Spontaneous carotid-cavernous fistulae in Ehlers-Danlos syndrome Type IV

Case report

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Spontaneous bilateral carotid-cavernous fistulae and cervical artery dissection is reported in a 20-year-old woman with Ehlers-Danlos syndrome Type IV. The clinical features of 16 previously published cases of spontaneous carotid-cavernous fistulae associated with Ehlers-Danlos syndrome Type IV are reviewed, for a total of 17 cases. The mean age of the 14 women and three men was 31.6 years. Only direct fistulae were encountered. Diagnostic neuroangiography carried morbidity and mortality rates of 36% and 12%, respectively; neuroradiological treatment resulted in death in one of six patients. The possible value of desmopressin in the management of these patients is discussed. In view of the risks of arterial puncture and surgery, the authors emphasize the importance of early recognition of Ehlers-Danlos syndrome.

Key Words: carotid-cavernous fistula • Ehlers-Danlos syndrome • collagen disorder • embolization • angiography

Spontaneous carotid-cavernous fistulae may be anatomically divided into direct and dural fistulae. Direct carotid-cavernous fistulae are usually high-flow direct communications between the internal carotid artery (ICA) and the cavernous sinus, and are commonly due to rupture of an intracavernous carotid aneurysm into the cavernous sinus. Dural carotid-cavernous fistulae consist of abnormal low-flow communications between dural branches of the internal and/or external carotid artery and the cavernous sinus, and presumably develop secondary to a previous cavernous sinus thrombosis.

Ehlers-Danlos syndrome is a group of hereditary connective-tissue disorders first described in 1668 and is currently divided into nine types. Ehlers-Danlos syndrome Type IV, the so-called "vascular type," was first described as a distinct clinical entity in 1967. The basic molecular defect of this condition is an abnormality of Type III collagen. This type of collagen is one of the three major fibrillar collagens and normally constitutes approximately 40% of blood vessel walls. The gene for the alpha-1 chain of Type III collagen is on the long arm of chromosome 2 (2q31-q32.3). In comparison to the other types of Ehlers-Danlos syndrome, Type IV is the most life-threatening because of its vascular manifestations which are characterized by rupture, dissection, or aneurysm formation affecting large or medium-sized arteries. Associated disorders of the cervicocerebral vessels include carotid-cavernous fistulae, intracranial aneurysms, extracranial cervical artery aneurysms, and carotid artery dissection.

In this paper we report one patient with Ehlers-Danlos syndrome Type IV who developed bilateral carotid-cavernous fistulae, and we review the literature on carotid-cavernous fistulae occurring in patients with this syndrome.

Case Report

This 20-year-old woman suddenly developed severe right retro-orbital pain, followed by a "thumping" noise in her right ear. She had no history of trauma.

Medical History. Since early childhood, the patient had frequently suffered severe spontaneous bruising. At 17 years of age, she had suffered a similar spontaneous
attack of left retro-orbital pain, followed by the signs and symptoms typical of a carotid-cavernous fistula. Evaluation carried out elsewhere included carotid arteriography by direct puncture which demonstrated a left-sided direct carotid-cavernous fistula. Following the procedure, a large hematoma developed in the patient’s neck, causing airway obstruction and requiring emergency evacuation and intubation. A femoral venipuncture resulted in local ooze for 2 days thereafter. To treat the fistula, the left ICA was ligated intracranially, embolized with a piece of muscle, and finally the left ICA was ligated in the neck (Hamby procedure). The patient made a full recovery with complete resolution of all symptoms. The following year she underwent surgery for spontaneous perforation of the colon.

The patient is of English, Irish, and German descent. When she was born, her father was aged 36 and her mother 31 years; the parents are not known to be consanguineous. The mother, who died of bilateral breast cancer at 44 years of age, was not known to have had any signs of Ehlers-Danlos syndrome. The father, sister, and brother were reported to be healthy, at ages 57, 24, and 23 years, respectively.

Examination. On admission, the patient exhibited a right ocular proptosis, periorbital ecchymoses, chemosis, and scleral hemorrhages; corrected visual acuity was 20/20 bilaterally. A bruit could be heard over the right carotid and orbital regions. There were no neurological deficits. Physical examination revealed mild scoliosis, a depressed sternum, and increased laxity of the finger joints. The patient’s skin was atrophic with a prominent venous pattern over the upper thorax, considerable keloid in the abdominal scar, several ecchymoses on the limbs, and numerous superficial varicose veins on both legs.

