**NOTE 5 \***

**PATIENT 1003**

**DATE: 5/21/21**

**Patient Summary**

Patient is a 2+mo old 36w1d 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, remains fluid restricted, intubated, on anti-hypertensives. Started PD 5/17 plan to continue over weekend. Starting to focus on weaning respiratory support, considered weaning rate and or PEEP over weekend.

**Events in Last 24 hours**  
Calcium, calcitriol d/c  
CVL sutures noted not intact, Surg aware  
CXR confirms CVL central SVC  
Bleeding from CVL site, re-dressed, Surg to re-suture  
Mild hypotension, amlodipine held  
CBC ok  
  
**Plan**  
CV: **Hypotension noted 5/21, likely in setting of dehydration s/p NS bolus x2 with improvement, d/w Renal**. HTN s/p amlodipine/labetalol (d/c 5/20, 5/21), and PRN isradipine, goal SBP <110.  Follow BPs and titrate as indicated. 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.  
Access: PD catheter (4/26-), DL L subclavian PICC placed by surgery on 5/8, central, **noted sutures not intact 5/21, still central on CXR, bleeding noted, Surg resutured 5/22.**  
  
Resp: Remains intubated, likely plan to remain intubated through PD.  **Goal weaning support over the weekend.** **Planning to get to minimal settings then work on pressure support sprints for several days before attempting extubation.** Trend blood gas and titrate support to adequate gas exchange.  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: Fluid restricted to TF 60ml/kg/d of 54 kcal MM/NeoSure vis GT. 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. Daily electrolytes while starting PD.  
GI: Asplenia and malrotation, 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.  Getting 18 cycles a day with 1.75% dex solution, 20/kg. **Given fluid-responsive hypotension 5/21, planned 8 cycles overnight, with close observation and more cycles if normotensive/tolerates, to goal 18 cycles.** Monitor urine output, lytes, and fluid status closely. Nephrology and Urology following.  
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ID: S/p treatment for staph epi bacteremia 3/16. Asplenia on amoxicillin ppx.  
  
Heme: H/o anemia requiring PRBCs, most recent 4/21. Last HCT 32% on 5/21. Continue Epogen M/Th - increased (5/16)- and trend CBC qweekly. On Fe.  
  
Neuro: HUS negative. Neurology consulted with abnormal neurological examination at HOSP 1 (slow to arouse after weaning all sedation, and had excessive tremors). EEG on 4/14 and Neurology evaluation, no seizure correlate. Monitor.  
  
Sedation: Currently on midaz, fentanyl and Precedex. Enteral clonidine  
  
Endo: H/o elevated TSH, improved and within normal range, no indication to repeat. Endocrine signed off.  
  
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.  
  
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 and initiation of peritoneal dialysis

**Weight**  
Last weight: 5.025kg (05/20/21)  
Weight change: +140g (05/19/21 to 05/20/21)  
  
Fluid Balance (5/20/2021 07:00 to 5/21/2021 06:59) In: 360 mL / Out: 339 mL / Balance: +21 mL