

Case Report

FAP Concurrent with Colon Cancer Managed by Total Proctocolectomy with Reverse Total Mesorectal Excision: A Case Report and Literature Review

Wen-Shih Huang^{1,2}

Tsan-Hung Lee¹

Chih-Jung Chen¹

Chih-Chien Chin^{1,2}

Yi-Hung Kuo¹

Cheng-Yi Huang¹

¹*Division of Colon and Rectal Surgery,
Department of Surgery, Chang Gung
Memorial Hospital, Chiayi,*

²*Graduate Institute of Clinical Medical
Science, College of Medicine, Chang Gung
University, Taoyuan, Taiwan*

Restorative total proctocolectomy (RTPC) is the surgical option for patients of familial adenomatous polyposis (FAP) with concurrent colorectal cancer (CRC) or at high risk of developing CRC in the future. Open or laparoscopy-assisted procedures have been proposed. We proposed, herein, a case of FAP with synchronous sigmoid colon cancer in a 17-year-old adolescent surgically treated as single-incisional total proctocolectomy combining with transanal endoscopic total mesorectal excision (TME) and inter-sphincteric resection (ISR) under the assistance of laparoscopic assistance and ileal-J-pouch anal anastomosis (IJPAA) approached by self-made multichannel port system. Literature review about minimal invasive surgery to manage FAP is also presented.

[J Soc Colon Rectal Surgeon (Taiwan) 2014;25:151-157]

Key Words

Familial adenomatous polyposis

(FAP);

Single-incisional laparoscopic surgery

(SILS);

Transanal total mesorectal excision

(TME)

Restorative total proctocolectomy (RTPC) with ileal-J-pouch anal anastomosis (IJPAA) for gastrointestinal tract reconstruction is the golden surgical treatment option for the patients of familial adenomatous polyposis (FAP) with concurrent colorectal cancer (CRC) or as prophylaxis to whom without underlying malignancy. It has advantages than conventional total proctocolectomy without restoring the gastrointestinal continuity on both improving the life quality and reducing the cancer-related mortality. Either

RTPC with IJPAA or total proctocolectomy (TPC) with permanent end ileostomy could be performed by laparotomy or laparoscopic approach procedures. Surgical expertise, the clinical facilities and patient's performance status would dictate outcomes. Herein, we proposed a 17-year-old male adolescent underlying FAP with synchronous sigmoid colonic cancer and profuse polyposis in the whole large intestine with villous transformation among the lowest located polyps just above the dentate line. We resumed self-

Received: March 3, 2014.

Accepted: July 29, 2014.

Correspondence to: Dr. Wen-Shih Huang, Division of Colon and Rectal Surgery, Department of Surgery, Chang Gung Memorial Hospital, No. 6, Sec. West, Chia-Pu Road, Putz City, Chiayi 613, Taiwan. Tel: +886-5-362-1000 ext. 2862; Fax: +886-5-362-3001; E-mail: wshuang77@hotmail.com

made multichannel port system approach both trans-abdominally and transanally to complete RTPC and JPAA procedures accessed via a single aperture. In particular, a novel device comprising an optimal cost with terrific efficacy was substituted to commercial ready-made access to fulfil transanal total mesorectal excision (TME) and inter-sphincteric resection (ISR).

Case Report

Brief medical history

A 17-year-old male adolescent without any previous medical history presented with a history of intermittent blood-tinged or mucus-mixing stool passage for more than six months. He visited out-patient department of proctology at our institute. Rigid sigmoidoscopy was performed and disclosed diverse-sized polyps distributing since dentate line upwards till 22 cm level and a sigmoid colon polypoid tumor hanging over 20 cm level from the anus. He has a familial history of FAP, inclusive of his mother, elder aunt and younger sister. Under the impression of FAP with highly suspicious of synchronous CRC, he was admitted for further workup. Abdominal, chest computed tomography (CT) and complete colonoscopy revealed more than one thousand polyps with possible sigmoid colonic cancer, but no distant metastasis was identified. Initial serum carcinoembryonic antigen (CEA) level was 2.28 ng/mL. He underwent synchronous SILS RPC with nature orifice transanal laparoscopic endoscopic TME accessed by self-made multichannel port system, ileal-J-pouch anal anastomosis and diverting loop ileostomy at incision site. Tumor size was 2.2×1.6 cm, and pathology showed carcinoma-in-situ with no metastasized lymph node in 39 lymph nodes harvested. He discharged smoothly at post-operatively day 10.

