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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 cases⁶ (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 tenuous 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 Read¹ 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 study³ we addressed the

question of how continence was maintained. A technique similar to that adopted in urodynamics was used. Rectal and anal canal pressures were measured together with puborectalis and external anal sphincter electromyogram whilst the subject carried out a Valsalva manoeuvre. The rectum was filled with a dilute barium solution until a sensation of fullness was perceived. During the Valsalva manoeuvre the rectum was visualised radiologically whilst measurements were made. We have now carried out over 50 such examinations and never observed a flap valve occlusion of the upper anal canal. Indeed the anterior rectal wall was always clearly separated from the top of the anal canal. Moreover rectal pressures never exceeded anal canal pressures. The conclusions of our study reported in the *British Journal of Surgery*³ were similar to those of Bannister *et al*¹—namely, that continence is maintained by sphincteric means.

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Extrahepatic portal venous obstruction

SIR,—We thank Dr Triger for commenting on our paper in his leading article.^{1,2} We would agree with him on most of the points except on the following issues. Our recent analysis of 521 patients with portal hypertension revealed cirrhosis of the liver in 215 (41%), EHPO in 213 (40%) and non-cirrhotic portal fibrosis (NCPF) in 93 (18%) patients. Only 17% of our patients with EHPO had their first bleed after the age of 20 years, compared with the series quoted by Webb and Sherlock where almost 50% were adults.³ Because patients with NCPF have a similar presentation as EHPO, namely, well tolerated bleeds with splenomegaly, the discriminant analysis of the two conditions (EHPO and NCPF) from our centre has shown NCPF to present mostly in 2nd or 3rd decade with gross splenomegaly usually more than 11 cm below the costal margin, while EHPO presents in the

first or second decade with the spleen size usually less than 7 cm below the costal margin.⁴ Our analysis of the size of spleen in EHPO has shown mild splenomegaly (5 cm) to be present in 42% of patients, moderate (6–10 cm) in 40% and gross splenomegaly (>11 cm) in only 18%. Thus gross splenomegaly, as mentioned in the leading article,¹ is not a very common feature in EHPO. Because of the size of the spleen, hypersplenism is more of a feature in NCPF than in EHPO.

As far as aetiology of EHPO is concerned, we have been unable to find any cause or association in 92% of our patients. Umbilical sepsis was responsible in only 13 (6%) of 213 patients. Congenital anomalies in the form of VSD, and mitral valve prolapse syndrome was noticed in only four patients (2%). We have never observed EHPO in a patient with cirrhosis, unlike the series quoted from Italy in the article.

In our series of 213 patients, obstruction in right and left portal vein branches was seen in 14.2% of patients, main portal vein obstruction in 51.7% and a total block of portal and splenic vein was seen in 28.9%. Isolated splenic vein thrombosis was seen in only one patient, unlike the series quoted by the leading article. Hence splenectomy alone is almost never done in a patient with EHPO.

With respect to the management of patients with EHPO, we agree with the feelings of Dr Triger that the first line of treatment should be sclerotherapy in this relatively benign condition with the hope that they will grow out of their bleed as the time passes. Indeed we have recently shown (under publication) that 40% of the patients developed large spontaneous/natural shunts after obliteration of oesophageal varices by endoscopic sclerotherapy. To date they have not rebled with a follow up of two years. These sclerotherapy induced spontaneous/natural shunts are significantly more common, than observed otherwise in EHPO without sclerotherapy².

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