CASE SERIES

Ascending aorta disease: a case series

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Thoracic aortic aneurysms have a prevalence of up to 0.16%, with fatal complications. *Objective.* To describe a series of cases in the treatment of thoracic aortic disease. *Material and methods.* We retrospectively reviewed patients undergoing surgery of the thoracic aorta during January 1, 2013 to December 31, 2017, documenting risk factors, functional class, type of aneurysm and / or dissection and surgical risk. *Results.* There were 6 patients, 4 of the male gender (66%), with an average age of 38 years (21-45). The cardiovascular risk factors were: systemic hypertension (50%), dyslipidemia (33%), smoking (66%). We found one case with Marfan Syndrome (16%), one as reoperation (16%) and 3 had type A dissection (50%). They were in preoperative functional class III of 66% and III for 33%; ejection fraction of the left ventricle higher than 50% in 83%, and moderate aortic insufficiency in 66%. The average EuroSCORE II was 6.9 (1.52-25.14). Operative mortality was 50%. *Conclusions.* Our hospital has a high early operative mortality due to the characteristics of the series of patients. However, even a more extensive series is needed to improve our experience while making comparison with other national and international centers.

Key words: **Thoracic Aneurysm; Thoracic Aorta; Ascending Aorta; Aortic Dissection.**

Los aneurismas de la aorta torácica tienen una prevalencia hasta del 0.16%, con complicaciones mortales. *Objetivo.* Describir una serie de casos en el tratamiento de la enfermedad de la aorta torácica. *Material y métodos.* Revisamos retrospectivamente los pacientes sometidos a cirugía de la aorta torácica durante el 1 de enero de 2013 al 31 de diciembre de 2017, documentando factores de riesgo, clase funcional, tipo de aneurisma y/o disección y riesgo quirúrgico. *Resultados.* Fueron 6 pacientes, 4 del género masculino (66%), con edad promedio de 38 años (21-45). Los factores de riesgo cardiovascular fueron: hipertensión arterial sistémica (50%), dislipidemia (33%), tabaquismo (66%). Encontramos un caso con síndrome de Marfan (16%), una reoperación (16%) y 3 tuvieron disección tipo A (50%). Tenían clase funcional preoperatoria en estadio II el 66% y 33% en estadio III; fracción de eyección del ventrículo izquierdo > 50% en 83% e insuficiencia aórtica moderada el 66%. El EuroSCORE promedio fue 6.9 (1.52-25.14). La mortalidad temprana fue del 50%. *Conclusiones.* Nuestro centro hospitalario tiene una mortalidad temprana elevada por las características de la serie de pacientes. Sin embargo, se necesita de más casos para comparar con otros centros nacionales e internacionales.

Palabras clave: **Aneurisma torácico; Aorta torácica; Aorta ascendente; Disección aórtica.**

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The aortic aneurysm, which is defined as the localized dilation of more than 50% of the normal diameter of the aorta, includes the entire arterial wall (intima, media and adventitia) [1,2]. This aneurysmal disease occurs with a low frequency, prevalence of up to 0.16% [3] and with incidences between 5.9 to 16.3 per 100,000 patients/year [3,4], depending on the series, mainly on male gender [2-4]. The most frequent location is in the thoracic aorta (60%), followed by the descending aorta (40%) and finally in the aortic arch (10%) [5]. Degenerative condition of the median layer leads to a wall weakening, while getting higher the risk for aneurysmal dilation. Systemic arterial hypertension accelerates this aforementioned process [2,5].

Aneurysmal dilation may remain totally asymptomatic, just before when compression symptoms over other surrounding structures appear. In such conditions, imaging approach become highly relevant. Transthoracic echocardiography, angiotomography and nuclear magnetic resonance are very useful tools to make clear diagnosis and opportune treatment [5].

All this above notwithstanding, aortic dissection may occur, a very dreadfully complication with fatal prognosis for life expectancy [6,7].

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MATERIAL AND METHODS

We collected all information from clinical records and files from patients treated in our institution since January 1, 2013 to December 31, 2017. Demographic data as age, sex, occupation, functional class, factors and associated comorbidities, whether or not it any dissection, type of operation, surgical risk and mortality were all obtained.

RESULTS

Six patients were operated on, out of them 4 were male (66%). Average age was 38 years-old (range, 21 - 45 years). Associated cardiovascular risk factors can be seen in **Table** 1.