Over the next few days a right sixth cranial nerve palsy developed. Cranial computerized tomography was noncontributory. Platelet count, bleeding time, prothrombin time, activated partial thromboplastin time, thrombin time, prothrombin consumption, Factor VIII-C, and von Willebrand factor:antigen were all within normal limits.

Transfemoral arteriography showed a direct right carotid-cavernous fistula (Fig. 1). The left common carotid artery was occluded. Marked changes were present in the tortuous extracranial right ICA with ectasias and aneurysm formation near the skull base, most likely due to previous carotid artery dissection (Fig. 1). An aneurysm and multiple wall irregularities, including an intimal flap indicating arterial dissection, were observed in the ecstatic extracranial portion of the left vertebral artery (Fig. 2). The cerebral circulation was mainly from the vertebrobasilar system (Fig. 2). After the angiographic procedure, a large hematoma developed at the puncture site in the groin and was treated with a pressure dressing.

Operations. Balloon embolization of the carotid-cavernous fistula was attempted via a transfemoral approach but was unsuccessful as the tortuosity of the right ICA prevented distal balloon placement. The following day, with an anesthesiologist in attendance and an operating room ready, a second attempt at embolization was made, this time through a percutaneous right carotid artery puncture; this resulted in immediate severe bleeding into the neck followed by circulatory collapse. The procedure was terminated and the patient was taken to the operating room where resuscitation was continued. She was intubated and adequate ventilation was achieved. The common carotid artery was exposed and temporarily clamped, the neck hematoma was evacuated, and the arterial puncture was repaired. The patient recovered well and was free of symptoms.
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2 months later. The bruit had disappeared, but slight diplopia upon right lateral gaze and mild dilatation of scleral veins persisted. The right-sided cervical incision healed with a considerable amount of keloid.

Postoperative Course. Four months later intravenous digital subtraction angiography (DSA) was unsuccessful due to extravasation of contrast material in the patient’s arm. An echocardiogram was normal. The following year intravenous DSA was performed without complications and showed no fistula. The patient has remained well during a 3-year follow-up period.

Discussion

Ehlers-Danlos syndrome Type IV is considered a rare disease but many unrecognized cases probably exist due to the variability of its signs. The external signs may be quite subtle. In addition to the vascular manifestations, rupture of the intestine or gravid uterus (tissues rich in Type III collagen) may also occur. Other features of Ehlers-Danlos Type IV usually limited to the digits, mitral valve prolapse, pheochromocytoma, and varicose veins. A characteristic facies with prominent eyes, pinched nose, thin lips, and lobeless ears may be observed.

Literature Review

A total of 16 patients with probable or definite Ehlers-Danlos syndrome Type IV and spontaneous carotid-cavernous fistulae were identified in the literature. The clinical features of these 16 patients and our patient are summarized in Tables 1 and 2. There were 14 women and three men, with a mean age of 31.6 years (range 17 to 52 years). A total of 20 carotid-cavernous fistulae were observed, with three patients having bilateral fistulae. The involved side was the left in 12 cases, the right in seven cases, and not reported in one case. The type of fistula was established as direct for 17 fistulae; three were not identified.
TABLE 2
Summary of the clinical course and treatment of 17 patients with Ehlers-Danlos syndrome Type IV and carotid-cavernous fistulas

