

# Wiskott Aldrich Syndrome - A Case Report

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**Abstract:** Wiskott Aldrich Syndrome (WAS) is an immunological disorder caused by a genetic abnormality (Brunner, 2017). WAS is a rare genetic disorder of the immune system that primarily affects boys. It is characterized by abnormal immune function and a reduced ability to form blood clots. It is one of the primary immune deficiency disorders (NIAID, 2015) which results in prolonged bleeding and recurrent infections. The platelet abnormality is typically present from birth and can lead to easy bruising or episodes of prolonged bleeding following minor trauma. Early correction with hematopoietic stem cell transplantation or gene therapy is necessary to prevent the risk of Severe Combined Immunodeficiency (SCID), autoimmune complications and Haematological malignancies. This article glimpses a case scenario with WAS disease and the medical and nursing management carried out in a tertiary care hospital in southern India which is one of the pioneer hospitals for Haematopoietic stem cell transplantation in India.

**Keywords:** genetic, bleeding, infection, hematopoietic stem cell transplantation

## Wiskott-Aldrich syndrome:

Wiskott-Aldrich syndrome (WAS) is an X-linked autosomal recessive disorder characterized by the clinical triad of microthrombocytopenia, eczema, and recurrent infections (e-medicine, 2019). Basically, the problem is with B and T cells and platelets resulting in prolonged episodes of bleeding, recurrent bacterial and fungal infections and increased risk of cancer and autoimmune disease (Hinkle, 2018).

## Background:

WAS was named after two paediatricians who described this disease. In 1937, Alfred Wiskott, a German paediatrician, first described three brothers who died before the age of 2 years with chronic bloody diarrhea, eczema and recurrent ear infections. In 1954, Robert Aldrich, an American pediatrician, reported Dutch kindred of boys who all died of similar clinical symptoms described by Wiskott, clearly demonstrating an X-linked mode of inheritance. Forty years later, the gene responsible for WAS was identified on the short arm of the X chromosome (Xp11.22-p11.23) by linkage analysis.

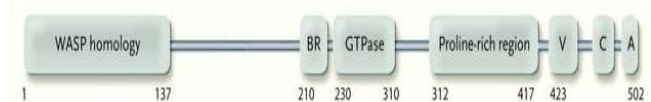
## Epidemiology:

In United States, the incidence of WAS is 1 in 1 million cases per live birth. WAS is found to be 3% of all primary immunodeficiency disorders. According to the National registries in Ireland, Italy, Japan, Switzerland, Sweden 4.1 cases per 1 million live births and occurs 2-8.8% of patients with primary immunodeficiencies (Huynh, Fscience 2019). In India, according to the 'Foundation for research on rare diseases and disorders' statistics in 2010, WAS is 0.2 per 1 million live births and 2400 population were affected with WAS.

## Genetics:

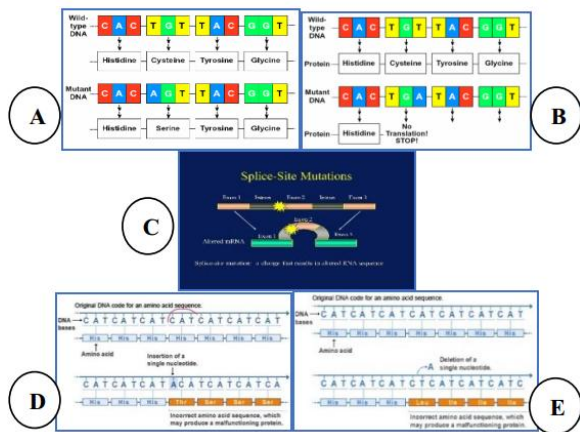
Depending on the mutations within the Wiskott-Aldrich Syndrome Protein (WASp) gene, there are various clinical disease. Alfred Wiskott described deletion of two nucleotides (AC73-74del) of the WAS gene as original family.

The gene product WASp is a 502 amino acid protein expressed within the cytoplasm of non-erythroid hematopoietic cells.

