Non-A Non-B Aortic Dissection: A Literature Review

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Abstract

Non-A non-B aortic dissections are an infrequent occurrence and represent a small proportion of aortic dissections. Treating this life-threatening medical emergency often requires surgeons to undertake some one of the most challenging surgical or endovascular cases in medicine. This literature review aims to define and classify non-A non-B dissections, describe their epidemiology as well as their pathology. This review also aims to discuss the range of surgical techniques employed in their treatment and management and to investigate the patient outcomes associated with each technique.

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Abstract

Non-A non-B aortic dissections are an infrequent occurrence and represent a small proportion of aortic dissections. Treating this life-threatening medical emergency often requires surgeons to undertake some one of the most challenging surgical or endovascular cases in medicine. This literature review aims to define and classify non-A non-B dissections, describe their epidemiology as well as their pathology. This review also aims to discuss the range of surgical techniques employed in their treatment and management and to investigate the patient outcomes associated with each technique.

1.0 Introduction

Non-A non-B aortic dissections are a rare occurrence and treating this life-threatening medical emergency often requires surgeons to undertake some one of the most challenging surgical or endovascular cases in medicine. In general, acute aortic dissections are a serious condition characterised by a tear in the aorta's intima allowing blood to enter the medial layer of the aorta therefore splitting the aortic wall into two layers, hindering blood flow and causing end-organ malperfusion (Hiratzka *et al.* , 2010; Czerny *et al.* , 2015). In 1994 von Segesser proposed the term the non-A non-B dissection for dissections in which an intima tear is localised beyond the ascending aorta (von Segesser *et al.* , 1994). In these forms, the dissection is limited to the aortic arch or can be described as a retrograde dissection arising from the descending aorta that extends into the arch and stops before the ascending aorta (Urbanski and Wagner, 2016).

In untreated acute type A aortic dissection, the rate of mortality within in the first 48 hours is greater than 50% and emergency open surgery is generally indicated (Gallo *et al.* , 2005; Erbel*et al.* , 2014). However, the progression of acute type B dissection is often uncomplicated and the generally accepted first line treatment for this consists of medical therapy (Erbel *et al.* , 2014; Hiratzka *et al.* , 2010; Riambau *et al.* , 2017). In complicated acute type B dissection however thoracic endovascular aortic repair (TEVAR) is the established treatment (Riambau *et al.* , 2017; Erbel *et al.* , 2014). Non-A non-B dissections exist in between these two entities and represents only a fraction of the established literature on aortic dissections with evidence for their optimum treatment and management thin.

2.0 Classification of Aortic Dissection

Several classifications of aortic dissection are classically used throughout the medical world, with the most significant of them being the DeBakey and Stanford classifications (Gawinecka, Schönrath and von Eckardstein, 2017). The DeBakey system consists of three types of dissection: Type I, II and III (Gawinecka, Schönrath and von Eckardstein, 2017). In Type I aortic dissection the tear arises in the ascending aorta and may include the arch and descending aorta. In Type II dissection the tear is confined to the ascending aorta whereas in Type III dissection the tear is limited to the descending aorta (DEBAKEY*et al.* , 1965). The Stanford classification system simplifies this and divides aortic dissection into two types: type A involving the ascending aorta and type B, involving the descending aorta distal to the left subclavian artery (Figure 1) (Rylski *et al.* , 2017; Gawinecka, Schönrath and von Eckardstein, 2017).

INSERT FIGURE 1

However, neither of these classifications address dissections involving the aortic arch alone or dissections comprising of the aortic arch and the descending aorta. When the dissection is limited to the aortic arch or can be described as a retrograde dissection arising from the descending aorta that extends into the arch and stops before the ascending aorta; these dissections are then termed as non-A non-B aortic dissections (Carino *et al.* , 2019) (Urbanski and Wagner, 2016). The Contemporary classifications such as the TEM (Type, Entry and Malperfusion) aortic dissection classification include non-A non-B dissections (Sievers *et al.* , 2020).

3.0 Epidemiology

According to several studies, the incidence of non-A non-B aortic dissection is lower than type A aortic dissection but is higher than type B dissection (Gawinecka, Schönrath and von Eckardstein, 2017; Lempel *et al.* , 2014). The frequency of non-A non-B dissection among all acute aortic dissection patients has been shown to vary from 3%-11% (Rylski *et al.* , 2017; Sievers *et al.* , 2020; Lempel *et al.* , 2014; Urbanski and Wagner, 2016) (Table 1).

