**NOTE 7**

**PATIENT 1001**

**DATE: 5/26/22**

**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 on CPAP, now s/p bowel perforation in setting of malrotation s/p repair, also with meckel's diverticulum. On antibiotics, bowel rest.

**Events in Last 24 hours**  
see event note from 5/25 - overnight with tachycardia, poor perfusion, increased respiratory support needs requiring intubation, sepsis workup, abd distention, then found to have hypoglycemia, KUB with NJ overlying lower quadrant, stat abd ultrasound showed free fluid and concern for perforation with nj tube, no free air, to OR for ex lap and repair:  
  
s/p ex lap and repair of bowel perforation, removal of ~ 150 of formula filled fluid from abdomen, appeared recent and not infected or highly inflamed appearing, patient found to be malrotated with perforation at site of malrotation. s/p Ladd's procedure, repair of serosal injury 2 layer repair, snaked NJ tube past anastomosis site, also found to have meckel's diverticulum 20 cm from IC valve, appendix remains, RIJ 4 F DL CVL at 6 cm. Plan CXR/gas, 7 days NPO on PN/IL and 7 days of antibiotics, If NJ comes out do not replace, plan UGI prior to feeding, keep NG to suction. Continue sedation and pain medications  
  
  
**Plan**  
*CV:* Hemodynamically stable, s/p aorta coarctation repair 5/11, monitor, cards following, last ECHO 5/25 with no evidence of re-coarc and good function.  ASA held, plan to restart 5/26 and continue  x3 months post-op. EKG 5/21 ok.  
Access:  s/p 2.6 Fr DL PICC - out 5/23. New R IJ 4 F DL CVL placed 5/25 - needed for sedation and nutrition.  
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*Resp:* Intubated, on moderate vent settings, wean as able. Hx of CPAP prior to decompensation, Known compromised resp status likely multifactorial, only mild malacia on bronch. On diuretics, atrovent nebs.   
  
*FEN:*s/p bowel perf in setting of malrotation, now s/p repair on 5/25, plan 7 days NPO on PN/IL, monitor lytes and replace as needed, plan UGI prior to any feeding to reassess. continue replogle to suction. Has meckel's diverticulum.  
Previously on NJ 150ml/kg/d.   
  
*Heme:* H/o anemia s/p PRBCs, monitor counts. Continue ASA x 3 months post-op (held 5/25).  
  
*ID:* sepsis w/u 5/25 on amp/cef/flagyl, peritonitis in setting of bowel perf, plan to continue for 7 days pending cultures and clinical course.  
  
*Neuro:* Has Dandy Walker malformation. HC daily, stable.  
  
Sedation: On IV precedex, IV clonidine, IV morphine and ativan gtt. adjust as needed. SBS 0 to -1 when 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).  
  
*Other*: PACT following, last meeting 4/5.  
   
*Social*: Updated on admission. Portuguese speaking. Continue to update and support. Family updated extensively on 5/25 with interpretor. will continue to update and support and plan for in person meeting on sunday 5/27 family meeting, per father request given inability to visit during week.   
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: 4.205kg (05/25/22)  
Weight change: +115g (05/23/22 to 05/25/22