

## What do we really know about Aortic Dissection?

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## ABSTRACT

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**Introduction:** Aortic dissection (AD) is a rare and severe condition caused by modified (smoking cigarettes, high BP) and fundamental factors (for example sex, age, genetic disorders). Classification considers location in respect to aortic arch – Type A is proximal to the arch whereas Type B is located distally. It is difficult to diagnose this illness due to many possible presentations and non-specific symptoms. It is not a frequently occurring illness. However, it is lethal and time plays a critical role. Thus, it is vital to be aware of it.

**Objectives:** The purpose of this article is to present the latest information concerning risk factors, symptoms, diagnosis, medical management and possible complications of Aortic Dissection.

**Material and methods:** The article is based on review of publications in PubMed database using the

terms: “Aortic Dissection”, “Risk factors of Aortic Dissection”, “Acute Type A Aortic Dissection”, “Acute Type B Aortic Dissection”, “Chronic Aortic Dissection”, “Treatment and complications of AD”.

**Summary:** Aortic Dissection is a life-threatening condition that must be considered in many cases because it is often misdiagnosed. Past medical history, genetic disorders and lifestyle choices needs to be known to make a correct diagnosis. Treatment is selected based on location, the patient's condition and illness phase. Knowledge of treatment methods and its possible complications are essential for patient's survival.

**Keywords:** Aortic dissection, risk factors, symptoms, diagnosis, medical management and possible complications

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## **INTRODUCTION AND GENERAL INFORMATION**

Aortic dissection (AD) is a severe medical condition caused by internal wall damage. This leads blood flow to come between the walls of aorta and creates additional lumen called false lumen. It can decrease life expectancy and cause many complications [1]. There are two main classifications of AD - Stanford Classification and DeBakey. Stanford Classification presents two types of aortic dissection – Type A, which always includes aortic arch and Type B which locates dissection below the arch. DeBakey classification divides dissection into 3 groups. Classification is based on whether dissection covers ascending aorta (I,II) or not (IIIa and IIIb –the dissection is below left subclavian artery) [2]. The studies show that in group of 100,000 people about 4 will suffer from AD. However, the incidence is higher in patients between 65 and 75 years (35 people with AD per 100,000) [1,3].

## **RISK FACTORS**

Statistically men are two to four times more likely to develop AD. Other factors include hypertension, cigarette addiction, genetic disorders (Marfan, Loeys-Dietz and Ehlers Danlos syndrome), bicuspid aortic valve and drugs (for example cocaine, amphetamine) are reported to increase the risk of aorta dissection [1,4]. Pregnancy can also affect aortic dissection as female with previous AD in past medical history should be discouraged from getting pregnant. The risk for another AD is higher during pregnancy [5,6]. It has also been noticed that acute type A aortic dissection is statistically more often in winter and influenza season. On the other hand acute type B is mostly observed during spring [7,8].

## **SYMPTOMS, DIAGNOSIS AND MEDICAL MANAGEMENT**

AD diagnosis can be demanding and time-consuming owing to non-specific symptoms such as chest, back or stomach pain and even loss of consciousness. Sudden chest pain is the most common symptom in approximately 80% of AD cases. Hypertension can also be observed as 33% patients manifest it. Neurological symptoms can appear due to malperfusion and cause lower or upper limb ischemia or paraplegia. Thus lack of pulse can be a significant sign [5,9]. This specific symptom is seen in 31% of acute A type AD [9]. Cardiac tamponade also can be seen as a symptom especially in acute type A [10]. All of these above can lead to misdiagnosis, which is estimated to affect 33% patients with AD and can cause severe complications because every hour chance of survival is decreased

by 1 % (in the first two days for acute patients) [11,12]. Laboratory tests have to be made such as Morphology, Procalcitonin, CPR, Glucose, Troponin I or T, Creatine Kinase, AST, ALT, Blood gases and D-dimer. It is essential to know that D-dimer is non-specific marker. However very high D-dimer level (the highest in the first hour after the presentation of symptoms) when admitted to the hospital can suggest AD in contrast with other conditions in which D-dimers increase slowly over a time period. Computer tomography, Magnetic Resonance Imaging or Echocardiography are essential to rule out diagnosis [13].

