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Aortic Dissection: Incidence, Natural History and Impact of Surgery

J. Auer, R. Berent, B. Eber

Acute ascending aortic dissection is included in the differential diagnosis of patients with acute chest pain. Recent literature discussing incidence and natural history of aortic dissection are reviewed. With respect to diseases of the aorta, dissection is considered as the most common disaster and is two to three times as frequent as a rupture of the infrarenal aorta. The most recent series of patients with untreated aortic dissection involving proximal and distal aorta revealed a rate of 50 % of patients dying within 48 hours, 84 % at one month and 90 % at three months.

History of surgical interventions, current surgical techniques and impact of surgery are discussed. Today, the two well-known surgical procedures for complete replacement of ascending aorta are the Bentall and the button technique. The third most commonly used technique, is the Cabrol method. The use of biological glue has greatly aided the performance of blood-tight-anastomosis on the aorta, particularly with acute dissection.

Stanford type A dissection is a surgical emergency that requires urgent and expeditious diagnosis and immediate surgical intervention. Mural haematoma, which has a similar natural history, also requires prompt surgical treatment. In contrast, Stanford type B dissection should be treated medically (nonsurgical), with surgery considered only when complications develop or conservative therapy fails. *J Clin Basic Cardiol 2000; 3: 151–4.*

Key words: aortic dissection, incidence, natural history, surgery, Stanford type

With respect to diseases of the aorta, dissection is considered as the most common serious complication and is more than two times as frequent as a rupture of the infrarenal aorta. Diagnosis of this disease has important prognostic implication. The prevalence of aortic dissection is less than 1 % in large series of autopsies. The most recent series of patients with untreated aortic dissection involving proximal and distal aorta revealed a rate of less than 10 % still alive at one year and nearly all patients died within ten years. Because of unfavourable prognosis of untreated aortic dissection aggressive medical therapy and surgery are recruited to improve survival rates. Stanford type A dissection is a surgical emergency that requires urgent and expeditious diagnosis and immediate surgical intervention. The best technique for surgical treatment of aortic dissection has to be determined by each surgeon, according to his own experience. Today the two well-known surgical procedures for complete replacement of ascending aorta are the Bentall and the button technique. The third most commonly used technique is the Cabrol method. The elephant trunk procedure is an alternative form of composite valve graft replacement of the proximal aorta. Glue aortoplasty is a useful technique but cannot be considered superior to the conventional models of dealing with proximal or distal aortic dissection.

Stanford type B dissection should be treated medically, with surgery considered only when complications develop or conservative therapy fails.

Incidence of aortic dissection

The true incidence of aortic dissection is difficult to determine, because of many not correctly diagnosed cases which escape notice. The prevalence of aortic dissection ranged from 0.2 % to 0.8 % in large series of autopsies [1–7]. A minority of series of necropsies, reported a prevalence of 0.1 % [8]. The percentage of correctly diagnosed patients *ante mortem* in large series of autopsies ranged from 40.4 % to 84 % [9, 10]. In an Italian series of necropsies the incidence of aortic dissection increased continuously over the years analysed [11]. In contrast, another large series of autopsies showed a peak of the incidence of aortic dissection in 1968, whereupon it has decreased continuously. A relationship to improvement of diagnosis, medical and surgical therapy was discussed [12].

The mortality from aortic dissection ranged between 0.5 and 2.7 % per 100,000 people from 1950 to 1981 [13]. The highest incidence was shown for black males [12].

Males are more frequently affected from aortic dissection than females, the rate is considered between 2:1 and 5:1 [9, 14, 15].

Young people affected from aortic dissection below the age of 40 are mostly patients with Marfan syndrome [2, 16]. In women aged under 40 years, about 50 % of all aortic dissections occur during pregnancy with a highest incidence in the third trimester and, less frequently, during or soon after labour [17, 18]. High blood pressure and left ventricular hypertrophy are closely related to the incidence of aortic dissection [9, 15, 19]. Other predisposing factors for aortic dissection are chromosomal aberrations (Turner's, Noonan's syndrome), hereditary abnormal connective tissue (Marfan, Ehlers-Danlos) [20], congenital aortic valve defects [21], coarctation of the aorta [22], inflammatory or infectious abnormalities [20, 23] and aortic aneurysm [24]. Fusiform aneurysms of the aorta often antedate or precede aortic dissections, particularly in patients with Marfan syndrome. The age-related peak of aortic dissection is the 5th decade, but there are remarkable differences in consideration of dissection type, concomitant diseases, gender and race [9].

The incidence of hospital admission for aortic dissection ranged between 1:5,335 to 1:16,550 [9]. The range of annual occurrence of aortic dissection was reported between 5 and 20 per million population [4, 8, 25]. The age related maximum for proximal dissection is between 50 and 55 years, for dissection beyond the origin of the left subclavian artery between 60 and 70 years [6, 25–27]. The incidence of hypertension is higher in distal dissection as compared with proximal (80 % vs. 50 %) [28, 29]. With respect to diseases of the aorta dissection is considered as the most common disaster and is two to three times as frequent as a rupture of the infrarenal aorta [4, 20, 25, 30].

