Carotid and Transcranial Color-Coded Duplex Sonography in Different Types of Carotid-Cavernous Fistula

Yu-Wei Chen, MD; Jiann-Shing Jeng, MD; Hon-Man Liu, MD; Bao-Show Hwang, BS; Win-Hwan Lin, RN; Ping-Keung Yip, MD

Background and Purpose—Patients with carotid-cavernous fistula (CCF) may undergo direct or indirect shunting. Ultrasonography has value that is complementary to angiography in the assessment and follow-up of these patients. The aim of this study was to characterize findings provided by carotid duplex sonography (CDS) and transcranial color-coded duplex sonography (TCCD) in patients with different types of CCF.

Methods—CDS and TCCD were independently performed by technologists and neurologists. Digital subtraction or MR angiography was interpreted by a neuroradiologist. Ultrasonographic studies were categorized into 4 types: I, direct shunting only; II, direct shunting with a carotid aneurysm; III, indirect shunting only; and IV, mixed (direct and indirect) shunting. In addition to carotid and intracranial flow velocities, volume, and pulsatility, other direct and indirect ultrasound signs of shunting were evaluated. The direct sign of CCF was a mosaic flash detected by TCCD. Alteration of hemodynamic parameters on CDS and demonstration of draining veins with the use of TCCD were considered indirect signs.

Results—Fifteen patients (8 men, 7 women) were included in the study. According to angiographic results, patients in ultrasonographic classification types I (n=7) and II (n=3) corresponded to type A of Barrow’s classification. Patients with type III (n=8) were Barrow’s type C. Type IV (n=1) had a combination of Barrow’s types A and C. On ultrasound, both direct and indirect signs were seen in types I, II, and IV CCF. The presence of a 2-colored oval mass divided by a zone of separation without turbulence differentiated type I from type II CCF. All patients with type III CCF had indirect signs, and only 1 patient had direct signs on TCCD. Abnormal TCCD findings were most commonly seen through the transorbital window (100%), followed by the transtemporal window (63%) and transforaminal window (40%).

Conclusions—If only indirect ultrasonographic signs of CCF are present, TCCD can be used to predict an indirect CCF type on the basis of the origin of the fistula. With direct communication between carotid artery and cavernous sinus, both direct and indirect ultrasonographic signs can be found. The combination of CDS/TCCD may provide a noninvasive and reliable way to classify patients with CCF. (Stroke. 2000;31:701-706.)

Key Words: aneurysm ■ cavernous sinus ■ fistula ■ ultrasonography, Doppler, duplex, transcranial

Carotid-cavernous fistula (CCF), which is the abnormal communication between the carotid arteries and the cavernous sinus (CS), can be classified into direct, indirect, and mixed types on the basis of the origin of the fistula. The direct type is characterized by a fistula between the internal carotid artery (ICA) and the CS. The indirect type shows a shunting flow between the meningeal branches of either the ICA or external carotid artery (ECA) and the CS. In addition, mixed types with contributions from both the ICA and ECA have also been reported. Barrow et al proposed an anatomic-angiographic classification for spontaneous CCF. Type A is a direct high-flow shunt between the ICA and CS. Type B is a dural shunt between meningeal branches of the ICA and the CS. Type C is a dural shunt between meningeal branches of the ECA and the CS, and type D is a dural shunt between meningeal branches of both the ICA and ECA and the CS. CCF can arise spontaneously or be a complication of head injury. Another kind of vascular anomaly around this area is aneurysmal formation of the ICA or its branches. Infrequently, CCF is associated with an ICA aneurysm after a severe head injury or due to congenital connective tissue defects. An aneurysm superimposed on a CCF may pose problems of diagnosis and management. Catastrophic hemorrhage can result from the aneurysm rupturing either spontaneously or during intervention. These conditions require rapid recognition and prompt treatment. The more complicated condition of an associated aneurysm with CCF was not
TABLE 1. Proposed Ultrasonographic Findings in Different Types of CCF

