CHAPTER 9
MANAGEMENT OF SICKLE CELL DISEASE

PAIN MANAGEMENT IN SCD

Sickle cell pain should be managed aggressively. Pain relief should be based on assessment and the analgesia ladder used accordingly. The treatment plan for each patient is individualised, chartered and monitored by using assesment tools as described previously (page 84-85). Management of sickle cell disease pain should include rest, hydration and pharmacological and non-pharmacological methods of pain relief.

- **Hydration**

  Fluid management is a very important aspect of management of sickle cell pain, as dehydration is known to precipitate a crisis. In addition, patients with sickle cell disease do not concentrate urine very well, which can lead to increased frequency of passing urine resulting in reduced fluid in both the vascular and tissue compartments, which in turn increases the problem of intracellular dehydration and increased plasma viscosity.

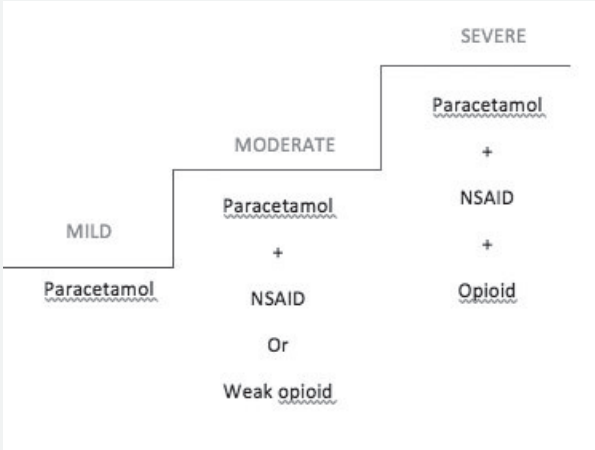
  Patients with vaso-occlusive crisis should be well-hydrated, through the oral or intravenous route. Venous access can often prove difficult in patients and great care should be taken in canulation. The oral route should be encouraged when possible. When patients have abdominal symptoms or are unable to tolerate oral fluids, the intravenous route should be used but discontinued once the patient is stable and the pain controlled.

  Patients with sickle cell disease often require 1.5 times the normal fluid requirement (2-3 litres over 24hours in adult patients), except when contra-indicated. However, fluid overload must be avoided as it can complicate an acute chest crisis, if present.

  The nurse should ensure that the patient has access to fluids at the bedside and document signs of dehydration such as dryness of the skin, cracked lips, sunken eyes, increased jaundice, particularly of the eyes, and increased or decreased urine output. An adequate fluid balance chart should be maintained, as this will guide the medical team in the management of the patient.

- **Pharmacological management**

  Analgesia should be tailored according to the needs of the individual patient, following a successful pain assessment, and should consist of opiates, non-opiates and adjuvant medication, which can be used singly or in combination to achieve adequate pain relief. The WHO guideline for analgesia drug therapy (shown below) suggests the administration of analgesia by the ladder, by the clock and by the appropriate route.



WHO Analgesic ladder

Analgesia

Entonox (nitrous oxide gas) - can be used for a short period of time in the ambulance/first 30-60mins in hospital (holding measure).

Paracetamol - this could be oral, by the intravenous route, or the rectal route (20mg/kg/ dose start, then 15-20mg/kg every 4-6hours in children. (It is essential to follow the manufacturer's guidelines.)

NSAIDs* - Ibuprofen or Diclofenac. (Caution in renal or liver impairment.)

Opiiods

- Intra-nasal Diamorphine-0.1mg/kg (max 6mg) given within first 5 minutes (one dose only, in children over 10kg).
- Codeine phosphate for oral use-available as a syrup 0.5-1mg/kg in children under 12 years and tablets 30-60mg above 12 years. Should be used with caution if the patient has decreased kidney or liver function.
- Dihydrocodeine tablets-30mg every 4-6 hours. Not recommended for children under 12 years.
- Morphine sulphate (oral, rectal, subcutaneous, intramuscular or intravenous route)-examples of available preparations are liquid Oromorph, tablet Sevredol, modified-release (MST). The oral dose for adults is 10-20mg every 4 hours (may go up to 50mg in severe pain). In children, 5-10mg according to age and severity of pain. The MST continuous release suspension or tablets are given every 12 hours. Parenteral forms SC, IM and IV are given in severe pain in adults, usually from 2.5-10mg to a maximum of 20mg, and in children from 1-12 years 200mcg/kg to a maximum of 2.5mg.

