Routine Procedures and Investigations for Adult Hemoglobinopathy Clinic Patients

To be performed when new patients are brought into the room (document on visit note sheet):
- Confirm diagnosis
- List of medications and doses
- Any symptoms today?
- Vital signs

To be performed when follow-up patients are brought into the room (document on visit note sheet):
- List of medications and doses (ask if any issues, any changes in dose)
- Any symptoms today?
- Any new concerns?
- Vital signs

Laboratory tests:
New sickle cell disease
- CBC, peripheral smear, reticulocyte count, ferritin, Hb electrophoresis, creatinine, AST, ALT, GGT, ALP, bilirubin, group and screen (with note in comments field to perform RBC phenotyping), urinalysis, calcium, vitamin D, chronic hepatitis serology (HBV and HCV), HIV, TSH, ft4, fasting glucose, FSH, LH, total testosterone (men), estradiol (women)

New thalassemia major and intermedia
- CBC, peripheral smear, reticulocyte count, ferritin, creatinine, AST, ALT, GGT, ALP, bilirubin, group and screen (with note in comments field to perform RBC phenotyping), urinalysis, calcium, vitamin D, hepatitis B & C serology, HIV, TSH, ft4, fasting glucose, FSH, LH, total testosterone (men), estradiol (women)

Sickle – follow-up visits
- CBC, peripheral smear, reticulocyte count, ferritin
- If monitoring or planning to start HU: MHS (“monitor Hb S”), creatinine, AST, ALT, GGT, ALP, bilirubin, urinalysis
- Yearly (all): calcium, vitamin D, TSH, ft4, fasting glucose, urinalysis, FSH, LH, total testosterone (men), estradiol (women)
- Yearly (if transfusion exposure in preceding year): chronic hepatitis serology (HBV and HCV), HIV

Thalassemia intermedia – follow-up visits
- CBC, peripheral smear, reticulocyte count, creatinine, AST, ALT, GGT, ALP, bilirubin, ferritin
- Yearly: calcium, vitamin D, chronic hepatitis serology (HBV and HCV), HIV, TSH, ft4, fasting glucose, FSH, LH, total testosterone (men), estradiol (women), urinalysis

Thalassemia major
- Monthly transfusion bloodwork – group & screen, CBC, peripheral smear, reticulocyte count, creatinine, AST, ALT, GGT, ALP, bilirubin, ferritin
- Yearly: calcium, vitamin D, chronic hepatitis serology (HBV and HCV), HIV, TSH, ft4, fasting glucose, FSH, LH, total testosterone (men), estradiol (women), urinalysis
Investigations:
Sickle cell disease
- **Echocardiogram**: baseline and then Q1-3yr, depending on results
- **Ophthalmology**: baseline and then Q6mos-2yr, depending on results
- **Ferriscan R2 liver MRI**: baseline if very high ferritin and heavy prior transfusion exposure and then Q1-2yr, depending on results, chelation and ongoing transfusion exposure
- **Cardiac T2* MRI**: baseline if very high ferritin and heavy prior transfusion exposure and then Q1-2yr, depending on results, chelation and ongoing transfusion exposure
- **BMD**: baseline and then Q1-5yr, depending on results

Thalassemia (major and intermedia)
- **Echocardiogram**: baseline and then Q1-3yr, depending on results
- **Ophthalmology**: baseline and Q1yr *if on desferal*
- **Audiology**: baseline and Q1yr *if on desferal*
- **Ferriscan R2 liver MRI**: baseline and then Q6mos-1yr
- **Cardiac T2* MRI**: baseline and then Q Q6mos-1yr (thal major) or Q6mos-3yrs (thal intermedia – depending on results, chelation and ongoing transfusion exposure)
- **BMD**: baseline and then Q1-5yr, depending on results