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The profile of Autosomal Dominant Polycystic Kidney Disease (ADPKD) patients in rural areas of North India

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Abstract

Aims & Objectives: To study the profile of Autosomal Dominant Polycystic Kidney Disease (ADPKD) in low lying areas of NorthIndia. This is a 2 year observation study of patients attending Nephrology department in National Institute of Medical Sciences, Shobha Nagar, Jaipur, Rajasthan, India. **Material & Methods:** This is a 2 year prospective study done in National Institute of Medical Sciences- a tertiary hospital in rural area close to Jaipur. All ADPKD patients attending Nephrology department were studied to know their complete profile. **Results:** A total of 54 ADPKD patients attended Nephrology department between Jan 2014-Jan 2016, Out of which 34 patients presented with pain abdomen, 22 patients with hypertension, 8 patients with chronic renal failure kept on medical management, 8 with Chronic Kidney Disease CKD-ESRD (End Stage Renal Disease) on maintenance hemodialysis(MHD). Out of these 8 CKD-ESRD, there were 4 incidents of AV fistula thrombosis and 1 died recently whereas 20 patients remained completely asymptomatic. **Conclusion:** Timely diagnosis, proper understanding of disease by patients will provide good quality of life in these patients. Low salt diet, good control of hypertension (<130/80 mm of hg) delays progression to ESRD in these patients.

Keywords: Autosomal Dominant Polycystic Kidney Disease, Chronic Kidney Disease, End Stage Renal disease, AV fistula thrombosis.

INTRODUCTION

Autosomal dominant polycystic kidney disease (ADPKD) is the most frequent inherited renal disease with an estimated prevalence of approximately 1 in 800 live births. It is caused by mutations in two genes: PKD1 in 85% and PKD2 in 15% of cases [1]. Clinical presentation ranges from asymptomatic cases to severe impairment due to enlargement of the kidneys and liver. ADPKD is characterized by increasing kidney size and gradually decreasing renal function. The majority of patients require dialysis by the age of 60 [2]. Diagnostic modality of choice is ultrasonography (Table 1) [3]. The progressive growth of renal cysts can cause tubular obstruction and renal ischemia, which increase activity of the renin-angiotensin system (RAS) with arterial hypertension [4]. Renal manifestations include hypertension, nephrolithiasis, cyst infection, urinary tract infection, hematuria and renal insufficiency. Hypertension and renal insufficiency are considered as serious markers for progressive renal damage which needs appropriate treatment, widely neglected in underdeveloped areas. Approximately 50% of individuals with ADPKD have end-stage renal disease (ESRD) by age of 60 years. Although during the first decades of life the renal function remains normal, the growth and development of renal cysts continues. After starting the detioration, the annual reduction of glomerular filtration rate (GFR) is 4.4-5.9 ml/ minute [5]. Total renal volume is the best predictor of progression to CKD. Kidney volume is probably the most important predictive factor for the loss of renal function. A measurement of kidney size is, therefore, recommended as soon as the diagnosis is made. ADPKD patients under age 30 with a combined kidney volume above 1500 mL and an estimated glomerular filtration rate (eGFR) below 90 mL/min are at high risk of needing kidney-replacement therapy (dialysis or transplantation) within 20 years, even if their renal function is normal [2,6]. The progression of CKD is hindered by control of blood pressure generally by ACE inhibitors with target blood pressure of <130/80 mm of hg [7,8]. Management includes low salt diet, good control of hypertension, V2 receptor antagonists (tolvaptan) [9], peritoneal dialysis [10], hemodialysis, nephrectomy and renal transplantation [11]. ADPKD patients' presents

with various manifestations like pain abdomen, urinary tract infections, hypertension and renal insufficiency etc., which needs detail evaluation and management to prolong quality of life. In underdeveloped areas, hypertensive patients are likely to be started on antihypertensives directly instead of evaluating the cause of hypertension which may miss causes like ADPKD. Improper control of hypertension and failure to assess renal function/volume is leading to rapid progression of ADPKD to ESRD, which is a point of interest in our study.

MATERIAL AND METHODS

This is a 2 year prospective observational study conducted in National Institute of Medical Sciences- a tertiary hospital in rural area close to Jaipur.

Statistical methods are calculated using chi-square test using SPSS software for extracting P value.

A total of 54 ADPKD patients attended Nephrology department between Jan 2014-Jan 2016, Out of which 34 patients presented with painabdomen- on further evaluating 10 patients had hematuria, 6 patients had cyst infection.

