The prevalence and clinical characteristics of adult polycystic kidney disease in Ilorin, Nigeria

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ABSTRACT

There is a dearth of information on the prevalence and clinical characteristics of adult polycystic kidney disease (APKD) in Nigeria. Earlier studies in the tropical developing countries have reported either the rarity or very low prevalence of this disease and more importantly as a cause of renal failure, but there are hardly any such studies focusing on the pattern of APKD in renal disease patients. The availability and increased use of imaging techniques have led to an increase in the recognition and diagnosis of cystic kidney disease. This longitudinal study was designed to determine the prevalence, clinical characteristics and prognosis of APKD in our environment.

All consecutive adult patients seen in the Renal Care Centre of the University of Ilorin Teaching Hospital over a 15-year period (July 1994-June 2009) were prospectively studied for the presence of APKD. It showed a progressive yearly increase in the number of cases. Seventy-eight out of 986 (8%) renal patients had polycystic kidney disease. The total age range was 25-60 years old with a mean of 49.8±3.6 years and male to female ratio of approximately 2:1. Mean age of males and females was 52±4.5 years and 45±5.97 years respectively. The mode of presentation was chronic renal failure (32%), hypertension (21%), abdominal pain (11%), abdominal swelling (4%) and urinary tract infection (3%) while 21% of cases were incidental findings. Six patients were lost to follow-up while 17 died, giving an approximate mortality rate of 24%. The majority of the mortality recorded in the study was due to sepsicaemia complicating terminal renal failure. Females presented a decade earlier than the males and with faster progression to ESRD.

It is concluded that APKD is not uncommon and is an important cause of morbidity and mortality in our environment. We therefore recommend that females with this disease should be monitored more closely.

Key-words: Adult polycystic kidney disease (APKD); clinical characteristics; prevalence; prognosis.

INTRODUCTION

Adult polycystic kidney disease is the most common life-threatening hereditary kidney disorder with extrarenal complications and a very important cause of end stage renal disease (ESRD) in Caucasians1-3. The two most common types of APKD are autosomal dominant polycystic kidney disease (ADPKD) and autosomal recessive polycystic kidney disease (ARPKD). Both are progressive bilateral disease that...
can occur in adults and may present with polycythae-
mia, subarachnoid haemorrhage, ruptured aneurysm, severe hypertension, significant enlargement of abdominal organs and often lead to renal failure in the majority of cases\textsuperscript{4,5}. There are at least three genetic mutations in ADPKD (PKD-1, PKD-2, PKD-3) that are responsible for polycystic kidney disease with the mutation of PKD-1 located on chromosome 16p while that of PKD-2 is on chromosome 4q. Some studies in the tropical environment, including Nigeria, have documented the rarity of polycystic kidney disease as a cause of chronic renal failure\textsuperscript{6,7} in contrast to other diseases in which the incidence in blacks and Caucasians are similar\textsuperscript{8,9}. The racial difference in the prevalence of APKD is difficult to explain as the disease is a genetic disorder.

The paucity of data on APKD in Nigeria may have been responsible for the notion that it is rare among our renal patients. The availability and increased use of ultrasonography in many of our hospitals within the last two decades prompted us to evaluate all patients seen in our centre for APKD. This longitudinal study was undertaken with the aims of defining the prevalence, clinical characteristics and prognosis of APKD in Ilorin.

\section*{Patients and Methods}

All consecutive adult patients seen in the renal care centre of the University of Ilorin Teaching Hospital (UITH) Ilorin, over a 15-year period (July 1994-June 2009) were prospectively studied. The presence of APKD was evaluated by detailed history, thorough physical examination, abdominal ultrasound and intravenous urogram. Blood and urine samples were collected from all APKD patients for full blood count, erythrocyte sedimentation rate (ESR), blood urea, serum creatinine and electrolytes, urine analysis, microscopy and culture. A 24-hour urine collection for protein estimation and creatinine clearance was performed. The criteria for the diagnosis of APKD in the study included most of the following: presence of at least three bilateral renal cysts as most of the patients were more than 25 years of age, presence of cysts in other intra-abdominal organs, positive family history of bilateral cystic kidney disease, documentation of extrarenal manifestation of the disease, presence of culprit gene by gene linkage analysis, enlarged kidneys with multiple echo-free areas in both kidneys, intravenous urographic demonstration of thinning and angulation of the collecting system with moth-eaten appearance of the cortex and impaired urine concentrating ability. There were logistic problems in screening most family members of patients for APKD either due to unwillingness or refusal of first degree relatives. The routine screening of family members still remain a contentious issue as the knowledge may likely evoke anxiety and decrease job opportunity in our environment.

\section*{Results}

Seventy-eight out of 986 (8\%) renal patients had polycystic kidney disease (Figure 1). It showed a progressive yearly increase over the preceding ten years (Figure 2). The total age range was 25-60 years with a mean of 49.8±3.6 years and male to female ratio of approximately 2:1. Males had age range of 30-60 years with a mean of 52±4.5 years while females had age range of 25-56 years with a mean of 45±5.97 years. The peak age incidence was in the 4\textsuperscript{th} and 5\textsuperscript{th} decade for females and males respectively. The most common mode of presentation (Table I) was chronic renal failure (32\%) followed by hypertension (21\%), abdominal pain (11\%), abdominal swelling (4\%) and urinary tract infection (3\%). About 21\% of cases were incidental findings referred to our centre from other departments and private hospitals. Six patients were lost to follow-up while 17 died, giving

\begin{figure}[h]
\centering
\includegraphics[width=0.5\textwidth]{Sonographic_picture_of_polycystic_kidney_disease.png}
\caption{Sonographic picture of polycystic kidney disease.}
\end{figure}
The prevalence and clinical characteristics of adult polycystic kidney disease in Ilorin, Nigeria

an approximate mortality rate of 24%. The majority of the mortality recorded in the study was due to septicaemia complicating severe uraemia as patients could not afford renal replacement therapy.

