

## Polycystic Kidney - A Cadaveric Study

<sup>1</sup>Eswari A.K., <sup>2</sup>Swayamjothi S., <sup>3</sup>Sathialakshmi V., <sup>4</sup>Hemanth Kommuru,  
<sup>5</sup>Sai Sucheethra D.  
SSSMC&RI & Jacintha Antony, Sree Balaji Medical College and Hospital, SBMCH

---

**Abstract:** It is a cystic genetic disorder of the kidneys. It is characterized by the presence of multiple cysts however 17% of cases initially present with the observable disease in one kidney, with most cases progressing to bilateral disease in adulthood.

---

### 1. Adult Polycystic Kidney (APKD)

This is relatively common, and varying degrees of the change may be seen from single cysts up to the classical type where the kidneys are converted into a mass of cysts. It is an inherited autosomal dominant trait due to two genes, PKD-1 (85% of cases) which encodes a protein polycystin-1, and PKD-2 (15%) associated with milder disease.

### 2. Infantile Polycystic Disease (Polycystic Kidney of the new born)

The nephrons are said to be normal in number and formation. Cystic dilatation is situated in the terminal branches of the collecting tubules. The disease, if severe, is incompatible with life, and death occurs shortly after birth. It is due to an autosomal recessive trait. Congenital hepatic fibrosis often coexists. Cystic change extends from medulla to cortex.

## I. Introduction

Autosomal dominant polycystic kidney disease (ADPKD) is a common genetic disorder which an incidence of 1 in 1000. It is characterized by progressive development and enlargement of renal cysts leading to end stage renal disease by late middle age (Gabow P.A et al (1992). Cysts are also found in other organs mainly in the liver. ADPKD is considered as systemic disorder (Gabow P.A et al (1992).) as it is usually associated with other conditions. ADPKD is genetically heterogenous (Kimberling W.J., et al (1993) ; Peters D.J., et al (1993). 85% of the cases are due to Polycystic kidney disease 1 (PKD1) locus (Peters D.J., et al (1993) located in 16p13.3. The recent identification (European Polycystic Kidney Disease Consortium (1994) and characterization of the PKD1 gene (International Polycystic Kidney Disease Consortium (1995); Hughes J., et al (1995) shows the primary defect is as a result of reduced or aberrant polycystin expression.

**Embryology:** The Kidneys develop from two sources. The Excretory tubules are derived from the **Metanephros** and the Collecting part is formed by ramification of the **Ureteric bud**.

**Genetic Aspect Of This Condition:** The epithelium of the Uretric bud from the mesonephros interacts with the mesenchyme of metanephric blastema. Genes involved in differentiation of the kidney - **WT<sub>1</sub>**, expressed by the mesenchyme, enables this tissue to respond to induction by the ureteric bud. **Glial derived neurotrophic factor (GDNF) and Hepatocyte growth factor (HGF)**, also produced by the mesenchyme, interact through their receptors, RET and MET, respectively, in the ureteric bud epithelium, to stimulate growth of the bud and maintain the interactions. The growth factors FGF2 and BMP7 stimulate proliferation of the mesenchyme and maintain WT1 expression.

WNT9B and WNT6 secreted by branches of the ureteric bud epithelium cause upregulation of PAX2 and WNT4 in the surrounding mesenchyme. In turn, these genes cause the mesenchyme to epithelialize (PAX2) and to then form tubules (WNT4). Changes in the extracellular matrix also occur, such that laminin and type IV collagen form a basement membrane (orange) for the epithelial cells

## II. Materials & Methods:

Polycystic kidney observed in a male cadaver

**Observation:** Multiple cysts were seen on the surface of both the kidneys(Fig.1)

Sagittal section of the kidneys also revealed the presence of cysts(Fig.2)

Histological section of the kidneys (H&E stain 10X) showed the presence of cysts of various shapes and size close to the cortex(Fig.3,3a,3b)

### III. Discussion & Conclusion

In ADPKD Kidneys cysts can develop from any of the part of the nephron (Baert, 1978). It is not known at what time the majority of cysts form, but rare analyses of renal tissue from PKD1 affected fetuses have shown tubular and glomerular microcysts as early as 12 weeks of gestation (Reeders S.T., et al (1986) Waldherr R et al (1989)), although there is evidence that cysts continue to form in adult life (Baert L (1978).

### IV. Extrarenal Manifestations:

The major extrarenal complications of ADPKD include cerebral aneurysms, hepatic cysts, pancreatic cysts, cardiac valve disease, colonic diverticula, and aortic root dilatation. Such findings were not present in the cadaver.

### References

- [1]. Baert L.(1978) *Kidney Int.* 13, 519-525
- [2]. European Polycystic Kidney Disease Consortium (1994) *Cell* 77. 881-894.
- [3]. Gabow P.A., Johnson A.M., Kachny w.d., Kimberling W.J., Lezotte D.C., Duley I.T. & Jones R.H (1992) *Kidney Int.* 41, 1311-1319.
- [4]. Gabow P.A. (1990) *Am. J. Kidney Dis* 16, 403-413
- [5]. Gabow P.A., Johnson A.M., Kachny w.d., Kimberling W.J., Lezotte D.C., Duley I.T. & Jones R.H (1992) *Kidney Int.* 41, 1311-1319.
- [6]. Hughes J., Ward C.J., Peral B., Aspinwall R., Clark K., San MILLAN J.L., Gamble V., Harris P.C (1995) *Nat Genet* 10, 151-160.
- [7]. International Polycystic Kidney Disease Consortium (1995) *cell* 81, 289-298.
- [8]. Kimberling W.J., Kumar S., Gabow P.A., Kenyon J.B., Connolly C.J & Somlo S. (1993) *Genomics* 18, 467-472.
- [9]. Peters D.J.M., Spruit L., Saris J.J., Ravine D., Sandkujil L.A., Fossdal R., Boersma J., Van Ejik R., Norby S., Constantinou-Deltas C.D., Pierides A., Brissenden J.E., Frants R.R., Van Ommen G.J.B & Breuning M.H (1993) *Nat Genet.* 5, 359-362.
- [10]. Peters D.J.M & Sandkujil L.A (1992) in *contribution to Nephrology: Polycystic kidney Disease.* Eds. Breuning M.H., Devoto M & Romeo G (Karger, Basel) Vol 97. pp. 128-139.
- [11]. Reeders S.T., Zerres K., Gal A., Hogenkamp T., Propping P., Schmidt W., Waldherr R., Dolata M.M., Davies K.F & Weatherrall D.J (1986). *Lancet* ii, 6-8.
- [12]. Waldherr R., Zerres K., Gall A., & Enders H. (1989) *Lancet* ii, 274-275.

**Fig.1-Multiple cysts were seen shows the presence of cysts**



**Fig.2- Sagittal section of the kidneys on the surface of both the kidneys**



**Fig-3**

**Fig-3a**

**Fig-3b**

Histological section of the kidneys (H&E stain 10X) showing the presence of cysts of various shapes and size close to the cortex(Fig.3,3a,3b)