SHORT REPORT

Family history of subarachnoid haemorrhage: supplemental value of scrutinising all relatives

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Abstract

***Objective and methods*—To assess the validity of the family history obtained at the bedside of patients with recent subarachnoid haemorrhage by subsequently contacting all first and second degree relatives, with verification from medical record data.**

***Results*—In a prospectively collected series of 163 patients with recent subarachnoid haemorrhage the history or cause of death could be ascertained in 1259 (98%) of the first degree relatives and in 3038 (85%) of the second degree relatives. For first degree relatives only, the sensitivity of the family history at the bedside was 0·75 (95% confidence interval (95% CI) 0·35–0·97) and the positive predictive value was 0·55 (95% CI 0·23–0·83); for first and second degree relatives together the sensitivity was 0·58 (95% CI 0·28–0·85) and the positive predictive value was 0·64 (95% CI 0·31–0·89).**

***Conclusion*—The accuracy of the family history taken at the bedside is modest; a more thorough collection of data is crucial if the decision is taken to screen relatives based on the family history.**

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In 6% to 9% of patients with subarachnoid haemorrhage the disorder is familial,[1] and in these familial cases outcome is worse.[2] If screening for and treatment of aneurysms in asymptomatic relatives is considered, it is important to be accurately informed about the family history. The most exact method to ascertain the number of relatives and the nature of any illnesses is to construct a pedigree for each patient to subsequently interview all relatives personally and to then verify this information with medical documents.

Because it is unknown whether this time consuming process yields more accurate information than a simple family history obtained at the bedside, we compared the two strategies in a prospective, hospital based series of patients with subarachnoid haemorrhage.

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Patients and methods

A series of 163 patients with aneurysmal subarachnoid haemorrhage established by CT, admitted to the University Hospitals in Utrecht and Rotterdam and the Academic Medical Center in Amsterdam, was prospectively collected from September 1991 to October 1992. In the same period 50 other patients with subarachnoid haemorrhage were admitted and excluded for the following reasons: three patients because a cause other than a ruptured aneurysm was found for the subarachnoid haemorrhage; 36 patients because the patient or the next of kin refused to participate; 10 patients because most relatives lived outside Europe; and one patient because she was adopted and knew nothing of her biological relatives.

Soon after admission patients were asked whether any of their relatives had had a subarachnoid haemorrhage or a stroke. For patients with a depressed level of consciousness, the family history was obtained from the partner, the next of kin, or in some instances from both at the same time. These data represent the standard strategy of collecting the "family history at the bedside". Our experimental and extensive strategy was as follows. A pedigree was drawn up for each family and all living relatives known to us were interviewed by telephone, by means of a standard questionnaire. For deceased relatives a next of kin was interviewed about the cause of death. If this informant mentioned a stroke or any other brain disease, all available medical documents were retrieved, including those from abroad. All histories and all medical documents with any relation to subarachnoid haemorrhage were classified independently by two observers (JECB and GJER) as definite subarachnoid haemorrhage, probable subarachnoid haemorrhage, or possible subarachnoid haemorrhage, according to criteria decided on in advance (table 1). A diagnosis of definite subarachnoid haemorrhage could be made only from medical records. In five cases the observers did not agree and in these instances the data were classified by a third observer (JvG) after which a decision was made by majority vote.

For the analysis we recorded as positive for the family history at the bedside all episodes

Table 1 Criteria for the diagnosis of subarachnoid haemorrhage in relatives

	Medical documentation	*History*
Definite subarachnoid haemorrhage	Clinical features *and* blood in basal cisterns on CT *or* xanthochromic CSF *or* aneurysm on angiogram or necropsy	—
Probable subarachnoid haemorrhage	Sudden severe headache *and* normal neurological examination *and* haemorrhagic CSF *and* sudden deterioration and death within four weeks	In first four weeks after "stroke" second ictus followed by death *and* age < 70
Possible subarachnoid haemorrhage	Sudden severe headache *and* normal neurological examination *and* angiography not performed *and* haemorrhagic CSF *or* Sudden severe headache *and* focal abnormalities and/or decreased consciousness *and* haemorrhagic or xanthochromic CSF *and* age < 70 *or* Sudden death *and* age < 40	"Stroke", no details *and* age < 50 *or* in first four weeks after "stroke" second ictus followed by death *and* age > 70 *or* Sudden severe headache necessitating bed rest *and* death within 4 weeks *and* age < 70 *and* no medical examination *or* Sudden severe headache followed by loss of consciousness and death *and* age < 70 *and* no medical examination

classified as probable or possible subarachnoid haemorrhage; most likely these include all instances of definite subarachnoid haemorrhage if records could be retrieved for these relatives. For the extensive search strategy we recorded as positive only episodes classified as definite or probable subarachnoid haemorrhage. The family history given by the patient or the next of kin at the bedside was compared with the information obtained by the extensive strategy; the extensive strategy was considered the "gold standard".