*NSAIDs : Nonsteroidal Antiinflammatory Drugs

- Other opioid base drugs are available, such as tramadol, oxycodone and fentanyl.
- Intravenous patient-controlled analgesia (PCA)/nurse-controlled analgesia (NCA), through an electronic infusion pump.

The advantages of PCA are
- Quicker onset of analgesia
- Patient controlled
- Smaller amount of total morphine used
- Potentially fewer side effects
- Safe

Disadvantages of PCA include
- Side effects
- Not suitable for all
- IV access required
- Attached to a machine
- Equipment malfunction

The possibility of complications increase when adding a continuous infusion
- Proxy use
- Patient risk factors
- Lack of training
- Where the chest syndrome is an opioid side-effect or because it is too painful to take deep breaths
- PCA used by the patient over sedation should not be a problem. (?)

When should PCA be stopped?
- When patient's condition improves
- When other, less invasive methods are available
- When side-effects are greater than pain relief

- **Non-pharmacological methods**
  Non-pharmacological methods of pain control are useful in alleviating stress, as well as increasing the ability to cope with acute and chronic pain and thereby improving sense of well-being.

  They include:
- Massage, which can relax the tension in muscle tissue around the joints and decrease muscle pain

- Warm bath
- Application of heat pads/tiger balm
- Positioning
- Diversionary therapy e.g. play, video, TV, etc.
- Input of psychologists to improve positive thoughts (see below)
- Involve play specialist
- Use of Tens machine (transcutaneous electrical nerve stimulation), which can improve blood circulation with subsequent vasodilatation and reduction in vaso-occlusion, as well as suppression of the transmission of painful stimuli via the A and C fibres.

Psychological intervention

- Key to helping the patient to self-manage the disease and its symptoms
- Not usually undertaken during the acute episode
- Helpful in:
  - Stress management
  - Distracting techniques
  - Treating clinical depression
  - Improving self-esteem, etc.

The tables A, B and C below provide guidance on the use of some pain management schemes (as described in the book: Sickle Cell Disease, by Adlette Inati-Khoriaty MD 2008).

Table A

ACETAMINOPHEN DOSAGE ACCORDING TO AGE AND APPROXIMATE WEIGHT					
		DOSAGE			
AGE	APPROXIMATE WEIGHT RANGE	ORAL DROPS	SYRUP	CHEWABLES 80 mg	CHEWABLES 100 mg
under 3 months	Under 4.55 kg	1/2 dropper	1/4 tsp.	-	-
3 to 9 months	8.18 - 10.45 kg	1 1/2 droppers	3/4 tsp.	-	-
2 to 3 years	10.9 - 15.9 kg	2 droppers	1 tsp.	2 tablets	-
4 to 5 years	16.36 - 21.36 kg	3 droppers	1 1/2 tsp.	3 tablets	1 1/2 tablets
6 to 8 years	21.8 - 26.8 kg	-	2 tsp.	4 - 5 tablets	2 - 2 1/2 tablets
9 to 10 years	27.27 - 32.27 kg	-	2 1/2 tsp.	6 tablets	3 tablets
11 years	32.7 - 43.18 kg	-	3 tsp.	6 tablets	3 tablets
12 years & over	43.64 kg & over	-	3 - 4 tsp.	6 - 8 tablets	3 - 4 tablets

