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# Real-world implications of IMACS malignancy screening guidelines for idiopathic inflammatory myopathies: An evaluation of compliance and economic impact at a tertiary referral center

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## Abstract

**Aim:** An inaugural set of consensus guidelines for malignancy screening in idiopathic inflammatory myopathy (IIM) were recently published by an international working group. These guidelines propose different investigation strategies based on “high”, “intermediate” or “standard” malignancy risk groups. This study compares current malignancy screening practices at an Australian tertiary referral center with the recommendations outlined in these guidelines.

**Methods:** We conducted a retrospective analysis of newly diagnosed IIM patients. Relevant demographic and clinical data regarding malignancy screening were recorded. Existing practice was compared with the guidelines using descriptive statistics; costs were calculated using the Australian Medicare Benefit Schedule.

**Results:** Of the 47 patients identified (66% female, median age: 63 years [IQR: 55.5–70], median disease duration: 4 years [IQR: 3–6]), only one had a screening-detected malignancy. Twenty patients (43%) were at high risk, while 20 (43%) were at intermediate risk; the remaining seven (15%) had IBM, for which the proposed guidelines do not recommend screening. Only three (6%) patients underwent screening fully compatible with International Myositis Assessment and Clinical Studies recommendations. The majority ( $N = 39$ , 83%) were under-screened; the remaining five (11%) over-screened patients had IBM. The main reason for guideline non-compliance was the lack of repeated annual screening in the 3 years post-diagnosis for high-risk individuals (0% compliance). The mean cost of screening was substantially lower than those projected by following the guidelines (\$481.52 [SD 423.53] vs \$1341 [SD 935.67] per patient), with the highest disparity observed in high-risk female patients (\$2314.29/patient).

**Conclusion:** Implementation of the proposed guidelines will significantly impact clinical practice and result in a potentially substantial additional economic burden.

Shereen Oon and Jessica Day contributed equally.

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**KEYWORDS**

compliance, costs, guidelines, idiopathic inflammatory myopathy, malignancy, myositis, screening

**Plain Language Summary**

1. The recently introduced malignancy screening guidelines for myositis are likely to significantly impact current practice. Notably, very few cases of full compliance with the recommendations were observed in the present study.
2. These guidelines have the potential to substantially increase healthcare costs associated with malignancy screening compared with current practice. The potential cost benefit of earlier malignancy detection should be determined with further economic studies.

## 1 | INTRODUCTION

The idiopathic inflammatory myopathies (IIM) are a collection of autoimmune diseases that predominantly affect skeletal muscles, but which can additionally present with extra-muscular manifestations including rash, inflammatory arthritis, and interstitial lung disease. This disease can result in significant disability, and carries a risk of permanent and life-limiting consequences if left untreated. Apart from the morbidity inherent to this disease, IIM has a strong association with malignancy within the first 3 years prior to and 3 years after IIM onset. The dermatomyositis (DM) subtype has the highest association with malignancy carrying a 5–14-fold increased risk.<sup>1,2</sup> The presence of certain myositis-specific antibodies also confers an increased risk of malignancy, most notably anti-TIF-1 $\gamma$  antibodies<sup>3</sup> followed by anti-NXP2 antibodies.<sup>4</sup> Understanding the risk factors for malignancy in IIM is important as malignancy is a leading cause of death for adults with this disease. Inclusion body myositis, however, has not been found to have an association with malignancy and malignancy is not a common cause of death in this IIM subtype.<sup>5</sup>

Despite the recognized association between IIM and malignancy, consensus guidelines for malignancy screening in individuals diagnosed with IIM were only recently issued by the International Myositis Assessment and Clinical Studies (IMACS) group.<sup>6</sup> These guidelines stratify patients into groups at high, intermediate, and standard risk of malignancy, based on the presence of factors such as myositis-associated antibodies, myositis-specific antibodies, demographics and clinical features. The recommended investigations for malignancy screening vary according to the identified risk group. Given the substantial risk of malignancy in IIM and the potential benefit of early detection, these guidelines are of considerable importance. Detection and treatment of an underlying malignancy could also lead to improvement in IIM disease activity, thereby reducing the need for prolonged immunosuppression which may increase the risk of developing or progressing the malignancy.<sup>7</sup> The recommendations outlined in the new IMACS guidelines may differ from established clinical practices and clinicians will need to adapt to the impending paradigm shift. The implementation of these guidelines into routine practice will therefore have implications for both clinical care and the associated health economic burden.

