginal bulbs) from the posterior aspect of the urogenital sinus below. These sinovaginal bulbs extend cranially to fuse with the caudal end of the vaginal cord, forming the vaginal plate. Subsequent canalization of the vaginal cord occurs, followed by epithelialization with cells derived mostly from endoderm of the urogenital sinus. [8]

Recent proposals hold that only the upper one third of the vagina is formed from the mullerian ducts and that the lower vagina develops from the vaginal plates of the urogenital sinus. [7,9] Most investigators now suggest

EMBRYOLOGY OF VAGINA



that the vagina develops under the influence of the mullerian ducts and estrogenic stimulation. [11]

The mesenchyme surrounding the mullerian ducts becomes condensed early in embryonic development and eventually forms the musculature of the female genital tract. The hymen is the embryologic septum between the sinovaginal bulbs above and the urogenital sinus proper below. It is lined by an internal layer of vaginal epithelium and an external layer of epithelium derived from the urogenital sinus (both of endodermal origin), with mesoderm between the two. It is not derived from the mullerian ducts. [7-10]

ANOMALIES IN ORGANOGENESIS OF THE VAGINA

If there is failure in the development of the mullerian ducts at any time between their origin from the coelomic epithelium at 5 weeks of embryonic age and their fusion with the urogenital sinus at 8 weeks, the sinovaginal bulbs will fail to proliferate from the urogenital sinus and the uterus and vagina will fail to develop. Congenital absence of the uterus and the vagina, known as the Mayer-Rokitansky-Kuster-Hauser syndrome, is the most common clinical example of this anomaly. [12-15]

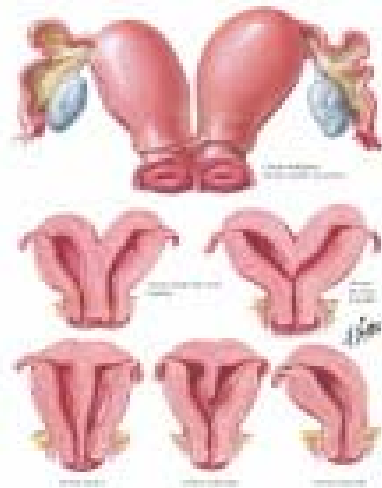
A transverse vaginal septum can develop at the junction between the vaginal plate and the caudal end of the fused mullerian ducts. This defect presumably is caused by failure of absorption of the tissue that separates the

VAGINAL AGENESIS



UTERINE ANOMALIES

TRANSVERSE VAGINAL SEPTUM



two or by failure of complete fusion of the two embryologic components of the vagina. A large segment of vagina can be atretic. In past reviews, this has been termed partial vaginal agenesis with a uterus present [16]. They have a low incidence of associated urinary tract anomalies.

Ineffective suppression of mullerian ducts causes ambiguous external genitalia, and is frequently accompanied by a small rudimentary uterus or a partially developed vagina. [7] When there is a genetic loss of cytoplasmic receptor proteins within androgenic target cells, as in the Androgen insensitivity syndrome the vagina is incompletely developed because the existing male gonads suppress the development of the mullerian ducts.

The Characteristics of the Mayer-Rokitansky-Kuster-Hauser syndrome are

- ❖ Congenital absence of the uterus and vagina (small rudimentary uterine bulbs are usually present with rudimentary fallopian tubes)
- ❖ Normal ovarian function, including ovulation
- ❖ Sex of rearing: female
- ❖ Phenotypic sex female (normal development of breasts, body proportions, hair distribution, and external genitalia)
- ❖ Genetic sex: female (46,XX karyotype)

- ❖ Frequent association of other congenital anomalies (skeletal, urologic, and especially renal)

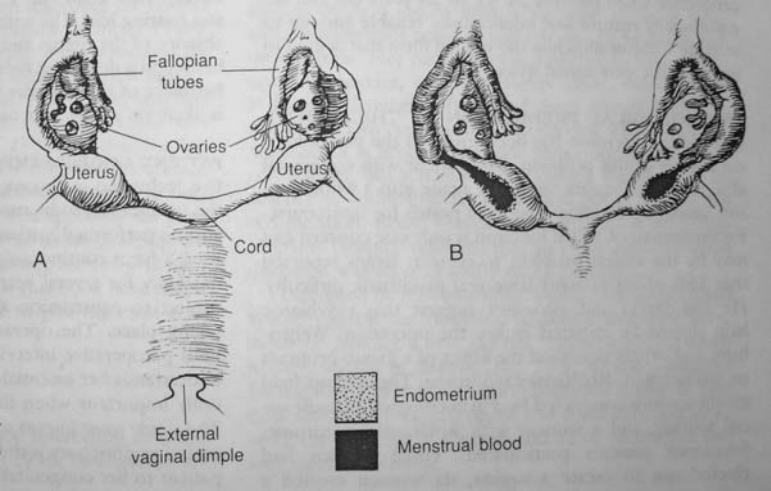
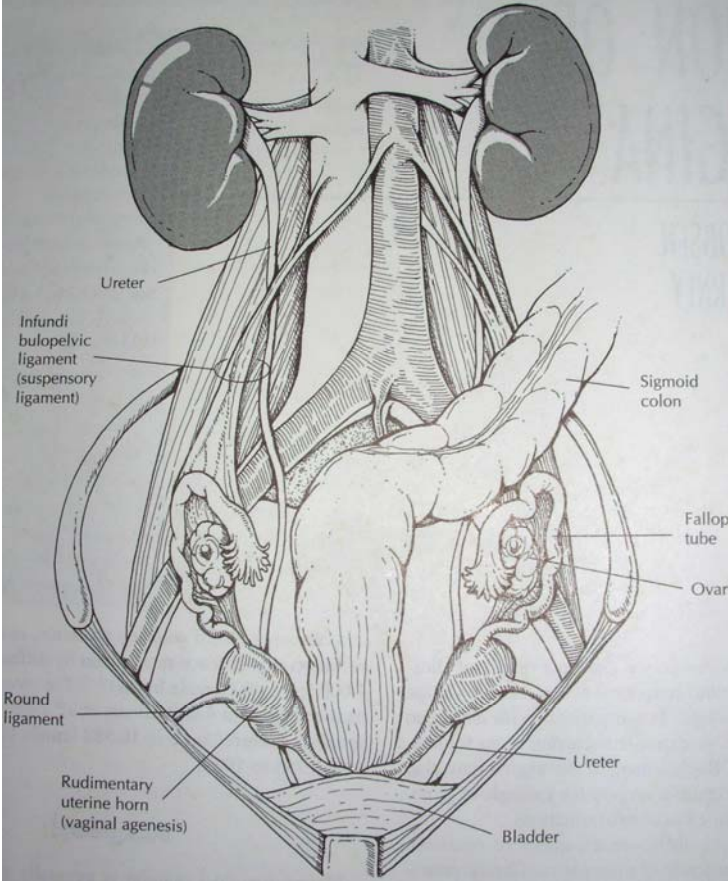
Realdus first described congenital absence of the vagina as one of the abnormalities found in stillborn infants with multiple birth defects.[7] Rokitansky in 1838 and Kuster in 1910 described an entity in which the vagina was absent, a small bipartite uterus was present, the ovaries were normal, and anomalies of other organ systems (renal and skeletal) were frequently observed.[13,14] Hauser and associates emphasized the spectrum of associated anomalies.[15] Pinsky suggested that congenital absence of the vagina is part of a symptom complex and not true syndrome.[28] Over the years, the disorder has come to be known as the Mayer-Rokitansky-Kuster-Hauser syndrome, the Rokitansky-Kuster-Hauser syndrome, or simply the Rokitansky syndrome. Counseller found that the condition occurred once in 4,000 female admissions to the Mayo clinic. [29] Evans estimated that vaginal agenesis occurred once in 10,588 female births in Michigan from 1953 to 1957. [2]

The patients usually are first seen by a gynecologist at age 14 to 15 years, when the absence of menses causes concern. They have a normal complement of chromosomes (46, XX) and usually have normal ovaries and secondary sex characteristics, including external genitalia. Menstruation

does not appear at the usual age because the uterus is absent, but ovulation occurs regularly. [7, 30] Some times polycystic ovaries and gonadal Dysgenesis have been reported in patients with vaginal agenesis. [31]

By gentle rectal examination, the physician can feel an absence of the midline mullerian structure that should represent the uterus. The physician instead feels a smooth band (possibly a remnant of the uterosacral ligaments) that extends from one side of the pelvis to the other. In MRKH syndrome, the uterus is represented by bilateral rudimentary uterine bulbs that vary in size, are not usually palpable, are connected to small fallopian tubes, and are located on the lateral pelvic side wall adjacent to normal ovaries. [32] Depending on their size, these rudimentary uterine bulbs may or may not contain a cavity lined by endometrial tissue. If present, the endometrial tissue can appear immature or, rarely, can show evidence of cyclic response to ovarian hormones. The endometrial cavity does not communicate often with the peritoneal cavity. Reports have described several patients with functioning endometrial tissue in one or both rudimentary uterine bulbs. The patient can develop a large hematometra due to cyclic accumulation of trapped blood with cyclic abdominal pain. [33-35] Endometrioma from the ovary and Myomas from the muscular wall were reported in addition to the Myomas in the inguinal canal and in the inguinal hernia sac. [7]

INTERNAL ANATOMY OF MRKH SYNDROME PATIENT



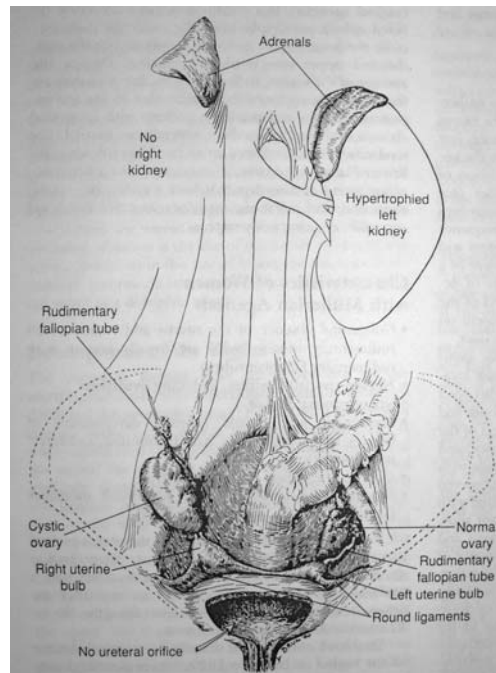
Chakravarty and colleagues and Singh and Devi have demonstrated that the rudimentary bulbs have the potential for function. They used these rudimentary uterine bulbs to reconstruct a midline uterus and connected to a newly constructed vagina. A surprising number of patients who have undergone this procedure have experienced cyclic menstruation, although recurrent stenosis and obstruction of the rudimentary horns are the most common results of such efforts. [36, 37]

ASSOCIATED UROLOGIC AND RENAL ANOMALIES

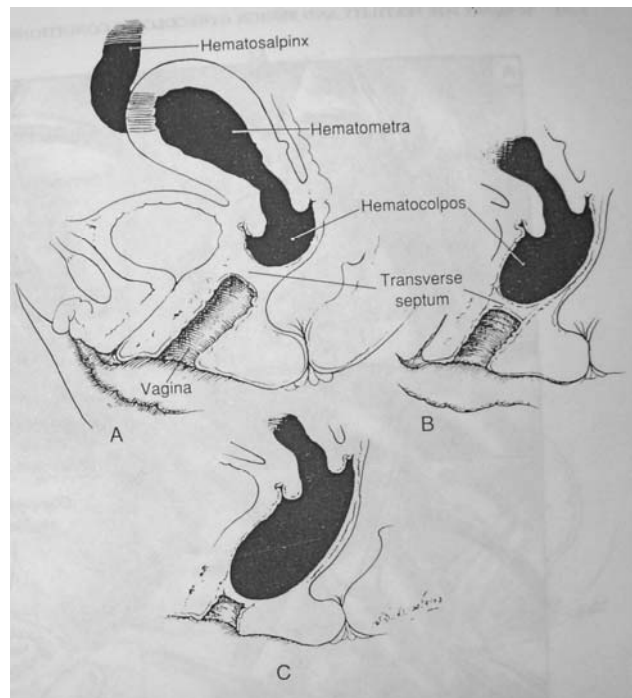
The close developmental relationships of the mullerian and wolffian ducts explain the frequency with which anomalies of the female genital system and urinary tract are associated with each other. The kidney can be absent on the side ipsilateral to the agenesis of a mullerian duct [17-19]. The kidneys can be absent, fused or in unusual locations in the pelvis. Ureters can be duplicated or can open in the vagina or uterus. Bilateral obstruction has not been observed clinically, because it would be associated with bilateral renal agenesis, which would not allow the embryo to develop. According to Thompson and Lynn, 40% of female patients with congenital absence of the kidney are found to have associated genital anomalies. [20]

Fore and associates reported that 47% of patients in whom evaluation of the urinary tract was performed had associated urologic anomalies. [38]

ASSOCIATED ANOMALIES IN MRKH SYNDROME



TRANSVERSE VAGINAL SEPTUM



In other studies, approximately one third of patients were found to have significant urinary anomalies. [40-54] A significant number of patients with partial vaginal agenesis also have associated urinary tract anomalies.

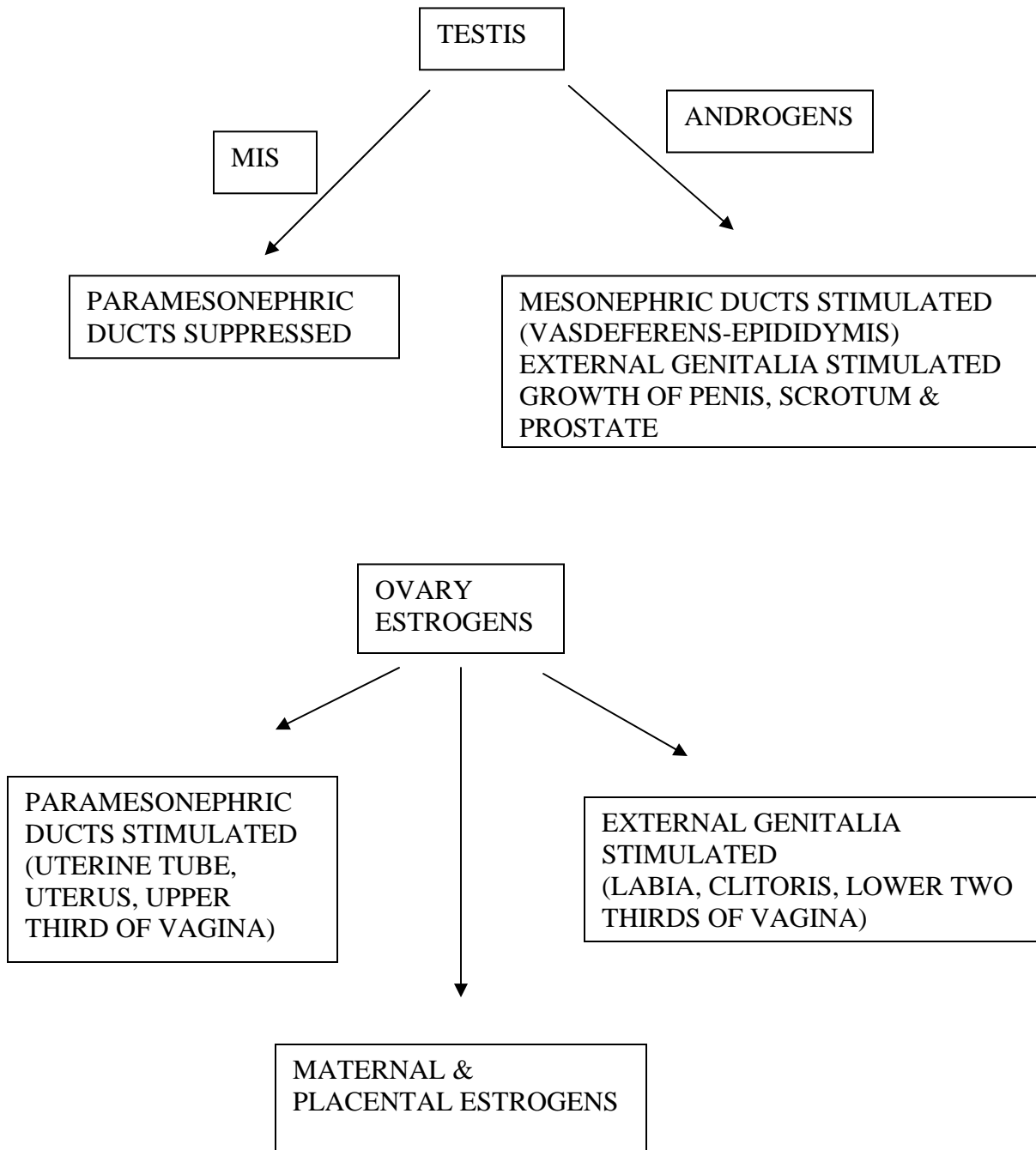
ASSOCIATED SKELETAL AND OTHER ANOMALIES

Griffin and associates found a 12% incidence of skeletal abnormalities. [23] Most of these abnormalities involve the spine (wedge vertebrae, fusions, rudimentary vertebral bodies, and supernumerary vertebrae), but the limbs and ribs also can be involved. Other anomalies include syndactyly, absence of a digit, congenital heart disease, and inguinal hernias.

ETIOLOGIC FACTORS

Much investigation has been undertaken to determine a genetic relationship in the development of disorders of the mullerian ducts. [21, 22] Familial aggregates are best explained on the basis of polygenic or multifactorial inheritance. [23] No information exists on the number and chromosomal location of responsible genes. Hand-foot-genital syndrome is a rare, dominantly inherited condition that affects both the distal limbs and the genitourinary tract. [24, 25] A nonsense mutation of the HOXA13 gene has been reported in several families with multiple mullerian abnormalities. [26] This may also be associated with other more complex malformation syndromes, in which the molecular basis remains unknown. [27, 28]

INFLUENCE OF SEX GLAND ON FURTHER SEX DIFFERENTIATION



An exclusively genetic etiology cannot be ascribed to vaginal agenesis because almost all patients have a normal karyotype (46, XX). [55] Shokeir investigated the families of 13 unrelated females with vaginal aplasia. [56] Similarly affected females were found in 10 families. Usually there was an affected female paternal relative, suggesting female-limited autosomal dominant inheritance of a mutant gene transmitted by male relatives. Knab believes that the teratogenic and the mutant gene etiologies are the most probable [57]. He has suggested the possible etiologic factors as:

1. Inappropriate production of mullerian regressive factor in the female embryonic gonad
2. Regional absence or deficiency of estrogen receptors limited to the lower mullerian ducts
3. Arrest of mullerian duct development by a teratogenic agent
4. mesenchymal inductive defect
5. sporadic gene mutation

PREOPERATIVE CONSIDERATIONS

If functioning endometrial tissue is present with the anlagen, then symptoms from cryptomenorhhea will begin shortly after female secondary sex characteristics develop. Prompt removal of the active uterine bulbs affords complete relief of symptoms.

Occasionally, older patients with the classic Mayer-Rokitansky-Kuster-Hauser syndrome consult a gynecologist because of difficult or painful intercourse. The indication for operations in these patients is obvious. Of all patients, they are the most satisfied with the operative results.

Most commonly, patients aged 14 to 16 years are seen by a gynecologist because of primary amenorrhea. In the past it was customary to advise delaying surgery to create a vagina for these young patients until just before their marriage. This led to difficulties, particularly when complications developed that required a delay in marriage until the vagina healed completely. More recently, it has become usual to perform the procedure when patients are 17 to 20 years old and are emotionally mature and intellectually reliable enough to manage without difficulty the vaginal form that is used to maintain the neovaginal space.

PSYCHOLOGICAL PREPARATION OF THE PATIENT

Insufficient attention has been given to the psychological aspects of this problem. Evans reported that 15% of his patients have real psychiatric difficulty. [2] He and David and associates suggest that psychiatric help should be initiated before the operation. [58] Weijenborg and others described the effect of a group program on women with Rokitansky syndrome. [59] They held group sessions conducted by a gynecologist, a

female social worker, and a woman with MRKH syndrome. Learning about this anomaly, especially at a young age, is a shock and is accompanied by diminished self-esteem.

When counseling patient, gestational surrogacy should definitely be included in the discussion. Beski and others confirmed the use of gestational surrogacy in a small population. [60]

PATIENT COOPERATION

Regardless of which operative technique is chosen, the patient must cooperate if the operation is to be successful. When a McIndoe operation is performed, patients must understand the need to wear a form continuously for several months and intermittently for several years until the vagina is no longer subject to constriction and until the regular intercourse is taking place.

LABORATORY AND DIAGNOSTIC TESTING

A complete chromosomal analysis should be performed in all patients. If there is a suspicion of ovarian Dysgenesis, androgen insensitivity syndrome, or some aberration of the classic Mayer-Rokitansky-Kuster-Hauser syndrome, then a consideration of additional SRY analysis should be entertained to assess the possible presence of any Y chromosome. An intravenous pyelogram should be done preoperatively. This also provides an adequate survey for anomalies of the spine. If a pelvic mass is present, then

additional special studies, including ultrasonography, should be performed to differentiate between hematometra, hematocolpos, endometrial and other ovarian cysts, and pelvic kidney. [7]

EVALUATION OF CYCLIC PAIN

Some patients without a pelvic mass complain of cyclic pain. This can be ovulatory or possibly a result of dysmenorrhea originating in well-developed rudimentary uterine bulbs. The physician can differentiate between the two by a basal body temperature chart. An intervening mass between the rectal finger and the instrument in the urethra can represent a hematocolpos behind an imperforate hymen.

