The Pattern of Paediatric Hepatobiliary Disorders in the Eastern Province of Saudi Arabia

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ABSTRACT

The pattern of paediatric hepatobiliary disorders in the Eastern Province of Saudi Arabia is reported. From May 1981 to December 1990, 112 patients were seen at King Fahd Hospital of the University in Al-Khobar, and retrospective review of medical records was possible in 82 patients. The age ranged from birth to 13 years, the male to female ratio was 2:1, and 83% of the children were Saudi nationals. Cholestatic syndrome of infancy was the commonest diagnosis occurring in 44 patients (68.2%). Of these, 21 patients had intrahepatic cholestasis, nine had extrahepatic biliary atresia, and the diagnosis of cholestasis was unknown in 14 patients. The pattern of other hepatobiliary diseases is also reported. It is concluded that major paediatric hepatobiliary disorders occur in the Eastern Province of Saudi Arabia. This information should help in the planning of patient care and in medical education.

The importance of information on patterns of disease in different populations is well recognised. Definition of priorities in patient care, teaching, and research depends on the pattern of problems in any given population. Although the pattern of paediatric hepatobiliary disorders is well established in the Western literature, it is possible that geographical variations exist.

In Saudi Arabia, information on paediatric hepatobiliary diseases is scarce[1,2]. The objective of this report is to define the pattern of these disorders in the Eastern Province of Saudi Arabia.

METHODS

From May 1981 to December 1990, a retrospective review of medical records of all children presenting with complaints related to the hepatobiliary system was carried out. During this period, all patients had been examined and managed either by a paediatric consultant, surgical consultant, or both, at King Fahd Hospital of the University in Al-Khobar, Eastern Province of Saudi Arabia. This hospital is the main referral centre in the area, but also provides care to patients who attend the emergency room without appointment or referral. In addition to complete history and physical examination, available investigations as and when necessary, were used in each case. These included all routine laboratory tests, liver function tests, serology of hepatitis; screening for congenital infections, galactosemia, alpha-1-antitrypsin deficiency and Wilson's disease. Ultrasound and computerised tomography were performed on all patients, whereas sweat chloride test and hepatobiliary scintigraphy were only occasionally performed. Closed liver biopsy, exploratory laparotomy with operative cholangiography and wedge liver biopsy were performed when deemed necessary by the consultant physicians.

Information retrieved from medical records included the hospital number, full name, age, sex, nationality, results of all investigations and final diagnosis. The combination of hospital number and names were used to safeguard against duplicate inclusion of patients who had several visits or admissions.

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RESULTS

From May 1981 to December 1990, 112 patients had been registered with complaints related to hepatobiliary disease. However, medical records were adequate for review in only 82 patients. The remainder of medical records either contained inadequate information or could not be located at the time of survey. The age range was from birth to 13 years with a mean of 30 months. The male to female ratio was 2:1, and 83% were Saudi nationals.

Table 1 indicates the pattern of infantile cholestasis in a total of 44 patients. The site of cholestasis was determined in 30 patients (68.2%). The remaining 14 patients (31.8%) could not be investigated fully. All cases of extrahepatic biliary atresia (9 patients) were diagnosed by laparotomy and operative cholangiography. The patency of extrahepatic biliary tract was documented in all 21 patients with intrahepatic cholestasis by either hepatic scintigraphy or laparotomy and operative cholangiography.

The pattern of other hepatobiliary diseases in the remaining 38 patients is shown in Table 2. The commonest diagnosis was hepatitis occurring in 15 patients (39.5%). There were two cases each of Hepatitis A, Hepatitis B, chronic active hepatitis, and schistosomiasis. The cause of hepatitis and cirrhosis was unknown in seven cases. Obstructive jaundice beyond infancy was the presenting symptom in nine patients; cholelithiasis was diagnosed in six patients (66.7%), and all required surgical treatment.

Table 1
Pattern of Infantile Cholestasis

Diagnosis	*Number (%) of cases*	*Nationality Saudis(%)*	*Sex M/F*
1. Extrahepatic biliary atresia	9 (20.5%)	100	6/3
2. Intrahepatic Cholestasis	21 (47.7%)	95.2	5/6
- Neonatal hepatitis	– 17	(94)	13/4
- Intrahepatic biliary hypoplasia	– 4	(100)	2/2
3. Unknown causes	14 (31.8)	71.4	9/5
Total	44 (100%)	—	30/14

Sickle cell disease or thalassaemia were underlying disorders in five patients (55.6%). No other predisposing conditions such as previous infection or surgery were identified in the remaining four patients. Familial hyperbilirubinemia was diagnosed in five patients including two cases of Criggler-Najjar Syndrome type I, two cases of Gilbert's disease, and one case of Dubin-Johnson Syndrome. Four cases have been diagnosed as glycogen storage disease. Hepatoblastoma was diagnosed in three patients and there were two cases of congenital hepatic fibrosis, confirmed by wedge liver biopsy.

