

EXTENT: A COMPLETELY NEW METHOD OF TREATMENT OF PATIENTS WITH MARFAN SYNDROME

Jan Pirk, Kočková R, Krebsová A and Malý J

IKEM, Czech Republic

Marfán syndrome is the most common genetic disorder of connective tissue. One complication that threatens the lives of patients is progressive dilatation of the ascending aorta with development of aortic valve regurgitation or the emergence of dissection, often leading to sudden death. Until now, these patients were operated only after dilatation of the ascending aorta causing hemodynamically significant regurgitation of the aortic valve. The surgery consisted of the replacement of the ascending aorta and aortic valve or valve sparing procedure. This new method is a preventive operation. The method involves creating a custom made external support of the root and the ascending portion of the aorta. Based on the CT examination, prosthesis Extent is created. The surgery is performed from the longitudinal median sternotomy without cardiopulmonary bypass. Entire aortic root is dissected to its origin from the left ventricle, ostia of the coronary arteries are encircled, the prosthesis is pulled underneath and fixed to the root and then sutured longitudinally. It is interesting that the prosthesis was developed and as the world's first has it sewn on himself (Mr Tal Goleworthy), 13 years ago in Oxford. Neither him nor the other 100 patients operated in this department with this disease had dilation or dissection throughout the study. It is because the prosthesis grows over time into the aortic wall thereby enforces it while maintaining the elastic properties of the wall. At our institute, we have so far experience with operations of 20 patients, with good results. This operation moves the care of patients with Marfan syndrome to qualitatively higher level

Biography

Jan Pirk has completed his Graduation from the Faculty of General Medicine, Charles University in Prague. After completion of his Graduation, he worked at the District Hospital in Nymburk from 1972 until 1974. He has been working in the Institute of Clinical and Experimental Medicine (IKEM) from 1974 until now. From 1991 to May 2017, he was the Head of the Clinic of Cardiovascular and Transplantation Surgery and since 1995 he has been the Head of the Cardiac Centre. In 1990-1991, he worked as a Consultant at Odense University Hospital in Denmark. He is a Member of a number of national and international scientific organizations. In his free time, he is mostly engaged in sports and likes theater.