Collaborative management of intracranial arteriovenous malformation

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ABSTRACT

Objectives: To study all the cases of intracranial arteriovenous malformation presented to a main neurosurgical center in Saudi Arabia and compare the results with reports from outside the Kingdom. To estimate the hospital-based prevalence of the disease which may reflect the frequency in the Saudi Arabia.

Methods: A retrospective study was conducted to analyze the cases of intracranial arteriovenous malformation admitted to King Faisal Specialist Hospital and Research Center from 1982 to 1996.

Results: A total of 97 patients were eligible for the present study. The characteristic aspects of intracranial arteriovenous malformation matched well with reports from other parts of the world. The hospital-based frequency was calculated to be 40 per 100,000 admissions. Most of the patients were men (2.5 men:1 woman). The average age at presentation was 29 years. More than half the patients presented with intracranial hemorrhage (52%); seizure was 2nd in frequency (32%). More than 80% of the arteriovenous malformations were in the supratentorial location, the rest were in the infratentorial location. According to the Spetzler and Martin grading system, 16 patients presented with grade I, 43 with grade II, 28 with grade III, 13 with grade IV, and one patient with grade V arteriovenous malformation. Three patients had double arteriovenous malformation. The patients were managed by single or combined procedures, which included surgery (28.9%), endovascular embolization (35.1%), and radiosurgery (16.5%). The complications included transient or persistent neurological deficit, status epilepticus, and immediate post-embolization intracerebral hemorrhage which proved fatal in one case. The different variables were tested for predictability of the risk of bleeding. The results were compared with those reported in the literature.

Conclusion: Intracranial arteriovenous malformation is not uncommon in Saudi Arabia. The patient presentation, arteriovenous malformation classification in each grade, and treatment outcome correlated well with other reports in the literature. The best obliteration rate was observed with surgical excision of the malformation (75%). No therapeutic modality was without complication.

Keywords: Arteriovenous malformation, endovascular embolization, intracranial vascular malformation, outcome, surgery, radiosurgery.


Arteriovenous malformation (AVM) is the most commonly encountered symptomatic vascular malformation. It is characterized by abnormal communication between the arterial and venous systems in the absence of normal capillary channels. According to the autopsy study by McCormick, the prevalence of AVM is 0.5 percent. Clinically, AVM is responsible for 8.6% of subarachnoid hemorrhage, with an annual incidence of 3 per 100,000. Most cases of intracranial AVM present between the 2nd and 5th decade of life, with less cases presenting in the extremes of age, and men

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are more affected than women.\textsuperscript{9,10} Intracranial hemorrhage is the most commonly reported mode of initial presentation, followed by seizure and other symptoms.\textsuperscript{6,11-14} In Saudi Arabia no study has addressed the problem of incidence and prevalence of AVM. The present study analyzed all the cases proved to have intracranial arteriovenous malformation presented to King Faisal Specialist Hospital and Research Center.

**Methods.** This is an observational, cross-sectional, analytical study of all cases of intracranial arteriovenous malformation (AVM) seen in King Faisal Specialist Hospital and Research Center, Riyadh, from 1982 to 1996. Medical records were retrieved and systematically studied. The clinical status of all patients on admission and at the end of the study period was documented and retrospectively graded using the Karnofski scale. Various radiological studies for all patients with AVM were reviewed. All cases had computed tomography (CT) scan with or without magnetic resonance imaging (MRI) of the brain, and were confirmed as AVM by cerebral angiography. The AVM was graded retrospectively according to the Spetzler and Martin classification. The data were registered in the computer using “Filemaker Pro-2” software program. Data were transferred to SPSS software computer program for statistical analysis of the results. The final clinical status of each patient was obtained from medical records and, when the telephone number was available, by directly contacting the patient or his/her family.

**Results.** **Hospital based prevalence.** From 1982 to 1996, 5708 patients with different pathological diagnoses were admitted to the Neurosurgery Department of King Faisal Specialist Hospital and Research Center. Of this number, 158 patients with the diagnosis of AVM were identified. Sixty-one patients whose charts were incomplete were excluded from the study, leaving a total of 97 eligible patients. The prevalence of AVM in all neurosurgery cases was 2.8%. During the same period the total number of admissions to the hospital was 394,927 patients. The hospital-based prevalence was therefore 40 cases per 100,000 admissions. The period of follow-up for each patient from the time of diagnosis until the end of this study or the time last seen or contacted ranged from 6 months to 21 years.

