



UZ  
LEUVEN



## Evolutie van 30 jaar opvolging: van chirurgie tot therapie

**Peter-Jan Vancoillie, MD**

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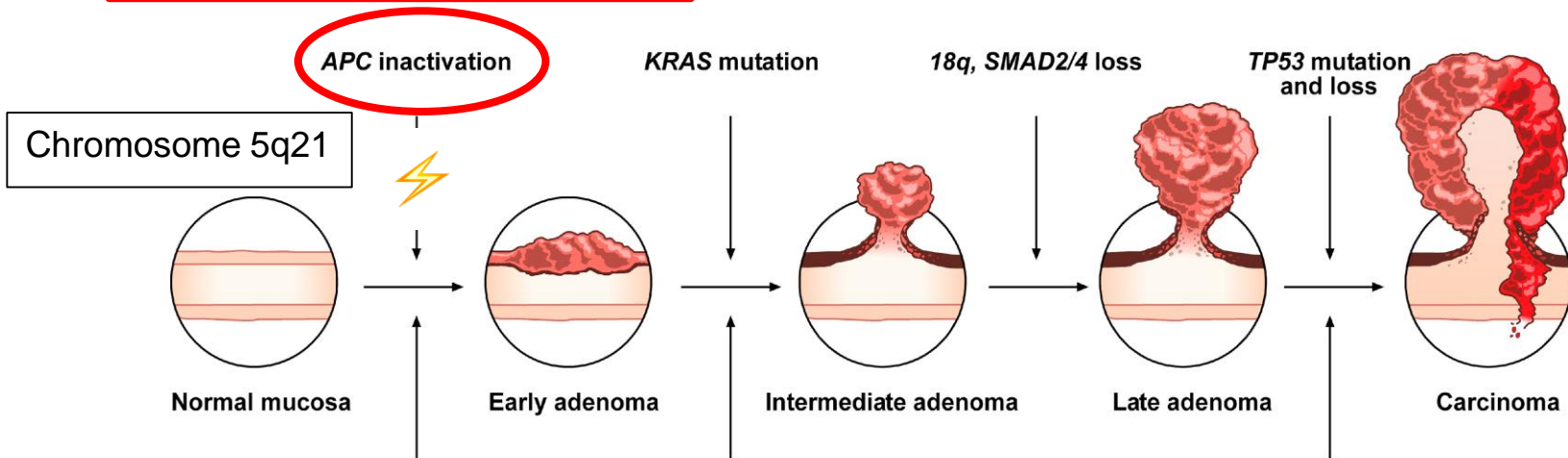
*Dienst Abdominale Heelkunde*

*Universitair Ziekenhuis, Gasthuisberg, Leuven, België*



# Familiale Adenomateuze Polypose (FAP)

CIN - Chromosomal Instability pathway

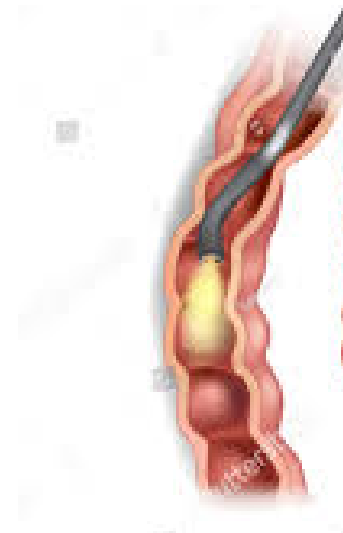


# Familiale Adenomateuze Polypose (FAP)

- FAP leidt 100% tot colorectale tumor
- Kanker ontwikkelt gemiddeld op de leeftijd van 39 jaar
- 'Attenuated' (=afgezwakte) FAP (<100 poliepen, 70% risico op kanker, gemiddelde leeftijd 58 jaar)
- Andere afwijkingen buiten het colon
  - *Poliepen thv maag en duodenum*
  - *Osteomen, epidermoïd cysten, tand afwijkingen, papillair schildkliercarcinoom*
  - *Desmoïdtumoren*
- Chirurgie is dé behandeling van FAP – preventief!

# Familiale Adenomateuze Polypose (FAP)

Van start screening tot timing heekunde



12-14j  
Coloscopie elke 1-3j



15-25j  
MAAR...

# Familiale Adenomateuze Polypose (FAP)

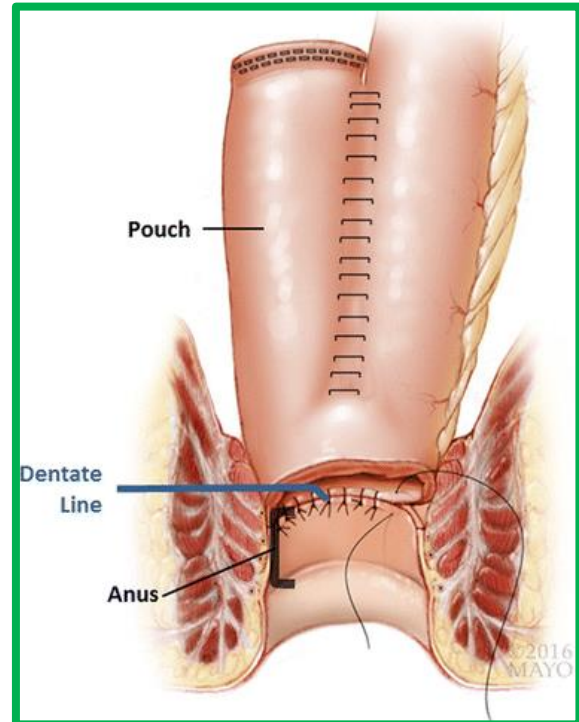
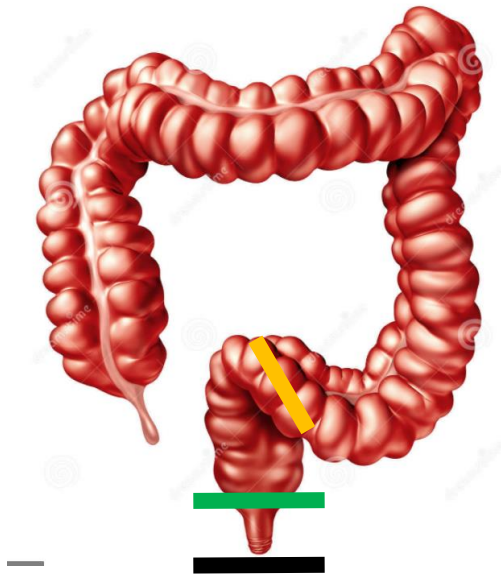
## Van start screening tot timing heelkunde

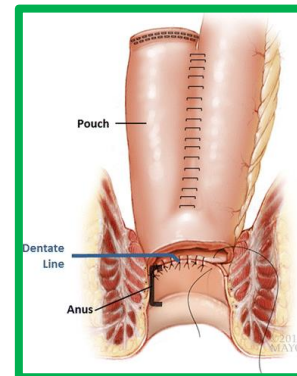
Heelkunde kan  
uitgesteld worden  
in ASYMPTOMATISCHE  
patiënten



- Beperkte hoeveelheid poliepen en
  1. Jonge vrouw met kinderwens
  2. Obese patiënten
  3. Hoog risico op desmoïd tumor







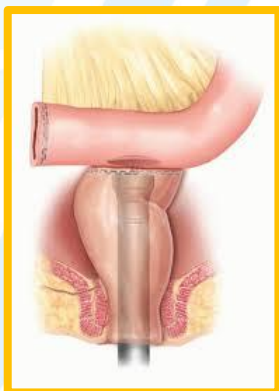
## Coloproctectomie + ileoanale pouch (IPAA)

