Scottish Intracranial Vascular Malformation Study (SIVMS) Evaluation of Methods, ICD-10 Coding, and Potential Sources of Bias in a Prospective, Population-Based Cohort

Rustam Al-Shahi, MA, MRCP(UK); Jo J. Bhattacharya, FRCR; David G. Currie, FRCS; Vakis Papanastassiou, FRCS; Vaughn Ritchie, MB, ChB; Richard C. Roberts, FRCP; Robin J. Sellar, FRCR; Charles P. Warlow, FRCP; for the SIVMS Collaborators

Background and Purpose—The rarity of intracranial vascular malformations (IVMs) and the infrequency of their outcomes make large, prolonged cohort studies the best means to evaluate their frequency and prognosis.

Methods—The Scottish Intracranial Vascular Malformation Study (SIVMS) is a prototype prospective, population-based study of adults resident in Scotland and diagnosed for the first time with an IVM after January 1, 1999. We evaluated the design of SIVMS using 2 complete years of data for adults with arteriovenous malformations (AVMs) of the brain.

Results—A collaborative network of clinicians, radiologists, and pathologists, combined with coding of hospital discharge data and death certificates, recruited a cohort distributed in proportion to the Scottish population. Coding (with International Classification of Diseases, 10th Revision [ICD-10] codes Q28.2 and I60.8) had a sensitivity of 72% (95% CI, 61% to 80%) and a positive predictive value of 46% (95% CI, 38% to 55%) for detecting incident brain AVMs. Adults who were detected by coding alone were significantly (<0.05) younger, more likely to present with hemorrhage, more frequently investigated with catheter angiography, and more likely to be treated. Adults recruited from tertiary referral centers were significantly more likely to be investigated with catheter angiography and to be treated. Using catheter angiography as a diagnostic requirement for brain AVMs significantly biases the cohort toward younger adults presenting with hemorrhage and receiving treatment.

Conclusions—Population-based studies of IVM frequency and prognosis should use multiple overlapping sources of case ascertainment, and such studies of brain AVMs should not require catheter angiography to be the diagnostic standard. (Stroke. 2003;34:1156-1162.)

Key Words: central nervous system ▪ cerebral arteriovenous malformations ▪ incidence ▪ prognosis ▪ registries ▪ vascular malformations

Intracranial vascular malformations (IVMs) are a morphologically and clinically heterogeneous grouping of arteriovenous malformations (AVMs) of the brain and dura, cavernous malformations (cavernomas), and venous malformations (developmental venous anomalies). IVM detection rates are rising over time, probably because of the widening availability and increasing use of noninvasive neuroimaging,1,2 such that they are now recognized to be the leading cause of spontaneous intracerebral hemorrhage in young adults.3

However, interest in using and reporting recent technological advances in both interventional and noninvasive treatments has largely overtaken the impetus to study other aspects of IVMs. There are few well-designed studies addressing frequency and prognosis;5,6 in fact, there is only one truly population-based study of frequency that was small and retrospective.1 Although there has been one randomized controlled trial comparing two embolic agents,5 there have been none comparing different treatment modalities against each other or placebo.6

Consequently, clinicians are frequently faced with various management dilemmas: what is the clinical course of IVMs, what is the prognosis for a particular person, and which treatment(s), if any, should be used?

There is therefore a pressing need for prospective, population-based observational cohort studies of IVMs.

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From the Department of Clinical Neurosciences, Western General Hospital, Edinburgh (R.A-S., R.J.S., C.P.W.); Institute of Neurological Sciences, Southern General Hospital, Glasgow (J.J.B., V.P.); Department of Neurosurgery, Aberdeen Royal Infirmary, Aberdeen (D.G.C.); Fauldhouse Health Centre, Fauldhouse (V.R.); and Department of Neurology, Ninewells Hospital and Medical School, Dundee (R.C.R.), Scotland.

