**NOTE 6**

**PATIENT 1002**

**DATE: 5/15/20**

**Patient Summary:**  
Patient (triplet A) is a 2 month old 27 4/7 wks GA boy with left CDH and malrotation s/p left-sided CDH repair and Ladd's (3/6). Post-op course c/b abdominal compartment syndrome and AKI/RF requiring ex-lap (3/8) found to have intestinal/colonic perforations s/p resection, ileostomy, and silo placement. Now s/p silo take down and Alloderm placement (3/11) and stoma revision (3/13). Also with evolving BPD + lung hypoplasia now extubated on CPAP after DART course. History of feeding intolerance, now NJ fed. Tolerating RAM CPAP 6 well with occasional spells and overall improving weight gain and now requiring treatment with Avastin injection for treatment of retinopathy of prematurity.   
  
**Events in Last 24 hours**  
- Stable on LFNC  
- Pulled out NJ, tried NG feeds , able to tolerate 3/4 volume feeds but became increasingly fussy w/ full volume feeds so continued 1/2 volume feeds and IVF  
  
**Weight**  
Last weight: 1.77kg (05/14/20)  
Weight change: +10g (05/13/20 to 05/14/20)

   
**Plan**  
*CV:* S/p SDS X24h on admission. Echo 2/18 with large PDA with bidirectional flow, PFO with left to right flow, small anterior mVSD with bidirectional flow, normal function. Repeat echo 2/24 showed large PDA with bidirectional flow s/p Tylenol 5d Tylenol course 2/28. Repeat ECHO 2/27, 3/3, 3/8, 3/21, 3/28 with closed PDA, no pHTN. Required pressors (dopa/epi/NE) and SDS during decompensation 3/7-10;3/13-14.  
- PHTN team following; echo 4/25 without pulmonary hypertension.  Per team OK to wean below 25% (though does not tolerate due to spells)  
**- Plan to repeat ECHO once off of CPAP, plan for echo on 5/15**  
- ContinueHCTZ BID  
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Access: None currently. S/p low lying UVC (2/17 -2/18), s/p UAC (2/17-3/10), SL PICC (2/18- 3/17), tunneled DL CVC (3/8 - 4/8).  
  
*Resp:*  Intubated in the delivery room with 2.5 ETT and started on high frequency ventilation (HFJV), s/p surf x1. Significant respiratory course on HFJV followed by CMV 3/24, extubated 3/25. Initially on NIPPV due to lack of effort but then transitioned to CPAP 3/27. s/p DART 3/22-3/31. Off caffeine since 4/18, s/p caffeine bolus 4/27.  
**-Continue LFNC 1L**  
  
*FEN:* H/o hyperglycemia improved off stress dose hydrocortisone. PN discontinued 4/8. TFG 140. Last weight adjusted feeds/meds on 5/10  
- NJ feeds at 150 ml/kg/day Elecare 30kcal, **plan to condense feeds in coming days**  
- NaCl supps started 4/24  
- Famotidine 4/24  
- Fe/VitD supps 4/26  
**- Trial paci dips  
- Feeding team evaluation week of 5/15 to assess readiness for trialing small PO intake**  
- monitor urine output and electrolytes closely  
  
*GI/Bili:* L sided CDH with liver up. S/p bedside open L CDH repair 3/6 via left subcostal incision, found to be large CDH (type C defect) containing left lobe of liver, stomach, spleen, small bowel and colon. S/p bedside reopening on 3/8 with findings of perforation and s/p ileocecectomy, end ileostomy, and silo placement. S/p OR 3/13 with reopening of recent laparotomy, removal of silo and Gore-tex mesh, Alloderm mesh closure of fascia, and partial skin closure. Stoma dilated and opened at bedside 3/20 by surgery. Stoma dilated bedside by surgery most recently 5/6, output improving.   
**- Monitor stoma output**  
- qMonday Chem while on diuretics  
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*Renal:* h/o AKI, Renal US 3/7 with "echogenic renal cortices with abnormal main renal arterial waveforms and elevated resistive indices; constellation of findings likely reflect medical renal disease/acute kidney injury"  
- Continue to monitor Uop  
  
*Heme:*History of thrombocytopenia of unclear origin, now resolved 3/29. S/p multiple pRBCs, most recently 4/15. Hematocrit improving on 5/8 at 36.3% with retics increasing at 10.5%  
- qMonday CBC  
  
*ID:*No risk factors for sepsis (delivered by C-section with intact membranes for twin reasons). Twin C (singleton, did not share a placenta or sac) developed coryneform bacteremia - being treated with amp/gent/vanc. Blood culture final NGTD. Urine CMV negative 3/3. S/p abx 3/7-3/22, S/p 48 r/o Cefepime/Vanc, cx NGTD, s/p 5day course for RUE cellulitis with cefazolin,  RVP negative 4/17. CBC, CRP 4/26 reassuring, spells not seemingly r/t infectious source  
-Continue to monitor clinically  
  
*Neuro:*HUS 2/18 with bilateral subdural hemorrhage, difficult to visualize anterior superior sagittal sinus. Repeat 2/20 with subdurals decreased in size. HUS stable 2/23. s/p Fentanyl --> transitioned to morphine gtt 2/28, OFF morphine 4/7. HUS 3/9, 3/22 stable w/o IVH, Term HUS 5/3 normal.  
  
*Immuno*: NBS x2 with low TRECs. Immunology added on lymphocyte subsets 3/7, repeat 3/27 with T cell memory panel: Evaluation not consistent with SCID or other T cell defect  
- Please hold all live vaccines (rotavirus, MMR, VZV) until patient can demonstrate adequate specific antibody response to inactivated vaccines (to be checked around ~6 months of age): Please contact immunology prior to discharge  
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*Genetics*  
- Consulted, rapid MCA and karyotype sent 3/14  
- Microarray with finding of unknown significant, copy gain of 14q11.2  
- Whole exome sent - findings of multi gene duplication, follow-up with genetics  
- Per genetics note 5/5 2020, likely paternal inheritance of this 14q11.2 duplication which is classified by GeneDx as likely pathogenic. Case reports suggest wide range of clinical manifestations; genetics team planning to review w/ patient’s parents. This result is not diagnostic for patient’s presentation of CDH.  
  
*Ophthalmology:*  
ROP (4/12): zone 2 immature, repeat in 2 week (4/26). 4/19: R stage 1, zone 2, L stage 1, zone 1-2. 5/2 Stage 2 Zone. **S/p Avastin injection 5/10.  
-** **Follow-up exam 5/15**  
  
*Social:* Continue to update and support family. Last family meeting 3/22.  
  
*Disposition:* 11 Hale for respiratory support and prematurity-related care  
  
*RHCM:*  
- Newborn screening:  
   - 2/17: multiple unsatisfactory, OOR SCID  
   - 2/19 (DOL2): OOR aminoacidopathies (methionine), OOR T4 (low), OOR SCID  
   - 3/2 (prior to transfusion): OOR aminoacidopathies (methionine and valine), AFT  
   - 3/19 (DOL30) OOR aminoacidopathies (leucine and hydroxyproline), Hemoglobin AT, OOR SCID  
   - 3/28: AFT (post-tx?)  
  - 2mo NBS sent 5/1, SCID (however, has already undergone negative evaluation)   
- Hepatitis B Vaccination: has never received  
**-** 2M vaccines: 4/19  
- CCHD: n/a, got echo  
- Hearing screen: Prior to discharge  
- Car seat testing: Will require prior to discharge  
- Circumcision: Will inquire  
  
PCP: To be determined

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