A novel device applied in transanal TME

Performing nature orifice transanal endoscopic TME with the assistance of laparoscopic instruments on this case, we abolished commercial ready-made

port system for this procedure, as transanal endoscopic surgery (TEM) equipment, SSL port (Ethicon Endo-Surgery Inc., Cincinnati, OH), Gelpoint path (Applied Medical, Rancho Santa Margarita, CA), SILS port (Covidien, Mansfield, Massachusetts) and TEO proctoscope device (Karl Storz, Tuttlingen, Germany). We applied a novel device to complete transanal TME procedure. This self-made device consisted of a surgical glove inserting through anal dilator, a part of the PPH-03 (Ethicon Endo-Surgery Inc., Cincinnati, OH), followed by the sleeve end of the glove was reverted to cover the bottom of the dilator (Fig. 1-a & Fig. 1-b). 3 pieces of trocar were inserted into finger part of the glove with silk thread tie-up for fixation.

Surgical technique

The patient with BMI of 18 kg/m^2 was placed in lithotomy position with the anus and perineum exposed well and operation table tilted as head-down position. 6 anchoring sutures were placed perianally for exposing the anal canal caudally as possible as could. Circumferential incision aimed at ISR was done over anorectal ring to mobilize the full-thickness of distal rectum cephalically about 4 cm above the incision line with the aid of PPH suture anoscope

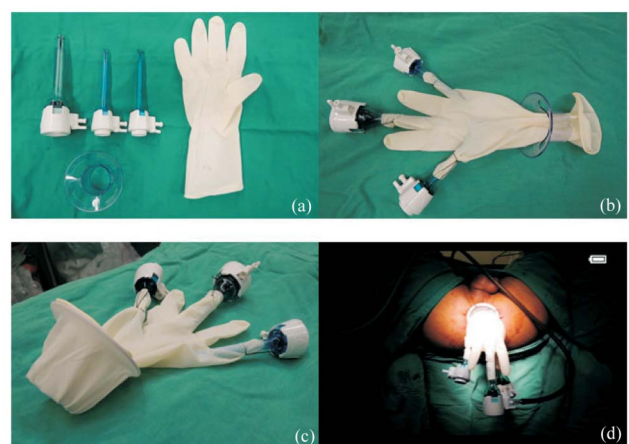


Fig. 1. (a/b) Components of a novel device for reverse TME, as PPH-anal dilator, a surgical glove and multiple trocars; (c) completion of the multichannel port system; (d) port system inserted into distal rectum.

(Ethicon Endo-Surgery Inc., Cincinnati, OH). Subsequently, occlusion of proximal rectal stump was done to prevent tumor implantation during the manipulation. With the aid of cartridge, such a self-made multichannel port system was inserted from the anal canal into the distal rectum. 4 stitches of anchoring suture were placed from the side-hole of the bottom of the anal dilator perianally with the intent of fixation of such a system. A 30-degree 10-mm laparoscopy A50002A (Endo-Eye, Olympus Inc., Japan) was inserted from the central trocar; one grasp and another energy device as Harmonic scalpel were inserted from the other trocars. Standard laparoscopic insufflator, for insufflating 1 to 3L of CO₂ per minute to a pressure of 20 mm-Hg to the extra-peritoneal perirectal space, and the transanal TME was practiced as bottom-to-up method (Fig. 1-c). TME plane was identified in a reverse direction initiating on top of the puborectal muscle. We performed anterior dissection firstly, under direct vision by using conventional laparoscopic instruments by dissecting on Denonvilliers fascia (retro-prostatic plane) as cephalad as possible, till the Douglas pouch was opened (Fig. 2-a). Lateral dissection involved dividing the middle hemorrhoidal artery was then performed with the mesorectal fascia preserved. Adequate lateral dissection was completed as could as possible, posterior TME plane was formed;

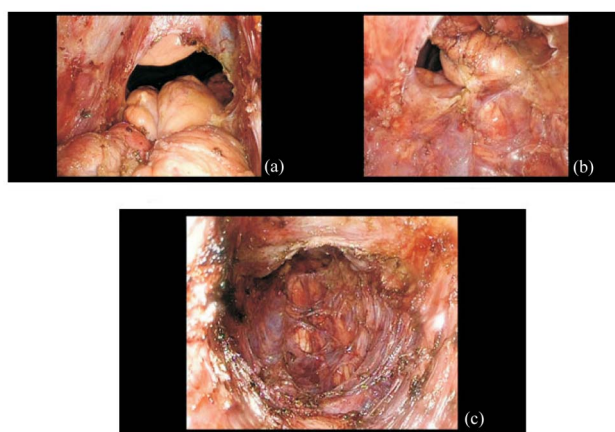


Fig. 2. (a) Anterior dissection plane of reverse TME with Douglas pouch opened; (b) posterior dissection plane of reverse TME at sacral promontory; (c) transanal bottom-to-top view with the whole rectum positioned in the peritoneal cavity.

