Outcome following open and endovascular intervention for carotid stump syndrome

Makinderjit Dulai1, Mohamed Elsherif1, Wael Tawfick1, Edel P Kavanagh1,2, Niamh Hynes1,2 and Sherif Sultan1,2

Abstract
Carotid stump syndrome is defined as the persistence of retinal or cerebral ischaemic events with complete occlusion of the ipsilateral internal carotid artery. The aim of this retrospective cases series was to assess the outcomes for patients with carotid stump syndrome managed with surgical intervention. A series of 11 cases of carotid stump syndrome in nine patients presented to our tertiary vascular centre from October 2004 to February 2016. Indications for intervention were amaurosis fugax, transient ischaemic attacks and stroke. In total, 11 procedures were performed on nine patients including carotid angioplasty and stenting or carotid endarterectomy with patching. The mean follow-up period was 56.6 months. One patient suffered a myocardial infarction 30 days, post-operatively, and one patient was lost to follow-up. In the remaining seven patients, there was a complete resolution of symptoms. There were no incidents of death, stroke, cranial nerve injury, wound haematoma or procedural bleeding. Surgical exclusion of carotid stumps combined with dual antiplatelet agents was found to be a safe and effective treatment method for carotid stump syndrome.

Keywords
Carotid stump syndrome, internal carotid artery, angioplasty, stents, endarterectomy

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Introduction
Carotid stump syndrome (CSS) is defined as the persistence of retinal or cerebral ischaemic events with complete occlusion of the ipsilateral internal carotid artery (ICA). Thromboemboli from a patent region of the ICA (carotid stump) passes through the ipsilateral external carotid artery (ECA) into the middle cerebral artery circulation through anastomotic channels.1 The incidence of CSS is unknown due to the relative rarity of the condition.2 Prompt diagnosis and management following the onset of ischaemic symptoms is crucial, as the overall risk of an ischaemic stroke from ICA occlusion is 30%.3 The gold standard treatment for CSS is a combination of medical therapy with dual antiplatelet agents and surgical intervention.2

Patients and methods
A series of 11 cases of CSS in nine patients that presented to our tertiary vascular centre from October 2004 to February 2016 were identified using our prospectively collated database (Vascubase, Consensus Medical Systems Inc., Richmond, BC, Canada). In total, 11 procedures were performed on nine patients, with two patients receiving bilateral interventions. Clinical findings were correlated with radiographic imaging (duplex ultrasound (DUS), computed tomography angiography (CTA), and magnetic resonance angiography (MRA)) of the carotid arteries to confirm complete occlusion of the ipsilateral ICA.

Cases 1 and 2
A 61-year-old male patient presented with a 5-year history of dizziness, intermittent headaches and recurrent transient ischaemic attacks (TIAs) with bilateral amaurosis fugax. He
had a history of hypertension, hypercholesterolaemia, ischaemic heart disease and was an ex-smoker with a 20 pack-year history. DUS showed bilateral ICA occlusion (Figure 1), 60%–70% left common carotid artery (CCA) stenosis, 99% left ECA stenosis, 40% right CCA stenosis and 80% right ECA stenosis. The vertebral arteries were patent bilaterally with antegrade flow. Clinical presentation and imaging suggested cerebral hypoperfusion syndrome and the patient was initially followed with best medical treatment for 4 years.

Due to disease progression and worsening of symptoms, a left CCA angioplasty and stenting (SMART vascular stent system, Cordis, USA) with a retrograde hybrid technique, and a left CCA to ECA endarterectomy and patching with an intervascular HemaCarotid patch (Datascope, Montvale, NJ, USA) were performed. He was managed with best medical therapy post-operatively. At a 6-week follow-up appointment, he still complained of dizziness. After 8 months, right proximal stenting of the CCA with a retrograde hybrid technique, and right CCA to ECA endarterectomy and patching with an intervascular HemaCarotid patch (Datascope, Montvale, NJ, USA) were performed. At 6-week and 1-year follow-ups, he still complained of dizziness; however, he did not have any new neurological deficits, including TIAs or amaurosis fugax. This remained at his 10-year follow-up.