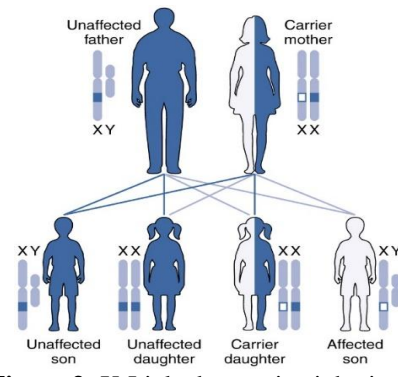


**Figure1:** WAS protein structure

WAS gene provides instructions for production of a protein called WASp. This protein plays an essential role in relaying signals from the surface of the blood cell to the cell's actin cytoskeleton, the network of fibers that make up the cell's structural framework. In T-cells, WASp is important because it is known to be activated via T cell receptors signalling pathways to induce cortical actin cytoskeleton rearrangements that are responsible for forming the immunological synapse. More than 300 unique mutations in the WAS gene have been identified. The most common mutations are missense mutations (change of a single base pair causes the substitution of a different amino acid in the resulting protein, render the protein non-functional), followed by nonsense mutations (substitution of a single base pair that leads to the appearance of a stop codon where previously there was a codon specifying an amino acid.), splice-site mutations (a genetic mutation that inserts, deletes or changes a number of nucleotides in the specific site at which splicing takes place during the maturation of messenger RNA) and frameshift mutation (a type of mutation involving the insertion or deletion of a nucleotide in which the number of deleted base pairs is not 'divisible by three'. "Divisible by three" means the cell reads a gene in groups of three bases. Each group of three bases corresponds to one of 20 different amino acids used to build a protein. If a mutation disrupts this reading frame, then the entire DNA sequence following the mutation will be read incorrectly).



**Figure 2:** Missense mutation (A), Nonsense mutation (B), Splice-site mutation (C), Frameshift mutation (D & E) (insertion and deletion)



**Figure 3:** X-Linked recessive inheritance

**Clinical manifestation:**

The classical features are low platelets, immunodeficiency and eczema. The defective platelets are removed from circulation by the spleen or liver, leading to low platelet counts. And immune deficiency has been linked to decreased antibody production and the inability of immune T cell to effectively combat infection.

- Bleeding: A reduction in the size and number of platelets is a hallmark of WAS. Bleeding into the skin may cause petechiae ranging in size from small pinheads to large bruises.

Defective gene in WAS shows;

- Paucity of microvillus on the surface of T-cells because of defective cytoskeleton organization
- Abnormality in the glycosylation of the cell surface
- Defective signaling
- Loss of T cell proliferation to antigenic stimulation

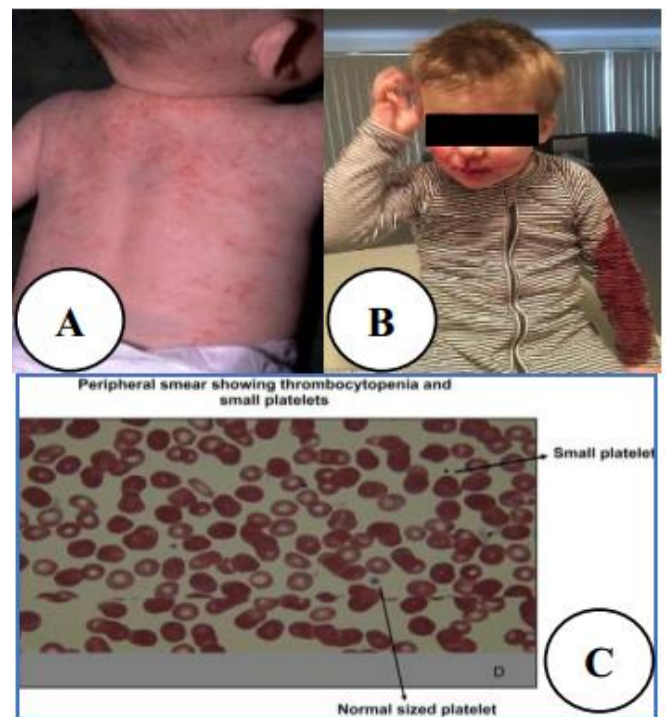
WASp gene is seen in spleen, thymus and lymphocytes. Cluster of differentiation 43 (CD43) or sialophorin absence is also linked to WAS. CD43 expressed in monocytes and lymphocytes are responsible for intracellular signalling and activation of the cell.

Generally, WAS gene mutations cause absent protein expression result in classic WAS. Reduced WAS protein expression results in X-linked thrombocytopenia (XLT). WASp activating gain-of-function mutations result in X-linked neutropenia (XLN).