METHODS OF CREATING A VAGINA

It is important for all surgeons doing this type of surgery to be familiar with several different techniques so that the operation most appropriate for the patient's particular situation will be chosen.

An ideal vagina must be

1. located at an appropriate place and directed postero superiorly,
2. of a adequate dimensions,
3. lined by elastile tissue either by full thickness skin or mucosa,
4. neither permanently moist nor malodours,
5. hairless,

6. sensate at least at the introitus level and
7. Constructed by a method that is simple, easily reproducible, and single staged with least morbidity.

HISTORY OF ATTEMPTS TO FORM A VAGINA

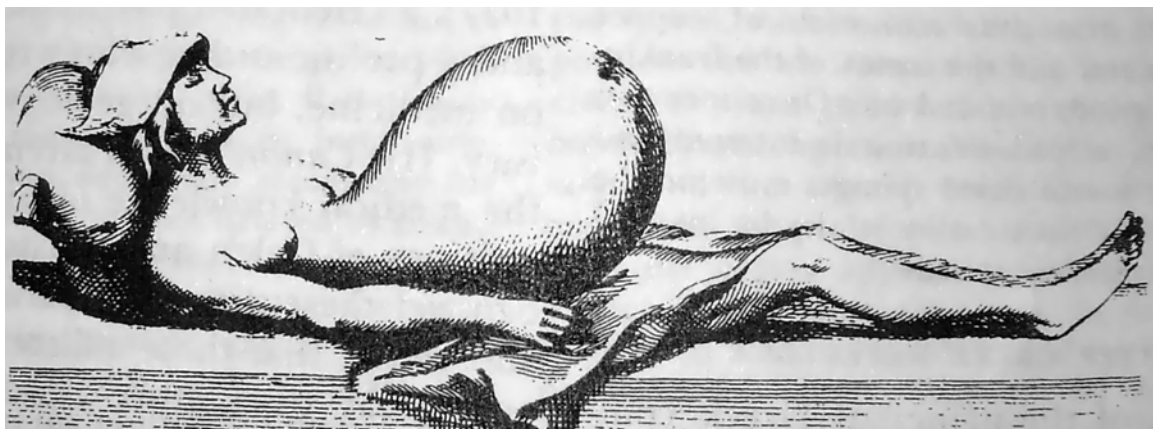
Hippocrates (? 460 – 370 B.C), had authentically wrote about the description of “membranous obstruction” of the vagina in the book “on the nature of women”. Celsus (first century A.D) was probably the first to describe imperforate hymen and possibly vaginal aplasia. His description of an operation to correct the latter sounds familiar to today’s surgeons, who find similarities in it to the first stage of reconstruction. He used lead tube smeared with an ointment and acting as a tamponade to keep the cut vaginal surfaces from reuniting.

Soranus (78-117 A. D.) of Ephesus first described the disease as “atresia” in his book of “diseases of women” which was an unquestionable reference for next 1500 years.

Aetios (502- 575 A.D.) Of Amida described as “There are three types of atresia. There are those in whom a membrane or an obstructing piece of flesh develops in the wings of the labia of the pudendum; in others the obstruction occurs about the mouth of the uterus itself”.



TREATMENT OF VAGINAL ATRESIA CHARAF ED-DIN (1465)



Abdominal swelling from retained menstrual fluid at autopsy on a young woman with vaginal atresia (Job J Van Meek'ren 1611-1666)



Robert Abbe



Archibald McIndoe



Charles Horton

Paulus Aegineta (625-690 A. D.) recommended that following removal of the obstruction, the vaginal surfaces be permitted to heal more naturally by the application of “a priapus-shaped tent covered with some epulotic medicine, in order that the parts may not unite again”.

Ibn sina or Avicenna (980-1036), the celebrated physician of Baghdad recognized the entity of imperforate vagina and noted that those afflicted had pain and great discomfort, abdominal swelling and could die if a passage is not created. In fifteenth century, Charaf ed-din (about 1465), a Turkish surgeon, was responsible for a beautiful illustrated surgical text.

Matteo Realdo columbo (1516-1559) is generally attributed the first recording of the absence of both the vagina and uterus. Fabricus of Acquapendente (1537-1619) had been a pupil of Gabriele Fallopio and the teacher of William Harvey at Padua treated two patients without an open vagina.

Job J. van Meek'ren's (1611-1666) illustration of abdominal distension, from retained menstrual fluid secondary to vaginal atresia was one of the earliest in the history.

During the last half of the eighteenth century and throughout the nineteenth century, numerous graduation theses were written on vaginal and uterine atresia. In 1764. Antoneus de Haen (1704-1776), published his “Ratio

Medendi in Noscomiio Practico”. He used two terms: imperforatorio or atresia, which is not attested in clinical usage. A more exact and scholarly description of absence of vagina and uterus was by Engel in a doctoral thesis (1781), containing the result of an autopsy on a young woman who died suddenly.

John syng Dorsey (1783-1818) of Philadelphia described in his 1818 text book of surgery the treatment of imperforate vagina as follows: “When the vagina is closed by a membrane, A straight incision, or if the membrane be very dense, a crucial incision should be made through it, and the wound kept from uniting by the introduction of a sponge tent, or roll of linen”. Perhaps the patient of Philip syng physick (1783-1837) of Philadelphia was the first in medical records to be successfully operated upon for vaginal agenesis.

According to Ricci, after DeHaen unsuccessfully operated for an absent vagina in 1761, Dupuytren in 1817 was the next to attempt a plastic surgical correction of this deformity. He, too, was unsuccessful, as was Villaume in 1826, Boyer in 1831, and Debrou in 1851, all of whom used DeHaen’s surgical method. This method apparently consisted of attempting to make an artificial vagina by creating an opening between the bladder and the rectum, kept patent by a tampon. Owens pointed out that this

fundamentally sound procedure was advocated by Dupuytren in 1817, but it was essentially forgotten until approximately 1936 when Monod and Iselin “improved Dupuytren’s original operation by applying Thiersch grafts over a hard stent and inserting the stent in to the newly formed vaginal canal”.

John Collins Warren (1778-1856) of Boston probably reported the first successful correction surgically for “non-existence of vagina”. His son, Jonathan Mason Warren (1811-1867), also reported surgery of three cases of vaginal atresia in 1851. The first plastic surgical operation following Warren’s operation for the reconstruction of an absent vagina in 1833 that proved successful was reported in 1870 by C. L. Heppner in St. Petersburg. He made an H-shaped incision in the rectovaginal septum and lined the walls of this cavity with tongue-shaped flaps of skin that he dissected from the adjacent thigh regions.

MODERN TIME TECHNIQUES

Even today there is still no consensus as to the best surgery. Treatments have varied considerably from the non operative approach of intermittent pressure and dilatation, the extreme of vaginal transplantation from mother to daughter. [61, 62]

TABLE-III

CLASSIFICATION OF METHODS TO FORM NEO VAGINA

NON SURGICAL

Frank's technique
Ingram's technique

SURGICAL

WITHOUT CAVITY DISSECTION

Constant pressure (vechiette)
Vulvovaginoplasty (William's, Obrien's)
Combined procedure

CAVITY DISSECTION AND LINING

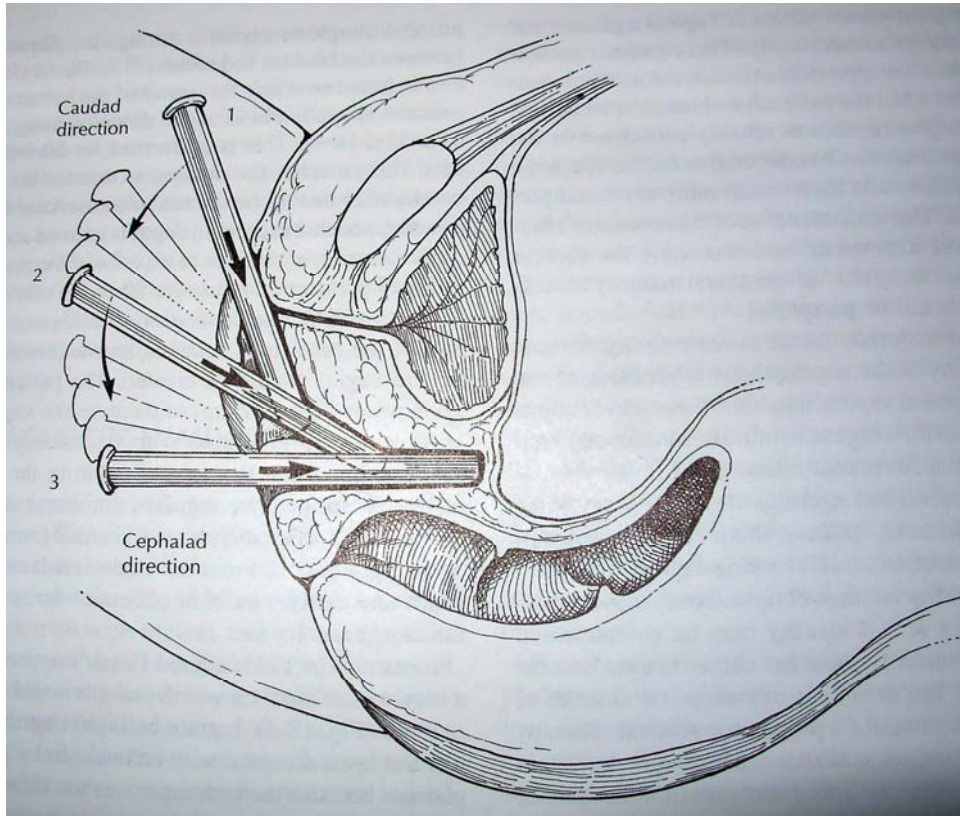
No lining
Lining with grafts
 Split thickness skin graft
 Full thickness skin graft
 Dermis graft
 Buccal mucosal graft
 Bladder mucosal graft
 Amnion
 Peritoneum
Lining with skin flaps
 Labial skin after tissue expansion
 Labia majora skin
 Labia minora skin
 Subcutaneous pedicled skin flaps
Lining with Facio-cutaneous flaps
 Perineal artery axial flap
 Malaga flap
 Neurovascular pudental thigh flaps
 Modified Singapore flap
Lining with musculo-cutaneous flaps
 Gracilis myocutaneous flap
 Rectus abdomis flap
 Vulvobulbocavernosus flap
Lining with intestine
 Jejunum
 Ileum
 Caecum
 Sigmoid colon
 Rectum
Micro vascular reconstruction
 Jejunum
 Scapular skin

NONSURGICAL METHODS

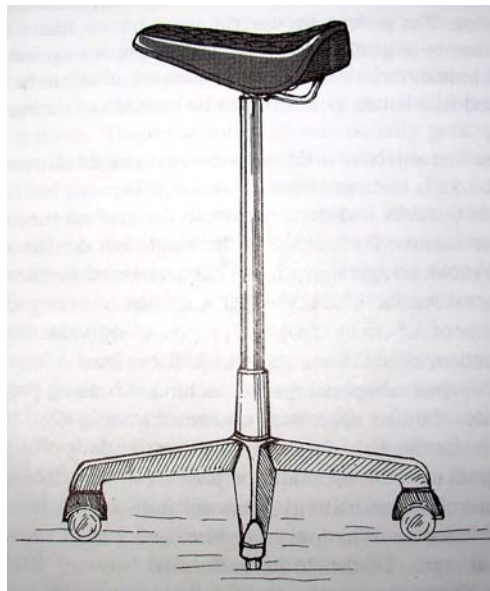
In 1938, Frank described a method of creating an artificial vagina without operation. In 1940, he reported remarkably satisfactory results in eight patients treated with this method. [63, 64] His follow up study showed that a vagina formed in this manner remained permanent in depth and caliber, even in patients who neglected dilatation for more than 1 year.

Prompted by the rewarding results of Broadbent and Woolf, Ingram has described a passive dilatation technique of creating a new vagina. [65, 66] Instructing his patient in the insertion of dilators specially designed for use with a bicycle seat stool, Ingram was able to produce satisfactory vaginal depth and coital function in 10 of 12 cases of vaginal agenesis and 32 of 40 cases of various types of stenosis. A series of graduated Lucite dilators slowly and evenly dilate the neovaginal space. The dilator may be held in place with a supportive undergarment and regular clothing worn over this. The patient is shown how to sit on a racing type bicycle seat that is placed on a stool 24 inches above the floor. She is instructed to sit leaning slightly forward with the dilator in place for at least 2 hours per day at intervals of 15 to 30 minutes. Follow-up is usually at monthly intervals and the patient can expect to graduate to the next size larger dilator about every month.

NON OPERATIVE TECHNIQUES



FRANK'S TECHNIQUE



INGRAM'S BICYCLE SEAT STOOL

An attempt at sexual intercourse may be suggested after the use of the largest dilator for 1 or 2 months. Continued dilatation is recommended if intercourse is infrequent.

Rock and Roberts reported the largest series of vaginal agenesis patients who used the Ingram method of dilatation to create a neovagina. [67] Functional success was achieved in 91.9% of those who attempted dilatation.

SURGICAL TECHNIQUES

COMBINED TECHNIQUE

Makinoda and colleagues reported 18 cases of two step non grafting method. First step is the Frank's dilation of the dimple. The second step is surgical via perineal approach. The apex of the dilated vaginal space is incised and further dissection is done up to peritoneal cavity. The uterine structures when present are pulled down and sutured to the newly created vaginal space. [68]

VECHIETTI PROCEDURE

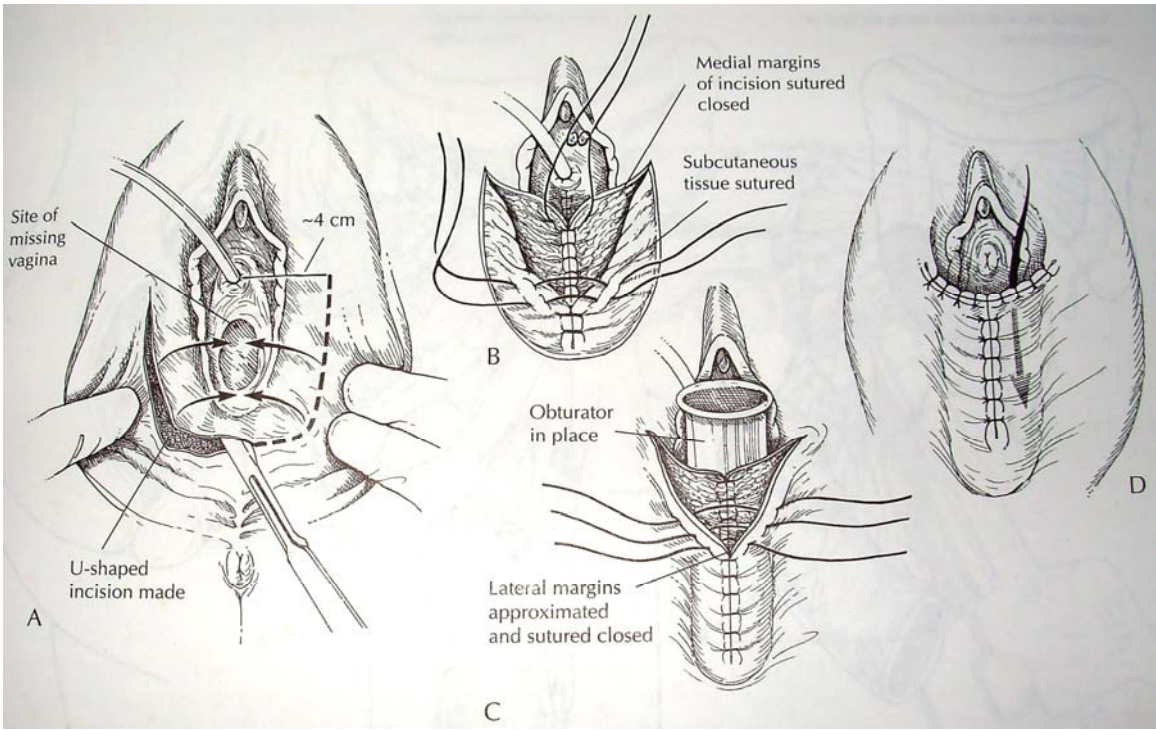
The vechiette procedure is used frequently in Western Europe and Asia, first described by Giuseppe vechiette in 1965. [69]A cavity is created using continuous progressive pressure to the vaginal dimple. Two threads are attached to a 2 cm acrylic olive and then passed through the pseudohymenal tissue at the introitus, up through the potential neovaginal

space and out through the abdominal wall. The threads are then connected to a traction device on the abdomen that draws the olive upward. The elastic skin stretches as the olive moves upward over the course of several days, creating a vaginal cavity of desired depth. Within 7 to 8 days in the hospital, a vagina of sexually usable depth is created, but regular sexual intercourse or the use of vaginal molds is necessary to maintain vaginal depth and patency. Gauwerky described a laparoscopic version of this procedure. [70] The vesico rectal space was dissected laparoscopically. The threads of olive device were positioned using a probe introduced into the abdomen through the perineum. In 1995, Laffarque and others described a laparoscopic intervention without creation of vesicorectal space. [71] Fedele used combined laparoscopic-ultrasonographic technique. The complications include bladder, rectal injuries, dysparenuia and vaginal vault bleeding. [72]

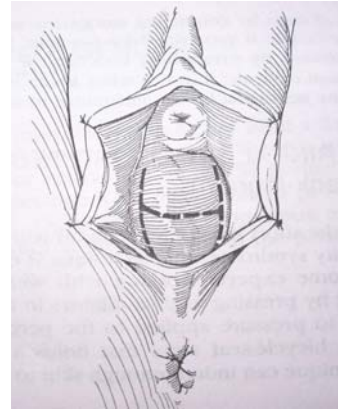
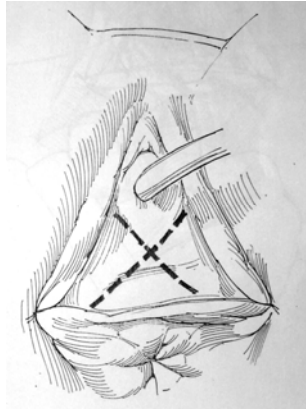
WILLIAMS VULVOVAGINOPLASTY

A U- shaped incision is made along the inner surface of the labia majora, as close to the hairline as possible, about 4 cm lateral to the urethra. The patient must have sufficient skin available to form a pouch. The skin is then undermined and, after it is well mobilized, the inner skin margins are sutured together with the knots tied inside the vaginal lumen. A second layer of interrupted sutures reapproximates the subcutaneous fat and perineal muscles for support. The external skin margins are then

WILLIAM'S VULVOVAGINOPLASTY



INCISIONS



approximated with interrupted sutures. The pouch should be at least 3 cm deep and wide enough to insert two fingers. These patients are placed on bed rest for 1 week to avoid tension on the suture line. Williams reported in 1976 that 51 of 52 patients on whom he performed this procedure had successful results. The simplicity of this surgery makes this option attractive. [73, 74]

The major drawback to this procedure is that the resultant canal is at a vertical angle, but it is reported to straighten out as it is gradually pressed posteriorly by the penis during intercourse. [75] This same result can be achieved if patients use dilators. Other drawbacks include a problem with urine collecting in the pouch. Some patients complain of a dry pouch and excessive stimulation against the clitoris. This procedure should not be done in patients with a patulous urethral meatus due to concern that the urethra might be stretched further by coitus.

In Obrien's vulvovaginoplasty, he uses the vulval tissue. He takes all the non hair bearing skin within the labia majora in the shape of a 'U' shaped flap based anteriorly and creates a new vagina. This flap divides all the neurovascular input coming from the internal pudendal system. [76] Case reports describe a satisfactory outcome of a variant of the Williams procedure combined with cavitations and subsequent dilation. [77, 78]

METHODS WITH CAVITY DISSECTION AND LINING

SURGICAL TECHNIQUE OF CREATING PERINEAL CAVITY

The procedure must be performed under general or spinal anesthesia. The perineum is shaved. However the thighs are not shaved in order to allow selection of hairless skin for inlay graft. A lower bowel preparation is given the night before and the morning of surgery. The lithotomy position allows access to the three areas required: the suprapubic area for laparotomy and gonadal biopsy, the vulva for vaginal reconstruction, and the inner aspect of thighs for skin graft.

A Y-shaped incision is made along the median raphe between the urethra and the anus. Originally Abbe described crescentic incision; the modifications include inverted V-shaped, H-shaped and X- shaped incisions. A catheter is placed in the urethra, and dissection is carried cephaloposteriorly. This dissection can be done relatively bluntly, and safety is ensured by keeping a gloved finger in the rectum. The Y incision allows three cutaneous flaps to be enfolded into the vaginal canal so that circumferential scar contraction can be avoided. The depth of dissection should be somewhat exaggerated and is in the range of 10 to 14 cm in the adult. The surgeon must overcorrect somewhat because of the expected subsequent contraction.