<table>
<thead>
<tr>
<th>Type</th>
<th>I (Direct CCF)</th>
<th>II (Direct CCF With Aneurysm)</th>
<th>III (Indirect CCF)</th>
<th>IV (Mixed CCF)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Direct signs*</td>
<td>+</td>
<td>+</td>
<td>–</td>
<td>+</td>
</tr>
<tr>
<td>Unique aneurysmal findings†</td>
<td>–</td>
<td>+</td>
<td>–</td>
<td>–</td>
</tr>
<tr>
<td>Indirect signs‡</td>
<td>– (ICA)</td>
<td>– (ICA)</td>
<td>+ (ECA)</td>
<td>+ (ICA + ECA)</td>
</tr>
</tbody>
</table>

*Viewing of mosaic flashes through TCCD windows.
†Viewing of 2-colored mass separated by a zone of separation without turbulence on TCCD.
‡Changes of RI and flow volume on CDS (arterial side) and viewing of draining veins of CCF on TCCD (venous side).

considered in Barrow’s classification. On the contrary, the low-flow or indirect dural CCF has a relatively high incidence of spontaneous resolution; therefore, invasive diagnostic measures and treatment might not be necessary. Our previous studies have clearly demonstrated that ultrasonography provides direct imaging of the glomus of CCF itself and differentiates the directness of origin, revealing a relationship between CCF and carotid/cerebral hemodynamics; ultrasonography is suitable for long-term follow-up after intervention. Recent reports have also described the application of ultrasonography in the detection of cerebral aneurysms. As a result of the invasiveness of angiography, some reports have described and advocated the application of carotid duplex sonography (CDS) and transcranial color-coded duplex sonography (TCCD) to insonate the basal cerebral arteries for diagnosis of CCF and other cerebrovascular diseases because of the safety and ease of repetition in follow-up. We tried to delineate further the role of ultrasonography in the diagnosis and classification of different conditions of CCF. The purpose of this study was to define the specific findings of CDS and TCCD in patients with different types of CCF with or without aneurysm formation.

Subjects and Methods
Fifteen patients (8 men, 7 women) with CCF, aged 23 to 60 years with a mean age of 43.2 years, were included in this study. Seven of them were included in our previous report. All patients except 1 had conventional cerebral angiography to prove and classify the vascular origins of CCF. The patient who refused conventional angiography underwent MR angiography instead. All patients received CDS and TCCD studies. CDS was performed with either the Diasonic VST MASTER Series, containing a 10-MHz real-time B-mode imaging transducer and a 6-MHz pulsed Doppler transducer, or the Aloka SSD-3000 Series, containing a 7.5-MHz real-time B-mode imaging transducer and a 6-MHz pulsed Doppler transducer. Arterial diameters, peak systolic velocity, end-diastolic velocity, time-averaged velocity, resistivity index (RI), and flow volume of the extracranial ECA, ICA, and vertebral artery were measured. Special emphasis was placed on RI and flow volume. RI was defined as [(peak systolic velocity) – (end-diastolic velocity)] / (peak systolic velocity). The flow volume was automatically calculated as the product of the time-averaged velocity and the cross-sectional area.

TCCD was performed with a Diasonic VST MASTER Series, which contains a 2.0-MHz real-time imaging transducer and a 2.0-MHz pulsed Doppler transducer. The maximum in situ Doppler energy output intensity was 89 mW/cm² I STPA (spatial peak time average intensity). In our laboratory, blue is traditionally assigned for the flow toward and red for the flow away from the transducer. Deep shades indicated slow mean blood velocities; lighter shades or a change from blue to green and from red to yellow indicated fast mean blood velocities. Every patient was comprehensively evaluated through the transorbital, transtemporal, and transforaminal windows. Transtemporal insonation of patients in the supine position revealed the ipsilateral distal ICA; the proximal parts of the anterior, middle, and posterior cerebral arteries of the circle of Willis; the CS; and sometimes the contralateral cerebral arteries. The examinations through the transorbital window consisted of 2 parts. We first examined the orbital cavity to see whether the ophthalmic veins were engorged and recorded the Doppler characteristics of these veins and then insonated more deeply to investigate the CS. The equipment was used to examine the retro-orbital vessels in the orbital cavity with reduced power for safety requirements. Through the transforaminal window, with patients in the decubitus position, we saw the intracranial vertebral and lower part of the basilar arteries. In addition to the normal intracranial vessels, we tried to insonate CCF and other abnormal vessels through these windows.