**Inheritance:**

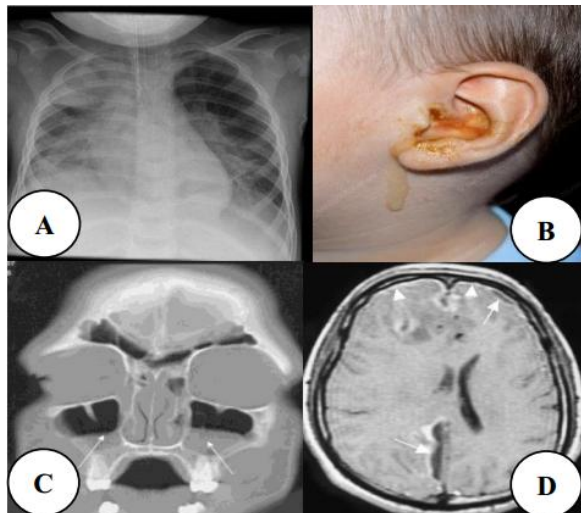
Wiskott-Aldrich syndrome follows an X-linked inheritance pattern. Each person has 23 pairs of chromosomes—one pair of sex chromosomes (XX for female and XY for male) and 22 pairs of numbered chromosomes, called autosomes. WAS gene is located on the X chromosome. Boys who inherit a disease-causing gene on their X chromosome are affected by the disease. Girls who inherit an X-linked mutation are “carriers”—meaning that they have one mutated copy of the gene and one normal copy.

All children of a carrier mother have a 50 percent chance of inheriting the mutation. This means that about half of the boys will be affected by the disease and about half of the girls will be carriers. Female carriers of a mutated WAS gene do not experience symptoms of the disease. This means that boys with Wiskott-Aldrich syndrome do not have other affected female relatives, but they may have brothers or male relatives on their mother’s side, such as uncles, who also have the disease. The disease cannot be passed from father to son.



**Figure 4:** Petechiae (A), prolonged obvious bleeding (B) and small platelets (C)

- Infections: Ear infections, sinus infections and pneumonia are common in WAS due to the deficiency of both B and T lymphocyte function. More severe infections of the bloodstream may also occur, as well as meningitis or severe viral infections.



**Figure 5:** Pneumonia (A), ear infection (B), sinusitis (C) and meningitis (D)

- Eczema: People with classic WAS frequently suffer from eczema. In infants, the eczema may resemble “cradle cap” or severe diaper rash; in older boys, it may be limited to the skin creases around the front of the elbow, around the wrist and neck, and behind the knee.



**Figure 6:** Skin lesions

- Autoimmune manifestations: A high incidence of “autoimmune-like” symptoms are common in both infants and adults with WAS. The most common of these are vasculitis, hemolytic anemia and immune thrombocytopenia purpura (ITP). Autoimmunity in WAS is due to the formation of autoantibodies or the presence of autoreactive T-cell clones.
- Malignancies: Cancer such as non-Hodgkin’s lymphoma or leukemia that involve B cells can occur more frequently in patients with WAS.

#### Classifications:

There are four types;

1. Classic WAS (most severe form)
2. X-linked thrombocytopenia
3. Intermittent thrombocytopenia
4. X-linked neutropenia

#### Diagnosis:

Evaluation of the signs of bleeding, infection, malignancy, general appearance, vital signs, height and weight, growth parameters, head and neck, dermatologic, pulmonary and neurologic functions.

#### Laboratory tests:

- Blood: Complete blood counts, Culture and sensitivity, Renal function test, Liver function test – to support the diagnosis
- Immunological skin test - to reveal hyposensitivity
- Quantitative serum immunoglobulin levels - IgM levels are low, IgA levels are elevated, and IgE levels may be elevated
- Specific antibody titres
- Genetic testing – decreased WASp, defect in the CD43 molecule
- DNA sequence analysis that detects WAS and the related disorders XLT and XLN
- Major histocompatibility tests of the patient, parents, and siblings to determine feasibility for stem cell transplantation
- Screening of patient and potential donor for infectious agents (e.g., HIV, CMV, hepatitis viruses)
- Bone marrow biopsy to rule out Leukaemia
- Imaging studies: Chest x-ray, CT and MRI studies as pre stem cell transplantation work up.

#### Management:

The **definitive treatment** is **Haematopoietic stem cell transplantation**.

