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# Demographic, imaging and disease patterns of moyamoya angiopathy in South Auckland: an observational study from electronic health records

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**Key words**
moyamoya, prevalence, Pacific, Māori, prognosis.

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## Abstract

**Background:** Prior reports show the occurrence of moyamoya angiopathy in the ethnically diverse South Auckland region, but little is known about the sociodemographic burden of the condition and clinical outcomes.
**Aims:** To determine the disease prevalence, age, sex and ethnic distribution, management and patient outcomes in adults residing in South Auckland (population 567 000). We also sought to determine associations with the outcomes of vascular events and functional independence following diagnosis.
**Methods:** We searched hospital medical records and radiology reports dating from 2008 to 2024 for the region using relevant ICD codes and the word 'moyamoya'. We used descriptive statistics and local population data to calculate point prevalence as of 19 September 2024. Statistical tests of association were performed for continuous, categorical and time-to-event data for the prespecified outcomes.
**Results:** We identified 49 patients with a total of 300 patient-years of follow-up. The point prevalence ranged from 4/100 000 in NZ Europeans and Asian people to 10/100 000 in Māori and 14/100 000 in Pacific Islanders. The mean age at diagnosis was 37 (standard deviation 16) years and females outnumbered males by 3:1. One-quarter had a vascular event within a median of 457 days from diagnosis and two-thirds of the overall cohort were independent on follow-up. Factors associated with a loss of independence included stroke/transient ischaemic attack (TIA) during follow-up, bilateral disease and severe hypoperfusion on imaging. The mortality rate was 14%.
**Conclusions:** In this New Zealand-based study, we found a wide ethnic variation in the prevalence of moyamoya angiopathy and an elevated early risk of TIA and stroke following diagnosis.

## Background

Moyamoya, the Japanese term for 'puff of smoke', refers to the angiographic appearances caused by stenosis of the supraclinoid internal carotid and circle of Willis with the development of a compensatory network of basal collaterals.[1] The disease was initially thought to affect East Asians exclusively but is now known to occur worldwide; lower prevalence rates are observed in the Western hemisphere, including Europe and North America.[2] Moyamoya may be unilateral but is more commonly bilateral and affects females twice as often as males, in whom it also follows a more aggressive course.[3] Clinical manifestations include ischaemic stroke from occluded intracranial vessels, cerebral haemorrhage from the rupture of friable collateral vessels and seizures. It is considered a progressive disease with a continued risk of recurrent stroke.[3]

Digital subtraction angiography (DSA) is considered the gold standard for the diagnosis of moyamoya angiopathy; however, it is more commonly diagnosed on

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computed tomography angiography (CTA) or magnetic resonance angiography (MRA). Guidelines recommend advanced haemodynamic imaging techniques to assess cerebrovascular reserve capacity and the haemodynamic status of brain parenchyma. The Ring Finger Protein 213 (RNF213) is a susceptibility gene for moyamoya disease.[4] The R4810K variant is particularly prevalent in Japanese and Korean populations, while other RNF213 variants are more commonly linked in non–East Asian and some Chinese populations.[5] The role of mutations in RNF213 in moyamoya is complex and may be associated with varying clinical manifestations such as extracranial arteriopathy (e.g. p.Arg4810Lys).[6] Treatments for moyamoya are limited, and the use of antiplatelet agents is generally extrapolated from more common conditions such as atherosclerosis.

Moyamoya can be categorised as either moyamoya disease when idiopathic or moyamoya syndrome when secondary to other pathological processes, such as brain radiation, haematological or genetic conditions (e.g. neurofibromatosis, Down syndrome). They are equivalent in radiological and in clinical presentation.[1,7] The updated Research Committee on Moyamoya Disease Guidelines now includes atherosclerosis, hyperthyroidism and head trauma within the moyamoya disease category as aetiological factors.[5] In this study, we will use the term moyamoya angiopathy (MMA) when referring to moyamoya disease or moyamoya syndrome.