The ICA and CS were insonated through the 3 windows to determine whether there was a heterogeneous color mosaic flash suggesting the glomus or the direct sign of CCF. The findings of a 2-colored oval structure divided by a dark zone of separation suggested the existence of an aneurysm. Draining veins could be found through the transorbital and transforaminal windows. Doppler spectral analysis of these vascular structures was recorded. The alterations of the carotid artery hemodynamics (decreased RI and increased blood flow volume) of the feeding arteries by CDS suggested arterial indirect signs. The demonstration of draining veins of CCF by TCCD was thought to be a venous indirect sign. The venous indirect signs were visual proof of reversed superior ophthalmic veins or other draining veins through the transorbital window and the engorged basilar plexus through the transforaminal window. These veins often exhibited a low-resistance and high-velocity turbulent flow pattern during the Doppler spectral analysis.

This was a blind study. Patients were referred to our neurovascular laboratory by clinicians on the suggestion of CCF. Two teams (B-S.H. and Y-W.C., and W-H.L. and J-S.J.) examined each patient, respectively. The results of 1 team were blind to the other, and the final reports were sent to the laboratory director (P-K.Y.). An experienced neuroradiologist (H-M.L.) reviewed the results of angiographic examinations. They were categorized into 4 ultrasonographic types according to the following findings. Type I included patients with direct signs of CCF and/or with indirect signs in the ICA. Type II shared the same findings with type I, plus additional aneurysmal findings. Type III consisted of patients with indirect ultrasound signs only. Type IV included patients with direct signs and indirect signs in both the ICA and ECA (Table 1).

Results
The hemodynamic parameters of CDS in each patient, including RI and flow volume of the bilateral ICAs and ECAs, in
combination with the direct findings of the mosaic flash and aneurysms on TCCD are summarized in Table 2. There were 7 arteries of 5 patients in type I and 3 arteries of 3 patients in type II corresponding to type A of Barrow’s classification. Eight arteries of 7 patients in type III were Barrow’s type C, and 1 patient in type IV had a combination of Barrow’s types A and C. One patient (patient 14) had type III CCF on the right side and type I CCF on the left side. Abnormal changes in the flow volume or RI, ie, arterial indirect signs, were detected in 71% (5/7) of type I CCF, 100% (3/3 and 1/1) of types II and IV, and only 25% (2/8) of type III.

The results of TCCD study are shown in Table 3. Various abnormal findings were discovered through the 3 separate windows with the use of TCCD. The transorbital window could show an engorged superior ophthalmic vein and other draining veins on color-coded imaging with reverse-flow direction and a turbulent flow pattern of low resistance and high velocity. The transtemporal window could demonstrate heterogeneous color mosaic flashes at a depth of approximately 7 cm, with the CS being located just above the bilateral carotid canals. Doppler spectral analysis showed unidirectional or bidirectional turbulent and low-resistance flow patterns within the flashes. Transforaminal examination could disclose engorged vessels leaving the cranium, which were adjacent to the verteobasilar arteries. Unidirectional, turbulent, and low-resistance flow patterns were shown on Doppler spectral analysis (Figure 1). The abnormal findings were more commonly found through the transtemporal and