**DISCUSSION**

Adult polycystic kidney disease is the most frequent genetic cause of renal failure in adults, accounting for 6-10% of ESRD cases in America and Europe. This hospital-based prevalence rate of 8% in our renal patients shows that it is not rare in our environment. It is likely to be higher if other imaging techniques are combined with ultrasound in community-based multicentre studies as black race and male gender are recognised risk factors for progressive disease in APKD. Ultrasoundography, the preferred diagnostic method in most studies, was used because of its high sensitivity, low cost and lack of exposure to radiation or contrast materials. In individuals with very small or indistinct cysts, T1-weighted magnetic resonance imaging (MRI) is more sensitive and can identify renal cysts as small as 3mm in diameter. This contrasts with ultrasonography which can reliably detect cysts that are 1cm or larger in diameter. The sensitivity of ultrasound for the diagnosis of APKD is almost 100% for those aged 30 years and above. The advantage of CT scan with contrast enhancement and magnetic resonance imaging is that it can pick out inherited renal cystic disease in persons aged under 30 years old who do not have ultrasound detectable renal cysts. MRI is also useful in the measurement of cyst volume, monitoring cyst growth and assessing progress of the disease. However, persons at risk of APKD without sonographically defined cyst can benefit from gene linkage technique and perinatal diagnosis is also possible, using DNA obtained by amniocentesis or chorionic-villus sampling. The mean age of 49.8 ± 3.6 observed in this study is comparable with the findings of other researchers. There was gender disparity in the mean ages and ratio of the population studied. The male to female ratio was approximately 2:1 while the mean ages were 52.5 ± 4.5 and 44.9 ± 5.97 for males and females respectively. The gender ratio of 2:1 noted in this population contrasted with the equal ratio finding in other studies. The reason for the gender disparity is not clear. This may be related to the group of patients studied as they were mainly renal patients seen in a hospital setting. Culture and healthcare seeking patterns of the population may have contributed to the disparity. The cultural preference for male children and the males being the predominant bread winners of the families puts the males at an advan-

<table>
<thead>
<tr>
<th>Presentation</th>
<th>No of patients</th>
<th>% of total patients</th>
<th>No of males(%)</th>
<th>No of females(%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Chronic renal failure</td>
<td>25</td>
<td>32.05</td>
<td>18 (40)</td>
<td>7 (21.2)</td>
</tr>
<tr>
<td>Hypertension</td>
<td>21</td>
<td>26.92</td>
<td>13 (28.9)</td>
<td>8 (24.2)</td>
</tr>
<tr>
<td>Abdominal pain</td>
<td>11</td>
<td>14.10</td>
<td>4 (8.9)</td>
<td>7 (21.2)</td>
</tr>
<tr>
<td>Abdominal swelling</td>
<td>3</td>
<td>3.84</td>
<td>-</td>
<td>3 (9.1)</td>
</tr>
<tr>
<td>Urinary tract infections</td>
<td>2</td>
<td>2.56</td>
<td>-</td>
<td>2 (6.1)</td>
</tr>
<tr>
<td>Incidental findings</td>
<td>16</td>
<td>20.51</td>
<td>10 (22.2)</td>
<td>6 (18.2)</td>
</tr>
<tr>
<td>Total</td>
<td>78</td>
<td>100</td>
<td>45 (100)</td>
<td>33 (100)</td>
</tr>
</tbody>
</table>

Figure 2
Yearly distribution of polycystic kidney disease.
Adult polycystic kidney disease is a common cause of ESRD and studies have shown that more than 50% would have reached ESRD by the age of 60 years. As routine screening of family members of patients could not be traced to the underlying disease. This is not unexpected as patients with unexplained abdominal pains may develop severe pains for reasons completely unrelated to their underlying disease.

The usual manifestations of APKD include chronic abdominal pains, haematuria, infections, nephrolithiasis, hypertension, valvular heart disease, aneurysm of blood vessels, liver and pancreatic cysts, chronic renal failure and diverticulitis of the colon. Even though our patients did not undergo gene linkage analysis, they met the clinical criteria for the diagnosis of autosomal dominant polycystic kidney disease. The most common mode of presentation in our patients was chronic renal failure, followed by systemic hypertension, abdominal pain, abdominal swelling and urinary tract infections.

An approximate mortality rate of 24% was recorded in this study. The majority of the deaths were due to sepsicaemia complicating terminal renal failure as patients could not afford renal replacement therapy.

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As routine screening of family members of patients with APKD that presented with abdominal pains could not be traced to the underlying disease. This is not unexpected as patients with unexplained abdominal pains are likely to be subjected to ultrasoundography. It is concluded that APKD is not common and is an important cause of morbidity and mortality worldwide. The incidence of the disease appears to be on the increase in our environment.
with APKD still remain a contentious issue, we recommend that symptomatic family members or those that develop hypertension, haematuria and proteinuria should be screened for APKD. The disease appears to run a more aggressive course in females resulting in its presentation at a younger age in females than males. The higher proportion of males than females that presented with advanced chronic renal failure in this study support the observation that male gender is a risk factor for progressive renal disease in APKD. It appears from the study that females with APKD may need closer monitoring than males in the early stages of the disease.

**Conflict of interest statement.** None declared.

**References**


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