POLYP BURDEN IN CHILDREN WITH PEUTZ-JEGHERS SYNDROME: IMPACT OF SURVEILLANCE DOUBLE-BALLOON ENTEROSCOPY
A SINGLE CENTER LONGITUDINAL STUDY

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Peutz-Jeghers Syndrome

- autosomal dominant
- 1 in 200,000 live births
- germline mutation in the $STK11$ (LKB1) gene (19p13.3) in 94%
- 95% of affected individuals with mucocutaneous pigmented lesions
- Polyps with characteristic histology found throughout the GI tract and other hollow viscera
Intestinal obstruction

Intestinal resection

Intussusception

Small Intestinal Polyp Growth

Short Bowel Syndrome

‘Clean Sweep’ / enteroscopy – polypectomy

You YN et al. Fam cancer 2010

Oncel M et al. Colorectal dis. 2004

61%

4%
Small bowel surveillance in PJS

Start screening *no later* than 8 years of age

Surveillance
- EGD Colonoscopy
- +/- polypectomy

SBC / MRE → SB polyp burden

Significant polyp burden → Enteroscopy + Polypectomy

Repeat SBC define post-enteroscopy polyp burden

- Single / double balloon enteroscopy
- Laparoscopic assisted enteroscopy

Determine timing for repeat EGC / COL / SBC (~ 2 years)

Latchford A et al. JPGN 2019
Belsha D et al. JPGN 2017

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Aims:

• SBC findings in pediatric PJS
  • Small bowel polyp burden: size and number of polyps in pediatric PJS

• Impact of DBE in SB polyp burden in PJS – polyp burden & surgery
SBC Studies performed in children - young adults (<21 years of age) 2010 - 2019

‘Filtered for ‘Peutz Jeghers syndrome’ or ‘polyp’

Report review

patients DXd with PJS (standard clinical criteria)

Detailed chart review

SBC report abstracted

Study:
• Quality
• Completeness
• Polyp burden (number and size cat.)

1033

287

57

55 studies

17pts

• Double Balloon Enteroscopy
• Surgery
Methods II

- **Polyp size** defined as fraction of lumen occlusion by polyp tissue expressed as a percentage
- **Polyp number** was categorized in increments of 5 and 10.
- **Completeness of study** was defined as SBC reaching / not reaching the cecum
- **Quality** of study was categorized as satisfactory / poor
Results

• 17 patients
• 12 M
• Age at 1st SBC: 12.4 (SD 4.2) yrs
• 55 SBC studies (Median 2/ptn)

Presentation:
• 73% asymptomatic at SBC
• Abdominal pain (18%)
• GI he (14%)
Results: SBC – SI polyp burden pediatric PJS

Polyp size (luminal occlusion%) by symptom

Polyp number by symptoms

- symptomatic
- asymptomatic
Results: SBC studies ↔ DBE: study completion, quality

• 4 (7%) incomplete study – (OR of incomplete study post DBE 6.3 (p=NS) )

• SBC quality reported as satisfactory in 41 (75%) of studies

• Poor quality study reported in 5/9 studies if performed immediately post-procedure (OR of poor-quality study in DBE compared with non-DBE studies 5.13 (p=0.03))
Results: SBC studies ↔ DBE ↔ polyp burden

Pre-DBE polyp burden (largest polyp size - bubble size)

Post-DBE polyp burden (largest polyp size - bubble size)
Results: SBC studies ↔ DBE ↔ surgery

• 55 SBC studies:

  • 17 pre – (DBE) (1.7 / ptn x 10 ptns)
    → 7 Laparoscopic SI polyp resection (2010 - 8/2015)
    + (2) Expl. laparotomy (jejunal intussusception) + SB resection
    + (1) Laparoscopic adhesiolysis

  • 38 post – (DBE) (3.4 / ptn x 11ptns)
    → 9 DBE procedures (post 8/2015)
    → no surgery
Conclusions

• The ideal modality and conventions to study and report SB polyp burden in PJS is unclear

• (Pilot study) DBE is safe and useful in decreasing elective and emergent surgery in adolescent PJS

• Follow-up SBC quality poor if immediate post-procedure
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