HEREDITARY CANCER IN CLINICAL PRACTICE

POSTER PRESENTATION **Open Access**

Validating the Family History Scoring System application to identify Lynch syndrome families

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Background

The FHSS (a points-based scoring system) was previously used to identify families at high risk of an inherited colorectal cancer syndrome. AM-I (Amsterdam-I) families scored high (≤12), but some scored low (≥8) when scored from unaffected relative's perspective [1]. The FHSS is being applied to Lynch families (MMR+) and familial colon cancer type X (Type X) to see if the scoring system would yield similar results.

Method

Family members (1 proband, 2 siblings and 2 children of the sibling) in Lynch and Type X families identified from the Jagelman Registry database were scored according to the score sheet described in Table 1.

They were scored from perspective of an affected proband (AP) or unaffected proband (UP), affected (AS) or unaffected (US) sibling or a child of each sibling (Child).

Results

91 probands (68 affected, 23 unaffected) in 48 AM-I, 14 Amsterdam-II (AM-II), 6 Amsterdam-Like (AM-Like), 10 Familial Colon Cancer (FCC), 6 no syndrome (syn) families and 7 Type X were scored. 197 relatives were scored (38 affected siblings, 23 Child AP/AS, 40 Child AP/US, 77 unaffected siblings, 9 Child UP/AS, 10 Child UP/US). AM-I and Type X median scores were higher than other syndromes (<12) and suggestive of HNPCC when scored from the perspective of the proband, sibling or child of AS. The median scores were lower in AM-II, FCC, AM-Like, and no syn families with fewer colon cancers (Table 2).

Conclusion

Findings were similar to validation study conducted on AM-I families. The FHSS is a reliable tool to determine familial risk of colorectal cancer especially in AM-I families and depends on the perspective of person being scored. In small families, those with predominantly extra colonic cancers or high-risk polyps, or if affected relatives are more than a generation from an individual, the FHHS will not always identify high risk or Lynch families.

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Table 1

	Points
Proband	
Affected	*3*
First Degree Relatives(FDR)	
Each FDR	3 each
1-FDR<50-yr, <u>including proband</u>)	3
1-FDR–same side family	Extra 3
Second Degree relatives(SDR)	
Each SDR	1 each
>1-SDR<50-yr	1
>1-SDR–same side family	Extra 1*
Combination FDR and SDR	
1-FDR and any SDR–same side family	2*

only if maximum 3 points per family not exceeded.

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Table 2 Median Scores

	AP	AP's AS	AP's US	Child AP's AS	Child AP's US	UP	UP's AS	UP's US	Child UP's AS	Child UP's US
Total Syndromes	14	18	14	12	4	12	15	12	7	4
AM- I	17	18	17	12	5	12	16	12	9	4
Type X	14	17	14	14	4	12	15	12		
AM-II	12	12	12	7	2	13	13	15	9	5
FCC	12	13	9	10	2	10	10	8	8	10
AM-LIKE	12		12		4	4		4		1
No-Syn	6		6		2	6	6		6	

Reference
1. Church JM: A scoring system for the strength of a family history of colorectal cancer. *Dis Colon Rectum* 2005, **48**(5):889-96.