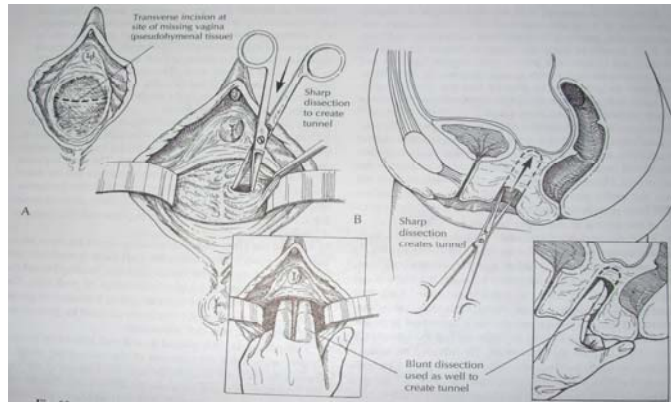
NO LINING

In 1938, Wharton combined adequate dissection of the vaginal space with continuous dilation by a balsa form covered with a thin rubber sheath. This was based on the principle that vaginal epithelium would proliferate and, in a short time, would cover the raw surface. Occasionally, however, the epithelium does not cover the entire vault and coital bleeding and leucorrhoea result from persistent granulation tissue. This leads to scarring of the vagina in the upper portion. [79]

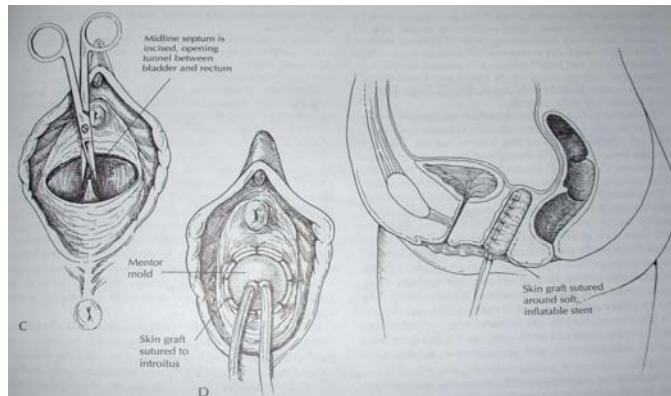
ABBE-McINDOE PROCEDURE

Over the years, the Abbe-McIndoe procedure evolved to become the easiest and most successful method of vaginal reconstruction, avoiding the risks and disadvantages of laparotomy. The use of inlay split-skin grafting is today's most common method of vaginal reconstruction, was pioneered by Robert Abbe (1851-1928) of New York city. He was a remarkable and resourceful surgeon who is known now for his treatment of ankylosis of the temporomandibular joint, tumors of the spinal cord, and his techniques of intestinal anastomosis and bile duct exploration-to cite a few examples of his versatility. Published in 1898, Abbe's article is a model of sound judgment and excellent technique. [80]

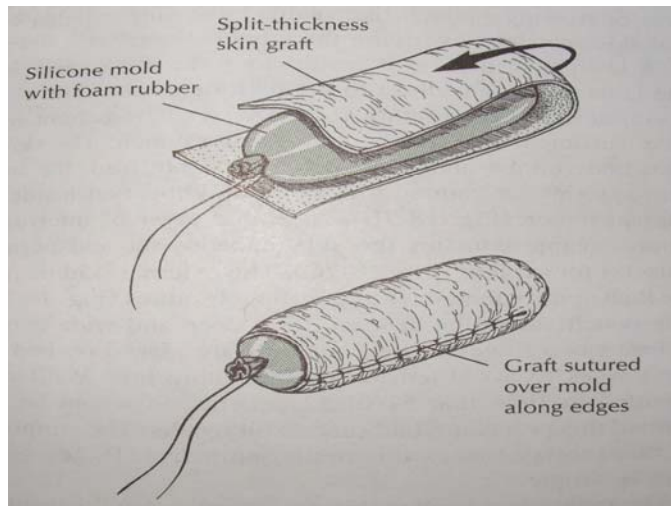
ABBE –McINDOE PROCEDURE



A: CREATION OF NEOVAGINAL SPACE
B: MEDIAN RAPHE CREATED



C: MEDIAN RAPHE IS CUT
D: SKIN GRAFT KEPT INSITU



SUTURING OF SKIN GRAFT OVER THE VAGINAL FORM

However, Abbe's report was lost for almost forty years until 1938, Sir Archibald McIndoe at Queen Victoria Hospital in England popularized a method that combined many of the principles developed before him. He emphasized, first, the dissection of an adequate space between the rectum and bladder. Second, an inlay split-thickness skin graft was placed so that the newly formed cavity would have an epithelial lining. Third, he recognized the most important principle of continuous and prolonged dilation during the contractile phase of healing of the skin graft. He advised wearing a mold continuously for 6 months following surgery. [81-83]

McIndoe reported an impressive array of 63 repairs, and subsequently Counsellor in 1948 reported 70 cases. [84] Counsellor and Flor modified this technique in 1958, by using a foam rubber mold covered by a condom instead of the acrylic shells used by McIndoe. [85]

After the dissection two or three sheets of skin graft are harvested from the medial aspect of thigh. Alternatively skin graft can be harvested from relatively hidden areas such as gluteal region, scalp. [86] The donor area is carefully selected to be hairless and hidden site.

A number of techniques have been developed to apply the skin graft to the vaginal canal. Historically, candle wax, carved balsa wood, gauze

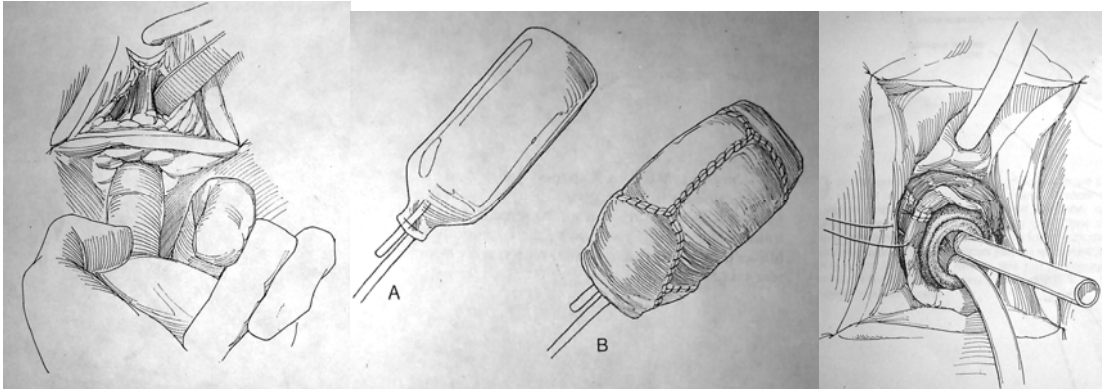
packing, syringe casing, dental wax, and hard plastic conformers have been used. Recently expandable vaginal conformer from the Heyer-Schulte company is also available.

Advantages of this procedure include simplicity, reliability and safety. [87, 88] In the report by Buss and Lee, 5 of 50 patients needed secondary operation due to stricture or contracture of the split-thickness skin graft-lined vaginas, and 15% cannot achieve successful function.[90] Following McIndoe's inlay split-thickness skin graft method, contracture and stenosis of the neovaginas also were noted in 42% of the 113 patients reported by Cali and Pratt. [90] OrtizMonasterio et al. followed their 21 patients from 1 to 16 years and found 4 cases of vaginal contracture (19%).[91] Moreover, the pigmented, irregular, and sometimes hypertrophic thigh scars after harvesting the split-thickness skin graft are also psychologically and socially unacceptable to those otherwise healthy young females in modern days of fashion consciousness.

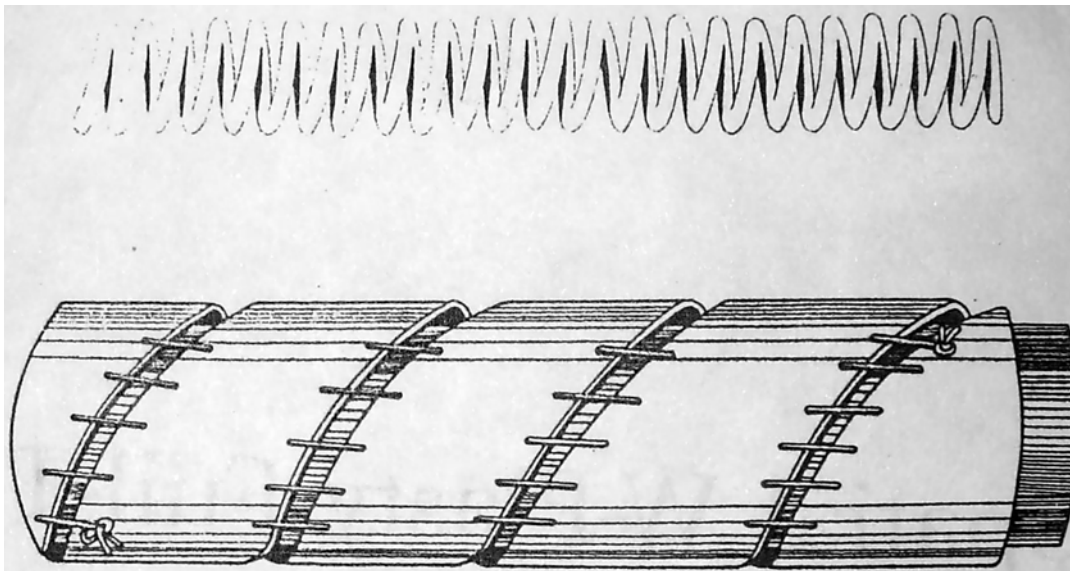
LINING OF CAVITY USING FULL-THICKNESS SKIN GRAFTS

Horton and colleagues utilized two pieces of full-thickness skin graft from both groin areas for neovaginal reconstruction. [92, 93] In Horton's series, there were no cases of stenosis, and the donor scars were inconspicuous. They reduce the amount of time required for postoperative stenting, allow reconstruction at an earlier age, and minimize the risk of

FULL THICKNESS SKIN GRAFTING



SPATIAL W- PLASTY



vaginal stenosis by decreasing postoperative vaginal contraction. Full-thickness skin grafts grow proportionately with the remainder of the body. This technique is applied primarily to vaginal aplasia, but is also useful in cases of male to female Trans-sexualism, unsuccessful transsexual operations resulting in vaginal stenosis, and certain other intersex conditions and cases of iatrogenic disease.

Full-thickness skin grafts are obtained from the lateral hairless groin areas bilaterally or from the gluteal crease. The donor sites are closed primarily. All extra fat is removed from the undersurface of the grafts which are sutured together over a stent. The foam rubber stent covered by the full-thickness grafts is then inserted into the new tract, and the proximal open edge of the skin graft is sutured to the vaginal mucosa and the V-flap with 3-0 interrupted sutures. The labia tissue is sutured together with 2-0 nylon mattress sutures to prevent extrusion of the stent and graft. However drawbacks of this technique are texture mismatch, dryness, desquamation, and hair growth, and it does not produce mucus.

Y.B.Chen et.al, has described a technique which combines the advantages of a full-thickness skin graft and Z-plasty principle in a series of four patients. This spatial W-plasty full-thickness skin graft is taken from one groin in the shape of a long strip instead of bilateral. The graft was placed on the stent inside out in a spiral fashion. [94]

LINING OF CAVITY USING BUCCAL MUCOSAL GRAFTS

Under general anesthesia, an elliptical full-thickness mucosal graft, 6 to 7 cm long and 2 to 3 cm wide, was harvested from the oral surface of both cheeks. The donor site was closed primarily with 4-0 chromic catgut interrupted sutures. After the submucosal fat undersurface of the grafts was cleaned, the mucosal grafts were expanded 2:1 by using multiple stab incisions and then divided into several smaller pieces 2 to 4 cm in area. Next, these graft pieces were sutured over the stent using 5-0 chromic catgut at a distance of 2 to 3 cm from one another. The grafts were placed in an inside out fashion with the dermis facing outward. A soft, condom-covered, foam rubber stent measuring 5 cm in diameter and 10 cm in length was used. The stent with its overlying patch buccal mucosal grafts was meticulously inserted into the neo vaginal cavity and both sides of labia minora were sutured together to retain the stent in position.

Four patients with congenital absence of vagina treated with this are presented. This surgical procedure created a mucus providing lining inside the neo vagina. Donor site is inconspicuous. Histologically, the neo vaginal lining was confirmed as mucosal, and the cream colored viscous fluid found in the neo vaginal cavity was confirmed as mucus. [95]

LINING OF CAVITY USING HUMAN AMNION

Amnion is an inexpensive allograft with low antigenicity, high antimicrobial activity, and the ability to enhance epithelialization. Membranes are taken immediately postpartum from a patient who has been afebrile and who has been ruptured for less than 12 hours. A woman undergoing an elective cesarean section is ideal. The amnion is then rinsed in sterile saline until free of contaminants such as blood, vernix, or meconium and stored at 4°C in saline containing 50,000 U/100ml of crystalline penicillin. It should be harvested close to the time it will be used for the neovaginal surgery, but it has been shown to last for days.

Once a cavity has been dissected, the amnion is draped over a mold with its chorion side outwards and inserted into the cavity. After 7 days, the mold is removed and a clean mold covered with a new piece of amnion is inserted. After 7 more days, the mold is removed and the patient is instructed to use a vaginal dilator at least three times daily for 15 minutes to keep the cavity open.

Morton and Dewhurst showed that by the time of the second mold change, epithelialization was complete in 24 of 27 patients undergoing this procedure. [96] By 4 weeks after the operation, healthy pink vaginal epithelium was visible. Dhall showed that the vaginal biopsies taken 4 to 6

weeks after operation showed early epithelialization, which was replaced by mature epithelium by the end of 8 to 10 weeks. [97] Postoperative dilation is essential to prevent vaginal contraction. Dhall reported that the entire procedure took 15 minutes. Other advantages to the use of amnion are that there is no painful buttock wound and no scar. The major disadvantage, which has limited the use of the procedure, is the concern about the transmission of the human immunodeficiency virus.

LINING OF CAVITY USING OTHER TISSUES

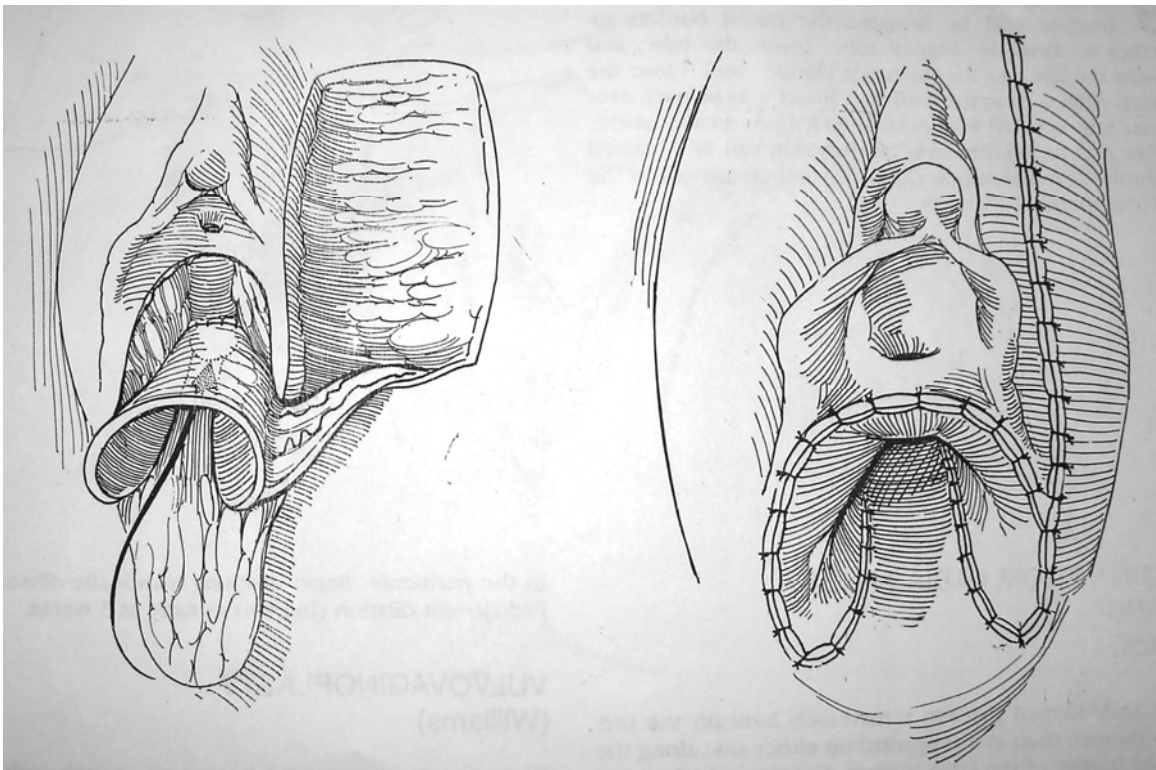
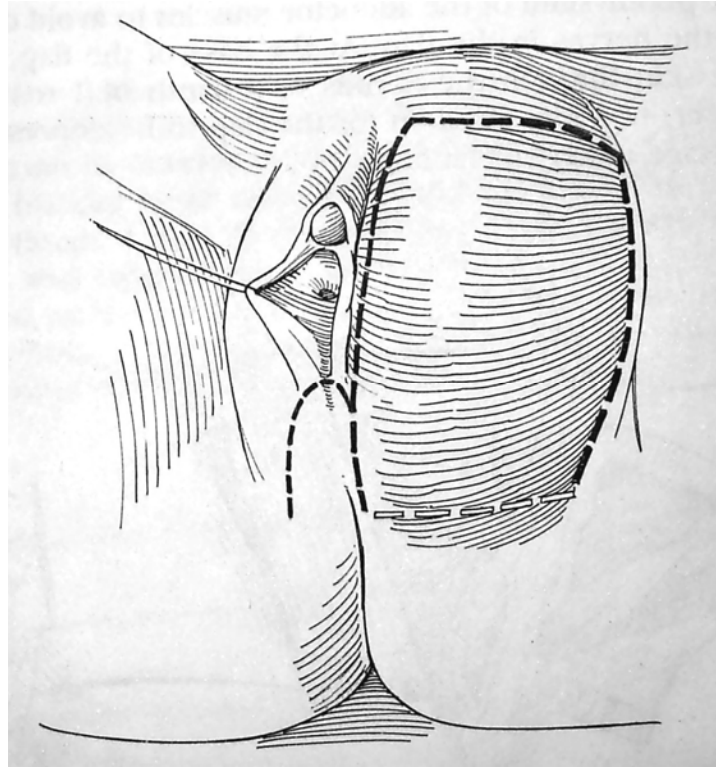
After creating the neo vaginal space, the cavity is lined with peritoneum hoping that cells may undergo metaplastic changes to form epithelium. [98, 99] Sometimes tissues like bladder mucosa and dermis were used to line the cavity.

LINING WITH SKIN FLAPS

TISSUE EXPANSION TECHNIQUE

The role of tissue expanders in vaginoplasty has been reviewed by Patil and Hixon. [100] Make a right inguinal incision, and digitally dissect a pocket in the right labium majus. Select a tissue expander of suitable size; 250 ml is suitable for an adolescent. Insert the tissue expander in the labial

TISSUE EXPANSION



pocket, and place the filling port subcutaneously in a (future) hair-bearing area, where it can be felt through the skin. Every 2 weeks add up to 20 ml of normal saline through a 25-gauge needle. Thus, well vascularized flaps of 10 cm long and 8 cm wide dimension are available for vaginoplasty.

Nicholas Johnson, Andrew Batchelor, R. J. Lilford reported a series of 17 cases in which the expansible balloons were inserted through bilateral groin incisions and placed slightly medial to labium minus. Then expanded skin is used for vaginoplasty. [101]

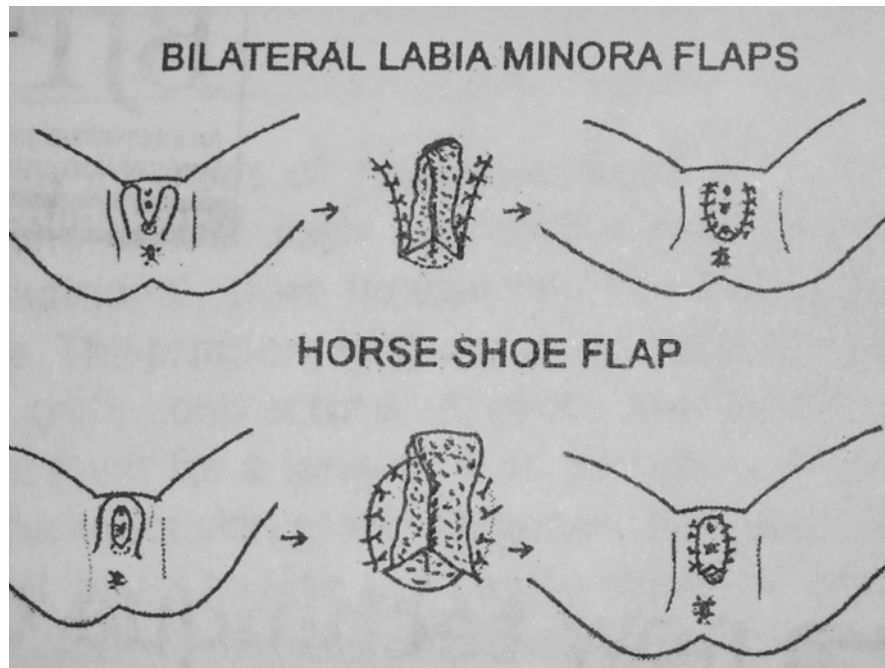
VAGINA FROM LABIA MINORA FLAPS

Flack C. E. et al described a flap using labia minora for vaginoplasty. [102] An M-shaped incision is made transversely beneath the urethral meatus, and then incised upward on either side along the medial border of the labia minora. The incision is continued caudally between the labia majora and minora. The paraurethral flaps are sutured together to form the floor of the new vagina, and then the caudal flaps are approximated to form the roof. The pocket so formed is rotated in to a cavity created in the perineum. Dilations were started as early as 3 weeks.

