**NOTE 4**

**PATIENT 1003**

**DATE: 4/20/21**

Patient is an ex 36w1d now 44w1d infant with 15q11.2 deletion and Prune Belly Syndrome most recently s/p right ureterostomy who remains admitted for nutritional optimization, sedation wean, and respiratory support. Patient was previously admitted to NICU from HOSP 1 and transferred to Nephrology Service on 4/11. He is now being transferred back to NICU for initiation of CPAP respiratory support in the setting of hypercarbia and increased WOB.  
  
**Nephrology Course by Systems (4/11-4/20)**  
CV: Last echo on 2/27 showed moderate PDA. Patient has a history of elevated blood pressures which were controlled initially with PRN isradipine and labetalol. However, on 4/20 he was started on q12hr amlodipine. He remains on PRN isradipine with labetalol as a second agent to maintain SBPs <110.  
  
RESP: History of intubation and bilateral pneumothoraces. Weaned to CPAP and then NC on 4/4. He continued on his supplemental oxygen which was weaned as tolerated. Pulmonary was consulted on 4/18 for outpatient discharge planning and they recommended getting a VBG which showed hypercarbia. The decision was made to transfer to NICU to initiate CPAP.  
   
FEN/GI: Currently receiving 150ml/kg/day of breast milk or Neosure 26cal/oz decanted with Kayexalate. He is able to PO up to 15ml per feed. Started on Vitamin D and calcitriol. Receiving calcium carbonate for phos binding. The plan had been for a modified barium swallow prior to decision to start CPAP. BMP obtained twice weekly.  
   
RENAL/GU: History of right nephrostomy. He underwent PUV ablation, right cutaneous loop ureterostomy, and circumcision on 4/7.  
   
ID: Asplenia, on amoxicillin prophylaxis. Infant has been afebrile.  
   
HEME: Receiving epoetin twice per week and ferrous sulfate supplementation. Hct 24.5 on 4/20.  
   
NEURO: He had EEG on 4/14 and evaluation with Neurology which said there was no seizure correlate. He continued on his morphine wean. He was initially weaned to PRN dosing but experienced signs of withdrawal, requiring resumption of scheduled morphine on 4/15. He was gradually weaned off of morphine on 4/19, and is continuing clonidine.  
   
ENDO: History of elevated TSH, but ultimately normalized TFTs on 4/11. PTH at that time elevated to 485.8 with Vit D 66.1.  
   
GENETICS: Cord blood micro array with abnormal result (chromosome 15 deletion - 15q11.2) and genetics was consulted. Genetic testing (Invitae's Expanded Renal Disease Panel + ACTA2, FLNA, and MYOCD) sent 3/17.  
  
RHCM:  
[ ] Hepatitis B Vaccine/2 month vaccines due 4/23 (parents previously declined Hep B vaccine)  
[x] CCHD screen (n/a - echo)  
[x] NBS 24 HOL sent on 2/24: normal  
[x] DOL 14 sent on 3/8: normal  
[x] DOL 30 sent 3/23: elevated TSH (14.5), normal T4 (15.3) - serum testing sent, see above  
[ ] Newborn Hearing screen: Will need prior to discharge  
[ ] Car seat test: Will need prior to discharge.  
[ ] PMD: Parents undecided.

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**Plan**

CV: Hemodynamically stable, with intermittent HTN on isradipine PRN. Echocardiogram 2/27 structurally normal heart with moderate PDA. Hypertension, started amlodipine 4/20 with isradipine/labetalol PRN with goal SBP <110. **Will need f/u ECHO**. Monitor closely  
Access: none  
  
Resp: New respiratory distress prompting admission to NICU 4/20 for initiation of CPAP. History of pulmonary hypoplasia secondary to renal abnormalities and history of RDS requiring intubation, HFOV, surf x2, and b/l pneumothorax s/p needle decompression and L CT, now resolved. Extubated 3/15, off CPAP 4/5. Titrate CPAP as needed while monitoring WOB, gas exchange and CXR as needed. Pulmonary consulting, potential sleep study in future  
  
FEN/GI: 150 mL/kg/day MM/Neo26. Had been trialing POs, will be NPO while on CPAP. Asplenia and malrotation, surgery following. Continue calcium carbonate, vitamin D, calcitriol. Monitor I/O, growth, electrolytes M/Th. Feeding team following, consider swallow study in future.  
  
Renal: Eagle Barrett/prune belly syndrome w/ right sided megaureter s/p right nephrostomy (2/26-4/7).s/p right ureterostomy, PUV ablation and circumcision 4/7. Nephrology and Urology following.  
  
ID: Asplenia on amoxicillin baseline, 10 mg/kg daily renally dosed. S/p staph epi positive blood culture treatment. No current infectious concerns.  
  
Heme: H/o anemia requiring PRBCs. Continue Epogen M/Th and trend CBC q2 weeks  
  
Neuro: HUS negative. Neurology consulted with abnormal neurological examination at HOSP 1 (slow to arouse after weaning all sedation, and had excessive tremors). He had EEG on 4/14 and evaluation with Neurology which had no seizure correlate. Monitor  
  
Sedation: On enteral clonidine, s/p morphine 4/19. Start weaning clonidine in future  
  
Endo: History of elevated TSH, now improving. Endocrine following.  
  
Genetics: Cord blood microarray with abnormal result (chromosome 15 deletion - 15q11.2) and genetics was consulted. They met with the family and recommended genetic testing for the parents (test for 15q11.2 microdeletion before next pregnancy and can self refer to a local geneticist). They also recommend Prevention Genetics Comprehensive inherited Kidney Diseases Panel (329 genes) with concurrent reflex of ACTA2, CHRM3, FLNA, HFN1B, and MYOCD. Plan to follow up outpatient with genetics clinic.  
  
Social: Family updated on admission. Continue to inform and support parents.  
  
Routine Health Care Maintenance  
[ ] Hepatitis B Vaccine/2 month vaccines due 4/23 (parents previously declined Hep B vaccine)  
[x] CCHD screen (n/a - echo)  
[x] NBS 24 HOL sent on 2/24: normal. Repeat next due on DOL60  
[x] DOL 14 sent on 3/8: normal  
[x] DOL 30 sent 3/23: elevated TSH (14.5), normal T4 (15.3) - serum testing sent, see above  
[ ] Newborn Hearing screen: Will need prior to discharge  
[ ] Car seat test: Will need prior to discharge.  
[ ] PMD: Parents undecided.  
  
Disposition: Pending stabilization of respiratory status