

be consulted before decisions are made. For smaller items of equipment the best management solution may well depend on local circumstances. But there is a clear need for purchasers to be more critical of the equipment they order and for details of equipment, once evaluated and found satisfactory, to be readily available centrally for reference.

¹ *Buying for the National Health Service*. Distributed by the Welsh Office of the DHSS with circular HBSS 93/13/2.

Mitral valve prolapse

The competence of the mitral valve depends on several features. Firstly, its structure must be normal. Secondly, there must be co-ordinated interaction of the mitral leaflets, annulus, chordae tendineae, papillary muscles, and left ventricular wall.¹⁻³ Mitral regurgitation may result from any condition which deforms or immobilises these structures.

Our increasing knowledge of mitral valve function has focused attention on a fascinating syndrome, recently reviewed by Devereux *et al.*⁴ Various named Barlow's syndrome, the late systolic click syndrome, floppy mitral valve, and mitral leaflet prolapse syndrome, it is not uncommon though readily missed; not overtly rheumatic in origin though this has been suggested. Occasionally familial, usually benign, in some patients it is associated with ill-explained complications such as chest pain, ventricular ectopic beats, angiographic abnormalities in left ventricular contractile pattern, and electrocardiographic abnormalities.

Gallevardin described the distinctive auscultatory features of mitral valve prolapse back in 1913, and he also observed the effects of posture on the timing of midsystolic clicks and the often associated late systolic murmurs. He mistakenly attributed these findings to pleuropericardial adhesions, and this was not refuted until 1961, when Reid suggested that the click was caused by abrupt tensing of the chordae and the late systolic murmur by late systolic mitral regurgitation.⁵ Barlow first showed angiographically that there was late systolic prolapse of the posterior mitral leaflet.⁶ In 1968 Engle drew attention to an abnormality of left ventricular contraction,⁷ and this has been extended^{8,9} to designate mitral valve prolapse as a segmental cardiomyopathy in which the altered shape of the ventricle in systole leads to slackening of the chordae and leaflets and consequent prolapse into the left atrium.¹⁰ The most recent theory suggests that the underlying abnormality is congenital or acquired dilatation of the mitral annulus¹¹ or defective systolic contraction of the annulus, possibly associated with defective contraction of the basal portion of the left ventricle.⁹

After a period of confusion and over-diagnosis echocardiography has greatly aided the recognition and understanding of mitral prolapse.¹² Prolapse of one or both leaflets into the left atrium can be directly visualised, and the click has been shown to occur at the moment of prolapse in late systole. Both the click and prolapse occur at a constant ventricular diameter, explaining the effect of posture and drugs on their timing.^{4,13} Prompt squatting or infusion of a pressor agent increases left ventricular volume and so reduces or abolishes prolapse, but they may induce chest pain.¹⁴ In one individual clicks may be multiple or absent and move their position in systole.¹⁵ The murmur may vary from absent to a honk or whoop which can be heard across the room.¹⁶ The auscultatory features are

highly variable, and prolapse which is evident angiographically or echocardiographically may occasionally be clinically silent. Other inconstant clinical features are jerky pulses and a frankly bifid left ventricular impulse.^{15,17}

Electrocardiographic abnormalities are common.^{8,15} There is T wave inversion in inferior and sometimes also lateral leads in about one-third of cases. Ventricular premature beats have also been reported and supraventricular tachycardia is common. Abnormalities of rhythm are often provoked by effort.

Most patients have no symptoms; in those who do these are often ill explained. Some patients are aware of an intermittent noise in the heart. Complaints of fatigue, faintness, shortness of breath, and decreased exercise tolerance may be due to exercise-induced dysrhythmias or to chest pain—which is common, often atypical, but sometimes highly suggestive of coronary origin. Wooley has recently suggested (in half fun, whole earnest) that a previously well-known but now extinct disease might in truth have been mitral valve prolapse syndrome¹⁸: the non-entity emotively described as “soldier's heart” or “the effort syndrome,” eponymously as Da Costa's syndrome, or simply as neurocirculatory asthenia or “disorderly action of the heart.” In the era of “soldier's heart” diagnosis had to remain conjectural unless the outcome of a disease was frequently fatal. Who can ever find out whether Wooley is right?

The incidence of mitral valve prolapse depends on how hard it is sought. The syndrome is uncommon in childhood, becomes more frequent in women than in men, but then gradually increases in frequency and in old age is probably equally common in either sex. Though mitral prolapse can result from ischaemic papillary muscle and segmental contraction abnormality, coronary artery disease is an uncommon association.

Nearly all patients with Marfan's syndrome have echocardiographic or clinical evidence of mitral prolapse, and Marfan's syndrome has been recognised in about 4% of reported cases.⁴ Myxomatous degeneration leads to ballooning of leaflets and thinning and elongation of chordae. The mitral annulus may also become appreciably dilated. Abnormalities of the thoracic cage without Marfan's syndrome have also been described in mitral prolapse: a narrow anteroposterior diameter to the chest with a straight back (making the auscultatory features much easier to recognise). The frequency of angiographic mitral prolapse in secundum atrial septal defect has only recently been fully recognised; it has been reported in up to one-third of patients in whom it has been specifically sought.

