Surveillance recommendations for patients with Lynch syndrome and FAP : a monocentric study

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Abstract

***Background and study aims* : The most important causes of hereditary colorectal cancer are Lynch syndrome (LS) and the adenomatous polyposis syndromes (familial adenomatous polyposis syndrome or FAP, attenuated FAP or AFAP and MUTYH associated polyposis syndrome or MAP). The aim of this study was to investigate whether all patients with a hereditary syndrome within one center receive uniform advice regarding surveillance and treatment.**

***Patients and methods* : A retrospective analysis was performed of all electronic patient health records of patients with LS, FAP, AFAP and MAP who received genetic counselling or were followed by a health care specialist at the University Hospital in Ghent.**

***Results* : Data from 122 patients were collected. For all patients, recommendations from the medical genetics department were highly consistent. Adherence to their recommendations was good within the center for the management of colon polyps. There was a lack of consistency in the screening and surveillance advice for other tumors in departments other than gastroenterology. Only 33 patients had systematic follow-up consultations to check results and organize surveillance.**

***Conclusion* : Previously, small studies have suggested that patients with hereditary gastrointestinal cancer syndromes infrequently have surveillance as specified in the guidelines. This study shows almost uniform recommendations and good adherence for surveillance of the colon, but incomplete or contradictory advice for surveillance of other organs. The need for an integrated approach from a multidisciplinary team will only increase in the future, because more families with hereditary cancer are likely to be found due to the increased use of next generation sequencing in cancer diagnostics.** (Acta gastroenterol. belg., **2020**, 83, **399-405**).

Key words : Lynch syndrome, FAP, surveillance.

Introduction

Colorectal cancer (CRC) is a frequent and potentially lethal disease. Most tumors are sporadic, but approximately 20 to 30% of patients with CRC have a family history of the disease. In 5 to 6% a germline mutation, diagnostic for a known hereditary cancer syndrome, can be identified (1,2). The most common syndromes are Lynch Syndrome (LS) (also called hereditary non-polyposis colorectal cancer or HNPCC) and the familial adenomatous polyposis syndromes.

LS is an autosomal dominant inherited disorder caused by a mutation in one of the mismatch repair (MMR: MLH1, MSH2, MSH6, PMS2) genes, leading to tumor DNA microsatellite instability (MSI). Patients with LS have an estimated lifetime cumulative incidence of 80% for CRC and 60% for endometrial cancer (EC). In addition, there is also an increased risk for other tumors such as stomach, small intestine, pancreas, bile duct, ovary, urinary tract and brain cancer. Unfortunately, this syndrome is often under-diagnosed, leading to significant morbidity and mortality (3).

The familial adenomatous polyposis syndromes consist of three hereditary syndromes in which the inheritance of a germline mutation causes accelerated colorectal carcinogenesis, which is manifested by the development of multiple adenomas at a young age with the potential for early development of CRC. We distinguish familial adenomatous polyposis syndrome (FAP), attenuated familial adenomatous polyposis syndrome (AFAP) and MUTYH-associated polyposis syndrome (MAP). FAP and AFAP have an autosomal dominant inheritance pattern. These disorders are often caused by a mutation in the APC gene (4). FAP is defined by the presence of more than 100 synchronous colorectal adenomas, whereas in AFAP there are typically fewer than 100 adenomas. It is characterized by a later onset of the disease and adenomas are often located more proximally. In addition to the increased risk for CRC, there is also an increased incidence of duodenal and ampullary tumors, thyroid cancer, hepatoblastoma and desmoid tumors.

MAP is caused by a biallelic mutation of the MUTYH gene and is inherited autosomal-recessively. The condition is characterized by the presence of multiple adenomatous polyps, often 20 to 99, similar to AFAP (4,5).

Patients with a hereditary gastrointestinal cancer syndrome are recommended to undergo surveillance in order to detect premalignant lesions and small tumors, thereby decreasing morbidity and mortality of associated cancers. Small studies have already suggested that many individuals do not have surveillance as specified in the guidelines and it has also been described that patients often receive different advice from their various health care providers (6,7). It is unclear whether inadequate surveillance is the result of patient non-compliance or rather incorrect recommendations from physicians.

The purpose of this monocentric study was to investigate whether all patients with a hereditary syndrome

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receive uniform advice regarding surveillance and treatment, or whether there are differences in the recommendations given by the various sub-disciplines where they are followed up.

