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Which carotid artery aneurysms need to be treated (and how)?

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ABSTRACT

Extra-cranial carotid artery aneurysms (ECAA) are uncommon and represent a therapeutic challenge for clinicians. An ECAA is generally defined as a dilation of the internal carotid artery (ICA) or common carotid artery (CCA) greater than 150% of the diameter of the normal healthy artery. The presence of an ECAA is usually found by coincidence in asymptomatic patients. Symptomatic patient may present with neurological dysfunction or symptoms of local compression. The initial diagnosis of ECAA is often by echo/duplex ultrasound imaging. However, computerized tomographic angiography (CTA), with 3-dimensional reconstruction of the images (3D) can provide additional and valuable information, especially when considering surgical exclusion of the aneurysm. Recently, vessel wall imaging using contrast enhanced magnetic resonance with gadolinium administration was explored, which could potentially provide valuable information regarding aneurysm wall changes during clinical follow up. Location and accessibility of an ECAA is key information when considering the appropriate treatment. With the lack of evidence based treatment guidelines, a conservative approach with or without medicinal treatment is currently the standard of care for asymptomatic non-growing ECAA. Open surgical repair has for long been the accepted treatment for patients with a growing aneurysms or aneurysm related symptoms. Endovascular interventions are increasingly applied, especially when surgical intervention is considered too risky or not possible due to patient comorbidities or anatomical restrictions. Data on the natural course, immediate and long-term results of surgical or endovascular therapy is scarce. Thus, there is a clear need for an international collaboration collecting data of ECAA within a registry.

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Extra-cranial carotid artery aneurysm (ECAA) is a rare vascular diagnosis that accounts for less than 1% of peripheral aneurysms.[1] ECAA's are more common in men with a male to female ratio of 2:1 and the mean age at presentation around 50 years (range 35-68 years).[2] The most common definition of an ECAA is a dilation of the carotid artery greater than 150% of the diameter of normal (uninvolved) internal carotid artery (ICA) or common carotid artery (CCA). Bilateral ECAA have been reported in 13% of the cases and 15-20% of patients with an ECAA have multiple aneurysms (predominantly intracranial).[2] Current knowledge on ECAA is limited and is based on small case series and case reports lacking long term follow up.[2] Presentation of an ECAA depends on the etiology, location and size. The natural course of an ECAA remains uncertain, but is believed that they rarely remain asymptomatic if they keep enlarging.[3] On the other hand, it is believed that non-growing ECAA will remain asymptomatic. Symptoms may include a palpable pulsating mass, peripheral nerve dysfunction, stridor, voice changes due to local compression, transient ischemic attacks (TIAs) or ischemic stroke. Morphologically there are two distinct types of carotid aneurysms which are most often located

in the ICA. The dilatation may be focal and saccular or fusiform and extensive. ECAA can be true or false aneurysms with the true aneurysms being caused by atherosclerosis, infection (HIV, tuberculosis syphilis and salmonella), arteritis, Marfan's syndrome, fibromuscular and medical degeneration. A false aneurysm or pseudo-aneurysm can be caused by trauma, iatrogenic, post carotid endarterectomy and post dissection. A recent histological analysis revealed two distinct types of ECAA, degenerative and dissection aneurysms with several inflammatory cells being found in some of the degenerative aneurysms.[4] A recent review described 281 published articles describing the management of ECAA.[2] Although several treatments have been developed over the last years, for any etiology, the preferred treatment is yet unknown due to the rarity of the lesion and the lack of evidence-based guidelines.

Diagnosis

The purpose of imaging is to identify and confirm the presence of an ECAA, classify the ECAA, and to assess the anatomy in order to plan a possible surgical intervention. Further, imaging can be applied to evaluate possible aneurysm growth during follow up. Aneurysms are mostly diagnosed with echo/duplex ultrasound imaging but additional computed tomography angiography (CTA) with 3-dimentional (3-D) reconstruction of the carotid arteries (Figure 1) seem to contribute to the diagnosis and therapeutic work up.[5] CTA provides additional information on location and especially accessibility, thus allowing the surgeon/interventionist to decide which intervention is appropriate when an intervention is indicated.