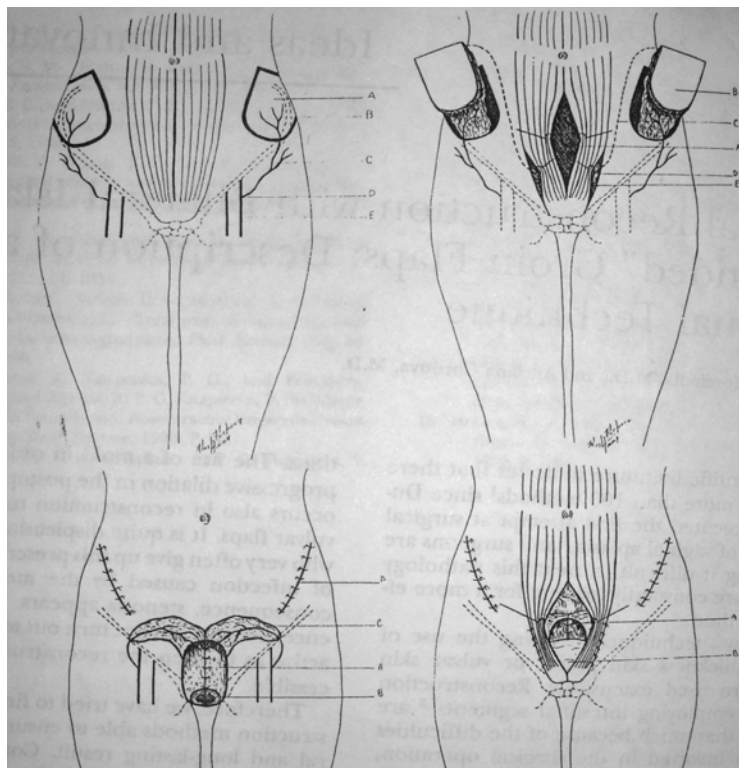
HORSE SHOE FLAP VAGINOPLASTY

A single horse shoe flap is marked involving both labia minora and the preputial skin. The base is close to the introitus posteriorly, at the

LABIA MINORA FLAPS



BILATERAL GROIN FLAPS



fourchette, where there is a rich vascular network. Both the medial incisions meet in the midline in a V fashion, where the future vagina is going to be. The flap is sutured together to form a tube with skin turned inwards. The tube flap is tunnelled into the space. A sponge stent is kept. [103]

MEDIAL THIGH SKIN FLAPS

The flaps are proximally based, and the proximal part is deepithelialized. A typical flap measures 15 cm in length and 5 to 6 cm in width. The flaps are rotated through 180 degrees and inserted through a subdermal tunnel into the vaginal cavity, where they are sutured. The donor site is closed in layers after wide undermining, with a gap left at the base of the flap, in order to avoid constriction. [104] In a series of 15 flaps, 3 suffered partial necrosis 1 total failure. In addition, revisional procedures to divide it have been necessary in some patients. [105]

PEDICLED ABDOMINAL FLAPS

The left inferior abdominal wall flap with subcutaneous pedicle containing superficial epigastric blood vessels and / or superficial circumflex iliac blood vessels external pudendal vessel and branches is raised and passed through an immediate extra peritoneal tunnel to the artificial vagina. [106]

BILATERAL ISLAND EXTENDED GROIN FLAPS

The technique requires the use of two island extended groin flaps designed over the iliac crests and vascularized by the superficial circumflex arteries. The flaps are elevated with the vascular pedicle. Through a subcutaneous tunnel the flaps are led straight toward the suprapubic area, before the rectus muscles. An incision is made along the lateral border of the recti muscle, and each flap by this way passes from before to behind through the fibro vascular tissue. Then the flaps are sutured together and to the vaginal ostium. [107]

LINING WITH MUSCULOCUTANEOUS FLAPS

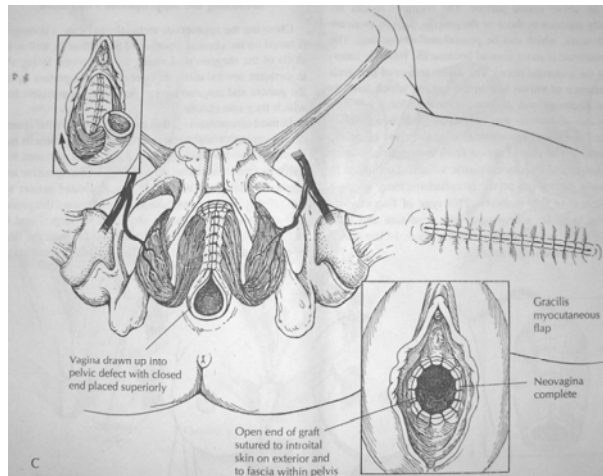
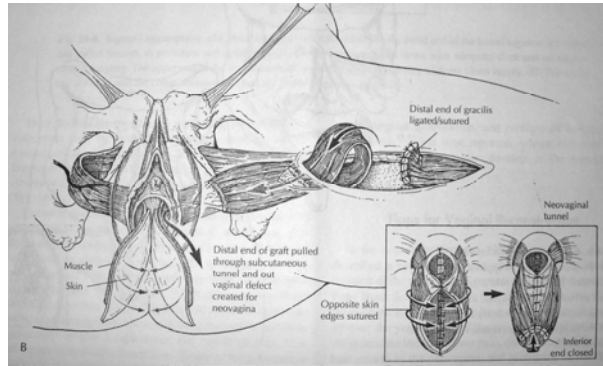
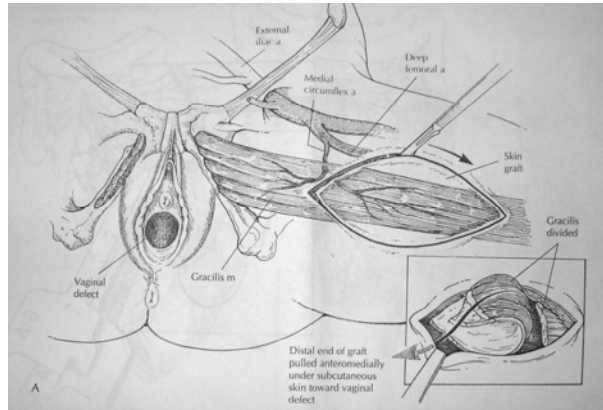
BILATERAL GRACILIS MYOCUTANEOUS FLAPS

Gracilis flaps are raised from both the thighs. A tunnel is formed to connect the flap incision with the vagina under the intact perineal skin. Flaps are rotated and drawn through the tunnel and sutured. The donor area is closed primarily. Although it appears much more reliable in the hands of Dibbell, who had no flap losses in 64 cases, In Mc Craw's series of 22 patients, 6 suffered partial flap loss and total loss in one patient. [108,109]

“SHORT” GRACILIS MYOCUTANEOUS FLAPS

In a modified short gracilis flap, a smaller skin island is developed and the primary vascular pedicle is deliberately sacrificed. An accessory

BILATERAL GRACILIS MYOCUTANEOUS FLAPS

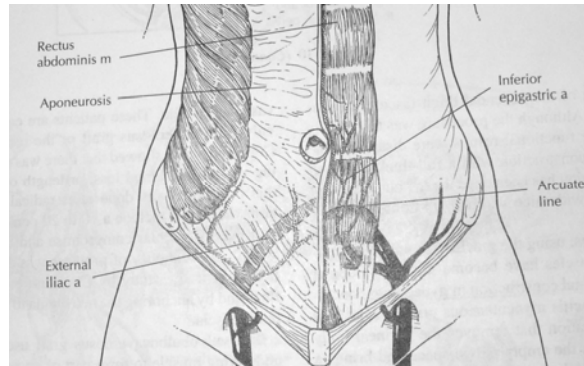


blood supply, derived from anastomotic terminal branches of the obturator and pudendal arteries, enters within the proximal 1 to 3 cm of the gracilis muscle can support the short gracilis myocutaneous flap. A line is drawn on from the pubic tubercle to the medial tibial plateau, along the margin of the adductor longus muscle. The skin island supplied by the gracilis will be located posterior to this line. A 10- to 14- cm long and 5- to 8-cm wide ellipsoid skin island centered over the proximal gracilis muscle will be used for the short flap, with the proximal margin located at the crural fold. [110] The short flap can be considered merely a modification or variation of the long flap, and both forms are often used.

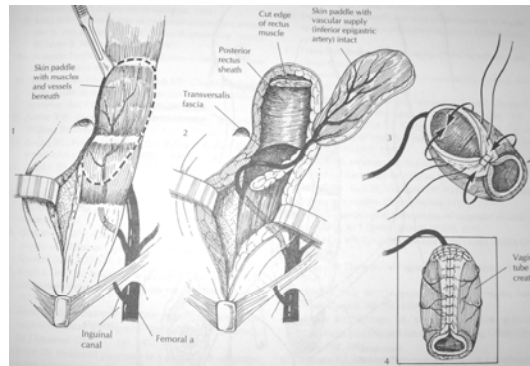
RECTUS ABDOMINIS FLAP

The cutaneous paddle for vaginal reconstruction is wide vertical ellipse centered over the upper half of the muscle. Skin incision is made along with corresponding anterior rectus sheath. The muscle is transected at level of skin paddle at the cephalic end and elevated off the posterior rectus sheath. The vaginal pouch is formed by the inverse tubing of the skin paddle. The flap is passed through the distal midline peritoneal incision into the pelvis, and open end of the vaginal pouch is inset into the vaginal cuff or introitus. [111, 112]

RECTUS ABDOMINIS MUSCLE FLAPS



FLAPS ARE BASED ON INFERIOR EPIGASTRIC VESSELS



THE PADDLE IS 10 TO 20 CM LONG. THE RECTUS ABDOMINIS MUSCLE IS TRANSECTED SUPERIORLY AND DISSECTED FREE FROM ITS FACIAL BED. THE VAGINAL TUBE IS FORMED BY INVERTING THE LATERAL AND INNER MARGINS.

VULVOBULBOCAVERNOSUS FLAP

This technique uses the skin, fat and underlying muscle to form part of a vaginal cylinder. The flaps one from each side are sewn together to form the posterior and lateral walls. The omental pedicle is to form the anterior wall of vagina. A split thickness skin graft is not reportedly required over the omental tissue, as long as the mould is kept in place while the normal epithelisation occurs. [113]

LINING WITH FACIOCUTANEOUS FLAPS

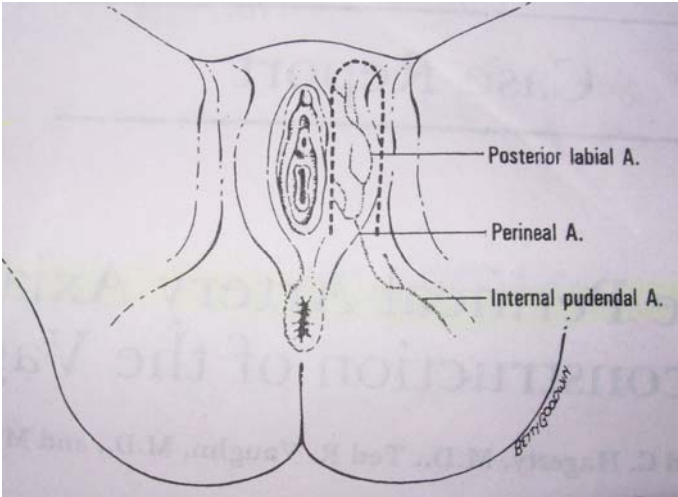
THE PERINEAL ARTERY AXIAL FLAP

By elevating a fasciocutaneous flap that incorporates the perineal artery, one can obtain an axially based flap of thin skin. The subsequent donor defect can be closed primarily. [114]

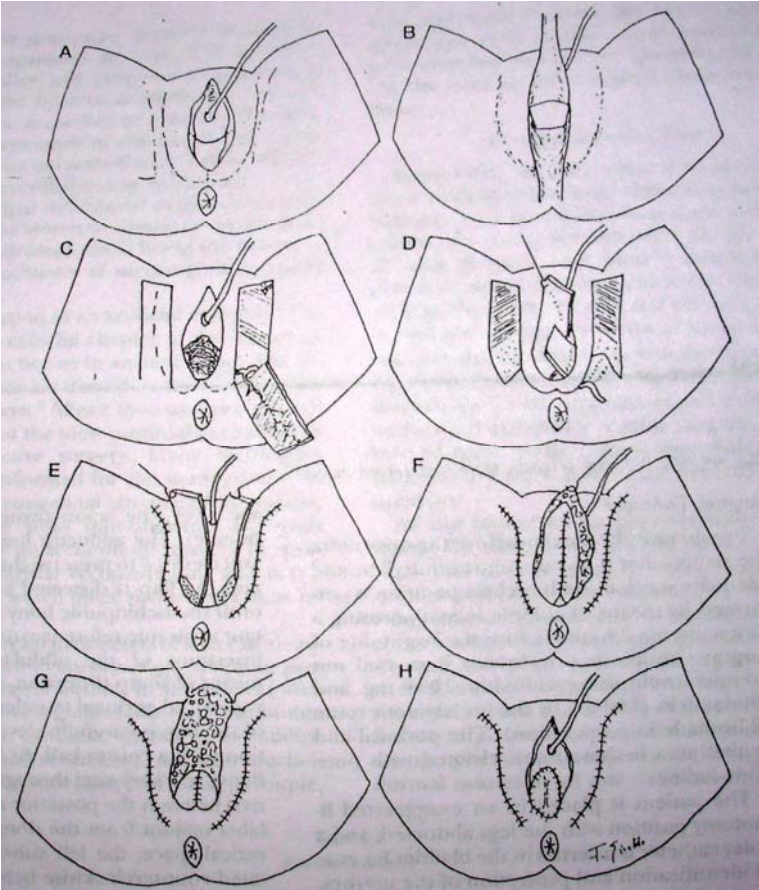
MALAGA FLAP

This vulvo perineal flap is 3-cm wide and 8 to 10 cm long with longitudinal axis centered over the lateral border of the labia majora. The flap is elevated at the lateral and superior margins. The adductor longus aponeurosis is incised and elevated up to the ischiopubic rami where the muscle fibers are sharply detached. The posterior half of the flap is elevated bluntly. Then the flaps are transposed to the defect and sutured together. A 7

PERINEAL ARTERY FLAP



MALAGA FLAP

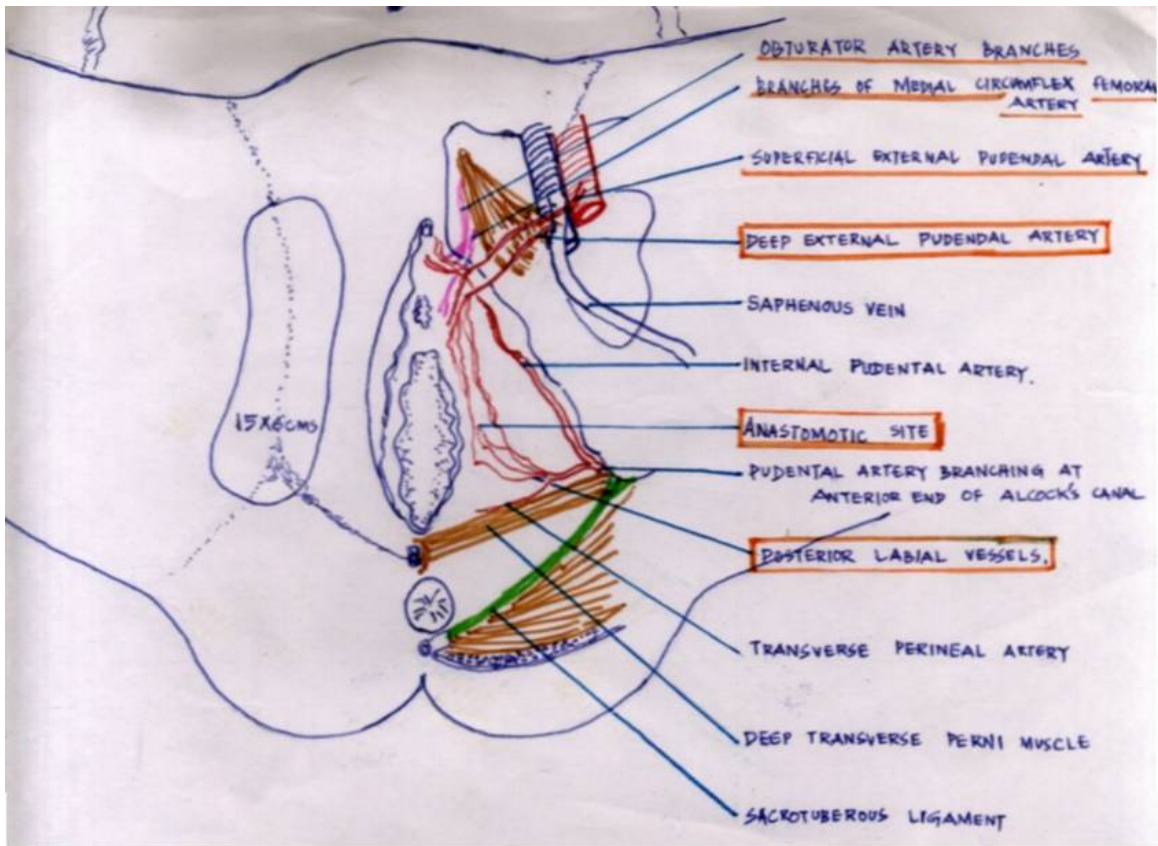


–mm strip of the flap pedicle is de-epithelialised beneath the labial skin and sutured. The donor area is primarily closed. [115-118]

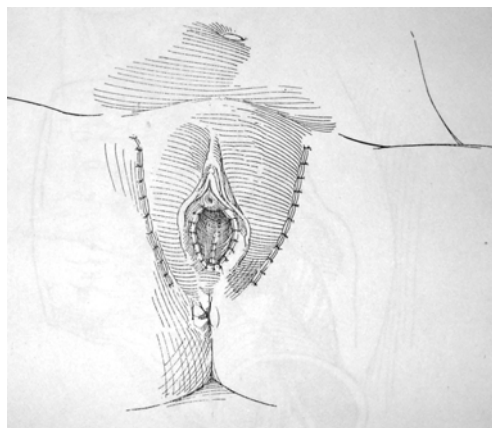
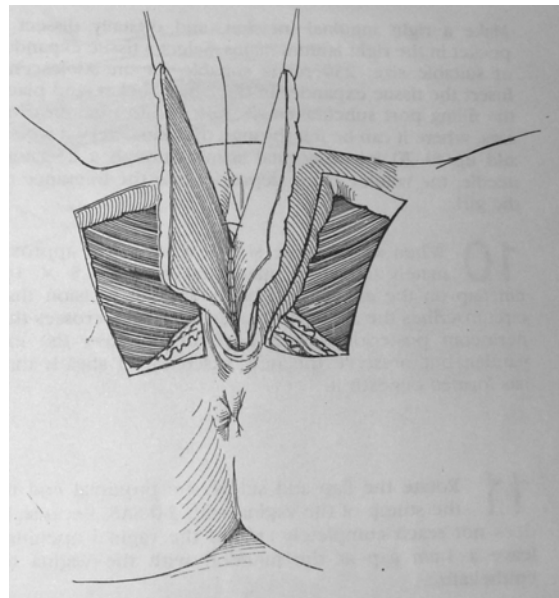
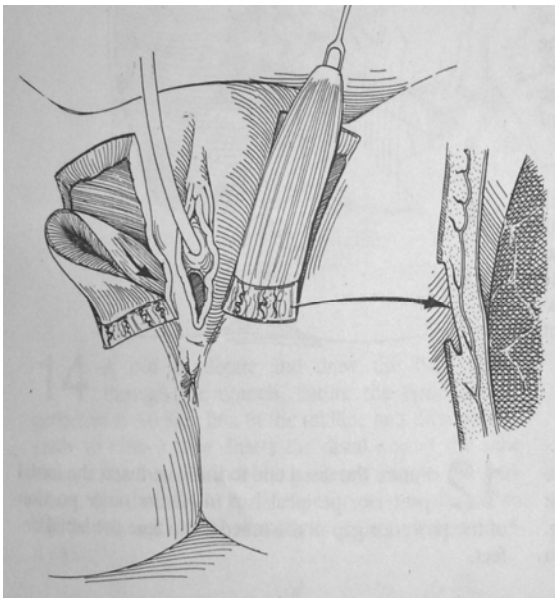
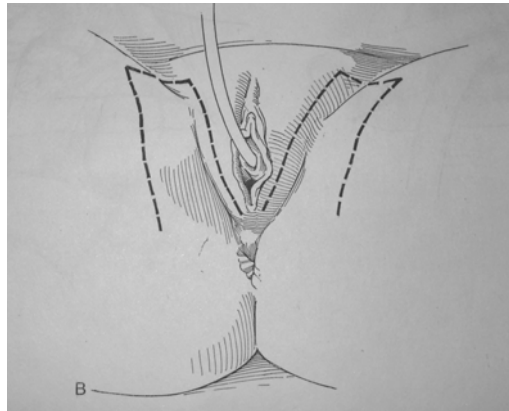
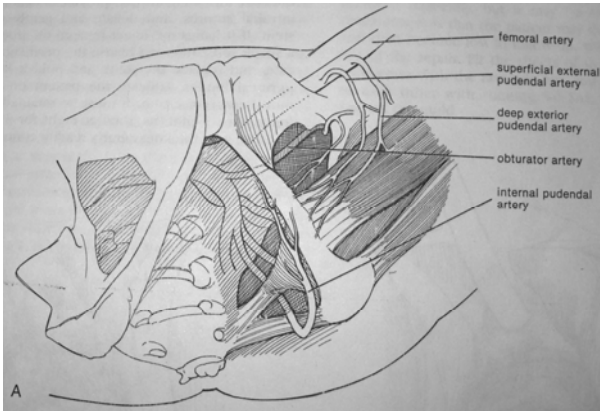
NEUROVASCULAR PUDENDAL-THIGH FLAPS

The internal pudendal artery supplies the perineum by means of its first branch, the inferior rectal artery, which courses through the anal region, and then by means of the perineal artery, which enters the superficial perineal pouch at the bases of the perineal membrane. The perineal artery, after giving off the transverse perineal, continues on as the posterior labial arteries. Pudendal thigh flaps are based on the posterior labial arteries, which are a continuation of the perineal artery. These arteries anastomose with branches of the deep external pudendal artery, as well as the medial femoral circumflex artery, and the anterior branch of the obturator artery over the proximal portion of the adductor muscle. The posterior part of the pudendal-thigh flap retains its innervation from the posterior labial branches from the pudendal nerve, as well as from the perineal rami of the posterior cutaneous nerve of the thigh when it is elevated. The anterior part of the flap near the medial corner of the femoral triangle, supplied by nerve twigs of the genitofemoral and ilioinguinal nerves, may be denervated in the process of elevation. Hence, sensation would be retained only in the lower part of the reconstructed vagina.

