

Incidence of Liver Cirrhosis in Infancy and Childhood

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ABSTRACT

Cirrhosis is the end result of chronic liver disease caused by the different pathological factors including congenital malformation, inflammation (hepatitis) and metabolic / storage disorders, leading to liver cell damage. To determine the incidence of liver cirrhosis in infancy and childhood with chronic hepatitis, we studied 41 biopsies of children for the presence of cirrhosis.

Objectives: To provide an overview of current childhood statistics of hepatitis and liver cirrhosis to facilitate analysis of the impact of past research discoveries on outcome and provide essential information for prioritizing future research directions.

Study Design: Retrospective Study.

Place and Duration of Study: This study was conducted at the Department of Basic Medical Sciences Institute, JPMC, Karachi from Jan. 2000 to Dec. 2007.

Materials and Methods: Slides / paraffin blocks of liver biopsies from patients under 15 years of age. The cases were of two categories i.e. retrospective & prospective. The distribution of 41 cases of hepatitis was according to Age & Sex. Total 22 (53.7%) cases were encountered in the youngest of 0-5 years age group, 13 (31.7%) cases in 6-10 years and only 6 (14.6%) cases in 11-15 years age group.

Results: The distribution of 41 cases of cirrhosis of liver, according to age & sex . the maximum 22 (53.77 %) youngest case in 0-5 years ,13 (31.7%) cases in 6-10 years & 6 (14.6%) cases were found in 11-15 year age group.

Conclusion: It is observed that the tendency of liver inflammation was decreased with increasing age and sexual differentiation shows male predominance with male to female ratio of 2.4:1. Liver cirrhosis discovered with increasing age in children. It is found to be a common cause for enlargement of liver with associated hepatitis and chronic liver diseases in infants and children. It can lead to higher risks of acute or chronic responses in adulthood and will require new treatment paradigms building on an increased understanding of the molecular processes for infancy and childhood liver cirrhosis.

Key Words: micro nodular cirrhosis, macro nodular cirrhosis, mixed type of cirrhosis, biliary cirrhosis.

INTRODUCTION

Liver Cirrhosis is a common disease¹. Fibrotic nodules formation in liver are associated with ascites, occurred frequently². In the presence of jaundice many cirrhotic livers appeared green in color instead of brown. The tawny color is due to a deposition of iron in the cirrhotic tissue. In the course of any liver inflammation, Cirrhosis can develop³. It has been customary to classify Cirrhosis in accordance with gross anatomic alterations⁴ occurred in Portal Cirrhosis and Biliary Cirrhosis⁵.

Cirrhosis is the end result of chronic liver disease, like congenital malformation, inflammation (hepatitis) and metabolic / storage disorders leading liver cell damage. Hepatic infection has become a major worldwide health problem due to potential natural course of the disease leading cirrhosis and then hepatocellular carcinoma^{6,7}. In chronic liver diseases, the regeneration of liver cells are partial and gradual with impairments of functions. Due to increased fibrosis normal lobulation of liver is lost and there is pseudolobules formation is taken place as cirrhosis. In focal and minor injuries the complete regeneration may be possible⁸. Liver cirrhosis is the end

result of protected liver damage⁹. Cirrhosis appears in compensated or decompensated state, in childhood compensated state of cirrhosis is compatible with normal growth and development for prolonged periods. An extensive list of causes is outlined in (Table 1)¹⁸. Researches demonstrate by sequential biopsy the progression from neonatal hepatitis to cirrhosis in a young child.

In Karachi, liver cirrhosis rate is increasing day by day. About 70% of the all new born have transplacental IgG antibodies against hepatitis¹⁰ that last about 8 months of age. Due to lack of proper health facilities or poor economical status and less public awareness about the transmission of major communicable diseases like hepatitis B, hepatitis C and Human Immunodeficiency syndrome causing the increase in cases of cirrhosis¹¹.

MATERIALS AND METHODS

Slides / paraffin blocks of liver biopsies from patients under 15 years of age. the cases were of two categories i.e. retrospective & prospective.

Retrospective:

1. Slides / paraffin blocks of liver biopsies received during last 10 years in the Department of

Pathology, Basic Medical Science Institute (BMSI), Jinnah Postgraduate Medical Center, Karachi.

- Slides / paraffin blocks of liver biopsies received in Department of Pathology, National Institute of Child Health (NICH) Karachi during last 7 years.

Prospective: Slides / paraffin blocks of liver biopsies received during last 10 years in the Department of Pathology, Basic Medical Science Institute (BMSI), Jinnah Postgraduate Medical Center and National Institute of Child Health (NICH) Karachi. A clinical protocol including the particulars about the patients name, age, sex and diagnosis were obtained from the surgical pathology registers, request cards and copies of report.

