Dr. D. Y. PATIL VIDYAPEETH, PUNE
(DEEMED UNIVERSITY)

CLINICAL MEET

DEPARTMENT OF PAEDIATRICS AND PAEDIATRIC SURGERY

Dr. D. Y. PATIL VIDYAPEETH, PUNE
(DEEMED UNIVERSITY)

Under the guidance

Dr.Shailaja Mane
PROFESSOR &HEAD
OF Department of Pediatrics ,
Dr. D.Y .Patil Hospital, Medical College & Research Centre,
Dr. D.Y .Patil VIDYAPEETH ,Pimpri Pune.

CASE – 3

BY DR.NEHA TYAGI. RESIDENT,DEPARTMENT OF PEDIATRICS

UNDER THE GUIDANCE OF

DR.SAMPADA TAMBOLKAR(PROFESSOR), DEPARTMENT OF PAEDIATRICS

DR.MANOJ PATIL (PAEDIATRIC INTENSIVIST), DEPARTMENT OF PAEDIATRICS

PATIENT DETAILS

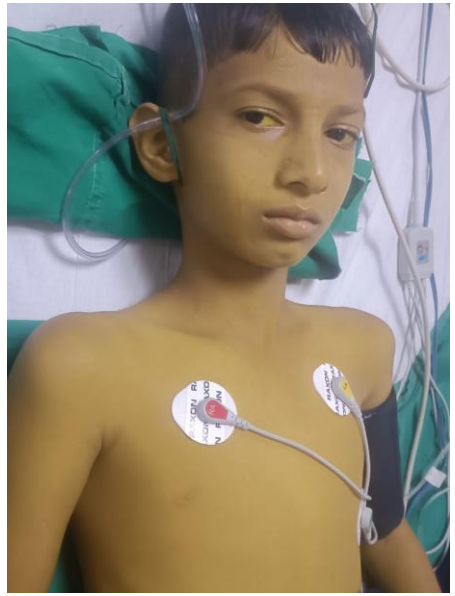
A 12 year old male child,
residing in Pimpri, Pune born in 2nd birth order in a non consanguineous marriage, informant being mother, reliable.

PRESENTING COMPLAINTS

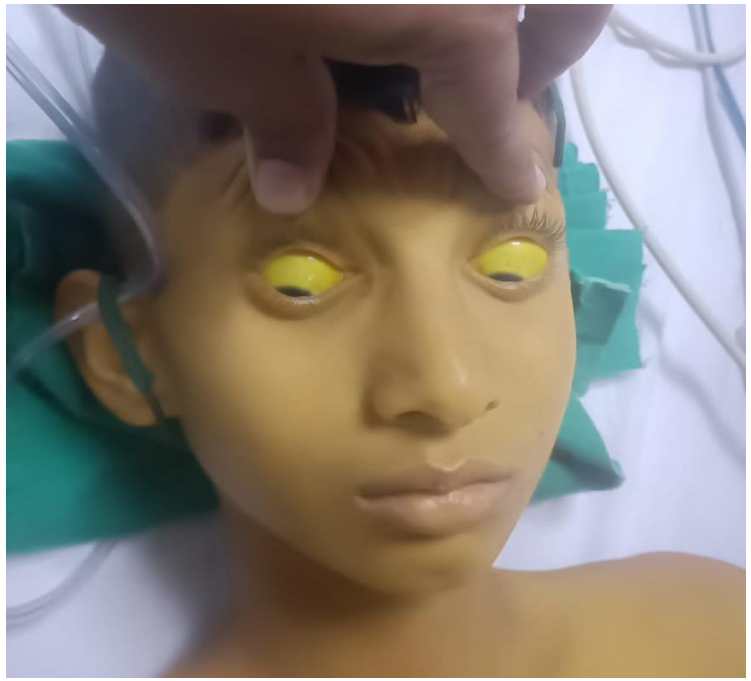
- The patient presented with complaints of vomiting and pain abdomen since 4-5 days.
- Complaint of yellowish discoloration of skin and sclera since 4 days.
- History of passing high colored urine since 4 days
- History of increased day time sleepiness and disturbance of sleep cycle since 3 days.
- No history of preceding fever spikes

History of Presenting Illness

- Vomiting- since 4-5days, nonprojectile, nonbilious, not blood tinged.
- Pain Abdomen- since 4-5 days, insidious in onset, intermittent localised to epigastrium, non radiating, not associated with altered bowel/bladder habits.
- Yellow discolouration- since 4 days, started with involvement of sclera followed by discolouration of entire body including palms and soles.
- High Coloured Urine- since 4 days, not associated with history of any ongoing medications, beetroot consumption. Not associated with passing clay coloured stools.

