

## Iron chelation

- Annual assessment of the effectiveness of the chelation program, including discussion of heart, endocrine and other organ dysfunction. Review of chelation options, dosing, and evidence of adequacy of chelation on the present regimen.

## NON-ANNUAL OR OTHER

### Immunizations

- Influenza annually
- Pneumococcal (Pneumovax and Prevnar) (for splenectomized patients)
- Meningococcal (Menactra) (for splenectomized patients)



### Pulmonary function studies - post splenectomy

- Every three years or as indicated.

### Genetics

- Globin genotype (once)
- HLA typing (once)
- HLA typing for new siblings after birth
- Genetic counseling

If you have questions concerning comprehensive care or the location of a thalassemia treatment Center of Excellence, please contact the Cooley's Anemia Foundation at (800) 522-7222.

**The Cooley's Anemia Foundation**  
330 Seventh Avenue, #900 \* New York, NY 10001  
(800) 522-7222 \* Fax: (212) 279-5999  
info@cooleysanemia.org  
www.cooleysanemia.org



## **Comprehensive Care Checklist**

### *Recommended Annual Comprehensive Evaluation Test for Thalassemia Patients*

The following tests are recommended for most patients with thalassemia on an annual basis. However, since each patient's case is unique, your doctor may recommend some of these tests be done more or less frequently. While each test result is important, their trends over time must also be analyzed to detect early problems.

These are tests that are recommended in addition to monthly CBC's and quarterly chemistry panels and Ferritin measurements.



### Cardiac Evaluation

The following tests should be performed at least once per year.

- Assessment by a cardiologist knowledgeable in problems of iron overload and thalassemia.
- Echocardiogram (with assessment of tricuspid regurgitant jet velocity after age 10 to diagnose pulmonary hypertension, and left ventricular ejection fraction to diagnose cardiomyopathy. Functional MRI of heart and MUGA scanning are useful assessments)
- T2\* MRI of heart for cardiac iron
- 24 Hour Holter Monitor or event recorder to diagnose cardiac arrhythmia (after age 12)
- ECG (electrocardiogram)

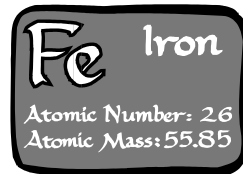
## Liver and Infection Evaluation

Annual testing for hepatitis, transfusion associated infections and response to vaccines are recommended.

- Annual Hepatitis C Antibody test if previously negative
- Annual hepatitis C RNA viral load (if not previously treated and antibody + )
- Liver Function Enzyme Screening (AST/SGOT, ALT/SGPT) every three months\*  
*If you have an elevated ALT, it is recommended you repeat the test every month. If ALT is persistently elevated for 6 months or more, a liver biopsy should be considered. All thalassemia patients should consider a liver biopsy to evaluate liver iron every two years.*
- Hepatitis B Panel
- Hepatitis A Panel (if not vaccinated or if hep A + in past )
- Annual HIV
- Serum AFP and abdominal ultrasound are annual screening tests for early liver cancer for patients with chronic hepatitis.

### *Iron Stores:*

- Annual or bi-annual quantitative liver iron assessment by liver biopsy or non-invasive measurement is recommended. As of 2006, the specialized Ferriscan technique is the only FDA approved non-invasive measure for hepatic iron; others are in development. SQUID (Superconducting Quantum Interference Device) is another non-invasive alternative but its availability to patients is limited.



## Endocrine Function Evaluation

Annual assessment in growing children should include growth and height velocity and puberty status. A bone age exam should be included every few years until puberty if low bone mass or slow growth is noted. After puberty, annual assessment should also include studies of gonadal function, fertility, impotence.

- TSH, Free T4, Parathyroid Hormone Level
- Fasting AM Cortisol
- Glucose Tolerance Test (for patients older than 10 yrs of age)
- Bone Density (for patients older than 8 years of age)
- Testosterone, FSH, LH, Estradiol and endocrinology consult recommended

## Ophthalmology Evaluation



- Annual evaluation by an ophthalmologist (for cataracts, night blindness, decreased visual field). (Especially recommended for patients on Desferal or with diabetes.)

## Audiological Evaluation

- Annual Audiological Evaluation for evaluation of hearing changes and Tinnitus (ringing in the ears)

## Transfusion Monitoring

- Annual assessment of number of red cell units transfused
- Review of transfusion complications (red cell antibodies, transfusion reactions)
- Review of the pre-transfusion hemoglobin values and frequency of transfusions