VASCULAR ANATOMY AND FLAP DESIGN OF NEURO-VASCULAR PUDENDAL THIGH FLAPS



NEUROVASCULAR PUDENDAL THIGH FLAPS



A skin flap measuring 15 X 6 cm should be essentially horn-shaped. It is planned lateral to the hair-bearing area of the labia major and centered on the crease of the groin. The base of the flap is marked transversely at the level of the posterior end of the introitus. In the adult, the base can be 6 cm wide. This will allow direct closure of the donor site without much undermining.

The flap can measure up to 15 cm in length.

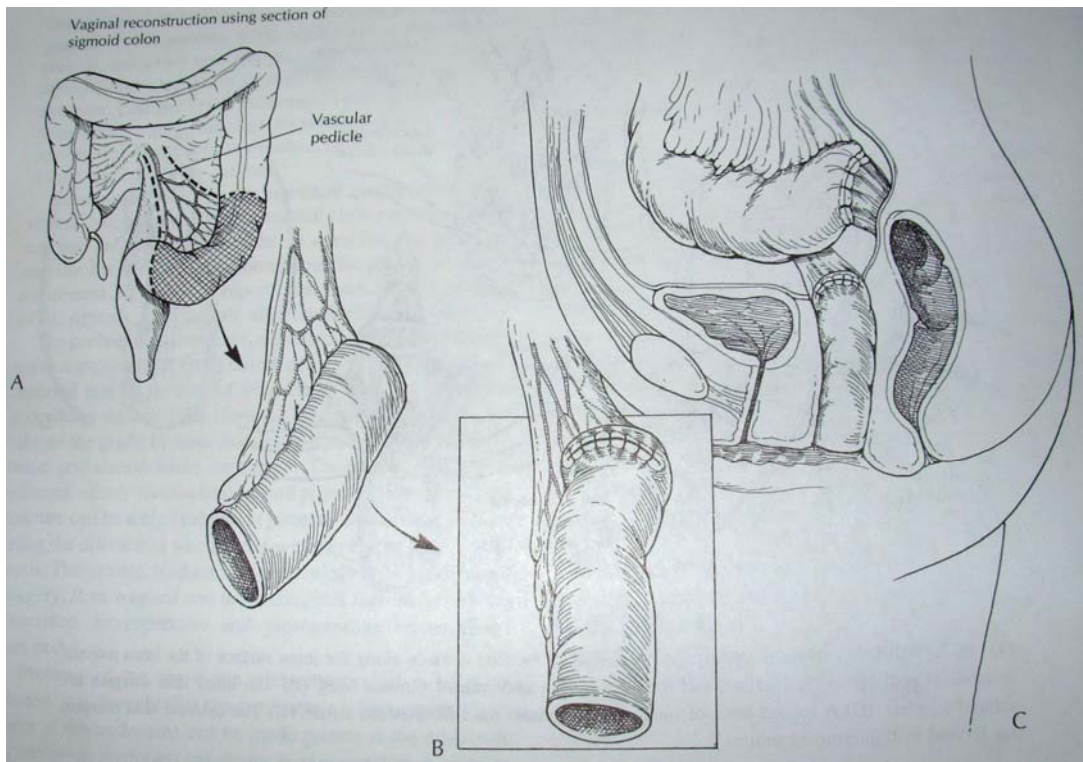
The advantages of this procedure are the following

1. This is the simple technique that can be completed in 2 or 2.5 hours with little blood loss.
2. The blood supply of the flaps is reliable and leads to early wound healing.
3. No stents are needed, since the reconstructed vagina is stable.
4. The angle of inclination of the vagina is physiologic and natural.
5. The linear scars of the donor sites are well –hidden in the groin crease.
6. The reconstructed vagina is sensate and retains the same innervation of erogenous zones of the perineum and upper thigh. [119, 120]

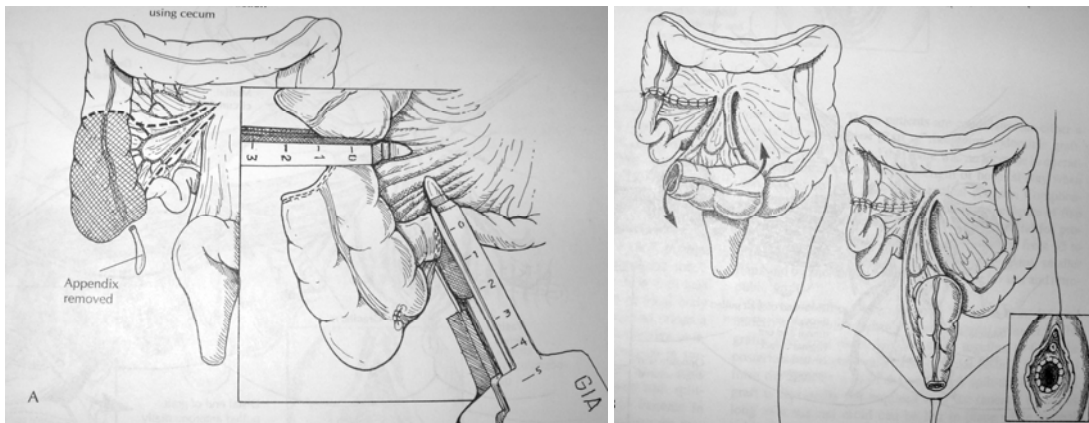
LINING WITH BOWEL SEGMENTS

In 1904, Baldwin described the use of a double loop of ileum to line the space between the bladder and rectum, leaving the mesentery connected to the bowel. [121] In 1910, Popoff constructed an artificial vagina using a

SIGMOID COLON VAGINOPLASTY



VAGINOPLASTY USING CAECUM



portion of the rectum that was moved anteriorly. [122] This operation was modified in 1911 by Schubert, who served the rectum above the anal sphincter and moved it anteriorly to serve as the vagina. The sigmoid colon was then sutured to the anus. The use of the colocecual reconstruction appears to be the most reliable and functional. The caliber is appropriate and it is easy to mobilize. [123-126] The colocecual interposition is performed by isolating the colocecum with its mesentery. An appendectomy is performed and the colonic segment is inverted and sutured to the vaginal introitus. An ileoascending enterocolostomy and closure of the mesenteric defect restores intestinal continuity. Turner-Warwick and Kirby achieved good results with no complications in the three patients in whom they performed this procedure.[127] In Freundt et al's group of patients, who received sigmoid transplants, 25 of 32 patients who were examined 6 weeks postoperatively were found to have a vagina suitable for intercourse. [128] The advantages to these techniques are that shortening does not seem to occur and stenting is not required. The major disadvantage is that these procedures involve a laparotomy and bowel surgery, with all the associated risks. In addition, the bowel tends to secrete a malodorous mucoid discharge and regular douching is required. Some patients develop introital stenosis that requires surgical intervention, and prolapse of bowel out of the introitus has been reported.

[129] Adenocarcinoma and ulcerative colitis can also develop in the transplanted colon. [130-132]

FREE JEJUNAL AUTO GRAFT

Two teams work simultaneously; one team prepares the neo vaginal cavity and recipient blood vessels in the right groin, deep inferior epigastric artery and saphenous vein. The deep inferior epigastric artery is clipped and turned down to the femoral triangle through the inguinal ligament. The jejunal segment is followed to approximately 40 cm below the ligament of Trietz, where the vascular arcades are evaluated by transillumination. Bowel segment is transected and bowel continuity restored. The bowel segment is brought to the vaginal cavity and the vascular anastomosis done. [133, 134]

MATERIALS AND METHODS

All the vaginal agenesis patients who attended our department during the period from August 2004 to April 2007 were included in our study. Only vaginal agenesis cases were included in our study, other cases such as testicular feminization syndrome, ambiguous genitalia, intersex and androgen insensitivity syndrome were excluded from our study. Thorough history taking and physical examination were done to find the cause of primary amenorrhea and associated anomalies. Then the necessary investigations such as Karyotyping, ultra sonogram of abdomen, kidneys and pelvis were done to confirm the diagnosis and to find associated anomalies. The diagnostic laparoscopy was done. Then the patients were given counseling regarding the diagnosis and possible treatment outcomes. After the investigations for the anesthetic assessment, surgery was done.

In our institution we perform three surgical procedures namely Abbe Mc Indoe, Neuro vascular pudendal thigh flaps and Horse shoe shaped labia minora flaps. Patient's abdomen, perineum and thighs were shaved. Lower bowel preparation was given the night before and the morning of the surgery. Under spinal anesthesia patient was placed in the lithotomy position. The whole abdomen perineum and both thighs were prepared, bladder was catheterized.

First step was the preparation of the vaginal cavity. An inverted “V” shaped incision was made in the vaginal dimple. Then a space between the bladder and rectum was created by both blunt as well as sharp dissection. A finger in the rectum will guide the correct plane of dissection. A cavity of size about 10 to 14cms length was created. Hemostasis achieved and the cavity was temporarily packed. Second step of surgery was done by the technique decided pre operatively according to surgeon and patient’s choice.

If the procedure selected was Abbe McIndoe, two sheets of skin grafts were harvested from the medial aspect of inner thigh. The skin grafts were placed inside out with raw area exposed outside over the conformer made of dental wax. Skin grafts were sutured to each other and to the wound edges. Then the labia majora were sutured together. Postoperatively patient was bedridden for five days. On the fifth post operative day under sedation labial sutures were cut, the stent was gently removed, leaving the skin graft intact. The stent was reapplied. The vaginal stent was removed, cleaned and reapplied every other day. The catheter was removed on 8th or 10th post operative day and patient was discharged.

If the Neurovascular pudendal thigh flap procedure was selected two flaps of size about 10 x 4cms was marked on the groin crease lateral to the hair bearing area of the labia majora. The base of the flap was marked

transversely at the level of the posterior end of the introitus. The incision was made through skin, subcutaneous tissue up to deep fascia over the adductor muscle starting from the tip of the flap. Flap was raised in sub fascial plane along with the epimysium of adductor muscle. The deep fascia was tacked to the deep fascia to prevent shearing. The posterior skin margin was incised through the skin only. The labia were elevated off the pubic rami and the flaps were tunneled underneath. Posterior suture line was completed first and then anterior suture line. The tip was anchored to the pelvic structures. The edges were sutured to the mucocutaneous edges to form the vaginal opening. Donor areas were primarily closed. Vaginal plug was kept. The labia were sutured together. Thighs were kept adducted. On the fifth postoperative day labial sutures were removed under sedation flaps were inspected. Vaginal conformer made of dental wax was kept. The patient was discharged on the 8th or 10th postoperative day after removing the catheter.

If the procedure selected was Horse shoe labia minora flap, the entire hairless skin of labia minora is used to create neo vagina. A horse shoe shaped fascio cutaneous flap is marked involving both labia minora and the preputial skin. The base of the flap is close to the introitus posteriorly. Both the medial incisions meet in the midline in a V fashion. The flap is elevated

as a fascio cutaneous flap and sutured together in the midline to form a tube with skin turned inwards using 4 0 vicryl. The tubed flap is tunnelled into the space and retained with an apical vicryl suture. A non adherent gauze stent kept. On the fifth post operative day labial sutures were removed, flaps inspected and stent kept. The patient was discharged on the 8th or 10th postoperative day after removing the catheter.

The patient was advised to remove, wash and reintroduce the conformer daily at home. The patient was advised to come for review at 1st, 2nd, 6th and 12th month postoperatively and then once in a year. The follow up was performed by physical examination and interview. We investigated the post operative results for a total of 14 points, composed of five points for the physical examination and nine points for the questionnaire. By physical examination we determine vaginal cavity depth and width, mucous discharge, malodour and cosmetic configuration. The questionnaire included sexual intercourse, use of lubricant, use of vaginal stent, abdominal pain, pain during intercourse, orgasm, problems in urination, problems in defecation and vaginal bleeding during intercourse.

The proforma for the collection of data was made. All the details of the patient during preoperative, surgical, postoperative and follow up periods were collected and analyzed.

OBSERVATIONS

A total of 24 patients underwent our study. The following observations were made from history taking, examination, investigations, treatment and follow-up.

In our study the youngest patient was aged 12 years and the oldest was aged 30 years (mean age 20 years). 4 patients belonged to 10 -15 age group, 12 patients belonged to 16-20 age group, 4 patients belonged to 21-25 age group, 4 patients belonged to 26-30 age group. The commonest age group was 16 to 20 years. All the patients in our study were reared as females. Phenotypically also they looked as females.

In our study 10 patients (42%) were married and mostly presented within 1 to 2 years after marriage. The longest gap was 8 years after marriage.

The presenting complaints were primary amenorrhea in 24(100%) patients, abdominal pain in 7(29.2%) patients of them 6 had cyclical pain and 1 had continuous pain. None of the patients had H/o vaginal spotting, inguinal swellings, urinary incontinence or hearing loss. 1 patient had H/o quick exhaustion.

All the patients had secondary sexual characters developed. Axillary and pubic hair growth was present. Breast was developed. None had voice change.

10 patients (42%) were married and Coitus was attempted. Coitus was possible in 9 patients. 4 patients complained of pain and 1 patient complained of spotting during coitus.

None of the patients were suffering from Diabetes mellitus, Hypertension, Epilepsy, or any other systemic illnesses. 1 patient underwent cleft lip and palate surgery in her childhood.

None of the family members of the patients were affected by similar illness in our study. History of consanguinity of parents was present in 7 patients (30%), of which 3 patients were born of II* consanguinity and 4 patients were born of III* consanguinity.

One patient had history of exposure to teratogens in the antenatal period (abortifacient? estrogens). All the patients were delivered normally at full term and had normal developmental milestones.

The built, stature and nourishment of the patients were found to be normal for their corresponding age group. 1 patient was 145cms height; 7 patients were between 146 to 150cms height; 7 patients were between 151 to 160cms height; and 9 patients were between 156 to 160cms height. The

CLINICAL FEATURES



ASSOCIATED ANOMALIES



mean height of the patients was 154cms. No patient had webbed neck, tall stature, or features suggestive of Turner's, Klienfelter's or Androgen insensitivity syndrome. Skeletal anomalies were found in 3 patients. (12.5%) 2 patients had thumb duplication and 1 patient had kyphoscoliosis. In our study 1 patient had cleft lip and palate. 1 patient had pre auricular skin tag.

Axillary and pubic hair growth was present. Breast was developed. All the patients had gynaecoid pelvis. External genitalia development was normal. Labia majora and Labia minora were normal. None of the patients had clitoral enlargement. Vagina was replaced by a dimple measuring 1 to 3cms. Urethral meatus was normal. None of the patients had features suggestive of inter sex such as scrotal or inguinal swellings, clitoral enlargement, or voice change.

Other systems and abdomen examination was normal. 1 patient had mass palpable per rectum. Vital signs were normal. None of the patients had hypertension in our study.

Karyotyping was done for all patients. (46xx) pattern was found in all patients. Ultrasound of abdomen and pelvis for evaluation kidney, ureter bladder and female genital tract was done for all the patients. Diagnostic laparoscopy was done for all patients. 8 patients (33.3%) had detectable

TABLE-IV
CLINICAL FEATURES

SL. NO	PATIENT NAME	VAGINA	UTERUS RIGHT	UTERUS LEFT	TUBES RIGHT	TUBES LEFT	OVARY RIGHT	OVARY LEFT	KIDNEY	OTHER ANOMALIES	MULLERIAN	MURCS	VCUAM
1	GOMATHY	A	A	A	H	H	P	P	N	ND	M2:M2	TYP	V5b C2b U4b A1b M0
2	SELVI	A	H	H	P	P	P	P	N	ND	M2:M2	TYP	V5b C2b U3 A0 M0
3	SAPNA	A	A	A	H	H	P	P	N	ND	M2:M2	TYP	V5b C2b U4b A1b M0
4	VIJAYALAKSHMI	A	H	H	P	P	P	P	N	L THUMB DUP & CLP	M2:M2	MU RCS	V5b C2b U3 A0 Ms
5	ALAMELU	A	H	A	H	A	H	A	N	ND	M2:M4	AT YP	V5b C2b U4a A3a M0
6	KOKILA	A	H	H	P	P	P	P	N	R THUMB DUP & PAT	M2:M2	MU RCS	V5b C2b U3 A0 Ms
7	AMBIGA	A	A	A	A	A	H	H	N	ND	M4:M4	AT YP	V5b C2b U4b A3b M0
8	REVATHY	A	P	P	P	P	P	P	N	ND	M1:M1	TYP	V5b C2b U0 A0 M0
9	REVATHY	A	A	A	A	A	H	H	N	ND	M3:M3	TYP	V5b C2b U4b A2b M0
10	ROJARANI	A	P	P	P	P	P	P	N	QUICK EXHAUSTION	M1:M1	TYP	V5b C2b U0 A0 M+
11	JOTHI	A	A	A	H	H	P	P	N	ND	M2:M2	TYP	V5b C2b U3 A1b M0
12	NIVEDHENE	A	A	A	H	H	P	P	N	ND	M2:M2	TYP	V5B C2b U4b A1b M0
13	SHAINAZ	A	H	H	P	P	P	P	N	ND	M2:M2	TYP	V5b C2b U3 A0 M0
14	MANJULA	A	A	A	A	A	H	H	N	ND	M4:M4	AT YP	V5b C2b U4b A2b M0
15	SABITHA	A	A	A	A	A	H	H	N	ND	M4:M4	AT YP	V5b C2b U4b A2b M0
16	VENDAARAM	A	H	H	P	P	P	P	N	ND	M1:M1	TYP	V5b C2b U3 A0 M0
17	SHANTHI	A	A	A	H	H	P	P	N	ND	M2:M2	TYP	V5b C2b U3 A1b M0
18	AKILA	A	P	P	P	P	P	P	N	ND	M1:M1	TYP	V5b C2b U0 A0 M0
19	SOWMIYA	A	A	A	A	A	H	H	N	ND	M4:M4	AT YP	V5b C2b U4B A2b M0
20	SELVI	A	A	P	A	P	A	P	N	KYPHOSCOLIOSIS	M4:M1	MU RCS	V5b C2b U4a A3a Ms
21	ROSSY	A	A	A	H	H	P	P	N	ND	M2:M2	TYP	V5b C2b U4b A1b M0
22	SUSEELA	A	A	A	A	A	H	H	N	ND	M4:M4	AT YP	V5b C2b U4b A2b M0
23	SHEELA	A	A	A	H	H	P	P	N	ND	M2:M2	TYP	V5b C2b U4b A1b M0
24	SUJA	A	H	H	P	P	P	P	N	ND	M2:M2	TYP	V5b C2b U3 A0 M0

TABLE-V

TREATMENT

SL. NO	PROCEDURE	NUMBER OF PATIENTS
1	Non operative	5
2	Abbe McIndoe	7
3	Pudental thigh flaps	10
4	Horse shoe vaginoplasty	2
	Total	24

anomalies in ovaries and 15 patients (62.5%) had anomalies in fallopian tubes, which are more than average percentage quoted in the literature.

5 patients (20.8%) did not want surgery. Of them youngest was 13 years and the oldest was 30 years old. 3 patients were not married and 2 patients were married. All the patients were explained about various treatment modalities available, but they refused surgery. They were very reluctant about trying dilation technique also. They did not come for follow-up after diagnosis. The mean vaginal dimple was 2cms in non married and 3cms in married group. The married patients had attempted sexual intercourse but painful in 1 patient.