TABLE 1. Cardiovascular risk factors associated with patients with ascending aortic disease (n = 6).

VARIABLE	n	%
Active smoking	4	66
Systemic Arterial Hypertension	3	50
Dyslipidemia	2	33

Systemic arterial hypertension in 3 patients (50%), dyslipidemia in 2 patients (33%), 4 patients (66%) with smoking. We found one case with Marfan syndrome (16%), one case for reoperation (16%) and 3 had Stanford type A dissection (50%). One patient presented a bicuspid aortic valve. The functional class of the New York Heart Association (NYHA) scale was stage II, 66% and stage III, 33% (**Table 2**).

TABLE 2. NYHA functional class (n = 6).

STAGE	n	%
I	0	0
II	4	66
III	2	33
IV	0	0

with left ventricular ejection fraction (LVEF) >50% in 83%, only one patient had LVEF between 20-30%; moderate aortic regurgitation was found in 66% of cases (**Table 3**).

The average EuroSCORE II was 6.9 (1.52-25.14), considering as "urgent" procedure because of the acute dissection. All cases undergone the Bentall-De Bono technique. Anterograde cardioplegia solution (Custodiol ®) was used as myocardial protection, with an average time of extracorporeal circulation of 165 ± 70 minutes, with a minimum time of 100 minutes and a maximum time of 240 minutes. Early operative mortality was 50%.

TABLE 3. Echocardiographic characteristics (n = 6).

LVEF	n	%
> 50%	5	83
50 - 31%	0	0
30-21%	1	16
≤ 20%	0	0
AORTIC INSUFFICIENCY		
Absent	0	0
Mild	2	33
Moderate	4	66
Severe	0	0

DISCUSSION

Aortic aneurysms are associated with high morbidity and mortality. According to international statistics, more than 17,000 deaths for this pathology are attributed annually in the United States [8]. Nevertheless, in our country Mexico we do not have accurate statistics coming from any reliable database for this purpose.

Several risk factors have been associated with aortic disease, according to the affected aorta segment. At the same time, some syndrome may be associated as well, most commonly to Marfan syndrome, Loeys-Dietz syndrome, and Ehlers-Danlos. In "non-syndromatic" aneurysms, abnormalities are limited to the cardiovascular system. Most of these conditions exhibit an autosomal dominant inheritance but affect individuals who do not have characteristics of cardiovascular disease or another recurrent phenotype. Among this type, dissection and familial aortic aneurysm are considered, also associated to bileaflet aortic valve [5,8].

Diagnosis approach must be made through imaging techniques such as such as echocardiography, magnetic resonance and mainly angiotomography with contrast [2,9,10], covering the internal and external diameters at different points. Especially care must be paid on the ascending aorta, where normal range goes from 3.5 cm up to 3.72 cm for women and from 3.63 cm up to 3.91 cm in men. Measurements at the level of the ascending aorta are about 2.86 cm for both cases [2].

Surgical treatment must be established accordingly with patients' special characteristics. Type of patient, asymptomatic, with genetic syndromes, with familial aortopathy, among many others, should be taken into account [2,10]. Thus, in asymptomatic patients with diameters in the ascending aorta equal to or greater than 5.5 cm or with growth larger than 0.5 cm / year or who present symptoms, the indicated treatment is operation. On the other hand, patients with an associated genetic anomaly (Marfan syndrome, bileaflet aortic valve, among others) should be considered. In this case, surgical option is reliable, with diameters in the ascending aorta equal to or greater than 5 cm or growing up to 0.5 cm / year [2,10]. Also, Canadian Cardiovascular Society proposes for the family aortopathy and other genetic syndromes other than Marfan the surgical correction with diameters between 4.2 cm to 5

