Surgical Challenges in Familial and Hereditary Colorectal Cancer

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Overview

- Definitions: Familial vs. Hereditary CRC
- Key syndromes: Lynch, FAP, MAP, FCC
- Surgical management challenges
- Decision-making frameworks

Defining Familial vs Hereditary CRC

- Hereditary CRC: Known gene mutations (e.g., Lynch, FAP, MAP)
- Familial CRC: Family history, no identifiable mutations (e.g., FCC Type X)
- Different implications for screening and surgery
- Genetic counseling essential

Lynch Syndrome

- MMR gene mutations (MLH1, MSH2, MSH6, PMS2)
- Lifetime CRC risk: up to 80%
- Right-sided colon predominance
- Risk of endometrial, ovarian, gastric cancers
- Colonoscopy every 1–2 years from age 20–25
- Consider extended colectomy

FAP

- APC gene mutation → hundreds to thousands of polyps
- CRC risk ~100% by age 40
- Surgery:
  Total proctocolectomy with IPAA
  Colectomy with IRA
  Desmoid tumor risk complicates decision-making

MUTYH-Associated Polyposis (MAP)

- Autosomal recessive, MUTYH bi-allelic mutations
- 10s–100s of polyps, CRC risk up to 80%
- Management like attenuated FAP
- Family screening is critical

Familial CRC Type X

- Meets Amsterdam criteria, no MMR defect
- Lower risk than Lynch
- Right-sided predominance
- No standard surgery guidelines
- Individualized approach

Surgical Challenges

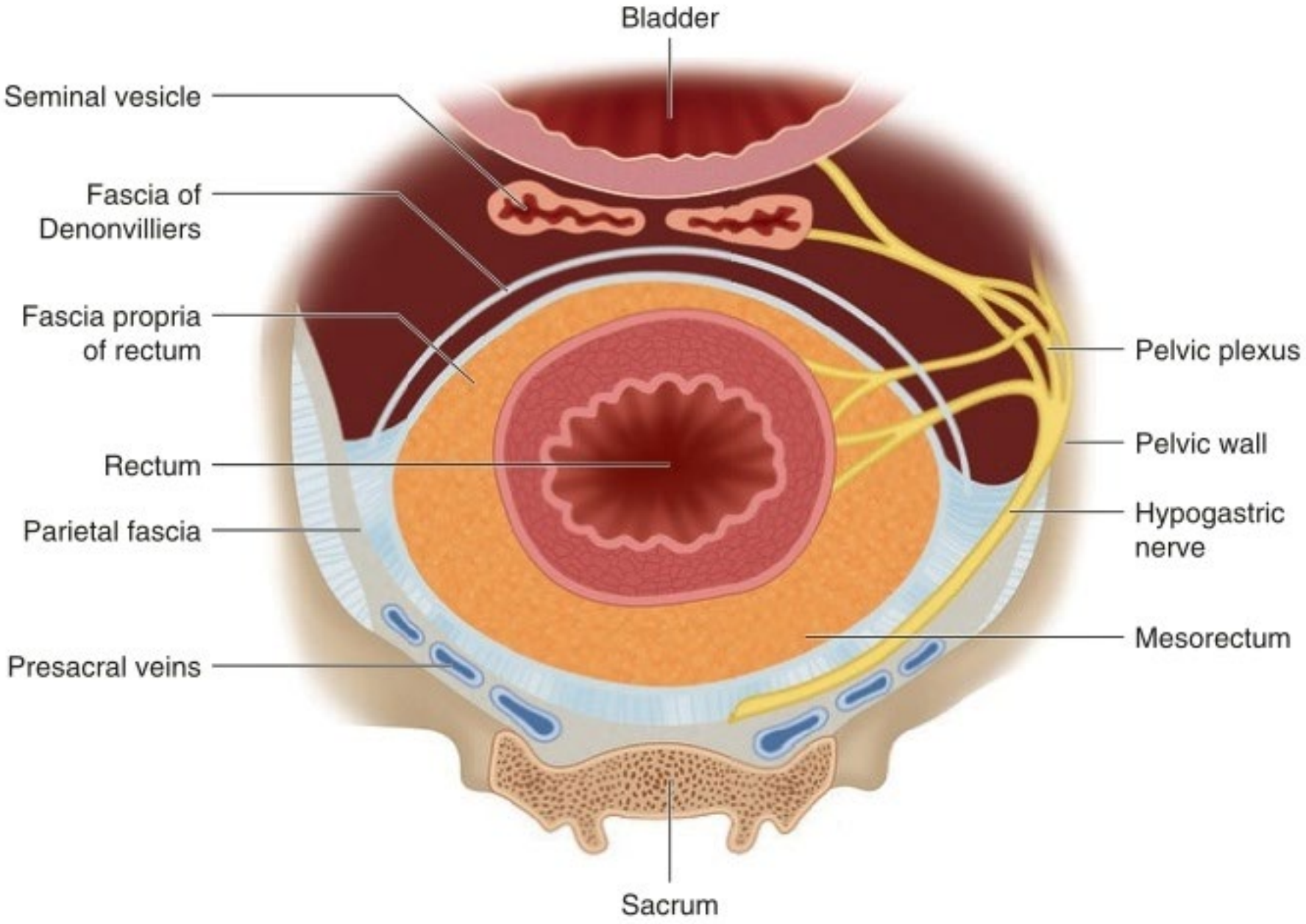
- When to perform prophylactic surgery
- Choosing segmental vs. total colectomy
- Rectal management
- Desmoid risks in FAP
- Quality of life, fertility, continence
- Long-term surveillance burden