Cases 3 and 4
A 72-year-old male patient presented with a 7-week history of bilateral amaurosis fugax. Episodes occurred approximately once per day, lasting between 10 s and 5 min and were more frequent in the left eye. He had a history of ischaemic heart disease, hypertension, hyperlipidaemia and paroxysmal atrial fibrillation. He was an ex-smoker with a 20 pack-year history. DUS, flow velocities suggested 1%–49% right ECA stenosis and 80%–99% left ECA stenosis. Both vertebral arteries were patent with antegrade flow. CTA of the carotid arteries showed significant calcification at the right and left carotid bulb with complete occlusion of the right ICA and left ICA at the level of the bifurcation (Figure 2).

A bilateral ECA angioplasty and stenting (XACT carotid stent system, Abbott, IL, USA) was performed (Figure 3). Post-operatively, 0–48 h, there was complete resolution of the amaurosis fugax. Post-operatively, 4-weeks, the patient presented with a non-ST segment elevated myocardial infarction (MI) with new onset atrial fibrillation. He subsequently recovered completely from the MI and had no other symptoms.

Case 5
A 65-year-old male patient presented with TIAs with amaurosis fugax. Episodes of amaurosis fugax occurred approximately once per day and resolved in less than 1 h. The patient had one episode of slurred speech prior to admission and a stroke 1-year prior. He had a history of hypertension, hypercholesterolaemia, chronic obstructive pulmonary disease (COPD), ischaemic heart disease and was a smoker with a 50 pack-year history.

DUS showed 60%–70% stenosis in the right ICA and a mixed plaque in an occluded left ICA with a patent proximal stump. There was mild proximal stenosis in the left and right ECA and patent vertebral arteries bilaterally with antegrade flow. A right carotid endarterectomy (CEA) was performed, but his symptoms persisted at a 6-week follow-up appointment. After 8 weeks, a left ECA endarterectomy with a CCA to ECA ultrathin HemaCarotid patch (Maquet Gentige Group, Rastatt, Germany) was performed with ligation of the left ICA. Post-operatively, the patient was still complaining...
of intermittent blurring of vision attributed to stenosis in the proximal part of the CCA. After 1 year, a left CCA angioplasty with stenting (XACT carotid stent system, Abbott, IL, USA) was performed (Figure 4). The patient was placed on best medical therapy and had complete resolution of ocular symptoms on follow-up appointments.

Case 6
A 79-year-old male patient presented with a 1-month history of recurrent episodes of postural hypotension. On assessment of blood pressure (BP), lying BP was 140/70 mm Hg and standing BP was 88/56 mm Hg. He had a history of hyperlipidaemia, severe peripheral vascular disease, prostate cancer and oesophageal cancer for which he received radiotherapy to the neck.

DUS showed a 40%–50% mid-distal right CCA stenosis, an occluded right ICA with a small patent stump at the origin (Figure 5) and moderate right ECA atheroma; 40%–50% distal left CCA stenosis, 50% left ICA stenosis and moderate left ECA atheroma. Angioplasty and stenting (XACT carotid stent system) of the right ECA was performed to completely exclude the stump and a thrombus in the right ICA was removed. At a 4-week follow-up appointment, there was complete resolution of symptoms, but DUS showed an occluded right CCA and ECA stent. There was 40%–50% left CCA stenosis, 50%–60% left ICA stenosis and moderate left ECA atheroma.

Case 7
A 68-year-old female patient presented with a 1-year history of bilateral headaches over the frontal and temporal areas with nuchal numbness. Headaches were 9/10 in severity,
waking her up at night and worsening on the Valsalva manoeuvre. They were increasing in frequency and did not improve with traditional analgesia. She also suffered from vertigo once per year with associated hearing loss on the right side and tinnitus bilaterally. She had a background history of hypertension, hypercholesterolaemia and is an ex-smoker with a five pack-year history.

