The Natural History and Management of Intracranial Dural Arteriovenous Fistulae

Part 2: Aggressive Lesions

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Summary

The natural history of aggressive intracranial dural arteriovenous fistulae (ICDAVF) is unknown. Despite this, the recently proposed classification scheme of Borden et al (Borden*) has the potential to predict aggressive lesion behavior after presentation for any lesion, but has so far been untested.

In addition, they discuss a new but logical treatment strategy for aggressive ICDAVF based on the elimination of retrograde leptomeningeal venous drainage (RLVD). Our similar philosophy and substantial experience with these lesions, provides a unique opportunity to test these hypotheses.

A cohort of 46 Borden* grade II and III ICDAVF was selected from a series of 102 ICDAVF seen at a single institution between 1984 and 1995. Patients with these lesions, presumed to have an aggressive course were all offered treatment. Conservative therapy was chosen by 14 (30%) patients, 22 (47%) had surgery, and 20 (43%) had embolisation either as sole treatment or prior to surgery. During the follow-up period (249 lesion months) for the conservatively treated group, four (29%) patients died. Excluding presentation, these patients were observed to have interval rates of intracranial hemorrhage (ICH), non haemorrhagic neurological deficit (NHND), and mortality, of 19.2%, 10.9%, and 19.3% / lesion year respectively.

The 11 patients who had embolisation alone were followed for a total of 344 months after treatment. All nine patients who had lesion obliteration, or subtotal obliteration with elimination of RLVD, as confirmed by angiography, experienced improvement or complete clinical recovery. Two patients had subtotal obliteration without elimination of RLVD. One died from interval ICH and the other experienced a delayed NHND.

Twenty-five surgical operations were performed on 23 ICDAVF in 22 patients. Resection of the ICDAVF was performed in 9 patients, and 16 patients were treated with surgical disconnection alone. Complications occurred in 3/9 (33%) patients who had their lesions resected and none of the disconnected group. Failure to achieve angiographic obliteration of RLVD in 2 patients treated with resection was associated with an adverse outcome in both cases (death, and interval NHND). All 16 (100%) of the disconnected group were shown to have undergone angiographic obliteration with excellent clinical outcome.

Untreated, Borden* grade II and III ICDAVF have a poor natural history. Also, persistence of RLVD after inadequate treatment results in adverse outcomes. Embolisation usually improves the safety of surgical access and may lead to obliteration on its own in some cases. For the aggressive ICDAVF, surgery is required in most cases, and our data confirm that surgical disconnection alone results in cure of all Borden* grade III ICDAVF, and in grade II lesions, if not cure, conversion to a benign grade I lesion.
Introduction

Aggressive intracranial dural arteriovenous fistulae (ICDAVF) present a particular diagnostic and management challenge for physicians involved in their care. The clinical hallmark of an aggressive ICDAVF is its tendency to cause intracranial haemorrhage (ICH), non-haemorrhagic neurological deficit (NHND), or death. Minor NHND such as ptosis or oculomotor paralysis are exceptions.²³

Although ischaemia from arterial steal may produce cranial nerve impairment, the role of the venous system is dominant in determining both the nature and site of both focal and global neurological manifestations because of retrograde venous flow, thrombosis, ischaemia or mass effect.¹⁴-¹⁵

Associations between aggressive clinical presentation and certain factors have been noted in the literature. These include: leptomeningeal venous drainage, venous ectasia, galenic venous drainage, dural venous sinus (DVS) restrictive disease, and nidus location.⁴,⁵,⁶,⁷,⁸,⁹,¹⁰,¹²,¹³,¹⁴,¹⁵

Classification schemes have also been developed in an attempt to identify patterns of venous anatomy seen on angiography that may suggest lesion aggressiveness. The original classification of Djindjian and Merland, has been subsequently expanded and refined by Cognard et al.² Based on the same model, Borden et al developed a more streamlined three tier classification in which Grade II and III lesions are proposed as dangerous because they possess retrograde leptomeningeal (or subarachnoid) venous drainage (RLVD).¹⁵