**Age and gender.** There were 69 men (71.1%) and 28 women (28.9%), giving a men:woman ratio of 2.5:1 (Table 1). Patients’ age at presentation ranged from 5 to 70 years (average 29). Only 23 patients were below the age of 20 years (Table 1).

**Presentation.** The initial presentation was intracranial hemorrhage in 52 patients (53.6%). Epilepsy was the 2nd most frequent presentation, occurring in 32 patients (33.0%). Other symptoms, including progressive neurological deficit, headache, dizziness, and trigeminal neuralgia, occurred in 13 patients (13.4%) (Table 1).

<table>
<thead>
<tr>
<th>Age</th>
<th>Hemorrhage (M/F)</th>
<th>Epilepsy (M/F)</th>
<th>Headache (M/F)</th>
<th>Other (M/F)</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>&lt;10</td>
<td>3 (0/3)</td>
<td>-</td>
<td>-</td>
<td>-</td>
<td>3</td>
</tr>
<tr>
<td>10-19</td>
<td>12 (7/5)</td>
<td>8 (2/6)</td>
<td>-</td>
<td>-</td>
<td>20</td>
</tr>
<tr>
<td>20-29</td>
<td>17 (13/4)</td>
<td>9 (9/0)</td>
<td>3 (3/0)</td>
<td>2 (2/0)</td>
<td>31</td>
</tr>
<tr>
<td>30-39</td>
<td>13 (10/3)</td>
<td>6 (5/1)</td>
<td>1 (1/0)</td>
<td>3 (2/1)</td>
<td>23</td>
</tr>
<tr>
<td>40-49</td>
<td>6 (5/1)</td>
<td>3 (2/1)</td>
<td>-</td>
<td>2 (2/0)</td>
<td>11</td>
</tr>
<tr>
<td>50-59</td>
<td>1 (0/1)</td>
<td>2 (2/0)</td>
<td>-</td>
<td>1 (0/1)</td>
<td>4</td>
</tr>
</tbody>
</table>

**TOTAL** | 52 (35/17) | 32 (23/9) | 4 (4/0) | 9 (6/3) | 97 |

**Hemorrhage.** Intracranial hemorrhage was the initial presentation in 52 patients (53.6%). Eight further patients had an episode of intracranial hemorrhage following a period of observation for seizure in 7 patients and for progressive neurological deficit in one patient. Intracerebral hematoma (ICH) was the predominant type of intracranial hemorrhage, occurring in isolation in 24 patients and in association with subarachnoid (SAH) or intraventricular hemorrhage (IVH) or both in another 21 patients. Subarachnoid hemorrhage occurred in 9 patients, and in combination with ICH or both IVH in 13 patients. Intraventricular hemorrhage occurred in 4 patients, and in combination with ICH or SAH or both in 17 patients. Among the 60 patients who had hemorrhage, 48 patients had a single attack. Twelve patients had recurrent bleeding episodes: 10 had 1 recurrence, and 2 had 2 recurrent bleeds. The average period between the first and 2nd attacks of hemorrhage in the 12 patients was 4.43 years, ranging from 1 week to 13 years. The period between the 2nd and 3rd hemorrhage in 2 patients was 6 years in 1 and 7 years in the other. The risk of bleeding from an AVM was tested statistically. A statistically significant increased risk was present in association with age of 30 years and below (P=0.035), with size of the malformation less than 3cm in its largest diameter (P=0.0026), ≤ 2 arterial feeders (P=0.0013) and high velocity flow (P=0.00078). There was no statistically significant relation between bleeding and the gender of the patient, the location of the AVM, the presence or absence of associated aneurysms, or whether the venous drainage was superficial or deep.

