# Evolution of Anorectal Malformation Management and Validation of Krickenbeck Classification in Operated Cases

## Thesis

Submitted for Partial Fulfillment of the Master Degree in General Surgery

## $\mathcal{B}y$

## Abd El-Rahman Azzam Ahmed Abd El-Baky

M.B.BCH

# Supervised by

## Prof. Dr. Osama Abdul Ellah El- Nagar

Professor of Pediatric Surgery
Faculty of Medicine, Ain Shams University

## Dr. Ahmed Bassiouny Arafa Radwan

Lecturer of Pediatric Surgery
Faculty of Medicine, Ain Shams University

## Dr. Mohamed Abdel - Latif Ayad

Lecturer of Pediatric Surgery
Faculty of Medicine, Helwan University

Faculty of Medicine
Ain Shams University
2016



- Thanks and for most thanks to ATTAH, the merciful of All, who helped me for accomplishment of this work.
- Twould like to express my deepest gratitude to Prof.

  Dr. Osama Abdul Ellah El- Nagar Professor of Pediatric Surgery, Faculty of Medicine, Ain Shams University, for his indispensable help and guidance which were essential for the accomplishment of this work.
- Im indebted a great deal to Dr. Ahmed Bassiouny Arafa Radwan Lecturer of Pediatric Surgery, Faculty of Medicine, Ain Shams University, for his kind guidance and cordial help. He gave much of his time and effort for supervision.
- Also, I can never forget Dr. Mohamed Abdel

  Tatif Ayad Tecturer of Pediatric Surgery, Faculty of
  medicine, Helwan University, for his precious guidance
  and close supervision of the final details.
- It is honorable to dedicate this work to my sweet family who was of great help, and gave me lots of guidance and support.





# **Contents**

Items	
➤ List o Figures	II
> Introduction	1
> Aim of the Work	3
> Review of Literature	
Surgery in Ancient Greek Era	4
Anew idea: CLOSTOMY	9
Surgery in the first half of 19th century	11
Surgery in the second half of 19th century	14
Surgery in the 20th century	16
Management in the newborn period	33
> Patients & methods	
> Results	46
> Discussion	57
> Summary	62
> References	64
> Arabic Summary	<u> </u>

# List of Table

Table	Title	Page
No.		No.
Table (1)	Kelly scoring system	22
Table (2)	Demographic data distribution of the	46
	study group.	
Table (3)	Standard for diagnostic procedures:	47
	International classification (krickenbeck)	
	distribution of the study group.	
Table (4)	Rare/ regional variants distribution of	48
	the study group	
Table (5)	International grouping (krickenbeck) of	49
	surgical procedures for follow up	
	distribution of the study group.	
Table (6)	Post- operative outcome of operated	50
	cases in the study group	
Table (7)	Perinal fistula	51
Table (8)	Recto urethral fistula	52
Table (9)	Rectovesical fistula	53
<b>Table</b> (10)	Vestibular fistula	54
<b>Table</b> (11)	Cloaca	55
<b>Table (12)</b>	No fistula	56

# List of Figures

Figures No.	Title	Page No.
<b>Fig.</b> (1)	triangular clave was used as the last step	5
	of cases having anus covered with a	
	thicker tissue	
Fig. (2)	An illustration from the Textbook of	8
	Sabuncuoglu. A surgical procedure	
	described for low type anal atresia in a	
	male patient. A female surgeon is noted	
Fig. (3)	The German surgeon, Heister	9
Fig. (4)	Some steps in the history of colostomy	11
	for anorectal malformations	
Fig. (5)	Jean Zule'ma Amussat (1796–1852)	13
	made important contributions for	
	Anorectal malformations and colostomy	
	techniques in pediatric surgery	
Fig.(6)	ladd's and gross's classification.	17
Fig (7)	Stephens' and smith's diagrammatic	19
	illustration of the normal levator ani	
	muscle.	
Fig. (8)	Rehbein's abdomino perineal operation.	20
Fig. (9)	Melbourne classification	24
Fig. (10)	Wingspread classification	25

