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Original Article

Outcomes of Treatment of Anorectal Malformations: A 7-year Review at Queen Sirikit National Institute of Child Health

Marin Pollakan, MD
Suranetr Laorwong, MD
Varaporn Mahatharadol, MD
Rangsan Niramis, MD
Department of Surgery, Queen Sirikit National Institute of Child Health, Bangkok, Thailand

Abstract

Background: **Anorectal malformations (ARM) are uncommon conditions and have the incidence of 1 in 4,000 to 5,000 live births. Many classifications and various operative procedures have been advocated and continuous improvement of postoperative outcome was reported.**

Objective: **The aim of this study was to review our experience in management of patients with ARM including classification, associated anomalies, surgical corrections and outcomes after definitive treatment.**

Materials and Methods: **A retrospective study was conducted by reviewing of medical records of the patients with ARM treated at Queen Sirikit National Institute of Child Health between 2006 and 2012. Patients' data were collected including demographics, types of ARM, associated anomalies, operative procedures and results of treatment. Wingspread classification was used to categorize types of ARM and Krickenbeck classification was used to evaluate postoperative results.**

Results: **A total of 365 patients (220 males and 145 females) were treated for ARM during the study period. The incidence of ARM at Rajavithi Hospital was 1: 2,820 live births. Over 70% of the patients were term babies with their birth weights over 2,500 g. The levels of ARM were categorized in low, intermediate and high types in 115 (52.3%), 75 (34.1%) and 30 (13.6%) for males and 74 (51.0%), 58 (40.0%) and 4 (2.8%) for females. Persistent cloaca was noted in 9 female patients (6.2%). Most of the patients with low anomalies were treated by cutback anoplasty. Alternative surgical treatment for low anomalies in 31 females were anoplasty by anal transfer and anterior sagittal anorectoplasty (ASARP) without preliminary colostomy. The most common operative procedures for intermediate anomalies were posterior sagittal anorectoplasty (PSARP) and ASARP. For high anomalies, PSARP and abdominoperineal pull-through operation (APP) were the definitive procedures. Laparoscopic assisted anorectoplasty (LAARP) was performed in seven males with intermediate and high anomalies and two cases in female with high anomalies. Nine cases with persistent cloaca underwent posterior sagittal anorecto - urethro - vaginoplasty and seven cases survived. The most common associated anomalies were genitourinary and cardiovascular abnormalities.Twenty-six patients (7.1%) with ARM died in immediate postoperative period due to congenital heart diseases, sepsis, respiratory and neurological problems. Long-term outcomes were evaluated in 335 patients with normal fecal continence between 51.7% and 71.8%, fecal soiling or incontinence between 2.8% and 22.2%, and constipation between 24.3% and 42.9%. Patients with low and intermediate anomalies had fecal continence approximately 70% and had long-term postoperative results better than the patients with high anomalies.**

Conclusion: **Approximately 70% of low and intermediate types of ARM had normal fecal continence and low incidence of fecal soiling. Every type of ARM was affected with constipation in a long-term period and required dietary, medical and toilet training therapies.**

Keywords: **Anorectal malformations, Wingspread classification, Krickenbeck classification, long-term outcomes**

Correspondence address: *Rangsan Niramis, MD, Department of Surgery, Queen Sirikit National of Child Health, 420/8 Rajavithi Road, Bangkok 10400, Thailand, Telephone/Fax: +66 2354 8095, E-mail: rniramis@hotmail.com*

Introduction

Anorectal malformations (ARM) are congenital abnormalities of the rectum and anus which embryological pathology is related to dysmorphogenesis of the cloaca and urorectum in early fetal life[1]. The anomaly may occur in isolation but many cases are commonly associated with other anomalies. Urogenital and musculoskeletal abnormalities are the most common association with ARM. The incidence of ARM was reported approximately 1 in 4,000 to 5,000 live births[2,3]. In Thailand, the incidence of ARM was 1 in 2,560 live births born at Rajavithi Hospital between 1977 and 1986[4].

Many classifications of ARM were proposed in order to be the guidelines of proper management. These classifications included International[5], Wingspread[6], Peña[7] and Krickenbeck classification[8]. Long-term outcomes of patients with ARM are concerned about fecal and urinary continence. At present, the outcomes of this disease have improved resulting from early diagnosis, management of associated anomalies, improved surgical techniques and meticulous postoperative care.

