Perspective

Strategies in the surgical treatment of type A aortic arch dissection

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Introduction

Aortic dissection involving the ascending aorta is a lethal condition, with mortality approaching 60% if surgical intervention is not performed early (1).

Stanford type A acute aortic dissection involves the ascending aorta, aortic arch and a variable extent of the descending thoracic aorta. Identification and resection of the primary intimal tear with re-approximation of the intima and adventitia remains the surgical principle of repair. Despite using this approach, a residual dissection flap persists in the arch and descending thoracic aorta in 64-90% of patients (2-5). This can lead to distal malperfusion in the acute setting (6), with a risk of aneurysm formation and rupture over the long term (7).

Despite the advancement of cardiac surgery techniques, employment of modern cerebral protection strategies, myocardial protection, and aortic graft development, the surgical mortality of these operations, even in centers with a specific interest in aortic surgery, is in the range of 25% (8). In the modern era of cardiac surgery, it is rare to undertake a "common" operation with a mortality of 25% and this serves as a challenge for the cardiac surgery community to improve. In order to reduce this high operative mortality, one first needs to understand the contributing factors.

Drivers of early mortality after surgical repair of type A dissection

Operative mortality for acute type A dissection is multifactorial, yet poorly elucidated. Review of our own database showed that the major classes of operative mortality are related to hemorrhage, cerebral ischemia, cardiogenic shock, and distal malperfusion. Perrault (2012, unpublished data) found cardiogenic shock, cerebral ischemia and massive hemorrhage to be responsible for almost 85% of perioperative deaths.

Visceral and renal malperfusion are often under-recognized at the time of presentation, with the emphasis placed on reconstructing the threatened proximal thoracic aorta and the associated valvular insufficiency. Distal malperfusion may manifest as ileofemoral, mesenteric, renal, cerebral or spinal cord in nature. The resulting lower limb ischemia, gut ischemia, renal failure and stroke significantly increases mortality.

It has been reported that as many as 20-30% of patients presenting with type A dissection may have one or more malperfusion syndromes (6,9). According to the data of the Pacini group (6), the overall mortality in type A dissection was almost 3 times higher in the group with malperfusion (43.7%) than with no malperfusion (15%).

This association of malperfusion and increased mortality is well documented (9-12). A recent prospective study on the use of hybrid operating rooms (OR) for the management of acute dissection found that a hybrid OR enabled the identification of downstream malperfusion sites (13). In this study, 23% of patients with acute type A dissection required primary endovascular intervention and 35% required descending aortic repair in addition to ascending aortic replacement. Visceral malperfusion, however, is challenging to diagnose in the acute and immediate post-operative setting and is most likely under-appreciated. Under-recognition of early visceral and renal ischemia may lead to mortality traditionally attributed to "multisystem organ failure".

Once we better understand the drivers of early mortality,

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therapies can be guided to the individual problems. Massive hemorrhage, cerebral ischemia and visceral/peripheral malperfusion may be areas that can be targeted by newer therapies.

Drivers of late mortality after surgical repair of type A dissection

Long-term survival and freedom from dissection related events are in the range of 50% at 10 years (14) after type A repair. The rate of re-intervention in this patient group is in the range of 18-22% (15-18). Recently, there have been several provocative studies identifying variables in the sub-acute phase that may portend a poor long-term outcome. These variables include:

(I) Size of the descending thoracic aorta at initial presentation: Sakaguchi (19) has found that significant medically treated pre-operative hypertension as well as a descending aortic diameter greater than 35 mm are risk factors for a patent false-lumen (a marker of aortic dilatation) postoperatively.

(II) False-lumen patency: patency of the false lumen has been implicated as a risk factor in the fate of the residual aorta (20,21). This may result from incomplete resection of the primary intimal tear or more likely secondary re-entry sites downstream in the arch and aorta. Some evidence suggests that a partially thrombosed false lumen may in fact be a worse prognostic factor than a completely patent false lumen (22).

(III) False lumen diameter: Song *et al*. (7) found that an initial false-lumen maximal diameter ≥22 mm in the proximal descending aorta was associated with accelerated aortic dilation and catastrophic clinical events.

(IV) Primary and residual intimal tear location and size: in a recent, thoughtful study, Evangelista *et al*. identified a large proximal intimal tear (>10 mm) as a significant risk factor for dissection related events and mortality (14).