Morbiditeit van de ingreep  
+  
Functionele Resultaten



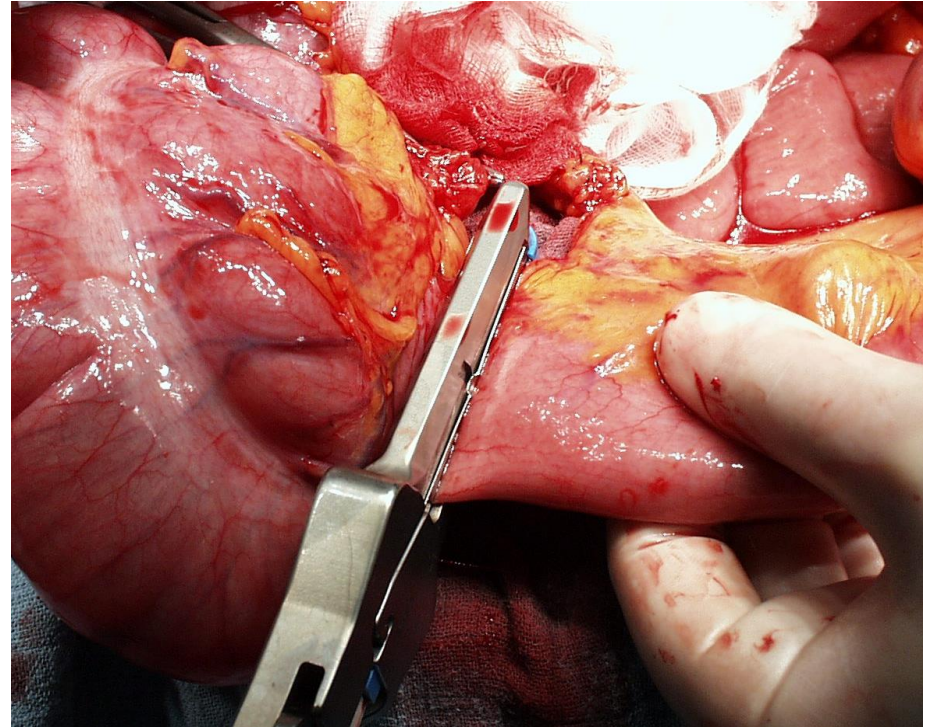
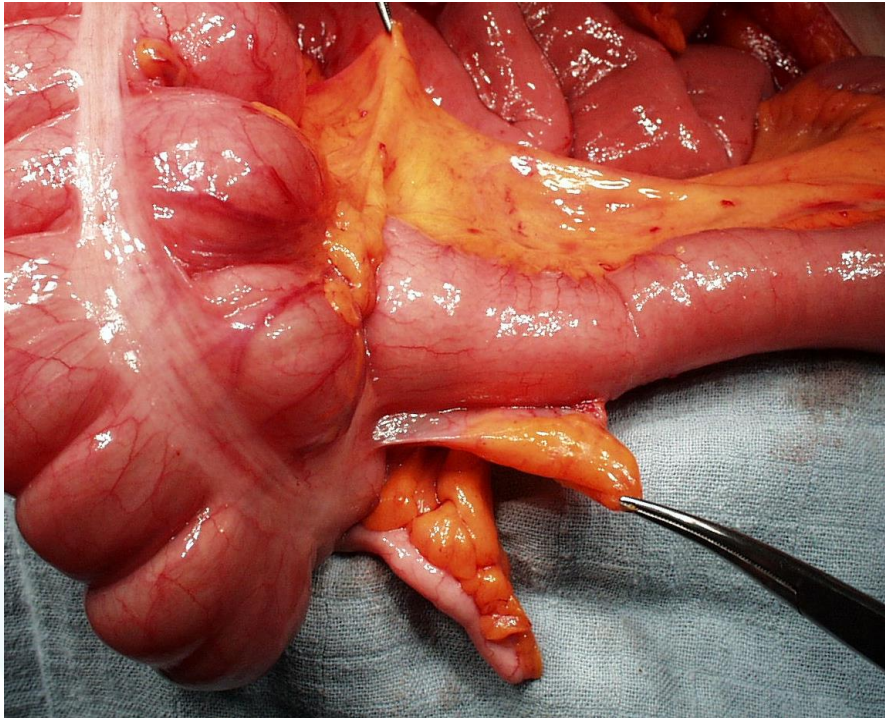
Preventie van kanker

**Totale colectomie + ileorectale anastomose (IRA)**

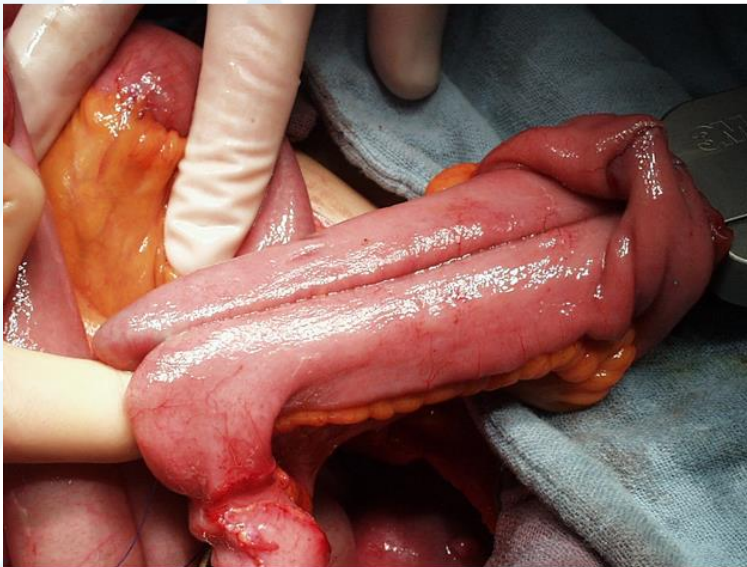
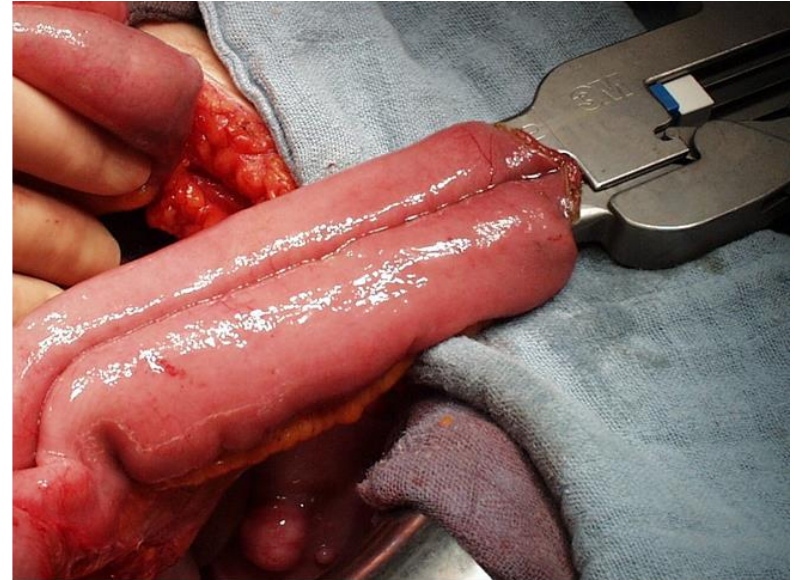
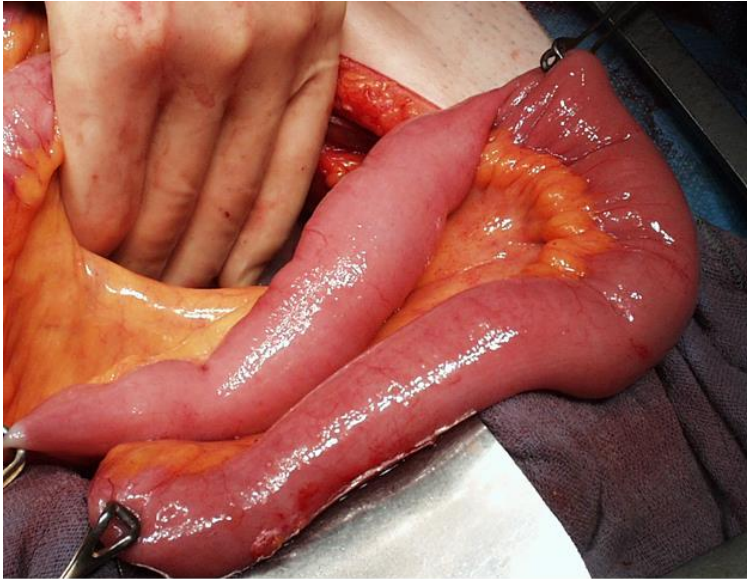


