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EDITORIAL

"The sciences alone, however, whether natural or physical, are not sufficient to understand the mystery contained within each person"

His Holiness Pope Francis, European Society of Cardiology Congress, Rome, Italy 31st August 2016

Dear Readers,

Unlike to previous Editorials, let's start with the last article in this new issue of the Journal – Report form the European Society of Cardiology Congress that took place in the last days of August in the everlasting city of Rome. Just a few days ago, we witnessed an unprecedented event in the whole history of not only European but world cardiology. It was that His Holiness Pope Francis, who is known for extraordinary gestures, attended Fiera di Roma – the venue where the annual European Society of Cardiology Congress has been held for several days. In the very emotional and yet sensible words, Pope Francis expressed His admiration for all health-care professionals for all their devotion for the sick and poor. Moreover, Pope Francis addressed particularly scientists with their ongoing quest for truth. Along the same line of thinking, Pope Francis reinforced the ultimate purpose of science that should be an improvement of care delivered to every human-being. Those wise words shall accompany us with all our daily struggle!

Coming back to the current issue of the Journal, there is a Review article on cardiac amyloidosis and Original article about exercise capacity in patients after Fontan operation. Moreover, as usual the middle part is composed of four exceptional clinical cases presented and commented by the managing teams. The Journal ends with the report from the annual European Society of Cardiology Congress that took place in August in Rome, Italy.

The state-of-the-art Review on cardiac amyloidosis has been authored by doctor Olga Kruszelnicka and colleagues from the Department Coronary Artery Disease, located in our hospital. The authors present most up-to-date and comprehensive facts on epidemiology, pathology, clinical presentation, diagnosis, risk stratification and therapy in cardiac amyloidosis. From the cardiological point of view, the authors describe in details the typical ECG and echocardiographic findings. Moreover, the particular attention was be paid to non-cardiac "red flags", including periorbital purpura, macroglossia, and carpal tunnel syndrome. The authors expertly describe the modes of treatment, including typical cardiac medications and as well as multi-drug non-cardiac therapy. Obviously, chemotherapy is reserved for hematology wards, nevertheless, cardiologists should be familiar with those complex regimens, especially in the view that those kinds of treatment carry the risk of subsequent cardiac adverse effects. Although rare, cardiac amyloidosis is a very serious medical condition with poor outcome if left untreated, therefore, as cardiologists we should stay vigilant and look for "red flags" that greatly help with the diagnosis. Therefore, this elegantly written review accompanied with a set of informative tables is definitely a considerable help and must-to-read.

Doctor Monika Smaś-Suska, under the supervision of dr Lidia Tomkiewicz-Pająk who is an expert in the field of grown-ups with congenital heart diseases (GUCH), published an original study on exercise capacity of patients long after Fontan operation. The authors recruited an impressive number of 37 patients after Fontan operation in whom they performed cardiopulmonary exercise tests to objectively study exercise tolerance. Depending on the time from the index procedure, the patients were divided into tertiles. Surprisingly, peak oxygen uptake and its derivates and peak heart rate were not different between groups, however, values of ventilatory equivalent for carbon dioxide were

significantly higher (indicating worse functional performance) in those patients who had the longest period of follow-up. The authors expertly explain their results and confront their findings with already accumulated knowledge in the informative discussion section.

And now comes the "salt" of the Journal in the form of four clinical cases of rare cardiovascular diseases. As the Journal gradually expands, this time we have three cases from the outside and only one case from our center. First case that is authored by doctor Marta Nowakowska and colleagues comes from pulmonary hypertension unit in Łodz. The authors describe their initial clinical experience with Macitentan that is a novel dual endothelin-receptor antagonist. The patient with pulmonary arterial hypertension who has been treated for a long time with Bosentan, an oral endothelin A and B receptor antagonist, developed a drug-induced hepatitis that is a well-documented complication of this therapy. The authors describe the course of this serious complication, which was eventually relived with drug withdrawal and initiation of Macitentan. The second case, authored by doctor Jacek Kuźma et al., comes for the pediatric center in Krakow and is about long-term observation of the cyst attached to the anterior mitral leaflet. At first, at the age of 6 years old the boy was completely asymptomatic and the managing physicians recommended the conservative approach. However, more than ten years later, the child developed minor symptoms of left ventricular outflow obstruction and the cyst doubled in size. Although, after long discussion the decision was made to proceed with the operation, the parents did not agree for the operation. In the short but well-written review of the existing literature, the authors comment on their strategy. The next case that is authored by doctor Sebastian Gurba and colleagues comes from Rzeszow in the south-eastern Poland. This case is truly exceptional as the authors describe an elderly female with pulmonary embolism with foreign material – cement used for previous percutaneous vertebroplasty. Probably only a few us would think of such an unusual source of pulmonary embolism, therefore, the authors should be congratulated twice. Once for confirming the diagnosis and for the second time as they shared their findings with us and in that way rise of awareness. Last case comes from our center and is authored by doctor Agnieszka Sarnecka et al. The authors describe very rare anomaly of pulmonary atresia with ventricular septal defect and subsequent development of major aortopulmonary collaterals (MAPCAs) that are primary responsible for supplying pulmonary circulation. The managing physicians report in details the results of step-by-step diagnostic approach and present high-quality diagnostic figures. After Heart Team discussions, the decision was made for conservative management and regular follow-ups.

The final part of the this issue of the Journal is the report, written by doctor Pawel Rubis, from the aforementioned annual European Society of Cardiology Conference.

We believe that also this time, the Readers find the current issue of the Journal interesting and worth-reading.

Piotr Podolec
Editor-in-Chief
Journal of Rare Cardiovascular Diseases