

به نام خدا

فرم اطلاعاتی بیماران نارسایی حاد کبدی

مرکز تحقیقات گوارش، کبد و تغذیه کودکان

*فیلد های اجباری

@ پاسخ های تائیدی

* کد پزشک معالج (فوق تخصص گوارش کبد کودکان): @

* کد ملی بیمار: @

* نام و نام خانوادگی بیمار: @

* تاریخ تولد: @

* شهر و استان محل تولد: @

* جنس: @

* نژاد: @

* شماره پرونده: @

* شماره تماس: @

* آدرس: @

۱ - سن تشخیص نارسایی حاد کبدی *

۲ - وضعیت رشد و تکامل * @

۱ - سن تشخیص نارسایی حاد کبدی *				
۲ - وضعیت رشد و تکامل * @				
Z score BMI (≥2 year)	Z-Score Wt/Lt (<2 year)	دورسر (سانتی متر)	قد (سانتی متر)	وزن (کیلوگرم)

Parameters of clinical interest in the history

Prenatal history (Yes/No)*

1. Prenatal ultrasonography (findings Presence of choledochal cyst, cholelithiasis, bowel anomalies or concern for syndromes)
2. Cholestasis of pregnancy
3. Acute fatty liver of pregnancy
4. Maternal infections
5. TORCH infections

Positive Family history (Yes/No)*

In (father/mother/sibling1/2/3.../grand mother/grandfather/ other)

1. Consanguinity
2. Neonatal cholestasis in the parents or siblings
3. History of repeated fetal loss or early demise
4. Gestational alloimmune liver disease
5. Spherocytosis and other hemolytic diseases
6. Autoimmune disorders
7. Viral hepatitis
8. Wilson disease
9. Hepatic mass
10. Hepatic cancers
11. Leukemia/lymphoma
12. Infiltrative disorders

Infant history*

1. Gestational age (>38wk/34-38/<34 wk)
2. SGA (small for Gestational Age)
3. congenital infections
4. Alloimmune hemolysis;
5. glucose-6-P-dehydrogenase (G6PD) deficiency;
6. hydrops fetalis
7. Neonatal infection (Urinary tract infection, sepsis related cholestasis, CMV, HIV, syphilis)
8. Newborn screen (Panhypopituitarism galactosemia, fatty acid oxidation defects, cystic fibrosis)
9. Source of nutrition: breast milk, formula, PN
10. Abnormal Growth
11. Abnormal Vision
12. Abnormal Hearing
13. Vomiting (projectile/non-projectile/ bloody/ bilious/post-prandial)
14. Stooling (Delayed stooling/ diarrhea)
15. Acholic stools
16. malodor urine/ Dark urine/
17. malodor sweat
18. Excessive bleeding

19. Disposition: (irritability, lethargy)
20. Abdominal surgery

Physical findings *

1. Assessment of general health (Ill appearance) **Yes/No**
2. General appearance (Dysmorphic features) **Yes/No**
3. Vision/slit lamp examination (cataract/septo-optic dysplasia/ posterior embryotoxon/ glaucoma/ cherry red spot)
4. Hearing
5. Cardiac examination: (murmur, signs of heart failure Congenital heart disease)
6. Abdominal examination (Presence of ascites; abdominal wall veins, hepatomegaly, splenomegaly, abdominal masses, umbilical hernia)
7. Stool examination (Achollic or hypopigmented stools)
8. Neurologic Examination
9. overall vigor and tone

@ * Targeted investigations

<i>Lab data</i>	<i>Date.....</i>	<i>Date.....</i>	<i>Date.....</i>
CBC			
WBC			
Neutrophil			
Lymphocyte			
Eosinophil/Monocyte			
RBC			
Hb			
Hct.			
Retic. count			
Platelet			
PBS			
Blood smear			
Blood culture			
PT			
Fibrinogen			
Coagulation factors(5,7,8)			
INR			
PTT			
AST			
ALT			
TB			
DB			
Total protein			
Albumin			
Globulin			
AlkP			
GGT			
Glucose			
Triglyceride			
Total Cholesterol			
LDL			
HDL			
VLDL			
Uric acid			
CPK			
Aldolase			
LDH			
Urine analysis	GFR:		
WBC			
RBC			
PH			
Reducing Substance			
Urobilinogen			
Bilirubin			
Urine culture			
Random Urine:			
Urine toxin screen:			

<i>Lab. data</i>	<i>Date.....</i>	<i>Date.....</i>	<i>Date.....</i>
BUN			
Urea			
Creatinine			
Na			
K			
Ca			
Phosphorus			
Mg			
Cl			
Blood Gas			
pH			
PCO2			
HCO3			
O2 Saturation			
PO2			
Base excess			
Ammonia			
Lactate			
Pyrovate			
Abnormal items in Serum amino acid chromatography HPLC (abnormal amino acids)			
1.			
2.			
3.			
Abnormal items in Urine amino-acid chromatography:			
1.			
2.			
3.			
Abnormal items in Urine Sugar chromatography:			
1.			
2.			
Abnormal items in Acyl Carnitine Profile:			
1.			
2.			
3.			
Abnormal items in Urine Organic Acid:			
1.			
2.			
3.			
RBC galactose-1-phosphate uridyltransferase (GALT)			
urine for succinylacetone			
Serum for succinylacetone			

