Surgical Management of Right Sided Aortic Arch Aneurysms

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Abstract

Aneurysms and dissections of the right sided aortic arch are rare and published data is limited to few case reports and small series. The optimal treatment strategy of this entity and the challenges associated with their management are not yet fully investigated and conclusive. We performed a systematic review of the literature to identify all patients who underwent surgical or endovascular intervention for right aortic arch aneurysms or dissections. We focused on presentation and critically assessed different management strategies and outcomes.

We identified 74 studies which reported 99 patients undergoing surgical or endovascular intervention for a right aortic arch aneurysm or dissection. Median age was 61 years. The commonest presenting symptoms were chest or back pain and dysphagia. 88 patients had an aberrant LSCA with only 11 patients having the mirror image variant of a right aortic arch. The commonest pathology was aneurysm arising from a Kommerell’s diverticulum occurring in over 50% of the patients. 28 patients had dissections, 19 of these were type B and 9 were type A. 81 patients had elective operations whilst 18 had emergency procedures. 67 patients underwent surgical treatment, 20 patients had hybrid surgical and endovascular procedures and 12 had totally endovascular procedure. There were 5 deaths, 4 of which were in patients undergoing emergency surgery and none in the endovascular repair group.

Aneurysms and dissections of a right sided aortic arch are rare. Advances in endovascular treatment and as well hybrid surgical and endovascular management is making this rare pathology amenable to these approaches and may confer improved outcomes compared to conventional extensive repair techniques.

Keywords: Right sided aortic arch; Kommerell’s diverticulum; Aneurysm; Dissection

Introduction

A right sided aortic arch was first described 250 years ago by Fioratti and Aglietti [1]. It results from alterations in the normal embryonic development with regression of the left fourth arch or the left dorsal aorta whilst the right dorsal aorta remains patent.

The commonest type of right sided aortic arch is the mirror image type where the first branch of the aortic arch is the left branchiocephalic artery then the right common carotid artery (RCCA) and finally the right subclavian artery (RSCA) [2]. The other main type of right sided aortic arch is an aberrant left subclavian artery (LSCA) originating from the diverticulum of Kommerell which is a remnant of the left arch. This can run posteriorly to the oesophagus 2. The branches originate from the aortic arch in the following order: left common carotid artery (LCCA), RCCA, RSCA and aberrant LSCA. If a left sided ligamentum arteriosum is present it will connect the left subclavian artery to the left pulmonary artery which forms a vascular ring with the potential to compress mediastinal structures [2].

A right sided aortic arch is found in 0.04–0.1% of autopsy studies [2]. There is an association with 22q11 deletion hence aortic arch laterality and branching can be a part of a spectrum of other cardiovascular anomalies [3]. Patients with right sided aortic arches are normally asymptomatic however symptoms can occur either due to the aberrant anatomy of the right sided aortic arch leading to compression of mediastinal structures or due to aneurysmal disease or dissection [4].
Compressive symptoms can lead to presentation, often in infancy, of dysphagia as a result of oesophageal compression or respiratory symptoms such as cough or stridor due to compression of the trachea or bronchi.

Aneurysms of a right sided aortic arch are rare with reports being limited to case reports and small case series. Aneurysmal disease often originates from Kommerell’s diverticulum (KD) resulting in Kommerell’s aneurysm located at the origin of the aberrant LSCA. This can also be a site for dissections to originate from [4]. Distinguishing between a true KD which is an embryonic phenomenon and an aneurysm arising from the origin of the aberrant LSCA is difficult due to atherosclerotic changes affecting the artery [48].

Right sided aortic arch aneurysms and dissections span two subspecialties of cardiac surgery. Right aortic arches are most commonly seen by congenital surgeons in patients who require surgery to relieve oesophageal or tracheal compression. However, aortic dissections and aneurysms are the remit of aortic surgeons. Management of right sided aortic aneurysms requires knowledge from both of these subspecialties to treat these complex pathologies.

Given the rarity as well as the heterogeneity of right aortic arch aneurysms and dissections there has been no accepted gold standard of treatment. Access to the right sided aortic arch is difficult [5]. Many different surgical and more recently endovascular techniques have been described.

We performed a systematic review of the literature to study all patients who underwent surgical or endovascular intervention for right aortic arch aneurysms or dissections. We focus on patient presentation and management strategies for addressing the different pathologies which bridge competencies from aortic and congenital subspecialisation. Finally we report short term outcomes from different management strategies.

**Methods**

Our search strategy was done in accordance with guidelines for the ‘Preferred Reporting Items for Systematic reviews and Meta-Analyses’ PRISMA [6]. We performed a PubMed search with the search criterion right aortic arch aneurysms. Articles were reviewed and if the article reported patient(s) with a right sided aortic arch aneurysm or dissection that underwent surgery on the thoracic aorta then it was included and the full text was retrieved. Our search was restricted to articles published in English only. We performed the search in October 2014 and looked at all historical articles (Figure 1). We also reviewed the references of all relevant articles and reference lists of review articles. Patient characteristics and pathologies, different treatment modalities and outcomes where examined and critically analyzed.

