



PART I

**Components of Comprehensive
Thalassemia Care**

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COMPONENTS OF COMPREHENSIVE THALASSEMIA CARE

1. Centres of Excellence: Providing a Network for Patient Care

Principles

- To provide thalassemia patients with comprehensive care delivered by a multidisciplinary network of specialized centres and satellite clinics.
- To ensure centres provide excellent routine care adhering to standards of clinical practice. This includes prevention and appropriate management of complications, in order to decrease morbidity and mortality, and improve quality of life for thalassemia patients.
- To ensure all thalassemia patients have access to the same quality of care, regardless of geographic location.
- To ensure patients receive their routine thalassemia care in a convenient, accessible manner with minimal disturbance to their everyday activities.
- To transition the care of thalassemia patients from pediatric clinics to adult clinics in a timely manner while providing high quality of care and support for patients and their families.

Recommendation

- Each program should consist of a collaborative regional network of one or more specialized thalassemia centres and affiliated satellite clinics. These facilities may also provide services for patients with other hemoglobinopathies and transfusion-dependent anemias if patient numbers do not justify the use of separate programs.
- Each program should have a formalized system for specialized thalassemia services that is appropriately configured to both the geographical location and the size of the patient population.
- Patients living within reasonable distance of a specialist centre should have access to this centre for both routine thalassemia-related care and specialized services.
- Patients living in remote areas should have access to a satellite centre for routine thalassemia care. They should access the affiliated specialist centre for services not available locally. This includes regular annual or semi-annual expert clinical review by a hematologist with expertise in treating thalassemia.
- Each program should have effective communication, documentation and accountability between satellite and specialized centres.
- Both the specialized centre and the affiliated satellite clinics should have regular opportunities for self-evaluation and staff development.
- Each program should have a transition plan from pediatric clinics to adult clinics. This includes transition clinics, education, activities planned in advance of the actual transition time.

Background

In order to provide comprehensive, high quality care to thalassemia patients, they must have easy access to both a trained hematologist and to subspecialists that can assist in treating thalassemia-related complications. A network of centres with specially trained staff ensures all patients have access to appropriate care, including those living outside major urban centres. This network consists of both specialized thalassemia centres in major urban areas, and affiliated satellite clinics in outlying areas.

The specialized centre is a facility with a multidisciplinary team of staff that are experienced and focused on the diagnosis, treatment and care of thalassemia. The team is lead by an adult or pediatric hematologist with a special interest or training in thalassemia, and dedicated nursing and subspecialty staff is available to monitor the patients' ongoing care

and complications. A satellite clinic is led by a physician with knowledge of thalassemia, and operates in conjunction with local outreach and hospital services. In this network model, the satellite clinic serves as the primary access point for the patient's routine thalassemia care, while the specialist centre manages the more complex issues that thalassemia patients experience. While the services provided by satellite clinics may vary, it is expected that each is able to adhere to the standards below when providing thalassemia care. Conversely, specialized centres are expected to provide routine thalassemia care to patients for whom this facility is the most easily accessible. By having a network of centres dedicated to providing high quality thalassemia care, patients are assured comprehensive services no matter their residential location.

Interventions

Role of the Specialized Centre

- Provide consultation at key thalassemia-related milestones including diagnosis, initiation of regular transfusions, initiation of chelation therapy, times of major complications, and transfer to an adult clinic. These consultations should be provided in addition to regular annual or semi-annual reviews.
- Provide specialist opinion on the management of complex issues including but not limited to: transfusion management; chelation therapy; phlebotomy management; compliance problems; peri-operative management; management of cardiac, liver, endocrine, and bone complications; fertility issues and genetic counseling; bone marrow transplantation and clinical trials if available; and complex psychosocial issues.
- Provide consultation, education, and training for staff at both the specialized centre and its affiliated satellite clinics.
- Participate in quality improvement including monitoring the program's adherence to standards of care, performance, and care outcomes.
- Be involved in clinical research studies and continuing education to improve overall patient care.
- Advocate for improved care and service delivery at the local, provincial and national levels.
- Provide transition care from pediatric to adult clinics, in a timely manner, planned in advance of transition time.

Role of the Satellite Clinics

- Provide regular transfusions, prescriptions for chelation therapy, and other necessary therapies that constitute routine thalassemia care.
- Monitor growth, development, and general health.
- Monitor psychosocial well-being and provide psychological assessment and treatment when necessary. This may require referral to the specialized centre.
- Organize the routine assessments and monitor tests when locally available. Referrals to the specialized centre should be made for investigations not available in locally.
- Be a local resource of information and support for the family.
- Provide treatment to the patient and family in a way that minimizes disturbance to normal, everyday activities.
- Communicate regularly with the specialized centre, and make referrals for consultation as needed.