Out of 54 patients, 22 were hypertensives, in which 8 diagnosed with early stages of chronic renal failure on medical management and 8 presented with CKD-ESRD on MHD, of these 8 ESRD patients, 4 had recurrent AV fistula thrombosis and 1 died eventually.

RESULT

Table 1: Complications seen in ADPKD patients

Complications	Number of patients presented with complications	Percentage
Pain abdomen	34	62.9%
Hematuria	10	18%
Cyst infection	6	11%
Hypertension	22	40%
CKD (stage 1-4)	8	14%
CKD (stage 5-ESRD)	8	14%
AV Fistula thrombosis	4	7%
Death	1	1.8%

Total no. of patients studied-(n=54)

Table 2: Number of ADPKD patients with hypertension

	Hypertensive	Normotensive	Total no. of patients
Symptomatic patients	20	14	34
Asymptomatic patients	2	18	20
Total no. of patients	22	32	54

Total no. of patients n= 54. p value equals to 0.0005, which is significant.

Hypertension (54%) is most commonly diagnosed in symptomatic patients, irrespective whether patient presented in early stage or end stage, with P value 0.0005 it is highly significant.

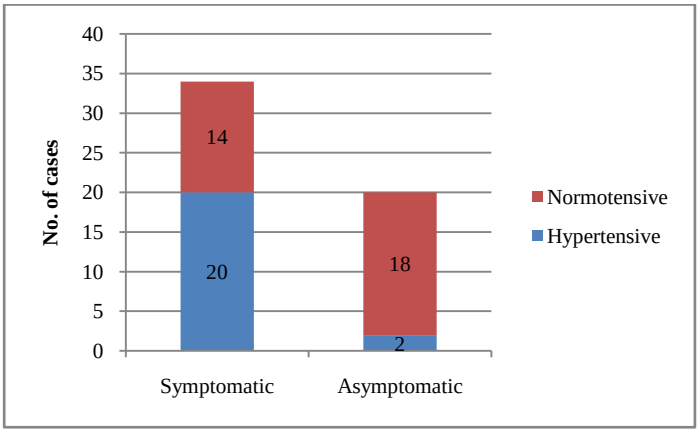

Figure 1: Comparision of asymptomatic and symptomatic patients in relation to hypertension

20 patients who were diagnosed incidentally remained asymptomatic with 2 out of them on anti-hypertensive.

Table 3: Statistical analysis of patients with and without hypertension

	No. of patients	% of patients	X^2
Hypertensive	22	40.74%	0.926
Normotensive	32	59.25%	0.926

$\sum X^2$= 1.852 and p value equal to 0.1736; which is not significant

In patients who were asymptomatic, hypertension was recorded in only 2 patients. With P value 0.1736, it is insignificant.

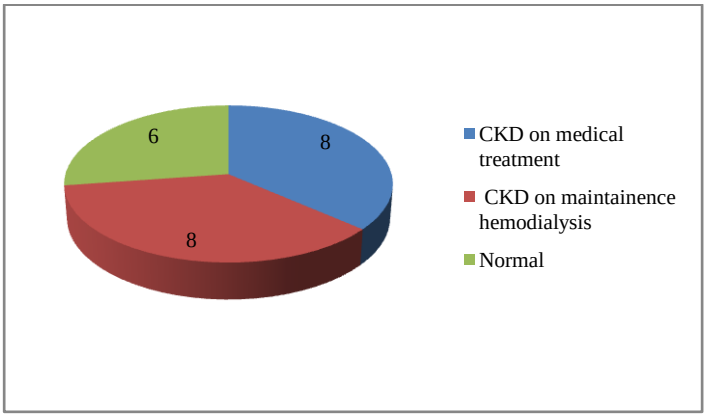

Figure 2: Total no. of hypertensive patients' n=22;

Among Hypertensives (n=22), 6 don't have any renal insufficiency, 8 were on medical treatment and 8 on hemodialysis.

DISCUSSION

The study is compared with last 5 year studies by searching the word ADPKD in PUBMED/Google Scholar.

Generally ADPKD is an incidental finding in sonography/ hypertension /pain abdomen workup. Out of 54 ADPKD patients 34 presented with pain abdomen, which is the most common presenting complaint in this disease. Patients mostly complain of recurrent episodes of pain abdomen localized to flanks radiating to back. Pain may be due to enlargement/compression of cyst, cyst infection, nephrolithiasis etc. In our patients we found 6 patients had cyst infection and 10 had hematuria, which are most common complications of ADPKD.