## 🕏 List of Figure 🗷

Figures No.	Title	Page No.
Fig. (11)	Posterior sagittal incision. Separation of	27
	the parasagittal fibers and exposure of	
	the muscle complex	
Fig. (12)	Some steps in the evolution process of	29
	anorectal malformations (PSARP	
	posterior sagittal anorectoplasty, AP	
	abdominoperineal)	
Fig. (13)	Decision-making algorithm for male	33
	newborns with anorectal malformations	
	(ARM). U/S Ultrasound, PSARP pos-	
	terior sagittal anorectoplasty, R/O rule	
	out	
Fig. (14)	Newborn anoplasty	36
Fig. (15)	Radiograph of cross-table lateral x-ray	36
Fig. (16)	Decision-making algorithm for female	37
	newborns with ARM. Urol. Urological	
Fig. (17)	Ideal colostomy	
Fig. (18)	Demographic data distribution of the	46
	study group.	
Fig. (19)	Standard for diagnostic procedures:	47
	International classification (krickenbeck)	
	distribution of the study group.	

## 🕏 List of Figure 🗷

Figures No.	Title	Page No.
Fig. (20)	International grouping (krickenbeck) of	49
	surgical procedures for follow up	
	distribution of the study group.	
Fig. (21)	Post- operative outcome of operated	50
	cases in the study group	
Fig. (22)	Perineal fistula	51
Fig. (23)	Recto urethral fistula.	52
Fig. (24)	Rectovesical fistula	53
Fig. (25)	Vestibular fistula	54
Fig. (26)	Cloaca.	55
Fig. (27)	No fistula	56

## Introduction

Anorectal malformations (ARMs) comprise a wide spectrum of diseases that affect boys and girls and can involve malformation of the distal anus and rectum, as well as the urinary and genital tracts. Malformations of the distal anus and rectum, as well as the urinary and genital tracts. Malformations range from minor, easily treated defects that carry an excellent functional prognosis, to complex defects that are difficult to treat, are often associated with other anomalies, and carry a poor functional prognosis (*Peña*, 2005)

ARMs are among the more frequent congenital anomalies encountered in paediatric surgery, with an estimated incidence ranging between 1 in 2000 and 1 in 5000 live births (*Levitt and Peña*, 2010).

ARMs are associated with a wide spectrum of other congenital abnormalities, with involvement of the genitourinary, spinal, cardiovascular, gastrointestinal, craniofacial and other systems (*Stoll et al.*, 2007).

The classification of ARMs was based upon sex and the position of rectum relative to the levatorani muscle in high,

intermediate and low (wingspread classification). Pena and colleagues suggested in the mid-90s a classification system based on the presence of a fistula (*Levitt and Peña*, 2006).

In 2005, a new international diagnostic classification system was devised by the Krickenbeck Conference on ARM in an attempt to design a standardised system for comparison of follow-up. This system incorporates an anatomic description of the ARM, type of surgical procedure done, and postoperative assessment of bowel movements, constipation and soiling (*Holschneider et al.*, 2005).

Management of these anomalies had been evoluted over years. In trials to obtain good results. Not only anatomical but also for functional point of view. The classic surgical approach consists of an early divergent stoma, later a surgical correction and finally closure of the stoma. The classic surgical treatment of intermediate and high ARM wan an abdomino perineal pull through technique. Later Peña and colleagues introduced posterior sagittal anorectoplasty (PSARP).

## Aim of the work

This study is conducted to assess post-operative outcome of anorectal malformations according to Krickenbeck classification in pediatric patients operated at pediatric surgery department of Ain Shams university hospitals and documentation of the main milestones in evolution of management of these anomalies from the literature review.

### **Review of literature**

ARMs remain one of the challenging topics in pediatric surgery. It's management have been evoluted through history from ancient Greek era up till now and different classification systems were supposed to aid in differentiation between different types of anomalies and to assess the outcome of cases underwent different surgical procedures (Yesildag et al., 2010).