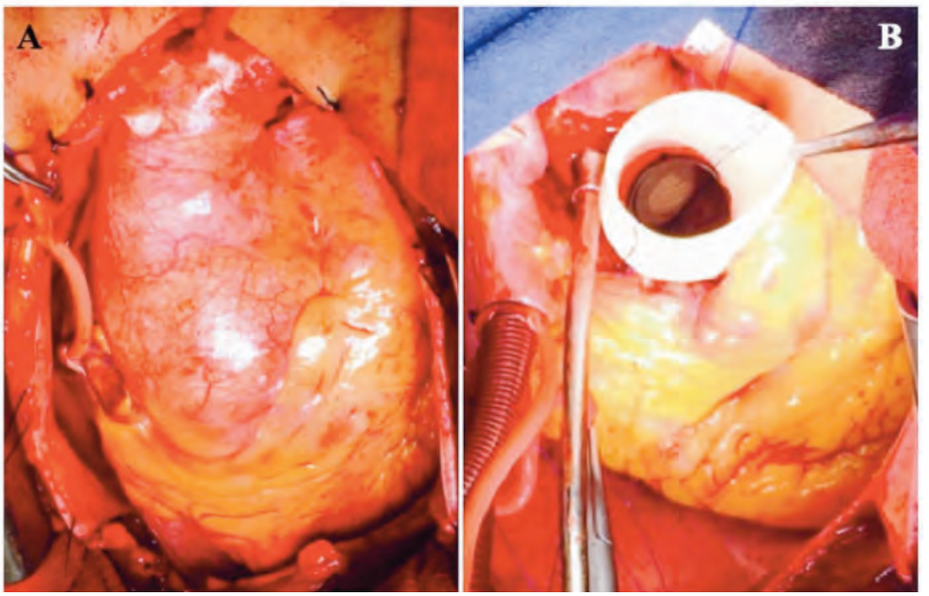

Figure 1. (A) Ascending aorta aneurysm in a patient with Marfan syndrome; (B) View at 30 cm, placement of a Dacron® valved graft with the Bentall - De Bono technique.

cm and with the aortic root with diameter of 4 cm, especially in pregnant patients carrying genetic syndromes [10].

There are different approaches, emphasizing always avoiding neurological complications such as deficit and decreasing mortality, thus Benedetto et al., demonstrated a lower mortality and neurological complications attributed to the reverse flow of central cannulation [11]. On the other hand, as there is also involvement of the thoracic aorta, the aortic arch compromise is added, with the possibility of selective or retrograde anterograde perfusion, in addition to the option of performing deep hypothermia and circulatory arrest, which Hu et al. found a significant difference in morbidity and mortality [12].

The procedure will depend on the area compromised, the conditions of the patient and, why not, the availability of hospital resources; having thus the Wheat technique, Bentall-De Bono technique, Cabrol technique, David technique, among others [13-15]; concomitantly to these is the graft material to be used, agreeing that Dacron® has shown a better result with respect to surgery of aortic root and replacement of ascending aorta [16].

Despite all therapeutic measures and in a timely manner, when developing an acute aortic dissection, associated comorbidities occur more frequently, so we have permanent cerebral vascular events, renal failure, prolonged mechanical ventilation and increased mortality, reaching up to 27,7% in centers of great experience [6-7].

In relation to the series of cases reported for the first time by our institution, it coincides with the literature in the predominance of male sex 66%, with an average age of 38 years, being somewhat younger population in relation to other results [9,13,14]. In this sense, the main associated comorbidity was arterial hypertension and the main genetic syndrome manifested was the Marfan syndrome, thus maintaining the relationship with that described in the literature [8,9,13,14].

If we consider that the factors that are most related to early mortality are age over 65 years, NYHA functional class stage III and stage IV, aortic dissection, emergency surgery, concomitant procedures and postoperative complications (bleeding, rhythm and neurological deficit) [13,14], our patients presented in a 50% Stanford A type aortic dissection and 33% were in NYHA stage III functional class, so we could infer an early mortality in our series that arrived to 50%, the latter being a higher percentage in relation to that presented by the Mayo Clinic [7]. Surgical correction in all cases was performed using the Bentall - De Bono technique with a valved graft Dacron ® (**Fig. 1**), with central cannulation in 66% of cases, without using selective anterograde cannulation.

In conclusion, our hospital has a high mortality in relation to that reported in the national and international literature, however this first reported series allows us to identify that half of it had at least one risk factor associated with mortality, in addition to when considering the socioeconomic characteristics of the patients who arrive at our institution and the advanced stages, they denote the delay in their timely diagnosis or the establishment of a therapeutics in time, which together with the costs of material and input could not be repaired in a timely manner. Finally, it is clear that it is necessary that a greater casuistry is needed for a comparison with other national and international reference centers.

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