Patients and Methods

This was a retrospective, single-center study regarding patients with a confirmed diagnosis of LS, (A)FAP or MAP, carried out at a tertiary hospital (Ghent University Hospital).

Data were collected from the database of the medical genetics department. A list of all patients who had undergone genetic screening for LS, (A)FAP or MAP during the period from 01/01/2007 up to 31/12/2016 was obtained. Four hundred and seventy patients with a confirmed genetic mutation (out of 4631 screening episodes) were identified. A review of the electronic patient files was performed and only patients that had received genetic counselling or were followed by a health care specialist within the university center were included in the study.

79 patients with LS, 35 patients with FAP, 7 with AFAP and 1 with MAP were included in the study. Some patients were index patients, others had been screened because of a known familial hereditary cancer syndrome. An overview of the departments where patients received follow-up and the recommendations for individual patients given regarding surveillance, prevention and treatment of tumors, was made. The number of patients who were lost-to-follow-up was also recorded. A summary of the number of patients, the reason for their diagnosis and the involved disciplines is shown in table 1.

This study was approved by the Ethics Committee of Ghent University hospital.

Results

Lynch syndrome

A total of 79 patients (40 female; 39 male) who received genetic counseling and/or sought specialist advice regarding surveillance following the diagnosis of LS were identified.

Advice in the department of medical genetics

In 73 out of 79 patients (37 female, 25 male), written advice from the medical genetics department could be found. For surveillance of the colon, a colonoscopy was recommended starting from the age of 20-25 years, every 1-2 years. In 2 out of 10 (20%) patients with an MSH6 mutation, it was specifically mentioned that surveillance could be postponed until the age of 30, as CRC occurs later in life than in other MMR gene mutations. In 20 of 73 patients (27%) it was stated that a total colectomy is preferable compared to a partial colectomy in the case of development of a CRC.

Screening for gynecological tumors was advised by means of a gynecological examination with transvaginal ultrasound and determination of CA 125, starting from the age of 30-35 years, to be repeated every 1-2 years. In only 11 of the 37 female patients (30%), an annual endometrial sampling was recommended. A prophylactic hysterectomy with bilateral salpingo-oophorectomy at the completion of child-bearing was recommended to 6 patients with an MLH1 mutation and 1 patient with a MSH6 mutation (7/37, 19%) with the specific advice that the risk of gynecological tumors is strongly increased in women with these mutations. No advice was given to the remaining 30 patients, but 7 (23%) of them had already undergone a prophylactic hysterectomy.

Surveillance for gastric tumors was recommended by means of an esophagogastroduodenoscopy (EGD) every 1-2 years, starting from the age of 30-35 years.

For the surveillance of urinary tract tumors, it was recommended to perform an abdominal ultrasound and a urinalysis, starting from the age of 30-35 years, every 1-2 years. To 1 patient, only annual urinalysis was recommended.

Two patients with a family history of brain tumors received advice to screen by means of a neurological examination and, optionally, an MRI of the brain. Finally, an annual dermatological check-up was recommended to 2 patients. One patient received the advice to seek optimal sun protection.

Advice in other departments

Data on follow-up was retrieved for 35 patients: 20 patients were seen exclusively in Ghent and 15 patients were also partially followed in an external center. For 27 patients at least one contact at the gastroenterology department was recorded. Five patients were seen at the surgical department and 21 patients had a contact in one of the following departments : gynecology, neurology,

Table 1. — **Overview of the patients followed in Ghent University Hospital**

	Total number of patients	Reason for diagnosis	Follow-up at the surgery department	Follow-up at the gastro-enterology department	Follow-up at other disciplines	Advice from the genetics department
LS	79	22 index patients, 21 with cancer, 3 metastatic 49 with known family history 8 without information	5	27	21	73
FAP	35	3 index patients, 1 with metastatic cancer 16 with known family history	14	13	12	27
AFAP/ MAP	8	2 index patients, both with cancer, 1 metastatic 6 without information	2	1	0	8

Table 2. — **Recommendations from the gastroenterology department regarding surveillance of patients with LS**

Exam	Number	Frequency and age of onset
Colonoscopy	21/21 (100%)	Every 1-2 years (n=10) Every year (n=10) Every 2 years (n=1) Age of onset: 20-25 years
Gynecological examination - Transvaginal ultrasound - CA125 - Endometrial sampling	8/11 (73%) 7 7 1	 Every 1-2 years (n=7) Every year (n=1) Age of onset: 30-35 years
EGD	21/21 (100%)	Every 1-2 years (n=10) Every year (n=10) Every 2 years (n=1) Age of onset: 30-35 years
Urinary tract - Ultrasound - Urine sediment - Urine cytology	19/21 (90%) 15 5 12	 Every 1-2 years (n=11) Every year (n=8) Age of onset: 30-35 years
Other recommendations: - Dermatologic examination - Neurologic examination	 2/21 (9.5%) 2/21 (9.5%)	 Every year No information

urology, dermatology, oncology, endocrinology or radiotherapy.