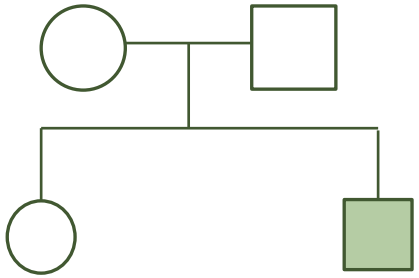


- History of eating outside food present.
- History of intake of herbal medications
- No History of hematemesis, Melena or haematochezia.
- No history of long term medications.
- No history of any blood transfusions.



- **Birth history**

  Full term /LSCS ivo CPD /2.5kg/no resuscitation required/ no NICU admission



- **Development history**

  All milestones attained according to age

- **Immunisation history**

  Immunized up to 18 months of age

EXAMINATION

Vitals :

- Temperature 98.6F
- Heart Rate 63/min
- Spo2 92%RA (taken on O2 by mask – Spo2 100%)
- Respiratory Rate 20/min
- PP well felt
- BP 95/47mmHg

GENERAL:

- Sclera- Icterus +
- Lower Palpable Conjunctiva – Pallor +
- Palms and Soles – Icterus +
- No Rash.

SYSTEMIC:

- P/A – Soft, Tender Hepatomegaly- 4cm below Right Costal Margin, No Splenomegaly
- CVS - S1 S2+, no murmurs
- RS - B/L air entry + , no added sounds
- CNS – conscious, alert, no focal neurological deficits

ON ADMISSION INVESTIGATIONS

Hb (g/dl)	7.40
TLC	15900
DLC (N/L)	75/19
PCV	22.1%
Urea	29
Creatinine	0.31
Sodium	131
Potassium	4.93
Chloride	94
Prothrombin Time (secs)	14.9
Activated Partial Thromboplastin (secs)	28.30
International Normalised Ratio	1.6

Total bilirubin (mg/dl)	66.1
Conjugated Bilirubin (mg/dl)	43.4
Unconjugated bilirubin (mg/dl)	22.7
AST (U/L)	1314
ALT (U/L)	2037
ALP (U/L)	350

HAV IgG	POSITIVE
IgM	POSITIVE

ACUTE LIVER FAILURE

3 criteria for ALF fulfilled:

a) Liver dysfunction within 8 weeks of onset of symptoms

b) Coagulopathy with- INR > 1.5 with Hepatic Encephalopathy
or
with INR> 2.0 without Hepatic Encephalopathy

c) No evidence of chronic liver disease neither at presentation nor in the past.

CLINICAL STAGING OF HEPATIC ENCEPHALOPATHY

Clinical grade	Clinical signs	Flapping tremor
Grade 1 (prodrome)	Alert, euphoric, occasionally depression. Poor concentration, slow mentation and affect, reversed sleep rhythm.	Infrequent at this stage
Grade 2 (impending coma)	Drowsiness, lethargic, inappropriate behavior, disorientation.	Easily elicited
Grade 3 (early coma)	Stuporose but easily rousable, marked confusion, incoherent speech	Usually present
Grade 4 (deep coma)	Coma, unresponsive but may respond to painful stimulus	Usually absent

7 December, 2014

The case was reviewed by the paediatric gastroenterologist-

- Suspecting Wilsonian crisis.
- Advised to send S. Ceruloplasmin levels and ophthalmology review for KF Rings.
- Avoid blood transfusions.