Authors are the SIVMS Research Fellow (R.A-S.), Principal Investigator (C.P.W.), and the rest of the SIVMS Steering Committee (alphabetical order by surname). A complete list of the SIVMS Collaborators (previously and currently active) appears in the Appendix, which is available online at http://stroke.ahajournals.org.

Correspondence to Rustam Al-Shahi, MA, MRCP(UK), Department of Clinical Neurosciences, Western General Hospital, Edinburgh EH4 2XU, Scotland. E-mail Rustam.Al-Shahi@ed.ac.uk

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1156
These studies should share a core set of methodological standards to minimize both bias and confounding to an acceptable level, to make studies comparable, and to speed the evaluation of prognosis with the greatest worldwide economy of research effort by cautiously synthesizing their results.3,7,8 We herein describe the design of a prototypical study set up in 1998, the Scottish Intracranial Vascular Malformation Study (SIVMS), evaluate it using 2 complete years of data for adults with brain AVMs, and quantify detection rates in an accompanying article.9

Subjects and Methods

People are included in SIVMS if they have a first-in-a-lifetime diagnosis of an IVM, whether symptomatic or not, made on or after January 1, 1999, when they are aged ≥16 years and are permanently resident in Scotland.

Diagnostic Criteria

Because IVMs are not always easy to distinguish from each other even with complete radiological or pathological examination10 and systems for their nomenclature and taxonomy have varied, SIVMS uses operational definitions and allocates a measure of certainty (definite/probable/possible) to each IVM diagnosis. Certainty of pathological diagnosis is determined from pathologists’ reports of autopsies and specimens from surgical excision. The final decision about certainty of radiological diagnoses made on CT, MRI, or catheter angiography, as well as the collection of detailed angiographic data, is arbitrated at consensus meetings between 2 consultant interventional neuroradiologists (J.J.B. and R.J.S.).

Brain AVM

A brain AVM is an anastomosis of nonnutritive blood vessels in the brain parenchyma, in which arteriovenous shunting occurs in a central nidus (the tangle of vessels in which usually multiple arteries and veins converge). They include brain (pial) arteriovenous fistulae (a single communication between 1 feeding artery and 1 draining vein) but exclude pure vein of Galen malformations and dural AVMs. Definite diagnoses were made by pathological examination, CT, MRI, or catheter angiography (Figure 1A to 1C).

Dural AVM

A dural AVM is an arteriovenous shunt through single or multiple fistulae within the dura mater or cavernous sinus (ie, including carotid-cavernous fistula) (Figure 1D). Definite diagnoses were made by pathological examination or catheter angiography.

Cavernous Malformation

A cavernous malformation is a well-circumscribed, lobulated, densely clustered mass of small vascular channels without intervening brain parenchyma. Definite diagnoses were made by pathological or MRI examination, the most distinctive appearance being a reticulated core of mixed signal intensity with a surrounding capsule of decreased signal intensity on T2-weighted MRI (Figure 1E).11

Venous Malformation

A venous malformation is a group of aberrant dilated veins that have an anomalous course, definitely diagnosed on the basis of enhancement of a curvilinear transcerebral draining vein or caput medusae on
CT, MRI, catheter angiography, or pathological examination (Figure 1F).

Inception
In SIVMS, an IVM is “incident” or “detected” on the date of a first-in-a-lifetime definite diagnosis. Exceptionally, a person may harbor multiple IVMS, necessitating separate incident dates for each IVM. Whether the clinical presentation leading to a medical evaluation that prompted investigation and a subsequent incident diagnosis was symptomatic of the IVM or not, the date of this clinical presentation is the point of inception (when prospective follow-up starts).

Study Setting

Demographics of Scotland
Scotland’s population denominator is a manageable size for surveys of a disease with an incidence of 1 to 2 per 100,000 per year. The most recent decennial census in Scotland was conducted in 2001. Between censuses the General Register Office (www.gro-scotland.gov.uk) estimates the size and age structure of the Scottish population annually using registration of births and deaths as well as data on immigration and emigration. The mid-2000 estimate of the population of Scotland was 5,114,600, of whom 4,114,052 were adults aged ≥16 years. There was only a small (0.3%) annual flux in the population (immigration of 6,400 and emigration of 6,600 people in the 12 months before June 30, 2000).