**Results**

**Reported studies and population characteristics**

We identified 74 studies which reported 99 patients undergoing surgical or endovascular intervention for a right aortic arch aneurysm or dissection. Most studies were case reports with some case series with the largest reporting 4 patients [13], 58.62 patients were male, 21 were female and for 16 patients the gender was not reported. The median age at presentation was 61 years (range 21-85 years).

The commonest presenting symptoms were chest or back pain in 31% (31 patients), dysphagia in 20% (20 patients), respiratory symptoms such as cough, stridor or haemoptysis in 10% (10 patients). 18% of patients (18 patients) were asymptomatic and the finding was incidental (Figure 2). The combination of dysphagia and cough are as frequent as the ‘typical’ symptoms of aneurysms, therefore high level of suspicion in this subgroup.

Diagnosis of a right sided aortic arch was most commonly made with a chest radiograph which was then confirmed with computed tomography.

88 patients had an aberrant LSCA with only 11 patients having the mirror image variant. The commonest pathology was an aneurysm arising from KD which occurred in 54 patients. 28 patients had dissections, 18 of these were type B and 9 were type A (Figure
3. Some of these type B dissections originated from KD. Other pathologies were aneurysms arising from elsewhere in the thoracic aorta.

**Interventional approaches**

81 patients had elective operations whilst 18 had emergency procedures. 67 patients underwent surgical treatment, 20 patients had a hybrid surgical and endovascular procedures and 12 had a totally endovascular procedure.

Of the hybrid procedures 6 patients had a total arch replacement with an elephant trunk graft formed by a thoracic endovascular aortic repair (TEVAR) with an endograft inserted into the elephant trunk [7]. 5 patients had a frozen elephant trunk and 1 patient had a modified elephant trunk. 8 patients had bypass procedures of the arch vessels followed by a TEVAR procedure [8,9].

For the surgical group the commonest way of accessing the aorta was via a median sternotomy or a right thoracotomy of the patients with information on their incision 32 patients had a sternotomy, 4 of whom had mini sternotomies. 22 patients had a right thoracotomy with information on their incision 32 patients had a sternotomy, 4 of whom had a modified thoracotomy. 8 patients had bypass procedures of the arch vessels followed by a TEVAR procedure [8,9].

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Table 1: Surgical complications for all patients in whom this information was available.

<table>
<thead>
<tr>
<th>Complication</th>
<th>Number of patients (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Death</td>
<td>5 (5%)</td>
</tr>
<tr>
<td>Hoarse voice</td>
<td>6 (6%)</td>
</tr>
<tr>
<td>Endoleak</td>
<td>5 (5%)</td>
</tr>
<tr>
<td>Respiratory complications</td>
<td>7 (7%)</td>
</tr>
<tr>
<td>Graft obstruction</td>
<td>1 (1%)</td>
</tr>
<tr>
<td>Intraoperative oesophageal tear</td>
<td>1 (1%)</td>
</tr>
<tr>
<td>Re-sternotomy for bleeding</td>
<td>1 (1%)</td>
</tr>
<tr>
<td>Mediastinitis</td>
<td>1 (1%)</td>
</tr>
</tbody>
</table>

Complications

The commonest complications reported were hoarse voice as a result of injury to the recurrent laryngeal nerve (6 patients, 6%) and respiratory complications (prolonged respiratory wean and chest infection (7 patients, 7%) (Table 1). There were 5 deaths, all of these except 1 were in patients undergoing emergency surgery and none in the endovascular repair group.

The first patient presented with a type B dissection and underwent emergency surgery with an interposition graft performed via a right thoracotomy. The LSCA was ligated. The patient never regained consciousness and died 3 days post operatively [10].

The second patient presented acutely with haemoptysis and was found to have fistula formation between a proximal right aortic arch aneurysm and the right upper lobe of the lung. The patient underwent emergency surgery and a knitted polyester patch (Gelseal™) closure of the fistula was performed as well as an interposition graft for an additional aneurysm of the DTA. He subsequently developed a peri-graft abscess that ruptured into the oesophagus that resulted in exsanguinations [11].

The third patient died of a massive pulmonary embolism after undergoing elective LSCA to carotid transposition and interposition graft to treat an aneurysm arising from KD [4].

The fourth patient presented acutely and was found to have rupture of an aneurysm arising from KD. They underwent emergency LSCA to RSCA bypass followed by total arch replacement. The cause of death was reported as mediastinitis post operatively [14].