**Seizure.** Thirty-two patients (33.0%) presented with seizure. In 2 patients it was a complex partial seizure, in 5 patients it was focal motor or sensory
seizure, and in the remainder of the group it was generalized tonic clonic. The period between the first seizure and establishing the diagnosis of AVM ranged from one week to 24 years, with an average of 4.5 years. Seven of the 32 patients (21.9%) who presented with seizures had hemorrhage later in their follow-up period; 4 of them were men and 3 were women. In 2 patients the hemorrhage occurred within 8 months, and in 5 patients it occurred within 1 to 9 years.

Other presentations. Thirteen patients had other presentations, including progressive weakness (2), headache (4), paresthesia (1), trigeminal pain (3), dizziness (2), and amaurosis fugax (1). Headache was the sole presentation in only one patient, the other patients had headache in association with other symptoms (Table 1).

Location and side. Eighty-eight AVMs were present in the supratentorial region: 67 in a superficial location and 21 deep within the cerebral hemispheres (Table 2); AVM was in the posterior fossa in 13 patients, the cerebellum in 11 and in the brain stem in 2.

Grading of AVM. The AVM was classified according to the Spetzler and Martin grading system (Table 3). Sixteen AVMs were grade I, 43 were grade II, 28 were grade III, 13 were grade IV, and one was grade V. The type of presentation in each grade is given in Table 3.

Associated aneurysms. The associated aneurysms were classified into pre nidal, nidal, and post nidal aneurysm on the basis of the 4 vessel cerebral angiography. Aneurysm within the nidus of the malformation was found in 14 patients (14.4%) at the time of diagnosis of AVM. Seven (50%) of the patients with intranidal aneurysm had intracranial hemorrhage either at presentation or during follow-up; 4 of these patients had recurrent intracranial bleeding. Post nidal aneurysm (venous ectasia) was present in 10 patients (10.3%). Intracranial hemorrhage occurred in 5 patients (50%), either at presentation or during follow-up. Two of the patients with post nidal aneurysm had recurrent intracranial hemorrhage. Arteriovenous malformation was in association with pre nidal arterial aneurysm in 7 patients (7.2%). Except for one patient where the arterial aneurysm was contralateral, all the aneurysms were ipsilateral to the associated malformation. The pre nidal arterial aneurysms were present in the following locations: On the posterior inferior cerebellar artery in 2 patients, middle cerebral artery aneurysm in 2 patients, ophthalmic segment of the internal carotid artery in 2 patients, and anterior cerebral artery aneurysm in one patient. Five patients (71.4%) had intracranial hemorrhage either at presentation or during follow-up, the AVM was always the source of bleeding. None of the associated pre nidal aneurysms ruptured during follow-up. Two arterial aneurysms were managed surgically: one posterior inferior cerebellar artery aneurysm and one ophthalmic–internal carotid segment aneurysm. One arterial aneurysm decreased in size after the associated AVM was occluded by endovascular embolization. Another arterial aneurysm remained unchanged 4 months after partial embolization of the associated AVM. The fate of the rest of the arterial aneurysms is not clear from the studied charts, and no follow-up angiography could
be found in the radiology records.

**Pregnancy.** Of the 28 woman patients, 14 were of childbearing age (15-40 years in the Saudi population) and each had at least one child (range 1-7 children). Three out of these 14 patients were pregnant when they presented with intracranial hemorrhage; 2 of them with subarachnoid hemorrhage and the 3rd with intracerebral hematoma. The attack of intracranial hemorrhage occurred during the last trimester in 2 of the patients, and it was not clear from the record when it occurred in the 3rd (no telephone number for contact). One patient had her first attack of intracranial hemorrhage from the AVM 3 weeks following spontaneous vaginal delivery of a normal baby. One patient had an abortion during follow-up with a residual AVM nidus after an attempt at surgical excision.

**Management.** A multi-disciplinary approach was used to manage the cases. The AVM was managed by surgical intervention in 28 patients, endovascular embolization in 34, and stereotactic radiosurgery in 16. Twenty-six patients did not have any treatment, either because it was so decided by the attending neurosurgeon, or treatment was refused by the patient or the family.