There are no data on the prevalence and management of MMA in New Zealand, and more specifically within South Auckland, despite there being recorded cases of the condition. Based on 2018 census population data, the resident population of South Auckland was estimated to be 567 000, representing 11.6% of New Zealand's population.[8] South Auckland is ethnically diverse, with 16% of the population identifying as Māori, 22% as Pacific peoples, 28% as Asian and 34% as New Zealand European or Other. This study aimed to determine the disease prevalence, age, sex and ethnic distribution, management and patient outcomes in those aged ≥16 years in the South Auckland region.

## Methods

We created a retrospective South Auckland moyamoya registry by searching for relevant admission codes ICD9/10 (437.5 and I67.5 respectively) at our institution and radiology reports and discharge letters for the word 'moyamoya' since 2008. Though New Zealand provides a universal public health system, we also searched reports of scans performed by local private radiology providers to maximise case ascertainment. Two investigators reviewed patient notes and images, and we sought advice from an independent radiologist to resolve any areas of disagreement. Imaging criteria for study inclusion required stenosis/occlusion of at least one terminal portion of the internal carotid artery and proximal branch of the Circle of Willis (e.g. M1 segment of the middle cerebral artery or A1 segment of the anterior cerebral artery) with an associated network of collateral (moyamoya) vessels demonstrated on brain imaging. While the initial use of the register was to retrospectively identify historical cases for the purpose of this study, it is now also available for the prospective collection of cases for future research.

The following information was collected from clinical records: demographic data (age at diagnosis, sex and ethnicity), vascular risk factors (e.g. hypertension, diabetes, hyperlipidaemia), moyamoya type (disease or syndrome), initial clinical presentation (e.g. transient ischaemic attack (TIA), ischaemic stroke, intracranial haemorrhage, seizures) and imaging findings. Co-morbidities were extracted from electronic clinical records, specifically from clinician-documented diagnoses in discharge summaries and outpatient letters, wherever available. Administrative codes and laboratory values were not used in isolation. We also collected follow-up data on vascular outcome events: TIA, ischaemic stroke and haemorrhagic stroke; multiple events were excluded to enable a time-to-first-recurrent-event analysis, which reduces complexity in modelling and aligns with standard practice in small retrospective cohorts. We also searched the most recent clinical records to find entries of suitable quality, which allowed us to determine the modified Rankin score. Data from all sources were collected at the individual level. We used self-reported ethnicity, and patients were allocated to a single ethnic group in accordance with the Ministry of Social Development hierarchy that aligns with Statistics NZ: Māori, Pacific Peoples, European, Asian, MELAA (Middle Eastern, Latin American and African) and Other.[9]

### Study timeline

Data were collected on subjects from January 2008, and follow-up outcome data were obtained from clinical records until 19 September 2024.

### Imaging data collection

We recorded the mode of neuroimaging performed (e.g. CTA, MRA, DSA and perfusion imaging), the location of intracranial arterial stenosis/occlusion and the presence of moyamoya collateral vessels. Bilateral disease describes the involvement of both internal carotid arteries. We also recorded the presence of linear high signal

intensities along cortical sulci (reflecting leptomeningeal collateral recruitment) on T2 MR fluid-attenuated inversion recovery (FLAIR) ('Ivy sign'). When present, cerebral hypoperfusion on MR or CT perfusion at the cortical level was considered severe if more than a 50% increase in Tmax (on CT) or mean transit time (on MR) was observed. Changes in follow-up imaging were also categorised as worsening, static or improved (based on angiographic or perfusion findings).

### Event definitions

Stroke was defined as clinical deficits lasting over 24 h or with evidence of acute changes on brain imaging (CT or MR) and categorised as ischaemic or haemorrhagic. Haemorrhage included intracerebral, intraventricular and subarachnoid haemorrhage. TIA was defined as neurological deficits of presumed vascular origin resolving within 24 h in the absence of acute changes on neuroimaging. Disability at follow-up was assessed using the Modified Rankin Scale (mRS), with a score of 0–2 representing functional independence.

### Clinical management

We recorded the date, type (direct or indirect) and side of the surgical revascularisation procedure. We also recorded the use of antithrombotic medication.

