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converting-enzyme inhibitors for those with symptomatic as well as asymptomatic reduction in left ventricular ejection fraction, in order to prevent recurrence of myocardial infarction. In this vulnerable subgroup of patients with left ventricular ejection fractions ranging from 20% to 45%, amiodarone plays an important role in suppressing co-existing ventricular arrhythmias without the penalty of an increase in mortality rate. These arrhythmic patients, in whom beta blockade is contraindicated by co-existing left ventricular systolic dysfunction, stand to gain comparable benefits from the actions of amiodarone, such as reduction in oxygen requirements. Additional benefits include coronary as well as peripheral vasodilatation. Finally, in metanalysis of published trials, this drug appears to confer survival benefit not only in its own right, but also in comparison with other anti-arrhythmic drugs prescribed in the post-myocardial infarction context.

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References

Secundum atrial septal defect repair

Sir,

I fully endorse the views expressed by Speechly-Dick and colleagues that early diagnosis and surgical repair of atrial septal defect is important to prevent long-term morbidity and to reduce mortality. However, in order to highlight the exceptional longevity and benign clinical course that can be associated with this lesion, I describe a patient who to my knowledge was the longest survivor of unrepaired atrial septal defect in the United Kingdom.

In 1986 an 89 year old man was admitted to the surgical ward of his local hospital for investigation of dysphagia. A routine chest X-ray showed cardiomegaly and prominent pulmonary vascular shadows. On further questioning he recalled that at the age of 9, he was investigated at St Bartholomew’s Hospital, London for ‘blue hands’ and a ‘hole in the heart’ was diagnosed.

On examination he was in atrial fibrillation with a slow ventricular response at a rate of 60 per minute. Blood pressure was 130/90 mmHg. Jugular venous pressure was elevated to 5 cm with a visible ‘V’ wave. Apex beat was displaced to the 6th intercostal space about 2.5 cm outside the mid-clavicular line. He had a loud pansystolic murmur audible at the apex and left sternal edge, and a mild systolic ejection murmur at the second right intercostal space. He had fixed splitting of the second heart sound.

A transthoracic echocardiogram showed gross right ventricular and right atrial enlargement with mild left atrial enlargement, and large secundum atrial septal defect. He was discharged home on a diuretic. He was reviewed on his 90th birthday when he was completely asymptomatic. He died of a cerebrovascular accident when 92.

It is suggested by Rodstein et al. that a well-defined clinical picture is often present in the elderly and that such patients experience repeated attacks of bronchopneumonia, dyspnoea and angina for many years before the onset of overt heart failure. The absence of pulmonary hypertension may be an important factor in their survival. Campbell et al. observed that atrial fibrillation was nearly always the major cause of heart failure in these patients owing to production of tricuspid incompetence which in turn elevates the right atrial pressure during systole. Although this makes the interatrial shunt diastolic rather than systolic, it does not materially alter the magnitude of the left to right shunt. The limited left ventricular filling and systemic output is further diminished by the lack of atrial contribution, as seen with atrial fibrillation in any condition.

This case highlights the fact that, in patients with atrial septal defect, not only is survival possible beyond the ninth decade, but freedom from all cardiac symptoms is also possible. Owing to the lack of thorough knowledge on the rate of progression of clinical and haemodynamic worsening of such a congenital lesion, it is not easy to explain the long-term survival of such patients.

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References
Survival without atrial septal defect repair

Sir,

I was interested in the report by Speechly-Dick et al.1 on the long-term surgical outcome of secundum atrial septal defect repair. The association of mitral valve prolapse and atrial septal defect is an interesting one, although the exact cause-and-effect relation is not well understood. The most likely explanation is the altered relation between the size of the left ventricle and that of the mitral valve apparatus in secundum atrial septal defect.2 Owing to the left-to-right atrial shunt in atrial septal defect, less blood enters the left ventricle than the right ventricle. Consequently, the left ventricle in subjects with an atrial septal defect tends to be underdeveloped. When the mitral valve becomes relatively ‘too big’ to be accommodated by the ‘small’ left ventricle during systole, mitral valve prolapse ensues.3 When one waits too long before repairing the atrial septal defect, mitral valve prolapse and the accompanying mitral regurgitation may become irreversible.

