

of ascertaining sibships with one affected child decline over time. If there had been a short-fall in ascertaining patients during the early years of this study, the effect would be evident as a change in immigrant group-specific incidence over time. No change of this nature was noted in the largest immigrant group in which parents were of Greek origin; there was no observed difference between observed and expected numbers of patients born in four cohorts during the years 1961-1976, ( $\chi^2 = 4.63$ ; not significant).

The incidence of homozygous beta-thalassaemia has increased with each time period (Table 1). Assuming that immigration policies do not change dramatically, the peak in incidence should have passed. Second-generation marriage rates within each immigrant group are lower than those in the first generation after immigration (although nearly twice as high for women of Greek and Italian origin than for second generation males from the same countries).<sup>9</sup> In addition, the availability of antenatal diagnosis (AND) in New South Wales since 1979 should contribute to the decline in birth of homozygous patients. In the United Kingdom, AND has become almost universally acceptable to parents of Cypriot origin who accounted for two-thirds of all homozygous patients. As far as is known, no affected child was born in the two years up to mid-1981 within the London Cypriot community.<sup>10,11</sup>

Approximately five homozygous patients were born annually in New South Wales between 1967 and 1976, suggesting that there may be at least 20 pregnancies to be monitored by AND each year.

About 70% of patients with the homozygous beta-thalassaemia born in New South Wales between 1961 and 1976 were of Greek or Greek-Cypriot origin. It appears probable that the majority of patients and couples who will require genetic counselling and AND will continue to come from this group.

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## Cerebral infarction caused by vasospasm

Gregory M. Stone and Richard J. Burns

**ABSTRACT:** Fifteen patients with cerebral infarction, the majority of them being under the age of 40 years, were reviewed. Patients in whom an underlying structural disease, such as atheroma or thromboembolism, was suspected on clinical grounds were excluded. On the basis of either a personal or family history of migraine, or a concomitant vascular

headache, or a migratory pattern of evolution of the neurological deficit, a migrainous disorder was considered possible in 14 patients. It is postulated that cerebral vasospasm is a cause of cerebral infarction, especially in younger people.

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CEREBRAL INFARCTION is a common disease in the elderly. Its occurrence in people under the age of 40 years is much less common, a conservative estimate being 5% of

all cases of infarction.<sup>1</sup> This still represents a considerable number of people. In the elderly, the cause is usually cerebral atheroma or thromboembolic disease from the carotid arteries, aortic arch or heart. On occasions, one finds less common conditions such as trauma, arteritis, or blood dyscrasias. In younger patients, degenerative arterial disease is a lesser factor.

Infarction resulting from temporary spasm of cerebral arteries can occur with subarachnoid haemorrhage caused by a berry aneurysm or by hypertensive encephalopathy. A

Faculty of Medicine, The University of Sydney, NSW 2006.

Gregory M. Stone, Final Year Medical Student.

Department of Medicine, Flinders Medical Centre, Bedford Park, SA 5042.

Richard J. Burns, FRCP, FRACP, Associate Professor of Medicine; Director, Neurology Unit.

Reprints: Dr R. J. Burns.

functional disturbance of cerebral arteries is also thought to be the mechanism of infarction which occurs in migraine.<sup>2</sup>

It is the purpose of this paper to discuss the clinical features of a group of 15 patients with cerebral infarction, in whom the most likely cause was considered to be cerebral vasospasm.

### Patients and methods

The patients selected were all seen by one of two neurologists of the Neurology Unit at the Flinders Medical Centre over a period of five years from 1976 to 1981. They were coded as cases of cerebral infarction, possibly due to migraine, cerebral vasospasm, or of unknown cause. Excluded from the study were those patients in whom the neurological deficit lasted for less than 24 hours and those in whom it was reasonable to assume that there was some underlying vascular disease. Patients with hypertension, diabetes

age of 40 years. Eight patients had a past history of migraine. All, except two, had a headache associated with the neurological episode. Seven patients had a family history of migraine.