**Gene therapy** using genetically modified Lentivirus is showing promising results recently.

#### Pharmacological managements are;

- Antibiotics
- Inhaled bronchodilators (eg, albuterol, salmeterol, beclomethasone, fluticasone)
- Hyperimmune globulins (eg, varicella-zoster immune globulin)
- Immunizations (eg, vaccines, including diphtheria and tetanus toxoids [DT or TD], acellular pertussis, conjugated HIB, conjugated pneumococcal vaccine, unconjugated meningococcal A and C, hepatitis B [HBV], influenza)
- Corticosteroids (eg, prednisone, methylprednisolone, flucinolone)
- Immunoglobulins (eg, immune globulin)

a) Surgical management is performed in case of **bleeding complications**.

- Neurosurgery if subdural hematoma
- Surgical evacuation of hematomas
- Surgical intervention to stop blood loss after any minor trauma
- Splenectomy in cases of coexisting severe thrombocytopenia and frequent bleeding

**Supportive cares** are transfusions of platelets, red blood cells, infusions of intravenous IgG

## B. Prevention:

**Primary Prevention:** Pneumococcal vaccine is recommended before splenectomy for all children < 2 years and for unvaccinated children between 24 - 59 months old who are at high risk for pneumococcal infections, if they are not on immunoglobulin therapy. Splenectomy necessitates penicillin prophylaxis for life.

**Secondary Prevention:** Mutation analysis for other family members, either if history suggestive of WAS or to detect carrier status.

## Case study:

Master M is a 10 month old male child born to parents from northern India. His parents had first degree consanguineous marriage and gave birth to his elder brother who died at 3 years of age due to epistaxis and thrombocytopenia. His mother gave no history of abortion in the past. Master M received only zero dose BCG & OPV, 1<sup>st</sup>, 2<sup>nd</sup> and 3<sup>rd</sup> doses of DPT & OPV immunization. His growth and development was appropriate to his age.

He presented with the history of frequent fever, flu like symptoms and petichael and purpuric rashes over the face and back which fades off spontaneously in couple of days. At his 8 months of age, he developed bloody diarrhoea which did not respond to antibiotics. Hence, he was evaluated in Mumbai and was found to have anaemia (Hb 8.8 g%), thrombocytopenia (8000/mm<sup>3</sup>) and leukocytosis (19500/mm<sup>3</sup>). Bone marrow analysis showed adequate megakaryocytes with no evidence of marrow disease. Immunoglobulin analysis showed decreased IgG (338 mg%), IgA (15.2mg%) and IgM (9mg%). He was treated with Inj. IVIG 2.5 g OD x 2 days and following which he was started

on syr. Omnacortil 5mL OD x 2 weeks and then alternate days. He was brought to CMC, Vellore in June 2019 for further evaluation and treatment. Gene analysis was done and he was diagnosed to have Wiskott Aldrich Syndrome with frame shift mutation in the WASp gene. As his diarrhoea was worsening, he was administered one dose of Inj. IVIG 2.5g and Haploidentical peripheral blood stem cell transplantation was done on 09.01.2020, his father was the donor. He was discharged from Bone marrow transplant unit on Day + 37. On Day + 94 after hematopoietic stem cell transplantation, he got admitted in Haematology ward with the complaints of loose stools 5-6 episodes for a day. He was evaluated for gut GvHD and diagnosed to have third degree gut GvHD. Later, his diarrhoea worsened and he was started on Total parenteral nutrition (TPN). He also developed a skin rash which was treated as atopic dermatitis. He was free from all the complications and was discharged on D+175 with 84% chimerism.

## Nursing Care:

### Application of Lydia Hall's theory on Master M.

#### Lydia Hall's theory:

Ms. Lydia Hall was born in New York City in the year 1906, derived a theory which consists of three independent but interconnected circles: the CORE, the CARE and the CURE.

1. **The core** is the person or patient to whom nursing care is directed and needed according to his feelings, and value system.
2. **The care** explains the role of nurses focused on performing noble task of nurturing the patient; meaning helping the patient to meet his needs and attain a sense of balance.
3. **The cure** is the attention given to patients by the medical professionals.

