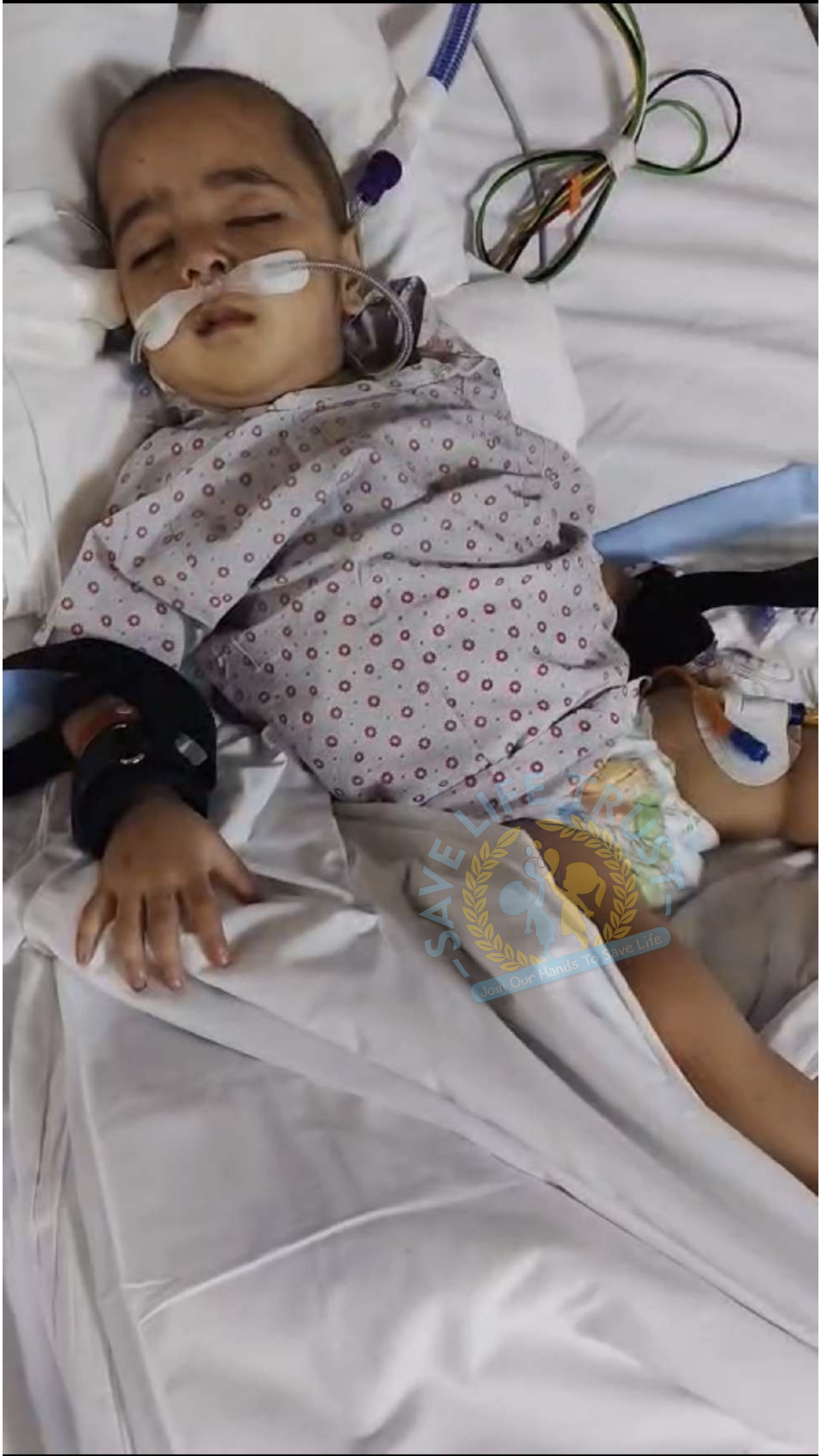
SAVE LIFE TRUST
Join Our Hands To Save Life

DEPARTMENT OF PAEDIATRIC EMERGENCY & CRITICAL CARE
DISCHARGE SUMMARY

Dr. Anil Sachdev
Dr. Suresh Gupta
Dr. Dhiren Gupta
Dr. Neeraj Gupta

Patient Name	Master Dakshu ,	**Registration No.**	3758377
Age	22 Mnths	**Episode No.**	IP01618349
Sex	Male	**Date of Admission**	16-Jun-26
Ward	OLD (OB)-PICU CHARITY WARD	**Bed**	M
Admitting Consultant	Dr. Anil Sachdev		
		Room Vacated on	**Date** **Time**

DIAGNOSIS

Non-Langerhans cell histiocytosis , Multisystem
CNS Risk lesion absent, Bone marrow positive
Secondary HLH
Started on LCH III protocol (22/06/26)
Started on Oral Trametinib (30/06/26)
Dicharged on Week 3 day 1

CLINICAL HISTORY

History:

Dakshu, 22 months old, male boy, r/o Haryana, presented with c/o

- Loose stools since last 6 months
- Fever since last 6 months
- Looking pale since last 6 months
- Lower limb swelling since last 1 week

Patient was apparently alright till January 2026, when he started having loose stools and fever. He was shown to nearby physician and received oral medications.

