

Homeless Health Care Case Report: Sharing Practice-Based Experience Volume 4, Number 2 December 2008

Decompensated Chronic Liver Disease with Comorbid Treatable Aplastic Anemia in a Homeless Adult Male

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This case report presents a cascade of life threatening complications arising from a chronic hepatitis C infection in a homeless man, originally contracted from contaminated tattoo needles. The patient's chronic liver disease progressed to cirrhosis and comorbid anemia secondary to hypersplenism, chronic GI blood loss, chronic liver disease, folate deficiency, and human parvovirus B19 infection resulting in aplastic anemia.

The medical challenges involved in the diagnosis, treatment, and management of this complex case were further complicated by psychosocial issues associated with the patient's history of prolonged homelessness and by the fact that the clinicians who cared for him had little prior experience with homeless people. The principal author contacted the National Health Care for the Homeless Council for guidance in describing best practices for the care of people experiencing homelessness, with the intent of educating other clinicians and patients faced with similar medical and psychosocial challenges. Clinicians with extensive experience in the care of homeless individuals with hepatitis graciously agreed to contribute to the development of this case report.¹

Background

Hepatitis C: Though rare in its acute form, hepatitis C virus (HCV) is the most common chronic blood-borne infection in the United States; 3.2 million people (1.3% of the population) are estimated to have chronic HCV infection (Armstrong, 2006). Prevalence is highest among non-Hispanic black males, aged 40–49 years. Of reported HCV cases, 60% were blood-borne infections, 20% were related to sexual exposures, and 10% through other known exposures (occupational, hemodialysis, nonsexual household, or perinatal); in the remaining 10% of cases without an identifiable source of infection, low socioeconomic level was an associated risk factor (Greer, 2004). Some homeless people are disproportionately affected by HCV infection, which is strongly associated with the sharing of needles, injection drug use (IDU), previous incarceration, veteran status, fair or poor health, and having multiple tattoos (Nyamanthi, 2006; Edlin & Carden, 2006). Indeed, the reported prevalence of HCV is 10 to 20 times higher in some homeless sub-groups than in the general US population, ranging from 22% to 80%, with higher rates among those with a history of IDU (Gish, 2005; Greer, 2004; Cheung, 2002).

Cirrhosis: Untreated chronic hepatitis C can lead to cirrhosis of the liver, in which normal liver cells are replaced by scar tissue. Alcoholism and hepatitis C, two major causes of cirrhosis, are overrepresented

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among homeless people. Loss of liver function secondary to cirrhosis can result in the buildup of toxins in the brain, dulling mental functioning and causing personality changes, coma, and even death. Symptoms include neglect of personal appearance, unresponsiveness, forgetfulness, trouble concentrating, and changes in sleep habits. (NIH, 2000)

Anemia: Chronic liver disease is also one of many causes of anemia, characterized by low concentration of hemoglobin in the blood. Poor nutrition (including iron deficiency), impediments to personal hygiene (increasing risk for infection/malnutrition secondary to infestation), frequent exposure to infection in overcrowded living situations, and financial barriers to medical care (including lack of health insurance) contribute to a higher prevalence of anemia among people living in poverty. Major causes of anemia include loss of blood (through heavy menstrual bleeding, wounds, or gastrointestinal ulcer), iron deficiency, chronic disease, kidney disease, pregnancy, poor nutrition (vitamin/mineral deficiency) and alcoholism. Uncommon causes of anemia include: bleeding disorders, liver disease, thalassemia, infection, cancer, arthritis, enzyme deficiency, sickle cell disease, hypothyroidism, and toxins (e.g., benzene exposure). (Naibili, 2008)

Failure of the bone marrow to produce blood cells results in *aplastic anemia*. Known causes include exposure to chemicals, drugs, viruses, radiation, immune conditions, pregnancy, paroxysmal nocturnal hemoglobinuria, and inherited disorders. In patients with hemoglobinopathies or hemolytic anemias, a decrease in the reticulocyte count to less than 1% (usually to 0%) may precipitate an aplastic crisis. Parvovirus B19 is the only known infectious cause of aplastic crisis. (Cunningham, 2006)