According to Lydia Hall, Nurses' role is more and emphasized in the CARE component

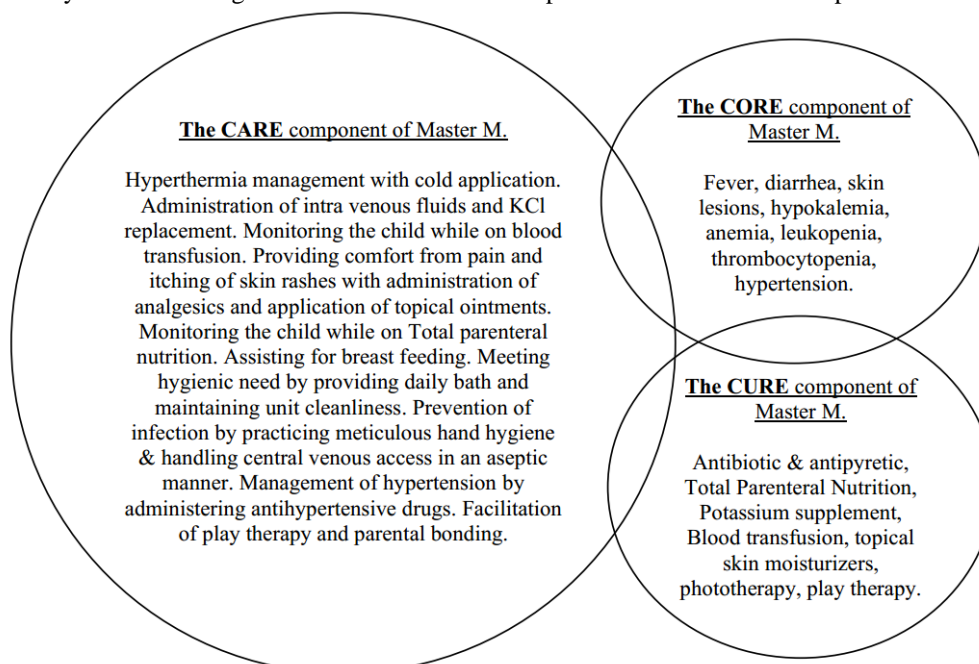


Figure 7: Lydia Hall's theory application on Master M

**Nursing Management:****Pre-stem cell transplantation:**

1. **Nursing diagnosis:** Diarrhoea (bloody) related to inflammation of the bowel and low platelet counts in the circulating blood as evidenced by multiple episodes of loose stools (bloody), low platelet counts and increased WBC counts.

**Goal:** Reduction in frequency of stool and normal stool consistency is attained.

**Nursing intervention:**

- Assessed bowel sounds, abdominal distension, rigidity. His abdomen was soft, no distension and non-tender.
- Monitored stool pattern and volume. He had multiple episodes of bloody diarrhoea per day.
- Assessed hydration status of the child, no signs of dehydration.
- Encouraged breast feeding and adequate oral fluid intake.
- Monitored urine output, it was normal.
- Assessed his perianal skin, was intact and not inflamed.
- Applied Zinc oxide ointment on the perianal region to prevent excoriation.
- Administered IV immunoglobulin 2.5g bd x 2days. His immunoglobulin levels were decreased; IgA was 15.2 mg% (27-66 mg%), 9 mg% (40-143mg%), WBC counts 19500/mm<sup>3</sup>.
- Transfused platelets. His platelet count was 8000/mm<sup>3</sup>.
- Educated his parents on spread and prevention of diarrhoea, importance of hand hygiene and personal hygiene.

**Evaluation:** His diarrhoea stopped after the administration of IV immunoglobulin and he passed stool with normal colour and consistency and twice daily.

2. **Nursing Diagnosis:** Deficient knowledge regarding new condition and treatment related to unfamiliarity of information and complexity of treatment as evidence by his father's verbalization of inaccurate information.

**Goal:** Learning needs on the knowledge of disease process and treatment is met.

**Nursing Intervention:**

- Assessed his parents' understanding of disease, treatment plan, complication and follow-up.
- Instructed about central venous catheter placement and care.
- Explained stem cell mobilization therapy with growth factor for the donor, peripheral blood stem cell collection via apheresis and storage.
- Discussed about conditioning regimen, side effects and measures to alleviate the toxicities such as administration of antiemetics, antipyretics, analgesics.
- Discussed the procedure for peripheral blood stem cell infusion and its complication.

- Explained that it would take up to 21 days for engraftment.
- Explained about the need for blood and platelet transfusion.
- Discussed about the importance of protective environment such as reverse barrier technique, isolation room, HEPA filter positive pressure room.
- Facilitated discussion about prophylactic anti-infective therapy, need for frequent blood sampling.
- Instructed about dietary modification with steamed food and low microbial diet.

