INTRODUCTION

Indian childhood cirrhosis of liver is almost exclusively confined to children in the Indian subcontinent. It is seldom reported from countries outside the India. Cirrhosis refers to the final common histologic pathway for a wide variety of chronic liver diseases. The term cirrhosis was first introduced by Laennec in 1826. It is derived from the Greek term *scirrhus* and is used to describe the orange or tawny surface of the liver seen at autopsy.

DEFINITION

Indian childhood cirrhosis is an auto immune disorder characterized by fever, abdominal distension and hepatosplenomegaly and most often seen in Indian children between the age of 6 months to 4 years.

EPIDEMIOLOGY

- Indian childhood cirrhosis is more common in the age group of 6 month to 4 years.
- It is more affected in male child than female child (1 : 4), the first born is at greater risk.
- Among twins, the member of the pair on mixed or artificial feeding in predisposed families was reported to have developed cirrhosis of liver. If the other twin was purely breast-fed for first six months, he or she escaped the illness.
- A definite family predisposition is the hallmark of ICC. Siblings and twins are affected. An increased prevalence of peptic ulcer, asthma, diabetes and migraine in the pedigrees affected by ICC has been observed.
- A large majority of cases belongs to middle class families.
- Majority of the patients have vegetarian dietetic background.
Recently, a significant decline in the incidence of Indian Childhood Cirrhosis has been observed in all parts of India since the use of copper and brass utensils for boiling milk is reduced.

ETIOPATHOGENESIS

It is an autoimmune disorder. ICC continues to be a disease of obscure etiology. The following factors may predispose to the illness:

1. **Toxic** (copper intoxication):
   - studies reported that there is an evidence of excess of copper binding proteins (orcein) in the liver of patient with ICC.
   - if the baby is weaned earlier and if the milk supplements were added to the breast milk often before 3 months of age.
   - use of copper or copper alloy pots for boiling milk and cooking food. During boiling of milk, copper is released from the copper pots and this is probably absorbed in excess from the gut by infant.

2. **Viral infection of the liver**
   - It is a consequence of neonatal or infective hepatitis.

3. **Immunologic factors**:
   - A variety of immunological disturbances were reported in patients with Indian Childhood Cirrhosis. High levels of circulating immune complexes may indicate that an environmental insult might alter the hepatocyte or tissue proteins and initiate an immune mediated injury to the liver.

4. **Nutritional factors**:
- it is believed that malnutrition is an important cause of cirrhosis.

The objections to this hypothesis are:

- Cirrhosis is rare in children suffering from kwashiorkor.
- Cirrhosis is rare in Africa, the home of kwashiorkor.
- Cirrhosis is uncommon in poor in whom malnutrition is common. On the contrary, the incidence of ICC is high in the middle class families.
- Fatty changes of liver in kwashiorkor is either absent or very insignificant in ICC.

- So, nutrition does not have clear role in its pathogenesis. All children are equally affected.

- Good nutrition does not protect the child from the disease.

5. Hepatotoxic agents:

Some hepatotoxic agents like Aflatoxin, produced by Aspergillus flavus that grows on ground nuts, maize, and rice can predispose to this disease. But the actual cause effect relationship is not established.

6. Familial history of the disease

A definite family predisposition is the hallmark of ICC. An increased prevalence of peptic ulcer, asthma, diabetes and migraine in the pedigrees affected by ICC has been observed.

7. Metabolic factors

A child with inborn error of tryptophan metabolism, aminoaciduria, aminoacidemia, disturbed lactose, zinc, copper and magnesium metabolism can predispose to the disease. So a child at risk should be put on low tryptophan diet.

To conclude, no single factor seems to be the cause of ICC. It is possible that a genetically prone child suffers from one or more of the superadded factors (viral, toxic, metabolic and
autoimmune) leading to the overt picture of ICC.

**PATHOPHYSIOLOGY**

**Etiologic factors**

Marked damage of the hepatocytes

Complete disorganization of liver architecture, that is, size of liver varies, and its color ranges from grey tan to frank green

formation of macronodules and micronodules in the surface of the liver. But the portal vein and the biliary passages are patent, the lymphatics appear normal.

Regenerating nodules in the liver are encircled by the bands of fibrous tissue

Absence of regenerative activity and manifesting degenerative changes in the liver

Necrosis and fibrosis of the hepatic lobules.

**CLINICAL FEATURES**

The onset is generally vague and ill defined, ranging from no symptoms to an icteric onset. Some infants show pre cirrhotic symptoms. Two modes of presentation are known:
a. Insidious which occurs in a large majority of the cases.
b. Acute which is less common

I. **Insidious onset** : In this, the disease will last for 6 months to 3 years. Symptoms are grouped under 2 headings.

1. **The pre-cirrhotic symptoms**
   - Irritability
   - disturbed appetite
   - chalky, pasty stools and distension of abdomen.
   - constipation or diarrhea
   - often slight irregular fever.

