Extracranial Carotid Artery Aneurysm: Optimal Treatment Approach

Extracranial carotid artery aneurysms (ECAA) are very rare, but exact data on the incidence are lacking. ECAA are mostly located at the carotid bifurcation or the distal part of the internal carotid artery. Presentation is usually around the age of 60, dependent on etiology, which is diverse and includes atherosclerosis, infection, fibromuscular dysplasia, connective tissue disease, and traumatic or spontaneous dissection. Most ECAA appear to be found by coincidence in asymptomatic patients. When symptomatic, cerebral thromboembolization and local compression are most often reported, while the risk of ECAA rupture, although a feared complication, is low. Local compression may lead to peripheral neurological dysfunction of the cranial nerves or dysphagia.

Data on natural follow up are scarce. In recent decades, mostly case reports and small case series have been published on ECAA. These observational studies suffer from incomplete data and most lack long-term follow up. The natural course of ECAA is poor, with a reported stroke rate of up to 50%. It has been suggested that asymptomatic ECAA could be treated conservatively with regular follow up. On the other hand, surgery is generally accepted as the treatment of choice for symptomatic ECAA, with complete resection of the aneurysm sac followed by arterial reconstruction being considered the gold standard approach. More recently, small case series advocating an endovascular approach to treat ECAA reported favorable procedural results but with a limited number of cases and no mid- or long-term follow up.

In a recent systematic review, 39 series were found describing 10 patients or more with a total of 1239 patients. However, most series reported their data only at group level, making the confounding by indication impossible to correct, and no comparison could be made between different treatment regimens. A systematic review of endovascular stenting in ECAA patients revealed that so far endovascular stenting has been applied, mainly in patients with traumatic ECAA, with a high success rate (92.8%). Despite this, the overall in hospital mortality was 4.1%. With a mean follow up period of 15.4 months, a high patency was reported and late stroke was <1%. The effect reported in this review remains uncertain while it is based only on records with a low quality of evidence. Furthermore, in a recent histological analysis, a clear shift was observed in the previously described etiological background of ECAA from atherosclerotic to post dissection carotid dilatation.

For a proper assessment of the benefit and complication risk from the different revascularization options for ECAA, a better insight in vascular procedural outcome is needed and especially in the natural follow up. Given the limited number of patients, randomized controlled trials are not feasible in rare diseases such as ECAA. Disease registries have become essential for the investigation of such diseases, thanks to their potential to describe the natural history. The importance of these disease registries has been acknowledged by EUCERD, an EU committee of experts in rare diseases that discusses policies and recommends activities in collaboration with EU agencies. To collect data, the Carotid Aneurysm Registry (CAR) was designed, a prospectively web based international registry assessing natural course, intervention results, and follow up data in patients with an ECAA.

The World Health Organization’s definition of a patient registry is “a file of documents containing uniform information about individual patients, collected in a systematic and comprehensive way, in order to serve a predetermined scientific, clinical or policy purpose.” Many registries begin on a voluntary and unfunded basis, but can develop into effective international disease registries. Two examples of successful registries are The Pompe Registry and The Cystic Fibrosis (CF) Registry.

The Pompe Registry is an ongoing long-term, international registry initiated in 2004, sponsored and administered by Genzyme. This registry contains the largest dataset on patients with Pompe disease and was started in order to gain information on the natural history of Pompe disease. Through electronic case report forms, information is collected on clinical manifestation, progression of the disease, and outcome. Analysis of data from this registry has resulted in many new insights, including timing and execution of diagnostic tests.

The CF Registry started in the USA in 1966 and in Canada in 1970. This combination of national CF registries was set up to describe population patterns of diagnosis, demographics, and mortality in CF patients. This registry demonstrated successful treatments which could be applied in specialist CF centers. Furthermore, the results from this registry, described in numerous publications, are used in the daily care for CF patients with positive consequences on quality of life and life expectancy.

International cooperation within a (web-based) registry has clear advantages. First of all, obviously, the number of patients increases. This results in detection of smaller effects and more strength to support detected effects within the
database. Working with different geographic groups also creates the possibility of comparing different treatment models by appropriate matching of cases. In rare diseases, because of lack of evidence based guidelines, expert opinion will influence the local treatment model creating different approaches within an international registry. Finally, bringing large groups of researchers together from different countries may result in new research ideas and strengthen further collaboration within the vascular surgical field.

The CAR is an international web-based prospective registry in which literally all patients with an extracranial aneurysm of the carotid artery can be included, independent of the treatment approach chosen. Also patients in whom a conservative approach is initiated are of highest interest for the registry.

The primary aim of this registry is to collect data to assess the natural history of ECAA. The further objectives of the CAR are to provide data on existing and evolving practice in ECAA diagnostics and treatment. Furthermore, it is intended to identify risk factors for thromboembolic complications related to the aneurysm and the safety and durability of different treatment strategies. All items that will be scored in the registry are listed in the CAR protocol and on the study website, www.carotidaneurymregistry.com. The primary endpoint in this registry is occurrence of aneurysm related symptoms at 30 days, 1, 3, and 5 years. The secondary endpoint in invasively treated patients comprises treatment safety. Complications may include peripheral nerve dysfunction (sensory and motor dysfunction), local hematoma or infection, local pain or headache, transient ischemic attack, ischemic stroke, local bleeding or aneurysm rupture, and all cause mortality. An endpoint adjudication committee will assess all endpoints reported.

Hopefully, the results from this prospective international registry (results will be analyzed in 1 and 5 years’ time) will provide greater clarity regarding ECAA diagnostics and treatment. The registry is now online and all vascular interventionists and specialists involved in the clinical care of patients with ECAA are invited to participate in this international collaborative project.

The project group of CAR consists of the following persons (in alphabetical order): A.F. Baas a, R.L.A.W. Bleys b, G.J. de Borst (chairman) c, M.L. Bots d, J. Hendrikse e, L.J. Kappelle f, T.H. Lo e, F.L. Moll c, G.J.E. Rinkel f, Y.M. Ruigrok f, M.D.I. Vergouwen f, J.C. Welleweerd f, H.B. van der Worp f

a Department of Medical Genetics, UMCU, Utrecht, The Netherlands
b Department of Anatomy, UMCU, Utrecht, The Netherlands
c Department of Vascular Surgery, UMCU, Utrecht, The Netherlands
d Julius Center for Health Sciences and Primary Care, UMCU, Utrecht, The Netherlands
e Department of Radiology, UMCU, Utrecht, The Netherlands
f Department of Neurology and Neurosurgery, UMCU, Utrecht, The Netherlands

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REFERENCES


J.C. Welteewerd, G.J. de Borst*, on behalf of the Carotid Aneurysm Registry Project Group

Department of Vascular Surgery, UMCU, Utrecht, The Netherlands

*Corresponding author. G.J. de Borst, University Medical Center Utrecht, Department of Vascular Surgery G04.129, PO Box 85500, 3508 GA Utrecht, The Netherlands.

Email-address: G.J.deBorst-2@umcutrecht.nl (G.J. de Borst)