How supplied:
Drops: Each 0.8-m1 dropper contains 80-mg acetaminophen.
Syrup: Each 5-ml teaspoon contains 160-mg acetaminophen.
Chewables: Regular tablets contain 80-mg acetaminophen each. Double strength tablets contain 160-mg acetaminophen each.
Dosage may be given every 4 hours as needed but not more than 5 times daily.
Adapted from http://www.drelizabethdickey.com/condition.aspx?condition_id=53

Table B

IBUPROFEN DOSAGE ACCORDING TO AGE AND APPROXIMATE BODY WEIGHT					
		DOSAGE			
AGE	APPROXIMATE WEIGHT RANGE	ORAL DROPS	SYRUP	CHEWABLES 80 mg	CHEWABLES 100 mg
5 - 11 months	4.55 - 7.73 kg	1 dropper	1/2 tsp.	-	-
12 - 23 months	8.18 - 10.45 kg	1 1/2 droppers	3/4 tsp.	1 1/2 tablets	1/2 tablet
2 to 3 years	10.9 - 15.9 kg	2 droppers	1 tsp.	2 tablets	1 tablet
4 to 5 years	16.36 - 21.36 kg	-	1 1/2 tsp.	3 tablets	1 1/2 tablets
6 to 8 years	21.8 - 26.8 kg	-	2 tsp.	4 tablets	2 tablets
9 to 10 years	27.27 - 32.27 kg	-	2 1/2 tsp.	5 tablets	2 1/2 tablets
11 years	32.7 - 40.45 kg	-	3 tsp.	6 tablets	3 tablets
12 years & over	40.9 kg & over	-	3 - 4 tsp.	8 tablets	4 tablets

Adapted from http://www.drelizabethdickey.com/condition.aspx?condition_id=53

Table C

QUANTITY OF FLUIDS NEEDED FOR A PERSON WITH SICKLE CELL DISEASE*			
BODY WEIGHT (kg)	LITRES (recommended range per day)	BODY WEIGHT (kg)	LITRES (recommended range per day)
5	0.5 to 0.7	35	1.8 to 2.7
10	1.0 to 1.4	45	2.0 to 3.0
15	1.2 to 1.8	55	2.3 to 3.4
20	1.4 to 2.2	65	2.5 to 3.8
25	1.5 to 2.3	75	2.8 to 4.1
30	1.7 to 2.5		

* May need more with fever, pain, exercise and hot water

New scientific data, information and use of drugs and combinations should certainly be taken into consideration when a clinic or unit develops its protocols and algorithms and all the tables above constitute only examples.

ORGANISATION OF CARE

The tables below (A & B) provide an example of the possible organisation of the care that can be delivered by the GP or the medical specialist in a SCD reference centre (from Sickle Cell Disease, by Adlette Inati-Khoriaty MD 2008).

Table A

REGULAR HEALTHCARE PLAN BY PAEDIATRICIAN/INTERNIST OR FAMILY DOCTOR		
AGE	FREQUENCY OF VISITS	EVALUATION
Birth - 6 months	Every month	Physical exam (PE) and developmental assessment, vaccines, monthly CBC and when needed
6 months - 1 year	Every 2 months	PE and developmental assessment, vaccines, PPD and urinalysis at one year, CBC every 2 months and when needed
1 year - 5 years	Every 4 to 6 months	PE and developmental assesssment, vaccines, CBC every 6 months and when needed, yearly PPD, yearly eye check-up > 3 years
6 - 18 years	Every 6 - 12 months	PE, vaccines, CBC every 6-12 months and when needed, yearly PPD, yearly eye check-up
Over 18 years	Every year	PE, CBC every year and when needed, other tests as advised by doctor