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Complications of Angiography</th>
<th>Treatment</th>
<th>Complications</th>
<th>Result &amp; Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>NA</td>
<td>conservative</td>
<td>NA</td>
<td>blindness</td>
</tr>
<tr>
<td>2</td>
<td>1st angio: hematoma at puncture site</td>
<td>graded occlusion</td>
<td>none</td>
<td>no change</td>
</tr>
<tr>
<td></td>
<td>2nd angio: none</td>
<td>extracranial ICA clamping</td>
<td>vascular fragility, blood loss</td>
<td>initial improvement; recurrence 1 yr later</td>
</tr>
<tr>
<td></td>
<td></td>
<td>CCA ligation</td>
<td>none</td>
<td>no change</td>
</tr>
<tr>
<td></td>
<td></td>
<td>ocular enucleation</td>
<td>blood loss</td>
<td>died 19 days later from cardiac rupture</td>
</tr>
<tr>
<td>3</td>
<td>epistaxis, hematoma at puncture site, blindness</td>
<td>CCA ligation</td>
<td>none</td>
<td>no change in vision, otherwise improved; temporary recurrence; died 6 yrs later from rupture of splenic artery</td>
</tr>
<tr>
<td>4</td>
<td>none</td>
<td>extracranial ICA ligation</td>
<td>none</td>
<td>good</td>
</tr>
<tr>
<td>5</td>
<td>1st angio: none</td>
<td>extracranial ligation of ICA, ECA, &amp; CCA</td>
<td>vascular fragility</td>
<td>initial improvement; developed contralateral CCF</td>
</tr>
<tr>
<td></td>
<td>2nd angio: none</td>
<td>attempt at extracranial ICA ligation</td>
<td>vascular fragility</td>
<td>no change in vision, otherwise improved</td>
</tr>
<tr>
<td>6</td>
<td>epistaxis, died</td>
<td>conservative</td>
<td>NA</td>
<td>spontaneous resolution; recurrence 10 yrs later</td>
</tr>
<tr>
<td>7</td>
<td>1st angio: none</td>
<td>attempt at extracranial ICA ligation</td>
<td>NA</td>
<td>no change</td>
</tr>
<tr>
<td></td>
<td>2nd angio: none</td>
<td>1st transvenous balloon embolization</td>
<td>vascular fragility</td>
<td>died 4 days later</td>
</tr>
<tr>
<td>8</td>
<td>hematoma at puncture site</td>
<td>2nd transvenous balloon embolization</td>
<td>none</td>
<td>no change</td>
</tr>
<tr>
<td></td>
<td></td>
<td>pontine bleeding</td>
<td>no change</td>
<td>spontaneous resolution</td>
</tr>
<tr>
<td>9</td>
<td>none</td>
<td>conservative</td>
<td>NA</td>
<td>spontaneous resolution</td>
</tr>
<tr>
<td>10</td>
<td>ICA ligation</td>
<td>NA</td>
<td>none</td>
<td>died 3 years later from hemoperitoneum</td>
</tr>
<tr>
<td>11</td>
<td>1st angio: cause of fatal arterial lacerations 12 days later</td>
<td>attempt at transarterial balloon embolization</td>
<td>hematoma at puncture site; CCA &amp; ICA dissection</td>
<td>NA</td>
</tr>
<tr>
<td></td>
<td>2nd angio: hematoma at puncture site</td>
<td>?</td>
<td>?</td>
<td>died 3 years later from renal artery rupture</td>
</tr>
<tr>
<td>12</td>
<td>?</td>
<td>?</td>
<td>?</td>
<td>died 3 years later from renal artery rupture</td>
</tr>
<tr>
<td>13</td>
<td>1st angio: none</td>
<td>transarterial balloon embolization</td>
<td>none</td>
<td>good</td>
</tr>
<tr>
<td></td>
<td>2nd angio: none</td>
<td>?</td>
<td>?</td>
<td>died 2 weeks later from ruptured ICA aneurysm</td>
</tr>
<tr>
<td>14</td>
<td>?</td>
<td>?</td>
<td>?</td>
<td>died 2 weeks later from ruptured ICA aneurysm</td>
</tr>
<tr>
<td>15</td>
<td>1st angio: arterial dissection</td>
<td>direct surgical repair</td>
<td>none</td>
<td>visual loss, ophthalmoplegia</td>
</tr>
<tr>
<td></td>
<td>2nd angio: arterial dissection</td>
<td>1st transarterial balloon embolization</td>
<td>none</td>
<td>initial improvement; recurrence</td>
</tr>
<tr>
<td></td>
<td>3rd angio: none</td>
<td>2nd transarterial balloon embolization</td>
<td>cavernous sinus &amp; ICA expansion</td>
<td>no change</td>
</tr>
<tr>
<td></td>
<td></td>
<td>transvenous injection of liquid adhesive</td>
<td>none</td>
<td>good</td>
</tr>
<tr>
<td>16</td>
<td>1st angio: arterial dissection</td>
<td>attempted transvenous balloon embolization</td>
<td>none</td>
<td>balloon placement failed</td>
</tr>
<tr>
<td></td>
<td>2nd angio: none</td>
<td>transarterial placement of coils &amp; suture material</td>
<td>none</td>
<td>good; died 7 mos later from ruptured viscus</td>
</tr>
<tr>
<td>17</td>
<td>1st angio: hematoma at puncture site</td>
<td>intracranial ICA &amp; CCA ligation &amp; muscle embolization</td>
<td>none</td>
<td>good</td>
</tr>
<tr>
<td></td>
<td>2nd angio: hematoma at puncture site</td>
<td>1st attempt at transarterial balloon embolization</td>
<td>none</td>
<td>balloon placement failed</td>
</tr>
<tr>
<td></td>
<td></td>
<td>2nd attempt at transarterial balloon embolization</td>
<td>hematoma at puncture site</td>
<td>resolution after temporary carotid artery clamping</td>
</tr>
</tbody>
</table>

