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ORIGINAL ARTICLE

Indigenous bronchiectasis assessment scale – the 'IBAS': a proposed new tool to assess bronchiectasis severity in adult Indigenous Australians

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Key words
assessment tools, disease burden, ethnic disparities, outcome assessment, risk assessment.

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Abstract

Background: There is a lack of a comprehensive bronchiectasis severity assessment tool specific for Indigenous people that corrects for normative references established for the non-Indigenous population.

Aims: An innovative bronchiectasis assessment tool is developed for use in adult Indigenous patients – the Indigenous bronchiectasis assessment scale '(IBAS)'.

Methods: A total of 454 adult Indigenous Australian patients, with chest CT confirmed bronchiectasis diagnosed between 2011 and 2020, were included. Age, sex, residence location, body mass index, radiological findings, sputum microbiology, lung function parameters and medical comorbidities were utilised to predict 5-year all-cause mortality and 5-year hospitalisations. Scores of parameters with $P < 0.20$ from univariate Cox regressions were derived.

Results: The resultant IBAS included age (<30, 30–50, 50–70 and 70+ years), urban residence, forced vital capacity (% predicted) (>50%, 30%–50% and <30%), right lower lobe involvement, history of *Haemophilus* spp., *Pseudomonas* spp., *yeast* spp. or *Moraxella* spp., 2-year respiratory condition hospitalisation history (<2, 2 and 3+ admissions), and comorbid chronic obstructive pulmonary disease, asthma and arterial hypertension. The maximum score was 18, with thresholds at 0–4 (mild, $n = 78$, 34.4%), 5–7 (moderate, $n = 111$, 48.9%) and ≥ 8 (severe, $n = 38$, 16.7%). The area under the curve for 5-year mortality was 0.743 (95% confidence interval (CI) 0.683, 0.803). The IBAS score demonstrated significant delineation in mortality between mild and moderate (moderate hazard ratio (HR) 3.45 (95% CI 1.57, 7.58)) and between moderate and severe (severe HR 2.43 (95% CI 1.45, 4.07)).

Conclusion: The proposed IBAS tool could be of aid in assessing bronchiectasis severity in Indigenous patients.

Abbreviations: AUC, area under the curve; BMI, body mass index; BSI, bronchiectasis severity index; CI, confidence interval; COPD, chronic obstructive pulmonary disease; CT, computed tomography; FACED, forced expiratory volume in one second (FEV_1), age, chronic colonisation, extension, and dyspnoea.; FEV_1, forced expiratory volume in 1 s; FVC, forced vital capacity; HRs, hazard ratios; IBAS, Indigenous bronchiectasis assessment scale; IQR, interquartile range; LFPs, lung function parameters; NT, Northern Territory; RLL, right lower lobe; ROC, receiver operating characteristic curve; SD, standard deviation; TEHS, Top End Health Service

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Introduction

Bronchiectasis is a chronic respiratory condition, clinically characterised by a disabling cycle of recurrent lower respiratory tract infections leading to high hospital admission rates and mortality.[1,2] There is substantial heterogeneity and geographic variation in the prevalence and in the clinical manifestations of bronchiectasis among global populations.[3] In the adult Indigenous population, bronchiectasis prevalence is reported to be much higher, estimated at 19.4/1000 people.[4] In addition to higher prevalence, adult Indigenous patients are observed to be significantly younger, have a higher burden of comorbidities, and display lower lung function parameters (LFPs) and a greater array of sputum microorganisms, including higher hospital admission rates and earlier mortality in comparison to most ethnically diverse global non-Indigenous populations.[5] The economic cost for Indigenous patients secondary to bronchiectasis exacerbations is estimated to be in excess of AU $2.3 million per year, and much more for patients residing in rural and remote communities, where equity of access to advanced health care is problematic.[6]

Two established tools are widely utilised to predict disease severity, exacerbations and mortality among patients with bronchiectasis – the forced expiratory volume in 1 s (FEV_1), age, chronic colonisation, extension and dyspnoea (FACED) score and the bronchiectasis severity index (BSI).[7,8] However, these tools were not developed using Indigenous patients normative reference values. Indeed, a recent study has demonstrated that the FACED score is not valid nor reliable in an Indigenous population, while the BSI's accuracy and validity are lower than that reported among non-Indigenous patients.[9]

Nevertheless, despite the known differences in the clinical manifestations and outcomes for Indigenous patients with bronchiectasis compared to non-Indigenous patients, there are no established tools that could be used to predict disease severity, exacerbation frequency and mortality specifically designed for adult Indigenous patients. About 3.2% of the population in Australia self-identify as being of Indigenous descent. However, in the Northern Territory (NT) of Australia, 26.3% of the population self-identify as Indigenous Australians, a larger proportion than in any other Australian state or territory.[10] In the recent past, several studies from our centre reported on various aspects of respiratory disorders in the adult Indigenous population residing in the Top End Health Service (TEHS) region of the NT of Australia.[11–19] In line with what is observed in other Organisation for Economic Co-operation and Development (OECD) countries among Indigenous people, bronchiectasis is also reported to be highly prevalent in the TEHS, NT adult Indigenous Australian population.[20,21] Therefore, our centre is ideally positioned to develop an Indigenous specific bronchiectasis assessment tool in a population with a high prevalence of bronchiectasis – the Indigenous bronchiectasis assessment scale (IBAS), utilising clinical data, laboratory parameters, hospitalisation rates and overall mortality among adult Indigenous patients diagnosed to have bronchiectasis over a 10-year period from the TEHS region of the NT of Australia.