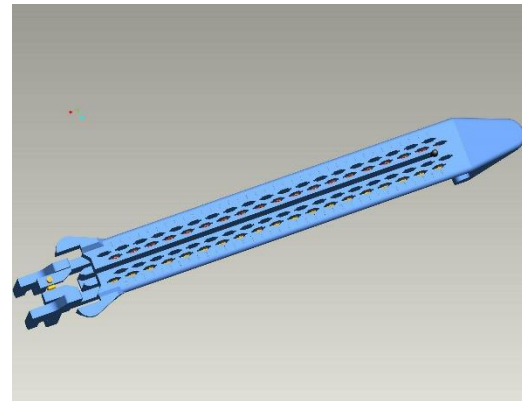
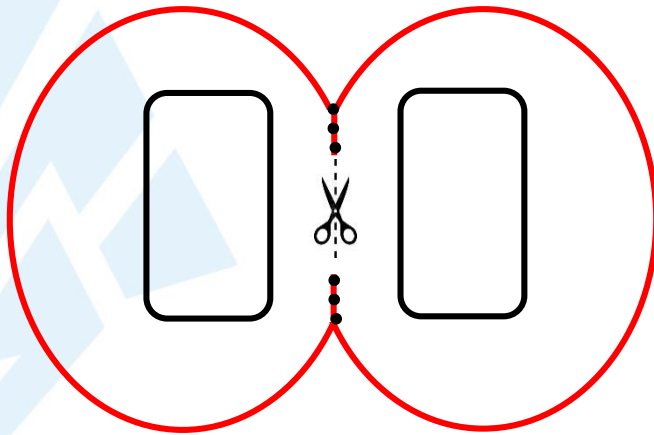
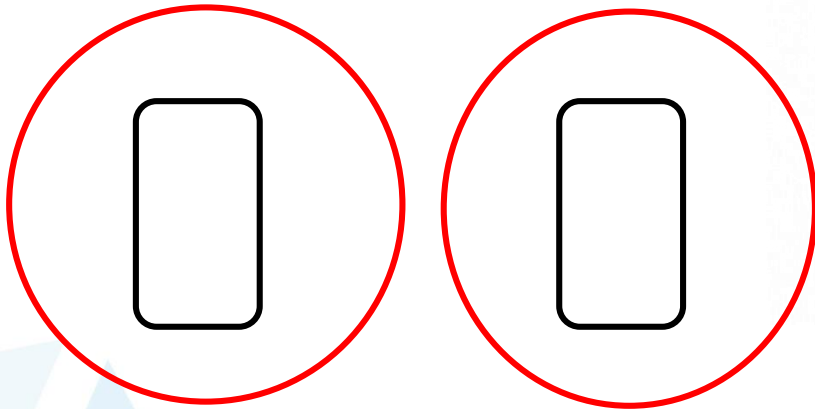
**Coloproctectomie + def ileostoma**