The aim of this study was to review our experience for management of patients with ARM during the recent 7-year period. The study stressed on types of ARM, associated anomalies, surgical procedures and results of treatment.

Materials and Methods

A retrospective study was conducted at Department of Surgery, Queen Sirikit National Institute of Child Health (QSNICH) after the proposal (Document No.58-063) had been approved by the Ethic Committees of the institute. Medical records of patients with ARM treated during January 2006 and December 2012 were reviewed. The patients who had surgical corrections of ARM done elsewhere were excluded from the study. Patients' data were collected on demographics, types of ARM, associated congenital anomalies, operative procedures and results of treatment. Various types of ARM were divided based on modified Wingspread Classification[5] that concerned the relationship of the terminal rectum to the levator ani muscle. Anomalies were grouped into high, intermediate, low, clocal and rare lesions with separate categories for male and female. We excluded rare anomalies from this study because patients' data ware not complete for the review.

Data were analyzed using SPSS version 20 (IBM® SPSS statistic). Correlations between categorical variables were evaluated by Chi-square test. A p-value of less than 0.05 was considered significant.

Results

Demographic data

A total of 365 patients with ARM (220 males and 145 females) was available for the study. Male and female ratio was 1.5 : 1. During the study period, 39,480 neonates were born at Rajavithi Hospital and 14 neonates were noted to have ARM[9]. Therefore, the incidence of ARM at Rajavithi Hospital was 1:2,820 live births. Gestational ages ranged from 27 to 42 weeks (mean 37.5 ± 2.4, median 38 weeks). Birth weight ranged from 1,060 to 4,800 g (mean 2,796.3 ± 2,850g). Over 70% of the cases were term babies (GA > 37 weeks) with their birth weights over 2,500 g (Table1).

Types of ARM

The levels of anomalies were categorized into low: intermediate: high types in 115 (52.3%) : 75 (34.1%) :30(13.6%) in males and 74 (51.0%) : 58 (40.0%) : 4 (2.8%) in females. Persistent cloaca was noted in 9 female patients (6.2%). Approximately

Table 1 Demographic data

Characteristics	Cases (Percent)
Gender	
Male	220 (60.3)
Female	145 (39.7)
Gestational age (weeks)	
< 32	21 (5.8)
32-36	60 (16.4)
37-40	233 (63.8)
> 41	51 (14.0)
Birth weight (grams)	
< 2,000	38 (10.4)
2,000-2,500	64 (17.5)
2,501-3,000	124 (34.0)
3,001-3,500	103 (28.2)
3,501-4,000	32 (8.8)
> 4,000	4 (1.1)