Parvovirus B19: Human parvovirus (B19V) is a treatable cause of aplastic anemia that can be easily missed by clinicians who are not looking for it. In individuals with an underlying hemolytic disorder (in which red blood cells are destroyed faster than the bone marrow can produce them), B19V causes transient aplastic crisis. In immunocompromised individuals, persistent B19V infection is manifest as pure red cell aplasia and chronic anemia that is potentially fatal (Heegaard & Brown, 2002). This comparatively benign etiology of aplastic anemia can be uncovered through a careful history, physical examination, and diagnostic testing. Treatment with intravenous immunoglobulins results in normalization of both the reticulocyte count and hematocrit within a few weeks.

A common infectious pathogen worldwide, B19V can be transmitted via the respiratory tract, through the transfusion of blood or blood components, or from mother to fetus. Homeless people living in congregate settings are potentially at high risk for contracting B19V infection, which has been reported in persons with chronic hepatitis B or C. There is no vaccine or medicine that can prevent B19V infection (Fifth's Disease). Frequent hand washing is highly recommended to decrease the risk of infection; but excluding infected individuals from work, child care centers, or schools is unlikely to prevent the spread of the virus, as people are contagious before they develop apparent symptoms (e.g., rash).

Case Description

This case involves Mr. X, a 48-year-old homeless man with decompensated chronic liver disease secondary to hepatitis C, complicated by comorbid anemia of multiple etiologies. Mr. X is an undocumented foreign national of hispanic origin (country unknown) with limited education who engaged in unskilled labor prior to coming to the United States. Since entering the U.S., he has experienced a long history of homelessness. After his health had deteriorated significantly, he finally sought help from a relative in Pennsylvania. The first clinical encounter with this patient occurred in September 2006.

Presenting complaints:

Mr. X presented to a primary care clinic in Pennsylvania with the following complaints: excessive fatigue, loss of appetite, abdominal pain upon eating, weight loss, and intermittent mild fever. The patient's affect was depressed.

Reported symptoms: severe, progressive fatigue of one month's duration; progressive weakness and shortness of breath while engaging in any kind of physical exertion (the patient had to crawl or walk with the support of a wall to get to the bathroom) and dizziness (whenever he tried to get out of bed). The patient's weakness and persistent fatigue progressed until he was unable to get to the bathroom at all and became permanently bed-ridden. (All of these symptoms are consistent with a diagnosis of anemia.)

Mr. X reported that he had not felt like eating for a month; whenever he ate something, he experienced severe abdominal pain (which may be a symptom of Cryoglobulinemia and associated vasculitis secondary to chronic hepatitis B or C). The patient could not point to the exact origin of pain, which he described as occurring all over the abdomen (like colic); it usually occurred during the evening hours and was always triggered by eating. The patient's discomfort was not increased by pressing on the abdomen and nothing seemed to relieve it. Hyoscine and a narcotic analgesic had minimal effect. The pain was associated with mild to moderate nausea.

The patient had lost 20 pounds during the previous month, which he attributed to poor intake due to abdominal pain, anorexia, nausea, and fever. He reported having a mild, intermittent fever during the last month, particularly during the evening, which he attributed to fatigue. He reported no recent history of cough, chest pain, diarrhoea, headache, confusion, or of muscle, bone, or joint pain.

Medical history:

Confirmed diagnoses: chronic liver disease with cirrhosis, hepatitis C, and hypertension.

The patient reported having 15–20 episodes of hematemesis (vomiting blood) and hematochezia (black stools) during the past 20 years. Ten years ago, he had surgery in his home country to insert a porto-caval shunt following multiple episodes of intractable bloody vomiting and black diarrhea. No medical records of this procedure were available. The surgery may have involved a blood transfusion. An obstruction of the blood flow in the constricted and cirrhotic liver caused a blood vessel to rupture, due to back flow in the gastrointestinal and portal vessels.