INSERT TABLE 1

Studies by Sievers et al. as well as Lempel et al. showcased that non-A and non-B dissection patients tend to be younger and have a lower mortality compared to type A dissection patients (Lempel *et al.* , 2014; Sievers *et al.* , 2020). The median age for non-A non-B dissection patients was 59 years compared to 65 and 67 years for type A and type B dissection respectively (Sievers *et al.* , 2020). A study by Rylski et al. divided non-A non-B dissection into descending-entry and arch-entry types with similar frequencies recorded in both types (Rylski *et al.* , 2017). However, a prospective study by Urbanski et al. revealed a higher case load in descending-entry type patients. This study also showed that surgery improved the outcomes of these patients compared to a more conservative approach. A multicentre study using the International Registry of Acute Aortic Dissection also reported that over 16% of type B aortic dissection cases had extension of the dissection into the aortic arch (Nauta *et al.* , 2016).

4.0 Pathophysiology of Non-A Non-B dissection

The wall of the aorta comprises three layers, the tunica intima, tunica media which largely is constituted of structural proteins including elastin and collagen and adventitia (Levy *et al.* , 2020; Frederick and Woo, 2012). These layers form a thick aortic wall able to withstand high pulsatile pressure and shear stress.

Aortic Dissection is a condition characterised by the separation of these aortic layers. Classically it involves the breaching of the tunica intima, resulting in blood being diverted into a newly created channel within the medial layer of the aorta, known as the false lumen. A tear in the intimal layer tends to arise in locations where the rise in blood pressure is the greatest, commonly 2-2.5cm above the aortic root (Levy*et al.* , 2020). The separation of these layers paves the way for the formation of a false lumen. Increase in size of the false lumen can lead to an aortic rupture which has a high mortality rate or a second intimal tear which allows blood to re-enter the intima to form a double-barrelled aorta (Gawinecka, Schönrath and von Eckardstein, 2017). This tear can occur in any part of the aorta including the ascending, arch and descending aorta.

Other origins of an aortic dissection include an intramural haematoma and aortic ulceration. The former occurs due to the formation of a haematoma in the media, as a result of bleeding into the aortic wall from the vasa vasorum (Alomari *et al.* , 2014; Nienaber *et al.* , 2016). The latter, also referred to as a penetrating aortic ulcer and linked to atherosclerotic disease, is a penetration of the elastic lamina and can result in a haematoma forming in the tunica media (Hayashi *et al.* , 2000). Such haematomas can contribute to an aneurysm forming prior to aortic dissection.

The formation of an aortic aneurysm is thought to be more likely as a consequence of weakening of the tunica media, through the degeneration of collagen and elastin, which in turn increases higher wall stress. This is explained by Laplace's Law which states that 'wall stress is directly proportional to pressure (i.e. hypertension) and radius, and inversely proportional to vessel wall thickness (Patel and Arora, 2008). Compromising the integrity of the aortic wall raises the risk of an aortic dissection. This is key to the risk factors associated with the condition.

Should an aortic dissection progress, through the passage of blood further down the false lumen of the tunica media, it has potential to stretch past the aorta and into the major blood vessels, leading to ischaemia. Dissections can progress in an anterograde or retrograde fashion. This progression of the dissection can result in pressure differences which may lead to the compression or obstruction of the true lumen by the false lumen. Following this, fenestration may re-communicate the false with the true lumen or there is a risk the dissection may rupture into the surrounding cavities (Patel and Arora, 2008).

Carino et al. in their systematic review demonstrated in their analysis that 88% of non-A non-B aortic dissection patients had a complicated disease course and that 29% of these patients had signs of malperfusion; defined as a loss of blood supply to vital organs resulting in end-organ ischaemia (Carino *et al.* , 2019; Deeb, Patel and Williams, 2010). This percentage formed a considerably larger proportion than observed in type B aortic dissections (Pape *et al.* , 2015; Estrera *et al.* , 2007; Ziganshin, Dumfarth and Elefteriades, 2014).

