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EDITORIAL

"One of the greatest disease is to be nobody to anybody"

Saint Mother Teresa of Calcutta

Dear Readers,

Before we go any further, try to ponder the quote once said by the Saint Teresa of Calcutta. She was by far one of the most entitled person in the world to say such an unpleasant truth. Certainly, for her entire life she had been working in extreme conditions in rural and underdeveloped areas in India. In the lands where bowl of hot soup, slice of whole-meal bread and clean bed sheets were unobtainable and priceless commodities. But is that all? No, she perfectly knew that above all, recognition of every human-being is the crucial thing. Those poor who were cared by her and her convention, could easily understand they are not treated as applicants but as subjects. This difference is of paramount importance and especially applies to the contemporary medicine. In the era of wealth, insurance, procedures, reimbursements, etc., we no longer spend a minute to see the patient in front us as a whole. Some would immediately start to think what is the best arterial access (either femoral or radial), some would argue that magnetic resonance is superior to echocardiography, others would say what guidelines recommend… It is a bit sad, indeed! In most cases, we really do not know what the true patient's needs are. As a poor excuse, we focus our attention on tiny fragments of patient's body and boldly claim that we cured him. Too often we try to fix one small body part (sometimes with too much risks and costs both from the patients and health service). How naïve it is! Then we send the patient home, as quickly as possible, and are surprised to learn that the patient is not feeling so great in the follow-up visits. Why is that? One might say, he needs one more procedure and it may be true but in the majority of cases, the patient needs our recognition, he wants us to see him as a human not as a diseased valve, atherosclerotic vessel or infarcted myocardium. Sometimes the honest conversation is much better that repeated (painful) examinations or procedures. Obviously, we cannot change our habits in a day but maybe it is a time think once more about the patient as a subject not an applicant.

In this last issue of the Journal in 2016, majority of articles is dedicated to inherited cardiac disease. We start with an interesting Review on ophthalmic manifestations in Kearns-Sayres syndrome. Further, there is the Original paper on long-term observation following Ross operation. Obeying our policy, the middle part is composed of four clinical cases of rare cardiovascular diseases that are presented in details and commented by the managing teams. In line with the foreword, this issue ends with the report from the joint meeting of patients with pulmonary hypertension and their families with managing physicians.

The Review article on ophthalmic manifestation in Kearns-Sayres syndrome is a little exotic for most of the readers as our ophthalmologic knowledge and skills are probably a little rusty. Thus, this is an extra opportunity for an refreshment. The authors focus on rare mitochondrial cytopathy, namely Kearns-Sayres syndrome. As any mitochondrial disease, also Kearns-Sayres syndrome is a multi-systemic disorder with the frequent cardiac and ophthalmic involvement. From the cardiologic point of view, the most frequent features are various conduction abnormalities. On the other hand, from the ophthalmologist perspective, the most common are: ptosis, chronic progressive external ophthalmoplegia, and pigmentary retinopathy. Those pathologies are described in details. The key message of the article is to rise suspicion of Kearns-Sayres syndrome in young patients with otherwise unexplained conduction disturbances and ophthalmologic problems.

Doctor Aleksandra Lenart-Migdalska, under the supervision of dr Lidia Tomkiewicz-Pająk who runs the service for adult patients with congenital heart diseases, studied the group of nine patients who underwent the Ross operation in childhood due to congenital aor-

tic valve disease. The authors focused on the long-term follow-up of those patients. Although, the Ross operation offers numerous advantages compared to aortic valve replacement, nevertheless, it also carries the risk of long-term serious complications. It was observed that majority of patients had some degree of post-operative problems, including dilatation of the aorta and pulmonary valve stenosis or regurgitation. Unfortunately, almost one-third of patients are currently considered for a reoperations, which is undoubtedly worrisome as redo-operations are always more risky and difficult then the index procedure. The high incidence of long-term serious complications puts into question the value of the original Ross operation. However, what is known for sure all those patients should be regularly monitored in the specialized centers.

Obviously, the cornerstone of the journal are cases of rare cardiovascular diseases and this issue is not an exception. The first case, that is authored by doctor Jacek Kuźma, comes from the pediatric cardiac center in Krakow. The authors present the complex case of the young woman with D-transposition of the great arteries, inflow ventricular septal defect and sub-pulmonary stenosis who developed pulmonary hypertension (PH) and eventually Eisenmenger syndrome. The patient underwent several palliative operations to be eventually stabilized with PH-specific medical therapy. This is a very interesting and well-written paper that surly needs further attention. The second case is written by doctor Monika Smaś-Suska and colleagues and touches the problem of unusual reason for supraventricular tachycardia, that turned out to be ruptured aneurysm of sinus of Valsalva. This is a perfect example of the importance of high-quality echocardiograms that enabled the correct diagnosis and further surgical treatment. The operation and recovery were uneventful and the patient was discharged being truly cured. The third case is authored by doctor Piotr Liszniański et al. from our long-term contributors in Chrzanów. The article is about the middle-aged male who following inferior wall myocardial infarction had ICD implanted as primary sudden cardiac death prevention. As we all have learned, ICD is not an universal panacea and sometimes it creates more problems than good. Over the years, the patient suffered from numerous adequate and inadequate ICD interventions. The authors describe in details their management strategies that eventually proved successful. Undoubtedly, the strong part of the paper is well-written and informative discussion. The last case is again from our center and is authored by doctor Katarzyna Holcman and colleagues. The title of the article is a little provocative – "Hypertrophic cardiomyopathy or hereditary hemochromatosis?" but the authors elaborate in details on the diagnostic pathways that were applied and eventually all becomes clear.

This issue of the Journal is closed with the report, written by doctor Jakub Stępniewski, and entitled "Highlights from the Polish Pulmonary Hypertension Patients and their Families Meeting", an event that took place this autumn in Krakow. The meeting organizers should be surly congratulated for their effort and enthusiasm that are crucial in making such ideas happen. Such meetings may not be always easy as physicians may be confronted with high patients' and their family's needs and expectations regarding treatment. Not always such problems are easy to solve. On the other hand, we should always try and put our patients first. Finally, coming back to our motto, patients will surly appreciate the extra recognition delivered by doctors.

It is hoped that the readers of this issue of the Journal will find it of great interest.

Lastly, please accept our greetings as the holiday season approaches soon. Best wishes for a Merry Christmas and prosperous New Year 2017!!

Piotr Podolec
Editor-in-Chief
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