

The role of wireless capsule endoscopy in gastrointestinal polyposis syndromes

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SUMMARY

Patients with hereditary gastrointestinal polyposis syndromes frequently develop polypoid lesions in the small bowel and are at increased risk for small bowel malignancy. In this setting small bowel surveillance is strongly recommended. Wireless Capsule Endoscopy is a new promising diagnostic method for the examination of the entire small bowel. It is non invasive, safe and can be repeated several times if necessary. Early experience from its use is promising. It has a high diagnostic yield for small bowel pathology and is clearly superior and safer compared to radiological methods. It tends to be the diagnostic method of first choice for the initial evaluation and surveillance in polyposis syndrome patients. Further investigation of the patients and therapeutic strategy can be tailored according to the results of video capsule examination.

Key words: Polyposis syndromes, Hereditary polyposis syndromes, Familial Polyposis Syndrome (FAP), Peutz – Jegher’s Syndrome, Wireless Capsule Endoscopy (WCE)

INTRODUCTION

Hereditary gastrointestinal polyposis syndromes (HPS) are characterized by the presence of numerous polyps, of various histology, distributed to one or more sites across the gastrointestinal (GI) tract. Sometimes they are combined with benign or malignant neoplasms of other organs (breast, uterus, ovaries, pancreas, CNS). HPS patients have a relatively high risk of small bowel (SB) malignancy (X

100-500 times) compared to the general population and need surveillance.

Surveillance is indicated in the following groups of patients:

- Familial Adenomatous Polyposis (FAP) and relative syndromes (Gardner, Turcot)
- Peutz Jegher’s syndrome (PJS)
- Familial Non Polyposis Colon Cancer (HNPCC)
- Familial Juvenile Polyposis Syndrome (FJP)

The risk for SB malignancy in these syndromes has been estimated as follows: for FAP relative risk (RR) is increased 100 times, and lifetime risk (LR) is 10%. For PJS RR is increased 500 times and LR is 13%, for HNPCC RR is X 100 and LR is 10%. Finally for FJP RR is increased 30 times and LR is that of the general population.

DIAGNOSTIC WORK UP AND SURVEILLANCE OF HPS PATIENTS

Small bowel has been characterized as the “black box” of the GI tract due to its inaccessibility. Diagnostic methods of the SB include:

Radiological: Small Bowel Follow Through (SBFT), Enteroclysis and recently CT/MRI Enteroclysis.

Endoscopic: Duodenoscopy (for and side viewing endoscope), Ileocolonoscopy, Push Enteroscopy, Double and Single Balloon Enteroscopy (DBE, SBE).¹⁻³

Wireless capsule endoscopy (WCE) is a new diagnostic method for SB evaluation. Although new, clinical experience from its use is large both worldwide as well as in our country. It is a friendly, safe test with high diagnostic sensitivity and accuracy for SB evaluation in various clinical settings (GI bleeding overt or occult with nega-

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tive upper and lower endoscopies, Crohn's and celiac disease, polyposis syndromes, etc).⁴⁻⁶ WCE in HPS is useful for identifying polyps in various sites of the SB and in surveillance of these patients. A report of WCE data in each syndrome will follow.

1. Familial Polyposis Syndrome (FAP)

It is characterized by numerous colonic adenomas which appear in early adolescence and the majority of patients develop colon cancer by the age of 30. Early surgical intervention is necessary. FAP patients have adenomas in the duodenum and in the stomach in 70% and 5-10% of the cases respectively. Duodenal distribution is usually around major papilla and the RR of cancer of the duodenum and the papilla is 300 and 120 times increased. Duodenal and papilla malignancies are the major causes of death in FAP patients who had a colectomy. Upper endoscopy (both for and side viewing) is the method of choice for duodenal evaluation and surveillance.⁷⁻¹²

The data for the presence of adenomas in other parts of the SB (Jejunum, ileum) before WCE was very poor. Radiological methods have limited diagnostic sensitivity for polyps, especially those under 0,5 cm in diameter. Enteroscopy data for this setting is very limited. Searching the data we found 8 studies in 143 FAP patients who had WCE. SB adenomas were present in 62/143 (43%). Figures were 30-60% in various studies. WCE was superior to radiological tests but inferior to enteroscopy in the part of the SB enteroscopy could check. In any case WCE was non invasive, friendly and safe for the patient. According to some studies the presence of adenomas in the duodenum is the only prognostic factor for adenomas in the jejunum and ileum. Major WCE drawbacks were the underestimation of number, overestimation of size, inaccuracy in identifying the exact anatomic location and low diagnostic accuracy for the duodenum and papilla area (30%).¹⁴⁻²¹

Conclusively SB evaluation in FAP patients is valuable due to the increased risk of malignancy. Duodenal evaluation includes endoscopy using both forward and side viewing instrument, as well as WCE to examine the rest of the duodenum, jejunum and ileum, especially in cases where duodenal adenomas are present. If WCE is positive the next step is enteroscopy and polypectomy. The time interval for this work up is not known at present.