The Scottish Health Service
Few patients receive healthcare outside the National Health Service (NHS), almost every patient is registered with a general (family) practitioner (GP), and there is negligible overseas and cross-border flow of the population for healthcare. The Information and Statistics Division (ISD) (www.show.scot.nhs.uk/isd) centrally registers and codes every death certificate and every hospital inpatient admission in the NHS in Scotland. There are 4 neuroscience centers in separate health boards (Glasgow, Greater Glasgow; Edinburgh, Lothian; Dundee, Tayside; Aberdeen, Grampian) staffed by, among others, 32 consultant neurologists, 19 consultant neurosurgeons, and 71 consultant radiologists based at a neuroscience center or affiliated with another hospital providing CT, MRI, or catheter angiography. Patterns of investigation of intracranial hemorrhage probably vary depending on each clinical scenario and the facilities available. Compared with previous estimates, autopsy rates are declining in the years 1999 and 2000, ISD data show that there were 118,080 adult deaths in Scotland, of whom 14,630 (12%) had a postmortem examination.

Sources of Case Ascertainment
SIVMS uses multiple overlapping sources of case ascertainment to identify adults meeting its inclusion criteria.

Collaborative Neuroscience Network
The primary source of ascertainment is an all-inclusive, collaborative network of clinicians, radiologists, and pathologists working in the clinical neurosciences and stroke medicine (see Appendix, available online at http://stroke.ahajournals.org). Collaborators are based not only at the 4 neuroscience centers serving the population of Scotland but also at the other hospitals in the country and at neighboring hospitals in the north of England, where brain imaging facilities are available. SIVMS Steering Committee members in Edinburgh and Glasgow also actively survey (and retrospectively check) the records of the 2 Scottish specialist AVM clinics for new cases. Although the vast majority of patients in Scotland receive healthcare through the state-funded NHS, we have ensured that our network also covers the few private facilities offering brain imaging and clinical consultations from relevant specialists. Collaborators receive monthly newsletters and reminders about the study by e-mail and use materials available on the study Web site (www.dcn.ed.ac.uk/ivm) to notify the study of incident cases. Up-to-date information about any potential new collaborators in each hospital is obtained quarterly from the Steering Committee.

General (Family) Practitioners
Prevalence data indicated that each general practice in Scotland would have, on average, at least 1 adult with an IVM. We supposed that GPs would find these people memorable, and therefore we contacted all 3,700 GPs in Scotland in January 2000, asking if any of their patients, who we did not already know about, were eligible for inclusion in SIVMS.

Centralized Coding of Hospital Discharge Data and Death Certificates
In Scotland, every episode of hospital care is coded with details of a patient’s main diagnosis, comorbidity (up to 5 subsidiary diagnoses), and any operations or procedures conducted. Diagnostic data from hospital discharges as well as death certificates are coded with the use of the International Classification of Diseases, 10th Revision (ICD-10). In Scotland, all hospital inpatient stays (known as Scottish Morbidity Records, SMR01) have been collated since 1980 and are linked with death records from the General Register Office (www.gro-scotland.gov.uk) by ISD. Several mechanisms for ensuring data quality are in place at ISD, including validation, accreditation, quality assurance and monitoring, and a national coding advice and training program. In August 2001, ISD provided SIVMS with records of adults dying or discharged from a hospital in 1999 and 2000 for the first time ever according to its records with an ICD-10 code for an IVM in any diagnostic position (although ICD-10 has dedicated codes only for brain AVMs [Table 1]).