Aneurysm volumetry using 3D images have been suggested to be of additional value next to diameter measurement for detecting changes in abdominal aneurysms.[6] This volumetry method to detect aneurysm growth could be used in the future for detecting ECAA changes and possibly predict the clinical course of an ECAA. More sophisticated imaging using 3 Tesla high-resolution contrast enhanced magnetic resonance imaging (3TMRI) with gadolinium administration is currently being investigated for vessel wall imaging. It is hypothesized that administering gadolinium during MRI results in enhancements of sites with inflammation [7] which could also be a marker for aneurysm growth. Also, in patients undergoing surgical resection the histological correlation of aneurysm wall enhancement with gadolinium and the degree of aneurysm wall inflammation could be investigated.

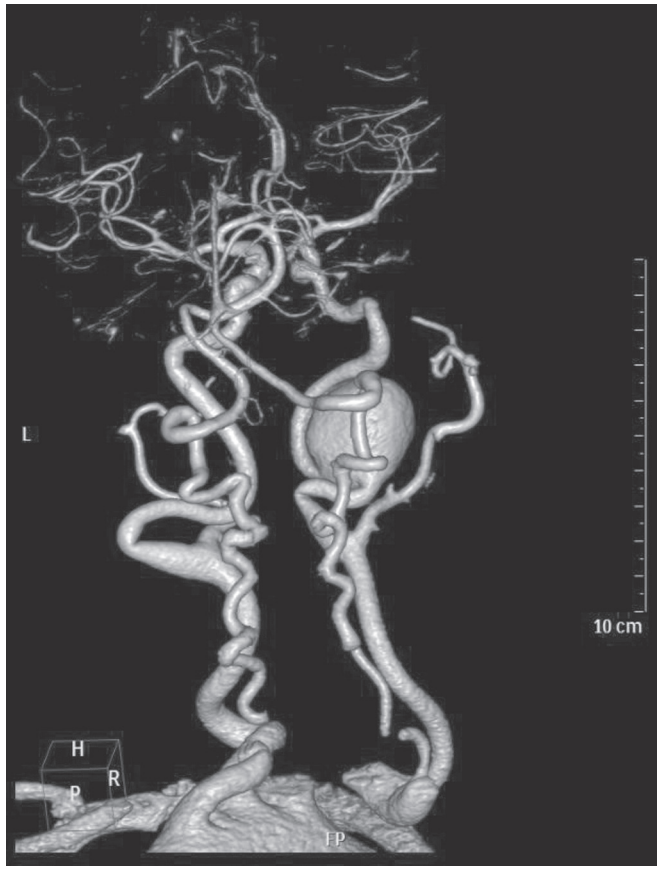

Figure 1.—3D CTA of a saccular aneurysm of the ICA. 3D: 3-dimesional, CTA: computerized tomographic angiography, ICA: internal carotid artery.

Treatment options

Current management of an ECAA is based on the clinical presentation, etiology and location of the aneurysm.[8] Treatment aims to relieve symptoms and/or prevent complications such as intra-cerebral thrombo-embolic events or pharyngeal compression. The treatment strategies currently available are conservative and interventions with an open surgical approach, an endovascular treatment or a hybrid approach, which combines an open and endovascular approach.[8] The conservative approach (antihypertensive medication, statin therapy

