**NOTE 6**

**PATIENT 1001**

**Date: 5/22/22**

**Patient Summary**

Patient is a 2+ month old term infant with Coffin Siris Syndrome, Dandy walker malformation, s/p coarct repair, s/p DLB without clear explanation for airway compromise when extubated.  CPAP, weaning, Full NJ feeds.

**Events in Last 24 hours**  
MSO4 gtt discontinued - enteral started  
colace to PRN  
  
**Plan**  
*CV:* Hemodynamically stable, s/p aorta coarctation repair 5/11, monitor, cards following, last ECHO 5/18. Continue ASA x3 months post-op. EKG 5/21 ok.  
Access:  2.6 Fr DL PICC found to be **non-central** 5/17, will use as non central line for sedation only-- will remove today  
  
*Resp:* CPAP, weaning as tolerated.  Known compromised resp status likely multifactorial, only mild malacia on bronch. On diuretics, atrovent nebs.   
  
*FEN:* Continue full NJ feeds, 150ml/kg/d, monitor I/Os, lytes. Trend weight. Spaced colace from Q12hr to Q24hr 5/21, changed 5/22 prn.  
  
*Heme:* H/o anemia s/p PRBCs, monitor counts. Continue ASA x 3 months post-op.  
  
*ID:* no active concerns.  
  
*Neuro:* Has Dandy Walker malformation. HC daily, stable.  
  
Sedation: On enteral ativan, morphine and clonidine.   
  
*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).  
  
*Other*: PACT following, last meeting 4/5.  
   
*Social*: Updated on admission. Portuguese speaking. Continue to update and support. Schedule family meeting week of 5/22.  
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 support

**Weight**  
Last weight: 4.025kg (05/22/22)  
Weight change: +20g (05/21/22 to 05/22/22)