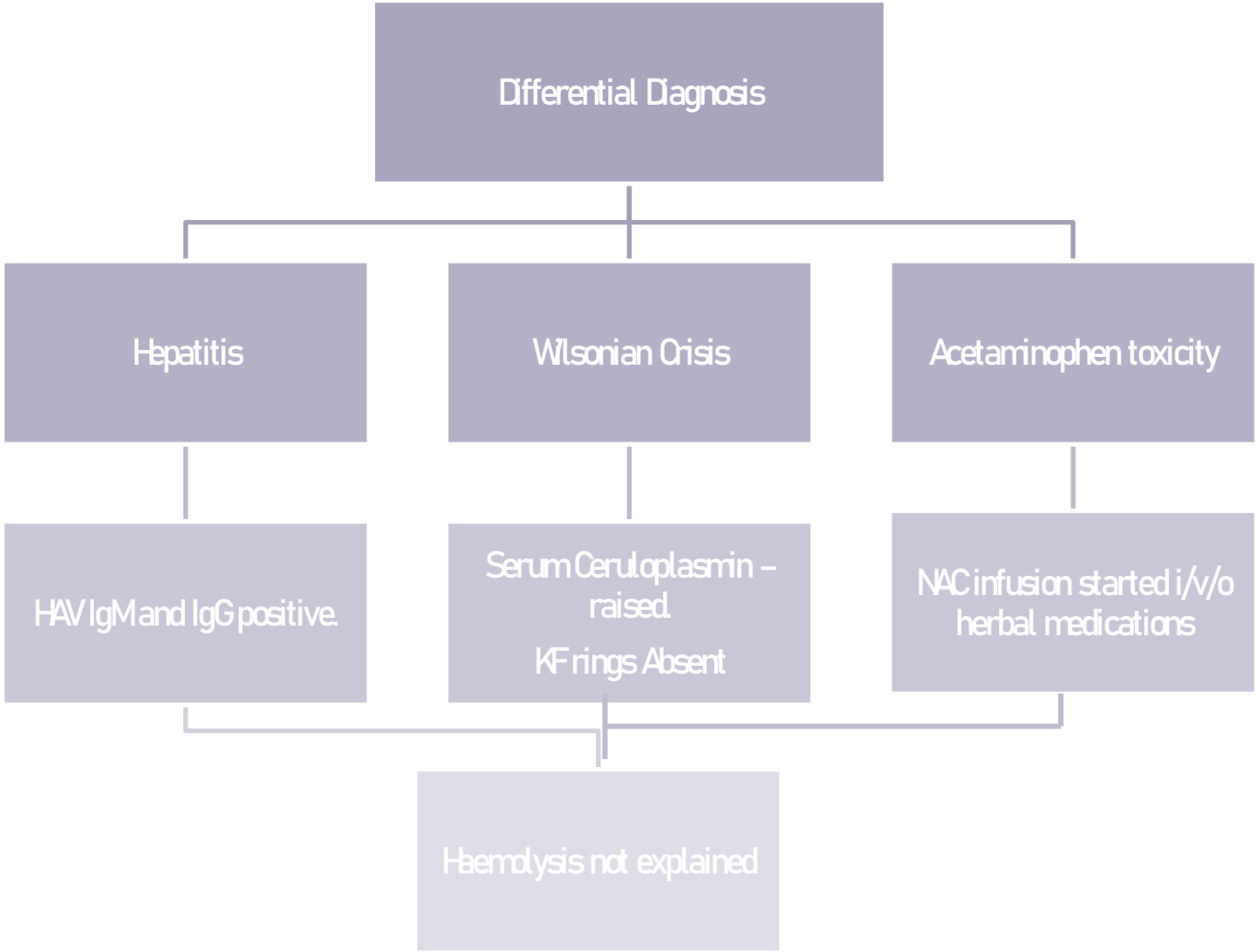
Labs sent:

S. Ceruloplasmin – Raised
KF Rings- Absent

On further Investigations

	6.2.23	10.2.23
Hb (g/dl)	7.40	6.8
TLC	15900	5600
DLC (N/L)	75/19	54/32
PCV	22.1%	20.8%
Retic count	8%	
Corrected Retic Count	3.9%	
LDH	863	712
Indirect Combs Test	POSITIVE	
Direct Combs Test	NEGATIVE	

There were signs of Haemolysis, on further evaluation, ICT was positive.