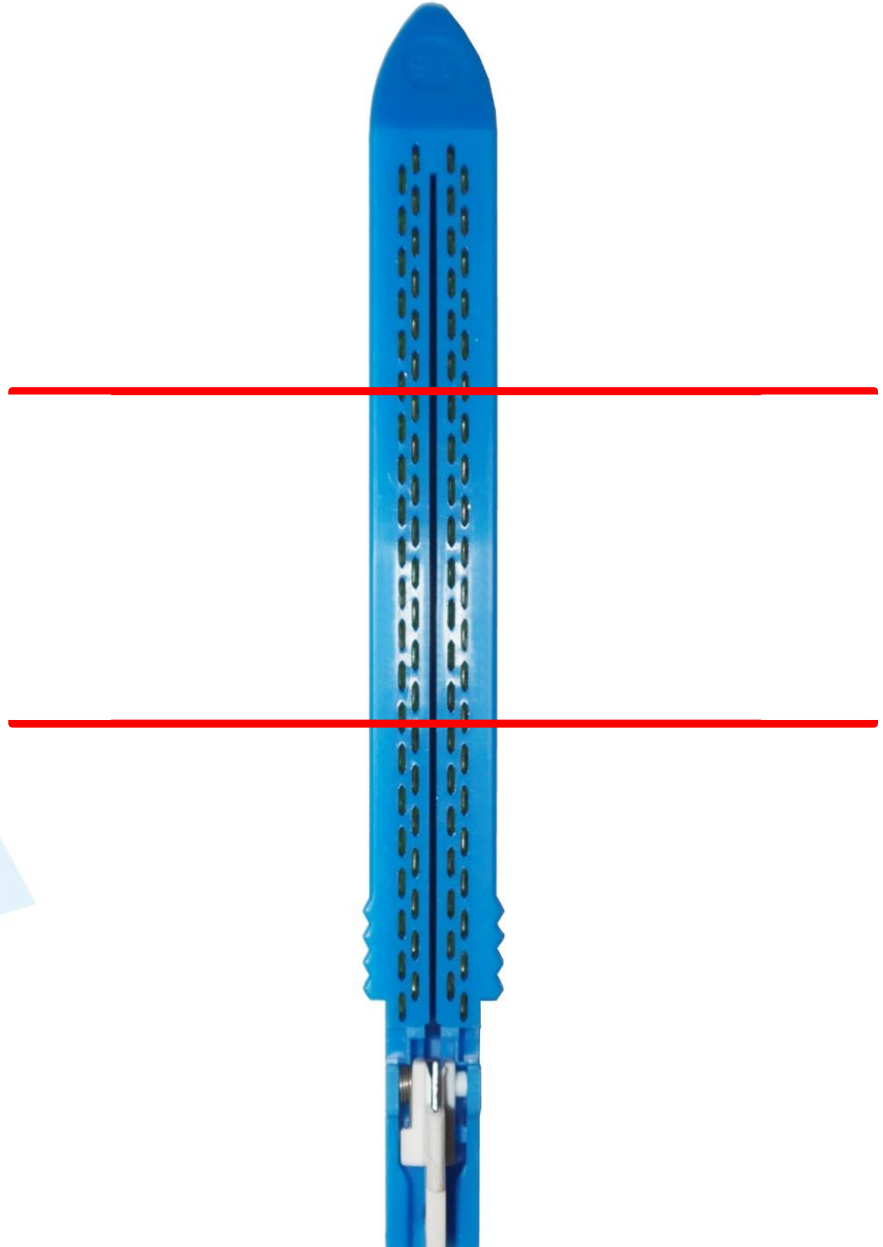


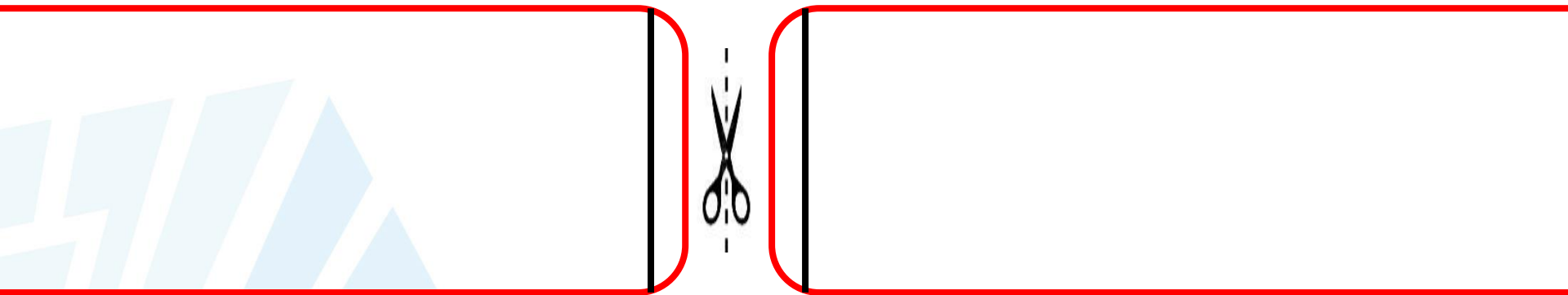




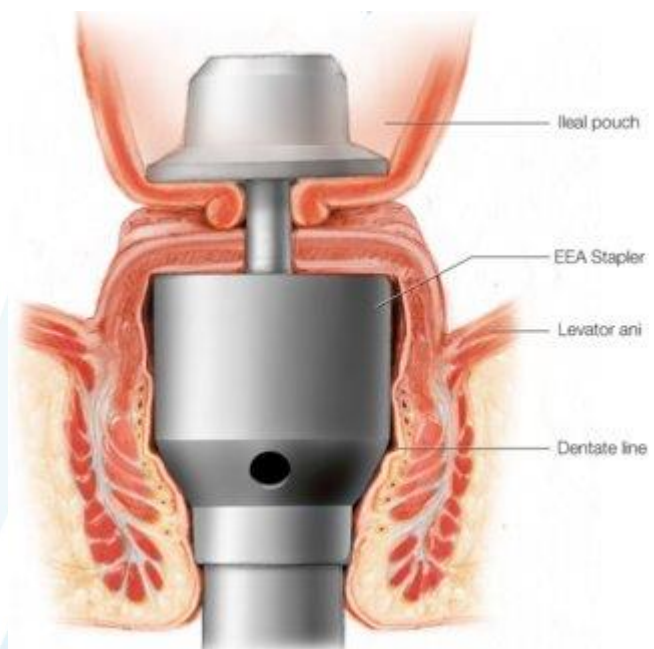




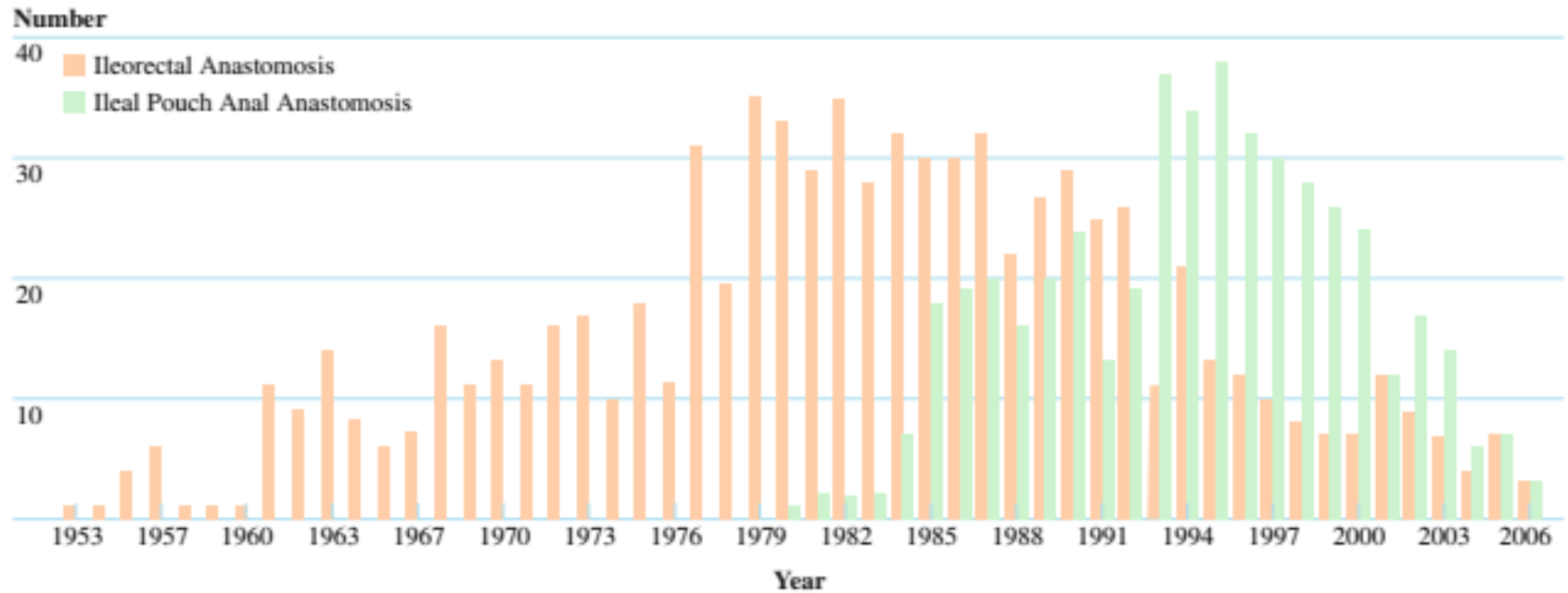




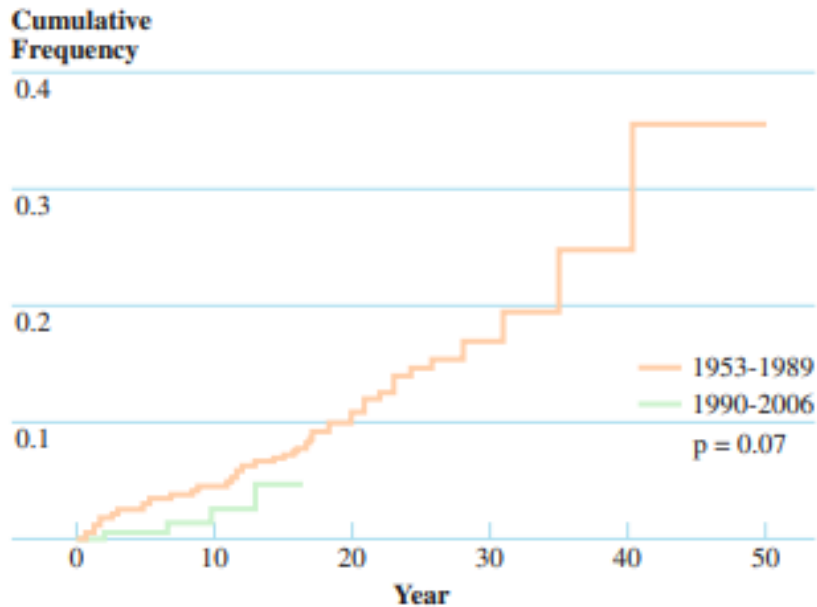




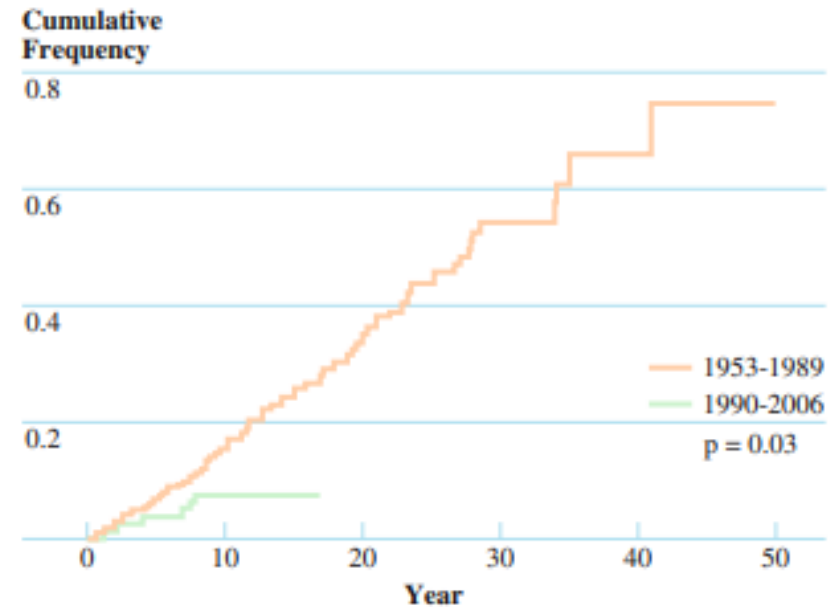
# Familiale Adenomateuze Polypose (FAP)



# Risico op kanker na IRA bij FAP



**FIGURE 2.** Cumulative frequency of rectal cancer after ileorectal anastomosis during the prepouch and pouch periods.



**FIGURE 4.** Cumulative frequency of proctectomy after ileorectal anastomosis in women during the prepouch and pouch periods.

# Familiale Adenomateuze Polypose (FAP)

## Rol van genetica binnen chirurgie

- **Secundaire proctectomie**
  - Mutatie analyse kan voorspellen of er een secundaire proctectomie zal nodig zijn (data na 20 jaar)
    - Attenuated genotype: 10%
    - Intermediate genotype: 39%
    - Severe genotype: 61%
- **Desmoiden**
  - APC mutatie tussen codon 1445 en 1578



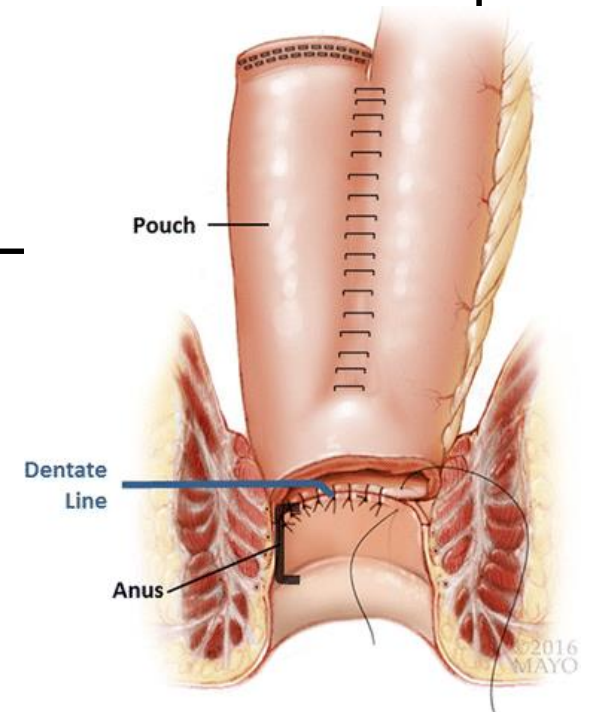
# Risico op Kanker

Review > Dis Colon Rectum. 2005 Sep;48(9):1708-13. doi: 10.1007/s10350-005-0057-1.