Follow-Up
Every participant is followed up on an annual basis. Their GP validates their survival, assesses their disability on the Rankin Scale, and comments on the occurrence of brain hemorrhage and epilepsy. GP and hospital case notes are also reviewed every year. Participants are contacted directly (if appropriate) for an assessment

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TABLE 1. Diagnostic Codes Pertinent to Intracranial Vascular Malformations in the International Classification of Diseases, 10th Revision

<table>
<thead>
<tr>
<th>Code</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>D18</td>
<td>Hemangioma and lymphangioma, any site</td>
</tr>
<tr>
<td>D18.0</td>
<td>Hemangioma, any site</td>
</tr>
<tr>
<td>I60</td>
<td>Subarachnoid hemorrhage</td>
</tr>
<tr>
<td>I60.8</td>
<td>Other subarachnoid hemorrhage</td>
</tr>
<tr>
<td>I60.80</td>
<td>Rupture of specified arteriovenous malformation</td>
</tr>
<tr>
<td>I67</td>
<td>Other cerebrovascular diseases</td>
</tr>
<tr>
<td>I67.1</td>
<td>Cerebral aneurysm, nonruptured (including acquired cerebral arteriovenous fistula)</td>
</tr>
<tr>
<td>I77</td>
<td>Other disorders of arteries and arterioles</td>
</tr>
<tr>
<td>I77.0</td>
<td>Acquired arteriovenous fistula (aneurysmal varix, acquired arteriovenous aneurysm; excluding cerebral [I67.1])</td>
</tr>
</tbody>
</table>

Q Congenital malformations, deformations, and chromosomal abnormalities
- Q27 Other congenital malformations of peripheral vascular system
- Q27.3 Peripheral arteriovenous malformation
- Q28 Other congenital malformations of the circulatory system
- Q28.2 Arteriovenous malformation of cerebral vessels
- Q28.3 Other malformations of cerebral vessels
of their health-related quality of life (on the Barthel Index, Short-Form 36, and Hospital Anxiety and Depression Scale). 16–18

Ethical Approval
The Multicenter Research Ethics Committee (MREC/98/0/48) and all local research ethics committees in Scotland approved this study of the adult population.19

Results
Such an exhaustive search for eligible adults would only be worthwhile if it resulted in a reasonably complete population-based sample. We herein evaluate the methods of SIVMS using data from the 92 adults who were incident in 1999 and 2000 with a definite brain AVM (data on the other IVMs will be published later).

Evaluation of Ascertainment

Completeness of Case Ascertainment
Figure 2 illustrates the overlap between the sources of ascertainment of brain AVMs. In the first year of the study, GPs only contributed cases already known to SIVMS, and therefore this labor-intensive and ineffective exercise was abandoned in the second year. The widespread collaborative network identified the vast majority of adults. However, coding (using I60.8 and Q28.2) yielded 4 adults (4% of the total) who were unidentified by the collaborative network.

Because it is virtually impossible for any survey to achieve complete ascertainment, an assessment of completeness would be desirable. A pragmatic approach is to observe the degree of overlap between the sources used (Figure 2). While prospective identification of every adult by both sources, although impossible, would be extremely reassuring, our approximately 50% overlap is adequate. Coding would never detect people diagnosed only as outpatients, although some may still be retrospectively ascertained in future years if they are admitted to a hospital with epilepsy or intracranial hemorrhage or for investigations or treatment, and therefore the degree of overlap will increase over time. Capture-recapture analysis estimates that the 2 major sources (Figure 2) missed 3 (95% CI, 1 to 9) adults with a brain AVM, which is <3% of the total.20

Representativeness of the Population
The adults incident in 1999 and 2000 were domiciled in proportion to the dispersion of the whole mid-2000 population estimate by health board (Figure 3), suggesting even and complete ascertainment. Although there appeared to be mild over-ascertainment in the 2 Scottish health boards where specialist brain AVM clinics operate (Lothian and Greater Glasgow), the 95% CIs of the age-standardized detection ratios for each health board all overlapped 1 (Figure I, available online at http://stroke.ahajournals.org).

