



PLAN OF CARE FOR PATIENTS REQUIRING TRANSFUSION

- I. Diagnosis- “day 1 talk”, including family studies.
- II. Start of Transfusions (Day One)
 - A. Provide information sheet and contract to family and patient
 - B. Check for hemochromatosis gene(s). Is patient heterozygous?
 - C. Extended phenotype
 - D. Obtain a baseline ferritin before transfusion.
 - E. Hepatitis, CMV and HIV testing.
 - F. All venous access ports placed should be plastic, if required
- III. Criteria for starting chelation
 - A. Transfusion #10-20
 - i. measure serum ferritin
 - B. Refer to audiology, ophthalmology and cardiology for echo/ EKG (if abnormal refer to cardiomyopathy clinic)
 - C. Between 10 and 20 transfusions with a serum ferritin greater than 1000-1500ng/ ml on 2 separate occasions, start chelation
 - D. Desferal 40 mg/kg starting dose 7 days/week, then giving a break
 - E. Exjade 20mg/ kg starting dose, may be escalated to 40 mg/kg max dose
 - F. Deferiprone (now available on compassionate use) if Exjade/ DFO not effective, **NOT to be used for DBA patients**
 - G. When patient is between 6 and 8 years old, consider a T2* to assess cardiac iron burden
- IV. First Year
 - A. Ferritin levels to be drawn every three months for first years, or more frequently as a compliance tool.
 - B. Urine dip for protein monthly, send for urine protein/creatinine ratio if UA positive for protein
 - C. If iron is in balance, (ferritin 1000 ng/ ml-1500 ng/ ml), continue without change. If serum ferritin is elevated, do an LIC by MR at age five, or when children can sit still without anesthesia. If abnormal, liver biopsy is indicated:
 - i. If LIC is greater than 7mg/gm dry weight (R2*) of liver, liver biopsy should be performed to look for fibrosis and cirrhosis.
 - ii. If LIC is greater than 7mg/gm dry weight, also investigate compliance, speculation of hemochromatosis, or incorrect dose of iron chelator



- V. Ongoing Care
- Monthly urinalysis
 - Ferritin every 3 months
 - Chemistry panel monthly
 - Thyroid function and glucose tolerance testing annually
 - Annual ophthalmology and audiology while on chelation
 - Yearly LIC by MR
 - First T2* same as LIC, then yearly thereafter for assessment of cardiac Status
 - Annual cardiac evaluation, including echo/ ECG, exercise stress test
 - Endocrine evaluation as needed
 - Annual Hepatitis and HIV testing

HEPATIC IRON CONCENTRATION IN IRON OVERLOAD

	mg/g dry weight	Micromoles/g dry weight	Micrograms/gram Dry weight	Hepatic Iron Index (NOT USEFUL IN PEDS)
Normal female	<7	< 30	400-2200	< 1
Normal Male	<7	< 40	400-2200	< 1
Mild		30/40-100	3000-10,000 (mild/moderate)	
Moderate		100-200		
Severe		> 200		> 1.9
Severe with fibrosis		Approx >=400	22000	

FORMULAS:

1) Micromoles/ g = n

Micrograms/ g = $\frac{n \text{ micromoles/ g} \times 55.8}{1000}$

Milligrams/ g = $\frac{n \text{ micromoles/ g} \times 55.8}{1000}$

2) Hepatic Iron index = Hepatic Iron in $\frac{\text{Micromoles/g}}{\text{Age in years}}$

3) Conversion of mg/g wet weight to dry weight:
 Dry = 0.1833 X wet weight value