Letters to the Editor

Doppler echocardiography in elderly patients with ejection systolic murmurs

Sir,

Whilst serving a useful function as sensitive markers for severe aortic stenosis, the symptoms mentioned by Bishop and Wilkinson, namely, angina, syncope and dyspnoea, all have relatively poor specificity. Isolated coronary heart disease is a much commoner cause of angina and the differential diagnosis of syncope is very wide, even in the presence of a systolic murmur. When dyspnoea and cardiac failure supervene, the murmur may itself become much softer or, as McKillop et al. point out, the patient may be too ill to cooperate with echocardiography. In both situations, misdiagnosis will occur by default. Although the 12-lead electrocardiogram is mentioned as a useful screening test for left ventricular hypertrophy, this is disappointingly insensitive, since as many as 52% of surgically eligible patients who die of aortic stenosis may not have electrocardiographic signs of left ventricular hypertrophy during life. Also disappointingly insensitive is the often quoted criterion of 50 mmHg for the mean aortic valve gradient signifying severe stenosis. In one study comprising 680 adults of mean age 78 years, with symptoms sufficiently severe to justify balloon valvuloplasty, the mean aortic valve gradient ranged as widely as 12–120 mmHg when measured by Doppler echocardiography.

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References


The genes for Noonan's syndrome, woolly hair and ulerythema ophryogenes

Sir,

The report of cerebral arteriovenous malformation in Noonan's syndrome1 is of interest to dermatologists because the photograph of the patient shows that his eyebrows are absent. This is probably due to a type of scarring alopecia called ulerythema ophryogenes (Greek; ophryos, eyebrows). This rare inherited condition usually presents at birth or infancy with erythema in the outer half of the eyebrows, associated with keratinous follicular plugs. The process spreads medially and eventually causes follicular atrophy and permanent loss of the eyebrows. It is thought to be a variant of keratosis pilaris.

Although not a widely recognized feature of Noonan's syndrome, ulerythema ophryogenes was present in five of nine cases examined by Pierini & Pierini3 and in a single case reported by Nield et al.4 The present case report5 thus supports the suggestion that these two rare conditions are associated.

Neither woolly hair (widely recognized as a feature of Noonan's syndrome) nor ulerythema ophryogenes is invariably present in Noonan's syndrome. The fact that these two conditions which affect hair follicles can be inherited separately or together, with or without Noonan's syndrome,4 suggests that the genes for curly hair and ulerythema ophryogenes are separate and both may be located close to the gene defect which causes Noonan's syndrome.

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References


Primary malignant neoplasms associated with chronic lymphocytic leukaemia

Sir,

We were pleased to read the scientific publication of Lishner and colleagues from Israel about chronic lymphoid leukaemia and the second tumours.1 We have made a study on the incidence rate of chronic lymphoid leukaemia and the second tumours. We should like to inform you about our findings, by sending you a brief report on our results, hoping that it will be of interest.

We diagnosed 231 chronic lymphoid leukaemia cases (147 men) between 1972 and 1988. During the 16 year period we lost 88 of our patients (38%). Their average survival time was 29.7 months. In this period we
The genes for Noonan's syndrome, woolly hair and ulerythema ophryogenes.

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