From: "ROOT" <root@sctimst.ac.in>
To: "ROOT" <root@sctimst.ac.in>
Date: 17/09/2025 08:57 AM
Subject: CPC Clinical Protocol 17.09.2025

Dear All

The next Wednesday CPC will be held tomorrow, **September 17, 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. Dr. Manjinder S Randhawa, Department of Pediatrics.*** Radiology will be presented by ***Dr. Dr. Madhurima Sharma.*** Autopsy pathology will be presented by ***Dr. Dr. Hemlata Jangir.***

Yours sincerely,

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Regional Resource Centre, North
Department of Telemedicine
PGIMER, Chandigarh

Staff CPC Clinical Protocol
17-09-2025

Name	Master A S			**Clinician In charge**	Prof Anju Gupta
Age/Sex	2 years / Male			**Clinical Discussant**	Dr. Manjinder S Randhawa
Cr. No.	202203771461			**Radiology Discussant**	Dr. Madhurima Sharma
Adm. No.	2024093518			**Pathology Discussant**	Dr. Hemlata Jangir
DOA	21/11/24	**DOD**	31/12/24		
Address				Mansa, Jatana, Punjab	
Chief Complaints:				Fever x 10 days Swelling over left side of neck and chin x 15 days	

HOPI - Child is a k/c/o LAD1 defect
Gradually progressive non purulent reddish-blue swelling over left neck for 15 days followed by another similar swelling below chin; Associated with Fever for 10 days, upto 102F, relieved by oral medications and vomiting for 2 days, non-bilious, non-projectile with lethargy and decreased appetite
No h/o swelling noted anywhere else on the body
No h/o abdominal pain /loose stools
No h/o jaundice
No h/o cough /fast breathing
No h/o altered sensorium/seizures
No h/o burning micturition
No h/o ear/eye discharge
No h/o TB contact

Past Admissions	Course
18/08/22 – 09/08/22	Fever since DOL3, delayed separation of Umbilical cord, Diagnosed as LAD-1 defect
14/10/22 – 25/11/22	Left femur osteomyelitis with left knee septic arthritis, surgical debridement + Abx for 6 weeks
Lost to follow-up for ~18 months	
02/04/24 – 15/04/24	SAM / AGE / Oral thrush – Nutritional rehabilitation, iv Abx and anti-fungals

Birth History – Born at Term, Cried immediately, AGA 3Kg, Smooth perinatal transition
Family History – 1st born to non-consanguineous couple, no family history of PID
Vaccination – Received only birth dose
Development – Delayed, DQ~60%
Socio-Economic – Lower Middle Class

Examination – Underweight (-5z), Stunted (-7z) with a small head (-2.5z)
Pallor+/No icterus, cyanosis, clubbing, edema, lymphadenopathy; BCG scar present and normal / No Rash/dermatosis
Wrist widening+, Rachitic Rosary +, Leukonychia+
Two ulcerated swellings over the chin and left submandibular sites, Max 2x2 cm, no pus
Systemic Examination – RS/CVS/Per Abdomen/CNS – NAD

Hemogram	**19/11/2024**	**26/11/24**	**06/12/24**	**16/12/24**	**24/12/24**	**28/12**	**30/12**
Hb	6.4	6.4	5.4	7.0	7.0	7.7	8.8
TLC	101300	93300	109280	104520	160000	81700	66670
N/L/M/E	78/17/3.5	78/19/2	85/9/5	81/14/4.2	90/7/3	91/7/1	94/5/0.8
Platelets	6.06 lac	5.5 lac	4.23 lac	1.59lac	1.14lac	16000	15000

