**From:** "ROOT" <root@sctimst.ac.in>
**To:** "ROOT" <root@sctimst.ac.in>
**Date:** 03/09/2025 03:35 PM
**Subject:** CPC Clinical Protocol 03.09.2025

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Dear All,

The next Wednesday CPC will be held tomorrow, **September 03, 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. Pankaj C Vaidya, Department of Pediatrics.*** Radiology will be presented by ***Dr. Anmol Bhatia.*** Autopsy pathology will be presented by ***Dr. Animesh Saurabh.***



Yours sincerely,

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Regional Resource Centre, North
Department of Telemedicine
PGIMER, Chandigarh

# STAFF CPC (03/09/2025)

D,5 months/ Mch; **DOB**: 24.08.2024; **R/O**Jind, Haryana
**CR No**.:202501240659; Adm no.: 2025006280
**DOA**: 22.01.2025
**DOD**: 22.02.2025 (Hosp. stay 1 month)

**ClinicianIncharge**: Dr.SadhnaLal
**Clinical Discussant**: Dr Pankaj C Vaidya
**Radiologist:** Dr Anmol Bhatia
**Pathologist**: Dr Animesh Saurabh

**Presenting Complaints:** 1. Yellowish discoloration of skinsince 2 month of age, 2. Abdominal distension since 2 month of age, 3. Breathing difficulty x 15 days, 4. Lethargy, poor feeding x 15 days, 5. Altered sensorium x 2 days.

**HOPI:** The child was apparently well till 2 months of age when parents noticed yellowish discoloration of skin. At 3 months of age, went for normal f/u, local doctor who also noticed yellowish discoloration and mild abdominal distension, told to have some problem in liver and referred to higher centre. Child then had gradually progressing abdominal distention-insidious in onset, generalized. On enquiring h/o passage of pale stools and diaper staining since birth. Parents noticed breathing difficulty since last 15 days in the form of fast breathing and subcostal retractions along with noisy breathing.Child also was lethargic and not feeding well for last 15 days. Also noticed to have altered sensorium for 2 days prior to presenting to PGI.No h/o fever, pruritis, feed intolerance/vomiting/loose stools, seizures/excessive irritability, CAM intake. No h/o melena/any bleeding manifestations.

**Treatment history:** Admitted twice before admission to PGI – jaundice, liver problem, sepsis (had resp. distress), anemia infantile cholestasis/early CLD/portal cavernoma – received symptomatic treatment IVAB, IVF, npO2, 1 PRBC → referred to PGI as decompensated COI (ascites+, coagulopathy+).

**Past history:** Nothing significant

**Birth history:** Antenatal: H/o 1 abortion, h/o pruritis since 6$^{th}$month of gestation.Natal: Child was born at Civil hospital, Jind–36wk late Preterm/NVD/1.5kg/CIAB/no h/o NNJ/no h/o NICU admission/smooth perinatal transition/no h/o delayed passage of meconium. Discharged on DOL 3.Postnatal: Uneventful. **Family history**: 2$^{nd}$ born to a non-consanguineously married couple. No h/o jaundice or similar illness. **Immunization**: only Birth dose taken. **Dietary history:** On exclusive breast feeding. No h/o top feed.

**Anthropometry:** Weight: 5kg (-3.51z), Length: 54cm (-5.61z), OFC: 36cm (-5.41z)

**G/E**: Conscious, GC-sick, Temperature: 98.6$^{0}$F, HR–70-85/min, RR-52/min, BP 137/74 mmHg. CP/PP – ++/++, CRT – <3seconds, SpO2 - 98% on npO2, Icterus +, Moderate-severe pallor/icterus+/ no cyanosis/ no rashes/ no lymphadenopathy, AF open at level – 1.5x2cm memberanous, rachiatic rosary+. RBS 35 → bolus → 132.

**P/A**: Inspection: Grossly distended, yellowish discoloured, umbilicus inverted, grossly dilated tortuous veins+ over abdomen, no prominent veins on back, left hemi-scrotum swelling. Palpation: Soft, non-tender, Liver 1 cm below right costal margins in MCL line, 2 cm in midline, firm-hard in consistency, sharp margins, irregular edges, granular surface. Spleen 3 cm below LCM, firm non reducible swelling Lt inguinal/scrotal region. Percussion: shifting dullness+. Auscultation: BS+

**RS**: B/L air entry present, equal. B/L crepts and conducted sounds present. **CVS**: S1, S2, continuous murmur present at interscapular region. **CNS**: GCS: E4M6V5, AF – 1.5x2cm memberanous.