**Surgery.** Surgical intervention was performed in 28 patients, either as a single treatment or in association with other therapeutic modalities. The average age of the patients in the surgical group was 30.6 years (range 2-66 years). Sixteen patients presented with intracranial hemorrhage, 10 with seizure, and 2 with progressive neurological deficit. The malformation was in the hemispheric supratentorial location in 24 patients: 14 on the right side and 10 on the left. In one patient the AVM was located deep within the thalamic-basal ganglia region. In 3 patients the malformation was located within the cerebellum. The AVM was grade I in 8 patients, grade II in 13, grade III in 6, and grade IV in one (Table 3). Two of the patients who presented with seizure were treated with Gamma Knife radiosurgery and endovascular embolization respectively, an attack of bleeding followed before they were treated by surgical excision. Thirty-six operations were performed primarily for the surgical excision of AVM, 3 operations were performed for evacuation of intracerebral hematoma caused by the AVM, 2 for clipping of associated arterial aneurysm, and one for the excision of associated hemangioma on the face. Three patients underwent more than one surgical attempt at obliteration and complete obliteration was confirmed in all. Complete obliteration was confirmed in 21 out of the 28 patients in the surgical group (75%) (Table 3). One patient presented with seizure and underwent partial resection; intracranial hemorrhage occurred after one year of follow-up. He improved to his pre-bleeding clinical condition and 2 years later he was treated with Gamma Knife radiosurgery; at 2 years’ follow-up, MRI of the brain suggested complete obliteration of the malformation. One patient with a large right-sided parietal AVM had preoperative stepwise reduction in the size of the malformation by endovascular embolization over 5 procedures, followed by uncomplicated complete surgical excision. Of the patients with residual AVM one had post-operative proton beam therapy, and in another patient preoperative embolization preceded the surgical attempts. There was no surgical mortality and morbidity occurred in 6 cases (21.4%) (Table 4).

The Karnofski score for the surgical group ranged from 20 to 90 on admission (average 60) and from 40 to 100 (average 85) at the end of the study period. The average Karnofski score on admission for the patients in the surgical group who presented with intracranial hemorrhage, and for those who presented with seizure was 50 and 80 respectively, and at the end of the study period was 87 and 86, respectively. The Karnofski score at the end of the study period had improved in 22 patients, stayed the same in 4 patients, and deteriorated in 2 patients (Table 4).

**Endovascular embolization.** Thirty-four patients underwent endovascular embolization; 2 patients had 2 simultaneous intracranial AVMs. The age of the patients ranged from 2 to 64 years (average 25.6 years). Six AVMs were in the infratentorial space, 4 in the cerebellum and 2 in the brain stem. Of the hemispheric supratentorial AVMs, 13 were on the right side and 7 on the left side. Ten patients had deeply located AVM, 9 in the thalamic-basal ganglia region, and one in the corpus callosum. Three patients had grade I AVM, 10 had grade II, 13 had grade III, and 10 had grade IV (Table 3). Fifteen patients presented with intracranial hemorrhage, 15 with seizures, 3 with progressive motor weakness, and one with progressive dizziness. In 3 out of the 15 patients presenting with seizure, the endovascular embolization was started only after an attack of intracranial hemorrhage. Intracranial hemorrhage occurred for the first time 7 months following the endovascular embolization in a patient who presented with seizure and underwent partial embolization of the AVM. Embolization was performed for 3 patients who presented with neurological deficit in the absence of seizure or intracranial hemorrhage. The clinical condition in 2 of them did not change in the follow-up period, the 3rd patient had increased

<table>
<thead>
<tr>
<th>Outcome</th>
<th>Surgery (n=28)</th>
<th>Embolization (n=34)</th>
<th>Radiosurgery (n=16)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Improved</td>
<td>22 (78.6%)</td>
<td>18 (52.9%)</td>
<td>13 (81.2%)</td>
</tr>
<tr>
<td>No change</td>
<td>4 (14.3%)</td>
<td>10 (29.4%)</td>
<td>2 (12.5%)</td>
</tr>
<tr>
<td>Deteriorated</td>
<td>2 (7.1%)</td>
<td>6 (17.6%)</td>
<td>1 (6.3%)</td>
</tr>
</tbody>
</table>