Methods

Ethics, setting and study participants

This study was approved by the Human Research Ethics Committee of the NT, Department of Health and Menzies School of Health Research (Reference: HREC; 2019-3547). The study patients included in this IBAS assessment tool are inclusive of patients represented in our previous report,[4] as a part of a larger project examining various aspects of bronchiectasis among adult Indigenous patients aged ≥18 years identified to have bronchiectasis via chest CT scan between 2011 and 2020 residing in the TEHS region of the NT of Australia. Further details on the population profiles of NT Australian Indigenous people and the study patients are available from our previous reports.[4,5]

Clinical data examined

In the current study cohort, due to the high prevalence of bronchiectasis among children in this region, all patients were assumed to have bronchiectasis at the beginning of the study period (1 January 2011) or, if they entered into the study at a later point, upon turning 18 years of age. Demographic and clinical data extracted from electronic medical records included age, sex, body mass index (BMI), medical comorbidities and residence location (urban or remote/very remote according to Australian statistical geography standard (ASGS) 3 and 4/5, respectively).[22] In addition, when available, smoking history, LFPs, radiological extent and type of bronchiectasis and sputum microbiology results were collected. Further, hospital admissions and overall, all-cause mortality were recorded.

Clinical data assessment

Age was categorised at thresholds of 30, 50 and 70 years due to a combination of prior research[7,8] showing

significant increases in risk above 70 years of age and the median age in this study cohort being 48 years (Table S1). LFPs were low, with only three patients recording a FEV_1 percentage predicted above 80%. Therefore, both FEV_1 and forced vital capacity (FVC) were assessed in three categories: mild (>50%), moderate (30%–50%) and severe (<30%). The presence of sputum micro-organism was defined as the recording of any positive culture results of *Pseudomonas* spp., *Haemophilus* spp., *Staphylococcus* spp., *Streptococcus* spp., *Aspergillus* spp., *Burkholderia* spp., *Klebsiella* spp., mycobacterium, non-*aspergillus* fungi or yeast between 2011 and 2020. The medical comorbidities considered included arterial hypertension, COPD, asthma, type 2 diabetes, chronic kidney disease and ischaemic heart disease. Hospitalisation data were restricted to those under the respiratory subcategory, international classification of diseases J code (ICD). To assess the severity of bronchiectasis, we considered a primary (5-year mortality) and secondary (respiratory hospitalisations in 5 years) outcome parameter. As with the BSI,[8] we considered a 2-year prior hospitalisation history as clinically significant. We defined a census date for our cohort as 1 January 2013, or when the patient reached 20 years of age if they were enrolled at the point when they turned 18.

Exclusion criteria

Participants were excluded from final analysis from the total cohort of 459 if (i) the reported date of death was earlier than the defined index date for the study ($n = 3$) and (ii) there was an excess of hospitalisations (>100 across the study period) ($n = 2$), which may have biased the results.

IBAS score development and statistical analysis

Utilising a random number generator within STATA (STATA 18, College Station, TX, USA), we split our cohort of patients randomly into either construction ($n = 227$) or validation ($n = 227$) cohorts. Initially, all continuous parameters were tested for normal distribution via Shapiro–Wilks W test within the construction and validation cohorts. Age, BMI, LFPs, number of lobes affected on chest CT and age of death were identified as non-normally distributed and, therefore, expressed as median (interquartile range (IQR)), while follow-up time was normally distributed and, thus, presented as mean ± standard deviation (SD), with categorical data presented as number (frequency (%)). Parameters were assessed within the construction cohort for their effect on the primary and secondary outcomes via Kruskal–Wallis rank-sum test (continuous outcomes) or chi-squared test (with two-tailed Fisher's exact test in the case of cells having <10 patients). No imputation of missing data was performed, and tests were only performed in patients with that respective parameter available.

In the second step of model construction, all parameters with a P-value <0.20 for either the primary or secondary outcomes were utilised in univariate Cox regression models reporting both hazard ratios (HRs) (95% confidence intervals (CIs)) and β coefficients (95% CIs) for their association with 5-year mortality and 5-year hospitalisations. To assign a score within the IBAS, we used the mean of the β coefficient for 5-year mortality and 5-year hospitalisation and rounded this to the nearest 0.50, as was done for the construction of the BSI scoring.[8] Parameters which yielded a score of 0 or a negative score following this process were excluded from the IBAS. We defined the IBAS as a total score which summed the score the patient received for each parameter. We defined thresholds of mild, moderate and severe for the total scores of the IBAS via visual inspection of receiver operating characteristic curve (ROC), area under the curve (AUC) which yielded the highest AUC for 5-year mortality. To validate the resultant total IBAS score, we used Cox regression models for 5-year mortality and 5-year hospitalisations in the validation cohort and the combined cohort. To check for differences in outcomes between severity levels, Cox regression models for 5-year mortality and hospitalisations were run testing mild against moderate and severe and testing severe against moderate. All analyses were conducted in STATA 18 (College Station, TX, USA).

