Role of Laparascopy in Diagnosis of Disorders of Sexual Development " DSD "

Thesis Submitted for Fulfillment of Master Degree

In General Surgery

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Table of Contents Page

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INTRODUCTION

Introduction

- The management of DSD patients is a challenge. Although in the majority of patients the diagnosis may be made on the basis of cytogenetic and biochemical tests, there is a selective group of patients with difficulties in the establishment of final diagnosis and gender assignment.

- Since laparoscopy has been used in the management of impalpable gonads in the normal male population, it may be an alternative method for the diagnosis and surgical management of DSD patients. Thus we have evaluated our experience with laparoscopy in DSD population.

- In order to establish a correct diagnosis some of these patients require gonadal biopsy in spite of the currently available genetic and biochemical evaluation. Furthermore in patients with impalpable gonads a precise localization of the gonads should be determined in order to proceed with orchidopexy or their removal when indicated.

- The laparoscopic biopsy has an important impact not only on the establishment of the final diagnosis but also on the decision of the appropriate rearing of a baby with ambiguous genitalia.

AIM OF WORK

Aim of Work

- The aim of this study is to evaluate our experience with laparoscopy in the diagnosis andmanagement including sex assignment of DSD patients.

REVIEW OF LITERATURE

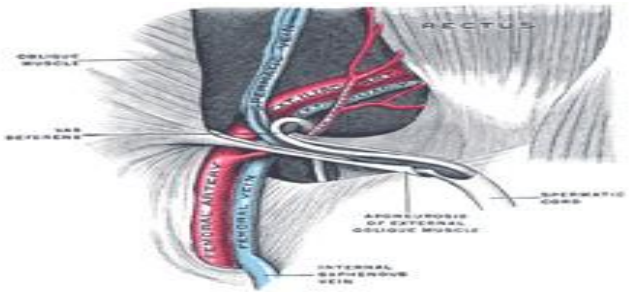
ANATOMY

Male internal genitalia

1-Testicular vessels:

- **The testicular arteries**:- are two long, slender vessels, which arise anteriorly from the aorta a little inferior to the renal arteries. Each passes inferolaterally under the parietal peritoneum on psoas major. The right testicular artery lies anterior to the inferior vena cava and posterior to the horizontal part of the duodenum, right colic and ileocolic arteries, root of the mesentery and terminal ileum. The left testicular artery lies posterior to the inferior mesenteric vein, left colic artery and lower part of the descending colon. Each artery crosses anterior to the genitofemoral nerve, ureter and the lower part of the external iliac artery and passes to the deep inguinal ring to enter the spermatic cord . " **Susan S. et al , 2008** "

- **The testicular veins** :-In the inguinal canal,the pampiniform plexus of veins is drained by three or four veins which run into the abdomen through the deep inguinal ring. Within the abdomen these veins coalesce into two veins, which ascend on each side of the testicular artery, anterior to psoas major and the ureter, and behind the peritoneum. The left veins pass behind the lower descending colon and inferior margin of the pancreas and are crossed by the left colic vessels, and the right veins pass behind the terminal ileum and horizontal part of the duodenum and are crossed by the root of the mesentery, ileocolic and right colic vessels. The veins join to form single right or left testicular veins: the right testicular vein opens into the inferior vena cava at an acute angle just inferior to the level of the renal veins, and the left testicular vein opens into the left renal vein at a right angle. " **Susan S. et al , 2008** "

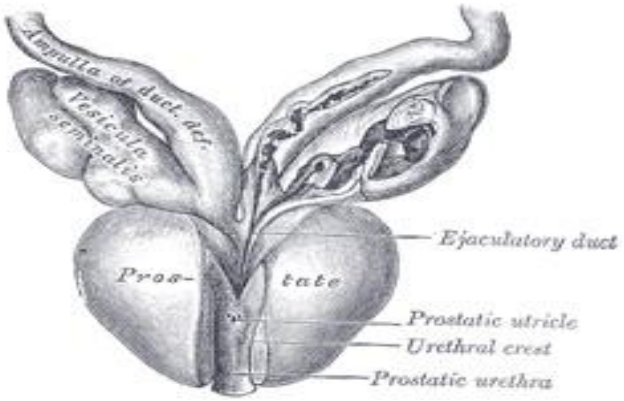


The spermatic cord in the inguinal canal. **(Poirier and Charpy.)**

2-Vas deferens

- At the internal (deep) inguinal ring the vas deferens leaves the cord, curves round the lateral side of the inferior epigastric artery and ascends for 2.5 cm anterior to the external iliac artery. It then turns back and inclines slightly down and obliquely across the external iliac vessels to enter the lesser pelvis, where, situated retroperitoneally, it continues posteriorly, medial to the obliterated umbilical artery, the obturator nerve and vessels, and the vesical vessels. " **Susan S. et al , 2008** "

- It crosses the ureter and bends acutely to pass anteromedially between the posterior surface of the bladder and the upper pole of the seminal vesicle.It then descends in contact with the seminal vesicle, gradually approaching the opposite duct.Here it lies between the base of the bladder and the rectum, from which it is separated by Denonvillier's fascia.It finally descends to the base of the prostate,where it joins the duct of the seminal vesicle at an acute angle to form the ejaculatory duct**.** " **Susan S. et al , 2008** "



Vesiculæseminales and ampullæ of ductusdeferentes, seen from the front. The anterior walls of the left ampulla, left seminal vesicle, and prostatic urethra have been cut away. " **Susan S. et al , 2008** "

Female internal genitalia

Uterus

- The uterus is a hollow, thick-walled and muscular organ. It is normally situated in the lesser pelvis between the urinary bladder and the rectum.