Another reason why atrial septal defect once diagnosed should be repaired as early as possible is the risk of atrial fibrillation which is a common late complication of atrial septal defect.4 Once atrial fibrillation develops in patients with atrial septal defect, it is usually permanent, as was well shown by Speechly-Dick et al. who reported that atrial fibrillation persisted postoperatively in all of their patients with the arrhythmia preoperatively. The association of atrial fibrillation and stroke is a distinct risk which threatens the long-term prognosis of patients with atrial septal defect,5 and lifelong anticoagulant therapy is indicated.6

Although operative mortality in surgical repair of secundum atrial septal defect is extremely low, it is not negligible, being 3.3% in the best hands.7 Furthermore, patients undergoing surgical repair of atrial septal defect must consider the extended hospital stay and expense, the need for blood, and all the morbidity and psychological trauma associated with open heart surgery. Fortunately, there is now on the horizon a non-surgical method of closure of atrial septal defect by means of transcatheter delivery of an umbrella device.8 This non-surgical technique is not only feasible but also promising, provided patients are properly selected.9 With increasing experience and longer follow-up, transcatheter closure may well become the standard of care for most patients with secundum atrial septal defects.9

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References


Idiopathic cranial diabetes insipidus associated with idiopathic hypoparathyroidism

Sir,

We report a case of idiopathic cranial diabetes insipidus associated with idiopathic hypoparathyroidism. With one exception,1 this clinical association has not been reported as a component of the autoimmune polyendocrine syndrome.

A 20 year old student presented with a history of polyuria, polydipsia, nocturia and nocturnal enuresis since childhood. He was born of a full-term normal pregnancy through vaginal delivery and there were no perinatal complications. There was no past history of head injury or family history of significance.

On examination, he was slim with an arm span of 71 inches and height of 68 inches. His weight was 50.0 kg. There was no abnormality noted on respiratory, cardiovascular, abdominal or neurological examination. Examination of the eyes and fundi was normal. There was no anosmia.

On 24 hours observation, his oral intake of fluids was 7.0 litres and urine output was 6.0 litres. Basal investigation showed haemoglobin 12.3 g/dl, serum sodium 139 mmol/l, potassium 3.7 mmol/l, creatinine 63 mmol/l, calcium 1.9 mmol/l (normal range: 2.15–2.62), phosphorus 1.63 mmol/l (0.86–1.53), alkaline phosphatase 103 IU/l and albumin 38 g/l.

Basal endocrine investigations showed serum thyroxine 138.8 nmol/l, tri-iodothyronine 1.93 nmol/l, thyroid stimulating hormone (TSH) 0.6 IU/l, follicle stimulating hormone (FSH) 9.4 IU/l, luteinizing hormone (LH) 3.4 IU/l, testosterone 8.0 nmol/l (9.6–38.2), cortisol 0900 hrs 320 mmol/l, growth hormone 5.0 IU/ml (<1.0–7.0), serum parathyroid hormone 4.7 pmol/l (0–5.6). To evaluate polyuria and polydipsia, a short water deprivation test was performed. After 5 hours of water deprivation, his serum osmolality rose from 293 to 299 mosmol/kg and the urine osmolality from 74 mosmol/kg to 182 mosmol/kg; then he was given DDAVP (desmopressin) 1 μg subcutaneously and the urine osmolality rose to 268 mosmol/kg. These results are consistent with a diagnosis of cranial diabetes insipidus.

A high resolution computed tomographic scan of the pituitary fossa did not reveal any space-occupying lesion in hypothalamus or pituitary region. He was treated with 10 μg desmopressin twice a day intranasally and is doing well with no polyuria or nocturia.
Secundum atrial septal defect repair.

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