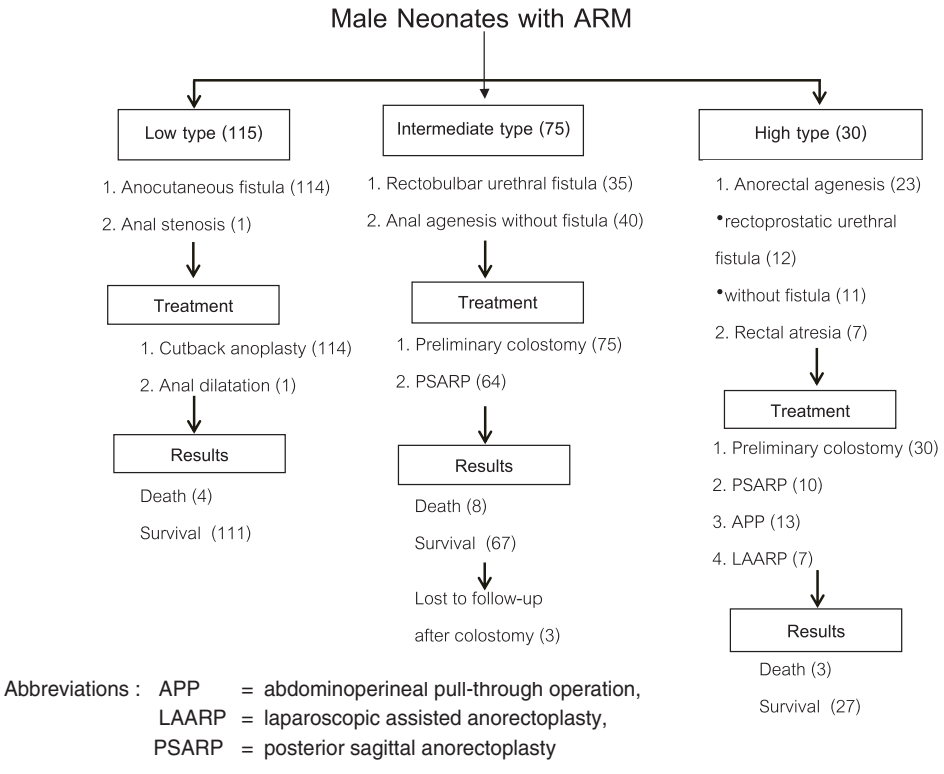

Figure 1 Schematic diagram of male neonates with ARM for the classification, operative treatment and outcomes

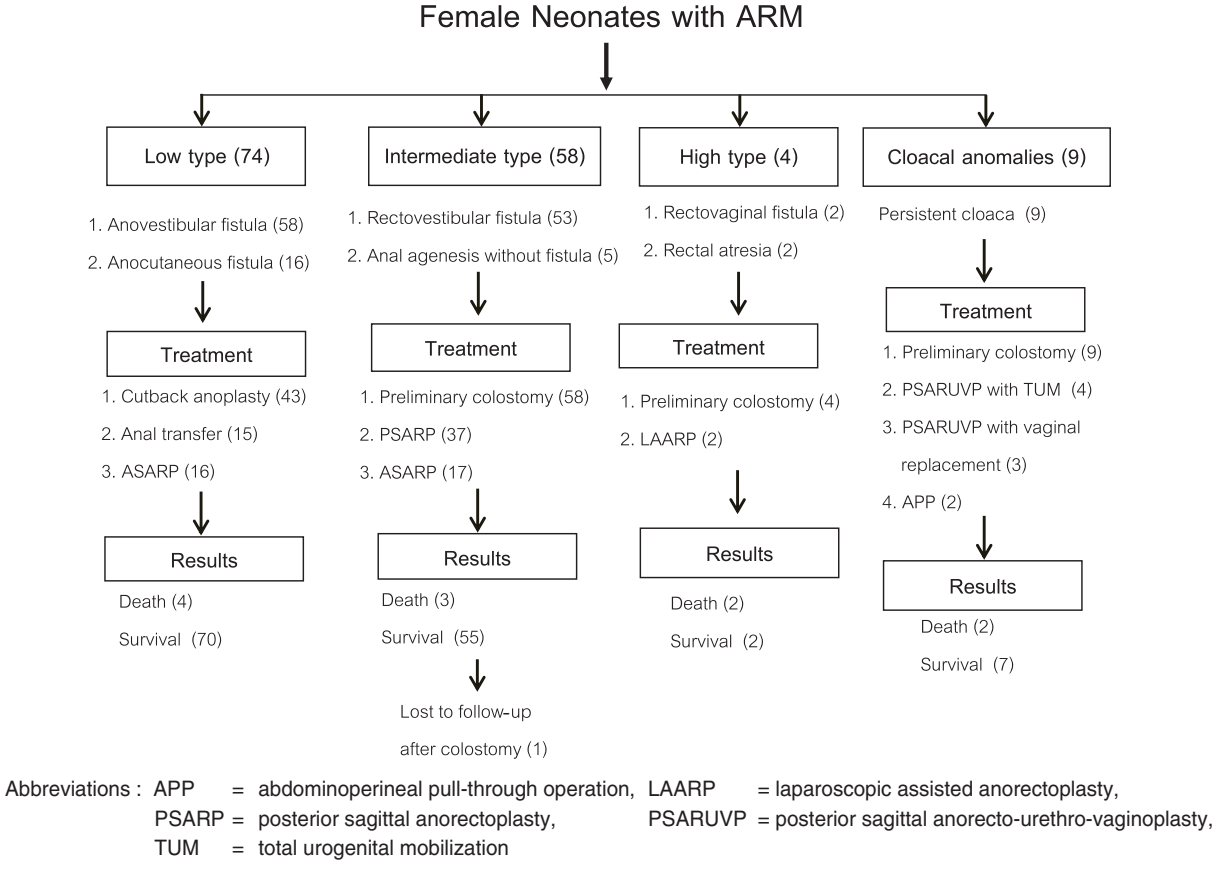

Figure 2 Schematic diagram of female neonates with ARM for the classification, operative treatment and outcomes

80% of the patients, both male and female, had fistulas (Figure 1, 2).