Utility of ICD-10 Coding
We evaluated the sensitivity and positive predictive value of the combination of the 2 ICD-10 codes for brain AVMs
Sensitivity

The search of coding identified 58 (63%) of the 92 incident adults (Figures 2 and 4). Of these 58 adults, coding contributed only 4 who were unknown to SIVMS because collaborators failed to notify us of them (C in Figure 4). To evaluate sensitivity, ISD used probability matching record linkage to detect records of any hospital admissions and/or death certificates of all 92 adults. Of the 34 adults missed by the search of coding, 11 would never have been detected because they had not died and were never hospital inpatients (D in Figure 4). This makes the true-positive denominator for the calculation impossible to quantify.


table.

<table>
<thead>
<tr>
<th>Characteristic</th>
<th>SVMS (n=92)</th>
<th>Coded (n=58)</th>
<th>Not Coded (n=34)</th>
<th>Test*</th>
<th>Significance</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age at presentation (mean±SD)</td>
<td>45±16</td>
<td>42±12</td>
<td>52±19</td>
<td>t=-2.8, df=1</td>
<td>P=0.007</td>
</tr>
<tr>
<td>Male (95% CI), %</td>
<td>53 (43–63)</td>
<td>59 (46–70)</td>
<td>44 (29–61)</td>
<td>χ² = 1.8</td>
<td>P=0.178</td>
</tr>
<tr>
<td>Mode of presentation</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Hemorrhage (95% CI), %</td>
<td>46 (36–56)</td>
<td>57 (44–69)</td>
<td>26 (15–43)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Epilepsy (95% CI), %</td>
<td>26 (18–36)</td>
<td>28 (18–40)</td>
<td>27 (15–43)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Incidental (95% CI), %</td>
<td>21 (14–30)</td>
<td>10 (5–21)</td>
<td>38 (24–55)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Other (95% CI), %</td>
<td>7 (3–14)</td>
<td>5 (2–14)</td>
<td>9 (3–23)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Presence of aneurysm(s) (95% CI), %</td>
<td>22 (15–31)</td>
<td>22 (14–35)</td>
<td>21 (10–37)</td>
<td>χ² = 0.04</td>
<td>P=0.838</td>
</tr>
<tr>
<td>Catheter angiography (95% CI), %</td>
<td>74 (64–82)</td>
<td>90 (79–95)</td>
<td>47 (32–63)</td>
<td>χ² = 20.2</td>
<td>P=0.000007</td>
</tr>
<tr>
<td>Treated with embolisation or surgical excision or radiotherapy (95% CI), %</td>
<td>61 (51–70)</td>
<td>76 (64–85)</td>
<td>35 (22–52)</td>
<td>χ² = 14.8</td>
<td>P=0.0001</td>
</tr>
</tbody>
</table>

*T tests of statistical significance compared adults who were coded with those who were not.
§ Significance was tested across all 4 modes of presentation in a 4×2 table.

Evaluation of Potential Biases

Simplified Case Ascertainment

Only 58 (63%) of 92 adults would have been detected by coding alone, thereby underestimating the overall detection rate if coding was used as a single source of case ascertainment. Moreover, such a cohort would be biased toward younger people, with more hemorrhagic and fewer incidental presentations, greater investigation with angiography, and a strong tendency toward treatment (Table 2). Furthermore, if SIVMS were to base its recruitment purely on collaborators at the 4 tertiary referral centers in Scotland (simulating a hospital-based cohort), it would have missed 8 (9%) of the 92 adults with a definite brain AVM. When these 8 adults were compared with the other 84, fewer had had an angiogram (38% versus 77%; χ² = 6 [df=1], P=0.014), and fewer had been treated (13% versus 66%; χ² = 8.6 [df=1], P=0.003). The numbers were too small to detect any other disparities, but these differences reflect how specialist hospital cohorts are generally unrepresentative of the population at baseline and are likely to remain so during follow-up.
Pattern of Diagnostic Investigation
In this population-based study, 68 (74%) of 92 adults with brain AVMs were investigated with angiography, which led to definite diagnoses in 66 (72%) of them (Figure II, available online at http://stroke.ahajournals.org). The other 28% were diagnosed by CT, MRI, or pathological examination. These investigation patterns resulted in a median time between clinical presentation and definite diagnosis of 14 days (range, 0 to 259 days), with delays reflecting the mode of presentation and occasional need for extensive and/or repeated investigation. Using catheter angiography as the diagnostic reference standard would have biased the cohort toward younger people (mean age, 42 versus 56 years; \( P = 0.002, 2\)-sample t test) who were more likely to have presented with hemorrhage (52% versus 29%; \( P = 0.011, \) Fisher’s exact test) and to have been subsequently treated (77% versus 17%; \( \chi^2 = 26.6 \) \([df = 1], P = 0.0000002\)).