The majority of infarcts were located in the left or right cerebral hemispheres; only two infarcts were in the posterior

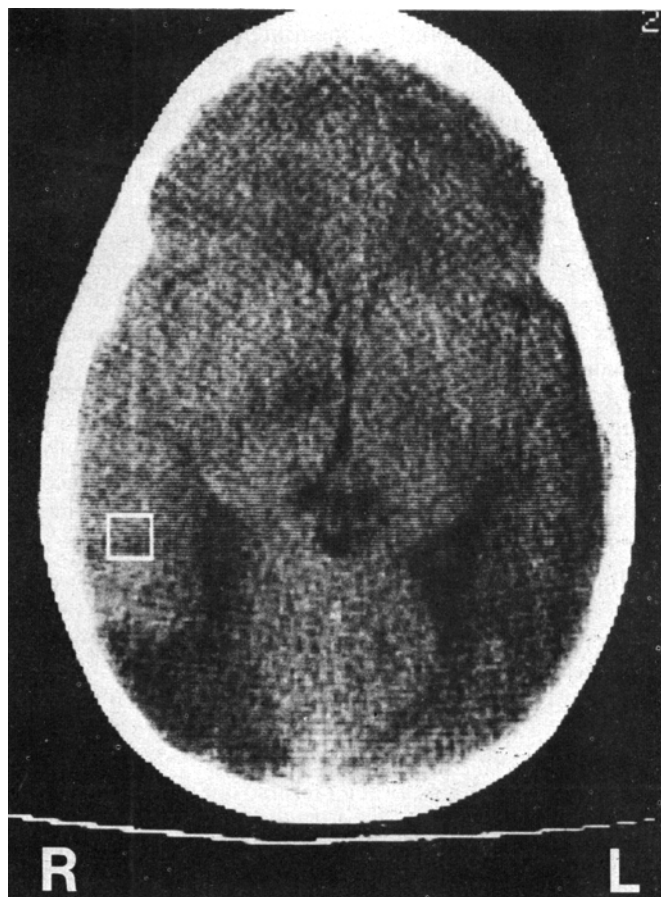


FIGURE 1: Plain CT head scan of the patient in Case 3 showing a low-density area in the right occipital region, and a smaller hypodense area (outlined) in the right internal capsule.

mellitus, evidence of a systemic vasculitis and hyperlipidaemia were excluded.

The medical records of 15 patients were retrospectively examined with particular reference to a personal and family history of migraine, the occurrence of headache in association with the acute neurological deficit and its temporal relationship to the deficit, the way in which the neurological deficits evolved, and the results of any special investigations which might have established a cause for the cerebral infarction. Other general information such as age and sex was also obtained.

### Results

The mean age of 15 patients (nine males) was 33 years (range, nine to 58 years); only two patients were over the

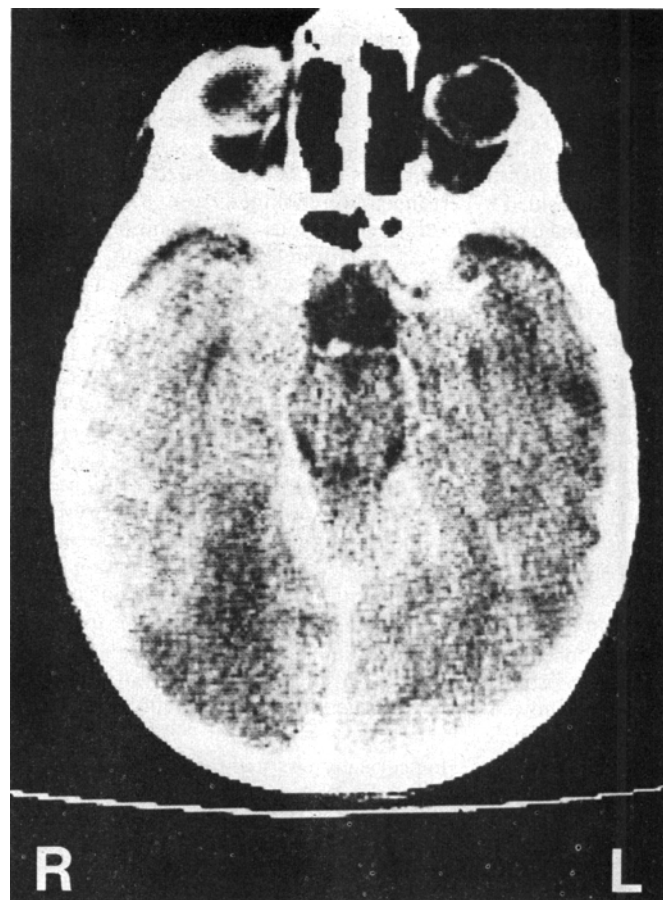


FIGURE 2: The enhanced CT head scan of the patient in Case 3 showing an infarct in the right occipital region.

fossa. In 10 patients evidence of a residual neurological deficit was present at three months after the infarction.

A CT scan was performed in 14 patients and, in nine of these, an area of low density consistent with cerebral infarction was found. In four patients, multiple infarcts were seen. Of the remaining six, in whom the results of the scan were reported as normal, two had early scans (that is, within the first three days of their neurological episode).