Associated anomalies

Table 2 showed congenital associated anomalies in 216 patients (59.2%) and some cases had more than one anomaly. Genitourinary abnormalities were the most common associated anomalies and were significantly different in female neonates with cloaca and high type of ARM ($p < 0.05$). Genitourinary anomalies were noted in 119 cases included hydronephrosis (43 cases), vesicoureteric reflux (33 cases), renal agenesis (22 cases), hypospadias (12 cases) and others (8 cases). Congenital heart diseases (CHD) were more common in intermediate and high ARM than low ARM ($p < 0.05$). CHD included patent ductus arteriosus (58 cases), atrial septum defect (53 cases), ventricular septal defect (37 cases), tetralogy of Fallot (14 cases) and complex heart diseases (25 cases). Esophageal atresia was the most common alimentary anomalies and was noted in 26 cases. Other alimentary abnormalities were gastroesophageal reflux (8 cases), duodenal atresia (6 cases) and malrotation of the intestine (2 cases). Musculoskeletal anomalies were noted in 77 cases included vertebral, radius and rib abnormalities in 49, 19 and 6 cases, respectively. Tethered cord syndrome was noted in 9 cases of neurological anomalies. Four cases of persistent cloaca had musculoskeletal anomalies, whereas tethered cord anomalies were mostly associated with high ARM. Chromosomal anomalies were noted in 23 cases and included trisomy 21 (12 cases), trisomy 13 (4 cases), Cornelia de Lange syndrome (2 cases), and chromosome 46xx del 10 (1 case).

Operative management and immediate outcomes

Low anomalies

Cutback anoplasty was the most common procedure for management of ARM with low anomalies, 114 cases in male and 43 cases in female (Figure 1, 2). The other procedures for anovestibular and anocutaneous fistula in females were anoplasty by anal transfer (15 cases) and anterior sagittal anorectoplasty or ASARP (16 cases). These operative procedures could be performed within three months of life without preliminary colostomy. The incidence of wound infection was approximately 10%. Immediate postoperative death (within one month) was noted in 8 cases (4.2% of all low anomalies).

Intermediate anomalies

All of the 75 males and 58 females with intermediate anomalies underwent preliminary colostomy within a few days after birth. Definitive operative procedures for intermediate anomalies were posterior sagittal anorectoplasty (PSARP) and ASARP. Laparoscopic assisted anorectoplasty (LAARP) were done in two males with rectobular urethral fistula (Figure 1, 2). Eleven cases in male and 4 cases in females were preliminarily treated colostomy without a definitive procedure because 11 cases died after colostomy and 4 cases lost to follow-up. Mortality rate of all intermediate anomalies was 8.3% (11 in 133 cases).

High anomalies

Preliminary colostomy were done in all 30 males and 4 females with high anomalies. PSARP and abdominoperineal pull-through operation (APP) were definitive procedures in high anomalies. Five cases of male and two cases of female were treated by LAARP

Table 2 Associated congenital anomalies of the 365 patients with anorectal malformations

Associated anomalies	Low type (n=189) (%)	Intermediate type (n=133) (%)	High type (n=34) (%)	Cloacal anomalies (n=9) (%)
Genitourinary system (119)	40 (21.2)	42 (31.6)	28 (82.4)	9 (100)
Cardiovascular system (111)	33 (17.5)	56 (42.1)	19 (55.9)	3 (33.3)
Alimentary system (39)	9 (4.8)	17 (12.8)	12 (35.3)	1 (11.1)
Musculoskeletal system (77)	34 (18.0)	30 (22.6)	9 (26.5)	4 (44.4)
Neurological system (9)	0	2 (1.5)	7 (5.2)	0
Chromosomal anomalies (23)	7 (3.7)	13 (9.8)	2 (5.9)	1 (11.1)

Table 3 Long-term postoperative outcomes of the 335 survivals based on Krickenbeck classification[8]*

