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Bicuspid aortic valve and aortopathy: two sides of the same coin?

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Objectives: We conducted a cohort study to assess baseline characteristics of aortopathy and progression to aneurysm formation in adults with bicuspid aortic valve (BAV) followed-up in our health network.

Methods: We recruited all consecutive asymptomatic patients (p) with definite diagnosis of BAV since 2009 after approval of informed consent. We analyzed baseline characteristics of aortopathy and incidence of aortic complications (aneurysm >45mm, dissection or aortic intervention) during follow-up.

We assessed aortic dimensions at 6 different levels and, incidence of aortopathy with 2 definitions (absolute ≥ 40 mm and indexed, $21\text{mm}/\text{m}^2$).

Results: The cohort included 220 p (43 \pm 15.2 y/o, 75% male, 7% with familial BAV). Age at diagnosis was 37 \pm 15.5 yrs. Most of the p had fusion of coronary cusps (Type I BAV: 175, 79.4%) and a raphe in 155 p (70.4%). 20 p (9.1%) had severe aortic valve dysfunction at baseline.

The mean maximum aortic diameter was 36 \pm 6.8mm and 1 out of 2.5 p (42%) had aortic dilatation at baseline. We found 24 patients (10.9%) with aneurysm criteria at BAV diagnosis with a mean diameter of 47.2 \pm 5mm. 63 patients (29%) had aortic diameters >21 mm/ m^2 , mostly smaller patients. $p < 0.0001$ (Figure 1). Tubular ascending aorta dilatation was present in 36 (41%) p, followed by root dilatation in 30 (33%) p. We found no association between aortopathy patterns and BAV phenotype (Table 1).

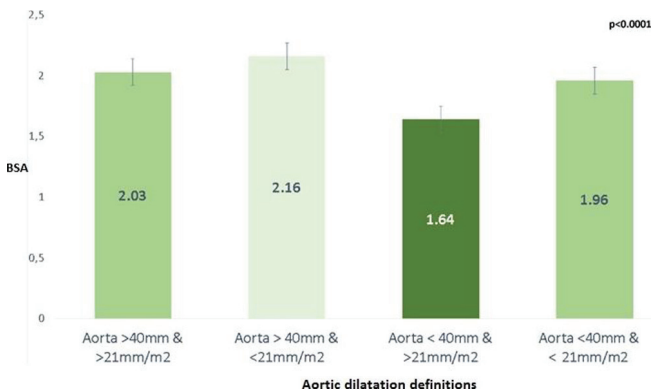
There were 2 deaths during follow-up (4.1 \pm 2.3y). The most frequent procedure (10, 55.5% of all interventions) was AVR, followed by Bentall de Bono (5, 27.7%). The primary indication for intervention was severe aortic stenosis (8, 44%), followed by asymptomatic aortic dilatation (4, 22%).

16 (8.2%) p developed aneurysm and there were no aortic dissections during follow-up.

Comparison of diameters between valve phenotypes I and II at six levels showed non-statistically significant differences

Aortic dimensions	Indexed diameters (cm/ m^2) (n=220)	BAV Type I indexed (cm/ m^2) (n=175)	BAV Type II indexed (cm/ m^2) (n=27)	p
Aortic annulus	1,19 (1,06–1,29)	1,22 (1,07–1,29)	1,19 (1,07–1,26)	NS
Sinus of Valsalva	1,89 (1,71–2,07)	1,91 (1,71–2,07)	1,91 (1,65–2,23)	NS
Sinotubular junction	1,62 (1,44–1,81)	1,62 (1,43–1,79)	1,63 (1,45–1,82)	NS
Proximal Ascending aorta	1,88 (1,67–2,09)	1,87 (1,67–2,05)	1,91 (1,77–2,06)	NS
Aortic arch	1,25 (1,11–1,55)	1,26 (1,10–1,51)	1,25 (1,18–1,56)	NS
Isthmus	0,93 (0,81–1,21)	0,93 (0,81–1,19)	0,96 (0,90–1,24)	NS

Data are expressed as median (IQR). * $p < 0.05$.



BSA and aortic dilatation definitions.

Conclusions: The incidence of intervention on aortic valve or ascending aorta and death over a mean of 4.1 y was high. We identified a clinical aortopathy associated to BAV, with excess risk of aneurysm formation. There were no aortic dissections during follow-up.

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Association of alcohol consumption with aortic root dilatation: Corinthia study

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Introduction: The harmful effects of excessive alcohol consumption on the cardiovascular system as well as on general health are already known. However, data on the incidence of alcohol in aortic diseases are isolated.