The potential impact on current clinical practice and the economic burden of these guidelines has not been previously evaluated. This study therefore aimed to compare existing practice for IIM-associated malignancy screening at an Australian tertiary public hospital with the proposed guidelines, and evaluate the difference in direct costs of existing screening practices to that of the proposed guidelines.

## 2 | METHODS

### 2.1 | Study population

A retrospective cohort analysis was conducted on all patients diagnosed with IIM between January 1, 2012, and April 1, 2023, at the Royal Melbourne Hospital in Victoria, Australia, a large tertiary metropolitan hospital providing specialized inpatient and outpatient rheumatology services. Patients were included if they were diagnosed with any IIM subtype, namely dermatomyositis (DM), polymyositis (PM), anti-synthetase syndrome (ASyS), inclusion body myositis (IBM), immune-mediated necrotizing myopathy (INMN), cutaneous amyopathic dermatomyositis (CADM), and connective tissue disease-related myositis.

The study cohort was identified from two primary sources. A search of the hospital's electronic medical records (EMR) for the following terms was conducted: "myositis," "dermatomyositis," "polymyositis," "anti-synthetase syndrome," "inclusion body myositis," "immune-mediated necrotizing myopathy," and "HMG-CoA reductase antibodies." In parallel, a search of the hospital's network drive containing historical rheumatology outpatient letters predating the implementation of the EMR system was performed. Patients for whom we were unable to locate sufficient information to confirm the diagnosis were excluded. Patients were included even if they did not fulfill the EULAR/ACR 2017 classification criteria of IIM<sup>8</sup> as these criteria do not account for all currently known myositis-specific antibodies. Patients not meeting EULAR/ACR criteria were diagnosed and subtyped on the basis of clinical, serological, and histological features.

Once patients were identified, information to risk stratify patients according the IMACS guidelines was extracted from their



medical records. Specifically, the subtype of IIM, antibody positivity, presence of clinical features of Raynaud's phenomenon, inflammatory arthropathy, interstitial lung disease, sex, age at IIM onset, and whether the patient was experiencing persistent disease activity despite immunosuppressive therapy were collected. Patients were then stratified into either a standard, intermediate, or high-risk group based on the IMACS criteria, that is, patients with  $\geq 2$  intermediate-risk factors or 1 high-risk factor were considered at intermediate risk of malignancy while high-risk patients for malignancy were those with  $\geq 2$  high-risk factors. Patients who did not fulfill these criteria were classified as standard risk. Patients with a diagnosis of IBM were recommended to not be screened for malignancy. According to the guidelines, all non-IBM groups are recommended to undergo "Basic Screening" at the time of diagnosis, consisting of a history, physical examination, full blood count, liver function tests, inflammatory markers, serum protein electrophoresis, urinalysis, and chest X-ray. Those stratified into intermediate- and high-risk groups are recommended to undergo additional "Enhanced Screening" tests at diagnosis, consisting of a CT neck, chest, abdomen, and pelvis, cervical screening, mammography, a pelvic/transvaginal ultrasound, fecal occult blood test, and tumor markers. It is recommended that consideration should be given to performing PET scans and endoscopy in high-risk patients if the initial work-up does not identify a malignancy and that high-risk groups undergo basic screening annually for 3 years following IIM diagnosis.