5.0 Treatment

The challenges associated with the treatment of non-A non-B aortic dissections are well recognised and in amongst this surgical field there is currently no gold standard consensus on how to treat acute non-a non-b. Due to the infrequency of presentation of non-a non-b dissections, the literature is currently significantly reduced on this topic, however various studies from across the world have utilised many of these techniques with a differing spectrum of intra-operative and post-operative results published.

Various surgical approaches exist however endovascular treatment has been shown in a recent systematic review and meta-analysis to be the most widely used technique (Brown *et al.* , 2020) (Carino *et al.* , 2019). In their study, TEVAR made up a significant percentage of surgical treatments for Non-A non-B dissections; TEVAR with extrathoracic surgical transposition of the supra aortic branches was adopted in 18% of surgeries and TEVAR with chimney stent graft in 36% of cases (Carino *et al.* , 2019). A growing proportion of studies also list TEVAR as the treatment of choice for acute complicated and many chronic type B and Non-A non-B aortic dissections (Shrestha*et al.* , 2015; Brown *et al.* , 2020). Success of an endovascular approach for this subset of dissection patients is with some growing consensus attributed to the closure of the primary entry tear (Erbel *et al.* , 2014). Application of different thoracic endovascular aortic repair (TEVAR) zones is used to enable effective entry tear closure. For descending entry patients that have an entry tear distal to the left subclavian artery, TEVAR zone 3 (landing zone that is distal to left subclavian artery) and TEVAR zone 2 (landing zone that is between the left subclavian and left common carotid) for more proximal entry tears just at the edge of the left subclavian artery (Rylski *et al.* , 2017). Despite TEVARs more widespread use, an endovascular approach of this type might not always be feasible due to the lack of an adequate proximal landing zone as well as an increased risk of retrograde type A dissection which is particularly apparent in patients with additional aortic pathologies in zones 1 to 3 (Shrestha*et al.* , 2015). Application of TEVAR in patients with connective tissue diseases is also controversially reported in the literature (Czerny *et al.* , 2019b; Czerny *et al.* , 2019a; Kreibich*et al.* , 2018; Shrestha *et al.* , 2015). Therefore, in a scenario in which an entry tear is located in the aortic arch, a more extensive arch repair such as a hybrid aortic repair involving rerouting of the supra-aortic arteries with TEVAR zone 0 (landing zone at the ascending aorta) or even a complete arch replacement utilising the frozen elephant technique (FET), could be necessary in order to close the primary entry tear (Czerny *et al.* , 2019b).

5.1 TEVAR

5.2 TEVAR with chimney stent graft

This surgical approach has been shown to be reliable in cases where a suitable proximal landing zone can be established, where a simple left carotid to left subclavian bypass or one chimney stent graft for the left subclavian in sufficient (Carino *et al.* , 2019). When the aortic pathology involves or is in close proximity to the aortic branches, it is paramount that the endografts must cross their ostia in order to produce an adequate seal (Malina, Resch and Sonesson, 2008). In this scenario, a standard angioplasty/stenting technique otherwise known as a chimney graft can make performing a TEVAR procedure possible using off the shelf devices (Greenberg *et al.* , 2003; Donas *et al.* , 2010). This technique, first reported by Greenberg et al. and subsequently described in detail by various other vascular groups, is a means of gaining additional fixation length in order to stabilise the aortic stent grafts whilst also safeguarding perfusion to the vital branches (Greenberg *et al.* , 2003; Baldwin *et al.* , 2008; Donas *et al.* , 2010; Criado, 2007). However, the addition of a chimney stent graft has been shown to increase the risk of type IA endoleak (Ahmad *et al.* , 2017).

A 2016 study by Huang et al. describes 27 consecutive non-A non-B patient outcomes without adequate proximal landing zones that were treated with the chimney stent graft endovascular technique with a mean follow up time of 17.6 months (Huang *et al.* , 2016). Chimney stents were deployed parallel to the main endografts in order to preserve blood flow while extending the landing zones. The technical success rate published was 100% with endografts deployed in zone 0 (3), zone 1 (18) and zone 2 (6). Proximal endoleaks were reported in 5 patients immediately after surgery and were treated with kissing balloon technique in order to minimise gutter formation. Computed tomography angiography showed all aortic stent and chimney stent grafts to be patent post-surgery. Huang et al. also report a 0% 30-day mortality rate and a 0% retrograde type A dissection, however 2 out of the 27 patients were reported to have suffered a stroke post operatively (Huang*et al.* , 2016).