DISCUSSION

Although the data presented do not reflect the incidence or prevalence of disease in the Eastern Province of Saudi Arabia, it provides overall information on the range of disorders encountered, and documents the occurrence of certain entities that have not been reported from the area. The pattern of hepatobiliary disorders in this report indicates a predominance of cholestatic disorders of infancy. Forty-four patients out of 82 (53.7%) had infantile cholestasis. This high proportion may be related to the referral nature of our institution which is the principal centre in the Eastern Province that provides care for these patients. Another possibility is that the prevalence of these diseases may be truly high in this area. Among infants presenting with prolonged jaundice, intrahepatic cholestasis was the commonest diagnosis in our patients, accounting for 47.7% of the cases, followed by extra-hepatic biliary

Table 2
Pattern of other Hepatobiliary Diseases

Diagnosis	*Number (%) of cases*	*Nationality Saudis(%)*	*Sex M/F*
1. Hepatitis + Cirrhosis	15 (39.5)	86	9/6
2. Obstructive Jaundice	9 (23.7)	78	7/2
3. Familial Hyperbilirubinemia	5 (13.1)	60	4/1
4. Glycogen Storage Disease	4 (10.5)	50	2/2
5. Hepatoblastoma	3 (7.9)	100	3/0
6. Congenital Hepatic Fibrosis	2 (5.3)	50	0/2
Total	38 (100)	—	25/13

atresia (20.5%), whereas 14 patients (31.8%) could not be diagnosed, either because of unavailability of appropriate investigations or because of objection of parents to the performance of certain procedures such as liver biopsy and minilaparotomy. Although this pattern is similar to that reported in the Western literature[3], the percentage of patients that could not be investigated is high (31.8%), and justifies the need for establishment of specialised centres. Physicians caring for young infants should be aware that infants presenting with prolonged jaundice require prompt referral to these centres in order to identify treatable causes as early as possible.

The pattern of the hepatobiliary disorders indicates the occurrence of some interesting entities. Hepatitis with or without cirrhosis was the commonest diagnosis occurring in 15 patients (39.5%). There were two cases each of hepatitis A, B, chronic active hepatitis and schistosomiasis. Four patients had hepatitis and 3 cirrhosis; all of unknown aetiology. The small number of cases of hepatitis A and B does not reflect the true proportion in the Region as these patients are usually admitted in isolation hospitals in the area. The two patients with hepatosplenic schistosomiasis were from the South-West of Saudi Arabia and Yemen respectively reflecting the known endemicity of these areas for schistosomiasis.[4] The limited resources in our institution combined with the reluctance of some parents for liver biopsy explain the high percentage of patients with hepatitis and cirrhosis that could not accurately be diagnosed (7/15 = 46.7%).

The finding of cholelithiasis in 66.7% of the children presenting with obstructive jaundice indicates that cholelithiasis is an important cause of obstructive jaundice in our patients even in the absence of underlying haemolytic disease. Such a pattern is consistent with the experience of others[5]. The occurrence of familial hyperbilirubinemia in Saudi children has not previously been reported with the exception of two cases of Dubin-Johnson Syndrome[6]. Awareness of this pattern should help in the differential diagnosis of jaundice in infants and children in the Region, thereby, avoiding unnecessary diagnostic procedures. All patients with glycogen storage diseases presented with hepatomegally and the diagnosis was confirmed by liver biopsy in three patients whereas the diagnosis in the fourth patient was based on clinical and biochemical findings. Unfortunately, the type of glycogen storage disease was not determined because of lack of diagnostic facilities in our institution. The documentation of the occurrence of hepatoblastoma in three patients indicates that this condition should be considered in the differential diagnosis of hepatomegally in infants and children in the Region. No cases of other liver tumours were documented during the period of this study. Congenital hepatic fibrosis was the final diagnosis in two patients who presented with massive hepatomegaly. Closed liver biopsy was inconclusive in both cases and open liver biopsy was necessary to confirm the diagnosis. The association with multicystic kidney disease was documented in one patient. These two cases and a previous report from Saudi Arabia[2] indicates that congenital hepatic fibrosis occurs in Saudi children and should be considered in the differential diagnosis of hepatomegaly. Finally, despite the search for Wilson's disease and alpha-1-antitrypsin deficiency, we did not document this diagnosis in any of our patients. However, we are aware of reports of cases of Wilson's disease from other centres in the Kingdom of Saudi Arabia indicating that this diagnosis should always be considered in Saudi children with liver disease[7-9].

Although our data does not reflect prevalence or incidence of these disorders in Saudi Arabia the pattern indicates that major paediatric hepatobiliary disease do occur in the Eastern Province and probably in other provinces as well. This information should help the planners of patient care and medical educators.

REFERENCES

1. Babiker MA, Bahakim HB, El-Hazemi MAF. Hepatitis B and Hepatitis A markers in children with thalassemia and sickle cell disease in Riyadh. Ann Trop Paediatr 1986;6:59-62.
2. Abdulla AMA, Nazer H, Attiyah M, Ali MA. Congenital hepatic fibrosis. [Abstract]. Ann Saudi Med 1990;10:225A.
3. Fitzgerald JF. Cholestatic disorders of infancy. Pediatr Clin North Am 1988;35:357-73.
4. Ghandour AM, Al-Robai AA, Doenhoff M, Banaja AEA. Epidemiology of Schistosomiasis in Western Saudi Arabia: 1. Prevalence, intensity and incidence in the human human population in the community. Saudi Medical Journal 1991;12:25-9.
5. Gryboski J, Allan Walker W. Acquired lesions of the biliary tree. In: Gastrointestinal problems in the infant. Gryboski, Walker, eds. Philadelphia: WB Saunders, 1983.
6. Yasawy MI, Al-Karawi M, Coode P. Dubin-Johnson Syndrome (black liver disease): Report of two cases. Ann Saudi Med 1988;8:382-5.
7. Nazer H. Wilson's disease: a plea for early diagnosis. [Abstract]. Ann Saudi Med 1988;8:88A.
8. Al-Mofleh IA, Hafeex MA, Bahemuka M, Karrar ZA, Bahakim H. Wilson's disease: A review of seven consecutive cases at the University Hospital, Riyadh. [Abstract] Ann Saudi Med 1988;8:78A.
9. Ahmad FER, El-Tayeb AA, Satti MB. Wilson's disease in Saudi Arabia: Report of a Saudi Arab Family. Ann Saudi Med 1991;11:605-10.