**Investigations:**

| Date | 24/01 | 25/01 | 28/01 | 29/01 | 03/02 | 06/02 | 10/02 | 13/02 | 17/02 | 20/02 |
|---|---|---|---|---|---|---|---|---|---|---|
| Hb | 12.9 | 12.2 | 12.2 | 12.7 | 8.5 | 8.1 | 6.8 | 6.4 | 6.3 | 6.5 |
| TLC | 23480 | 17940 | 17380 | 18260 | 10670 | 15000 | 5860 | 5860 | 7060 | 7620 |
| DLC | | 50/38 | 55/35 | | 34/49 | | 49/39 | | 49/40 | 46/45/2.2/6.6 |
| Plat | 1.48L | 1.18L | 1.18L | 1.03L | 1.1L | 1.03L | 75k | 90k | 59k | 71k |
| MCV | 83.2 | 81.9 | 77.3 | 78 | 79 | 78.3 | 78.4 | 81 | 87 | 94 |
| MCH | 25.8 | 26 | 25.8 | 25.9 | 26 | 27.4 | 27 | 32.4 | 27.5 | 28.5 |
| MCHC | 31 | 31.8 | 33.4 | 33.1 | 33 | 35 | 35 | 40 | 31.7 | 30.4 |
| PBF | fragmented cells | | | | | | | | | |

| Date | 01/01 | 25/01 | 27/01 | 28/01 | 29/01 | 04/02 | 06/02 | 09/02 | 13/02 | 17/02 | 20/2 | 22/02 |
|---|---|---|---|---|---|---|---|---|---|---|---|---|
| Na/K/Cl | 143/4.60/112 | 139/4.6/108 | 132/4.36/95 | 127/5.22/94 | 123/4.9/90 | 123/3.63/76 | 116/4.56/69 | 111/3.79/70 | 132/5.12/97 | 140/3.56/102 | 140/4.55/108 | 138/4.57/108 |
| Urea/creat | 34/0.20 | 31/0.25 | 37/0.12 | 36/0.18 | 23/0.16 | 26/0.19 | 31/0.1 | 35/0.22 | 20/0.02 | 29/0.26 | 31/0.23 | 31/0.21 |
| Ca/PO4 | 8.2/2.5 | 9.3/3 | | | | 9.3/3.8 | | | | | | |
| Tb/cb | 19/9.3 | 18.7/8.8 | 17/9 | 17.7/7.2 | 20.6/9.6 | 25/10 | 24.3/12.7 | 23.24/15 | 26.56/12.35 | 26.42/16.62 | 29/19 | 28.6/13.3 |
| TP/Alb | 4.5/2 | 4/1.8 | 4.2/1.6 | 4.5/2.6 | 3.8/1.8 | 3.8/2.9 | 4.6/3.68 | 3.4/3.0 | 3.7/3.3 | 4.2/3.8 | 4.5/4.1 | 4.6/4.2 |
| AST/ALT | 414/264 | 301/230 | 260/204 | 282/188 | 231/155 | 421/150 | 350/135 | 325/97 | 246/94 | 329/121 | 246/128 | 230/122 |
| ALP | | 348 | | 380 | 407 | 344 | 295 | 239 | 188 | 233 | 252 | 286 |
| CRP | 7.4 | 6.8 | | | 10 | | | | | | | 9.6 |

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|---|---|---|---|---|---|---|---|---|---|---|---|---|
| Procal | 2.6 | | | | 0.8 | | 1.5 | | 2.5 | | 1.8 | |
| PT/PTI | 25.7/44 | 26.7/42 | | | 30.2/37 | 31.1/36 | 38.3/29 | 44.5/25 | 42.4/26 | 37.5/30 | | >2min |
| INR/aPTT | 2.29/45.9 | 2.38/54 | | | 2.70/78.1 | 2.78/82.6 | 3.42/>2 min | 3.97/>2 min | 3.78/>2 min | 3.35/>2 min | | >2min |
| Fib/d-dimer | | 0.52/4370 | | | 0.89/3096 | 0.83/3237 | | 0.32/2307 | | 0.351/2933 | | |