### Statistical analysis

Categorical data are presented as numbers and frequencies, and continuous data are summarised as means (standard deviation (SD)) for normally distributed or median (interquartile range (IQR)) for non-normally distributed data. Categorical data were analysed using the chi-squared test or Fisher's exact test as appropriate, and continuous data were analysed using a *t*-test or analysis of variance (ANOVA) if normally distributed or Mann–Whitney if non-normally distributed. Point prevalence rates were calculated using the number of MMA patients alive on 19 September 2024 as the numerator and the South Auckland population as the denominator (567 000) and presented using a binomial (exact) confidence interval. Ethnicity-specific prevalence rates were calculated using 2018 data provided to Te Whatu Ora (Health NZ) by Stats NZ, based on the prioritised ethnicity framework. Although 2023 Census data are available, they report total response ethnicity, which is not directly comparable. Time-to-event data were analysed by Cox proportional regression analysis to test for associations; censoring occurred at the time of death or the last documentation, which allowed a mRS to be calculated. Differences are shown as hazard rate (HR) and 95% confidence interval (CI). Multivariable models were used to adjust for potential confounders using prior clinical knowledge, clinically relevant variables and those with a *P*-value ≤0.2. The proportional hazards assumption was assessed for both univariate and multivariate Cox models using Schoenfeld residuals, and graphical methods were used for categorical covariates. We used complete case analysis, and a *P*-level of ≤5% was considered statistically significant.

### Bias minimisation

Since the researchers were unblinded to the diagnosis, to minimise information bias, we collected clinical data from electronic medical records, prioritising documentation written by treating clinicians, such as discharge summaries and clinic notes. We minimised measurement bias in image interpretation using radiology reports first and secondly our combined observations. Where doubt persisted, we requested the opinion of an independent radiologist with an interest in neuroimaging. We attempted to minimise selection bias using the same search strategy across multiple different sources: hospital records and public and private radiology services.

### Standard protocol approvals, registration and patient consent

This was a low-risk observational study, using patient data routinely collected during routine clinical care. We were granted a waiver for obtaining informed consent, and this study was approved by the Health and Disability Ethics Committee (ref: 2024 EXP 20125). This manuscript was prepared according to the Strengthening the Reporting of Observational Studies in Epidemiology (STROBE) statements.[10] Statistical analysis was performed using Stata BE/17.

## Results

We identified 190 potentially eligible cases. This reduced to 52 after removing duplicates, patients under 16 and patients not residing in the study district. Following a formal review of clinical notes and images three non-moyamoya cases were removed, leaving 49 patients with confirmed MMA who were included in this study. We had 300 patient-years of follow-up with a median follow-up time of 3.5 years (range 20 days to 25 years, IQR 1–9 years).

Based on local population data, the point prevalence (95% CI) in South Auckland is 9/100 000 (6 to 11/100 000), with the following ethnicity distribution: NZ

Europeans/Others 4/100 000 (2 to 8/100 000), Māori 10/100 000 (5 to 19/100 000), Pacific peoples 14/100 000 (9 to 23/100 000) and Asians 4/100 000 (2 to 9/100 000). Baseline characteristics are highlighted in Table 1.

Moyamoya syndrome was associated with meningitis, brain irradiation and neurofibromatosis.

There are reports of extracranial internal carotid artery involvement in MMA in the literature; we observed this in one-quarter of our cohort.[11] Though this study is underpowered for subgroup analysis, there was a trend towards more extracranial carotid involvement in Māori and Pacific peoples (e.g. Fig. 1) and family history in Asian and Pacific peoples ($P = 0.13$ and $P = 0.08$ respectively).

Of patients with perfusion imaging, half had features of severe hypoperfusion, an example demonstrated in Figure 2 (cerebral blood volume (CBV) and CTP tmax). Other imaging findings are reported in Table 1. Ivy sign, a radiological finding of slow flow in pial vessels, was seen in 27 patients: In two patients, it reduced following surgical revascularisation, a finding described in other studies.[12] Twenty-two (45%) patients were referred for neurosurgery, and revascularisation was performed in one-third of these.