subsequently, posterior dissection was performed by bottom-to-top orientation along with the Waldeyer's plane till the level of sacral promontory (Fig. 2-b). After transanal TME was done adequately (transanal bottom-to-top view, Fig. 2-c), then, transabdominal approach was practiced as SILS accessed by glove-port multichannel system with incision over abdominal right low quadrant (planned ileostomy site). Standard procedure of laparoscopic total colectomy was initiated by medial-to-lateral mobilization of the left-sided colon to divide inferior mesenteric vessels and to mobilize the splenic flexure colon. Lesser sac was opened and transverse mesocolon was mobilized and perfusion vessels were then divided. Subsequently, right-sided colon was dissected from retroperitoneal plane and relative vessels were divided but ileocolic vessels were preserved at their origin. Total colorectum was retrieved from the abdominal incision wound due to bulky specimen relative to narrow anal orifice and divided at distal ileum (Fig. 3-a). The specimen was opened at the operation setting later (Fig. 3-b). After creation of 16-cm sized ileal-J-pouch was completed, pneumoperitoneum was constructed again, the ileal-J-pouch was then pulled up at the anal orifice under the laparoscopic view intended to maintain adequate pouch orientation and axis. Reconstruction of intestinal tract by ileal-J-pouch anal anastomosis was

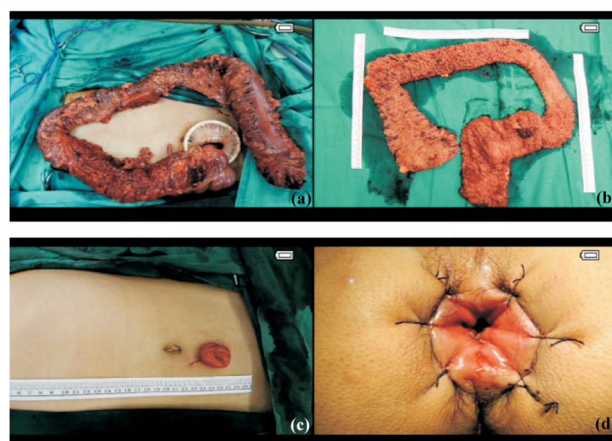


Fig. 3. (a) Total colorectum retrieved from the positioned ileostomy site; (b) the opened specimen; (c) abdomen with an incision wound and ileostomy matured; (d) coloanal anastomosis completion.

created by handsewn continuous suture with 3-0 V-Loc thread (Covidien, Mansfield, Massachusetts). Due to bloodless course in this surgical course, neither pelvic nor abdominal drainage tube was placed. Eventually, the abdomen with an incision wound and ileostomy matured (Fig. 3-c), and the ileal-J-pouch anal anastomosis was completed without a drainage tube left (Fig. 3-d). The operation time consumption was described as followings: (1) ISR under direct vision: 45 minutes; (2) transanal TME under assistance of laparoscopic device: 80 minutes; (3) transabdominal single-incisional laparoscopic total colectomy with ileal-J-pouch anal anastomosis: 275 minutes. In sum, it took 400 minutes to resume single-incisional total proctocolectomy combining with transanal endoscopic total mesorectal excision under the assistance of laparoscopic instruments approached by self-made multichannel port system.

Discussion

FAP is characterized by the presence of multiple colorectal adenomatous polyps (typically more than one hundred), which occurs in approximately 1/10,000 to 1/30,000 live births, and accounts for less than 1 percent of all colorectal cancers in the United States.¹ The increased risk of CRC in FAP is due to the vast number of adenomas that occur at an early age; with time, one at least among these adenomas inevitably progress to CRC.² Polyposis generally develops in the second or third decade of life. The mean age of polyp crisis is sixteen years; but the onset of polyps has been identified in patients between the ages of 8 and 34 years.³ Colectomy is strongly reserved for patients with concurrent CRC; in addition, the patients with multiple large (> 1 cm) adenomas or adenomas with villous histology and/or high-grade dysplasia. The preferred operation in children with extensive polyposis included the rectum is a RTPC with IJPAA. A subtotal colectomy with ongoing surveillance or a total colectomy is reasonable in patients with attenuated FAP having minor rectal involvement (fewer than 20 around adenomas in the rectum).⁴ This presented 17-year-old male adolescent underlying FAP with syn-

chronous sigmoid colonic cancer and profuse polyposis in the whole large intestine with villous transformation among the lowest located polyps just above the dentate line; in hence, RTPC with IJPAA was recommended for his surgical intervention.

