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Date of Service: 04/10/2013 0:00:00
Authored By: Hsu, Evelyn Kanyu, MD

GI Consultation

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GI CONSULTATION

XIE, JIANHUA DRACO
DOB: 09/16/2012 M -MR #: 01-27-55-67

DATE OF ADMISSION: 04/09/2013 DATE OF CONSULTATION: 04/10/2013

REQUESTING PROVIDER: Lincoln Smith, MD, of the Pediatric Intensive Care Unit.

REASON FOR CONSULTATION: We are asked by Dr. Smith of the Pediatric ICU to consult on the care of Jianhua Xie regarding coagulopathy, hypoalbuminemia, ascites, fever and jaundice in a 6-month-old male.

HISTORY OF PRESENT ILLNESS: Jianhua is a 6-month-old male who was previously healthy up until several days ago, when he developed periorbital edema, jaundice and abdominal distension following 3 days of diarrhea. Also, since about a month ago, his weight has gone from the 50th percentile up to the 90th percentile. Parents noted that prior to the development of the diarrheal illness he did not have any jaundice. He was brought first to the primary care provider for evaluation, and subsequently was referred to the SCH emergency room. In the emergency room he developed a fever of 40, Blood cultures were drawn and he was given ceftriaxone. He also had 1 episode of emesis, nonbilious, nonbloody. He received a total of 30 mL/kg of normal saline. Upon laboratory evaluation, he was found to be coagulopathic, with an INR of 5.6, conjugated bilirubine was 4, AST and ALT only mildly elevated at 107 and 97 respectively. He was subsequently admitted to the Pediatric ICU for further workup as well as transitioned to piperacillin-tazobactam for antibiotic coverage.

In retrospect, parents felt that he has become more weak over the last week, and has definitely gained weight faster over the past month. Otherwise, they do not feel that he is any more irritable than normal.

MEDICATIONS:
Home medications: None.
He is currently receiving:
1. Piperacillin-tazobactam 945 mg every 8 hours.
2. Albuterol nebulizer once daily as needed for wheezing.

ALLERGIES: No known drug allergies.

PAST MEDICAL HISTORY: Patient was born 10 days early, but essentially at term. Pregnancy was uncomplicated. He is the 4th child in his family. He did have some issues with diarrhea and vomiting early on. He was switched from cow's milk-based formula to soy milk-based formula and has been on that since 2

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months of age. He also had RSV (respiratory syncytial virus) in January, but did not require, but has had some chronic wheezing since that time. He has otherwise not had any hospitalizations or surgeries.

SOCIAL HISTORY: Patient lives at home with his mother, father, and 3 older siblings, all of whom are healthy. He has had no travel, no exposures, and he is currently at home with his grandmother, and parents deny vehemently that he has received any herbal or eastern medicine, no supplements. The only medications that he may have had access to were vitamins.

FAMILY HISTORY: Noncontributory other than that his father had had some symptoms of vomiting and diarrhea that were consistent with a rotavirus infection over the weekend. There is no other illness. His father brought up that in the last 20 days Jianhua has been taking formula that was ordered online through an independent seller through Amazon. They are concerned that this formula may have been tainted in some way, and would like for it to be examined.

REVIEW OF SYSTEMS: Positive for a history of wheezing following RSV infection. He has had relatively good weight gain. He has jaundice, coagulopathy, hypoalbuminemia, some irritability today. Parents also note that ever since birth he has had what they feel to be greasy stools. No bruising, no bleeding, no petechiae, no skin rash. A 14-element review of systems is otherwise negative except for those mentioned in the HPI.