Half of the patients had follow-up angiographic or perfusion imaging. Of these, changes were static in one-third, progressive in approximately 40% and improved in one-quarter. Of those with improved follow-up imaging, this was after a thrombectomy (for acute thrombotic intracranial occlusion) in one patient, improved spontaneous collateral flow in one, post-surgical changes in two and unknown in two. Findings are described in Table 2.

One-quarter of our cohort had follow-up vascular events, predominantly TIA or ischaemic stroke (85%, Table 2), at a median of 457 days (IQR 109–1639) from diagnosis. Beyond this median time point, six further recurrent vascular events were observed over 10 years. At follow-up (median of 3.5 years from diagnosis), almost two-thirds were independent for activities of daily living (mRS 0–2) and seven (14%) had died. Table 2 summarises the interventions and outcomes.

**Table 1** Baseline characteristics

| Baseline characteristics | Number (%) $n = 49$ |
|---|---|
| Male | 13 (26.5) |
| Female | 36 (73.5) |
| Age at diagnosis (SD), years | 37 (16) |
| Ethnicity | |
| European | 8 (16.3) |
| Māori | 11 (22.4) |
| Pacific Islander | 20 (40.8) |
| East Asian (e.g. Taiwan, China, Korea) | 8 (16.3) |
| Central/South Asian (e.g. India) | 2 (4.1) |
| Family history of moyamoya | 7 (14.9) |
| Moyamoya disease | 46 (93.9) |
| Diabetes | 19 (38.8) |
| Hyperlipidaemia | 27 (55.1) |
| Hypertension | 30 (61.2) |
| Smoking | 24 (49) |
| Migraine | 11 (22.4) |
| Presenting symptoms | |
| TIA | 10 (20.4) |
| Ischaemic stroke | 22 (44.9) |
| Haemorrhagic stroke | 6 (12.3) |
| Seizure | 6 (12.3) |
| Headache/migraine | 4 (8.2) |
| Imaging performed | |
| CTA | 37 (75.5) |
| MRA | 41 (83.7) |
| DSA | 14 (28.6) |
| MR/CT perfusion | 25 (51) |
| Imaging findings | |
| Extracranial internal carotid artery stenosis | 15 (28.6) |
| Unilateral disease | 15 (28.6) |
| Ivy sign present (MRI) | 27 (65.9) |
| Hypoperfusion (on perfusion imaging) | 24 (96) |
| Severe hypoperfusion | 11 (44) |
| Antithrombotic treatment | |
| Aspirin | 29 (59.2) |
| Clopidogrel | 9 (18.4) |
| Aspirin + clopidogrel | 1 (2) |
| Warfarin | 3 (6.1) |
| Nil | 7 (14.3) |