7 patients (29.1%) underwent Abbe-McIndoe procedure in our study. The youngest patient was aged 18 years and oldest 22 years. 3 patients were married before surgery. 4 patients were not married and 1 patient got married 3 months after surgery. All 7 patients did not have functioning uterus segment proximally. All the patients underwent surgery under spinal anesthesia and lithotomy position. Split thickness skin graft was harvested from medial side of thigh. The operating time was between 75 to 120 minutes. None required blood transfusion. Graft take was full in all cases. There were no significant postoperative complications except in one patient contracture of cavity occurred which necessitated manipulation. Vaginal

ABBE –McINDOE PROCEDURE

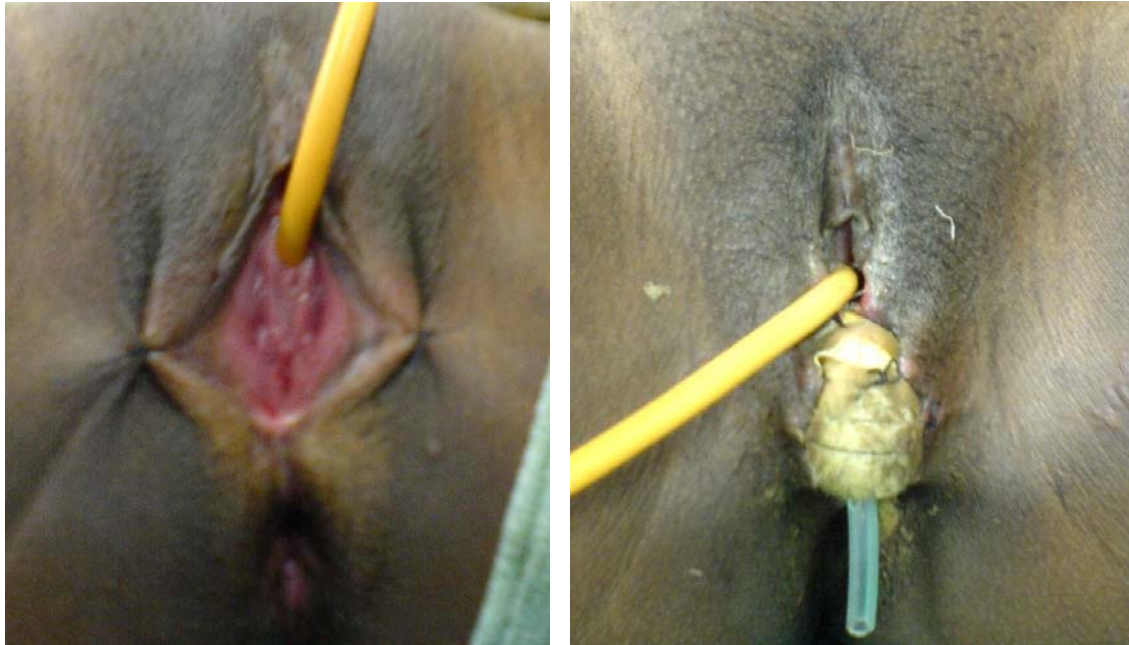
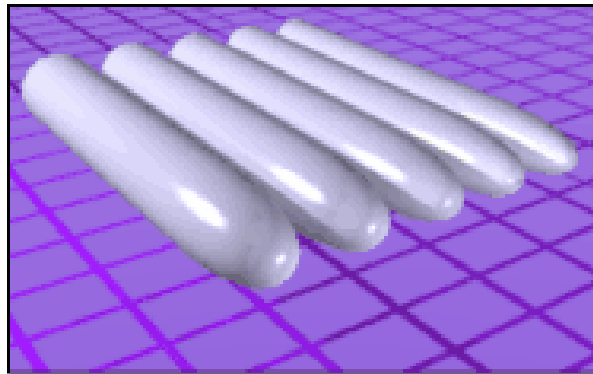


FIG – I NEOVAGINAL SPACE CREATED

FIG-II SPLIT THICKNESS SKIN GRAFT APPLIED



VAGINAL CONFORMER

cavity width obtained was 2cms in all cases. Vaginal cavity depth were 10cms in 1 case, 9cms in 2 cases, 8cms in 3 cases and 7.5cms in 1 case (mean depth- un married 7cms & married 8cms). Mucus discharge and malodor were present immediately after surgery, but reduced to minimum with a month. Cosmetic configuration was good in all cases. All 3 married patients and 1 patient got married 3 months after surgery started sexual intercourse 3 months to 6 months post operatively. Lubrication was needed in all cases. All married patients used stent for 3 to 5 months postoperatively. All married patients discontinued stent at 6 months postoperatively. None of the married patients had abdominal pain or pain or bleeding during inter course. 1 patient only had orgasm during inter course. None had difficulties in defecation or urination. All 3 unmarried patients used lubricants and stent at 6 months post operatively. None had difficulties in defecation or urination. 1 patient, who had abdominal pain preoperatively, had pain after surgery. 1 patient only came for follow-up after 12 months, 1 patient came up to 12 months, and remaining 5 patients came only up to 6 months.

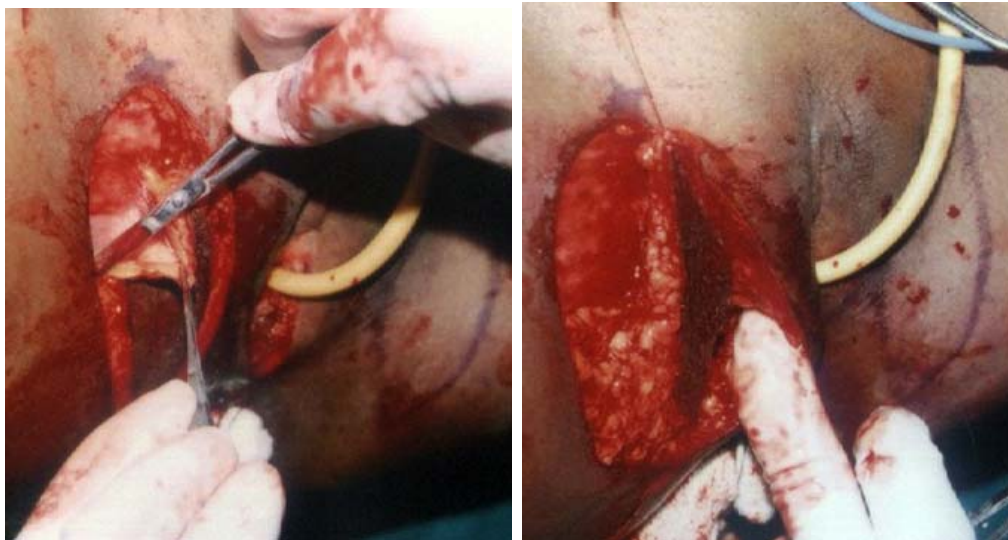
10 patients (41.6%) underwent pudendal thigh flap surgery. The youngest patient was 12 years and the oldest was 30 years old. 6 patients were unmarried and 4 patients were married. All the patients underwent surgery under spinal anesthesia in lithotomy position. The operating time

was between 120 to 150 minutes. Blood transfusion was not necessary. Flaps were posteriorly based in 9 patients and anteriorly based in 1 patient. Flaps measured 8 to 10cms in length and 3 to 4cms in breadth. The smallest flap was 8cms x 3cms and the largest flap was 10cms x 4cms in dimension. All the flaps survived no flap necrosis. Complications were 1 case of donor wound dehiscence for which secondary suturing was done, 1 patient had hematocolpos which required drainage and 1 patient developed pelvic abscess two months later which required drainage. 1 patient had menarche 3 months after surgery. The mean vaginal depth obtained was 8cms and width was 2cms. 1 patient had persistent mucus discharge and malodor. The cosmetic appearance of the neo vagina was fair, the color of the flap skin was obviously noticeable, the hair growth was not appreciable and the donor scar was noticeable. All 4 married patients started sexual intercourse 4 to 5 months after surgery. All were using lubricants but discontinued stenting. None of the married patients had abdominal pain, pain or bleeding during intercourse, orgasm or difficulties in urination and defecation. 3 unmarried patients had abdominal pain post operatively. All the patients came for follow-up at 6 months, but only one patient came for follow up after that time.

NEURO VASCULAR PUDENDAL THIGH FLAPS



FLAP MARKING



RIGHT SIDE FLAP ELEVATION



CREATION OF NEOVAGINAL SPACE

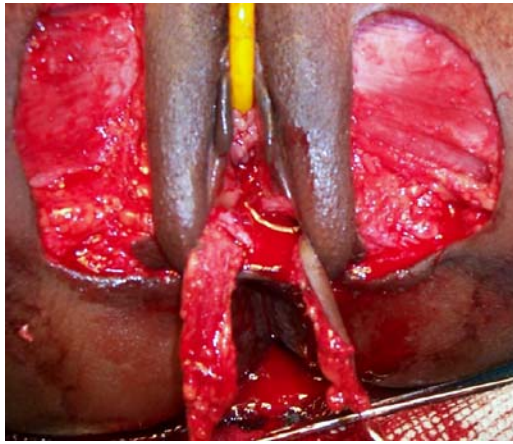


FLAPS SUTURED



DONOR AREA PRIMARILY CLOSED

MORE CASES

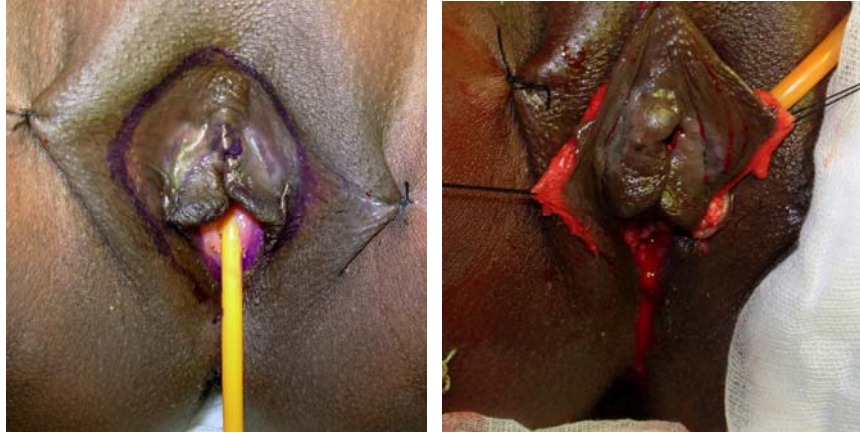


FLAP MODIFICATION - ANTERIORLY BASED FLAPS

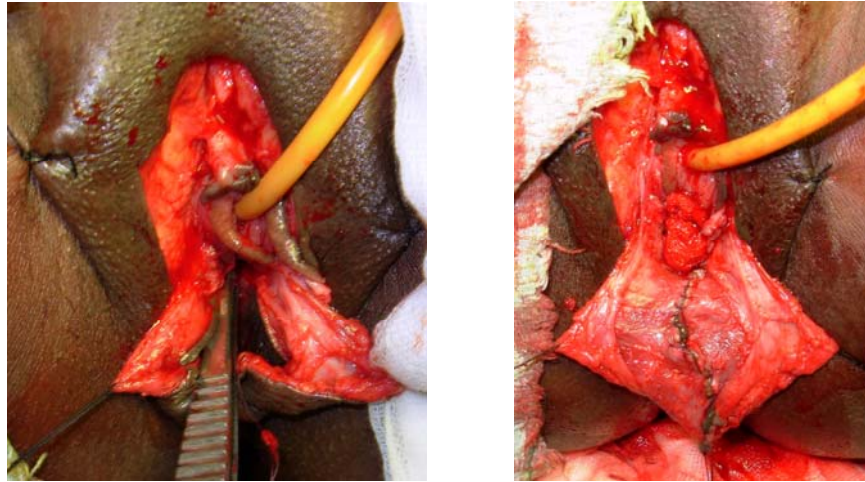


2 patients (8.3%) underwent horse shoe shaped labia minora flaps. 1 patient was unmarried and aged 20 years, and 1 patient was married and aged 28 years. Both patients underwent surgery under spinal anesthesia in lithotomy position. The operating time was between 150 to 180 minutes. None required blood transfusion. For the unmarried patient 8cms x 3cms flap was taken and the flap survived without any complications. The postoperative vagina was 8cms deep and 2cms wide. Mucus discharge and malodor was present. The cosmetic appearance was fair because of the distortion of the labia but no donor scar was seen. The patient was using the lubricant and stent. For the married patient 8cms x 3cms flap was taken but post operatively contracture of vagina occurred. Mucus discharge and malodor was present. The depth of the resultant vagina was 6cms and width was 2cms. The cosmetic appearance was fair because of the distortion of the labia but no donor scar was seen. The patient was using the lubricant and stent. These patients were recently operated and under follow up care.

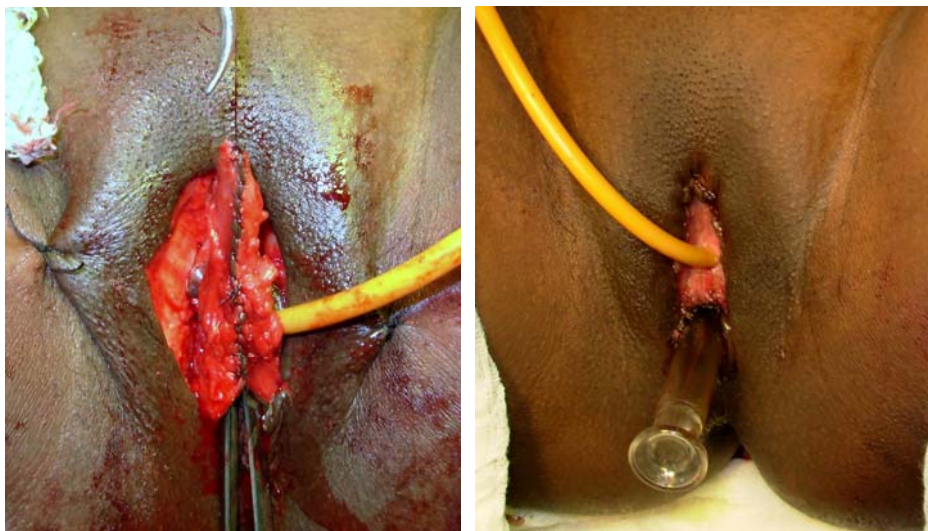
HORSE SHOE SHAPED LABIA MINORA FLAPS



FLAP MARKING AND ELEVATION



NEOVAGINAL SPACE CREATION



FLAPS TUBULARISED & INSERTED, DONOR AREA CLOSED

FOLLOW UP RESULTS



ABBE- Mc INDOE AT 24 MONTHS POST OPERATIVELY



PUDENDAL THIGH FLAPS AT 12 MONTHS POST OPERATIVELY

TABLE-VII

COMPARISION OF CLINICAL FEATURES

STUDY	PATIENTS	OVARY	FALLOPIAN TUBE	RENAL SYS	SKEL SYS	NERVE SYS	INGUINAL HERNIA	HEART	TYP	ATYPICAL	MURCS
Plevraki et.al. (2004)	6	3	4	3	6	ND	ND	ND	0(0)	1(17)	5(83)
Griffin et.al.(1976)	14	0	0	7	10	ND	ND	1	5(36)	2(14)	7(50)
Chervenak et.al.(1982)	7	1	2	7	3	ND	ND	ND	3(43)	1(14)	3(43)
Hauser & schreiner (1961)	21	0	0	3	2	0	4	1	15(72)	3(14)	3(14)
Schmid-tannwald & hauser (1977)	33	1	12	7	ND	ND	ND	ND	26(79)	7(21)	0(0)
Seifert et al.(1974)	34	ND	ND	7	4	ND	5	ND	25(74)	5(15)	4(11)
Wabrek et.al.(1971)	10	0	0	3	ND	ND	ND	ND	7(70)	3(30)	0(0)
Yenen (1957)	18	ND	ND	ND	ND	ND	1	ND	18(100)	0(0)	0(0)
Reindollar et. al. (1981)	37	ND	ND	12	3	ND	2	1	23(62)	10(27)	4(11)
Smith (1983)	22	ND	ND	11	3	ND	8	ND	10(45)	9(41)	3(14)
Darai et .al. (2003)	7	ND	ND	3	ND	ND	ND	ND	4(57)	3(43)	0(0)
Brun et.al (2002)	25	0	0	7	1	0	ND	ND	18(72)	6(24)	1(4)
Fedele et.al.(2000)	52	0	ND	16	5	ND	ND	ND	34(65)	13(25)	5(10)
Creatsas et.al.(2001)	111	1	ND	43	9	5	ND	ND	63(57)	39(35)	9(8)
Gauwerky et.al.(1997)	47	ND	ND	11	ND	ND	ND	ND	36(77)	11(23)	0(0)
Palatynski(1986)	24	3	9	3	ND	ND	ND	ND	21(87)	3(13)	0(0)
Oppelt et.al.	53	6	6	23	19	2	7	3	25(47)	11(21)	17(32)
Total ref	521	15 (3)	33(6)	166 (32)	65 (12)	7(1)	27 (5)	6(1)	333(64)	127(24)	61(12)
OUR STUDY	24	8 (33.3)	15(62.5)	ND	3(12.5)	ND	ND	ND	15(62.5)	6(25)	3(12.5)

DISCUSSION

A total of 24 patients underwent our study. In our study the youngest patient was aged 12 years and the oldest was aged 30 years (mean age 20 years). The commonest age group was 16 to 20 years. John A Rock describes the commonest presentation are usually at the age of 14 to 15 years by the gynecologist. [7] Lisa Jane describes the commonest age of presentation is 15 to 18 years. [30] The average of discovery is 17 years in other studies [135]. Whatever may be the age of presentation, the referral to plastic surgeon is usually from the gynecologist as in our study. Previously the opinion regarding the ideal age for surgery was considered as just before marriage or when regular sexual activity was anticipated. The introduction of flap procedures had changed the management to an earlier age, probably during her teenage itself so as to psychologically adjust to the world and also to improve the self esteem of the patients.

As Secondary sexual characters and external genitalia were feminine, all the patients were reared as females. Psychologically also the patients felt themselves as females. Similar observation was made by the other studies also. [7, 4, 30] The importance of psychological aspects of understanding the patient's emotions and psychological preparation of patients and their parents was elaborately discussed in various studies. [2, 58, 59, 60]

In our study 10 patients (42%) were married and mostly presented within 1 to 2 years after marriage. The longest gap was 8 years after marriage. Primary infertility was the main concern which led them to the treatment. This is in contrast with the western world, where the commonest age of diagnosis is 14 to 15 years when the patient is investigated by the gynecologist for primary amenorrhea. Coitus was attempted in 10 patients (42%) who were married. Coitus was possible in 9 patients. 4 patients complained of pain and 1 patient complained of spotting during coitus.

Primary amenorrhea was the presenting complaint of all 24 patients in our study. John A Rock, Lisa Jane, Peter Oppelt, Tarry W.F and others have stated that primary amenorrhea is the presenting symptom of MRKH syndrome. Sometimes abdominal pain and mass may present the disease earlier than usual age. [7, 4, 30, 135, 136]

Cyclical lower abdominal pain was present in 7 patients (29%). On subsequent investigations these patients were commonly found to have developed upper 2/3rd of uterus and sometimes functioning ovaries. John A Rock describes that the abdominal pain without pelvic mass could be an ovulatory pain or possibly as a result of dysmenorrhea originating in well developed rudimentary uterine bulbs. [7, 4, 30, 135, 136]

History of consanguinity of parents was present in 7 patients (30%), of which 3 patients were born of II* consanguinity and 4 patients were born of III* consanguinity. None of the family members of the patients were affected by similar illness in our study. Occasional familial occurrence of vaginal agenesis was reported but that was explained by Griffin as could be due to different interpretation. Some cases may be due a hereditary abnormality or may be due to a mutant gene transmission. [21, 22, 23, 25, 26, 55, 56] Alternatively this may be only one manifestation of a variably expressed genetic defect. So the real frequency of familial involvement has been under estimated. Petrozza and others in a retrospective study in 1997, attempted to determine an inheritance pattern of the syndrome. They concluded that congenital absence of the uterus and vagina was not commonly inherited in a dominant fashion and is likely via a polygenic mechanism. [21]

One patient had history of exposure to teratogens in the antenatal period (abortifacient? estrogens). Some investigators had suggested possible association of antenatal exposure to Di ethyl stilbestrol with vaginal agenesis. [137] This supports the ideological concept of effect of teratogenic exposure between 37th and 41st gestational day during which vagina is formed.

The mean height of the patients was 154 cms. The built, stature and nourishment of the patients were found to be normal for their corresponding age group. No patient had webbed neck, tall stature, or features suggestive of Turner's, Klienfelter's or androgen insensitivity syndrome.

Skeletal anomalies were found in 3 patients. (12.5%) 2 patients had thumb duplication. 1 patient had kyphoscoliosis. This is comparable with other studies which also show an average of 12% skeletal anomalies. (Table-v) The skeletal malformations observed in other studies include spina bifida, sacralisation of sacral bone and malformations of cervical vertebrae. Similarly syndactyly, rib deformities, shoulder and pelvic anomalies were described in various studies. Ventricular Septal defects, hearing loss, situs inversus were also reported in literature. [40-54]

In our study 1 patient had cleft lip and palate. 1 patient had pre auricular skin tag. 1 patient had history of quick exhaustion. No patient had urinary incontinence or hearing loss.

All the patients had secondary sexual characters developed. Axillary and pubic hair growth was present. Breast was developed. All the patients had gynaecoid pelvis. External genitalia development was normal. Labia majora and Labia minora were normal. None of the patients had clitoral enlargement. Vagina was replaced by a dimple measuring 1 to 3 cms.

