

“There is no failure except no longer trying” Elbert Hubbard

Dear Readers,

The way of time fly is inevitable and the end of 2015 is slowly approaching. Not long ago we were celebrating its onset but now it is time for a farewell. This time of the year is just perfect for a few reflections. Firstly about this Journal. After obtaining the Copernicus Index earlier this year, we have made some progress but yet we are impatiently awaiting another decision from the Ministry of Science regarding formal recognition of the Journal in their database. We know that our march will be long but every milestone is a source of joy and extra strength. We believe in newly-started initiative of early education of rare diseases in medical college. The curriculum has been officially approved and our first students have been already thought basic aspects of rare diseases. The idea planted several years ago starts slowly to blossom as more and more physicians are aware of rare diseases. It is reflected by the growing number of hospital and outpatients referrals. Obviously, this is all we wanted, isn't it? However, all those and other initiatives are not paralleled with the formal help from the governing bodies, including reimbursement of costly procedures, medications or long hospital stay. We strongly believe that the incoming year 2016 will bring us not only extra work but also extra understanding and funding from the authorities.

The 13th issue of the Journal traditionally begins with the informative Review on the tricky topic of cardiopulmonary exercise testing (CPET). For decades CPET has been a gold standard for objective assessment of functional capacity, not only in heart failure area but also in other fields, such as congenital heart diseases or cardiomyopathies. The optimal performance but more importantly interpretation of CPET results requires in-depth knowledge of exercise physiology both in healthy and diseased subjects. To distinguish cardiac or non-cardiac source of disprove or exercise intolerance can be tricky and not necessarily straightforward. Relying on few CEPT parameters may not be optimal in every case, therefore constant education and discussion is of paramount importance. The combination of

rare cardiovascular diseases and CEPT poses difficult yet possible to overcome challenge. Hopefully, after reading this interesting paper, this topic will be more friendly-user.

Since the beginning, this Journal tackled difficult and controversial topics. Therefore, the Original Article authored by doctor Monika Komar and colleagues from our Centre, on the safety, feasibility and efficacy of trans-catheter closure of secundum atrial septal defect (ASD) in elderly patients is another perfect example of such approach. There are numerous pros and cons regarding closure of ASD in the elderly. As the controversies exist and the debate is far from over, those who feel up to the task have their say. Doctor Komar presents high-quality data on 15 consecutive patients over 70 years old who had successful ASD closure in our Centre. Not only the procedure was successful in all subjects without any major complications but more importantly provided long-term improvement in all three aspects of clinical symptoms, cardiac morphology and function, and quality of life. These findings are reassuring and once again confirm the truth that age alone cannot simply disqualify from any procedure per se.

Clinical cases of rare cardiovascular diseases are the cornerstones of this Journal. Likewise, the previous issues, there are four exceptional cases that are presented and discussed in-details by the managing teams. All cases are presented in the context of current literature and provide an accurate view on the commented topics. First two cases come from our long-term collaborators in Bialystok in eastern Poland. In the first article doctor Katarzyna Ptaszyńska-Kopczyńska and colleagues present a series of four patients with severe pulmonary arterial hypertension (PAH) with significant component of lung diseases. After exhaustive diagnostic work-ups, which are perfect examples of how patients with suspicion of PAH should be managed, the authors concluded that those patients despite significant hemodynamic imbalance and lung disorders, have PAH out-of-proportion. Unfortunately, the of-

ferred complex and costly treatment to those patients did not have much effect on their symptoms. In the interesting Discussion, the authors comment on their management strategy and describe the possible sources of problems. In her second case doctor Katarzyna Ptaszyńska-Kopczyńska et al. describe equally interesting topic of rough clinical course, leading ultimately to premature death, of a middle-aged woman suffered from AL-amyloidosis. Sadly, shortly after the correct diagnosis has been made and before initiation of chemotherapy the patient deceased. The authors expertly discuss this case in the view of the current literature in the Discussion section and provide explanations why the diagnosis of amyloidosis is frequently made (too) late. The third case comes from our Centre and is authored by doctor Agnieszka Żygadło and colleagues and is about a rare case of constrictive pericarditis. The managing physicians found and described all characteristic features of constrictive pericarditis, therefore, this text has high educational value. Although the ultimate treatment of severe, drug-resistant constrictive pericarditis, is surgical, this procedure is difficult and associated with high mortality due to uncontrolled bleeding complications, which took place in this poor patient. The last case, authored by professor Zbigniew Gąsior et al. from Katowice, focus on arrhythmogenic right ventricular cardiomyopathy (ARVC) complicated with thrombus. On the contrary to the Mediterranean countries, ARVC is infrequently diagnosed in Poland. Therefore, every case coming from our background is valid and worth reading. The presented young male

had all necessary features to clearly diagnose ARVC. Moreover, there was thrombus present in the right ventricle, which fortunately was dense and did not cause embolic complications. The short Discussion section underlines typical characteristics of ARVC.

This issue of the Journal is closed by the report, written by doctor Pawel Rubis, from the 12th Meeting of the Myocardial and Pericardial Diseases Working Group of the European Society of Cardiology that took place in October in Florence. The annual Working Group meeting is an important and influential event for those particularly interested in the field of cardiomyopathies.

We hope that the current issue of the Journal is at least as good as the previous ones and will be warmly welcomed in our community. However, as humans we are aware of our imperfections and if You find anything that needs to be separately commented or improved, please do not hesitate but contact us at your earliest convenience.

And finally, on behalf of the Editorial Team of the Journal of Rare Cardiovascular Diseases, I would like to wish all our Readers and Families Merry Christmas and a Very Happy and Prosperous New Year 2016!!

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Piotr Podolec
Editor-in-Chief
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