Table B

CARE PLAN IN A COMPREHENSIVE SICKLE CELL DISEASE CENTRE	
EVALUATION	INTERVAL
Physical Exam • Birth - 6 years • 6 - 18 years • Over 18 years	 Every 2 - 4 months Every 4 - 6 months Every 6 - 12 months
Family genetics	At diagnosis
Genetic Counselling and education	At diagnosis and 1 - 2 times per year
Social Worker visits (home,school and work site)	At diagnosis and once a year
Laboratory tests: Blood (CBC, Kidney, liver tests and iron test) and urine (urinalysis)	Every 6 - 12 months
Other special laboratory and x ray studies	When needed
Abdominal echo (to look for gallstones and spleen/liver size)	Every 2 years (>6 years) and when needed
TCD (to study stroke risk)	Once a year > 2 years and more if indicated
Cardiac echo (studies heart function)	Once a year > 10 years
Evaluation (eyes, lungs, neurological)	Once a year and when needed
Psychological/Family/Therapy Consultation	Once a year and when needed
Physical Therapy Assesment (for joint problems and after surgery)	When needed
Developmental Screen	Once a year and when needed
Dietician	Once a year and when needed
Adolescent Centre Evaluation	At least once/year and when needed

MONITORING AND NURSING CARE

Nurses play a vital role in the management of sickle cell patients in pain. Continuous assessment and close monitoring are essential. Specific measures include:

- Close monitoring of observations, temperature, pulse, respiration, blood pressure and oxygen saturation.
- Give oxygen if below 95% in air.
- Document pain, sedation and nausea scores 1-2 hourly if on opiates.
- Monitor for drug side effects, such as constipation, pruritus, over-sedation, respiratory depression (if on opioids), nausea and vomiting, and ensure adequate treatment is given.
- Use of incentive spirometry in patients above 6 years, if pain is in the chest or the abdomen, or in the back above the diaphragm.
- Adequate education of the patient on home management and coping mechanisms.
- Seek multidisciplinary approach for complicated cases.
- Use hydroxyurea and blood transfusion therapy where patient quality of life is affected by frequent painful episodes.

TREATMENT OF SICKLE CELL DISEASE COMPLICATIONS

Sickle cell disease (SCD) is a chronic, life-long disease. However, with early diagnosis and treatment, together with parental education and involvement, affected people can survive into middle and late adulthood.

Treatment for SCD is usually aimed at avoiding crises, relieving symptoms and preventing complications. A major part of the care of sickle cell disease can be handled at home by parents. Responsible, dedicated and knowledgeable parents make a great difference to a child's survival and quality of life. The nurse plays a critical part in enabling and supporting parents in that role.

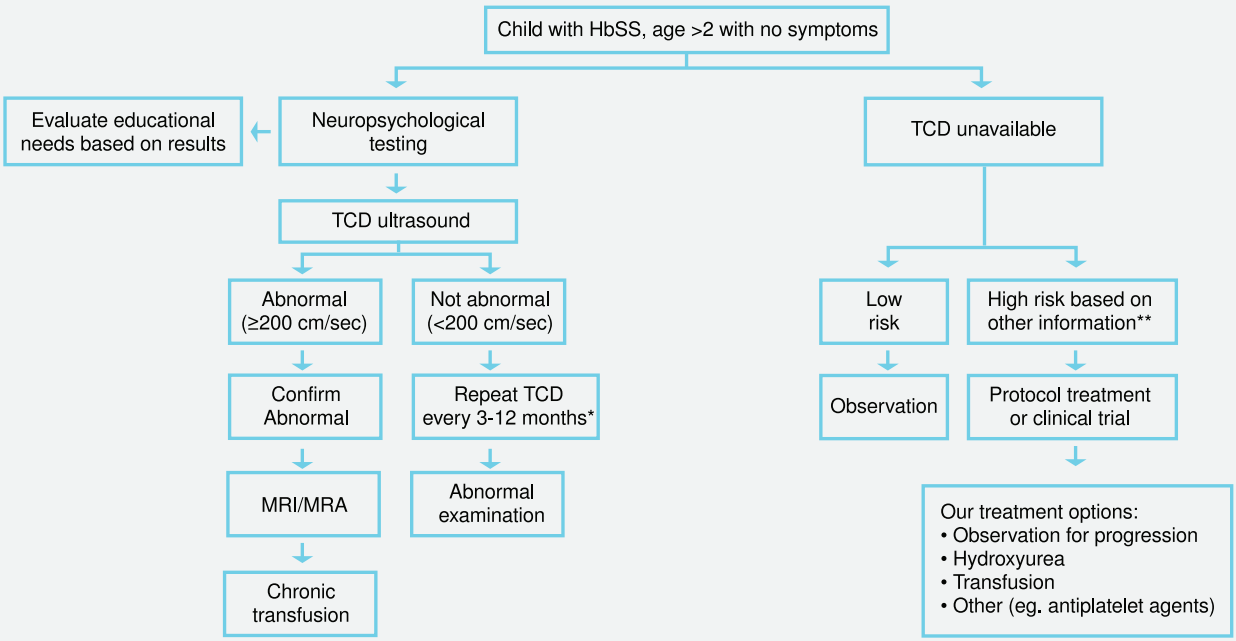
Once a child has been diagnosed with sickle cell disease, parents should be advised where possible to enrol at a specialised (comprehensive) sickle cell disease centre, where a team of experienced health professionals can oversee the child's care. Such teams include doctors of multiple specialities, nurse practitioners, genetic counsellors, nurses, social workers, play and occupational therapists, psychologists and dieticians. Adolescents and adults with sickle cell disease also benefit greatly from enrolment in comprehensive sickle cell disease centres, where regular screening for possible complications can be carried out, and new treatments and recent research discussed.