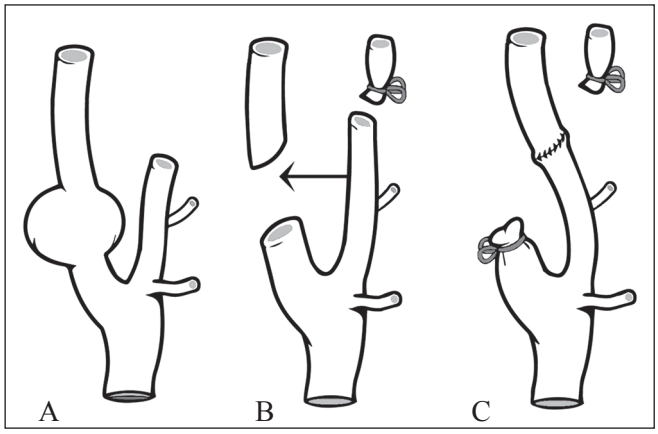

Figure 2.—Aneurysm resection with transposition of the ECA to ICA. A) ECAA located in the ICA. B) Resection of the aneurysm and ligation of the distal ECA. C) Transposition of the proximal ECA to the distal ICA and ligation of the proximal ICA. ECAA: extracranial carotid aneurysm, ICA: internal carotid artery.

and/or antiplatelet therapy) may be appropriate in asymptomatic non-growing aneurysms, inoperable cases and patients with life limiting comorbidities. However, very little data are available on the natural follow-up of asymptomatic patients. Currently patients with a symptomatic ECAA are considered best treated by invasive treatment. Surgeon/interventionist experience certainly plays a role in the type of treatment offered to the patient. Surgical resection of the complete aneurysm sac with direct reconstruction or an interposition graft to restore the blood flow is currently the gold standard.[1-3, 5] Access to the ICA is the most important factor in planning a surgical intervention. The location of an ECAA has been used in different classifications. Most studies refer to the cervical vertebrae to indicate the ECAA location. The Bouthillier classification describes the cervical part as C1, the petrous part as C2 and the intracranial part as C3-C7.[9] According to the classification of Attigah and Malikov this line of Blaisdell is used to determine if accessibility can be achieved. This line is a "virtual" line between the mastoid and the mandible angle. If the carotid artery is located above this line it is considered less accessible by a standard surgical approach, making alternative exposure techniques and/or endovascular assistance necessary.[10, 11] Operative resection of an ECAA with or without arterial replacement graft has been associated with the risk of stroke and cranial nerve damage.[2] A recently published review, although based on small series, demonstrated low stroke numbers in both surgical and endovascular treatment group.[2]

Treatment option: surgery

If there is any doubt about accessibility of the distal ICA, the surgeon must decide whether certain additional procedures (e.g. mandibular subluxation) are necessary preoperatively. Most surgeons perform carotid aneurysm surgery with the patient under general anesthesia. With the patients neck slightly extended and rotated, the surgeon can gain access to the carotid bifurcation, internal jugular vein and the ICA by careful dissection, paying special attention to the hypoglossal and the vagus nerve. Vascular clamps can obtain proximal and distal carotid control but, in the case of distal aneurysm extension, a Fogarty catheter might also be helpful. Before the artery is clamped, the patient should be systemically heparinized to prevent thrombotic occlusion. Perfusion of the brain could significantly decrease by temporary clamping the carotid artery; therefore an intraluminal shunt may be used either in a standardized fashion or selectively when based on intraoperative neuromonitoring.[12] After gaining access to the ECAA one of these options may follow; 1) ECAA resection with ligation of the proximal (inflow) and distal (outflow) part of the extracranial ICA; 2) bypass without resection; and 3) resection of the aneurysm with direct or indirect reconstruction of the blood flow.[8] Sir Astley Cooper was the first to perform ligation of the carotid artery and it is still performed in selected cases such as patients with a ruptured or mycotic an ECAA.[8, 13] Although ligation is associated with an increased risk of stroke, it is believed that it can be safely performed if the backpressure in the carotid stump exceeds 70 mmHg, suggesting an intact contralateral blood flow and an intact circle of Willis.[5] In some cases, an aneurysm cannot be resected, due to the large size, extension towards the base of the skull or because of adherence to adjacent structures. In these cases it may be possible to balloon occlude the aneurysm and then bypass the aneurysm by creating an extracranial-intracranial (EC-IC) bypass. By not excluding the aneurysm from the circulation, the ECAA could still cause neurological symptoms due to embolization.[14] Following complete resection of the ECAA, the ICA can be reconstructed in different ways. In case of an elongated carotid artery adjacent to the ECAA (Figure 1), it is possible to perform an end-to-end anastomosis or the external carotid artery can be used as a proximal transposition site (Figure 2).[5] Another reconstruction method is using an autologous saphenous vein as a graft to cre-

