Chapter 1

The Rare Diagnosis and Operations Register

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This book is born out of an idea conceived during a Joint Vascular Research Group meeting in the mid 1990s. At the time, the project was called the Rare Diagnosis and Operations Register.

The Rare Diagnosis and Operations Register is a collection of unusual vascular diagnoses and operations seen or performed by members of the Joint Vascular Research Group. Members contribute cases to the author who stores them in a database, which is then available to any member of the group. It was set up in May 1995 following the spring meeting of the Joint Vascular Research Group.

Aims

The database was set up with a number of aims in mind. These are as follows:

- To create a collection of unusual cases.
- To provide members with the opportunity of publishing, separately or jointly, collective reports of unusual vascular conditions.
- To allow expertise and advice to be disseminated between members for the management of unusual cases.
- To encourage contribution to the JVRG generally by promoting a competitive culture.

Method

From the outset, members were asked to contribute any cases they thought unusual. In order to encourage as many members to contribute as many cases as possible, no preconditions were set.

To facilitate data collection, a minimum of data were required on each case. These are summarised in Table 1. Latterly, hospital number has become the only patient identifier collected.

Each case submitted was entered in a database with the date of submission. At the beginning, there were a small number of categories that we particularly wanted to collect, though submissions were not confined to those groups. When more than three similar non-categorised cases were submitted, a new category was created.

Table 1. Data required on each case.

Surgeon data	Patient data
Name	Surname
Centre	First name
	Date of birth
	Hospital number
	Case details

Table 2. Categories of cases submitted.

Aortocaval fistula	Major vessel arteritis
Aorto-enteric fistula	Mesenteric ischaemia
Arm ischaemia	Mycotic aneurysm
Arteriovenous malformation	Other odd aneurysms
Brachial embolus	Paget Schroetter syndrome
Leg ulcer calcification	Popliteal entrapment
Carotid artery surgery	Profunda femoris aneurysms
Carotid body tumour	Right to left shunt or paradoxical embolism
Cystic degeneration	AAA with renal abnormality
Deep vein thrombosis	Subclavian steal syndrome
Radiotherapy arteritis	Thoracic outlet compression
Ehlers-Danlos syndrome	Vascular trauma
Klippel-Trenaunay syndrome	Visceral aneurysms
Systemic lupus erythematosus	Vena caval obstruction
Mid-aortic syndrome	Arterial disease in patients under 40 years

To start there were six categories. These included: popliteal entrapment, major vessel arteritis, carotid body tumour, mesenteric ischaemia, cystic degeneration and lupus. At the time of writing there are 30 categories and these are shown in Table 2.

The reports

Regular reports have been produced which are circulated to the membership. Reports are in several parts:

- A table of numbers submitted by category and by centre in total and since the last report.
- A list of all new cases showing contributing surgeon, category and details of case.
- A list of non-categorized cases with their details by centre.
- A newsletter.
- A proforma for subsequent case submissions.

Early on the cases were attributed to individual surgeons but as the size of the group has increased cases are now classified by referring centre. There are now 22 centres with approximately 30 contributing surgeons. Sent with the report are a newsletter and a proforma for completion during the next time interval. More recently, submissions directly by email have become possible.

The results

The Rare Diagnosis and Operations Register has been running for ten years. During that time 1322 cases have been submitted.

This book has chapter headings that reflect the categories in the Register. Chapter authors have been encouraged to contact other members of the group for material for their chapters as has previously been done for scientific publications [1,2]. The anonymised database is available on the JVRG website as a resource for members and others needing information and advice on how to manage that very unusual case, and to disseminate information.

References

1. Lambert AW, Wilkins DC. Popliteal artery entrapment syndrome: collaborative experience of the Joint Vascular Research Group. *Br J Surg* 1998; 85: 1367-8.
2. Varga ZA, Locke-Edmunds JC, Baird RN. A multicentre study of popliteal aneurysms. Joint Vascular Research Group. *J Vasc Surg* 1994; 20: 171-7.

Chapter 2

Anatomical variation for the vascular specialist

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Regional arterial variants

Aortic arch and carotid arterial anatomy

The derivation of a single aortic arch from six embryological arteries gives huge scope for the development of variant anatomy. Many such variants are relevant to endovascular procedures of the carotid or thoracic aortic territory. Those of the supracervical carotid segment are largely the concern of neuro-interventionists and neurosurgeons. Variants of the cervical carotid impact on vascular surgical practice when pathology presents fortuitously, or because the variant itself generates disease.

The bovine arch

A common origin of both common carotids and the right subclavian artery occurs in 10% of people. As five of six primary sources of cerebral perfusion arise from a single aortic branch, disease of this common trunk is a threatening disease pattern offering substantial surgical challenges; thankfully, it is uncommon. A large vascular unit accrued only six such patients over two decades [1]. Revascularisation strategies include: prosthetic bypass graft from the ascending aorta to the innominate or left common carotid arteries or both, or endarterectomy to include branch vessels. This small series reported two neurological complications, one fatal ischaemic stroke and one hyperperfusion syndrome, attesting to the threat posed by this pattern of disease. Furthermore, this pattern does not easily lend itself to a less invasive (endovascular) solution.

Variations in the position of the carotid bifurcation

The carotid bifurcation may lie higher than usual, at the level of the hyoid bone, or more rarely in a lower position than normal, level with the middle of the larynx or the lower border of the cricoid (Figure 1). Alternative patterns include absence of a recognisable bifurcation, when both internal (ICA) and external carotids (ECA) arise directly from the aortic arch, or failure of common carotid branching in the neck.

If the bifurcation is high, it may be difficult to expose a sufficient length of ICA to permit safe carotid endarterectomy (CEA). Adequate exposure may require division of the posterior belly of digastric and occasionally, disarticulation of the mandible. Endarterectomy in the setting of a high carotid bifurcation is associated with a greater incidence of