Despite the value of individual factors and grading scales in predicting presenting clinical manifestations, little is known about the natural history of these lesions after diagnosis. Just as concerning is the lack of consensus regarding the indications for treatment of ICDAVF which range from active management of all cases²⁴ to a more selective approach based on known risk factors.⁴,⁵,¹⁰,¹⁴,¹⁸. In addition, a confusing range of therapies are available including intermittent manual compression, transarterial or transvenous embolisation and surgery. Surgical resection is often associated with high morbidity and mortality even in experienced hands.¹⁹,²⁰ Recently, Borden et al suggested that surgical disconnection of the retrogradely filling leptomeningeal vein should be an effective treatment for their grade II and III (aggressive) ICDAVF.¹⁵ The efficacy of this same approach in the management of spinal dural arteriovenous fistulae would suggest disconnection to be a logical treatment strategy for intracranial lesions also.²³

The aims of our study were therefore (i) to examine the behaviour after presentation, of a cohort of aggressive ICDAVF chosen on the basis of the classification of Borden et al (Borden*) (grade II and III) from a large heterogeneous single institution experience, and (ii) to assess the efficacy of elimination of RLVD, in particular by way of surgical disconnection, as the simple and logical treatment for grade II and III ICDAVF.

Materials and Methods

One hundred and two ICDAVF in 98 patients were seen by the University of Toronto Brain Vascular Malformation Study Group between 1984 and July 1995. Forty-seven of these were classified as Borden* grade II and III lesions and included in this study. Retrospective data were obtained by review of the hospital files and collected prospectively from 1991.

Clinical information was collected on each patient including age, date of presentation and onset of symptoms or signs, presenting clinical features, prior treatment, and potential predisposing factors. Aggressive clinical features were considered to be ICH, NHND (except orbital venous hypertension and local cranial nerve phenomena secondary to a cavernous sinus lesion), or death. Each patient's angiographic studies were independently reviewed by two experienced neuroradiologists. A grading according to the Borden* classification was assigned, in addition to recording precise details of the nidus location, arterial supply, and venous morphology.

An ICDAVF was considered to be a grade II lesion if the nidus drained into leptomeningeal veins in addition to draining into a dural venous sinus or meningeal vein. The nidus of a grade III lesion drained only into leptomeningeal veins. Grade I ICDAVF, which drained only into a dural venous sinus or meningeal vein were not considered in this study.

All grade II and III lesions were considered to be dangerous and to warrant treatment. The goal of treatment was to eliminate RLVD, which in grade III lesions was expected to result
in cure, and in grade II lesions, cure in many cases, or conversion to a benign grade I lesion. Treatment was refused by 14 patients. Embolisation was chosen as the primary mode of treatment only if angiographic features were conducive to obliteration with safety.

That is, easy and close access to a small nidus with via a small number of large external carotid feeders. Transarterial embolisation was otherwise recommended as a preoperative measure to reduce blood supply to facilitate surgical access. Surgery was recommended in the majority of cases. Total nidus excision was performed nine times on eight patients. Surgical disconnection, used exclusively in the latter half of our review period, was performed in 16 cases, one of which had had an attempted resection in the past. This procedure involved division of the retrogradely filling leptomeningeal vein(s) only, between clips placed just distal to its exit from the nidus.

Clinical follow up was available on 42 (91%) patients. The majority of patients returned to the clinic for personal assessment on a regular basis, whilst data on others were collected via phone interviews corroborated by family physician assessments. Interval events or changes in clinical status were recorded. Outcome was also graded as clinical recovery, major and minor improvement, no change, deterioration and death. Post treatment angiography was performed on all patients who received active treatment except one.

For statistical analysis of data we used chi square testing of contingency tables.