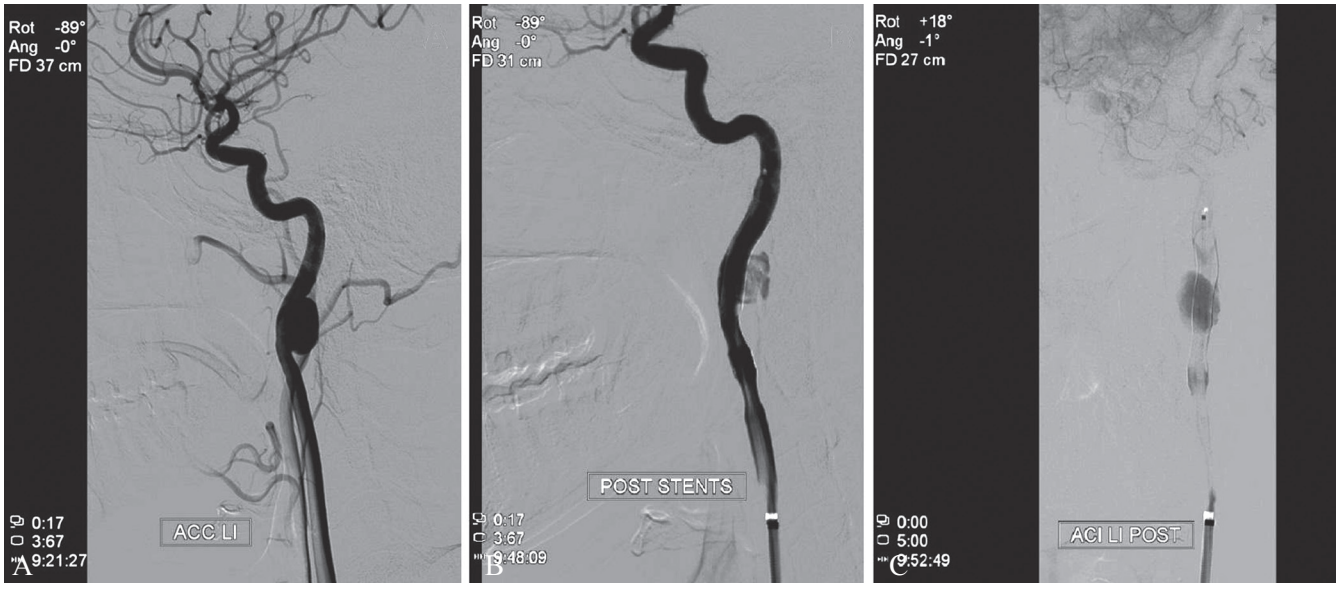

Figure 3.—Flow diverting stent. A) Angiogram of the left ICA with an ECAA prior to stenting. B, C) Angiogram post stenting with reduced flow in the aneurysm. ECAA: extracranial carotid aneurysm, ICA: internal carotid artery.

ate an interposition bypass. If a vein is unavailable, a polytetrafluoroethylene (PTFE) or Dacron interposition graft can be used. An end-to-end or end-to-side anastomosis can be made between the native artery and the graft. Partial resection of an aneurysm can be performed in selected cases, when complete resection is not possible. Placing a patch or direct closure can repair the remaining defect**.** This method should be considered as the last option, because it leaves a part of the aneurysm wall behind which is prone to dilation and thrombus accumulation.[14]