Purpose: The aim of this study was to investigate the possible association of

excessive alcohol consumption with aortic root dilatation.

Methods: We examine 1043 participants (467 men, aged between 30 and 98) of the Corinthia study. Among other tests, echocardiography was performed and subjects were asked to complete questionnaires. Several demographic and clinical characteristics were recorded. Alcohol consumption was assessed on the basis of the questionnaire replies. Subjects with daily consumption of alcohol were categorized as habitual alcohol consumers (HAC) and those consuming alcohol occasionally were categorized as social drinkers (SD).

Results: Of the total studied population, 448 individuals (43%) were found to be HAC. Between HAC and SD there was no difference in age (65 \pm 12y vs. 64 \pm 11y, $p=0.12$) and the incidence of hypertension, whereas there was an increased prevalence of male gender in the HAC groups compared to SD (60% vs. 40%, $p<0.001$). Also, HAC had an increased aortic root diameter (AoRD) (33.61 \pm 4.13mm vs. 31.83 \pm 4.41mm, $p<0.001$) and an elevated AoRD index (17.43 \pm 2.23mm/ m^2 vs. 16.84 \pm 2.32mm/ m^2 , $p<0.001$). Interestingly, individuals with an aortic root aneurysm (AoRD>40mm) were more often HAC (61% versus 39%, $p=0.01$). In addition, depending on the daily alcohol consumption, a gradual increase in the dimensions of AoRD (0–1glass: 31.99 \pm 3.65mm vs. 1–2 glasses: 33.65 \pm 3.73mm vs. >3 glasses: 34.58 \pm 4.44mm, $p<0.001$) and in AoRD index (0–1 glass: 16.95 \pm 2.18mm/ m^2 vs. 1–2 glasses: 17.45 \pm 2.07mm/ m^2 vs. >3 glasses: 17.39 \pm 2.39mm/ m^2 , $p<0.04$) was observed (Figure 1, panel A, B). Even after adjustment for many confounders such as age, gender, smoking, hypertension and body surface area, the amount of alcohol consumption appeared to be significantly associated with aortic root dilation (B coefficient = 0.55, 95% CI: 0.09–1.01, $p=0.02$) (Figure 1, panel C).

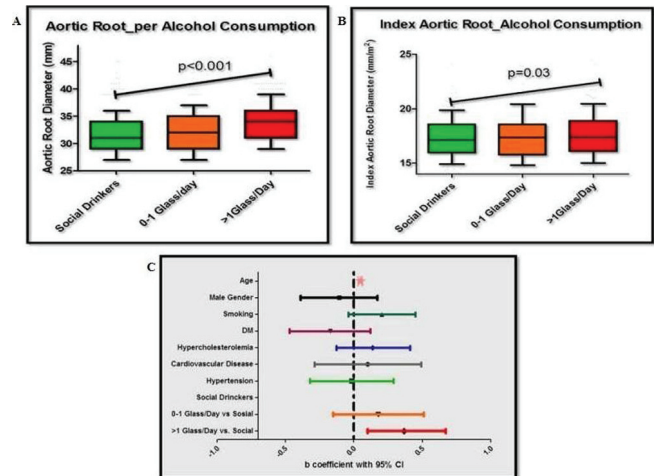


Figure 1

Conclusion: Systemic alcohol consumption is associated with increased aortic root diameter. These findings shed light on the pathophysiologic mechanisms underlying dilatation of ascending aorta and may be used to highlight novel risk factors.

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Long-term efficacy of losartan vs atenolol for the prevention of aortic dilation and clinical complications in Marfan syndrome

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Background: Beta-blockers are the standard treatment for preventing aortic dilation in Marfan syndrome (MFS). Recent clinical trials on the effectiveness of Losartan in MFS have resulted in conflicting results. The LOAT trial did not show benefit from losartan compared to atenolol in these patients. However, all these studies have a limited follow-up (3 to 3.5 years). Therefore, the aim of the present study was to evaluate the benefit of losartan compared to atenolol for the prevention of aortic dilation and aortic complications in MFS patients over a longer period of observation (>5years).

Methods: The MFS patients included in the previous LOAT clinical trial were followed up over a non-blinded extension of the study, maintaining the initial treatment (losartan or atenolol). Clinical events were registered and, after at least 5 years from the start of the clinical trial, a new MRI study was performed.

