**From:** "ROOT" <root@sctimst.ac.in>
**To:** "ROOT" <root@sctimst.ac.in>
**Date:** 07/01/2025 03:12 PM
**Subject:** CPC Clinical Protocol 08.01.2025

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Dear All,
**Happy New Years 2025**. Welcome to the New Academic Session, 2025.
The first Wednesday CPC of the session will be held tomorrow, **January 8, 2025** at 08.00 hours (IST) in **Lecture Theatre 1**, Nehru Hospital, PGIMER, Chandigarh.

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

https://india-edu.webex.com/india-edu/j.php?MTID=m1cb4b75b29a2cff53f63e5991b9bab7b

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. Shiv Sajan Saini, Department of Pediatrics.*** Radiology will be discussed by ***Dr. Anmol Bhatia.*** Autopsy pathology will be presented by ***Dr. Nandita Kakkar.***



Yours sincerely,

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Regional Resource Centre, North
Department of Telemedicine
PGIMER, Chandigarh

**STAFF CPC (08.01.2025)**

**Name**: D, **Age**: 7 months, **Sex**: Male

**CR no**: 4282559

**PGIMER DOA**: 31 October 2011

**DO Demise**: 2 November 2011.

**Clinician incharge**: Prof. R Marwaha/ Prof Inusha

**Clinical discussant**: Dr. Shiv Sajan Saini

**Radiology discussant**: Dr Anmol Bhatia

**Pathology discussant**: Prof. Nandita Kakkar

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**Presenting complaints**:

- Fever × 2 days
- Cough × 2 days
- Rapid breathing × 1 day
- Altered sensorium × 1 day

**History of present illness**

The child was apparently well two days back, when he developed mild to moderate grade fever—intermittent pattern, not associated with rigors & chills and was responding to antipyretics. Simultaneously, he also developed non-paroxysmal cough. On the next day, the child developed rapid breathing and increased respiratory efforts. The child also developed drowsiness/altered sensorium as he did not respond to the parents' voice. The parents also doubtful abnormal body movements (possible seizures) preceding the onset of altered sensorium.

There was no history of skin rash, discoloration, pyoderma, ear discharge, loose stools, oliguria, jaundice or bleeding from any site.

**Past history**: No major illness or previous hospitalizations.

**Family history**: First child. No history of any sibling or infant death in the family. There was no history of tuberculosis or any other chronic illnesses in the family.

**Birth history**: FTNVD at PHC in the village with a smooth transition in the perinatal period.

**Development history**: Appropriate for age (At 7 months the child had neck holding, was able to sit, hold objects and was able to produce sounds)

**Immunization history**: Immunized for age

**EXAMINATION**

**Vitals**: HR: 118/min, RR: 38/min, CFT 2 sec, BP: 96/60 mmHg, PP/CP- adequately palpable, warm, and pink peripheries, GCS- E4M3V1, AF at level, pupils- NSNR, SpO2 (room air)- 97%

**RBS** (admission)- **low**, increased to 174 mg/dL after dextrose bolus.

**Anthropometry**: Wt- 8 kg (36$^{th}$ centile), length 63 cm (**<1 centile**), OFC 43 cm (21$^{st}$ centile)

**GPE**: Pallor +nt, no cyanosis, clubbing, edema, icterus, or lymphadenopathy

**Head to toe exam**: Rounded chubby face, no apparent facial dysmorphism.

**Chest**: Bilateral air entry was equal, fine crepitations all over the lung fields

**CVS**- S1 and S2 normal, no adventitious sounds

**P/A**- Distended, no skin discoloration, liver- 10 cm below right costal margin, (span 14 cm), firm consistency, smooth surface, rounded margins (right lobe reaching up to right iliac fossa, left lobe enlarged), spleen 3 cm below the costal margin, flanks normal.

**CNS**: lethargic, drowsy, brisk DTR, planters upgoing, normal cranial nerves examination

**INVESTIGATIONS**

| Characteristics | 31.10.11 | 01.11.11 |
|---|---|---|
| Hb | 9.4 | 7.1 |
| TLC | 6000 | 11500 |
| Platelets | - | 314000 |
| $Na^+/K^+$ | 139/4.4 | 139/5.8 |
| BU/Cr | 75/0.8 | -/0.3 |

**liver function tests**: (31.10.11) Samples not received

**Coagulation profile**: (31.10.11) PT 36 sec, APTT 36 sec, PTI- 39%, INR- 2.57

| Characteristics | 31.10.11 (5 pm) | 01.11.11 (3 AM) | 01.11.11 (9 AM) | 01.11.11 (8 PM) |
|---|---|---|---|---|
| pH | 7.45 | 7.45 | 7.317 | 7. 399 |
| PO2 | 177.6 | 43.7 | 100.8 | 148 |
| PCO2 | 20.4 | 17.2 | 35.1 | 21.4 |
| HCO3 | 13.9 | 11.7 | 18.1 | 12.9 |
| BE | -7.4 | -9.2 | -6.6 | -9.4 |
| SaO2 | 99.6 | 80.7 | 97.3 | 99.2 |

**Card test for malaria**- negative

**CSF examination**: colorless, cells in ill, protein-36 mg%, sugar 42 mg%, gram stain -ve, culture sterile

**RADIOLOGY**

- **CXR** (31.10.11): diffuse bilateral infiltrates seen involving whole of lung fields
- **AXR** (31.10.11): bowel loops gas present till end, not dilated no air fluid levels
- **USG abdomen**: liver span 13.1 cm, mildly heterogenous eco-texture, normal IHBR, normal gallbladder, CBD obscured due to gas, normal venous system; Right kidney 7.74 cm, left kidney 7.2 cm, normal echogenicity, Spleen 8.34 cm, no free fluid. Impression- hepato-splenomegaly with mildly enlarged bilateral kidneys.

**Post-mortem liver biopsy (02.11.11) Biopsy no S-16987/2011)**

The microscopic section studied from liver is composed of two linear cores, showing maintained lobular architecture. The hepatocytes showed diffuse macro-vesicular and microvascular steatosis. The portal tracts are unremarkable. Overall features are suggestive of fatty liver. The second section labelled as lung showed liver tissue. Impression- Steatosis

**Course and management**

The child was started on injection ceftriaxone for suspected pneumonia. For low RBS at presentation, dextrose bolus was given. For low GCS and poor respiratory efforts, the child was intubated and put on manual IPPR. After 12 hours, the respiratory support was changed to ET T-piece for maintenance of airway. The child showed improvement in sensorium. Therefore, he was extubated and put on nasal prong CPAP and IV Antibiotics were continued. RBS was within normal limits on maintenance fluids. He was afebrile and hemodynamically stable. Investigations were planned to find out the cause of large hepatosplenomegaly. After 34 hours of hospital stay, the child developed sudden cardiac arrest. CPR was performed as per protocol for 30 minutes (three doses of adrenalin, slow calcium bolus, and dextrose bolus). However, he could not be revived. The RBS after dextrose bolus was 450 mg%. Sudden cardiac arrest was unexplained.

**Final diagnosis**: Pneumonia with sepsis with underlying storage disorder- ?Glycogen storage disorder ?Tyrosinemia ?Niemann Pick disease