Table 4 - Clinical status at the end of the study period.
hemiparesis immediately following embolization; in all 3 patients there was residual AVM nidus at the end of the study. In all except 3 of the 34 patients the endovascular obliteration represented the only method of treatment. In 2 patients with a right-sided large AVM who presented with seizure, endovascular embolization was performed as a preoperative adjuvant therapy, after which they underwent surgical intervention. Another patient with a right-sided fronto-parietal grade II AVM presented with dizziness and some headache; he underwent partial obliteration with the endovascular technique, and after 11 months the residual AVM was radiated by Gamma Knife. This patient is in the beginning of his 2nd year of follow-up and obliteration is still not confirmed. Complications occurred in 20 patients (58.8%) with a mortality rate of 2.9% (one patient) and a morbidity rate of 55.9%. Early (≤ 24 hours) post-embolization bleeding occurred in 2 patients, causing death in one of them. Technical error without any additional morbidity occurred in 2 procedures (9.53%): advancement of the glue in a branch of the anterior cerebral artery supplying normal surrounding brain parenchyma in one, and slippage of a coil into the deep femoral artery causing partial obstruction in the other.

Complete obliteration of the AVM nidus was documented in 6 patients in the embolization group (16.7%) (Table 3). The Karnofski score for the endovascular embolization group ranged from 20 to 90 (average 65) on admission and from 0 to 100 (average 80) at the end of the study. The Karnofski score at the end of the study had improved in 18 patients, stayed the same in 10 patients, and deteriorated in 6 patients (Table 4). There were 4 deaths in this group: one was related to the embolization, one died from a recurrent hemorrhage, one had brain stem AVM, and one died from liver failure 5 years following attempted embolization.

**Radiosurgery.** Radiosurgery treatment was provided to 16 patients (16.5%), as a single treatment in 10 patients and in combination with surgery or endovascular embolization in 6 patients. As Gamma Knife is not available in this institution, the radiosurgery was performed elsewhere and the patients returned for follow-up. The average age of the patients in this group was 29 years (range 14-54 years). The AVM was in the hemispheric supratentorial location in 11 patients (7 on the right side and 4 on the left), in the corpus callosum in 2, in the thalamic-basal ganglia region in one, and in the infratentorial location of the cerebellum in two. The AVM was grade I in 3 patients, grade II in 10, grade III in one, and grade IV in 2 (Table 3). The presentation was with intracranial hemorrhage in 10 patients, seizure in 3 patients, and 3 patients presented with progressive neurological deficit and/or progressive dizziness. One of the patients who presented with seizures underwent surgery with partial excision of the AVM; one year later he had an attack of intracranial bleeding and he was treated with Gamma Knife radiosurgery. One other patient with seizure presentation had Gamma Knife radiosurgery treatment for 8 months followed by complete surgical excision. One patient with progressive neurological deficit was treated with endovascular embolization followed by Gamma Knife radiosurgery 10 months later. Another patient with progressive neurological deficit had an intracranial hemorrhage before he was subjected to Gamma Knife therapy; 12 years later he presented with another intracranial hemorrhage and after the hematoma resolved he underwent endovascular embolization. Complete obliteration was confirmed in 7 of the 16 patients in the radiosurgery group (43.8%), in 2 of them obliteration was suggested on the MRI and/or MR angiography but with no confirmation by conventional angiography (Table 5). There was no mortality related to the radiosurgery treatment. Morbidity occurred in 4 patients (25%): 2 patients had decreased visual acuity, one had memory impairment, and one had transient diplopia (Table 5).

The Karnofski score for the radiosurgery group ranged from 30 to 90 (average 60) on admission and 40 to 100 (average 85) at the end of the study. The clinical condition at the end of the study had improved in 13 patients, stayed the same in 2 patients, and deteriorated in one patient (Table 4).