Zhu et al. has also previously reported similar results in terms of success and also the incidence of endoleak whilst utilising this technique in a cohort of 34 patients (Zhu *et al.* , 2013). The technical success was reported at 82% and immediate type I endoleaks were reported in 5 patients all of which underwent bare chimney stent techniques (Zhu *et al.* , 2013). No perioperative death or strokes were observed however one perioperative morbidity included an ST-elevation myocardial infarction. The mean follow-up for this study was 16.3 months and primary patency was maintained in all the chimney stents as well as in the surgical bypasses across this period with no incidence of stent fracture or chimney-related endoleak observed in addition. The authors conclude that this technique provides a minimally invasive way of the preservation of arch branch blood flow with favourable mid-term outcomes. However, the study also concluded that the application of the bare chimney stents seemed to be associated with a higher incidence of immediate type I endoleaks and suggests that balloon-expandable stents should be regarded as the first choice because of their greater radial strength (Zhu *et al.* , 2013).

An earlier study by Shu et al. reported outcomes of the chimney stent-graft technique on 8 patients treated for Non-a non-b aortic dissections with no adequate proximal sealing zones (Shu *et al.* , 2011). Covered stents were placed parallel to the aortic stent grafts in order to restore flow to the left common carotid artery while extending the proximal fixation zones; the left subclavian arteries were also intentionally covered after cerebrovascular assessment. All 8 procedures were completed successfully with one main aortic stent graft deployed alongside one chimney graft implanted in the left common carotid artery. The authors report two retrograde type II endoleaks that were identified perioperatively but were left untreated but followed closely using computed tomography (Shu *et al.* , 2011). They also report no instances of any puncture site complication, strokes, death or paralysis during the hospital stay and a 30-day mortality of 0%. Mean follow-up was 11.4 months and during this time there was no mortality with duplex ultrasound and computed tomography displaying patency of stent grafts, enlargement of the true lumen and compression of the false lumen (Shu*et al.* , 2011). One of the type II endoleaks disappeared in two weeks post-operatively while the other faded gradually until almost disappearance at 11 months post-operatively.

A 2015 study by Liu et al. reported outcomes of 41 consecutive patients treated with the chimney stent graft technique for Non-a non-b aortic dissection including 8 emergent repairs (Liu *et al.* , 2015). This technique was utilised to reconstruct the left subclavian artery in 5 patients and the left common carotid artery in 34 patients. In 2 cases the double chimney technique was used in order to simultaneously reconstruct the innominate artery and the left common carotid artery. The mean follow-up period for this cohort was 17.3±6.1 months and the authors reported a 0% 30-day mortality rate (Liu *et al.* , 2015). None of the patients were reported to have a type I endoleak however four had type II endoleak. During the follow-up no patients were reported to have suffered severe neurological complications, migration or occlusion of any stent grafts. Similar results including 0% 30-day mortality, 0% stroke and 0% retrograde type A dissection were reported in a study by Zou et al. while using the chimney stent graft technique in a Non-A non-B dissection cohort (Zou *et al.* , 2016).

5.3 TEVAR with extrathoracic surgical transposition of the supra-aortic branches

A 2017 study by Rylski et al. reported surgical outcomes on 43 non-A non-B aortic dissection patients repaired using TEVAR with extrathoracic surgical transposition of the supra-aortic branches (Rylski *et al.* , 2017). In this study endovascular treatment involved TEVAR with or without carotid-subclavian bypass or transposition of both left carotid and left subclavian artery as well as isolated stenting of dissected visceral vessels (Rylski *et al.* , 2017). The authors classified Non-A non-B dissections as descending entry type with entry distal to the left subclavian artery and dissection extending into the aortic arch, and arch entry type with entry between the innominate and left subclavian arteries. These two groups were then compared in terms of presentation, treatment and outcomes with 21 patients forming this descending entry group and 22 the arch entry cohort (Rylski *et al.* , 2017). The cardiovascular risk profiles of these groups did not differ and the overwhelmingly majority of aortic segments were not dilated in patients from both groups. Across both groups the 30 day mortality rate was 9%, one patient suffered a stroke and two patients suffered a retrograde type A dissection (Rylski *et al.* , 2017). Aortic repair due to new organ malperfusion, rapid aortic growth or persisting pain was performed in 43% of descending entry patients and 36% arch entry patients with a 0% in hospital mortality.

