Phone/Email/Verbal Communication

Date of Service:	1/2/2014 08:58 PST
Authored By:	Kuiper,Victoria ,RN (1/2/2014 09:08 PST)
Result Status:	Auth (Verified)
Document Type:	Phone/Email/Verbal Communication

General Phone Note

Type of call	X	**Incoming call. Date/Time:** 1/2/14 @ 0900		**Outgoing Call. Date/Time:**			
		Follow-Up of visit or test result	X	**Parent Question**		**Forms needed**	

Name of caller: (Name, relationship to patient, phone#)	Yong Xie, father, (425)214-1910
Reason for call:	Dad called and e-mailed this RN with concerns of fever, coughing, congestion, and trouble sleeping. Dad reports that Jianhua started having fevers the evening of 12/24 and has had them off and on since then. He has also had viral cold symptoms (coughing, conjection, etc) and has not been sleeping well. Dad is concerned that this illness could damage his liver again, since these symptoms are similar to the symptoms that brought Jianhua into the hospital in April 2013 with RSV/liver failure. Dad had also started giving Jianhua eggs since he last met with Dr. Horslen last month, when the results of the whole exome sequencing was discussed. Dad was also worried that this is, in part, causing Jianhua's illness.
Discussion/update:	Discussed the above/e-mail with Dr. Horslen. There are currently no openings in txp clinic this afternoon, and dad was advised that we do not recommend that patients present to clinic with viral symptoms/fevers, so as to protect our other immunosuppressed patients in our clinic and the surrounding clinics. Dad was advised to return either to his pediatrician or to present Jianhua to the ED for evaluation if he worsened or dad was worried about him. Dr. Horslen is on service and he is available to answer questions the ED may have if he does present there. Dad verbalized understanding and stated that for now, he would continue to watch Jianhua's symptoms and if they worsened, he would bring him to the ED as he did not feel that the PMD was appropriate for this illness at this time. Dad also stated that he would stop giving him eggs for now and only feed him formula until he was seen in txp clinic next week.
Plan:	Dad to bring Jianhua to the ED if symptoms persisted or worsened. Otherwise, we will see him in clinic next week as previously planned.

Chemistry

LEGEND: *=Abnormal, C=Critical, ^ = Interpretive Data, c=Corrected, L=Low, H=High, f=Footnote, T=Textual Result

Result Comments
f17: Zinc Level

Test Performed by:
Mayo Clinic Laboratories - Rochester Superior Drive
200 First Street SW, Rochester, MN 55905
Laboratory Director: Franklin R. Cockerill, III, M.D.

Collected Date	1/22/2014	1/7/2014		
Collected Time	11:50 PST	14:03 PST		
Procedure			Units	Reference Range
Sodium Level	140	140	mEq/L	[135-145]
Potassium Level	5.0	4.7	mEq/L	[3.5-5.5]
Chloride Lvl	104	105	mEq/L	[96-109]
CO2 Lvl	25	22	mEq/L	[18-27]
Anion Gap	16.0	17.7	mEq/L	[8.0-22.0]
Glucose Level	78	80	mg/dL	[60-105]
BUN	11	16	mg/dL	[6-20]
Creatinine	**0.5** H	**0.7** H	mg/dL	[0.1-0.4]
Estimated GFR (Schwartz Bedside)	**63** L H f6	**44** C f16	mL/min/1.73 m2	
Calcium Level,Total	10.4	10.1	mg/dL	[8.7-10.7]
Magnesium Serum	2.2	2.2	mg/dL	[1.8-2.4]
Phosphorus Serum	5.7	6.2	mg/dL	[3.9-6.5]
Bilirubin Total	0.2	0.2	mg/dL	[0.1-1.3]
Bilirubin Conjugated	0.0	0.0	mg/dL	[0.0-0.3]
Bilirubin Unconjugated	0.0	0.0	mg/dL	[0.0-1.1]
Bilirubin Delta	0.2	0.2	mg/dL	[0.0-0.2]
AST (SGOT)	**78** H	**60** H	IU/L	[5-41]
ALT (SGPT)	**49** H	38	IU/L	[6-40]
Alkaline Phosphatase,Serum	**458** H	**408** H	IU/L	[95-380]
Gamma Glutamyltransferase	22	20	IU/L	[15-85]
Ammonia,Blood Venous	-	**46** H	mcmol/L	[9-35]
Albumin Level	4.7	4.7	g/dL	[3.8-5.4]