## The Beginning:

#### **Surgery for survival:**

Hippocrates (ca. 460-370 BC), was aware of Ano rectal fistula and tried to interpret their mechanism of origin. He attempted to treat them, either conservatively, using laxatives and purgative medications, or surgically, via an anoscope. He had very well understood the importance of the surgical option which is now a routine choice. In fact a variety of operations exist mainly aiming to decrease recurrence and incontinence rates (**Fig. 1**) (*Jacob et al., 2010*).

*Soranus*, a Roman physician, stressed the importance of anal examination in all new born. He changed the concept of leaving neonates with congenital anomalies to death (*Yesildag et al.*, 2010).

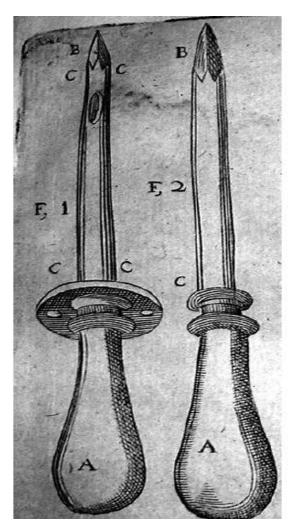


Fig. (1): A triangular clave was used as the last step of cases having anus covered with a thicker tissue (Yesildag et al., 2010).

In Aristotle's (384-322BC) works. He recorded failed attempts to restore an ano\_rectal ring atresia. In particular he stated: "some animals have already been diagnosed with anal symphysis. Someone even pointed out, in the village of Perinthos, the excretion of fine food with the urine in a cow, due to its infiltration from the intestine to the bladder, and when a membrane incision was performed, soon adhesions emerged once again and food was excreted via the bladder". The principles of operative technique resemble modern surgical methods (Bischoff et al., 2011).

Oribasius (ca 320-400) suggesting a manipulation of opening up by hand prior to surgery .This opening up seems valid up until now, though, instead of hands, scalpel and special surgical retractors are used in order to achieve better control of the area for detection of the imperforate part which may be behind the anal membrane or even higher (*Grosfeld et al., 2006*).

Paul of Aegina (ca.625-690) suggested opening of imperforate ring by hand prior to the surgical intervention. In cases of recurrence in infants, he recommended the use of a small lead tube until recovery (*Tsoucalas et al.*, 2010).

In addition, Leo the philosopher (ca. 790-869) suggested repair of imperforate anus by a special instrument resembling a sickle (*Tsoucalas et al.*, 2012).

The main objective of treatment during many centuries was only to create an orifice in the perineum but many times this was not enough for the survival (*Grosfeld et al.*, 2006).

#### Arabic era:

Abu Kasim Al-Zahrawi (Abulcasis) an arabic surgeon his procedure was doing perineal perforation by using finger then using leaden tube for dilatation or stenting in cases of stricture (*Montagnani*, 1986).

Serafeddin in the fifteenth century in Central Anatolia (Turkey) Besides making different surgical contributions, colored picture of operation performed by him also placed (**Fig. 2**) He was the first one who mentioned the importance of differing low and high anomalies (*Yesildag et al., 2010*).



**Fig. (2):** An illustration from the Textbook of Sabuncuoglu. A surgical procedure described for low type anal atresia in a male patient. A female surgeon is noted (*Yesildag et al.*, 2010).

# **Surgery from 14<sup>th</sup> to 18<sup>th</sup> century:**

Galen was a prominent Greek physician, surgeon and philosopher in the Roman Empire described the anal sphincters, levator muscles, and coccyx in 1576. Scultet was German surgeon treated an infant with anal stenosis by dilating the anal orifice nearly 1000 years after Paul of Aeginata. Cook British surgeon dilated the anal canal with an instrument after making an incision over the blind pouch in a baby with low type anal atresia in 1676. (*Kiely & Pena, 1998*) Saviard was the first to plunge a trocar through the perineum to treat a high termination of the bowel in 1693 (*Grosfeld et al., 2006*).