### Table 2. Findings of CDS and TCCD in Patients With Different Types of CCF

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Age, y/Sex</th>
<th>Cause</th>
<th>Side</th>
<th>ICA FV</th>
<th>ICA RI</th>
<th>ECA FV</th>
<th>ECA RI</th>
<th>Mosaic Flash</th>
<th>Aneurysm</th>
<th>Ultrasound Classification</th>
<th>Barrow’s Classification</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>38/M</td>
<td>Trauma</td>
<td>L</td>
<td>13%</td>
<td>2%</td>
<td>22%</td>
<td>0</td>
<td>+</td>
<td>–</td>
<td>–</td>
<td>I</td>
</tr>
<tr>
<td>2</td>
<td>42/F</td>
<td>Trauma</td>
<td>R</td>
<td>118%*</td>
<td>36%*</td>
<td>66%</td>
<td>24%</td>
<td>–</td>
<td>+</td>
<td>–</td>
<td>I</td>
</tr>
<tr>
<td>3</td>
<td>40/M</td>
<td>Trauma</td>
<td>R</td>
<td>153%*</td>
<td>25%*</td>
<td>15%</td>
<td>9%</td>
<td>+</td>
<td>–</td>
<td>–</td>
<td>I</td>
</tr>
<tr>
<td>4</td>
<td>58/F</td>
<td>Trauma</td>
<td>R</td>
<td>60%*</td>
<td>52%*</td>
<td>25%</td>
<td>9%</td>
<td>+</td>
<td>–</td>
<td>I</td>
<td>A</td>
</tr>
<tr>
<td>5</td>
<td>31/M</td>
<td>Trauma</td>
<td>L</td>
<td>194%*</td>
<td>38%*</td>
<td>48%</td>
<td>12%</td>
<td>+</td>
<td>+</td>
<td>–</td>
<td>I</td>
</tr>
<tr>
<td>6</td>
<td>40/F</td>
<td>Trauma</td>
<td>L</td>
<td>42%*</td>
<td>14%</td>
<td>15%</td>
<td>13%</td>
<td>+</td>
<td>+</td>
<td>–</td>
<td>I</td>
</tr>
<tr>
<td>7</td>
<td>45/F</td>
<td>Spontaneous</td>
<td>L</td>
<td>690%</td>
<td>31%*</td>
<td>27%</td>
<td>3%</td>
<td>+</td>
<td>+</td>
<td>II</td>
<td>A</td>
</tr>
<tr>
<td>8</td>
<td>53/M</td>
<td>Spontaneous</td>
<td>R</td>
<td>5%</td>
<td>0</td>
<td>48%</td>
<td>11%</td>
<td>–</td>
<td>–</td>
<td>–</td>
<td>III</td>
</tr>
<tr>
<td>9</td>
<td>24/F</td>
<td>Spontaneous</td>
<td>R</td>
<td>1%</td>
<td>8%</td>
<td>83%</td>
<td>–</td>
<td>–</td>
<td>–</td>
<td>–</td>
<td>III</td>
</tr>
<tr>
<td>10</td>
<td>60/F</td>
<td>Spontaneous</td>
<td>R</td>
<td>13%</td>
<td>5%</td>
<td>13%</td>
<td>7%</td>
<td>–</td>
<td>–</td>
<td>–</td>
<td>III</td>
</tr>
<tr>
<td>11</td>
<td>58/F</td>
<td>Spontaneous</td>
<td>L</td>
<td>23%</td>
<td>11%</td>
<td>9%</td>
<td>10%</td>
<td>–</td>
<td>–</td>
<td>–</td>
<td>III</td>
</tr>
<tr>
<td>12</td>
<td>37/M</td>
<td>Trauma</td>
<td>L</td>
<td>22%</td>
<td>7%</td>
<td>77%*</td>
<td>1%</td>
<td>–</td>
<td>–</td>
<td>–</td>
<td>III</td>
</tr>
<tr>
<td>13</td>
<td>27/M</td>
<td>Trauma</td>
<td>L</td>
<td>Occluded</td>
<td>45%</td>
<td>–</td>
<td>1%</td>
<td>–</td>
<td>–</td>
<td>–</td>
<td>III</td>
</tr>
<tr>
<td>14</td>
<td>44/M</td>
<td>Trauma</td>
<td>R</td>
<td>10%</td>
<td>20%</td>
<td>15%</td>
<td>0</td>
<td>–</td>
<td>–</td>
<td>–</td>
<td>III</td>
</tr>
<tr>
<td>15</td>
<td>52/M</td>
<td>Trauma</td>
<td>L</td>
<td>425%*</td>
<td>36%*</td>
<td>389%*</td>
<td>14%</td>
<td>–</td>
<td>+</td>
<td>–</td>
<td>IV</td>
</tr>
</tbody>
</table>

FV indicates flow volume; R, right; L, left; +, increase; –, decrease.
*Values = (mean ± 2 SD) or = (mean – 2 SD).
†Case 14 had both type III and type I on either side.

