FAMILIAL ADENOMATOSIS POLYPOSIS COLI

REVIEWED BY : Dr.YOGESH WAIKAR.

Inheritance: Autosomal dominant

AGE: 10 YEARS.

SIZE :< 1 CM

TYPE: ADENOMA.

Progression:carcinoma, 10 years. More in the rectal segment

Family h/o: 20% of patients with FAP have a negative **family** history

Family screen: testing at-risk children be delayed until age 10 to 12 years

If the gene test is negative, perform sigmoido-scopy after adolescence

Treatment OPTIONS:

1. proctocolectomy with ileal pouch-anal anastomosis: less bowel frequency, nocturnal defecation, and use of incontinence pads, but more fecal urgency
2. total proctocolectomy either with a conventional ileostomy
3. negative consent for above 2: subtotal colectomy with ileorectal anastomosis

ASSOCIATIONS:

Colonic adenomas (thousands)

Duodenal, periampullary adenomas: 60% to 90%

Gastric fundic gland Polyp: 0% to 100% of patients/non-neoplastic

Gastric adenoma: antrum.

Jejunal 40 % and ileal adenomas 20%

Ileal polyp: lymphoid.

Mandibular Osteomas 90%: no malignant potential.
Dental abnormalities

SYNDROMES:

Gardner:

Osteomas

CHRPE

Desmoid tumors: diffuse mesenteric fibromatosis: 4 to 32%

Epidermoid and sebaceous cysts

Fibromas, lipomas

Thyroid: papillary , adrenal tumors

Turcot :

Medulloblastoma

Glioblastoma multiforme

CHRPE

Attenuated FAP

Colonic adenomas (<100; proximal colon)

Duodenal, periampullary adenomas

Gastric POLYP: fundic gland

Familial tooth agenesis:

COLONIC POLYP: Hyperplastic

Bloom's syndrome:

Small stature

Facial erythema/telangiectasia

Male sterility

Adenocarcinomas, leukemia, lymphoma.

PEDGIHEP PROTOCOL:

PREOP INVESTIGATIONS:

1. **Colonoscopy**
2. **Upper Gi endoscopy: SOS BIOPSY OF POLYP**
3. **Enteroscopy.**
4. **XRAY SKULL AP /LATERAL:R/O OSTEOMA**
5. **SERUM ALPHAFETOPROTEIN.: R/O HEPATOBLASTOMA:>10-200NG/ML**
6. **SERUM CEA: INSITU CA.> 10-20 NG/ML**
7. **CBC/LFT/SR CREAT/PT/INR/HIV/HBSAG: R/O LEUEMIA**
8. **USG ABDOMEN:R/O HEPATOBLASTOMA OR DESMOID**
9. **THYROID USG:R/O PAPILLARY THYROID CA.**
10. **FUNDUS EXAM.: R/O CHRPE**
11. **TAKE B.P R/O ADRENAL.**
12. **CXRAY PA VIEW.**
13. **SIGMOIDOSCOPIC POLYPECTOMY AND HISTOPATH: PROVE ADENOMA/HAMARTOMA/JUVENILE**
14. **DERMAT REF SOS**
15. **DENTAL EVALUATION.**

Post op:

1. Day 1: AG MONITORING/VITALS /FEVER SPIKES/LOCAL WOUND EXAM/STOMA 0R STOOL OUTPUT
2. DAY 2 CBC/SERUM Na/SERUM K/SERUM CREAT/ AG MONITORING/VITALS /FEVER SPIKES/LOCAL WOUND EXAM/ STOMA 0R STOOL OUTPUT
3. DAY 3:START LIQUIDS /CLEAR FLUIDS/ AG MONITORING/VITALS /FEVER SPIKES/LOCAL WOUND EXAM/ STOMA 0R STOOL OUTPUT
4. DAY 4: START SEMISOLIDS/SERUM Na/SERUM K/SERUM CREAT/ AG MONITORING/VITALS /FEVER SPIKES/LOCAL WOUND EXAM/ STOMA 0R STOOL OUTPUT/SEMISOLID DIET/STOMA CARE/LOCAL WOUND CARE.
5. DAY 5: SOLIDS/ AG MONITORING/VITALS /FEVER SPIKES/LOCAL WOUND EXAM/ STOMA 0R STOOL OUTPUT/SEMISOLID DIET/STOMA CARE/LOCAL WOUND CARE.
6. DAY 6: TRAINING PARENTS / VITALS /FEVER SPIKES/LOCAL WOUND EXAM/ STOMA 0R STOOL OUTPUT/SEMISOLID DIET/STOMA CARE/LOCAL WOUND CARE.

7. DAY 7: DISCHARGE: SIMYL MCT/CA/VIT C/SOS SULDINAC/MVBC/PEPTAMEN15 KCAL =30ML/SALT/FATTY ENERGY DENSE FOODS.
8. AVOID :HIGH FIBER /FRUCTOSE/SORBITOL/COMPLEX CARB/AERATED DRINKS/

FOLLOW UP:

14 DAYS/1MONTH : REGULAR FOLLOW UP …3MONTHS : ANOSCOPY/POUCHOSCOPY.

EGD: 6 MONTHS.

1 YEAR: EGD with side-viewing endoscope.MUST START AT 20 YRS

periodic abdominal US/ thyroid examination MUST START AT 10 YEARS

head CT/MRI / hepatic US/serum α-fetoprotein for first decade of life.

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Spigelman Classification of Duodenal Polyposis (Adenomas in FAP)

	Number of Points		
	1P	**2P**	**3P**
Number of polyps	1–4	5–20	>20
Polyp size (mm)	1–4	5–10	>10
Histology	Tubulous	Tubulovillous	Villous
Dysplasia	Mild	Moderate	Severe
Stage	Spigelman score*		
0	0		
I	1–4		
III	5–6		
III	7–8		
IV	9–12		

Table 2 Proposed programme for surveillance and treatment of duodenal adenomatosis

Spigelman stage 0	Endoscopy* at intervals of 5 y
Spigelman stage I	Endoscopy† at intervals of 5 y
Spigelman stage II	Endoscopy† at intervals of 3 y
Spigelman stage III	Endoscopy† at intervals of 1–2 y
Spigelman stage IV	Endoscopic ultrasonography Consider pancreas sparing or pylorus sparing duodenectomy

*Including multiple random biopsies from mucosal folds in patients without visible polyps.
†Including multiple biopsies from polyps.