Results

Of the total 454 participants included, the median (IQR) age of the overall sample was 48.8 (41.3, 58.2) years. A greater proportion (56%) were female, and *Pseudomonas* spp. was cultured in 17.4%. LFPs were available for 169 patients (37.2%), of whom 94.7% scored <50% predicted FVC. Five-year mortality was 14.5%, and 71.8% recorded at least one respiratory hospitalisation in the 5-year follow-up. Other relevant clinical data are detailed in Table S1. In the construction cohort, the median (IQR) and mean ± SD follow-up time were 120 (80.8, 120) and 98.2 ± 32.9 months, respectively. The 5-year mortality rate was 15.9%, and the median age of death 57.26 years (IQR 47.78, 68.08).

5-year mortality

Increasing age, urban residence, prior hospitalisation and comorbid asthma were all significantly associated with

5-year mortality ($P < 0.018$ for all) (Table S2). Other parameters, which met criteria for inclusion in the IBAS but were not statistically significant included smoking history ($P = 0.058$), right lower lobe (RLL) involvement on CT ($P = 0.083$), cylindrical/tubular type bronchiectasis ($P = 0.098$), a history of *Pseudomonas* spp. ($P = 0.092$) and comorbid systemic hypertension (HTN) ($P = 0.054$).

5-year hospitalisation

Prior hospitalisations and comorbid COPD were significantly associated with 5-year hospitalisation ($P = 0.001$ and $P = 0.014$, respectively) (Table S2). Other parameters, which met criteria for inclusion in the IBAS but were not statistically significant, included age ($P = 0.076$), residence location ($P = 0.084$), RLL involvement on CT ($P = 0.174$), a history of *Haemophilus* spp. ($P = 0.060$), *Pseudomonas* spp. ($P = 0.060$), *yeast* spp. ($P = 0.159$), *Streptococcus* spp. ($P = 0.187$), *Moraxella* spp. ($P = 0.087$) and comorbid asthma ($P = 0.070$).

Therefore, for the IBAS development, we included age category, residence location, smoking history, RLL involvement, cylindrical/tubular bronchiectasis, previous hospitalisations, history of *Haemophilus* spp., *Pseudomonas* spp., *yeast* spp., *Streptococcus* spp., *Moraxella* spp. and comorbid asthma, COPD or HTN. We additionally included FEV_1 and FVC based on previous research demonstrating the importance, despite the lack of significance in this study. Table S2 shows the full list of parameters assessed and their impact on mortality and hospitalisations within the development cohort.

Second stage of IBAS model construction

In the univariate Cox regression model, the following parameters were associated with significantly increased HRs for 5-year mortality (Table 1): urban residence, a history of prior hospitalisations and comorbid asthma. A history of prior hospitalisations and comorbid COPD were associated with significantly increased HR for 5-year hospitalisations. The means of the βs for 5-year mortality and 5-year hospitalisations are shown in Table 1. Both cylindrical/tubular bronchiectasis on CT and *Streptococcus* spp. were excluded from the final model as the mean βs rounded to zero. The rounded βs for FVC were higher than those for FEV_1; thus, FVC was included in the final model in preference. The final scorings and contributions of each parameter to the IBAS are detailed in Table 2. For ease of use, we doubled the score assigned to each parameter, so that only whole numbers were used.

IBAS severity categorisation

ROC analysis of the total IBAS score showed an AUC of 0.777 (95% CI 0.701, 0.854) for 5-year mortality and 0.737 (95% CI 0.67, 0.805) for 5-year hospitalisations (Figure 1). Cut-off points were defined as mild (0 to 4) ($n = 78$, 34.4%), moderate (5 to 7) ($n = 111$, 48.9%) and severe (≥8) ($n = 38$, 16.7%). The AUC for the total score and the categorised scores was equal for 5-year mortality; however, for 5-year hospitalisation, the categorised scores were lower, although not statistically significantly so.

IBAS validation

In the validation cohort, 97 patients (42.7%) were categorised as mild, 76 (33.5%) as moderate and 54 (23.8%) as severe via the total IBAS score. The total IBAS score was significantly associated with 5-year mortality (HR 1.55 (95% CI 1.22, 1.96)) and 5-year hospitalisation (HR 1.28 (95% CI 1.15, 1.42)) (Table 3). The AUC for 5-year mortality of the total IBAS score was 0.711 (95% CI 0.617, 0.804) and for 5-year hospitalisation was 0.774 (95% CI 0.714, 0.835). Patients categorised as severe had a significantly higher HR for 5-year mortality compared to those categorised as mild (HR 4.26 (95% CI 1.62, 11.21)), but not those categorised as moderate. The AUC for 5-year mortality of the categorised score was 0.664 (95% CI 0.567, 0.762) and for 5-year hospitalisation was 0.741 (95% CI 0.682, 0.8).

