HEMOLYTIC UREMIC SYNDROME

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INTRODUCTION

Hemolytic Uremic Syndrome, a disease that destroys red blood cells, is the most common cause of sudden, short term acute renal failure in children. This disease occurs primarily in infants and smaller children between the ages of 6 month and 5 years. The syndrome usually occurs in previously healthy child and often preceeded by gastrointestinal enteritis.

DEFINITION

HUS is defined as combination of symptoms which is characterised by haemolytic anemia with variable degree of thrombocytopenia and renal failure.

CLASSIFICATION

Hemolytic uremic syndrome is mainly classified as

- D⁺ Hemolytic Uremic Syndrome
  - D⁺HUS is the classic form, accounting for 95% of cases in children. The epidemic type or prodromal type is more common and is accompanied by an enteritis.
- D⁻ Hemolytic Uremic Syndrome
  - D⁻ is account for remaining 5% of cases of HUS. This is a sporadic or non prodromal type and is not accompanied by an enteritis.

INCIDENCE

- The overall incidence of D⁺ HUS is estimated to be approximately 2.1 cases per 100,000 persons per year, with a peak incidence in children who are younger than 5 years (6.1 cases per 100,000 per year).
- Incidence tends to parallel the seasonal fluctuation of E coli infection, which peaks between June and September.
- Incidence of D⁻ HUS in children is approximately 2 cases per year per 100,000 total population.

ETIOLOGICAL FACTORS

- D⁺ HUS Type
  - This type of HUS is mainly caused by bacteria such as Ecoli, shigalle, pneumococci and salmonelle.
  - Rarely is caused by viruses such as adenoviruses and echoviruses.

These infectious agents are acquired by the:

- Consumption of raw or undercooked meat especially ground beef
- Un pasteurized milk or fruit juice, especially apple.
- Drinking or swimming in contaminated water
- Person to person transmission

**D-HUS Type**
- Ideopathic
- Causative organism is streptococcus pneumoniae

**Family history haemolytic uremic syndrome**
- Drug toxicity: chemotherapy drugs, immune mediators such as quinines, tilopidine.
- Use oral contraceptives
- Malignancies
- Vascular diseases such as varicosities, DVT
- Connective tissue diseases eg. SLE, systemic sclerosis

**PATHOPHYSIOLOGY**

Due to the etiological factors

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Invasion of micro organism into the GI tract

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Toxins are released from the microorganism mainly from Ecoli and shigelle

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The toxin bind and destroy the colonic epithelial mucosal cells resulting in bloody diarrhoea.

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Toxin is enter into the blood stream

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Toxin is attach to the endothelial lining of the small glomerular arterioles

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Glomerular arterioles become injured, swollen and occluded with platelet deposits and fibrin clot

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The circulating RBC cells are forced through the occluded arterioles
Deformed and fragmentation of red blood cells

Anemia and thrombocytopenia

CLINICAL MANIFESTATION

- Abdominal pain
- Diarrhoea
- Fever
- Vomiting
- Irritability
- Marked pallor

➢ Haematological manifestations
  - With in 5-7 days patient exhibits the signs of anemia
  - Thrombocytopenia
  - Bruising, bleeding from nose or mouth
  - Petechiae
  - Bloody diarrhoea

➢ Renal manifestation

More than half of the children with haemolytic uremic syndrome develop acute kidney failure.

- Hematuria
- Oliguria or anuria
- Proteinuria
- Hypertension
- Generalized edema: edema in face, hands, and feet
- Metabolic acidosis

➢ CNS Manifestation
  - Irritability
  - Extreme fatigue
  - Hemiparesis
  - Seizure
  - Stupor or coma

➢ Other manifestation
  - Hyperbilirubinemia
  - Spleenomegaly
  - Pancreatic insufficiency which can lead to diabetes mellitus
  - Myocarditis, cardiomyopathy
  - Death

DIAGNOSTIC MEASURES

❖ History
❖ Physical examination
Blood investigation such as
- Haemoglobin level: it should less than 8g/dl
- Hamatocrite: it is less than 24%
- CBC including total platelet count
- Coagulation test including BT, CT, PPT
- Blood urea nitrogen
- Serum creatinine
- Serum electrolytes
- Serum bilirubin

Urine analysis
Renal biopsy helps to determine the chronic injury
Stool culture typically detects the Ecoli

THERAPEUTIC MANAGEMENT
The goal of therapy are early diagnosis and supportive care of ARF and haemolytic anemia. Their is no specific management. only symptomatic management.

1. DRUG THERAPY
   - Administration of antiplatelet drugs, intravenous immunoglobulin and anticoagulants
   - Diuretic therapy
     - calcium channel blockers to maintain the normal blood pressure
     - Administration of alkalicyc agents such as sodium bicarbonate or combination of sodium and potassium citrate to correct the metabolic acidosis.

2. FLUID MANAGEMENT
   It is depend on the patient current hydration status. In case of diarrhoeal diseases fluid resuscitation is necessary, although one must avoid fluid overload.

3. DIALYSIS
   The most constantly effective treatment of HUS is hemodialysis or peritoneal dialysis.
   - Indication for dialysis
     - Fluid overload resistant to diuretic therapy
     - hyperkalemia
     - symptoms of uremia
     - intractable acidosis

4. Blood transfusion with fresh, washed packed cell are administered for severe anemia
5. Pasmapherisis is benefit in in case of A tupical HUS.
6. Nutritional management
   - Administer high calorie with minimum protein containing diet. Salt containing diet should be restricted.

7. Avoid unnecessary use of antibiotics and antimotility drugs.

8. Kidney transplantation: kidney transplantation is new acceptable and effective means of therapy in paediatric age group. Kidney for transplant are available from two sources, a living related donor and cadaver donor
NURSING MANAGEMENT

- Assess the general condition of the child
- Check the vital signs
- Reassure the child
- Check the fluid status and electrolyte status
- Maintain the intake and output chart
- Nutritional management: high calorie, low sodium and low protein diet.
- Maintain the optimal thermal environment
- Prevent injury
- Provide proper skin care and hygienic care.
- Blood transfusion, if the child is having severe anemia
- Emotional support and education to the child and family.

NURSING DIAGNOSIS

- Hyperthermia related to inflammatory process.
- Ineffective tissue perfusion related to sodium and water retention
- Fluid volume excess related to accumulation of fluid in the tissues.
- Imbalanced nutrition less than body requirement related to restricted diet.
- Risk for injury related accumulated electrolytes and low platelet count.

COMPLICATION

- Chronic renal failure
- Hypertension
- Central nervous system disorders
- Coma
- Death

PROGNOSIS

With prompt treatment recovery rate is about 95%, but residual renal impairment is ranges from 10-50%. The overall prognosis for D+ HUS is better than for D- HUS. The early mortality rate, incidence of ESRD, frequency of renal failure and hypertension are higher in this group.

BIBLIOGRAPHY