**NOTE 6**

**PATIENT 1003**

**DATE: 6/3/21**

**Patient Summary**

Patient is a 3mo old 36w infant with 15q11.2 deletion, Prune Belly Syndrome, CKD (no function in left) s/p right ureterostomy with declining renal function s/p PD cath and GT 4/26. On PD since 5/17. Extubated on 5/26, on CPAP.

**Events in Last 24 hours**  
Remains on CPAP 7 and hold for wekeend   
On cycler 160 for 12 cycles, UF 230  
Ongoing sedation management  
Dl CVL line out today (leaking)  
In afternoon found to have "pus: like material coming from uterostomy tube. Stable CBC, blood culutre sent, and UA showed 100 WBC. Urology consulted and ? concern for fungal etiology. Initiated abx  
  
  
**Plan**  
CV: HDS. HTN s/p amlodipine/labetalol (d/c 5/20, 5/21), off all HTN meds now that on PD.  Monitor BO closely.  
Most recent echo (4/21) w/ LVH and dynamic LVOT 2/2 cHTN and anemia, cardiology following, plan to repeat in 1-2 weeks after starting PD per cardiology. **Plan for echo week of 6/5.**  
  
Access: PD catheter (4/26-), s/p DL L subclavian PICC placed by surgery on 5/8-6/3.  
  
Resp: Extubated 5/26 to CPAP, weaning gradually. **Remain on CPAP 7 through the weekend. Next wean week of 6/5.**  H/o pulmonary hypoplasia 2/2 renal abnormalities and RDS requiring intubation, HFOV, surf x2, and b/l pneumothorax s/p L CT, now resolved.  
  
FEN: TFG 80 ml/kg/d 46 kcal MM/Sim Advance via GT. Potassium slightly uptrending today 6/2, will d/w renal. S/p Kayexalate, d/c 5/18 given better potassium. Calcium carbonate and calcitriol, adjusting based on levels, continue glycerin PRN.  Monitor I/O, UOP, weight, growth. End stage renal following. Monitor lytes closely.   
GI: Asplenia, malrotation is possible with his syndrome but this has not been studied, suspicion low per surgery, surgery consulted in past.  
  
Renal: Eagle Barrett/prune belly syndrome w/ right sided megaureter s/p right nephrostomy (2/26-4/7), right ureterostomy, PUV ablation and circumcision (4/7), and PD catheter placement (4/26). PD started started 5/17.  Monitor urine output, lytes, FB closely. Nephrology and Urology following.  
  
ID: S/p treatment for staph epi bacteremia 3/16. Asplenia on amoxicillin ppx. Now with concern for pus coming from uterotomy tube. On cefepime and Flagyl. Urology and Renal involved  
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Heme: H/o anemia requiring PRBCs, most recent 4/21. Last HCT 32% on 5/21. Continue Epogen M/Th - increased (5/23)- and trend CBC qweekly. On Fe.  
  
Neuro: HUS negative. Neurology consulted previously (at HOSP 1) due to slow to arouse after sedation wean/tremors. EEG (4/14) no seizure correlate. Continue to monitor.  
  
Sedation: **Currently on morphine s/p conversion 6/2**Enteral clonidine and ativan. Increase clonidine 6/2.  
  
Endo: H/o elevated TSH,  now improved and within normal range, no indication to repeat.  
  
Skin: Small red spot on right side of head, continue to monitor site, CNS 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: Continue to inform and support parents. Family meeting 5/8. Now able to hold baby, d/w dialysis team. Parents updated on rounds 6/2.  
  
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.  
[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  
[ x] DOL 60 - elevated C5DC, metabolism recs: acylcarnitine profile, free/total carnitine, urine organic acids, and urine acylglycines - will review with metabolism  
[ ] Newborn Hearing screen: Will need prior to discharge  
[ ] Car seat test: Will need prior to discharge.  
[ ] PMD: Parents undecided.  
  
Dispo: requiring ICU care for chronic respiratory failure

**Weight**  
Last weight:  5.51kg (06/02/21)  
Weight change:  +140g (06/02/21 to 06/02/21)  
Fluid Balance (6/2/2021 07:00 to 6/3/2021 06:17) In: 473 mL / Out: 340 mL / Balance: +133 mL