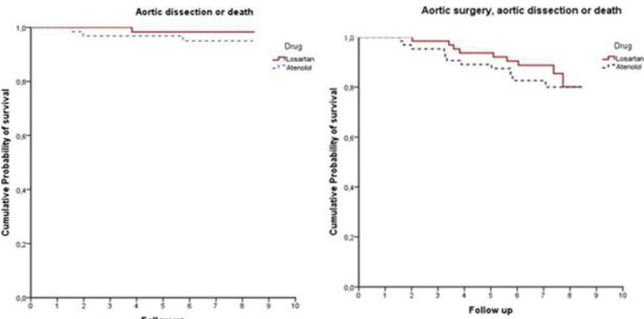
Results: A total of 128 patients were included in the study (64 in the atenolol and 64 in the Losartan group). Mean clinical follow-up was 6.7 \pm 1.5yrs. A total of 9 (14.1%) events occurred in the losartan group and 12 (18.8%) in the atenolol group. Survival analysis showed no differences in the combined end-point of need for aortic surgery, aortic dissection or death ($p=0.462$) or for the

Abstract P723 – Table 1. Aortic dilation outcome by MRI

Aortic dilation rate by MRI	n	Losartan, mean (95% CI)	n	Atenolol, mean (95% CI)	Inter-group difference, mean (95% CI)	p
Aortic root						
Diameter (mm/yr)	61	0.4±0.6 (0.2 to 0.5)	58	0.4±0.5 (0.3 to 0.6)	0.0 (–0.25 to 0.17)	0.754
Diameter/BSA (mm/m ² /yr)	61	–0.1±0.4 (–0.2 to 0.0)	58	–0.1±0.4 (–0.2 to 0.0)	0.0 (–0.2 to 0.1)	0.803
Z-score (per year)	61	–0.1±0.4 (–0.2 to –0.0)	57	–0.2±0.4 (–0.3 to 0.1)	0.1 (–0.1 to 0.2)	0.236
Ascending aorta						
Diameter (mm/yr)	61	0.2±0.4 (0.1 to 0.3)	58	0.1±0.6 (0.0 to 0.3)	0.1 (–0.1 to 0.2)	0.551
Diameter/BSA (mm/m ² /yr)	61	–0.2±0.4 (–0.3 to –0.1)	58	–0.2±0.4 (–0.3 to –0.1)	0.0 (–0.1 to 0.2)	0.745

Results are annual change expressed as mean ± SD (95% CI). p represents t-test. BSA: body surface area.

combined end-point of acute aortic syndrome or death (p=0.305) (Figure). Furthermore, no statistically-significant differences were observed in aortic dilation progression between groups (Table).



Cumulative survival curves

Conclusions: In Marfan patients, losartan was not superior to atenolol for the prevention of aortic dilation or aortic events in a long period of observation.

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Techniques and outcomes of trans-catheter closure of aortic anastomosis leak after aortic replacement

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Background: Aortic anastomotic leak (AAL) is a rare but intractable problem after surgical aortic replacement and patients are at high risk. Surgical repair of AAL remains the most common therapy, but with high morbidity and mortality rates. Trans-catheter closure has emerged as an alternative approach for such patients. However, the available data are limited to case reports or small series.

Purpose: The aim of this research was to describe the procedural techniques of aortic anastomotic leak (AAL) closure and evaluate the feasibility and efficacy of this procedure.

Methods: From October 2015 to November 2017, twenty consecutive patients (13 males, mean age 47.1±11.8 years) were referred to our center for possible trans-catheter closure. After transesophageal echocardiography and computed tomography angiography evaluation, AAL was defined as three types. Type I AAL resulted in aorta-to-right atrium fistula. Suture line dehiscence led to a pseudoaneurysm was defined as type II. Type III led to aortic false lumen patency and procedure inefficiency for aortic dissection. The procedure and clinical outcome were assessed.