Traditional surgical intervention

The current best practice guidelines for type A dissection repair involve ascending aortic replacement and open distal anastomosis (hemiarch) under circulatory arrest with varying degrees of hypothermia and selection of cerebral protection techniques including antegrade or retrograde cerebral perfusion, or deep hypothermia alone. Although short periods of circulatory arrest are very well tolerated (23), the prolonged periods of circulatory arrest that are occasionally required to reconstruct a dissected arch may account for the high incidence of neurologic complications following this operation. Cerebral ischemia may be present pre-operatively or can be a complication of the operation itself. As previously noted, cerebral ischemia is one of the drivers of this early mortality.

Another hypothetical cause of high mortality with traditional surgery is that the primary intimal tear is not always addressed. Patients who undergo sternotomy for repair are occasionally discovered to have a primary intimal tear in the arch with a flap that extends both retrograde and antegrade. In these cases, the option of emergency total arch replacement may be viewed as prohibitively risky and the primary intimal tear is not resected. Careful interpretation of the pre-operative CT, however, can identify the culprit primary intimal tear in 40-100% of patients (24,25).

More aggressive strategies to address the arch at the time of type A repair

For years, several reports from Japan have advocated a formal total arch and elephant trunk repair in acute dissection to mitigate the long-term complications of the residual dissection in the arch and more distal aorta (26,27).

Over the last decade, various strategies have been employed to decrease the risk of total arch replacement. Open total arch replacement and ascending aortic replacement with a "frozen" elephant trunk technique have been developed and advocated (28). Pochettino and colleagues (29) have advocated the placement of an antegrade stent-graft into the proximal descending aorta during open type A dissection repair.

This hybrid approach, of conventional arch replacement with downstream placement of a stent-graft through an open arch, has stimulated the development of novel grafts such as the E-vita prosthesis, which is composed of a Dacron segment for replacement of the ascending aorta and arch, together with a stent graft segment, which is positioned in the proximal descending thoracic aorta. The group at Essen (30) has developed the International E-vita open registry with promising early results.

Although these approaches have been shown to promote false lumen thrombosis and may decrease long-term complications, they require systemic hypothermia and prolonged periods of circulatory arrest to perform the conventional total arch replacement. While early results

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indicate the techniques to be "safe" when compared to conventional surgery for acute aortic dissection, operative morbidity and mortality remain substantial.

Novel and innovative hybrid strategies to address the arch at the time of type A repair

Several groups have recently published novel single-stage hybrid techniques, involving ascending aortic replacement, arch de-branching and antegrade endovascular stent deployment (31-34). Based on a skill-set derived from our experience with elective thoracic aortomegaly patients (35), we have recently been performing surgery for acute type A dissection without circulatory arrest and consequently using mild hypothermia (32) (*Figures 1-4*). Using a very similar strategy, Chang *et al*. operated on 21 consecutive type A dissection patients with one hospital mortality (34).

Marullo and colleagues (33) have employed an interesting staged approach whereby the ascending aorta is replaced and the arch is de-branched. An endograft is then deployed in the arch and proximal descending thoracic aorta if clinically indicated by the presence of a malperfusion syndrome or early evidence of distal aortic dilation. Using this strategy they deployed an endograft in 62.5% of patients, resulting in false lumen thrombosis and no resultant endoleaks.

Figure 5 shows an intra-operative aortogram of a 54-year old male who presented with an arch dissection starting at the left carotid artery. The pre-operative CT revealed the common finding of a severely effaced true lumen in the distal descending and thoraco-abdominal aorta. The clinical significance of this finding of an "effaced true lumen" remains unclear. After undergoing ascending aortic replacement and arch de-branching, and before thoracic endovascular aneurysm repair (TEVAR) placement, an intra-operative angiogram was done showing severely compromised flow to visceral vessels and kidneys (*Figure 5A*). Placement of an endovascular device across the arch improved flow to the abdominal aorta and visceral organs (*Figure 5B*). Traditional hemiarch replacement would have left the patient with compromised flow to the viscera.

There are no randomized studies comparing "extended arch" strategies to conventional "open distal anastomosis" technique for acute type A dissection surgery. However, potential advantages of a hybrid repair technique include:

- Absence of deep hypothermia and associated coagulopathy/hemorrhage
- Arch reconstruction is made into an easier, more "proximal" operation, thus potentially decreasing hemorrhagic complications
- Complete resection/exclusion of primary intimal tear
- Improved recognition and treatment of acute malperfusion
- Decreased cerebral ischemia by avoiding prolonged circulatory arrest
- Elimination of residual tears in the arch which are a risk factor for aortic complications
- Provides a suitable proximal landing zone if future distal endovascular treatment is required

Future strategies to address the arch at time of type A repair

The development of endovascular techniques has allowed surgeons to pursue novel strategies in an effort to further reduce the risk associated with surgery for acute type A dissection, and to address the residual type B dissection.

