JOINT DECISIONS

The admitting diagnosis was septic arthritis. Bilateral arthrocenteses appeared inflammatory. Naproxen and low-dose prednisone were added.

Cultures and liver function were normal, as were WBC and platelet counts. However, WBC bands increased to 56% from 15%, and hemoglobin dropped from 3g to 8.6 g/dL in the first 3 days of admission. Serologies for rheumatic diseases, HIV, and other viruses were negative.

Acute renal failure and liver function abnormalities developed concurrently. Naproxen was discontinued.

After admission to the ICU, "in the course of 10 days, everything worsened," noted Paulo B. Pinho, M.D., a Staten Island, N.Y., pediatrician and internist. Dr. Pinho reported the case in a poster session at the annual meeting of the Federation of Clinical Immunological Societies in Boston.

The patient's first few days in the ICU were marked by hypotension (BP 90s/30s), tachycardia (110-115 bpm), persistent fevers (>101° F), pancytopenia with bands of 88%, and coagulopathy. Her erythrocyte sedimentation rate (ESR) was low (5 mm/hr), but C-reactive protein (CRP) was elevated (24 mg/dL). Transaminases had climbed to an aspartate transferase level of 2653U/L and alanine transferase of 504 U/L. Ferritin increased to >2,000 ng/mL (fractionated 152,197).

Given right upper quadrant pain and hepatosplenomegaly, an abdominal ultrasound was performed to look for intra-abdominal causes of presumed septic shock. It revealed only sludging.

Admitting diagnosis to the ICU was septic shock. Other diagnoses considered included adrenal insufficiency (potentially induced by corticosteroid withdrawal), thrombotic thrombocytopenia purpura (TTP, worsening renal function and altered mental state), and macrophage activation syndrome (MAS).

All cultures were negative; sepsis was rejected. The absence of microangiopathic hemolytic anemia ruled out TTP. Cortisol levels were consistent with the level of systemic inflammation, so adrenal insufficiency was also abandoned.

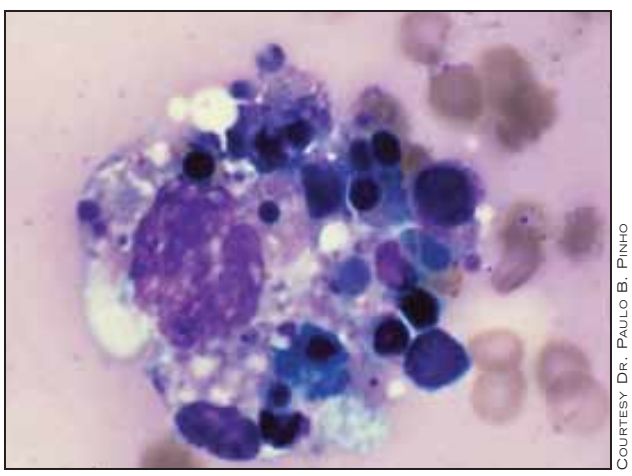
A bone marrow biopsy showed variable cellularity (40%-60%) with prominent hemophagocytosis. This finding, along with the markedly elevated ferritin, low ESR, yet elevated CRP suggested that the patient's previously uncharacterized febrile illnesses may have been a subtle presentation of systemic onset juvenile idiopathic arthritis (S-JIA).

The physician team entertained a diagnosis of JIA flare at this point, but the decreased counts of platelets and other cell lines, massive coagulopathy, and markedly elevated ferritin suggested macrophage activation syndrome (MAS), according to Dr. Pinho.

MAS is a relatively rare but potentially fatal complication of S-JIA and other chronic rheumatologic diseases. Its incidence is uncertain; the literature documents only hundreds of cases in patients with S-JIA, said Dr. Pinho. Reported mortality rates range from 8% to 22%.

The cause of MAS is also unclear, but it may be precipitated by infection or medication changes. "The clinical signs and symptoms can be easily explained by the surge of numerous cytokines and chemokines produced by activated macrophages and T cells," said Dr. Pinho.

MAS presents as a confusing constellation of signs and symptoms. It is often characterized by well-differentiated hallmark macrophages showing hemophagocytosis in the bone marrow. But there are no firm diagnostic criteria.



COURTESY DR. PAULO B. PINHO

A bone marrow biopsy done at ICU admission showed variable cellularity with prominent hemophagocytosis.

Angelo Ravelli, M.D., and colleagues recently proposed preliminary diagnostic criteria for differentiating MAS from a disease flare in patients with S-JIA.

The researchers performed a systematic evaluation of the sensitivity and specificity of findings in 74 patients with MAS and 37 patients with S-JIA who had disease flares (J. Pediatr. 2005;146:598-604).

Their criteria focus on laboratory findings, since clinical symptoms of MAS often arrive late. They suggest the presence of any two of the following four criteria is diagnostic: decreased platelet count ($\leq 262 \times 10^9$/L); elevated levels of aspartate transferase (>59 U/L); decreased WBC count ($\leq 4.0 \times 10^9$/L); or hypofibrinogenemia (≤ 2.5 g/L).

Hyperferritinemia may also be highly diagnostic. Bone marrow biopsy for macrophage hemophagocytosis should be reserved for confirmation of only doubtful cases because hemophagocytosis is not always present in MAS, they wrote.