## Ileoanal pouch neoplasia in familial adenomatous polyposis: an underestimated threat

James Church<sup>1</sup>

- Risico op adenoma (poliepen) in de pouch → 50% op 10j
- Risk of carcinoma (kanker) 1,9% op 10j
- 25% in de pouch mucosa en 75% ATZ



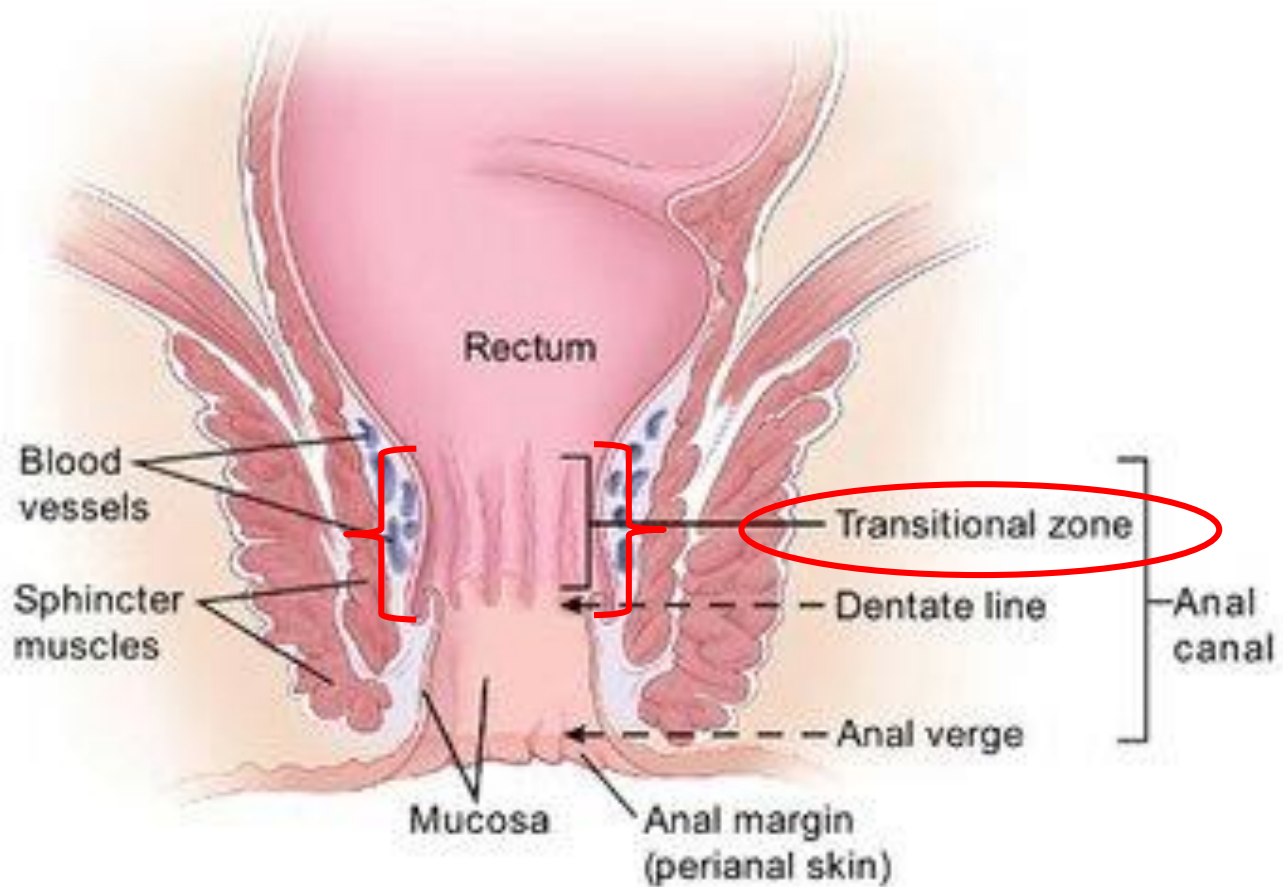
Von Roon AC, Ann Surg 2011

Van Duijvendijck P, J Gastroint Surg 1999

Friederich P, Clin Gastroenterol Hepat 2008

Smith JC, J Cancer Ther 2013

# Anale Transitie Zone - ATZ

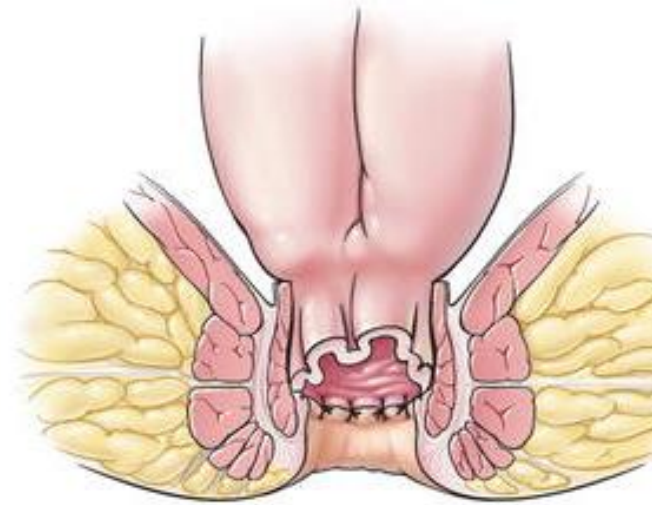


# Familiale Adenomateuze Polypose (FAP)

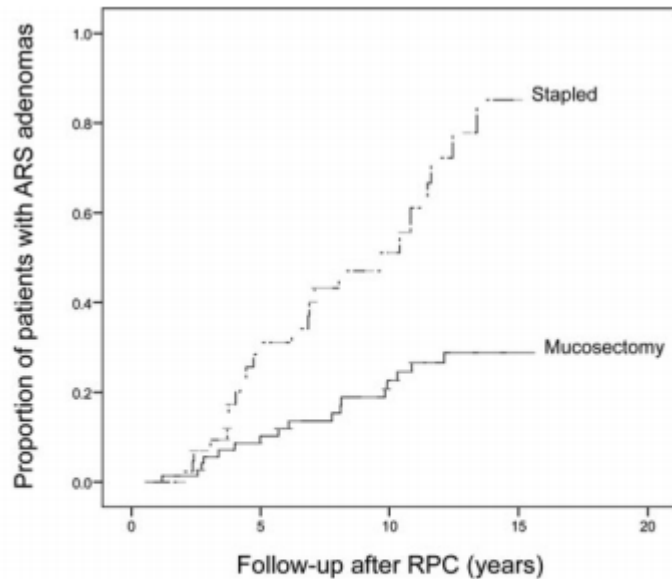
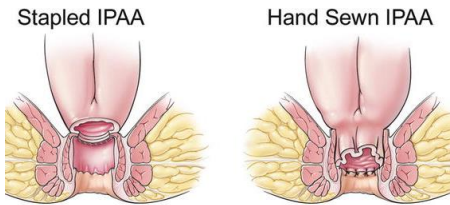
Stapled IPAA