Discussion
Evidence for this study’s comprehensive case ascertainment comes from recruitment in proportion to the distribution of the Scottish population, sizable overlap between the sources of cases, and capture-recapture analysis. We also found that streamlining the sources of case ascertainment to rely on less onerous methods such as coding alone or on tertiary referral center–based collaborators alone would have resulted in the recruitment of a biased cohort, as would reliance on angiography for definitive diagnosis of a brain AVM. Coding of hospital discharge and death certificate data had reasonable sensitivity and moderate positive predictive value for detecting incident adults but was clearly insufficient by itself. These explorations of sources of bias illustrate just how vulnerable studies of frequency and prognosis are to imperfections in study design.

Diagnostic Criteria
Definitions and diagnostic standards alone can introduce considerable bias. We used working definitions dependent only on adequate investigations. These are generalizable to routine clinical practice in which extensive investigation may not be performed if it is thought to be clinically inappropriate, because the patient declines it, or because it is not available. For brain AVMs in particular, a definition reliant on catheter angiography will miss diagnoses made on CT, MRI, or pathological examination alone and ignores the existence of angiographically occult brain AVMs. One quarter of adults with brain AVMs would have been excluded from this study had we required them to have been diagnosed with catheter angiography.

Although a diagnostic standard based on catheter angiography might foster complete collection of variations of vascular anatomy (angiarchitectural) that are of great interest as prognostic factors, \( \text{SIVMS} \) has shown that this biases studies toward younger people with more disabling modes of presentation who are more likely to receive interventional treatment. These studies would underestimate the frequency of brain AVMs, underestimate their importance as a cause of sudden death by missing postmortem diagnoses, and produce an unrepresentative cohort for follow-up.

Case Ascertainment
To ensure identification of a complete sample representative of the population, we used several sources of case ascertainment, which overlapped considerably. Regional healthcare systems will dictate which sources are most appropriate for other studies, but additional potential secondary data sources include databases of imaging studies performed in the geographic area of interest \( \text{2} \) and surveillance systems such as the British Neurological Surveillance Unit (www.theabn.org/academic/bnsu.html).

Population-Based Versus Hospital-Based Studies
SIVMS has avoided many of the disadvantages of hospital-based studies. Hospital-based cohorts tend to represent local referral practices and the beliefs and treatment preferences of research groups with a special interest in brain AVMs rather than describe the behavior of a more representative population-based sample. Hospital-based studies are more likely to detect people with a disabling (yet nonfatal) mode of presentation, and SIVMS would have been biased toward people with more extensive investigation and treatment if simply relying on specialists. This hospital-based sampling bias also affects prognostic subgroups unequally, makes the comparison of separate hospital-based cohorts inappropriate, and militates against their meta-analysis. This has been confirmed by an analysis of 5 single-center brain AVM cohorts, which were found to have significant differences in both demographic and clinical characteristics as well as in brain AVM angiography.\( \text{22} \)

Utility of Coding
The NHS has a long-established coding infrastructure, and the median accuracy of a wide variety of \text{ICD} codes in the United Kingdom has been found to be 84%.\( \text{23} \) However, we knew from previous experience in Scotland that coding inaccuracies made a large impact on the detection and mortality rates of motor neuron disease, a disease with a detection rate similar to that of brain AVMs.\( \text{24} \) Regrettably, we found the sensitivity and positive predictive value of \text{ICD-10} coding of brain AVMs to be moderate in practice. Coding missed 37% of adults with brain AVMs, although it alone contributed 4% of the whole cohort.