**Results**

Forty-four patients harboured 47 Borden* grade II and III ICDAVF. There were 32 men and 12 women, with mean age of 60 years. Twenty-nine ICDAVF (62%) were grade III and 18 (38%) were grade II. For this group, the lesion nidus was most commonly found to be cavernous sinus (CS), tentorial, or transverse sinus (TS) in location (table 1). There were no grade III CS lesions.

When considered in terms of the full spectrum of ICDAVF (including grade I lesions not included in this study), the locations most likely to harbour grade II and III lesions were the anterior cranial fossa (100%), tentorium (100%), and foramen magnum (60%) 8

A potential etiologic factor was found in the past history of 16 patients. These included significant antecedent trauma in 11, local surgery in one, tumour involving the sinus in two, and infection in two others.

**Presentation**

A total of 30 (64%) grade II and III ICDAVF presented in an aggressive fashion. Intracerebral haemorrhage was a presenting clinical feature in 2/18 (11%) of grade II, and 14/29 (48%) of grade III ICDAVF. NHNDs were found in 5/18 (28%) grade II, and 9/29 (32%) grade III patients. Focal NHNDs were consistent with brain dysfunction in the region normally drained by the leptomeningeal vein now subjected to retrograde flow.

**Treatment and Outcome**

Thirteen patients refused treatment of any kind, 22 had surgery, of which nine received preoperative embolisation, and eleven patients received embolisation alone.

**Conservative Group**

An additional patient was followed for 61 months prior to consenting to embolisation therapy and is included for this period to give a total of 14 ICDAVF in 14 patients. These lesions were followed for a total of 249 lesion months.

<table>
<thead>
<tr>
<th>Borden* Grade</th>
<th>II</th>
<th>III</th>
</tr>
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<tbody>
<tr>
<td>Location</td>
<td></td>
<td></td>
</tr>
<tr>
<td>ACF</td>
<td>4</td>
<td>0</td>
</tr>
<tr>
<td>Cavernous sinus</td>
<td>30</td>
<td>7</td>
</tr>
<tr>
<td>Foramen magnum</td>
<td>5</td>
<td>3</td>
</tr>
<tr>
<td>SSS</td>
<td>2</td>
<td>1</td>
</tr>
<tr>
<td>MCF</td>
<td>2</td>
<td>1</td>
</tr>
<tr>
<td>Tentorium</td>
<td>14</td>
<td>3</td>
</tr>
<tr>
<td>Transverse sinus</td>
<td>45</td>
<td>7</td>
</tr>
<tr>
<td>Total</td>
<td>102</td>
<td>18</td>
</tr>
</tbody>
</table>

ACF = anterior cranial fossa; SSS = superior sagittal sinus; MCF = middle cranial fossa.
(mean 25). During this time four (29%) patients died and one deteriorated after further adverse interval events (e.g. ICH or NHND), one was unchanged and four patients experienced improvement with no new interval events. Excluding presentation an interval ICH rate of 19.2% / lesion year was observed. The interval NHND rate was 10.9% / lesion year. The mortality was calculated to be 19.3% / lesion year.

Emboliisation

Emboliisation was the sole treatment modality used on 11 ICDAVF in 11 patients. These were followed for a total of 359 months (mean 33), of which 344 months were post treatment. All nine patients who had complete angiographic obliteration or subtotal obliteration with elimination of RLVD experienced improvement or complete clinical recovery (table 2). Worsening of short term memory in one patient was felt to be the only complication.

<table>
<thead>
<tr>
<th>Grade (n)</th>
<th>Angiographic result</th>
<th>Clinical result</th>
</tr>
</thead>
<tbody>
<tr>
<td>II (8) AO</td>
<td>STO/RLVD NEG</td>
<td>SR/IMP DET/DEATH</td>
</tr>
<tr>
<td></td>
<td>STO/RLVD POS</td>
<td></td>
</tr>
<tr>
<td>III (3) AO</td>
<td></td>
<td></td>
</tr>
</tbody>
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AO = Angiographically obliterated; STO = subtotal obliteration; RLVD = retrograde leptomeningeal venous drainage; SR = symptom/sign resolution; IMP = improved; DET = deteriorated; POS = positive; NEG = negative.

