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Familial adenomatous polyposis syndrome in children: Evidence based screening program and intervention

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Abstract

 ${f F}$ amilial adenomatous polyposis (FAP) is an autosomal dominant polyposis syndrome classically characterized by mutation in the APC gene on the long arm of chromosome 5. These mutations results in hundreds to thousands of adenomatous polyps in the colon and rectum early in life. Progression to colorectal cancer occurs by age 40-50 with rare malignancy occurring in teenagers. Thus, screening protocols have been developed to reduce the risk of colorectal and other associated malignancy in these people. Review of our history with children with FAP revealed approximately 30% of children had extraintestinal manifestations including papillary thyroid cancer and hepatoblastoma. The mean age of polyp detection was 12 years; however, 37% of our patients were younger than 10 years of age when polyps were discovered. Thus, we have recommended beginning screening at 7 years of age or at same age of other family members if polyps detected younger than 7 years of age. Once polyp burden is >30 or if symptoms develop, consideration for total proctocolectomy with ileal pouch anal anastomosis (IPAA) is advised. Our data shows that the mean age at IPAA is 15 years of age. Older children tend to undergo 2 stage procedures versus younger children tend to have more 1 stage procedures. Early postoperative complications occurred in 20% of patients with 10% requiring a reoperation. 20% of patients developed polyps in the rectal cuff, thus continued monitoring even after IPAA is indicated. 99% of patients have a functioning IPAA at the time of last follow up. Thus, we advocate for early, routine screening for polyps in children with FAP. Once the polyp burden is high, then IPAA is the procedure of choice.



Biography:

D Dean Potter Jr completed his MD at the University of Iowa College of Medicine. His General Surgery training was completed at Mayo Clinic Rochester, followed by a Pediatric



Surgery Fellowship at the University of Colorado Denver College of Medicine. He is the Chair of Pediatric Surgery at the Mayo Clinic Rochester. He has published 48 papers in peer reviewed journals and has given many presentations.

Speaker Publications:

1. "McKenna NP, Potter DD, Bews KA, Glasgow AE, Mathis KL, Habermann EB. Ileal-pouch anal anastomosis in pediatric NSQIP: Does a laparoscopic approach reduce complications and length of stay?" J Pediatr Surg. 2019 Jan; 54 (1):112-117 Epub 2018 Oct 05.

2. "Turner AE, Hollier LH Jr. Review of "Assessing the Safety of Overlapping Surgery at a Children's Hospital" by Hyder JA, Hanson KT, Storlie CB, Madde NR, Brown MJ, Kor DJ, Potter DD, Cirna RR, Habermann EB in Ann Surg 268: e24-e27; 2018. J Craniofac Surg. 2018 Dec 28.

3. "Lightner AL, Tse CS, Potter DD Jr, Moir C. Postoperative outcomes in vedolizumab-treated pediatric patients undergoing abdominal operations for inflammatory bowel disease. J Pediatr Surg. 2018 Sep; 53 (9):1706-1709 Epub 2017 Oct 09.

4. "Hyder JA, Hanson KT, Storlie CB, Madde NR, Brown MJ, Kor DJ, Potter DD, Cima RR, Habermann EB. Assessing the Safety of Overlapping Surgery at a Children's Hospital. Ann Surg 2018 Aug; 268 (2):e24-e27.

5. "Marek AP, Nygaard RM, Cohen EM, Polites SF, Sirany AE, Wildenberg SE, Elsbernd TA, Murphy S, Dean Potter D, Zielinski MD, Richardson CJ. Rural versus urban pediatric non-accidental trauma: different patients, similar outcomes. BMC Res Notes. 2018 Jul 28; 11 (1):519 Epub 2018 July 28.

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