**Evaluation:** His parents verbalized understanding of their son's disease, treatments, possible complications and follow up care.

3. **Nursing diagnosis:** Anxiety (parental) related to situational crisis and stress secondary to disclosure of their son's diagnosis, treatment plans and prognosis as evidenced by his father blaming health care system and being hostile.

**Goal:** Parental anxiety is minimized.

**Nursing intervention:**

- Assessed the level of his parents' anxiety. Father was in anger and bargaining stage.
- Acknowledged awareness about the parental anxiety.
- Maintained calmness while interacting with the family, especially with his father.
- Oriented the parents to the disease, treatment plans and prognosis by facilitating family meeting with the treating physician regularly.
- Assisted the parents to develop new anxiety-reducing strategy such as relaxation, deep breathing and positive strokes.
- Encouraged his parents to verbalize anxiety provoking situations/incidents.
- Used simple words in his mother-tongue (Hindi) to communicate any information.

**Evaluation:** Parents expressed reduced anxiety as evidenced by his mother being cheerful and his father communicating in friendly manner with the health care team.

**Post-stem cell transplantation:**

4. **Nursing diagnosis:** Actual infection related to presence of pathogens and poor immune response of the child as evidenced by fever on Day + 142, diarrhoea, increased WBC counts, immune compromised state.

**Goal:** Infection is minimised.

**Nursing Intervention:**

- Monitored vital signs. Body temperature was 100.20F – 103.80F, heart rate was 130/mt. - 144/mt., Respiratory rate was 28/mt. - 34/mt.
- Followed meticulous hand hygiene.
- Maintained strict aseptic technique for all invasive procedures.
- Performed central line dressing regularly.

- Hydrated him adequately with intravenous fluids, oral fluids and total parenteral nutrition.
- Monitored total blood counts and blood culture. WBC was 18000/mm<sup>3</sup>.
- Monitored blood culture and sensitivity of blood, urine and stool as and when necessary, there was no growth except in his urine culture Acinetobacter species was present. He was negative for the presence of Clostridium difficile in his stool.
- Evaluated for viral and fungal infections. Negative for CMV, BK, Serratia, procalcitonin was 0.12ng/mL in the blood.
- Administered Inj. Meropenem 250mg IV Q8h X 7 days, syrup Levofloxacin 75mg OD X 7 days, Inj. Ganciclovir 30mg IV BD (3/7) and Tab. Pentid 2L unit BD X 3 days and then syrup Septran-p 30mg BD (2/7).
- Administered Inj. Grafeel 30mcg S/C OD (2/7) since D+158.
- Educated his parents on prevention of further infection.

**Evaluation:** Infection was treated body temperature of the child was 97.8<sup>0</sup>F - 98.4<sup>0</sup>F for a week. Child was normothermic on discharge.

**5. Nursing Diagnosis:** Decreased cardiac output related to drug induced systemic vascular resistance as evidenced by BP of 160/100mmHg on D+98 and his Hb ranged between 6g% - 11.4g%.

**Goal:** Normal cardiac output is maintained.

**Nursing intervention:**

- Assessed his blood pressure Q4h. It was between 140/90 mmHg -160/100 mmHg.
- Monitored body weight daily. His weight was between 6Kg - 6.8Kg.
- Monitored his urine output. It was 30-40mL/hr.
- Assessed for signs of fluid overload. He had only facial puffiness.
- Administered Tab. Aldactone 6.25mg OD.
- Inj. Cyclosporin 15mg IV BD was reduced to 10mg IV BD and then was stopped and Tab. Sirolimus 0.5mg OD was started on D+154.
- Administered Tab. Amlodipine 1.25mg BD and Tab. Labetalol 25 mg BD.
- Monitored haemoglobin level alternate days. Transfused packed cells 50mL-100mL to maintain Hb >8g%. Administered Tab. Folic acid 2.5mg OD (1/7).

**Evaluation:** His blood pressure was maintained between 84/44mmHg - 92/46 mmHg with Tab. Amlodipine 1.25mg BD, Tab. Labetalol 25mg OD and Tab. Aldactone 6.25mg OD, at the time of discharge (D+175). Facial puffiness reduced.