The patient's current medications and dietary supplements include Atenolol (for hypertension), vitamin K, vitamin E, and multivitamins. He has not adhered well to treatment because of lack of resources to obtain prescribed medications, and depression.

Psychosocial history:

Mr. X is currently unemployed. After a period of prolonged homelessness, he has been living in his relative's home for one month in an over-crowded environment. He does not have a partner or children, which he attributes to his liver disease. There is no reported history of sexual promiscuity (important to rule out because hepatitis B and C are transmitted by sexual contact and are prevalent in homeless populations). He does not like doctors very much.

The patient reports mild to moderate depression, difficulty sleeping, and irritability, which he attributes to his long history of liver disease. He does not like being homeless and chronically ill, but he reports no suicidal intentions. Lack of employment has also made his life worse. The reasons for unemployment mentioned are physical incapacity and lack of vocational skills.

There is no reported history of exposure to occupational or environmental hazard (relevant to the etiology of aplastic anemia) or to illicit drug or alcohol use (which are strongly associated with hepatitis

and cirrhosis in the US). The patient admitted to being an occasional smoker but said he quit 20 years ago after he began to vomit blood. Many smokers with liver disease have a distaste for tobacco and quit smoking (McPhee & Papadakis, 2009).

Mr. X did not have health insurance and was reluctant to seek available social services. He did not agree to have a psychiatric evaluation. His clinicians did not refer him to a mental health professional because of the urgency of addressing his medical conditions.

Physical examination:

Appearance: Bedridden, middle-aged man looking tired; tattoo mark visible on his left upper arm (possible cause of hepatitis C). Signs of pallor, jaundice, palmar erythema, spider nevi, ecchymosis, and cheilosis. Brown coated tongue with no atrophy or inflammation. No signs of clubbing, koilonychia, splinter hemorrhages, oslar nodes, cyanosis, or lymph nodes enlargement evident.

Temperature: 100.4°F; blood pressure: 140/90

Cardiovascular system: bounding pulse, evidence of 3rd heart sound, no murmur.

Central nervous system: higher mental function, cranial nerve normal, gait normal.

Gastrointestinal system: Upper border level of liver normal; lower border is 1 inch down the costal margin; surface is rough; no liver tenderness. The spleen is 2 inches below the costal margin; no clinical ascites, no general tenderness.

Respiratory system: B/L normal vesicular breathing.

Diagnostic test results:

Complete blood count: hemaglobin very low, hematocrit low, RBC mass low, WBC count low, platelet count low, reticulocyte count low, corrected reticulocyte index low, MCV 95 fl, ESR slightly increased.

Peripheral smear examination: peripheral smear anisocytosis with increased red cell distribution width, multilobular polymorph on smear

Serum iron studies: ferritin normal, transferrin and iron low.

Serology and specific blood studies: IGM parvovirus B19 positive. RBC folate low. B12 normal, PT40 sec. Normal copper profile, ANA, and TSH. Blood culture showed no evidence of bacterial infection.

Liver function tests: albumin low, other liver function tests indicate viral hepatitis.

Kidney function tests: mild derangement. Liver biopsy: histology of viral hepatitis.

Bone marrow biopsy: hypocellular with normal iron store.

Treatment/intervention:

Treatment goals:

Medical: Recovery from life threatening condition, improvement of energy level.

Psychosocial: Improvement of depression, physical and social rehabilitation; end homelessness through social, occupational and psychological rehabilitation.

The patient was hospitalized and admitted to the ICU where the following treatments were administered: Red cell transfusion, iron, folic acid, oral B12, IV immunoglobulin for parvovirus B19, IV vitamin K, erythropoietin (expansive), respiratory isolation/prophylactic antibodies when WBC count too low, acetaminophen as needed, proper diet/intake. The patient's aplastic crisis was resolved following treatment and he was discharged to the home of relatives in the U.S. who arranged for him to return to his country of origin. The clinicians caring for him did not know about medical respite care or its availability in the community. The only treatment option to restore liver function is a liver transplant, which is unavailable to persons without a stable living situation, even if financial barriers were overcome.