An earlier study by Lu et al. retrospectively analysed 22 consecutive patients treated with extrathoracic surgical transposition of the supra aortic branches for Non-A non-B dissection (Lu *et al.* , 2011). Hybrid, scalloped or fenestrated endovascular stent grafts were selected based on dissection characteristics and median follow up time was 27.1 months with patients assessed with computed tomography angiography. Primary end points of the study included pathology, complications and survival rates (Lu *et al.* , 2011). The authors reported surgery was successful in all patients except one with an operative complication and they report a 30-day mortality rate of 9% (Lu *et al.* , 2011). Thrombosis had formed in the aortic false lumen of the graft exclusion segment in all patients however the maximum diameter of this segment was shown to be decreased in 18 patients and stable in two (Lu *et al.* , 2011). Patency was observed at both mid- and long-term follow-up and no proximal endoleak, graft displacement or deaths were reported in this period.

5.3 Hybrid aortic repair

When the entry tear occurs in the proximal aortic arch more invasive solutions can be necessary which include hybrid repair that involves rerouting of the arch branches and zone 0 TEVAR or arch replacement. A recent systematic review and meta-analysis by Carino et al. demonstrated that these hybrid techniques have been utilised for Non-A non-B dissections in 21% of total surgical cases (Carino *et al.* , 2019). Zone 0 TEVAR use in hybrid procedures for acute dissection has been shown to be considered dangerous with a significantly higher risk of retrograde dissection in comparison to a more distal landing zone (Cao *et al.* , 2012; Canaud *et al.* , 2014; Czerny *et al.* , 2012).

A 2020 study by Wang et al. describe the outcomes of 28 patients with non-A non-B dissection who underwent a novel hybrid surgery (Wang*et al.* , 2020). The novel hybrid surgery, also termed the 'inclusion aortic arch technique' involves initially a transverse incision of the aortic arch wall and bilateral antegrade cerebral perfusion was accomplished after cannulating the left common carotid artery (Liu *et al.* , 2020). Then the intimal tear of the dissection was identified and sealed using mattress sutures of 4-0 Prolene. An appropriate stent graft was subsequently introduced into the descending aorta and after stent graft released where its proximal edge was just distal to the left subclavian artery ostium with double check, the vascular graft was trimmed into an elliptical shape around the left common carotid and left subclavian artery orifices. Following this, pledgetted stitches were placed at the left subclavian artery orifice's lower margin with a 4-0 polypropylene double armed suture needle therefore immobilising the vascular graft and aortic arch tissue. A single suture was then used to stitch the vascular graft to the anterior aortic arch wall from outside to inside the aortic arch and then reverse the direction through the aortic arch wall and vascular graft layers. Another suture is then added to accomplish the continuous suture by the anastomosis of the posterior aortic arch wall and vascular graft as deep as possible. Following this the trimmed vascular graft was attached firmly to the aortic wall. The initial transverse aortic arch wall incision was then closed with 4-0 Prolene sutures. Arterial cannulation blood was then used for de-airing through the aortic arch incision before the last sutures, antegrade systemic perfusion was resumed and the patient rewarmed (Wang *et al.* , 2020).

All patients in this cohort of 28 non-A non-B dissection patients underwent an emergency operation (Wang *et al.* , 2020). The authors reported no early adverse event such as in-hospital mortalities, re-explorations for haemorrhage, paraplegia, stroke, endoleak or left subclavian artery occlusions (Wang *et al.* , 2020). Mean follow-up time was 39.12±15.04 months, however one patient was lost during this time and another died suddenly due to false lumen patency in the aortic arch and descending aorta without any symptoms (Wang *et al.* , 2020). At 6 months the computer tomography angiography showed significantly smaller distal aortic arch diameters and descending aorta diameters than were measured pre-operatively (Wang *et al.* , 2020). No incidences of paraplegia, cerebral infarction, upper limb ischemia or left subclavian artery ischemia events were reported during the follow up period. The authors concluded that their inclusion aortic arch technique is both safe, effective and simple treatment for non-A non-B dissections which avoids endoleak, requires no blood products and demonstrates satisfactory early outcomes (Wang *et al.* , 2020).

