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Bicuspid Aortic valve in children with Marfan Syndrome: high appearance without correlation to severity of clinical symptoms.

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Introduction: Due to age dependent development of most pathologies in Marfan syndrome (MFS) estimation of clinical outcome in childhood is difficult. Bicuspid aortic valve (BAV) is correlated with MFS and prevalence is up to 4.7% in adult Marfan patients. Data in childhood about appearance and correlation with severity of phenotype is missing. Subsequently it may be a predictor for onset of pathologies and helpful to determine individual patient care. We hereby evaluated correlation of present BAV with other Marfan pathologies in childhood.

Methods: Since 2008 we investigated 395 patients (11.4±5.5 y) with suspected MFS. In 145 patients MFS was diagnosed, thereby 16 patients showed BAV. We retrospectively analyzed correlation of prevalence and age of onset of cardiovascular pathologies (Dilatation sinus of valsalvae (SV), Mitral valve prolaps (MVP)), systemic manifestation of Ghent Criteria and *FBN1* mutation with appearance of BAV.

Results: Prevalence of BAV in pediatric Marfan patients was 11.0 % whereas age of patients with and without BAV did not differ significantly. There is no correlation of BAV with prevalence or age of onset of pathologies of MFS. (Table1).

Table1: Correlation of Marfan pathologies with BAV appearance.

Pathology	**Prevalence**		**Age of onset[years]**		**P(Prevalence)**	**P(Age of onset)**
	BAV	**noBAV**	**BAV**	**noBAV**		
SV	12/16 (75.0%)	71/129 (67.8%)	11.8±1.5	9.5±0.7	ns	ns
MVP	11/16 (68.8%)	76/129 (58.9%)	11.1±1.7	9.8±0.7	ns	ns
Systemic manifestation	8/16 (50.0%)	62/129 (48.1%)	12.4±1.8	12.9±0.6	ns	ns
Ectopia lentis	3/16 (18.8%)	25/129 (19.4%)	7.2±1.8	9.1±1.1	ns	ns
FBN1	12/16 (75.0%)	88/129 (68.2%)	-	-	ns	-

Conclusions: In our large collective prevalence of BAV in MFS was surprisingly higher compared to adult. The reason is unknown, but can possibly be an effect of selection. Indeed, occurrence of BAV did not correlate with severity of clinical phenotype. Especially SV dilatation did not appear earlier or more often in BAV patients. Thus BAV is present frequently in MFS but is no predictor for clinical outcome and estimation of need for therapy. Development of pathologies in those patients in adulthood is unknown but may be relevant for course of disease. Good predictors for severity of phenotype in childhood are unfortunately still missing.