Percutaneous atrial septal defect closure in patients with pulmonary hypertension

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To the editor: We read with great interest the article by Balint and coworkers,[1] in which they reported the results of catheter closure of an atrial septal defect (ASD) in 54 patients with associated pulmonary arterial hypertension (PAH). The authors described, on the one hand, successful closure and a decrease in pulmonary artery pressure and New York Heart Association (NYHA) functional class at 1 year follow-up in the majority of patients. On the other hand, more than half of the patients had persistent PAH (moderate or severe) after closure and two patients died during follow-up. The authors' interpretation of these data is that catheter closure of ASD in patients with PAH is successful and has a good outcome. In our opinion, however, this conclusion is not justified and we feel that it is important to add some considerations on PAH associated with congenital pulmonary-to-systemic shunts, in order to put the interpretation of the data, as presented by the authors, in a broader perspective.

PAH in congenital heart defects associated with a systemic-to-pulmonary shunt, including ASD, is a progressive pulmonary vascular disease. Its progressive course is characterised by a reversible phase early in the disease and a progressive, irreversible phase in the advanced stage of the disease.[2] When the heart defect is adequately corrected in the early, reversible phase of the pulmonary vascular disease (either surgically or by catheter intervention), the PAH will disappear and remodelled pulmonary arteries will normalise.[1 2] However, when the heart defect is corrected late, in the advanced stage of the pulmonary vascular disease, and characteristic vascular lesions have developed, including concentric laminar intimal fibrosis and plexiform lesions, the pulmonary vascular disease will be not only irreversible, but will also progress in time despite closure of the shunt.[2 3] In this latter situation, closure of the heart defect will eventually lead to deterioration of the clinical condition and a decreased survival in these patients. This is because when right ventricular failure occurs the defect can no longer serve as an escape mechanism to maintain cardiac output at the cost of cyanosis, as is the case in Eisenmenger's syndrome.[3 4]

The considerations above lead to two important comments on the data presented by Balint and coworkers and to the interpretation of these data by the authors. The first question that should be asked in a patient with ASD and PAH, before attempting to close the defect is: How far has the pulmonary vascular disease progressed in this patient? The authors provide no data about assessment of the progression of the pulmonary vascular disease in the patients studied. Determination of the acute response to pulmonary vasodilator testing, with nitric oxide, oxygen or other agents, has been generally used to assess this progression in patients with congenital heart disease and to assess the possibility of closing the defect.[3] Although it is recognised that there is a grey area in which it may be difficult to distinguish between reversible and irreversible disease, most patients with advanced pulmonary vascular disease can be identified, in whom closure of the defect will have a detrimental effect in time.[3]

The second comment relates to the suggestion in the paper that a decrease in pulmonary artery pressure early after closure of the ASD is reassuring and can be regarded as a successful outcome of the procedure for the patient. In our opinion this is definitely not the case. If an increased pulmonary blood flow is diminished by closure of a heart defect, the pulmonary artery pressure will always decrease, independently of pulmonary vascular resistance. This will also occur in patients with advanced, progressive, pulmonary vascular disease. In these latter patients, however, pulmonary artery pressure will gradually increase again in parallel with the progression of the vascular disease, and eventually the outcome will be worse than that with an unclosed defect.[4]

In their paper, Balint and coworkers report that PAH was still present after 1 year in more than half of their patients. In the light of the considerations described above, one might ask whether the conclusion of the authors, ''Transcatheter closure in patients with secundum ASD and PAH is associated with good outcomes'', is justified. We think it is not: there are concerns about the outcome of these patients at long-term follow-up (which is more than 1 year) compared with similar patients in whom the ASD is not closed. We do agree completely with the authors that follow-up studies in this respect are needed.

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The authors' reply: We thank Dr Berger and colleagues for their interest in our paper and their important comments. We would first like to clarify the results of the study. We reported that 57% of patients with pulmonary arterial hypertension (PAH) did not have complete normalisation of right ventricular systolic pressures (RVSP <40 mm Hg as measured by echocardiography) in late follow-up. Although six of the patients had persistent severe PAH (range of RVSP 62–89 mm Hg), the majority of the patients with residual PAH had only mild PAH (range of RVSP 40–49 mm Hg). Second, the two late deaths that occurred were not clearly related to the intervention; one patient had a bowel obstruction and one patient with a previously undiagnosed clotting disorder had multiple pulmonary emboli. For the latter patient, it is difficult to know if the outcome would have been different, either better or worse, if the atrial septal defect had not been closed. Thus, we felt that overall, the outcomes at this duration of follow-up were reassuring for many patients.

Their letter deals with a number of important aspects of the interpretation of our results. First, and perhaps not highlighted adequately in our paper, we had an obvious selection bias in our patient group. Only patients felt by the interventional staff to be optimal candidates for transcatheter closure and who underwent device closure were reported on in this study. Furthermore, at our centre, patients with Eisenmenger's syndrome (intracardiac or extracardiac shunts with right-to-left shunt flow) are not considered candidates for device closure and therefore the conclusion reported in this study would not pertain to that specific subgroup of patients with PAH.

Second, Dr Berger and colleagues discuss the importance of cardiac catheterisation in understanding the mechanism of this disease process. We agree, but these data were not available for most patients with milder forms of PAH as this group of patients does not routinely undergo a complete invasive haemodynamic assessment before device closure at our centre.

Finally, Dr Berger and colleagues discuss the potential for adverse outcomes with additional follow-up. We agree that the condition of some of these patients may deteriorate over time with the loss of an