Results: Type I AAL demonstrated in 6 (30%) patients, type II demonstrated in 4 (20%) patients and type III demonstrated in 10 (50%) patients, respectively. Successful occluder deployment was accomplished in 17 cases (85%). Reduction in AAL to mild or less was achieved in 15 patients (88%). A second closure was carried out for residual shunt in 2 patients. There were no procedure-related death, myocardial infarction or stroke. Complete thrombosis rate was 100% in the 10 patients (type III) with false lumen patency. The NYHA cardiac function improved

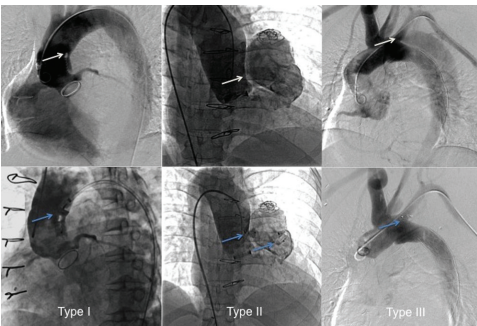


Figure 1. Three types of AAL

in 6 patients (type I) with aorta-to-right atrium fistula, the right atrium systolic pressure (25.3±4.1 mmHg to 7.0±1.2 mmHg, p=0.003) was significantly reduced. The diameter of pseudoaneurysm was decreased (4.95±1.8 mm to 2.0±1.8 mm, p<0.001) in 4 patients of type II.

Conclusion: Trans-catheter closure appears to be a reasonable alternative approach for AAL in patients who were high-risk candidates for redo-surgery.

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Effect of renin-angiotensin system blockade in prevention of thoracic aortic aneurysm progression

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Introduction: Beta blockade is a viable option in treating patients with thoracic aortic aneurysms (TAA), particularly in Marfan syndrome. However, the data on efficacy of potentially beneficial angiotensin-converting enzyme inhibitor (ACEI) or angiotensin II receptor blocker (ARB) therapy are limited. The aim was to evaluate the effect of adding ACEI or ARB to baseline beta-blocker therapy in patients with non-syndromic TAA.

Methods: 50 consecutive patients (mean age: 57.9±14.9 years) with TAA were enrolled in our retrospective, non-randomized, single-center study. Most patients (74%) according to the case reports were receiving a beta-blocker at baseline. 17 patients (34%) were receiving ACEI (perindopril, enalapril or fosinopril), 18 patients (36%) – ARB (losartan or valsartan) and 15 patients (30%) didn't receive any ACEI or ARB medications. Repeated (in 2.39±1.73 years) computed tomography with three-dimensional reconstruction used to evaluate aortic diameters and aortic dilatation rate. Left ventricular function and dimensions were examined during transthoracic echocardiography.

Results: The mean aortic diameter at the level of the sinuses of Valsalva in the overall group didn't change significant during the follow-up (from 44.6±7.2 mm to 44.9±6.3 mm, p=0.86).

The mean increase in aortic root diameter was 0.17±1.33 mm/year in patients receiving ARB, and significantly higher – 2.0±0.81 mm/year in ACEI patients (p=0.04), and 3.0±1.58 mm/year (p=0.01) in patients without ARB or ACEI therapy.

Global systolic LV function was depressed in all groups (EFs Simpson: 53.1±10.3% in ACEI, 55.5±10.2% in ARB, and 58.6±8.0% in control patients, p>0.05 for all). No interaction was found between aortic valve morphology and aortic dilatation rate. Bicuspid aortic valve was identified in 18 patients (37.5%) with 1.0±2.9 mm/year aortic root dilatation, tricuspid – in 32 patients (62.5%) with 1.85±1.21 mm/year aortic growth (p=0.50).

Conclusions: In contrast to results of Marfan syndrome studies, additional ARB to beta-blocker therapy may have beneficial effect on aortic growth in patients with non-syndromic thoracic aortic aneurysm.

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Predictors of early and late complications after valve-sparing and total aortic root replacement surgery in patients with non-syndromic non-familial aortic root aneurysms

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Background: Ascending aortic aneurysm is a life-threatening disease, and the only way to manage patients with this pathology is surgery. Possible options are complex aortic root replacement surgery or aortic valve-sparing aortic root replacement. Despite the accumulated experience, some factors influencing the early and late outcomes remain unclear.

Purpose: To assess predictors of early and late complications after valve-sparing and total aortic root replacement surgery in patients with non-syndromic non-familial aortic root aneurysms.

Methods: From January 2009 to December 2015, aortic root replacement was performed in 253 consecutive patients. Patients with Stanford type A aortic dissections, connective tissue disorders, familial and inflammatory aortic aneurysms were excluded. The remaining 88 patients were divided into 2 groups: valve-sparing (David I) aortic root replacement (Group 1, 30 patients, males – 21, mean age 59.4±10.4 years, tricuspid aortic valve – 100%) and total aortic root replacement with a mechanical composite valved graft (Group 2, 58 patients, males – 55, mean age 53.5±11.2 years, tricuspid aortic valve – 39.7%, bicuspid aortic valve – 60.3%). We analyzed early and late outcomes.