RESULT

The distribution of 41 cases of cirrhosis of liver, according to age & sex. the maximum 22 (53.77 %) youngest case in 0-5 years, 13 (31.7%) cases in 6-10 years & 6 (14.6%) cases were found in 11-15 year age group.

Table No.1: Showing causes of Liver Cirrhosis

Presenting in Infancy Viral	Presenting in Childhood
Cytomegalovirus	Infectious
Rubella	Chronic Hepatitis B+ _ delta
Herpes simplex	Chronic Hepatitis C
Hepatitis B	Metabolic/genetic
Delta Hepatitis	Alpha-I-antitrypsin deficiency
Bacterial	Cystic fibrosis
Syphilis	Wilson disease
Metabolic/Genetic	Indian childhood cirrhosis
Alpha-Lantitypsin deficiency	Hepatic porphyria
Galactosemia	Hepatobiliary anatomic
Frucllosemia	Choledochal cyst
Tyrosinosis	Intrahepatic cystic biliary
Glycogen storage disease type 3,4	Dilation (Caroli disease)
Niemann-Pick disease	Congenital hepatic fibrosis
Wolman disease	Sclerosing cholangitis
Idiopathic	Toxic/drug
Neonatal hepatitis	Malnutrition
Familial intrahepatic cholestasis	Hepatotoxic drug
Biliary tree abnormalities	
Extrahepatic biliary atresia	
Arteriohepatic dysplasia	
Intrahepatic duct paucity	
Choledochal cyst	
Vascular	
Congestive cardiac failure	
Constrictive pericarditis	
Veno-occlusive disease	
Budd-Chiari syndrome	
Toxic	
Parenteral nutrition	

Table No.2: Showing age and sex Differentiation in Young Population

Age	Male	%	Female	%	Total	%
0-5	16	55.8	6	50.0	22	53.7
6-10	7	24.1	6	50.0	13	31.7
11-15	6	20.1	0	0	6	14.6
Total	29	100	12	100	41	100

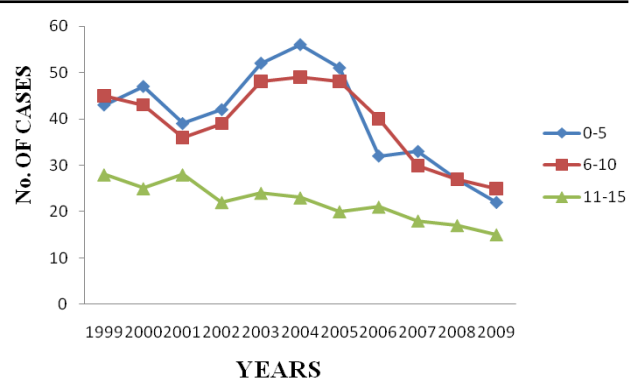


Figure 1: Showing year wise distribution of 41 cases reported for Physiological Liver Cirrhosis and Hepatitis

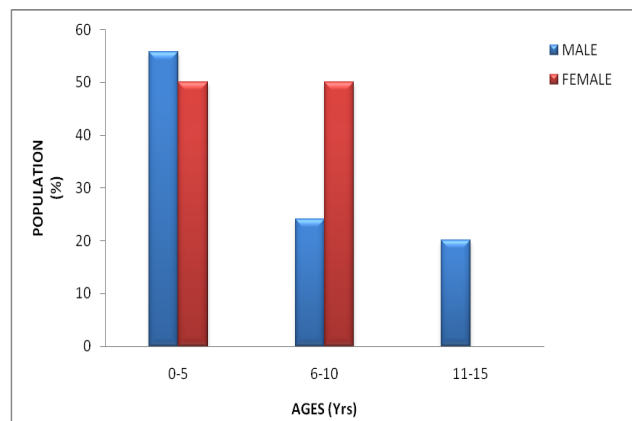


Figure 1: Showing year wise distribution of 41 cases reported for Physiological Jaundice and Hepatitis

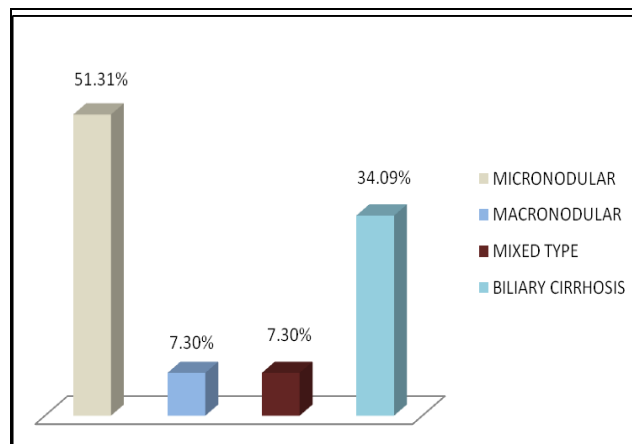


Figure 2: Showing distribution of the different types of Liver Cirrhosis in 41 cases