In the combined cohort, for both 5-year mortality and hospitalisations, patients categorised as moderate revealed significantly increased HRs compared to those categorised as mild, and participants categorised as severe showed significantly increased HRs compared to both mild and moderate patients (Table 3, Figure 2). As can be viewed in Figure 2, there were marked differences in survival rates over time in terms of both hospitalisation and mortality between patients categorised as mild, moderate and severe, with these differences becoming most evident from approximately 20 months onwards. The AUC for the total score on 5-year mortality was 0.743 (95% CI 0.683, 0.803) and for hospitalisations 0.759 (95% CI 0.714, 0.804). Using the categorised score these were 0.714 (95% CI 0.653, 0.775) and 0.714 (95% CI 0.7, 0.758), respectively.

Discussion

Our proposed IBAS assessment tool demonstrates high accuracy and delineation of mild, moderate and severe categorisation. The authors believe this is of use in day-to-day clinical practice to estimate bronchiectasis

Table 1 Hazard ratios (HRs) and β coefficients from univariate Cox regression on 5-year mortality and hospitalisation outcomes for each of the considered predictors within the construction cohort

Clinical parameters	Unit/Category	5-year mortality HR ($n = 227$)		5-year hospitalisation ($n = 227$)		β mean	Rounded score
		HR (95% CI)	β (95% CI)	HR (95% CI)	β (95% CI)		
Age category	30.1 < 50	1.88 (0.24, 14.55)	0.63 (−1.42, 2.68)	1.54 (0.74, 3.2)	0.43 (−0.30, 1.16)	0.53	0.5
(<30 reference)	50.1 < 70	3.35 (0.45, 25.06)	1.21 (−0.80, 3.22)	1.66 (0.80, 3.46)	0.51 (−0.22, 1.24)	0.86	1
	>70.1	5.82 (0.68, 49.85)	1.76 (−0.39, 3.91)	2.33 (0.98, 5.54)	0.84 (−0.02, 1.71)	1.3	1.5
Residence	Urban	2.89 (1.20, 6.95)*	1.06 (0.18, 1.94)*	1.62 (0.94, 2.80)	0.48 (−0.06, 1.03)	0.77	1
FEV_1 (% predicted)	<50%	0.93 (0.13, 6.62)	−0.07 (−2.03, 1.89)	1.29 (0.70, 2.37)	0.26 (−0.35, 0.86)	0.1	0
	<30%	2.15 (0.39, 11.75)	0.77 (−0.93, 2.46)	1.31 (0.70, 2.46)	0.27 (−0.36, 0.90)	0.52	0.5
FVC (% predicted)	<50%	1.89 (0.42, 8.43)	0.63 (−0.86, 2.13)	1.05 (0.63, 1.77)	0.05 (−0.46, 0.57)	0.34	0.5
	<30%	3.07 (0.32, 29.54)	1.12 (−1.14, 3.39)	1.55 (0.61, 3.97)	0.44 (−0.50, 1.38)	0.78	1
CT chest – Right lower lobe	Yes	0.75 (0.11, 5.33)	0.67 (−0.09, 1.42)	1.25 (0.91, 1.72)	0.22 (−0.10, 0.54)	0.45	0.5
Cylindrical/tubular bronchiectasis	Yes	0.37 (0.11, 1.2)	−1.00 (−2.18, 0.19)	0.89 (0.6, 1.32)	−0.12 (−0.51, 0.28)	−0.56	0
Haemophilus spp.	Yes	1.95 (0.92, 4.14)	0.24 (−0.49, 0.97)	1.38 (0.99, 1.93)	0.32 (−0.01, 0.66)	0.28	0.5
Pseudomonas spp.	Yes	0.38 (0.09, 1.57)	0.63 (−0.10, 1.36)	1.43 (0.98, 2.08)	0.36 (−0.02, 0.73)	0.5	0.5
Yeast spp.	Yes	1.27 (0.61, 2.64)	0.41 (−0.30, 1.12)	1.28 (0.91, 1.82)	0.25 (−0.10, 0.60)	0.33	0.5
Streptococcus spp.	Yes	1.87 (0.90, 3.88)	−0.05 (−1.00, 0.89)	1.31 (0.88, 1.94)	0.27 (−0.13, 0.66)	0.11	0
Moraxella spp.	Yes	1.5 (0.74, 3.05)	0.54 (−0.29, 1.36)	1.43 (0.95, 2.17)	0.36 (−0.05, 0.77)	0.45	0.5
Prior respiratory	1 admission	0.93 (0.27, 3.2)	−0.08 (−1.32, 1.16)	1.39 (0.92, 2.11)	0.33 (−0.09, 0.74)	0.13	0
hospitalisations	2 admissions	3.86 (1.57, 9.47)*	1.35 (0.45, 2.25)*	1.79 (1.09, 2.92)*	0.58 (0.09, 1.07)*	0.97	1
(No admissions reference)	≥3 admissions	7.11 (3.26, 15.52)*	1.96 (1.18, 2.74)*	2.82 (1.76, 4.53)*	1.04 (0.56, 1.51)*	1.5	1.5
COPD	yes	1.39 (0.49, 3.94)	0.33 (−0.71, 1.37)	1.85 (1.07, 3.19)*	0.61 (0.07, 1.16)*	0.47	0.5
Asthma	yes	2.3 (1.19, 4.47)*	0.83 (0.17, 1.50)*	1.37 (0.97, 1.92)	0.31 (−0.03, 0.65)	0.57	0.5
Systemic arterial hypertension	yes	2.16 (0.99, 4.75)	0.77 (−0.01, 1.56)	1.17 (0.85, 1.62)	0.16 (−0.16, 0.48)	0.47	0.5
Maximum score							9

*Significance at $P < 0.05$.