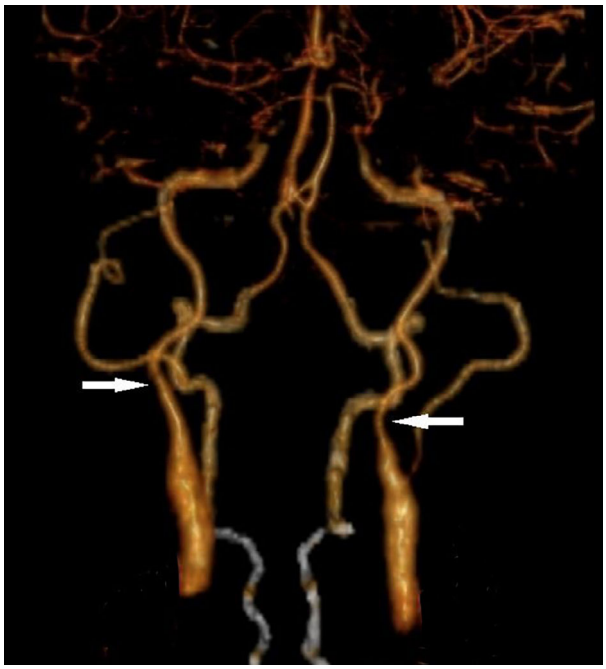

**Figure 1** Computed tomography angiography demonstrating bilateral proximal internal carotid artery stenosis.

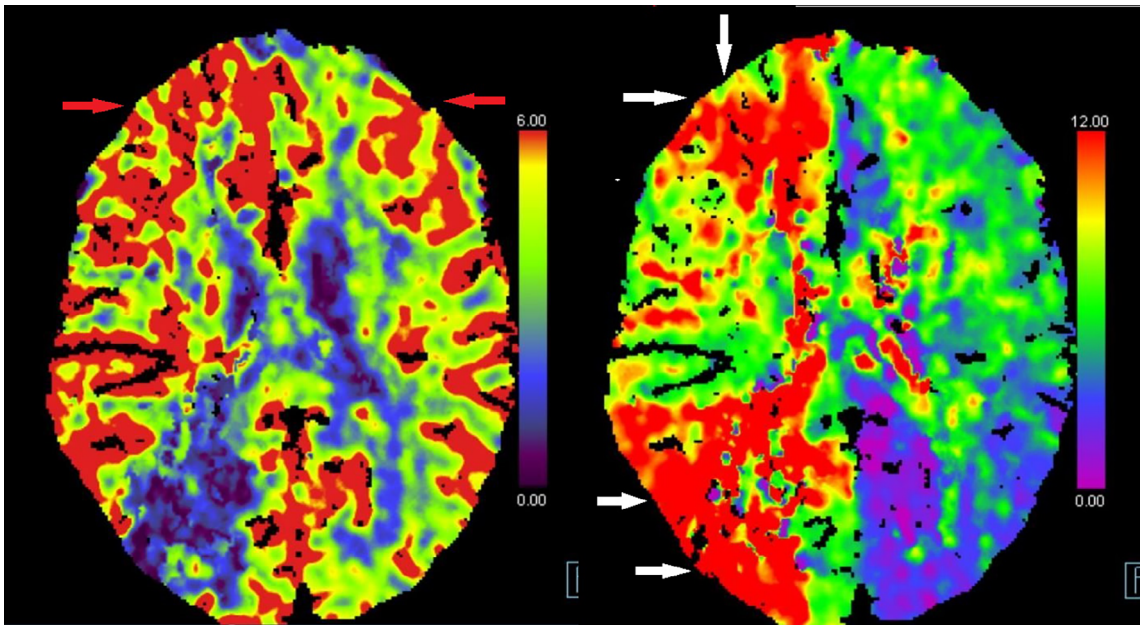


**Figure 2** Computed tomography perfusion imaging. Left: Cerebral blood volume demonstrating increased blood volume in the right frontal cortex compared with the left frontal cortex, caused by the recruitment of leptomeningeal collaterals (red arrows). Right: Tmax showing severe hypoperfusion in the right frontal and parietal lobes (white arrows) and a mild degree of hypoperfusion in the left frontal lobe. This patient was severely disabled (mRS 5) on follow-up.

**Table 2** Interventions and outcomes

| Outcome/Intervention | Number (%) $n = 49$ |
|---|---|
| Reperfusion therapy | 1 (2) |
| Surgery | 7 (14.3) |
| EDAS† | 3 (42.9) |
| STA-MCA bypass‡ | 2 (28.6) |
| STA-MCA bypass + EDAS†,‡ | 2 (28.6) |
| Recurrent stroke/TIA on follow-up | 13 (26.5) |
| TIA | 5 (38.5) |
| Ischaemic stroke | 6 (46.2) |
| Cerebral haemorrhage | 2 (15.4) |
| Final mRS | |
| 0–2 | 31 (63.7) |
| 3–5 | 11 (22) |
| 6 | 7 (14.3) |
| Follow-up imaging (angiography/perfusion) | 24 (49) |
| Improved | 6 (25) |
| Static | 8 (33.3) |
| Worsening | 10 (41.7) |

†EDAS: Encephaloduroarteriosynangiosis.‡STA-MCA: Superficial temporal artery to middle cerebral artery bypass.