Results	Low type (n=181) (%)	Intermediate type (n=118) (%)	High type (n=29) (%)	Cloacal anomalies (n=7) (%)	*P*-value
1. Voluntary bowel movement or continence (212)	130 (71.8)	83 (70.3)	15 (51.7)	5 (71.4)	0.04
2. Soiling or incontinence (27)	5 (2.8)	13 (11.0)	7 (24.1)	2 (28.6)	< 0.01
3. Constipation (97)	44 (24.3)	37 (31.4)	12 (41.4)	3 (42.9)	0.39

*Krickenbeck classification for postoperative results

1. Voluntary bowel movement (continence); feeling of urge, capacity to verbalize, hold the bowel movement
2. Soiling: grade 1 occasionally (once/twice per week), grade 2 everyday, no social problem, grade 3 constant, social problem
3. Constipation: grade 1 manageable by change of diet, grade 2 requires laxatives, grade 3 resistant to laxatives and diet

(Figure 1, 2). Only colostomy was done in 2 females without definitive treatment because of immediate postoperative death. Mortality rate of all high anomalies was 17.6% (6 in 34 cases).

Cloacal anomalies

All of the nine cases with persistent cloaca underwent posterior sagittal anorecto-urethro-vaginoplasty (PSARUVP) with total urogenital mobilization (TUM), vaginal replacement and APP in 4, 3 and 2 cases, respectively. Two cases died after definitive surgical correction. The mortality rate was 22.2% (2 in 9 cases).

Long-term outcomes

Of the 365 patients with ARM, 26 cases (7.1%) succumbed after operation due to CHD, sepsis, respiratory and neurological problems and chromosomal anomalies. Four cases were lost to follow-up after preliminary colostomy without definitive procedure. The remaining 335 patients were available for evaluation of bowel function. Modified Krickenbeck classification[8] for postoperative results was used to evaluate these patients for fecal continence, soiling or incontinence and constipation without grading of the functions because of incomplete detailed information in the medical records (Table 3). Voluntary bowel movement or continence were noted in approximately 70% of low, intermediate and cloacal anomalies, while patients with high anomalies had normal bowel function in 51.7% (70% vs 51.7% p = 0.04). Fecal soiling or incontinence was found in every type of ARM, but increased incidence over 20% in high and cloacal anomalies (p<0.01). Constipation that required dietary, medical and toilet training therapies was noted over 20% in all types of ARM and also increased incidence over 40% in high and cloacal anomalies (p = 0.39). Follow-up period ranged from 1 to 8 years (average 5 years 3 months).

Discussion

The incidence of ARM in Thailand studied at Rajavithi Hospital was similar between the 2 periods, 1 : 2, 560 live births in the previous study in 1988[4] and 1 : 2,820 live births in the present study. In addition, associated anomalies with ARM were also similar, compared between the previous study in 2008[10] and the present study. Three major anomalies associated with genitourinary, cardiovascular and musculoskeletal system. In contrast, many investigators reported that genitourinary and spinal abnormalities were the most common anomalies association with ARM and CHD were less common than vertebral abnormalities[11-15].

At our institute, pediatric surgeons categorize ARM based on Wingspread Classification[6]. The present study revealed the incidence of low (51.8%), intermediate (36.4%), high (9.3%) and cloacal anomalies (2.5%), respectively. Endo[14] analyzed 1992 cases with ARM and found low (57.2%), intermediate (10.7%), high (26%), miscellaneous (4.5%) and unclassified anomalies (1.8%), different from our study. Endo[14] reported high type more common than intermediate type, whereas the present study revealed the lowest incidence in high type. We found that anocutaneous fistula in males and anovestibular fistula in females were the most common anomalies, similar

to Endo's report.

The same type of ARM could be treated by different operative procedures and obtained different outcomes, anovestibular fistula (low type) in female could be treated by cutback anopasty, anal transfer and ASARP. The intermediate and high anomalies could be chosen PSARP, ASARP, LAARP and APP as the surgeons prefer. The overall mortality rate in this study was 7.1% that was similar to the report of Bhargava[16]. Major causes of death included CHD, sepsis, respiratory and neurological complications and associated with chromosomal anomalies.