<table>
<thead>
<tr>
<th>Complication</th>
<th>Number</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>Related to Surgical Intervention:</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Moderate motor weakness</td>
<td>2</td>
<td>6</td>
</tr>
<tr>
<td>Status epilepticus</td>
<td>2</td>
<td></td>
</tr>
<tr>
<td>Infection</td>
<td>1</td>
<td></td>
</tr>
<tr>
<td>Subdural collection</td>
<td>1</td>
<td></td>
</tr>
<tr>
<td>Related to Embolization:</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Early I.C. bleed</td>
<td>2</td>
<td></td>
</tr>
<tr>
<td>Permanent neurological deficit</td>
<td>7</td>
<td></td>
</tr>
<tr>
<td>Transient neurological deficit</td>
<td>3</td>
<td></td>
</tr>
<tr>
<td>Status epilepticus</td>
<td>2</td>
<td></td>
</tr>
<tr>
<td>Transient severe headache</td>
<td>3</td>
<td></td>
</tr>
<tr>
<td>Persistent trigeminal pain</td>
<td>1</td>
<td></td>
</tr>
<tr>
<td>Technical error</td>
<td>2</td>
<td>20</td>
</tr>
<tr>
<td>Related to Radiosurgery</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Impaired vision</td>
<td>2</td>
<td></td>
</tr>
<tr>
<td>Impaired memory</td>
<td>1</td>
<td></td>
</tr>
<tr>
<td>Transient diplopia</td>
<td>1</td>
<td>4</td>
</tr>
<tr>
<td>TOTAL</td>
<td></td>
<td>30</td>
</tr>
</tbody>
</table>

The presentation was with intracranial hemorrhage in 10 patients, seizure in 3 patients, and 3 patients presented with progressive neurological deficit and/or progressive dizziness. One of the patients who presented with seizures underwent surgery with partial excision of the AVM; one year later he had an attack of intracranial bleeding and he was treated with Gamma Knife radiosurgery. One other patient with seizure presentation had Gamma Knife radiosurgery treatment for 8 months followed by complete surgical excision. One patient with progressive neurological deficit was treated with endovascular embolization followed by Gamma Knife radiosurgery 10 months later. Another patient with progressive neurological deficit had an intracranial hemorrhage before he was subjected to Gamma Knife therapy; 12 years later he presented with another intracranial hemorrhage and after the hematoma resolved he underwent endovascular embolization. Complete obliteration was confirmed in 7 of the 16 patients in the radiosurgery group (43.8%), in 2 of them obliteration was suggested on the MRI and/or MR angiography but with no confirmation by conventional angiography (Table 5). There was no mortality related to the radiosurgery treatment. Morbidity occurred in 4 patients (25%): 2 patients had decreased visual acuity, one had memory impairment, and one had transient diplopia (Table 5).