Data regarding the screening investigations performed for each patient in accordance with the IMACS recommendations were collected, including blood tests, imaging, and procedural interventions such as endoscopy. We did not collect data regarding age-appropriate cancer screening tests mandated by national guidelines (e.g., PAP smear, mammogram) as these were presumed to have been completed by the patient's primary care physician in the community setting. In assessing compliance with the proposed guidelines, we considered high-risk patients who were within 3 years of their IIM diagnosis to be compliant with the IMACS guidelines if they had undergone the entire guideline-recommended work-up up to the date of their last review. Retrospectively determining the purpose of some investigations was challenging—certain investigations were likely performed as part of routine clinical monitoring (such as basic blood tests), however were assumed to have been conducted for malignancy work-up for the purposes of assessing compliance with the IMACS guidelines. We did not include screening for nasopharyngeal carcinoma in our analysis given that this cancer is not at epidemiologically elevated risk in our Australian setting.<sup>9</sup>

## 2.2 | Analysis of direct costs of screening

The direct costs of each investigation performed were determined using the Australian MBS (Medicare Benefits Schedule) billing codes on 01/04/2023, and are reported in Australian dollars. The cost of screening for high-risk patients included FDG-PET CT and gastroscopy and colonoscopy, if the patient did not have a

malignancy identified on initial screening, in line with the IMACS guidelines.

## 2.3 | Statistical analysis

Continuous variables were expressed as a median with interquartile range. Costs were expressed as a mean.

## 2.4 | Ethics

This study was conducted with approval from the Melbourne Health Office for Research Ethics & Governance (QA2022003).

# 3 | RESULTS

## 3.1 | Demographics

A total of 47 patients were identified. The most common subtype of IIM was dermatomyositis, diagnosed in 18 patients (38%). This was followed by polymyositis ( $n$ : 9, 19%), IMNM ( $n$ : 8, 17%), IBM ( $n$ : 7, 14%), and anti-synthetase syndrome (ASyS) ( $n$ : 5, 10%). That DM was the most common IIM subtype within this cohort is notable. This may reflect the advances in serological profiling that enable more refined classification of PM into distinct entities such as ASyS, overlap myositis, and IMNM. Historically, these conditions may have been categorized as polymyositis. Additionally, the observed distribution of IIM subtypes may be skewed by the fact that neurologists at our institution also treat IMNM and IBM, potentially contributing to their under-representation within this rheumatology-derived cohort. Patient demographics reflected the known sex-based prevalence of IIM with a ratio of 2:1 female ( $n$ : 31) to male ( $n$ : 16) patients (Table 1). The median age was 63 (IQR: 55.5–70) years. The most common high-risk factor was age  $>40$  years, with only 3 patients younger than 40. Age was also the most common reason patients were stratified into a higher risk group. Notably, all patients with only two high-risk factors were aged above 40 years; had they been younger than 40 years of age, and they would have been classified as intermediate risk. Similarly, in the intermediate-risk group, 10 (50%) of the patients would have been classified as standard risk had they been younger than 40 years old. The median length of follow-up was 4 (IQR 3–6) years. Ten patients (21%) were within the first 3 years of their IIM diagnosis.

## 3.2 | Malignancy risk stratification per IMACS guidelines

Stratified by sex and risk of malignancy, the most common risk group was female patients with intermediate risk of malignancy ( $n$ : 15, 32%) (Table 2) followed by female patients with high

**TABLE 1** Demographics and malignancy risk factors as per the International Myositis Assessment and Clinical Studies (IMACS) screening recommendations.