Long-term outcomes after definitive surgical correction were focused on voluntary bowel movement or continence, soiling or incontinence and constipation based on Krickenbeck classificaltion[8]. The overall incidence of voluntary bowel movement or continence in the present study was 65% (233 in 335 patients). Low and intermediate types had the continence rate approximately 71% which were better than 51% continence in high type. However, soiling and constipation occurred in patients with low, intermediate and high ARM. Rintala[17] suggested that long-term follow-up of patients with low ARM was to treat the main functional problem, constipation, which also was the cause of soiling. Rintala[18] reported the outcomes of high ARM, 35% excellent, 35% good and 30% fair and poor continence outcomes. Presence of sacral or intraspinal anomalies affected the significant cause of incontinence in intermediate and high ARM. From the literature review, many articles revealed improvement in sphincter function in terms of decreasing quantity of soiling with increasing age of the patients[19-22]. Improvement of the overall clinical outcome was related to successful treatment or spontaneous resolution of constipation and the complications of constipation including fecal impaction and over flow soiling[23]. Templeton[24] suggested that patients with a poor or fair fecal continence result and having the problems of social life should be offered a secondary repair.

Peña classification[7] has been preferable and his operative technique, PSARP, has been generally accepted amongst pediatric surgeons[15,25,26]. PSARP can be performed as a secondary operation for the treatment of fecal incontinence[27]. The functional outcomes of repair ARM have significantly improved since PSARP has been advocated[15,26,28,29]. In the present study, PSARP was the most common procedure for intermediate and high ARM. ASARP, which modified from PSARP, was performed in some female cases with intermediate anomalies (rectovestibular fistula).

The present study has some limitations because it was a retrospective review. Patients' data were obtained from the medical records which were incomplete, in some details such as scoring or grading of constipation and functional bowel control. This review could be a baseline for future study, planning health care services for patients with ARM.

Conclusion

Patients with ARM in this study were orderly categorized into low, intermediate, high and cloacal anomalies. Approximately 80% of the cases had fistulas, either perineal or genitourinary fistula. Anoplasty was the definitive procedure for correction of low ARM, while PSARP was the most common in intermediate and high ARM. Mortality of the patients was affected from congenital anomalies, especially CHD, chromosomal anomalies, neurological and respiratory problems. Low and intermediate ARM obtained a good result, whereas high anomalies had a fair result for continence. Constipation requiring toilet training, dietary or medical treatment was found in every type of ARM and complicated fecal impaction overflow soiling. Long-term follow-up of these patients to manage constipation, the main functional problem, is warranted.

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บทคัดย่อ **ผลการรักษาความผิดปกติของทวารหนักและไส้ตรง: การศึกษาในระยะเวลา 7 ปี ที่สถาบันสุขภาพเด็กแห่งชาติมหาราชินี**

มารินทร์ พลขันธ์, พ.บ.*, สุรเนตร ลออวงศ์, พ.บ.*, วราภรณ์มหธราดล, พ.บ.*, รังสรรค์ นิรามิษ,พ.บ.*
**กลุ่มงานศัลยศาสตร์ สถาบันสุขภาพเด็กแห่งชาติมหาราชินี กรุงเทพฯ*

ความเป็นมา: ความผิดปกติของทวารหนักและไส้ตรงเป็นภาวะที่พบได้ไม่บ่อยนัก อุบัติการณ์โดยทั่วไปพบ 1 ใน 4,000 ถึง 5,000 ทารกเกิดมีชีพ การแบ่งชนิดและการผ่าตัดแก้ไขมีหลากหลายวิธีที่ถูกนำเสนอและผลการผ่าตัดรักษาโรคนี้ถูกรายงานว่าดีขึ้นอย่างต่อเนื่อง

วัตถุประสงค์: เพื่อศึกษาถึงประสบการณ์ในการรักษาผู้ป่วยที่มีความผิดปกติของทวารหนักและไส้ตรงในเรื่องของการแบ่งชนิด ความผิดปกติร่วม การผ่าตัดแก้ไข และผลของการรักษาหลังการผ่าตัด

วัสดุและวิธีการ: เป็นการศึกษาย้อนหลังจากการทบทวนเวชระเบียนของผู้ป่วยที่ความผิดปกติของทวารหนักและไส้ตรงที่เข้ามารักษาในสถาบันสุขภาพเด็กแห่งชาติมหาราชินี ระหว่างปี ค.ศ. 2006 ถึง ค.ศ. 2012 ข้อมูลของผู้ป่วยที่รวบรวมประกอบด้วย ข้อมูลทั่วไป ชนิดของความผิดปกติ ความผิดปกติร่วม วิธีการผ่าตัดและผลของการรักษา การแบ่งชนิดของความผิดปกติใช้ตาม Wingspread Classification และประเมินผลหลังการผ่าตัดรักษาตาม Kickenbeck Classification

