A healthy 14-month-old girl was referred for evaluation of hepatomegaly found during a physical examination prompted by accidental trauma of her left wrist. She had no history of jaundice, weight loss, night sweats, fever, or decreased activity. Her medical history was negative for perinatal difficulties, including cardiac abnormality. There was no recent foreign travel, and the family had no pets. The family history was negative for hepatitis, Wilson disease, α1-antitrypsin deficiency, storage diseases, or hepatic tumors.

Findings from physical examination revealed a well-developed infant with height and weight in the 50th percentile. Heart rate, blood pressure, and respiratory rate were normal, as were the retinal examination. No cutaneous lesions or cardiac murmur were present. The liver extended 5 cm to 6 cm below the right costal margin. The spleen was not palpable. The stool was negative for occult blood. Results of laboratory examination showed reference values for hemoglobin, hematocrit, platelet count, white blood cell count, erythrocyte sedimentation rate, alanine aminotransferase, aspartate aminotransferase, alkaline phosphatase, γ-glutamyltranspeptidase, bilirubin, and albumin. Abdominal ultrasonogram and computed tomographic scan (with and without intravenous contrast medium) were obtained (Figure 1 and Figure 2). Serum α1-fetoprotein levels were normal. A radiolabeled red blood cell scan was unhelpful for diagnosis because of technical difficulties. Repeated ultrasonography and computed tomography were performed 6 months later with no change seen.

From the Department of Pediatrics, University of Mississippi Medical Center, Jackson.
Denouement and Discussion

**Hepatic Hemangiomatosis**

Figure 1. Abdominal ultrasonogram reveals numerous, 1- to 4-cm ovoid hypoechoic hepatic lesions.

Figure 2. Abdominal computed tomographic scan shows numerous, spherical intrahepatic lesions (left) that enhance with intravenous contrast medium (right).

Hepatic hemangiomatosis (HH) is a rare condition of infancy with the potential for severe, life-threatening complications. Hepatic hemangiomatosis may occur as part of diffuse neonatal hemangiomatosis, characterized by hemangioma of the skin and at least 2 visceral organs. Hepatic involvement in diffuse neonatal hemangiomatosis occurs in nearly two thirds of cases. Less commonly, HH may be present without any other organ involvement.\(^1\) Most patients have signs and symptoms of high-output congestive heart failure (CHF). The hemangioma serves as multiple arteriovenous fistulae, decreasing peripheral vascular resistance and increasing cardiac output. Other complications of HH are jaundice, Kasabach-Merritt syndrome, hemorrhage, and hypofibrinogenemia.\(^6\) Hepatic hemangiomatosis is clinically and radiographically indistinguishable from hepatic hemangioendothelioma, and differentiation depends on histologic evaluation. Hemangioendotheliomas have a predominance of cellularity to vascular space.\(^6\) Hemangioendothelioma is the most common benign hepatic tumor of childhood and the most common hepatic tumor of the first year of life.\(^7\) The importance of distinction between HH and hepatic hemangioendothelioma rests on the potential for sarcomatous change in the latter condition.\(^8\)

The natural history of HH parallels that of cutaneous hemangiomas, with spontaneous involution during early childhood. Asymptomatic patients require no specific treatment. However, follow-up imaging studies are indicated to evaluate resolution of the lesion and to exclude hepatic endothelioma and the potential for malignant change.\(^9\)

In patients with CHF resulting from HH, therapeutic intervention is indicated. Initial treatment of CHF in patients with HH with diuretics and digitalis was disappointing, with mortality rates of nearly 90%.\(^3\) Treatment with corticosteroids and digitalis improves the outcome of CHF in patients with HH\(^10-12;\) however, this treatment is not universally effective.\(^1,13-16\) Surgical removal is prohibited by the extensive hepatic involvement. Interruption of blood flow to the hemangioma may be successfully accomplished by ligation of the hepatic artery;\(^1,3-15,15\) though ligation of the main hepatic artery at its origin from the celiac axis does not interrupt all arterial flow to the liver as there is collateral flow from the left gastric artery and the superior mesenteric artery. More recently, embolization has been accomplished with a variety of agents such as polyvinyl alcohol, silicone balloons, metal coils, and Spongostan (Ferrosan Co, Copenhagen, Denmark).\(^17-19\) Radiotherapy has been used for children who have not responded to medical therapy or have been too ill for surgery\(^16,20-22;\) however, the possibility of long-term sequelae of hepatic irradiation makes radiotherapy an undesirable form of treatment in children.

**REFERENCES**