The last patient presented acutely with a type A dissection. Total arch replacement was performed, however they were unable to resect KD and so this was left. On post-operative day 30 the patient had massive haematemia leading to death. Post mortem examination revealed a fistula between KD and the oesophagus [13].

**Discussion**

Our systematic review found 99 patients with right aortic arch aneurysm or dissection, highlighting the rarity of this condition. A higher proportion of patients were male and the median age of presentation was 61 years old. Pain was the commonest presenting symptom and was either felt in the chest, epigastrium or back. Compression of the trachea, bronchi or the oesophagus leading to respiratory symptoms or dysphagia, respectively, was responsible for most of the other presentations. 89% of patients had an aberrant LSCA and only 11% had a mirror image variant. This is differs from pathological studies of the right aortic arch where 85% of patients have the mirror image variant and 15% have an aberrant LSCA2. This difference is likely the result of KD predisposing to aneurysm formation as this area constitutes an area of weakness in the aortic wall that is a potential area for aneurysmal dilatation and dissections 49. Additionally the median age at presentation was 61 years which differs from patients with a left sided aortic arch in whom the median age of presentation is about 65 years [14]. When there is a left sided aortic arch it is the ascending aorta that accounts for about 70% of the pathology [14]. These differences further highlight the significance of KD on causing aortic pathology in a right sided arch.

CT was the commonest imaging modality for identifying right aortic arch pathology. This is due to the availability of CT scanning as well as the speed of scanning and high spatial resolution (Figure 4). Magnetic resonance imaging (MRI) has also been used in assessing aortic pathology in patients with right sided aortic arches. It has the disadvantage of being less available than CT and the increased time taken to obtain images excludes its use in emergency settings. However it useful in assessing the pulsatile nature of tracheal and oesophageal compression as well as the advantage of not using ionising radiation. A case report from almost 20 years ago in a patient with a type B dissection in a right aortic arch found that differentiation between a dissection and an aortic aneurysm with mural thrombus may be difficult on CT compared with MRI, however CT imaging has improved greatly in that time [15].

The decision of when to operate on a patient with an aneurysm arising from KD in patients with a right sided aortic arch is not straightforward. Clearly troublesome symptoms caused by compression of mediastinal structures are an indication to intervene. Advising on
surgical intervention based on size is hampered by the paucity of data. Balancing the risks of surgery against the risk of potential rupture if left untreated is difficult. Some researchers have advised to intervene when the diameter is greater than 3 cm whilst others recommended 5 cm as the point for intervention [4,16]. However rupture has been described in aneurysms measuring as little as 2 cm [4]. In the absence of significant comorbidities and with advent of less invasive techniques it may be that the size threshold for aneurysms of the right aortic arch is lower than the ones arising from the left.

The surgical or endovascular approach depends on the pathology and site of the aorta affected. We discuss the approaches used depending on the pathology.

**Kommerell’s aneurysm**

Aneurysms arising from KD were the commonest pathology, occurring in 54% of patients (n= 54). The treatment varied depending upon the extent of the aneurysm.

For Kommerell’s aneurysms that involved the arch and descending thoracic aorta (DTA) repair could either be performed as a single procedure or staged procedure. The frozen elephant trunk graft, which has been described previously, allows this to be performed as a single procedure (Figure 5). Idrees et al. [14] reported doing this through a mini J sternotomy. The alternative is a 2 stage procedure where the arch is replaced in the first instance with an elephant trunk graft followed by either a TEVAR endograft inserted into the elephant trunk8 or surgical replacement of the DTA via a right thoracotomy [17].

For more localized aneurysmal dilatation of KD this could be addressed by a number of surgical techniques. Options included transposition of the LSCA to the LCCA followed by endoaneurysmorrhaphy5 or ligation of the proximal and distal ends of the aneurysm followed by bypass between the aorta and LSCA with a synthetic graft [18]. All of the described approaches persevered blood flow through the LSCA. Access was achieved via a median sternotomy, right or left thoracotomy. These procedures were performed either without bypass or with partial bypass from the left atrium to the femoral artery or to the DTA. Left heart bypass allows isolation of an aneurysm arising from KD as well as part of the aortic arch and DTA. An aortic cross is placed between either the RCCA or RSA and the aneurysm as well as a cross clamp on the DTA distal to the aneurysm. Cina et al. [4] used left heart bypass in 3 patients who had aneurysms arising from KD with or without arch involvement and required LSCA to LCCA transposition and endoaneurysmorrhaphy. If a cross clamp can be placed between the aneurysm arising from KD and the aorta then this could potentially treatment without bypass, however if there is more extensive involvement of the arch then a period of circulatory arrest is required.

When the aneurysm affected the arch then this could be addressed either with a total arch replacement either by anastomosing all 4 branches separately to the graft [19], alternatively some surgeons transposed the LSCA to the LCCA or RSA first and then performed total arch replacement [12]. All of these procedures were performed via a median sternotomy.