### Table 3. Frequencies of Abnormal Findings Using TCCD Through Different Windows in Patients With CCF

<table>
<thead>
<tr>
<th>Type</th>
<th>No. of Patients</th>
<th>Lesion Sides</th>
<th>Transtemporal Window</th>
<th>Transorbital Window</th>
<th>Transforaminal Window</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>5†</td>
<td>7</td>
<td>7 (100)</td>
<td>7 (100)</td>
<td>2 (40)</td>
</tr>
<tr>
<td>II</td>
<td>3</td>
<td>3</td>
<td>3 (100)</td>
<td>3 (100)</td>
<td>3 (100)</td>
</tr>
<tr>
<td>III</td>
<td>7†</td>
<td>8</td>
<td>1 (12.5)</td>
<td>1 (100)</td>
<td>0 (0)</td>
</tr>
<tr>
<td>IV</td>
<td>1</td>
<td>1</td>
<td>1 (100)</td>
<td>1 (100)</td>
<td>1 (100)</td>
</tr>
<tr>
<td>Overall</td>
<td>15</td>
<td>19</td>
<td>12 (63)</td>
<td>19 (100)</td>
<td>6 (40)</td>
</tr>
</tbody>
</table>

*Percentage based on number of patients.
†Case 14 had both type III and type I on either side.
transorbital windows than the transforaminal window in the type I, II, and IV patients.

In addition to the common abnormal findings as in type I, there were other discoveries in type II patients. Aneurysms arising from the ICA were found in patients 5 and 6 as another complication of head injuries. TCCD showed an additional oval mass with 2 different colors, either red or blue, in different scanning planes. In the plane that transected through the mid aneurysm level, 2 different colors (red and blue) divided by a dark zone of separation were shown simultaneously. Spectral analysis showed no turbulence within either the red or blue area, which was different from the turbulent flow shown in CCF (Figure 2).

The universal findings of type III were engorged superior ophthalmic veins through the transorbital window in all patients. The CCF was directly shown as a color flash in the transtemporal window only in patient 9. None showed abnormal findings through the transforaminal window.

**Discussion**

There are many classification systems for CCF. Barrow et al² provided a detailed anatomic classification of CCF into 4 types on the basis of angiographic findings and origins of shunting. This classification is pertinent to ultrasonography since the hemodynamic changes of CCF can be assessed and expressed with the Doppler flow signals. With the upgrading of cerebrovascular ultrasonography, there are increasingly more applications of CDS and TCCD in the study of intracranial vascular anomalies, which could previously only be definitely diagnosed with cerebral angiography.

Additional findings of a 2-colored oval mass distal to CCF shown on TCCD separated the cases from the commonly occurring direct fistula. These unique findings suggested that these patients suffered from aneurysmal formation of the ICA as well. In most of the reported cases of such combinations, they were secondary to head injury and were usually located distal to the fistula. An aneurysm superimposed on CCF may pose problems of diagnosis and management since early opacification of the CS on angiography may mask the presence of an aneurysm, and its presence may be overlooked unless good-quality subtraction films are routinely obtained. Catastrophic hemorrhage could result from the aneurysm rupturing either spontaneously or during intervention. Although this combination is rare, it requires rapid
recognition and prompt treatment. Therefore, we propose to divide Barrow’s type A into 2 types on the basis of ultrasonographic findings, ie, type I, direct CCF only, and type II, direct CCF with an aneurysm. The findings of our 3 type II patients included the characteristic findings of both CCF and carotid aneurysms. The 2-colored oval masses were located approximately 1 cm distal to the mosaic mass of CCF in the transtemporal window. The spectral analysis showed laminal flow in the former and turbulent flow in the latter. The reported detection rate of aneurysms with the use of TCCD was variable, ranging from 47% to 85%. If the aneurysms were small (<5 mm), thrombosed, or located with an unfavorable scanning plane, the ability of TCCD to detect them was limited. Intensity-dependent color-coded sonography (power Doppler mode) provided supplementary information through the detection of slow blood flow in the aneurysm. Administration of contrast medium may also increase the detection rate. Although TCCD should not be used as a screening procedure for the aforementioned reasons, being familiar with the characteristic findings of aneurysms can help physicians to find more aneurysms. This procedure also offers a noninvasive method for monitoring progressive intra-aneurysmal thrombosis after coil embolization and during the follow-up period of untreatable fusiform aneurysms.