2. Peutz Jegher's syndrome

Patients have numerous amartomatous polyps in the GI tract, and characteristic melanchrosis of the skin and lips. They have an increased risk of SB cancer (RR:X500, LR:13%), higher than any other HPS. Cancer usually de-

velops at the age of 40-50. Apart from the SB, patients have an increased risk for Ca of the colon, stomach, breast, pancreas, and gonads. Polyps are present in the SB, colon and stomach in 78%, 45% and 25% of the cases respectively. Whether malignant transformation is just from the amartoma or from the amartoma – adenoma – adenoCa sequence is unknown.²²⁻²⁴

The only method for SB surveillance of PJS patients was enteroclysis till recently. Enteroclysis has limited diagnostic value, is invasive, and exposes the patient to radiation. WCE is simple, non invasive, friendly and safe. It has a very good diagnostic accuracy and can be repeated several times without the disadvantage of additional radiation. There are 7 studies in 53 PJS patients in the literature. WCE is superior to all radiological tests (enteroclysis, CT/MRI enteroclysis). If WCE is positive (SB polyps) the next step is enteroscopy (DBE or surgical) and excision of at least the bigger lesions. If WCE is negative the test is repeated (unknown when).

Conclusively WCE is the test of choice for PJS patients for SB evaluation and surveillance. It is a highly diagnostic, non invasive, safe test and can be repeated several times. Disadvantages are the underestimation of number, overestimation of size and inaccuracy in the anatomic determination of lesions.^{15-21,25,26}

3. Hereditary Non Polyposis Colon Cancer (HNPCC)

Patients have an increased risk for malignancies: colon (80%), uterus (40-60%), and SB (4%). The relative risk for SB Ca is 100 times higher than the general population. Data for WCE is poor, only two references as case reports.^{27,28}

4. Familial juvenile polyposis syndrome (JPS)

Patients have a considerable amount of juvenile polyps in their colon. SB polyps are present in 16% of the cases. JPS has an increased risk for Ca of the colon, stomach, duodenum and papilla but not of the rest of the SB. There are no reports for WCE in this setting. Screening and surveillance of patients with WCE may not be needed as the risk of SB cancer is very low and does not differ from the general population.

CONCLUSIONS

Hereditary polyposis syndrome patients have an increased risk for small bowel malignancy and need to be under surveillance from early adulthood. The precise surveillance scheme has not been established. The increasing use of WCE and its growing data have established its

central role in SB investigation in HPS patients. WCE is effective, non invasive and safe and will change the work up for this group of patients. Future goals should include the more precise evaluation of size, morphology and anatomic distribution of lesions as well as the ability to obtain samples for histology.

