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Atrial septal defect: waiting for symptoms remains an unsolved medical anachronism

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This editorial refers to ‘Benefit of atrial septal defect closure in adults: impact of age’, by M. Humenberger et al., on page 553

During the past decade a ‘more modern’ natural history of atrial septal defect (ASD) in adults has been written, but still the ASD remains by far the most underdiagnosed congenital heart disease in the adult age group. The moment of diagnosis within the adult population is mostly tantamount to the initiation of treatment. Due to the superiority of closure over medical treatment alone,1 the consequence is closure either by intervention in selected patients or by surgery for all unselected or at least for the remaining patients not treatable by intervention. Meanwhile supported by direct comparison of the two methods, interventional treatment in selected patients has been accepted as the standard treatment and the treatment of choice due to its lesser invasiveness and lower complication rate.2,3 The study by Humenberger et al.4 shows nicely in a relatively large cohort of adult patients with an age range of 20–80 years that the defects were able to be closed successfully by intervention regardless of age, with excellent results and no significant complications. New York Heart Association functional class improved in almost all patients and rapid right ventricular remodelling occurred with a statistically significant improvement in right ventricular measurements and a decrease in pulmonary artery (PA) pressure levels in those with formerly elevated pressures and resistances. The authors again affirm the findings of numerous publications during the past decade and also uniformly conclude that the best outcome is to be expected if patients are free of symptoms at the time of treatment.5–7 The authors selected their patients rather conservatively in that they excluded patients with defects > 36 mm and those with multiple defects and/or atrial septal aneurysm, even though interventional closure may also be successful in these patients, and they should not be excluded if an interventional treatment option may be evaluated.8,9 However, even though presenting excellent data the authors fail to clearly highlight the seriousness of this disease despite their results containing definitely supporting data (Figure 1).10 The majority of patients remain free of symptoms up to their mid-forties or the beginning of their fifties and the onset of symptoms is proved to be a function of time expressing the duration of volume load (Figure 2). By at least 50 years and older patients became clinically symptomatic and show a higher incidence of atrial tachyarrhythmia (Figure 3) and increased PA pressures.11–13 This demonstrates again that as a matter of the permanent persistence of the left-to-right shunt and therefore the volume load of the right ventricle (RV) and the pulmonary vasculature the disease advances to a higher severity code. Both the surgical and the interventional treatments seem to be comparable with regard to their results, if the patients undergoing interventional closure have been strictly selected within the limits of the interventional methods. Even though closure of the ASD seems unable to prolong life expectancy significantly on the one hand, or to restore sinus rhythm, especially not in older patients suffering from atrial fibrillation, on the other, the improvement in symptoms and the right heart load conditions justifies closure at any age due to a significant improvement in quality of life.14 The authors omit also to report the importance of the concomitant left ventricular remodelling that underlines the close RV–left ventricle (LV) interaction also in this relatively uncomplicated congenital heart disease. With ongoing age beyond 60 years the restrictive properties of the LV myocardium with increasing diastolic stiffness remain underestimated in some patients and may be totally masked by the presence of an ASD.15 This diastolic stiffness, if not to be called dysfunction, may also eventually be a reason for increased pulmonary vascular pressures and resistance in the elderly. Fortunately almost all of them show a decrease in PA pressure levels and pulmonary vascular resistance after closure of the ASD. This may be due to the disappearance of the shunt alone but also to the remodelling and adaptation of the LV, which follows the same time course as those of the RV, and may be supported by the delayed and sometimes prolonged time course of normalization of PA pressures. Although the authors report a relatively large cohort of patients successfully treated by interventional closure of the ASD, there are still limited data available on surgical or catheter closure of ASDs in older patients.