Surgery

Twenty five surgical operations were performed in 22 patients with 23 ICDAVF. Sixteen of these operations were surgical disconnection (SD) only, and nine were surgical resection (SR). In two patients, initial SR failed to eliminate RLVD. One of these required a second SR, and the other went on to SD. Three other operative complications occurred in the SR group. These were a lower motor neuron facial nerve palsy following a subtemporal approach and SR of a tentorial / petrous ridge lesion, a left homonymous hemianopia after SR of a tentorial lesion, and meningitis in association with a cutaneous CSF fistula after SR of a foramen magnum lesion. There were no complications in the six patients who underwent SD.

Nine of these patients received emboliisation prior to surgery. This occurred immediately before the procedure in three cases, or between four and 60 months earlier in six cases. These later cases were delayed because of initial patient reluctance to proceed to surgery despite persistent RLVD. Angiographic transformation from a grade II to a grade III lesion following emboliisation was noted in one ICDAVF, and accompanied by progressive visual deterioration in that patient over the subsequent month.

Clinical follow-up was available in all surgical cases for a total of 734 months (mean 33) from presentation, and 531 months (mean 24) from surgical intervention. Delayed angiographic follow-up was obtained in all patients except one SR patient. All 16 patients (100%) who ultimately had SD were demonstrated to have had angiographic obliteration of their lesions. In addition, all had a favorable outcome with follow-up assessment demonstrating clinical improvement.

<table>
<thead>
<tr>
<th>PROCEDURE</th>
<th>N</th>
<th>ANGIOGRAPHIC OUTCOME</th>
<th>CLINICAL OUTCOME</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td>AO / RLVD PERSISTS</td>
<td>GOOD INTERVAL ICH</td>
</tr>
<tr>
<td>Disconnect</td>
<td>16</td>
<td>16</td>
<td>16</td>
</tr>
<tr>
<td>Resection</td>
<td>9</td>
<td>6</td>
<td>7</td>
</tr>
</tbody>
</table>

AO = Angiographically Obliterated; RLVD = Retrograde Leptomeningeal Venous Drainage; ICH = Intracranial Haemorrhage; NHND = Non Haemorrhagic Neurological Deficit; Complic. = Complications.
improvement or recovery in 21/22 patients. One patient was unchanged with a static neurological deficit from a previous haemorrhage. Of nine SR procedures performed, seven had a favorable clinical outcome. Six of these patients (6/7) had angiographic follow-up, which demonstrated lesion obliteration in all cases. Two patients had incomplete SR procedures with failure to eliminate RLVD. Both had adverse interval events: one an ICH, and the other a progressive NHND (table 3).

Presence of Persistent RLVD

Those patients (Group A) who had persistent RLVD after presentation, either because of conservative treatment, or in association with subtotal therapeutic intervention, were presumed to pose a special risk. They were compared with patients (Group B) who had had successful elimination of RLVD at surgery or embolisation with resulting complete lesion obliteration or conversion to a grade I lesion. In group B (RLVD eliminated) there were no adverse interval events and no mortality. In group A (RLVD persisting), there were five deaths, and non fatal ICHs and NHNDs occurred in two and one patient(s) respectively, giving an adverse interval event rate of 27.3% / lesion year (figure 1). The interval haemorrhage rate in group A was 20.5% / lesion year.

Discussion

Conservative Management, Natural History, and RLVD

The natural history of Borden* grade II and III ICDAVF is dismal. We have already established the accuracy of this classification scheme in predicting behavior at presentation, with 2% grade I, 39% grade II, and 79% grade III ICDAVF presenting in an aggressive fashion 30. The more important issue, that is, the nature of their behavior after presentation, has until now not been established clearly. Cognard et al followed no more than 50% of their 94 grade IIb to V patients (Borden* II and III) for a mean of 52 months. Although providing good overall frequency data for individual aggressive fea-
tures, they did not discriminate for post presenta-
tion events nor indicate details of numbers treated. Fernand et al were able to contribute
to the natural history of benign ICDAVF only, with their long term follow-up of 43 lateral
sinus lesions.

