NHS
Ealing Community Partners

Management of Sickle Cell Disease

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NATIONAL STANDARDS

- **All babies born in the UK are offered testing via the Newborn Screening Programme**
  - Blood test between 5-7 days after birth
- **Positive Result**
  - Carrier status –parents informed by 6 weeks old
  - Confirmed SCD- parents are informed by 28 days old of the result
- **Registered in local hospital by 8 weeks old**
- **Seen in clinic by 12 weeks old**

RECOMMENDED MEDICATIONS & IMMUNISATION

- **Penicillin Prophylaxis**
  - Infant prescribed Penicillin V (or alternative) by 3 months old
  - Prophylactic Penicillin given BD
- **Folic Acid Daily**
- **Pneumococcal Immunisation**
  - Commenced at age 2 years and every 5 years thereafter
- **Hepatitis B**

PAIN MANAGEMENT

- **Depending on location & severity**
- **Home Medications**
  - **Adult**-Codeine Phosphate, Naproxen, Co-codamol
  - **Children**-Paracetamol, Ibuprofen
- **Hospital Admission**
  - If pain cannot be controlled at home
- **Psychological Interventions**
  - Cognitive behavioural therapy
- **Complementary Therapies**
  - Tens machine, Massage, warm bath

PREVENTION OF ILL HEALTH

- **Education of Parents / Patients**
  - Recognising signs and symptoms of sickle cell crisis
  - Knowing when to seek medical help
- **Health & Lifestyle**
  - Try to prevent Infection
  - Smoking
  - Alcohol consumption
  - Prevention of dehydration
  - Avoid Stress
  - Appropriate clothing

Social & Psychological-1

- **School**
  - Education of teachers/welfare officers
  - Providing individual school healthcare plan
  - Choosing suitable career
- **Travel**
  - Immunisations
  - Insurance
- **Relationship & Marriage**
  - Preconception counselling
  - Contraception & Family Planning
  - Prevention of infection

SOCIAL & PSYCHOLOGICAL-2

- **Referrals**
  - Social Care
  - Housing department
  - Voluntary organisations
  - Patient support groups

SPECIALIST THERAPIES

- **Hydroxycarbamide**
  - Recommended for all patients including infants to reduce sickle cell crisis and organ damage
- **Blood transfusion**
  - Strokes
  - Recurrent splenic sequestration
  - Chronic complications e.g. cardiac, respiratory
  - Failed hydroxycarbamide or refused
- **Bone Marrow Transplantation**
  - Stem cells
  - Not yet standard treatment for adults

CLASSIFICATION OF THALASSAEMIA

- **Beta Thalassaemia Trait (Carrier/Minor)**
  - No treatment required
- **Thalassaemia Intermedia**
  - May require blood transfusion
- **Beta Thalassaemia Major**
  - Blood transfusion dependant

SIGNS & SYMPTOMS OF THALASSAEMIA MAJOR

- Severe Anaemia
- Hb 20-60g/l around 6/12 months old
- Failure to thrive
- Skeletal Changes
- Prone to Infection

TREATMENT FOR THALASSAEMIA MAJOR

- **Regular Blood Transfusions** to:
  - Correct anaemia
  - Enable growth and normal activity
  - Prevent enlargement of the spleen and inhibit the erythroid marrow expansion
  - Iron chelation when ferritin levels are >1000

COMPLICATIONS OF IRON OVERLOAD

- Delayed growth/ puberty
- Liver damage
- Diabetes
- Cardiac impairment/failure
- Erectile dysfunction
- Amenorrhoea

COVID-19 RECOVERY PLAN

- **From 1st August Shielding Paused**
- **Advice is to:**
  - Keep 2 metres away, if not possible wear face mask
  - Can go to work, as long as the workplace is COVID-secure but carry on working from home if possible
  - Children can go back to school
  - Can go outside to buy food, to places of worship and for exercise
  - To follow all government recommendations