Serrated Polyposis Syndrome (SPS)

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Pathophysiology and Presentation

Historically Known as "Hyperplastic Polyposis Syndrome"

Genetic Mutations

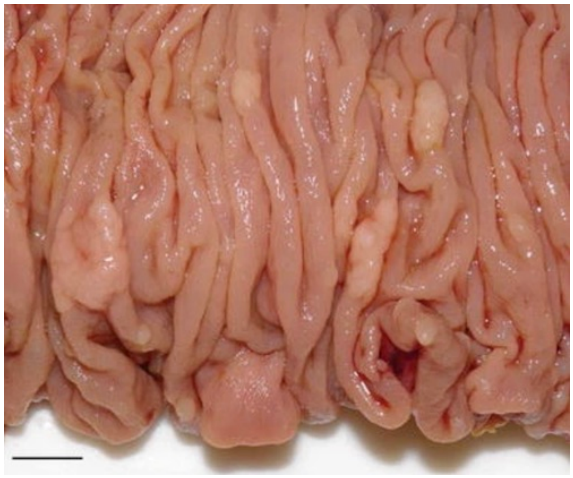
- Genetic Basis is Mostly Unknown [1]
- Mutations in RNF43 are Most Closely Related [2]
- Most Patients Have No Predisposing Mutation (97%) [2]

Colorectal Cancer

- Lifetime Risk: 25-70% [3-5]
- 5-Year Incidence: 1.5% [6]

Extracolonic Manifestations

- Overall Incidence: [6]
  - 54% Have a Family History of Extracolonic Malignancy
  - 24% Develop an Extracolonic Malignancy
- Exact Associations are Debated [7,8]



Colectomy Specimen with Multiple Sessile Polyps in a Patient with SPS [9]

Diagnosis and Management

Diagnosis (WHO 2019 Criteria) [10]

- Either:
  - ≥ 5 Serrated Lesions/Polyps Proximal to the Rectum
    - All ≥ 5 mm in Size
    - Two ≥ 10 mm in Size
  - > 20 Serrated Lesions/Polyps Throughout the Colon
    - ≥ 5 Polyps Proximal to the Rectum
- Polyp Count is Cumulative Over Multiple Colonoscopies [5]
- Criteria are Not Systematically Applied and the Diagnosis is Often Missed [11]
- *Historical 2010 WHO Criteria Also Included "Any Number of Serrated Polyps Proximal to the Sigmoid Colon with a Family History of SPS in a First-Degree Relative", Now Removed

Screening/Surveillance [1]

- Colonoscopy Every 1-3 Years
  - May Consider Shorter Intervals of 1-2 Years [12]
  - Starting Age Not Clearly Defined
- No Extracolonic Cancer Surveillance Indicated

Colectomy

- Indications for Colectomy: [1]
  - Unable to Control the Growth of Serrated Polyps
  - Development of Colorectal Cancer
- Prophylactic Colectomy Generally Not Indicated
- Generally Recommend Total Abdominal Colectomy with Ileorectal Anastomosis (TAC/IRA) [1]
  - May Consider Segmental Colectomy or Proctocolectomy if Indicated

Other Colorectal Cancer and Polyposis Syndromes

Syndromes

- *Familial Adenomatous Polyposis (FAP)*
- *Lynch Syndrome*
- *Juvenile Polyposis Syndrome (JPS)/Familial Juvenile Polyposis*
- *MUT Y Homolog (MUTYH)-Associated Polyposis (MAP)*
- *Peutz-Jeghers Syndrome (PJS)*
- *Serrated Polyposis Syndrome (SPS)*
- PTEN Hamartoma Tumor Syndromes: (PHTS)

Comparisons

- *See Familial Colorectal Cancer and Polyposis Syndromes

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