ผล: ผู้ป่วยทั้งหมด 365 ราย (เพศชาย 220 ราย เพศหญิง 145 ราย) ได้รับการรักษาโรคนี้ระหว่างปีที่ทำการศึกษา อุบัติการณ์ความผิดปกติของทวารหนักและไส้ตรง พบ 1 : 2,820 ทารกเกิดมีชีพที่โรงพยาบาลราชวิถี มากกว่าร้อยละ 70 ของผู้ป่วยเป็นทารกที่คลอดครบกำหนดและน้ำหนักแรกเกิดมากกว่า 2,500 กรัม ระดับความผิดปกติของทวารหนักและไส้ตรงพบระดับต่ำ ระดับกลาง และระดับสูง 115 (ร้อยละ 52.3), 75 (ร้อยละ 34.1) และ 30 ราย (13.6) ในเพศชาย และพบ 74 (ร้อยละ 51.0), 58 (ร้อยละ 40.0) และ 4 ราย (ร้อยละ 2.8) ตามลำดับในเพศหญิง พบ persistent cloaca 9 ราย (ร้อยละ 6.2) ในเพศหญิง เกือบทั้งหมดของผู้ป่วยที่มีความผิดปกติระดับต่ำได้รับการรักษาโดยวิธี cutback anoplasty การผ่าตัดของความผิดปกติชนิดต่ำในเพศหญิงมีทางเลือกอื่นคือ anoplasty by anal transfer และ anterior sagittal anorectoplasty (ASARP) ในผู้ป่วย 31 ราย โดยไม่มีการทำ colostomy ไว้ก่อน การผ่าตัดที่ทำมากที่สุดในผู้ป่วยที่มีความผิดปกติระดับกลางคือ posterior sagittal anorectoplasty (PSARP) และ (ASARP) ในเพศหญิง สำหรับการผ่าตัดแก้ไขในความผิดปกติระดับสูงคือ PSARP และ abdominoperineal pull-through operation (APP) การใช้การส่องกล้องช่วยในการผ่าตัดแก้ไขทำในผู้ป่วยที่มีความผิดปกติระดับสูง ในเพศชาย 7 ราย และ 2 รายในเพศหญิง ผู้ป่วยเด็กหญิง 9 รายที่มี persistent cloaca ได้รับการผ่าตัด posterior sagittal anorecto-urethro-vaginoplasty และมีผู้ป่วยรอดชีวิต 7 ราย ความพิการแต่กำเนิดที่พบร่วมมากที่สุดคือ ความผิดปกติของระบบสืบพันธุ์ร่วมกับทางเดินปัสสาวะและความพิการของระบบหัวใจและหลอดเลือด มีผู้ป่วยเสียชีวิตหลังผ่าตัด 26 ราย (ร้อยละ 7.1) สาเหตุจากความพิการของหัวใจแต่กำเนิด การติดเชื้อในกระแสโลหิต การหายใจผิดปกติและปัญหาระบบสมองและประสาท ผลการรักษาในระยะยาวประเมินในผู้ป่วย 335 ราย พบว่ามีการถ่ายและกลั้นอุจจาระได้เป็นปกติ ร้อยละ 51.7-71.8 กลั้นอุจจาระไม่ได้ร้อยละ 2.8-22.2 และมีอาการท้องผูกร้อยละ 24.3-42.9 ผู้ป่วยที่มีความผิดปกติระดับต่ำและระดับกลางมีผลการรักษาระยะยาว หลังผ่าตัดดีกว่าผู้ป่วยที่มีความผิดปกติระดับสูง

สรุป: ประมาณร้อยละ 70 ของความผิดปกติของทวารหนักและไส้ตรงระดับต่ำและระดับกลางมีการถ่ายและกลั้นอุจจาระได้ปกติและมีการกลั้นอุจจาระไม่ได้ในอัตราต่ำ ความผิดปกติของทวารหนักและไส้ตรงทุกระดับในระยะยาวมีปัญหาเรื่องท้องผูกและต้องการรักษาด้วยเรื่องการปรับอาหาร ยาระบายและฝึกการถ่ายอุจจาระ