From: "ROOT" <root@sctimst.ac.in>
To: "ROOT" <root@sctimst.ac.in>
Date: 10/09/2024 09:44 AM
Subject: CPC Clinical Protocol 11.09.2024

Dear All,
Season's Greetings.
The next Wednesday CPC of the session will be held on **September 11, 2024** 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://telemedicine.webex.com/telemedicine/j.php?MTID=mb8f5c93d0efe86e2ec7df721b3c21227

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. Naveen Sankhyan, Department of Pediatrics.*** Radiology will be discussed by ***Dr. Anuj Prabhakar.*** Autopsy pathology will be presented by ***Dr. Nivetha.***

Yours sincerely,

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Regional Resource Centre, North
Department of Telemedicine
PGIMER, Chandigarh

Staff CPC: (11-09-2024) Clinical Protocol

Name: L, Age: 19 months, Gender: Female CR no. 202304846767 **Clinician Incharge**: Prof Sadhana Lal

DOB: 24-6-2022, Date of Admission-8.1.24 , DOD- 14.1.24

Clinical discussant: Dr. Naveen Sankhyan, **Pathologist**: Dr Nivetha **Radiologist**: Dr Anuj Prabhakar

Presenting complaints

Progressively worsening

- Involuntary limb movements/posturing- 2 days
- Altered sensorium: 1 day

History:

April 23- Sep 23: (Age 10-15 months) Recurrent vomiting, food, non-bilious, non-projectile, no hematemesis, one-two times/ month, no accompanying complaints
Sep 2023: Exacerbation, vomiting 10-15 times/day, lasting 3-4 days, required hospital admission – treated with i/v fluids and medications. Lethargy present , No fever, no seizures, hypoglycemia. After that: Similar episodes occurred almost every day of the week, food contents, No h/o visible gastric peristalsis/abdominal distension/borborygmi. H/o multiple hospital admissions – almost every 4 days.
Oct-Dec 2023: PGI follow-up, OPD - Lansoprazole and Ondansetron- no benefit
Under Gastro, since Nov 23, detected to have hyperammonemia, treated as UCD? Arg + Sod Benzoate
Jan 5, 2024: Progressive Irritability, abnormal body movements, including clenching of teeth, tightening of limbs, and extension of lower > upper limbs. Progressive loss of awareness, no visual fixation. Next day: The frequency of these episodes increased to every 2 – 3 mins. Insomnia and worsening sensorium -brought to PGI.
H/o constipation- 2 days.
No h/o fever, bleeding manifestations, jaundice/high colored urine/pale stools/pruritis, abdominal distension/ reduced urine output, respiratory distress/fast breathing/ cyanosis.
Past history: Admitted at age 6 months for 5 days. pneumonia/bronchiolitis
Immunization History: Immunized till 1.5 years of age.
Developmental History:
GM –sit - 6 months, stand with support at 9 months- no further gain
FM –scribbling at >1 year / pincer grasp +
Social – Bye-bye at 8 months, recognizes family, few 1-step commands- like" ram ram"
Language – mummy/papa at 7 months, 5-6 words with meaning.
Slow gain after the onset of illness at 10 months.
No regression of previously acquired milestones.
Dietary History: Exclusively breastfed till 6 months. Complementary feeding introduced after 6 months
Family history: Firstborn to a non-consanguineous couple. No h/o similar illness
Natal history: Term / NVD / 3 kg / CIAB / discharged on DOL – 3. H/o oligohydramnios acc. to mother. No h/o jaundice/pruritis in mother during antenatal period

Examination:

GPE: PR -173/min, RR- 56/min, BP-140/109, Wt: 7.5kg (-2.75), L-82cm(0.2), HC-44 cm (-1.71), No LAP/Ict/Cyn/Clb/edm. **P/A** – non-distended, liver 2-3 cm BCM, non-tender, spleen -NP, no free fluid, Bowel sounds present. **CVS:** S1, S2 normal, no murmur, **Resp:** NVBS, No added sounds **CNS**: E4 (M)VTM1, CN's N, Hypertonia, Brisk DTR, Clonus.

Investigations:

Prior to the PGI hospitalization

- 27-9-2023- RBS- 98, CRP-11, Hb-10.9, TLC-20580 (N39 L54), plt 6.48 lakhs
- 1-10-2023- Hb: 12.5, TLC- 26400 (N-59, L36), Plt- 3.16Lakhs, Urea- 23, Cr-0.03

- (15/11) AST / ALT / ALP – 318 / 400 / 186, urea-13, Bil-1.29, FBS-82
  - Hb-11.9, TLC-10.64, DLC- 66/22/4.7,7.5, Plt – 3.94
- 23-11-23 : Vit D-14.0, T3-13.7,T4-2.11,TSH-1.02, GGT-30(8-61), HBA1c-4.1,tTg-<0.1
- 29-11-2023; Ammonia- 358, CK-MB-42(7-25), Lactate – 4.5, AST / ALT: 55/73, bil- 1.19, urea-17, Uric acid 2.8
- 14-12-23: Ammonia- 233, Lactate –3.0
- 16-12-2023: Ammonia: 232, 239, Ck MB- 3 (initiated on ARG and Sod Benzoate)