As experience with deploying stents in the proximal aorta increases, isolated endovascular repair of type A dissection may become a realistic possibility. Two recent studies evaluated pre-operative CT findings and found that up to 30-50% of patients may be candidates for an isolated endovascular approach (24,25). Anecdotal case reports of isolated TEVAR for acute type A dissection have been published (36), leading to the possibility of isolated ascending aortic TEVAR becoming a viable treatment option for patients previously considered too high risk for surgery.

Availability of off-the-shelf branched endovascular grafts and a hybrid suite may further advance the management of acute aortic dissection. In the future, we envision a strategy of complete endovascular repair in a proportion of cases. Whether this can improve short- and long-term results will need to be determined. We propose that a second, more likely option might involve open ascending aortic replacement with Dacron graft, and formal surgical attention to valve reconstruction, coronary arteries and pericardial effusion followed by branched endovascular repair of the arch. Timing of endovascular deployment could be immediate at the time of ascending aortic replacement or delayed based on a more refined understanding of risk factors for early and late complications. This type of strategy would simplify the current hybrid operations by removing the need to further de-branch the arch. Strategies such as these emphasize the need for cardiac surgeons to be trained in endovascular

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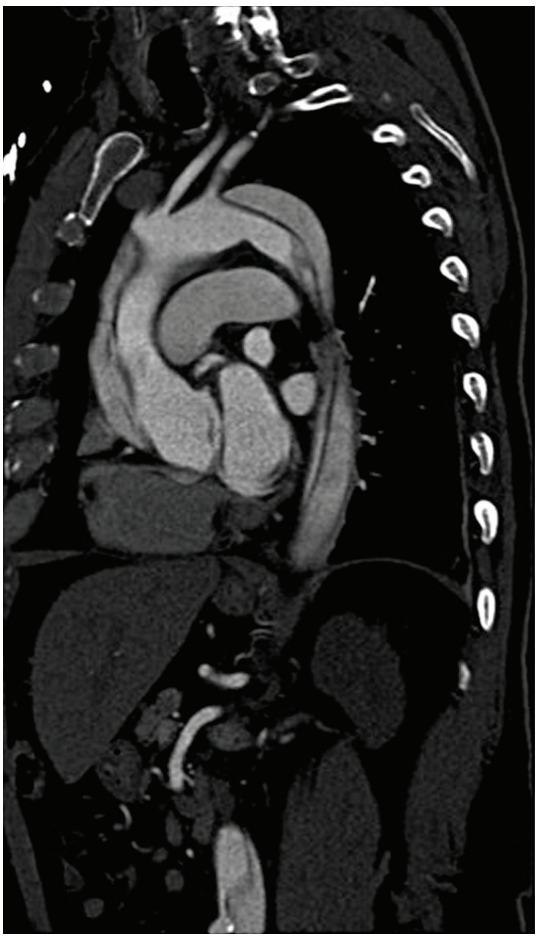


Figure 1 A preoperative CT angiogram demonstrating a type A dissection. The primary intimal tear was identified in the arch

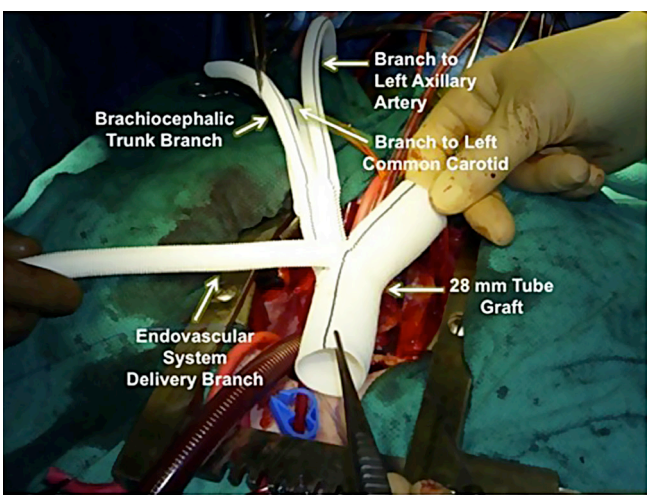


Figure 2 A Bavaria Graft with 4 side branches is used for replacement of ascending aorta and aortic arch debranching