In the Cox regression univariate analysis, there was a trend towards fewer recurrent vascular events in males. For the outcome of functional independence (mRS 0–2), recurrent vascular events, severe hypoperfusion and bilateral disease were associated with a significantly lower likelihood of remaining functionally independent. There was a trend towards improved outcomes with the use of antithrombotics. Detailed findings from the univariate analysis are presented in Table 3.

In the multivariable regression model adjusting for potential confounders of age, sex and bilateral disease, only recurrent cerebrovascular events (HR: 0.32 (95% CI: 0.11–0.94), $P < 0.05$) were associated with a significantly lower likelihood of achieving functional independence. Harrell's c-statistic (a measure of the predictive accuracy of the risk model) was 0.72, indicating reasonable discrimination. The proportional hazards assumption was assessed for the univariate and multivariate models using the global Schoenfeld residuals, and no significant violation of the assumption was detected.

## Discussion

In this New Zealand-based study, we found a high burden of MMA in South Auckland, particularly in Māori (10/100 000) and Pacific peoples (14/100 000). While these rates are lower than those reported in Japan (17.6/100 000) and South Korea (16.1/100 000), this suggests an important burden of disease in New Zealand's indigenous and Pacific populations.[13] Our findings also highlight a relatively higher female-to-male ratio (3:1) compared to prior cohorts (2:1).[1]

**Table 3** Cox proportional hazard regression of variables associated with follow-up events and functional independence

| | Follow-up stroke/TIA $n = 13$ (26.6%) HR (95% CI) | *P*-value | Final mRS 0–2 $n = 31$ (63.7%) HR (95% CI) | *P*-value |
|---|---|---|---|---|
| Male sex | 0.18 (0.023–1.43) | 0.11 | 0.76 (0.31–1.85) | 0.54 |
| Age | 1.02 (0.98–1.05) | 0.42 | 1.02 (0.99–1.04) | 0.2 |
| Ethnicity | | | | |
| • NZ European | Reference | - | Reference | - |
| • Māori | 1.37 (0.25–7.51) | 0.72 | 1.12 (0.35–3.55) | 0.86 |
| • Pacific Islander | 0.96 (0.19–4.97) | 0.96 | 1.29 (0.49–3.42) | 0.6 |
| • Asian | 0.83 (0.16–5.92) | 0.85 | 1.1 (0.33–3.62 | 0.88 |
| Presenting complaint | | | | |
| • TIA | 0.43 (0.06–3.39) | 0.43 | 2.9 (1.32–6.38) | 0.01* |
| • Ischaemic stroke | 2.9 (0.78–10.9) | 0.11 | 1.16 (0.47–2.65) | 0.8 |
| • Haemorrhagic stroke | 1.1 (0.1–12.2) | 0.94 | 0.42 (0.08–2.06) | 0.28 |
| Vascular risk factors | | | | |
| • Diabetes | 1.84 (0.62–5.48) | 0.27 | 0.69 (0.30–1.57) | 0.38 |
| • Hypertension | 1.75 (0.53–5.78) | 0.36 | 0.69 (0.33–1.42) | 0.31 |
| • Hyperlipidaemia | 1.42 (0.46–4.34) | 0.54 | 1.34 (0.63–2.86) | 0.45 |
| • Smoking | 0.78 (0.26–2.34) | 0.65 | 0.72 (0.35–1.49) | 0.38 |
| • Stroke/TIA during follow-up | | - | 0.32 (0.12–0.84) | 0.02* |
| Imaging | | | | |
| • Severe perfusion mismatch | 1.84 (0.53–6.33) | 0.34 | 0.15 (0.03–0.69) | 0.01* |
| • Ivy sign | 1.47 (0.39–5.57) | 0.57 | 1.57 (0.63–3.92) | 0.33 |
| • Bilateral disease | 1.54 (0.34–7.02) | 0.58 | 0.39 (0.19–0.83) | 0.01* |
| • Extracranial internal carotid disease | 1.66 (0.54–5.1) | 0.38 | 0.64 (0.26–1.55) | 0.32 |
| • Progressive disease | 2.67 (0.89–8) | 0.08 | 0.73 (0.21–2.53) | 0.63 |
| • Number of vessels involved | 1.18 (0.83–1.69) | 0.35 | 0.83 (0.69–1.01) | 0.06 |
| Treatment | | | | |
| • Antithrombotics | 1.88 (0.41–8.63) | 0.41 | 2.9 (0.83–10.2) | 0.1 |
| • Surgery | 2.51 (0.77–8.24) | 0.13 | 0.73 (0.29–1.81) | 0.49 |

*$P < 0.05$.

