From: "ROOT" <root@sctimst.ac.in>
To: "ROOT" <root@sctimst.ac.in>
Date: 14/10/2025 03:36 PM
Subject: CPC Clinical Protocol 15.10.2025

The next Wednesday CPC will be held tomorrow, **October 15, 2025** at **08.00 hours** (IST) in **Lecture Theatre 1**, Nehru Hospital, PGIMER, Chandigarh under the Chairmanship of **Prof. Sanjay Jain.**

The session will also be available on the Webex platform. Kindly follow the link below to join.

https://pgitelemed.webex.com/pgitelemed/j.php?MTID=md00288711fdbf94321a0819943e426a2

In case you join in thru WebEx, kindly **ensure that your microphone and camera are switched off** and **PLEASE DO NOT SHARE YOUR SCREEN**.

The Clinical handout of the case to be discussed is attached herewith.
The clinical protocol will be discussed by ***Dr. Bijaylaxmi Behera, Department of Pediatrics.*** Radiology will be presented by ***Dr. Akshay Saxena.*** Autopsy pathology will be presented by ***Dr. Nandita Kakkar.***

Yours sincerely,

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Regional Resource Centre, North
Department of Telemedicine
PGIMER, Chandigarh

CPC CLINICAL PROTOCOL 15-10-2025

Name: B/O S **Age/Sex:** 31 days,Female baby **CR No:202501438361**

Clinician In charge: Prof Saurabh Datta **Clinical Discussant**: Dr Bijaylaxmi Behera

Pathology Discussant: Prof Nandita Kakkar **Radiology Discussant:** Prof Akhshay Saxena

Pediatric Haematology Discussant: Prof Prateek Bhatia

Date of Admission: 07.02.2025 **Date of Demise:** 14.02.2025

Presenting Complaints:

Fever, lethargy, poor feeding X 2 days

Jaundice with high colored urine and pale stool X 2 days

Skin peeling, dry, progressing to entire body X 2 days

History of present Illness –

Baby was born on 08/01/2025, at full term (39+2 weeks), by NVD(hospital), with birth weight of 3.4 kg. Initiated breastfeeding after birth and was discharged on the same day. On day three of life baby developed poor feeding, lethargy, respiratory distress and yellowish discolouration of body, for which the baby was admitted in a private hospital. Was treated as EOS with antibiotics, received phototherapy of NNJ. Baby had seizure episode on day four of life, received Inj Midazolam and Phenobarbitone. Baby was initially started on nasal prongs oxygen and later given CPAP support for respiratory distress. CXR was s/o pneumonia. Baby also had hypoglycemia, received GIR and shock treated with Dopamine and Dobutamine. Baby was discharged on 01/02/25. Baby was on mixed feeds(breastfeeds and formula feeds)at home. Developed poor feeding, lethargy, fever, icterus and skin peeling three days after discharge and went to the nearby hospital, later referred to PGIMER.

Clinical Examination:

Baby was lethargic

Generalized skin peeling ++ , nails -N

Vitals: HR- 136/min, RR- 36/min, BP- 70/ 42mm Hg, Spo2: 96 % on RA

Respiratory system: BLAEE,NVBS

Cardiovascular: S1S2 -N, No murmur

P/A-distended, liver 4 cm below RCM

Neurological Examination: Lethargic , tone decreased

HEMOGRAM

DATE	7/02	8/02	9/02	11/02	12/02
Hemoglobin	13.6	12.8	9.8	10.2	6.9
Platelets	36,000	54,000	15,000	103,000	75,000
TLC	56800	65200	44000	38750	22,000
N%	-	24	25	27	
L%	-	52	40	30	
M%	-	10.3	19	23.9	
E%	-	11.8	13.6	17	
CRP	-	58.05	44	15	

Peripheral smear: The polymorph and eosinophils show variably sized vacuoles consistent with Jordans anomaly.r/o Channarin Dorfman Syndrome

RFTs

DATE	7/02	8/02	9/02	12/02
Na^+	157	160	154	128
K^+	5	7.08	4.97	5.61
Cl^-	122	130	121	105
Urea	111	135	132	100
Creatinine	0.88	1.77	1.81	1.77

LFTs

Date	07/12	08/02	09/02	12/02
Bilirubin (T)	24	25.94	26	26

Bilirubin (C)	13	18	18	17
AST	527	1154	387	272
ALT	372	470	322	131
TP (Total Prot.)	4.0	3.8	3.8	3
Albumin	2.66	2.2	2.4	1.9
Ca^{2+}		7.8	8.7	9.5
PO_4		5.1	4.7	7.3
ALP		123	114	128

COAGULOGRAM

Test	07/02	08/02
PT	39.6	>1
PTI	28	
APTT	69.3	>2
INR	3.54	
Fibrinogen	0.45	

- **Ammonia**: 275,**Lactate**: 9.9,**AFP**: 458
- **GGT**-340, **Ferritin:** 398
- **USG abdomen:** moderate ascites,periportal edema, GB wall edema(s/o infective aetiology), B/L kidney-raised echogenicity
- **Skin Biopsy-** Epiptosis shows apoptotic keratinocytes, parakeratosis with irregular acanthosis. There is no interface activity. Dermis shows mild lymphomononuclear inflammation in the perivascular and periadnexal region

COURSE IN PGIMER:

At admission, the baby presented with conjugated hyperbilirubinemia, markedly elevated transaminases, and coagulopathy suggestive of severe hepatic dysfunction.Supportive management was initiated with ursodeoxycholic acid (UDCA), fat-soluble vitamins (A, D, E, K), and intravenous vitamin K for correction of coagulopathy and pediatric gastroenterology consultation was taken. In view of ongoing fever, elevated CRP, and leukocytosis, the baby was started on intravenous Cefotaxime, which was subsequently escalated to Colistin, Teicoplanin, and Fluconazole. Peripheral smear revealed Jordan's anomaly, raising the possibility of a neutral lipid storage disorder such as Channarin-Dorfman syndrome, and a whole exome sequencing (WES) test was sent. Dermatology consultation was obtained for generalised skin desquamation, and a skin biopsy was sent. Baby developed hypoglycemia managed with a glucose infusion rate of 10 mg/kg/min and coagulopathy was managed with transfusions of fresh frozen plasma. Ultrasonography of the abdomen showed moderate ascites, gallbladder wall and periportal oedema, and increased renal echogenicity. Baby had gradual worsening of ascites and developed respiratory distress on day three of admission, for which initially CPAP and later went into respiratory failure for which baby was ventilated. On day

three of admission baby developed features of shock for which she was started on Inj Dopamine later was escalated to Inj Adrenaline, Noradrenalin and Vasopressin and Inj Hydrocortisone for refractory shock. Baby had acute kidney injury and gradually progressed to anuria for which baby was put on restricted fluid. But despite all measures baby's condition continued to deteriorate, and she succumbed to illness following cardiopulmonary arrest and was declared dead on 14/02/25 at 05:45 pm.

UNIT FINAL DIAGNOSIS	CAUSE OF DEATH
Term/39+2 wk/3.4 Kg / AGA/Neonatal cholestasis/ ALF/Sepsis/ AKI/ MODS/ Icthyosis/Refractory Shock	Refractory shock