Differential Diagnosis
Hepatitis
Wilsonian Crisis
Acetaminophen toxicity
HAV IgM and IgG positive.
Serum Ceruloplasmin – raised.
KF rings Absent
NAC infusion started i/v/o herbal medications
Haemolysis not explained

Haemolysis Workup

- Haptoglobin < 7.25 (normal – 30–200) – Sign of Intravascular haemolysis.
- With suspicion of male child, haemolysis exacerbated due to infective aetiology– G6PD suspected
- G6PD decolourisation method >120 mins (normal – 30–60 min)
- G6PD qualitative kinetic method – 4.0 (normal 5–13.5)

DIAGNOSIS

Acute Liver Failure Secondary to Hepatitis A Infection Exacerbating G6PD Deficiency.

TREATMENT

- Started treatment as for Acute Hepatic Failure
- Started on N-Acetyl Cysteine Infusion for first 24hrs.
- Started liver supportive measures- Vitamin ADEK supplements, Oral Ursodeoxycholic Acid.

Response following Treatment

LFTs and Coagulation markers seen in falling trend after treatment of infection

SERIAL LFTs

	6.2.23	7.2.23	8.2.23	9.2.23	10.2.23	12.2.23	14.2.23	16.2.23
Total bilirubin (mg/dl)	66.1	59.6	47.9	30.2	17.19	10.8	9.7	7.9
Conjugated Bilirubin (mg/dl)	43.4	38.5	32	20.36	12.04	8.11	7.35	5.95
Unconjugated bilirubin (mg/dl)	22.7	21.1	15.9	9.82	5.15			
AST (U/L)	1314	740	367	142	84	69	72	98
ALT (U/L)	2037	1530	1252	817	559	333	234	152
ALP (U/L)	350	315	315	262	219	192	212	183

SERIAL PT,aPTT,INR

	6.2.23	7.2.23	8.2.23	9.2.23	10.2.23
Prothrombin Time (secs)	14.9	11.4	17.7	19.0	15.7
Activated Partial Thromboplastin (secs)	28.3	31.3	33.6	35.5	33.6
International Normalised Ratio	1.6	1.69	1.54	1.65	1.37

On Discharge

Patient was Given a list of medications and food items to be avoided and Risks associated with their consumption explained.

High Risk	Medium/Low Risk
Aspirin	Acetaminophen
Chloramphenicol	Acetazolamide
Chloroquine	Ascorbic Acid
Dapsone	Benzhexol
Doxorubicin	Colchicine
Nitrofurantoin	Isoniazid
Primaquine	Phenytoin
Quinolones	Procainamide
Sulfamethoxazole	Quinidine
Sulfasalazine	Trimethoprim
FAVA BEANS	Sulphonylurea
	Vitamin K

DISCUSSION

Causes of ALF

CATEGORY	ETIOLOGY	
VIRUS	Hepatitis A virus	
	Hepatitis B virus +/- Delta virus	
	Hepatitis E	
	HSV/ Varicella Zoster/ Cytomegalovirus	
	EBV/ Adenovirus/ HHV-6	
DRUGS	Idiosyncratic reactions	Isoniazid, NSAIDs, Carbazepine
	Dose dependant hepatotoxicity	Acetaminophen, Sulfonamides, tetracycline
	Herbal supplements	
Vascular Diseases	Right Heart Failure	
	Budd-Chiari Syndrome	
Toxins	Amanita Phalloids	
	Bacillus Cerreus	
	Yellow Phosphorous	
	Carbon Tetrachloride	
Metabolic Diseases	Wilson Disease	
	Reye Syndrome	
Autoimmune Diseases	Autoimmune Hepatitis	
Malignant	Lymphoma	

- Neonatal screening could have helped in reaching the child's current diagnosis faster, with fewer diversions on the way.
- A latent or theretofore asymptomatic deficiency of G6PD may clinically manifest under the appropriate conditions, even when evidence of past or current clinical evidence of G6PD deficiency is lacking.

THANK YOU