Hand Sewn IPAA



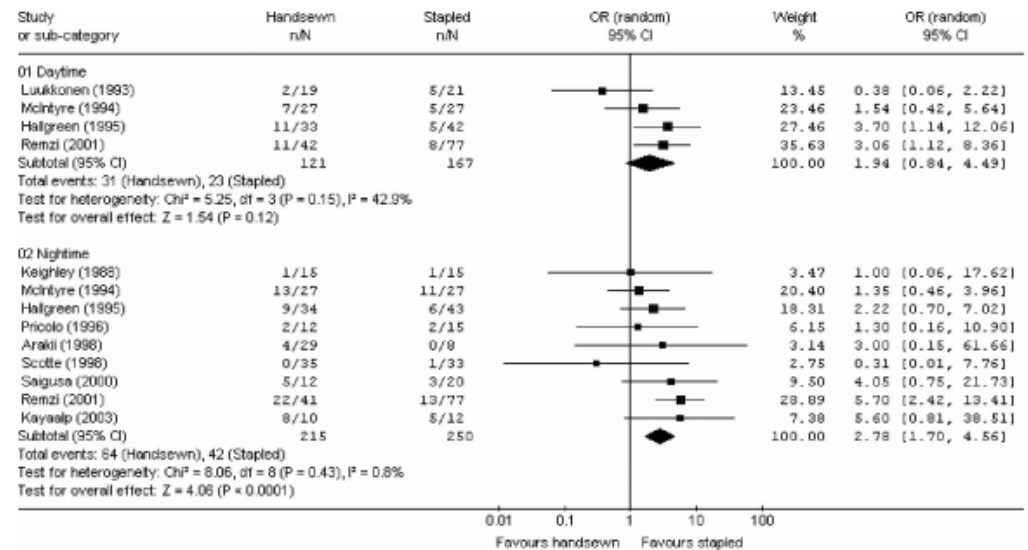
# Familiale Adenomateuze Polypose (FAP)



Number of at-risk individuals entering time interval:

	0-5	5-10	10-15	15-20
Stapled	44	24	12	2
Mucosectomy	76	55	41	23

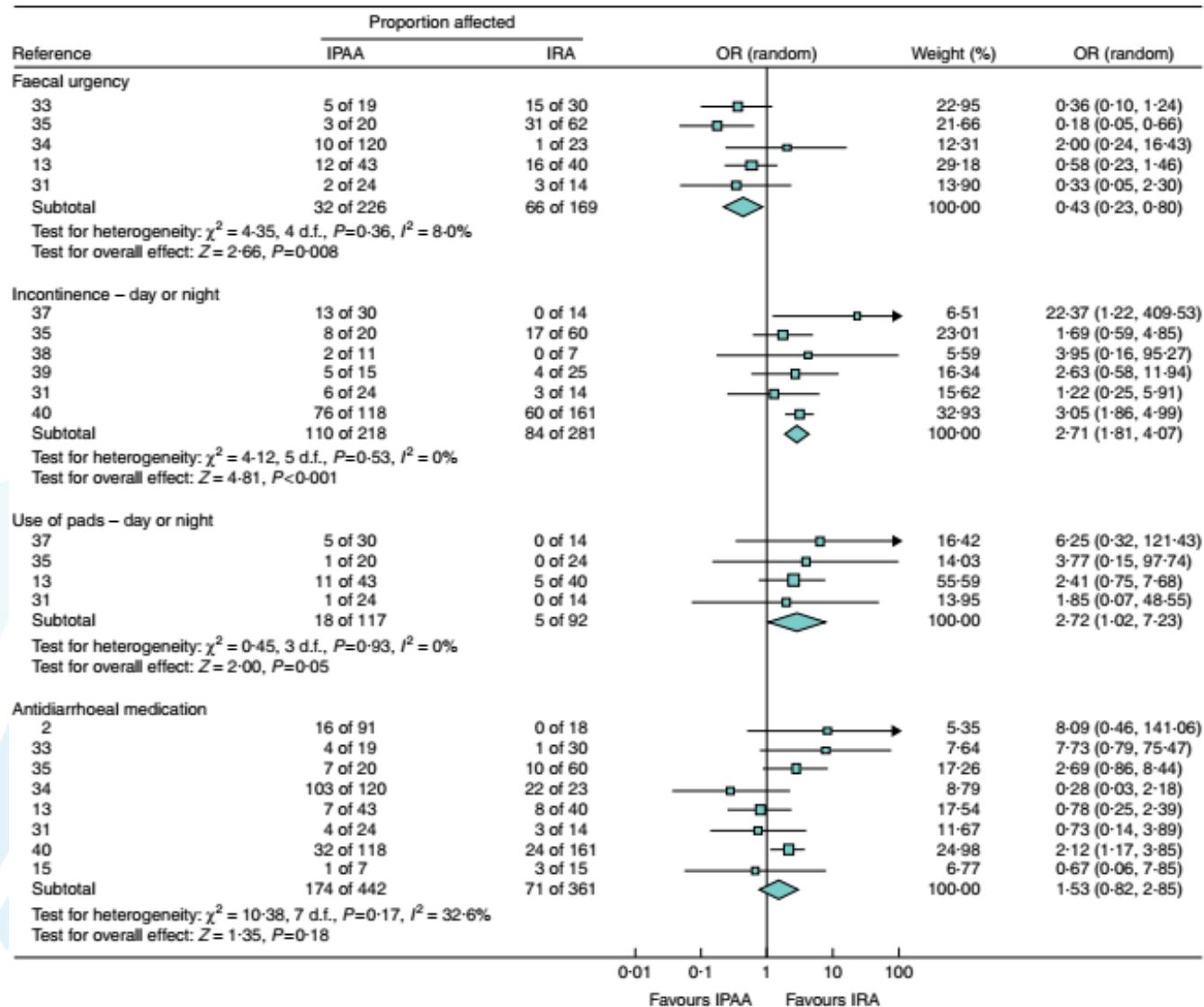
**FIGURE 1.** Cumulative risk of adenoma in the anorectal segment after restorative proctocolectomy for familial adenomatous polyposis.



**FIGURE 1.** Meta-analysis illustrating seepage of stool during the day and at night.



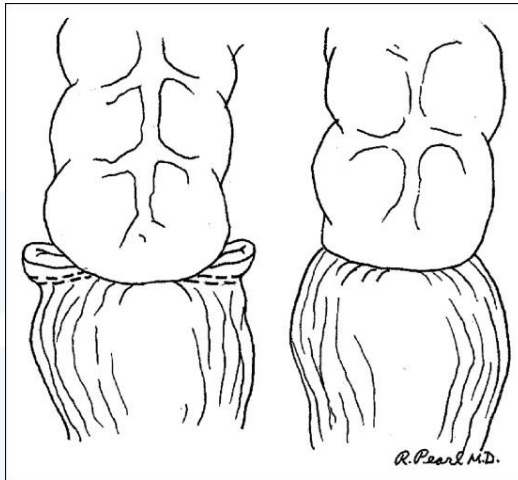
# Functionele resultaten IPAA vs IRA



# Double stapled vs TTSS

Increasing experience of modified two-stage transanal ileal pouch–anal anastomosis for therapy refractory ulcerative colitis. What have we learned? A retrospective analysis on 75 consecutive cases at a tertiary referral hospital

G. Bisleghi<sup>\*</sup>, B. Martin-Perez<sup>\*</sup>, S. Fieus<sup>†</sup>, A. Wolthuis<sup>\*</sup> and A. D'Hoore<sup>\*</sup>