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Natural history

In a large series of necropsies of acute aortic dissections, more than 40 % of patients with proximal dissection died immediately, the rate of death ranged between 1 and 3 % per hour. Within 24 hours 70 %, within 1 week 94 % and within 5 weeks 100 % of people with proximal aortic dissection died [14]. The highly lethal nature of acute aortic dissection was confirmed by studies analysing both proximal and distal untreated dissection published during the following decades. 30 % of patients were dead within 24 hours and a rate of 50 % mortality at 48 hours and about 95 % at one month was reported. The reported deaths were related to 75 % to dissection into the mediastinum pleural cavity and pericardium. High incidence of rupture is also reported in patients with chronic aortic dissection and the 5-year survival is approximately 10–15 % [31].

The most recent series of patients with untreated aortic dissection involving the proximal and distal aorta revealed again a rate of 50 % of patients dying within 48 hours, 84 % at one month and 90 % at three months. At one year only 8 % were still alive and at nine years all patients were dead. Because of unfavourable prognosis of untreated aortic dissection aggressive medical therapy and surgery are retried to improve survival rates [32]. In patients with distal dissection, the survival rate at one month was 75 % [31]. Pharmacological interventions to reduce aortic wall tension in aortic dissection have shown lowered early death rates by stabilization long enough until operation can be performed [33–35].

Impact of surgery, surgical techniques and glue aortoplasty

Acute proximal dissection and intramural haematoma as a variant of dissection involving the ascending aorta is generally considered a surgical emergency [36–42]. Surgical intervention improves the natural history of the disease in proximal aortic aneurysm and dissection [43]. After surgical treatment of proximal aortic dissection the survival rate is approximately 70 % after 3 years [44].

In chronic proximal dissection with presentation weeks or months after the acute event or in limited dissections occurring in Marfan's syndrome, treatment depends on the condition of the patient. In critical valve regurgitation or rupture, surgery is performed promptly. Elective aortic replacement because of weakening of the aortic wall, if the vessel diameter exceeds 5 cm, should be performed in general consideration [45]. The critical diameter necessitating surgery is still under discussion [24, 37, 46]. Rapid expansion of vessel diameter and eccentricity of the affected segment are factors that increase the risk of rupture. In the case of a family history of dissection in patients with Marfan's syndrome, elective surgery should be performed if the vessel diameter has reached 4 cm [47]. In stable patients with acute distal aortic dissection medical management is considered justified. Conservative therapy has shown to be effective in preventing death in this situation [38, 48, 49]. The operative mortality and the surgical complications are higher in the acute phase in this setting [15, 33, 48]. Because of very high surgical mortality in Stanford type B acute aortic dissection, medical treatment is the therapy of choice unless there are major complications [50]. After *acute distal* aortic dissection, mortality rates range from 21 % to 67 % and a mid-time survival rate of 30 % or 40 % is reported for patients with medical management [42, 48, 49, 51, 52]. Operative intervention in *acute distal* aortic dissection should be considered in rupture, ischaemic complications of visceral organ systems, limbs, rapidly expanding diameter and pain refractory to intensive medical therapy. Operations of *acute distal* aortic dissection carry a high risk of mortality (35–75 %) which exceeds the risk rate of operated proximal aortic dissection [40, 48, 52, 53]. The mortality rate depends on occurrence of ischaemic organ dysfunction (renal, visceral) and on the age of the patients [52, 53]. The perioperative risk and operative mortality for *chronic distal* aortic dissection correlates with the operative risk for thoraco-abdominal aneurysm [45, 54–56]. The operative risk of spinal cord complications occurring with elective surgery of chronic distal aortic dissection is higher than with replacement of descending and thoraco-abdominal aneurysms [54]. Surgical and medical treatment has influenced the prognosis of patients with aortic dissection. Currently more than 70 % of patients with aortic dissection recover from an acute event if it is treated aggressively [25, 33]. Overall survival rates at ten years are approximately 5 % for untreated patients and 50 % for treated patients [49]. Operative mortality has decreased to about 20 % [57].