## **ACUTE TYPE A AORTIC DISSECTION**

Acute A type AD is usually managed surgically, because the mortality rate is 2-3 times higher without this approach. Types of procedures are picked individually, and the surgery may involve for instance ascending aorta, aortic root, aortic valve, total arch replacement, descending aorta replacement, open procedure or even cerebral perfusion [9,10]. Usually the operation is under hypothermic circulation [7]. The need for operation must be assessed as not all patients will qualify. Some symptoms question the operation to be the first choice for patients that are physically enabled (before acute A type AD diagnosis). Age is not considered to be a predictor for higher mortality for acute A type AD patients as this age group have the same outcome as younger patients. Malperfusion, tamponade, neurological presentation of symptoms increase the mortality for operated patients but the surgery is the only way to survive and the time plays a vital role [10]. For instance, the study presents the benefits of operation for patients with cerebral malperfusion (Acute A Type Dissection) treated surgically and it shows a 75% survival rate in comparison to conservative approach which was about 25% [14,15]. Patients in stable state also have to be urgently operated on as it is impossible to predict the rupture of aortic wall [10]. In theory acute AD type A can be cured if there is no false lumen after the operation. However, this cannot be achieved in most cases, and patients usually enter the chronic phase [17]. Trimachi et al. shows that mortality rate for surgically treated patients is 25% for unstable acute AD type A, and for stable acute AD type A patients it is 16%. This data only concerns patients that underwent operation in hospital. A different study presents 58% mortality rate for those not receiving surgery in contrast to 26% mortality rate for those who underwent the operation (Acute A type Dissection) [16]. Another report by Westaby et al. presents other data that the mortality rate for A type acute AD was 5.3-6.3% (in-hospital patients treated surgically). In this study there was a different approach and the operation was focused on the complete resection of the entry tear [7,17].

## **ACUTE B TYPE AORTIC DISSECTION**

Type B can be categorized into 3 groups based on the duration of the symptoms –acute (less than 2 weeks), subacute (between acute and chronic phase) and chronic (over 3 months) [18]. Acute AD is clinically grouped as complicated (for example rupture of the wall, malperfusion) or uncomplicated. The latter can be treated pharmacologically (oral medical therapy). The goal is to reduce maximum pressure in the left ventricle and also in false lumen and systolic blood pressure (SBP) needs to be lowered to less than 120mm/Hg. Minimalizing the aortic wall stress and heart rate (HR <70/min) is also essential. Beta blockers, alpha and beta blockers, sodium nitroprusside, analgesics (to control the pain) are used to treat acute phase. If the HR and SBP are controlled, oral hypertension medications and painkillers have to be taken for the rest of one's life [2].

For uncomplicated patients, oral medical therapy compared to endovascular stent-grafting shows no difference concerning mortality in study conducted for over 2 years. However, uncomplicated patients with endovascular treatments are observed to have an advantage like aorta remodeling [19]. Other studies reveal that remodeling was shown to influence 5-year survival positively, but no sooner than 2 years after the procedure. It suggests that endovascular approach should be considered for those with longer than 2 years life expectancy. Thus, they are said to experience the long-term benefits and potentially delay complications [20].

Adherence to medications must also be considered in uncomplicated cases when treated pharmacologically only. It was observed that less than 50% of examined patients were taking medications as prescribed. Not only aortic surgery in the past but also understanding the condition and possible risks, taking numerous tablets were reported, to increase patients' compliance [21].

Endovascular method could also be considered for those (uncomplicated acute aorta dissection) with wider false lumen than true lumen, as well as for those with aortic diameter longer than 40mm [22]. Open surgical repair, TEVAR and hybrid surgery are the main methods used in complicated Acute Aortic Dissection - B type. Endovascular treatment in Acute type B was proven to have lower mortality rate than open surgery and was associated with fewer medical complications afterwards [23]. In other words, it is suggested to take on this approach whenever an endovascular method is possible [24]. Mortality rate for patients after TEVAR is estimated at 7.3%, for uncomplicated patients treated pharmacologically the rate is 10% and open surgical repair has a value of 30% in hospital mortality rate (acute AD type B) [9,23,25].