CI, confidence interval; COPD, chronic obstructive pulmonary disease; CT, computed tomography; FEV_1, forced expiratory volume in one second; FVC, forced vital capacity; HR, hazard ratio; spp., species.

severity, as a risk stratification tool to predict future morbidity and mortality among Indigenous patients with bronchiectasis. Worldwide, Indigenous people have a higher burden of chronic respiratory disorders, including bronchiectasis.[20,21] In addition to geographical isolation, inequitable access to specialist heath care, and the presence of multimorbidity perpetuate higher hospitalisation rates and mortality at a much younger age in Indigenous people.[23] There is a lack of disease assessment tools specifically designed to address diagnosis and management strategies for Indigenous people, despite many decades of research demonstrating a significant disease burden among the Indigenous population.[24] Utilising tools derived from non-Indigenous populations in the Indigenous population may explain the ongoing observed health disparity between the populations.[25] For this reason, it is imperative to develop appropriate normative reference ranges and management tools tailored to the Indigenous populations. In comparison to existing tools, the IBAS displayed an AUC for 5-year mortality of 0.714 (95% CI 0.653, 0.775) compared to the AUCs of the BSI and FACED on 4-year mortality of 0.703 (95% CI 0.636, 0.770) and 0.515 (95% CI 0.454, 0.575), respectively.[9] For 5-year hospitalisation the IBAS AUC was 0.714 (95% CI 0.7, 0.758), while for 4-year hospitalisations the BSI and FACED AUCs were 0.788 (95% CI 0.750, 0.825) and 0.559 (95% CI 0.524, 0.594), respectively.[9]

For many Indigenous patients, bronchiectasis is a lifelong disease, presenting from early childhood on the background of high early childhood respiratory infections, poor access to healthcare and worsened with an often-transient remote health workforce.[26] Due to this early onset of disease among Indigenous people, the median age of all-cause mortality is observed to be around 60 years.[4,12] The application of non-Indigenous reference ranges, defining patients as 'at risk' only if they are over or less than 70 years of age, as in the FACED assessment tool, would spuriously underestimate the severity of bronchiectasis. In our study, we have utilised significantly younger thresholds and included more accurate age categories in Indigenous patients for

Table 2 Parameters included in Indigenous bronchiectasis assessment scale (IBAS) and their contribution to the total score

Clinical parameters	Clinical threshold	IBAS score
Age range (years)	30.1 < 50	1
	50.1 < 70	2
	>70.1	3
Residence	Urban	2
Forced vital capacity (FVC) (% predicted)	30% – 50%	1
	<30%	2
Computed tomography (CT) location of bronchiectasis	Right lower lobe	1
Positive micro-organism cultured	*Haemophilus* spp.	1
	Pseudomonas spp.	1
	Yeast spp.	1
	Moraxella spp.	1
Previous hospitalisation history in the 2 years prior to index year	2 admissions	2
	≥3 admissions	3
Comorbidities	Chronic obstructive pulmonary disease (COPD)	1
	Asthma	1
	Systemic arterial hypertension (HTN)	1
Maximum score		18

risk stratification. Utilising this methodology there is already an increase in both mortality and hospitalisations in the 30- to 50-year age category (11.3 and 73.2%, respectively) compared to the under 30 years age category (6.3 and 50%, respectively).

In the proposed IBAS tool, the FVC% predicted values with a threshold at 30% or more showed higher utility than the FEV_1 parameter utilised in the FACED and BSI tools. This may be an artefact of the reduced proportion of our patients who had LFPs available, and indeed, the FVC HRs did show wider confidence intervals than FEV_1 for both mortality and hospitalisations. However, among adult Indigenous people, a restrictive impairment, with lower FVC values, appears to be the predominant lung function impairment observed.[27] There is ongoing debate and controversy regarding the ideal LFT parameter that could be utilised among patients with bronchiectasis.[28,29] Although LFT parameters did not demonstrate significant correlations to outcomes in this study, inclusion of LFT parameters in the bronchiectasis assessment among Indigenous populations would be useful. As such, LFT parameters may have greater implications in the overall disease course, especially in the presence of

