dictor of mortality in patients with severe aortic stenosis (AS) and preserved LV ejection fraction (LVEF). However, its quantification using echocardiography may be subject to error measurement. The aim of this study is to determine the prevalence and impact on long-term survival of high Zva, purposely measured by cardiac catheterization.

**Methods and Results:** 768 patients with preserved LVEF (>50%) and severe AS (valve area ≤1cm²) underwent cardiac catheterization. Zva was derived from catheterization data and calculated using validated formula. Zva was considered high when > 5 mmHg/mL/M². Overall, high Zva was found in 42% of all AS patients.

Patients with high Zva were significantly older (p<0.0001), and more often female (p<0.0001), they had significantly smaller aortic valve area (p<0.0001), higher mean gradient (p=0.001), lower indexed stroke volume (p<0.0001) and cardiac output (p<0.0001), significantly higher LVED filling pressures (p=0.03), systolic pulmonary artery pressure (p=0.003), higher capillary wedge pressure (p=0.006), reduced systemic arterial compliance (p<0.0001), but higher systemic vascular resistance (p<0.0001).

Ten-year survival was significantly reduced in patients with higher Zva (50±5%) as compared to those with lower Zva (67±3%; p=0.01). After adjustment for all other risk factors, Zva was independently associated with reduced long-term survival (hazard ratio [HR] = 1.12 95% CI: 1.009-1.22; p=0.03). Of interest, high Zva remains associated with reduced survival as compared to low Zva in patients with normal LV stroke volume, but was no longer significant in low flow patients (>60mL: 49±8Vs. 69±4%, p=0.012; ≤60mL: 49±7 vs. 53±13%; p=0.96).

**Conclusion:** In this large cardiac catheterization-based study, high Zva estimated invasively is frequent in patients with severe AS, and appears as a robust and independent predictor of survival.

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**Marfan syndrome diagnosed during childhood: focus on cardiac events in the French database**

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**Life expectancy of patients with Marfan syndrome has increased, due to earlier diagnosis, better familial screening, regular follow-up (FU) and timely prophylactic aortic surgery (PASu). Incidence of aortic events in affected patients recognized during childhood is unknown.**

**Methods:** 465 patients with Marfan syndrome, diagnosed before 18 year-old between 1993 and 2013 were included in the French multicenter database. Cardio-vascular events (death, aortic dissection or PASu) were recorded.

**Results:** FU was complete for 69.5%. A cardio-vascular event occurred in 25 patients (5.4% 95CI 3.5-7.8%), including PASu in 12 (4.3% 95CI 2.5-6.2%), aortic dissection in 3 (0.6% 95CI 0.0-1.4%) and deaths in 2 (0.2% 95CI 0.0-1.0%). 16 events (64%) occurred before 19 year-old (Median 15.0, min 2.8, interquartile 11.7-16.3; PASu n=12, deaths n=2 and dissection n=2). One sudden death occurred in a 18 y.o. girl followed until the age of 14.3 under beta-blockade treatment. A 3.4 year-old boy with a FBN1 mutation diagnosed at the age of 1.1 died from respiratory distress and viral myocarditis.

An aortic surgery was performed in 23 patients (4.9%, 95CI 3.0-6.9%), including a Bentall procedure with mechanical aortic valve in 10 (43.5%), a valve sparing surgery in the remaining 13 (56.5%) and a supra-coronary graft in 4 (17.4%, dissection: n=2 and PASu: n=2). Mean age at the date of PASu was 17.1±5.5 year-old.

Events occurred before or at inclusion in the database in 8 patients (32.0%) (PASu n=5, dissection n=2, death n=1). Dissection was observed before inclusion in 2 patients out of 3 and during pregnancy in 1 patient aged 25 and lost of FU until 19 year-old. Kaplan-Meier survival estimate indicates that 95% of patients remained free from events at eighteen and 78% at thirty year-old.

**Conclusion:** Prophylactic surgery for enlarged aorta is the main cause of cardiac events in patients with Marfan syndrome diagnosed during childhood. A quarter of them have a cardiac event before thirty year-old (figure next page).