payment for phonocardiography, and lastly but certainly not least, the decline in the presence of senior teachers and master clinicians.7–9 As stated by Adolph,10 “We must remedy the situation before it is too late. When the senior teachers are gone, who will then teach?” In an excellent review on this subject, Craige11 stated, “The present trend toward the denigration of auscultation may soon leave us with a whole new generation of young physicians who have no confidence in their own ability to make worthwhile bedside diagnoses.” Unfortunately, his prophetic wisdom has come to pass. Cardiac auscultation is an art and a science. This is one of the few pleasures that can be derived from the ever-changing practice of medicine. It stimulates acquisition of a good doctor-patient relationship and provides a satisfying alternative to many of the idiopathic inconsistencies and inadequacies of medical practice. It is unquestionably an art and skill that must not be allowed to succumb to the way of the impossible dream of Don Quixote.

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Epoprostenol and Nesiritide in Pulmonary Hypertension

To the Editor:

The case report by Kurian et al1 highlights several important misconceptions regarding the management of pulmonary hypertension that warrant attention. Firstly, one cannot address the optimal approach to therapy of pulmonary hypertension without first delineating its etiology. In the case presented, the disease process was postcapillary in origin, and therefore should have been treated as such. Although the authors refer to epoprostenol (prostacyclin) as “traditional therapy,” this drug is approved by the Food and Drug Administration for the treatment of patients with pulmonary arterial hypertension (PAH) who fall in to New York Heart Association functional classes III or IV; indeed, epoprostenol therapy is considered to be contraindicated in patients with pulmonary venous hypertension due to left-sided heart disease.2 At best, this would have to be considered off-label use that carries considerable risk without evidence of benefit for this condition.

Whether nesiritide may offer benefit in patients with PAH will require carefully performed clinical trials in a well-defined population using established criteria for disease definition. The current management of patients with PAH is presented comprehensively in the American College of Chest Physicians evidence-based guidelines, which accompanied the July issue of CHEST.3

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The Importance of Clinical Registries for Pulmonary Epithelioid Hemangioendothelioma

To the Editor:

We read with great interest the article by Cronin et al1 in CHEST (February 2004) entitled “Pulmonary Epithelioid Hemangioendothelioma: An Unusual Case and Review of the Literature.” In their article, the authors commented on several aspects of pulmonary epithelioid hemangioendothelioma, including the rarity of epithelioid hemangioendothelioma (EHE) to the lung, the ability to make this diagnosis by transbronchial biopsy, the fact that EHE can affect various organs, the varied prognosis for patients with EHE, the varied response with chemotherapy, and, notably, the fact that some patients have shown full regression of EHE without any treatment. We agree with the authors that the rarity of this condition, the lack of clear standards for treatment, and the partial-to-complete spontaneous regression of EHE seen in some patients up to 15 years from initial detection makes it difficult to decide on the most appropriate treatment.2 However, we do not necessarily think that the involvement of other organs such as the liver translates into poor survival, as commented on by the authors. Long-term follow-up of ≥20 years has been reported in many cases of EHE involving both the lung and liver, with a survival time of up to 27 years reported.

Partial hepatectomy and liver transplantation for diffuse involvement is now a standard treatment for localized and diffuse involvement of the liver with EHE. Marino et al3 reported a 76% 5-year survival rate for 10 patients with EHE involving the liver who underwent liver transplantation. Five patients who received transplants were free of metastatic disease, although two patients

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died at 3 months and 16 months. Surprisingly, four of those patients who had undergone transplantation had microscopic metastasis to the hilar lymph nodes, chest wall, lung, pleura, or diaphragm. All patients with metastasis were alive at the time of reporting after a mean (±SD) duration of 40.6 ± 22 months. The authors concluded that liver transplantation was a reasonable procedure for bulky, otherwise unresectable EHE even in the presence of metastatic disease.

The treatment for localized lung lesions involves local resection, whereas diffuse lung lesions have historically been treated with chemotherapy in many instances. Reports on the use of intraarterial 5-fluorouracil, doxorubicin, and newer agents like interferon-α, b and interleukin-2 for chemotherapy have provided mixed results. High-dose chemotherapy with autologous stem-cell rescue also has been used with mixed results. Radiotherapy is widely considered to be ineffective and has been used to palliate pain from bone involvement. Progestrone and estrogen receptors have been expressed on tumor cells, and we think that there may be a role for hormonal therapy in patients with diffuse pulmonary disease.

Due to the inconsistent therapeutic results with surgery, the lack of proven effective chemotherapy, and the possibility of regression in some patients with multorgan involvement from EHE, treatments should still be individualized. Multisystemic involvement should not equate necessarily to a poor prognosis. Hormonal therapy should be studied closely to see whether there is any beneficial effect on progestrone and estrogen receptor-positive tumor cells. Due to the rarity of this disease, double-blind randomized controlled studies investigating at various management strategies will not be possible. Clinical registries such as the Armed Forces Institute of Pathology Registry and the International Hemangioendothelioma, Epithelioid Hemangioendothelioma and Vascular Disorders Registry are places where physicians as well as patients and their families can share their symptoms and treatments, along with response rates and relapses. Such registries may help both patients and physicians alike in following the natural history of the disease and its response to different therapeutic regimens.

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To the Editor:

Kpodonu and others have written regarding our report in CHEST (February 2004) of a patient with epithelioid hemangioendothelioma of the lung, and they have cited additional information about this very rare condition. They have correctly pointed out the range of prognoses that are associated with patients who receive this diagnosis and added liver transplantation to the possible list of treatments that one might consider in cases of extensive hepatic involvement. We thank them for bringing this to our attention and acknowledge the reports cited in their correspondence. We agree that it is impossible to point to any accurate standard of care for such a rare condition, especially one with a natural history that appears to be so variable from patient to patient. Kpodonu and colleagues advocated for the use of registries to assist patients and clinicians in decision making about this and other rare conditions, in which randomized trials are not feasible, an idea that we support enthusiastically.

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