Cerebral angiography was performed in five patients. In four, the results were normal. The abnormal angiogram showed a possible mild stenosis of the supraclinoid portion of the internal carotid artery, which was thought to be irrelevant.

Examination of the cerebrospinal fluid (CSF) was performed in six patients; the results were normal in four patients, and showed a mild increase in protein level in two patients.

### Illustrative case reports

**Case 1.**—A 33-year-old manager presented with retrosternal pain, typical of angina pectoris. No abnormalities were found on

coronary angiography. Several months later, he had an episode of dysphasia which resolved in a few hours. One month after this episode, he presented with a sudden headache and right-sided motor and sensory signs without dysphasia. A CT scan showed an area of low density in the left temporal region, consistent with infarction. The patient subsequently had episodes of right-sided visual hallucinations which lasted for 30 minutes and were followed by hemispheric headache. No abnormality was found on a left carotid angiogram.

**Case 2.**—An 18-year-old man had classic migraine every five or six weeks for two years. His mother also suffered from migraine. On this occasion, a headache of three days' duration led to the development of weakness and clumsiness of his left arm. Within hours, the left leg became weak, he complained of diplopia, and his left arm then felt numb. His headache persisted. On examination, a left-sided hypertonia and hypokinesia were found, and, in addition, he had a horizontal nystagmus. These signs persisted for several days. A CT scan performed on admission, an electroencephalogram (EEG), and a right carotid angiogram showed no abnormalities. A slight increase in protein level was found on examination of his CSF.

**Case 3.**—A nine-year-old boy awoke with vomiting, headache, fever, abdominal pain and lethargy. There was a strong family history of migraine. He was admitted to hospital, where, on examination, no neurological deficit was found. Twenty-four hours later, it was noted that his left pupil was dilated and reacted sluggishly to light. CSF examination at that time showed no abnormality. Next day, he developed left-sided facial weakness, a left-sided hemiparesis, a left-sided homonymous hemianopia, and a left-sided hemisensory inattention. He also had dressing apraxia.

A CT scan was performed, which showed a low density area in the right occipital region and a smaller hypodense area in the right internal capsule, which were considered to be areas of infarction. An EEG showed a very distinct focal abnormality in the right occipital region.

The patient's neurological signs persisted for five days, and then slowly began to resolve.

## Discussion

A diagnosis of vasospastic infarction was considered possible in 14 of the 15 patients. This was made on the basis of a number of factors, including a past history of migraine, a family history of migraine, headache which occurred either before, during, or after, the neurological deficit, and neurological symptoms which evolved after a period of time. In the one patient, in whom migraine could not be implicated, the oral contraceptive pill could have been a contributing cause.

In the many clinical and pathological studies performed in cerebral infarction, there has always been a group of patients in whom it has not been possible to determine the exact cause.<sup>1,3</sup> In our patients, it was not possible to completely exclude cerebral atheroma, cerebral thromboembolism or arteritis. Even if four-vessel angiography and echocardiography had been performed on every patient, such diagnostic possibilities would still exist. Nevertheless, we feel that the young age of the patients makes the presence of a significant degenerative vascular disease most unlikely.

CT-scan evidence of cerebral infarction was not present in all cases. However, because the occurrence of headache was thought to be an important symptom in establishing the diagnosis, the scan at least excluded an intracerebral haemorrhage, and this was one of the reasons why it was performed early rather than late. It is possible that follow-up scans might have shown infarction in some of the patients in whom the result of initial scan was normal.

Headache is not a prominent feature of cerebral infarction caused by cerebral atheroma or thromboembolism, although it is present on occasions.<sup>4</sup> Thirteen of the 15 patients experienced headache temporally, related to the neurological deficit. The description of each was consistent with vascular origin, rather than a tension headache, or that related to intracranial hypertension. Seven of the patients had no past history of migraine. However, previous studies have stressed that a first migraine attack can be complicated by infarction.<sup>4</sup>

The manner in which the neurological deficit evolves will also help in the diagnosis of the underlying disorder. The neurological deficit in cerebral vasospasm, particularly in migraine, may evolve over a period of minutes or hours with resolution of some symptoms as new ones appear.

In a recent review of vasospasm, Miller *et al* have suggested that there might be a link between migraine, angina and Raynaud's phenomenon.<sup>5</sup> Even though angiographic studies of patients performed during the course of migraine with prominent ischaemic symptoms have been very infrequent,<sup>11</sup> there exists a real possibility that cerebral vasospasm, with or without the traditional migraine symptoms of headache and vomiting, does occur. It is now accepted that coronary artery vasospasm is an explanation of angina pectoris when angiographic studies show normal coronary arteries. Peripheral vasospasm has long been accepted as the cause of Raynaud's phenomenon.