The advice given at the gastroenterology department is shown in table 2. Written advice on surveillance was found in 21 out of 27 patients (78%). Of the remaining 6 patients, 3 were only seen at the endoscopy department, with no further advice being prescribed. Two patients were only seen after surgery for a CRC, setting the indication for chemotherapy, after which they returned to an external center. One patient died before formal advice could be given.

Five patients with a history of CRC were followed up at the surgical department after resection of their primary tumor. In 3 patients, no advice for further surveillance was written down, but 1 of them had a metastatic tumor. The 2 remaining patients were advised to perform an annual colonoscopy. Gynecological examination and an EGD was recommended to the one female patient. No advice was given regarding surveillance of urinary tract tumors.

Twenty-one patients were also regularly seen by health care professionals of other disciplines. The majority of these patients only received advice about surveillance within the practitioners discipline and the recommendations were variable (see table 3).

Follow-up

Out of 35 patients with at least some record of follow-up, 30 patients were followed at the gastroenterology and / or surgery department. Of these 30 patients, only 15 (50%) had an annual or 2- yearly appointment in the hospital, where all results and necessary examinations were discussed and planned. Four patients were only seen at the endoscopy department during their follow-up, two of whom also had surveillance of the other organ systems. In total, 6 patients (20%) were lost to follow-up.

FAP, AFAP and MAP

A total of 43 patients with adenomatous polyposis syndromes were identified: 35 patients with FAP, 7 with AFAP and 1 with MAP.

Advice in the department of medical genetics

Thirty-five patients received genetic counseling regarding surveillance and treatment (27 patients with FAP, all with AFAP and MAP).

In patients with an APC mutation, an annual colonoscopy was recommended, starting from the age of 10-12 years. Only 1 (3.7%) of the 27 FAP patients was advised to start with a flexible sigmoidoscopy and only to proceed to a total colonoscopy if polyps were detected. In 5 patients with AFAP a total colonoscopy was recommended starting from the age of 10-12 years, the other 2 were recommended to start at 18-20 years. To the patient with MAP, a total colonoscopy was recommended starting from the age of 25-30 years, every 2-3 years. Twenty-six of the 27 (96%) FAP patients were advised to undergo a prophylactic colectomy with ileoanal pouch

Table 3. — **Recommendations from various departments regarding surveillance of patients with LS**

Discipline	Number	Recommendations
Neurology	2	- Annual neurological examination and MRI of the brain (n=1) - 2-yearly MRI of the brain (n=1)
Dermatology	2	- Annual examination is possible but not necessary (n=1) - 6-monthly follow-up given the history of skin tumors (n=1)
Endocrinology	2	- No systematic surveillance for thyroid cancer (n=1) - Annual surveillance after thyroidectomy (n=1)
Urology	4	- Annual urinalysis, cystoscopy/ CT scan if necessary (n=1) - No routine surveillance recommended, only if hematuria (n=1) - 6- monthly follow-up given the history of urological tumors (n=1) - No advice (follow-up prostate cancer) (n=1)
Gynecology	6	- No written advice (n=1) - Annual follow-up with gynecological examination (n=3), vaginal ultrasound (n=2), CA 125 (n=4) and endometrial sampling (n=2) starting at 35-40 year - Hysterectomy (n=3), bilateral salpingo-oophorectomy after completion of childbearing - 6- monthly gynecological examination, annual colonoscopy and ultrasound of the kidneys (n=1)
Radiotherapy	4	- No written advice
Medical Oncology	1	- EGD every 3 years, annual colonoscopy, urine analysis and ultrasound (n=1)

(IPAA) or ileorectal anastomosis (IRA), to be carried out 'at relatively young age'.

Surveillance of the stomach and duodenum was recommended to all patients by means of an EGD at the age of 20-25 years, to be repeated every 1-3 years.

