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Thalassemia Foundation of Canada
Anemia Institute for Research & Education
Thalassemia Clinicians and Nurses
Patients/Families

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Goals and Scope

Improving overall health and quality of life for thalassemia patients

   Essential components of multidisciplinary team approach

   Current evidence-based best practice

   Guide to thalassemia healthcare professionals & patients/families

Chapters

   Principles- desirable goals of centre
   Guidelines- requirements of the centre
   Interventions- specific steps to implement each guideline
Guidelines for the Clinical Care of Patients with Thalassemia in Canada
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<tr>
<th>Team Members</th>
<th>Satellite Clinic</th>
<th>Specialist Centre</th>
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<tbody>
<tr>
<td><strong>Physicians</strong></td>
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<tr>
<td>Consultant pediatric or adult hematologist (depending on age of patients) with experience in thalassemia</td>
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<tr>
<td>Pediatrician, internist, hematologist or general practitioner/internist with knowledge of thalassemia care</td>
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<td>On-call physicians for after-hours issues</td>
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<td><strong>Nursing</strong></td>
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<tr>
<td>Nurse specialist for thalassemia service: training, monitoring, co-coordinating and auditing</td>
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<tr>
<td>Registered nurses in outpatient day care unit area who can perform intravenous (I.V.) cannulation and supervise transfusions</td>
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<td>Nurse outreach into the community: home visits, teaching pump use, etc. This may be coordinated with local home care programs</td>
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<td>Nurse contact for patient and family</td>
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<td><strong>Access to other multidisciplinary team members</strong></td>
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<tr>
<td>Clinical psychologist</td>
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<td>Psychologist</td>
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<td>Social worker</td>
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<td>Dietician</td>
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<td><strong>Access to specialist consultants</strong></td>
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<td>Designated pediatric or adult cardiologist</td>
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<td>Designated pediatric or adult endocrinologist</td>
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<td>Designated pediatric or adult hepatologist</td>
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<tr>
<td>Genetic counselling</td>
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<td>Designated obstetrician and fertility program</td>
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<td>Bone marrow transplant service</td>
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<td><strong>Other support services</strong></td>
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<td>Appropriate laboratory support (transfusion, diagnostic), diagnostic imaging</td>
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<td>Access to translation services</td>
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<td>Administrative support sufficient to ensure record maintenance and proper communication between patient and family with clinic, centre, family doctor and all services involved</td>
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Clinical and Laboratory Aspects When Deciding Appropriateness of Initiating a Transfusion Program

New diagnosis of β-thalassemia

- At least monthly assessment until phenotype determined
- Complete assessment with emphasis on:

Indications for transfusions

History
- Symptoms of anemia
- Abnormal growth and development

Physical Examination
- Height and weight
- Facial deformities
- Bone deformities
- Hepatosplenomegaly

Laboratory Investigations
- Hemoglobin

Laboratory evaluation pre-transfusion initiation

Haematology
- Serial hemoglobin
- G6PD screen
- Rule out other causes of anemia

Transfusion
- Full red blood cell phenotype
- Ferritin

Biochemistry
- Liver enzymes
- Hepatitis B serology
- Hepatitis C serology

Virology
- HIV serology

Initiation of a transfusion program

Blood product
- Matched for major Rh and Kell antigens

Transfusion
- Pre transfusion target: Hb 90 – 100 g/l
- Approximately every 3 – 4 weeks
- Total needs < 200 – 250 ml/kg/yr
Monitoring of Iron Load and Appropriate Chelation Therapy

Start Transfusions

Iron Load Monitoring
After 10 - 20 transfusions
Ferritin 1000 - 2500 µg/ml OR
LI C 7 - 15 mg Fe/g dry weight

Intolerant
Ineffective
Non-compliant

Start Chelation
Deferoxamine
20 - 40 mg/kg/d over
8 - 10 hours on 5 - 7 days

Deferasirox
10 - 30 mg/kg/d
- Optimize/increase chelation treatment
- Encourage compliance
- Re-assess every 6 months

Iron Load Monitoring
Ferritin 1000 - 2500 µg/ml AND
LI C 7 - 15 mg Fe/g dry weight AND
Cardiac T2* 10 - 20 msec

Cardiac function assessment
Normal
Abnormal

Aggressive Chelation
1. Deferoxamine
   Continuous infusion > 50 mg/kg/d
   (max 6 g/24 hours), OR
2. Change to oral iron chelator, OR
3. Consider combination therapy with deferiprone and deferoxamine
Next steps

Implementation of guidelines

- Guide for healthcare professionals
- Tool to advocate for resources
- Benchmark to gauge success
Acknowledgments

Dr. Farzana Sayani
Dr. Molly Warner
Dr. John Wu
Dr. Durhane Wong-Rieger
Kimberley Humphreys

Reviewers- Canada and International

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