

MANAGEMENT OF CHILDREN WITH SYSTEMIC DISEASES

1. Cardiovascular disorders
2. Disorders of the blood.
 - a. Disorders of homeostasis
 - b. Hematological disorders.
3. Respiratory system disease.
4. Convulsive disorders.
5. Metabolic and endocrine.
6. Hepatic disorders.
7. Renal disorders.

1. Cardiovascular disorders.

a. Congenital Heart Disease

Congenital heart disease is due to structural abnormalities of the heart. The cause is rarely known in individual cases but multi-factorial inheritance patterns are mainly responsible.

The main type of congenital heart disease are:

- * Ventricular septal defect
- * Atrial septal defect
- * Pulmonary stenosis
- * Patent ductus arteriosus
- * Tetralogy of fallot
- * Aortic stenosis
- * Coarctation of the aorta
- * Transposition of great arteries
- * Rare / diverse

Acquired Heart Disease

* **Rheumatic heart disease.**

It is the most significant pathologic sequela of rheumatic fever and may affect mitral, aortic, tricuspid and pulmonary valves.

* **Infective bacterial endocarditis**

Microbial infection of the heart valves or endocardium.

Acute: caused by staphylococcus aureus, group A streptococci and pneumococcus. The microorganisms are of high pathogenicity attacking a normal heart, causing erosive destruction of the valves.

Sub-acute: caused by viridians streptococci usually develops in persons with pre-existing congenital cardiac disease or rheumatic valvular lesion.

Dental care for children with cardiovascular disorders.

- * Prevention of dental disease (dietary, fluoride therapy, fissure sealant, and oral hygiene instruction).
- * Regular monitoring, both clinically and radiographically.
- * Active dental disease should be treated before cardiac surgery is undertaken.
- * In case of an emergency dental situation or if the physician cannot be reached, it is prudent to use antibiotic coverage.
- * Pulp therapy for primary teeth is not recommended, because of the high incidence of associated chronic infection.
- * Endodontic therapy in permanent dentition is not to be performed except in selected cases and if adequately performed.

Antibiotic Prophylaxis

There is still some controversy over which conditions to do or do not require prophylactic antibiotic therapy.

- * If any doubt exists then the paediatrician or cardiologist should be consulted before invasive dental procedures are undertaken.
- * The dentist should discuss the treatment with the child's physician or cardiologist, and a written statement identifying the nature of the disease and need for antibiotic therapy and type of premedication if any should be obtained and placed in dental record.

2. Disorders of the Blood

a. Disorders of homeostasis

Hemophilia, Thrombocytopenia and Van Willebran's disease.

*** Hemophilia**

It is congenital blood coagulation disorder inherited as an X linked recessive (affected males, female are carriers).

*** Thrombocytopenia**

This is caused by a reduction in the number of circulating platelets in the blood stream.

Normal levels are between 150 and 400 x 10⁹ per liter. The platelet count should be at least 50 x 10⁹ per liter before surgery is attempted.

*** Van Willebran's Disease**

This is dominantly inherited, complex, and variable condition characterized by a vascular abnormality of large irregular capillaries, defective platelets which do not adhere to each other, and decreased levels of factor VII.

Common clinical manifestations are nosebleeds and spontaneous gingival hemorrhage (1:1000)

Classification of Bleeding Disorder

1. Coagulation defects

a. Inherited

- i. Hemophilia A: Factor VIII deficiency.
- ii. Hemophilia B: Factor IX deficiency (Christmas disease)

b. Acquired

- i. Liver disease
- ii. Vitamin deficiency
- iii. Anti-coagulant drugs (heparin, warfarin).
- iv. Disseminated intravascular coagulation (DIC).

Thrombocytopenic purpuras

a. Primary

- i. Idiopathic thrombocytopenic purpura.
- ii. Pancytopenia, fanconi syndrome.

b. Secondary: Systemic disease – Leukemia

- i. Drug induced
- ii. Physical agents radiation

Non-thrombocytopenic purpuras

a. Vascular Wall alteration

- i. Scurvy
- ii. Infections
- iii. Allergy

b. Disorders of platelet function

- i. Inherited Van Willebrand's disease.
- ii. Drug, aspirin, non-steroidal anti-inflammatories, alcohol, penicillin.
- iii. Allergy
- iv. Auto-immune disease
- v. Uraemia

Dental management of bleeding disorders.

- * A good history is the best screening device.
- * Effective communication with the child's physician or hematologist is important.
- * Strict preventive dentistry program for hemophiliac patients to minimize the needs for hazards of restorative treatment or tooth extraction.
- * Local anaesthetic is contraindicated to hemophiliac children except when pain is extreme (infiltration or intra ligamentous injections).
- * Regional anaesthesia, such as an inferior dental block, is contraindicated as bleeding in the pterygomandibular region may result in asphyxia.
- * Pulp treatment of primary molar teeth may be required to avoid extractions.
- * If dental extraction or surgery does become necessary then the patients are usually best managed in the hospital situation.

b. Hematological Disorders

There are several relatively common disorders of the red and white blood cells that may influence dental care in the child.