Table 1. Aortic dissection: Techniques and grafts used for surgical interventions (for references see text)

- Local fenestration procedure
- Transthoracic fenestration technique
- Insertion of an aortic valve and separate ascending aortic grafts
- Compositive valve graft
- Bentall technique
- Button technique
- Cabrol method
- Elephant trunk procedure
- Homograft or pulmonary autografts
- Repairs of the distal aorta combined with repairs of the proximal aorta
- Descending thoracic aorta replacement
- Thoraco-abdominal aortic repair
- Second stage elephant trunk procedure
- Glue aortoplasty
- Sutureless intraluminal grafts
- Cylinder-type balloon catheters
- Intraluminal sutureless prothesis

Surgical techniques (Table 1)

The objective in operation on patients with dissections is not the removal of the entire dissected aorta, but the excision and replacement of the segment containing the size of origin of the dissection [43]. Formation of thrombus in the false lumen seems to be a good prognostic sign and surgery should be considered as only a part in treatment of aortic dissection [58]. Surgical repair of the proximal tear is the surgical technique of choice in treatment of thoracic aortic dissection. In about 50 % of patients treated surgically, the dissection and false lumen persists [59, 60]. All patients with a history of dissection require close long-term follow-up, in part to control persistent hypertension, but also in part to watch for recurrent dissection. In many operated patients the false lumen remains open with the possibility of further extension of vessel diameter and the consequent requirement of further surgery [60].

A *local fenestration procedure* [61] was first described in 1935, DeBakey, Cooley and Creech reported about a *transthoracic fenestration technique* [62] in 1955. This procedure was a cornerstone in surgery for aortic dissection. Today, treatment by fenestration of the intimal wall of the aneurysm is considered in persistent organ ischaemia after surgical correction of the proximal tear [63]. In the mid 1950s first attempts to perform a definitive repair of the affected part of the aorta with resection of aneurysm and closing of false channel and interposition of tube graft were reported [64, 65].

Another method in surgical treatment of proximal aortic aneurysm is *insertion of an aortic valve and separate ascending aortic*

grafts [66]. *Compositive valve graft* insertion has been recommended in patients with Marfan's syndrome.

Today the two well-known surgical procedures for complete replacement of ascending aorta are the *Bentall and the button technique* [67, 68].

The third most commonly used technique, is the *Cabrol method* [63, 69].

The *elephant trunk procedure* is an alternative form of composite valve graft replacement of the proximal aorta [68].

Aortic root replacement with a *homograft or pulmonary autografts* was reported to be a possible surgical method in 1987 [70, 71].

Antegrade brain perfusion is usually performed in aortic arch replacement [72].

A two-stage technique by which the ascending aorta and aortic arch were replaced first, leaving a segment of distal tubular graft in the descending thoracic aorta. In a second stage the distal aorta is repaired beyond the subclavian artery. This technique is called "*elephant trunk technique*" [73].

Further surgical procedures

- *Repairs of the distal aorta combined with repairs of the proximal aorta* through the left chest.
- Descending thoracic aorta replacement.
- Thoraco-abdominal aortic repair.
- Second stage elephant trunk procedure.

Glue aortoplasty

The common goal of all surgery of the dissected aorta, the complete elimination of the diseased segment in a single operation cannot be achieved in many cases. Conjoining the acutely and chronically dissected aortic wall layers with gelatin-resorcin-formalin (GFR) tissue adhesive was a general idea for development of glue aortoplasty. The use of biological glue has greatly aided the performance of blood-tight-anastomosis on the aorta particularly with acute dissection. The adhesive consisting of GFR has added important contribution to modern aortic dissection surgery [74, 75]. The potentially toxic formalin was recently replaced with a mixture of glutaraldehyde and glyoxal [76]. The dissected space in the ascending aorta and in the arch can be obliterated with the glue, if required. The glue has also been used in descending thoracic aorta to fuse the dissection [77–80]. Complete disappearance of the false channel was achieved in more than 50 % of the patients [77, 81]. Glue aortoplasty is a useful technique [82] but cannot be considered superior to the conventional models of dealing with proximal or distal aortic dissection [83]. There are a few alternative procedures in surgical treatment of aortic dissection like thrombo-exclusion and aortic tailoring. The value of these methods in clinical practice requires determination in further examinations [84, 85].

The use of inlay grafts has furthermore influenced operative tactics in surgical treatment of acute proximal aortic dissection [86].

Sutureless intraluminal grafts have been introduced in aortic dissection surgery to avoid a non-stenotic complication and long cross-clamp times [87].

In the management of acute aortic dissections *cylinder-type balloon catheters* have been used for minimal invasive procedures to close the entry side of the dissection [88].

An innovative technique for high risk is the utilization of an *intraluminal sutureless prosthesis* in patients with friable thoracic aortic tissue [89].

Another alternative surgical technic is *bypassing of the dissecting aorta with a sleeve of Dacron* to create reversal blood flow in the distal aorta to perfuse the major arterial branches arising from the dissected segment [84].

Percutaneous stent insertion to open an included aortic branch has been reported recently [90].

Another percutaneous procedure is fenestration of the intimal septum using an angioplasty balloon to re-establish blood flow to visceral arteries and to support lower limb perfusion [91].

The best technique for surgical treatment of aortic dissection has to be determined, every surgeon has his own experience.

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