The prognosis is usually benign, but four major complications may supervene. Sudden chordal rupture may lead to the abrupt development of severe mitral regurgitation, and this medical emergency is now well recognised. Gradually progressive mitral regurgitation due to increasing prolapse accounts for about 10% of valve replacements for isolated mitral regurgitation in adults. Infective endocarditis is rare, though an undoubted hazard, and this too can be responsible for the advent of a serious leak. Sudden death is the least common complication; it may be related to ventricular dysrhythmias¹⁹ or to an associated re-entry pathway leading to a catastrophic rapid ventricular rate should atrial fibrillation develop.²⁰

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Polyposis coli and the stomach

Among the varieties of gastrointestinal polyposis the most common and most important is polyposis coli (adenomatous polyposis of the colon and rectum). Its study has contributed much to our knowledge of the pathogenesis of colorectal cancer.¹

Polyposis coli is a precancerous genetic disorder with a dominant mode of inheritance. In affected individuals there are hundreds to thousands of adenomas in the large bowel.² Early colectomy and ileorectal anastomosis may anticipate the development of colonic adenocarcinoma in a high proportion of patients; but careful follow-up examinations of the remaining rectum are essential. Some time ago we emphasised³ the differences between this disease and the non-adenomatous types of polyposis such as the Peutz-Jeghers syndrome and juvenile polyposis. As experience of polyposis coli has increased so reports of upper gastrointestinal manifestations of the disease have accumulated.⁴ Of these, the most important clinically is the association with periampullary carcinoma: at least 17 cases have been reported.⁵ Indeed, periampullary carcinoma may be the first indication of an underlying polyposis coli.⁶

Polyps of the small bowel are rare. There have been reports of polyps in the terminal ileum and duodenum in patients with polyposis coli, but often insufficient detail was given to know whether these were adenomas.⁷ Certainly some growths in the terminal ileum are, in fact, benign lymphoid lesions.⁴ On the other hand, in the St Mark's Hospital register genuine adenomas have been recorded in the terminal ileum in two patients and in the duodenum in three patients.⁸ A recent upper gastrointestinal endoscopic study from Japan recorded small bowel adenomas in five of ten patients examined⁹; these also appeared to be genuine examples.

True gastric adenomas are just as rare. Hyperplastic or regenerative polyps of the stomach are common and may be misinterpreted as adenomas. Another Japanese paper¹⁰ has raised the possibility of an association between polyposis coli and benign gastric tumours. The authors correctly comment that polyposis coli may not only be a disease of the large intestine. They add, however, that the high incidence of gastric cancer in Japan makes their positive findings in 10 of 15 patients difficult to interpret. Similar caution is necessary when an even higher incidence of gastric polyps is reported in association with polyposis coli.⁹ In this latter series all but one example were regarded as hyperplastic fundal glands and not as adenomas. In the polyposis families seen at St Mark's

there are no records of gastric adenomas, but hyperplastic foci have been observed in one patient.⁸

A Swiss report¹¹ described a clustering of gastric carcinomas in one family with minor polyposis coli. These patients, who had small numbers of large bowel adenomas, probably represented Veale's "recessive adenoma expression," in which many fewer adenomas are found than in classical polyposis coli.¹² Outside Japan there is only one recorded example of gastric carcinoma in a patient with polyposis.¹³

Certainly the physician or surgeon caring for a patient with polyposis coli should also investigate the upper gastrointestinal tract. Clinical symptoms should be evaluated together with air contrast barium meal studies. One barium meal early in the disease and a repeat later, along with gastroscopy and duodenoscopy, may be the minimum investigation required. Because the various types of polyp have a different prognosis careful histological study of any biopsies is essential. But even when upper gastrointestinal disease is established—whether as adenomas, hyperplastic, or hamartomatous lesions—it is difficult to advise what further can be done. There is no evidence, as yet, to suggest that duodenal adenomas progress to periampullary carcinoma or that the gastric polyps are precancerous. If there is such progression, experience with the large bowel suggests that the history of malignant change may be measured in years.¹ For the future, upper gastrointestinal endoscopic polypectomy may resolve some of these problems. For the present, the major preoccupation with a patient with polyposis must remain the risk of rectal carcinoma after the colon has been removed. The significance of upper gastrointestinal tract malignancy has still to be defined.

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Irritant woods

Charles Darwin would doubtless have been upset had he known of the Coco de mono tree of Venezuela.¹ It apparently bears pods of such complexity that only the most dexterous of monkeys can open them and obtain the tasty almond-like nut. Once the nuts have been eaten the monkey's hair drops out and he soon expires—thus ensuring the survival of the least fit members of each generation.

We are all conditioned to be suspicious of trees with strange fruits, but the fact that the timber itself may be toxic in a number of ways is perhaps less well known. Woods and Calnan² have done well to draw attention in great detail to the large range of trees used in manufacturing processes in the context of their potential as irritants and sensitizers. The substances responsible may be present in bark, wood, or pollen, while allergies to lichens growing on the bark pose an added hazard to the unwary and unfortunate.