Screening for thyroid cancer by performing an annual clinical examination and an ultrasound was recommended in FAP and AFAP patients, starting from the age of 10-12 years. This advice was uniform except for one patient, in whom a 3-monthly clinical examination with evaluation of TSH and free thyroxine value was recommended starting from birth.

A final recommendation concerns the screening for hepatoblastoma. Overall, a clinical examination with liver palpation, ultrasound and determination of the AFP value was advised annually, starting from birth up to the age of 7 years. To one patient it was recommended that surveillance could be stopped at the age of 5 years. To another patient a more intensive screening was proposed with a three-monthly determination of the AFP value and an ultrasound; for a third patient it was stated that there was no consensus about the screening frequency, ranging from 1 time every 3 months to annually.

Advice in other departments

Eighteen out of 43 patients had surveillance in an external center. The remaining 25 had at least one follow-up contact : 18 were seen exclusively at the Ghent university hospital and 7 partly in an external center. Sixteen patients had at least one contact at the surgical department, 14 at the gastroenterology department, 10 at the department of pediatrics and 2 patients were followed at other departments. The advice given at the gastroenterology, surgery and pediatrics department is shown in table 4 and 5.

Two out of 14 patients who had at least one contact at the gastroenterology department (13 with FAP, 1 with MAP) were only referred for a second opinion regarding the treatment of duodenal polyps, after which they were referred back to an external center. Sixteen patients (14 with FAP, 2 with AFAP and 1 with MAP) were seen at the surgery department. Two patients had a metastatic tumor and surveillance measures were deemed inappropriate. No written advice was recorded in 7 out of 14 remaining patients (50%). Six of these patients were postoperatively referred to another department or an external center. One patient was only seen pre-operatively.

Follow-up

Out of the 25 patients with at least one contact at the gastroenterology, surgery or pediatrics department, 18 patients had a regular (annual to 2-yearly) follow-up consultation. A total of 8 patients (32%) were lost-to-follow-up: 4 children at the pediatrics department, 1 patient who did not receive follow-up after surgery (neither at the department of surgery, nor at the pediatrics department), 2 patients who were followed at the gastroenterology and surgery department and 1 patient from the gastroenterology department. Of these, only one patient was picked up for further surveillance after a number of years.

Discussion

Present guidelines regarding the surveillance and treatment of patients with LS, (A)FAP and MAP vary to some extent between countries and organisations (tables 6 and 7). The differences in recommendations regarding screening, surveillance and surgery for polyps and colon

Table 4. — **Recommendations from the gastroenterology and surgery department for patients with FAP/AFAP/MAP**

Exam	Gastroenterology department (n = 14)		Surgery department (n = 7)	
	Number	Frequency	Number	Frequency
Colonoscopy	11 (79%)	Yearly	7 (100%)	1-2 yearly (6), no interval reported (1)
EGD	12 (86%)	Yearly (3), 2- yearly (1), according to Spigelman stage (6); not reported (2)	5 (71%)	Yearly (3), 2- yearly (1), no interval reported (1)
Thyroid examination and ultrasound	5 (36%)	Yearly (5)	1 (14%)	2- yearly

Table 5. — **Recommendations from the pediatrics department for patients with FAP**

Exam	Number	Frequency and age of onset
Clinical examination	5 (50%)	Every year (n= 4) or every 3 months (n= 1) Starting from birth
Liver palpation, ultrasound and AFP	6 (60%)	Every 3 months (n=1), every year (n= 4) or every 6 months (n= 1) Starting from birth up to the age of 7 years (n= 5) or 5 years (n = 1)
Colonoscopy	9 (90%)	Every year (n= 9) Age of onset: 10-12 years
EGD	5 (50%)	Every 1 – 3 year (n= 4) Age of onset: 20-25 years (n= 4) or 15 years (n= 1)
Palpation and ultrasound of the thyroid gland	5 (50%)	Every year (n = 4) or every 3 months (n= 1) Age of onset: 10-12 years (n= 4) or 16–18 years (n = 1)
Abdominal ultrasound for detection of desmoid tumors	1 (10%)	Every year

Table 6. — **Recommendations in literature for surveillance in FAP/AFAP/MAP patients**

