**NOTE 5**

**PATIENT 1001**

**Date: 5/12/22**

**Chief Complaint**  
Coarctation  
   
**History of Present Illness:**  
Patient is a 8-week-old infant male with Coffin-Siris syndrome and an atypical coarctation of the aorta presenting for coarctation repair.  
  
Prior to presenting to the CV OR, he was admitted in the NICU since birth. Two attempts of trial off of PGE resulted in severe coarctation despite absence of a patent ductus arteriosus, suggestive of circumferential ductal tissue responsive to PGE. Supported with CPAP, with frequent intermittent desaturations thought secondary to upper airway obstruction; DLB 4/25/22 unremarkable, no significant malacia. Plan per Pulmonology to pursue sleep study following cardiac repair.  
  
**Anatomy:** {S,D,S} Normal intracardiac anatomy, napkin ring-like coarctation of the aorta. LAA with NB. Normal coronary artery origins.  
  
**Problem List:**  
1) Coarctation  
2) Coffin-Siris syndrome  
3) Dandy-Walker malformation, partial agenesis of the corpus callosum.  
4) NG dependent.  
5) History of direct hyperbilirubinemia, improved. Hepatic cyst on ultrasound.  
6) Bilateral cryptorchidism and inguinal hernias  
  
**Primary Cardiologist:** Dara,S  
**CV Surgeon:** Cord, B  
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**Procedure:**  
**OR 5/11/22**  
Anesthesia: Grade 1 view, 3.0 cuffed ETT @ 9.5cm.  
Procedure: Left thoracotomy, PDA ligation and division, coarctectomy and extended end-to-end repair  
Tubes/Drains/Packing: One 15Fr L pleural blake drain  
Findings: There was a ductal ligament/PDA (unclear if it was open) that was divided after double ligation. There was a coarctation of the aorta centered around the take off of the L subclavian. Total clamp time was 19 minutes. Gradient between UE and LE at end of case (off PGE) was 7 mmHg       
Patient Condition: Stable for ICU level of care      
   
**CICU Course:**  
5/11: admitted to the CICU s/p CV OR ~2230  
   
   
**Impression/Plan:**  
Patient is a 8-week-old infant male with Coffin-Siris syndrome, Dandy Walker malformation, and an atypical coarctation of the aorta who went to the OR for repair.  To OR 5/11 Left thoracotomy, PDA ligation and division, coarctectomy and extended end-to-end repair. Had good surgical result, but currently has atelectesis with intermittent desaturations.  Goal to optimize respiratory function prior to extubation back to cpap.    
   
CV:  
- EKG now, monitor rhythm  
- Goal BPs: SBP 70s-90s  
-Nicardipine for BP goals  
-Diuretic: Diuril with goal FB -50 to  100  
- Trend MV and lactate/UOP/CT output  
   
RESP:  
- Ventilate to normal gas exchange  
- On CPAP pre-op  
--- dynamic bronch on 5/11 pre CV OR, see report for full details  
- CXR daily  
   
NEURO:  
- SBS goal: -2  
- Sedation:  precedex infusion  
- Analgesia: ATC tylenol  
- PRNs: MS04, midazolam  
*-* Hx Dandy walker malformation and agenesis of corpus callosum confirmed on MRI (3/17), neurology following. Daily HC.  On clonidine and ativan, precedex pre-op  
--- restart intermittent ativan  
--- will need to restart clonidine when receiving enterals  
   
FEN/GI:  
- NPO, mIVF  
- Famotidine ppx  
- Trend chemistry panel  
- 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:  
- CBC and coags  
- Anticoagulation plan per CV surgery:  intermediate heparin and ASA  
- Monitor CT output   
   
ID:  
**-** vanc/cef (5/10 - ) for septic rule out in setting of hypothermia, monitor blood/urine, change to kefzol post-op ppx once r/o is complete- d/c 5/12  
- Trend indices of infection and WBC  
-UC frm 5/9 with less than 4000 CFUs of GNRs (no tx)  
   
*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.  
   
Access:  
- dl PICC  
- PAL  
- CT x1  
- Foley  
- ETT  
  
SOC:  
- Family lives in MA. Preferred language is Portuguese.  
   
Rounds completed at the bedside with CICU team, Dr. Sam attending