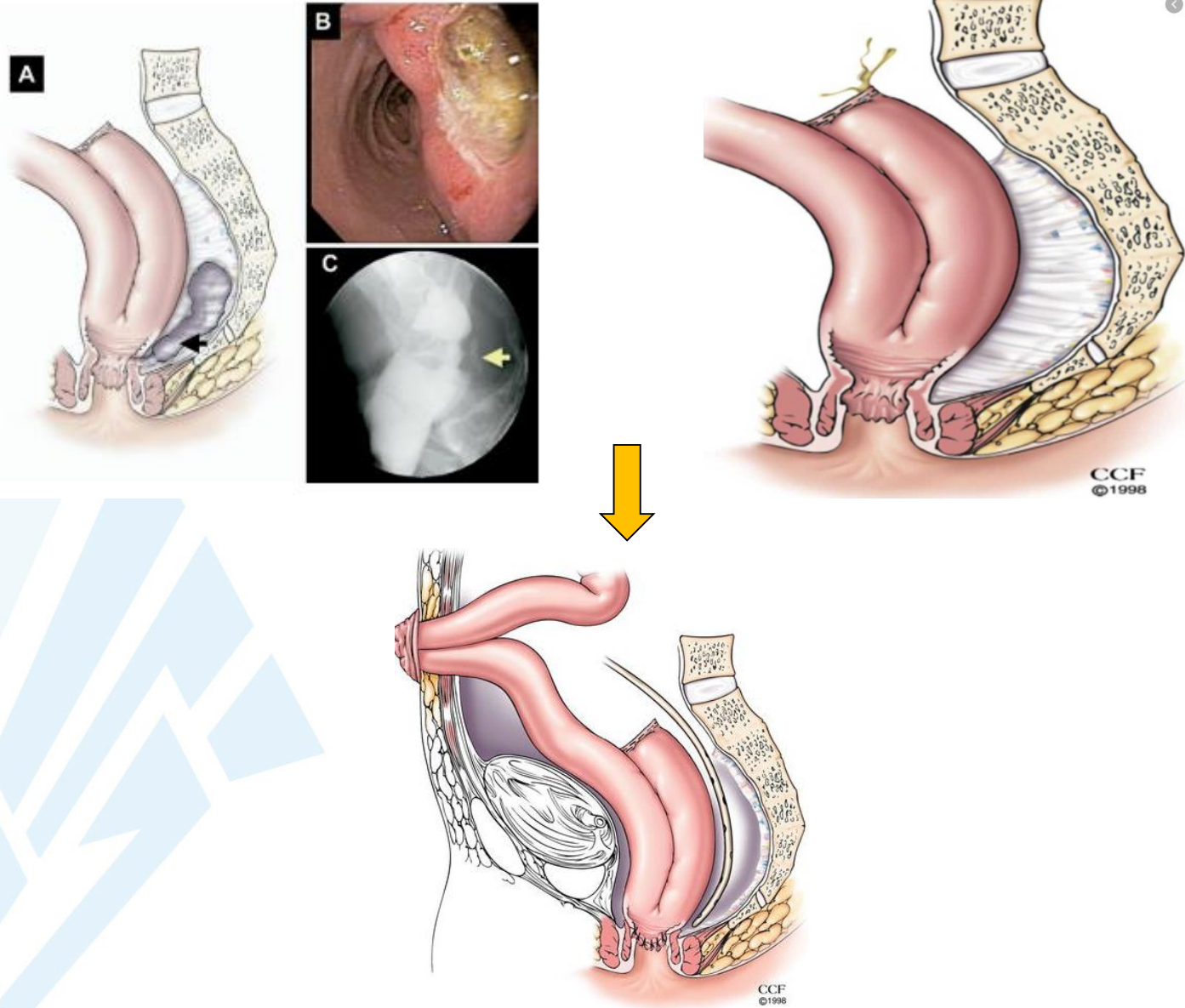
- ✓ Purse-string placement, rectal transection and bottom up TME



- ✓ Modified SINGLE STAPLED anastomosis



# Complicaties van IPAA



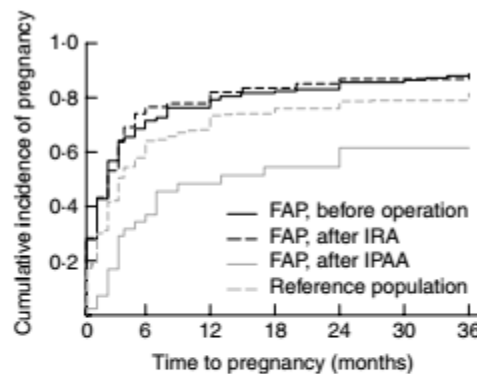
# Fertilität

Original article

## Female fecundity before and after operation for familial adenomatous polyposis

K. Ø. Olsen<sup>1</sup>, S. Juul<sup>2</sup>, S. Bülow<sup>3</sup>, H. J. Järvinen<sup>4</sup>, A. Bakka<sup>5</sup>, J. Björk<sup>6</sup>, T. Öresland<sup>7</sup> and S. Laurberg<sup>1</sup>

BJS, 2003



No. at risk				
FAP, before operation	171	39	25	14
FAP, after IRA	81	15	8	6
FAP, after IPAA	42	17	12	7
Reference population	914	202	108	72

**Fig. 1** Cumulative incidence of pregnancy within 3 years in patients with familial adenomatous polyposis (FAP), before operation, after ileorectal anastomosis (IRA) and after ileal pouch–anal anastomosis (IPAA), and in a reference population

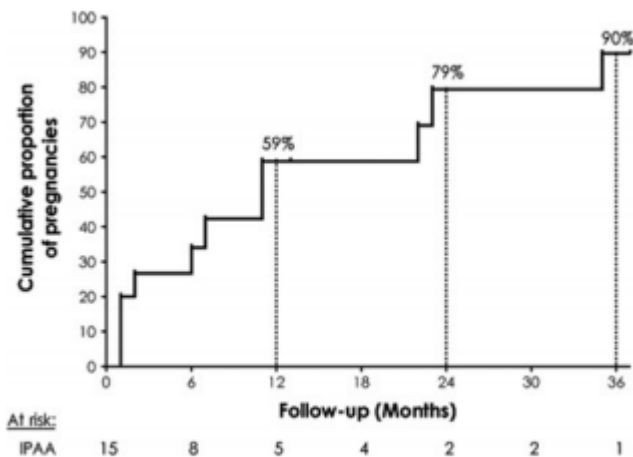
# Fertilität

## A Total Laparoscopic Approach Reduces the Infertility Rate After Ileal Pouch-Anal Anastomosis

### A 2-Center Study

*Laura Beyer-Berjot, MD,\*† Léon Maggiori, MD,\* David Birnbaum, MD,† Jérémie H. Lefevre, MD,\*  
Stéphane Berdah, MD, PhD,† and Yves Panis, MD, PhD\**

Ann Surgery, 2013



**FIGURE 2.** Time until pregnancy in 15 women attempting pregnancy after undergoing laparoscopic IPAA (Kaplan-Meier).



# Fertiliteit

## Female Fertility After Colorectal Surgery for Familial Adenomatous Polyposis

### *A Nationwide Cross-sectional Study*

Marry H. Nieuwenhuis, MD, MSc,\* Kirsten F. Douma, MSc, PhD,† Eveline M. Bleiker, PhD,‡  
Willem A. Bemelman, MD, PhD,‡ Neil K. Aaronson, PhD,† and Hans F. Vasen, MD, PhD\*§

Ann Surgery, 2010

**TABLE 2.** The Frequency of Fertility Problems in Relation to Surgery and Comorbidity and Mean Ages at Diagnosis of FAP and at Primary and Secondary Surgery (n = 138)

	With Fertility Problems (n = 23) n (%)	Without Fertility Problems (n = 115) n (%)	P		
Type of last surgery					
IRA	9 (39)	49 (43)	0.56		
IPAA	9 (39)	51 (44)			
Proctocolectomy and ileostomy	5 (22)	15 (13)			
Indication first operation					
Prophylaxis	22 (96)	110 (96)	1.00		
Cancer	1 (4)	5 (4)			
No. operations					
1	16 (70)	90 (78)	0.37		
>1	7 (30)	25 (22)			
Complications (adhesions, bleeding, abscess)					
Yes	12 (52)	44 (38)	0.22		
No	11 (48)	71 (62)			
Desmoid tumour					
Yes	4 (17)	14 (12)	0.50		
No	19 (83)	101 (88)			
Cancer (colorectal, thyroid, skin, cervix, non-Hodgkin lymphoma)					
Yes	3 (13)	13 (11)	0.73		
No	20 (87)	102 (89)			
Comorbidity (Cardiovascular or pulmonary diseases, cerebrovascular accidents, diabetes, osteoarthritis, kidney failure, malignant tumors, psychological complaints)					
Yes	12 (52)	62 (54)	0.81		
No	11 (48)	53 (46)			
	n	Mean (Range)	n	Mean (Range)	
Age at diagnosis of FAP	23	20 (6–36)	115	27 (9–58)	0.01*
Age at first operation	23	22 (10–36)	114	28 (10–59)	0.01*
Age at second operation	7	36 (26–54)	18	39 (17–59)	0.67

Pearson  $\chi^2$ . Fisher exact test was performed for Desmoid tumour and Cancer, because of small numbers.

