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Small intestinal adenocarcinoma, duodenal carcinoid tumour, and von Recklinghausen's neurofibromatosis

SIR,—We would like to comment on the report by Jones and Marshall (*Gut* 1987; **28:** 1173–6) of small intestinal adenocarcinoma occurring in neurofibromatosis. On the basis of their own and four additional published cases, they suggest the existence of a specific association between these two conditions. Surprisingly, as two of the five tumours were duodenal, they did not mention the now established link between duodenal carcinoid and neurofibromatosis.

In recent years we[1 2] and others[3 4] have drawn attention to an association between von Recklinghausen's disease and a distinctive carcinoid tumour of the duodenum – a review of 27 such cases has recently been published.[5] This duodenal carcinoid tumour is distinguished by containing somatostatin, and on microscopy commonly has psammoma bodies and a glandular growth pattern. The latter feature of the tumour makes it easily confused with duodenal adenocarcinoma by the histopathologist – indeed of nine duodenal carcinoids studied by ourselves, four had not previously been investigated with immunocytochemistry and were diagnosed originally as adenocarcinomas.[5] In two of the cases of small bowel adenocarcinoma in neurofibromatosis cited by Jones and Marshall the intestinal tumour was in the duodenum, yet the authors do not appear to have considered the possibility that they might be glandular carcinoids. In fact, we have studied personally the duodenal tumour from one of the cases[6] (Jones and Marshall's fourth case) and have indeed found it to be a somatostatin containing glandular carcinoid; this case has also been studied by Dayal *et al*[4] who agree with this interpretation.

Jones and Marshall base their suggestion of an association between neurofibromatosis and small bowel adenocarcinoma on five cases – now known to be four. While there may be such a link, it remains a tenous one, particularly since rare conditions occurring together are much more likely to be reported than when they occur singly. We feel that it is important for clinicians to be aware of the more certain association of neurofibromatosis with duodenal glandular carcinoids, so that this diagnosis is the first to be considered in a patient with von Recklinghausen's disease and a duodenal (especially a periampullary) tumour. The histopathologist should be aware of the association also, the mimicry of adenocarcinoma may otherwise lead to misdiagnosis. The distinction is an important one – duodenal carcinoids are much less aggressive than duodenal adenocarcinomas. Consequently their surgical extirpation is far more likely to lead to cure.

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Preservation of faecal continence during rises in ultra-abdominal pressure

SIR,—I was pleased to see the paper by Bannister, Gibbons, and Read[1] confirming that continence is sphincteric and not as previously thought dependent on an anorectal flap valve. Parks popularised the flap valve theory of continence which has gained wide acceptance.[2] In a recent study[3] we addressed the

Reply

SIR,—We thank Drs Kochlar, Rajwanshi, and Mehta for their interest in our paper, and we look forward to reading their results when published. They correctly point out that since our paper was accepted Lange *et al* have published the results of a similar technique, although with a poor yield of readable samples. We believe our method of gentle movement of the needle tip in the specimen is helpful in increasing the amount of sample obtained, and may account for the difference in results.

We are also grateful to the writers for drawing our attention to abstracts of other work in this field which do not appear in *Index Medicus* as no full paper has apparently been published. Clearly several workers have found endoscopic needle aspiration cytology of the stomach to be practicable. We hope our evidence of its value in the diagnosis of gastric malignancy will encourage others to adopt the technique.

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Staffing and administration of endoscopy

SIR,—Your occasional report, 'Results of a questionnaire concerning the staffing and administration of endoscopy in England and Wales' highlights many inadequacies in the provision of endoscopy services in England and Wales. In particular I was concerned to read of the ongoing deficiencies in training for those working in endoscopy units and the apparent unwillingness of anyone in the NHS to pick up this responsibility and run with it.

My company as the major suppliers of endoscopes to the NHS has not neglected its responsibility in this area and is running training courses regularly which have now accommodated an aggregate of over 3000 nurses and technicians over 14 years. In addition, it has supported, with financial and practical aid, the only regular advanced endoscopy course for nurses. The importance to the NHS of these courses has been two-fold: better understanding of the instrumentation has both cut the cost of repairs (very considerably) and thereby helped in providing uninterrupted services for patients.

Your readers will be interested to compare the reported attendance at the ENB course with the current booking position for our primary course 'Care and maintenance' which is 85% fully booked three weeks after announcement, and has a waiting list for a suitable place of over 1500 potential delegates.

The procurement Directorate of the DHSS is encouraging more aggressive purchasing on value for money grounds, but in practice this rarely seems to take into account such 'invisibles' as user training. If the NHS wants such services then it must either be prepared to pay for them or to make the necessary very substantial investment in providing the courses that, as our waiting lists suggest, are in demand.

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Addendum to letter on p. 553 entitled 'Small intestinal adenocarcinoma. . . .' The authors wish to add the following: Jones and Marshall's paper prompted a further report by Kingston (Letter, *Gut* 1988; **29:** 134) of a periampullary tumour in a patient with neurofibromatosis. We have kindly been allowed to study this tumour which again proves to be a somatostatin-rich glandular carcinoid.

Books

Inflammatory bowel disease: a personal view by Henry D. Janowitz, M.D. (Pp. 179; illustrated; $42·95.) Chicago, USA, Field, Rich, 1985.

This is not a textbook but a sharing of unrivalled experience. Dr Janowitz began work at the Mount Sinai Hospital, New York, in 1939 and has devoted much of his professional life to inflammatory bowel disease ever since. He has worked with Crohn, Ginzberg, and Oppenheimer and many other distinguished colleagues, he has lead a distinguished research team himself, and he has experience of countless patients. The recurring problems of the patients with inflammatory bowel disease are dealt with practically and sympathetically. The rare occurrences in a long busy career, such as massive haematuria caused by bladder involvement in Crohn's disease, add particular interest. A well produced set of small intestinal radiographs sharpen our knowledge of differential diagnosis, and remind us that Crohn's disease has many mimics.

Medical practice in a sophisticated tertiary referral