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particularly in those with chronic atrial fibrillation. However, all studies have been able to show that even in these elderly patients closing the defect is not only feasible and safe but also leads to an impressive improvement in haemodynamic measurements and adaptation of both the right and left ventricles. The key issue of the dramatic improvement in functional class of nearly all patients is the disappearance of the volume load, although almost all of the patients with chronic atrial fibrillation seem to remain in atrial fibrillation even after closure, at least those above the age of ~60 years. Knowing the importance of a sinus rhythm with regard to quality of life and exercise capacity we still lack data on whether the surgical approach combined with a MAZE procedure...
may potentially be superior in these patients. The situation for patients suffering from paroxysmal atrial tachycardia remains even more unclear. There are limited data suggesting that these patients may benefit from closure also with regard to prevention of arrhythmia, but in the era of electrophysiology and all the ablational options we do lack data on whether an ablational treatment should be performed before closing the ASD because access to the LV after implantation of the device may be complicated, if not impossible.16 We also lack information on whether the incidence of atrial tachyarrhythmia that could probably benefit from ablational procedures may also influence our judgment on the implantation of large devices leading to difficult access to the left atrium in the long term. Nevertheless, in the current era the interventional approach for selected patients remains the method of choice, but all efforts have to be undertaken to invent totally resorbable devices capable of occluding defects of up to 40 mm in diameter, to overcome the theoretical disadvantages of failing access to the left atrium in the long term.

Optimally children should not reach school age untreated when suffering from a haemodynamically significant ASD. The aim must be complete closure of the defect regardless of the treatment modality, surgery or intervention, for selected patients. Therefore we should not only report the optimal results of interventional or surgical closure, but should try to establish intelligent screening programmes to detect all the patients suffering from asymptomatic but haemodynamically significant ASD before they reach adulthood and start to develop clinical symptoms with ongoing age. The knowledge that increasing adult age is accompanied by increasing symptoms and physical findings remains the main concern. ASD is not a benign disease and waiting for symptoms to prompt diagnosis is definitely unsatisfying due to the progress of the untreated disease.

References

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Tri-leaflet mitral valve in combination with hypertrophic cardiomyopathy

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A female octogenarian with no prior cardiac history was referred for stress echocardiography to evaluate exertional breathlessness. Resting images revealed asymmetrical septal hypertrophy and direct papillary muscle continuation into the presumed anterior mitral leaflet (top row, centre image). Subvalvular systolic anterior motion, significant left ventricular outflow tract obstruction, and moderate mitral incompetence were noted (see Supplementary material online, Video S1). The mitral valve demonstrated an unusual ‘Mercedes-star’ appearance in short axis (top left; see Supplementary material online, Video S2). Three-dimensional transthoracic echocardiography provided further evidence of tri-leaflet morphology, the absence of an endocardial cushion defect, and papillary muscle insertion into the ‘septal’ leaflet (middle row; see Supplementary material online, Video S2). Cardiac magnetic resonance imaging corroborated the echocardiography findings (bottom row; see Supplementary material online, Video S4), and further supported hypertrophic cardiomyopathy by antero-apical displacement of papillary muscle and late gadolinium enhancement of the antero-septal right ventricular insertion point.

The tri-leaflet mitral valve may be distinguished from isolated leaflet cleft by three evenly spaced commissures, central leaflet coaptation, and the absence of a typically positioned antero-lateral commissure (illustration bottom right). It is noteworthy how well this unique leaflet arrangement has functioned throughout the life of the patient—the clinical presentation being most likely a result of outflow tract obstruction rather than valve dysfunction per se. We conclude that the use of the latest imaging techniques can be invaluable in the assessment of unusual cardiac lesions.

Top row. Left image shows ‘Mercedes-star’ mitral valve appearance. Centre and right images show abnormal papillary muscle insertion (red arrows) using echocardiography and cardiac magnetic resonance, respectively.

Middle row. Three-dimensional transthoracic echocardiographic imaging orientated from a ventricular aspect in diastole (left) and systole (right).

Bottom row. Steady-state free precession cardiac magnetic resonance sequence. Mitral valve ‘en face’ in diastole (left) and systole (right). (Bottom right) Illustration of mitral valve morphology.

Supplementary material
Supplementary material is available at European Heart Journal online.

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