MP3-16

Health Related Quality of Life in Pediatric Patients with Marfan´s syndrome

Steiner K.J., Wild J.M., Stark V., Mueller G.C., Arndt F., Weil J., Mir T.S.
Clinic for Pediatric Cardiology, University Heart Centre Hamburg-Eppendorf, Germany

Introduction:
The “Health Related Quality of Life” (HRQOL) is a term that became increasingly important during the last 15 years. It is not yet known however if children with Marfan’s syndrome are particularly impaired in different aspects of their Quality of life (QoL). We hypothesized that the QoL of patients with MFS is age-dependent due to increasing clinical features, and, compared to healthy children, reduced due to skeletal abnormalities, aortic root dilatation with consecutive need for drug therapy and regular medical checkup.
Understanding the relationship between subjective QoL in children with MFS has important implications for preventive care strategies that may impact the outcome of HRQoL.

Methods:
220 children between 4-16 years were included in this study. 46 patients (4-16 years, mean age 10.8 years) of our outpatient clinic completed the KINDL-R survey to assess life quality. Patients were included if MFS was determined genetically or clinically according to Ghent criteria, 39.1% were female, 60.8 % male. Additionally data of 174 kindergarten and school children were collected (4-16 years, mean age 10.9 years), 48.8% were female, 51.1% male. Samples were divided into 4 age groups and subjective QoL was compared. Additionally we created subgroups based on the main diagnostic criteria of the revised Ghent nosology.

Results:

QoL total score	4-7 years	MFS 77.6 (95% ci 70.9-84.3)	control group 77 (95% ci 73.1-80.8)	p>0.05
QoL total score	8-11 years	MFS 75.3 (95% ci 68.5-82)	control group 74 (95% ci 70.8-77.1)	p>0.05
QoL total score	12-16 years	MFS 75 (95% ci 70.9-79.1)	control group 68.4 (95% ci 66.2-70.6)	p<0.05
QoL total score	8-16 years	MFS 75.1 (95% ci 71.5 –78.8)	control group 70.4 (95% ci 68.6-72.3)	p<0.05
QoL total score	8-16 years	MFS patients with lens luxation 81.6 (95% ci 75.2-88),	control group 70.5 (95% ci 68.6-72.3)	p<0.05

Conclusions:
We report, that pediatric MFS patients showed partially higher HRQoL than children in the control group. MFS patients aged 12-16 years showed significant higher QoL than children in the control group although clinical features develop gradually. Analysis on different subgroups based on Ghent nosology confirmed these results. The results suggest that children with MFS experience increased QoL compared to children without this genetic predisposition. It can be speculated that parents may facilitate a stable social environment in the face of a children’s disease. Furthermore children with MFS may be schooled through their environment to engage in more positive coping skills that increases their subjective experience of life quality.