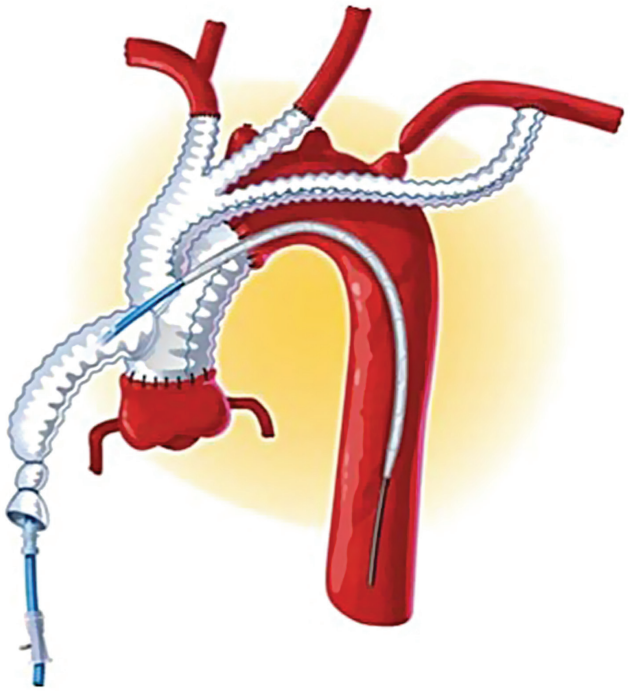


Figure 3 A schematic demonstrating an endovascular prosthesis being deployed in an antegrade manner to promote thrombosis of the false lumen in the arch and proximal descending aorta

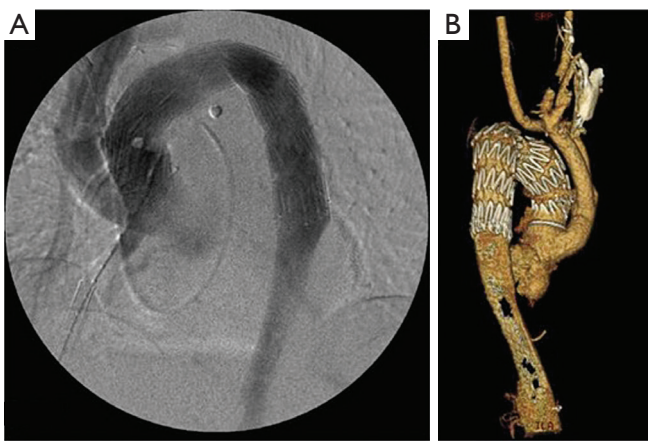


Figure 4 A. Intra-operative angiogram after stent-graft deployment; B. Post-operative CT angiographic reconstruction showing completed hybrid arch repair

aortic surgery and help define future treatment algorithms. At our center, all thoracic endovascular cases are done by a collaborative team consisting of interventional radiology, cardiac surgery, and cardiac anesthesiology. We have found benefits to having more than one perspective in the

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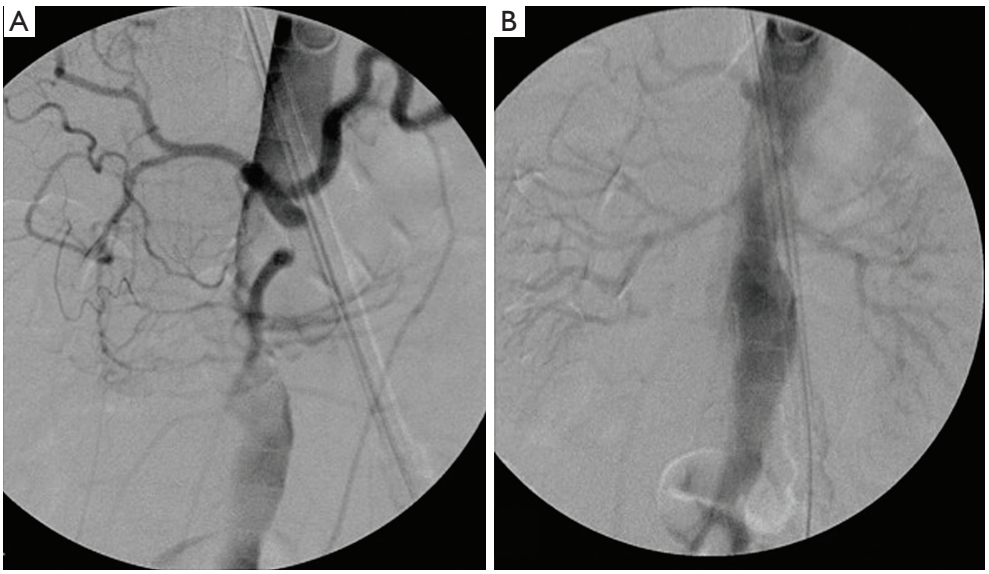