TABLE-VII

COMPARISION OF CLINICAL FEATURES

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Urethral meatus was normal. None of the patients had features suggestive of inter sex such as scrotal or inguinal swellings, clitoral enlargement, or voice change.

Other systems and abdomen examination was normal. 1 patient had mass palpable per rectum. Vital signs were normal. None of the patients had hypertension in our study.

Karyotyping was done for all patients. (46xx) pattern was found in all patients. Various articles quote that, Karyotyping will be (46xx) for these patients. [4, 7, 30, 54] Ultrasound of abdomen and pelvis for evaluation kidney, ureter and bladder region and female genital tract development was done for all the patients. Diagnostic laparoscopy was done for all patients and the ultra sonographic examination findings were correlated. Various anomalies were found in the development of ovaries, fallopian tubes, uterus and vagina. But none of our patients had detectable anomalies of urinary tract. 8 patients (33.3%) had detectable anomalies in ovaries and 15 patients (62.5%) had anomalies in fallopian tubes, which are more than average percentage quoted in the literature. (Table-v) Female genital tract anomalies were detected more but no urinary tract anomalies were detected in our study, probably because we did not use any MRI examinations or Intra venous pyelogram examinations specifically for urinary tract. Urinary tract

TABLE-VIII

**NEW RECOMMENDED INVESTIGATIONS FOR DIAGNOSIS OF
MRKH SYNDROME (Peter Oppelt et. al.)**

Essential examinations

- Chromosome analysis
- MRI of the kidneys and pelvis
- Hormone status (LH, FSH, estradiol)

Additional examinations

- Ultrasound of the vaginal vestibule, rectum
- Diagnostic laparoscopy
- Ovarian biopsy

Supplementary examinations

- Urinary incontinence-urodynamics
- Quick exhaustion-myography, echocardiography
- Skeletal malformations-radiography, computed tomography

Hearing loss-audiography

anomalies on an average of 32% were reported in the literature. (Table-v)
Newer recommendations of investigations are proposed by Peter Oppelt for complete evaluations of associated anomalies (Table-VI). [54]

Many different names for this disease were given by primary referring physicians. So we analysed the comprehensiveness of various classification systems in the description of disease status and its usefulness in diagnosis, comparison and management of the disease. Monie and sigurdson proposed a classification system called “M” system and Tarry WF made modifications in 1986. [4, 138] It codes each anomaly of corpus and cervix of uterus and vagina. Second system of classification is called as MURCS classification proposed by Schmid-Tannwald and Hauser (1977) and Duncan et al. (1979). (42, 139) Third classification system used was called as VCUAM classification by Peter Oppelt et al. [54, 140]

After the evaluation with investigations we classified the patients according to all three systems. We found that in “M” classification the finer differentiation between cervix, upper and lower parts of uterus were not possible; all of them were collectively called as M2 stage. Some times uterine hypoplasia was associated with ovarian hypoplasia, which could not be differentiated by this method. Associated renal anomalies could not be noted.

In our study 15 patients (62.5%) belonged to typical group, 6 patients (25%) to atypical and 3 patients (12.5%) to MURCS group according to MURCS classification. This is comparable with the incidence of this combination in literature (Table-V) This system is easier to use but it has similar disadvantages of “M” classification. Various combinations of anomalies and differences between uterus, cervix and tubes could not be made out. Even though it includes other anomalies it puts everything in one group.

We used VCUAM classification in our study. Like TNM classification for malignancy, this system can be used to describe the anomaly. It is possible to obtain a precise description of the condition and associated anomalies. It is useful in the diagnosis and management of vaginal agenesis. This VCUAM classification gives even more details than diagrammatic representation of the anomalies.

24 patients under went the study. 19 patients under went surgical treatment. All patients were operated under spinal anesthesia in lithotomy position in our study.

5 patients (20.8%) were not willing for surgical management, non operative methods were offered for these patients. But none of these 5 patients came for follow-up. Ingram had 20 successful and 6 failed cases

between 1975 and 1980. The technique took 4 to 6 months to achieve an adequate vagina. [66] Roberts CP et al in a study reveal that passive dilation with Ingram method is capable of creating an adequate vaginal canal in patients with vaginal agenesis, with respect to both function and anatomy even those patients with a previous hymenectomy and resultant scar. 91.9% patients achieved anatomic and functional success. Passive dilation failed in 8.1% patients. The mean time to successful dilation was 11.8 +/- 1.6 months with a range of 3 to 33 months. [67] Usually there are no complications with this technique. Prolapse of neo vagina and enterocele has been reported. [141]. We found this technique is not acceptable procedure in our patients, as this need a highly motivated patient with good intelligence. More over it takes considerable time to form vagina.

7 patients (29.1%) underwent Abbe-McIndoe procedure in our study. The operating time was between 75 to 120 minutes. Graft take was full in all cases. Graft take were 96% in Alessandrescu et al 1996 study [88] and 90.6% in Seccia et al 2002 study [142]. Our study results were compared with other studies in Tables- IX & X. The mean vaginal depths were 7cms in unmarried group and 8cms in married group which is also comparable to other studies. [81- 91, 153] This procedure is used as life boat in failures of dilation therapy [67, 153] and labia minora flaps. [152]. This procedure was

TABLE- IX

COMPARISION OF McIndoe PROCEDURE

Sl. No	Study [ref no]	No of cases	Graft take in %	complications
1	Alessandrescu et al 1996 (88)	201	96	Rectal perforation-1% Graft infection-4% Donor infection-5.5%
2	Seccia et al 2002(142)	32	90.6	Graft partial take -9.4% Anxiety -6.3% Donor site keloids- 3.1%
3	Cali & Prat 1968(90)	113	-	Contracture- 42%
4	Ortiz & monasterio1972 (91)	21	-	Contracture-19%
5	Buss & Lee 1989(89)	50	-	Revision procedure-10% Contracture- 15%
6	OUR STUDY 2007	7	100	Contracture- 14.3%

also compared with other procedures like Williams procedure [75]. In one study, the conversion to full thickness skin graft harvested from excess abdominal skin during Abdominoplasty was done in 6 male to female transsexuals to tackle contracture. [154] New innovations in McIndoe procedure are the use of vacuum assisted closure [155], vacuum expandable mould [156], fibrin glue [157] and inflatable stent [158].

10 patients (41.6%) underwent pudendal thigh flap surgery. The operating time was between 120 to 150 minutes same as Wee's study [119]. Flaps were posteriorly based in 9 patients and anteriorly based in 1 patient. In anatomical study of 10 cadavers (20 sides) found that there are multiple blood supplies to the flap. Anterior cutaneous branches of obturator artery spread out on middle portion of the flap. Piercing site of it is (3 +/- 0.5) cm apart from the middle line of perineum, (1.7 +/- 0.4) cm from the anterior margin of vaginal introitus, (0.6 +/- 0.2) cm from outer fringe of the lower pubic rami and (0.8 +/- 0.1) mm in diameter. Posterior labial arteries mainly supply the labia majora and constantly anastomose with superficial external pudendal arteries in direct vascular anastomosis in sub cutis of labia majora. The main stems send out 2-3 lateral branches posterior labial arteries which distribute to posterior portion of the pudendal thigh flaps. Their conclusion was the major vessels are the lateral branches of posterior labial arteries and

TABLE- XI

COMPARISION OF PUDENDAL THIGH FLAP PROCEDURE

SL NO	Study [ref no]	No of cases	Flap survival (%)	complications
1	Wee & Joseph 1989(119)	3	100	Nil
2	Li et al 2000(146)	12	91.6	Total flap necrosis-8.3%
3	Chen 1991(147)	4	100	Nil
4	Gurlek 2002(148)	31	100	Wound dehiscence-6.4%
5	Li & cheg 2003(149)	47	100	nil
6	Selvaggi2003(150)	2	100	Granulamatus polyp-50% Stenosis- 50%
7	Sanchas 2006(151)	33	100	Hematometra-9%
8	OUR STUDY 2007	10	100	Wound dehiscence-10% Hematometra-10% Pelvic abscess-10%

TABLE- XII

COMPARISION OF HORSE SHOE FLAP PROCEDURE

SL NO	Study [ref no]	No of cases	Flap survival (%)	complications
1	Purushothaman2005(103)	15	100	Nil
2	Hwang 1985(152)	10	100	Additional SSG-30%
3	Flack 1993(102)	3	100	Nil
4	OUR STUDY 2007	2	100	Stenosis- 50%

not the main stem itself. Flaps pedicled on the anterior cutaneous branches of the obturator arteries may be elevated to repair rectovaginal fistula or small defects of perineum and are not suited to reconstruct a neo vagina as its piercing point of blood vessels is much higher and the location of the pedicle is more fixed. [159] But in our study we did 1 anteriorly based flap of size 8cms x 3 cms for vaginal reconstruction. Flap survived well without any complication. The mean vaginal depth obtained was 8cms and width was 2cms. A flap of size was 14cms x 5cms demonstrated by Li et al [146, 160]. Laparoscopy can be used to protect vaginal depth [161]. All the flaps in our study survived. 1 patient had persistent mucus discharge and malodor. The cosmetic appearance of the neo vagina was fair, the color of the flap skin was obviously noticeable, the hair growth was not appreciable and the donor scar was noticeable. Complications were 1 case of donor wound dehiscence for which secondary suturing was done, 1 patient had hematocolpos which required drainage and 1 patient developed pelvic abscess two months later which required drainage. Our study was compared with other studies in Table-XI. Hair growth in our series of was not noticeable, but other studies have shown obvious appearance. [162, 163] 1 patient had menarche 3 months after surgery. Restoration of utero vaginal

continuity is possible with flap procedures [150]. New innovations like use of fibrin glue has been described [164]

2 patients (8.3%) underwent horse shoe shaped labia minora flaps. The operating time was between 150 to 180 minutes. The vagina depth was 8cms in 1 patient and 6cms in 1 patient in which contraction has occurred. Purushothaman has advised the use of dilator for 3 months post operatively for satisfactory vaginal depth. Flack et al had vaginal depths of size 7, 5 & 7cms in three patients immediately after surgery and achieved 10, 9 & 7cms after dilation [102]. Because of smaller size of flaps Hwang et al suggested additional use of split skin graft with labia minora flaps [152].

The operating times were 75 to 120 minutes for McIndoe procedure, 120 to 150 minutes for pudental thigh flaps and 150 to 180 minutes for labia minora flaps. The operating time is least for McIndoe procedure because of simplicity and longer for horse shoe flaps because of difficult dissection.

The mean vaginal depth was 7cms for unmarried and 8cms for married patients who under went McIndoe procedure. The mean vaginal depth was 8cms for patients who under went pudental thigh flaps. The vaginal depth were 8cms and 6cms for patients who under went labia minora flaps. A survey of text books and medical papers in the mid-1990's revealed average quoted lengths ranging from 9 to 11cms for the longest (posterior)

TABLE- XII

COMPARISION OF OUR SURGICAL PROCEDURES

Sl no	Observation	McIndoe(7)	Pudental(10)	Horse shoe(2)
1	Operating time in minutes	75 to 120	120 to 150	150 to 180
2	Mean vaginal depth in cms	UM-7,M8	8	UM-8,M-6
3	Cosmetic appearance	Good	Fair	Fair
4	complications	Contracture-1	Wound dehiscence-1 Hematometra -1 Pelvic abscess-1	Stenosis- 1

wall in normal XX women. Shah and Woolley give the normal length as greater than or equal to 6cms. In our study satisfactory vaginal length was achieved. The vaginal depth tends to decrease over time in unmarried group especially when the patient was irregular in using vaginal stent. For patients who underwent labia minora flaps, regular dilation programme is a must.

Cosmetic appearance was good in patients who under went McIndoe procedure as the genitalia was not distorted. Donor area scar was acceptable or skin graft can be taken from hidden regions. Cosmetic appearance was fair in patients who under went pudental thigh flaps because the donor area scars and flap's skin color were noticeable. Cosmetic appearance was fair in patients who under went horse shoe flap procedure because the external genitalia was distorted.

Contracture rate was more in patients who under went McIndoe procedure especially who were irregular in using stents. No other significant complications occur with this procedure. For well executed pudental thigh flaps complications were very minimal. Horse shoe flaps are technically difficult to execute and also the depth obtained was not satisfactory. Contracture rate was very high as well as patients need to be well motivated for post operative dilation programme.

CONCLUSION

After this study the following conclusions were made.

1. Our patients present little later than the western world and a considerable number of patients present late even after marriage.
2. All the patients were phenotypically and genetically 46XX females with normal secondary sexual characters.
3. The renal anomalies were not detected as frequently as in the literature.
4. The terminology used for diagnosis by the primary referring physicians was not consistent.
5. The VCUAM classification gives better diagnosis and useful in the management.
6. Only 79.2% of patients accepted surgical treatment, the remaining 20.8% patients did not accept surgical treatment or dilation therapy.
7. The Abbe-McIndoe procedure was simple, safe procedure with good aesthetic results and had least complication rate.
8. The pudendal thigh flap procedure was simple, safe procedure with fair aesthetic results and had acceptable complication rate.
9. The Horse shoe flap procedure was technically difficult procedure with fair aesthetic results and had high contraction rate.

10. Most of our patients were very irregular in follow up and stent usage.

So, flap procedure especially pudendal thigh flaps which has the least contraction rate is the preferable treatment option for such patients.

BIBLIOGRAPHY

1. Fligner, J. R. congenital atresia of the vagina. *Surg.Gynecol. Obstet.* 165: 387, 1987.
2. Evans TN, Poland ML, Boving RL. Vaginal malformation. *Am J Obstet Gynecol* 1981; 141: 910-20.
3. Jones, K. L. *Smith's Recognizable patterns of Human Malformation.* Philadelphia: Saunders, 1988.
4. Tarry, W. F., Duckett, J. W., and Stephens, F. D. The Mayer-Rokitansky syndrome: Pathogenesis, classification and management. *J. Urol.* 136: 648, 1986.
5. Huffstadt A. J. C. *congenital Malformations.* Amsterdam: Excerpta Medica, 1980. P. 303.
6. American fertility society classification of mullerian anomalies. *Fertile steril* 1988; 49:952.
7. John A Rock., Lesley L Breech. *Surgery for the anomalies of the mullerian ducts.* Te Linde's operative Gynaecology 9th edition. Lippincott Williams & Wilkins, Philadelphia. 2003.
8. Thompson JD, Wharton LR, TeLinde RW: Congenital absence of the vagina: an analysis of 32 cases corrected by the McIndoe operation. *Am J obstet Gynecol* 74:397, 1957
9. Sadler TW: *Langman's medical Embryology.* Williams & Wilkins, Baltimore, 1990
10. Ulfelder H, Robboy SJ. The embryologic development of the human vagina. *Am J Obstet Gynecol* 1976; 126:769.
11. Boving RL, Pelusa JJ, Boving BG: Estrogen binding sites in genital tracts of human female fetuses. *Anat Rec* 190:344,1978
12. Mayer, C. a. J.: *Über verdoppelungen des uterus und ihre Arten, nebst Bemerkungen Über Hasenscharte und Wolfsrachen.* *J. Chir. Auger,* 13:525, 1829.
13. Rokitansky, K.: *Über die sogenannten verdoppelungen des uterus.* *Med. Jahrb. Ost, Staat.* 26:39,1838.
14. Kuster, H; *uterus bipartitus solidus rudimentarius cum vagina solida.* *Z. Geb. Gyn.,* 67: 692, 1910.
15. Hauser GA, Keller M, Koller T. *Das Rokitansky-Kuster syndrome. Uterus bipartitus solidus rudimentarius cum vagina solida.* *Gynecologia* 1961; 151:111.
16. Jones HW: *Reconstruction of congenital uterovaginal anomalies.*p. 246. in Rock JA (Ed): *Female Reproductiva Surgery.* Williams & Wilkins, Baltimore,1992
17. Rock JA, Jones HW. The double uterus associated with an obstructed hemivagina and ipsilateral renal agenesis. *AM J Obstet Gynecol* 1980; 138:339.
18. Singh M, Gearheart JP, Rock JA. Double urethra, double bladder, left renal agenesis, Persistent hymen, double vagina and uterus didelphys. *Adolesc Pediatr Gynecol* 1993; 6:99.
19. Pumphong V, Pruksananonda K, Taneepanichskul S, et al. Double uterus with unilaterally obstructed hemivagina and ipsilateral renal agenesis: a variety presentation and a ten year review of the literature. *J Med Assoc Thai* 2000; 83:569.

20. Thompson DP, Lynn HB. Genital anomalies associated with solitary kidney. *Mayo Clin Proc* 1966; 41:538.
21. Petrozza JC, Gray MR, Davis AJ, et al. Congenital absence of the uterus and vagina is not commonly transmitted as a dominant genetic trait: outcomes of surrogate pregnancies. *Fertil Steril* 1997; 67:387.
22. Simpson JL. Genetics of the female reproductive ducts. *Am J Med Genet* 1999; 89:224.
23. Griffin JE, Edwards C, Madden JD, et al. Congenital absence of the vagina. *Ann Intern Med* 1976; 85:224.
24. McKusick VA, Weilbaccher RG, Gragg Gw. Recessive inheritance of a congenital malformation syndrome. *JAMA* 1968; 204:111.
25. Goodman FR, Bacchelli c, Bradley AF, et al. Novel HOXA13 mutations and the phenotypic spectrum of hand-foot-genital syndrome. *Am J Hum Genet* 2000; 67:197.
26. Goodman FR, Scambler PJ. Human HOX gene mutations. *Clin Genet* 2001;59:1
27. Wharton LR. Congenital malformations associated with developmental defects of the female reproductive organs. *Am J Obstet Gynecol* 1947; 53:37.
28. Pinsky L. A community of human malformation syndromes involving the mullerian ducts, distal extremities, urinary tract, and ears. *Teratology* 1974; 9:65.
29. Counsellor V. S. and Davis C. E. Atresia of the vagina. *Obstet Gynecol* 32,528-536.1968.
30. Lisa Jane Jacobsen, Alan H. Dechrney creation of a neo vagina *Gynecologic Surgery* Churchill Livingstone 1996
31. Rock JA, Parmley T, Murphy AA, et al. Malposition of the ovary associated with uterine anomalies. *Fertil Steril* 1986; 45:561.
32. Hauser, G. A. and Schreiner, W. E.: Mayer- Rokitansky- Kuster syndrome. Rudimentary solid bipartite uterus with solid vagina. *Schweiz Med. Wschr.*, 91:381, 1961.
33. Maciulla GJ, Heine M. W, Christian C. D. functional endometrial tissue with vaginal agenesis. *J Reprod Med* 21:373.1978.
34. Jeffocoate TNA. Advancement of the upper vagina in the treatment of the haematocolpos and haematometra caused by vaginal aplasia. Pregnancy following the construction of an artificial vagina. *J Obstet Gynaecol Br Comm* 1969; 76:961.
35. Murphy AA, Krall A, Rock JA. Bilateral functioning uterine anlagen with the Mayer- Rokitansky- Kuster-Hauser syndrome. *Int J Fertil* 1987; 32:296.
36. Chakravarty BN: congenital absence of the vagina and uterus – simultaneous vaginoplasty and hysteroplasty. *J Obset Gynecol (India)*. 27:627, 1977
37. Singh J, Devi YL: pregnancy following surgical correction of non fused mullerian bulbs and absent vagina. *Obset Gynecol* 101:267,1983
38. Fore SR, Hammond CB, Parker RT et al: Urologic and genital anomalies in patients with congenital absence vagina. *Obstet Gynecol* 46:410, 1975
39. Genest D, Farber M, Mitchell GW, et al. Partial vaginal agenesis with a urinary-vaginal fistula. *Obstet gynecol* 1981; 58:130.
40. Plevraki E kita M bilateral ovarian agenesis and the presence of the testis specific protein1 –y-linked gene: two new features of MRKH syndrome. *Fertil Steril* 81,689-692

41. Chervenak FA, Stangel JJ MRKH syndrome: congenital absence of vagina. NY state J Med 82,23-26.(1982)
42. Schmid –Tannwald and Hauser (1977) Atypical forms of MRKH syndrome. Geburtshilfe frauenheilkd 37,386-392.
43. Seiferrt B, Woraschk HJ Construction of an artificial vagina in MRKH syndrome. Zentrabl Gynaecol 96,1034-1039.(1974)
44. Waberk AJ Millard PR (1971) creation of a neo vagina by the frank non operative method. Obstet Gynaecol 37,408-413.
45. Yenen E (1957) surgery of atresia and aplasia of the vagina; experience with 45 cases and description of new method. Zentrabl Gynaecol, 79, 1641-1647.
46. Reindollar RH Byrd JR (1981) delayed sexual development; a study of 252 patients. Am J Obstet Gynaecol, 140,371-380.
47. Smith MR (1983) vaginal aplasia: therapeutic options Am J Obstet Gynaecol, 146, 488-494.
48. Darai E, Toullalan O (2003) Anatomic and functional results of laparoscopic perineal neovagina construction by sigmoid coloplasty in with MRKH syndrome. hum reprod, 18, 2454-2459.
49. Brun JL, Belleanne G (2002) long term results after neo vagina creation in MRKH syndrome by Vecchietti's operation. Eur J Obstet Gynaecol Reprod Biol 103,168-172.
50. Fedele L, Bianchi S (2000), Laparoscopic creation of new vagina in patients with Rokitansky syndrome; analysis of 52 cases. Fertil Steril 74,384-389.
51. Creatas G, Deligeoroglou E (2001) Creation of new vagina following Williams's vaginoplasty in 111 cases of MRKH syndrome. Fertil Steril, 76, 1036-1040.
52. GauwerkyJFH, ReinhardB, Oppelt P (1997) Rekonstrukton der vagina unter besonderer berucksichtigung neur endoskopischer techniken, Gynakolge 30,507-514.
53. Palatynski (1986) a laparoscopic diagnosis in MRKH syndrome Zentralbl Gynaekol 108, 1130-1134.
54. Peter oppelt, Stefan P.Renner, clinical aspects of MRKH syndrome: recommendations for clinical diagnosis and staging. Human reproduction vol.21, No.3pp.792-797, 2006. Advance access publication November 10,2005.
55. Lischek JH: Discordance of vaginal agenesis in monozygotic twins. Obstet Gynecol 41:920, 1973
56. Shokeir MHK. Aplasia of the mullerian system: evidence for probably sex-limited autosomal dominant inheritance. Birth Defects 1978; 14:147.
57. Knab DR. Mullerian agenesis; a review. Bethesda, MD: Department of Gynecology/obstetrics, Uniformed services University School of Medicine And Naval Hospital, 1983.
58. David A , Carvil D, Bar-David E, et al. Congenital absence of the vagina: Clinical and psychological aspects. Obstet Gynecol 1975; 46:407.
59. Weijenborg PT, Terkuile MM. The effect of a group programme on women with the Mayer- Rokitansky- Kuster-Hauser syndrome. Br J Obstet Gynecol 2000; 107:365.
60. Beski S, Gorgy A, Venkat G, et al. Gestational surrogacy: a feasible option for patients with Rokitansky syndrome. Hum Reprod 2000; 15:2326.

