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From neonates to adults in Marfan syndrome- Diagnosis, progress and transition to adult care.

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Objectives:
Medical care for chronic diseases like Marfan syndrome (MFS) is a big challenge. Thereby long-term outcome and quality of life (QoL) are essential and highest aim for patients. Early diagnosis, due to improvement of genetics and clinical judgment, allows insights of disease development and possible preventive strategies in childhood. In consequence, patients require lifelong follow-up and need an effective transition to adult medicine. We hereby present more than 10 years of follow-up with a distinguish insight of all aspects of diagnosis and prophylaxis in MFS.

Methods:
Since 2008 we investigated 424 patients in our specialized pediatric Marfan clinic whereas in 149 MFS was diagnosed (9.5±5.8y; m:62%) according Ghent Criteria (GC). We attended 978.5 patient years. We analyzed criterions of GC, requirement of prophylaxis and QoL in children and adults and evaluated effectiveness of transition to adult care.

Results:
Analysis of GC showed age dependent increase of prevalence (Table1). Medical prophylaxis was initialized in 80 patients (<18y 52%; >18y 62%; 10.2±5.1y). Significant z-score reduction could be achieved with sartan and/or betablockers (BB). Seven patients required aortic root replacement (11.5±5.6y) in childhood. There was no dissection or mortality. Compared to adults QoL did not show significant impairment in children. From 34 adults 59% were accompanied to adult care, 35% are still in pre-transition section, 5% are lost in follow-up.

Table 1: MFS Pathologies in children/ young adults.

Pathology	**Prevalence(<18y)**	**Prevalence(>18y)**	**P**
Sinus valsalvae	73/115 (63%)	24/34 (70.6%)	ns
MVP	59/115 (51%)	19/34 (55.9%)	ns
Systemic manifestation	54/115 (46%)	18/34 (52.9%)	ns
Ectopia lentis	21/115 (18%)	8/34 (23.5%)	ns
Dura ectasia	37/69 (53%)	19/31 (61.2%)	ns

Conclusions:
Age dependent onset of symptoms remains a challenge in MFS. Especially onset of cardiovascular abnormalities in young adults is very dynamic. To prevent complications early medical prophylaxis is effective and essential in childhood (Sartan and/or BB). Lifelong follow-up examinations need to be standardized and a safe cooperation of pediatric cardiologists and adult cardiology is important. Patients in transition progress require special attention, assistance and a close contact to the patient and family. Finally, success of transition needs to be reevaluated before pediatric patient care ends.