The findings of type III were quite different from those of types I and II. The changes of flow volume and RI were not as conspicuous as with a direct type of CCF. Abnormal values of the feeding ECA were shown in patients 9 and 12 (25%). In the TCCD study, the consistent findings were the abnormal draining of ophthalmic veins through the transorbital windows (100%). Viewing of the mosaic mass via transtemporal windows was only noted in patient 9 (13%, 1/8). Since the shunting flow of the indirect type of CCF is usually less than that of direct ones, the alterations of flow volume in CDS and of Doppler signals in TCCD are therefore less distinct. Patient 9 had increased flow volume in the feeding of ECA, which was also a rare finding in CDS among this group (Table 2). It was the remarkable increase in blood flow that demonstrated CCF with direct signs. On the basis of these findings, we concluded that direct signs on TCCD were uncommon in the patients with indirect CCF and could only be seen when the flow volume of the feeding artery was markedly increased, which could be measured with CDS. This extraordinary condition was not reported in our previous study because of the limited number of cases.

CDS and TCCD may be used as an aid to plan the optimal treatment for each type of CCF patient. In type I patients, coil embolization should be considered first. There is still controversy about the ideal treatment for type II patients. Reddy and Sundt believed that CCF with concomitant aneurysms required direct surgical exploration in most cases. Other investigators believe that a traumatic ICA aneurysm should be treated conservatively because of the high probability of intraoperative rupture and the difficulty in clipping the aneurysm because of a broad neck or a fibrous wall. In type III, an initial conservative approach is indicated because of the high incidence of spontaneous resolution. De Keizer even proposed that angiography may be deferred, considering that a complication rate of 10% must be accounted for in arteriography. In such cases, CDS and TCCD are good tools for follow-up of patients on the basis of changes of symptoms.

The limitations of ultrasonographic classification of CCF include the following. (1) Ultrasonographic examinations are dependent on technology, especially in small and angle-dependent lesions, eg, small CCF or aneurysms. (2) The ability of CDS to differentiate various types of indirect CCF is not perfect when the alteration in hemodynamic parameters is not apparent. In patient 14, the indirect shunting from the
left ECA was not recognized because the changes of hemodynamic parameters of the ECA were not prominent. The limitations of this classification are also true for other indirect ICA shunting lesions (type B of Barrow’s classification), and it cannot be used to differentiate type C from type D because the same changes in hemodynamic parameters occurred.\(^3\) However, the additional tool of TCCD is sensitive in detecting the presence of CCF if venous indirect signs are present. Engorged ophthalmic veins with reverse flow were universal findings in previous reports and the present study,\(^4,10\) and this examination can also be performed with TCCD and even with CDS (with reduced power). Therefore, we suggest that ultrasonographic classification of CCF may be used as a screening procedure in suspected patients to select patients for further investigation.

On the basis of our previous and present findings, when only indirect ultrasonographic signs of CCF are present, TCCD is predictive of an indirect CCF type. When direct signs and arterial indirect signs in the ICA are present, this is indicative of a direct CCF type. The addition of a 2-colored oval mass divided by a dark zone of separation differentiates type II from type I. When the arterial indirect signs are present in the ICA and ECA with the direct signs of CCF, a mixed type of CCF should be considered. These findings can help us in the diagnosis and classification of patients with different types of CCF. The combination of CDS and TCCD provides an excellent method for follow-up after intervention because of CCF and aneurysms.\(^3,11,14,18,19\)

References

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