Demographics	
Median (IQR) years	
Age at IIM diagnosis (median [IQR])	63 (55–70)
Length of follow-up	4 (3–6)
Sex	n (%)
Female	31 (66%)
Male	16 (34%)
Race	n (%)
White/Caucasian	41 (87%)
Asian	3 (6%)
Indigenous Australian	1 (2%)
Black/African descent	1 (2%)
Pacific Islander	1 (2%)
Risk Factors	n (%)
High Risk	
Dermatomyositis	18 (38%)
Anti-TIF1 $\gamma$ positivity	4 (9%)
Anti-NXP2 positivity	3 (6%)
Age > 40 years at time of onset	43 (92%)
Features of persistent high disease activity despite immunosuppressive therapy	1 (2%)
Dysphagia	16 (34%)
Cutaneous necrosis	16 (34%)
Intermediate risk	
Clinically amyopathic dermatomyositis	2 (4%)
Polymyositis	9 (19%)
Immune-mediated necrotizing myopathy	8 (17%)
Anti-SAE1 positivity	4 (9%)
Anti-HMGCR positivity	7 (15%)
Anti-Mi2 positivity	2 (4%)
Anti-MDA5 positivity	2 (4%)
Male Sex	16 (34%)
Low risk	
Anti-synthetase syndrome	5 (11%)
Overlap IIM/Connective tissue disease-associated myositis	0
Anti-SRP positivity	0
Anti-Jo1 positivity	2 (4%)
Non-anti-Jo1 anti-synthetase syndrome antibody positivity	2 (4%)
Myositis-associated antibody positivity (anti-PM-Scl, anti-Ku, anti-RNP, anti-Ro/La (SSA/B))	0
Raynaud's	2 (4%)
Inflammatory arthropathy	12 (26%)
Interstitial lung disease	8 (17%)
Not for screening	
Inclusion Body Myositis	7 (15%)

malignancy risk ( $n = 11$ , 23%). None of the patients met the criteria for the standard-risk group, and seven individuals (14%) had a diagnosis of IBM. Female patients outnumbered male patients in each risk group.