DUS showed an occluded right ICA with a short patent stump at the origin (Figure 6), 0.5 cm in length, and 40%–50% stenosis in the left ICA. Vertebral arteries were patent bilaterally with antegrade flow. MRA confirmed complete right ICA occlusion. A right carotid angioplasty and stenting (XACT carotid stent system) to close the stump with aspiration of thrombus from the stump was performed to prevent any thromboembolic events through the ECA via the supraorbital and supraorbital artery. The patient was placed on best medical therapy post-operatively and at a 6-week follow-up appointment, her symptoms resolved with no new neurological deficits. At 2-year follow-up, the patient was completely asymptomatic. DUS showed a patent proximal right CCA and an occlusion in the mid-distal vessel. The stent was occluded throughout (Figure 7).

Case 9
A 70-year-old male patient presented with sudden onset of headache, difficulty speaking, confusion, unsteady gait and weakness in the right arm. A computed tomography (CT) brain showed a left orbitoparietal infarct. He had a significant history of hypertension, hyperlipidaemia, type II diabetes mellitus (DM), peripheral vascular disease, COPD, prostate cancer and was an ex-smoker with an 80 pack-year history. He suffered from recurrent TIAs for 3 years prior to admission. DUS showed an occluded left ICA and an 80%–99% stenosis of the left ECA with a haemorrhagic plaque. The right ICA was occluded with 1%–49% stenosis of the right ECA. The vertebral arteries were patent bilaterally with antegrade flow. MRA confirmed bilateral ICA occlusion and 99% left ECA stenosis (Figure 8).

A left CCA to ECA endarterectomy with patching (ultrashin HemaCarotid patch, Maquet Getinge, Rassatt, Germany) was performed. He was asymptomatic post-operatively. DUS at
3-year follow-up showed mild atheroma in the left CCA–ECA patch and mild increased flow in the distal left ECA.

**Case 10**

A 65-year-old male patient presented with a 1-month history of amaurosis fugax in the right eye. He had a history of hypertension, hyperlipidaemia and ischaemic heart disease and was an ex-smoker with a 60 pack-year history. DUS showed an occluded right ICA with mild proximal atheroma in the right ECA and 20%–30% left ICA stenosis with a mild proximal atheroma in the left ECA. A right CCA to ECA endarterectomy with patch closure was performed (ultrathin HemaCarotid patch, Maquet Getinge, Rassatt, Germany). This patient was lost to follow-up.

**Case 11**

An 81-year-old male patient presented with three episodes of crescendo amaurosis fugax of the left eye over 72 h. DUS showed complete occlusion of the left ICA with a stump in the proximal portion of the ICA and a patent ECA. The right ICA was 80%–99% stenosed and there was no significant stenosis in both CCA’s. MRA demonstrated patent left supraorbital and supratrochlear branches. He had a normal echocardiogram and computed axial tomography (CAT) scan.

A left CCA to ECA endarterectomy was performed with obliteration of the carotid stump from within using prolene sutures (Ethicon Inc., New Jersey, USA). The patient was discharged home 3 days post-operatively. At 13 months follow-up, DUS showed normal flow through the left CCA–ECA patch and contralateral 80%–99% stenosis. The patient was asymptomatic and on best medical therapy.

**Results**

Clinical details for patients are shown in Table 1. In total, 11 cases (n = 11) of ICA occlusion in nine patients (eight males, one female) were managed by either carotid angioplasty and stenting (45.5%; n = 5) or carotid endarterectomy with patching (54.5%; n = 6). Indications for intervention were amaurosis fugax (54.5%; n = 6), TIAs (18.2%; n = 2) and stroke (9.1%; n = 1).

Patient surgical outcomes are shown in Table 2. One patient (11.1%) suffered a MI 30 days, post-operatively. There were no incidents of death, stroke, cranial nerve injury, wound haematoma or procedural bleeding. Mean hospital stay was 2.8 days. Mean follow-up was 56.6 months (range 1–144 months). Eight patients (88.9%) had complete resolution of neurological symptoms and one patient (11.1%) was lost to follow-up. There were three incidences (60%) of complete occlusion of the stent during follow-up.