Discussion

This case focuses on the experience of a 48-year-old Hispanic male with decompensated liver disease secondary to chronic hepatitis C which was severely complicated by comorbid anemia and treatment barriers related to the patient's homelessness and undocumented status. The case report vividly illustrates the potentially life-threatening consequences of tattoo marking with unsterilized needles exacerbated by aplastic anemia secondary to parvovirus B19 infection, poor diet, financial barriers to health care, and poor adherence to medical treatment.

Differential diagnosis of anemia:

ETIOLOGY OF ANEMIA

Iron deficiency anemia is the most common anemia worldwide. Its primary causes are decreased intake, poor diet, and worm infestations. A retrospective review of the hospital records of New York City children aged 3 months through 6 years showed that homeless children were more likely than others to have an elevated erythrocyte protoporphyrin (EP) level, consistent with iron deficiency (Fierman et al., 1993).

Folate deficiency anemia can result from the lack of folate (folic acid), which is necessary for red blood cell formation and growth. Folate is destroyed during the process of cooking food) and must be replenished continuously. Folate deficiency is compounded by any bodily process that increases metabolism.

Anemia of chronic disease vs. anemias of liver/ kidney/ endocrine system: Anemia of chronic disease, the most common normocytic anemia (MCV 80-100 fl), is distinct from anemias of the liver, kidney, or endocrine system. Pathogenic serum levels that are pathognomic of anemia of chronic disease—i-e, increased or normal ferritin, decreased iron, decreased transferrin—are sometimes lacking in anemias associated with the liver, kidney, or endocrine system. Liver, kidney or endocrine diseases also decrease the life span of red blood cells by increasing harmful metabolites as well as decreasing production or increasing resistance of erythropoitin.

Other causes of normocytic anemia are aplastic anemia, hemolysis, hemorrhagic diseases, fluid overload, or the combination of microcytic and macrocytic anemia. Examples of microcytic anemia (<80 fl)12 are iron deficiency anemia and thalassemia. Examples of macrocytic anemia (>100 fl) In early stages all anemia could present as normocytic). Medicines could be responsible for anemias, so drug history is important.

One of the more serious complicating factors was the development of anemia of multiple etiologies, rarely seen in a single patient. Thus, the differential diagnosis of this patient's multiple and complex health problems was especially challenging.

Proper diet is essential to the prevention and management of anemia. Malnutrition can lead to iron deficiency anemia, folate deficiency anemia, or b12 deficiency anemia, as well as to further complications.

Ordering appropriate diagnostic tests is important to assure the most cost-effective treatment strategy. If anemia is suspected in a patient with decompensated chronic liver disease, the recommended workup is to get the corrected reticulocyte index, peripheral smear examination, MCV, and red blood cell distribution width, and then proceed according to test results.

- 1) One of the first tests performed should be the **corrected reticulocyte index**; if elevated, the cause is increased destruction/loss of RBC. If the corrected reticulocyte index is normal or low, then look for a cause related to decreased RBC production such as bone marrow insufficiency, iron deficiency, or anemia of chronic disease. In iron deficiency anemia, serum iron is low, ferritin is low, and transferrin is high.
- 2) Next perform the Coombs test; if positive, the cause is immune mediated hemolysis. If negative, look for mechanical damage (RBC membrane defect, hemoglobinopathy, RBC enzyme deficiency or Paroxysmal Nocturnal Hemoglobinuria) or loss of RBC.