The ileal-J-pouch could be served as an internal pelvic reservoir for intestinal contents.⁵⁻⁷ Friederich et al. proposed despite the high risk of adenomas, the risk of malignant transformation appeared to be low from a report of 212 patients.⁸ The cumulative risk of developing a pouch cancer was estimated to be 1 percent at 10 years. Regardless of the precise risk evaluation, regular surveillance of any remaining rectum or the ileal pouch is recommended after colectomy.⁸

Minimal invasive surgery to practice RTPC is encompassed to fast-track protocols with merits of a decrease in inflammatory mediators, improved pulmonary function with oncological outcomes comparable to the open access, faster return of bowel function, reduced hospital length of stay, and decreased operation-related morbidities.⁹⁻¹² Our colectomy team has practiced more than 300 single-incision laparoscopic surgery (SILS) for colectomies by glove port since 2011 May. To our surprise, SILS for our previously performed colectomy in CRC with conventional straight laparoscopic instruments were not a difficult procedure. We found that the glove port had some advantages that the instruments and the laparoscope are not confined by rigid laparoscopic ports because of the optimal distensibility with tactility of the surgical glove and the port system could be anchored at the abdominal wall from the Alexis wound retractor (Applied Medical, Rancho Santa Margarita, CA) inlayed extra-/intra-peritoneally. In addition, the port system we using has an extremely low cost than other commercially available access devices. Moreover, we develop "substitution method", it is, withdrawal of the jerkiest instrument except the scope and reinserting from the trocar to overcome the restrained motion of the instrument. Besides, opening of bilateral jaws of the bowel grasper to form 2-point traction site in the dissection plane or tissue could also facilitate dissection. Straight laparoscope usually creates the so-called end-on view. We used a 30 degree laparoscope to avoid such a view and help us get a more compre-

hensive view to make dissection easier. Surprisingly, by way of the home-made multichannel port system, 30 degree laparoscope and conventional straight instruments, we could perform this patient without conversion to laparotomy or multiple site laparoscopic surgery (MSLS) either transabdominal or transanal approach.

In this present report, we describe a hybrid method of combining transabdominal with transanal approach by the access of single-incisional multichannel port system and straight laparoscopic instruments to complete the procedures of patient necessitating TME with ISR of the rectum or total proctocolectomy. In particular, we performed nature orifice transanal endoscopic TME approached by a novel surgical platform of advantages of economics and no compromising such a novel procedure. Such procedures convey the merits of SILS and transanal TME. In view of SILS, it carried the advantages of some reports as cosmetic profit, fewer length of hospital stay, decreased operation-related morbidity from laparotomy or conventional MSLS such as incisional hernia and adhesion formation either benign or malignant abdominal diseases.¹³⁻¹⁷ Nature orifice transanal endoscopic TME with laparoscopic instruments assistance could overcome some hurdles from transabdominal laparoscopic TME in cases of narrow pelvic anatomy, bony landmarks, and nonadapted instruments, by far in an obese male patient. Transanal TME could also provide the patients requiring manual coloanal anastomosis or intersphincteric proctectomy. Furthermore, it could achieve optimal circumferential and distal section margin free of rectal malignancy.¹⁸⁻²¹

Our procedure was similar to the method Tuech proposed in 2011.²² Either transabdominal or transanal access was approached by single-port, different from the remaining two groups resumed multiport-site approach transabdominally. A purely transabdominal SILS approach could avoid the complications of pain and hemorrhage with improved cosmetics from multiple-site. But, distinct from the former three reports, we applied self-made multichannel port system transabdominally or transanally, not commercial ready-made port system as they used. The major component of self-made port system we used was the sur-

gical glove composing of rubber. Owing to its content of rubber, the surgical glove has the ideal tactility and distensibility. Maintaining the presumed pneumoperitoneal or pneumoperirectal pressure was also reached. In addition, anchoring at the abdomen or anus, the Alexis wound retractor or PPH anal dilator was hardly dislodging out of the fixation site. Moreover, these devices had the economic advantages of low cost other than easy introduction and removal of instruments or devices.