Among asymptomatic patients who were diagnosed incidentally on ultrasonogram, most commonly we found increase kidney size and

volume which is in accordance with US Consortium for Radiologic Imaging Studies in Polycystic Kidney Disease (CRISP) [12].

There were 22 hypertensives among 54 patients, which accounts for 40% which is in line with Ecder T *et al* [13].

Among these 22, 8 diagnosed with early stages of CKD kept on medical management with proper hypertensive control, low sodium diet and regular followup, whereas 8 patients presented to us with ESRD who were diagnosed retrospectively as having ADPKD were kept on hemodialysis. 4 among them had recurrent history of AV fistula thrombosis and 1 of them died eventually. This shows the spectrum of ADPKD with Hypertension, 8 out of 22 hypertensives (36%) are in need of renal replacement therapy, which shows seriousness of disease.

Early diagnosis of hypertension and suspicion of secondary hypertension is needed to diagnose ADPKD. Early and effective treatment of hypertension is very important in ADPKD in order to slow down the progression to ESRD and prevent cardiovascular complications. This is in accordance with HALT-PKD study which supports strict blood pressure control with significantly lower target levels [14].

CONCLUSION

1. When ever a case of hypertension is diagnosed, it is must to search for secondary cause of hypertension which is easily missed in underdeveloped areas.

2. ADPKD patients irrespective whether symptomatic or asymptomatic should be screened for hypertension, assessed for renal volume, glomerular filtration rate and managed likewise with regular follow ups, as good control of hypertension prevents disease progression and prolongs quality of life.

3. It is always advised to screen all the family members of an ADPKD patient as this disease runs in families, which not only helps in early diagnosis and management but also in study of progression of disease in various geographical locations.

4. Patients with ADPKD with progressive loss of renal function should be planned for renal transplantation which provides near normal quality of life.

REFERENCES

1. Hateboer N et al. Comparison of phenotypes of polycystic kidney disease types 1 and 2. Lancet. 1999; 353(9147): 103-7.
2. Kühn EW, Walz G: The treatment of autosomal dominant polycystic kidney disease. Dtsch Arztebl Int 2015; 112: 884–90.
3. Pei Y et al. Unified criteria for ultrasonographic diagnosis of ADPKD. J Am SocNephrol. 2009; 20(1):205-12.
4. Victor Martinez. Autosomal dominant polycystic kidney disease: review and management update; EMJ Neph. 2014;1: 61-66.
5. Klahr S et al. Dietary protein restriction, blood pressure control and the progression of polycystic kidney disease. Modification of Diet in Renal Disease Study Group. J Am SocNephrol. 1995; 5(12):2037-47.
6. King BF et al. Magnetic resonance measurements of renal blood flow as a marker of disease severity in autosomal dominant polycystic kidney disease. Kidney Int. 2003; 64(6):2214-21.
7. Ecder T, Schrier RW. Hypertension in autosomal-dominant polycystic kidney disease: early occurrence and unique aspects. J Am Soc Nephrol. 2001; 12(1): 194-200. 34. Grantham JJ. Clinical practice. Autosomal dominant polycystic kidney disease. N Engl J Med. 2008; 359(14): 1477-85.
8. Grantham JJ. Clinical practice. Autosomal dominant polycystic kidney disease. N Engl J Med. 2008; 359(14): 1477-85.
9. Torres VE et al. TEMPO 3:4 trial investigators. Tolvaptan in patients with autosomal dominant polycystic kidney disease. N Engl J Med. 2012; 367(25): 2407-18.
10. Li L et al. Peritoneal dialysis as the first-line renal replacement therapy in patients with autosomal dominant polycystic kidney disease. Am J Kidney Dis. 2011; 57(6):903-7.
11. Alam A, Perrone RD. Management of ESRD in patients with autosomal dominant polycystic kidney disease. Adv Chronic Kidney Dis. 2010; 17(2):164-72.
12. Chapman AB, Bost JE, Torres VE, et al.: Kidney volume and functional outcomes in autosomal dominant polycystic kidney disease. Clin J Am Soc Nephrol 2012; 7: 479–86.
13. Ecder T, Schrier RW. Hypertension in autosomal-dominant polycystic kidney disease: early occurrence and unique aspects. J Am Soc Nephrol 2001; 12: 194–200.
14. Schrier RW, Abebe KZ, Perrone RD, et al.: Blood pressure in early autosomal dominant polycystic kidney disease. N Engl J Med 2014; 371: 2255–66.