Staffing

- The specialized centre should be led by a pediatric or an adult hematologist with experience in thalassemia care, and the satellite clinic by one or more clinicians with knowledge of or interest in thalassemia.
- Each site should have a designated primary nurse to provide support and guidance on routine care and assist in accessing local services. This nurse should be the key contact for the patient and family.
- A psychologist and social worker should be integral members of the interdisciplinary team.
- Members of the interdisciplinary team should meet on a regular basis to discuss patients with emphasis on the medical, nursing, and psychosocial needs of the patients and family.
- Staff should be well-trained in the different aspects of thalassemia, and maintain their knowledge through

various means of continuing education.

- Staff providing routine thalassemia care should have well-developed intravenous insertion skills.
- Staff turnover should be minimized to allow for the development of effective, long-term relationships between the team and patients.
- Other crucial members of a multidisciplinary team at the specialized centre and satellite clinic should be present, as outlined in Table 1.

Facilities

- Designated facilities with adequate space should be provided for staff-patient consultation and the provision of routine thalassemia care.
- Equipment used in these facilities should be maintained in accordance with each centre's policies.
- Several specialties should be available for consultation at the specialized centre, and if possible, at the satellite clinic. These specialties include but are not limited to audiology, cardiology, endocrinology, genetic counseling, interventional radiology, nuclear medicine, obstetric and fertility medicine and ophthalmology. Multiple consultants from each should be available to assess and provide care for the patient.
- Both specialized centres and satellite clinics should have access to a transfusion medicine laboratory for the provision of correctly collected and matched blood products.
- Facilities at specialized centres should provide special investigations such as MRIs, CT scans, and bone density testing.
- Effort should be made to provide transfusions and other routine care at times that are convenient to patients and families.

Quality Assurance

- Each formalized regional network should have systems in place for regular self-assessment of clinical practice and services.
- Each centre, either specialized or satellite, should design and implement internal audits of clinical practice, adherence to practice guidelines, and services available to patients and families.
- When available, each centre should participate in external audits of their clinical practices, adherence to practice guidelines, and service available to patients and families.

Team Members	Services	Sattelite Clinic	Specialized Centre
Physicians	<ul style="list-style-type: none"> Consultant pediatric or adult hematologist (depending on age of patients) with experience or training in thalassemia 		●
	<ul style="list-style-type: none"> Pediatrician, internist, hematologist or general practitioner with knowledge of thalassemia care 	●	●
	<ul style="list-style-type: none"> On-call physicians for after-hours issues 	●	●
Nursing	<ul style="list-style-type: none"> Thalassemia nurse specialist to provide training, monitoring, co-coordinating and auditing of patients and program 		●
	<ul style="list-style-type: none"> Primary nurse contact for patient and family (such same as a nurse specialist) 	●	●
	<ul style="list-style-type: none"> Registered nurses in outpatient day care unit area who can perform intravenous (I.V.) cannulation and supervise transfusions 	●	●
	<ul style="list-style-type: none"> Nursing services for community outreach if necessary: home visits, central line care, teaching pump use, etc. 		●
Access to other multidisciplinary team members	<ul style="list-style-type: none"> Clinical psychologist 		●
	<ul style="list-style-type: none"> Social Worker 	●	●
	<ul style="list-style-type: none"> Dietician 	●	●
Access to specialist consultants	<ul style="list-style-type: none"> Designated endocrinologist 		●
	<ul style="list-style-type: none"> Designated cardiologist 		●
	<ul style="list-style-type: none"> Designated hepatologist 		●
	<ul style="list-style-type: none"> Designated ophthalmologist 		●
	<ul style="list-style-type: none"> Designated audiologist 		●
	<ul style="list-style-type: none"> Genetic counselling 		●
	<ul style="list-style-type: none"> Designated obstetrician and fertility program 	●	●
	<ul style="list-style-type: none"> Bone marrow transplant service 		●
Other support services	<ul style="list-style-type: none"> Appropriate laboratory support (transfusion, diagnostics) and diagnostic imaging 	●	●
	<ul style="list-style-type: none"> Access to equipment for specialized investigations such as MRIs and bone density testing 		●
	<ul style="list-style-type: none"> Access to translation services 	●	●
	<ul style="list-style-type: none"> Administrative support sufficient to ensure proper record maintenance and communication between patient and family with clinic, centre, family doctor and all services involved 	●	●

Table 1: Staffing Recommendations for Sattelite Clinics and Specialized Centres