Treatment option: endovascular

Stent placement instead of surgical treatment can be considered in patients with comorbidities that make surgery too risky. Endovascular treatment is also favorable in patients with a "hostile neck" due to e.g. radiation, patients who have had previous surgery in the neck area or when the aneurysm is considered to be inaccessible (above the Blaisdell line). Endovascular procedures are typically performed under local anesthesia. Access to the ICA is obtained through a percutaneous common femoral artery puncture. An alternative approach is through a direct puncture of the proximal common carotid artery (CCA). A cerebral protection device may be deployed in the distal ICA to prevent embolization complications. When placing the stent it must cover the entire aneurysm with the proximal and distal landing zones on "lesion-free" arterial wall to secure a "healthy-to-healthy" bridge. Stent choice depends on the arterial anatomy and aneurysm characteristics. Different stents are available including bare metal stents, balloon-expendable stents, self-expandable stents, covered stents and tapered and non-tapered stents. Self-expandable stents are mostly applied in stenotic carotid artery lesions. Bare-metal stents are usually the stent of choice for ECAA. They alter the blood inflow resulting in thrombosis of the aneurysm and at the same time preserving the vessel patency. Bare-metals stents (BMS) can also be used in combination with coil embolization if complete embolization with a BMS alone does not occur.[5, 15, 16] Detachable coils are inserted into the aneurysm with a micro-catheter that is placed through the struts of the uncovered stent. These coils obstruct the blood inflow in the aneurysm cavity, which induces thrombus formation.[5] In non-branching arteries, wide-necked aneurysms and pseudo-aneurysms a covered stent can be used. These stents might reduce the risk of embolization during the procedure by trapping debris in the aneurysm sac that would otherwise protrude through the orifices of a bare-metal stent.[16] A disadvantage of covered stents is the need for a large delivery system, making the procedure technically challenging.[5]

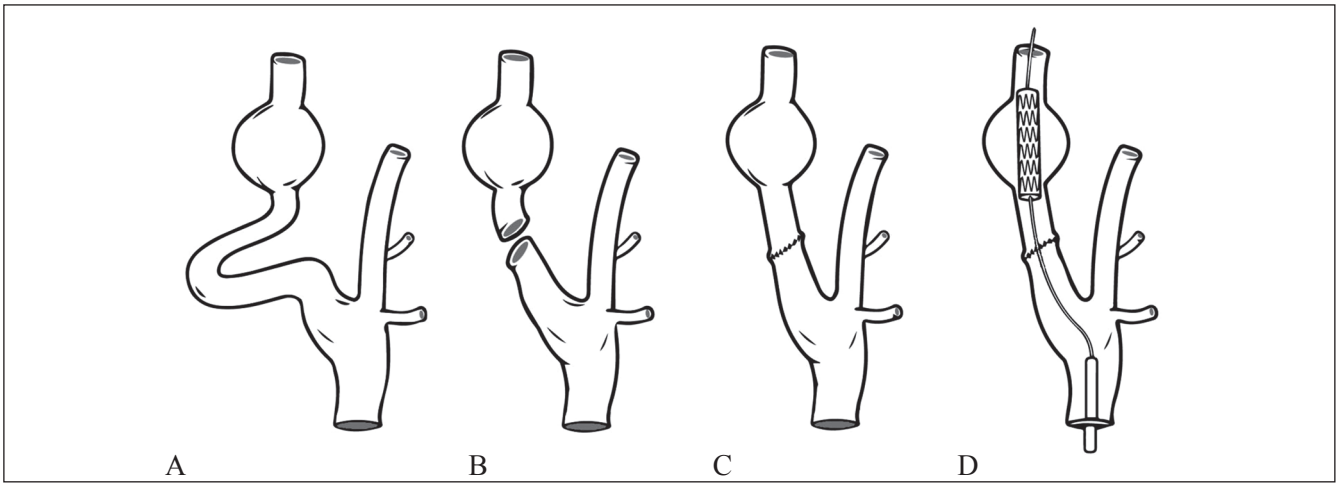

Figure 4.—Hybrid approach. A) Aneurysm located in the distal ICA. B) Resection of a loop in the ICA. C) Primary end-to end anastomosis. D) Endovascular stent placement over the ECAA. ECAA: extracranial carotid aneurysm, ICA: internal carotid artery.