**Discussion**

CSS is a rare condition that was first described in 1978.1 It is defined as the persistence of retinal or cerebral ischaemic events due to microembolisation from a patent proximal remnant of a completely occluded ipsilateral ICA.1 A diagnosis of CSS is made by correlating clinical symptoms with radiographic imaging. A DUS is diagnostic, but a CT scan or MRA may be performed for confirmation.3 Timely diagnosis and management is essential as one-third of patients with ICA occlusion experience disabling symptoms or die.3 Due to its relative rarity, the incidence rate of CSS is undocumented in current literature.2 Out of 9585 patients with carotid disease referred to our tertiary vascular centre over 12 years, 691 carotid interventions were performed. In total, 11 of these cases (1.59%) had CSS.
The standard treatment of CSS is exclusion of the stump by surgical intervention followed by dual antiplatelet therapy. Open intervention includes either oversewing the origin of the ICA or by obliterating it with a large metallic clip with an endarterectomy of the ECA. Endovascular exclusion with placement of a covered stent is an alternative to open surgery. No current evidence suggests that one method is more effective than the other.

The literature on CSS is limited. In a study by Kumar et al., 22 patients underwent stump exclusion by oversewing the ICA origin. All but one patient remained asymptomatic at follow-up. Barnett et al. reported on seven patients treated by open intervention. Six cases had complete resolution of neurological symptoms and one patient passed away from an MI 2 weeks post-operatively. At our tertiary vascular centre, six carotid endarterectomies with patching were performed with oversewing the ICA occlusion. All patients had complete resolution of symptoms.

Our literature review yielded two case reports documenting endovascular treatment of CSS. In one case, the patient suffered a TIA 24 h post-operatively with complete resolution of symptoms. He was asymptomatic at a 12-month follow-up appointment. The second patient suffered a brief episode of expressive dysphagia, with no hemimotor or sensory symptoms, 7 days post-operatively. There was complete resolution of symptoms within 30 min and he was

### Table 1. Clinical details and imaging results.

| Case | Age (years) | Sex | Symptoms | DUS |
|------|-------------|-----|----------|-----|
| 1    | 61          | M   | Headaches, TIA, amaurosis fugax | L and R ICA occlusion, L ECA 99% stenosis, R ECA 80% stenosis |
| 2    | 61          | M   | TIA, amaurosis fugax | L and R ICA occlusion, L ECA 99% stenosis, R ECA 80% stenosis |
| 3    | 72          | M   | Amaurosis fugax | L and R ICA occlusion, R ECA 1%–49% stenosis, L ECA 80%–99% stenosis |
| 4    | 72          | M   | Amaurosis fugax | L and R ICA occlusion, R ECA 1%–49% stenosis, L ECA 80%–99% stenosis |
| 5    | 65          | M   | Headaches, dizziness, amaurosis fugax | L ICA occlusion, R ICA 60%–70% stenosis |
| 6    | 79          | M   | Light-headedness, collapse | R ICA occlusion, 40%–50% R CCA stenosis, L CCA 40%–50% L ICA 50% stenosis |
| 7    | 68          | F   | Headaches | R ICA occlusion, L ICA 40%–50% stenosis |
| 8    | 68          | M   | Headaches, dizziness, amaurosis fugax | R ICA occlusion, L ICA 40%–50% stenosis |
| 9    | 70          | M   | Asymptomatic from CVA 6 months pre-operatively | L ICA occlusion, L ECA 80%–99% stenosis, R ICA occlusion, R ECA 1%–49% stenosis |
| 10   | 65          | M   | Amaurosis fugax | R ICA occlusion, L ICA 20%–30% stenosis |
| 11   | 81          | M   | Amaurosis fugax | L ICA occlusion, R ICA 80%–99% stenosis |

DUS: duplex ultrasound; TIA: transient ischemic attack; ICA: internal carotid artery; ECA: external carotid artery; CCA: common carotid artery; CVA: cerebrovascular accident.