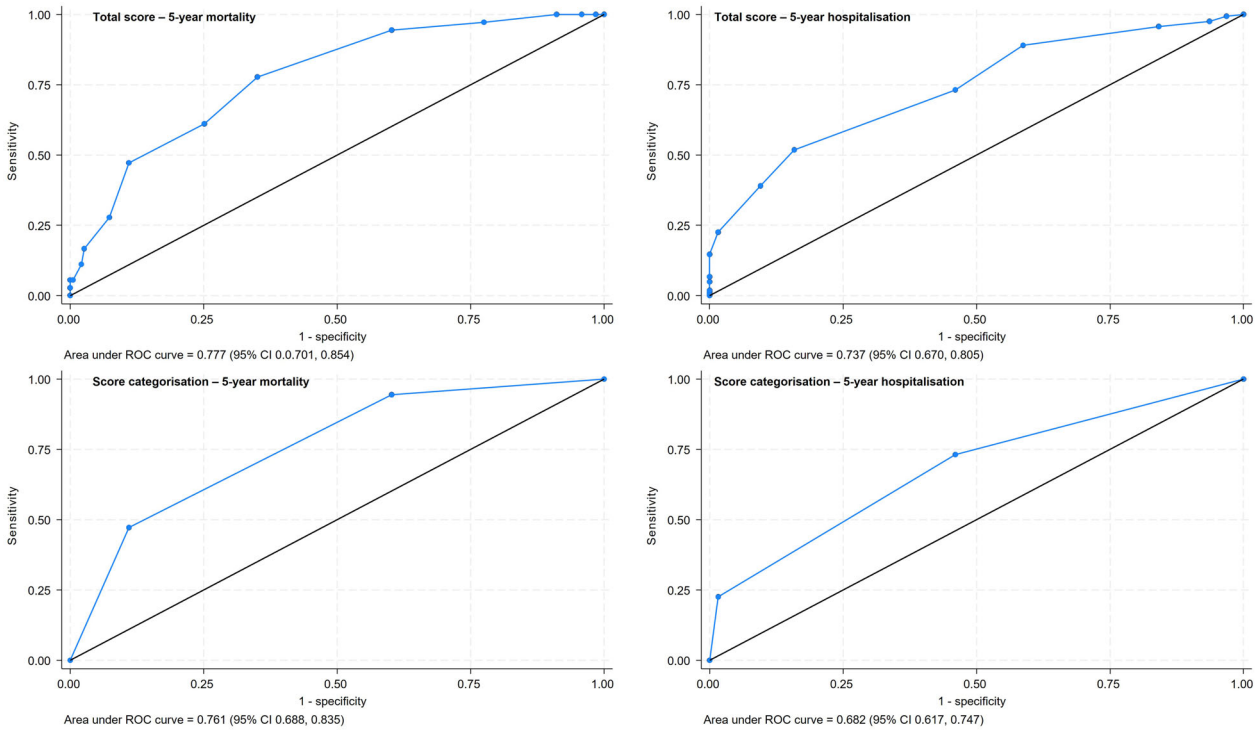

Figure 1 AUC ROC curves for continuous and categorised severity scores of IBAS total score for 5-year mortality (left) and 5-year hospitalisation (right). AUC, area under the curve; IBAS, Indigenous bronchiectasis assessment scale; ROC, receiver-operating characteristic curve.

Table 3 Validation of Indigenous bronchiectasis assessment scale (IBAS) scoring system within construction, validation and combined cohorts

	Construction ($n = 227$)		Validation ($n = 227$)		Combined ($n = 454$)	
	5-year mortality	5-year hospitalisation	5-year mortality	5-year hospitalisation	5-year mortality	5-year hospitalisation
Total score						
IBAS total score	2.05 (1.60, 2.61)*	1.41 (1.24, 1.61)*	1.55 (1.22, 1.96)*	1.28 (1.15, 1.42)*	1.74 (1.47, 2.06)*	1.33 (1.22, 1.44)*
IBAS moderate vs mild	6.48 (1.50, 28.04)*	1.51 (1.05, 2.18)*	2.40 (0.89, 6.5)	1.66 (1.14, 2.42)*	3.45 (1.57, 7.58)*	1.58 (1.22, 2.05)*
IBAS severe vs mild	22.92 (5.30, 99.30)*	2.89 (1.87, 4.48)*	4.26 (1.62, 11.21)*	2.26 (1.53, 3.33)*	8.40 (3.85, 18.32)*	2.47 (1.85, 3.30)*
IBAS severe vs moderate	3.54 (1.80, 6.94)*	1.91 (1.30, 2.82)*	1.77 (0.79, 3.96)	1.36 (0.94, 1.97)	2.43 (1.45, 4.07)*	1.56 (1.20, 2.04)*

*Significance at $P < 0.05$. Data displayed as HR (95% CI).

CI, confidence interval; HR, hazard ratio; IBAS, Indigenous bronchiectasis assessment scale.

multimorbidity, more specifically for FVC values.[30] It is therefore reasonable and appropriate to use FVC spirometry values for bronchiectasis severity assessment in Indigenous patients until and unless future studies indicate otherwise.

There is minimal published evidence detailing the radiological characteristics of bronchiectasis among Indigenous patients.[31] However, with the available evidence, bilateral lower lobe involvement appears to be common,[32] and indeed, in our study we observed that factoring in RLL involvement on CT improved the predictive capacity for bronchiectasis severity. Previous literature demonstrated cystic-type bronchiectasis and greater radiological extent to be associated with greater and earlier mortality, and indeed, cystic bronchiectasis is considered within the BSI.[8]

Similarly, to the radiological evidence, there is limited literature on the microbiological profile of bronchiectasis among Indigenous populations, including Australian Indigenous populations. Previous research has noted geographic differences in the microbiological profile in Indigenous patients.[12] In our study context, *Haemophilus* spp., *Pseudomonas* spp., yeast spp. and *Moraxella* spp. were observed to be the most cultured organisms. Furthermore, yeast species have been associated with increased hospitalisation rates in this study cohort, in addition to *Pseudomonas* spp.[33,34] In the current study, each of *Moraxella*, *Pseudomonas*, yeast and *Haemophilus* were given equal weighting in this IBAS tool, due to similar effects upon mortality and hospitalisations, instead of more importance to *Pseudomonas* as in the FACED and BSI tools.