Result Comments
f16: Estimated GFR (Schwartz Bedside)
Calculated by a Discern Rule
Estimated GFR(Schwartz Bedside) = 0.413 * Height(cm)/SCr(mg/dL)

Collected Date	11/14/2013	8/15/2013		
Collected Time	13:30 PST	15:30 PDT		
Procedure			Units	Reference Range
Sodium Level	141	143	mEq/L	[135-145]
Potassium Level	4.6	4.5	mEq/L	[3.5-5.5]
Chloride Lvl	107	-	mEq/L	[96-109]
Chloride Lvl	-	105	mEq/L	[96-110]
CO2 Lvl	18	23	mEq/L	[18-27]
Anion Gap	20.6	19.5	mEq/L	[8.0-22.0]
Glucose Level	**112** H	62	mg/dL	[60-105]

Glomerular Filtration Rate (GFR)

kidney.org /atoz/content/gfr

12/24/2015

What is GFR?

GFR - glomerular filtration rate is the best test to measure your level of kidney function and determine your stage of kidney disease. Your doctor can calculate it from the results of your blood creatinine test, your age, body size and gender. Your GFR tells your doctor your stage of kidney disease and helps the doctor plan your treatment. If your GFR number is low, your kidneys are not working as well as they should. The earlier kidney disease is detected, the better the chance of slowing or stopping its progression.

What are the Stages of Chronic Kidney Disease (CKD)?

Stage	Description	(GFR)
At increased risk	**Risk factors** for kidney disease (e.g., diabetes, high blood pressure, family history, older age, ethnic group)	More than 90
1	Kidney damage with normal kidney function	90 or above
2	Kidney damage with mild loss of kidney function	89 to 60
3a	Mild to moderate loss of kidney function	59 to 44
3b	Moderate to severe loss of kidney function	44 to 30
4	Severe loss of kidney function	29 to 15
5	Kidney failure	Less than 15

Your GFR number tells you how much kidney function you have.
As kidney disease gets worse, the GFR number goes down.

What happens if my test results show I may have chronic kidney disease?

- A GFR below 60 for three months or more or a GFR above 60 with kidney damage (marked by high levels of albumin in your urine) indicates chronic kidney disease. Your doctor will want to investigate the cause of your kidney disease and continue to check your kidney function to help plan your treatment.
- Typically, a simple urine test will also be done to check for blood or albumin (a type of protein) in the urine. When you have albumin in your urine it is called albuminuria. Blood or protein in the urine can be an early sign of kidney disease.
- People with a high amount of albumin in their urine are at an increased risk of having chronic kidney disease progress to kidney failure. (See chart below)

				Albuminuria categories		
				A1	**A2**	**A3**
				Normal to mildly increased	Moderately increased	Severely increased
				<30 mg/g <3 mg/mmol	30-299 mg/g 3-29 mg/mmol	≥300 mg/g ≥30 mg/mmol
GFR Stages	**G1**	Normal or high	≥90			
	G2	Mildly decreased	60-90			
	G3a	Mildly to moderately decreased	45-59			
	G3b	Moderately to severely decreased	30-44			
	G4	Severely decreased	15-29			
	G5	Kidney failure	<15			

Key to Figure:
Colors: Represents the risk for progression, morbidity and mortality by color from best to worst.
Green: Low Risk (if no other markers of kidney disease, no CKD)
Yellow: Moderately Increased Risk
Orange: High Risk
Red: Very High Risk
Deep Red: Highest Risk

Your doctor may also suggest further testing, if necessary, such as:

- Imaging tests such as an ultrasound or CT scan to get a picture of your kidneys and urinary tract. This tells your doctor whether your kidneys are too large or too small, whether you have a problem like a kidney stone or tumor and whether there are any problems in the structure of your kidneys and urinary tract.
- A kidney biopsy, which is done in some cases to check for a specific type of kidney disease, see how much kidney damage has occurred and help plan treatment. To do a biopsy, the doctor removes small pieces of kidney tissue and looks at them under a microscope.