The choice of surgery must balance risk reduction with patient-specific concerns. These include lifestyle, function, and tolerance of surveillance. Many patients prioritize bowel function over absolute cancer risk reduction.

Decision-Making Factors

- Mutation type and cancer phenotype
- Age, fertility, comorbidities
- Polyp burden and cancer stage
- Functional status and patient preference
- Psychosocial impact
- Multidisciplinary input

Surgical decisions should never be gene-based alone. Consider age, comorbidities, family plans, and social context. Multidisciplinary care is essential to align technical decisions with life goals.


Bladder
Seminal vesicle
Fascia of Denonvilliers
Fascia propria of rectum
Rectum
Parietal fascia
Presacral veins
Pelvic plexus
Pelvic wall
Hypogastric nerve
Mesorectum
Sacrum

Surgical Challenges in Elderly CRC Patients

- Physiology: Reduced reserve, cognitive issues
- Cancer biology: Slower growth, atypical presentation
- Surgical decisions: Avoid aggressive surgery if risk > benefit
- Geriatric input: Multidisciplinary approach essential
- Focus: Function, safety, life quality

Special Considerations

- Pregnancy
- 2nd trimester = safest for surgery
- Delay prophylactic procedures unless urgent
- Multidisciplinary

Intellectual Disability

- Use legal guardians/surrogates
- Avoid complex surveillance (favor total colectomy if needed)
- Ethics and simplified care models essential

Severe Comorbidities

- Use risk scores (ASA, CCI)
- Tailor surgery (e.g., local resection vs. full colectomy)
- Consider non-op if localized and stable
- Enhanced recovery protocols

Surgical Decision-Making Summary

- Risk stratify carefully
- Avoid overtreatment in elderly or comorbid
- Consider fertility and mental capacity
- Adapt surgery type to polyp burden and anatomy
- Use MDT + patient values = ideal care

References

- NCCN Guidelines 2024
- ESMO Hereditary CRC Guidelines 2023
- ASCRS Practice Parameters 2023
- Syngal et al., JAMA Oncol 2024
- Vasen et al., Gut 2020
- Church et al., DCR 2023