2. **The cirrhotic symptoms**

   Cirrhotic symptoms are grouped under 3 stages.

   a. Stage I:
      - Slight fever
      - Liver is enlarged to 3-5 cm, edges become sharp and giving an appearance of leafy boarder.
      - Children exhibits jaundice
      - Poor growth
      - Anorexia
      - Constipation/ diarrhea.
      - Clay colored stools.
      - Growth failure

   b. Stage II:
      - Diffuse hepatomegaly
      - Splenomegaly
c. Stage III

- It is the terminal stage of the disease
- Restlessness
- Confusion
- Dyspnea and cyanosis on exertion.
- Evidences of hepatocellular failure in the form of palmar erythema and spider nevi appearance on the upper torso
- A peculiar garlic odor is present in patients with impending liver cell failure.
- Enlarged and hard spleen
- Terminally, there is jaundice and hepatic coma and is often associated with gastrointestinal bleeding.
- Child may die at this stage either from hepatic failure or intercurrent infections.

II. Acute onset: sudden onset of disease and sometimes child becomes symptomatic for a variable period and then shows the manifestations of insidious onset. The symptoms includes:

- Sudden onset of fever
- Jaundice
- Clay-colored stools
- Hepatomegaly
- Child may die with hepatic coma
DIAGNOSTIC EVALUATION

1. History collection
2. Complete physical examination
   Liver can be palpable, very firm in consistency and its boarders will be sharp. On auscultation hepatic bruit in severe cases.
   If there is ascites, fluid thrill test can be done.
3. Liver function test
   -increased ALT(alanine transaminase, an enzyme present in hepatocytes.)
   -increased GGT( gama glutamyl transpeptidase)
4. Prothrombin time, clotting time and bleeding time should be assessed.
   PT will be prolonged
5. Liver biopsy – to find out the sclerosis of liver. It is a reliable method of arriving at a foolproof of diagnosis.
6. Cupriuresis : testing the presence of copper in urine after administration of d-penicillamine.

TREATMENT

Until recently, ICC was dubbed as a “frustrating situation” for which no specific treatment was available. If the diagnosis is made at an early stage( before the development of jaundice and ascites) , ICC is potentially treated.

1. **Initial stage:**
   - Adequate diet with enough of good quality proteins, vitamins and minerals is desirable.
   - Antibiotics should be given to treat the intercurrent infections / infestations.
   - The drug of choice is d- pencillamine (which chelate copper) in a dose of 20- 40 mg/kg/day for 12 to 18 months, leads to marked improvement and even total reversal in the histopathologic picture.
   - Symptomatic treatment should be given.
- Immuno modulators such as levamisole can be used
- Corticosteroids and gammaglobulins are also helpful.
- administer IV fluids if there is dehydration
- Prevention of infection: follow aseptic techniques
  - Prophylactic antibiotics can be given to prevent infection

2. Terminal stage:

- If the patient has entered precoma or coma, the protein intake should be reduced.
- administration of neomycin by gavage and 20% IV glucose drip are helpful.
- oxygen can be administered if necessary.
- exchange transfusion to remove the circulating toxins.

SURGICAL MANAGEMENT

➢ No specific surgical correction. The only successful treatment for end stage liver disease is liver transplantation.
➢ If there is portal hypertension with hematemesis, Sengstaken tube may help to control the esophageal bleed.
➢ A portocaval anastamosis may be done to relieve the portal hypertention and complications of hypersplenism

NURSING MANAGEMENT

➢ Provide symptomatic treatment to the child.
➢ Provide adequate rest and semi fowler’s position.
➢ Check and record the abdominal girth every 4th hourly.
➢ Administer IV fluids if needed.
➢ Provide small and frequent diet.
➢ Provide protein rich food and massive doses of vitamin B₆.
➢ Follow aseptic techniques to prevent infection.
➢ Intake and output chart should be maintained.

➢ **Provide parental education**

- explain the cause, symptoms and management of the disease
- avoid food rich in copper like dry nuts, chocolates, liver etc
- provide small and frequent diet to the child.
- Advise the mother to breast feed their baby for longer period and not to introduce food supplements beyond the age of 6 months.
- Milk used for infant should not be boiled and stored in copper and copper alloy pots.
- reduce the use of brass and copper vessels.
- use aluminium and steel utensils.
- foods rich in tryptophan (milk, eggs, meat, nuts, beans, fish, and cheese) should be reduced.
- provide more vitamin B₆ foods like potato, banana, spinach, soya bean, fruits and vegetables. (vitamin B₆ help to convert tryptophan to niacin)

**Nursing diagnosis**

1. Hyperthermia related to inflammatory process in the liver.
2. Impaired breathing pattern related to pressure on diaphragm secondary to ascites.
3. Imbalanced nutrition less than body requirement related to anorexia.
4. Diarrhoea or constipation related to acute abdominal condition.
5. Parental anxiety related to management of the disease condition.
CONCLUSION

So, ICC is a fatal disease and may result from various diseases such as atresia of bile duct, neonatal hepatitis, cystic fibrosis, galactosemia etc. The parents should be educated about its symptoms, complications and its prevention. ICC has a good prognosis if the liver transplantation is successful.

BIBLIOGRAPHY