The 14 patients in our series who survived presentation and subsequently refused treat-
ment have provided a unique insight into the behavior of Borden grade II and III ICDAVF. Untreated, our data suggest patients harbouring these aggressive ICDAVF will face a post presentation interval ICH rate of 19.2% / lesion year, NHND rate of 10.9% / lesion year, and mortality of 19.3% / lesion year.

Borden et Al deliberately structured their classification scheme based on the central theme of RLVD. They recognize, along with many authors, that RLVD is the essential element leading to ICH, NHND, and death. We believe that persistent occurrence of aggressive interval events in untreated grade II and III patients is closely linked to this key issue.

Even more conclusive data regarding the importance of RLVD in influencing outcome, come from those patients who had treatment which has failed to eliminate RLVD despite major changes to either the nidus or arterial input. Cognard et Al reported three fatal haemorrhages during the follow-up period in 19 patients with grade III or IV (Borden grade III) lesions who had embolisation which was subtotal and had failed to obliterate RLVD.

In our series, all four patients (100%) who had persistence of RLVD after unsuccessful treatment, went on to have an adverse clinical event: two NHND, and two ICHs; one fatal. In contrast, three patients who had their RLVD eliminated despite incomplete lesion obliteration, all had favorable courses. Examining all patients with persistent RLVD either because of inadequate or conservative treatment, the calculated incidence of death or non fatal ICH / NHND was 27% / lesion year.

Emboliostion

Emboliostion has a major role to play in the management of Borden grade II and III IC-
DAVF either as a sole treatment modality or as an adjunct to surgery.

The most appropriate method, be it transar-
terial or transvenous is dependent upon the les-
ion anatomy. Transarterial external carotid emboliostion is an effective technique with high
reported cure rates of 72% reported by Picard et Al and 78% by Halbach et Al, however
many of these patients were Borden grade I lesions, or cavernous sinus lesions, which have a
tendency to thrombose with flow reduction.

Particulate emboli such as polyvinyl alcohol or liquid adhesive may be selected based on the
treatment goal. Particulate emboli are recognize
ted to be safe, but treated patients have a
higher recurrence rate with lesion recanaliza-
tion. Liquid adhesives (cyanoacrylates) tend
to produce higher and more permanent cure
rates but with more morbidity, usually because
of reflux into significant anastomoses between
the external carotid and anterior or posterior
circulations, or with inadvertent sinus penetra
tion which may direct greater flow back into a
patent leptomeningeal vein. The transvenous
route, using either liquid adhesives or thrombo
genic coils is also appropriate in selected cases.

Some authors have advocated that complete occlusion of the involved segment of dural sinus
will result in cure in many cases. Patient selec
tion is important and requires the fistula to be
in the wall of a patent sinus with which it
communicates (Borden grade II), but the sinus must not contribute to venous drainage of the brain.

This technique can be hazardous if it leads to
inadvertent trapping of the involved sinus seg-
ment resulting in diversion of the entire shunt
into the draining leptomeningeal vein. One of
our grade II CS patients was converted to a
grade III following his third transvenous emboliostion and required urgent surgical discon
nection one week later, because of progressive
venous congestive encephalopathy.