1. Red Blood cell disorders

a. Anemia

- i. Iron deficiency
- ii. Glucose 6-phosphate dehydrogenate deficiency
- iii. Sickle cell
- iv. Thalassaemia

b. Polcythaemia

2. White Blood cell disorders

a. Leucocytosis

- i. Infectious mononucleosis (glandular fever).
- ii. Neplasia

b. Heucopenia: neutropenia

- i. Congenital\
- ii. Drug induced

c. Leukemia's

- i. Acute lymphocytic (ALL)
- ii. Acute myeloid (AML)
- iii. Chronic

d. Lymphomas

- i. Hodgkins's
- ii. Non-Hodgkin's
- iii. Burkett's

- **Sickle-Cell Anemia**

This is an inherited autosomal-recessive disorder that results in the substitution of a single amino acid in the hemoglobin chain.

The red blood cells containing hemoglobin S have a life of only 30-60 days and become clumped together under certain conditions, thus blocking small blood vessels and leading to pain and necrosis.

- *** Thalassaemia**

This is an inherited disorder of hemoglobin synthesis and may occur as a heterozygous trait or homozygous thalassaemia major. It results in a severe progressive haemolytic anemia.

- *** Leukemia**

Leukemia is a malignant proliferation of white blood cells. The general clinical features of all types of leukemia are similar as all involve a severe disruption of bone-marrow functions.

Dental Management of Anemia.

- All anemic children have a greater tendency to bleed after invasive dental procedures.
- The hemoglobin level and haematocrit tests used for screening and a white blood cell and platelet count should also be obtained.
- A family history of conditions such as sickle-cell anemia and thalassaemia is significant and all black patients should be tested routinely for sickle-cell disease prior to a general anaesthetic.
- General anaesthetics should be avoided in preference to the use of local anesthesia for sickle-cell anemic patient.

Dental Management of Leukemia

- For a Leukemia Patient – Unless there is a major dental emergency no active dental treatment should be carried out until the child is in remission.
- Any dental pain should be treated conservatively by the use of antibiotics and analgesics.
- Current haematological information is required to assess bleeding risks.
- Prophylactic antibiotic therapy may be used in some case.

- Regional block anaesthesia may be contraindicated due to the risk of deep hemorrhage

3. Respiratory System Disease

Asthma – is a diffuse obstructive lung disease which causes breathlessness, coughing, and wheezing it is associated with hyper-reactivity of the airways to a variety of stimuli and a high degree of reversibility of the obstructive process.

Dental Management of Asthma.

- Dental treatment itself can cause emotional stress which may precipitate an attack.
- General anaesthesia for severe asthmatics usually requires in-patient hospital admission.

4. Convulsive Disorders

Epilepsy – it is a symptom that is characterized by recurrent seizures caused by abnormal neuronal discharge in the brain, unknown origin (idiopathic epilepsy) or due to congenital or acquired brain lesions (secondary epilepsy).

Dental Management of Epilepsy.

- The dentist should contact the child's physician if the child is taking seizure control medication.
- Care should be taken to avoid inducing seizures e.g. anxiety intense light and intravascular local anaesthesia are seizure triggers.
- The child with good control of seizures needs a minimum of restrictions, although the possibility of attack occurring in the dental chair should be considered.
- A very high standard of oral hygiene is required.
- **If the seizure occur during dental treatment:**
 - A mouth prop, consisting of tongue blades wrapped in gauze and heartily taped can be used to prop open the mouth and prevent tongue biting.
 - Place the patient on the floor away from any instruments or glass to avoid hurting himself during the seizure.

- The patient should be turned to one side to keep an open airways and allow secretions to drain.

5. Metabolic and Endocrine Disorders

Diabetes Mellitus

Type I (insulin dependent) – mostly in children.

Type II (non-insulin dependent) – after 40 years of age.

Type III (secondary diabetes).

Diabetes

Is due to the deficiency of insulin and abnormal metabolism of carbohydrate.

Type I. Juvenile Diabetes

It is insulin-dependent and usually of juvenile onset. It is highly age related with peaks of presentation between 5 and 7 years and puberty.

Dental Management of Diabetes.

- The well-controlled diabetic child with no serious complications can have any dental treatment but should receive preventive care as a priority.
- Dental appointment should be arranged at time when the blood sugar levels are well controlled (morning appointments are better).
- General anaesthetics are a problem because of pre-anaesthetic fasting that is required.
- Strict oral hygiene.

6. Hepatic Disorders

Viral hepatitis is a common infection that produces inflammation of liver cells, which may lead to necrosis.

- Hepatitis A virus infection.
- Hepatitis B virus infection.
- Hepatitis D virus infection.
- Non-A, non B (Hepatitis C) – parenterally transmitted.
- Non A, non B (Hepatitis E) – enterically transmitted.
- **Potential problem related to dental care**
- **Hepatitis infection control protocol.**
 - It is important to liaise directly with the patient's physician in the first instance when planning dental treatment.

7. Renal Disorders

End-stage renal failure leads to a drop in glomerular filtration rate which results in progressive hypertension, fluid retention and build-up of metabolites that are not excreted normally.

Conditions Affecting the Kidneys.

- Ureteric reflux causing reflux nephropathy or hypoplasia.
- Obstructive uropathy.
- Glomerulonephritis / glomerulosclerosis.
- Medullary cystic disease.
- Systemic lupus erythematosus.
- Cystinosis

Dental Management of Renal Disorders.

- Consultation with renal physician.
- Prevention, this is of great importance before transplantation.
- Extraction of pulpally involved teeth. Treatment should be aggressive.
- Admission is required for general anaesthesia.