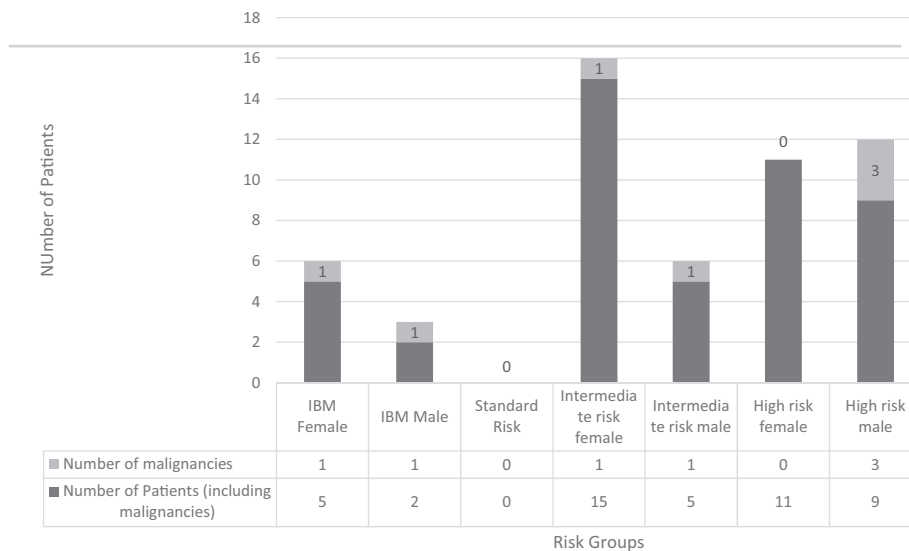
### 3.3 | Detected cancers

Cancers were identified in seven patients (Table 2), six of whom were diagnosed with malignancies via targeted screening directed by symptoms suggestive of a malignancy; the remaining patient was diagnosed via blind screening not targeted to any specific symptom (Table 3). Of the six patients diagnosed via targeted screening, two had an IIM diagnosis of anti-TIF1 $\gamma$  positive DM. One such patient was diagnosed with metastatic prostate cancer 2 years after their DM diagnosis, while the other was diagnosed with tonsillar squamous cell carcinoma 2 years prior to their DM diagnosis. Interestingly, two of the patients with IBM were diagnosed with a hematological malignancy, one with Hodgkin's lymphoma 2 years after their diagnosis of IBM and the other patient with splenic marginal zone lymphoma 3 years post their IBM diagnosis, acknowledging that patients with IBM were not recommended to undergo any cancer screening due to a lack of association with malignancy. The patient diagnosed via blind screening presented with a 3-month history of a heliotrope rash, proximal muscle weakness, and bilateral pleural effusions. A skin biopsy showed a lichenoid reaction pattern with superficial and deep perivascular inflammation, dermal mucin, and bullous formation, consistent with dermatomyositis. Cytology of the pleural fluid identified malignant cells consistent with an upper gastrointestinal/pancreaticobiliary or lower gastrointestinal malignancy; however, a specific tissue diagnosis of the primary malignancy was unable to be obtained.

### 3.4 | Compliance with IMACS malignancy screening recommendations

Only three patients were completely compliant with the IMACS screening recommendations. Two of these patients were diagnosed with IBM and did not receive any malignancy screening with the exclusion of basic blood tests which were performed for routine monitoring. The remaining patient was a high-risk male patient diagnosed with anti-NXP2 positive DM who at the time of data extraction was 1 year post IIM diagnosis and was compliant with all screening requirements to date.

Many patients underwent at least part of the recommended screening investigations at the time of diagnosis; however, high-risk patients rarely underwent repeated annual screening as recommended (Table 4). While all patients completed basic blood tests (FBE, LFT, CRP, ESR) at each clinic review, only a small percentage of patients received the complete annual work-up recommended by the IMACS guidelines, consisting of an annual serum electrophoresis ( $n = 0$ , 0% compliance), urinalysis ( $n =$



**TABLE 2** Malignancy risk stratification of patients with idiopathic inflammatory myopathy (IIM) per International Myositis Assessment and Clinical Studies (IMACS) guidelines.

**TABLE 3** Characteristics of patients with idiopathic inflammatory myopathy (IIM) in whom a malignancy was detected. Targeted screening denotes screening targeted to concerning clinical features. Blind screening denotes screening not guided by specific clinical features.

IIM subtype	Risk factors	Risk group	Type of screening	Cancer	Time to onset of cancer
Dermatomyositis	Age >40years, Dermatomyositis, anti-TIF1 $\gamma$ positive, dysphagia, male	High-risk male group	Targeted	Metastatic prostate cancer	2 years prior to IIM
Dermatomyositis	Age >40years, Dermatomyositis, anti-TIF1 $\gamma$ positive, dysphagia, male	High-risk male group	Targeted	Tonsillar squamous cell carcinoma	2 months prior to IIM
Dermatomyositis	Age >40years, DM, anti-Mi2 positive	Intermediate-risk female group	Targeted	Breast cancer	1 year prior to IIM
Dermatomyositis	Age >40, DM, male	High-risk male group	Blind	Metastatic malignancy with unknown primary	5 days after IIM
Immune-mediated necrotizing myopathy	Age >40years, male, IMNM, anti-HMG-CoA reductase positive	Intermediate-risk male group	Targeted	Metastatic prostate cancer	2 years after IIM
Inclusion body Myositis	Age >40years, dysphagia, male	Not indicated for screening	Targeted	1. Non-small cell lung cancer 2. Hodgkin's lymphoma	1. 1 year prior to IIM 2. 2 years after IMM
Inclusion body Myositis	Age >40years	Not indicated for screening	Targeted	Splenic marginal zone lymphoma	3 years after IIM

2, 10%), and chest X-rays ( $n = 2, 10\%$ ) (Table 4). There were also significant gaps in compliance between existing practice and the recommended guidelines for the enhanced screening regimen, particularly with regard to the use of CT imaging (Table 5). While more than half the patients underwent a CT chest, abdomen, and pelvis ( $n = 25, 63\%$ ), only two patients (5%) received the IMACS-recommended additional CT neck along with the CT chest, abdomen, and pelvis. Imaging for detection of gynecological malignancies was similarly infrequently performed in only two eligible patients (8%).