The utility of routinely collected data depends on the precision of diagnosis at death or at discharge from the hospital. This is affected by the variable nomenclature used by clinicians, inadequate completion of the discharge summary and SMR01 form, and inaccuracies in transcription to ISD and its record linkage. Twelve percent of adults with brain AVMs in SIVMS were not admitted to a hospital in the year of their diagnosis. Coding might only identify these people some time after inception, depending on whether they die and whether they are admitted to hospital for further investigation or treatment.

Moreover, the adequacy of \text{ICD} coding depends on the existence of relevant codes. The \text{International Classification of Diseases, Ninth Revision (ICD-9)} used a single code (747.8) for “cerebrovascular anomalies” (encompassing a heterogeneous spectrum of IVMs and other lesions), which was shown to have a 94% (95% CI, 91% to 96%) sensitivity but poor specificity in a New York hospital.\( \text{25} \) The transition...
to ICD-10 has merely created an explicit code for brain AVMs but not for other IVMs (Table 1).

**Bias in Simplification of Ascertainment**

The administrative burden of maintaining a collaborative network and following people prospectively through their medical records questions whether coding alone would be a satisfactory means of ascertaining and following up incident adults. Sadly, we have found that adults who are coded are a biased group. Using coding for follow-up would also subject this group to the accuracy of record linkage and coding of death certificates, and it would miss any important outcomes (such as epilepsy) not resulting in hospital admission.

**Conclusions**

We have found that, just as for stroke in general, the sensitivity and positive predictive value of coding are poor26–28 and that there are important differences in the composition of a brain AVM cohort according to whether population-based or purely hospital-based sources are used. Our study illustrates the potential inaccuracies of studying brain AVMs in single settings (such as individual tertiary referral centers) or with reliance on secondary data (such as coding).25,29

We hope that this article and the accompanying article on IVM detection rates,9 in conjunction with a complementary report by the Joint Writing Group,21 will encourage other researchers to set up similarly designed, potentially collaborative, multicenter, population-based studies. These studies will need to be based on large populations (because of the infrequency of outcomes attributable to IVMs). The ultimate goal is to establish the comparative epidemiology and prognosis of IVMs and to provide a permanent infrastructure for mounting large multicenter trials of treatments.

**Acknowledgments**

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**References**


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In the article entitled “Scottish Intracranial Vascular Malformation Study (SIVMS): Evaluation of Methods, ICD-10 Coding, and Potential Sources of Bias in a Prospective, Population-Based Cohort” by Al-Shahi et al, a reference to information in Figure 4 was printed incorrectly. The paragraph from page 1160 including the corrected text (in bold) appears below:

**Sensitivity**

The search of coding identified 58 (63%) of the 92 incident adults (Figures 2 and 4). Of these 58 adults, coding contributed only 4 who were unknown to SIVMS because collaborators failed to notify us of them (C in Figure 4). To evaluate sensitivity, ISD used probability matching record linkage to detect records of any hospital admissions and/or death certificates of all 92 adults. Of the 34 adults missed by the search of coding, 11 would never have been detected because they had not died and were never hospital inpatients (D in Figure 4). This makes the true-positive denominator for the calculation of coding sensitivity 81 (A–D in Figure 4), resulting in a sensitivity of 72% (95% CI, 61% to 80%). Coding missed the false-negatives (E in Figure 4) because of the incorrect allocation of Q27.3 to 8 adults and I67.1 to 3 adults (Table 1); the remainder were due to brain AVM diagnoses not appearing on the discharge summary or death certificate or missing records in the ISD data set.

The publisher apologizes for the error.