61. Goldwyn RM. History of attempts to form a vagina. *Plast Reconstr Surg* 1977; 59: 319-29.
62. Ricci J.V: one hundred years of Gynaecology, pp. 433-442. Blakiston Co., Philadelphia. 1945
63. Frank RT: The formation of an artificial vagina without operation. *AM J Obstet gynecol* 35:1053, 1938
64. Frank RT: The formation of an artificial vagina without operation. *NY State J Med* 40:1669, 1940
65. Broadbent Tr, Woolf RM. Congenital absence of the vagina: reconstruction without operation. *Br J Plast Surg* 1977;30:118.
66. Ingram JM: The bicycle seat stool in the treatment of vaginal agenesis and stenosis: a preliminary report. *AM J obstet Gynecol* 1981; 140:867.
67. Roberts CP, Haber MJ, Rock JA. Vaginal creation for mullerian agenesis. *Am J Obstet Gynecol* 2001; 185:1349-52.
68. Makinoda S , Nishiya M, sogame M, et al., Non grafting method of vaginal reconstruction for patients of vaginal agenesis without functioning uterus *Int Surg* 1996; 81:385
69. Vecchietti G. Neovagina nella sindrome di Rokitansky- Kuster- hauser. *Attual Ostet Ginecol* 1965;11:129.
70. Gauwerky JFH, Wallweiner D an endoscopically assisted technique reconstruction of a neovagina. *Arch Gynaecol Obstet* 1992; 252:59.
71. Laffarque f, Giacalone PL, Boulot P, et al. A Laparoscopic procedure for the treatment of vaginal aplasia. *BR J Obstet Gynaecol* 1995; 102:565.
72. Fedele L: Laparoscopic creation of a neovagina in Mayer-Rokitansky-Kuster-Hauser syndrome by modification of Vecchietti's operation. *Am J obstet Gynecol* 171:268
73. Williams EA. Congenital absence of vagina simple operation for relief. *J Obstet Gynaecol Br Comm* 1964; 71:511
74. Williams EA. Uterovaginal agenesis *Ann R Coll Surg Eng l* 1976: 58: 266
75. Feroze RM Dewhurst CJ , Wephy G : Vaginoplasty at the Chelsea hospital for women : a comparison of two techniques *Br J Obset Gynaecol* 82: 536, 1975
76. O'Brien BM Mellow CG Mac Isaac IA : treatment of vaginal agenesis with a new vulvovaginoplasty *Plastic Reconstr Surg* 85: 942,1990
77. Creatasas GC. Creatasas modification of Williams's vaginoplasty. *J Gynecol Surg* 1991; 7:219.
78. Creatasas G Deligeoroglou E et.al., creation of neo vagina following Williams vaginoplasty and creatasas modification in 111 patients with MRKH syndrome *Fertil Steril* 2001 nov 76(5): 1036-40
79. Wharton LR: A simple method of constructing a vagina. *Ann Surg* 107:842, 1938
80. Abbe R. New method of creating a vagina in a case of congenital absence. *Med Rec* 1898; 54:836.
81. McIndoe AH, Banister JB. An operation for the cure of congenital absence of the vagina. *J Obstet Gynaecol Br Emp* 1938; 45:490.
82. McIndoe AH: The treatment of congenital absence and obliterative conditions of the vagina. *Br J plast surg* 2:254, 1950

83. McIndoe AH. The treatment of congenital absence and obliterative conditions of the vagina. *Br J Plast Surg* 1950; 2:254.
84. Counsellor VS: Congenital absence of the vagina. *JAMA* 136:861, 1948
85. Counsellor VS, Flor FS: Congenital absence of the vagina: further results of treatment and a technique. *Surg clin North Am* 37:1107, 1957
86. Michael Hockel Henrik Menke Vaginoplasty with split skin grafts from the scalp: optimization of the surgical treatment for vaginal agenesis *Am J Obstet Gynaecol* 2003 1100-1102.
87. Strickland JL, Cameron WJ, Krantz KE. Long-term satisfaction of adults undergoing McIndoe vaginoplasty as adolescents. *Adolesc Pediatr Gynecol* 1993; 6:135.
88. Alessandrescu D, Peltecu GC, Buhimschi CS, et al. Neocolpoptosis with split-thickness skin graft as a surgical treatment of vaginal agenesis: retrospective review of 201 cases. *Am J obstet Gynecol* 1996; 175:131.
89. Buss J, Lee R McIndoe procedure for vaginal agenesis: results and complications. *Mayo Clin Proc* 64:758, 1989.
90. Cali RW Prat JH congenital absence of the vagina: long term results of vaginal reconstruction in 175 cases, *Am J obstet Gynecol*.100:752, 1968.
91. Ortiz- Monasterio F, SerranoA congenital absence of the vagina: long term follow up in 21 patients treated with skin grafts. *Plast Reconstr Surg* .49:165, 1972.
92. Horton. F., sadove, r. C., and McCraw, J. B. Reconstruction of Female Genital Defects. In J. G. McCarthy (Ed.), *plastic surgery*, Vol. 6. Philadelphia: Saunders, 1990. Pp. 4203-4208.
93. Sadove, R. C., and Horton, C. E. Utilizing full-thickness skin grafts for vaginal reconstruction. *Clin. Plast. Surg.* 15: 443, 1988.
94. Chen, Y. B., Cheng, T., Lin, H. H., and Yang. Y. S. Spatial W-plasty full-thickness skin graft for neovaginal reconstruction. *Plast. Reconstr. Surg.* 94: 727. 1994
95. Guzin Yesim Ozgenel, M.D., Mesut Ozcan, M.D. Neo vaginal construction with buccal mucosal grafts. *Plastic and Reconstructive surgery* June 2003 Vol 111 No 7. Pp 2250-2254.
96. Morton KE, Dewhurst CJ: Human amnion in the treatment of vaginal malformations *Br J obstet Gynecol* 93:56,1986
97. Dhall K: Amnion graft for treatment of congenital absence of the vagina. *Br J obstet Gynecol* 91:279, 1984
98. Templeman CL, Hertweck Dp, Levine RL, et al. Use of laparoscopically mobilized peritoneum in the creation of a neovagina. *Fertil Steril* 2000; 74:589.
99. Davydov SN. Colopoptosis from the peritoneum of the uterorectal space. In: *Proceedings of the Ninth World Congress of Obstetrics and Gynecology*, Tokyo, 1979. Amsterdam, Excerpta Medica, 1980:793.
100. Patil V, Hixon FP The role of tissue expansion in vaginoplasty for congenital malformations of vagina, *Br J Urol* 1992; 70:554.
101. Nicholas Johnson, Andrew Batchelor, Lilford RJ Experience with tissue expansion vaginoplasty *Br J obstet Gynecol* 98, 564-568,1991.

102. Flack CE, Barraza MA, and Stevens PS. Vaginoplasty: combination therapy using labia minora flaps and Lucite dilators- preliminary report J Urol 1993; 150:654-6.
103. Purushothaman V Horse shoe shaped vaginoplasty – a new technique of vaginal reconstruction with labia minora flaps for primary vaginal agenesis. Br J Plast Surg .2005 58(7):934-9.
104. Pakiam AI medial thigh skin flaps for repair of vaginal stenosis Grabb's encyclopedia of flaps pp 1458-1460.
105. Wang TN, Whetzel T A fascio-cutaneous flap for vaginal and perineal reconstruction Plastic and Reconstructive surgery1987; 80:95-102.
106. Zong-ji Chen, Mei-yun Chen, Vaginal reconstruction with axial subcutaneous pedicle flap from the inferior abdominal wall: a new method. Plastic and Reconstructive surgery1989, 83:1005-1012.
107. Francesco Moschella, Adriana Codova Vaginal reconstruction with bilateral island extended groin flaps: description of a personal technique Plastic and Reconstructive surgery1994, 94; 1079-1084.
108. McCraw JB, Massey FM, Shanklin KD, et al. Vaginal reconstruction with gracilis myocutaneous flaps. Plast Reconstr Surg 1976; 58:176.
109. Copeland LJ, Hancock KC, Gershenson DM: Gracilis myocutaneous vaginal; reconstruction concurrent with total pelvic ex-entiation. Am J Obstet Gynecol 160:1095,1989
110. Soper JT Short gracilis myocutaneous flaps for vulvo vaginal reconstruction Grabb's encyclopedia of flaps pp 1471-1475.
111. Benson C, Soisson AP, Culbertson G: Neovaginal reconstruction with a rectus abdominis myocutaneous flap. Obstet gynecol 51:871, 1993
112. Lilford RJ Johnson N Batchelor A A new operation for vaginal agenesis: construction of neo vagina from a rectus abdominus musculocutaneous flap.British J of Obstet Gynaecol 96; pp1089-1094.1989.
113. Hatch KD: Construction of a neovagina after exentiation using the vulvobulbocavernosus myocutaneous graft. Obstet Gynecol 63:110, 1984
114. Hagerty RC, VuaghnTR, the perineal artery axial flap in reconstruction of vagina Plastic and Reconstructive surgery1988; 82(2):344-5.
115. Giraldo F, SolanoA the Malaga flap for vaginoplasty in the MRKH syndrome experience and early term results. Plast reconstr surg 1996; 98(2):305-12.
116. Giraldo F, Mora MJ. Anatomic study of the superficial neuro vascular pedicle; implications in vulvoperineal flap design Plastic and Reconstructive surgery1997; 99(1):100-8.
117. Giraldo F, Gaspar D. treatment of vaginal agenesis with vulvoperineal fasciocutaneous flap Plastic and Reconstructive surgery1994; 93(1):131-40.
118. Giraldo F cutaneous neovaginoplasty using Malaga flap: a 12 year follow up study Plastic and Reconstructive surgery2003; 111(3); 1249-55.
119. Wee and Joseph of Singapore. A new technique of vaginal reconstruction using neurovascular pudendal thigh flaps. Plastic and Reconstructive surgery1989; 83(4):701-9.

120. Woods JE, Alter G, experience with vaginal reconstruction utilizing the modified Singapore flap. *Plastic and Reconstructive surgery* 1992; 90(2):270-4.
121. Baldwin JF: The formation of an artificial vagina by intestinal transplantation. *Ann Surg* 40:398,1984
122. Popoff DD. Utilisation of the rectum in construction of the functional vagina. *Russk Virach St Peter* 1910; 43:1512.
123. Hendren WH: Use of bowel for vaginal reconstruction. *J Urol* 152:752,1994
124. Freundt I, Toolenaar TA, and Jeekel H: Prolapse of the sigmoid neovagina: report of three cases. *Obstet Gynecol* 83:876
125. Pratt JH. Vaginal atresia corrected by use of small and large bowel. *Clin obstet Gynecol* 1972;15:639
126. Pratt JH: Vaginal reconstruction with a sigmoid loop. *Am J obstet Gynecol* 96:31,1966
127. Turner- Warwick R, Kirby RS: The construction and reconstruction of the vagina with the colocecum. *Surg Gynecol obstet* 170:1132, 1990
128. Freundt I, Toolenaar TA, Huikeshoven FJ: A modified technique to create a neovagina with an isolated segment of sigmoid colon. *Surg Gynecol obstet* 174:11,1992
129. Freundt I, Toolenaar TA, and Jeekel H: Prolapse of the sigmoid neovagina: report of three cases. *Obstet Gynecol* 83:876
130. Froese DP, Haggitt RC, and Friend WG: Ulcerative colitis in the auto transplanted neovagina. *Gastroenterology*. 100:1749, 1991
131. Munkarah A, Malone JM, Budev HD: Mucinous Adenocarcinoma arising in a neovagina. *Gynecol oncol* 52:272, 1994
132. Rotmensch J, Rosensheim N, Dillon M, et al. Carcinoma arising in the neovagina: case report and review of the literature. *Obstet Gynecol* 1983; 61:534.
133. Johnson N, Lilford RJ, Batchelor A: The free-flap vaginoplasty: a new surgical procedure for the treatment of vaginal agenesis. *Br obstet Gynecol* 98:184,1991
134. Sakurai H Nozaki M the use of free jejunal autograft for the treatment of vaginal agenesis: surgical methods and long term results. *British journal of plastic surgery* 2000:319-323.
135. Info @ www.mrkh.or
136. ACOG Committee Opinion No. 355: Vaginal Agenesis: diagnosis management and routine care. *Obstet Gynecol*. 2006 Dec; 108(6): 1605-9.
137. Kaufmann RH, Binder GL. upper genital tract changes associated with exposure in utero to diethyl stilbestrol. *Am J Obstet Gynecol* 1977; 128:51.
138. Monie I Sigurdson L A. A proposed classification for uterine and vaginal anomalies. *Am J Obstet Gynecol* 59: 696, 1950.
139. Duncan PA Shapiro LR The MURCS association: Mullerian duct aplasia, renal aplasia, and cervico thoracic somite dysplasia *J Pediatr* 95,399-402.1979
140. Peter Oppelt Stefan p. Renner The V-C-U-A-M classification the vagina uterus cervix adnexa associated malformation classification: a new classification for genital malformations. Personal communication from p.oppelt@web.de

141. Peters WA: prolapse of neo vagina created by self dilation. *Obstet Gynecol* 76:904, 1990.
142. Seccia A Salgarello M neo vaginal reconstruction with modified McIndoe technique : a review of 32 cases. *Ann Plast Surg* 2002, 49:379-84.
143. Hojsgaard A Villadsen I McIndoe procedure for vaginal agenesis: complications and results *Br J Plast Surg* 1995, 48:97-102.
144. Evans TN The artificial vagina. *Am J Obstet Gynecol* 99:44,1967
145. Seok Kwun Kim, Ji Hoon Park, Long term results in patients after rectosigmoid vagino plasty. *Plastic and Reconstructive surgery*, 2003, 112:143-151.
146. Li S, Liu Y, Li Y. Twelve cases of vaginal reconstruction using neurovascular pudental-thigh flaps. *Zhonghua Fu Chan Ke Za Zhi*. 2000; 35(4):216-8.
147. Chen Z, Chen C. Vaginal reconstruction using perineal-thigh flaps with subcutaneous pedicle. *Chin Med Sci J*. 1991; 6(1):14-7.
148. Gurlek A, Monstrey S prof S. The versatility of the pudental thigh fasciocutaneous flap used as an island flap. *Plast Reconstr Surg*. 2002; 109(1):402-3.
149. Li GZ, Cheng XD. Experience of vaginal reconstruction by using a pudental – thigh island flaps. *Zhonghua Zheng Xing Wai Ke Za Zhi*.2003; 19(3):183-5.
150. Selvaggi G, Monstrey S, Depypere H, Blondeel P, Van landuyt K, Hamdi M, Dhont M. Creation of a neovagina with use of a pudental thigh fasciocutaneous flap and restoration of uterovaginal continuity. *Fertil Steril*. 2003;80(3):607-11
151. Sanchez Contreras J, Pasos Romero I, surgical correction of vaginal agenesis. *Ginecol Obstet Mex*.2006; 74(1):37-47.
152. Hwang WY, Chang TS, Sun P. Vaginal reconstruction using labia minora flaps in congenital total absence. *Ann Plast Surg*. 1985; 15(6):534-7.
153. Rock JA, Reeves LA. Success following vaginal creation for mullerian agenesis. *Fertil Steril*. 1983; 39(6):809-13.
154. Hage JJ Karim RB Abdominoplastic secondary full- thickness skin graft vaginoplasty for male to female transsexuals. *Plastic Reconst Surg* 1998; 101:1512-5.
155. Hallberg H Holmstrom H Vaginal reconstruction with skin grafts and vacuum assisted closure. *Scand J Plasst Surg Hand Surg* 2003;37:97-101.
156. Adamson CD, Naik BJ. The vacuum expandable condom mold: a simple vaginal stent for McIndoe-style vaginoplasty. *Plast Reconstr Surg*. 2004; 113(2):664-6.
157. Tercan M, Balat O. The use of fibrin glue in the McIndoe technique of vaginoplasty. *Plast Reconstr Surg*. 2002; 109(2):706-9.
158. Barutcu A, Akguner M. McIndoe vaginoplasty with the inflatable vaginal stent. *Ann Plast Surg*. 1998; 41(5):568-9.
159. Liu Y LI S Applied anatomical study of the pudental thigh flap and establishment of flap model. *Zhonghua Zheng Xing Wai Ke Za Zhi*.2001;17:272-5.

160. Khazanchi RK, Takkar D. Vaginal depth following reconstruction with pudendal thigh flaps in congenital vaginal atresia. *Plast Reconstr Surg.* 1997; 99(2):592-3.
161. Copcu E, Odabasi AR, Sivrioglu N, Yuksel H. Protection of vagina depth by laparoscopy in vaginal reconstruction with pudendal thigh flaps. *Plast Reconstr Surg.* 2005; 115(2):663-4.
162. Giraldo F, Solano A. Hair growth in the vagina after reconstruction with pudendal thigh flaps in congenital vaginal agenesis. *Plast Reconstr Surg.* 1998; 102(3):924-5.
163. Karacaoglan N. Hair growth in the vagina after reconstruction with pudendal thigh flaps in congenital vaginal agenesis. *Plast Reconstr Surg.* 1997; 100(6):1618.
164. Bazan A, Samper A. The use of the fibrin glue in vaginal reconstruction with a pudendal thigh flap. *Ann Plast Surg.* 1999; 43(5):576.
165. Shah R Woolley MM : testicular feminization : androgen insensitivity syndrome. *J Pediatric Surgery* ,27, 1992: 757-760.

PHYSICAL EXAMINATION

General examination

Built

Stature

Nourishment

Anemic/ not anemic

Phenotype- masculine/ feminine

Skeletal/ limb anomalies

Secondary sexual characters

Breast development- Tanner's stage

Axillary hair

Pubic hair

Moustache/ beard

Pelvis

Voice

Vital signs

Systemic examination

External genitalia

Pubic hair

Clitoris

Labia majora

Labia minora

Vaginal introitus

Hymen/septum

Scars

Urethral meatus

Features of inter sexuality- scrotal/ inguinal swelling, clitoral enlargement/
phallus

Pelvis, sacrum & pubic bones

Inguinal region- hernia, swellings

Per rectal examination- rectal anomalies, cervix, uterus, ovaries, mass.

Abdomen

Spine and cranium

Cardio vascular system

Respiratory system

Central nervous system

INVESTIGATIONS

Routine- Blood Hb%- TC DC ESR

Urine – alb sug dep

Blood sugar urea creatinine

Chest X- ray

ECG in all leads

Karyotyping

USG of abdomen, kidneys & pelvis

Per vaginal/ per rectal USG

Hormone status- LH

FSH

Diagnostic laparoscopy

Diagnosis

Treatment: operative/ non operative

Surgery:

Date of surgery:

Anesthesia: GA/ spinal

Post operative period:

Graft/ Flap:

Catheter removal:

Complications:

Discharged on:

FOLLOW UP:

Points of physical examination:

<i>POINTS</i>	<i>AT DISCHARGE</i>	<i>1ST MONTH</i>	<i>2ND MONTH</i>	<i>6TH MONTH</i>	<i>12TH MONTH</i>	<i>LATER</i>
Vaginal cavity depth						
Vaginal cavity width						
Mucus discharge						
Malodor						
Cosmetic configuration						

Points of questionnaire:

<i>POINTS</i>	<i>AT DISCHARGE</i>	<i>1ST MONTH</i>	<i>2ND MONTH</i>	<i>6TH MONTH</i>	<i>12TH MONTH</i>	<i>LATER</i>
Sexual intercourse						
Lubricant						
Vaginal stent						
Abdominal pain						
Pain during intercourse						
Orgasm						
Urination						
Defecation						
Vaginal bleeding during intercourse						