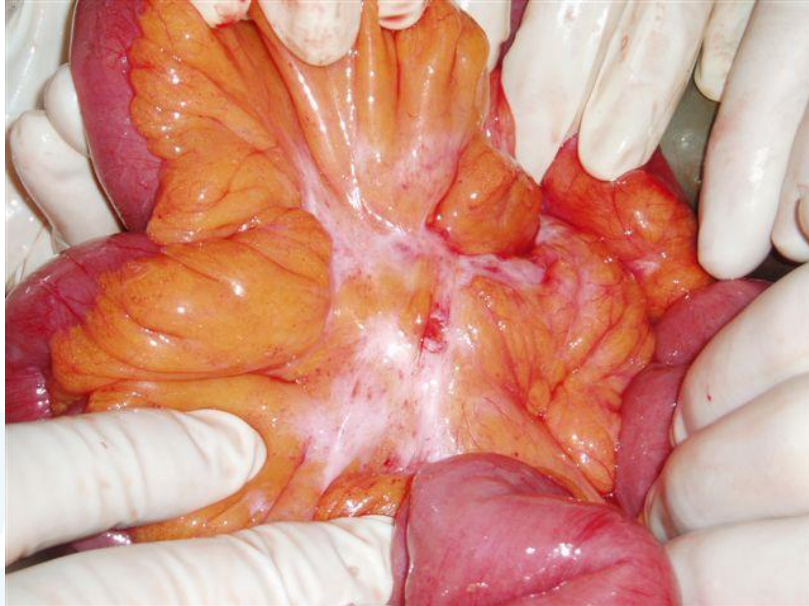
\*Significant at 0.01 level (2-tailed).

IRA indicates ileorectal anastomosis; IPAA, ileal pouch-anal anastomosis.

# Desmoïdtumor

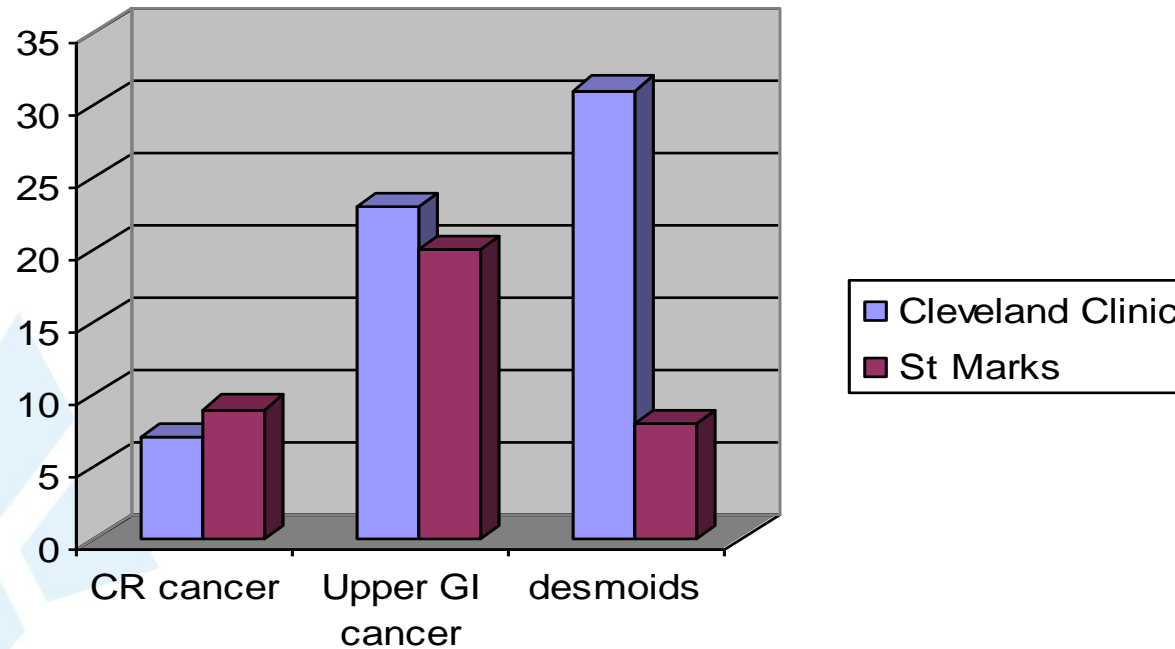
- Lokaal destructief - geen uitzaaiingen
- 15% van alle FAP patienten
- 50% intra-abdominale (mesenterium) en 40% buikwand
- Compressie of invasie van omliggende organen/structuren
  
- Erfelijk
- V>M
- Specifieke mutaties (codon 1399-1400)
- Heelkunde (chronische omsteking/trauma na chirurgie)
  
- Heelkunde op vroege leeftijd
- Open > laparoscopie
- Aard van heelkunde (CPR > totale colectomie+IRA)

# Desmoidtumor

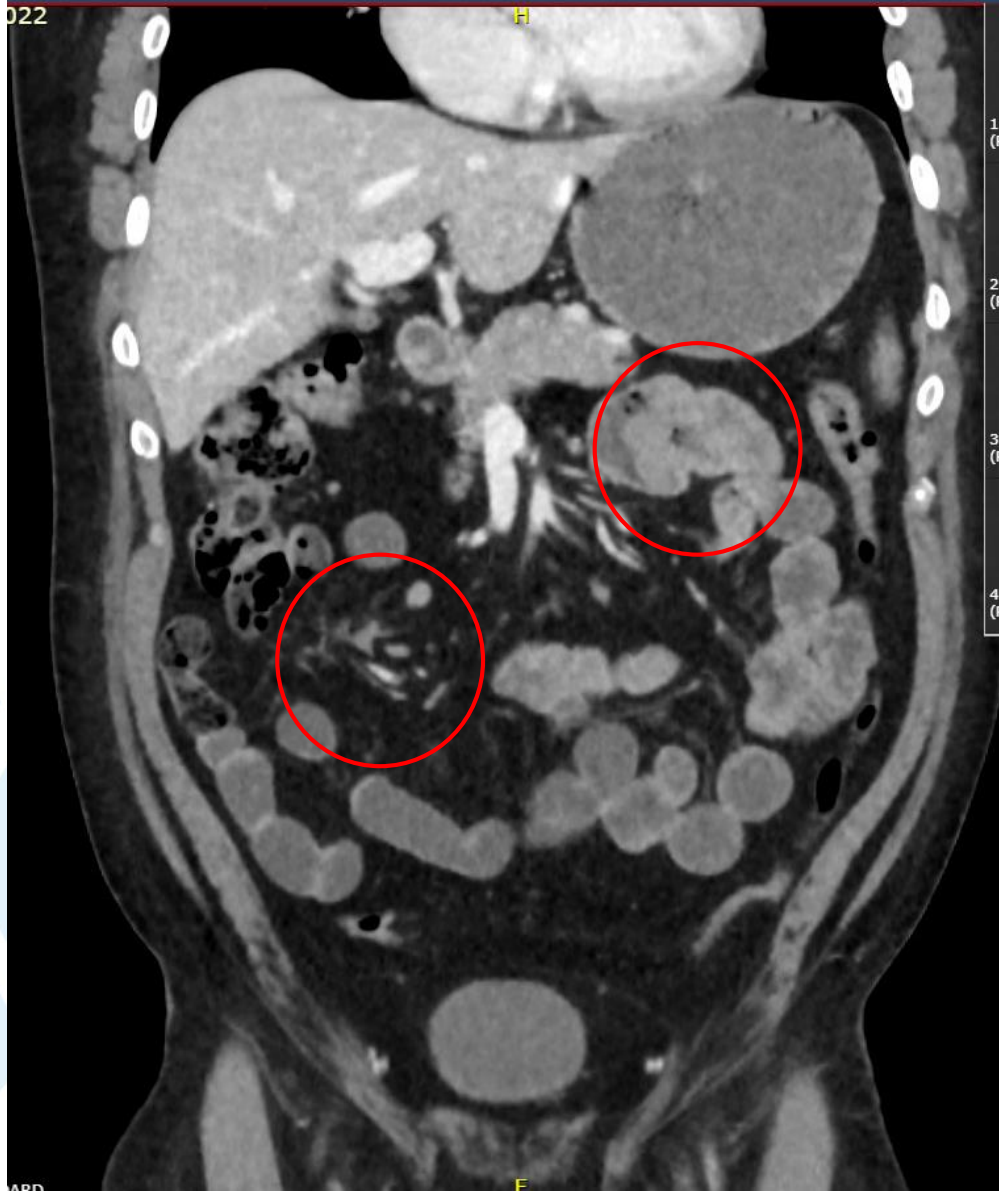


# Desmoidtumor

FAP-gerelateerde mortaliteit na profylactische colectomie



022



ARD

F





# Familiale Adenomateuze Polypose (FAP)



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*TOPIC HIGHLIGHT*

**WJG 20<sup>th</sup> Anniversary Special Issues (20): Gastrointestinal surgery**

## **Surgical treatment of familial adenomatous polyposis: Dilemmas and current recommendations**

Fábio Guilherme Campos



**Bedankt voor de aandacht**

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