**6. Nursing diagnosis:** Hyperthermia related to immature thermo regulation as evidenced by skin was warm to touch and body temperature of 102.6<sup>0</sup>F.

**Goal:** His body temperature will be reduced to normal.

**Nursing Intervention:**

- Assessed his body temperature and related vital signs. It was 100.20F – 103.80F, heart rate was 130/mt. - 144/mt., Respiratory rate was 28/mt. - 34/mt.
- Removed excess clothing from the child.
- Tepid sponging was done during increased body temperature.
- Made him drink more oral fluids.
- Administered Syr. Paracetamol 100mg prn (antipyretic) and Inj. Meropenem 250mg IV Q8h, Tab. Penicillin (Pentid) 2L unit BD X 3 days and then syrup Septran-P 30mg BD (2/7) (antibacterials).
- Educated his mother on fever management.

**Evaluation:** His body temperature was brought down to 97.8<sup>0</sup>F - 98.4<sup>0</sup>F and WBC of 8,200/mm<sup>3</sup> on the day of discharge (D+175).

**7. Nursing diagnosis:** Imbalanced electrolyte; hypokalaemia related to intake of intravenous antifungal therapy as evidenced by his serum potassium was 2.2mmol/L – 3.4mmol/L.

**Goal:** His serum Potassium level is maintained within normal limits.

**Nursing Intervention:**

- Assessed signs of hypokalaemia such as muscle twitching, weakness, arrhythmia, nil.
- Monitored serum potassium level daily, value was between 2.2mmol/L – 3.4mmol/L.
- Monitored vital signs. Had regular heart rhythm and heart rate was 110/mt.-116/mt. RR 30/mt.
- Administered Inj. KCl 0.75g to 1.5g in 100 mL of normal saline over 3 – 4 hours when there was hypokalemia.
- Administered Inj. Liposomal Amphotericin 25mg IV OD X 15 days and then it was stopped.
- Administered total parenteral nutrition 650mL/day.
- Taught his mother about rich sources of potassium (fruits, vegetables, tender coconut water, whole grains and nuts).

**Evaluation:** His serum potassium level maintained between 3.5mmol/L to 3.8 mmol/L without potassium supplement for three days prior to discharge.

**8. Nursing Diagnosis:** Imbalanced nutrition; less than body requirement related to inability to ingest food as evidenced by refusing to take food orally.

**Goal:** Optimal nutritional requirement per day is met.

**Nursing intervention:**

- Assessed his diet pattern. He refused breast feeding and other oral fluids.
- Monitored his weight daily. He maintained weight between 6Kg to 6.5Kg.
- Provided oral care (gauze sweep wipe) thrice using 0.2% Chlorhexidine mouth wash solution.

- Monitored serum electrolytes. He had hypokalaemia which was corrected with Inj. KCl infusion.
- Administered Total parenteral nutrition (TPN) 650mL/day.

**Evaluation:** He was breastfed, taking food orally and maintained his weight of 6.8Kg at the time of discharge (D+175).

**9. Nursing Diagnosis:** Impaired skin integrity related to immunological reactions as evidenced by presence of papular skin lesions over the face, arms and back and scratch marks on the arms.

**Goal:** Optimal skin integrity is maintained.

**Nursing intervention:**

- Assessed his skin, it was dry and patchy over his arms, face and back.
- Identified signs of itching such as scratch marks.
- The child was given bath daily with luke warm water.
- Applied skin moisturizer (Cetaphil baby lotion) twice daily and steroid (Eumosone) cream once daily, which were prescribed by the Dermatologist.
- Assessed for skin GvHD through skin biopsy which revealed mild spongiotic dermatitis with neutrophil rich intraepidermal vesicles with yeast within it.
- Administered syrup Atarax 0.5mg TID (Anti-histamine), syrup Zincovit 5mL OD (Vitamin supplement), Cap. Budesonide 3mg OD (steroid), syrup Prednisolone 1 mg alternate days (steroid), Inj. Artesunate 15mg IV OD (anti tumor and anti-inflammatory), Tab. Ruxolitinib 2.5 mg BD (JAK1&2 inhibitor) and Inj. Liposomal Amphotericin 25mg IV OD.
- She underwent whole body phototherapy once on alternate days for two weeks since D+155.

**Evaluation:** Child had no itching and skin lesions during discharge (D+175).

**10. Nursing Diagnosis:** Impaired comfort; pruritus related to presence of eczema as evidenced by skin rashes over his face, arms and back, dry skin and scratch marks on the arms.