- The uterus is divided into two main regions. The body of the uterus (corpus uteri) forms the upper two-thirds, and the cervix (cervix uteri) forms the lower third.

- The body of the uterus extends from the fundus at its uppermost part to the cervix inferiorly. Near its upper end, the body receives uterine tubes on both sides.

- The point of fusion between the uterine tube and body is called the uterine cornu. Inferoanterior to the cornu is the round ligament and inferoposterior is the ovarian ligament. The fundus is superior to the entry points of the uterine tubes, and the uterine body narrows as it extends towards the cervix.

- The lateral margins of the body are convex, and on each side their peritoneum is reflected laterally to form the broad ligament, which extends as a flat sheet to the pelvic wall.

- The anterior surface of the uterine body is covered by peritoneum, which is reflected onto the bladder at the uterovesical fold. This normally occurs at the level of the internal os, which is the most inferior margin of the body of the uterus. Between the bladder and uterus there is the vesico-uterine pouch.

- The posterior surface of the uterus is convex transversely. Its peritoneal covering continues down to the cervix and upper vagina and is then reflected back to the rectum along the surface of the recto-uterine pouch (of Douglas), which lies posterior to the uterus**.** " **Susan S. et al , 2008** "

- The uterus is continuous with a number of 'ligaments':

1. *Uterovesical ligament*:-

   The uterovesical ligament or anterior ligament consists of peritoneum reflected onto the bladder from the uterus at the junction of its cervix and body.

2. *Rectovaginal ligament*:-

   The rectovaginal ligament or posterior ligament consists of peritoneum reflected from the posterior vaginal fornix on to the front of the rectum, thereby forming the deep recto-uterine pouch.

3. *Broad ligament:-*

   The broad ligaments extend, one from each side of the uterus, to the lateral walls of the pelvis, where they become continuous with the peritoneum**."Gray's anatomy"**

Uterine tubes

- The two uterine (Fallopian) tubes lie on each side of the uterus in the upper margin of the broad ligament. The medial opening of the tube (the uterine os) is located at the superior angle of the uterus. The tube passes laterally and superiorly and consists of four main parts: intramural; isthmus; ampulla; and infundibulum.

1- Intramural part:-

It is the part between the end of the isthmus and the uterine cornu, located within the uterine wall.

2- Isthmus:-

The isthmus extends from the tubocornual junction to the most proximal part of the ampulla. It represents the medial one-third of the tube.

3- *Ampulla:-*

The ampulla constitutes the remaining two-thirds of the uterine tube. Anatomically it extends from the distal part of the isthmus to the fimbriated vestibule.

4- *Infundibulum:-*

The infundibulum is the funnel-shaped part of the ampulla, related to the ovary. It is characterized by a peritoneal opening 2 mm in diameter at its proximal end, surrounded by fimbria**."Skandalakis JE et al , 1983 "**

Ovaries

- The ovaries are traditionally described as almond-shaped. They are white in color. The ovaries are asymmetrical, the right larger than the left.

- Each ovary is located in the ovarian fossa on the lateral pelvic sidewall, and is attached to the posterior and superior aspect of the broad ligament by a double peritoneal fold, called the mesovarium. The mesovarium does not cover the ovaries; it only "attaches" to their anterior borders. The mesovarium is a reduplication of the posterior lamina of the broad ligament.

- The position of the ovaries is variable. Normally each is located on the lateral pelvic sidewall on either side of the uterus, below and posterior to each uterine tube, resting within the ovarian fossa. The suspensory ligament is a good anatomic landmark for localization of the ovaries and lymph nodes related to ovarian tumors**."Bazot M et al , 1999 "**

1- *Borders:-*

The anterior ovarian border is related to the mesovarium, which contains the vessels and nerves for the hilum. The posterior ovarian border is free.

2- *Poles:-*

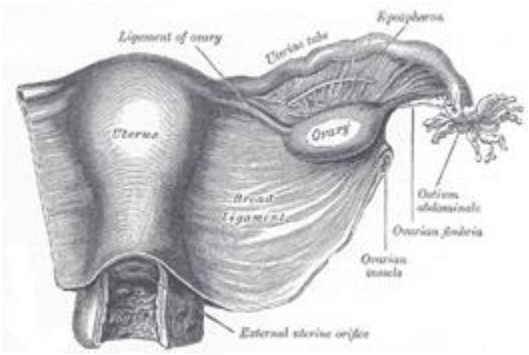
The medial surface is related to the uterine tube.The medial surface is related to the fimbriated end of the uterine tube which practically covers this surface. The lateral surface is related to ovarian fossa.

3- *Ligaments:-*

(a) The infundibulopelvic ligament (suspensory ligament of the ovary) is a fan-shaped band of fibromuscular visceral connective tissue containing arteries, veins, lymphatics, and visceral nerves extending from the upper ovarian pole to the lateral pelvic wall.

(b) The utero-ovarian ligament (proper ligament of the ovary) is a cordlike structure invested with the posterior layer of the broad ligament. The ovarian ligament extends from the lower ovarian pole to the lateral uterine wall.

(c) The mesovarium is a short peritoneal fold from the posterior surface of the broad ligament to the anterior ovarian wall. It facilitates the passage of ovarian vessels and nerves into the ovarian portae (hila**)."Skandaliks JE et al , 1983 "**



Uterus and right broad ligament.The broad ligament has been spread out **."Susan S et al , 2008 "**