DISCUSSION

Hepatitis is associated with the necrosis of liver cells with fibrosis and is a major cause of liver diseases^{12,13}. Severity of inflammation and necrosis with parenchymal fibrosis was published in Hepatology as histology activity index (HAI) in 1981¹⁴. Chronic persistent hepatitis has a generally good prognosis as

compare to Chronic aggressive hepatitis which can cause liver cirrhosis¹⁵. This may lead to portal hypertension and liver failure. The primary causes of cirrhosis of the liver in infants and children in many parts of the world are hepatitis and congenital malformation of the biliary tree¹⁶. There are various types of liver cirrhosis reported in children (Fig.2) in which most of the cases were of macronodular type. This type of cirrhosis can occur after any disease that causes liver cell death. Exactly why the regenerating nodules are not able to replace the function of the liver is not clear, although it may be the scarring itself that prevents the functional recovery. The disease is not entirely benign in children, more clinical cases are being diagnosed due to the increased age of those susceptible, which is paradoxical to childhood infection where the majority of infections are subclinical¹⁷.

CONCLUSION

There was over all decrease in tendency of the disease with increased age & male to female ratio was 2.4:1, with male predominance. Our results indicate that the incidence of cirrhosis is high in children with chronic hepatitis, especially of the autoimmune type, and that cirrhosis may occur early, irrespective of cause.

REFERENCES

1. Mathur PS. Some Clinical Observations on the Use of Liv. 52 (An Indigenous Drug) in Cases of Cirrhosis of Liver in Children. Current Medical Practice 2nd ed. 1957.p.107.
2. Laennec RJH. Troite de l'insulation. Paris JS Clande 1926;2:96.
3. Rokitansky C. A manual of Pathological Anatomy. 2nd ed. London: Sydenham Society Atlas Plate 3; 1849. p.125.
4. Connor CL, Quart J. Studies in Alcohol 1940;6: 195-208.
5. Alter MJ, Margolis HS, Krawczynski K, et al. The natural history of community-acquired hepatitis C in the United States. N Engl J Med 1992; 327:1899-905.
6. Tsukuma H, Hiyama T, Tanaka S, et al. Risk factors for hepatocellular carcinoma among patients with chronic liver disease. N Engl J Med 1993;328:1791-801.
7. Kelly DE, Wood RL, Enders AC. Gastrointestinal tract. Baileys textbook of microscopic anatomy. 18th ed. 1984.p.590-611.
8. Thomas HC. chronic hepatitis and cirrhosis. Med Intl. Pakistan 1986;2(10):1181-5.
9. Victor JC, Monto AS, Surdina TY, Suleimenova SZ, Vaughan G, Nainan OV, et al. Hepatitis A vaccine versus immune globulin for postexposure prophylaxis. N Engl J Med 2007;357(17):1685-94.
10. Wong DC, Purcell RH, Sreenivasan MA, Prasad SR, Pavri KM. Epidemic and endemic hepatitis in India: Evidence for a non-A, non-B hepatitis virus aetiology. Lancet 1980;2:876-9.
11. Mowat AP. Chronic Hepatitis. liver disorders in Childhood. Butter worth Heinemann Ltd.: Lincare House Jordan Hill Oxford 1994.p.180-6.
12. Shakoar KA. Histological diagnosis of pediatric liver disease. Pakistan Pediatr J 1987;11(2):73-80.
13. Knodell RG, Ishak KG, Black WC, Chen TS, Craig R, Kaplowitz N, Kiernan, et al. Formulation and application of a numerical scoring system for assessing histological activity in asymptomatic chronic active hepatitis. Hepatology 1981;1: 431-435
14. Bianchi L, de Groote J, Desmet VJ, Gedigk P, Korb G, Popper H, Poulsen H, et al. Morphological criteria in viral hepatitis. Lancet 1971;1:333-337.
15. Mews C, Sinatra F. Chronic Liver Disease in Children. Pediatr Rev 1993;14:436-443.
16. Gust ID. Epidemiological patterns of hepatitis. A in different parts of the world. Vaccine 1992; 10 Suppl 1: S56-S58
17. Purcell RH, Mannucci PM, Gdovin S, Gringeri A, Colombo M, Mele A, et al. Virology of the hepatitis A epidemic in Italy. Vox Sang 1994;67 Suppl 4: 2-7; discussion 24-26.

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