Overscreening also occurred in five (10%) patients, especially in patients diagnosed with IBM, where all male patients underwent a CT chest, abdomen, and pelvis (Table 5).

### 3.5 | Cost comparison between existing and recommended practice

Consistent with the previous findings of a trend toward under-screening patients in existing practice compared with the recommended guidelines, the actual costs of screening incurred for existing practice were less than the costs associated with the proposed guidelines. This pattern held true for each risk group, except for IBM, where overscreening occurred. Overall, there was a mean actual spend of \$481 (SD 423.53) per patient which was below the mean proposed costs of \$1341 (SD 935.67) per patient.

Examination of the costs associated with each risk group revealed that the largest disparity between actual spending versus proposed



**TABLE 4** Compliance of existing practice against the proposed International Myositis Assessment and Clinical Studies (IMACS) guideline's basic screening regimen (*n*, %).

Risk Group	Basic bloods (FBE, LFT, CRP, ESR) <sup>a</sup>		Serum Protein Electrophoresis		Urinalysis		Chest X-ray	
	At diagnosis	Annually 3 years	At diagnosis	Annually 3 years	At diagnosis	Annually 3 years	At diagnosis	Annually 3 years
IBM female ( <i>n</i> =5)	5 (100%) <sup>a</sup>	5 (100%) <sup>a</sup>	1 (20%) <sup>a</sup>	NR	1 (20%) <sup>a</sup>	NR	1 (20%) <sup>a</sup>	NR
IBM male ( <i>n</i> =2)	2 (100%) <sup>a</sup>	2 (100%) <sup>a</sup>	1 (50%) <sup>a</sup>	NR	0 (0%)	NR	1 (50%) <sup>a</sup>	NR
Low risk ( <i>n</i> =0)	-	-	-	-	-	-	-	-
Intermediate-risk female ( <i>n</i> =15)	15 (100%)	15 (100%)	3 (20%)	NR	6 (40%)	NR	10 (66%)	NR
Intermediate-risk male ( <i>n</i> =5)	5 (100%)	5 (100%)	2 (40%)	NR	1 (20%)	NR	5 (100%)	NR
High-risk female ( <i>n</i> =11)	11 (100%)	11 (100%)	3 (27%)	0 (0%)	3 (27%)	2 (18%)	7 (64%)	2 (18%)
High-risk males ( <i>n</i> =9)	9 (100%)	9 (100%)	2 (22%)	0 (0%)	4 (44%)	0 (0%)	7 (78%)	0 (0%)
<b>Compliance</b>	40 (100%)	40 (100%)	10 (25%)	0 (0%)	14 (35%)	2 (10%)	29 (73%)	2 (10%)

Abbreviation: NR, not required.

<sup>a</sup>Denotes overscreening.

**TABLE 5** Compliance of existing practice against the proposed International Myositis Assessment and Clinical Studies (IMACS) guideline's enhanced screening regimen (*n*, %).

Risk Group	Tumor Markers (PSA or CA125)	CT Neck/Chest/Abdomen/Pelvis	CT Chest/Abdomen/Pelvis	Pelvic/Transvaginal Ultrasound	PET scan	Gastroscopy and Colonoscopy
IBM female ( <i>n</i> =5)	0 (0%)	0 (0%)	1 (20%) <sup>a</sup>	1 (20%) <sup>a</sup>	0 (0%)	0 (0%)
IBM male ( <i>n</i> =2)	2 (100%) <sup>a</sup>	0 (0%)	2 (100%) <sup>a</sup>	N/A	0 (0%)	0 (0%)
Low risk ( <i>n</i> =0)	-	-	-	-	-	-
Intermediate-risk female ( <i>n</i> =15)	3 (20%)	0 (0%)	8 (53%)	2 (13%)	2 (13%)	1 (6%)
Intermediate-risk male ( <i>n</i> =5)	1 (20%)	0 (0%)	3 (60%)	N/A	1 (20%)	1 (20%)
High-risk female ( <i>n</i> =11)	1 (9%)	1 (9%)	7 (64%)	0 (0%)	1 (9%)	3 (27%)
High-risk males ( <i>n</i> =9)	2 (22%)	1 (11%)	7 (78%)	N/A	5 (56%)	3 (33%)
<b>Compliance</b>	7 (18%)	2 (5%)	25 (63%)	2 (8%)	9 (23%)	8 (20%)