The 163 patients had 1290 first degree and 3588 second degree relatives. The medical history or cause of death was confirmed in 1259 (98%) first degree relatives and in 3038 (85%) second degree relatives.

Results

The family history obtained at the bedside identified 11 families in which subarachnoid haemorrhage had previously occurred in one of the relatives (table 2). In seven cases this was correct: in six of these patients it concerned a first degree, in one a second degree relative. In five of these seven patients the relative had died.

For four of the 11 relatives purported to have had a subarachnoid haemorrhage this diagnosis could not be confirmed: one (first degree) relative proved to have had a pontine haemorrhage, another had had a "stroke" but no details could be retrieved. In the other two instances our search strategy did not confirm a subarachnoid haemorrhage in any of the relatives.

In the remaining 152 families the history at the bedside was negative for familial subarachnoid haemorrhage. In five of these 152 families scrutinising the relatives disclosed unreported instances of subarachnoid haemorrhage: in one family a sister, in another family a half brother; and in three families a second degree relative. Three of these five relatives had died. Apart from the 12 relatives with definite or probable subarachnoid haemorrhage there were 12 relatives with possible episodes of subarachnoid haemorrhage that could not be confirmed because medical records were no longer available.

For first degree relatives only, the predictive value of a bedside history positive for familial subarachnoid haemorrhage was 0·55 (95% confidence interval (95% CI) 0·23–0·83) and the sensitivity was 0·75 (95% CI 0·35–0·97). For the first and second degree relatives combined the predictive value of a positive family history of subarachnoid haemorrhage was 0·64 (95% CI 0·31–0·89) and the sensitivity was 0·58 (95% CI 0·28–0·85).

Table 2 Number of families with or without familial subarachnoid haemorrhage (SAH)

Standard family history at bedside	*Interview of all relatives plus medical data*		
	SAH	*No SAH*	
SAH	7 (6)	4 (5)	11
No SAH	5 (2)	147 (150)	152
	12 (8)	151 (155)	163

The numbers in parentheses represent the data when only first degree relatives are taken into account.

Discussion

In our study one quarter of the families with a positive history for subarachnoid haemorrhage in a first degree relative would have been undetected without the information provided by scrutinising all individual relatives; if second degree relatives were also taken into account, the proportion of undetected families rose to almost a half. The poor sensitivity of family history for subarachnoid haemorrhage shows that the frequency of "familial subarachnoid haemorrhage" in other studies has probably been underestimated as in none of these studies were the relatives contacted systematically.[3-5]

Because subarachnoid haemorrhage is a dramatic event it should be easily remembered by relatives, but apparently it is not. In a recent study on the reliability of the family history for myocardial infarction, sensitivity was comparably poor, but in that study the family history was verified only by contacting the

general practitioners of the living relatives.[6] Even when the analysis was restricted to deceased relatives, sensitivity for family history of stroke in general proved to be low.[7] These data corroborate our present finding that the sensitivity of the family history is low, even for well known emergencies, and that an accurate family history requires verification of the family history by medical record data.

Several factors may have influenced our results. Firstly, in a minority of the relatives who had died, the cause of death could not be retrieved. In some families relatives were no longer in contact with one another and their whereabouts or even their being alive could not be ascertained. In other families relatives declined to cooperate, including one family in which the index patient had died and one first degree relative had previously had a subarachnoid haemorrhage. In addition, when relatives were willing to provide information, medical reports had sometimes been destroyed if the event had occurred more than five or 10 years previously so that the information could not be verified. Thus even by contacting all relatives we probably have still underestimated the familial occurrence of subarachnoid haemorrhage. Secondly, this study has been carried out in centres specialising in care of patients with subarachnoid haemorrhage. Attending physicians in these centres may obtain the family history more accurately than physicians in general hospitals; the family history may have been collected more thoroughly than usual as some attending physicians were aware of our study being in progress, but we do not think that these two phenomena have had a major influence on our results. Thirty six families (18%) could not be included because the patient or next of kin refused participation in the study. In most instances the reason for refusal was that the patient had died; we consider it unlikely that this has introduced an important bias. A third factor that should be taken into account in the interpretation of our results is that we accepted even the slightest suspicion of a subarachnoid haemorrhage as a positive family history at the bedside, whereas for the extensive strategy only episodes of definite and probable subarachnoid haemorrhage were counted as positive. The sensitivity of the bedside history decreases even further if only highly suggestive histories are considered positive.

In conclusion, our study shows that a considerable proportion of familial cases of subarachnoid haemorrhage will be missed if the medical history of all relatives is not scrutinised. Family history has become important in subarachnoid haemorrhage, because non-invasive imaging methods allow screening of asymptomatic relatives in familial subarachnoid haemorrhage. If screening is based on a positive family history, we advise a more thorough collection of data than a routine conversation at the bedside.

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