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Angiotension II Inhibitor therapy in pediatric patients with Marfan-Syndrome (MFS) - Update after six years of experience with particular regard to tolerability

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Introduction: MFS is an inherited connective tissue disorder in which aortic root dilatation remains the significant indicator for morbidity and mortality. After recent publication of the pediatric multicenter study and publication of our mono-centric data concerning the effectiveness and tolerability of therapy with ARB vs. beta blocker (BB) we would like to present an actual update after six years of experience.

Methods: We identified 126 children with confirmed MFS. Indication for prophylaxis was found in 64 patients and was subjected to standardized diagnostic program including echocardiography, MRI and clinical examination. We examined the effectiveness of therapy with ARB (n=44) on the growth of the sinus valsalvae (SV) with comparison of z-scores of SV before treatment and during follow up and compared these data to patients treated with BB (n=20) only.

Results: Treatment by ARB and BB leads to significant reduction of SV dilation ($p < 0.05$). The deviation of SV enlargement from normal as expressed by the rate of change in z-scores was significantly reduced by a mean difference of -0.57 ± 0.65 z-scores ($p < 0.05$) under ARB therapy and by a mean difference of -0.42 ± 0.55 z-scores ($p < 0.05$) under BB therapy. The effect of ARB and BB on aortic root dilation is similar in both groups ($p > 0.05$). Therapy with BB was discontinued in 20% (4/20). No discontinuation in the ARB group (0/34).

Conclusion: The prophylactic effect of ARB and BB in pediatric patients with MFS is similar but tolerability of ARB is clearly superior. In addition inhibition of TGF- β signaling by ARB which is supposed to contribute to the pathogenesis of MFS has to consider.

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Risk management & patient safety in pediatric and adolescent surgery

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How to increase patient safety and quality of care, or how to implement an effective risk management in our health care setting, are one of the most burning questions currently discussed within the public, among politicians and the medical community. Globally, first aspects have been founded in the Millennium Development Goals (MDG) of the year 2015 or in the Global Initiative for Emergency and Essential Surgical Care (GIEESC) in 2008. In addition, guidelines and recommendations for such an institutional and clinical risk management in surgery have been issued in first world countries, like Germany, by professional risk managers, patient safety experts and medical chambers. By tradition, they focus more on adults than on children, and of course, more on public health issues than on surgery. In this communication well known patient safety and risk management goals are re-focused and proposals for their adaptation for the specific needs in pediatric and adolescent surgery are made. It has been especially considered, if safety tools and the safety culture out of the aviation sector or other High Reliability Organizations (HRO) are already have been implemented or could be implemented in the near future. The expertise of the author and his all day clinical practice are discussed in front of a literature review to define our current standing and possible impact for the future. Finally, we will conclude, if our evaluation is sound enough to serve as a basis for a commonly accepted accreditation tool in pediatric & adolescent surgery.

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