We observed a trend towards higher rates of extracranial vessel involvement in Pacific peoples and Māori. Routine genetic testing could be pursued in the future to shed more light on this issue. Functional outcomes in our cohort were largely favourable, with two-thirds of patients remaining independent at follow-up. However, there was an early peak in cerebrovascular events, consistent with international cohorts from Denmark and China.[3,16] Unsurprisingly, follow-up vascular events were associated with a significantly lower likelihood of remaining functionally independent.

MMA is characterised by the progressive stenosis of intracranial vessels and compensatory formation of basal and leptomeningeal collaterals via the external carotid artery. As the steno-occlusive changes of the bilateral internal carotid arteries progress, moyamoya vessels eventually regress, and the entire cerebral hemisphere is perfused by the external carotid and vertebrobasilar artery systems.[14] Our follow-up vascular event and functional independence rates suggest that this physiological compensatory strategy is adequate in many individuals. However, the challenge lies in the early identification of individuals who would be most likely to benefit from surgery. MMA revascularisation is complex and associated with significant complications such as cerebral hyperperfusion syndrome (5.7%–16.5%) and graft occlusion (2.9%–6.4%).[15] While no harm was demonstrated, our sample was too small to draw firm conclusions around the benefits of revascularisation in the New Zealand setting.

Other findings also show that moyamoya does not progress linearly; there is a critical period in the first 2 years from diagnosis when patients are at higher risk of vascular events, presumably due to a mismatch between demand and supply as collaterals develop. Interestingly, in other observational studies of moyamoya patients from Denmark and China, the median time to vascular events from diagnosis was 2 and 3 years respectively.[3,16] If the patient remains event-free during this critical period, their disease course is likely to be more stable.

We found an association between severe hypoperfusion and loss of independence. Similar to our findings, previous studies found that cerebral hypoperfusion and reduced cerebrovascular reserve were associated with

clinical deterioration and also identified patients most likely to benefit from revascularisation.[17,18]

The strengths of this study include the individual review of patient-level data obtained from multiple sources, population-based data and long-term follow-up. We used strict inclusion criteria and a wide search strategy reducing selection bias. Although a single-centre study with relatively small numbers, the varied ethnicity in our study supports sufficient generalisability to inform an update of New Zealand MMA treatment recommendations incorporating broader consensus discussions between stroke specialists, neuroradiologists and neurosurgeons.

Limitations of our study include the relatively small sample size, which limits our ability to draw further associations, and inherent biases associated with the use of retrospective data. Records that could be searched for ICD codes were limited, and we relied on radiology reports, meaning variations of moyamoya (e.g. moya moya) or typos (e.g. m oyamoya) would not be detected, leading to potential selection bias and perhaps an underestimate of MMA in South Auckland. We were unable to access clinic letters from private clinicians, so follow-up stroke/TIA may also be underestimated. Our strategy for determining mRS at follow-up could introduce misclassification since follow-up assessments were inconsistent. Limitations of our statistical analysis included the potential for type I error inflation due to multiple statistical testing, though our analyses were hypothesis-driven based on prior literature (e.g. hypoperfusion, bilaterality), rather than exploratory data dredging. Further, the results in Table 3 should be interpreted with caution and viewed primarily as hypothesis generating. Finally, the relatively small sample size may mean non-significant findings represent type II error.

## Conclusion

We found a high burden of MMA across a range of ethnicities residing in South Auckland, with a particularly high prevalence in Māori and Pacific peoples. In line with other cohort studies, we found an elevated early risk of TIA and stroke following diagnosis.

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### Data availability statement

Anonymised data will be shared on reasonable request from researchers.

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