Figure 5 Visceral ischemia with type A dissection. A. Abdominal angiogram after ascending aortic replacement and prior to stent deployment. There is pseudocoarctation of the perivisceral aorta and lack of filling of renal parenchyma; B. Abdominal angiogram after arch stent deployment. There is marked improvement of filling of true lumen and renal parenchyma

treatment of these patients.

Potential indications for arch replacement in acute type A dissection

The nuances of aortic dissection and risk factors for acute and long-term complications will continue to evolve. Based on the current understanding of aortic dissection, the following might be conditions where we would consider arch replacement at time of type A surgery:

- Pre-existing arch aneurysm
- Primary intimal tear identified on pre-operative CT in the distal arch or descending thoracic aorta
- Secondary intimal tear in the arch measuring >10 mm
- Clinical signs of visceral or peripheral extremity malperfusion
- Radiologic signs of potential visceral, renal and peripheral compromise such as a severely effaced true lumen in descending thoracic aorta
- False lumen diameter > than 22 mm (20)
- Descending thoracic aorta diameter >35 mm

It is also worth questioning whether young age should be an indication for a more aggressive approach to the arch. On one hand, these patients have a more aggressive disease process and are at risk for complications over the years, while on the other hand, the long-term outcome of an endovascular prosthesis in the angulated arch has not been delineated. Review of our local database revealed one late complication in 15 patients with Zone 0 TEVAR at up to 47 months of follow up, with mean radiological follow up of 545 days and cumulative follow up of 8,176 days (*in submission*).

Conclusions

Strides made in understanding causes of early and late mortality following conventional type A repair will serve to reduce the morbidity and mortality associated with this lethal condition. Continued substantial advancement in outcomes will be made when risk factors for operative mortality and long-term complications are further elucidated, understood and validated.

A detailed and precise analysis of pre-operative CT will become the cornerstone in the assessment of these risk factors. Specifically, the determination of the site and size of the primary and secondary intimal tears, as well as characterization of distal perfusion and the size of both true and false lumens, will be critical in pre-operative planning.

A more nuanced approach to the aortic arch may be justified in those patients that are at risk for complications, with the axiom that any adjunctive procedure would substantially reduce mortality. However, much yet needs to be discerned regarding the optimal techniques to address the arch, methods of cerebral protection and perfusion,

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and the types of de-branching and endovascular procedures chosen.

Theoretically, aortic arch replacement at the time of type A dissection repair has the advantage of reducing early (malperfusion and addressing arch tears) and late (aneurysm expansion) morbidity and mortality. There is no unequivocal evidence to support this assertion and further research is required to better understand which patients would benefit.

A hybrid arch repair approach has the potential to reduce operative morbidity and mortality as compared to a formal surgical total arch replacement technique by moving suture lines more proximally, and avoiding prolonged circulatory arrest and profound hypothermia. These hybrid techniques and future endovascular and branched endovascular techniques need to be studied for their potential impact on short- and long-term mortality rates.

We believe that the arch should very likely be addressed at the time of type A surgery, with the caveat that the aortic surgery community is able to identify which patients are at risk for both poor short- and long-term outcomes and if the adjunct procedure can be performed without increased risk and preferably a reduction in mortality. In early 2013, the answers to the above questions are not completely understood. Thus, the arch should only be replaced at time of type A dissection in certain centers with experience in aortic surgery, interest in verifying the known risk factors, curiosity to assess new prognostic indicators and an ability to academically report their findings.

Our continued mandate should be to further elucidate which patients will benefit from arch replacement and assess these results in the context of the various techniques available to us as they evolve, with the ultimate goal of decreasing short- and long-term mortality rates in those with type A dissection.

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