Your doctor may also ask you to see a kidney specialist called a nephrologist who will consult on your case and help

manage your care.

What is a normal GFR number?

In adults, the normal GFR number is more than 90. GFR declines with age, even in people without kidney disease.See chart below for average estimated GFR based on age.

Age (years)	Average estimated GFR
20–29	116
30–39	107
40–49	99
50–59	93
60–69	85
70+	75

For more information:

Glomerular Filtration Rate (GFR) Brochure

Calculate your Glomerular Filtration Rate (GFR)

If you would like more information, please contact us.

Clinic Note

Date of Service:	1/7/2014 00:00 PST
Authored By:	Chan,Wai as proxy for Hsu,Evelyn Kanyu,MD (4/8/2014 15:16 PDT); Hsu,Evelyn Kanyu,MD (1/9/2014 10:25 PST)
Result Status:	Modified
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Addendum by Chan, Wai on 08 April 2014 15:16

TRANSCRIPTION ERROR PER REVIEW OF THE DICTATION

Replace the incidents of "macronodular" to "mirconodular":
A 15-month-old male with a history of **mirconodular** cirrhosis of unknown etiology.

He is again referred by Peggy Kulpa, MD for evaluation and therapy of his chronic liver disease secondary to **mirconodular** cirrhosis of unknown etiology.

Transplant Clinic Outpt Report

Document may Not be Signed/Finalized. See End of report for Electronic Authentication of Signature.

OUTPATIENT NOTE

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

CLINIC: TRANSPLANT

DATE OF SERVICE: 01/07/2014

CHIEF COMPLAINT AND IDENTIFICATION: A 15-month-old male with a history of ~~macronodular~~ cirrhosis of unknown etiology.

HISTORY OF PRESENT ILLNESS: Jianhua is a 15-month-old Asian male seen today in clinic accompanied by his father and his paternal grandmother. He is again referred by Peggy Kulpa, MD for evaluation and therapy of his chronic liver disease secondary to ~~macronodular~~ cirrhosis of unknown etiology.

Jianhua is 15 months old and has the following problem list:
1. Chronic liver disease secondary to micronodular cirrhosis of unknown etiology, presented with acute on chronic liver failure at 7 months of age. Complete workup including liver biopsy was not revealing of a diagnosis, but we did rule out autoimmune, metabolic, or infiltrative disease. His biopsy showed cirrhosis with bile duct proliferation and electron microscopy did not show any bile consistent with a PFIC (progressive familial intrahepatic cholestasis). Genetic workup has included jaundice chip that showed no mutations and a clinical exome sequencing did not reveal any findings that were consistent with his diagnosis.
2. History of possible pancreatic insufficiency with elevated stool elastase, however, stool elastase was done when he had excessive diarrhea. His baseline stooling is about 3-4 times per day and recently has been increased.
3. Portal hypertension with splenomegaly and a history of ascites, currently on spironolactone; however, splenomegaly has largely resolved and he has not had any ascites or peripheral edema.

4. Fat soluble vitamin deficiency, currently on vitamin supplements, although levels have been increasing.
5. No abnormalities in the NICCD gene with full exome sequencing was done.

Jianhua was last seen in clinic by myself on 11/14/2013. Dr. Horslen met with the family to discuss results of the clinical exome sequencing in December. After meeting with the family to discuss the clinical exome sequencing results, our team felt that there with no sufficient evidence to limit the protein that Jianhua was consuming. He was liberalized from half an egg per day to a full egg each day.

Jianhua was recently ill with fever, cough and wheezing symptoms over the last several weeks and his father has attributed this likely viral illness to the onset of liberalization of protein intake. He was given ibuprofen and Tylenol which seemed to help his fever. He was evaluated by Dr. Kulpa, who was not able to find a bacterial source for his fever. Parents are very concerned that this elevation in his temperature is directly related to introducing eggs into his diet and so had subsequently stopped giving him egg on New Year's Day. Since then, he has had no fever. He has had no vomiting, although he has had increased stooling the last 10 days for his stooling has gone from a baseline of 2-3 times per day to 6-7 times per day. It is not watery, but it is loose. He is eating well, taking a total of 35 ounces of Pregestimil 20 kcal/ounce at 1-2 ounces of water per day. He is taking all manner of foods but is largely restricted. His parents are worried that his illness was brought on by food allergy. Now he intermittently will have wheezing with agitation.

