**NOTE 3**

**PATIENT 1001**

**Date: 5/7/22**

**Patient Summary**

Patient is a 1+ month old term infant with Coffin Siris Syndrome, Dandy walker malformation, aortic coarct on PGE, on CPAP s/p DLB without malacia, ongoing spells, and planning for coarct repair. 

**Events in Last 24 hours**  
started precedex; ectopy. weamed precedex  
spell when mask malpositioned  
  
**Weight**  
**Last weight:**3.855kg (05/06/22)  
**Weight change:**+15g (05/06/22 to 05/07/22)  
  
**Plan**  
*CV:* HDS. Napkin ring coarctation of the aorta, **continue PGE 0.05mcg/kg/min**. CT surgery and cardiology following. **Currently undergoing planning for surgical repair.**  
Access: RUE PICC (4/29-) central in RA/SVC junction, needed for PGE infusion; powerflushed on 4/29  
  
*Resp*: H/o stridor, desaturation events, secretions, DLB (4/25) w/o significant malacia, no indication for pexy, VC mobile.  Currently stable in CPAP. **Discussing possible repeat bronch, tracheopexy with cardiac repair.** Continue Atrovent TID.    
   
*FEN*: Tolerating enteral feeds of Sim 20 at TF160cc/kg/day via NJ. Continue vitamin D. Monitor I/O, feeding tolerance, weights.  
   
*GI/Bili*: History of direct hyperbili i/s/o prolonged PN, follow LFTs/GGT qMonday. AUS (3/15) with liver cyst and submucosal nodule in bladder, liver cyst seen again on hepatic US 5/2.  
   
*GU:* Undescended testes high in inguinal canal and bilateral inguinal hernias confirmed on US (3/15), follow clinically, will need surgical evaluation for hernias after cardiac repair. 5/2 US showed R duplex kidney system.  
  
*Heme*: Last HCT 32.3 (4/24), monitor.  
   
*ID*: No active infectious concerns, monitor.  
   
*Neuro*: Dandy walker malformation and agenesis of corpus callosum confirmed on MRI (3/17), neurology following. Daily HC.  On clonidine and ativan and Precedex at 0.2  
   
*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. Last family meeting 4/19.   
   
*Disposition*: In NICU while requiring CPAP and during on-going surgical planning.