The Karnofski score for the radiosurgery group ranged from 30 to 90 (average 60) on admission and 40 to 100 (average 85) at the end of the study. The clinical condition at the end of the study had improved in 13 patients, stayed the same in 2 patients, and deteriorated in one patient (Table 4).
Discussion. Arteriovenous malformation (AVM) is the most commonly encountered symptomatic vascular malformation. It is characterized by abnormal communication between the arterial and venous systems in the absence of normal capillary channels. According to the autopsy study by McCormick, the prevalence of AVM is 0.5%. Clinically, AVM is responsible for 8.6% of subarachnoid hemorrhage, with an annual incidence of 3 per 100,000. No study has been reported on the incidence and prevalence of AVM in Saudi Arabia. From the analysis on subarachnoid hemorrhage by Ammar et al., which was conducted in a single hospital, AVM was the 2nd most common cause of subarachnoid hemorrhage (23%) after aneurysm (41%). Because of the small number of cases and the biased population in the present analysis, and because it is a retrospective study, the incidence rate was not considered. The hospital-based prevalence was found to be 40 per 100,000 admissions. These figures cannot be taken to reflect the true rate of occurrence of the disease in Saudi Arabia. King Faisal Specialist Hospital and Research Center is a tertiary care referral hospital. This may reflect a falsely high incidence among the Saudi population. On the other hand, most of the AVMs are in grade I and grade II lesions, which can be managed surgically with minimal surgery-related morbidity and very low, if any, mortality. This can give a falsely low incidence. Most of the higher grades AVMs were referred to this hospital for endovascular embolization as it was the only center for such therapy in the Kingdom. In later years the Armed Forces Hospital in Riyadh used both the endovascular embolization techniques and Linear accelerator stereotactic radiotherapy for the management of AVM. This may have led to the splitting of the referral pattern to both these centers, and so gives a falsely low incidence. Although the difference between the number of patients of Saudi nationality and those of non-Saudi nationality is highly significant, this result does not reflect the truth. Admission to King Faisal Specialist Hospital and Research Center is, with very few exceptions, restricted to Saudi patients. Most cases of AVM present between the 2nd and 5th decade of life, with less cases presenting in the extremes of age. The Chi square test was applied to verify the age group most at risk of bleeding from AVM. The cut-off point of 30 years of age or less was highly significant (P=0.035). This significance was lost when the age group was increased to 35 or above. When the group below 30 years of age was divided into adults and children the result was statistically non-significant. In this report, men are more affected than women in the ratio of 2.5:1, which matches that in most of the reported series, although equal sex distribution has also been reported in the literature. More men presented with intracranial hemorrhage than women (40:20). This is not a true risk factor for bleeding but rather the overall trend of more men than women being affected with AVM (P=0.216). Intracranial hemorrhage is the most commonly reported mode of initial presentation, followed by seizure and other symptoms. This mode of clinical presentation was reproducible in the present population of patients. The intracerebral location was the most common type of intracranial hemorrhage in our series as well as in other reported studies. The incidence of hemorrhage in relation to each AVM grade is: 75% in grade I, 55.8% in grade II, 39.3% in grade III, and 46.2% in grade IV. Only one case of AVM was in grade V and the presentation was with seizure. These figures indicate that the risk of bleeding is greater with smaller lesions than with larger ones, a fact noted by some other observers. Others do not agree on the difference in risk of hemorrhage regardless of the size. When the size was tested separately to verify the risk of bleeding, it was highly significant (P=0.0026). The mean size of AVM which bled is 2.4 cm, and with other presentations is 3.3 cm. To classify the AVM in our cases, the Spetzler and Martin grading system was used for its simplicity, and its popularity enabled us to compare our findings with other studies in the literature. According to this grading system, 16 AVM were grade I, 43 grade II, 28 grade III, 13 grade IV, and one in grade V. From the literature, the majority of AVM present in grade I or II. Recurrent bleeding occurred in 12 of the 60 patients who had an initial attack of intracranial hemorrhage, giving a rate of re-bleed of 20%. The period of follow-up of the cases varied significantly, ranging from 2 to 21 years (one patient was lost to follow-up with no contact telephone number), with an average of 7.6 years. Because of this difference in duration of follow-up it is impossible to calculate an accurate risk of re-bleeding in our patients. Applying the calculating formula for the risk of re-bleed per year will be as follows: number of patients with 2nd bleed (12) divided by the number of patients with 1st bleed (60) x 100, divided by the number of years of the follow-up period (7.6). The yearly risk of re-bleed is 2.6% per year. Although we have disparity in the follow-up period, we have calculated the annual risk of re-bleeding to fall within the international rate of 2%-4% per year. A common factor in all cases of recurrent bleeding is the delay between presentation and therapeutic management, ranging from 3 months to 9 years with a median of 2 years. The AVM in 7 out of the 12 patients was located in the thalamus and/or basal ganglia, this meant that most of them were of higher grade, making complete obliteration of the nidus difficult to accomplish the first time. Therefore, these patients were kept in the circle of the natural history of the AVM with a gradual increase in the annual risk of bleeding. In this study the presence of...
Intracranial and/or prenal aneurysm appeared to increase the risk of bleeding, an observation shared by others. AVM with associated intranidal aneurysm had greater recurrence of bleeding than those without intranidal aneurysm or with other associated aneurysms. Furthermore, all cases with associated prenal aneurysm that had bleeding were associated with intranidal aneurysm. This clinical observation of increased risk of bleeding in association with intranidal aneurysms was statistically non-significant (P=0.615). The association between AVM and arterial aneurysm is discussed. The cases reviewed in this study may represent both theories. The association of AVM with aneurysm on the same side and on the feeding artery of the malformation seen in 6 out of 7 patients may represent the hyperdynamic theory and the increased flow through the feeding artery leading to weakness of the arterial wall and resulting in aneurysmal dilatation. The single patient with AVM in association with aneurysm on different sides and different arterial territory may represent the theory of the chance association of more than one congenital anomaly at the same time.