REFERENCES

- Buckley JA, Fishman EK. CT evaluation of small bowel neoplasms: spectrum of disease. *Radiographics* 1998; 18:379-392.
- Debatin JF, Patak MA. MRI of the small and large bowel. *Eur Radiol* 1999; 9:1523-1534.
- North JH, Pack MS. Malignant tumors of the small intestine: a review of 114 cases. *Am Surg* 2000; 66:46-51.
- Rastogi A, Schoen RE, Slivka A. Diagnostic yield and clinical outcomes of capsule endoscopy. *Gastrointest Endosc* 2004; 60:959-964.
- Enns R, Go K, Chang H, et al. Capsule endoscopy: a single center experience with the first 226 capsules. *Can J Gastroenterol* 2004; 18:555-558.
- Pennazio M. Small bowel endoscopy. *Endoscopy* 2004; 36:32-41.
- Kurtz RC, Sternberg SS, Miller HH, DeCosse JJ. Upper gastrointestinal neoplasia in familial polyposis. *Dig Dis Sci* 1987; 32:459-465.
- Jagelman DG, DeCosse JJ, Bussey JJ. Upper gastrointestinal cancer in familial adenomatous polyposis. *Lancet* 1998; 1:1149-1151.
- Church J, McGannon E, Hull-Boiner S, et al. Gastroduodenal polyps in familial adenomatous polyposis. *Dis Colon rectum* 1992; 35:1170-1173.
- Matsumoto T, Iida M, Nakamura S, et al. Natural history of ampullary adenoma in familial adenomatous polyposis: reconfirmation of benign nature during extended surveillance. *Am J Gastroenterol* 2000; 95:1557-1562.
- Spigelman AD, Williams CB, Talbot IC, et al. Upper gastrointestinal cancer in patients with familial adenomatous polyposis. *Lancet* 1989; 11:783-785.
- Griffioen G, Bus PJ, Vasen HF, et al. Extracolonic manifestations of familial adenomatous polyposis: desmoids tumors and upper gastrointestinal adenomas and carcinomas. *Scand J Gastroenterol* 1999 suppl 1998; 225:85-91.
- Ianquinto G, Fornasarig M, Quaia M, et al. Capsule endoscopy is useful and safe for small bowel surveillance in familial adenomatous polyposis. *Gastrointest Endosc* 2008; 67:61-67.
- Wong RF, Tuteja AK, Haslem DS, et al. Video capsule endoscopy compared with standard endoscopy for the evaluation of small bowel polyps in persons with familial adenomatous polyposis. *Gastrointest Endosc* 2006; 64:530-537.
- Barkay O, Moshkowitz M, Fireman Z, et al. Initial experience of video capsule endoscopy for diagnosing small bowel tumors in patients with GI polyposis syndromes. *Gastrointest Endosc* 2005; 62:448-462.
- Burke CA, Santisi J, Church J, Levinthal G. The utility of capsule endoscopy in small bowel surveillance in patients with polyposis. *Am J Gastroenterol* 2005; 100:1498-1502.
- Mata A, Liach J, Castells A, et al. A prospective trial comparing wireless capsule endoscopy and barium contrast series for small bowel surveillance in hereditary GI polyposis syndromes. *Gastrointest Endosc* 2005; 61:721-725.
- Schulmann K, Hollerback S, Kraus K, et al. Feasibility and diagnostic utility of video capsule endoscopy in patients with hereditary polyposis syndromes. *Am J Gastroenterol* 2005; 100:27-37.
- Caspari R, vonFalkenhausen M, Krautmacher C, et al. Comparison of capsule endoscopy and magnetic resonance imaging for the detection of the small intestine in patients with familial adenomatous polyposis or with Peutz-Jeghers' syndrome. *Endoscopy* 2004; 36:1054-1059.
- Rossini FP, Risio M, Pennazio M. Small bowel tumors and polyposis syndromes. *Gastrointest Endosc Clin N Am* 1999; 9:93-114.
- Matsumoto T, Esaki M, Moriyama T, et al. Comparison of capsule endoscopy with the double balloon method in patients with obscure bleeding and polyposis. *Endoscopy* 2005; 37:827-832.
- Kitagawa S, Townsend BL, Hebert AA. Peutz-Jeghers syndrome. *Dermatol Clin* 1995; 13:127-133.
- Westerman AM, Wilson JH. Peutz-Jeghers syndrome: risks of a hereditary condition. *Scand J Gastroenterol Suppl* 1999; 230:64-70.
- Westerman AM, Gutius MM, debar E, et al. Peutz-Jeghers syndrome: 78 year follow up of the original family. *Lancet* 1999; 353:1211-1215.
- Pennazio M, Rossini FP. Small bowel polyps in Peutz-Jeghers syndrome: management by combined push enteroscopy and intraoperative enteroscopy. *Gastrointest Endosc* 2000; 51:304-308.
- Brown G, Fraser C, Scholfield G, et al. Video capsule endoscopy in Peutz-Jegheers syndrome: a blinded comparison with barium follow through for detection of small bowel polyps. *Endoscopy* 2006; 38:358-390.
- Marmo R, Rotandano G, Riccio G, et al. Small bowel adenocarcinoma diagnosed via capsule endoscopy in a patient found to have hereditary non polyposis colorectal cancer. *Gastrointest Endosc* 2007; 65:524=525/
- Yamagami H, Oshitani N, Kamata N, et al. duodenal cancer diagnosed by capsule endoscopy and double balloon enteroscopy in a patient with hereditary non polyposis colorectal cancer. *Digestion* 2006; 74:126-127.