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CLINICAL ARTICLE

# Availability and scope of integrated screening for patients with Lynch syndrome☆

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ABSTRACT

*Objective:* To assess the availability and capacity of US-based integrated centers for the management of Lynch syndrome. *Methods:* A cross-sectional survey of practice patterns in the care of patients with Lynch syndrome was conducted at 33 National Cancer Institute-designated cancer centers in the USA from March 1 to June 1, 2013. Each cancer center was contacted by telephone and the caller used a uniform scripted greeting and survey format. *Results:* All centers routinely recommended colonoscopy. Other recommended screening modalities were hysterectomy and bilateral salpingo-oophorectomy (29/33; 88%), endoscopy (27/33; 82%), urinalysis (23/33; 70%), endometrial sampling (21/33; 64%), dermatologic examination (19/32; 59%), pelvic ultrasonography (18/33; 55%), serum CA125 level (14/33; 42%), urine cytology (14/33; 42%), computed tomography (1/33; 3%), and magnetic resonance imaging (1/33; 3%). Each center had a multidisciplinary team but the composition varied. A designated team leader was present at 21 centers (64%). Having a team leader was associated with an increased likelihood of recommending endoscopy ($P = 0.04$) and dermatologic surveillance ($P = 0.01$). Only 23 centers (70%) had a system in place for communicating follow-up with patients. *Conclusion:* The lack of consensus in practice patterns recorded among participating centers probably reflected the limited existing evidence on the usefulness of most screening modalities.


## 1. Introduction

Lynch syndrome is an autosomal dominant disorder caused by germline mutations in genes encoding DNA mismatch repair proteins. The loss of these mismatch repair proteins results in unrepaired errors during DNA replication and a phenomenon known as microsatellite instability. Patients with Lynch syndrome have a substantially elevated risk of endometrial and colon cancer compared with the general population. Patients with Lynch syndrome also have an elevated risk of ovarian, stomach, hepatobiliary tract, pancreatic, small bowel, urinary tract, and central nervous system cancers [1]. Studies have shown that approximately 2% of all endometrial cancers and 3% of all colon cancers can be attributed to Lynch syndrome [1,2].

Prospective studies have indicated that cancer surveillance can help to reduce the morbidity and mortality of colorectal cancer among patients with Lynch syndrome [3,4]. In addition, a retrospective study of 315 women with Lynch syndrome found that performing prophylactic hysterectomy with bilateral salpingo-oophorectomy could prevent ovarian and endometrial cancer in this population [5]. Nevertheless, despite the documented benefits of screening and risk-reducing surgery, such approaches have not been fully integrated into the clinical management of families with Lynch syndrome. Indeed, some research suggests that the rates of inadequate compliance with cancer screening could be as high as 50% [3].

The underlying causes of poor screening compliance remain unclear; however, the lack of consensus screening guidelines and difficulty in accessing coordinated care are possible contributing factors. Numerous expert groups have published screening recommendations but these differ in their scope, frequency, and age of initiation for cancer screening [6]. Furthermore, although Lynch syndrome is included in medical education curricula, there remains a disproportionate emphasis on colorectal cancer and little attention is given to the other associated tumors [7]. In one study, only 36% of surveyed obstetrician gynecologists

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reported feeling comfortable counseling women with Lynch syndrome about endometrial cancer screening [8]. Healthcare providers have the ability to optimize adherence to surveillance and encourage patients to become involved in the management of their disease. However, there is evidence that healthcare systems might actually create barriers to screening through ineffective coordination of care, lack of continuity of care, inadequate access to specialty services, and disparate recommendations [9,10].

The aim of the present study was to evaluate the availability and scope of integrated centers for Lynch syndrome in the USA, with an emphasis on the homogeneity of screening recommendations and the methods used to deliver care.

## 2. Materials and methods

A cross-sectional study was conducted from March 1 to June 1, 2013, to assess practice patterns in the care of patients with Lynch syndrome at US-based cancer centers designated by the National Cancer Institute (NCI). Approval was obtained from the institutional review board of the New York Presbyterian Hospital-Weill Cornell Medical College, New York, NY, USA.

The present study targeted the NCI-designated cancer centers because these institutions are at the forefront of cancer care, and both patients and physicians often claim that they represent the current standard of care in the USA. The NCI website was searched on March 1, 2013, to extract the names and contact information of all NCI-designated cancer centers [11]. Each cancer center was then contacted by telephone using the number provided on the NCI website. The caller (S.J.P.) used a uniform scripted greeting, which stated that she was calling from New York Presbyterian Hospital-Weill Cornell Medical College as part of an approved study to evaluate the care available for patients with Lynch syndrome at NCI-designated cancer centers. Participation was voluntary and anonymous; no incentive was provided.