Stroke

Children who have had a stroke and those whose tests (TCDs) show they are at high risk for developing a stroke need to be treated with monthly blood transfusions for at least 5 years (if not indefinitely, as indicated by recent studies). These transfusions help prevent more strokes. Treatment for minor strokes is often the same as treatment for other strokes.

At the end of the treatment period, the patient will undergo special tests to determine if the risk of developing a stroke in the future is still present. If this risk is still present, then transfusion needs to be continued. If not, transfusion will be stopped and the child will be monitored very carefully. The doctor can explain the benefits of transfusion therapy in stroke and the treatment of transfusional iron overload, with the added support of the nurse.

A child that has had a stroke is best treated in a comprehensive sickle cell disease centre, under the supervision of an expert multidisciplinary team, able to offer neuropsychological testing and rehabilitation, in addition to advice about iron overload and chelation. A serious outcome of sickle cell strokes and other brain problems is the development of learning problems in some affected children. To identify these learning problems early, all children should be screened with routine exams, starting at 6 years (as shown in the below algorithm). If there are learning problems, these need to be managed from the outset.



*Optimal frequency of rescreening not established. Younger children with velocity closer to 200 cm/sec should be rescreened more frequently
** Prior transient ischemic attack, low staedy-state Hb, rate and recency of acute chest syndrome, elevated systolic blood pressure

Identification and management of stroke risk in children with sickle cell disease. http://www.nhlbi.nih.gov/health/prof/bloods/sickle/sc-mngt.pdf 2004.

Acute chest syndrome

This extremely common complication requires hospitalisation and urgent treatment. Any delay in treatment will expose a child to unfavourable consequences. The treatment of acute chest syndrome consists of broad spectrum antibiotics intravenously and by mouth, in addition to medication to open up the airways (bronchodilators). Early transfusion has been shown to improve recovery and shorten hospital stay. Administration of oxygen, medications to control fever and pain and incentive spirometry to prevent lung collapse (atelectasis) are important parts of treatment. Over-hydration can be dangerous and should be avoided.

Acute splenic sequestration

Acute splenic sequestration is a medical emergency. Failure to recognise the symptoms and signs of this dangerous condition can result in death within a few hours. The treatment of acute splenic sequestration is an immediate blood transfusion. Acute splenic sequestration can recur in a high percentage of patients and there are various treatment options for this condition, including chronic transfusion and complete or partial removal of the spleen (splenectomy). The doctor can outline treatment options for parents, explaining their pros and cons, with the support of the nurse.