Flow diverting stents are a more recent development in stent designs. These laser-cut stents are designed to alter blood flow in the longitudinal axis (Figure 3). Like BMS, flow diverters promote thrombosis in the aneurysm sac and have been described to be effective in the exclusion of intracranial aneurysms.[17] One recently published case report described the successful treatment of an ECAA with a flow-diverting stent without any procedural or follow up complications.[17]

Treatment option: hybrid approach

A seldom performed procedure is the hybrid approach, which combines open surgical exposure of the proximal CCA and endovascular stent placement. During this procedure kinking and arterial loops that hinder endovascular stent placement, can be removed (Figure 4). Resection of the artery loop is followed by an arterial anastomosis (or anastomosis is made after stent placing), then a stent is placed in the more distally located ECAA.[15] This approach has been performed on distally located ECAA with proximal loops or kinks according to a recent published case series.[15] No procedural complications like cerebral ischemia occurred and no deaths or strokes were reported within 30 days of intervention. These patients had no local or neurological symptoms during follow up, demonstrating good clinical and technical results of the hybrid procedure with placing bare metal.[15]

Treatment option: endovascular embolization

The placement of a coil in an aneurysm sac as a treatment for an ECAA or any other peripheral aneurysm is rarely performed. During the procedure a micro-catheter is passed through the neck of the aneurysm where the coil will be placed, which has a high risk of coil migration. Therefore coils are used in combination with a bare metal stent. Percutaneous thrombin injection in the aneurysm, a treatment applied to pseudo-aneurysm in the femoral artery, is considered technically difficult and too risky in ECAA because of the mostly fusiform shape of ECAA and the possibility of thrombin migrating into the intracranial system causing ischemic complications.[18]

Discussion

ECAA is a rare condition and if left untreated or not treated on time when indicated, it may cause serious morbidity. Little is known about the natural course of both asymptomatic and symptomatic ECAA. Further, several treatment options exist but there is no clear guidance on the optimal type of treatment. Imaging modalities like aneurysm volumetry with a CT scan and the correlation between MRI and the histology of an aneurysm are believed to help predict the natural course of an ECAA. Future studies with long term follow up are needed to demonstrate if these imaging modalities can help physicians to decide whether an aneurysm should

be treated or not. However, with the lack of evidence based treatment guidelines physicians are faced with a therapeutic dilemma. Therefore a web-based international registry has been designed to collect data on ECAA (www.cartoidaneurysregistry.com).[19, 20] This registry opts to prospectively analyze the available data of both conservative and invasively treated ECAA in 1 and 5 years' time.[20] Subsequently, physicians could be accurately advised which patient with an ECAA should be treated and how.

Conclusion

There is no evidence based guidance to indicate which patient needs to be treated nor is there an unequivocal recommendation for the optimal type of treatment for ECAA. A conservative strategy may be warranted for the asymptomatic, non-growing aneurysms, with or without medication. The current treatment of choice for symptomatic or growing aneurysm is surgical repair, with resection of the aneurysm and reconstruction of the blood flow the considered gold standard. Endovascular techniques with stent placement for exclusion of an aneurysm is the accepted alternative treatment for patients that cannot undergo surgery due to different comorbidities, high risk of complications, or patients with anatomical variations. The optimal treatment of patients presenting with ECAA is yet to be investigated. Important insights may be gained within a few years with the help of an international multicenter registry on this pathology.

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