*Angio = angiogram; NA = not applicable; ICA = internal carotid artery; CCA = common carotid artery; ECA = external carotid artery; CCF = carotid-cavernous fistula; ? = no data available. For origin of cases see Table 1.
Three additional cases of carotid-cavernous fistulae associated with probable or definite Ehlers-Danlos syndrome Type IV reported in the literature were not included because of the lack of basic clinical information (for example, age and sex).6,27

Angiography. Conventional diagnostic neuroangiographic examinations were performed 25 times in 14 patients. No angiogram was obtained in one patient, while insufficient details were given in two other cases. Nine patients suffered one or more complications resulting in a morbidity rate of 36% for 25 angiographic procedures. Six angiographic examinations were complicated by severe bleeding at the site of arterial puncture causing tracheal compression in the four instances in which the carotid artery was punctured. On three occasions, transfemoral angiography resulted in iliac arterial dissections. Massive epistaxis occurred in two cases. In one patient, the signs and symptoms of the fistula became much worse following angiography, resulting in permanent blindness.

Three of the 25 angiographic examinations resulted in death, for a mortality rate of 12%. One patient died during transfemoral angiography as a result of a tear in the ascending aorta. Another death occurred 16 days after transfemoral arteriography because of multiple arterial lacerations and dissections. The third patient died during massive epistaxis following direct carotid angiography; an autopsy was not performed.

Conservative Treatment. Three patients were treated conservatively. One patient experienced spontaneous resolution with a good final outcome. In another patient, the fistula resolved but recurred 10 years later, resulting in permanently impaired vision. Another patient lost her vision because of retinal thrombosis and detachment.

Surgical Treatment. Eight patients underwent 10 surgical procedures consisting of (attempted) ligation or clamping of one or more cervical carotid vessels in seven procedures, one Hamby procedure, one direct surgical repair, and ocular enucleation in one patient. Unusually severe blood loss occurred during two surgical procedures. Vascular fragility was noted during three operations, preventing completion of the procedure in one patient. The patient undergoing ocular enucleation died from cardiac rupture 19 days after the operation. Three patients improved following cervical carotid artery ligation, with one instance of recurrence. The patient who had undergone a direct surgical repair of the fistula suffered visual loss and ophthalmoplegia. In our patient, the Hamby procedure was successful in treating the initial fistula.

Balloon Embolization. Balloon embolization of the fistula was either attempted or carried out in six patients. One patient who underwent transvenous balloon embolization suffered a fatal pontine hemorrhage 4 days after the procedure, presumably due to diversion of arterialized blood into cortical veins. In another patient, embolization via the transarterial route was attempted through direct puncture of the carotid artery, but this produced a dissection of the common and internal carotid artery and a massive hematoma at the puncture site necessitating intubation. This patient died several days later from arterial lacerations caused by previous diagnostic transfemoral arteriography (see above). In two patients the fistulae could be cured only after multiple attempts at transarterial and transvenous closure of the fistula. Expansion of the cavernous sinus and carotid artery following balloon placement prevented complete closure of the fistula in one patient. In our patient, two attempts at transarterial balloon embolization were unsuccessful. Embolization was successful in only one previously reported patient; in that case the transarterial route was used, and the patient experienced a good outcome after a single attempt at closing the fistula.