Hand-foot syndrome

This early sign of sickle cell disease in children is a self-limiting condition without adverse resulting conditions (sequalae). Its treatment consists of hydration, analgesics such as acetaminophen or ibuprofen, and observation. If the pain is severe and non-responsive to the usual analgesics and/or if there is fever, the child's treating physician should be alerted. In such cases the patient may need hospitalisation for administration of stronger analgesics and intravenous hydration.

Surgery

People with sickle cell disease may need various types of surgery. The three commonest types are removal of the spleen (splenectomy) and removal of the gall bladder (cholecystectomy), and orthopedic surgery for problems like osteomyelitis, and avascular necrosis. The spleen is removed when it starts trapping blood suddenly, resulting in life-threatening anaemia (acute splenic sequestration). The commonest reason for removing the gallbladder is for gallstones. It is advised that transfusion be given prior to surgery to decrease the percentage of sickle cells (see below). This will improve oxygenation and may minimise complications. During surgery, the patient needs to be kept warm and must receive adequate hydration. Following surgery, blowing into a small mouthpiece (spirometry) will help decrease lung collapse and lung infection, which are the two commonest complications of surgery in sickle cell disease. Early mobilisation is essential and helpful to recovery.

BLOOD TRANSFUSION

This is not a lifelong component of treatment in SCD, as it is in β-thalassaemia major. However, it is indicated in SCD in some situations, including:

- **Severe anaemia**
- **Prevention and treatment of stroke**
- **Prolonged, painful erection of the penis (priapism)**
- **Lung infarction or pneumonia (acute chest syndrome)**
- **Surgery**
- **Frequent and severe painful episodes**

Below is a table describing in more detail the indication for transfusion therapy in SCD.

Indications for transfusion therapy in sickle cell disease[7,24]

Acute/episodic	■ Anemia ■ Splenic sequestration ■ Severe or long-lasting aplastic crises ■ Stroke ■ Acute chest syndrome ■ Multiple-organ failure syndrome ■ Pre-operative (in select cases) ■ Malaria-associated severe hemolytic anemia with impending cardiac decompensation
Chronic	■ Heart failure ■ Prophylaxis against recurrent stroke ■ Stroke prevention when transcranial doppler velocities are abnormal ■ Chronic pulmonary hypertension (unresponsive to other approaches) ■ Refractory congestive heart failure ■ Severe recurrent vaso-occlusive crises ■ Previous splenic sequestration in a child aged 2–3 years (in anticipation of later splenectomy) ■ Chronic pain

Stuart MJ, Nagel RL. Sickle-cell disease. Lancet 2004;364:1343-1360
24:Vichinsky E. Consensus document for transfusion-related iron overload. Semin Hematol 2001;38:2-4

In sickle cell disease, blood transfusions increase the number of normal red blood cells in circulation and decrease the number of rigid sickle cells, thus helping to relieve anaemia and improve blood flow to tissues. In children with sickle cell anaemia at high risk of stroke, regular blood transfusions can decrease the risk of stroke. If used appropriately

transfusion therapy can prevent or minimise organ damage in many people with sickle cell disease.

There are two types of transfusions a patient may receive: simple or exchange. Below is a table describing the various approaches to transfusion therapy.

APPROACHES TO TRANSFUSION THERAPY			
APPROACH	APPLICATIONS	ADVANTAGES	DISADVANTAGES
Simple transfusion Patients are given additional units of blood without removal of sickle cell blood	Severely anemic patient Hb levels 5-6 g/dl	• Simple and effective • Widely available	• Iron Loading
Automated exchange transfusion (erythrocytapheresis) Sickle cells are removed and replaced with normal red cells	Prefered when rapid alteration of Hb is required	• Rapid • Little net iron gain • Decreases HbS while leaving haematocrit and whole blood viscocity unchanged	• Increased red cell utilization • Increased rate of donor exposure, therefore, increased risk of infection or alloimmunisation
Rapid exchange transfusion Whole blood removed from one arm while donor cells are transfused in the other	Widely appropriate	• Little net iron gain	• Somewhat slower • Careful control of blood removed versus blood infused required

A simple transfusion involves the transfusion of a set amount of blood into the patient. An exchange transfusion involves giving a certain amount of blood while removing the same amount. This type of transfusion is used mostly in sickle cell disease in order to replace sickle cells with normal cells. It will also increase the haemoglobin level without increasing blood viscosity, and may result in less iron accumulation than regular (simple) transfusions.