- 3) A peripheral smear may reveal the cause of anemia by the morphology of red blood cells (RBC).
- 4) A bone marrow biopsy diagnoses bone marrow infiltration, iron deficiency, ineffective erythropoiesis of megaloblastic anemia (Hyper cellular marrow), and aplastic anemia (Hypo cellular marrow). Erythropoietin is effective most of time in all anemias, where the cause is decreased production of erythropoietin.
- 5) Confirmatory tests for hemolytic anemias: The confirmatory test for sickle cell disease is electrophoresis; for G6PD deficiency: RBC G6PD (may be normal in the acute phase); for DIC: coagulation profile; for pyruvate kinase deficiency: RBC P-50 screening and RBC pyruvate kinase (confirmatory); for hereditary spherocytosis: theosmotic fragility test; for mechanical hemolysis: schistocytes in the peripheral smear. (In severe hypersplenism, a splenectomy is done, if the patient's condition permits.)
- 6) Confirmatory tests for parvovirus B19 infection: If diagnostic test results confirm a diagnosis of aplastic anemia, consider B19V infection as a possible cause. The treatment of aplastic anemia is entirely changed if there is comorbid parvovirus. The result of missing the parvovirus diagnosis could be lifelong aplastic anemia rather than aplastic anemia that can be resolved. If the patient was exposed to benzene and got aplastic anemia, it is untreatable; but aplastic anemia secondary to parvovirus is treatable.

In this case, there were indications that parvovirus B19 infection was a likely cause of the patient's aplastic anemia. Use the IgM test to confirm a B19V infection in an immunocompetent patient whose infection remains detectable for 3 months; use PCR for an immunocompromised patient. Giant pronormoblasts on the blood smear or in the bone marrow aspirate are sensitive, not specific. Intravenous immune globulin and erythropoietin are effective in resolving aplastic anemia secondary to B19V infection.

HCV progressing to cirrhosis with some degree of anemia (and often co-occurring renal failure) is not an unusual scenario for providers working with homeless patients. The main medical lesson of this case study is to determine the etiology of comorbid anemia because of its important implications for treatment and prognosis. For patients with a history of homelessness, chronic liver disease and comorbid anemia, clinicians are advised to have a high index of suspicion for parvovirus B19, which is a treatable cause of aplastic anemia.

Recommended practice adaptations for homeless patients:

Individuals experiencing homelessness have psychosocial issues in addition to medical issues that need to be factored into the plan of care. These patients often benefit from the services of a multidisciplinary clinical team that includes behavioral health care providers and case managers. Clinicians experienced in the care of homeless persons with complex medical conditions report that they frequently see patients with decompensated chronic liver disease and comorbid anemia, and fear they will be seeing significantly more such patients in the future, given the high incidence and prevalence of HCV infection among people with a history of incarceration, injection drug use, and homelessness who have limited access to antiviral treatment.

Health Care for the Homeless providers recommend the following practice adaptations to prevent, resolve, or ameliorate chronic liver disease secondary to hepatitis C infection—including HCV prevention education, harm reduction, screening, referral to specialists and treatment if warranted, early discharge planning from institutional care, integration of primary and behavioral health care, and palliative care for patients with advanced disease.

Take Home Messages for Primary Care Providers

- 1. Fundamental issues that threaten the health of people experiencing homelessness and compromise their capacity to recover from serious illnesses (Bonin et al., 2004):
 - Unstable housing lack of privacy, risk of abuse, theft of medications, and no place to lie down during the day.
 - Limited access to nutritious food and water, increasing their risk for malnutrition and dehydration;
 - Higher risk for physical and sexual abuse (prior to and after becoming homeless).
 - Higher prevalence of behavioral health problems, serious and complex medical conditions, and physical/cognitive impairments.
 - Limited access to immunizations and other preventive health care.
 - Health care (especially specialty care) is inaccessible and/or lacks continuity and consistency (due to lack of health insurance/transportation/housing, transience).
 - Barriers to disability assistance.
 - Cultural and linguistic barriers.
 - Limited education/literacy.
 - Lack of social supports.
 - Chronic stress.