- CBC (26/01) Hb- 5.1 gm/dl, TLC-11k/cumm, Platelets-46k/cumm MCV 78.2 fL, RDW 28.3 %
- CBC (01/02) Hb- 6.6 gm/dl, TLC-6100/cumm, Platelets-15k/cumm
- CBC (15/02) Hb- 4.9 gm/dl, TLC-7300/cumm, Platelets-31k/cumm, Ferritin 100 ng/dl, LDH 960 mg/dl

Symptoms did not resolved completely hence admitted at private hospital (23/02 to 27/02) where CBC showed Hb- 5.5 gm/dl, TLC-7200/cumm, Platelets-13k/cumm. He received IV antibiotics, IV dexa, IV iron and PRBC and RDP transfusion. Fever resolved for few days and then reappeared. He was again admitted and received transfusion.

In April 2026, patient was admitted with c/o seizure episode lasting for more then 20 minutes. He received IV AED but attendant took LAMA and did not continue followup there. Fever was persistent with max spike 102-103 F, 1-2 spike/day. Child was active in interfebrile period but gradually developed generalized weakness requiring need for sitting up and stopped walking on its own.

Now presented with new onset lower limb swelling and facial puffiness since last 1 week. He was admitted at nearby healthcare centre (06/06 to 09/06). Received PRBC transfusion and referred to higher centre.

Outside labs

Date	05/04	12/05	06/06	08/06	09/06
Hb (g/dl)	7.1	6.35	4	8.1	6.5

Resident Doctor

Consultant

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Paediatrics

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nt Name Master Dakshu , **Episode No.** IP01618349

TLC (/cumm) 9500 11k 11400 8300 5900
Platelet (/cumm)1.19 L 93k 35k 25k 20k
ANC (/cumm) - 4000 5130 4537 2242
MCV (fL) 89 87.9
LFT- total bilitubin-1.2 mg/dl, Direct bilirubin 0.2mg/dl, Albumin-1.8 mg/dl
Retic count 1.8 %
Ferritin 181 ng/dl
Peripheral smear- Anisopoikilocytosis, tear drop cells, nucleated RBCs +
No h/o chronic diarrhea
No h/o swelling elsewhere
No c/o bleeding manifestation
No symptoms s/o focal neurological deficits or poor vision
No family h/o recurrent transfusion
BIRTH HISTORY-
FT/NVD/CIAB/birth weight- 2.6. No h/o NICU stay.
FAMILY HISTORY
Second by birth order born from non-consanguineous union out of 2 siblings. No h/o malignancy in family. H/o missed abortion +
DEVELOPMENTAL HISTORY-
Appropriate for age till 1.5 years of age. No not able to walk or even sit by himself.
IMMUNIZATION HISTORY-
Complete till 9 months of age as per NIS.

PHYSICAL EXAMINATION

Wt –9.8 kg Height: 80cm
On admission, febrile, HR 170/min with good peripheral pulses, RR-54/min, BP- 100/60 mmHg, spo2 – 90 % RA. Pallor present, No icterus.
Seborrhic dermatitis present, Frontal bossing +, occipital protruberance+
Piiting edema in bilateral lower limb
Systemic examination: P/A: soft and distended; non tender, liver- 12 cm palpable below RCM, smooth in consistency with sharp margins, spleen- 8 cm palpable along splenic axis, firm in consistency
Testis Normal
RS- Chest clear
CVS- S1S2 heard, CNS- No focal neurological deficits.

CLINICAL SUMMARY

Dakshu was admitted. CBC showed Hb – 6.8 g/dl , TLC- 7220/cumm and PLT – 29k/cumm, ANC- 261/cumm, AMC – 866/cumm, MCV- 83%, RDW-18%.
After sending Blood culture, he was started on IV antibiotics (Inj Magnex). PRBC transfusion was given in aliquots along with Inj Lasix over period of 3 days.

Work up done:

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Peripheral smear - RBCs – Mild anisocytosis showing normocytic normochromic cells for age. Few macrocytes, tear drop cells and polychromatophils seen. 02nRBC/100 WBC seen.WBCs – No significant abnormality seen.Platelets - Reduced, verified on the smear.

- Ferritin – 239 ng/ml
- PT/INR/ aPTT- 16.5/1.46/42.6
- LFT- total bilitubin-3.25 mg/dl, Direct bilirubin 2.24 mg/dl, SGPT- 15 IU/dL, SGOT-6 IU/dL, Albumin- 2.93 mg/dl
- Trispot- Negative
- Enzyme assay for Beta glucosidase and Neimann pick disease- WNL
- USG whole abdomen (11/06)- Hepatosplenomegaly with minimal ascites.
- X-ray skull s/o lytic lesions.
- Bone Marrow Aspiration and biopsy (11/06/26)-

After taking informed consent, Bone marrow aspiration and biopsy was done. Bone marrow aspirate was sent for smear examination and chromosomal analysis. He tolerated the procedure well.
Bone marrow aspirate showed marked increase in histiocytes with prominent and florid hemophagocytosis. No infiltrate/ granuloma/ hemoparasite seen in the smears examined.
Bone marrow biopsy showed : Clusters of histiocytes are seen, with many showing hemophagocytosis.