The incidence of hospital admission rates, especially due to exacerbation of airway disease, including bronchiectasis, is significantly higher for Indigenous patients.[35] The IBAS tool showed two or more admissions in the past 2 years to be a reliable parameter to assess the severity of bronchiectasis. However, in contrast to the BSI tool, the impact of prior hospitalisations upon future outcomes was not nearly as strong. This may be

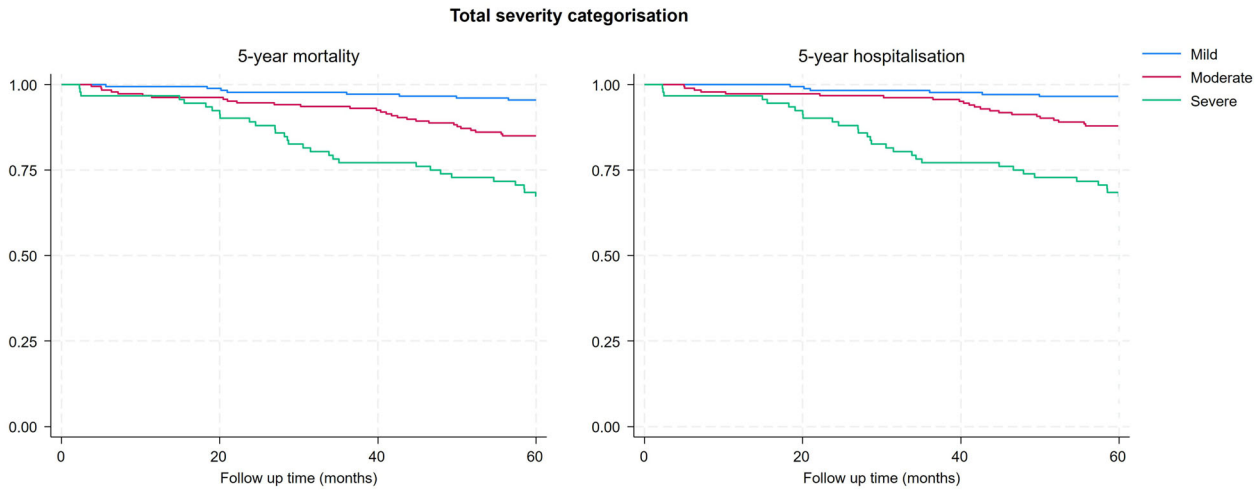

Figure 2 Kaplan–Meier survival graphs for the combined cohorts showing the survival rate until mortality or hospitalisation over 5 years by the Indigenous bronchiectasis assessment scale severity category.

due to the definition used in this study, which included any respiratory-related hospitalisation, whereas the BSI defines only hospitalisation related to significant exacerbation of bronchiectasis. However, among Indigenous patients, alongside bronchiectasis, multimorbidities, including the presence of concurrent COPD, are highly prevalent.[5,36] In non-Indigenous patients with bronchiectasis, it may be reasonable not to factor in other medical comorbidities as in both FACED and BSI tools. However, among Indigenous patients, it is hard to differentiate whether bronchiectasis *per se* is driving higher overall morbidity and mortality or the concurrent presence of other significant medical comorbidities. Hence, it is paramount that this factor be considered. The proposed IBAS tool showed COPD, asthma and the presence of systemic arterial hypertension were important other medical comorbidities to be considered.

A notable omission from the current study and proposed IBAS is the lack of symptom assessment, such as dyspnoea, as utilised in FACED and BSI tools. Despite dyspnoea being one of the common respiratory symptoms, including among Indigenous patients with airway disease, such as bronchiectasis,[13] the utility and validity of the Medical Research Council dyspnoea scale, which is adopted in both the FACED and BSI tools, have never been assessed or validated within an Indigenous population, to the best of the authors' knowledge. Therefore, it may not be applicable to Indigenous populations due to the absence of reliability assessment and validation. Indeed, dyspnoea could be related to the concurrent presence of COPD, asthma or frequent exposure to landscape/bushfire smoke and other potential non-tobacco exposure among Indigenous people on the background of having bronchiectasis.[37,38] One other intriguing observation in this study was that urban-residing patients showed a higher risk compared to rural-residing patients. This is despite the fact that the vast majority of Indigenous people reside remotely and are known to have poorer access to health care with remote residence. In the authors' opinion, the higher risk observed among the urban patients in this study setting may be related to Indigenous patients relocating to urban areas permanently in order to have improved access to health care, more specifically in the case of having multiple comorbidities and needing frequent specialist assessment. We did not further explore the reason for the difference (urban vs remote), as it was beyond the scope of this study. Nonetheless, in other Indigenous populations settings, there may be differing urban and rural factors leading to different risks and results compared to what is seen in the current study, highlighting the need for local area and context expertise when assessing the severity of bronchiectasis.