In two reported patients no details were given about the management of their carotid-cavernous fistulae.

Mortality. Ten of the 17 patients died, for a mortality rate of 59%. Four deaths were a direct result of diagnostic or therapeutic procedures. Five additional patients died from vascular catastrophes 2 weeks to 6 years after the fistula occurred. One patient died from a perforated viscus 7 months after treatment of a carotid-cavernous fistula.

Pathogenesis

Spontaneous carotid-cavernous fistulae appear to be a relatively frequent complication of Ehlers-Danlos syndrome Type IV. The mean age of patients who develop spontaneous carotid-cavernous fistulae is considerably lower for patients with the syndrome (31.6 years) than that reported for patients apparently without the syndrome (58.3 years).5,12,24,46,51 The male:female ratio is similar in those with and without Ehlers-Danlos syndrome Type IV (1:4.7 vs. 1:4).5,12,24,46,51

Spontaneous direct carotid-cavernous fistulae account for approximately 10% of all spontaneous carotid-cavernous fistulae in the general population.5,11,24,51 In patients with Ehlers-Danlos syndrome Type IV, however, only direct fistulae have been observed. Spontaneous direct carotid-cavernous fistulae may be caused by the rupture of a pre-existent intracavernous aneurysm or by dissection or rupture of the carotid artery as it traverses the cavernous sinus. In four patients with Ehlers-Danlos syndrome Type IV presenting with carotid-cavernous fistulae, such ruptured intracavernous aneurysms were detected angiographically.6,16,20,43 A 24-year-old woman with bilateral unruptured intracavernous aneurysms recently underwent genetic evaluation at our institution and was diagnosed as having Ehlers-Danlos syndrome Type IV. An autopsy in another patient with this condition showed a small transmural tear in the intracavernous portion of the ICA causing a fistula and a small intact lateral aneurysm of the contralateral intracavernous ICA.28 In
yet another patient, autopsy also revealed a small perforation in the intracavernous ICA.97 Ehlers-Danlos syndrome Type IV may predispose to the carotid artery abnormalities that can result in spontaneous direct fistulae due to structural weaknesses of the vessel wall caused by Type III collagen defects. Type III collagen abnormalities should be suspected in patients with spontaneous carotid-cavernous fistulae, especially in young and middle-aged patients with a direct fistula.

Early clinical recognition of Ehlers-Danlos syndrome Type IV is important in view of the hazards of arterial puncture and surgery associated with this condition.

Angiography

Complications of diagnostic angiography in patients with Ehlers-Danlos syndrome Type IV may occur at the puncture site or anywhere along the route of the catheter. In general, angiography should be avoided in patients with this syndrome, if at all possible. However, angiography is essential for differentiating direct from dural fistulae, assessing the hemodynamic characteristics, and visualizing the precise vascular architecture so that the optimal management strategy may be selected. Careful insertion and manipulation of the smallest possible soft-tipped angiographic catheter may decrease some of the risk. If a false passage is suspected, or if other difficulties occur with manipulation of the catheter, the procedure should be terminated. Magnetic resonance-assisted angiography may be helpful in the evaluation of these patients but will not likely define the pathology to the degree necessary for therapeutic planning.

Treatment

The optimal therapy for spontaneous carotid-cavernous fistulae remains a matter of some debate. A spontaneous carotid-cavernous fistula is rarely a life-threatening condition, although there is a definite threat to the patient's vision. Although none of the patients we reviewed died as a direct result of the fistula, the lethal potential of carotid-cavernous fistulae may be higher in patients with Ehlers-Danlos syndrome Type IV.

While about 50% (range 9% to 73%) of dural carotid-cavernous fistulae resolve spontaneously,5,12,24,34,37,46 direct fistulae undergo spontaneous resolution much less frequently. Balloon occlusion of the fistula is usually the treatment of choice. If balloon occlusion proves to be impossible or unsuccessful, its use in combination with sacrifice of the ICA or surgical trapping may be necessary. Direct surgical approach and repair of the fistula in the cavernous sinus is now another alternative.13 These same therapeutic alternatives apply to patients with Ehlers-Danlos syndrome Type IV, although the decision to treat a carotid-cavernous fistula in such a patient should be made only after cautious consideration of each alternative including conservative treatment.