CURRENT MEDICATIONS:
1. Vitamin E 50 units once daily.
2. Ursodiol 80 mg 2 times daily.
3. Cholecalciferol 1000 units once daily.
4. SourceCF 1 mL once daily.
5. Spironolactone 12 mg once daily.

The father told Dr. Hsu that it was FAT many times!!! Not protein.
The father told her that it was FAOD, not allergy.

ALLERGIES: No known drug allergies.

PAST MEDICAL HISTORY: As detailed as above.

SOCIAL HISTORY: Unchanged since his last clinic note dated 11/14/2013.

FAMILY HISTORY: Noncontributory.

PHYSICAL EXAMINATION: Weight today is 9.52 kg, height is 76.15 cm. BMI (body mass index) 16.42. He is very watchful and appropriately tearful during his exam. HEENT exam is normal. Sclerae are white, conjunctivae clear. Pupils equal and round. Nasal septum midline. Oral mucosa normal. Neck is supple; no lymphadenopathy. Chest is clear to auscultation in all lung fields with good air movement bilaterally without wheezes, rhonchi, or crackles. Cardiac exam is regular rate and rhythm without any murmurs, gallops, or rubs. Extremities are normal without edema, erythema, swelling, or deformation. Skin is without rashes. Gross and fine motor control are normal, and affect is appropriate. Abdomen is soft, nondistended, nontender, no evidence of any ascites. I am not able to palpate any hepatosplenomegaly. There is no evidence of caput medusae. The umbilical hernia has greatly improved.

LABORATORY EVALUATION: Today shows sodium level 140, potassium 4.7, chloride 105, bicarbonate 22, BUN of 16, which is increased from previously and creatinine 0.7, GFR is calculated at 44, calcium level 10.1,

magnesium 2.2, phosphorus 6.2, conjugated bilirubin zero, AST 60, ALT 38, GGT of 20, albumin level 4.7, white blood cell count of 15.0, platelet level of 286, hemoglobin 13.1, INR is 1.1. Prothrombin time 13.6. His last vitamin D level 23 from 11/14, vitamin A, vitamin E were replete at that time.

ASSESSMENT AND PLAN: Jianhua is a 15-month-old Chinese male known to have micronodular cirrhosis of unknown etiology, likely a congenital finding. Labs continue to improve. He has a recent viral illness the severity of which his parents are ascribing to his recent introduction of increased protein in his diet. He continues on Pregestimil plus MCT, as well as a number of different foods. They have not increased the amount of carbohydrates that he is getting, despite his demands for more food. Parents are extremely worried that giving him increased food or giving him more protein will result in a recurrence of his decompensation, although we have no evidence of metabolic inborn error, and a full genome exome sequencing that has failed to discover mutations in the genes for PFIC (progressive familial intrahepatic cholestasis) disease and NICCD.

1. Management of chronic liver disease and fat soluble vitamin deficiency: I would like to continue his ursodiol, but with resolution of his ascites, I would like to stop the spironolactone, especially in the setting of increased creatinine. I would like to continue vitamin E, cholecalciferol and SourceCF. We will follow his vitamin levels and perhaps consider decreasing this in the next month.
2. Increased BUN and creatinine. I urged increased hydration at this visit. We will follow up the labs again in 1 week's time and consider further nephrology workup of his elevated creatinine and decreased GFR if these are not resolved.
3. Jianhua should return to clinic in 3 months' time for his next visit with labs prior to this visit.

Electronically Authenticated by
Evelyn K Hsu, MD 01/09/2014 10:25 A

Evelyn K Hsu, MD , Attending Physician, Gastroenterology

EKH/trl Doc #3277281 d: 01/07/2014 05:28 P t: 01/08/2014 08:11 A (1789926-)
Location: TXP
cc: Peggy J Kulpa, MD