This study has demonstrated that differing clinical parameters need to be considered in assessing bronchiectasis in an Indigenous population context. The authors do acknowledge that one of the drawbacks to the proposed IBAS tool is the missing LFP data for a number of patients in the study cohort. Further, the CT scans were not specifically conducted to assess the severity of bronchiectasis, and the authors relied on the reporting radiologists' report for the presence and the location of bronchiectasis. However, it is imperative to acknowledge the inequity of access to health care, regardless of where Indigenous people reside across countries and continents. Access to investigations such as LFT/CT may not be viable – especially in remote and resource-poor settings – and clinicians may thus not have all the relevant data available for bronchiectasis assessment.[39] This means that clinicians must make the most of the limited data available and take up differing and innovative approaches in managing chronic respiratory disorders in these populations.[40]

In this vein, alongside the proposed IBAS tool, we propose a relative scoring system for when all clinical parameters are not readily available to measure the severity utilising the IBAS tool. We suggest that the relative score is calculated by dividing the total individual score by the total available score (i.e. if a patient scored 11 out of a possible 18 but did not have FVC data available (which in the IBAS is worth a maximum of 1 point), the potential maximum score would be only 17, thus $11/17 = 64.7\%$), as demonstrated in a previous report.[9] Indeed, the relative IBAS scoring system also showed high accuracy in the prediction of mortality and hospitalisations, similar to that of the 'total' IBAS score (Figures S1 and S2, Table S3), highlighting its utility in contexts where not all data are readily available. Nevertheless, our study fills a gap in non-biased comprehensive bronchiectasis assessment that corrects for the normative references established for non-Indigenous patients. Therefore, the IBAS scoring tool as illustrated in Table 4 and Data S1 details the proposed standard reference score and the absolute and relative score that could be considered while assessing bronchiectasis among adult Indigenous patients in day-to-day clinical practice. We hope this study will be of interest for clinicians and researchers caring for Indigenous people globally. Among other global Indigenous populations, certain demographics and social determinants may be similar to what is represented in this study, including comorbidities, geographical isolation and limited access to advanced specialist health care. However, the proposed IBAS tool could be improved upon or modified to suit/fit the local Indigenous populations' clinical profile/settings. Therefore, further studies are warranted to assess the applicability and suitability of this IBAS tool in the wider Indigenous populations in Australia and globally.

Table 4 Indigenous bronchiectasis assessment scale (IBAS)

Clinical parameters	Clinical threshold	IBAS standard reference score	Patients absolute score	Patients relative score
Age	30.1 < 50 years	1		
	50.1 < 70 years	2		
	>70.1 years	3		
Residence	Urban residence	2		
Forced vital capacity (FVC) (% predicted)	FVC 30% – ≥ 50%	1		
	FVC < 30%	2		
Computed tomography (CT) location of bronchiectasis	Right lower lobe	1		
Positive micro-organism cultured	*Haemophilus* spp.	1		
	Pseudomonas spp.	1		
	Yeast spp.	1		
	Moraxella spp.	1		
Previous respiratory condition hospitalisations	2 admissions in past 2 years	2		
	≥3 admissions in past 2 years	3		
Comorbidities	Chronic obstructive pulmonary disease (COPD)	1		
	Asthma	1		
	Systemic arterial hypertension (HTN)	1		
	Total score	18		

Absolute score indicates if all clinical parameters are available against the standard reference score. Relative score is calculated by dividing the total individual score by their total available score if some clinical parameters are not available. Severity category: mild (0–4), moderate (5–7) and severe (≥8).

Limitations

The study cohort to develop the IBAS tool included only adult Indigenous Australian patients residing in the TEHS region of the NT of Australia; hence, the utility of the IBAS tool and its applicability to wider Indigenous Australian populations or to Indigenous people globally is not known. Moreover, since this was a retrospective study, there could have been some bias in the outcomes, as not all parameters were available for all the study's patients, more specifically for LFPs. In the absence of any other similar studies in any other Indigenous populations, we were unable to compare our data to determine whether it was similar to other Indigenous patients or tools. Furthermore, the mortality data presented are all-cause related and, as such, we do not know whether the mortality was directly related to bronchiectasis. Nonetheless, this is the first study to develop a bronchiectasis assessment tool specifically for an adult Indigenous population, opening up avenues for prospective studies for the future.

Conclusion

This study has demonstrated that the proposed IBAS tool is of aid in assessing bronchiectasis severity in Indigenous patients. However, further prospective studies are warranted to assess the applicability and the utility in wider Indigenous populations globally.

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Supporting Information

Additional supporting information may be found in the online version of this article at the publisher's web-site:

Figure S1: AUC ROC curves for the continuous and categorised severity scores of the relative IBAS score for 5-year mortality (left) and 5-year hospitalisation (right). AUC, area under the curve; IBAS, indigenous bronchiectasis assessment scale; ROC, receiver-operating characteristic curve.
Figure S2: Kaplan–Meier survival graphs for combined cohorts showing survival rate until mortality or hospitalisation over 5 years by relative Indigenous bronchiectasis assessment scale severity category.
Data S1: Indigenous bronchiectasis assessment scale (IBAS).
Table S1: Demographic and clinical details.
Table S2: Influence of potential predictor parameters on primary and secondary outcomes within construction cohort via Kruskal–Wallis rank-sum test or chi-squared test.
Table S3: Validation of Indigenous bronchiectasis assessment scale (IBAS) relative scoring system within construction, validation and combined cohorts.