Before any transfusion, the donor blood as in every case of transfusion is fully matched with that of the recipient, taking all precautions to eliminate contaminants. The blood for transfusion as for β-thalassaemia major, is filtered and leukodepleted and pre-warmed to body temperature.

In an exchange transfusion, the patient's blood is withdrawn 5-20ml at a time (according to the patient's size), discarded and replaced by donor blood. In sickle cell anaemia this means that sickled cells will be replaced by healthy red cells, which have a normal level of haemoglobin, thus correcting the anaemia. In addition, the increase in haemoglobin will suppress the bone marrow's production of more sickle cells.

The exchange may be carried out through a single vein using a three-way tap, with manual removal and replacement. Another technique is to remove whole blood from one arm and at the same time to transfuse the donor blood through the other arm, at the same rate of flow. Automated systems for exchange transfusions are also available.

The total volume of blood exchanged depends on the patient's weight and haematocrit. In children the volume is 50-60ml/kg. In adults 6-8 units of blood are usually needed.
All transfusions carry the risk of transmission of bacterial or viral agents and more frequent than in β-thalassaemia major the risk of allo-immunisation. In the case of exchange transfusions, there are a number of additional risks that must be considered:

- Volume overload, if the amount transfused is greater than the amount removed. This is especially dangerous in patients with pre-existing heart complications, in whom heart failure can easily be precipitated.
- Conversely, inadequate replacement of blood may lead to hypotension and shock. Increasing the haemoglobin level above 12g/dl may lead to hyperviscocity, which may precipitate a vaso-occlusive crisis and stroke.
- Blood clots may disturb the acid base balance. Hypoglycaemia and other metabolic disturbances are also a danger, mainly in children.

Complications of Transfusion Therapy

While transfusion therapy provides considerable clinical advantages, it also presents a number of challenges, some of which are greater in patients with SCD than other populations:

- Volume overload - can result in congestive heart failure and pulmonary oedema in patients with cardiac dysfunction.
- Iron overload - iron loading and subsequent accumulation is potentially toxic and debilitating, unless effective iron chelation therapy is administered.
- Alloimmunisation and delayed haemolytic transfusion reactions - 20-30% of patients with SCD who receive transfusion therapy become alloimmunised. The delayed transfusion reaction occurs 5-20 days after transfusion and can cause severe anaemia, painful crisis or death.
- Viral infection - hepatitis and other viral infections are particularly problematic in patients with SCD, due to pre-existing organ damage. Bacterial infections are rare.

Below is a table that provides recommendations based on UK, NHS on the protocol to be used for assessing and monitoring iron overload.

IRON OVERLOAD ASSESSMENT AND MONITORING PROTOCOL				
CONDITION	S. FERRITIN	MYOCARDIAL T2*MRI	FERRISCAN Measures Liver Iron Content (LIC)	LIVER
SCD on regular top-up/exchange transfusion	Before staring transfusion and then every 3 months	Every 5 years after starting transfusion	1 year after staring transfusion then annually unless 20mg/g dw when every 6 months	Only if indicated for histology or if laparotomy

TREATMENT OF LEG ULCERS

It is important to treat ulcers at any early stage, when they are small and not infected. This treatment is difficult and entails good compliance from the patient and family. It consists of: leg-elevation, cleaning and covering the ulcer, wearing comfortable flat shoes and clean white cotton socks until the ulcer heals. If the ulcer is surrounded by red, painful skin and there is pus oozing from it or its surroundings, indicating that it is infected, you must consult a doctor for antibiotics. If in 2 or 3 weeks after the above treatment the ulcer is getting larger or has not shown signs of healing, hospitalisation is required for intensive ulcer care, strict bed rest and transfusion. Transfusion brings more oxygen to the tissues and may aid healing. If, despite this, treatment the ulcer still doesn't heal, then a skin graft will be used to cover the ulcer. Sometimes, more than one skin graft is needed. Care of ankle ulcerations in haemoglobinopathy patients can be divided into topical measures, in which the nurse plays an active role, and systemic measures, which depend on medical decisions and are related to the failure of local measures to heal the ulcer over time.