A 2014 study by Kefeng et al. describes the use of a hybrid procedure for 15 patients with non-A non-B aortic dissections (10 acute, 5 chronic) (Kefeng *et al.* , 2014). The hybrid procedure performed in these patients comprised of 7 patients with zone 1 inclusion and 8 patients with zone 2. The authors report a technical success rate of 100% and no incidences of paraplegia were reported (Kefeng *et al.* , 2014). 30-day mortality and incident of stroke were 0%. However, during the follow-up period (median follow-up of 12 months) a stroke and death occurred in one patient who was not associated with an endograft complication. During this follow-up period, overall mortality was 6.7% and the overall late endoleak rate was 7.7% however no retrograde dissection occurred across the cohort (Kefeng *et al.* , 2014). The authors also report no differences in outcome between acute and chronic dissection or proximal landing zones except for proximal endograft dimension (Kefeng *et al.* , 2014).

An earlier paper published by Bünger et al. reported outcomes of 75 consecutive patients of which a subgroup of 45 patients underwent hybrid aortic repair for non-A non-B aortic dissection (Bünger *et al.* , 2013). Complete supra-aortic debranching was performed on 6 patients in zone 0, and partial debranching in 39 patients (16 in zone 1 and 23 in zone 2). Technical success was reported at 86.7% and the 30-day mortality rate at 4.4%. The in-hospital mortality was 11.1% following the deaths of 3 patients after days 33, 35, and 111 (Bünger *et al.* , 2013). After a median follow up of 20.8 months, the overall mortality reported was 13.3% (Bünger *et al.* , 2013). Additionally, the stroke rate recorded was 8.8% and paraplegia developed in one patient with complete recovery following a spinal drainage. Retrograde dissection also occurred in one patient 14 days after complete debranching and zone 0 TEVAR with a fatal outcome. The authors report the overall early and late endoleak occurrence rates were 27% and 43% respectively (Bünger *et al.* , 2013). Reintervention was required in 8 patients and freedom from reintervention was reported at 91% at 1 year and 81% at 2 years (Bünger *et al.* , 2013). Bünger et al. concluded that hybrid repair in zones 1 and 2 proved a viable alternative to conventional aortic arch surgery in these patients despite persistent issues with stroke and endoleak rate. Treatment of non-A non-B dissection patients with supra-aortic debranching and TEVAR in zone 0 however is associated with high mortality (Bünger *et al.* , 2013).

5.4 Frozen elephant trunk

Another surgical treatment utilised in the surgical management of non-A non-B aortic dissection with proximal entry tear is the frozen elephant trunk technique (FET). This procedure involves ascending aorta and arch replacement in combination with antegrade stent graft implantation in the descending thoracic aorta while using a single hybrid prosthesis (Shrestha *et al.* , 2015). A recent systematic review and meta-analysis by Carino et al. found that 7% of patients were treated with this technique and that FET may also be an important option in cases of malperfusion syndrome as it can potentially open the compressed true lumen and cover any additional entry tears that could be positioned in the proximal descending aorta so that pressurisation the false lumen is maintained (Di Bartolomeo *et al.* , 2017; Shrestha *et al.* , 2015; Carino *et al.* , 2019). FET is also recognised in the literature to promote favourable remodelling in the distal aorta (Dohle*et al.* , 2016; Iafrancesco *et al.* , 2017). Another advantage of FET is no type I endoleak as well as its ability to establish a highly stable proximal landing zone for the eventual stent graft implantation in the descending thoracic aorta (Berger *et al.* , 2019; Berger *et al.* , 2018; Dohle *et al.* , 2016). However, the main limitation associated with FET is the increased surgical trauma secondary to the necessarily prolonged periods of extracorporeal circulation, circulatory arrest as well as myocardial ischemia (Carino*et al.* , 2019). In addition, the technical demand of this procedure requires experienced surgeons in high volume aortic centres (Shrestha *et al.* , 2015).