Conclusion

Transabdominal and transanal approach by single-port access for this presented patient of FAP with concurrent sigmoid colonic cancer necessitating TME, ISR and total proctocolectomy under laparoscopy assistance is a feasible and safe procedure. A larger clinical trial of such a procedure in meticulously recruited patients is mandatory to evaluate long-term functional and oncological outcomes.

References

1. Wennstrom J, Pierce ER, McKusick VA. Hereditary benign and malignant lesions of the large bowel. *Cancer* 1974;34: suppl:850-7. doi: 10.1002/1097-0142(197409)34:3+<850::AID-CNCR2820340711>3.0.CO;2-S
2. Burt RW1, DiSario JA, Cannon-Albright L. Genetics of colon cancer: impact of inheritance on colon cancer risk. *Annu Rev Med* 1995;46:371-9. doi: 10.1146/annurev.med.46.1.371
3. Petersen GM1, Slack J, Nakamura Y. Screening guidelines and premorbid diagnosis of familial adenomatous polyposis using linkage. *Gastroenterology* 1991;100:1658-64.
4. Nieuwenhuis MH, Mathus-Vliegen LM, Slors FJ, Griffioen G, Nagengast FM, Schouten WR, et al. Genotype-phenotype correlations as a guide in the management of familial adenomatous polyposis. *Clin Gastroenterol Hepatol* 2007;5:374-8. doi: 10.1016/j.cgh.2006.12.014
5. Parks AG, Nicholls RJ, Belliveau P. Proctocolectomy with ileal reservoir and anal anastomosis. *Br J Surg* 1980;67:533-8. doi: 10.1002/bjs.1800670802
6. Utsunomiya J, Iwama T, Imajo M, Matsuo S, Sawai S, Yaegashi K, et al. Total colectomy, mucosal proctectomy and ileoanal anastomosis. *Dis Colon Rectum* 1980;23:459-66. doi: 10.1007/BF02987076
7. Sagar PM, Pemberton JH. Intraoperative, postoperative and