<i>Lab. data</i>	<i>Date.....</i>	<i>Date.....</i>	<i>Date.....</i>
Alfa-Fetoprotein			
Alfa 1 AT (level)			
Alfa 1 AT (Genotyping/phenotyping)			
Abnormal items in Serum Bile acids/ bile salt species profiling:			
Serum cortisol (8:00 am)			
Insulin-like GF1			
TSH			
T4/free T4			
T3/free T3			
Abnormal items in Other endocrine panel:			
1.			
2.			
3.			
Sweat Chloride Test			
Cl			
Na			
Stool fat droplet (Sudan)			
Fecal Elastase-1			
serum immunoreactive trypsinogen level			
Stool Trypsin activity			
Infections			
CMV (IgM/IgG/PCR)			
HSV (IgM/IgG/PCR)			
EBV (IgM/IgG/PCR)			
Toxoplasmosis (IgM/IgG/PCR)			
HIV (IgM/IgG/PCR)			
HAV (IgM/IgG)			
HBs Ag			
HBs Ab			
HBe Ag			
HBe Ab			
HBc Ab			
HBV PCR (viral load)			
HCV Ab			
HCV PCR (viral load)			
HEV PCR			
HDV PCR			
Other virus			
Ascitic Fluid			
Color			
WBC/Neutrophil			
RBC			
LDH			
Sugar			
Gram Stain			

<i>Lab. data</i>	<i>Date.....</i>	<i>Date.....</i>	<i>Date.....</i>
Culture			
Cytology			
ADA			
TG			
Other			
Autoimmune markers			
ANA			
ASMA			
AMA			
Anti-LKM1			
Anti SLA/LP			
Anti LC			
Total IgG			
Total IgE			
Total IgM			
Total IgA			
Anti TTg (IgA)			
Anti TTg (IgG)			
Anti EMA titer			
C3			
C4			
CH50			
NBT			
DHR			
NK cell function			
Perforin			
Granzyme B			
sIL2			
Triglyceride			
Other Immunologic work up			
Filter Paper for			
Gaucher disease			
NP-C			
Other			
Iron Storage Disorders			
Ferritin			
Transferrin Saturation			
Iron			
TIBC			
Copper Storage disorders			
Ceruloplasmin			
24- hour urine Copper			
24- hour urine Copper (challenge test)			
Serum copper			
Hepatic copper concentration			
25-OH Vit D			
Serum acetaminophen			

level			

Ophthalmologist Examination *		
	Date.....	Date.....
Cataract		
Septo-optic dysplasia		
Glaucoma		
Cherry-Red spots		
Posterior embryotoxon		
Scleritis/episcleritis		
Congenital toxoplasmosis		
Congenital infections		
K-F ring		
Other		

Imaging@ *		
	Date.....	Date.....
CXR		
Spine X-Ray		
Abdominal Ultrasonography		
Doppler Sonography of hepatic and mesenteric vasculature		
	Date.....	Date.....
Brain Sonography		

Echocardiography		
Barium study (Meal/Small Bowel follow through)		
HIDA scan		
Abdominal CT scan		
MRCP		
Intra operative cholangiography		

Liver Biopsy*

	Date.....	Date.....
Ductular proliferation		
Bile plug		
Portal fibrosis		
Perilobular fibrosis		
Inter portal fibrosis		
Edema		
Lobular disarray		
Inflammatory cells in portal areas		
Intact bile ductules		
Giant cell hepatitis		
Feathery degeneration		
Interface hepatitis		
emperipolesis;		
hepatic rosette		
Bridging fibrosis		
Micro vesicular steatosis		
Macro vesicular steatosis		
Glycogenated nuclei		
Mallory body		
Cirrhosis		
Other @		

Other work up @
ECG
Extremity Radiography
Brain MRI
EEG

Complications @ *		
	Date.....	Date.....
Need PICU admission?	Yes	No
Acute kidney Injury/Renal Failure?	Yes	No
Cardiovascular dysfunction?	Yes	No
Need norepinephrine?	Yes	No
Relative Adrenal Insufficiency	Yes	No
Acute Respiratory Failure?	Yes	No
ARDS?		
Need Mechanical ventilation?	Yes	No
Mode:		
PEEP:		
TV:		
FiO2:	Yes	No
FLUID/Electrolyte/Acid-base abnormality?	Yes	No

Type of abnormality:		
NPO? Type of Nutrition?	Yes	No
Infections? SIRS? Type of infection?	Yes Yes	No No
Coagulopathy? Type of Coagulopathy:	Yes	No
Hepatic Encephalopathy? Grade:	Yes	No
Intracranial hypertension? Cerebral Edema? Received hypertonic saline? Received mannitol?	Yes Yes Yes Yes	No No No No
Seizure? Treatment?	Yes	No
Need any sedation/analgesic? Type of sedation/analgesic	Yes	No

PELD score:
MELD score:
Child score:

Transfer for liver Transplantation?	Yes	No
Need Transplantation?	Yes	No
Type of liver Tx?		

Final outcome	
1 month Tx free survival	Alive/expired
6 month Tx free survival	Alive/expired
12 month Tx free survival	Alive/expired
1 month post Tx Survival	Alive/expired
6 month post Tx Survival	Alive/expired
12 month post Tx Survival	Alive/expired