**NOTE 8**

**PATIENT 1001**

**DATE: 6/6/22**

**Patient Summary**

Patient is a 2 month old term infant with Coffin Siris Syndrome, Dandy walker malformation, s/p coarct repair, s/p DLB with mild malacia and multifactorial respiratory failure, with abdominal perforation on 5/25 in setting of malrotation and NJ tube placement, now s/p ex lap, repair, Ladd's procedure, NPO, s/p antibiotics, intubated and weaning vent support and sedation. Now with new yeast UTI.

**Events in Last 24 hours**  
Worked toward extubation yesterday, however postponed in setting of fever 38.6.  
UCx, BCx, SCx collected at time of fever - UA and KOH positive for yeast - started on vancomycin, cefepime, and fluconazole  
  
  
**Plan**  
*CV:* Hemodynamically stable, s/p aorta coarctation repair (5/11). Last ECHO (5/25) with no evidence of re-coarc and good function. Cardiology following closely.  
  
Access:  R IJ CVL (5/25), central in location, needed for medications and nutrition. s/p 2.6 Fr DL PICC (removed 5/23).  
  
*Resp:* Intubated on relatively low vent settings, previously on CPAP prior to perforation, known compromised resp status likely multifactorial, with mild malacia on DLB. S/p Lasix x 3 days and then resume Diuril once tolerating enteral feeds. Continue Atrovent nebs.  
- ERT done successfully  
- consider extubation again after treating infection and monitoring cultures 4-48 hours  
  
*FEN/GI:* Bowel perforation + malrotation s/p repair and Ladd's 5/25 UGI (6/2) without leak, enteral feeds started. TF 130 mL/kg/day PN/IL, now on 40 ml/kg/d enteral, advancing 10ml/kg/d BID, Sim via NJ. Previously tolerating full NJ feeds. Continue to monitor electrolytes, i/o's, and growth.    
also with Meckel's and still has appendix.  Attempt to d/c Vygone while preparing for extubation if needed replace OG.  
-----------------------------------------------------------------------------------------------------------------------------------------  
*Heme:* H/o anemia s/p PRBCs, most recent Hct 33.2%. Continue ASA x 3 months post-op from coarc repair, will restart once tolerating enteral feeds.  
  
*ID:*  Bowel perf and Enterococcus UTI s/p amp/cef/flagyl 10 days total treatment (ended 6/4).  
  
*Neuro:* Has Dandy Walker malformation. HC daily, stable.  
  
Sedation: On clonidine patch, morphine and midazolam gtt. Weaning. SBS 0 to -1 while intubated.  
  
*Genetics:*Congenital disorders of glycosylation and karyotype normal. Microarray XY male. WES - heterozygous for de novo variant in SMARCA4, Coffin Siris syndrome, heterozygous for a VUS in CHD7. Genetics following. Will need follow up with cancer dept regarding screening for tumors given CSS.  
  
*Endo:* DOL 30 abnormal TFTs, labs 4/17 reassuring (TSH 4.5, FT4 1.1). Most recent NBS TSH elevated - **will send TFTs.**  
  
*Other*: PACT following, last meeting 4/5.  
Re-involve following extubation  
   
*Social*: Parents updated regularly by phone and at bedside using Portuguese interpreter. Continue to update and support.  
NBS, f/u  
CCHD had echo  
Hearing screen: determine results, will need repeat  
Car seat test: will need  
PCP: determine and update  
  
*Disposition:* Requires ICU care while on advanced resp sup, recovering from GI perforation

**Relevant Labs**

**Blood Culture Routine, Aerobic:** (Blood) 05/25/2022.***Final Report:***No growth  
**KOH Fungal Stain:** (Urine) 06/06/2022.***Final Report:***Moderate Yeast cells seen  
**Respiratory Culture and Gram Stain:** (Endotracheal Tube Aspirate) 06/06/2022.***Gram Stain Report:***No polys seen No organisms seen

**Weight**  
Last weight: 4.49kg (06/05/22)  
Weight change: +65g (06/04/22 to 06/05/22)  
  
Fluid Balance (6/5/2022 07:00 to 6/6/2022 06:59) In: 601 mL / Out: 377 mL / Balance: +224 mL