		Colon				Hepatoblastoma		
		What	**Start (y)**	**Frequency (y)**	**Treatment**	**What**	**Start (y)**	**Frequency (y)**
ESMO (5)	FAP	Sigmoidoscopy Colonoscopy when adenoma	12-14	2 1 when adenoma	Colectomy (IRA) Proctocolectomy (IPAA)	/		
	AFAP/MAP	Colonoscopy	18-20	2 1 when adenoma				
ASCO (2)	FAP	Sigmoidoscopy Colonoscopy when adenoma	10-11	1-2 1 when adenoma	Colectomy (IRA) Proctocolectomy (IPAA)	/		
	AFAP	Colonoscopy	18-20	1-2 1 when adenoma				
ACG (7)	FAP	Sigmoidoscopy/colonoscopy	Puberty	1	Colectomy	AFP and ultrasound	0 till 7	2
	AFAP/MAP	Colonoscopy		1				
Mallorca (16)	FAP	Sigmoidoscopy Colonoscopy when adenoma	10-12	2 1 when adenoma	Colectomy (IRA) Proctocolectomy (IPAA)	/		
	AFAP/MAP	Colonoscopy	18-20	2 1 when adenoma				
ASCRS (17)	FAP	Sigmoidoscopy à colonoscopy	10	2 1 when adenoma	Colectomy (IRA) Proctocolectomy (IPAA)	/		
	AFAP	Colonoscopy	20	2				
	MAP	Colonoscopy	20	1				

Table 6 (continued)

		Stomach			Thyroid		
		What	**Start (y)**	**Frequency**	**What**	**Start (y)**	**Frequency (y)**
ESMO	FAP/AFAP	EGD with duodenoscope Consider videocapsule	25-30	According to Spigelman stage	US	25-30	1
	MAP				/		
ASCO	FAP/AFAP	EGD with duodenoscope	25-30	According to Spigelman stage	Consider US	25-30	1
	MAP				/		
ACG	FAP/AFAP	EGD with duodenoscope	25-30	According to Spigelman stage	US		1
	MAP	Random biopsies fundic gland polyps					
Marllorca	FAP/AFAP	EGD with duodenoscope	25-30	According to Spigelman	/		
	MAP						
ASCRS	FAP/AFAP	EGD with duodenoscope	20-25	According to Spigelman stage	Consider US		1
	MAP	EGD with duodenoscope	30		/		

Abbreviations : IRA : ileorectal anastomosis ; IPAA : Ileo-anal pouch anastomosis ; EGD : esophagogastroduodenoscopy ; US : ultrasound.

cancer are small, but advice regarding risk-management for associated tumors is variable and mainly based on expert opinion given the lack of high-quality studies.

Previously, small studies have already suggested that many individuals with a hereditary gastrointestinal cancer syndrome do not have surveillance as specified in the guidelines and it has also been described that patients often receive different advice from their various health care providers (6,7). In this study, little variation was seen in the initial proposal for surveillance, as written down by the department of genetics in the 73 patients with LS and 35 patients with polyposis syndromes. Whatever small variation was noted, was not due to changes in institutional guidelines over time, but more to an accent given by the physician at the time of the consultation. Despite clear written recommendations from the medical genetics department, follow-up was not uniformly organized and sometimes different advice was given in the various departments were patients received their follow-up. The fact that some patients combine exams in different hospitals also makes it difficult to maintain an overview and have a coordinated follow-up. Overall, advice on screening and surveillance for colon polyps as well as surgery were in accordance with international guidelines – both for LS and polyposis syndromes. However, surveillance for other tumors, most importantly endometrial and ovarian cancer, but also urinary tract in LS and thyroid cancer in polyposis, is less well organized. It is possible that we underestimated the number of patients that undergo the proper examinations in other hospitals, but for those that have surveillance in our center, advices vary considerably between departments. Again, this is not due to a change in institutional guidelines over time. The variability in advice may be explained by the fact that evidence in the medical literature on the benefit of surveillance for these tumors is less specific and recommendations in different guidelines also vary. Consequently, the advice from the different departments are not in themselves 'wrong', but ideally they should be uniform and in accordance with the recommendations given at the medical genetics department, which can easily be found in the electronic patient files.