- reoperative problems with ileoanal pouches. *Br J Surg* 2012; 99:454-68. doi: [10.1002/bjs.8697](https://doi.org/10.1002/bjs.8697)
8. Friederich P1, de Jong AE, Mathus-Vliegen LM, Dekker E, Krieken HH, Dees J, et al. Risk of developing adenomas and carcinomas in the ileal pouch in patients with familial adenomatous polyposis. *Clin Gastroenterol Hepatol* 2008;6:1237-42. doi: [10.1016/j.cgh.2008.06.011](https://doi.org/10.1016/j.cgh.2008.06.011)
 9. Lourenco T1, Murray A, Grant A, McKinley A, Krukowski Z, Vale L. Laparoscopic surgery for colorectal cancer: safe and effective? - A systematic review. *Surg Endosc* 2008;22: 1146-60. doi: [10.1007/s00464-007-9686-x](https://doi.org/10.1007/s00464-007-9686-x)
 10. Novitsky YW1, Litwin DE, Callery MP. The net immunologic advantage of laparoscopic surgery. *Surg Endosc* 2004; 18:1411-9. doi: [10.1007/s00464-003-8275-x](https://doi.org/10.1007/s00464-003-8275-x)
 11. Shea JA, Berlin JA, Bachwich DR, Staroscik RN, Malet PF, McGuckin M, et al. Indications for and outcomes of cholecystectomy: a comparison of the pre and postlaparoscopic eras. *Ann Surg* 1998;227:343-50. doi: [10.1097/00000658-199803000-00005](https://doi.org/10.1097/00000658-199803000-00005)
 12. Fleshman J, Sargent DJ, Green E, Anvari M, Stryker SJ, Beart RW Jr, et al. Clinical Outcomes of Surgical Therapy Study Group. Laparoscopic colectomy for cancer is not inferior to open surgery based on 5-year data from the COST Study Group trial. *Ann Surg* 2007;246:655-62. doi: [10.1097/SLA.0b013e318155a762](https://doi.org/10.1097/SLA.0b013e318155a762)
 13. Maggiori L, Gaujoux S, Tribillon E, Bretagnol F, Panis Y. Single-incision laparoscopy for colorectal resection: a systematic review and meta-analysis of more than a thousand procedures. *Colorectal Dis* 2012;14:e643-54. doi: [10.1111/j.1463-1318.2012.03105.x](https://doi.org/10.1111/j.1463-1318.2012.03105.x)
 14. Makino T, Milsom JW, Lee SW. Feasibility and safety of single-incision laparoscopic colectomy: a systematic review. *Ann Surg* 2012;255:667-76. doi: [10.1097/SLA.0b013e31823fbae7](https://doi.org/10.1097/SLA.0b013e31823fbae7)
 15. Boni L, Dionigi G, Cassinotti E, Di Giuseppe M, Diurni M, Rauseri S, et al. Single incision laparoscopic right colectomy. *Surg Endosc* 2010;24:3233-6. doi: [10.1007/s00464-010-1100-4](https://doi.org/10.1007/s00464-010-1100-4)
 16. Vestweber B, Alfes A, Pual C, Haaf F, Vestweber KH. Single-incision laparoscopic surgery: a promising approach to sigmoidectomy for diverticular disease. *Surg Endosc* 2010; 24:3225-8. doi: [10.1007/s00464-010-1090-2](https://doi.org/10.1007/s00464-010-1090-2)
 17. Diana M, Dhumane P, Cahill RA, Mortensen N, Leroy J, Marescaux J. Minimal invasive single-site surgery in colorectal procedures: Current state of the art. *J Minim Access Surg* 2011;7:52-60.
 18. Wolthuis AM, Cini C, Penninckx F, D'Hoore A. Transanal single port access to facilitate distal rectal mobilization in laparoscopic rectal sleeve resection with hand-sewn coloanal anastomosis. *Tech Coloproctol* 2012;16:161-5. doi: [10.1007/s10151-011-0795-0](https://doi.org/10.1007/s10151-011-0795-0)
 19. Velthuis S, van den Boezem PB, van der Peet DL, Cuesta MA, Sietses C. Feasibility study of transanal total mesorectal excision. *Br J Surg* 2013;100:828-31. doi: [10.1002/bjs.9069](https://doi.org/10.1002/bjs.9069)
 20. Sylla P, Bordeianou LG, Berger D, Han KS, Lauwers GY, Sahani DV, et al. A pilot study of natural orifice transanal endoscopic total mesorectal excision with laparoscopic assistance for rectal cancer. *Surg Endosc* 2013;27:3396-405. doi: [10.1007/s00464-013-2922-7](https://doi.org/10.1007/s00464-013-2922-7)
 21. Wolthuis AM, de Buck van Overstraeten A, D'Hoore A. Dynamic article: transanal rectal excision: a pilot study. *Dis Colon Rectum* 2014;57:105-9. doi: [10.1097/DCR.0000000000000008](https://doi.org/10.1097/DCR.0000000000000008)
 22. Tuech JJ, Bridoux V, Kianifard B, Schwarz L, Tsilivlidis B, Huet E, Michot F. Natural orifice total mesorectal excision using transanal port and laparoscopic assistance. *Eur J Surg Oncol* 2011;37:334-5. doi: [10.1016/j.ejso.2010.12.016](https://doi.org/10.1016/j.ejso.2010.12.016)

病例報告

家族性腺性息肉症候群併乙狀結腸癌以單切口腹腔鏡手術切除全結腸直腸合併經由自製之多通道埠系統及腹腔鏡器械輔助下經肛門行全直腸系膜切除：病例報告與文獻回顧

黃文詩^{1,2} 李讚紘¹ 陳志榕¹ 靳志堅^{1,2} 郭益宏¹ 黃政義¹

¹嘉義長庚紀念醫院 外科部 大腸直腸肛門外科

²長庚大學 臨床醫學研究所

恢復性全結腸直腸切除術是家族性腺性息肉症候群 (FAP) 併發結直腸癌 (CRC) 或在將來可能發展結直腸癌的高風險病患之手術選擇。已有開腹或藉由腹腔鏡輔助之方式進行此項手術。此病例報告為一 17 歲青少年患有家族性腺性息肉症候群併乙狀結腸癌，我們建議，以單切口腹腔鏡手術切除全結腸直腸合併經由自製之多通道埠系統及腹腔鏡器械輔助下經肛門行全直腸系膜切除合併回腸 J 型與肛管吻合術 (ileal-J-pouch anal anastomosis)。此外介紹了微創手術來治療 FAP 與其文獻回顧。

關鍵詞 家族性腺性息肉症候群 (FAP)、單切口腹腔鏡手術 (SILS)、經肛門直腸全系膜切除 (transanal TME)。