The exact incidence of cerebral infarction occurring in association with migraine has not been determined.<sup>6,7</sup> Since the advent of CT scanning, there have been reports to indicate that cerebral infarction in patients known to have migraine is much more common than had been previously realised.<sup>8,9,10</sup> In some instances, the infarction is not accompanied by a demonstrable neurological deficit or even a history of transient neurological symptoms.<sup>9,10</sup>

Until a method is devised by which one can safely study the calibre of cerebral blood vessels in humans under physiological conditions and during the phases of migraine, the exact mechanism of cerebral infarction in migraine will remain unproven. However, we believe that, in our current state of knowledge, it is reasonable to assume that cerebral infarction results from vasospasm.

There are probably other factors, apart from migraine, which might trigger cerebral vasoconstriction. Migraine is a common condition and transient ischaemic symptoms occurring with it are also common,<sup>12,13</sup> and, yet, on most occasions infarction does not occur. Presumably the duration, severity and extent of the spasm are of critical importance. As yet, there seems no way in which one can predict in any person, migraine sufferer or not, when this type of infarction is likely to happen.

As far as we are aware, no drug has been found capable of reversing the process of vasospasm, and the efficacy of migraine prophylaxis in preventing cerebral infarction has not been established.

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# Survival of South Australian cancer patients

## A study of the State's Cancer Registry data

Anton Bonett and David M. Roder

**ABSTRACT:** Survival rates for cancer patients derived from population-based data are a fundamental means of monitoring the effectiveness of treatment for the community at large. We used South Australian Cancer Registry data to study survival by cancer site from date of diagnosis in 1977-79 to August 1980. One-year and

two-year cumulative percentage survival rates for all types of invasive cancer were 64% ( $\pm 0.5\%$ ) and 54% ( $\pm 0.5\%$ ) respectively. We anticipate that these findings will provide a yardstick for intra-hospital survival analyses.

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INFORMATION on the survival of cancer patients may be obtained from several sources, including: (i) individual clinicians' records; (ii) hospital-based registries; and (iii) a population-based registry which covers the total population of a geographic region. Each source is well-placed to provide a unique contribution. In general, the broader the population coverage, the greater the emphasis placed on data with an epidemiological application. Conversely, clinical records and hospital-based registries are generally directed at monitoring treatment.

The South Australian Cancer Registry covers the total population of the State. Cancer cases are notified at diagnosis by all hospitals and histopathology laboratories. Deaths of cancer patients are identified through: (i) monthly reports of all deaths from cancer, as provided by the Registrar of Deaths; (ii) notifications from hospitals of deaths from cancer and (iii) computer linkage of Registry files with State death records to detect deaths of cancer patients from causes other than cancer.

In this report, we provide information on the survival of patients from the date of diagnosis in 1977-79 to August 1980. This period is sufficient to calculate one-year and two-year survival rates for most cancer patients.

### Methods

We calculated rates by means of the non-parametric technique described by Cutler and Ederer.<sup>1</sup> By using a life-table approach, this technique enables one-year and two-year survival rates to be based on information for all cases of cancer, not simply cases diagnosed one or two years previously. Inherent in the approach is the assumption that the continued survival of patients not followed for the entire period will be the same as for patients followed throughout.

Standard errors are provided with all rates to indicate the level of imprecision from sampling variability. It can be assumed that about 95% of samples of the stated sizes would yield survival rates within two standard errors of the rates now presented.

In general, there was little difference between male and female survival rates, and so the rates are presented for both sexes combined.<sup>2</sup>

### Results

One-year and two-year cumulative percentage survival rates for all types of invasive cancer (including melanoma) collectively were 64% ( $\pm 0.5\%$ ) respectively. In other words, it follows from our series that 36% of patients die within one year of diagnosis and 46% die within two years.

Whereas the two-year survival rate for patients with cancers of the buccal cavity was higher than for all cancer patients collectively, lower rates applied to patients with lesions of the oropharynx, nasopharynx, and hypopharynx (Figure 1).

Lower rates were also apparent for patients with cancers of the oesophagus, stomach, small intestine, liver, gall-bladder and bile ducts, and pancreas, but not for patients

South Australian Health Commission, G.P.O. Box 1313, Adelaide, SA 5001.

Anton Bonett, MD, DPH, Head, Central Cancer Registry Unit.  
David M. Roder, BDS, MPH, DDSc, FRACDS, Director of Epidemiology Services.  
Reprints: Dr Bonett.