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| TC/TG/HDL/LDL | 53/77/13.8/72 |
| Uric acid | 3.4 |
| ABG (admission) | pH 7.29, PCO2 50.8, HCO3 23.6 |
| Ammonia (23/01) | 59 |
| Lactate (23/01) | 2.8 |
| AFP (23/01) | 239 |
| DCT | Negative |
| Gamma-glutamyltransferase (GGT) | 30 ↓ (>125 U/L [2.0 µkat/L]) |
| G6PD | Normal |
| Plasma Hb/urine Hb | 98/nil |
| TFT | T3 0.380 ↓, T4 1.91 ↓, TSH 2.28 N |
| LDH | 861→670 |
| Urine non-glucose reducing substance (NGRS) (27/01) | Positive |
| Bile acids (27/01) | 64 ↑ (19.6 +/- 5.2 mumol/l) |
| CK-NAC | 112 |
| GALT (galactose-1 phosphate uridyltransferase) assay | couldn't be done as child had received BT outside a week prior |

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| **Blood culture** (24/01) | Sterile |
| 10/02 | Sterile |
| 22/02 | Sterile |
| Resp. viral panel (24/01) RSV, hMPV, Adeno | Neg |

| **Ascitic fluid** | **25/01** | **27/01** | **05/02** | **09/02** |
|---|---|---|---|---|
| Cells (N/L) | 453 (N69/L39) | 4 | 10 (10/90) | 81 (N38 L62) |
| S/P | 60/0.3 | 105/0.3 | -/0.5 | 96/0.4 |
| Gram stain | Neg | Neg | | |
| Culture | Sterile | Sterile | Sterile | Sterile |
| SAAG | 0.7 | | | |

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| Urine RME | RBC-189, WBC-12 |
| UP:UC | 0.56 |
| Urine Culture (27/01) | Growth of yeast |
| Urine Culture (03/02) | Growth of yeast |
| Urine culture (07/02) | Sterile |
| Urine osmolarity | 209 |
| Serum osmolarity | 246 |
| Spot Urine electrolytes | Na: 6 mmol/L, K: 12mmol/L, Creat: 42.05mg/dL |

**Imaging:**

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|---|---|
| ECG | Normal |
| ECHO (29/01) | Small PDA (3-4mm), L → R shunt, PFO 3mm, Trivial TR, mild PR, normal biventricular function |
| Rotational thromboelastometry (ROTEM) | Increased clotting time |
| USG Abdomen with doppler (12/02) | Gross ascites, Liver heterogenous echotexture, irregular outline, no focal lesion, HV normal colour/flow/direction, Spleno-portal confluence patent, multiple collaterals joining to form main portal vein at porta. Multiple collaterals along the fissure of ligamentumteres. Kidneys raised echotexture, mild HDN right kidney. |
| CXR (24/01, 12/02) | Bilateral infiltrates – viral pneumonia |
| NCCT Head (10/2/25) | Normal (PG 448278) |

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| UGIE (14/02) | E: leash of vessels+, S: severe PHG+, altered blood + in pylorus (?NG trauma) → after wash 50% clot dislodged, 50% adherent clot still + |
| Fundus | No cataract, No TORCH stigmata |

**Course and Management:**

Admitted as decompensated cholestasis of infancy with SBP and pneumonia. Multiple therapeutic ascitic taps (to alleviate the respiratory distress) and repeated episodes of hypoglycemia. Coagulopathy continued to worsen - received FFP transfusion. Also on supplements, rifaximin, lactulose, udiliv, carvedilol. Also daily albumin infusions, diuretics, salbutamol nebulization. Hyponatremic - transient encephalopathy, ?seizure → symptomatic management and oral sodium supplementation. Started fluconazole → worsening LAMB. Preterminal events: D30 RD → CXR b/l diffuse infiltrates → CPAP and Colistin. D31 child found unresponsive, in arrest, AF tense bulging → Laryngoscopy vocal cord edema, but no blood oozing from airway → CPR as per protocol → could not be revived.

Cause of death: Intracranial bleed secondary to coagulopathy and thrombocytopenia in a child with Decompensated CLD.

**Units Final Diagnosis: Cholestasis of infancy (Decompensated - Ascites); Co-morbidities: PDA 3-4mm; Complications: Sepsis (SBP, pneumonia, fungal UTI), Symptomatic hyponatremia, Portal cavernoma, Thrombocytopenia, Coagulopathy, Anemia**