

ulation in our study was selected from 273 consecutive patients with carcinoid tumors who were referred for echocardiography because of suspected carcinoid heart disease. Thus, a high frequency of carcinoid heart disease and high 5-HIAA excretion would be expected. However, we did not find any difference in peak 5-HIAA levels between the patients who were included in the study (median, 222 mg per 24 hours; interquartile range, 148 to 345) and those who were excluded (median, 238 mg per 24 hours; interquartile range, 131 to 362) ($P=0.79$). In contrast to the data presented by van der Horst-Schrivers and colleagues, we found that 5-HIAA levels decreased during follow-up, probably as a result of an aggressive therapeutic strategy. The 5-HIAA levels in our study were similar to those measured in previous studies of similar populations.^{1,2} Thus, we believe that the population we studied is representative of patients with clinically significant carcinoid heart disease.

It is possible that somatostatin analogues may slow the progression of carcinoid heart disease. However, the available data indicate that current methods of treatment are inadequate to prevent the development and progression of carcinoid heart disease. Larger, prospective studies are needed to define the optimal treatment strategy.

Jacob E. Møller, M.D.
Heidi M. Connolly, M.D.
Patricia A. Pellikka, M.D.

Mayo Clinic
Rochester, MN 55905
pellikka.patricia@mayo.edu

1. Denney WD, Kemp WE Jr, Anthony LB, Oates JA, Byrd BF III. Echocardiographic and biochemical evaluation of the development and progression of carcinoid heart disease. *J Am Coll Cardiol* 1998; 32:1017-22.
2. Westberg G, Wängberg B, Ahlman H, Bergh CH, Beckman-Suurkula M, Caidahl K. Prediction of prognosis by echocardiography in patients with midgut carcinoid syndrome. *Br J Surg* 2001;88: 865-72.

Genetics of Colorectal Cancer

TO THE EDITOR: Lynch and de la Chapelle (March 6 issue)¹ emphasize the screening of high-risk patients who have a mutation in the adenomatous polyposis coli (APC) gene or who have one or more first-degree relatives with familial adenomatous polyposis. However, the importance of ophthalmic examination in screening for and diagnosis of familial adenomatous polyposis is not noted. Congenital hypertrophy of retinal pigment epithelium is the most prominent extracolonic manifestation of familial adenomatous polyposis and is present in about 90 percent of patients.²⁻⁴ This condition can be identified by noninvasive methods even in infants and young children by simple fundus examination with the pupils dilated. A combined approach involving the detection of an APC mutation and detection of congenital hypertrophy of retinal pigment epithelium for presymptomatic diagnosis of familial adenomatous polyposis is highly recommended.⁵

Wai Man Chan, M.R.C.P., F.R.C.S.
Chi Pui Pang, D.Phil.
Dennis S.C. Lam, F.R.C.S., F.R.C.Ophth.

Chinese University of Hong Kong
Kowloon, Hong Kong
dennislam@cuhk.edu.hk

1. Lynch HT, de la Chapelle A. Hereditary colorectal cancer. *N Engl J Med* 2003;348:919-32.
2. Pang CP, Fan DS, Keung JW, et al. Congenital hypertrophy of the

retinal pigment epithelium and APC mutations in Chinese with familial adenomatous polyposis. *Ophthalmologica* 2001;215:408-11.

3. Valanzano R, Cama A, Volpe R, et al. Congenital hypertrophy of the retinal pigment epithelium in familial adenomatous polyposis: novel criteria of assessment and correlations with constitutional adenomatous polyposis coli gene mutations. *Cancer* 1996;78:2400-10.
4. Lam DSC, Kwok SPY, Kwok AKH, Liew CT, Lau JWY, Pang CC. Incidence and predictive value of congenital hypertrophy of retinal pigment epithelium in Chinese familial adenomatous polyposis patients. *Chin Med J (Engl)* 1998;111:278-81.
5. Pang CP, Lam DS. Differential occurrence of mutations causative of eye diseases in the Chinese population. *Hum Mutat* 2002;19: 189-208.

TO THE EDITOR: In Table 3 of the review article by Lynch and de la Chapelle, microcephaly is listed as one of the phenotypic features of the Bannayan-Ruvalcaba-Riley syndrome. In fact, patients with this syndrome have macrocephaly¹ (with normal-size ventricles). Typically, their birth weight is greater than 4 kg and their birth length above the 97th percentile, but their final height as adults is within the normal range.² In addition, 50 percent of the patients have hypotonia, delayed gross motor or speech development, or mental retardation.² In about 60 percent of the patients, a myopathic process affecting the proximal muscles is present.²

Karen W. Gripp, M.D.
A.I. duPont Hospital for Children
Wilmington, DE 19803
kgripp@nemours.org

1. Gorlin RJ, Cohen MM Jr, Condon LM, Burke BA, Bannayan-Riley-Ruvalcaba syndrome. *Am J Med Genet* 1992;44:307-14.
2. Jones KL. *Smith's recognizable patterns of human malformation*. 5th ed. Philadelphia: W.B. Saunders, 1997.