Biochemistry	**19/11/2024**	**26/11/24**	**06/12/24**	**16/12/24**	**24/12/24**	**28/12**	**31/12**
Na/K/Cl	122/2.6/77	122/4.3/89	128/2.32/91	132/4.39/100	131/2.4/101	148/2.7/108	145/4.36/110
Urea/Creat	11/0.2	12/0.20	14/0.14	21/0.13	46/0.19	121/0.5	134/0.42
AST/ALT	13/10	54/5	17/2	12/7	9/27	82/22	14/15
ALP	263	312	218	213	207	204	--
TB/CB	0.48/0.11	0.12/0.03	0.5/0.11	0.56/0.08	0.75/0.35	2.46/1.61	2.97/2.15
TP/Alb	8.8/2.9	8.2/2.4	8.2/2.4	6.8/2.0	4.8/0.19	5.5/2.1	5.0/2.25
Ca/PO4	9.3/3.4	8.2/3.8	8.3/5	8.1/5.3	6.9/9.6	7.5/4.6	7.1/--
CRP	365	315	371	307	271	293	
Procalcitonin	70	10	0.85	3.3	15.5	110	110
PT/INR/PTI				25/2.28/44/32.9	18.2/1.6/62/33.4	26.1/2.3/43/	19.7/1.76/51

Fib/D-dimers					4.2/2669	3.5/5027	
Ferritin					1650	>19500	16325

Date	**Blood cultures**
11/11/24	Sterile
19/11/24	Sterile
25/11/24	Staphylococcus aureus (MRSA)
02/12/24	Sterile
13/12/24	Sterile
01/01/24 (Collected posthumously)	**Proteus Mirabilis :** S- Amikacin, Cefoperazone-sulbactam, piperacillin tazobactam R- Cefotaxime, Ceftazidime, Cefepime, ciprofloxacin
23/12/24	Bone Marrow culture - sterile

Tuberculosis Workup

GL for AFB (18/12/24): No AFB Seen

Bone marrow CBNAAT (23/12/2024): M.Tb. not detected

Bone marrow MGIT (23/12/2024): No growth

Bone Marrow Biopsy – 21/12/24 – Normocellular marrow with granulocyte hyperplasia; No atypical cells

PET CT Scan – 13/12/24	FDG avid soft tissue lesion in pancreas, mural thickening in the bowel loops, GEJ, and esophagus, Increased FDG uptake in the spleen, marrow; likely lymphomatous etiology
CECT Abdomen 19/12/24	Circumferential thickening of lower esophagus and GE junction, Clumping of the small bowel loop in the right iliac fossa, ill-defined lytic lesions in the vertebrae and bilateral pelvic bones, possibility of lymphoma

Course during Hospital Stay –

Child presented with fever and impetigo like lesions. He was empirically started on Ceftriaxone and Cloxacillin. There was improvement in the skin lesions however child had persistent fever spikes, and all inflammatory markers were high, so antimicrobials changed to Meropenem and Vancomycin and subsequently Colistin, Vancomycin and Liposomal Amphotericin B (because procalcitonin had reduced, CRP remained high, child had prolonged hospital stay, fungal sepsis was also considered). Child extensively evaluated for cause of fever, serial blood cultures were sterile, infective work-up sent from bone marrow was also not suggestive of any etiology. PET -CT was done s/o FDG avid soft tissue lesion in pancreas, mural thickening in the bowel loops, GEJ, and esophagus , increased FDG uptake in the spleen and marrow. Biopsy was not feasible. Pre-terminally, empirical ATT was also added.

Child had abdominal distension with bilious vomiting around D26 of hospital stay, CECT showed intestinal mucosal thickening and clumping with osteolytic lesions in vertebrae and pelvis. TPN was initiated. Exploratory laparotomy was done s/o multiple small intestinal ulcers with inter-bowel fistulae. Resection of affected small bowel with jejunostomy and colostomy was done.

Child had high grade fever spikes, progressive thrombocytopenia, catecholamine refractory shock and had a cardiac arrest. CPR was done as per protocol but he could not be revived.

Unit's Final Diagnosis - K/C/O Leukocyte Adhesion Defect-1

GI sepsis with intestinal perforation (s/p ileostomy and jejunostomy) with catecholamine resistant septic shock with Disseminated Intravascular coagulation (DIC)