Other clinical criteria include CNS dysfunction, hemorrhages, and hepatomegaly.

The patient was bolused with intravenous methylprednisolone and started on cyclosporin A. Intense blood product, ventilatory, and fluid support were essential. Not until day 14 did she begin to improve. By day 18 she was extubated and on day 31 discharged from the hospital with a prednisone taper and cyclosporin.

The extraordinary and swift transformation of a healthy-appearing, young female with a sore knee and sore throat to a desperately ill patient with multiple organ failure points out the need for early recognition and prompt action, two essential components to good outcomes in this disease, according to Dr. Pinho.

His colleagues included Miguel Rodriguez, M.D.; Davida Menasha, M.D.; Manal Youssef-Bessler, M.D.; Bhavna Suri, M.D.; Allen Blaivas, M.D.; Ralph K. Messo Jr., D.O.; Rajendra Kapila, M.D.; and Leonard Bielory, M.D., of New Jersey Medical School and Staten Island University Hospital, New York.

—Colin Nelson

Concierge Care Can Work for Wealthy or Indigent Patients

BY JENNIFER SILVERMAN
Associate Editor, Practice Trends

Garrison Bliss, M.D., doesn't believe that so-called concierge care has to involve a $4,000 yearly fee.

The Seattle internist charged patients only $65 a month when he opened his practice in Washington state in 1997—one of the first practices in the country to adopt the concierge care model. His current monthly fee is $85. Patients who receive medical services don't get a bill and neither do their insurance companies, he said.

Traditionally associated with high fees and a limited and wealthy patient base, concierge care—now often called "retainer care"—is morphing into a number of different types of practices, according to Matthew Wynia, M.D., an internist and director of the American Medical Association's Institute for Ethics. So many different types of retainer care have emerged that the trade association for concierge care practices changed its name to "the Society for Innovative Medical Practice Design," he said.

There is a misconception that retainer care is elitist, that patients don't want to pay for it, and that it's something they can't afford, said Marcy Zwelling-Aamot, M.D., an internist who runs a retainer practice in Long Beach, Calif. The 460 patients who belong to her "choice care" program pay a $1,500 yearly fee—but can pay it in monthly installments. "That's less than what they pay for car insurance, a little more than $100 a month," she said in an interview.

For patients who cannot afford the retainer, she provides free care in exchange for volunteer work at a 501(c)3 organization such as a cancer foundation. "It really is a nice exchange. Some of my patients have gotten really involved in the volunteer work—one who was volunteering at the hospital called me and said she wanted to work there."

About 10% of her patients take part in the program.

Dr. Bliss also cares for indigent patients. Those who can't pay the monthly fee fill out a form indicating what fee they can afford. "Whatever their answer is, that's the price they pay," he said.

From the start, he assumed that 10%-15% of his patients would be indigent, he said.

"If every doctor had 10%-15% of their practices with people who couldn't afford it, that would go a very long way toward solving the problem" of the poor getting health care, he said. Plus, there would be no government programs to supervise the practice, no insurance costs, and no billing involved.

Some retainer practices cater to specific segments of the population. John Levinson, M.D., a cardiologist at Massachusetts General Hospital in Boston, runs a "hybrid" hospital/office-based practice that includes both retainer and nonretainer patients.

"The way my day works is I drive to the hospital at 5 in the morning, see my inpatients until 8 a.m., then have a regular office day," where he sees patients that are on Medicaid and other types of insurance, and his retainer patients. At the end of the day, he goes back to the hospital to check on in his inpatients.

There are two groups of patients within the small group of 40 retainer patients he sees. Most see Dr. Levinson as their primary care physician. However, a smaller group sees him for cardiac care only. "Some—about 25—use me for primary and cardiology care, and the others are just cardiology patients."

Those who want comprehensive care pay a higher retainer fee than the cardiology only patients, he said. He would not disclose the fee.

Pediatrician Scott Serbin, M.D., who established a retainer practice for children in December 2004, decided to "tier" his fees based on the age of the child.

Some physicians who spoke with this newspaper said they doubted that any type of retainer medicine would become a major trend.

"It doesn't bode well for medicine, and it smacks of elitism, but in its defense, it is what America has pushed some doctors into doing," Charles Scott, M.D., a pediatrician in Medford, N.J., said in an interview.

"Without a doubt [retainer] physicians know that there are ethical dilemmas associated with their practices, that colleagues are really scrutinizing them for their ethics," Dr. Wynia said.

In a recent survey of 83 retainer practices, he found that retainer physicians reported better quality of care and fewer hassles, but they also saw fewer minorities and Medicaid patients, and fewer patients with chronic illnesses than regular practices.

The physician's role "is to provide 24/7 access for our patients—all patients, whether they're on Medicaid, have special health care needs, etc. That's what the medical home is all about," said Garry Gardner, M.D., a pediatrician in Darien, Ill.

Dr. Zwelling-Aamot, who is trained in emergency medicine, said her patients are not compromised by her "round the clock" hours. Her office is next to the hospital, and she always carries her electronic medical records with her. She uses a variety of specialists in the area to cover for her.

This is how medicine used to work, when physicians volunteered at the local hospitals and free clinics, she said. ■