Immunohistochemistry:
CD1a: Negative.
Langerin: Negative.
S-100: Positive in small clusters.
CD71: Positive on erythroid precursors.
CD34: No increase in blasts seen.
AFB Stain: Negative.
GMS Stain: Negative.
BRAF V600E : Positive clusters noted.

Impression:
In correlation with immunohistochemistry, a diagnosis of BRAF V600E positive Non LCH type histiocytosis alongwith hemophagocytic histiocytosis is suggested.

Bone marrow chromosomal analysis : 46 XY.

- Whole body FDG PET CT scan done on 12/06/26 –
- Diffuse FDG uptake is seen in marrow of axial and appendicular skeleton.
- Mildly FDG avid (SUVmax –3.9) lytic lesions are seen involving skull bones and right scapula.
- Subtle lucency is seen in the manubrium.
- Hepatosplenomegaly with patchy FDG distribution in liver parenchyma.
- Mild ascites.
- No other significant FDG avid lesion in the body surveyed.

- IR Guided skull lesion biopsy (13/06/26)-

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After taking informed consent, under sedation, tissue biopsy was taken from skull bone lesion by IR team. The procedure was uneventful. Skull lesion biopsy was inconclusive.

- Liver Biopsy showed :

The biopsy consists of three linear cores showing predominantly preserved lobular architecture with mild lobular disarray.
Portal tracts: Up to 15 portal tracts identified, with mild fibrous expansion. A mild portal-based lymphomononuclear infiltrate is present, admixed with scattered eosinophils and occasional plasma cells, with focal periportal spillover.
Bile ducts: Mild lymphocytic duct injury with degenerative epithelial changes affecting native ducts. Mild peripheral ductular reaction with neutrophilic ductulitis is noted. Occasional portal tracts show markedly degenerated duct profiles, confirmed as a keratin-positive epithelial remnants on CK7 IHC. No ductopenia is identified. Periportal hepatocytes show biliary metaplasia, highlighted on CK7.
Hepatocytes: Regenerative nuclear changes with focal rosette formation, focal centrizonal and canalicular cholestasis, and mild focal nuclear ballooning. Occasional hepatocytes contain fine refractile golden-brown cytoplasmic pigment.
Lobular inflammation: Scattered foci of lobular inflammation (~1/10 hpf) with an occasional neutrophilic microabscess.
Sinusoids: Marked sinusoidal dilation and congestion. Numerous sinusoidal histiocytes are present, singly and in small aggregates, with ovoid to reniform nuclei, finely dispersed chromatin, small distinct nucleoli, and abundant pale eosinophilic to foamy cytoplasm. Occasional multinucleated forms are seen. Hemophagocytosis (erythrocytes, neutrophils, and lymphocytes within histiocytic cytoplasm) is identified, including engulfed cells within clear cytoplasmic vacuoles.
Immunohistochemistry (histiocytic population)
CD68: Positive
S100: Positive
BRAF V600E (VE1): Strong positive
Cyclin D1: Positive
OCT2: Positive
CD1a, Langerin: Negative in the sinusoidal histiocytic population (focal non-specific portal stromal background staining noted with CD1a and Langerin)
ALK (D5F3): Negative
Special stains:
Masson trichrome: Mild portal fibrous expansion; mild perisinusoidal (pericellular) fibrosis noted.
Perls' iron stain: Focal fine granular cytoplasmic hemosiderin deposition, Scheuer grade 1.
CK7: Confirms bile duct injury with ductular reaction and periportal hepatocyte biliary metaplasia.

DIAGNOSIS
Liver biopsy:
1. Sinusoidal infiltrate of CD1a-negative, Langerin-negative, BRAF V600E-positive histiocytes with hemophagocytic activity, consistent with a non-Langerhans cell histiocytosis (see comment).
2. Bile duct injury with ductular reaction and focal centrizonal cholestasis.
3. Mild portal fibrosis (Ishak-equivalent stage 1).

-The histiocytic infiltrate is CD1a-, Langerin- and ALK-negative, excluding Langerhans cell and ALK-positive histiocytosis.

-It is S100- and OCT2-positive with occasional emperipolesis and an immunophenotype characteristic of Rosai-Dorfman disease (RDD), however the infiltrate is also strongly BRAF V600E-positive, which is usually wild type in RDD.

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30-06-2026

To,
Save Life Trust
New Delhi

This is to certify that Mr. Dakshu, 1 yr 10 months old boy, Reg. No. 3758377, is a known case of Non-LCH type Histiocytosis multisystem. This is a serious disease and he will require chemotherapy. The approximate duration of treatment is 2 years. The expected cost of treatment for the same is around INR 15 lakhs.

Kindly consider to extend all help for the family.

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