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Original Research Article.

Clinical Features of Autosomal Dominant Polycystic Kidney Disease (ADPKD) Among Bangladeshi Patients in a Tertiary Care Center

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ABSTRACT

To observe the clinical nature of ADPKD in Bangladeshi patients we studied 40 cases, among them 16 (40%) were male and 24 (60%) female. A higher proportion of younger than older patients were affected (40% Vs 10%, P<0.05). Hypertension and loin pain was present in 30 (75%) and 22 (55%) cases respectively. Renal function at presentation was normal in 20 (50%) cases, with mild to moderate and severe renal failure was present in 16 (40%) and 4 (10%) cases respectively. Bilateral enlarged kidneys found in 30 (70%) cases. Size of kidneys varies from 12.1cm to 25.6cm. Multiple cysts in both kidneys were present in 36 (90%) patients, with hepatic and pancreatic cyst was present in 15 (37.5%) and 3 (7.5%) cases respectively. Much younger patients are diagnosed as ADPKD in our population, so every effort should be made for early diagnosis in suspected cases so that need for dialysis may be reduced by retarding rate of progression by conservative measures.

Keywords: Autosomal Dominant Polycystic Kidney Disease, Hypertension, Renal Function.

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INTRODUCTION

Autosomal dominant polycystic kidney disease (ADPKD) is a multisystem disorder characterized by multiple bilateral renal cyst associated with cysts in other organs such as liver, pancreas and arachnoids membrane. It occurs worldwide and in all races and is considered the most common inherited cystic disease of kidney with a prevalence of genetically affected individuals at birth estimated at 1:400 to 1:1000.[1] In the United States, approximately 500000 peoples are affected and ADPKD accounts for 5% of end-stage Renal disease (ESRD) in developed countries.[2] One study on dialysis patients[3] in Bangladesh showed that 2.55% cases were suffering from ADPKD. Another study[4] among Bangladeshi ESRD patients on dialysis showed that ADPKD contributes 13% of cases. So, in Bangladesh, it is one of the important causes of ESRD. But unfortunately, no paper regarding the nature of the disease in our country is available, so we became interested to study the nature of the disease in our population. Probably this is the first paper on ADPKD among Bangladeshi patients.

METHODOLOGY

We studied forty (40) patients who were observed in outpatient and inpatient department of Nephrology in Sir Salimullah Medical College and Midfort Hospital, Dhaka. Suspected patients were examined clinically with special attention to family history of kidney disease and the kidney status was evaluated with the following investigations: Urine R/E, CBC, S Creatinine., Serum electrolytes, Ultrasonogram of whole abdomen and in special cases X- ray KVB and urine for culture and sensibility was done.

For diagnosis of autosomal dominant polycystic kidney disease (ADPKD), Ravine Revised Criteria[5] was adapted in this study. According to Ravine criteria at least three (Unilateral or bilateral) renal cysts and two cysts in each kidney are sufficient for diagnosis of at-risk individuals age15 to 39 and 40 to 59 years respectively. For at-risk individuals age 60 and older, four or more cysts in each kidney is considered diagnostic.[5]

In our study, bilateral enlarged kidneys with multiple renal cysts were considered diagnostic. Patients with a positive family history and unilateral renal cysts with enlarged kidney were considered as ADPKD. Diagnosis was more firmly accepted when these finding was associated with cysts in other organs like liver, pancreas.

Statistical Analyses

Statistics for Continuous Variables were reported as mean ± standard deviation (SD) and for categorical variables as frequency (percent). The chi-squared test was used to compare categorical variable. P-value <0.05 was considered statistically significant.

Table 1: Age & Sex distribution among the study population

Criteria	N (%)	P-value
Age		
≤40 years	16(40%)	<0.05
≥60 years	6(15%)	
Sex		
Male	16(40%)	<0.05
Female	24(60%)	

Table 2: Ultrasonographic Findings

Criteria	Findings (n=40)
Bilateral enlarged kidneys	30 (70%)
Kidney size (pole to pole)	12.10cm to 25.6cm (mean 18.9 ± 5.32cm)
Multiple cysts in Both Kidneys	36 (90%)
Cysts in other organs	
Liver	15(37.5%)
Pancreas	3(7.5%)

RESULTS

Total forty (40) patients were studied among them 16(40%) were male and 24 (60%) female. Age ranged from 22-65 years (Mean 43.51± 12.32). Younger patients≤40 years was 16 (40%) and older patient's ≥ 60 years were 6 (15%), showing a higher population of younger than the older patients (40% Vs 15%; p<0.05). Hypertension was present in 30 (75%) patients studied. Hemoglobin ranged from 5-13 gm/dl with 20 (50%) were anemic having hemoglobin level ≤ 10 gm/dl. Loin pain was present in 22 (55%) patients and only 6(15%) patients have concomitant renal stone disease. About renal function, renal function was normal in 20 (50%) patients and mild to moderate function impairment was present in 16 (40%) and 4 (10%) patients respectively. 4 (10%) patients have severe renal failure (CKD4 and CKD-5) at presentation. 16 (40%) patients suffering from the disease for the last 5 years among them 8 (20%) were on dialysis during the study period. Ultra-sonogram (USG) showed bilateral enlarged kidneys in 30(70%) patients. Sizes of kidneys varies from 12.10 cm -25.6 cm from pole to pole (mean 18.9±5.32cm).

Multiple cysts in both kidneys were present in 36 (90%) patients and 18 (45%) patients bearing cysts in other organs like liver 15 (37.5%) and pancreas 3 (7.5%) patients.