The subject of AVM during pregnancy has been discussed in the literature. There is no agreement on whether or not pregnancy increases the risk of bleeding. In the present study, although 3 out of the 14 women of childbearing age had bleeding during pregnancy, the remainder had at least one child during their follow-up period with no bleeding during pregnancy. Moreover, those who had bleeding during pregnancy completed their pregnancy with no further bleed episodes. Certainly no conclusion can be drawn from this small number of patients.

Surgical intervention was only applied to 29% of the patients in this series. Compared with the literature this is a small number. The preferred and first line of therapy for AVM regardless of the grade is surgical excision, either alone or in conjunction with other therapeutic modalities. The use of therapeutic modalities such as endovascular embolization and radiosurgery is usually reserved for those cases that cannot be surgically obliterated, mostly because of their high grade or their situation in an eloquent area of the brain. In the present study almost 50% of the cases subjected to endovascular obliteration and/or radiosurgery were grade I or II. The surgical resection in the present study gave the highest obliteration rate (75%) of the different therapeutic modalities. Results as high as 97% obliteration regardless of the grade have been reported. The obliteration rates after radiosurgery are time-dependent. The obliteration rate reported in the literature after a latent period of three years ranged from 73% to 86%. In the present study the obliteration rate with radiosurgery at the end of the study period was 43.8%. This cannot be compared with the rate reported in the literature as only a few of our patients are three years or more after the radiosurgery while the others are still in the latency period. The obliteration in the group in the present study treated by endovascular obliteration is 16.7%. This is comparable with the rate of obliteration in reported series composed of a similar collection of different AVM grades, ranging from 7.2% to 20%. The clinical status of the patients at the end of the study period generally improved regardless of the treatment modality used. This may be explained by the natural improvement resulting from resorption of the intracerebral hematoma, which has also been documented in the literature. The complication rate from the different therapeutic modalities was as follows: 21% surgical morbidity, compared with 14% in the literature, 59% complication rate following endovascular embolization. This included the cases with technical problems, which did not give rise to deterioration in the clinical status of the patients, and transient severe headache which was not secondary to cerebrovascular accident. If these are excluded the complication rate will be about 40%, still high compared with 14%-24% in the literature. 18% complication rate following radiosurgery in 16 patients, a high rate compared with that given by Steiner et al of 3.6% over 247 patients.

In conclusion, Arteriovenous malformation is not an uncommon disease in Saudi Arabia. The hospital-based frequency was established in this study to be 40 per 100,000 admissions. Men are affected more than women (more than twice as many men as females in the present study). The most common mode of presentation is with intracranial hemorrhage and in the younger age group, which was also the case in the present study. Although some patients have presentation other than hemorrhage, they still have the risk of bleeding from the malformation if complete obliteration is not secured. Although several therapeutic modalities are available to treat such a disease, the ideal solution should be tailored for each case. The management is best when the case is discussed by a team comprising neurosurgeon, interventional neuroradiologist, and radiation therapist. Regardless of the clinical presentation, whenever possible active management should aim for complete obliteration of the malformation. The surgical resection in this study gave the highest obliteration rate, which concurs with the literature. We agree with the general opinion that the first line of treatment of AVM should be surgical excision, either alone or in various combinations with other therapeutic modalities. However, the development of new microcatheters (whether flow guided or directed via a guidewire) and new embolic material (especially the liquid agents IBCA and NBCA) have extended the scope in endovascular embolization to accommodate AVM which were considered.
dangerous to embolize because of inability to catheterize the feeding artery to AVM without injecting into an artery supplying the adjacent brain tissue. Similarly in radiosurgery: because of the development of new collimators, which can accommodate different AVM shapes and angles without subjecting the surrounding brain structure to the same dose of radiation, larger-sized AVM are being considered eligible for radiosurgery therapy.

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References