2. Recommendations for the initial clinical evaluation of a homeless individual in whom chronic liver disease is suspected:

- a. Explore the following issues in the history (Bonin et al., 2004):
 - Basic living conditions: access to food, water, rest rooms, place to store medications; exposure
 to toxins, allergens, infection; threats to health/safety, place to lie down in the day time.
 - Possible substance use (alcohol is poison for an already diseased liver and can cause anemia).
 - Diet and nutrition/hydration.
 - Sexual history and sexually transmitted diseases, needle sharing, transfusion and tattoos (hepatitis is blood borne).
 - Family history of liver disease/anemia.
 - Whether sexual partner has been informed of the patient's STD.

b. Perform a comprehensive physical and dental examination (Bonin et al., 2004):

- Give patients the option of deferring a genital examination until comfort level allows, recognizing that many homeless patients have been victims of physical and sexual violence.
- c. Diagnostic tests should include (Greer, 2004; Bonin et al., 2004):
 - Screening for exposure to HCV, recognizing that homeless people are at extremely high risk for HCV infection and that its consequences are significant and long term.
 - Simple Screening Instrument for Alcohol and Other Drug Use (SSI-AOD).
 - PHQ-9 depression screen and GAD 7 anxiety screen for homeless patients and all patients with chronic disease.
 - Mini-Mental State Examination (MMSE) to identify cognitive impairment.
 - STD screening: for chlamydia, gonorrhea, syphilis, HIV, HBV, HCV, trichomonas, bacterial vaginosis, and monilia.
 - Baseline labs: including liver function tests and tuberculin test (PPD).

d. Counsel the patient about (Greer, 2004; Ed Farrell, MD):

- Early prevention of HCV: education about transmission of infection from needles
- Reduction of alcohol/other substance use/smoking
- Nutrition
- Trauma/interpersonal violence
- Obesity
- Depression and other mental health problems
- Stress management
- Entitlements and other benefits
- Available social services
- Meeting basic needs
- Treatment adherence and immunization

3. Recommendations for the treatment and management of hepatitis C in homeless patients:

- a. Treatment & management of HCV infection (Greer, 2004, Ed Farrell, MD):
 - Vaccinate all HCV patients against hepatitis A and B.
 - Perform a careful and thorough medical and neuropsychiatric evaluation for all potential candidates for anti-viral therapy.
 - For patients with HCV infection, initiate antiviral treatment if not contraindicated.
 - Contraindications to antiviral treatment: cirrhosis, active alcohol use.
 - Active IVDU is not a contraindication to treatment, but may compromise adherence.
 - Regular attendance at clinic visits and adherence to therapy are essential for treatment.
 - Side effects of HCV treatment are significant and appropriate management is essential.
 - Refer patients with cirrhosis to a hepatologist and other specialists as needed (e.g., infectious disease specialist, gastroenterologist), recognizing that lack of health insurance coverage is a serious obstacle to specialty care in many parts of the U.S.

b. Integration of primary and behavioral health care (Jan Caughlan, LCSW-C):

- Facilitate the resolution of psychosocial issues by arranging a "warm hand-off" from the primary care provider to a licensed behavioral health care provider who assesses the patient's psychiatric and social service needs and works with the provider to incorporate these issues into the medical plan of care.
- The behavioral health care provider should work with clinicians on relationship building and enhancing the patient's motivation to address psychiatric, housing and income issues.
- This integrated model of care will provide additional support to patients with advanced disease who require palliative care.

c. Discharge planning (Bechara Couchair, MD; James O'Connell, MD):

- Recognize the importance of early hospital discharge planning for homeless patients. Work
 with the hospital discharge planning team, beginning at admission and daily thereafter, to
 identify housing/long-term care options and establish linkage to essential community services
 and supports following discharge.
- If possible, contact family members in the patient's State or country of origin to participate in discharge planning; consider contacting the embassy of the country of origin if outside the US.
- Refer patients without homes to a medical respite care facility, if available, where they can find a safe place to heal and recover, instead of being discharged to a shelter or the streets.

"Homeless patients face enormous obstacles to specialty and other health care clinics, including language barriers and a lack of insurance, transportation, and housing.... Much work remains to be done, especially in screening and development of appropriate systems of care for homeless people."

- Pedro Greer, MD, Boston Health Care for the Homeless Program

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