History reveals 12 (30%) patients have a positive family history of cystic kidney disease. Total family members affected were 25. Among them brothers/sisters were more commonly affected. Other patients failed to give any history regarding kidney disease of their family members.

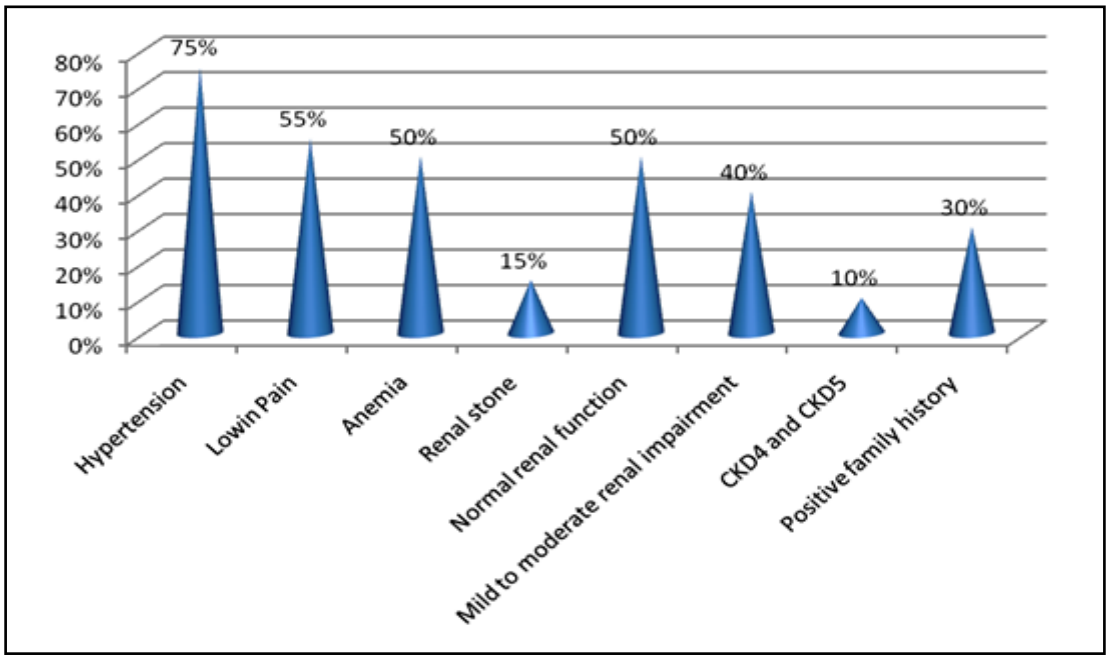

Figure 1: Study results

DISCUSSION

This is the first study on ADPKD among Bangladeshi patients. Among 40 patients there was female predominance in this study (60% Vs 40%;p<0.05). This female predominance is probably due to more USG examination done in female populations in our country for various conditions like pregnancy, Gynecological and various Medico-surgical reasons. Age study shows that younger age group (≤40 years) is more than older (>60 years) patients (40% Vs 15%; p<0.05). This may be due to wider use of sonographic study now-a-days in our younger population for various medico-surgical check-up in comparison to far-past. Regarding symptoms it was observed that symptoms typically begin between 30-50 years of age. Hypertension is the commonest clinical manifestation with some series representing its presence in up to 80% of patients.[6]

Early detection and treatment of hypertension is important because cardiovascular disease is the main cause of death in ADPKD patients.[7] Our study shows 30(75%) of patients were

hypertensive, this is consistent with other studies. Bellp et al showed that even before the presence of significant renal impairment, a large population of patients with ADPKD has high blood pressure, 60% of men & 45% of women.[8]

Gabow PA mentioned that pain is a very prevalent symptom (60%) that often leads to the diagnosis of ADPKD.[9] Our study shows, 22 (55%) patients were suffering from loin pain, this finding is consistent with the abovementioned study done by GabowPA.[9]

About 20-30% of ADPKD patients develop renal stone disease.[10] Rafti UB showed that renal stone occurs in about 20% of patient with ADPKD.[11] Our study shows renal stone affecting 6(15%) of patients which is very similar to the other studies as mentioned above. Our study shows that multiple cysts in both kidneys were present in 36 (90%) of patients. This finding is consistent with the diagnostic criteria for ADPKD as mentioned by PeiYerd[5] and Ravine et al.[12] In our study, 18 (45%) patients having cysts in other organs. Among them 15 (37%) having hepatic and 3 (7%) were having pancreatic cysts. Fick GM[13] showed that hepatic cyst occurs in up to 60% of ADPKD patients. These findings show a much higher occurrence of hepatic cyst than our study (60% Vs 45%). In another study[14], overall prevalence of hepatic cysts in patients with ADPKD was 83% using MRI. This study also showed a much higher occurrence of hepatic cysts than our study (83% Vs 48%). This difference in probably due to more accurate detection of cysts by MRI in that study, as we used only USG study.

Our study shows 20 (50%) of patients have normal renal function, 16 (40%) have CKD and 6 (10%) patients having severe renal impairment. ADPKD ultimately progress to ESRD and it occurs for 10-15% of patients requiring dialysis in USA.[15,16]

CONCLUSION

Much younger patients in our population are diagnosed as ADPKD due to multidisciplinary use of USG study. Among them 16% have severe renal failure; so every attempt should be mode for early diagnosis in suspected cases to reduce the need for dialysis by retarding progression by conservative measures.

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