PHYSICAL EXAMINATION: Temperature at admission was 40. Heart rates have ranged 130-183, respiratory rate ranging 27-60, blood pressures ranging 74-135 over 20-85. His weight on admission was 9.98 kg. In general, this is a crying, vigorous infant male. He has periorbital edema and some lower extremity edema. HEENT exam: He has some mild scleral icterus. Pupils equal, round, and reactive to light. Extraocular movements intact. Cardiovascular: He has regular rate and rhythm. He does have a 2/6 systolic ejection murmur heard best at the left upper sternal border. There is no thrill, no hyperdynamicity. Respiratory exam: Clear to auscultation bilaterally, no wheezes, no rales, no increased work of breathing, no retractions. He does have intermittent coughing. Abdomen is soft, moderately distended, with an easily reducible umbilical hernia. No palpable hepatomegaly; however, I do feel a spleen down to 2 cm below the left costal margin. He does not have any increased superficial vasculature over his abdomen. GU exam does not have any scrotal edema. He is a normal Tanner I male. There is no evidence of any diaper rash. Extremities are prominent for lower extremity edema, no pitting. Neurologically, patient is moving all 4 extremities and his neurologic exam is grossly normal. He does seem irritable, but will calm when held.

LABORATORY EVALUATIONS: Labs are significant for a blood test showing a pH of 7.39, pCO2 28, pO2 venous of 74, bicarbonate level of 16.3. Initially on presentation his sodium level was 134, potassium 5.0, chloride 110, bicarbonate 17. Anion gap was 1. Glucose level has been normal, ranging above 80-120. BUN 9, creatinine 0.2, calcium 1.28, magnesium 2.4, phosphorus low at 3.4, lactates ranging 2.1-3.1. White blood cell count high at 27.0, platelets 146, hemoglobin 8.1, hematocrit 24.8. INR 5.6 with a prothrombin time of 48.6. Fibrinogen is 60, PTT 75. Urinalysis showed a urine specific gravity of 1.021 but a urine pH of less than 5, negative for nitrites, leukocyte esterase, and glucose, positive for trace protein, positive for trace ketones, negative for blood. Stool occult blood was negative.

Blood cultures have been drawn and are still negative. Stool cultures have also been done. Rotavirus was found to be negative. Respiratory viral PCR was positive for rhinovirus/enterovirus. Acetaminophen level was

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less than 10. B natriuretic peptide was 33. Liver enzymes show AST of 107, ALT of 97, alkaline phosphatase of 1270. Uric acid 4.3, triglycerides low at 47, LDH 810 and normal, albumin level 2.3, conjugated bilirubin 4.0, unconjugated 10.3, ferritin 93, total protein low at 4.1, ionized calcium 1.15. Ammonia level 40. PT/INR following 1 dose of vitamin K showed a prothrombin time of 46.3, INR 5.3, PTT 69, fibrinogen 80, and a thrombin time of 19. Bordetella pertussis PCR was negative.

Abdominal ultrasound with Doppler study shows edematous liver exhibiting decreased through-transmission, small echogenic foci throughout the right lobe of the liver, contracted gallbladder; spleen is slightly enlarged, measuring 8.4 cm; patent IVC, hepatic veins, portal vein. There was an incidental note of recanalization of the umbilical vein. There was some small amount of anechoic free intraperitoneal fluid. Differential diagnosis for echogenic hepatic foci includes septic emboli, metastatic lesions, and the possibility of hemorrhagic infarcts.

An echocardiogram done today in the afternoon showed no intracardiac vegetation, no intracardiac shunting, normal valvular structure and function, no pericardial effusion, some mild increase in velocity across aortic, pulmonary, and mitral valves, indicating hyperdynamic or fluid overloaded status.