### Table 2. Outcome and follow-up after surgical intervention.

| Case | Interval from latest episode to surgery | Duration of Symptoms | Surgery | Outcome | Follow-Up |
|------|----------------------------------------|----------------------|---------|---------|-----------|
| 1    | 0                                      | 5 years              | L CCA-ECA CEA | Complete resolution | 10 years |
| 2    | 8 months                               | 5 years              | R CAST, R CCA-ECA | Complete resolution | 10 years |
| 3    | 0                                      | 7 weeks              | R ECA CAST | MI 4 weeks post-op | 4 weeks |
| 4    | 0                                      | 7 weeks              | L ECA CAST | MI 4 weeks post-op | 4 weeks |
| 5    | 0                                      | 5 months             | L CCA-ECA CEA | Complete resolution | 12 years |
| 6    | 0                                      | 1 month              | R ECA CAST | Complete resolution | 1 year |
| 7    | 0                                      | 1 year               | R CCA CAST | Complete resolution | 2 years |
| 8    | 0                                      | 9 months             | R CCA CAST | Complete resolution | 9 years |
| 9    | Asymptomatic                           | 6 months             | L CCA-ECA CEA | Complete resolution | 3 years |
| 10   | 0                                      | 1 month              | R CCA-ECA CEA | Complete resolution | Lost to follow-up |
| 11   | 0                                      | 72 h                 | L CCA-ECA | Complete resolution | 13 months |

CCA: common carotid artery; ECA: external carotid artery; CEA: carotid endarterectomy; MI: myocardial infarction.
asymptomatic at 1 and 3-month follow-up appointments. Both case reports do not report on the patency of the stent, post-operatively. At our tertiary vascular centre, five angioplasty and stenting procedures were performed. There was complete resolution of neurological symptoms in all cases. Eventually, three of the five stents completely occluded, but the patients remained asymptomatic at follow-up.

It was originally believed that a completely occluded ICA did not produce any symptoms. However, several mechanisms have been proposed to explain the recurrent embolic symptoms in those with CSS. For one, they may be haemodynamic attacks caused by alterations in blood flow through anastomotic channels. This includes changes in cardiac output or a postural decrease in BP. Possible support for this hypothesis was seen in case six who presented with recurrent episodes of light-headedness, collapse and postural hypotension on physical examination. Another theory is that ischaemic events may result from embolisation of material from the soft distal end of the thrombus located in the totally occluded ICA. This should be considered when both the ipsilateral ICA and ECA are occluded and when ischaemic symptoms persist after surgical exclusion of the proximal stump. There were no such cases in this series.

Carotid artery pseudo-occlusion is defined as segmental occlusion at the origin of the ICA with very thin distal flow. It may be falsely diagnosed as total ICA occlusion by means of DUS, CTA or MRA. Therefore, cases of segmental ICA occlusion may be misdiagnosed as CSS. The neurological symptoms in these patients may originate from an atherosclerotic lesion or acute partial thrombosis with a tail that continues to embolise. There is no established criteria for the optimal management for ICA pseudo-occlusion, partly because its natural history is unknown. Asymptomatic patients may be managed on best medical treatment alone. However, operative intervention in addition to best medical management is preferred in most centres. For patients with TIAs or strokes, several studies demonstrated that surgical intervention by means of ligation of the ICA, endarterectomy with patch closure, eversion endarterectomy, carotid bypass and carotid angioplasty and stent (CAS) to be safe and effective.

The carotid stump may not be the sole potential source for emboli when there is significant stenosis in other arteries. These include the ipsilateral CCA or ECA, the carotid bifurcation via a patent ECA and contralateral carotid vessels via the circle of Willis. If there is a significant contralateral disease, an endarterectomy to the side is recommended. However, symptoms usually do not resolve because the symptomatic hemisphere does not receive significant blood supply from the contralateral carotid artery. Nine cases in this series had significant contralateral stenosis and so radiographic imaging is essential to ascertain the potential sources of emboli. The most widely accepted hypothesis is that emboli from the patent portion of the occluded ICA pass through the ipsilateral ECA into the middle cerebral artery circulation through anastomotic channels.

Conclusion

Our case series of 11 patients illustrates the safety and efficacy of both open and endovascular intervention for CSS. Timely diagnosis and management resulted in complete resolution of neurological symptoms in all cases.

Declaration of conflicting interests

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Ethical approval

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Informed consent

Written informed consent was obtained from the patient(s) for their anonymised information to be published in this article.

ORCID iD

Edel P Kavanagh https://orcid.org/0000-0001-6003-944X

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