## **CHRONIC AORTIC DISSECTION**

Chronic phase patients are predominantly the ones that overcame the acute phase. Also, the chronic phase can be subclinical, diagnosed by accident with no symptoms. In other words, the chronic group consist of patients who were operated after A type AD diagnosis and survived, patients with uncomplicated acute type B AD who were treated conservatively and, finally, complicated type B AD patients that underwent surgery [26]. Blood pressure control is essential for AD chronic treatment. Beta-blockers lowers BP, decrease aortic wall stress and left ventricular pressure [27]. Furthermore, the enlargement of aorta is lower in patients taking beta-blockers compared to the other medications, thus they lower the potential risk of reoperation [28]. Medicaments that lower lipids level are said to be beneficial as they prevent faster collagen breakdown, limit the inflammation and reduce the increase rate of aortic measures [29]. 20-50% of type B patients that were previously treated with medications (in acute phase and have to continue taking antihypertensive medications in chronic phase) will need invasive treatment further in life because of an aneurysm or fast aorta enlargement that is why they need to control aortic measurements and condition. Fast aorta enlargement, false lumen increased measures, aneurysm, pain and signs of malperfusion are symptoms in chronic phase that need to be treated invasively [28].

## **FOLLOW-UP AND POSSIBLE COMPLICATIONS**

Follow-up methods includes Computer Tomography Angiography (CTA), Magnetic Resonance Imaging (MRI), Ultrasound. CTA is a gold standard. It shows accurate measurements of aorta, density of aneurysm and a possible blood leak outside the stent graft. On the other hand, in CTA patients are exposed to ionizing radiation and are given nephrotoxic contrast. MRI is also an excellent method for aneurysm and endoleak detection. However, it is less available, more costly and can cause problems with stainless and nickel stent grafts analysis because of many artifacts. Ultrasound or duplex ultrasound can only be a great addition to CTA as they do not reveal mechanical complication within stent graft, which is a disadvantage [30]. About 40% of patients develop an aneurysm after diagnosis and treatment in Type B AD [31]. Aneurysm is the most frequent sequelae and is also a cause of reoperation. Control of blood pressure reduces the risk of aneurysm. Studies show that uncontrolled BP can lead to an aneurysm and increased the size of aorta. 50% of patients with uncontrolled BP develop an aneurysm in contrast with patients that have controlled BP, as there is 15% chance of aneurysm creation within this group. In

other words, adequate control of BP decrease the chance of reoperation [32,33]. After diagnosis and treatment of acute A type AD, dilatation of aorta can occur. It was showed that that the time frame for this complication is mainly 24 months. In other words, if there is no increase in aorta diameter, and there is no obstruction in false lumen there is a meagre chance of further complications and the duration for the next follow-up can be prolonged (follow-up can be planned once in 2 or 3 years instead of annual follow-up) [26]. Statistically, type A is more associated with complications such as a rupture of aorta before treatment and further aorta dissection (further dissection concerns 50% of acute A type AD patients) [9,10,16,31]. In a group of acute A survivals reoperations concern aortic valve, root or distal aorta especially if there is enlargement of aorta (it is considered that 1mm growth per year increases the potential chance of reoperation). Reoperations after TEVAR also applies for type B group as about 30% of survived will be operated again [9,10]. New entry after TEVAR is caused by stent graft or iatrogenic induced. However, it mostly concerns dissection type A rather than B and the ones with acute presentation of symptoms. The described state is most likely to be observed 19 +/- 7 months after the operation and is due to the aortic walls' fragility. That shows importance of CT follow-up scans as only 5% of patients present the symptoms such as back or chest pain [34]. Moreover, studies show that too big stent-graft can cause retrograde dissection. This is not an often complication in type B, but very fetal one and thus needs reoperation [35].

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