**Goal:** Optimum comfort is provided.

**Nursing intervention:**

- Assess his comfort level. He was having mild restlessness as he had itching and scratching.
- Provided bath daily using luke warm water.
- Moistened his skin with skin moisturizer Cetaphil lotion twice daily.
- Applied Eumosone steroid cream once daily.
- Administered syrup Atarax 0.5 mg TID and syrup Paracetamol 100mg OD prn.
- Encouraged his mother to engage him in activities as diversional therapy.

**Evaluation:** He was able to sleep and play comfortably with no itching, during discharge.

**11. Nursing Diagnosis:** Deficient fluid volume related to loss of fluid as evidenced by increased frequency and quantity of watery stool.

**Goal:** Adequate fluid volume is maintained.

**Nursing Intervention:**

- Assessed his weight daily. He maintained weight between 6 – 6.8 Kg.
- Assessed for signs of dehydration. He did not have any signs of dehydration.
- Monitored vital signs. HR was 120 – 130/mt. RR was 30-34/mt.
- Hydrated him with IV fluids and ORS.
- Monitored intake and output balance. It was within 100-150mL of positive balance/24 hr.
- Assessed his stool. It was watery, yellow in colour, 6 -7 episodes (300 – 600mL/ day).
- Monitored blood culture and sensitivity, urine and stool alternate weeks, there was no growth. He was negative for the presence of Clostridium difficile in his stool.
- Rigid sigmoidoscopy was done and rectal biopsy was obtained on D+114, revealed third degree gut GvHD.
- Administered Inj. Basiliximab 10mg od (2/7) X 6 weeks and then Tab. Ruxolitinib 2.5 mg BD (JAK1&2 inhibitor), IVIG 5g and Inj. Methylprednisolone 2mg IV OD increased to 3mg IV OD.

**Evaluation:** His diarrhoea stopped after 8 days and he was adequately hydrated throughout his hospitalization.

**12. Nursing Diagnosis:** Defensive coping (father) related to financial crisis(father was on loss of pay leave for three months) as evidenced by projection of blame on nurses regarding his child's treatment related bills and difficulty in establishing good interpersonal relationship with nurses.

**Goal:** Father will demonstrate appropriate interaction with nurses and adhere to the plan of his child's care with more control over the situations.

**Nursing Intervention:**

- Assessed the reason for his defensive approach and learnt his defensive behaviour.
- Explained each plan of care in simple words in his language clearly.
- Considered and met his reasonable requests.
- Initiated friendly and supportive conversation.
- Maintained a neutral tone with consistent positive regard while interacting with his father.
- Conversed in simple, goal-directed manner when his father was defensive.
- Presented positive options for his financial support such as PM, CM, NGO funds.

- Did not react or defend to his father's negative displacements on nurses.
- Reinforced on adaptive coping (problem solving, rationalization) that would help him to achieve the established goal.
- Demonstrated honest, non-judgmental, non-defensive, neutral approach with his father.

**Evaluation:** His father demonstrated adaptive coping with calm and neutral manner of communication with the nurses and expressed his financial concerns and requested for prayer support.

**13. Nursing Diagnosis:** Readiness for enhanced knowledge regarding home care as evidenced by verbalizing interest to know the information to be followed at home.

**Goal:** Learning needs regarding home care is met.

#### Nursing Intervention:

- Assessed his parents' understanding about home care, identifying complication & follow-up.
- Explained about the importance of follow up check-up, monitoring blood counts.
- Discussed about measures to alleviate the mild health problems such as self-administration of antiemetics, antipyretics and analgesics.
- Discussed the need for procedure such as regular central venous catheter care, blood sampling and bone marrow aspiration and biopsy during follow up.
- Explained about the need for blood and platelet transfusion after doctor visit in OPD.
- Instructed about the importance of prevention of infection; personal hygiene, perineal hygiene, oral hygiene and protective environment.
- Educated the parents about food hygiene.
- Facilitated discussion about prophylactic anti-infective therapy, immune suppressive therapy and growth factor.
- Explained about resuming activities of daily living gradually.
- Encouraged to report warning signs such as fever, breathlessness, large episodes of loose stools, severe head ache, abdominal pain, immediately.

**Evaluation:** His parents verbalized understanding of their son's home care, possible complications identification and follow up.

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