If percutaneous vascular balloon occlusion is planned, it should be carried out with complete anesthetic and surgical services ready for emergency interventions should severe bleeding occur as happened with our patient. In the patient who died 4 days after transvenous balloon embolization, the transvenous route had been selected because a transarterial approach was considered to be too dangerous for a patient with Ehlers-Danlos syndrome Type IV.15,23 The increased vascular distensibility associated with Type III collagen defects7,35 may have accounted for the expansion of the cavernous sinus and carotid artery observed in one patient following endovascular balloon placement.22 An additional problem is posed by the ectasias and tortuosities of extracranial carotid arteries which are a common manifestation of this syndrome,6,10,20,22,25,26,45,47 and may impede successful placement of the balloon.

Vascular surgery in general is hazardous in patients with Ehlers-Danlos syndrome Type IV.8,33,48,53 Both arteries and veins are abnormally friable and may tear easily. The slightest force may cause arteries to separate from their parent vessel. Clamps and ligatures often do not hold well and may cause extensive arterial lacerations. Sutures may easily tear through the vessel walls. Hemostasis is difficult and large postoperative hematomas are common. The use of interrupted buttressed sutures for arterial reconstruction, heavy nonabsorbable sutures or umbilical tape for vessel ligation, and the strict avoidance of hypertension may lessen some of the surgical complications.

Coagulopathy and the Potential Applicability of Desmopressin

An important and frequent presenting symptom of patients with Ehlers-Danlos syndrome Type IV is easy bruising. Often, a tendency toward continued oozing is observed during surgery. Standard coagulation tests are usually normal, as they were in our patient. The etiology of this bleeding tendency has not been firmly established, but is probably a defective interaction of platelets with the vascular subendothelium. Type III collagen is the major component of the subendothelium16 and strongly promotes platelet aggregation in vitro.1 The synthetic vasopressin analog 1-desamino [8-D-arginine] vasopressin (DDAVP, or desmopressin) stimulates the release of von Willebrand factor,26,30 which in turn promotes adhesion of platelets to the subendothelial connective tissue.43 Desmopressin might therefore be of benefit in the management of patients with Ehlers-Danlos syndrome Type IV.44,52 Due to its hemostatic effects, desmopressin may prevent oozing from small vessels. In addition, it has been postulated to have a direct effect on the vessel wall.4 If defective platelet interaction with the Type III collagen-deficient vascular subendothelium indeed plays a role in the pathogenesis of vascular dissection or rupture, desmopressin may reduce the risk of these vascular complications.44 One patient with Ehlers-Danlos syndrome Type IV underwent a successful Caesarean section after receiving desmopressin to reduce perioperative blood loss.52 It should be noted, however, that desmopressin has a potential
thrombotic effect; the degree of this has not been clearly established\textsuperscript{14} but appears to be minimal.\textsuperscript{26,29,30} Intravenous desmopressin administration may be considered in patients with a definite diagnosis of Ehlers-Danlos syndrome Type IV who must undergo angiography or surgery, especially vascular surgery.

**Genetic Counseling**

Most patients with Ehlers-Danlos syndrome Type IV have a noncontributory family history. These "sporadic" cases most likely represent new autosomal mutations with dominant effect, although in some families autosomal-recessive inheritance can be demonstrated. All children and siblings, and preferably the parents, of patients with Ehlers-Danlos syndrome Type IV should therefore be examined for this condition. In family planning discussions, the patient with Ehlers-Danlos syndrome Type IV not only should be told about the genetic prognosis, but should also be advised to consider the personal prognosis that might prevent the patient from adequately supporting a family. Women with Ehlers-Danlos syndrome Type IV should also be advised about the possible obstetrical complications of this condition.

**Addendum**

Subsequent to the submission of this manuscript for publication, Halbach, et al.,\textsuperscript{22} reported on treatment of four patients with Ehlers-Danlos syndrome and carotid-cavernous fistulae. Their Case 1 is the patient summarized in this case report; the percutaneous transluminal therapeutic attempts were carried out at the Mayo Clinic with the assistance of Dr. Grant Hieshima.

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**References**


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