If a center agreed to participate, a set of predetermined questions was posed according to a uniform script: (1) "Do you recommend the following for patients with a known Lynch syndrome mutation: colonoscopy, endometrial sampling, pelvic ultrasonography, serum CA125 level, hysterectomy and bilateral salpingo-oophorectomy, urine cytology, urine analysis, endoscopy, dermatologic examination, computed tomography, or magnetic resonance imaging?"; (2) "Which services are involved in the care of a patient with Lynch syndrome?"; (3) "Is there a designated team leader or single service in charge of the care of patients with Lynch syndrome. If so, who is this leader?"; and (4) "Is there a system in place to keep track of screening and results for patients with Lynch syndrome? If so, who is responsible and how are patients reminded to follow-up with recommended screening?"

The data were analyzed using SAS version 9.3 (SAS Institute, Cary, NC, USA). Bivariate analyses were conducted to determine whether having a designated team leader and the number of services involved in care increased the likelihood of recommendation of individual screening modalities and decreased the likelihood of patients being responsible for their own follow-up. Associations between categorical variables were evaluated by $\chi^2$ or Fisher exact tests as appropriate for the category size. Prevalence ratios and 95% confidence intervals were also calculated for each outcome. Owing to the small sample size and the distribution of the number of services involved and the number of screenings recommended, these variables were dichotomized. The number of services involved was dichotomized into less than and greater than or equal to the median number of services involved. The number of screenings was dichotomized into less than or equal to and greater than the median number of recommended screenings. The acceptable $\alpha$ error level was set at a $P$ value of 0.05 using two-tailed tests.

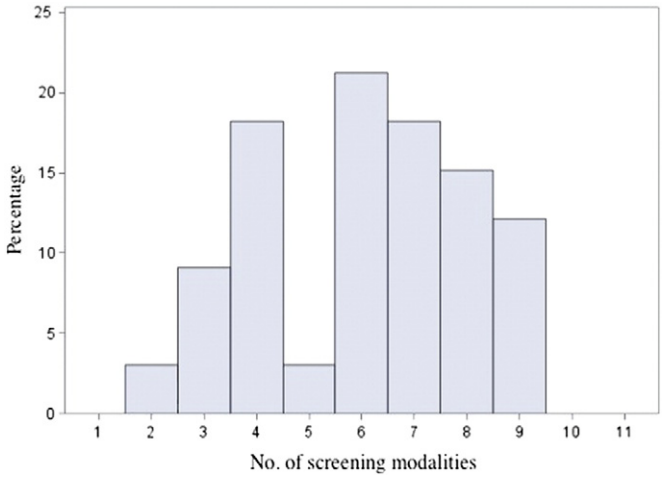


**Fig. 1.** The number of screening modalities used by the 33 National Cancer Institute-designated cancer centers for patients with Lynch syndrome.

## 3. Results

The search of the NCI website identified 60 NCI-designated cancer centers. All of these centers were contacted and 33 (55%) agreed to participate in the present study.

The participating centers recommended a median of six screening and/or risk-reducing interventions (range 2–9) (Fig. 1). All centers routinely recommended colonoscopy for patients; however, recommendation of the other interventions varied widely (Table 1). In all, 29 (88%) centers recommended risk-reducing hysterectomy and bilateral salpingo-oophorectomy at the completion of child-bearing. All interventions were statistically significantly less likely to be recommended than colonoscopy, with the exception of hysterectomy ($P = 0.11$).

Each center had a multidisciplinary team for the management of patients with Lynch syndrome; however, the composition of individual teams differed (Fig. 2). All 33 participating centers included representatives from the departments of gynecologic oncology, gastroenterology, and genetics. Other services included were general surgery and/or colorectal surgery (n = 28; 85%); social work and dermatology (n = 25; 76%); psychiatry (n = 20; 61%); medical oncology (n = 7; 21%); reproductive endocrinology (n = 3; 9%); primary-care physicians and chaplain services (n = 2; 6%); and pathology, a dietician, and a sexual health counselor (n = 1; 3%). The median number of services involved was 6 (range 2–11) (Fig. 3).

The presence of a designated team leader was hypothesized to be associated with more thorough and organized screening systems than

**Table 1**
Frequency of screening and risk-reducing surgery recommendations at 33 National Cancer Institute-designated cancer centers compared to colonoscopy.[a]

| Measure | Recommended | Not Recommended | $P$ value[b] |
|---|---|---|---|
| Colonoscopy | 33 (100) | 0 | Reference |
| Endometrial sampling | 21 (64) | 12 (36) | <0.001 |
| Pelvic ultrasonography | 18 (55) | 15 (45) | <0.001 |
| Serum CA125 level | 14 (42) | 19 (58) | <0.001 |
| Hysterectomy and bilateral salpingo-oophorectomy | 29 (88) | 4 (12) | 0.11 |
| Urine cytology | 14 (42) | 19 (58) | <0.001 |
| Urine analysis | 23 (70) | 10 (30) | <0.001 |
| Endoscopy | 27 (82) | 6 (18) | 0.02 |
| Dermatologic examination[c] | 19 (59) | 13 (41) | <0.001 |
| Computed tomography | 1 (3) | 32 (97) | <0.001 |
| Magnetic resonance imaging | 1 (3) | 32 (97) | <0.001 |

[a] Values given as number (percentage).
[b] Fisher exact test.
[c] Data available for 32 centers.