Abbreviation: N/A—not applicable.

<sup>a</sup>Denotes overscreening.

spending was in high-risk female patients, with a difference of \$2314.29 (Table 6) per patient. This observation is particularly notable given that this risk group had the highest proposed spend of all the risk groups at \$2853.15 per patient. In contrast, the high-risk male group had a lower proposed spend at \$2631.80 per patient, but a higher actual mean spend compared with the high-risk female group (\$790.71 versus \$534.86, respectively). A similar underspend on female patients compared with male patients was seen the intermediate-risk group though this was a difference of only \$67.80. Only in the IBM group was there higher expenditure directed toward screening female patients as opposed to male patients, with an overspend of \$138.45 and \$96.55 for female and male patients, respectively.

## 4 | DISCUSSION

Our results indicate that stringent implementation of the proposed IIM malignancy screening guidelines would substantially change

current practice. Key findings were that very few patients at the study center were fully compliant with the proposed guidelines, largely due non-compliance with repeated annual screening for the full set of recommended bloodwork, and lack of a CT neck in addition to CT chest, abdomen and pelvis. This was associated with an underspend compared with the proposed guidelines of just under a mean of \$1000 per patient. An important modification to existing practice will be the performance of screening investigations yearly over a period of 3 years. As established in previous studies, IIM-associated malignancies commonly occur within the 5 years period before or after a diagnosis of IIM.<sup>10</sup> Hence, clinicians should be mindful that while investigations at time of diagnosis may be reassuring with a negative result, malignancies could become evident years after the initial IIM diagnosis. Additionally, care must be given to ensure that bias does not influence the application of appropriate cancer screening. Notably, transvaginal/pelvic ultrasound to screen for gynecological malignancies was one of the least performed screening tests and female patients were found to have had less expenditure allocated



**TABLE 6** Costs of screening for each risk group compared with proposed costs.

Risk Group	Actual cost Mean (SD) \$AUD	Proposed cost \$AUD	Difference in cost (proposed-actual) \$AUD
IBM female <sup>a</sup> (n=5)	138.45 (44.93)	0	-138.45
IBM male <sup>a</sup> (n=2)	96.55 (25.54)	0	-96.55
Low risk (n=0)	No patients	134.45	Not applicable
Intermediate-risk female <sup>a</sup> (n=15)	296.00 (406.10)	985.35	689.35
Intermediate-risk male <sup>a</sup> (n=5)	363.8 (400.16)	764	400.2
High-risk female <sup>b</sup> (n=11)	534.86 (417.17)	2853.15	2314.29
High-risk males <sup>b</sup> (n=9)	790.71 (509.20)	2631.8	1840.29

<sup>a</sup>Costs of screening for the intermediate-risk and IBM groups were calculated over 1 year post-diagnosis.