A 2020 study by Kreibich et al. reports outcomes of the FET technique to treat 41 patients presenting with acute complicated or chronic type B or non-A non-B aortic dissection (Kreibich *et al.* , 2020). FET was implemented when supra-aortic vessel transposition would not suffice to create a satisfactory proximal landing zone for TEVAR, when a concomitant ascending or arch aneurysm was present or if any patients suffered with any connective tissue disorders. Of these 41 patients, 23 presented with a non-A non-B dissection (Kreibich *et al.* , 2020). In the 41-patient overall cohort one patient was reported to have died intra-operatively secondary to an aortic rupture in downstream aortic segments however no other post-operative deaths occurred. 4 patients suffered a non-disabling stroke post-operatively and were subsequently discharged with no clinical symptoms (1 patient), no significant disability (2 patients) or with slight disability (1 patient) (Kreibich*et al.* , 2020). The authors report one patient death during follow-up after two years (not aorta related) and 16 patients subsequently underwent an aortic re-intervention after 7.7 months (Kreibich *et al.* , 2020). Kreibich et al. concluded that FET is an effective treatment option for acute complicated and chronic type B as well as non-A non-B aortic dissection patients in whom primary endovascular was not deemed as feasible. They also conclude that this study underlines the considerable need for aortic re-interventions and the importance of continuous follow-up of patient after undergoing FET procedures (Kreibich *et al.* , 2020).

An earlier study by Zhao et al. assessed 24 consecutive patients with non-A non-B aortic dissection treated with the FET technique (Zhao*et al.* , 2012). This cohort also included concomitant procedures including the Bentall procedure in 3 patients, David procedure in 1 patient and ascending aortic replacement in 7 patients (Zhao *et al.* , 2012). The in-hospital mortality rate was recorded at 4.1% with one patient dying of multi-organ failure after surgery (Zhao *et al.* , 2012). No incidences of paraplegia were reported (Zhao *et al.* , 2012). During follow-up one patient was reported to have died following gastrointestinal bleeding 2 months after surgery and type II endoleak occurred in 1 patient. The 5-year survival rate was 91.7% (Zhao *et al.* , 2012). The authors concluded that the application of this technique was safe and feasible for non-A non-B aortic dissection with a low rate of mortality and morbidity as well as a satisfactory 5-year survival rate (Zhao *et al.* , 2012).

A 2016 study by Urbanski et al. surgically treated 8 patients with a non-A non-B aortic dissection, with 4 patients treated surgically and 4 patients conservatively (Urbanski and Wagner, 2016).

Amongst the surgically treated patients, 1 patient underwent a partial arch replacement, and the remaining patients received a complete arch replacement via a modified elephant trunk technique (Urbanski and Wagner, 2016; Urbanski *et al.* , 2010). In the surgically treated cohort, there were no reported deaths or relevant clinical events recorded during a median follow-up time of 40 months (Urbanski and Wagner, 2016). In the conservatively treated patient group, 3 patients had died by 28 months of follow-up, 1 from an aortic rupture and 2 due to the progression of the dissection and subsequent malperfusion (Urbanski and Wagner, 2016). The authors concluded that surgical treatment of acute aortic dissection involving the arch but sparing the ascending aorta seemed to offer improved clinical outcomes (Urbanski and Wagner, 2016).

5.5 Medical replacement

When considering a suspect aortic dissection, it is important to provide haemodynamic support. While this resuscitation protocol may vary locally, medications such as noradrenaline and dobutamine, often administered intravenously, have been indicated in the treatment of aortic dissections (Feldman, Shah and Elefteriades, 2009). Carino et al. includes studies focusing on medical treatment of non-A non-B dissections which show admission to intensive care units for monitoring of blood pressure, analgesia and antihypertensive therapy to be indicated as a method of medical treatment (Carino *et al.* , 2019). Their analysis showed that the proportion of medically treated patients ranged from 5 to 54% of patients (Carino *et al.* , 2019). The 30-day mortality rate for these patients was recorded as 14% in comparison to 3.6% in patients who had undergone intervention (Carino*et al.* , 2019).

6.0 Conclusions

Non-A non-B aortic dissections represent a small proportion of the total number of dissections that occur annually, and this is reflected in the quantity of studies published in the literature. By definition non-A non-B dissections fall outside of the standardised classifications for aortic dissections despite the literature showing that a significant majority of these patients have a complicated disease course which has been shown to occur more frequently than other types of aortic dissection. Studies from around the world have demonstrated that surgical intervention when compared to medical management produce far more satisfactory 30-day mortality rates, with both open surgical and endovascular techniques providing good short-term results. Despite these published findings, further large-scale prospective studies are required in order to improve our understanding of best treatment and to provide more substantial evidence for the application of different surgical techniques for this subset of aortic dissections.

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