Table 7. — **Recommendations in literature for surveillance in Lynch patients**

	Colon				Ovaries/endometrium		
	What	**Start (y)**	**Frequency (y)**	**Alternative**	**What**	**Start (y)**	**Frequency (y)**
ACG (7)	Colonoscopy	20-25	1-2	Colectomy if CRC	TVUS+ES	30-35	1
					Hysterectomy and BSO	40-45	
MST (1)		20-25 2-5y before youngest family member if < 25	1-2	(partial) colectomy if not controlable	TVUS+ES	30-35	1
					Hysterectomy	40	
Mallorca (8)		20-25	1-2	(partial) colectomy if CRC	TVUS+ES	35-40	1-2
					Hysterectomy and BSO	40	
NCCN (3)		20-25 2-5y before youngest family member if < 25	1-2		TVUS+ES+CA125	After completion child-bearing	1
					Hysterectomy and BSO		
ESMO (5)		20-25 <5y before youngest family member if < 25	1-2	(partial) colectomy if CRC, to discuss with patient	TVUS+ES+CA125	30-35	1
					Hysterectomy and BSO	35	
ASCO (2)		20-25 <5y before youngest family member if < 25	1-2		TV US + ES, not CA125	30-35	
					Hysterectomy and BSO	After completion child-bearing	1
AGA (9, 10)		20-25 <5y before youngest family member if < 25			/		

Table 7 (continued)

	Stomach			Urinary tract		
	What	**Start (y)**	**Frequency (y)**	**What**	**Start (y)**	**Frequency (y)**
ACG	EGD + biopsy Screening for HP	30-35	3-5 if family history	No surveillance recommended		
MST	EGD + biopsy Screening for HP	30-35	2-3 if risk factors	Urinalysis	30-35	1
Mallorca	EGD in countries with high ncidence One- off screening for HP	25	1-2	Urine cytology + US if MSH2	30-35	1
NCCN	EGD + biopsy if high incidence Consider screening for HP	30-35	3-5	Urinalysis	30-35	1
ESMO	EGD if high incidence Screening for HP		1-3	No surveillance recommended		
ASCO	EGD of high incidence Screening for HP		1-3	No surveillance recommended		
AGA	/			/		

Abbreviations : TV US : transvaginal ultrasound ; ES : endometrial sampling ; BSO: bilateral ; EGD : esophagogastroduodenoscopy ; HP : Helicobacter Pylori ; US : ultrasound.

Most importantly, only a low number of patients (15 patients with LS and 18 with FAP/AFAP/MAP) were seen at consultation to discuss the results of the exams and to organize the next surveillance. These patients did not organize all surveillance examinations in our hospital, but are the only ones for whom we can be sure that all exams were performed regularly. In addition, 20% of patients with LS and 32% of patients with FAP were lost to follow-up, which is a fairly high percentage. Whether this is due to insufficient compliance of the patient or caused by incomplete or incorrect recommendations from the health care professionals is unclear.

The problem of variable recommendations and non-compliance has already been described in literature and there is some evidence that healthcare systems might actually create barriers to screening through ineffective coordination of care, lack of continuity and disparate recommendations (18). This failure in the patient care pathway has multiple causes. First, there is often a lack of responsibility assumed for surveillance follow-up, allowing individuals to be overlooked by the different caring systems (19). The content of information that patients receive, a good communication and knowledge about the disease are important factors that contribute to compliance and especially a good guidance from health care professionals appears to be very important (20-22). It has already been hypothesized that the presence of a designated team leader for these patients is associated with more thorough and organized screening systems and leads to a better compliance (18). Second, specialists themselves confirm that they often have a lack of knowledge regarding oncogenetics and express educational needs regarding this. Indeed patients often describe important differences in the support they receive from their doctors and up to one third experienced difficulties in finding health care providers who were

knowledgeable about Lynch syndrome (6). Additional training for doctors and better awareness of hereditary cancer syndromes seems appropriate. Lastly, there often seems to be a lack of information exchange between health care professionals (23). Given that patients with LS and FAP have an increased risk of multiple tumors, a multidisciplinary approach seems legitimate. The importance of good integrated care has already been confirmed in the literature, where it was shown that compliance was significantly higher in specific colorectal cancer clinics than in other centers (19).

This study has some limitations. First, the cohort was relatively small. Second, it was a retrospective analysis, based on the information that was found in the electronic patient record, so verbally delivered information could not be included.

In conclusion, a number of recommendations can be made with regard to the follow-up and surveillance of patients with hereditary cancer syndromes. There is a need for in integrated approach from a multidisciplinary team and uniform recommendations. The patient should be well informed regarding surveillance and a health care practitioner should take responsibility to coordinate all investigations. An annual, preferably multidisciplinary, outpatient check-up with discussion of all results and the planning of future investigations should be offered to reduce the risk of missing important investigations as much as possible.

Conflict of interest : none

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