Pathogenesis

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Historically , tumours arising from the neuroendocrine cells were referred to as ‘karzinoide’ (carcinoma-like) tumours by Oberndorfer. Now they are termed neuroendocrine tumours (NETs). NETs can be either benign or malignant. In the gastrointestinal tract, especially the small intes tine, they secrete excess 5-HT , which is rapidly metabolised by the liver. In the presence of liver metastases, NET metas tases secrete 5-HT into the IVC, av oiding hepatic first-pass metabolism and leading to symptoms of 5-HT excess, known as carcinoid syndrome . The cardinal features are sweat ing, flushing, bronchospasm and restrictive cardiomyopathy secondary to fibrotic right heart valvular disease (Hedinger syndrome). The overall incidence of GI-NETs is 2.5–5.0 per 100 /uni00A0 000 people per year, with a much higher prevalence of 35 per 100 /uni00A0 000. Their relative distribution in the gastrointestinal tract is shown in Table 57.6 . Nikolai Kulchitsky , 1856–1925, Professor of Histology , Kharkov Imperial University , Ukraine. Siegfried Oberndorfer , 1876–1944, Professor of Pathology , Munich, Germany . Christoph Hedinger , 1917–1999, pathologist, Zurich, Germany .

tumours in different organs. Site Distribution (%) Lung 10 Stomach 5 Duodenum 2 Small bowel 25 Appendix 40 Colon 6 Rectum 15

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