We maintain that the goal of treatment for
grade II and III ICDAVF is elimination of
RLVD. It may be possible to “disconnect” RLVD via a transvenous route in a grade II pa
tient, with conversion of the lesion to a benign
grade I ICDAVF or cure in some patients
(three in our series) with successful complete
sinus occlusion. We would advocate the goal of
elimination of RLVD only with DVS preserv-
alection. If this is not possible using endovascular
means, then surgical disconnection of the
RLVD can achieve the goal simply without
sinus sacrifice.
Figure 2  A) Axial CT brain image with intravenous contrast demonstrating multiple dilated intraparenchymal vessels within the right occipital lobe (arrows). B) Selective right internal maxillary artery (wide arrow) injection demonstrating a transverse sinus ICDAVF with significant supply from a posterior branch of the middle meningeal artery (thin arrow). Blood exits via a single leptomeningeal vein (curved arrow) which in turn reflexes into multiple intraparenchymal venous tributaries. C) Same as figure 2B but venous phase, demonstrating the progress of retrograde venous flow into multiple intraparenchymal veins (arrowheads), and from these into the occipital (open arrow) and transverse (curved arrow) sinuses (anterogradely), and the straight sinus / vein of Galen (very curved arrow) (retrogradely). D) Selective right internal maxillary artery (arrow) injection performed postoperatively demonstrating complete angiographic obliteration after surgical disconnection.
Transarterial embolisation was particularly helpful for preoperative nidal or prenidal occlusion to reduce blood supply and facilitate surgical access to the draining vein. In three grade III lesions it was possible to obtain a cure using a combination of particles and glue, with penetration into a small nidus via the transarterial route.

**Surgery**

The goal of complete surgical excision for ICDAVF has been plagued by extensive blood loss, and high morbidity and mortality. Sundt and Piepras, in their series of 27 surgical resections, calculated blood loss of 300 mL/min on several occasions, with two deaths on the operating table from blood loss, and two postoperative haemorrhagic venous infarctions requiring surgical evacuation.

Despite this they stressed that “every attempt should be made to excise the malformation as completely as possible with the initial procedure.” Surgery was ultimately required as the definitive procedure in 23 (70%) of the grade II and III ICDAVF actively treated in our series. Eight of these underwent surgical resection, from which there were three complications, and two patients required second procedures to eliminate RLVD.

Surgical disconnection should be the surgical procedure of choice for grade II and III ICDAVF. Preoperative transarterial embolisation facilitates a relatively bloodless approach to the nidus. With good superselective preoperative diagnostic angiography, a small dural opening is made to gain access to the arterialized leptomeningeal vein(s). An aneurysm clip or titanium hemostatic clip is then used to occlude the vein(s) just as it exits the nidus. The distal vein is then coagulated and divided (figure 2).

Surgical disconnection of the draining leptomeningeal vein for treatment of spinal dural arteriovenous malformations was used successfully in 1916 by Elsberg. Eventually, this became the standard simple treatment for these lesions. It was not until recently that a similar treatment strategy consisting of disconnection alone was reported for intracranial dural arteriovenous fistulae. Grisoli et al described four tentorial lesions consistent with Borden grade III ICDAVF which were treated successfully using this technique. Borden et al in 1995 proposed the logic of surgical disconnection of RLVD and reported five cases where it was applied as sole treatment.

To our knowledge, we are the first group to evaluate this technique in a significant number of patients with complete angiographic and clinical follow-up. In our hands the procedure was safe (0% morbidity and mortality), clinically efficacious, and resulted in a 100% obliteration proven by delayed angiography. In addition, there was almost no blood loss and operative times were extremely short. By definition, as Borden et al predicted, all grade III lesions should go on to obliteration. For grade II lesions, just as with embolisation, surgical disconnection will result in conversion to a benign grade I lesion.

In our experience, effective preoperative transarterial embolisation in grade II patients combined with surgical disconnection, also resulted in cure.

In summary, lesions which have RLVD (Borden II and III) have a poor natural history. Death, NHND, and ICH will continue to occur when RLVD persists as a result of subtotal treatment. All Borden grade II and III ICDAVF should be treated and the goal of therapy is elimination of RLVD. Embolisation itself may be successful in achieving this goal. Otherwise, embolisation plays a vital role in flow reduction for safe surgical access. Rather than surgical excision, the simple and logical treatment for these lesions is surgical disconnection, which will lead to cure or conversion to a benign grade I lesion.

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*See also editorial comment: P. Lasjaunias; Angioarchitecture and Natural History of dural arteriovenous shunts pags. 313-317.*
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