

Many years ago it was suggested that sarcoidosis was caused by a transmissible agent.<sup>11,12(p20)</sup> Recently disseminated granulomatous inflammation developed in two patients without sarcoidosis who received heart transplants from donors who had sarcoidosis.<sup>13</sup> Granulomas have also occurred in heart and lung allografts in patients with sarcoidosis who received heart and lungs from nonsarcoidotic donors. These observations raise the possibility of a transmissible causative agent. Furthermore, the agent(s) remaining sequestered in the body is able to withstand aggressive immunosuppression and is capable of attacking transplanted lungs that have normal ventilation and perfusion.<sup>14</sup> If, indeed, there is a transmissible agent, where does it reside? Interestingly, a group of patients with sarcoidosis who had liver transplants for sarcoidosis or other causes of liver disease showed regression of the multisystem disease.<sup>15</sup> Did sarcoidosis subside in these patients because of immunosuppression or because of the removal of the diseased liver, the storehouse of the causative agent?

It is widely accepted that there is a sarcoid diathesis or constitution, but no definite genetic factors relating to the cause have so far been detected. The disease is clearly a worldwide phenomenon. Numerous contributing factors, including occupation, hobbies, pets, alcohol, tobacco, place of residence, family history, and use of drugs, have been analyzed, but no relationship has been found. It is relevant that the disease mostly affects persons between 20 and 50 years in age. Are these mobile, healthy, working people at a constant risk of inhaling an agent that is present universally? Is there another unidentified virus lurking around? The search continues.<sup>16</sup>

OM P. SHARMA, MD  
Department of Medicine  
University of Southern California  
School of Medicine  
Los Angeles, California

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## Portal Hypertension— The Surgical Pendulum

THE PENDULUM HAS SWUNG to and fro as to the role of surgical management for portal hypertension since Nicolai Eck first advocated his portacaval fistula. Eck's justification to use this shunt in humans was based on an 87% mortality in dogs. His bravado was rapidly countered by Pavlov's systematic assessment of the risks of liver failure with the deprivation of portal venous flow to the liver. A flurry of surgical activity in the early 20th century was characterized by Vidal's "forced" shunt and attempts at partial decompression by Morison's omentopexy.<sup>1</sup> Poor results led to abandoning the use of surgical therapy until the 1940s when a group at Columbia-Presbyterian Medical Center (New York, NY) popularized total portal systemic shunting. Initial enthusiasm, based on excellent control of variceal bleeding, soon waned when randomized trials documented the side effects of total portal diversion.<sup>1</sup> Surgeons sought a better way to manage variceal bleeding by selective variceal decompression<sup>2</sup> and extensive devascularization procedures<sup>3</sup> in the 1970s. Although popularity for these operations largely disappeared in the 1980s under the acclaim of sclerotherapy, by the late 1980s and 1990s, liver transplantation has swung the pendulum once more towards surgical therapy for some patients.<sup>4</sup> In addition, the realities of bleeding through sclerotherapy have brought back some of the previous surgical approaches in good-risk patients who do not need transplantation.

In their excellent review of surgical options in the management of portal hypertension in this issue of the journal, Collins and Sarfeh carefully outline the goals and outcomes of the various surgical possibilities.<sup>5</sup> How does a physician really decide what to do with a patient who presents with variceal bleeding? Clearly, variceal bleeding is one of those sentinel events in medical practice that requires full evaluation and definitive management decisions. Let us examine this question in a little more detail.

### Acute Variceal Bleeding

Acute variceal bleeding is currently managed by endoscopic sclerotherapy. A new challenger, however, somatostatin or its analogue octreotide, is gaining popularity, with two randomized trials in acute bleeding documenting equal efficacy to sclerotherapy.<sup>6,7</sup> To practitioners who see few patients with variceal bleeding and who do sclerotherapy only occasionally, these data support the

use of octreotide (50 µg per hour) as a reasonable alternative. The popularity of the use of balloon tamponade has diminished, and its use should only be considered if a patient fails to stabilize with somatostatin or sclerotherapy, and even then only be used as a temporizing measure either to transport the patient or to arrange an alternative treatment modality.

I would take issue with the authors in advocating an emergency portacaval shunt for a patient in whom sclerotherapy for acute variceal bleeding fails. Transjugular intrahepatic portacaval shunt (TIPS) provides a less invasive alternative in such patients who are invariably high operative risks, and it provides portal decompression without an operation. Currently most centers accept this as an established role for TIPS.

Is there a role for emergency surgery within six hours of admission for variceal bleeding? A recent randomized trial provides data in support of this and, indeed, has led to a call for the further evaluation of this approach.<sup>8,9</sup>

#### Patient Evaluation

Patient evaluation is the key to making definitive treatment choices in a patient stabilized from an acute variceal bleed. The first question to be answered is whether the patient has end-stage liver disease that can only be managed by transplantation. If that is the case and the patient is otherwise an appropriate candidate for transplant, the choice is clear. At the other end of the spectrum, early identification of a patient with an extrahepatic cause for portal hypertension and a normal liver is important in further management of that patient. More difficult to sort out are the patients with cirrhosis with some impairment of hepatic function, but in whom the episode of variceal bleeding makes it difficult to judge the true severity, activity, and likely natural history of underlying liver disease.

#### Prevention of Rebleeding From Varices

Surgical decisions in the 1990s are not made in isolation, but as part of a team approach to total patient care. Once a patient is stabilized from an acute bleeding episode and has been evaluated, the surgeon has two questions to address:

- Is the patient now or at some time in the future likely to be a candidate for liver transplantation? If the patient has end-stage disease at this time and is a suitable candidate for transplantation, he or she should be transplanted. If the patient has advanced liver disease and is likely to come to transplantation in the next three to four years, the surgeon is less likely to advocate an alternative operative procedure, but would rather manage that patient with sclerotherapy, TIPS, or both. If the patient has a disease that may bring the patient to transplantation in the next five to ten years but has adequate hepatocellular reserve at this time, the decision may be that an alternative procedure such as a selective or partial shunt may provide good management for that patient for at least five years.<sup>10,11</sup> Such patients remain candidates for transplantation if and as their liver disease progresses.

A difficult patient is one with advanced disease but contraindications to transplantation, such as active alcoholism or substantial other-system disease. The management of variceal bleeding in such a patient may encompass any of the therapeutic modalities outlined in the current review.<sup>5</sup> Equally important, however, is to try and correct any associated concurrent underlying problems, as a proportion of these patients may ultimately become transplantation candidates.

- Which of the nontransplantation options provides the best surgical management? There is no single answer to this, but, rather, factors such as disease cause, portal venous anatomy, the availability of local skills, and the preference of the patient and surgeon weigh in this decision. There are advantages and disadvantages to each of the surgical therapies outlined. Total shunts control bleeding well, but the diversion of portal flow from a cirrhotic liver is associated with accelerated liver failure and a higher incidence of encephalopathy. Partial shunts and selective shunts have a lower incidence of hepatic failure and encephalopathy and control bleeding equally as well as total shunts. These are probably the most popular approaches at this time. Devascularization procedures have a higher risk of rebleeding but do not interfere with portal perfusion and liver function.<sup>12</sup>

In the mid-1990s, the pendulum has swung back in favor of surgical intervention for some patients with portal hypertension. Clearly the major surgical procedure in this field is liver transplantation, but for patients in whom there is preserved hepatic function in the face of variceal bleeding, other alternatives provide a reasonable approach for patient management.

J. MICHAEL HENDERSON, MD  
Chair  
Department of General Surgery  
The Cleveland Clinic Foundation  
Cleveland, Ohio

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