- TMS(28-12-23)- blood acylcarnitine and Amino acid profile: Elevated Alanine [714(74-613)]
- GCMS- urine(6-1-24): Moderate excretion of 2-ketoglutaric acid, Mild to moderate elevation of Orotic Acid
- USG abdomen- Grossly normal

Hemogram

Date	8/1	9/1	10/1	11/1	12/1	13/1
Hb	11.5	11.2	10.6	11.7	7.7	8.1
TLC DLC	34200 P-71, L-22	28180 P-47,L-39	19600 P-64, L-31	11100 P65,L32	6500 P42,L-56,	14300 P41, L56 M2
Plat(X10^3	9.14 lakhs	2.69 lakhs	68000	15000	33000	27000

Biochemistry

Date	8/1	9/1	10/1	11/1	12/1	13/1
Na	143	145				
K	4.7	5.3	2.44	6.6	2.63	4.4
Cl	110	112	128	146	134	125
Ur	22	32	21	12	11	13
Creat	0.35	0.35	0.5	0.32	0.24	0.29
Bill(T/C)	1.39/0.29	1.86/0.33	4.05/1.36	3.03/1.1	5.11/1.64	2.93/1.73
AST	98	115	146	858	1602	2761
ALT	133	148	162	944	1037	1344
SAP	197				208	
Prot/alb	7/4.8	6.4/4.35	5.7/3.5	4.8/3.1	4.5/2.8	3.3/1.8
Ca/P	10.7/3.9	8.4/4.2	9.6/2.8	9.3/2.2	9.6/3	9.5/3.1
Mg	2.46		1.86		1.72	
Uric acid			2.3		2.9	3.1
AMMONIA	520.3	262	181	137.6	190.4	124
LDH		652	543	626		
CK-NAC/MB	1770	622	229/121		572	
CRP				26.1		20.42
RBS	No hypoglycemia,120-333					
Urine ketones						

	8/1	9/1	11/1	12/1	13/1
PT:	21.1	42.1	51.1	56	>2 MIN
INR	1.78	3.52	4.27	4.67	
APTT	32.2	39.5	40.9	50.6	> 2 MIN
d-dimer +ve,			13705	10464	12777(0-240)
Fibrinogen		5.0	0.61(1.57-3.6)	0.55	

Date	pH	pCO_2	HCO_3	Lac
8-1	7.426	30.9	20	5.1
9-1	7.45	30.7	23.5	4.4
10-1	7.25	55.2		5.6
11-1	7.5, 7.266	33.7, 30.9	26.6, 13.6	5.6, 7.1
12-1	7.189	32.9	12	10.2

- Serum lipids: (9-1-2024): TG-104, CHOL-108, HDL-34, LDL-74
- Urine cs- sterile(9/1), Urine RE/ME- (9/1)- ++ Albumine, RBC 12 PHF
- Blood CS- 15-1-24: Enterococcus Faecium
- Fungal cs (23-1-24): Candida Albicans (blood)
- POCUS(11-1-24): poor cardiac contractility, ONSD 0.56 and 0.52

Course and management:

The child arrived in triage with poor GCS Initially put on O2 support by CPAP, IEM cocktail and 3% NaCl started along with other 1st line anti-RICP measures. I/v/o poor GCS child was intubated in triage and started on IPPR. The child was shifted to the PGE ward at 9 pm on 8/1/24 and was put on volume control ventilation.

Problems:

1. Raised ICP: First line anti-RICP measures followed by 3% NaCl .

2. Hyperammonemia: Child had ammonia of 520.3 on admission hence peritoneal dialysis was started Hyperammonemia reduced to >50% over 24 hours and continued to gradually reduce.

3. Shock : By 18 hours of hospital stay, tachycardia persisted, however child developed shock with feeble pulses, cold extremities, and low diastolic BP. Fluid bolus was given f/b noradrenaline was started and ionotropic support hiked to Adr @0.2 and Norad @0.2. The child further had worsening of shock and Vasopressin was also added @0.0004. I/v/o inotrope refractory shock, hydrocortisone was given @5 mg/kg stat by DOA – 2.

4. Hyperglycemia: Following hydrocortisone injection, the child developed hyperglycemia RBS = 333. Insulin infusion started, hiked to a maximum of 0.5 U/kg/hr.

5. Cytopenias: The child initially had neutrophilic leucocytosis with normal Hb and platelets; however, by DOA—2, the child started having thrombocytopenia—the lowest was 15000, which was supported with 1 PC. The child did not have clinical bleeds.

6. Dyselectrolytemias: Child had persistent hypokalemia for which i/v correction was given along with maintenance K+ added to fluid and PD fluid. Child also had asymptomatic hypernatremia, maximum 167 after which 3% NaCl was stopped.

7. Infection: The child was initially started on Cefotaxim; however, due to a high-grade fever, the antibiotics were upgraded to meropenem, which was continued.

8. Terminal events : The child had raised ICP with ALF with Inotrope refractory shock. Parents requested 'do not resuscitate' and 'do not escalate treatment' in the event of a cardiac arrest. The child had bradycardia was 10 hours on the day of demise with NR pulse and SpO2. Gradually HR decreased and death was declared on 14/1/24

Units Diagnosis: Urea Cycle disorder ?, hyperammonemia, Raised ICP, Multiorgan Dysfunction

Attachments:

File: CPC Clinical Protocol 11 Sept 2024.pdf	Size: 202k	Content Type: application/pdf