<sup>b</sup>Costs for high-risk group was calculated over 3 years post-diagnosis with PET scan, gastroscopy, and colonoscopy. The proposed cost without a PET scan, gastroscopy, and colonoscopy is \$1388.70 and \$1167.35 for female and male patients, respectively.

toward them compared with their male counterparts. This observation raises the possibility of a gender bias and is particularly pertinent considering that female patients are more commonly affected by non-IBM forms of IIM, as evidenced in both this study and others.<sup>11</sup>

Despite not being included in the proposed screening guidelines due to its purported lack of association with malignancy, we included IBM in our analysis to assess the number of patients who nonetheless did undergo some form of screening, and they were found to have been overscreened as a group. However, despite existing consensus that IBM does not portend a risk of malignancy, some evidence suggests that IBM may have an association with hematological malignancy, particularly T cell large granular lymphocytic leukemia (TLGLL).<sup>12</sup> Interestingly, the patients in this study who developed a malignancy concurrent to their IBM were diagnosed with hematological malignancies, although not TLGLL specifically. Should further research validate a link between IBM and TLGLL, or hematological malignancies more generally, a revision of the screening protocols for IBM patients may be necessary.

This study is the first assessment of the potential impact of the proposed IMACS malignancy screening guidelines on current practice and the healthcare economy. It was conducted in a real-world setting and reflects actual clinical practice. Nonetheless, there are some limitations to this study. Data regarding age-appropriate cancer screening were assumed to have been completed but not specifically collected for analysis as these investigations are usually conducted in the community and this information is therefore not reliably documented in hospital medical records. Further, the study comprises a relatively small cohort size in a single center. A more comprehensive evaluation will require a multicenter and ideally multinational approach, which will have an additional advantage of further diversifying the study cohort.

The implementation of any malignancy screening regimen should be evaluated for both its impact on clinical outcomes and economic burden. Such assessments have been carried out for large-scale age-appropriate screening programs for ovarian cancer, colon cancer, and breast cancer.<sup>13,14</sup> The challenges inherent to screening these large cohorts of patients may not fully apply to the IIM group given the low prevalence of IIM relative to a comparatively high incidence rate of associated malignancy. Hence, while screening costs maybe high, the absolute number of patients requiring screening investigations are relatively fewer and the likelihood of detecting cancer higher. As acknowledged by the guideline authors themselves, the guidelines will require such evaluation as they are largely consensus-based and evidence-based screening should be investigated.<sup>15</sup> Additionally, low-income countries or countries without a subsidized healthcare system may encounter financial barriers accessing more costly investigations such as PET scans. Economic considerations for malignancy screening also extend beyond simple cost barriers. The screening studies themselves may not be viable to be performed in under-resourced countries,<sup>16</sup> for example, the challenges faced in Malawi of implementing cervical cancer screening due to the low specificity of cytology testing as a result of technical and infrastructure limitations.

## 5 | CONCLUSION

Malignancy is a well-recognized association of idiopathic inflammatory myositis, and the introduction of consensus screening guidelines is a welcome development. Our study highlights the large discrepancy between current screening practices at a tertiary center and the recommendations outlined in the proposed guidelines. Non-compliance resulted primarily from a lack of annual screening involving urine and CXR, and the omission of a CT neck. Given these diagnostic procedures are generally accessible in Australia, achieving full compliance with the guidelines is feasible at our center. However, it is essential to acknowledge that this may not be universally applicable, as the availability and accessibility of diagnostic tools can vary across different healthcare settings globally. Our results additionally suggest that implementing the proposed guidelines will confer a notable increase in healthcare costs. However, this cost should be considered in light of the relatively low absolute number of patients with an IIM diagnosis and the potential cost benefits associated with earlier detection of malignancy. The economic implications and benefits of adhering to these guidelines will need thorough evaluation on a global scale in future studies.

## AUTHOR CONTRIBUTIONS

J.D. and S.O. conceptualised the study. Data was acquired and analysed by I.T. and V.H. The manuscript was written by I.T. with input from all authors. J.D. and S.O. contributed equally.

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## CONFLICT OF INTEREST STATEMENT

No conflicts of interest are identified.

## DATA AVAILABILITY STATEMENT

The data that support the findings of this study are available on request from the corresponding author. The data are not publicly available due to privacy or ethical restrictions.

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