

Table S1: Medical treatment and doses in HTADs (order by year of publication).

Author	Type of aorthopathy	Design	Primary outcome	Age (mean)	Medical treatment/comparison	Mean dose	Ref
<i>Shores, J. et al.</i>	MFS	Open label randomized trial	ARD and clinical event	14.5-15.4	Propranolol vs placebo	212 +/- 68 mg given, four divided doses per day	1
<i>Yetman et al.</i>	MFS	Prospective, non-randomized, open-label	ARD rate of change per year; Aortic stiffness index and distensibility	12.8 ± 7.8	Propranolol/ atenolol/ enalapril	Propranolol: 1 mg/kg twice a day Atenolol: 25 mg twice a day Enalapril: 5 mg twice a day	2
<i>Selamet, T. et al.</i>	MFS	Observational, Retrospective	Rates of change in ARD. Cardiac complications (death, AD or CS)	9	Atenolol vs propranolol	0.92 mg/kg/day for propranolol Metoprolol: dose not reported	3
<i>Ong KT</i>	vEDS	Multicenter, randomized, open trial	Arterial events (rupture or dissection, fatal or not)	36	Celiprolol vs placebo	Dosage error in the original article.	4
						100 mg celiprolol, titrated to 200 mg, 300 mg, and 400 mg per day	4
<i>Chiu HH</i>	MFS	Randomized, open-label, active-controlled trial	ARD	13.1± 6.3	Losartan vs BB (atenolol or propranolol)	Losartan: start 0.7 mg/kg daily → 50 mg/day or the maximum tolerable dose BB: 50 mg of atenolol/day; propranolol twice daily for adults and 1 mg/kg/day for children	5
<i>Sandor, G.G. et al.</i>	MFS and LDS	Clinical trial randomized, double-blind study	Vascular function	17	Atenolol vs losartan	Atenolol: 25–50 mg Losartan 25 mg daily	6
<i>Mullen, M. et al.</i>	MFS	Placebo-controlled, double-blind randomized trial	ARD	18	Irbesartan vs placebo	75 mg for all, and after randomization, 300 mg once daily (as tolerated) vs placebo	7
<i>Frank et al.</i>	vEDS	Retrospective cohort study	vEDS-related events and deaths	34,5	Celiprolol vs usual care	≤400 mg/day	8
<i>Van Driest et al.</i>	Variants in ADRB1 and CYP2C9 genes in MFS	Randomized trial	Annual rate of change in the maximum ARD ( z-score)	11.5	Atenolol vs losartan	Atenolol: maximum tolerate dose. Daily dose max 250 mg Losartan: 1.4mg/kg (maximum daily dose 100 mg)	9
<i>Olfe, J. et al.</i>	Genetic aortopathies	Retrospective study	Aortic root dimension	8-10	Predominantly metropolol vs valsartan	Metropolol:1–2 mg/kg bw/day. Valsartan:1-3mg/kg bw/day	10

**Table Legend:** MFS, Marfan's syndrome; vEDS, vascular Ehlers–Danlos syndrome; LDS Loeys–Dietz syndrome; AD, aortic dissection; CS, cardiac surgery; CV, cardiovascular; ARD, aortic root dimension; BB, b-blockade; NA not available.

## References table

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