The Electrocardiogram in Acute Myocardial Infarction

TO THE EDITOR: The review article by Zimetbaum and Josephson (March 6 issue)¹ on the usefulness of the electrocardiogram in the diagnosis of acute myocardial infarction is essential reading for anyone who cares for patients with a suspected acute myocardial infarction. Some qualifications regarding the diagnosis of acute myocardial infarction in patients with left bundle-branch block are in order. Although proposed criteria² facilitate the diagnosis, an occasional problem arises when reliance on the criterion of an ST-segment elevation of more than 5.0 mm in leads with primarily negative QRS complexes leads to a false positive diagnosis of acute myocardial infarction. This criterion is non-specific for acute myocardial infarction, and some patients who have left bundle-branch block without an acute myocardial infarction, but with left ventricular hypertrophy or dilatation, have electrocardiograms with ST-segment elevations that are much larger than 5.0 mm.^{3,4} An association between large ST-segment elevations and large QRS complexes has been reported.³ Repeating electrocardiography may also be of value, since a change in the amplitude of the ST-segment elevation suggests an acute myocardial infarction.⁵ Finally, since measurements of ST-segment elevations are being used for the diagnosis of acute myocardial infarction in patients with left bundle-branch block, it is prudent to mark the thorax in order to ensure reproducible serial electrocardiograms.⁵

John E. Madias, M.D.

Mount Sinai School of Medicine
New York, NY 10029
madiasj@nychhc.org

1. Zimetbaum PJ, Josephson ME. Use of the electrocardiogram in acute myocardial infarction. *N Engl J Med* 2003;348:933-40.
2. Sgarbossa EB, Pinski SL, Barbagelata A, et al. Electrocardiographic diagnosis of evolving acute myocardial infarction in the

presence of left bundle-branch block. *N Engl J Med* 1996;334:481-7. [Erratum, *N Engl J Med* 1996;334:931.]

3. Madias JE, Sinha A, Ashtiani R, Agarwal H, Win M, Narayan VK. A critique of the new ST-segment criteria for the diagnosis of acute myocardial infarction in patients with left bundle-branch block. *Clin Cardiol* 2001;24:652-5.

4. Madias JE, Sinha A, Agarwal H, Ashtiani R. ST-segment elevation in leads V1-V3 in patients with LBBB. *J Electrocardiol* 2001;34:87-8.

5. Madias JE. Serial ECG recordings via marked chest wall landmarks: an essential requirement for the diagnosis of myocardial infarction in the presence of left bundle branch block. *J Electrocardiol* 2002;35:299-302.

THE AUTHORS REPLY: We agree entirely with the comments of Dr. Madias. The criteria for a diagnosis of myocardial infarction in a patient with left bundle-branch block are flawed. Not only is it difficult to interpret ST-segment changes in a patient with left bundle-branch block and preexisting left ventricular hypertrophy or dilatation, but also the coexistence of multivessel coronary artery disease or left ventricular aneurysm will most likely limit the ability to diagnose acute myocardial infarction electrocardiographically. We chose to present data from the largest series of patients with documented acute myocardial infarction.¹ The editorial by Wellens that accompanied the report on that series further highlighted the difficulties involved in assessing acute myocardial infarction in the presence of left bundle-branch block.²

Peter Zimetbaum, M.D.

Mark E. Josephson, M.D.

Beth Israel Deaconess Medical Center
Boston, MA 02215
pzimetba@bidmc.harvard.edu

1. Sgarbossa EB, Pinski SL, Barbagelata A, et al. Electrocardiographic diagnosis of evolving acute myocardial infarction in the presence of left bundle-branch block. *N Engl J Med* 1996;334:481-7. [Erratum, *N Engl J Med* 1996;334:931.]
2. Wellens HJJ. Acute myocardial infarction and left bundle-branch block — can we lift the veil? *N Engl J Med* 1996;334:528-9.

Vestibular Neuritis

TO THE EDITOR: Droperidol (2.5 mg to 10 mg) was mentioned as a treatment option in the Clinical Practice article on vestibular neuritis by Baloh (March 13 issue),¹ with “liver or kidney disease” mentioned as a precaution. In December 2001, the Food and Drug Administration (FDA) strengthened its warnings about the use of droperidol, specifically add-

ing a “black box” warning about deaths associated with prolongation of the QT interval and torsade de pointes. It was recommended that droperidol be reserved for the treatment of patients whose condition was unresponsive to other therapies and that it be used only if the benefit outweighed the risk; it was also recommended that monitoring of vital