When aneurysmal dilatation was more localised to the distal arch +/- the DTA then this could be addressed with surgical, endovascular or hybrid procedures.

Surgical options were LSCA to LCCA transposition followed by resection of Kommerell’s aneurysm and the affected part of the aorta followed by an interposition graft [20]. If the DTA was also affected then this was also be replaced with a graft [20].

Endovascular options were TEVAR with the graft covering the LSCA [21], as well as endovascularly occluding the LSCA prior to TEVAR [22]. Alternatively to preserve flow through the LSCA the aortic graft could be deployed along with another LSCA covered stent directed parallel and extending more proximally to the main aortic graft (Chimney stent) [23].
Hybrid options depended on the extent that the aneurysm affected the aortic arch. For aneurysms more localized around the LSCA these could be treated with LSCA to LCCA bypass with ligation of the proximal LSCA followed by TEVAR with the proximal land zone just distal to the RSCA [8]. If the aneurysm extended towards the origin of the RSCA then RSCA to RCCA and LSCA to LCCA bypass with ligation of the proximal ends of both SCAs followed by TEVAR with the proximal land just distal to the RCCA could be performed [9]. The options for managing a patient with an aneurysm arising from KD are shown in Figure 6.

Type B dissection

18 patients had a type B dissection. Type B dissections could originate from distal or proximal to the LSCA. In some patients KD was the area from which a tear occurred resulting in a type B aortic dissection. Management can be surgical or endovascular. Surgical options if the dissection originates distally to the LSCA are with an interposition graft from the distal arch to the DTA [24]. If the dissection was proximal to the LSCA then an interposition graft between aorta just distal to the origin of the RSCA and DTA can be placed. The LSCA can either be ligated [10] or an interposition graft with a side branch that LSCA anastomoses to can be used to preserve blood flow can be used [25]. These procedures were performed via a right thoracotomy using CPB via the femoral artery and vein.

Endovascular treatment of type B dissection was performed in 6 patients and 3 of these patients had the mirror image variant. 3 patients had TEVAR procedures with an aortic stent alone whilst the other 3 had an aortic stent followed by a Chimney stent. One patient had the chimney stent for the RSCA into the aortic stent, one patient and a Chimney stent deployed in the RCCA into the aortic stent with no stent for the RSCA [26] and the final patient had chimney stents in both the LSCA and RSCA [27].

Type A dissection

9 patients had a type A dissection. 8 of these presented acutely and one was chronic [28]. Surgical strategies were similar to patients with a type A dissection of a left sided aortic arch. They included: aortic root replacement [29], interposition graft of the ascending aorta [30], total arch replacement [31], total arch replacement with elephant trunk and TEVAR into the elephant trunk [32] and frozen elephant trunk [28]. There was one death in this group that has been described above [13].

Descending thoracic aorta aneurysms

These were either managed surgically via a left thoracotomy or right thoracotomy and placing an interposition graft under circulatory arrest 19 or using left heart bypass [33] or endovascularly with a TEVAR stent deployed distally to the LSCA [34].

Specific surgical considerations in right arch

Division of ligamentum arteriosum is required in all cases. The right aortic arch can be retroesophageal where the right aortic arch passes behind the trachea and oesophagus as opposed to the standard configuration where it is in front of them [2]. This requires careful mobilisation of the retroesophageal portion, an area which is often less familiar for the general cardiac surgeon.

The means of accessing a right sided aortic arch depends on the pathology being addressed. A right thoracotomy gives better access to the right arch behind the oesophagus but hinders complex distal arch and descending repair (where a left thoracotomy is the preferred approach). Hybrid approaches allow for median sternotomy where central bypass techniques can also be safely utilized.

Other cardiac anomalies are commoner in patients with a right sided aortic arch. Focused echocardiography and coronary angiography are required due to association with septal defects and coronary anomalies (high take off of the RCA which is very relevant in the case of type A dissection) [2,35].

Limitations

Given the rarity of aneurysms and dissections of the right aortic arch the evidence is limited to case reports and small case series with short follow up. The lack of available long term data hinders...
to recommend what is the optimal approach for managing these pathologies.

**Conclusion**

Aneurysms and dissections of a right sided aortic arch are rare. The anatomy of the right sided aortic arch means that symptoms can be atypical for aneurysms as the predominant symptoms can be due to compression of the esophagus or the airways. KD appears to be significant in causing aortic pathology in a right sided arch and is the reason for a younger age of presentation in patients with a right aortic arch. Advances in endovascular treatment and as well hybrid surgical and endovascular management is making more pathology amendable to these approaches and could reduce morbidity. Preoperative planning and careful delineation of anatomy and possible associated cardiac anomalies is essential.

**References**