- Dressings are usually applied to keep the ulcer clean and allow healing. The affected area is cleaned with warm water or normal saline, removing dead tissue (debridement) and then applying a dressing. There are several types of dressing but there does not seem to be an advantage of one over the others. A simple non-sticky dressing is sufficient for most cases. Dressings are changed weekly.
- Elastic compression bandages may help with oedema and venous congestion.
  If there are signs of infection such as pus, then antibiotic ointments will help. Occasionally systemic antibiotics are necessary.
- Surgical interventions may be necessary, such as debridement and autologous skin grafting.

ADDITIONAL ROLES OF THE NURSE

Other aspects of the care of SCD patients in which the nurse may play a significant role are quite similar to those of the care of thalassaemia patients and include:

- Integration into society
- Issues related to school/university
- Pregnancy

Integration into society

Adolescents and adults with sickle cell disease may experience difficulties integrating with their peers and wider society, sensing a feeling of rejection and marginalisation. Instilling self-confidence and independence in patients is extremely important. Support groups are a very effective tool for enhancing patients' life-skills. Programmes aimed at educating society about sickle cell disease and the rights of affected people to a happy, healthy and fulfilled life are also important. Patients with sickle cell disease often have fears of dying, while their parents live in constant fear of losing their child. The nurse should encourage patients and parents to share these fears with a healthcare provider, a social worker or a close friend. The message must always be that talking can help patients and parents to contain and live with their fears, so that death does not dominate the life of parents or child.

Issues related to school/university

Children and adolescents with sickle cell disease may often be absent from school/university days due to recurrent health problems. This, combined with poor self-esteem and difficulty coping, can result in poor academic achievement and a further sense of depression and hopelessness, particularly in adolescence.

The nurse can support the patient and/or parents, ensuring that teachers are fully informed about sickle cell disease and patient needs, and encouraging open discussion between patient and parents about difficulties faced at school/university. It is important to support parents in insisting that their child stays in school, and setting goals and plans for a fulfilling career in the future. At the same time, it may also be necessary to consult a specialist for a possible learning disorder.

Pregnancy

Women with sickle cell disease can get pregnant and deliver healthy babies. However, it is important to ensure that a patient with SCD visits a genetic counsellor, who will clarify the chances of having a child with sickle cell disease and the various options available for having healthy children.

Prior to pregnancy and during pregnancy and labour, a pregnant woman with SCD will need close monitoring to minimise and prevent complications for both her and her baby. Early and regular prenatal care is important. Routine pregnancy care includes a healthy diet, vitamin and folic acid supplements, increased fluid intake, stopping alcohol, smoking and medicines that can be harmful for the baby, in addition to foetal growth and heart rate testing.

A fall in oxygen saturation is common in any woman in labour. If the mother has sickle cell disease this can precipitate a vaso-occlusive crisis, thromboembolism and acute chest syndrome. Apart from close monitoring of oxygen saturation there should be readiness to resuscitate and provide intensive care.

During labour, intravenous (IV) fluids, oxygen and close foetal monitoring are needed. Close monitoring by a team of medical specialists, including an obstetrician trained in high risk and sickle cell disease pregnancies, can help early detection and treatment of complications, resulting in a better pregnancy outcome. Complications for the mother include high blood pressure, urinary and lung infections, gallbladder problems, heart failure. Other risks include poor baby growth, premature birth, miscarriage or newborn death. Blood transfusion, which helps the blood carry more oxygen, is indicated for hypertension, severe anaemia, increased frequency of pain crisis and previous foetal loss.