ASSESSMENT AND PLAN: Jianhua is a 6-month-old previously healthy male with a history of some milk protein sensitivity, who presents with of coagulopathy, jaundice, hepatitis, abdominal distention, and hypoalbuminemia in the setting of fever and diarrhea. Imaging findings, coagulopathy unresponsive to vitamin K, and lack of improvement with antibiotic treatment suggests that there is intrinsic liver dysfunction and possible acute liver failure. What remains perplexing is that his degree of transaminase elevation is not as high as we would typically see with a viral hepatitis-induced liver failure, yet the coagulopathy, jaundice and presence of portal hypertension suggests possible chronic liver disease. Also within our differential, and thinking outside of the realm of liver disease, we continue to worry about sepsis/DIC (disseminated intravascular coagulation) like picture also leading to coagulopathy and elevated unconjugated bilirubin. Hemolysis (although LDH is low) remains a concern. A haptoglobin level may help to evaluate this. . Our differential for acute liver failure would include a viral hepatitis, metabolic disease (tyrosinemia, galactosemia, hereditary fructose intolerance, cystic fibrosis associated liver disease, alpha-1 antitrypsin liver disease), toxin induced disease, autoimmune liver disease, malignancy. Vascular obstruction or cardiac associated liver disease unlikely, as we have normal imaging of the heart and no obstruction seen on Doppler study of the liver. Within the differential for infectious causes of acute liver failure we should evaluate for hepatitis A, EBV (Epstein-Barr virus), CMV (cytomegalovirus), adenovirus, enterovirus, hepatitis B surface antigen, HSV, HHV-6, parvovirus, coxsackie, and varicella. HIV test should be done for pre-transplant work-up. With regards to possible autoimmune processes, his ferritin and triglycerides are not consistent with an HLH (hemophagocytic lymphohistiocytosis) like picture. We will be sending factor levels as well as an alpha fetoprotein. We have an autoimmune panel which is pending. With regards to possible metabolic causes of liver disease, we have requested urine organic acids, urine reducing substances, and urine succinylacetone, as well as a galactosemia screen and serum amino acids and acylcarnitine profile. It is possible that this could be some toxicity or ingestion although extensive questioning has not yielded any untoward exposures. We will be in contact with our Toxicology colleagues regarding father's request to test the formula that he had ordered online.

FAOD not in her scope.

Jianhua's condition is currently guarded. I spent an extensive amount of time discussing with his parents our current plan for workup and possible need for increased intervention and life-sustaining measures should his

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liver failure worsen. We will need to monitor Jianhua very closely with regards to his coagulopathy and support him as needed. We would also need to be very judicious in the amount of IV fluids that he gets and allow him to take enteral intake as long as he is able. I would continue his antibiotics until our blood cultures and urine cultures are known to be negative, as the greatest risk to his life right now would be infection. We will involve the Liver Transplant Team should Jianhua's clinical status start to deteriorate. We should monitor his weights on a twice daily basis and monitor his ins and outs carefully. We should monitor liver function and coagulation panel every 12 hours.

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Evelyn K Hsu, MD 04/11/2013 12:52 P

Evelyn K Hsu, MD , Attending Physician, Gastroenterology

EKH/jmc Doc #2932273 d: 04/10/2013 05:46 P t: 04/10/2013 06:33 P (1554684-)
cc: cc Pediatric Assoc Redmo
Julie E Wen, MD

Child Protective Team

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CHILD PROTECTION TEAM REPORT

XIE, JIANHUA
DRACO

DOB: 09/16/2012 M -MR #: 01-27-55-67

DATE OF CONSULTATION: 10/09/2014

SCAN CONSULTATION

Today, I received a call from Evelyn Hsu, MD, Gastroenterology Transplant attending. She and her team have been concerned that Jianhua initially presented about 7 months ago with acute liver failure. On biopsy, he had micronodular cirrhosis. Extensive testing was done at that time for a cause, and none determined; however, subsequently the clinical evidence of his liver disease has improved. He was last seen last spring, with the plan that he would follow up this fall.

During the course of his evaluations, his father developed concerns that he had a fatty acid oxidation defect. Extensive metabolic testing has been negative so far, yet dad has been unwilling to drop this diagnostic concern. Due to conflicts with the Gastroenterology Service, he also has indicated that he is seeking care elsewhere, including at Pittsburgh and Portland, Oregon.

The Gastroenterology Service is concerned that it would be important to document that, whatever the cause of the liver failure was, it is no longer active. I discussed with Dr. Hsu that, since Jianhua has a very serious medical problem, that failure of his father to seek appropriate follow-up care would constitute medical neglect. I recommended that Gastroenterology connect with dad and set a date this fall by which he must provide documentation that Jianhua is receiving appropriate care elsewhere. Short of that documentation, a Protective Services referral for medical neglect should be made.

Electronically Authenticated by
Kenneth W Feldman, MD 10/10/2014 11:19 A

Kenneth W Feldman, MD , Attending Physician

KWF/rll Doc #3649198 d: 10/09/2014 01:44 P t: 10/09/2014 02:31 P (2034547-)

cc: Evelyn K Hsu, MD
cc SCH Children's Protec